SURGERY for CONGENITAL HEART DEFECTS

Third Edition

Editors

J STARK, M. de LEVAL and VT TSANG

Great Ormond Street Hospital
for Children NHS Trust, London

Illustrated by

MICHAEL COURTNEY

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Contents

Preface to the Third Edition . . . . . **xix**
Acknowledgements . . . . . **xxi**
Contributors . . . . . **xxiii**
Foreword to the Third Edition . . . . . **xxix**
Foreword to the Second Edition . . . . . **xxxi**
Foreword to the First Edition . . . . . **xxxiii**
Abbreviations . . . . . **xxxv**

GENERAL CONSIDERATIONS . . . . . .

1: Classification and Nomenclature of Congenital Heart Defects, *R. H. Anderson and F. J. Macartney* **3**

Identification of the Cardiac Segments, 4 *Atrial Situs (Atrial Arrangement) and Morphology*, 4 *Ventricular Morphology*, 5 *Muscle Bundles Within the Right Ventricle*, 6 *Great Arterial Morphology*, 7 Junctions Between the Cardiac Segments, 7 *Types of Atrio-ventricular Connection*, 7 *Modes of Atrio-ventricular Connection*, 8 *Types of Ventriculo-arterial Connection*, 8 *Modes of Arterial Connection*, 9 Relations and Cardiac Position, 9 *Implications for Reporting Surgical Results*, 9 Additional Cardiovascular Anomalies, 9

2: Surgical Anatomy, *R. H. Anderson and A. E. Becker* **11**

Cardiac Shape and Position, 11 Pericardial Cavity, 11 Relationships of the Heart, 13 Anatomy of Cardiac Chambers, 15 *The Right Atrium*, 15 *The Left Atrium*, 19 *The Ventricles*, 20 *The Ventricular Septum*, 27 The Atrio-ventricular Junctions, 28 The Cardiac Skeleton, 29 The Atrio-ventricular Conduction System and Bundle Branches, 29 The Coronary Circulation, 30

3: Echocardiography, *I. D. Sullivan and V. M. Gooch* **33**

Basic Principles, 33 *M-Mode and Two-dimensional Echocardiography*, 33 *Doppler Flow Echocardiography*, 33 *Colour-flow Doppler Interrogation*, 35 *Tissue Doppler Imaging*, 35 *Three-dimensional Echocardiography*, 35 *Quantitative Measurement*, 35 Practical Considerations, 37 *Image Orientation*, 37 *Trans-oesophageal Echocardiography*, 38 *Archiving*, 40 Sequential Segmental Anatomy, 41 *Atrial Situs (Arrangement) and Venous Connections*, 41 *Atrio-ventricular Connection*, 44 *Ventriculo-arterial Connection*, 45 *Atria and Atrial Septum*, 48 *Atrio-ventricular Septum*, 48 *Ventricles and Ventricular Septum*, 48 *Cardiac Valves*, 51

The Great Arteries, 51 *Coronary Arteries*, 53 Intraoperative Echocardiography, 54 Postoperative Assessment, 57 Conclusion, 60 Appendix, 60 *Abbreviations in Figures Specific to This Chapter*, 60

4: Foetal Echocardiography, *L. D. Allan* **63**

Organization of Foetal Echocardiography, 63 Normal Foetal Cardiac Anatomy, 64 *The Four-chamber View*, 64 *Imaging the Great Arteries*, 64 Cardiac Malformations, 66 *Abnormalities of Connection*, 66 *Additional Abnormalities*, 68 Implications of Foetal Echocardiography, 69 Conclusions, 70

5: Magnetic Resonance Imaging and Computed Tomography in the Diagnosis of Congenital Heart Defects, *V. Mutharangu, A. M. Taylor and R. Razavi* **73**

Magnetic Resonance (MR) Imaging, 73 Basics, 73 Techniques, 73 *Spin-echo Imaging*, 73 *Gradient-echo Imaging*, 74 *Two-dimensional Gradient-echo Imaging*, 74 *Three-dimensional Gradient-echo Imaging*, 74 *Velocity-encoded Phase Contrast MR*, 74 *Gadolinium Contrast-enhanced MR Angiography*, 74 Acyanotic Heart Defects, 74 *Atrial Septal Defect*, 74 *Ventricular Septal Defect*, 75 *Coarctation of the Aorta*, 76 Cyanotic Heart Defects, 77 *Tetralogy of Fallot*, 77 *Pulmonary Atresia*, 78 *Transposition of the Great Arteries*, 80 *Congenitally corrected transposition (ccTGA)*, 81 *Double-outlet Right Ventricle*, 82 *Hypoplastic Left Heart Syndrome*, 82 Conclusion, 83 Computed Tomography (CT), 84 *MDCT Technique*, 84 *Indications for Cardiovascular MDCT*, 84 Vascular Anatomy, 85 Cardiac Anatomy, 85 *MRI vs. MDCT*, 86 Future Prospects, 87

6: Invasive Investigation and Exercise Testing, *G. Derrick and J. F. N. Taylor* . . . **91**

Invasive Investigation, 91 *General Conduct of an Invasive Investigation*, 91 *Vascular Access*, 91 *Blood Oxygen Saturation Measurements*, 93 *Pressure Measurements*, 93 *Blood Gas Measurements*, 93 *Measurements of Pulmonary Vascular Resistance*, 94 *Testing the Pulmonary Vascular Response*, 94 *Catheter Placement and Haemodynamic Measurement*, 94 *Angiography*, 95 *Postoperative Investigation*, 96 *Risks of Invasive Investigation*, 97 Summary, 97 Cardiopulmonary Exercise Testing in Children and Adolescents, 98 *Limitations*, 98 *Potential Impact*, 98

7: Interventional Catheterization in Congenital Heart Defects, *P. Bonhoeffer and Y. Boudjemline* **101**

Indications, 101 Opening of Abnormally or Normally Closed Structures, 101 *Balloon Dilatation*, 101 *Valvuloplasty for Congenital Valvular Diseases*, 101 *Angioplasty*, 104 *Atrial Septum*, 106 *Endovascular Stents*, 106 *Limitations and General Conclusions on Stent Technology*, 109 Closure of Abnormally Open Structures, 109 *Patent Ductus Arteriosus (PDA)*, 109 *Atrial Septal Defect*, 110 *Muscular Ventricular Septal Defects*, 112 *Perimembranous VSD*, 113 *Other Vascular Abnormalities and Surgically Created Structures*, 114 Trans-catheter Pulmonary Valve Insertion, 114 Hybrid approaches, 116

8: The Anatomy of Ventricular Septal Defects and Their Conduction Tissues, *R. H. Anderson and A. E. Becker* **121**

Basic Rules Regarding Disposition of the Conduction System, 121 Anatomy of Isolated Ventricular Septal Defects, 122 *Defects with Fibrous Continuity Between the Leaflets of the Aortic and Tricuspid Valves*, 123 *Defects with Aortic to Pulmonary*

Fibrous Continuity, 127 *Defects with Entirely Muscular Rims*, 128 Anatomy of the Ventricular Septal Defect in Tetralogy of Fallot, 130 Ventricular Septal Defects in Hearts with Concordant Atrio-ventricular and Discordant Ventriculo-arterial Connections, 131 The Interventricular Communication in Double-outlet Ventricle, 133 Ventricular Septal Defects in Single Outlet of the Heart, 136 Ventricular Septal Defects with Discordant Atrio-ventricular Connections, 136 Hearts with Straddling Atrio-ventricular Valves, 139 *Straddling Left Atrio-ventricular Valves*, 140 *Straddling Right Atrio-ventricular Valve*, 141

9: Use of Pacemakers and Defibrillators in Children, *P. G. Rees* **145**

Pacemaker Terminology, 146 Indications for Pacemaker Implantation in Children, 146 Choice of System to be Implanted, 148 Wire Implantation, 148 Implantation Techniques, 149 *Temporary Systems*, 149 *Permanent Systems*, 149 Pacemaker Problems, 156 *Electrode Dislodgement*, 156 *High Pacing Threshold*, 156 *Lead Fracture*, 156 *Muscle Twitching*, 157 *Erosion*, 157 *Left Ventricular Lead Malposition*, 157 *Infection*, 157 *Non-functioning Wire*, 157 *Endocardial Lead Extraction*, 157 *Elective Generator Replacement*, 158 Pacemaker Follow-up, 158 Conclusion, 158 Implantable Defibrillators, 158 Technique, 159

10: Catheter Ablation of Arrhythmias, *M. Lowe* **161**

Conditions, 161 *Atrial Septal Defect (ASD)*, 161 *Tetralogy of Fallot (TOF)*, 161 *Transposition of the Great Arteries*, 162 *Functional Single Ventricle and Fontan Operation*, 162 *Other Defects*, 163 Surgical Anatomy, 163 Case Selection and Preparation, 163 Electrophysiological Study, 163 *Equipment*, 163 *Conventional Contact Mapping*, 163 *Advanced Mapping Techniques*, 164 Ablation Strategies, 164 *Atrial Flutter and IART*, 164 *Atrial Fibrillation*, 165 *Ventricular Tachycardia*, 165 Complications of Catheter Ablation, 165

11: Perfusion Techniques, *T. Jones and M. J. Elliott* **167**

Pathophysiology, 167 Hypothermia, 168 Altered Perfusion, 169 Haemodilution, 169 Inflammatory Response, 170 *Emboli*, 171 The Extracorporeal Circuit, 171 *Prime*, 171 *Venous Reservoirs and Venous Drainage*, 172 *Blood Pumps*, 173 *Oxygenators*, 173 *Heat Exchanger*, 173 Conduct of CPB, 174 *Heparin Dosage and Heparin Neutralization*, 174 Surgical Aspects, 174 Cannulation, 174 *Arterial Cannulation*, 174 *Venous Cannulation*, 176 *Venting*, 178 *Intracardiac Suction*, 179 Blood Gas Management, 179 Hypothermic Circulatory Arrest and Low-Flow CPB, 180 Myocardial Preservation, 181 Management of Ventilation, 182 Special Circumstances, 182 *Left Superior Vena Cava*, 182 *Other Cannulation Techniques*, 182 *Control of Shunts*, 183 Ultrafiltration (UF), 183 Conclusion, 184

12: Anaesthesia, *E. Sumner* **187**

Cardiac Defects, 188 *Defects with High Pulmonary Blood Flow*, 188 *Defects with Low Pulmonary Blood Flow*, 189 *Transposition of the Great Arteries*, 189 *Left Ventricular Outflow Tract Obstruction*, 189 Systemic and Pulmonary Vascular Resistances, 189 *Pulmonary Vascular Resistance*, 189 *Systemic Vascular Resistance*, 191 Preoperative Management, 191 Induction, 192 *Vascular Access Lines*, 193 *Maintanance of Anaesthesia*, 194 Cardiopulmonary Bypass, 194 Post-bypass, 196 *Fast-tracking*, 197 Pulmonary Hypertension, 197 Re-do Surgery, 197 Special Considerations, 197 *Norwood Operation*, 197 *Univentricular Heart*, 198 *Transplantation*, 198 Closed Cardiac Operations, 199

13: Intensive Care Management of the Postoperative Patient, *D. Macrae and J. Larovere* **203**

Cardiovascular System, 203 *Monitoring*, 203 *Vascular Access*, 204 *Low Cardiac Output*, 204 *Pharmacological Management of Low Cardiac Output (LCO)*, 205 *Non-pharmacological Strategies in the Management of Postoperative Low Cardiac Output*, 208 *Other Cardiovascular Drugs*, 209 *Pulmonary Hypertension*, 209 *Cardiac Tamponade*, 210 Cardiopulmonary Resuscitation, 210 *Postresuscitation Stabilization*, 211 Fluid Therapy after Cardiac Surgery, 211 *Volume Replacement*, 211 Bleeding and Blood Transfusion, 211 Arrhythmias and Pacing, 211 *Early Postoperative Tachyarrhythmias*, 212 *Postoperative Bradycardia and Pacing*, 214 Respiratory System, 214 *Oxygen Therapy*, 214 *Positive Pressure Ventilation*, 215 *Weaning and Extubation*, 215 *Ventilation in Special Situations*, 216 *Analgesia and Sedation*, 216 Gastrointestinal System, 217 *Nutrition*, 217 *Gastroduodenal Paralysis*, 218 *Stress Ulceration*, 219 *Chylothorax*, 219 *Necrotizing Enterocolitis*, 219 Renal System and Fluid Therapy, 219 *Postoperative Fluid Management*, 219 *Electrolytes*, 220 *Capillary Leak Syndrome*, 220 *Perioperative Renal Dysfunction*, 220 *Acute Renal Failure (ARF)*, 221 Sepsis, 221 Neurological Dysfunction, 222 Tracheostomy, 223 *Surgical Technique*, 223 *Posttracheostomy Care*, 225 *Decannulation*, 226

14: Paediatric Cardiac Mechanical Support, *A. Karimova and A. Goldman* **229**

Aims of Cardiac Mechanical Support, 229 Choice of Mechanical Assistance Device, 229 Extracorporeal Membrane Oxygenation (ECMO), 230 Ventricular Assistance Devices (VADs), 230 Intra-aortic Balloon Pump (IABP), 231 Indications and Timing for Mechanical Support, 232 *Postcardiotomy Circulatory Failure*, 232 *Circulatory Failure Due to Myocarditis or Cardiomyopathy*, 232 *Rapid Deployment ECMO or Extracorporeal Cardiopulmonary Resuscitation (ECPR)*, 233 Patient Selection Criteria, 233 Practical Aspects of Mechanical Support, 233 *Cannulation*, 233 *Management of Bleeding*, 234 *Determining Adequacy of ECMO*, 234 *Management of ECMO in Patients with Systemic–Pulmonary Shunt*, 235 *Management of Patients on Mechanical Support*, 235 *Weaning from ECMO*, 235 *When to Stop Mechanical Support (How Long Is Long Enough?)*, 235 Complications of Mechanical Support, 236 Outcomes, 236 Future Developments, 236

15: Surgical Approaches, *J. Stark and V. T. Tsang* **239**

Midline Sternotomy, 239 *Cannulation*, 242 *Closure of Sternotomy*, 243 Posterolateral Thoracotomy, 244 Muscle-sparing Incision, 246 Bilateral Thoracotomy, 246 Endoscopic Approach, 247

SURGICAL PROCEDURES . . . . . .

16: Systemic-to-Pulmonary Artery Shunts, *C. van Doorn and M. R. de Leval* . . . **251**

Indications, 251 Disadvantages, 251 Surgical Technique, 251 *Classical Blalock–Taussig Shunt*, 251 *Potts Shunt and Waterston Shunt*, 254 *Prosthetic Systemic–Pulmonary Artery Shunt*, 255 Results, 259

17: Pulmonary Artery Banding, *V. T. Tsang and J. Stark* **261**

Classical Indications, 261 Current Indications, 261 *Ventricular Septal Defect*, 261 *Transposition of the Great Arteries*, 262 *Double-inlet Ventricle and Tricuspid Atresia without Pulmonary Stenosis*, 262 *Other Indications*, 263 *Hypoplastic Left Heart (HLH)*, 263 Operative Technique, 263 *Approach*, 263 *Technique*, 263

The Degree of Pulmonary Artery Banding, 265 *Material Used for Pulmonary Artery Banding*, 267 *Other approaches to PAB*, 267 Special Considerations, 267 *Transposition of the Great Arteries with Ventricular Septal Defect*, 267 *Double-inlet Ventricle and Tricuspid Atresia*, 267 *Absent Left Atrio-ventricular Connection*, 268 *Banding in Very Small/Young Infants*, 268 Complications, 268 Pulmonary Artery Reconstruction after Banding (Debanding), 268 Results, 268

18: Atrial Septectomy, *V. T. Tsang and J. Stark* **271**

Indications, 271 *TGA*, 271 *Univentricular Heart*, 271 Surgical Technique, 271 Result, 274

19: Persistent Ductus Arteriosus, *J. Stark and V. Hjortdal* **275**

Diagnosis, 275 Indications, 275 Methods for Closure of Persistent Ductus Arteriosus, 276 *Medical*, 276 *Surgical Technique*, 276 Complications, 282 Results, 282

20: Coarctation of the Aorta, *V. T. Tsang and J. Stark* **285**

Diagnosis and Medical Management, 285 *Symptomatic Infants*, 285 *Older Children*, 286 Surgical Technique, 286 *Resection and End-to-end Anastomosis*, 287 *Subclavian Flap Technique*, 289 *Reversed Subclavian Flap*, 290 *Extended Resection and Anastomosis of the Distal Aorta to the Undersurface of the Aortic Arch*, 291 *Patch Graft Aortoplasty*, 292 *Simultaneous Repair of Coarctation of the Aorta and Intracardiac Defects*, 293 Early Complications, 293 *Bleeding*, 293 *Paradoxical or Rebound Hypertension*, 293 *Paraplegia*, 293 *Renal Failure*, 293 *Abdominal Pain*, 293 *Chylothorax*, 294 Late Complications, 294 *Re-coarctation*, 294 Repair of Re-coarctation, 294 Results, 295 Long-term Follow-up, 296 *Hypertension*, 296 *Aneurysm*, 296

21: Interruption of the Aortic Arch, *J. L. Monro* **299**

Anatomy, 299 Diagnosis and Medical Management, 299 Surgical Technique, 300 *Midline Approach*, 300 *Repair of the Aortic Arch through the Left Chest*, 302 Results, 303 Late Follow-up, 303 *Anastomotic Obstruction*, 304 *Left Ventricular Outflow Tract Obstruction*, 304 Summary, 304

22: Vascular Rings, *M. R. de Leval and M. J. Elliott* **307**

Surgical Anatomy, 307 *Double Aortic Arch*, 307 *Vascular Compression Associated with a Left-sided Aortic Arch*, 307 *Vascular Compression Associated with Left-sided Aortic Arch and Right-sided Upper Descending Aorta*, 308 *Vascular Compression Associated with Right-sided Aortic Arch*, 308 *Cervical Aorta*, 309 *Pulmonary Artery Sling*, 309 Diagnosis, 309 Surgical Technique, 310 *Double Aortic Arch*, 310 *Vascular Compression Associated with Left-sided Aortic Arch*, 311 *Vascular Compression Associated with Right-sided Aortic Arch*, 311 *Cervical Aorta*, 311 *Pulmonary Artery Sling*, 311 Tracheal Stenosis and Malacia Associated with Vascular Rings, 312 *Assessment of the Airway*, 313 *Management of the Major Airways in Association with PA Sling*, 313 Postoperative Surveillance, 316 Management of Tracheobronchomalacia, 316 Results, 317

23: Anomalies of the Systemic Venous Return, *C. van Doorn and M. R. de Leval* . . **319**

Anomalous Venous Return to the Systemic Venous Atrium, 320 *Anatomy*, 320 *Surgical Technique*, 320 Specific Surgical Situations, 321 *Atrial Switch (Mustard or*

Senning), 321 *Atrio-ventricular Septal Defect*, 321 *Fontan Operation*, 321 Anomalous Venous Return to the Pulmonary Venous Atrium, 322 *Anatomy*, 322 Surgical Technique, 323 *Drainage of the Left-sided Superior Vena Cava to the Left Atrium*, 323 *Unroofed Coronary Sinus without Left-sided Superior Vena Cava*, 324 *Drainage of Inferior Vena Cava or Hepatic Veins to the Left Atrium*, 325 Anomalous Venous Return to Both Atria, 325 Combined Anomalous Systemic and Pulmonary Venous Return, 325 Results, 326

24: Total Anomalous Pulmonary Venous Return and Cor Triatriatum, *V. T. Tsang and J. Stark* **327**

Total Anomalous Pulmonary Venous Return, 327 Diagnosis, 327 Indications, 327 Surgical Technique, 328 *Vertical Vein*, 329 *Supracardiac Type*, 329 *Cardiac Type*, 331 *Infracardiac Type*, 332 *Mixed Type*, 334 Early Complications, 335 *Pulmonary Oedema*, 335 *Pulmonary Hypertensive Crises*, 335 *Phrenic Nerve Damage*, 336 Late Complications, 336 *Pulmonary Venous Obstruction*, 336 *Anastomotic Stricture*, 336 *Pulmonary Venous Stenosis*, 336 Repair of Pulmonary Venous Obstruction (PVO), 336 *Congenital Stenosis of Pulmonary Veins*, 336 *Heterotaxy*, 337 Results, 337 *After Primary Repair*, 337 *Results after Reoperations for Pulmonary Venous Obstruction (PVO)*, 338 Cor Triatriatum, 338 Diagnosis, 338 Indications, 339 Surgical Technique, 339 Results, 339

25: Secundum Atrial Septal Defect and Partial Anomalous Pulmonary Venous Return, *J. Stark and V. T. Tsang* **343**

Diagnosis, 343 Indications, 343 Types of Atrial Septal Defect (ASD), 344 Types of Partial Anomalous Pulmonary Venous Drainage, 344 Surgical Technique, 345 *Direct Closure*, 345 *Patch Closure*, 346 Techniques for Special Conditions, 347 *Sinus Venosus Defect*, 347 *Repair of Coronary Sinus Atrial Septal Defect*, 347 *Repair of Partial Anomalous Pulmonary Venous Return*, 347 *Repair of Drainage from the Right Pulmonary Veins to the Right Atrium*, 348 *Repair of Drainage from the Right Pulmonary Veins to the Inferior Vena Cava (Scimitar Syndrome)*, 349 *Anomalous Drainage from the Left Lung into the Innominate Vein*, 351 ''Residual'' ASD, 352 Complications, 352 *Late Pericardial Effusion and Tamponade*, 352 *Arrhythmias*, 352 Results, 352 *ASD Closure*, 352 *Closure via Right Thoracotomy*, 352 *Device Closure of ASD*, 353 *Repair of Sinus Venosus Defect*, 353 *Repair of Scimitar Syndrome*, 353

26: Ventricular Septal Defects, *C. van Doorn and M. R. de Leval* **355**

Diagnosis, 355 Indications, 355 *Large VSD*, 355 *Moderately Large VSD*, 356 *Small to Moderate VSDs*, 356 *Doubly Committed Subarterial VSDs*, 356 Surgical Techniques, 356 *Closure through the Right Atrium*, 357 *Closure through the Right Ventricle*, 364 *Closure through a Great Artery*, 366 *Closure through the Apex*, 369 *Ventricular Septal Defect and Aortic Arch Anomalies*, 369 *Trans-catheter Device Closure of Ventricular Septal Defects*, 370 Results, 370 *Residual VSD*, 370 *Conduction Disturbances*, 370 *Hospital Mortality*, 370 Long-term Results, 370

27: Atrio-ventricular Septal Defects, *A. D. Pacifico* **373**

Diagnosis, 374 Partial Form, 374 *Indications*, 374 *Surgical Technique*, 375 *Results*, 377 *Complications and Late Results*, 377 Complete Form, 377 *Indications*, 377 *Surgical Technique*, 378 *Results*, 384 *Complications and Late Results*, 384

28: Right Ventricular Outflow Tract Obstruction with Intact Ventricular Septum, *C. van Doorn and M. R. de Leval* **387**

Anatomical Substrate, 387 *Pulmonary Valve*, 387 *Pulmonary Arteries*, 387 *Right Ventricle and Tricuspid Valve*, 387 *Right Atrium*, 388 *Coronary Arteries*, 388 Principles of Management, 388 *Symptomatic Neonate with Critical Pulmonary Stenosis or Pulmonary Atresia*, 389 *Definitive Repair of Patients with Pulmonary Atresia with Intact Ventricular Septum*, 390 *Repair of Right Ventricular Outflow Tract Obstructions in Children*, 391 Operative Techniques, 392 *Pulmonary Valvotomy/Valvectomy under Inflow Occlusion*, 392 *Relief of Right Ventricular Outflow Tract Obstruction on Cardiopulmonary Bypass*, 392 *Repair of Supravalvar Pulmonary Stenosis*, 396 Results, 397 *RVOTO with Normal Sized Right Ventricle*, 397 *Pulmonary Atresia with Intact Ventricular Septum*, 397

29: Tetralogy of Fallot, *E. L. Bove and J. C. Hirsch* **399**

Diagnosis, 400 Indications for Operation, 401 Surgical Technique, 402 *Anaesthesia*, 402 *Surgical Technique*, 402 Special Circumstances, 406 *Pulmonary Artery Abnormalities*, 406 *Pulmonary Atresia with Multiple Aortopulmonary Collateral Arteries (MAPCAs)*, 407 *Tetralogy of Fallot with Complete Atrio-ventricular Septal Defect*, 407 Intensive Care Unit Management, 407 Results, 407 Late Complications, 408

30: Pulmonary Atresia and Ventricular Septal Defect, *W. J. Brawn and M. R. de Leval* **411**

General Considerations, 411 Morphology of the Intra Pericardial and Intra-pulmonary Arteries, 411 Investigations, 412 Pulmonary Atresia and VSD, Confluent Normal-sized or Slightly Hypoplastic Central Pulmonary Arteries, 413 *Surgical Techniques*, 413 Pulmonary Atresia with Markedly Hypoplastic Central Pulmonary Arteries without MAPCAs, 414 Pulmonary Atresia with VSD and MAPCAs, 416 *Morphology of the Pulmonary Arteries and MAPCAs*, 416 General Considerations, 417 *Surgery in Infants and Young Children*, 417 *The Young Adult with Pulmonary Atresia, VSD and MAPCAs*, 419 Surgical Comments, 419 *Can the VSD be Closed?*, 420 Postoperative Management and Problems, 420 Combined Management with Surgery and Interventional Cardiology, 421 Valved Conduits and Patches, 421 Results, 421 *Pulmonary Atresia and VSD*, 421 *Pulmonary Atresia, VSD and MAPCAs*, 421

31: Absent Pulmonary Valve Syndrome, *M. J. Elliott* **425**

Diagnosis, 425 Indications for Surgery, 426 Surgical Technique, 427 *Preparation for Repair*, 427 *Repair of Intracardiac Defects*, 428 *Approaches to Repair the Pulmonary Arteries*, 428 Complications, 431 Results, 431

32: Double-outlet Right Ventricle, *V. T. Tsang and J. Stark* **435**

Diagnosis, 436 Indications, 436 Surgical Technique, 436 *Double-outlet Right Ventricle with Subaortic Ventricular Septal Defect without Pulmonary Stenosis*, 436 *Double-outlet Right Ventricle, Ventricular Septal Defect and Pulmonary Valvar or Subvalvar Stenosis*, 437 *Double-outlet Right Ventricle with Doubly-committed Ventricular Septal Defect*, 438 *Double-outlet Right Ventricle with Subpulmonary Ventricular Septal Defect (Taussig–Bing Type)*, 438 *Double-outlet Right Ventricle with Uncommitted Ventricular Septal Defect*, 439 Complications, 441 Results, 441 Double-outlet Left Ventricle, 442

33: Mustard Operation, *J. Stark* **443**

Diagnosis, 443 Indications, 443 Surgical Technique, 443 Results, 448 Complications, 448 Re-operations after Mustard Operation, 448 *Right Thoracotomy Approach*, 448

34: Senning Operation, *A. D. Pacifico* **451**

Indications, 451 Surgical Technique, 451 *Systemic Venous Pathway*, 452 *Pulmonary Venous Pathway*, 454 Results, 456 Late Results and Complications, 456

35: Surgery for Complex Transposition, *V. T. Tsang and J. Stark* **459**

TGA and VSD, 459 TGA and Pulmonary Vascular Obstructive Disease, 460 *TGA, VSD and Pulmonary Vascular Disease*, 460 *TGA, Intact Ventricular Septum and Pulmonary Vascular Disease*, 460 TGA, VSD and LVOTO, 461 *Rastelli Operation*, 462 *REV Procedure (''Reparation á l'Etage Ventriculaire'')*, 465 *Aortic Translocation*, 466 Results, 466 TGA, LVOTO and Intact Ventricular Septum, 466 *LV–PA Conduit*, 467 Results, 468

36: The Arterial Switch Operation, *R. B. B. Mee* **471**

Indications for the Arterial Switch Operation, 472 *TGA and VSD or Double-outlet Right Ventricle with Subpulmonary VSD*, 472 *Simple TGA*, 472 *Double-inlet Univentricular Heart of LV Type, TGA with Severe Subaortic Stenosis, Coarctation, Severe Arch Hypoplasia, or Interruption with Duct-dependent Descending Aortic Flow in Neonates*, 473 *RV Dysfunction and Tricuspid Incompetence after Mustard or Senning Operation*, 473 *Progressive Dynamic LVOTO after Inflow Atrial Repair*, 474 *Patients with Congenitally Corrected TGA and VSD, Unrestricted Pulmonary Blood Flow with or without Ebstein's Anomaly of the Morphological RV and Tricuspid Valve*, 474 Management Methodology for Arterial Switch, 474 *Diagnosis*, 474 *Preoperative Preparation*, 474 *Anaesthesia*, 475 *Surgical Exposure*, 475 *Cannulation*, 475 *Perfusion*, 475 *Cardioplegia*, 476 Surgical Technique, 476 *Coronary Artery Translocation*, 477 *The Neo-aorta*, 479 *Repair of Aortic Sinus Defect*, 480 *Intracardiac Repair*, 481 *Completing the Operation*, 481 *Bleeding*, 482 *Chest Closure*, 482 Postoperative Management, 482 *General*, 482 *The Peritoneal Dialysis Catheter*, 482 Results, 483 *Indication 1: Complex TGA and DORV*, 483 *Indication 2: Simple TGA, Including Some with Small or Tiny VSDs that Were Not Closed*, 484 *Indication 3: Double-inlet Ventricle or LV Type with TGA and Severe Subaortic Stenosis, Severe Arch Hypoplasia and Coarctation of the Aorta, or Interrupted Aortic Arch*, 485 *Indications 4 and 5: Conversion of Previous Mustard and Senning Operations to Arterial Switch*, 486 *Indication 6: Congenitally Corrected TGA with Morphological RV Dysfunction and Tricuspid Valve Incompetence*, 486

37: Surgery of the Left Ventricular Outflow Tract, *V. T. Tsang and M. R. de Leval* . **489**

Anatomical Substrate, 489 *Normal Left Ventricular Outflow Tract*, 489 *Abnormal Left Ventricular Outflow Tract*, 490 Clinical Presentation and Diagnosis, 491 Indications for Operation and Interventions, 492 Surgical Techniques, 493 *Valvotomy in Neonates and Infants*, 493 *Valvotomy in Childhood*, 493 *Aortic Valve Repair in Children*, 494 *Aortic Valve/Root Replacement in the Young*, 494 *Aortic Root Replacement*, 498 *Relief of Subvalvar Aortic Stenosis*, 502 *Small Aortic Roots and Diffuse Left Ventricular Outflow Tract Obstructions*, 504 *Relief of Supravalvar Aortic Stenosis*, 510 Results, 511 *Aortic Valvar Stenosis in Neonates*, 511 *Aortic Valve Repair*, 511 *Aortic Valve Stenosis in Children*, 511

38: Persistent Truncus Arteriosus, *M. R. de Leval* **515**

DiGeorge's Syndrome, 516 Diagnosis and Indications for Surgery, 516 Surgical Techniques, 517 *Repair of Truncus Arteriosus*, 517 Special Considerations, 520 *Repair of Truncus Arteriosus with Truncal Valve Regurgitation*, 520 *Repair of Truncus Arteriosus with Interrupted Aortic Arch*, 520 *Repair of Truncus Arteriosus with Discontinuous Pulmonary Arteries*, 521 *Repair of Truncus Arteriosus with Prior Banding of the Pulmonary Arteries*, 522 Postoperative Management, 522 Results, 523

39: Atrio-ventricular Discordance and Congenitally Corrected Transposition, *R. B. B. Mee and J. Stark* **525**

Diagnosis, 525 Indications, 526 Surgery for Congenitally Corrected Transposition of the Great Arteries, 526 *Surgical Technique*, 526 *Systemic-to-Pulmonary Artery Shunting*, 527 *Left Ventricular Outflow Tract Obstruction (mLVOTO)*, 527 *VSD Closure*, 527 *Persistent Morphological Left Superior Vena Cava (mLSVC)*, 528 *Separate Drainage of the Hepatic Veins*, 530 *Reconstruction of Distorted Pulmonary Arteries*, 530 *The RV–PA Conduit*, 530 *Arterial Switch Procedure*, 531 *The Senning Operation with Modifications*, 531 Surgical Preparation and Method, 535 Atrio-ventricular Discordance with Double-outlet Right or Left Ventricle, 537 Isolated Ventricular Inversion, 538 Results, 538 *Physiological Repair*, 538 *Anatomical Repair (Double Switch)*, 538 Late Results, 539

40: Single Ventricle, *C. van Doorn and M. R. de Leval* **543**

Historical Note, 543 Indications for the Fontan Operation, 544 Selection Criteria for Fontan Operation, 544 Surgical Preparation for the Fontan Circulation, 545 *Preparation for the Fontan Circulation in the Neonatal and Early Infancy*, 545 Physiology of Right Heart Bypass Operations, 545 *Atriopulmonary Connection*, 547 *Total Cavopulmonary Connection (TCPC)*, 547 Surgical Techniques, 547 *Superior Cavopulmonary Anastomosis*, 547 *Total Cavopulmonary Connection*, 548 Additional Procedures, 551 *Fenestration of the Intra-atrial Baffle*, 551 *Relief of Subaortic Stenosis*, 552 TCPC in Isomeric Hearts, 553 Results, 553 *Death before or Unsuitability for the Fontan Procedure*, 553 *Outcome after the Fontan Operation*, 553 Management of the Failing Fontan, 555

41: Hypoplastic Left Heart Syndrome, *C. Pizarro and W. I. Norwood* **559**

Diagnosis, 560 Preoperative Considerations, 561 Norwood Procedure (Stage One), 562 *Anaesthetic Considerations*, 562 *Surgical Technique*, 563 Hemi-Fontan (Stage Two), 567 Modified Fontan Procedure, 568 Results, 569

42: Congenital Malformations of the Mitral Valve, *A. Carpentier and C. Brizard* . . **573**

Clinical Presentation, 573 *Mitral Valve Stenosis*, 573 *Mitral Valve Regurgitation*, 573 Preoperative Valve Analysis. The Functional Classification, 573 Surgical Technique, 574 *Approach*, 574 *Cardiopulmonary Bypass and Myocardial Protection*, 574 *Intraoperative Valve Analysis and Repair*, 574 Mitral Valve Stenosis, 575 *Congenital Mitral Valve Stenosis with Normal Papillary Muscles*, 575 *Mitral Valve Stenosis with Abnormal Papillary Muscles*, 576 Mitral Valve Regurgitation, 578 *Normal Leaflet Motion*, 578 *Leaflet Prolapse*, 580 *Restricted Leaflet Motion*, 581 Assessment of Valve Repair, 583 Valve Replacement, 583 Congenital Mitral Valve Anomalies in Newborns and Infants, 583 *Anatomy*, 584 *Clinical Presentation and Investigations*, 584 *Medical Treatment*, 584 *Surgical Treatment*, 584 Results, 585 Conclusion, 588

43: Malformations of the Tricuspid Valve and Ebstein's Anomaly, *A. Carpentier and C. Brizard* **591**

Congenital Tricuspid Valve Stenosis, 591 Congenital Tricuspid Valve Regurgitation, 591 Ebstein's Anomaly, 591 Surgical Anatomy, 592 Diagnosis, 593 Presentation, 593 Medical Management, 593 General Operative Considerations, 594 Cardiopulmonary Bypass and Myocardial Protection, 594 *Assessment of Repair*, 594 *Indications*, 594 Surgical Technique, 595 Results, 597

44: Aortopulmonary Window, *J. Stark and V. T. Tsang* **599**

Diagnosis, 599 Indications, 599 Surgical Technique, 599 *AP Window Associated with Interrupted Aortic Arch*, 601 Results, 601

45: Congenital Anomalies of Sinuses of Valsalva and Aorticoventricular Tunnel, *V. T. Tsang and J. Stark* **603**

Aneurysm and Fistula of Sinus of Valsalva, 603 Diagnosis, 603 Surgical Treatment, 603 *Results*, 605 Aortico–Left Ventricular Tunnel, 605 *Diagnosis*, 605 Indications, 606 *Surgical Technique*, 606 *Results*, 606

46: Congenital Coronary Artery Fistula and Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery, *V. T. Tsang and J. Stark* **609**

Congenital Coronary Artery Fistula, 609 *Diagnosis*, 609 *Surgical Technique*, 610 *Results*, 612 Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA), 612 *Physiology and Diagnosis*, 612 *Indications for Operation*, 613 *Surgical Technique*, 613 *Results*, 617

47: Surgery for Arrhythmias, *Y. Ishii, J. L. Cox and R. J. Damiano* **619**

Surgical Anatomy, 619 Electrophysiological Evaluation, 622 *Preoperative Mapping*, 622 *Intraoperative Mapping*, 623 Surgical Techniques, 623 *Supraventricular Tachyarrhythmias Atrial Fibrillation*, 623 Indications, 623 Surgical Technique of the Cox–Maze III Procedure, 624 *Accessory Atrio-ventricular Connections in Infants and Children*, 627 Ventricular Tachyarrhythmias after Repair of Congenital Heart Defects, 631

48: Paediatric Heart and Heart–Lung Transplantation, *J. Wallwork, A. R. Hosseinpour and M. Burch* **635**

Heart Transplantation, 635 Indications, 635 Contraindications, 636 Preoperative Evaluation, 636 Infant Transplantation and ABO Mismatch, 637 Pretransplantation Management, 637 Surgical Techniques, 638 *Donor Considerations*, 638 *Donor Heart Excision*, 638 *Recipient Operation*, 639 Technical Aspects in Complex Congenital Heart Defects, 640 *Pulmonary Venous Return*, 641 *Systemic Venous Return*, 641 *Atrial Morphology*, 641 *Ascending Aorta and Aortic Arch*, 641 *Main Pulmonary Artery and Its Two Main Branches*, 642 *Spatial Relationship between the Aorta and the Pulmonary Artery*, 642 Postoperative Management, 642 *Immunosuppression*, 642 *Surveillance and Treatment of Acute Rejection*, 643 *Cardiac Allograft Vasculopathy*, 643 *Infection*, 644 *Lymphoproliferative Disease and Tumours*, 644 Outcome, 644 *Survival*, 644 *Morbidity*, 644 Heart–Lung Transplantation, 646 Selection of Patients, 646 Donors, 646 *Donor Procurement*, 646 Recipient Operation, 647 *Explantation*, 647 *Implantation*, 648 Postoperative Management, 649 *Diagnosis of Acute Rejection*, 649 *Infection*, 650 Outcome, 650 *Chronic Rejection*, 650 The Future, 650

49: Paediatric Lung Transplantation, *J. J. Nigro, R. D. Bart and V. A. Starnes* . . . **653**

Indications, 653 Contraindications, 655 Psychosocial Evaluation, 656 Types of Lung Transplants, 656 *Heart–Lung Transplant*, 656 *Bilateral Lung Transplant*, 657 *Single Lung Transplant*, 657 *Living Lobar Lung Transplant*, 657 Donor Selection, 658 Organ Procurement, 658 Graft Preparation, 659 Living Lobar Lung Transplantation, 660 *Technique*, 660 Recipient Operation, 661 *Anaesthesia and Positioning*, 661 *Cardiopulmonary Bypass*, 662 *Operative Technique*, 662 *Living Related Lobar Transplantation*, 664 Postoperative Management, 664 Immunosuppression, 664 Complications, 665 *Acute Rejection*, 665 *Infection*, 666 Outcomes, 666 Summary, 667

50: Miscellaneous: Straddling Atrio-ventricular Valves, Pericardium, Tumours, Diverticula and Ectopia Cordis, *J. Stark and V. T. Tsang* **669**

Straddling Atrio-ventricular Valves, 669 *Surgical Implications*, 670 *Results*, 670 Pericardium, 671 *Congenital Defects*, 671 *Cysts*, 671 *Tumours*, 671 *Tamponade*, 671 *Pericarditis*, 672 Tumours, 674 Surgical Technique, 674 Diverticula, 675 Ectopia Cordis, 676

51: Data Collection and Assessment of Performance in Congenital Heart Surgery, *S. Gallivan, J. Stark, N. Kang and V. T. Tsang* **679**

Introduction, 679 Issues Associated with Data Collection, 679 *Golden Rules of Data Collection Studies*, 679 *Specific Purposes for Collecting Data Relating to CHS*, 680 Issues Associated with the Scale of Data Collection Related to CHS, 680 Nomenclature and Mapping, 680 Errors and Database Validation, 681 Existing Systems, 682 Risk Stratification, 682 *The Rationale for Risk Stratification*, 682 Categorizing Congenital Heart Operations According to Their Complexity, 683 Assessment of Outcomes, 684 *Graphical Methods for Assessing Outcome*, 684 Limitations of Risk Modelling, 686

Index **689**

Preface to the Third Edition

The second edition of *Surgery for Congenital Heart Defects*, published in 1994, has been out of print for some time. We did not think that enough changes had occurred during the past 10 years to warrant another edition. However, our trainees and attendants of courses, particularly those attending the European School of Cardiothoracic Surgery in Bergamo, Italy, were urging us to do so.

When we reviewed the progress of the past decade, many new diagnostic techniques had been introduced, several aspects of anaesthesia, perfusion and postoperative care had been refined and a number of operative procedures had been modified, with emphasis on complex small infant surgery. For all these reasons we agreed to update the last edition.

Some chapters are completely new, such as those on MRI and CT scanning, interventional catheterization, electrophysiology of arrhythmias, paediatric cardiac mechanical support, lung transplantation and assessment of surgeons' performance. The last chapter was added to help readers to understand the difficulties and pitfalls of data collection and the importance of accurate data for everyday work.

All other chapters were revised to reflect the current practices of the editors and leading specialists in the field of congenital heart disease contributing to this edition. Major changes and updates were made in the chapters on echocardiography, invasive investigation and exercise testing, anaesthesia, postoperative care, vascular rings, anomalous pulmonary venous return, tetralogy of Fallot, pulmonary atresia with ventricular septal defect, surgery of left ventricular outflow tract atrio-ventricular discordance, hypoplastic left heart syndrome and paediatric heart and heart lung transplantation.

The format of the book follows that of the two previous editions. The emphasis remains on the details of operative procedures. Diagnostic criteria are briefly discussed in each chapter. Results are summarized for completeness. Fortunately the same artist, Michael Courtney, provided most of the additional illustrations. In the general section, readers should find information about nomenclature, anatomy, diagnosis, including invasive investigation, exercise testing, echocardiography and foetal echocardiography, anaesthesia, perfusion, postoperative care, pacing, electrophysiology, cardiac mechanical support and approaches to the heart. In the section on surgical procedures, we have kept some classical operations, even though some of them are currently performed only rarely or even exceptionally. Because of the rare use of these procedures, it is not easy for young surgeon to familiarize themselves with such procedures during their training.

We hope that this updated edition will again be helpful to trainees in paediatric cardiac surgery and allow them to start their independent careers without learning curves. It may also assist established surgeons when dealing with rare defects. Paediatric cardiologists, anaesthetists, intensivists, nurses and technicians may find the book a useful source of information.

Jaroslav F. Stark
Marc R. de Leval
Victor T. Tsang

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We are grateful to all the contributors for their outstanding work. We are particularly indebted to Michael Courtney, whose illustrations are such an integral part of the book.

We would like to thank our research secretary Faith Hanstater, and in particular our secretary Juliet Aghion for her help with the manuscripts, organization of all the details of the work and her general support. Our researcher, Katy Burke, helped greatly with the sourcing and organisation of the literature.

We thank our team of cardiologists, anaesthetists, intensivists, nurses, physiotherapists and technicians, who have participated with us over the years in the care of our patients.

Finally, we are extremely grateful to Haymans Trust for their generous financial support, which helped with the secretarial support and additional illustrations required for this up-dated edition.

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Foreword to the Third Edition

The Hospital for Sick Children at Great Ormond Street (GOS) in London, England (currently Great Ormond Street Hospital for Children) has been at the forefront of paediatric cardiac surgery since its inception. Throughout much of GOS history, two of its surgeons, Jaroslav F. Stark and Marc R. de Leval, have contributed enormously to the clinical practice, research and teaching of congenital heart surgery.

Their textbook *Surgery for Congenital Heart Disease* was first published in 1983, was revised in 1994, and again revised in 2006 as this 3rd edition. The book has become a reliable and trusted friend. It has a proven ''track record'' and contains sage and practical advice for the present and future progress of our specialty.

The original publication grew from the authors' teaching sessions. In the preface to the 1983 edition, they state; ''*The successful outcome of many operations depends to a large extent on the careful planning and precise execution of a series of minor technical steps*''. Stark and de Leval provide a recipe for successful surgical outcomes by clearly defining the technical details that are often overlooked or assumed in other textbooks. In the current edition, Stark and de Leval have added a new editor, their younger colleague Victor Tsang, who continues their tradition of excellence in clinical surgery and clear unambiguous description of the essence of surgical care for children with congenital heart disease.

Core authorship of the book is the outstanding faculty at GOS. In addition, selected colleagues outside GOS have provided contributions in their areas of special expertise, thereby adding to the book's international scope and authority. The format of each chapter is consistent with emphasis on the practical. The artwork by Michael Courtney complements the text and greatly facilitates comprehension of the essential operative details which are the stated mission of the book.

The past few decades have produced remarkable improvements in the care for patients with congenital heart disease. Newborns with cardiac lesions that were rapidly fatal when the two previous editions of this textbook were conceived are now successfully palliated into adulthood. The functional ability and quality of life for all of our patients is improving. Yet much remains to be accomplished and will be, as we incorporate new science and technology into surgical practice. The essence of good surgical practice will remain the same and is clearly described in this text. The GOS tradition of excellence in clinical surgery and teaching lives on in this 3rd edition of *Surgery for Congenital Heart Disease*. It is a privilege to review the final draft of this valuable contribution to the care of children with congenital heart disease.

WILLIAM G. WILLIAMS, MD, FRCSC
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Foreword to the Second Edition

This second edition of the masterful *Surgery for Congenital Heart Defects* is a worthy successor to the first edition of that great work, which appeared in 1983. Drs Stark and de Leval have designated themselves 'editors,' but in fact they clearly are both the stimuli for the second edition and the main contributors to it. In fact, this important book could be considered a detailed summary of all aspects of the experience of their prestigious unit, The Hospital for Sick Children at Great Ormond Street, London, with congenital cardiac disease over a period of at least 40 years. This alone recommends this text to all serious students and practitioners of the art and science of managing patients with these conditions. The fact that the book describes far more than simply operations and their results emphasizes the multidisciplinary nature of all serious endeavors in this area; good surgery alone can no longer suffice as evidence of institutional expertise in the area of congenital cardiac disease.

The book is clearly more broadly based than it would be had it been written only by the staff at the Great Ormond Street hospital. In a sense it could be considered a contribution of London, England, to the expanding knowledge of all aspects of congenital heart disease; the major contributions from this area over a period of many years also make the text invaluable. In addition, however, Drs Stark and de Leval have selectively tapped the resources of the rest of the medical world in developing this superb text.

This book becomes available at a particularly important and probably pivotal time in the area of congenital heart disease. In some parts of the world, there now exists the capability of eliminating congenital heart disease from the population, using prenatal echocardiographic diagnosis and abortion of an affected fetus. Ironically, this comes at a time when the results of many types of interventions for congenital cardiac disease are outstandingly good. For some types of congenital heart disease, literal cures by surgery have been documented. In contrast, it is now known that some conditions (for example, coarctation of the aorta) that seemed to be cured by intervention are only palliated, but this palliation may last for 50 years or more. Certain congenital cardiac anomalies that only a few years ago were essentially untreatable, such as interrupted aortic arch and aortic atresia, now can be at least extremely well palliated by one of several methods, and this palliation can be expected to last for a number of years even though not for the usual lifetime of a normal individual. In view of the diversity in the time-related quality of the outcomes, the variety of the interventions available for various types of congenital heart disease, and the many decisions that currently must be made in managing patients with congenital heart disease, it now becomes essential to quantify and compare outcomes. Also, it may become necessary somehow to pick and choose between institutions because of the growing suspicion that a concentration of relatively large numbers of such patients in relatively few institutions may improve outcomes. This again should involve quantification of outcomes, sophisticated methods of analyses, and factual (rather than simply political) comparisons of institutions and physicians.

Were this not enough to justify the extensive material presented in this text, society in general has added quality of care, appropriateness of care, effectiveness of care, and cost of care to its jargon of qualifications to be met by all of us in our hospital wards, laboratories, operating rooms, and intensive care units. It is this, plus the alleged imperfections in the outcomes we obtain and their high costs, along with the changing lifestyles and philosophies of our times and the strong emotions that accompany all discussions of health care (let alone those of abortion, nonuseful extension of life, and physician-assisted death), that makes so necessary the gathering together of the varied and extensive information presented in this second edition of *Surgery for Congenital Heart Defects*.

I look forward to detailed study of each of the wonderful chapters that have been presented in this book. Indeed, I recommend such study to everyone involved with decisions and management for that part of our populations afflicted with congenital heart disease.

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Foreword to the First Edition

The entire discipline dealing with the surgical treatment of congenital heart disease is unique in retaining much of its original awesomeness. Perhaps this is because the act of opening a child's chest to manipulate in or about the heart still evokes the feeling that this is an incredibly impertinent thing to do. Even after all these years, the act seems to fly in the face of that which is natural and possible. Perhaps it is because of the great gravity of the procedure, the sometimes narrow margin between failure and success, with an entire unlived lifetime in the balance. Or perhaps it is because the surgical capability to invade the heart developed more recently than for any other organ—indeed, within the professional lifetime of some of its still-pioneering pioneers.

It is unquestionably true of each of the surgical disciplines that success or failure is related to the availability of fundamental knowledge, advanced technology and artistic skill. But perhaps those who are involved in surgery of congenital heart defects could be forgiven for observing that this axiom relates most particularly of all to this discipline. The structural complexity and functional intricacy of the cardiovascular system are exceeded by only one other, the central nervous system, which is so exalted in these respects as to remain relatively intractable to all but ablative surgical interventions. Of all other systems, with regard to complexity, none seem to compare embryologically, anatomically, physiologically, or electronically with the heart. And within the cardiovascular system itself, the subclass of conditions which has a congenital aetiology greatly surpasses other forms of heart disease with respect to multiplicity of pathological presentations, diversity of combinations of associated lesions, and requirements for ingenuity and precision of surgical technique and patient care.

The congenitally deformed heart poses special obstacles to corrective operation. The necessity for growth is particularly important, especially when prosthetic valves or conduits are required, and true also for cardiac transplantation or perhaps ultimately the use of an artificial heart. Pertinent also, is the problem of secondary effects of the congenital anomaly on pulmonary or peripheral vasculature, or on the myocardium itself.

A situation *par excellence* which demonstrates the necessity of sophisticated teamwork in modern medical practice is provided by groups which are successful in the management of patients with congenital heart disease. Collaboration, coordination, communication, cross-fertilization and mutual esteem should be in evidence among participants of this team, which includes a cardiologist, radiologist, physiologist, pathologist, anaesthetist, nurse, technician, surgeon and several other partners. Perhaps as well as being the most ''incisive'', the surgeon's role in this team is also highly decisive. It is necessarily upon the surgeon's shoulders that the principal burden of the operative intervention must primarily rest. He or she must feel the weight of having accepted from the parent that strongest of all possible human obligations—the profound, primordial, instinctive identification and responsibility for an offspring. Surely no more depth of sincerity can be imagined than that expressed in the ''Thank you'' of a parent for a safe and successful surgical endeavour, nor more pain, frustation and despair if the outcome should be the opposite. By virtue of the complexity of the many problems to be dealt with, the precision required during all phases of patient care, and the immense parental trust resting upon them, the team dealing with congenital heart disease sometimes finds the demands on reserves of physical, emotional and spiritual strength almost overwhelming. Yet the incomparable sense of reward which is frequently achieved compensates and justifies all.

Some 40–45 years are all that have been required for progression from that first adventure of cardiac surgery to the present when virtually every anomaly is amenable to some form of surgical intervention. During this time, the advance of one ingenious approach after another has stirred excitement and expectation, and the prospect of further refinements and innovations continues. However, at this point, one senses that a landmark may have been

reached. Techniques, approaches, results and remaining issues of concern all seem to have reached a tentative landing on the staircase of progress. This pause presents, perhaps for the first time, a splendid opportunity to survey the subject broadly and thoroughly. The authors of this book are well suited to the task from the standpoint of their multi-disciplinary diversity, their uniformly superb qualifications and their substantial individual contributions to this body of knowledge. The challenge of condensing the mass of information surrounding each aspect and anomaly, while retaining comprehensiveness, has, I think, been expertly accomplished.

It gives me honour to have the opportunity to present these reflections as a preliminary to this fine text. It gives me pleasure to congratulate all of those who contributed in either small or large measure to the remarkable achievements to which it attests. To its authors, I heartily extend my respect and gratitude.

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Abbreviations

This is a list of abbreviations which occur frequently in the text. Those less commonly used are explained where they occur.

Ao	aorta
A-P	aorto-pulmonary
ARF	acute renal failure
ASD	atrial septal defect
A-V	atrio-ventricular
BSA	body surface area
CHD	congenital heart defect
CPAP	continuous positive airway pressure
CS	coronary sinus
CVP	central venous pressure
DAo	descending aorta
FiO_2	fraction of inspired oxygen
IPPV	intermittent positive pressure ventilation
ITU	intensive therapy unit
IVC	inferior vena cava
LA	left atrium
LAA	left atrial appendage
LBB	left bundle branch
LCA	left coronary artery
LPA	left pulmonary artery
LSA	left subclavian artery
LV	left ventricle
LVOT	left ventricular outflow tract
LVOTO	left ventricular outflow tract obstruction
MAPCA	major aorto-pulmonary collateral artery
MPA	main pulmonary artery
MV	mitral valve
PA	pulmonary artery
PAPVD	partial anomalous pulmonary venous drainage
PAB	pulmonary artery banding
$PaCO_2$	carbon dioxide tension (partial pressure) in arterial blood
PDA	persistent ductus arteriosus
PEEP	positive end expiratory pressure
PaO_2	oxygen tension (partial pressure) in arterial blood
PV	pulmonary valve
PVOD	pulmonary vascular obstructive disease
PVS	pulmonary valve stenosis
Qp	pulmonary blood flow
Qs	systemic blood flow
RA	right atrium
RBB	right bundle branch
RCA	right coronary artery
RPA	right pulmonary artery
RV	right ventricle
RVOT	right ventricular outflow tract
RVOTO	right ventricular outflow tract obstruction
SVC	superior vena cava
TAPVD	total anomalous pulmonary venous drainage
TGA	transposition of the great arteries
TV	tricuspid valve
V-A	ventriculo-arterial
VSD	ventricular septal defect

Part I
General Considerations

1

Classification and Nomenclature of Congenital Heart Defects*

R. H. Anderson and F. J. Macartney†

In the previous edition of this book, we emphasized that one of the first tasks given by God to Adam was to name the animals. Thus began natural science. The population was studied, and the observation of similarities and differences between individuals permitted groups to be identified. These groups formed the basis of a classification and, once this had been established, nomenclature followed. A fundamental purpose of nomenclature, therefore, in addition to providing names, is to recognize similarities and differences, and to give them weight in proportion to their importance. Superficially, a caterpillar resembles a centipede more than it does a butterfly. As far as classification is concerned, however, the fact that caterpillars turn into butterflies, whereas centipedes do not, is much more important. Caterpillars, therefore, are classified with butterflies. In a subject that developed as rapidly as the surgical correction of congenital cardiac malformations, changes of emphasis as to what is or is not important were inevitable. It followed inexorably that nomenclature would also change, albeit that recent years have seen more consensus amongst differing ''schools of thought''. In this respect, what is fundamental to an embryologist may be of very limited importance to a surgeon. Should each choose to ignore the other's point of view, nonetheless, two different nomenclatures will inevitably emerge, and the already existing failure of communication will be exacerbated. Happily, advances over the period since the appearance of the previous edition have seen welcome agreement in the approach to nomenclature. It is now agreed that, when the overall spectrum of congenital cardiac abnormalities is surveyed, the heart is made up of atrial, ventricular and great arterial segments, each of which can vary independently of the others. Thus, it is identification of the chambers and great arterial trunks that is the foundation of nomenclature of congenital cardiac disease. That fact has been accepted for many years. It was at the next step that divergent opinions tended to appear. The essence of the original segmental approach was to take the relations between and within the segments as fundamental (Van Praagh, 1972). When we sought to improve the segmental approach, we took the stance that relationships were relatively unimportant when compared to the way the segments were joined together (Macartney *et al*., 1976; Shinebourne *et al*., 1976; Tynan *et al*., 1979; Anderson and Ho, 1997). Relations describe the way that cardiac structures are arranged in space. These do not necessarily reflect that fashion in which two structures are joined one to the other, this latter feature being described by us as a ''connection''. Abnormalities of relations may modify the surgical approach, but they do not require surgical treatment. Abnormalities of connections, in contrast, need surgical correction, unless the effects of two abnormal connections cancel each other out. For this reason, the approach emphasized in this chapter is in spirit identical to that put forward by Kirklin *et al*. (1973), although Kirklin and his colleagues did not distinguish specifically between connections and relations. It is now noteworthy that Van Praagh has recognized the way the segments join to each other, albeit that he chooses to describe this feature as ''alignments'' (Van Praagh, 2000). We continue to place our emphasis on the union between the cardiac segments rather than their relations, since this approach

* Editors' note: The editors have decided to use Latin anatomical terminology to maintain uniformity throughout this book. This was done with the agreement of Professors Anderson and Macartney. They wish to emphasize, however, their preference for usage of English rather than Latin words.

† It was the Reverend Professor Fergus Macartney who produced this chapter for the initial editions of this book, and who collaborated with RHA on the revision of the chapter for this edition. Sadly, Fergus died before this edition could be printed. The editors of the book join with the surviving author of this chapter in dedicating these pages to his eternal memory.

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is relevant not only to surgeons but also to embryologists, pathologists, cardiologists and radiologists. In short, it is pertinent to all those involved in the nomenclature of cardiac disease. Once the heart is divided into segments and the junctions between the segments are properly analysed, then most problems previously perceived to be ''complex'' turn out to be rather simple. It is probably fair to say that the surgical correction of complex cardiac abnormalities was held up no more by imperfections in surgical technique than by imprecision in preoperative diagnosis. This, in turn, may have resulted from inadequate application of sequential segmental analysis.

IDENTIFICATION OF THE CARDIAC SEGMENTS

Atrial Situs (Atrial Arrangement) and Morphology

Sequential segmental analysis can only work if structures are defined morphologically (Van Praagh, 1972; Anderson and Ho, 1997). This presents a problem only in relation to nomenclature of ventricles and the atrial chambers. Strict clarity demands that the words morphologically ''left'', or ''right'', ''ventricle'' or ''atrium'' should be used whenever these chambers are discussed and described, but this is somewhat cumbersome. For this reason, throughout this book the term ''left ventricle'' is used to indicate the morphologically left ventricle, and so on. In all other circumstances, left and right are used to indicate position, not morphology. It is the appendages of the atrial chambers, and their relationship to the remaining atrial components, that serve as the guide to the basic viscero-atrial situs. The right atrial appendage is broad and blunt, extending from the orifice of the superior vena cava almost to the orifice of the inferior vena cava. It contains parallel trabeculations, the musculi pectinati, which run all the way round the atrio-ventricular junction, and insert at right angles into a rather thicker muscular band, the crista terminalis, inserting in the fashion of the teeth of a comb (**Figure 1.1**). The left atrial appendage, in contrast, is narrow and pointed, often looking like a crooked finger. The musculi pectinati are confined within the appendage, so that the left atrial vestibule is smooth (**Figure 1.1**). The presence of a morphologically left appendage on one side, and a morphologically right appendage on the other, constitutes lateralization. This can be seen in the situs solitus, or situs inversus. In other situations, however, there can be isomeric situs with duplication of morphologically right or morphologically left structures on each side of the body. Because the spleen is the only organ in the body that is left-sided from its inception, right isomerism tends to be associated with asplenia, while left isomerism is usually accompanied by polysplenia, in which the spleens arise on either side of the dorsal mesogastrium (Van Mierop *et al*., 1972). In right isomerism, both lungs tend to have three lobes, and to be fed by eparterial bronchi, whereas in left isomerism, both lungs tend to be bilobed, and to have hyparterial bronchi. The remainder of the organs is haphazardly distributed; an arrangement often termed visceral heterotaxy, albeit that, taken literally, ''heterotaxy'' describes any departure from the normal (Macartney *et al*., 1978). It is the appendages, nonetheless, that provide the best guide to isomerism within the heart. Thus, when musculi pectinati encircle both atrio-ventricular junctions, there is right isomerism, whereas bilaterally smooth vestibules are indicative of left isomerism (Uemura *et al*., 1995b). The method of distinction between lateralized and isomeric situs should not depend on autopsy. The traditional division of complex malformation with jumbled-up bodily organs into the subsets of ''polysplenia'' or ''asplenia'' is unsatisfactory for a number of reasons. First, there is no real consensus on the distinction between polysplenia and one large spleen plus multiple small spleens (Moller *et al*., 1967; Landing *et al*., 1971; Van Mierop *et al*., 1972). Second, the so-called ''asplenia syndrome'' has been described in the presence of a spleen, albeit rudimentary (Layman *et al*., 1967). Most importantly, there are marked discrepancies in classification when features are analysed in the post mortem room on the basis of splenic morphology, as opposed to the morphology of the atrial appendages (Uemura *et al*., 1995a). If such uncertainties exist at autopsy, they are hardly likely to be clarified by investigation in a living patient. In the living patient, it is simplest to infer the atrial situs having visualized the situs of the tracheobronchial tree by plain chest radiography or bronchial tomography (Van Mierop *et al*., 1970; Partridge *et al*., 1975; Macartney *et al*., 1978). A ratio between the lengths of the two main bronchi that is less than 1.6 indicates isomerism, whereas a

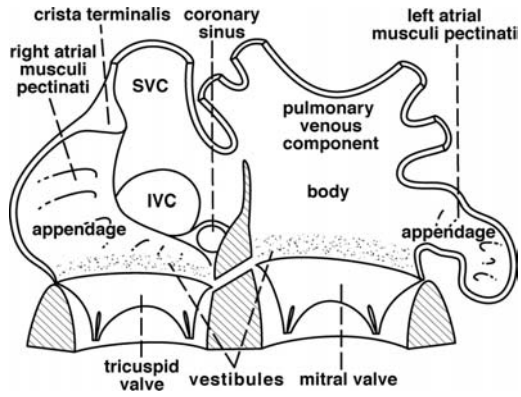

Figure 1.1

ratio higher than 1.6 indicates lateralization, with the morphologically left bronchus being longer. Although discordance between bronchial and atrial situs is also recognized (Caruso and Becker, 1979), the rule of broncho-atrial concordance is accepted as one of the most reliable to be found in paediatric cardiology, given that all rules have exceptions (Macartney *et al*., 1980). Routine cross-sectional echocardiographic scanning of the descending aorta and inferior caval vein also permits accurate inference of atrial arrangement, with the usual situation found when they lie apart with the aorta to the left, mirror imagery when they lie apart but with the aorta to the right, and right isomerism when they run together (Huhta *et al*., 1982). Interruption of the inferior vena cava is suggestive, but not diagnostic, of left isomerism. Because of the lack of complete harmony between the situs of the atrial appendages and the rest of the organs (Uemura *et al*., 1995a), the term ''viscero-atrial situs'' has limited meaning. It is the arrangement of the appendages that is the key to the appropriate description of the atrial segment (Uemura *et al*., 1995b). When in doubt, all systems should be described in independent fashion.

Ventricular Morphology

Because the ventricles develop in series rather than in parallel, there is very limited opportunity for development of ventricular isomerism, although a solitary case has been described with two right ventricles (Rinne *et al*., 2000). The nomenclature of the heart characterized by defective septation, however, has been bedevilled by failure to define precisely what is meant by a ventricle, specifically, how much of a ventricle may be missing before it no longer merits the name ''ventricle'' (Anderson and Ho, 1998). In the past, we argued that chambers lacking an atrial input should be denied ventricular status (Wilkinson *et al*., 1979). We now recognize the futility of this approach (Anderson and Cook, 2004). The key to appropriate analysis is to recognize that normal ventricles have three major components (**Figures 1.2 and 1.3**):

- The inlet component, upstream to the origin of the tensor apparatus.
- The outlet component, or the outflows to the two great arteries.
- The apical trabecular component, found toward the apex of the ventricles.

The septum normally separating the ventricles may be identified by its characteristic tendency to run inferoposteriorly towards the crux of the heart. This external landmark is marked by the intersection of one line formed by the coronary sinus with another formed by the interatrial groove superiorly and the inferior interventricular coronary artery. The septum can also run to the crux, however, when both atrio-ventricular junctions are connected to the morphologically right ventricle (**Figure 1.4**). In some situations, the ventricular septum is malaligned relative to the atrial septum, as in double-inlet left ventricle, and then the ventricular septum runs toward the acute margin of the heart (**Figure 1.4**). A similar situation, with antero-cephalad malalignment of the ventricular septum, is seen in the setting of straddling of the tricuspid valve (Milo *et al*., 1979). In this setting, the tension apparatus of the atrio-ventricular valve is attached to both sides of the

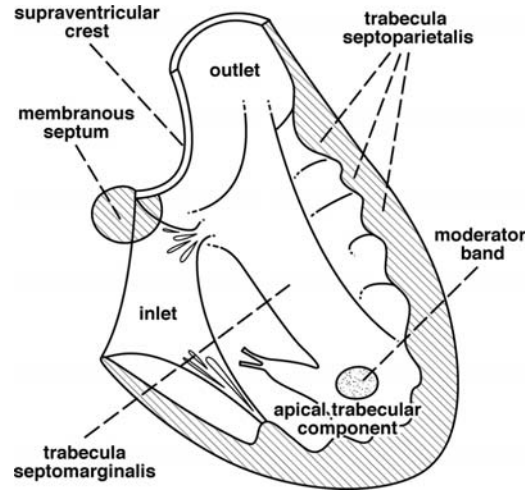

Figure 1.2

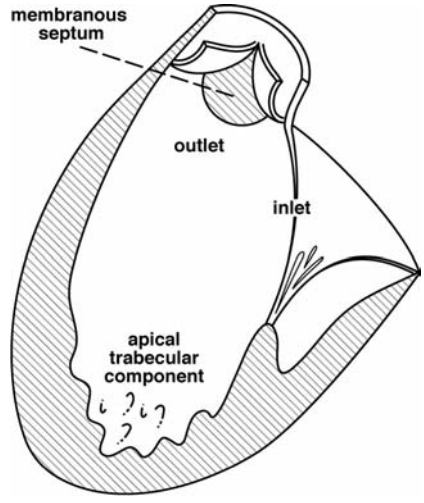

Figure 1.3

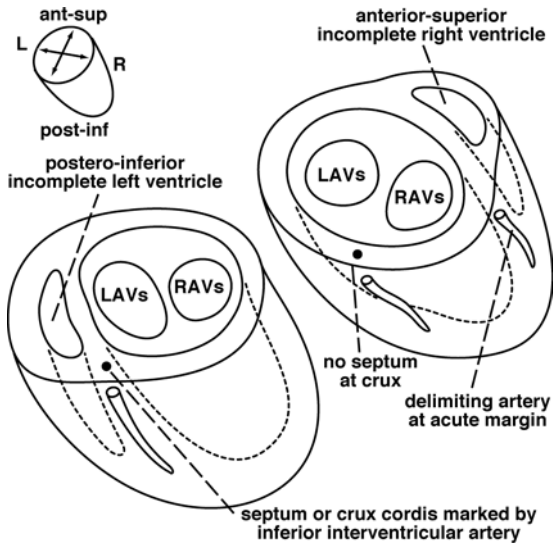

Figure 1.4

septum. The mitral valve can also straddle in this fashion, and straddling is usually associated with overriding of the valvar orifice. On occasion, straddling occurs without overriding, or overriding without straddling (Milo *et al*., 1979). Pathological observations indicate a continuous spectrum of the degree of straddling, from the situation in which one cord merely crosses the septum, to one in which tensor apparatus originates equally from either side. There is a similar spectrum of overriding. In order to determine the atrio-ventricular junctional connections, the valve must be arbitrarily assigned to one ventricle or the other, just as will be done for overriding arterial valves. The valve is assigned to the ventricle that supports the majority of the circumference of the atrio-ventricular junction. Irrespective of these junctional arrangements, however, the presence of valvar overriding has no influence of the apical trabeculations within the ventricles, albeit that the chambers tend to be hypoplastic when they receive only part of an inlet or outlet. In the past, these situations demanded that we change the name of ventricles and rudimentary chambers according to whether or not they possessed an inlet component. Now, we recognize that all chambers within the ventricular mass possessing apical components can be named as ventricles (Anderson and Cook, 2004). They can be of right, left, or indeterminate morphology. Right and left ventricles coexist, with the apical parts separated by the apical septum. This septum carries the conduction system, and is perfused by the septal perforating arteries. Ventricles of indeterminate morphology lack an apical septum, and are the only examples of anatomically single or solitary ventricles.

The apical septum is further characterized by having a morphologically right side that is heavily trabeculated, and a morphologically left side that is finely trabeculated and usually completely smooth in its superior portion. The other important right ventricular characteristic is the presence of a well-developed infundibulum, which interposes between the leaflets of the atrio-ventricular and arterial valves, these leaflets being typically in fibrous continuity in the roof of the left ventricle. This rule, however, has some well-known exceptions. When the atrio-ventricular valves are normally incorporated into the ventricles, a mitral valve, with a solitary zone of apposition between two leaflets, and two papillary muscles not attached to the septum, reliably identifies the left ventricle. A tricuspid valve, with three leaflets, three zones of apposition, and a septal leaflet attached by short cords and multiple discrete papillary muscles to the septum, indicates the right ventricle.

Muscle Bundles Within the Right Ventricle

In the normal heart, the ‘‘supraventricular crest’’ (crista supraventricularis), separates the attachments of the leaflets of the tricuspid and pulmonary valves (**Figure 1.2**). At the point where this crest fuses with the rest of the septum, a small part of muscle can be identified by dissection that interposes between the subaortic and subpulmonary outflow tracts. Without dissection, however, it is not possible to distinguish between these two components. In hearts with anomalies involving the outflow tracts, the two component parts are able to achieve their own identity. Instead of one muscular structure in the ventricular roof, there are two. One, the ventriculo-infundibular fold, separates the attachments of the leaflets of one or both atrio-ventricular valves from the leaflets of one or both arterial valves. The other, the muscular outlet, or infundibular, septum separates the subarterial ventricular outflows to the two great arteries. It is confusing, therefore, to use the term ‘‘crista supraventricularis’’ in an attempt to describe these muscle bundles when the outflow tracts are themselves malformed (Anderson *et al*., 1977). The terms ‘‘parietal band’’ and ‘‘septal band’’, still popular in certain parts of North America, are even more confusing. This is because most surgeons use ‘‘septal band’’ in malformed hearts to describe the septal attachment of the muscular outlet septum. In the normal heart, however, the term is also used to denote an entirely different structure. This is the prominent Y-shaped muscular strap that reinforces the right ventricular septal surface. This strap is best described as the trabecula septomarginalis (Vricella *et al*., 2004). It has two limbs, the postero-inferior, which supports the medial papillary muscle complex or the papillary muscle of the conus, and the

anterosuperior limb, which runs in cephalad direction to support the leaflets of the pulmonary valve. In tetralogy of Fallot, for example, these limbs form a Y-shaped configuration and constitute the inferior rim of the ventricular septal defect. The stem of the Y is the body, which gives off a series of trabeculae septomarginales, one of which, the moderator band, extends to join the anterior papillary muscle of the tricuspid valve (**Figure 1.2**). These considerations are particularly pertinent to tetralogy of Fallot. Thus, in this setting, it is the septal and parietal extensions of the muscular outlet septum that are mobilized to relieve infundibular obstruction, not the ''septal'' and ''parietal'' bands. And the defect is certainly not ''infracristal'', since the components of the normal supraventricular crest have sprung apart as a result of the malalignment between the muscular outlet septum and the rest of the ventricular septum. Because of this septal malalignment, the aortic valve overrides the septal defect, with biventricular connection of the aortic valve (Becker *et al*., 1975).

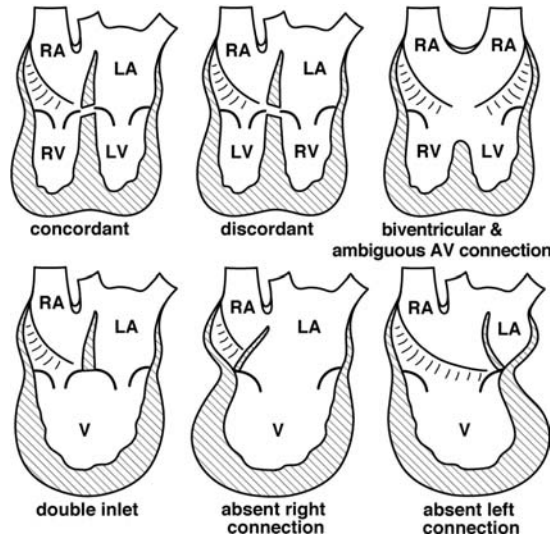

Figure 1.5

Great Arterial Morphology

There is in general little difficulty in distinguishing the two great arteries, in as much as the aorta gives the origin to the coronary and brachiocephalic arteries, while the pulmonary trunk divides into right and left pulmonary arteries. On occasion, one coronary artery or, more rarely, both coronary arteries, can originate from the pulmonary trunk. A brachiocephalic artery may appear to arise from the left or right pulmonary arteries, but in such cases, careful analysis will show that the communication is via a persistently patent ductus arteriosus. No case has thus far been described, to the best of our knowledge, in which there was real difficulty in distinguishing the two great arteries.

JUNCTIONS BETWEEN THE CARDIAC SEGMENTS

Types of Atrio-ventricular Connection

There are several ways in which the atrial chambers can be joined to the underlying ventricular mass (Tynan *et al*., 1979), as illustrated in **Figure 1.5**. We describe the way in which the atrial and ventricular muscles masses are joined together as the ''types of atrio-ventricular connection''. In this respect, it should be noted that the normal heart, and many malformed hearts, possess two atrio-ventricular junctions. Thus, connections are usually plural. Should there be overriding of an atrio-ventricular valve, or the more frequent common atrio-ventricular valve, or rarely both atrio-ventricular valves, the overriding valvar orifice is assigned to the ventricle supporting the greater part of its circumference. When the right atrium drains to the right ventricle, and the left atrium to the left ventricle, the atrio-ventricular connections are said to be *concordant*. When the connections are *discordant*, the right atrium is joined to the left ventricle, and the left atrium to the right ventricle. In the setting of isomeric atrial appendages, then, of necessity, if each atrium is joined to its own ventricle, the connections will be biventricular and *ambiguous*. When both atrial chambers are connected to only one ventricle, there is a *double-inlet* connection. This is the paradigm of the univentricular atrio-ventricular connection, but absence of one atrio-ventricular connection also fulfils the definitions for this final category (Anderson and Cook, 2004). This last condition, therefore, is the only one needing further explanation. It is still frequently not appreciated that the majority of patients with classical tricuspid atresia do not possess an imperforate tricuspid valve, but rather have no tricuspid valve at all, nor any muscular right atrio-ventricular connection (Anderson *et al*., 1979). There is a plane of cleavage containing extracardiac adipose tissue interposed between the right atrium and ventricular mass that can be revealed by blunt dissection in the right atrio-ventricular groove. This plane extends to the central fibrous body, where it achieves continuity with the left atrio-ventricular junction. In this instance, therefore, there is only one atrio-ventricular junction present. The same applies to the left atrio-ventricular groove when the left atrio-ventricular connection is absent. If one atrio-ventricular

connection is *absent*, of necessity there must be a solitary atrio-ventricular valve. In most instances, this solitary valve is exclusively connected to one ventricle, thus producing the second variant of the univentricular atrio-ventricular connection. Infrequently, however, the solitary valve can itself straddle and override. This then produces a uniatrial but biventricular connection, giving one form of so-called ''double-outlet atrium'' (Anderson and Cook, 2004). When describing hearts with absence of one atrio-ventricular connection, it is not always possible to use the adjectives ''tricuspid'' or ''mitral'' with confidence. It is much more accurate, and less confusing, simply to describe the side on which the connection is absent.

Modes of Atrio-ventricular Connection

The *modes* of connection are quite separate from the *types* of connection (Tynan *et al*., 1979). They are illustrated in **Figure 1.6**. The mode of connection is a way of describing the morphology of the atrio-ventricular valves. When both atrial chambers are joined to the ventricular mass, in other words, in the setting of two atrio-ventricular junctions, the mode of connection may be via *two perforate valves*, a *common atrio-ventricular valve*, or *one perforate and one imperforate valve*. Common atrio-ventricular valves normally, but not invariably, *straddle*. The common valve guards both atrio-ventricular junctions. In contrast, the single perforate valve in any of the combinations described earlier, or even the one that is associated with absence of one connection, guards only a solitary atrio-ventricular junction. Such a valve, nonetheless, may also straddle. An imperforate valve is recognized as a translucent membrane blocking the junction between an atrium and its underlying ventricle. Because this can occasionally be confused with the membranous atrio-ventricular septum, the presence of any kind of tension apparatus, be it only a single chorda tendinea inserting into the membrane from its ventricular aspect, assists greatly in recognition of an imperforate valve. Hitherto, the difficulty of distinguishing preoperatively between an absent connection and an imperforate valve has tended to blur the distinction between the two. The ability to make this distinction has now been greatly enhanced by techniques such as cross-sectional echocardiography and other tomographic modalities.

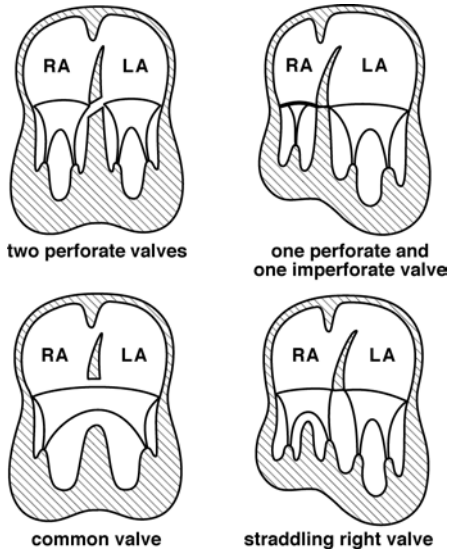

Figure 1.6

Types of Ventriculo-arterial Connection

These are analogous to the types of atrio-ventricular connection, except that there is no biventricular and ambiguous connection. Categorization may also necessitate assignment of an overriding arterial valve to the ventricle, supporting the greater part of its circumference (Kirklin *et al*., 1973; Shinebourne *et al*., 1976; Tynan *et al*., 1979). This assignment is somewhat more arbitrary than for atrio-ventricular valves because arterial overriding is far more common. When overriding approaches 50%, it often helps to assign each leaflet of the arterial valve in turn to a ventricle. The valve is then assigned to supporting the most leaflets or portions thereof. The situation, in which the aorta arises from the left ventricle or its rudiment, and the pulmonary trunk from the right ventricle or its rudiment, is described in terms of concordant ventriculo-arterial connections. With discordant connections, often called ''transposition'', these sites of origin are reversed. Exactly analogous to the double-inlet atrio-ventricular connection is double-outlet ventriculo-arterial connection. When only one great artery is connected to the heart, there is a single outlet from the heart (**Figure 1.7**). This can be via a common arterial trunk, an aorta with pulmonary atresia, a pulmonary trunk with aortic atresia, or via a solitary arterial trunk when there is complete absence of the intrapericardial pulmonary arteries. Should it be possible to trace the atretic trunk to its ventricle of origin, however, it is justifiable to describe the connection accordingly, such as double-outlet right ventricle with pulmonary atresia. It is necessary also to describe a solitary arterial trunk, since it is not possible to determine, had intrapericardial pulmonary arteries been present, whether they would have taken origin from the pulmonary trunk or from the ventricular mass.

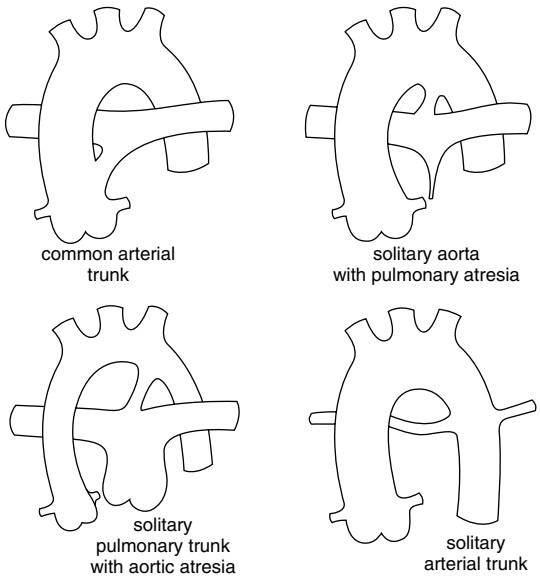

Figure 1.7

Modes of Arterial Connection

The mode of ventriculo-arterial connection includes common, overriding and imperforate valves. Because potential connections of the atrial chambers or great arteries to the ventricular mass by imperforate valves may be difficult to discern in living patients, the absence of the right or left atrio-ventricular connections, and single outlet from the heart, are the designations most likely to be changed by post mortem examination. One of the strengths of this classification, nonetheless, is that even if, for example, a concordant ventriculo-arterial connection with aortic atresia is technically misclassified as single outlet via the pulmonary trunk, a great deal of useful information will have been given.

RELATIONS AND CARDIAC POSITION

These may need to be specified, particularly if they are unpredictable from knowledge of atrial situs and segmental connections. Although an unpredictable relation or position may affect the surgical approach, it does not alter the nature of the operation in the way that abnormal connections do. For example, if the atrio-ventricular connections are discordant, whilst the ventriculo-arterial connections are concordant, it is fundamental to appreciate that the pulmonary and systemic circulations require reversal. Whether the heart is in the right or left side of the chest, whether the ventricles are side by side or one is above the other, and whether the aorta is anterior or posterior to the pulmonary trunk all constitute important pieces of information, but none in itself is fundamental. It is particularly confusing to use a relationship to indicate a connection. Above all, the term ''d-transposition'' should not be used to imply concordant atrio-ventricular connections, neither should the term ''l-transposition'' be used to imply discordant atrio-ventricular connections.

Implications for Reporting Surgical Results

In our opinion, although often held up to ridicule, the so-called ''50% rule'' as applied to determination of atrio-ventricular and ventriculo-arterial connections has solved many more problems than it has created. The rule does have implications, nonetheless, as used currently, for reporting of results of surgery on conditions that form part of a spectrum of anomalies, such as tetralogy of Fallot and double-outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis. Results for both conditions should best be presented simultaneously. Indeed, it could be argued that the term ''double-outlet'' should be used only to account for the ventriculo-arterial connection, and not to describe a phenotypic entity.

ADDITIONAL CARDIOVASCULAR ANOMALIES

These are most helpfully listed in an order, starting with the great veins and passing through the heart to the great arteries. In the majority of cases, these abnormalities are the only ones present. Consequently, most conditions continue to be described by extremely familiar terms.

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2

Surgical Anatomy

R. H. Anderson and A. E. Becker

Cardiac surgery on neonates and infants is severely limited in access by the small size of the organ, and by the necessity of returning the organ in a functioning state to its owner. In cardiac surgery, it is also possibly more important to know the precise locations of the cardiac conduction tissues in infants than in adults, because even a minimal deviation in such a miniature field can result in a potentially fatal arrhythmia. This chapter* therefore describes cardiac anatomy and, in particular, the disposition of the conduction tissues, in a manner that we hope is helpful to surgeons who operate on the hearts of very young children. Two points must be made clear. In this chapter, we assume the presence of concordant atrio-ventricular and ventriculo-arterial connections. Deviations from this normal pattern produced by abnormal connections and communications between chambers are discussed in Chapter 8. Second, because dissection is much easier in adult hearts and permits higher quality photographs, most of the illustrations depict adult hearts.

CARDIAC SHAPE AND POSITION

The heart is shaped like a rounded three-sided pyramid that occupies the lower part of the mediastinum (Anderson *et al*., 2004). It possesses a base, which lies in an oblique plane behind the sternum and angles as it descends to the right, and an apex, which extends outward into the left hemithorax (**Figure 2.1**). The pyramidal shape of the heart is best appreciated by viewing it from the apex along its long axis (**Figure 2.2**). Its three surfaces are an anterior sternocostal surface that abuts against the rib cage and curves upward and backwards; an inferior diaphragmatic surface; and the posterior and more rounded supero-posterior surface, which lies within the cardiac notch of the left lung. The junction between the sternocostal and diaphragmatic surfaces is sharp, the so-called ''acute margin'', whereas the junction between the sternocostal and posterior surfaces is more an area than a margin, but is termed the ''obtuse margin''. When viewed in terms of its thoracic location, the heart occupies the mediastinum. In this position, one-third of its mass is to the right, and two-thirds are to the left of the midline. Its long axis is orientated from the right shoulder toward the left hypochondrium (**Figure 2.1**). The surface projection of the heart is approximately trapezoidal. The base forms the right and superior edges of the trapezoid. It is composed of the atrial chambers, together with the great arteries that leave the ventricular mass. The acute and obtuse margins of the ventricular mass form the left and inferior edges of the silhouette. The two margins come together to form the apex.

PERICARDIAL CAVITY

The heart is enclosed by a firm, fibrous sac, the pericardium. In functional terms, the pericardium consists of a tough bag that encloses the heart. The heart is placed into it from the base toward the apex, and the bag is subsequently closed by the fibrous layer merging at the cardiac base with the adventitial layers of the veins that enter and the arteries that leave the heart. In anatomical terms, the pericardium is a much more complex structure. This is because a second sac, the serous pericardium, is contained within the fibrous pericardium. The serous pericardium is a double-layered membrane that is invaginated by the heart during its development. Because of this invagination, the inner layer of the serous pericardium is fused to the surface of the heart, forming the epicardium, or visceral pericardium (**Figure 2.3**). The outer layer of the serous pericardium is fused to the fibrous pericardium, and forms the parietal pericardium. Because of the attachments of these membranes, the pericardial cavity, which separates the two layers of

* Editors' note: The editors have decided to use Latin anatomical terminology to maintain uniformity throughout this book. This was done with the agreement of Professors Anderson and Becker. They wish to emphasize, however, their preference for usage of English rather than Latin words.

Surgery for Congenital Heart Defects, Third Edition. Edited by J. F. Stark, M. R. de Leval and V. T. Tsang
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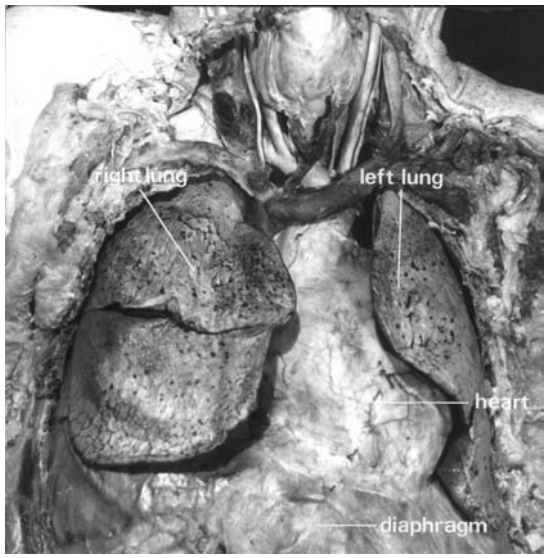


Figure 2.1

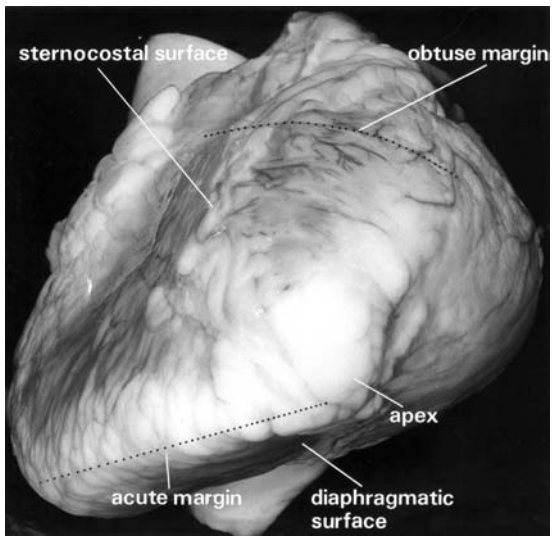


Figure 2.2

Figure 2.3

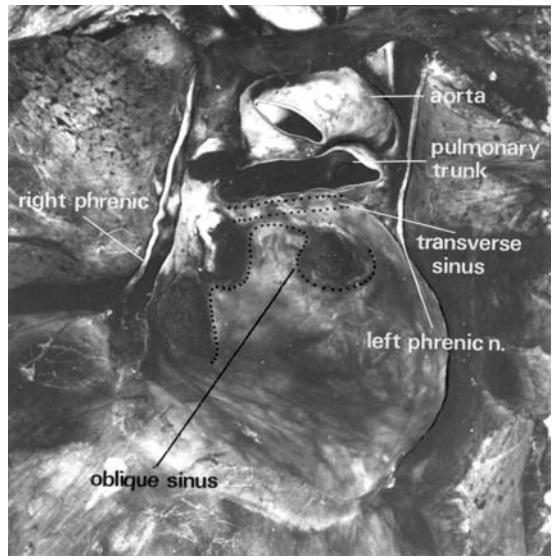


Figure 2.4

serous pericardium, is also therefore the space between the fibrous pericardium and the surface of the heart. The two layers of serous pericardium are reflected on one another at the entrances and exits of the great vessels so as to enclose the pericardial cavity. In contrast, the fibrous pericardium extends for some distance along the arteries and veins beyond the confines of the serous pericardial sac before merging with the walls of the vessels. By virtue of the arrangement of entry and exit of the great vessels, two recesses exist within the confines of the pericardial cavity. One of these, the oblique sinus, is a cul-de-sac behind the left atrium (**Figure 2.4**). It is limited to the right by the inferior vena cava (IVC) and right pulmonary veins, and to the left by the left pulmonary veins. Its right wall is a fold of tissue extending from the right pulmonary vein around the IVC. If it is necessary to encircle the

IVC within the fibrous pericardium during surgery, this fold must be divided. The reflection between the right and left pulmonary veins forms the roof of the sinus, and separates the oblique and the transverse sinuses, the small bare area of the left atrium being between the sinuses. To the left of the roof, between the left pulmonary artery and the subjacent pulmonary vein, is the fold of serous pericardium known as the ligament of the left superior vena cava (SVC), or Marshall's ligament (Marshall, 1850). In the presence of a persistent left vein, the pericardial fold is more extensive and surrounds it. The transverse sinus is an extensive recess between the layer of serous pericardium surrounding the anterior aspect of the atrial chambers and the cuff of pericardium that encloses the aorta and the pulmonary trunk. This sinus lines the important inner curvature of the heart, marking the site of the ventriculo-infundibular fold. Although a probe can be passed through the sinus to encircle the aorta, such a probe would also encircle the pulmonary trunk (Wilcox *et al*., 2004). In order to separate the aorta and the pulmonary trunk within the fibrous pericardium, it is necessary to incise the cuff of serous pericardium that surrounds both great arteries.

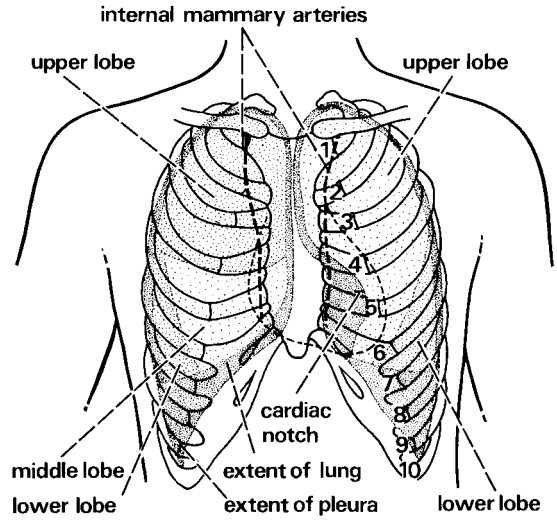
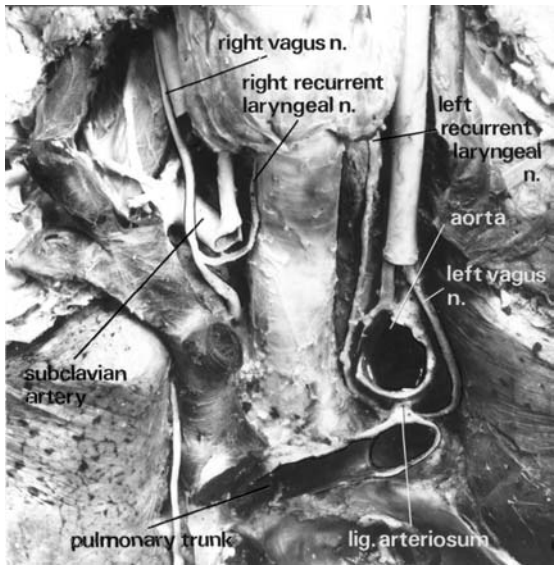


Figure 2.5

RELATIONSHIPS OF THE HEART

The anterior surface of the heart in its midpart is directly related to the underside of the thoracic skeleton, but to the right and left it is covered by the lungs and pleural cavities. The lungs are enclosed by invaginated double-layered sacs of pleura in the same way as the heart is enclosed by the pericardium. The line of reflection of the visceral and parietal layers extends down well beyond the edge of the lungs themselves (**Figure 2.5**). It is therefore easy to open the pleural cavity during sternotomy. The other major structures to be avoided during anterior thoracotomy are the internal thoracic arteries. These arteries originate from the first part of the subclavian artery on either side of the thoracic inlet, and pass down to either side of the sternum, branching as they descend to supply the anterior intercostal arteries. The right pleural cavity overlies the right side of the heart almost to the midline. The left pleural cavity, in contrast, extends away from the midline in the cardiac notch, leaving a bare area of the pericardium that projects directly onto the underside of the thoracic cage (see **Figure 2.5**). Reflection of the pleural cavities reveals the hilar regions of the lungs, the areas in which the parietal and visceral pleural layers reflect on each other, and through which the pulmonary vessels and airways enter and leave the lungs. The thymus is situated between the hilar regions above the heart. This structure is prominent at birth, and continues to grow until puberty, but regresses thereafter. In early life, it is made up of two unequal-sized pyramidal lobes that are connected by areolar tissue. The lower borders of both lobes extend onto the surface of the pericardium, and the tapering apices extend into the neck, sometimes reaching up as far as the thyroid gland. Each lobe is surrounded by a delicate fibrous capsule and can be separated from the other lobe along the midline. Two important nerves descend through the mediastinum posterior to the thymus and in relation to the hilar regions, viz. the vagus (10th cranial) and phrenic nerves. Both nerves enter through the thoracic inlet. The vagus nerve runs along the carotid arteries, but the phrenic nerve, descending from the cervical plexus, enters the thoracic cavity on the surface of the anterior scalene muscle, covered by the prevertebral fascia. As they pass toward the heart, the phrenic nerve is anterior to the vagus nerve on both sides. On the right side, the vagus nerve gives off its recurrent laryngeal branch high up, with the recurrent nerve then encircling the right subclavian artery before passing upward and out of the thoracic cavity (**Figure 2.6**). On the left side, the phrenic and vagus nerves extend forward across the aortic arch, and the superior intercostal vein is insinuated between them as it passes forward to drain into the brachiocephalic vein (**Figure 2.7**). The vagus nerve then gives off its recurrent laryngeal branch, which, on the left side, curves around the ductus arteriosus or its ligamentous remnant, passing inward between the aortic arch and left pulmonary artery before ascending to reach the larynx (see **Figure 2.6**). Another structure recurs around the subclavian artery on both sides but at the level of the thoracic inlet. This structure is the subclavian loop, or ansa subclavia, and should always be identified and



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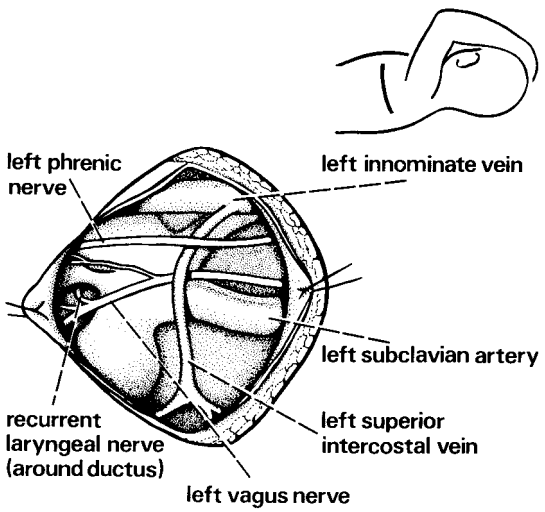


Figure 2.7

avoided when the subclavian arteries are liberated for usage in shunt procedures. As the vagus and phrenic nerves approach the heart, they diverge. The vagus nerve on each side passes posteriorly into the hilar regions, ramifying to form the pulmonary plexuses and rejoining to form the vagal trunks, which then leave the thorax along the oesophagus. In contrast, the phrenic nerves swing forward and arch across the pericardial sac as they pass down to ramify on the domes of the diaphragm.

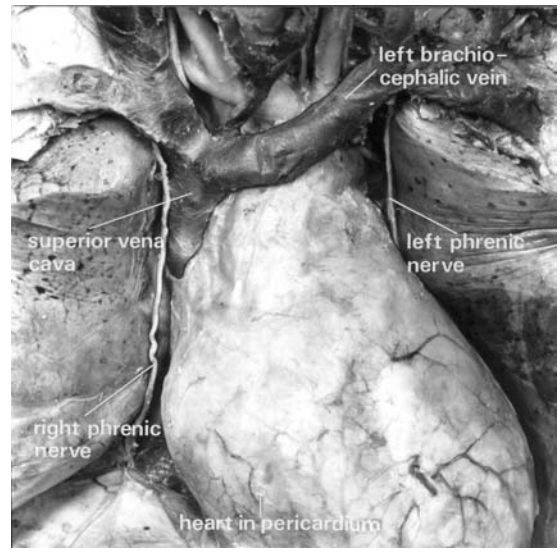


Figure 2.8

They are in potential danger of surgical trauma during the removal of part of the pericardium to harvest a patch, and during dissection around the SVC, especially at reoperations. At the base of the heart, after retraction of the pleura, the relationships of the great vessels are easily appreciated. On the right side, the SVC enters the superior aspect of the base of the heart (**Figure 2.8**). The right and left brachiocephalic veins, or innominate veins, lying superficial to their accompanying arteries, join to form the SVC, which then receives the azygos vein to its right side. This structure drains most of the blood from the thoracic cage. Sometimes the venous channel passes behind the arterial structures, but this is rare. The IVC is a very short channel within the thorax, entering the inferior aspect of the cardiac base almost as soon as it has penetrated the diaphragm. The great arteries leave the left side of the cardiac base, the pulmonary trunk being anterior to and to the left of the aorta. The aorta arises from its deeply positioned valve and ascends with a sweep to the right before traversing backwards and leftward to descend to the left of the vertebral column. Ascending from the arch are the brachiocephalic trunk, or innominate artery, soon to divide into right subclavian and common carotid arteries, and the left common carotid and subclavian arteries. The pulmonary trunk passes leftward from the upper margin of the cardiac base before dividing into right and left pulmonary arteries, which diverge toward the hilar regions of the lungs. The right pulmonary artery passes beneath the arch of the aorta. The trachea runs down into the thoracic cage behind the great arteries, and divides at the level of the

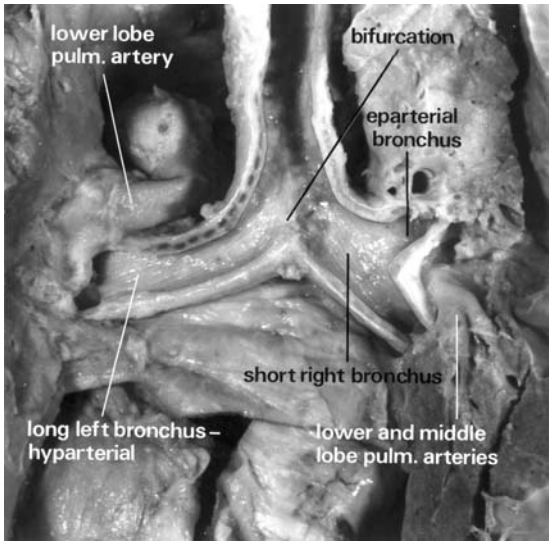


Figure 2.9

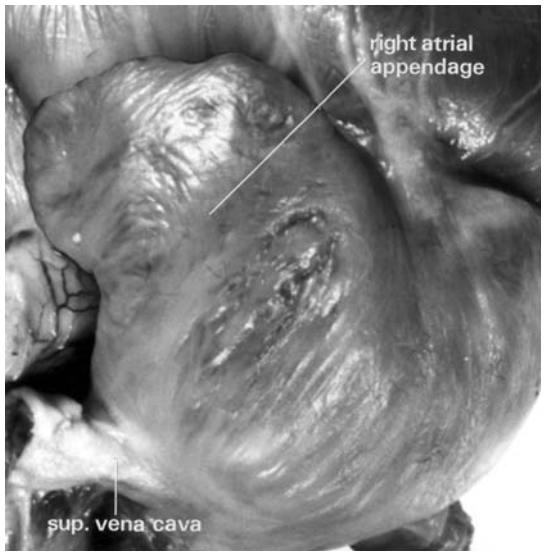
aortic arch. The left bronchus passes beneath the aortic arch, where it is joined by the left pulmonary artery. The branch from the artery to the left lower lobe curves over the left main bronchus before it divides. In contrast, on the right side, the right main bronchus divides early, and the artery to the right middle and lower lobes passes beneath the first division of the bronchus. The artery to the right upper lobe ascends to run into the lobe alongside and beneath this bronchus, which is therefore an eparterial bronchus, since it is above the artery. In contrast, the first branch of the left main bronchus is a hyparterial bronchus, since it runs underneath the branch of the pulmonary artery. Note that it is the posterior view of bronchial anatomy that is shown in **Figure 2.9**. The pulmonary veins return from the hilar regions to the posterior aspect of the cardiac base, sandwiching the bronchi between themselves and the pulmonary arteries. The oesophagus also has a direct relationship with the heart. Descending into the thoracic cavity with the trachea, it continues beyond the tracheal bifurcation and runs directly behind the left atrium, leaving the thoracic cavity through the right crux of the diaphragm. Being in such a posterior position, it is unlikely to be encountered during cardiac operations except for repair of infradiaphragmatic anomalous pulmonary venous connection, in which the descending channel runs along the oesophagus to reach the abdomen.

ANATOMY OF CARDIAC CHAMBERS

To understand the spatial relationships of the different cardiac chambers, three basic rules of cardiac anatomy should be remembered. First, because of the orientation of the cardiac long axis, the ventricles are more or less to the left of their corresponding atrial chambers. Second, the right atrium and the right ventricle are relatively anterior to their left counterparts. Third, because of the anterior position of the chambers of the right heart, the aorta and its valve have a central position in the heart, being wedged between the atrio-ventricular valves and posterior to the infundibulum of the right ventricle. The aortic valve is, therefore, related to all four cardiac chambers.

The Right Atrium

The right atrial chamber forms the right border of the cardiac silhouette, and is the most superficial chamber to the right of the midline. It has several anatomical components (Ho and Anderson, 2000a). The more posterior component, the venous sinus, or sinus venarum, has smooth walls. As usually viewed by the surgeon at operation, it receives the SVC to the left side and the IVC to the right side of its cavity. It communicates through a wide and unrestricted channel with the second, anterior, component of the chamber, the atrial appendage. The walls of the appendage are rough internally because of the multiple musculi pectinati that cross its surface. It extends forward in a pouch-like manner to clasp the right side of the aorta. When viewed externally in surgical orientation, the appendage is triangular but blunt (**Figure 2.10**) in comparison with the left atrial appendage, as viewed through a left thoracotomy (**Figure 2.11**). This distinction between the atrial appendages is of major significance because their shape is the best guide to atrial morphology, and hence to atrial situs. The junction between the venous part of the right atrium and the atrial appendage is marked externally by the sulcus terminalis. This sulcus marks the site of the crista terminalis, the prominent muscular structure that separates the venous sinus and appendage internally. The position of the sinus node as seen by the surgeon is shown in **Figure 2.12**. A second groove is seen when the heart is tilted leftward from the initial surgical orientation. This second groove is between the SVC and the pulmonary veins and continues toward the IVC, separating it from the left atrium. It marks the site of the interatrial fold, and is known as Waterston's groove (see **Figure 2.12**). When the right atrium is opened, a muscular prominence is seen in its floor; this prominence overlies the root of the aorta and the central fibrous body (**Figure 2.13**). To the right of, and superior to, this aortic mound is the tricuspid valve, which leads to the right ventricle. The myocardium inserting into the valvar leaflets is smooth, and forms the atrial vestibule. To the left and below is the septal surface of the chamber, including the fossa ovalis. At some distance beneath the mound is the orifice



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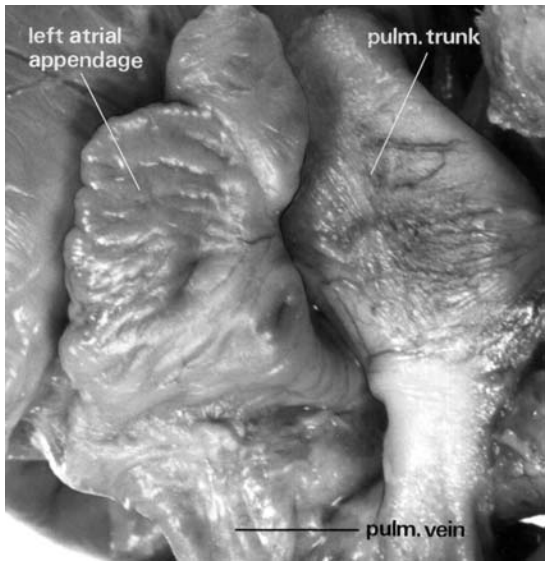

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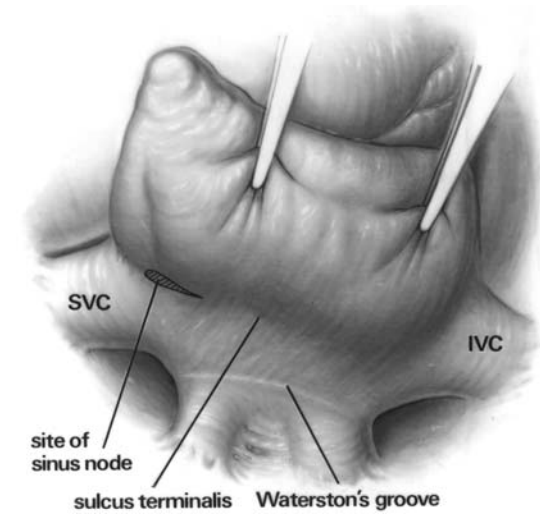


Figure 2.12

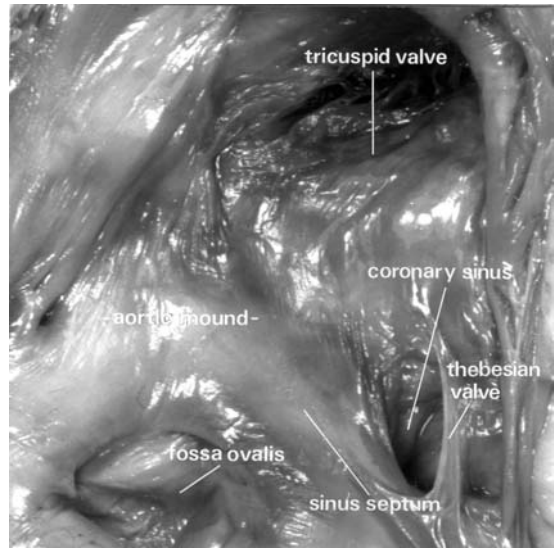
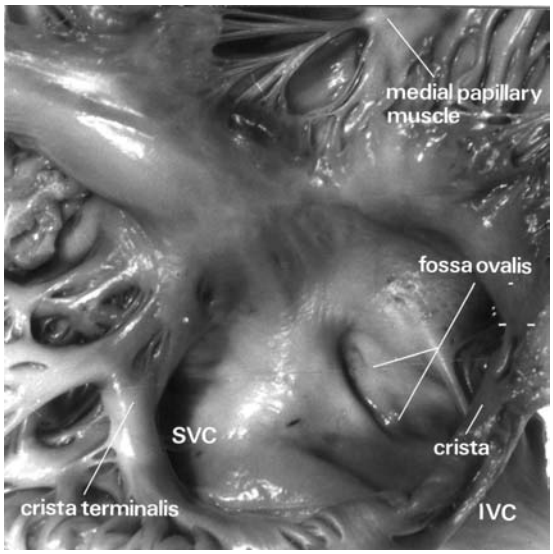


Figure 2.13

of the coronary sinus, which drains most of the venous return from the heart itself. The crista terminalis springs from within the atrial appendage. It extends onto the septal surface to become continuous with the superior rim of the fossa ovalis (**Figure 2.14**). The main part of the crista, however, swings upward and encircles the orifice of the SVC. It then passes in front of the orifice of the IVC and dips to the right of and beneath the mouth of the coronary sinus. At that point, the muscle bundle widens into a broad strip that runs into the septal leaflet of the tricuspid valve, forming a muscular area of variable dimensions that separates the attachment of the tricuspid leaflet from the coronary sinus and the fossa ovalis. This is the inferior isthmus, which is continuous superiorly with the septal part of the tricuspid vestibule that overlies part of the ventricular septum, and therefore separates the right atrium from the left ventricle. We



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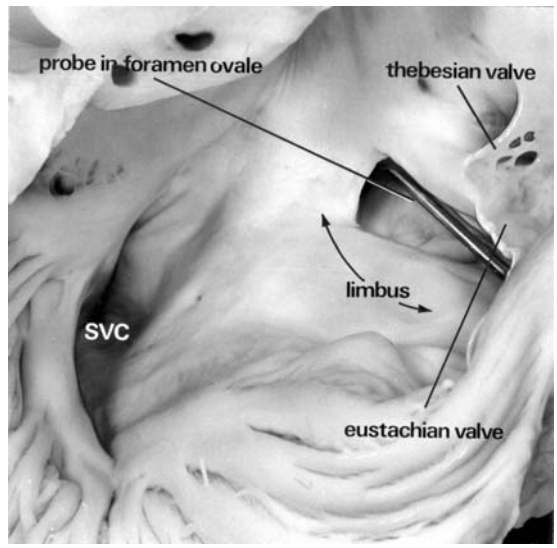


Figure 2.15

used to call this area the ''muscular atrio-ventricular septum''. We now know it is a muscular sandwich, since a superior continuation of the adipose tissue of the inferior atrio-ventricular groove is interposed between the atrial and ventricular muscular layers. Beneath the mouth of the coronary sinus, the transitional area from terminal crest to muscle strip is frequently pouched out to form a diverticulum, the sub-Thebesian sinus. To the left of this sinus, another muscle band, the so-called Eustachian ridge, springs from the crista terminalis and passes toward the aortic mound, separating the orifices of the IVC and the fossa ovalis from the mouth of the coronary sinus. This area also forms the inferior margin of the fossa ovalis (**Figure 2.15**). Fibrous membranes are attached to the crista terminalis at the entrance of the IVC (the valve of the IVC, or Eustachian valve), and above the entrance of the coronary sinus (the valve of the coronary sinus, or Thebesian valve). Well formed in infancy, these valves regress in adulthood, but frequently persist as filigreed networks that extend across the venous orifices. The junction of these valvar structures, however, is almost always present, forming a fibrous strand that passes forward toward the aortic mound, burying itself in the musculature of the antero-inferior rim of the fossa ovalis, and running forward and upward to insert into the central fibrous body. This structure is the tendon of Todaro, and can easily be brought into prominence by placing tension on the Eustachian valve (Ho and Anderson, 2000b). It is of major significance because it forms one of the boundaries (**Figure 2.16**) of the triangle that contains the atrio-ventricular node, the so-called

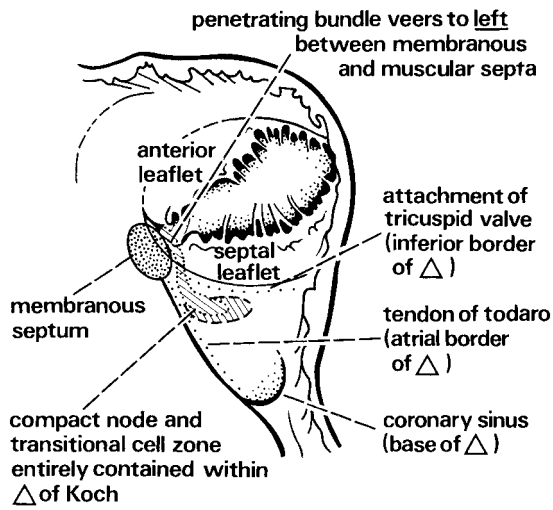
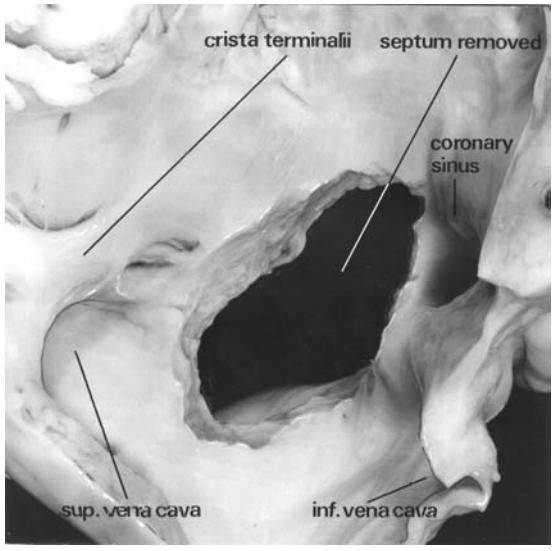


Figure 2.16

triangle of Koch. The tendon forms the left boundary of the triangle, meeting the right boundary, which is the septal attachment of the septal leaflet of the tricuspid valve, at the aortic mound. The base of the triangle is the orifice of the coronary sinus. In the view down into the right atrium, an extensive muscular area is seen abutting on the tricuspid and venous orifices. It is important to appreciate that not all of this is atrial septum. The floor of the fossa ovalis, confined by the superior and inferior rims, and the variably developed antero-inferior rim, are

indeed septal structures. Much of the superior rim, the so-called ''septum secundum'', however, is a fold between the right pulmonary veins and the SVC. This is evident externally as Waterston's groove. Incisions through this rim take the surgeon outside the heart, as do incisions in the area overlying the aortic mound, which is the superior atrial wall. Frequently, a small muscular fold produces a crevice in the anterior wall in this position. This crevice is of importance because it may catch a catheter passed into the atrium and aimed for the fossa ovalis. It is important to distinguish crevice from fossa ovalis when septal puncture is attempted, because the former overlies the transverse sinus, and is not an interatrial structure. Part of the area of the antero-inferior rim itself is an atrio-ventricular structure rather than an interatrial structure, because the tricuspid valve is attached to the septum more apically than is the mitral valve. The area between the septal attachments of the two atrio-ventricular valves is the atrio-ventricular muscular sandwich, and is described in more detail in the section on the atrio-ventricular junction (Anderson *et al*., 2000). The area of atrial floor that is truly interatrial is best demonstrated by removal of the interface between the atrial chambers (**Figure 2.17**). The right atrial myocardium contains and overlies two of the most vulnerable parts of the conduction system: the sinus and atrio-ventricular nodes. The sinus node is a small spindle-shaped structure that lies directly subepicardially in the sulcus terminalis (**Figure 2.18**). In about 90% of people, the node is entirely lateral to the junction of the SVC with the right atrium, but in the remaining 10% it may pass



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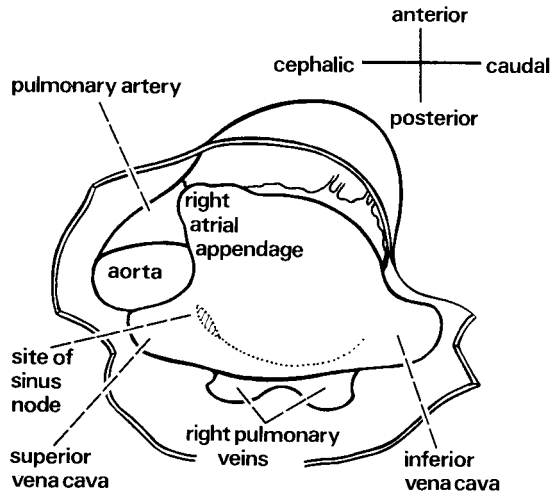

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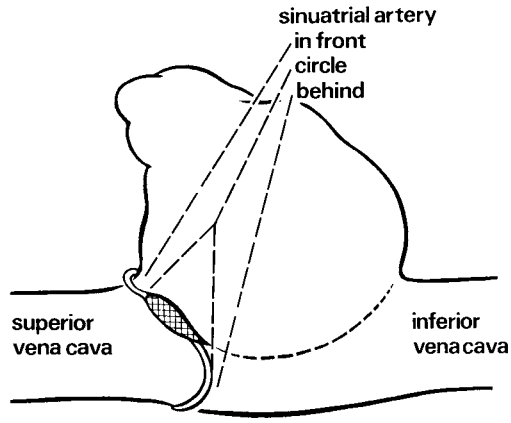


Figure 2.19

across the crest of the atrial appendage, thus sitting on the junction of crest and SVC in a horseshoe manner (Anderson *et al*., 1979). Because of its immediately subepicardial position, the node is particularly vulnerable to surgical trauma. The considerable variability found in the vascular supply to the node (**Figure 2.19**), together with the lesser variability in nodal position, compounds the vulnerability of the entire junction of the SVC with the right atrium during cardiac surgery. The nature of the right and interatrial myocardium, and its relationship to interatrial conduction, has been a controversial subject. Some authors have suggested that specialized tracts of conduction tissue extend through the walls of the right atrium and the atrial septum to connect the sinus

node with the atrio-ventricular node. We have searched diligently for such tracts by serially sectioning the entire atrial chambers of infant hearts in a single block of tissue. Our results show that the tissue connecting the nodes is the plain atrial myocardium of the right atrial walls and the interatrial septum. There are no tracts of conduction tissue that are insulated from the myocardium in a manner comparable with that of the ventricular conduction pathways (Anderson *et al*., 1981). The geometrical configuration of the right atrium is such that its walls are divided into bands of muscular tissue by the venous orifices and the fossa ovalis. This atrial morphology more than adequately accounts for the routes of preferential conduction found in electrophysiological mapping. In our opinion, histologically specialized tracts do not exist between the nodes. The atrio-ventricular node, together with its transitional cell zones, is contained entirely within the triangle of Koch (Anderson and Ho, 2001; see **Figure 2.16**). If all incisions and procedures avoid this triangle, damage cannot be inflicted on the node except in the rare circumstances in which atrio-ventricular conduction occurs through an aberrant conduction system (see Chapter 8). The node, located within the interatrial septum, penetrates into the central fibrous body to become the penetrating atrio-ventricular bundle at the apex of the triangle. The bundle passes from the right atrial tissues into the left ventricular outflow tract on the ventricular aspect of the point of insertion of the tendon of Todaro into the central fibrous body. The detailed anatomy of the penetrating bundle and the ventricular conduction tissues are described in the sections devoted to the atrio-ventricular junction and the left ventricular outflow tract.

The Left Atrium

The left atrium is the most posterior of the four cardiac chambers. Like the right atrium, it has several components: a smooth-walled part that receives the pulmonary veins, an appendage with muscli pectinati on its inner surface, a smooth vestibule inserting to the left atrio-ventricular junction, a septal component, and then an obvious body that supports the other components (Anderson and Ho, 2000a). **Figure 2.20** shows the dissected chamber as it may be viewed through a left thoracotomy. In contrast to the right atrium, there is no prominent muscle band between the smooth component and the pectinate part, and the appendage is much smaller and not as well trabeculated. Its shape is characteristic, being hooked and usually constricted at several points along its extensive but narrow length (**Figure 2.11**). The appendage extends around the left border of the pulmonary trunk, and is the only part of the left atrium to appear on the cardiac silhouette. The smooth-walled

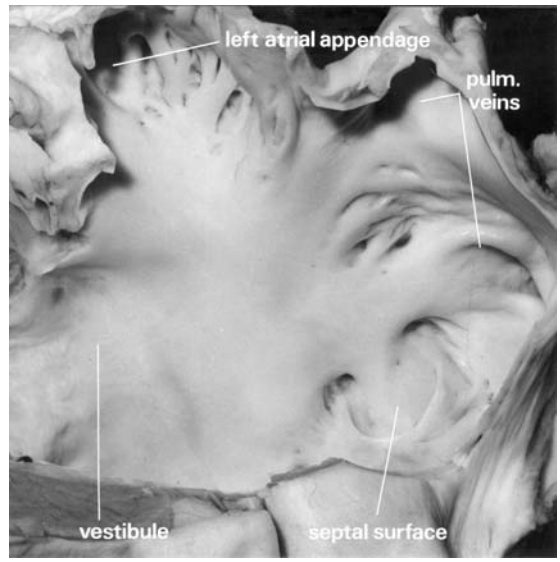


Figure 2.20

roof of the left atrium receives a pulmonary vein at each of its four corners. Running inferiorly beneath the posterior surface, and occupying the left inferior aspect of the atrio-ventricular junction, is the coronary sinus, which drains to the right atrium. When a left SVC is present, it usually drains to the coronary sinus, extending downward between the left atrial appendage and the left pulmonary veins. More usually, the left SVC is not present, being represented by its vestige, the oblique vein of the left atrium, itself related to a fold of pericardium in the oblique sinus. Surgical access to the left atrium is usually obtained either through the atrial septum, in which case its confines must be remembered (**Figure 2.17**), or through the left atrium between the interatrial groove and right pulmonary veins. A discrete furrow exists between the entrance of the right pulmonary veins and the SVC. This furrow, known as Waterston's or Sondergaard's groove, forms the greater part of the superior rim of the fossa ovalis. It is no more than an infolding of the atrial roof and, if necessary, can be dissected to produce additional atrial tissue, as, for example, for use in the Senning procedure. The atrial roof itself is extensive between the entrances of the right and left pulmonary veins. Having opened the left atrium through an incision in either the septum or the roof, the surgeon is confronted with the vestibule of the mitral valve in the floor of the atrium, and with the much smaller orifice to the appendage on the left. The septal surface of the left atrium is not visible. When seen in isolation, the characteristic roughened aspect is, in reality, part of the anterior wall rather than the septum and overlies the

transverse sinus. It is not the left atrial counterpart of the fossa ovalis.

The Ventricles

Although the right and left ventricles exhibit important differences, they are comparable units. Each ventricle is composed of an inlet portion, which contains an atrio-ventricular valve, and is limited by the attachments of its tension apparatus, an apical trabecular portion, and an outlet portion, which supports the leaflets of an arterial valve. The atrio-ventricular and arterial valves similarly are of comparable structure in each ventricle, albeit that each valve possesses significant individual differences. Both sets of valves are complex structures. The leaflets are the primary units of the valve, meeting centrally when closed. The leaflets do not meet at their free edges; rather, they close at about one-third of the distance from the free edge to the attached margin. In the atrio-ventricular valves, the leaflets originate at the atrio-ventricular junction, in which fibrous tissue is formed to a variable extent, and which is usually termed the annulus (Angelini *et al*., 1988). Supporting the ventricular aspect of the leaflets are the chordae tendineae, which attach mostly to the so-called ''rough zones''. In some places, however, a single chorda divides in fan-like manner, and the subsidiary cords are then attached to the free edge of two adjacent leaflets. Each of the single chorda branching in a fan-like manner usually arises from the apex of a papillary muscle, and they tend to mark the ends of the zones of apposition between the leaflets. The chordae that are attached to the free edges of the leaflets may also branch in fan-like fashion to support the spaces between scallops in some of the leaflets. These are referred to as cleft chordae. In addition to the chordae supporting the free edges and rough zones, the leaflets are also supported by chordae that pass directly from the ventricular myocardium to the underside of the leaflets. These are the basal chordae. In contrast to the atrio-ventricular valves, the arterial valves lack any tension apparatus. The overall architecture, nonetheless, is surprisingly complex. The interrelationship between the aortic sinuses and the leaflets of the arterial valves is such that a marked discordance is produced between the haemodynamic and anatomical ventriculo-arterial junctions (Anderson, 2000; **Figure 2.21**). The haemodynamic junction is dictated by the attachment of the leaflets. This attachment is arranged in a semilunar manner, in which the attachment of each leaflet arises to the peripheral attachment of the zone of apposition with the adjacent leaflet, and sinks to the mid-point of the leaflet. As the attachment rises toward the sinutubular junction, it crosses the anatomical ventriculo-arterial junction. This latter junction is the circle over which the fibrous wall of the arterial trunk is supported by the respective ventricle. As a consequence of this arrangement, three triangles of arterial wall, thinner than the adjacent walls of the arterial sinuses, are incorporated within the structure of the ventricular outflow tracts, and three scallops of ventricle are incorporated at the base of each arterial sinus (**Figures 2.22 and 2.23**). It follows from this complex geometrical arrangement that there is no collagenous ring, or annulus, supporting the semilunar leaflets of the arterial valves in any sense comparable with the way in which the leaflets of the atrio-ventricular valves are attached in a circular manner at the atrio-ventricular junction. The only ring-like structure at the ventriculo-arterial junction is the line of fusion of the fibrous arterial wall with the supporting ventricle, viz. the anatomical ventriculo-arterial junction (Anderson, 2000). It is the crown-like configuration of the attachments of the leaflets, nonetheless, that is referred to by surgeons as the ''annulus''.

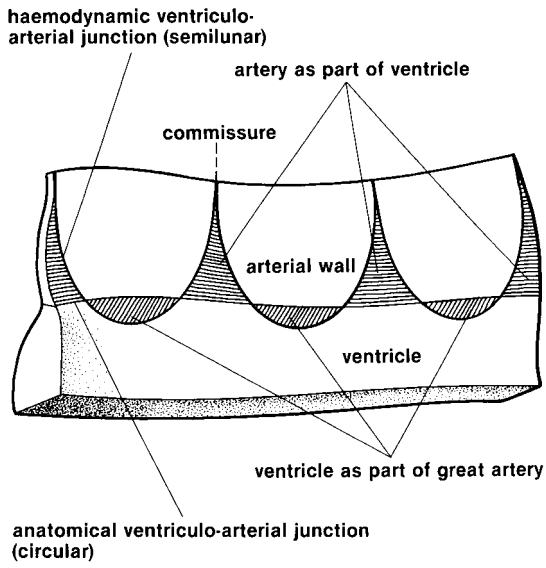


Figure 2.21

The Morphologically Right Ventricle

This chamber forms the greater part of the anterior surface of the ventricular mass, extending from its more or less vertically positioned atrio-ventricular valve, the tricuspid valve, and running behind the sternum to the horizontal arterial valve, the pulmonary valve, at the left border of the cardiac base (Anderson *et al*., 2004). The characteristic morphological features of the right

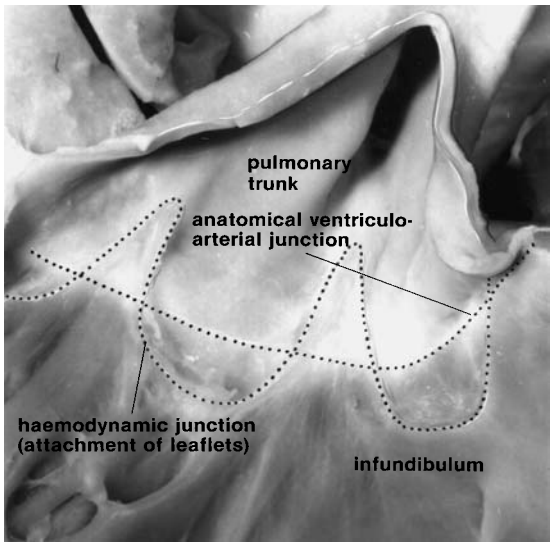


Figure 2.22

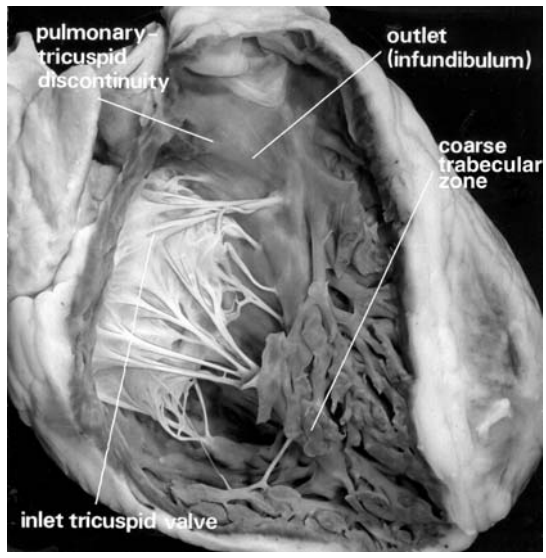


Figure 2.24

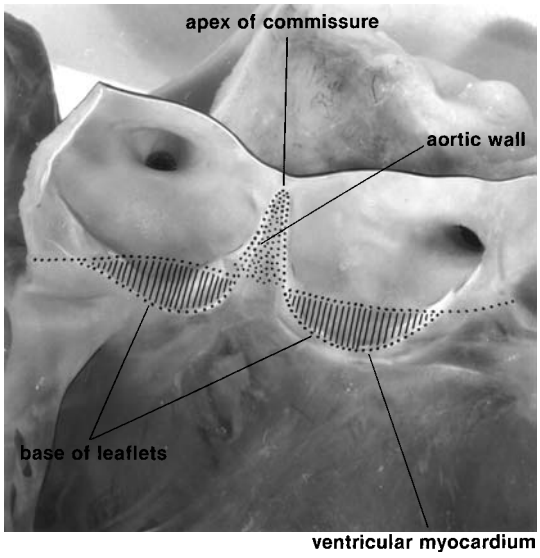


Figure 2.23

ventricle, apart from the features of its atrio-ventricular valve, are that, first, the apical trabecular zone has coarse trabeculations, and second, the atrio-ventricular and arterial valves are separated by a prominent muscular shelf in the ventricular roof, the supraventricular crest (crista supraventricularis) (**Figure 2.24**). The inlet portion of the right ventricle extends from the junctional attachment of the leaflets of the tricuspid valve to the ventricular attachments of their papillary muscles. The funnel thus formed is more extensive inferiorly and laterally than septally and superiorly. The three leaflets of the tricuspid valve are positioned septally, anterosuperiorly and inferiorly. The zones of apposition separating the leaflets have anteroseptal, supero-inferior and inferior ends. The leaflets are illustrated in **Figure 2.25**, with the heart in anatomical position, but with wide retraction of the valve opened through its inferior commissure. When the surgeon approaches the valve through the right atrium, the orientation is altered (**Figure 2.26**). The anteroseptal zone of apposition is on the left, the inferior zone of apposition is on the right, and the septal leaflet extends between them. The anteroseptal zone of apposition is supported by the almost constant, but small, medial papillary muscle, known also as the muscle of Lancisi, which springs from the inferoposterior limb of the trabecula septomarginalis (**Figure 2.27**). A cleft extending to the area of the membranous septum is frequently seen in the septal leaflet of the valve (**Figure 2.28**). The area around both the cleft and the zone of apposition should be avoided at all costs because it is the location of the atrio-ventricular conduction tissues contained within the triangle of Koch (**Figure 2.28**). The morphology of anterosuperior and inferior leaflets and their zone of apposition is more variable. This zone of apposition is frequently supported by the prominent anterior papillary muscle, which originates from the apical body of the trabecula septomarginalis. In many hearts, however, this muscle supports the middle of the anterosuperior leaflet,

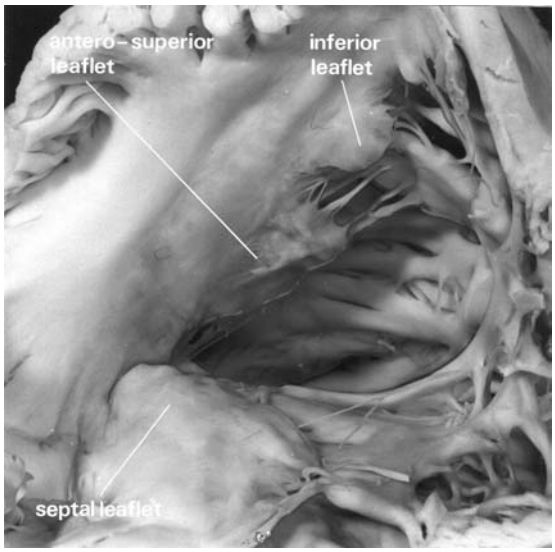


Figure 2.25

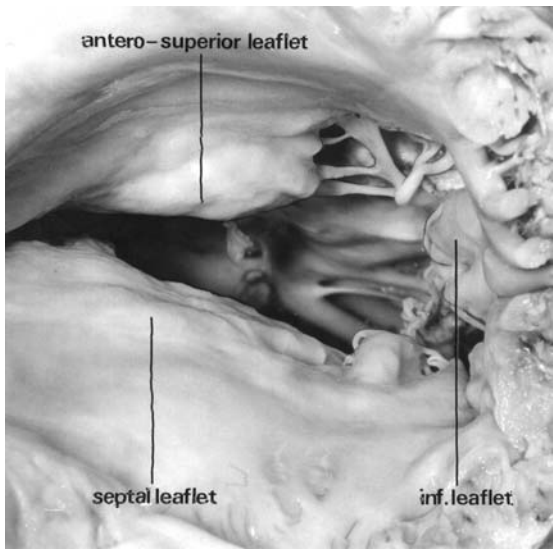


Figure 2.26

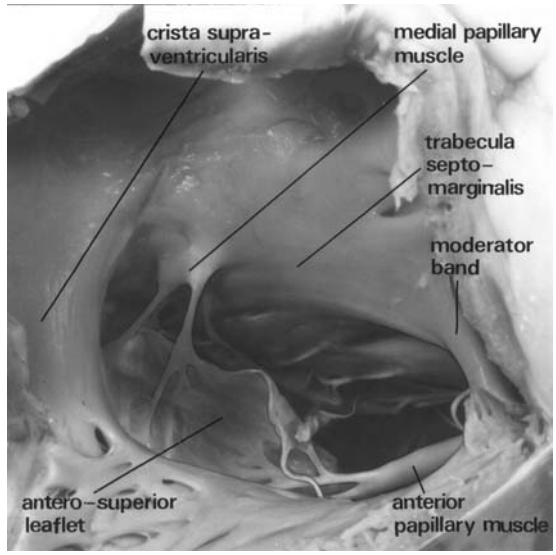


Figure 2.27

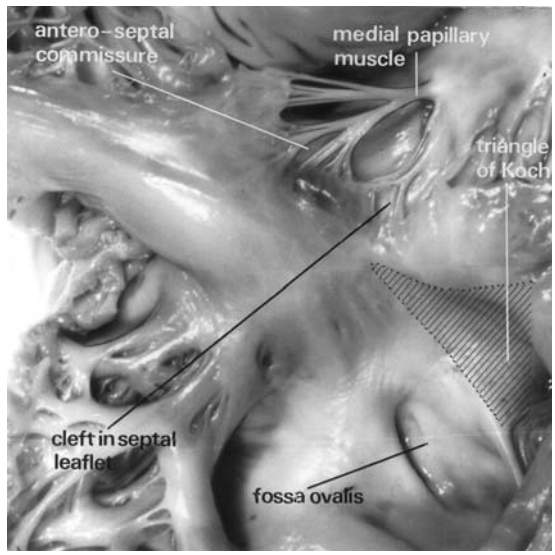


Figure 2.28

and a second, smaller, anterior papillary muscle supports the zone of apposition. The inferior zone of apposition is supported by a small inferior papillary muscle, but several other muscles spring like a sheaf from the apical ramification of the trabecula septomarginalis. When approached from the infundibulum, it is the anterosuperior leaflet of the tricuspid valve that is visible to the surgeon. This leaflet is supported superiorly and to the left by the medial papillary muscle and to the right by the anterior papillary muscle (**Figure 2.27**). A muscular ring demarcates the approximate limits of the ventricular inlet portion. In the ventricular roof, the ring is formed by the supraventricular crest, which represents the inner curvature of the heart wall (Vricella *et al*., 2004). Septally, the crest inserts between the limbs of the trabecula septomarginalis. The trabeculation is

extensive, and is characteristic of the right ventricle. The anterocephalad limb of the trabeculation runs upward toward the attachments of the pulmonary valve, and the posterocaudal limb extends variably into the inlet portion, giving rise to the medial papillary muscle (pm in **Figure 2.29**), and other chordae that support the leaflets of the tricuspid valve. The body of the trabeculation runs apically, and ramifies at the apex into the leash of papillary muscles that support the inferior and septal leaflets of the tricuspid valve. It also gives rise to the prominent moderator band that crosses to the anterior papillary muscle, forming the effective floor of the muscular ring. The parietal rim of the ring is another prominent trabeculation, viz. the downward continuation of the crest opposite the trabecula septomarginalis. The trabecular zone of the ventricle extends from this muscular ring, confining the inlet portion toward the ventricular apex, and being characteristically coarse. It extends well up toward the infundibulum, and additional bands from the trabecula septomargionalis, the trabeculae septoparietales, cross freely through it. The infundibulum, or ventricular outlet portion, is the muscular funnel that supports the leaflets of the pulmonary valve and the wall of the pulmonary trunk. Its margin with the trabecular zone is indistinct. The walls become smoother immediately beneath the valve. When viewed from the anterior aspect, the posterior wall of the infundibulum is made up for the larger part by the supraventricular crest. A small part of the crest that inserts between the limbs of the trabecula septomarginalis can be identified by dissection to represent a true muscular outlet septum,

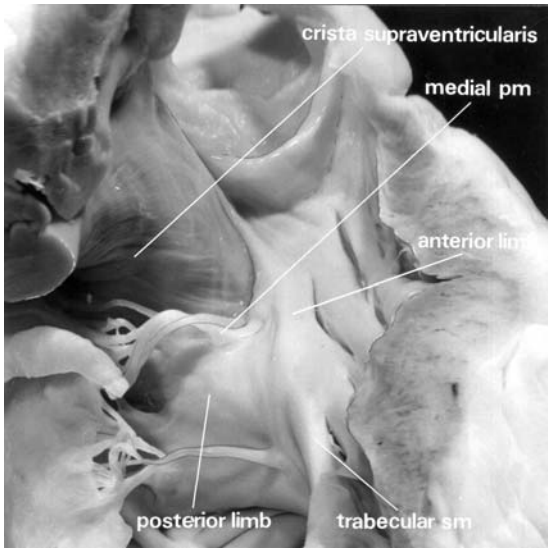


Figure 2.29

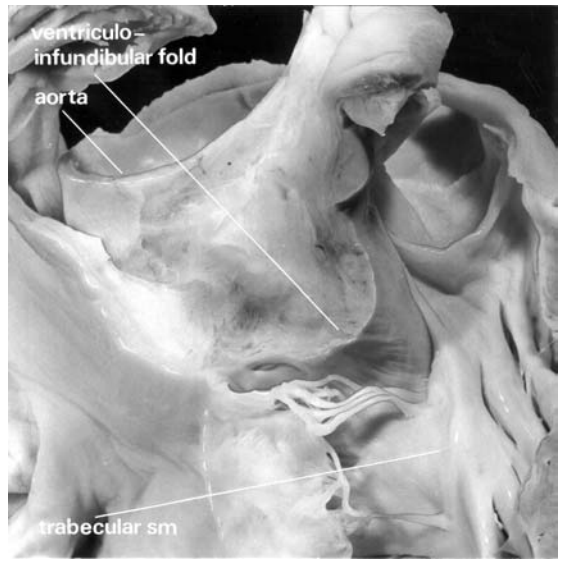


Figure 2.30

but this is of minimal dimensions in the normal heart (**Figure 2.30**) and cannot be identified with certainty without resorting to dissection. This anatomical structure, however, is of considerable importance with regard to controversies concerning the nature and description of the supraventricular crest. The crest in the normal heart is the extensive muscular structure that separates the attachments of the leaflets of the tricuspid and pulmonary valves in the roof of the right ventricle (**Figure 2.27**). Some authors categorize the trabecula septomarginalis (sm in **Figure 2.30**) as the ''septal band'' of the normal crest, but the trabeculation is purely a septal structure, and is in no way ''supraventricular''. The crest itself has two distinct components that are discrete structures only in malformed hearts. The larger part, the ventriculo-infundibular fold, represents the inner curvature of the heart wall (**Figure 2.30**). Incisions through this part take the surgeon outside the heart. Only a very small part of the crest, the muscular outlet septum, is located between the subpulmonary and subaortic outflow tracts.

The leaflets of the pulmonary valve are delicate structures attached in a semilunar way in part to the muscular wall of the infundibulum and in part to the wall of the pulmonary trunk. This arrangement (**Figure 2.22**) means that, as in the aortic root, part of the right ventricular muscular infundibulum is incorporated within the arterial sinus, while triangles of arterial wall are included within the ventricular outlet. Furthermore, careful dissection (**Figure 2.31**) reveals that the entire subpulmonary infundibulum is a free-standing structure that can be removed in one piece from the ventricle

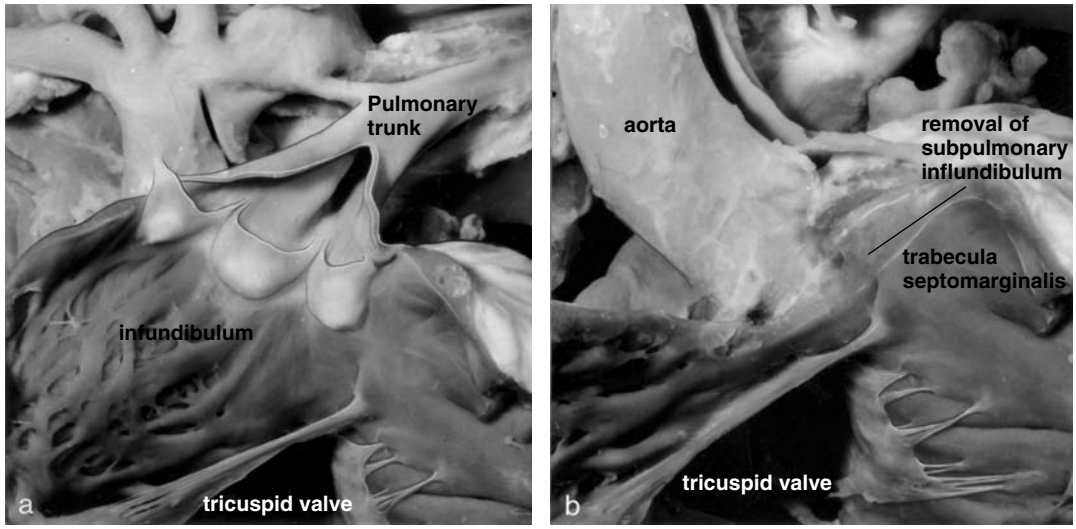


Figure 2.31

without disturbing the musculature of the left ventricle (Merrick *et al*., 2000). This has important consequences concerning the morphology of the ventricular septum (to be described). When the sinuses of the pulmonary trunk, and the leaflets of the pulmonary valve that they contain, are named and described, advantage can be taken of the fact that two sinuses of the pulmonary trunk almost always ''face'' the sinuses of the aorta that give rise to the coronary arteries. These sinuses can, therefore, be termed the right and left facing sinuses. The zone of apposition between the leaflets that they support is almost always opposite the zone of apposition between the corresponding facing leaflets of the aortic valve. The other leaflet of the pulmonary valve is thereby described as the non-facing leaflet, and is supported by the non-facing sinus.

The Morphologically Left Ventricle

The left ventricle is located almost entirely behind the right ventricle; only its most leftward rim is visible on the cardiac silhouette. The major features that differentiate it from the right ventricle (**Figures 2.24 and 2.32**) are that: first, the apical trabecular zone of the left ventricle has characteristically fine trabeculations; second, the leaflets of its atrio-ventricular and arterial valves are in fibrous continuity, whereas in the right ventricle the supraventricular crest separates them; third, the septal surface is smooth, lacking any structure comparable with the trabecula septomarginalis of the right ventricle. The inlet portion of the left ventricle extends from the atrio-

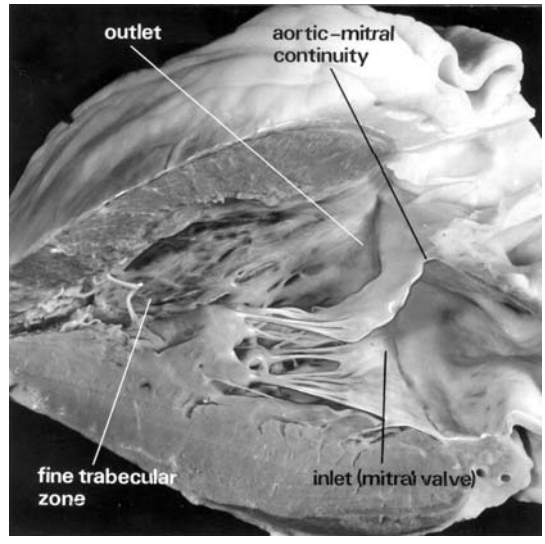


Figure 2.32

ventricular junction to the attachment of the papillary muscles, and contains the mitral valve. This valve has two major leaflets, separated by the solitary zone of apposition between them, and is supported by paired papillary muscle groups (Kanani and Anderson, 2003). Although usually described as being ''anterolateral'' and ''posteromedial'', in reality they are positioned anteroseptally and posterolaterally. Surgeons usually gain access to the mitral valve through the left atrium

behind the interatrial groove, via the right atrium and the atrial septum, or superiorly through the left atrial roof. When seen through any of these approaches, the aortic, or anterior, leaflet is superior and to the right of the mural, or posterior, leaflet (**Figure 2.33**). The aortic leaflet, so-called because it is in fibrous continuity with the leaflets of the aortic valve, occupies only one-third of the circumference of the mitral orifice, but the leaflet is much deeper than the mural one, so that the area occupied by the leaflets is about the same. The mural leaflet is usually divided into a series of mini-leaflets, or scallops, by marked slits. Three scallops are usually seen, along with two further components adjacent to the ends of the zone of apposition between the leaflets. The inferoseptal end of the zone of apposition, along with its so-called commissural leaflet, abuts against the atrial septum. This is the area of the valve most vulnerable to surgical trauma because the atrio-ventricular (av in **Figure 2.33**) node and bundle occupy the septum at this point. The attachment of the valve laterally may be closely related to the left coronary artery, a feature that is discussed in the section on the atrio-ventricular junction. The apical trabecular zone extends from the attachments of the papillary muscles, which are adjacent when the heart is unopened, to the ventricular apex. The trabeculations are characteristically fine, and a moderator band is not found. The outflow tract of the ventricle extends from the apical trabecular component to the attachments of the leaflets of the aortic valve. The margins between the apical trabecular and outlet portions are indistinct.

Posteriorly, the aortic leaflet of the mitral valve separates the inlet and outlet portions. Laterally, the left margin of the inner heart curvature supports the aortic valve, and the ventricular parietal wall forms the margin of the outflow tract, merging anteriorly with the ventricular septum. The septal surface of the outflow tract is characteristically smooth. If the surgeon requires direct access to the outflow tract through the anterior wall, as in corrected or complete transposition, care must be taken not to damage the papillary muscle when the ventricular incision is made. A discrete plane usually exists between the septum, marked by the anterior descending coronary artery, and the supero-lateral papillary muscle, through which good access can be gained to the outflow tract (**Figure 2.34**). The leaflets of the aortic valve, forming the distal margin of the left ventricular outflow tract, have partly muscular and partly fibrous support. The leaflets are supported over half their circumference by muscular tissues, viz. by the ventricular septum anteriorly and by the ventricular free wall and inner heart curvature laterally. This can be seen by transecting the outflow tract and viewing its anterior part from behind (**Figure 2.35a**). The rest of the circumference of the leaflets is supported by fibrous tissue, specifically the membranous septum and right fibrous trigone, together making up the central fibrous body, and the area of continuity between the leaflets of the aortic and mitral valves, with its thickened leftward extremity, forming the left fibrous trigone. This is best demonstrated by viewing the posterior part of the transected outflow tract from the front (**Figure 2.35b**). The three leaflets of the aortic valve are named the right coronary, left coronary and non-coronary leaflets. When the aortic valve is approached through the aorta, as seen

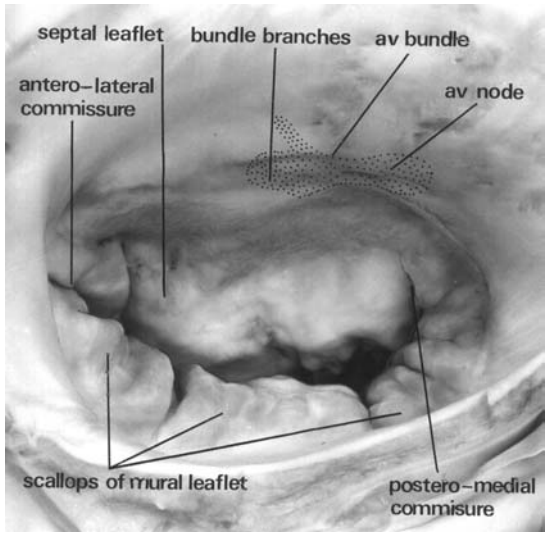


Figure 2.33

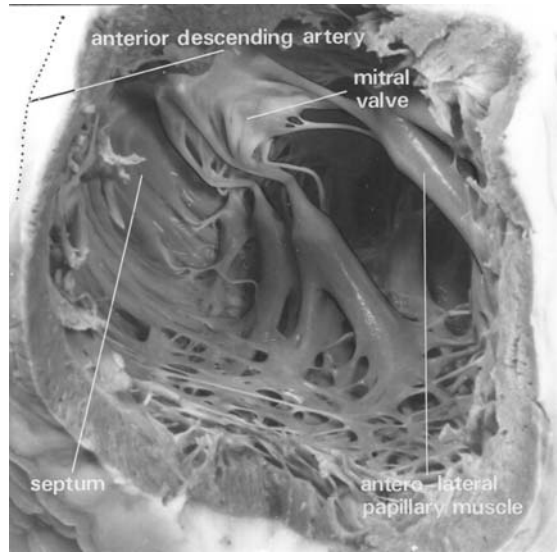


Figure 2.34

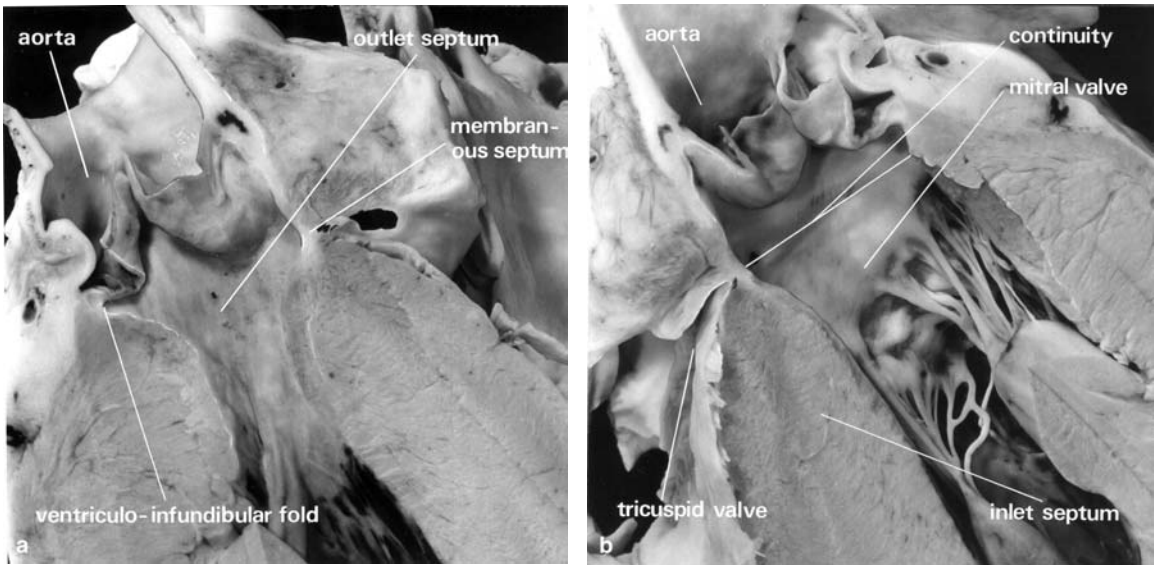


Figure 2.35

by the surgeon, the left coronary leaflet is on the left, the right coronary leaflet is anterior, and the non-coronary leaflet is posterior and to the right (**Figure 2.36**). The zone of apposition between the leaflets arising from the two aortic sinuses giving rise to the coronary arteries always faces a zone of apposition between the leaflets of the pulmonary valve (PV in **Figure 2.36**). The right coronary leaflet extends from this point to the area of the central fibrous body. The peripheral attachment of the zone of apposition between it and the non-coronary leaflet usually overlies the penetrating atrio-ventricular bundle. The non-coronary leaflet, at its attachment to the membranous septum, is in potential connexion with the right atrium and right ventricle. The attachment of its zone of apposition with the left coronary leaflet is normally positioned along the area of fibrous continuity, but this positioning is variable. At the peripheral attachment of the zones of apposition, the outflow tract is tented up, and the attachments reach onto the wall of the aorta. As in the right ventricle, this arrangement incorporates part of the arterial wall within the ventricular outflow tract. This is well shown by a dissection in which the left atrium and the tubular part of the aorta are removed (**Figure 2.37**). The tip of the tent in the area of fibrous continuity is separated from the transverse sinus of the pericardium. Aneurysms in this area can herniate directly into the pericardial cavity. Of equal significance, the apices of the other zones of apposition extend upward, so that the interleaflet triangles are again in potential continuity with the pericardial cavity. The site of the triangle separating the non-coronary and right coronary leaflets of the aortic valve is particularly important because it incorporates the area of the membranous septum. This, in turn, overlies the point of penetration of the atrio-ventricular conduction axis (**Figure 2.38**). When approached from the aorta, therefore, the area most vulnerable, from the standpoint of the conduction tissues, to surgical trauma is the area related to the zone of apposition between the right coronary and non-coronary

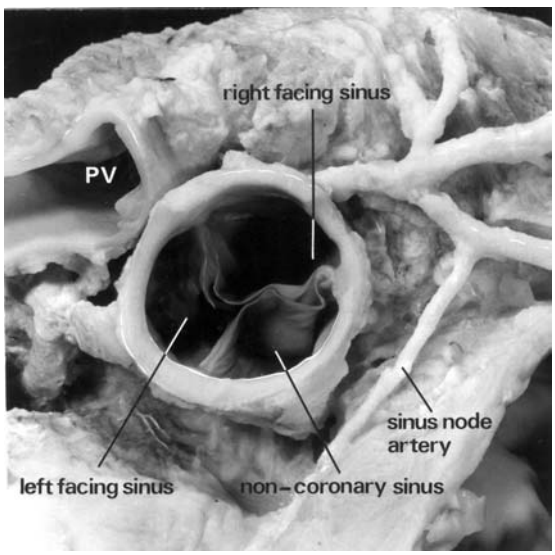


Figure 2.36

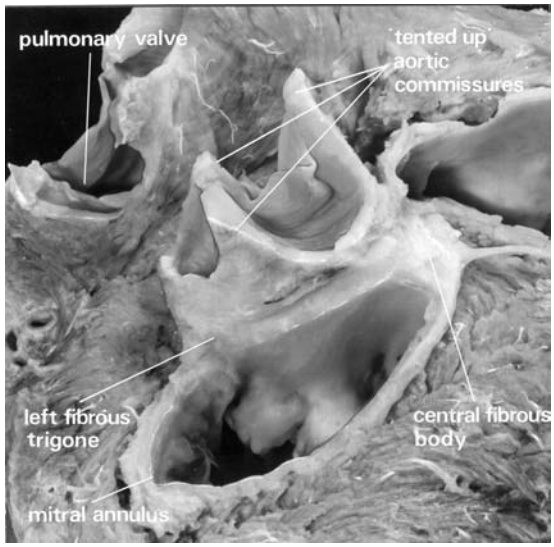


Figure 2.37

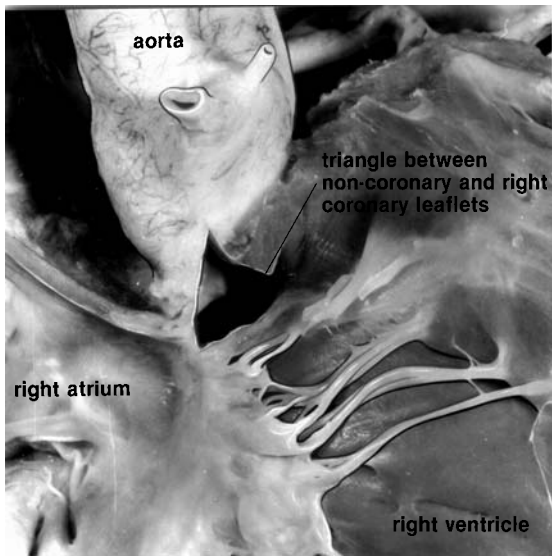


Figure 2.38

leaflets. The apex of the zone of apposition between the facing, or coronary, leaflets of the aortic valve is also related to the outside of the heart, specifically to an extensive tissue plane separating the front of the aorta from the back of the pulmonary trunk (**Figure 2.39**). The extent of this interleaflet triangle is best seen anatomically after the subpulmonary infundibulum is removed from the right ventricle.

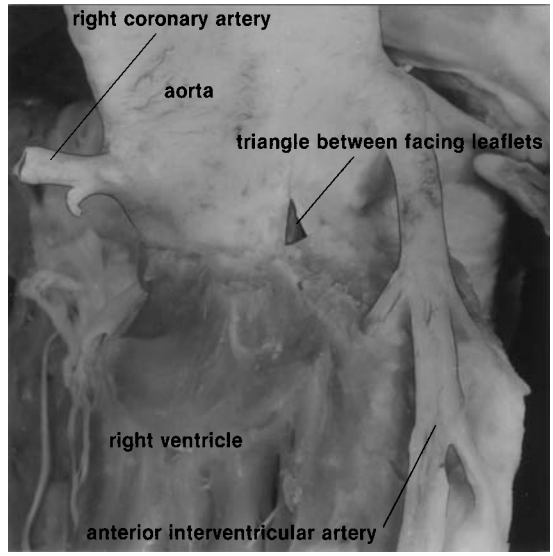
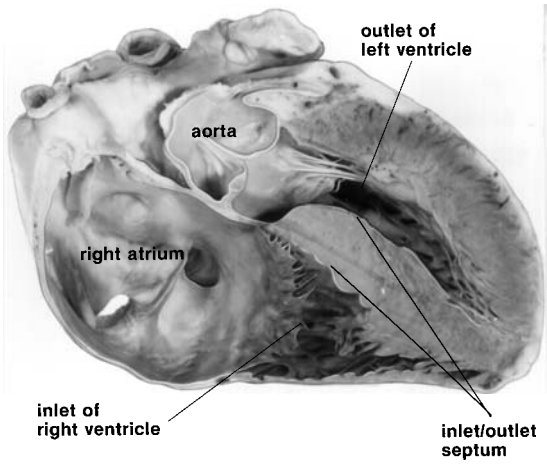


Figure 2.39

The Ventricular Septum

Although the ventricles can appropriately be divided into inlet, trabecular and outlet zones, it is over-simplistic to consider the muscular ventricular septum to be similarly divided into inlet, trabecular and outlet portions, although in the First Edition of this book we did propose those conventions. We now appreciate that our earlier concept is incorrect, having taken note of the location of the subaortic outflow tract between the mitral valve and the septum, and its relation to the right ventricle (Anderson *et al*., 2004; **Figure 2.40**). Thus, most of the inlet of the right ventricle is divided from the outlet of the left ventricle by this muscular ventricular septum. Furthermore, because of the difference in the level of attachment of mitral and tricuspid valves, part of the muscular septum is overlapped by atrial myocardium, separating the cavity of the right atrium from the left ventricle (LV in **Figure 2.41**). The situation is then further complicated because, as discussed earlier, the subpulmonary infundibulum is a free-standing sleeve of musculature, separating the inside from the outside of the heart (**Figure 2.42**; RV, right ventricle). Very little discrete outlet septum is recognizable as such in the normally structured heart. In the normal heart, therefore, it is most accurate to recognize the septum as having an extensive muscular part and a very small fibrous or membranous component. Most of this membranous component is itself positioned in an atrio-ventricular location because it is crossed on its right-sided aspect by the attachment of the septal leaflet of the tricuspid valve (**Figure 2.43**).



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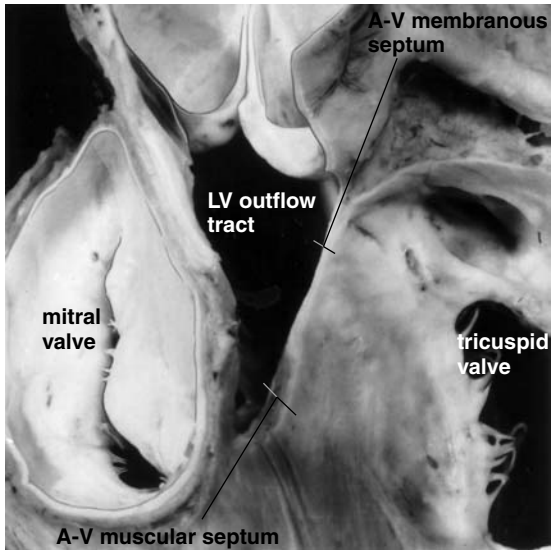

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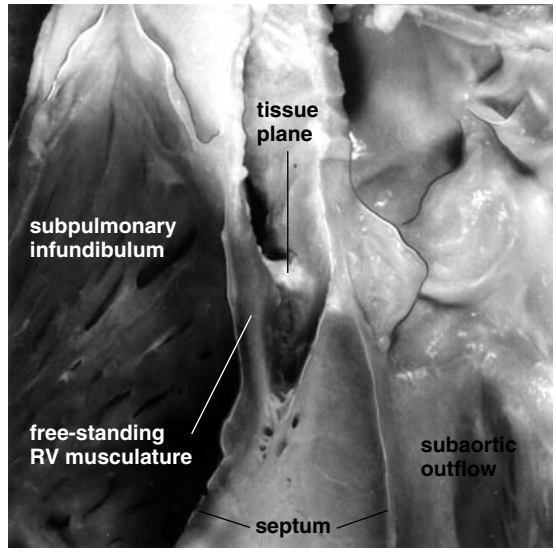

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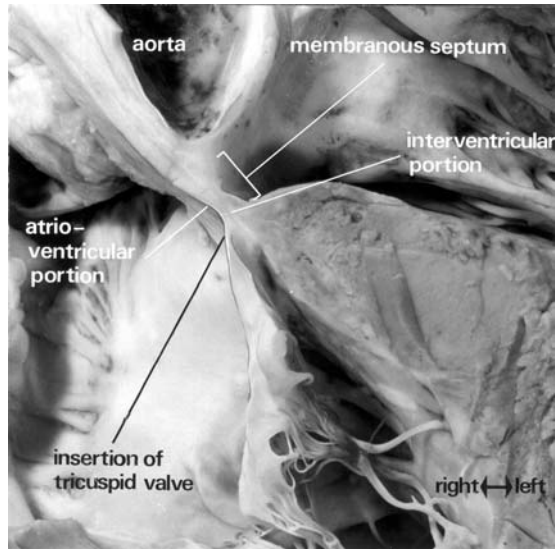

Figure 2.43

THE ATRIO-VENTRICULAR JUNCTIONS

The most vulnerable and complicated regions of the normal heart are the atrio-ventricular junctions, because they contain the origins of the leaflets of the atrio-ventricular valves, the atrio-ventricular conduction system and the major branches of the coronary arteries (Anderson *et al*., 2000). The areas are further complicated anatomically because the junction of the right heart chambers occupies a spatial plane different from that of the left heart chambers, the attachment of the leaflets of the tricuspid valve being situated more toward the ventricular apex than that of the leaflets of the mitral valve. The subaortic outflow, being firmly wedged between the orifices of the tricuspid and mitral valves, also forms an integral part of the junction. The attachments of the leaflets of the aortic, mitral and tricuspid valves form the fibrous skeleton of the heart.

THE CARDIAC SKELETON

The fibrous skeleton is variably developed around the atrio-ventricular junctions, being strongest at the central fibrous body (**Figure 2.37**). There, the attachments of the leaflets of the mitral, tricuspid and aortic valves all meet together. Part of the fibrous body in this position also forms the membranous portion of the cardiac septum. Because of the differing positions of attachments of the leaflets of the mitral and tricuspid valves to the central fibrous body, and because of the wedge position of the subaortic outlet, part of this septum is interposed between the aortic outflow tract and the atrium. This is best shown by a dissection in which the non-coronary sinus of the aorta is removed (**Figure 2.41**). Part of the fibrous body usually extends into the ventricles beneath the attachment of the tricuspid valve, and forms the interventricular component of the membranous septum. This component is tiny in comparison with the area of the muscular ventricular septum, but it is important to recognize it because the branching atrio-ventricular bundle is sandwiched between the membranous and muscular parts of the ventricular septum, so that the membranous septum can be an excellent landmark for the site of the branching bundle. The atrio-ventricular part of the membranous septum is interposed between the right atrium and the portion of the aortic outflow tract wedged between the tricuspid and mitral valves. Posterior to the membranous septum is a small area of the right atrial wall that is in potential communication with the inlet portion of the left ventricle, again by virtue of the difference in level of attachment of the leaflets of the tricuspid and mitral valves to the septum. This area of muscular overlapping, forming the muscular atrio-ventricular sandwich, is relatively small. This is because, posterior to it, the attachments of the leaflets of the atrio-ventricular valves diverge from one another and rapidly achieve attachments to the free parietal ventricular walls. In this region of valvar divergence, the coronary sinus extends along the postero-inferior aspect of the left atrio-ventricular junction and empties into the right atrial cavity, being very much a part of the left atrio-ventricular junction (**Figure 2.44**). The aortic and mitral valves form a very strong unit in themselves, and their adjacent parts form part of the fibrous skeleton. The skeleton is further strengthened at their left junction to form the left fibrous trigone (**Figure 2.37**). The aortic–mitral unit can easily be dissected free from the ventricular myocardium as a complete block of fibrous tissue. In contrast, the morphological features of the fibrous skeleton related to the tricuspid valve are much more variable. It varies in thickness from point to point in a single heart. Indeed, it is rare to find a complete fibrous ring that can be removed in a manner comparable with the removal of the supporting

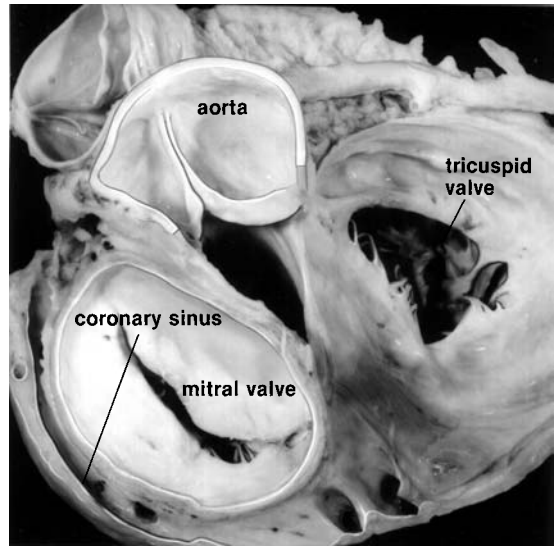

Figure 2.44

leaflets of the mitral valve. The leaflets of neither the aortic nor the pulmonary valves are supported by any annular structure in a way comparable with that of the mitral valve. The only circular structure is the anatomical ventriculo-arterial junction. As discussed earlier, this is crossed in semilunar fashion by the attachments of the leaflets of the arterial valves, so that it is the crown-like hinge points of the leaflets that form the surgical ‘‘annulus’’.

THE ATRIO-VENTRICULAR CONDUCTION SYSTEM AND BUNDLE BRANCHES

The atrial components of the atrio-ventricular conduction tissues are contained entirely within the area of right atrial wall known as the triangle of Koch. This is, in essence, the atrial component of the atrio-ventricular muscular sandwich, and the penetrating atrio-ventricular bundle is found at the apex of this triangle. If surgical procedures are conducted so as to avoid the confines of this triangle, it should not be possible to damage the atrial segments of the atrio-ventricular conduction tissues. The penetrating atrio-ventricular (av in **Figure 2.45**) bundle passes through the central fibrous body at the apex of the triangle and reaches the crest of the trabecular septum immediately beneath the interventricular component of the membranous septum. Here it becomes the branching bundle (bb in **Figure 2.45**). In normal hearts, the bundle branches as soon as it has penetrated, but in hearts with ventricular septal defects there may be an extensive non-branching bundle. In normal hearts, the right bundle

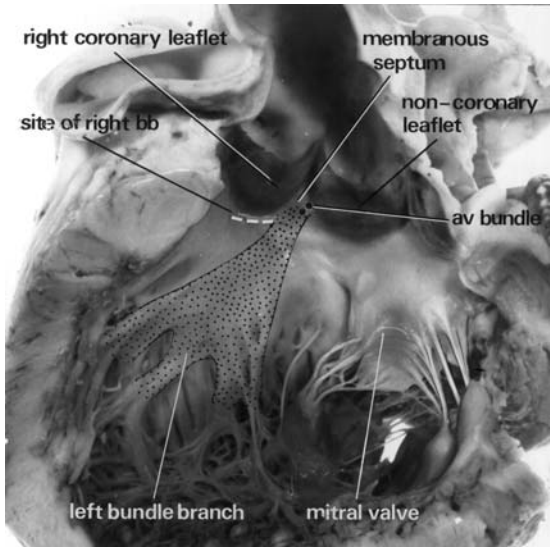

Figure 2.45

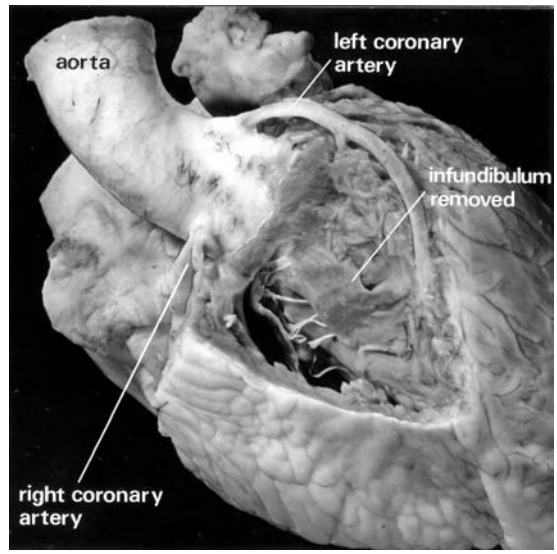

Figure 2.46

branch runs toward the base of the medial papillary muscle in an intramyocardial position and then descends toward the ventricular apex in the substance of the trabecula septomarginalis. It ramifies at the apex of the ventricle and part of it crosses the ventricular cavity in the moderator band. The left bundle branch descends from the crest of the ventricular septum as a sheet of cells over the smooth upper part of the septum. The point of penetration of the bundle, when viewed from the left, is usually adjacent to the interleaflet triangle between the right coronary and non-coronary sinuses of the aorta, but the branching bundle is always found immediately beneath the membranous septum (**Figure 2.45**). As it descends, the left bundle branch breaks up into a sheet of fascicles with interconnecting divisions. Three main divisions can usually be distinguished: anterior, middle and posterior. The anterior and posterior divisions extend toward the superolateral and inferoseptal papillary muscles, respectively, where they ramify into the ventricular myocardium.

THE CORONARY CIRCULATION

The major branches of the coronary arteries also run in the area of the atrio-ventricular junction. The right and left coronary arteries are the first branches of the aorta (**Figure 2.46**), emerging from the right and left coronary sinuses of the aorta, respectively (**Figure 2.47**; PV, pulmonary valve). The left coronary artery usually has a single orifice, but the first branches of the right coronary artery frequently originate directly from the right sinus as

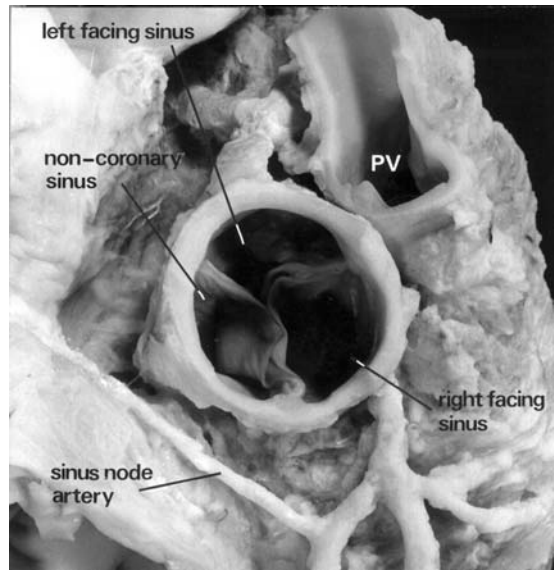

Figure 2.47

accessory orifices, particularly the infundibular and sinus nodal branches. In most hearts, the coronary orifices are inferior to the junction of aortic sinus with the tubular part of the ascending aorta, and are positioned mid-way between the two zones of apposition of the aortic leaflets, but in some hearts the arteries may originate above the junction or perforate close to a zone of apposition

(Muriago *et al*., 1997). Having emerged into the atrio-ventricular groove, the right coronary artery encircles the tricuspid orifice (**Figure 2.48**), whereas the left coronary artery divides immediately into anterior descending and circumflex arteries (**Figure 2.49**). The latter then encircles the mitral orifice. The arteries give off down-going branches to the ventricles and up-going branches to the atria as they pass around the atrio-ventricular junctions. The anterior descending coronary artery, the other main branch of the left coronary artery, descends

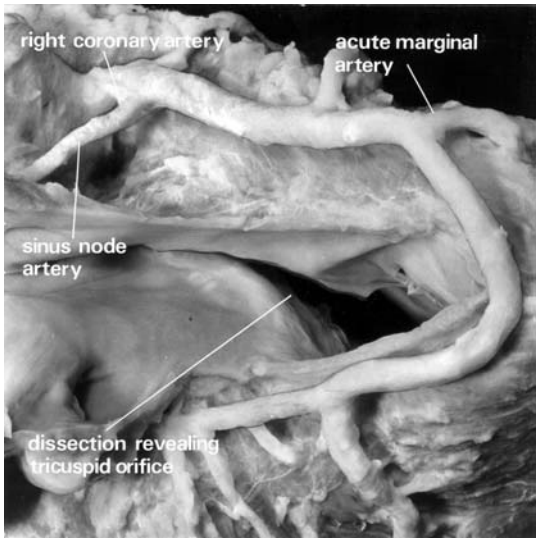

Figure 2.48

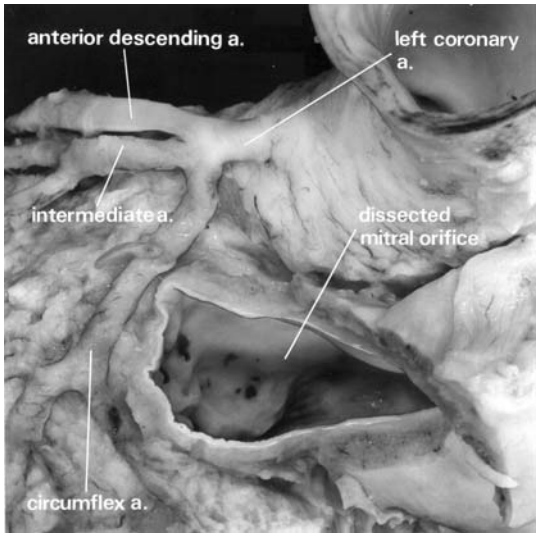

Figure 2.49

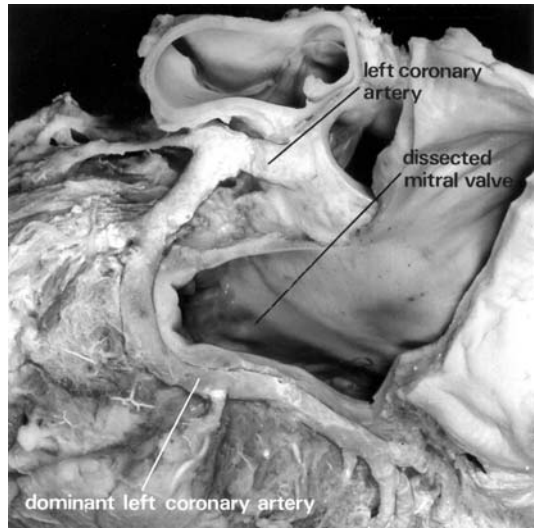

Figure 2.50

along the anterior interventricular groove. Another major artery, the inferior interventricular artery, descends through the inferior interventricular groove, but its site of origin varies. In the majority of hearts, approximately 90%, it is a branch of the right coronary artery, and this arrangement is termed ''right dominance''. In approximately 10% of hearts, in contrast, the inferior interventricular artery is the continuation of the left coronary artery, and this arrangement is called ''left dominance''. In a small proportion of hearts, both arteries give rise to interventricular branches. This arrangement is called a balanced circulation. Several of the atrial and ventricular branches are sufficiently constant to receive names. For the paediatric cardiac surgeon, the arteries to the sinus and atrio-ventricular nodes are important branches but have variable sites of origin. The artery to the sinus node arises as one of the first branches of the right coronary artery in 55% of humans, and from the left circumflex artery in most of the remainder. In a very small minority, it may originate as a lateral branch of either the right or the circumflex coronary artery. Unless it originates laterally, the artery ascends through the interatrial groove, frequently running intramyocardially, and then reaches the terminal groove by passing in front of, or behind, or by encircling the origin of the SVC. The artery to the atrio-ventricular node arises from the dominant coronary artery as it makes a U-turn in the atrio-ventricular groove at the crux. It then extends forward, running in the tissue plane in the atrio-ventricular muscular sandwich, to enter the node and then perforate the central fibrous body to supply part of the ventricular septum. The first descending

branch of the right coronary artery is the relatively constant infundibular branch, which may be multiple. Another constant branch, the acute marginal branch, descends as the right coronary artery bends around the tricuspid orifice at the acute margin (**Figure 2.48**). The main stem of the left coronary artery is usually short, rarely exceeding 1 cm in length (**Figure 2.49**). The length of the left circumflex artery depends on whether the left coronary is dominant (**Figure 2.50**) or, if not dominant, on the extent to which the right coronary artery supplies the diaphragmatic surface of the left ventricle. The left circumflex frequently gives rise only to an obtuse marginal branch. The major branches of the anterior descending coronary artery are the perforating arteries, which pass perpendicularly into the ventricular septum, supplying it and giving off small tributaries to the ventricular conduction tissues (Hosseinpour *et al*., 2001). Another branch that usually arises from the anterior descending coronary artery is an oblique branch that passes backwards to supply the obtuse area of the ventricular mass. Most of the coronary arterial branches, except for the perforating branches, occupy directly epicardial positions. The myocardial branches of the epicardial coronary arteries turn perpendicularly into the muscle and penetrate toward the endocardium. During this penetrating course, further branches arise at right angles to the perpendicular branches and ramify among the fibre bundles. An extensive network of branches is found in the subendocardial layer running parallel to the endocardial surface. The blood supply from the coronary arteries is collected by the coronary veins, which drain either into major venous channels or directly to the cardiac chambers. The major venous channels run along with the coronary arteries and drain into the coronary sinus. The middle cardiac vein ascends into the coronary sinus from the inferior interventricular groove, and the small cardiac vein runs up alongside the marginal coronary artery before turning to enter the inferior atrio-ventricular groove with the right coronary artery. Veins from the atrial chambers also drain into the coronary sinus. The oblique vein of the left atrium, and the ligament of Marshall, are vestiges of the embryonic left sinus horn and persist as the termination of a left SVC when such a channel draining to the coronary sinus is present. The heart also possesses an extensive lymphatic network, divided into deep, middle and superficial plexuses. All drain eventually into collecting channels that accompany the major arterial stems and finally into primary lymph nodes in the anterior mediastinum.

Acknowledgements

We are indebted to Siew Yen Ho for her help in preparing the illustrations, and to Wilfred Meun and Ruud Verhoeven for their photographic skills. We also thank Dr James R Zuberbuhler for permitting us to use photographs of normal hearts from the cardiopathological museum of the Children's Hospital of Pittsburgh, PA, USA.

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3

Echocardiography

I. D. Sullivan and V. M. Gooch

Analysis of structural heart disease in infants and children is ideally suited to two-dimensional (2D) and Doppler echocardiography. Infinite imaging planes are available for displaying anatomy, which is frequently complex. In comparison with cardiac structures in adults, those in infants and children are close to the body surface, which means that higher-frequency ultrasound, which has poorer tissue penetration but provides better image resolution, can be used. Doppler signals of better quality can also be obtained more easily in infants and children because of the shorter distances involved. The technique is non-invasive, painless and safe; these considerations are important in all patients but especially so in neonates and infants who are haemodynamically unstable. Echocardiographic studies can be performed in the intensive care unit, operating theatre or catheter laboratory, serial studies are easy to conduct, and the technique is affordable in comparison with other forms of imaging and haemodynamic measurement.

BASIC PRINCIPLES

M-Mode and Two-dimensional Echocardiography

An echocardiography transducer converts electrical energy to ultrasound waves by stimulation of a crystal. The emitted signal reflects off tissue and tissue interfaces, and the returning ultrasound wave is converted back to electrical energy. Cardiac ultrasound was introduced to clinical practice in the early 1970s with the development of single-line echocardiography. This requires a motion (M)-mode transducer to emit and receive bursts of ultrasound waves in a single beam. Because the velocity of ultrasound waves through tissue is known, the depth from which the reflected ultrasound wave originated can be determined. The M-mode signal is recorded on an oscilloscope or a strip chart recorder to display cardiac motion over time, and such recording is a good technique for measuring cavity dimensions and wall thickness and their rates of change, and can be recorded simultaneously with other physiological measurements (**Figure 3.1**). However, it was superseded in the diagnosis of structural heart disease by 2D echocardiography, developed in the late 1970s, which has since been also used to select the sites for M-mode sampling (**Figure 3.1**). 2D echocardiography provides images of a sector of the heart. The ultrasound beam is swept through an arc to produce a sector image made up of many adjacent lines of M-mode information. Each set of lines constitutes a single frame. Under most circumstances, sufficient frames per second are obtained to give the visual illusion of continuous motion of the heart. In recent years, increasingly sophisticated formation of the transmitted ultrasound waveform and processing of the received echo has been developed. This has led to improved spatial resolution, greatly increased frame rates and progressive improvement in image quality. Multiple frequency transducers have been developed, allowing operator selection of a range of transmit and receive ultrasound frequencies. This includes harmonic imaging, which is the capability of a transducer to transmit an ultrasound pulse of a fundamental frequency but to analyse only the returning second harmonic frequency echo. For example, the transmitted fundamental frequency may be 2 mHz but suitable filtering allows analysis of the weaker second harmonic echo received at 4 mHz, which improves the signal-to-noise ratio and produces improved images. The improved ultrasound tissue penetration provided by harmonic imaging is not often needed in infants or small children, but may be valuable in larger subjects, or in foetal echocardiography.

Doppler Flow Echocardiography

Ultrasound waves backscattered by red blood cells are of much lower amplitude than those reflected from tissue and are usually filtered out of the signals used to display the 2D image. Sound emitted by or reflected off a moving object has a frequency change that depends on the velocity and direction of the object relative to the observer. This change in frequency is the Doppler

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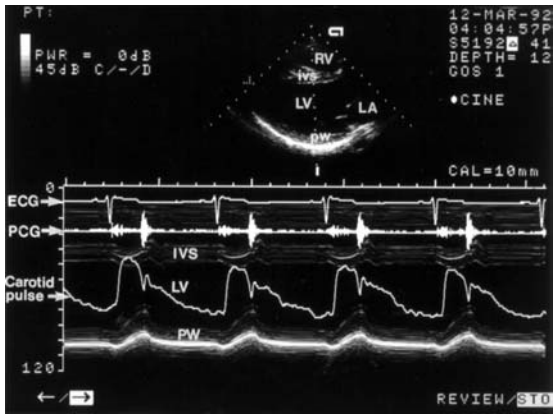


Figure 3.1

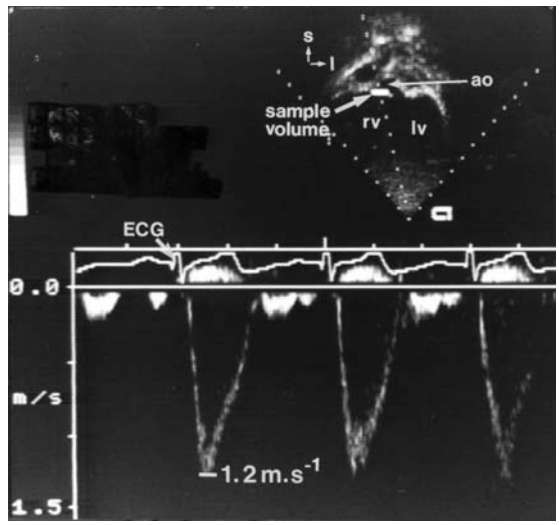


Figure 3.2

shift. The phenomenon is experienced when a speeding vehicle, such as a racing car or train, passes an observer. As the vehicle approaches at high speed the sound wave frequency perceived is increased, but as the vehicle passes and moves away the perceived frequency abruptly decreases. The Doppler principle enables information about velocity and direction of blood flow to be obtained by ultrasound. The frequency of the reflected ultrasound wave is compared with that of the transmitted signal: the frequency of the signal reflected from stationary objects is unchanged, whereas the frequency of ultrasound waves reflected off red blood cells moving toward the transducer is increased and that of ultrasound waves reflected off red blood cells moving away from the transducer is decreased. The frequency shift is proportional to the velocity of the moving red blood cells.

The frequency shifts that are observed in Doppler examination of normal or disturbed blood flow occur within the audible range of frequencies, so Doppler audio signals are helpful to assist alignment of the ultrasound beam with either normal or disturbed flow. Spectral display of the signal is obtained from sophisticated mathematical processing of the complex Doppler waveforms and enables quantitative analysis of Doppler information. The intensity of each part of the spectral signal is related to the quantity of red blood cells flowing at a particular velocity and direction. The range of velocities for normal flow is narrow (**Figure 3.2**), whereas that for disturbed or turbulent flow is wide (**Colour Plate 1**). By convention, flow away from the transducer is displayed below the zero line, whereas flow towards the transducer is displayed above the zero line.

A continuous-wave Doppler system emits ultrasound waves of known frequency continuously from one crystal and receives reflected ultrasound waves on an adjacent crystal. Motion at any part of the course of the transmitted beam contributes to the frequency shift in the reflected signal. High-frequency shifts, and hence high blood flow velocities, can be recorded accurately because of the continuous sampling (**Colour Plate 1b**).

A pulsed Doppler system is similar to M-mode echocardiography, in that the same crystal emits and receives ultrasound waves. Pulsed Doppler and 2D imaging capability are combined in the same transducer, so that the anatomical image can be used to position the Doppler sample volume (**Figure 3.2**). A burst of ultrasound waves is transmitted and, after a delay, the reflected ultrasound waves are received before the next burst is transmitted. The delay is determined by the depth of the sample volume from the scan head; it is the time required for transmitted ultrasound waves to make the round trip from the transducer to the region of interest and back again. Consequently, the flow velocities detected are exclusively from this selected site. The rate at which the bursts of ultrasound are emitted is the sampling rate, or pulse repetition frequency, and is determined by the depth of the sample volume; the greater the depth, the lower the pulse repetition frequency. The maximal frequency shift that can be determined unambiguously is half the pulse repetition frequency, because above this limit (Nyquist's limit) frequency shifts in the opposite direction are interpreted. This phenomenon is termed ''aliasing'', and it accounts for the apparent changes in direction of rotation perceived as a spoked wheel or fan accelerates. The resulting inability to quantify high flow velocity is the main disadvantage of conventional pulsed Doppler examination.

Colour-flow Doppler Interrogation

Colour-flow Doppler interrogation is performed over a selected portion of the anatomical 2D image, whereby pulsed Doppler sampling occurs at many points on each line of the ultrasound beam within this chosen portion. The mean velocities at each sampling site are colour-coded according to the direction and velocity of flow. Normally, shades of red are assigned for flow toward the transducer, and shades of blue are assigned for flow away from the transducer. This spatially orientated colour-flow information is superimposed on the anatomical image (**Colour Plate 2**), (a) systolic and (b) diastolic frames after repair of tetralogy of Fallot, showing diastolic pulmonary regurgitation). Frequency aliasing occurs at lower flow velocities than with conventional pulsed Doppler interrogation because the need to interrogate multiple sampling sites means that the sampling rate for each site is reduced. However, in contrast to conventional pulsed Doppler interrogation, this is not necessarily a disadvantage, because the spatial display of flow velocities enables easy recognition of aliasing and helps identify the fastest flow region (**Colour Plate 1**).

Colour-flow Doppler imaging is largely a semi-quantitative technique (Thomas, 2002), but its value is in the eye-catching demonstration of abnormal flow. Interrogation of the selected site of disturbed flow can be undertaken by conventional spectral analysis by superimposing a pulsed Doppler sample volume or a beam of steerable continuous-wave Doppler onto the colour-flow image (**Colour Plate 3**, right ventricular outflow obstruction and pulmonary incompetence in ''absent pulmonary valve syndrome'', showing diastolic colour aliasing and continuous wave Doppler spectral analysis).

Tissue Doppler Imaging

The Doppler ultrasound signals backscattered from moving red blood cells are of relatively high velocity but low amplitude, compared to the high-amplitude, low-velocity signals reflected from the moving myocardium or atrio-ventricular valve rings. Doppler flow systems are designed to filter low-velocity signals but can be adjusted to detect only low-velocity, high-amplitude signals, so that the Doppler signals from the moving blood are filtered out. Myocardial velocities can then be measured from specific myocardial sites using pulsed Doppler. This myocardial motion can be colour-coded, analogous to colour-flow Doppler, and visualized in ''real'' time or displayed as spectral pulsed Doppler signals. Tissue Doppler imaging has been used to assess both systolic and diastolic ventricular performance (Weidemann *et al*., 2002; Pellerin *et al*., 2003), although this has been confined mainly to research applications to date.

Three-dimensional Echocardiography

During the past decade, attempts have been made to acquire three-dimensional (3D) ultrasound data sets using a conventional echo transducer mounted on a mechanical rotational device. The data have been awkward to acquire, and have then required time-consuming off-line analysis to construct ''blocks'' of ultrasound data, using extensive signal processing to smooth the displayed image. The recent development of transducers which allow electronic acquisition of 3D data sets in real time make the technique much more attractive for potential clinical use. This allows immediate or off-line ''virtual dissection'' by electronic slicing of a block of acquired ultrasound data, which can be orientated on the screen according to operator choice [**Figure 3.3**, (a) systolic and (b) diastolic frames in atrio-ventricular septal defect]. This means, for example, that an atrio-ventricular valve can be viewed from the ventricular aspect, and the ultrasound block then reorientated and sliced to the level where valve closure can be observed from the atrial aspect, analogous to the surgeon's view. This is a developing field, but the most exciting prospect in the assessment of congenital heart defects is the ability to ''view'' cardiac structures from perspectives not available with conventional 2D echocardiography. This may be helpful in situations such as planning atrio-ventricular valve repair or possibly complex intraventricular repair, such as may be required in some forms of double-outlet right ventricle.

Quantitative Measurement

Echo measurement of ventricular shortening, wall thickness and cavity size remains a cornerstone of the assessment of ventricular function, hypertrophy or volume load. (**Figure 3.4**, parasternal long axis in diastole, left, and systole, right, showing systolic apposition of the mitral valve and the septum as the mechanism of left ventricular outflow obstruction in hypertrophic cardiomyopathy). Contemporary clinical surgical decision making in many situations relies heavily on relatively simple M-mode (Cheung *et al*., 2003) or 2D (Lofland *et al*., 2001) measures of chamber, artery or valve dimensions indexed to body size. When pulsatile structures are measured for serial comparison, such as segmental aortic root dimensions in Marfan syndrome, it is important to take account of the phase of the cardiac cycle at which the measure is made. The same applies to measures made for patient or equipment selection for various interventional catheter procedures, and preoperative selection of prosthetic or homograft valve size.

Left ventricular fractional shortening measured by M-mode (**Figure 3.1**) is the most convenient clinical

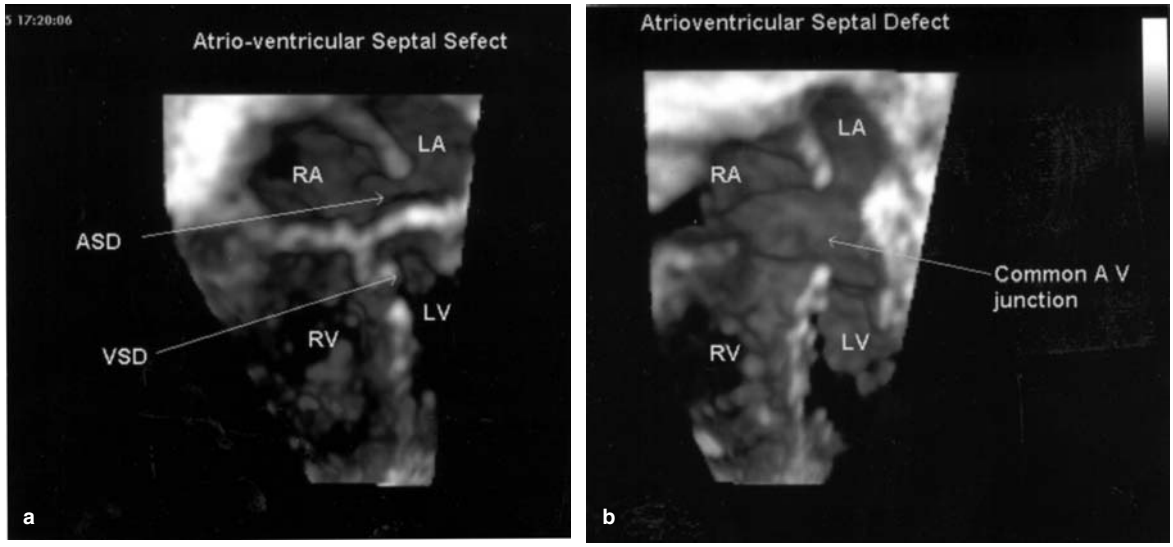


Figure 3.3

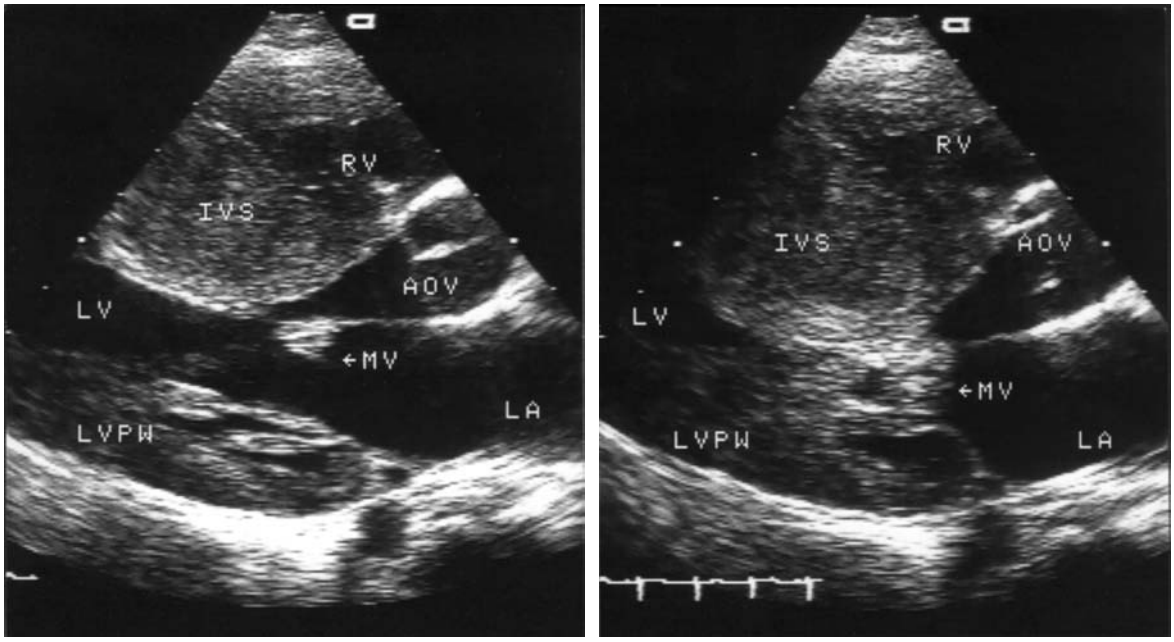


Figure 3.4

estimate of ventricular function:

$$\text{fractional shortening} = \frac{\text{diastolic dimension} - \text{systolic dimension}}{\text{diastolic dimension}}$$

Normal left ventricular fractional shortening is > 28% and is essentially independent of age. This value can be converted to an estimate of ejection fraction by formulae which make assumptions about left ventricular geometry. This may be useful for comparison with data from angiography, magnetic resonance (MR) or radionuclide imaging. Measures of left ventricular function, such as fractional shortening or ejection fraction, are highly dependent on ventricular loading conditions (preload, afterload, heart rate). Methods to measure left ventricular intrinsic contractility, independent of load using M-mode echo, have been developed. These have been useful in many clinical studies, but the methods are cumbersome and time consuming, and depend on relatively normal left ventricular shape and absence of wall motion abnormalities, which limits the suitability for many patients with congenital heart defects. Consequently, the search for a convenient measure of ventricular performance has received much attention. A non-invasive measure recently described is isovolaemic acceleration, measured by tissue Doppler at the very onset of ventricular contraction so that the influence of loading conditions should be minimized (Vogel *et al*., 2003).

Doppler measurement of flow volume requires calculation of the mean velocity of flow obtained by integration of the spectral signal, together with the 2D cross-sectional area of the site at which flow is measured. Cardiac output can be measured in this way, but accurate measurement of the area of flow is particularly difficult, limiting this application in practice. Conversely, one of the most practical benefits of Doppler echocardiography is the ability to measure pressure gradients. The peak flow velocity through a stenosis is related to the drop in pressure. For most discrete narrowings, the systolic pressure gradient can be estimated from the simplified Bernoulli equation ($p = 4v^2$, where p is peak instantaneous gradient in millimetres of mercury and v is maximal trans-stenotic velocity in metres per second). Colour-flow demonstration of the jet is very helpful in guiding optimal alignment of a continuous-wave Doppler beam with the area of fastest flow (**Colour Plates 1 and 3**), and the audio signal assists in fine-tuning of transducer position. Doppler imaging enables estimation of the peak instantaneous pressure gradient. In comparison, prior to the availability of reliable Doppler information, peak-to-peak systolic pressure differences measured at cardiac catheterization were conventionally assessed. In practice, the two measurements are similar in many, but not all, circumstances. Typically, the simplified Bernoulli equation is used to assess the peak and/or mean systolic pressure gradient across a discrete ventricular outflow tract obstruction. Ventricular systolic pressure can be estimated by determining the maximal velocity of an atrio-ventricular valve regurgitant jet (**Colour Plate 1**). The simplified Bernoulli equation cannot be applied indiscriminantly. For example, in long tubular narrowings, such as prosthetic systemic-to-pulmonary artery grafts, the simplified Bernoulli equation typically underestimates the systolic pressure difference, whereas the systolic pressure difference may be overestimated if flow velocity immediately proximal to the obstruction is increased, as may occur in multi-level right ventricular outflow tract obstruction or coarctation of the aorta. Persistence of flow in diastole (**Colour Plate 4**) is an important indicator of obstruction in the latter situation. The peak flow velocity through a ventricular septal defect (VSD) will provide an estimate of the systolic pressure difference between left ventricle and right ventricle, but interpretation must take into account that left ventricular systolic pressure usually increases faster than right ventricular systolic pressure. Left ventricular systolic pressure also rises faster than ascending aorta pressure, which must be considered in the interpretation of Doppler-derived peak instantaneous left ventricular outflow tract systolic pressure difference. It may be helpful to estimate mean systolic pressure gradient in this situation. Mean gradient should be estimated in atrio-ventricular valve stenosis or venous narrowing, such as pulmonary venous pathway obstruction after a Mustard or Senning operation.

PRACTICAL CONSIDERATIONS

Image Orientation

The anatomical coronal, sagittal and transverse planes of the body cannot be used conveniently in the description of tomographic cardiac structure, and so the axis of the heart is used to define the conventional long-axis, short-axis and four-chamber echocardiographic views (**Figure 3.5**). Images in each of these orthogonal planes can be obtained from several transducer locations. For example, the four-chamber plane may be imaged from a number of subcostal, apical or oesophageal transducer positions. Transducer locations must often be adapted in congenital heart defects to take account of abnormal cardiac positions in the thorax and complex intracardiac anatomy. In order to obtain a 3D description of cardiac structure from a series of 2D images, the scan head is moved in sweeps from each transducer position, so that the real-time image conveys information about the relation of adjacent structures that is not apparent from images of single frames. The operator must correlate the

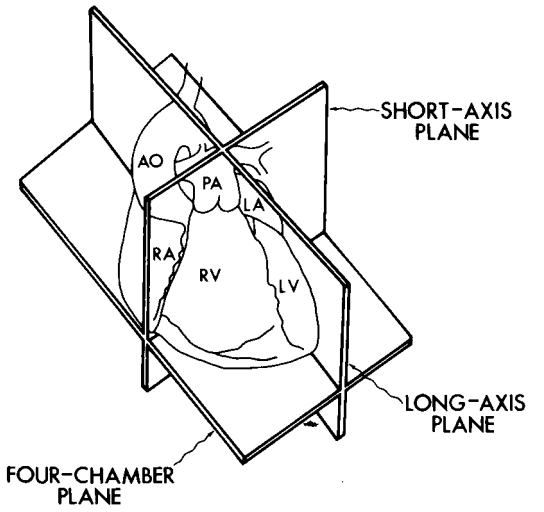


Figure 3.5

real-time images with the movement of the scan head, so as to demonstrate and describe the anatomy.

The presentation format of echocardiographic images can be confusing. Early 2D echocardiography machines did not include the ability to invert the sector image, and so the part of the heart closest to the transducer was always at the top of the screen. Consequently, it was necessary to present the heart in an upside-down format when imaged from a subcostal or an apical location. This is not necessarily a disadvantage with conventional cardiac anatomy, but it may make the analysis of complex structural abnormalities more difficult. Electronic inversion of the sector meant that views from subcostal and apical locations subsequently could be presented in an anatomical format (Gutgesell, 1985). This undoubtedly makes echocardiography of complex congenital heart defects an easier skill to acquire, particularly because the best acoustic window in infants and small children is usually subcostal, and allows easier correlation with conventional angiographic, MR or CT images. It must be remembered that some manufacturers do not invert the colour-flow velocity indicator when the image is inverted, so that the direction of flow is the opposite to that displayed in the colour flow map (**Colour Plates 1 and 3**), but this is usually obvious in real-time display. Parasternal long-axis views are conventionally presented with the great arteries to the observer's right on the screen, which may be thought of as examining slices of the heart from the supine patient's left side, and parasternal short-axis views are presented with the patient's left side to the observer's right on the screen, which is like viewing the heart from below. Suprasternal images are usually referred to the anatomical planes and to the long axis of the aortic arch.

Trans-oesophageal Echocardiography

Inserting an echocardiography transducer mounted on a flexible endoscope into the oesophagus and stomach provides a posterior window to the heart. Until the late 1980s, only transverse imaging planes at right angles to the shaft of the probe were available through trans-oesophageal echocardiography (TOE). Biplane TOE transducers, which utilize two separate ultrasound crystals adjacent to each other to also permit imaging in planes parallel to the shaft of the probe, were next developed (**Figure 3.6**, TOE images of a previously undiagnosed large ASD in an adult, with bidirectional flow, upper right panel. Parasagittal imaging in the plane of the caval veins, lower panels, shows that the inferior rim is deficient, lower left, making the hole unsuitable for device closure). Subsequently, biplane imaging was superseded by multiple plane transducers, in which the ultrasound crystal is mounted on a motorized dial to allow imaging in an infinite number of planes, and the display of sweeps of imaging in a manner analogous to trans-thoracic echo. Suggested orientations of these views vary (Stumper *et al*., 1990b; Seward *et al*., 1993). However, inversion of the image, with the optional exception of transverse four-chamber views from the mid-oesophagus (**Figure 3.6**), enables easier correlation with conventional praecordial echo images and with other forms of cardiac imaging. This format, popularized by the Mayo Clinic (Seward *et al*., 1993), is our preference and that of most congenital heart units, even if image orientation differs from that still employed by most adult cardiologists.

Assessment of posterior cardiac structures by conventional echocardiography may be limited by poor ultrasound wave penetration. This limitation is exacerbated when prosthetic material produces an acoustic shadow that prevents acquisition of imaging or Doppler information. The additional problem with colour-flow mapping is that long distances from the skin surface result in low frame rates. Consequently, TOE rapidly acquired a role in adults, for example in the assessment of mitral regurgitation and left atrial appendage thrombus, but similar applications may apply in children (**Figure 3.7**, thrombus in left atrial appendage after atrio-ventricular septal defect repair, not obvious on trans-thoracic imaging, left, but readily apparent on TOE, right). However, ultrasound penetration from conventional transducer locations is excellent in infants

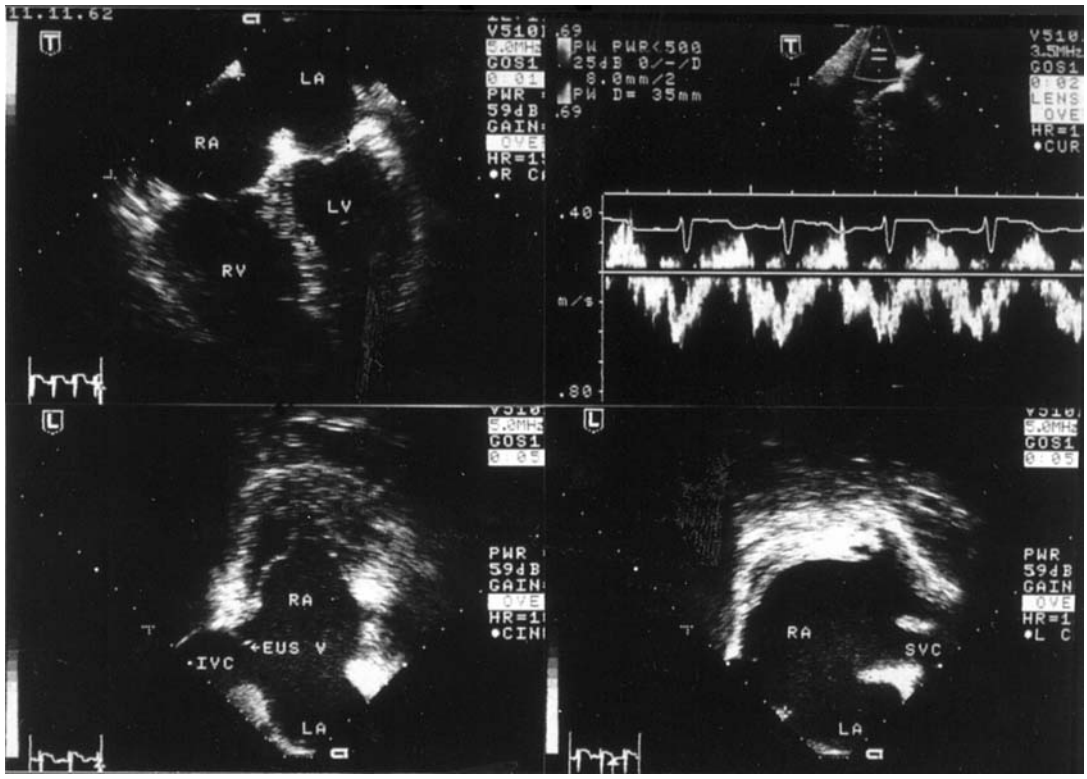


Figure 3.6

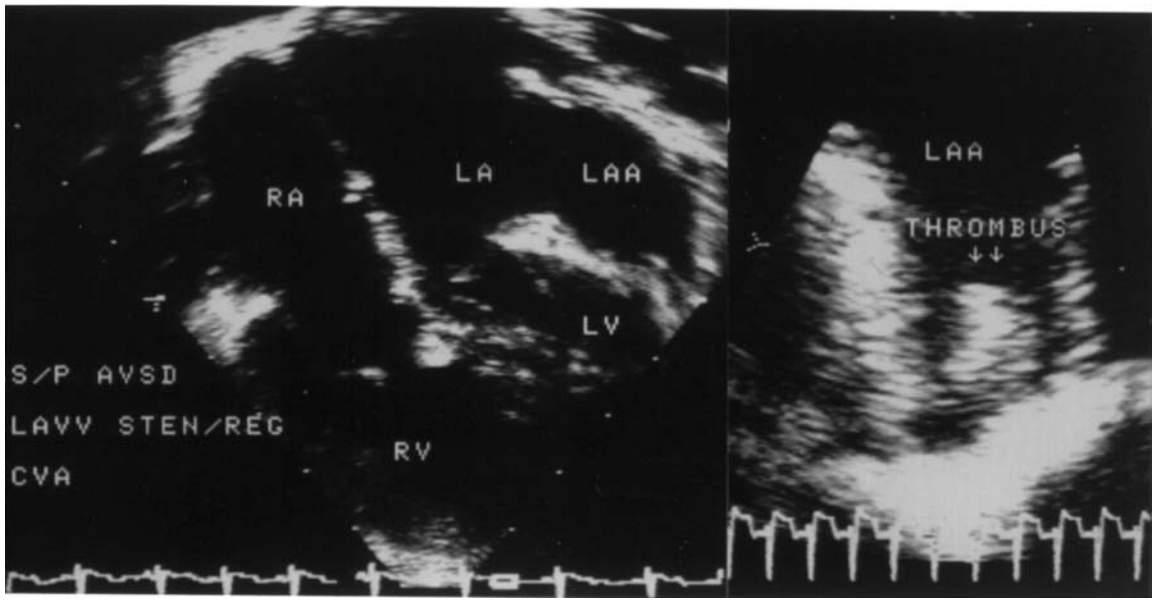


Figure 3.7

and small children, and even posterior mediastinal structures are not far from the scan head. Until the late 1980s, only adult-sized TOE transducers were available, which made the technique potentially more dangerous in children, and especially in infants. It is also uncomfortable, indicating the use of general anaesthesia in children. Increasing miniaturization of TOE probes, even multiplane probes, extended the technique to small children and infants, particularly for intraoperative and early postoperative studies and simultaneous studies, are used to guide many interventional catheterization procedures [**Figure 3.8**, (a) short and (b) long axis TOE views of large perimembranous VSD, closed by device (c) using TOE guidance].

Archiving

M-mode echo images were originally stored using strip chart recorders. The advent of 2D echo was accompanied by the wide availability of videotape, which provided an

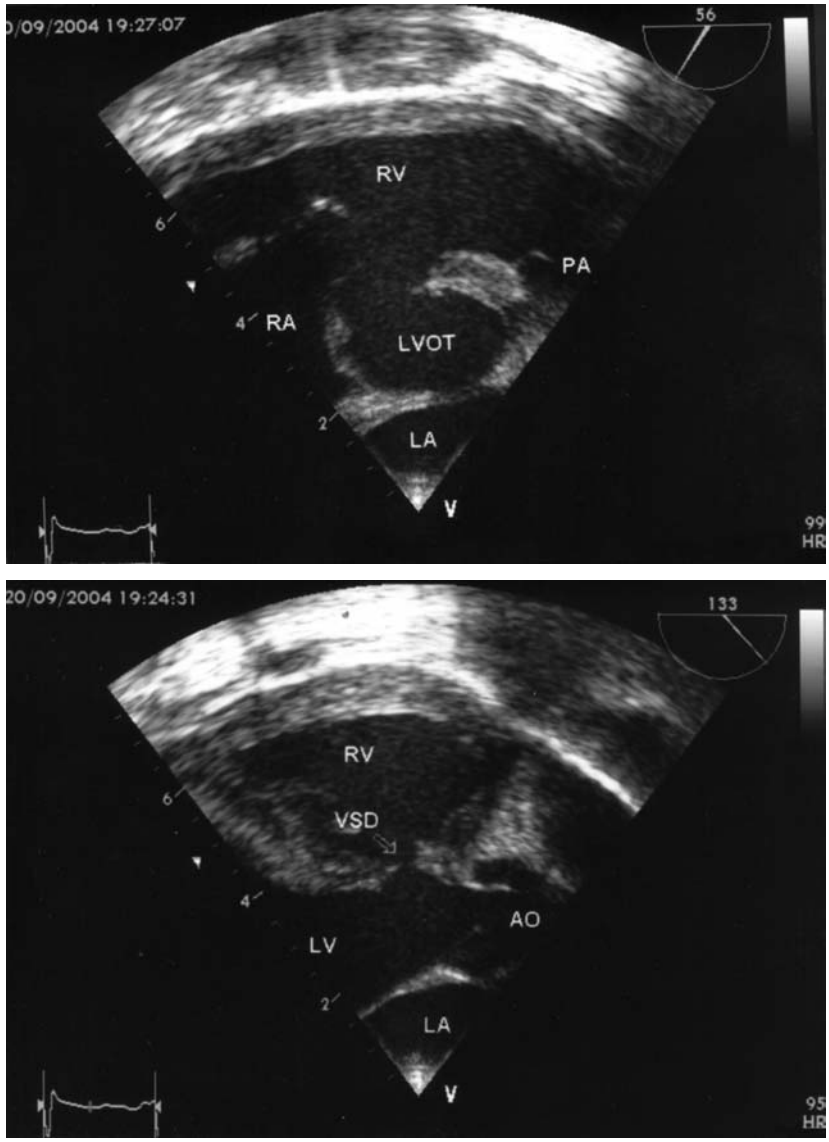


Figure 3.8

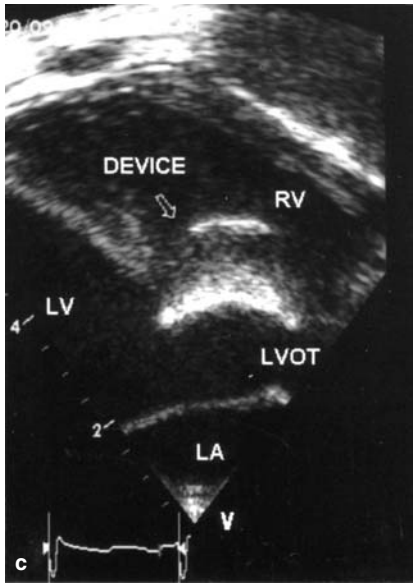


Figure 3.8 (*continued*).

ideal medium for archiving of moving cardiac images. Recently, digital archiving and storage has gradually been superseding videotape. This trend has been slower in paediatric than in adult cardiology, because complex structural heart disease is less well suited to archiving in a clip format of 1 or 2 cardiac cycles than most applications in adult cardiology, where the emphasis will typically be detecting or comparing regional wall motion abnormalities. Nevertheless, multiple beat or several seconds of time-compressed information can now be archived, which diminishes this problem. Advantages of digital archiving include convenient storage, postprocessing of data, and remote, readily accessible viewing in venues such as the operating theatre or intensive care unit. Continuing problems include the major capital outlay required in a field with rapid technology changes and issues of compatibility between equipment manufactured by many different vendors.

SEQUENTIAL SEGMENTAL ANATOMY

The sequential segmental approach to cardiac morphology provides a convenient framework on which to base the description of cardiac anatomy and also facilitates a systematic approach to echocardiographic examination. This makes it less likely that important lesions receive detailed attention but that subsidiary additional abnormalities are overlooked. The heart is normally positioned in the left side of the chest, with the cardiac apex directed to the left. An abnormal position of the heart should be described. For example, this might be dextrocardia, meaning that the heart is situated predominantly in the right side of the thorax, with the cardiac apex directed to either right or left. Examples of abnormalities that might be detected at each stage of segmental echocardiographic examination are described below.

Atrial Situs (Arrangement) and Venous Connections

In most patients, atrial situs (arrangement) is normal (situs solitus). However, the abnormalities of cardiac position and intracardiac anatomy that typically accompany atrial isomerism are easier to analyse if the abnormal atrial situs is ascertained at the outset of the examination. The atrial appendages can be imaged (**Figure 3.7**), but in practice it is much easier to infer atrial arrangement from the anatomy of the abdominal great vessels just inferior to the diaphragm. In situs solitus the inferior vena cava (IVC) is positioned to the right of the spine, and the descending aorta is to the left, usually quite close to the midline (**Figure 3.9a**). In situs inversus (mirror-image atrial arrangement) the IVC is to the left of the spine (**Figure 3.9b**). The morphological right atrium is lateralized to the same side as the IVC. Hepatic veins join the IVC just below its junction with the right atrium. In right isomerism, both the descending aorta and the IVC are to the same side of the spine, and the IVC is anterior to the aorta (**Figure 3.9c**). In left isomerism, the suprarenal portion of the IVC is usually absent, and there is azygos or hemi-azygos continuation of the IVC behind the heart to join either the right- or left-sided superior vena cava (SVC); the azygos continuation lies posterolateral to the descending aorta below the diaphragm (**Figure 3.9d**), and the hepatic veins usually drain separately to the heart (**Figure 3.10**, hepatic veins draining to atrial mass in left isomerism).

In the superior mediastinum, the innominate and right internal jugular veins join to form the SVC (**Figure 3.11**). The innominate (brachiocephalic) vein normally passes anterior to the proximal part of the innominate (brachiocephalic) artery. In occasional patients, especially when tetralogy of Fallot is present, it may pass posterior to the ascending aorta (**Figure 3.12**). It is important to be aware of this abnormal anatomy before operation, so as to avoid inadvertent damage to the innominate vein during dissection of the great arteries. Knowledge of the presence of a left-sided SVC is necessary for planning venous cannulation techniques, and its presence does of course modify surgical strategies in some complex heart defects. A left-sided SVC ordinarily drains to the coronary sinus (**Figure 3.13**) but typically connects directly to the left-sided atrium in the presence of atrial isomerism. When

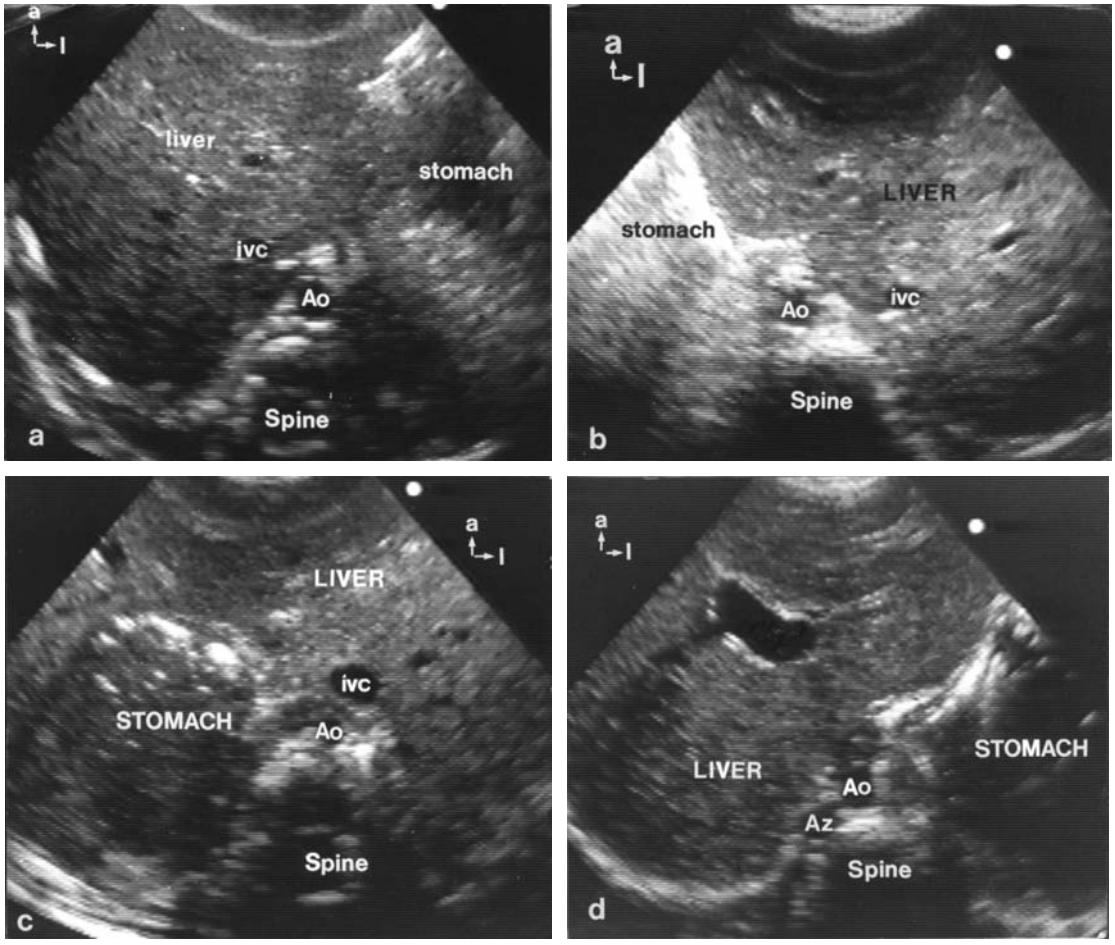


Figure 3.9

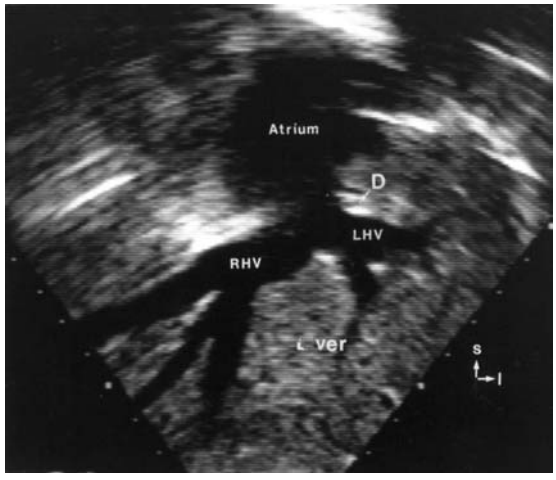


Figure 3.10

there is a right-sided SVC, the presence of an additional left-sided SVC should be suspected when the coronary sinus is large and the innominate vein is absent or small.

Identification of each pulmonary vein draining into the left atrium (**Figure 3.11**) may be difficult, but has been made easier by colour-flow mapping and TOE in selected cases. In totally anomalous pulmonary venous drainage (TAPVD), there is typically a pulmonary venous confluence separate from the left atrium (**Figure 3.14**), with volume overload of the right side of the heart and right-to-left atrial flow. This was among the first abnormalities in which echocardiography provided complete preoperative diagnosis, and improved surgical results were attributed to the avoidance of invasive preoperative investigation. Obstructed infradiaphragmatic TAPVD may be apparent as soon as the transducer is placed on the abdomen, because of the

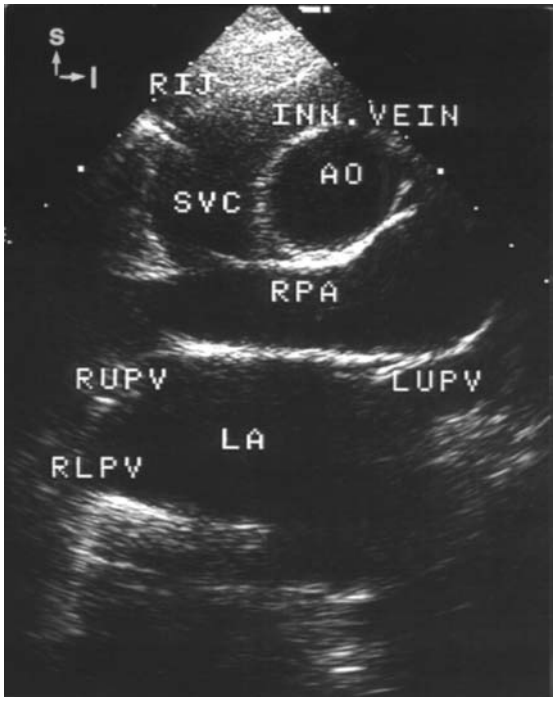


Figure 3.11

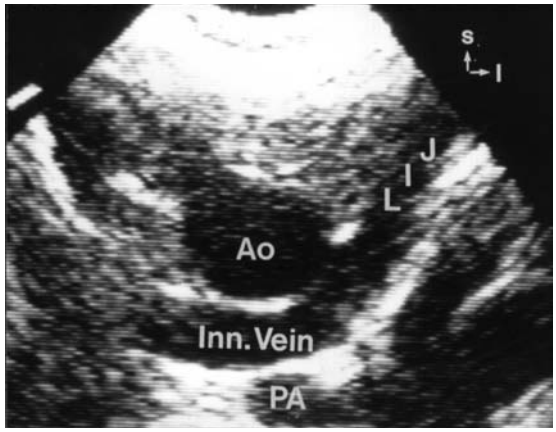


Figure 3.12

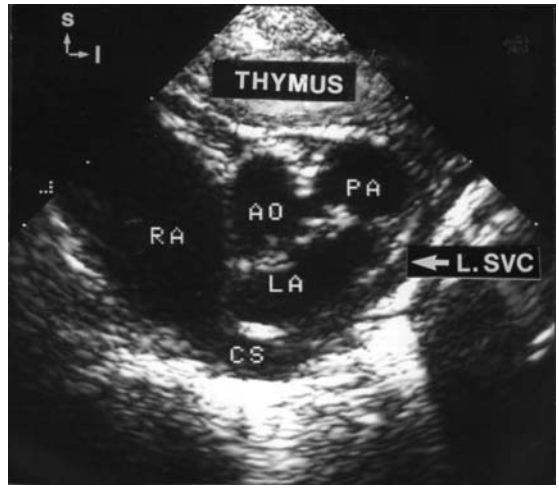


Figure 3.13

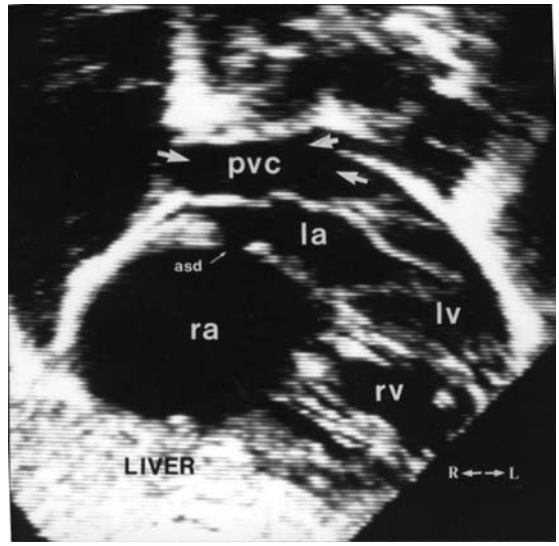


Figure 3.14

gross distension of the portal venous channels within the liver (**Figure 3.15**, pulsed Doppler in descending vein at site x demonstrates low-velocity flow, phasic with respiration rather than the cardiac cycle, indicative of downstream obstruction to anomalous pulmonary venous flow). In contrast, when TAPVD is to the coronary sinus, pulmonary venous flow is unobstructed, and there is marked enlargement of the coronary sinus rather than a separate pulmonary venous confluence (**Figure 3.16**). Patterns of pulmonary venous drainage are especially important in atrial isomerism. In left isomerism, the most common pattern by far is connection of the ipsilateral pulmonary veins to the ipsilateral atrium. In right isomerism, TAPVD is by definition present, and there are a variety of patterns. The assessment of actual or potential obstruction to pulmonary venous drainage is especially important in neonates with right isomerism and

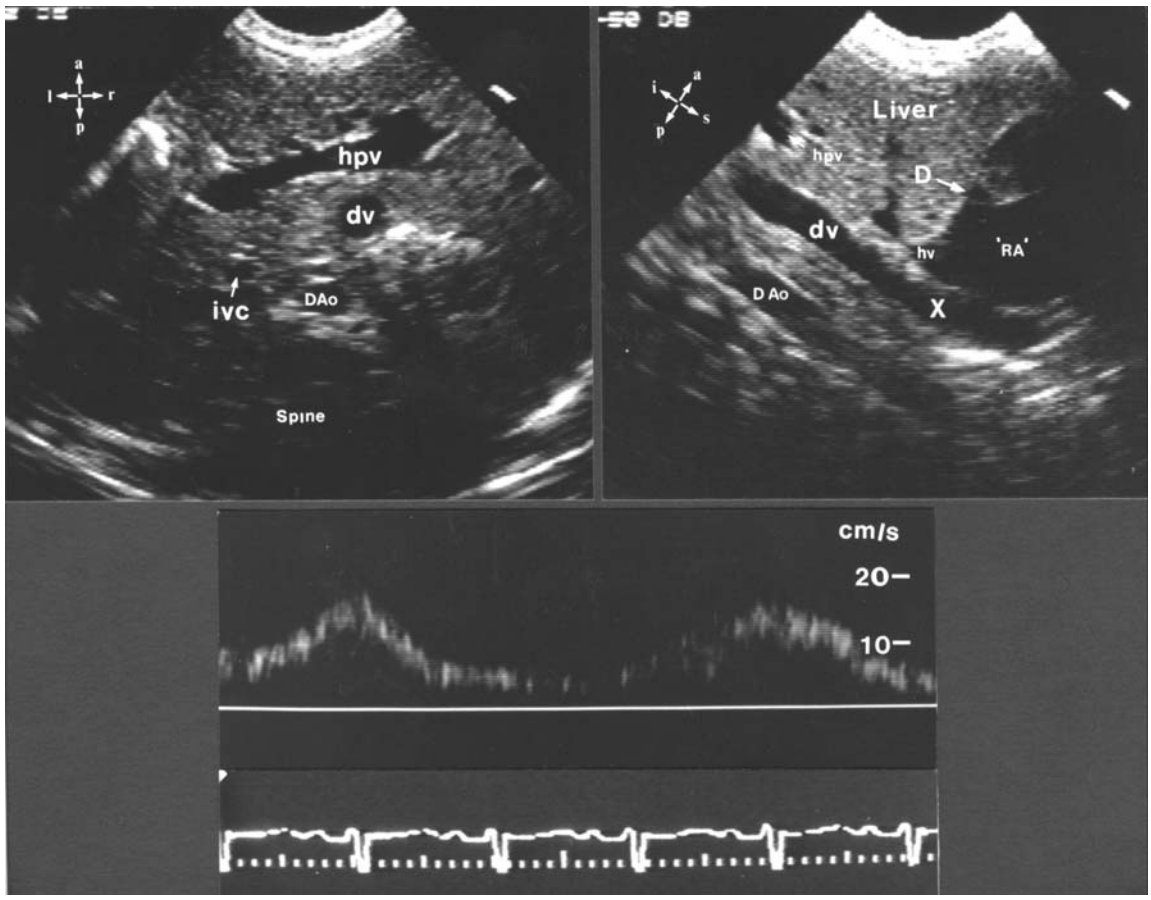


Figure 3.15

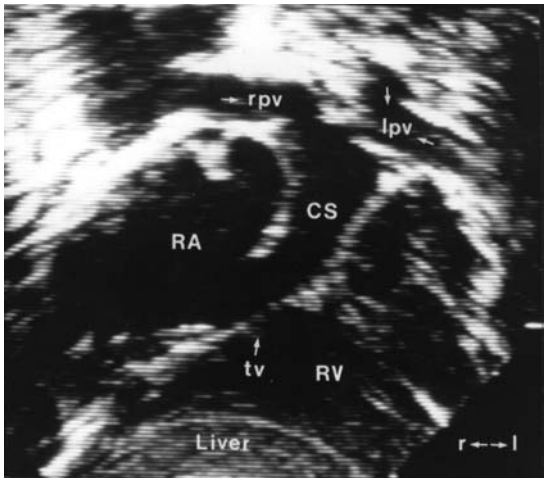


Figure 3.16

severe pulmonary stenosis or pulmonary atresia before construction of a systemic-to-pulmonary artery shunt.

Atrio-ventricular Connection

Once atrial situs has been defined, it is necessary to recognize ventricular structure in order to assess the atrio-ventricular connection. When there is a biventricular atrio-ventricular connection, the morphological right ventricle can be recognized by the coarse apical and septal trabecular pattern and from the fact that the septal origin of the tricuspid valve is positioned slightly more apically than that of the mitral valve (**Figure 3.17**, (a) concordant and (b) discordant atrio-ventricular connection), part of its tension apparatus being attached to the ventricular septum (**Figure 3.18**, discordant atrio-ventricular connection). A univentricular atrio-ventricular connection is present when the atria are connected to only one ventricle. When both atria connect

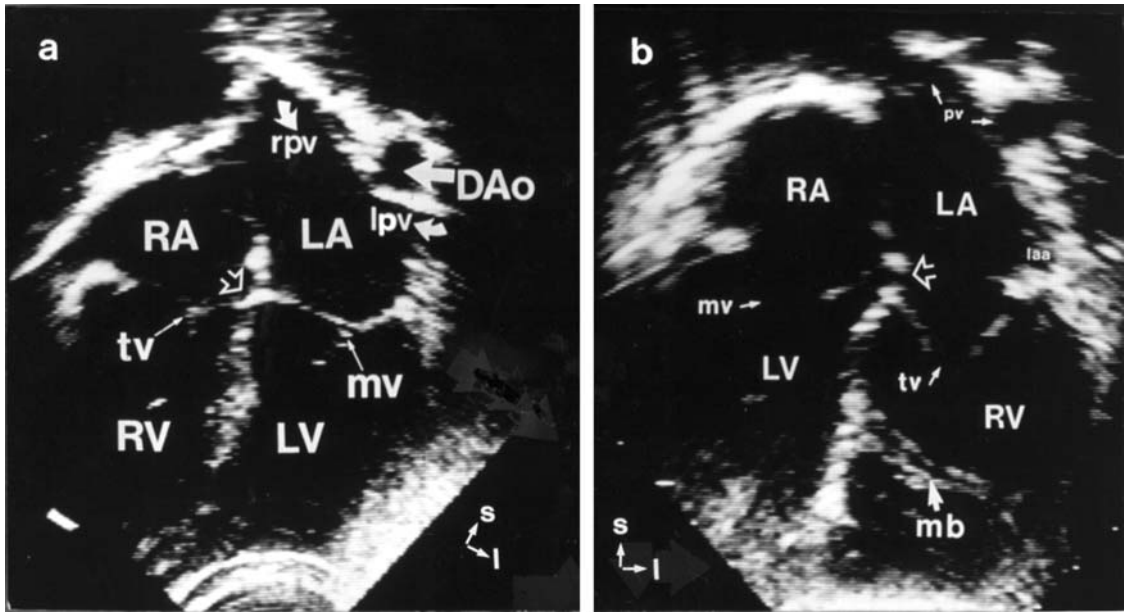


Figure 3.17

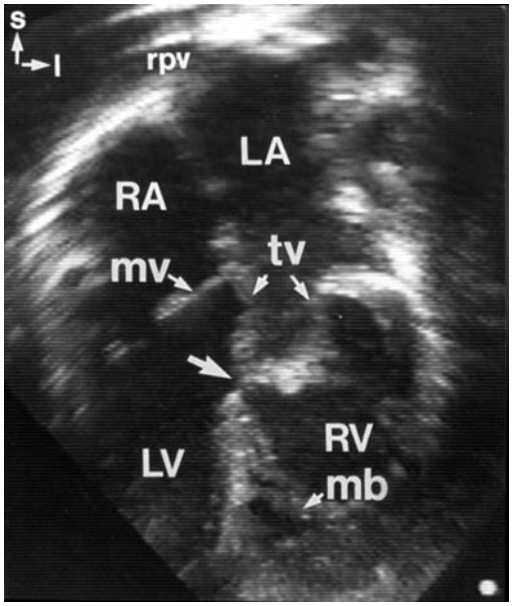


Figure 3.18

to one ventricle, there are usually two atrio-ventricular valves (**Figure 3.19**) although there may be a common atrio-ventricular orifice (**Figure 3.20**). Alternatively, one or another atrio-ventricular connection may be absent, as in tricuspid atresia (**Figure 3.21a**) or mitral atresia (**Figure 3.21b**). In the presence of a univentricular atrio-ventricular connection, nearly always an anterior ventricle is a morphological right ventricle and a posterior ventricle is a morphological left ventricle, regardless of which is the main chamber and which is the rudimentary chamber.

Ventriculo-arterial Connection

The great arteries can be distinguished by early branching of the pulmonary trunk and origin of the head and neck vessels from the aorta. Normally, the right ventricular outflow tract and proximal pulmonary trunk cross over the proximal aorta. In contrast, complete transposition of the great arteries (TGA) (**Figure 3.22a**), congenitally corrected TGA (**Figure 3.22b**) and some forms of double-outlet ventricle are characterized by a parallel course of the proximal great arteries. When an arterial valve overrides the trabecular part of the ventricular septum, the great artery is judged to connect to whichever ventricle it overrides by more than 50%. This should be assessed from the parasternal long-axis plane rather than a subcostal or an apical plane. When the relevant great artery overrides the right ventricle by more than 50% in tetralogy of Fallot (**Figure 3.23a**) or in the presence of a subpulmonary VSD with an anterior aorta (**Figure 3.23b**), the ventriculo-arterial connection

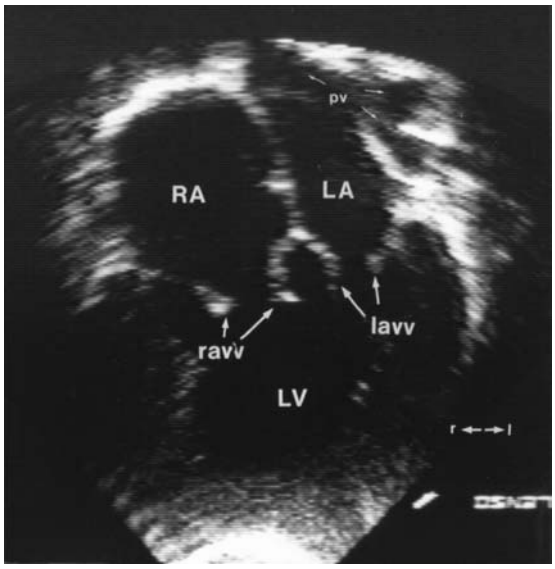
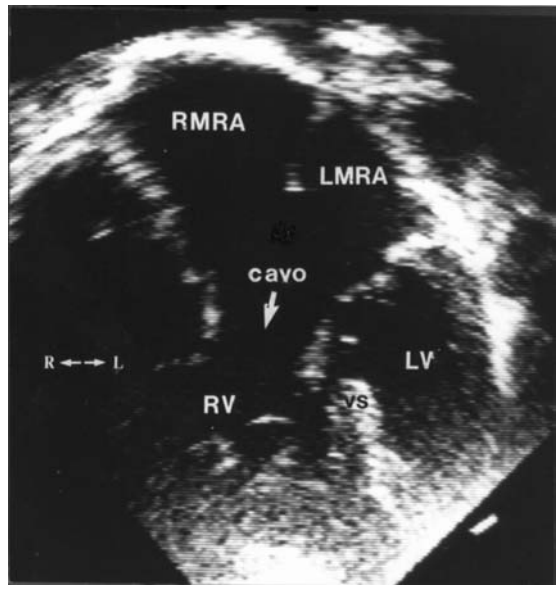


Figure 3.19



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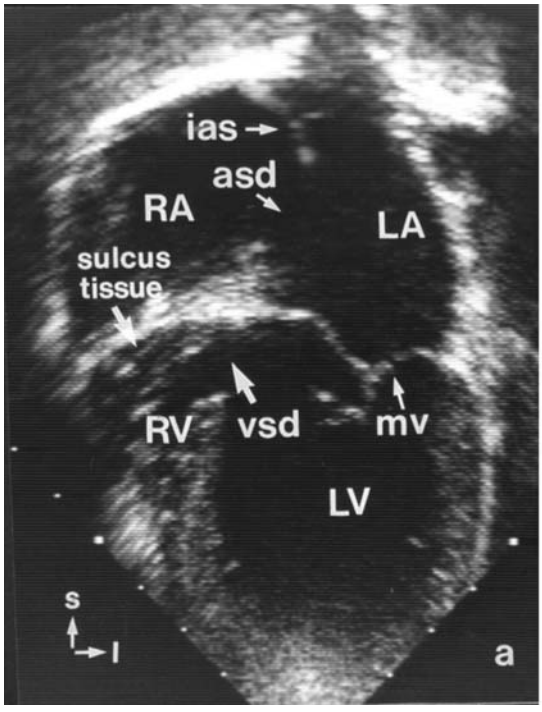

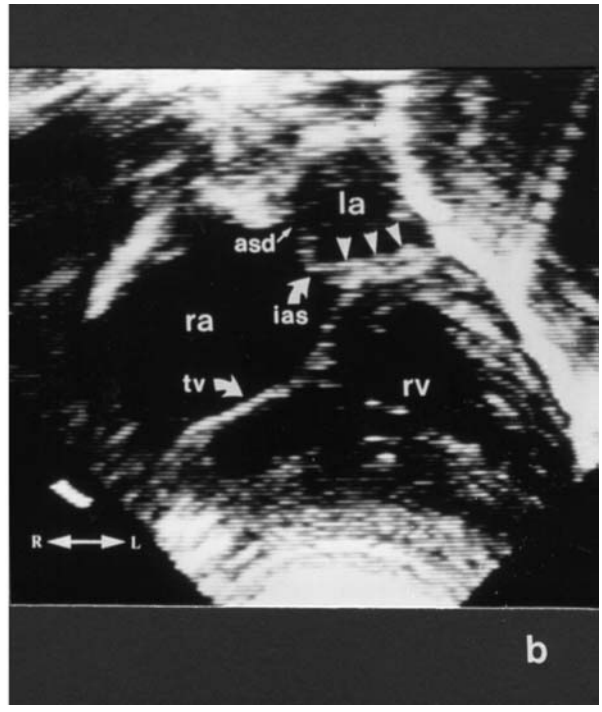

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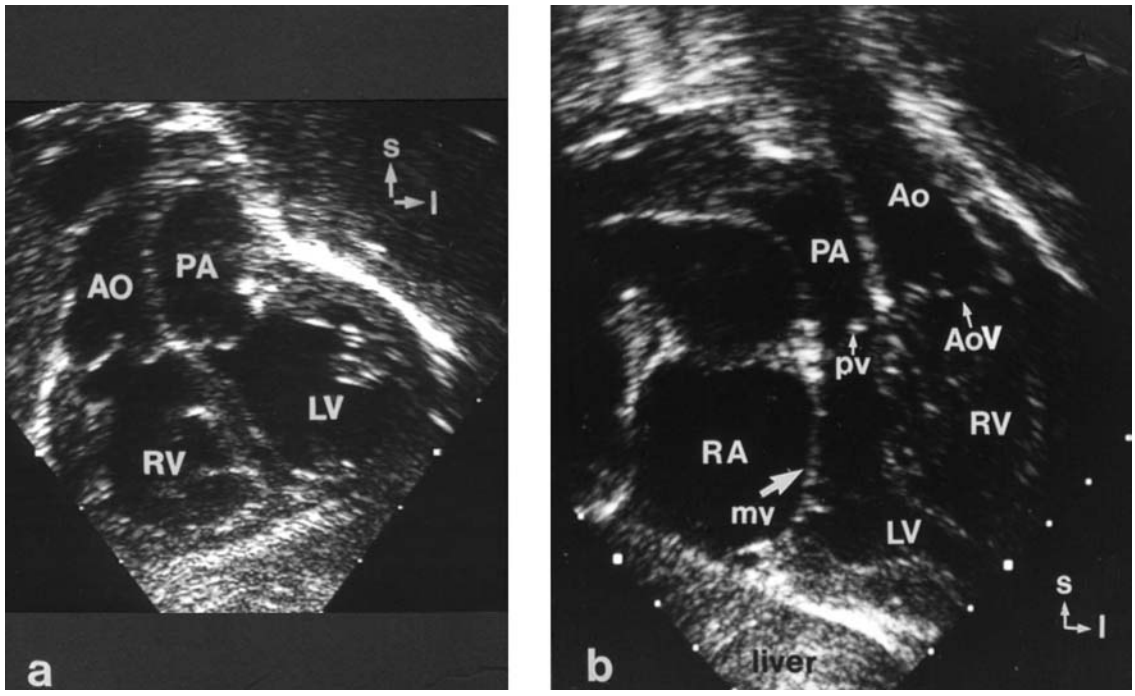


Figure 3.22

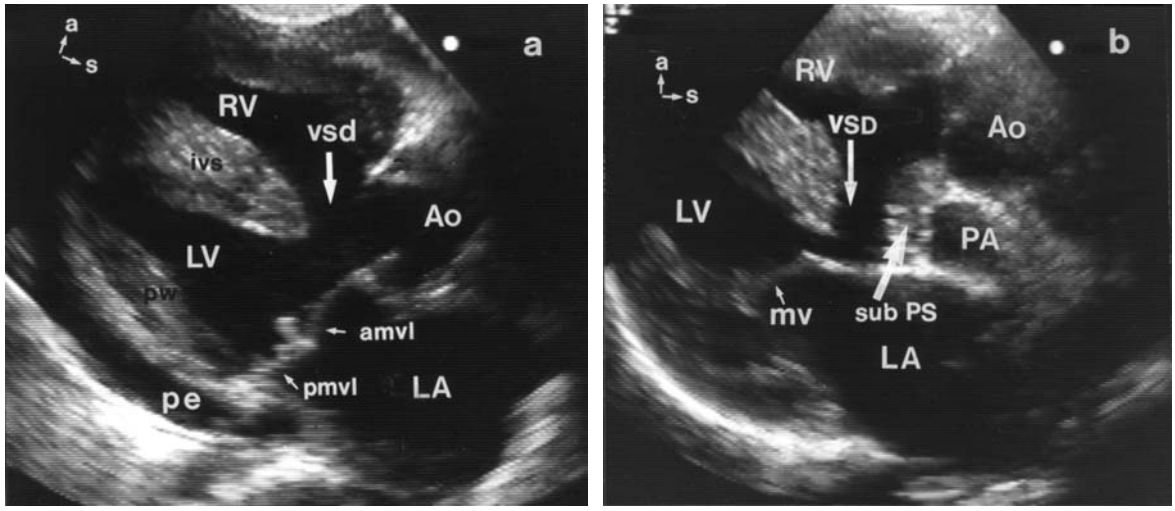


Figure 3.23

is double-outlet right ventricle. A single outlet from the heart is present in truncus arteriosus and in some cases of tetralogy of Fallot with pulmonary atresia.

Atria and Atrial Septum

The two atria are normally of similar sizes. Bowing of the atrial septum into the right (**Figure 3.24a**) or left atrium (**Figure 3.24b**) suggests volume overload, pressure overload or both of the left or right atrium, respectively. A secundum atrial septal defect (**Colour Plate 5**) is the most common type of interatrial communication. This is usually best visualized in children from a subcostal position. Delineation of the margins of the defect in relation to other structures is crucial in the assessment of suitability for percutaneous device closure (**Figure 3.6**). A sinus venosus defect is an interatrial communication at the junction of a vena cava (almost always the SVC) and the heart. This communication is best identified from a subcostal sagittal view in the plane of the venae cavae.

Atrio-ventricular Septum

In normal hearts, the atrio-ventricular septum effectively separates the left ventricle from the right atrium. It comprises a muscular portion posteriorly, which is the result of the offsetting of the septal attachments of the atrio-ventricular valves (**Figure 3.17**, open arrows), and a membranous portion anteriorly, to which the base of the septal leaflet of the tricuspid valve is normally attached. Failure of normal development of these structures occurs in atrio-ventricular septal defects, which are characterized by a common atrio-ventricular junction, usually with an interatrial communication below the true atrial septum and, typically, an interventricular communication immediately below the atrio-ventricular valve leaflets. The atrio-ventricular valve morphology is best assessed from subcostal views in the plane of the common atrio-ventricular junction (**Figure 3.25**). The left side of the zone of apposition between the superior and inferior bridging leaflets is sometimes referred to as a ''cleft'' but should not be confused with a true cleft in the anterior leaflet of a morphological mitral valve.

Ventricles and Ventricular Septum

Ventricular volumes, wall thickness and wall movement demonstrate the haemodynamic consequences of structural abnormality. For example, there is reduced cavity volume and increased wall thickness of the right ventricle in pulmonary atresia with intact ventricular septum (**Figure 3.24b**) and of the left ventricle in hypoplastic left-heart syndrome. Right ventricular volume overload occurs most commonly in left-to-right shunting at atrial level (**Colour Plate 5**), but is increasingly seen during late follow-up after repair of tetralogy of Fallot because of pulmonary incompetence (**Colour Plate 2**), and when

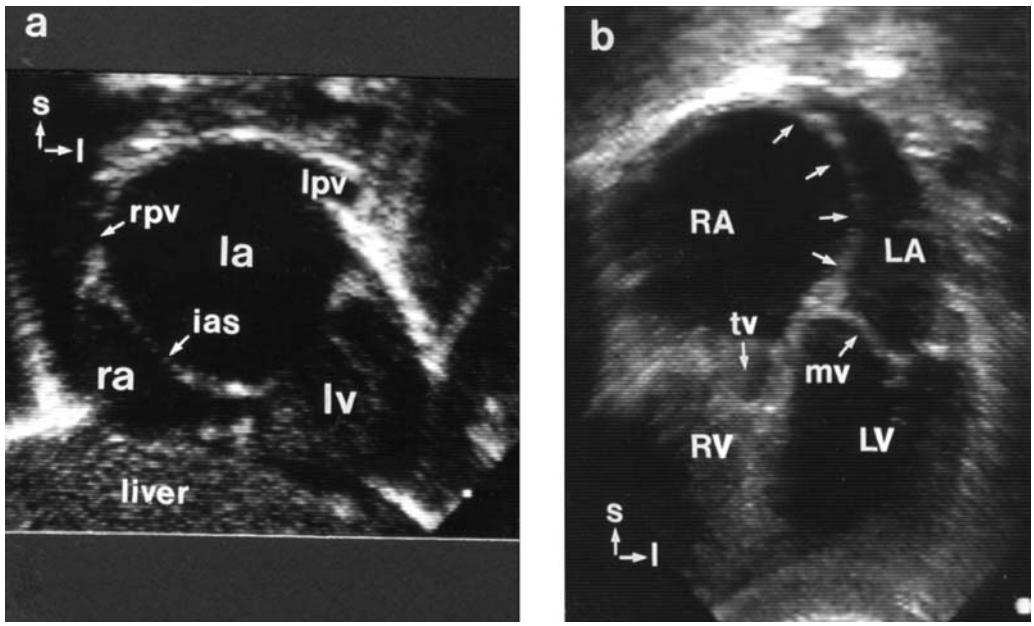


Figure 3.24

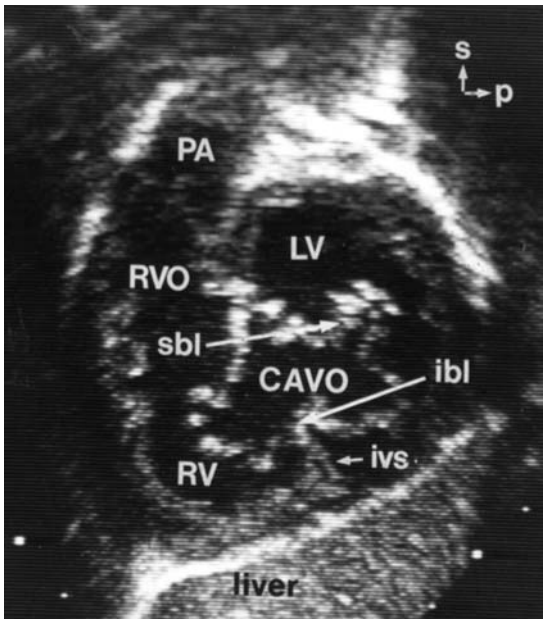


Figure 3.25

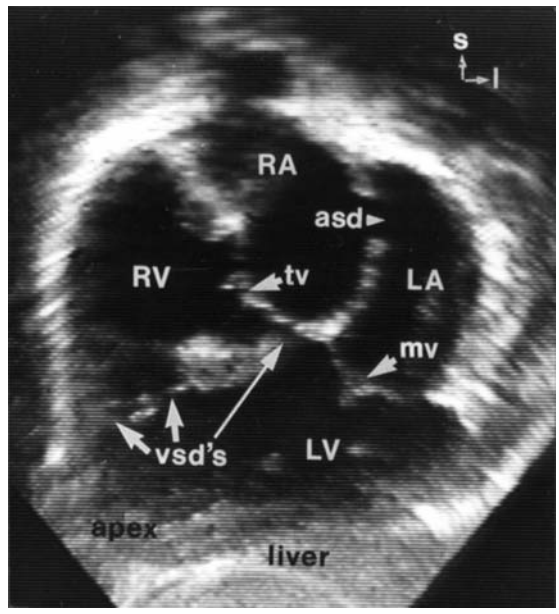


Figure 3.26

this overload is severe, there is paradoxical motion of the ventricular septum into the left ventricle in diastole because of increased diastolic flow into the right ventricle. Left ventricular volume overload occurs typically with left-to-right shunts at ventricular (**Figure 3.8**) or great artery level and with aortic and mitral regurgitation. In normal hearts, the small membranous and much larger muscular portion of the ventricular septum separates the ventricles as the right ventricle folds around the anterior aspect of the left ventricle. Differences in septal geometry, however, occur in many congenital heart defects. For example, in congenitally corrected TGA, the ventricles may be side-by-side, so that the ventricular septum can be in a more or less sagittal plane (**Figure 3.17b**), whereas there may be a superior–inferior ventricular relationship, so that the ventricular septum is relatively horizontal. A univentricular atrio-ventricular connection clearly produces major abnormality of ventricular septal anatomy.

VSDs may be perimembranous, muscular or subarterial in position (Baker *et al*., 1988). A perimembranous VSD is adjacent to the membranous portion of the septum; part of its margin is composed of fibrous tissue. There is fibrous continuity between the tricuspid and aortic valves at the margin of a perimembranous VSD in a heart with normal connections (**Figure 3.8a**). Fibrous tissue related to the tricuspid valve on the right ventricular aspect of the VSD is a mechanism of spontaneous closure of the hole. Tethering of the tricuspid valve may sometimes promote left ventricle to right atrium flow in the presence of a perimembranous VSD.

Defects with entirely muscular rims are typically in the trabecular septum and may be multiple (**Figure 3.26**, there is also dextrocardia with apex to the right). Echocardiography with colour-flow Doppler imaging is a sensitive technique for detection of additional VSDs, even in the presence of a large, non-restrictive defect (Chin *et al*., 1990). A major consideration with large muscular VSDs is surgical accessibility. It is important to identify the precise location of the defect in relation to other structures to aid intraoperative identification, which may be very difficult in the empty heart. A muscular VSD near the cardiac apex will quite often have a single left ventricular entry site, but may have multiple sites of entry into the right ventricle because the flow is divided by right ventricular trabeculations. Apical muscular VSDs may require division of the moderator band or other right ventricular trabeculations to allow visualization through the tricuspid valve, or occasionally an apical right ventriculotomy. Percutaneous device closure of such defects may sometimes be an attractive option.

A doubly-committed subarterial VSD is roofed by the aortic and pulmonary valves, which are in fibrous continuity rather than being separated by the septal aspect of the subpulmonary infundibulum. Many doubly-committed VSDs are also perimembranous. In infancy, the arterial valves are typically at the same level. Large defects are best managed by early surgical closure via

the pulmonary trunk. Offsetting of the arterial valves is the substrate for, or consequence of, prolapse of the right coronary cusp of the aortic valve into the VSD. Prolapse of the right coronary cusp and occasionally the non-coronary cusp into a perimembranous VSD that is not doubly committed can also occur (**Colour Plate 6**). The concern in this situation is that progressive distortion of the aortic valve will lead to increasingly severe aortic regurgitation.

A malalignment VSD occurs when the plane of the outlet septum is deviated with regard to that of the trabecular septum. The antero-cephalad deviation of the outlet septum in tetralogy of Fallot causes the aortic valve to override the crest of the trabecular septum and contributes to right ventricular outflow tract obstruction (**Figure 3.23a**). Lesser degrees of malalignment may be associated with progressive development of right ventricular muscle bundles, sometimes referred to as double-chambered right ventricle when the right ventricular outflow obstruction becomes important. Left ventricular outflow tract obstruction may also occur with this VSD morphology, because of the growth of a fibromuscular subaortic ridge or shelf of tissue on the left ventricular aspect of the crest of the trabecular septum at the margin of the defect (**Figure 3.27**). Posterior deviation of the outlet septum may result in left ventricular outflow tract obstruction and is typically associated with aortic arch obstruction in hearts with normal connections.

In double-outlet right ventricle the size of the VSD and its relationship to the great arteries and the tension apparatus of the atrio-ventricular valves (**Figure 3.28**, tricuspid valve attachment to outlet septum) may be crucial in planning surgical management. When double-inlet left ventricle or tricuspid atresia is

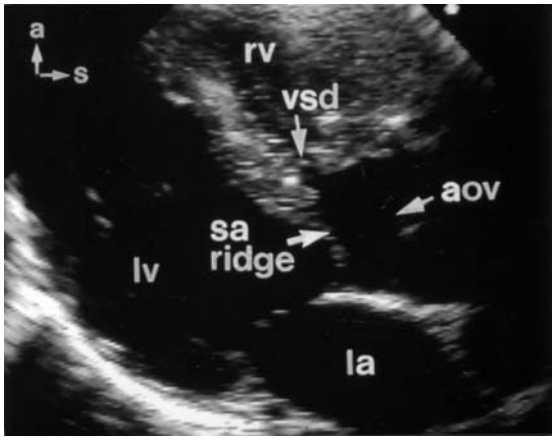


Figure 3.27

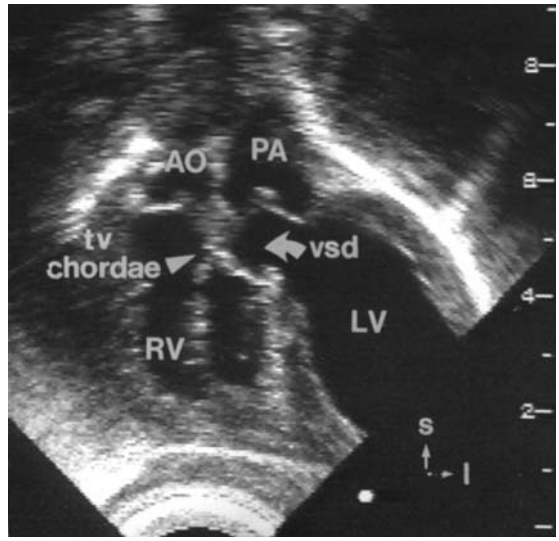


Figure 3.28

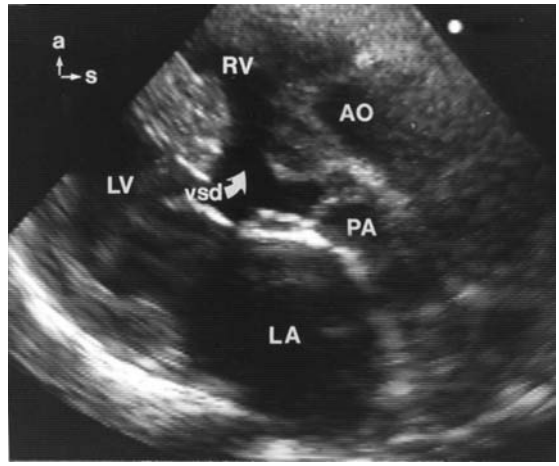


Figure 3.29

associated with ventriculo-arterial discordance, the VSD itself is an actual or potential site of subaortic stenosis (**Figure 3.29**). If the aortic arch is not obstructed, the relative sizes of the VSD and ascending aorta, as measured on echocardiography, may be used as a guide to the appropriateness or not of pulmonary artery banding as initial palliation. Serial echocardiography is ideal for assessing other forms of fixed subaortic stenosis in hearts with normal connections (**Figures 3.27** and **3.30**) because there exists the potential for changes in form as well

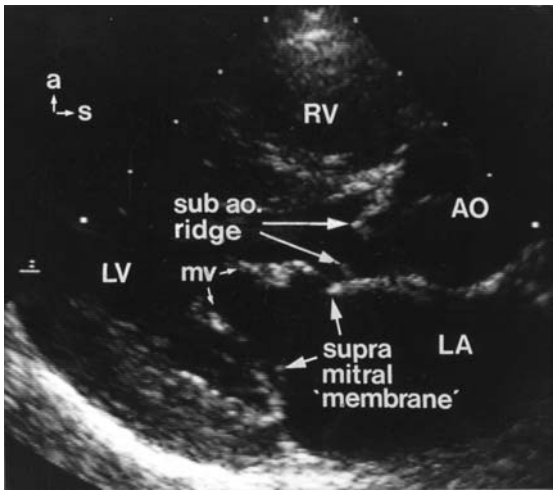


Figure 3.30

as progression in severity of obstruction (Choi and Sullivan, 1991).

Cardiac Valves

Aspects of atrio-ventricular valve morphology have been discussed in relation to the atrio-ventricular connection. Echocardiography is a good technique for characterizing atrio-ventricular valve abnormalities, such as Ebstein's anomaly of the tricuspid valve (**Figure 3.31** left, arrowheads indicate septal leaflet attachment to ventricular septum). Mechanisms of mitral regurgitation, such as mitral valve prolapse, chordal rupture and endocarditis, may be accurately defined. Left ventricular inflow obstruction at valvar (**Colour Plate 7**) or supravalvar level may be identified (**Figure 3.30**) (Tulloh *et al*., 1995). Detailed preoperative assessment of the atrio-ventricular valve tension apparatus is imperative. Atrio-ventricular valve attachments may complicate or contraindicate VSD closure (**Figure 3.31** right). Attachment of tricuspid valve chordae to the infundibular septum may prevent construction of an intraventricular tunnel from the left ventricle to the aorta as part of a Rastelli procedure for hearts with TGA, VSD and left ventricular outflow tract obstruction, or intraventricular repair in double-outlet right ventricle with subpulmonary VSD (**Figure 3.28**, arrowhead). Abnormal mitral valve attachments to the septum may result in left ventricular outflow tract obstruction, especially in hearts with ventriculo-arterial discordance.

Abnormalities of arterial valve morphology are also well defined. Most congenitally stenotic aortic valves are bicuspid. Aortic valve prolapse in the presence of a VSD has been described above, but it may also be the consequence of endocarditis or aortic root abscess. Pulmonary valve morphology determines the likelihood of successful balloon dilatation in pulmonary valve stenosis, and it is important to confirm normal pulmonary valve function if pulmonary autograft replacement of the aortic root is contemplated.

The Great Arteries

Details of aortic, pulmonary arterial and PDA anatomy are especially important when a palliative systemic-to-pulmonary artery shunt, surgery for aortic arch obstruction, relief of vascular compression of the trachea, or repair of other arterial anomalies, such as aortopulmonary window, is required. The relationship of the aortic arch to the trachea determines the side of the aortic arch. This can usually, but not always, be determined from the scanning plane of the long axis of

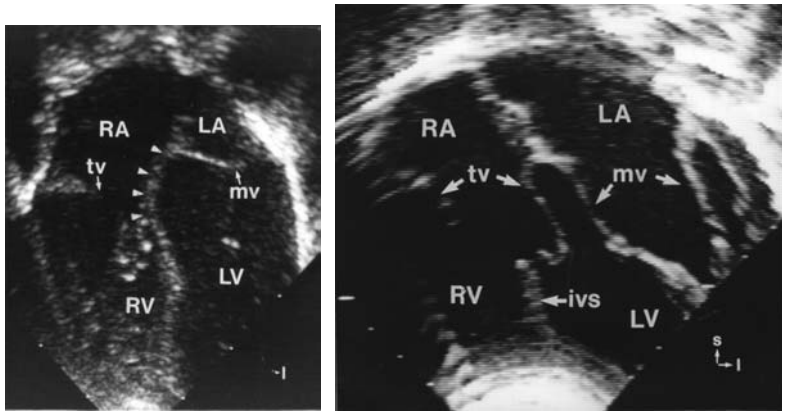


Figure 3.31

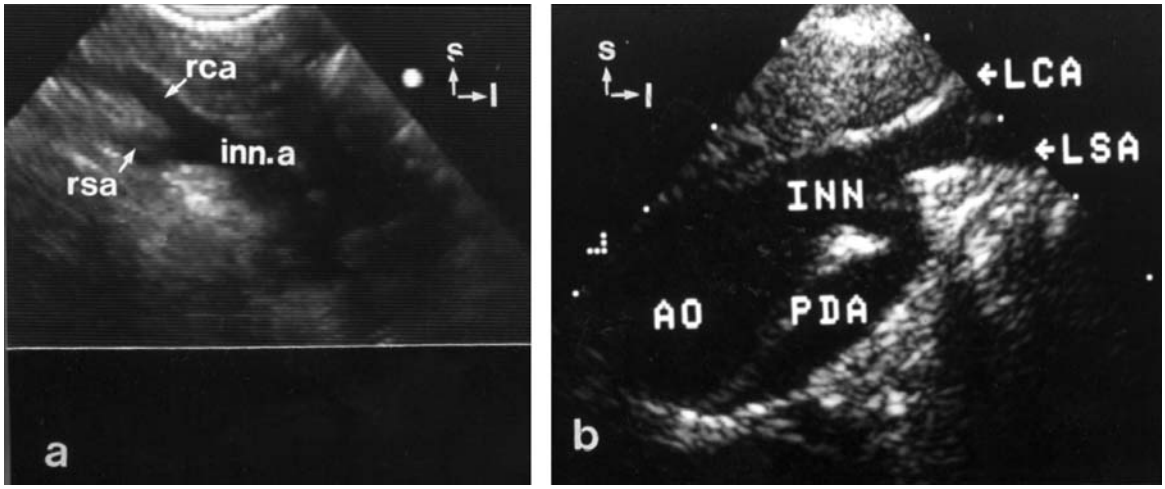


Figure 3.32

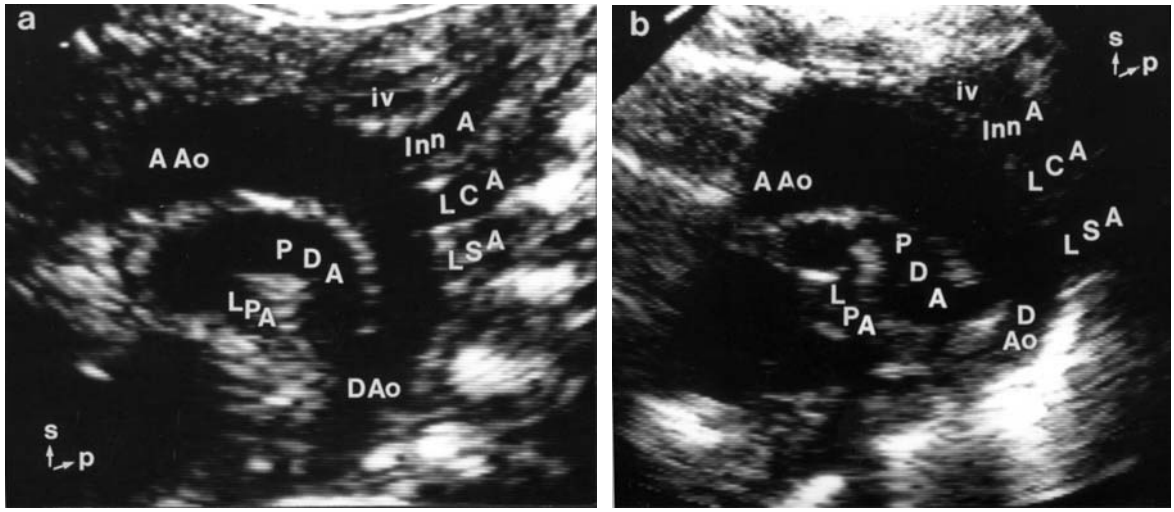


Figure 3.33

the arch. The branching pattern of the aortic arch is a better guide; a left arch has the first branch to the right (**Figure 3.32a**); this branch is usually the innominate (brachiocephalic) artery with a proximal bifurcation. When there is a right aortic arch the first branch is to the left and usually gives rise to the ductus arteriosus (**Figure 3.32b**). If the first branch is smaller than might be expected and does not bifurcate, an anomalous origin of the ipsilateral subclavian artery is likely. This artery may be difficult to image because of its posterior origin, but flow in the anomalous vessel can be identified using colour-flow mapping.

A persistent ductus arteriosus ordinarily arises from the undersurface of a left-sided aortic arch (**Figure 3.33a**). The aortic end of the duct often has a more vertical origin, and the duct has a tortuous course, when there is duct-dependent pulmonary blood flow (**Figure 3.33b**). It is important to ascertain the site of PDA entry before the construction of a palliative shunt. Clamping the contralateral pulmonary artery to construct the shunt does not abolish pulmonary blood flow when this flow is duct-dependent. However, in this situation, it may be preferred to construct a systemic-to-pulmonary shunt via median sternotomy, as this allows ligation of

the PDA at the same procedure, rather than depending on spontaneous closure of the PDA after prostaglandin withdrawal if a lateral thoracotomy approach has been used for the shunt. Colour-flow mapping has facilitated the interpretation of patterns of pulmonary blood flow, including the identification of non-confluent pulmonary arteries and the presence of major aortopulmonary collateral arteries, because continuous-flow patterns in these locations are especially eye-catching in real-time imaging.

When obstruction to aortic flow occurs at the aortic valvar level, there is often poststenotic dilatation of the ascending aorta (**Figure 3.34**), but obstruction to ascending aortic flow may also be supravalvar (**Figure 3.35**). Planning the optimal repair for coarctation of the aorta (**Colour Plate 4**) requires analysis of the arch anatomy on a segmental basis. Neonates with isolated coarctation have variable hypoplasia of the transverse arch. Which patients require extensive repair can be determined on the basis of echocardiographic demonstration of important hypoplasia of the transverse arch. Interruption of the aortic arch, most commonly distal to the left carotid artery, is recognized from the smooth sweep of the ascending aorta into its terminal branch. The characteristic PDA flow pattern in neonates with interruption of the aortic arch or significant coarctation of the aorta demonstrates systolic flow from the pulmonary artery to the descending aorta with diastolic flow reversal, because pulmonary vascular resistance remains lower than systemic vascular resistance, although similar flow patterns are seen in the presence of high pulmonary vascular resistance in the newborn period (**Figure 3.36**).

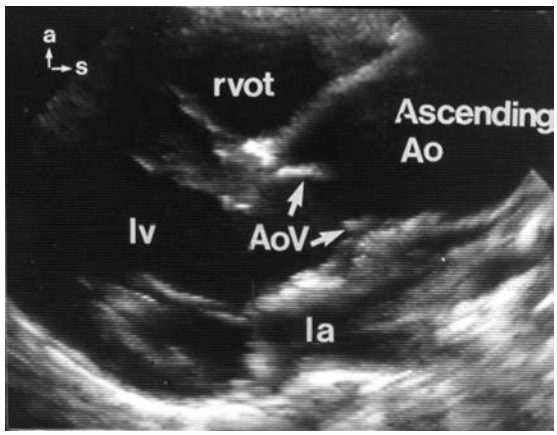


Figure 3.34

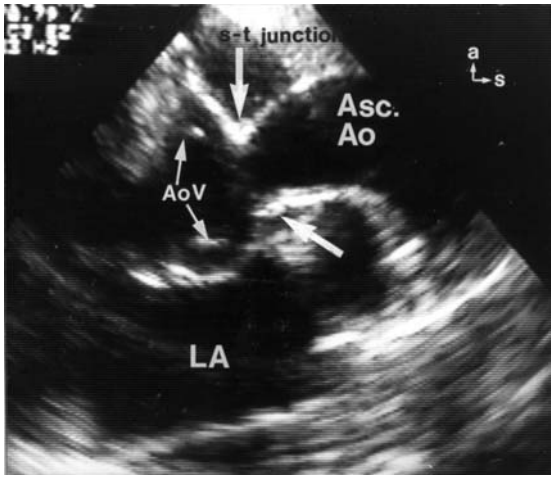


Figure 3.35

Vascular compression of the trachea and oesophagus may be the result of a vascular ring or sling. Barium oesophagogram and echocardiography are complementary investigations in this situation: the barium study can confirm extrinsic compression, and echocardiography can delineate the vascular anatomy in defects such as double aortic arch (**Figure 3.37**). A pulmonary artery sling may be associated with airway abnormality, such as complete tracheal rings and separate origin of the right upper lobe bronchus in about 50% of cases, and may be a difficult echocardiographic diagnosis, especially if there is associated mediastinal displacement. Multislice CT scanning has probably become the definitive imaging to define arterial and airway anatomy in these patients.

Coronary Arteries

Knowledge of coronary artery anatomy is important in the assessment of many congenital heart defects. In complete TGA (Pasquini *et al*., 1987), the coronary arteries normally arise separately from the aortic sinuses that face the pulmonary trunk. The most important variant to identify is an intramural proximal coronary artery course—this is usually first suspected by identifying the proximal left coronary artery coursing between the aorta and pulmonary trunk (**Figure 3.38**); adjustment of the scanning plane should then identify the origin (**Figure 3.39**). Other abnormalities of the coronary origins (**Figure 3.40**, single origin from aortic sinus 1) and proximal course (**Figure 3.41**, right coronary and left circumflex coronary arteries arising from aortic sinus 2), should be recognized preoperatively. A major coronary artery coursing anterior to the pulmonary outflow tract in tetralogy of Fallot may be a relative contraindication to early surgical repair. When anomalous origin of the left coronary artery from the pulmonary artery is suspected,

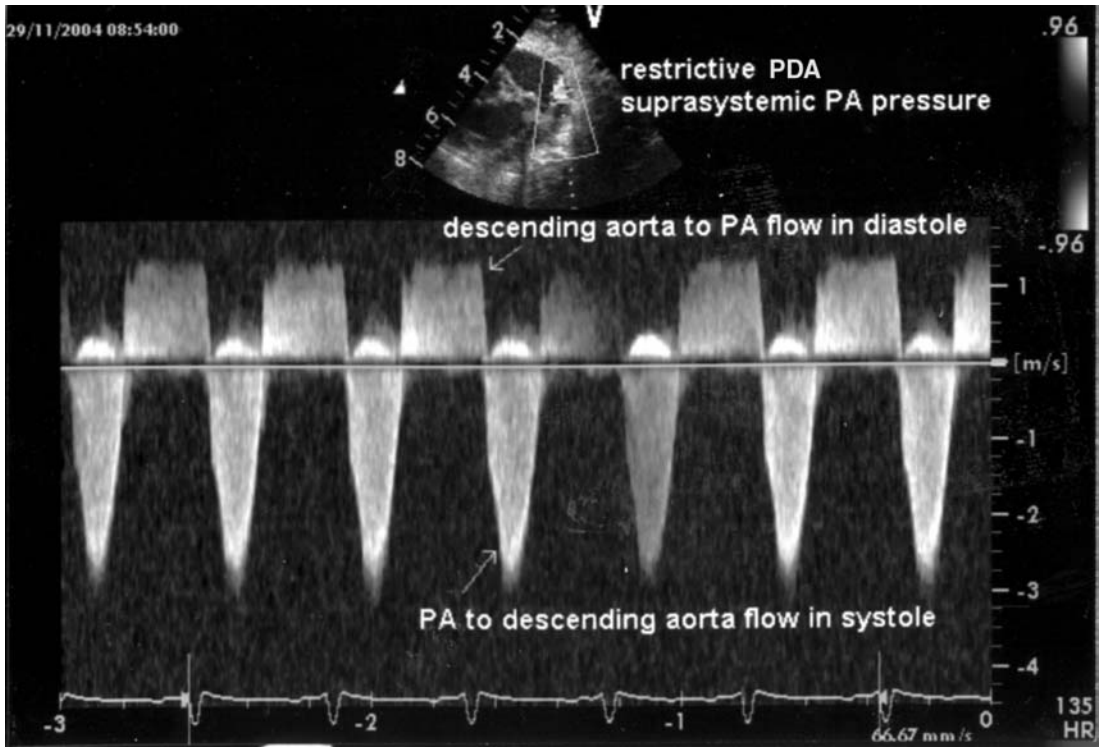


Figure 3.36

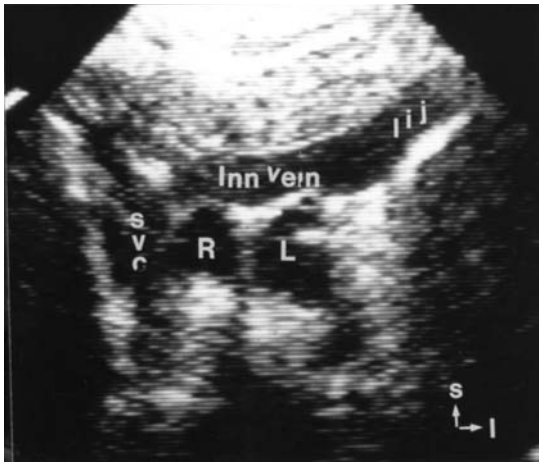


Figure 3.37

the direction of flow in the proximal left coronary artery may confirm or refute the diagnosis, although image acquisition may be technically difficult. A large proximal coronary artery may suggest a communication between the coronary artery and a cardiac chamber. This may be an isolated coronary cameral fistula, or be indicative of a communication between right ventricle and coronary artery in pulmonary atresia with intact ventricular septum. Acquired coronary artery dilation and aneurysm formation is characteristic of Kawasaki disease.

INTRAOPERATIVE ECHOCARDIOGRAPHY

Both epicardial and trans-oesophageal ultrasonic windows may be used for intraoperative echocardiography. When the epicardial approach is used, the ultrasound transducer is enclosed in a sterile plastic sleeve, contact being maintained between the face of the scan head and the plastic by ultrasound jelly. The pericardial cavity may be filled with saline, and the transducer is held gently against the epicardial surface.

Epicardial and trans-oesophageal echocardiography may be of complementary value when undertaken intraoperatively in patients with congenital heart disease. The main advantage of the trans-oesophageal approach is the potential for continuous monitoring of morphology and ventricular function without

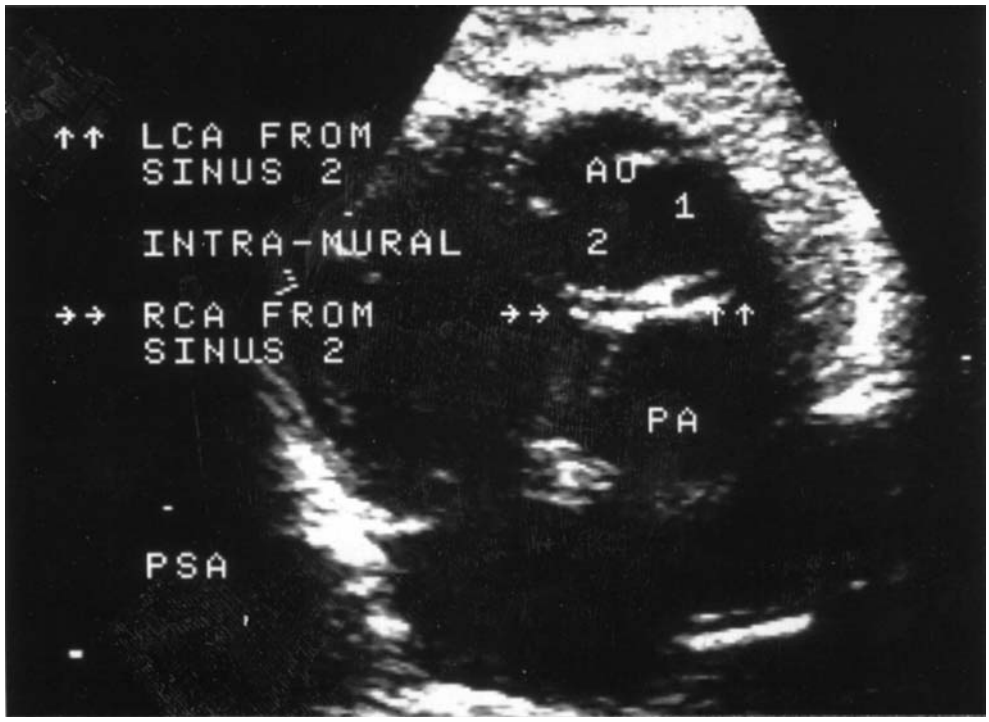


Figure 3.38

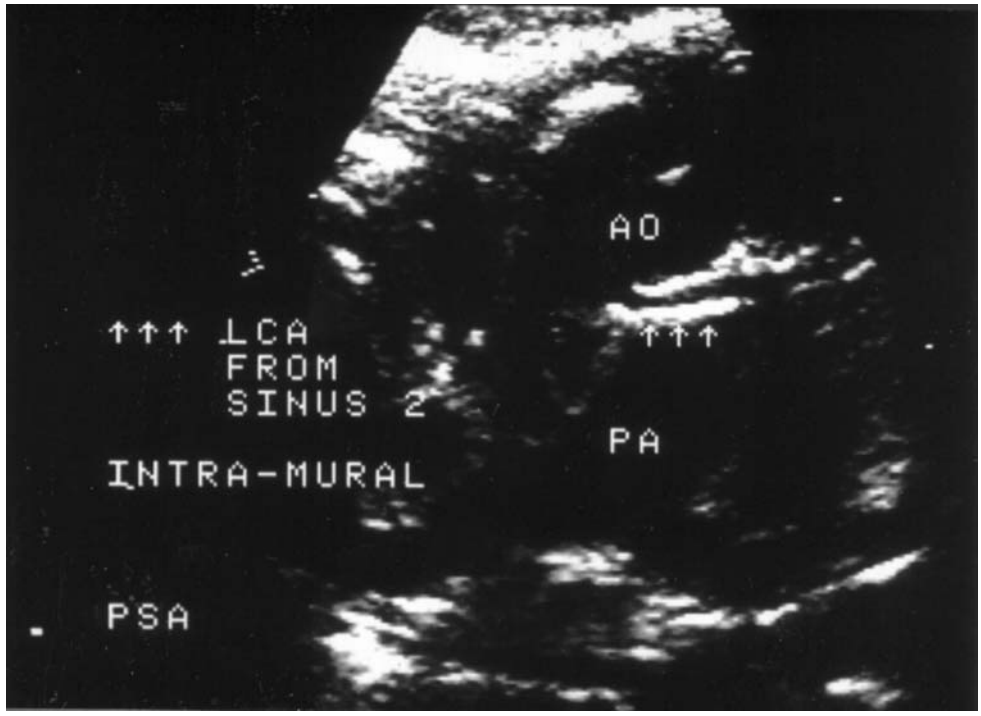


Figure 3.39

Figure 3.40

interrupting or prolonging the surgical procedure. The epicardial approach necessitates interruption of surgery but provides a much higher number of scanning planes, can be undertaken in the smallest patients, and may enable more complete haemodynamic evaluation. Intraoperative echocardiography before cardiopulmonary bypass adds little information to adequate preoperative evaluation, and it did not modify the surgical approach in any of a series of 50 patients who were selected because it was thought that the intraoperative study might yield additional information (Hsu *et al*., 1991). The major advantage of the detection of residual correctable abnormality by intraoperative echocardiography after cardiopulmonary bypass is that an immediate surgical revision can be undertaken. It is important, however, not to overinterpret information obtained. Colour-flow mapping is a very sensitive technique for the detection of disturbed flow. For example, residual VSD flow is identified in about 50% of patients who have undergone VSD closure, and in the absence of multiple defects, the residual flow is rarely of haemodynamic significance and frequently resolves postoperatively.

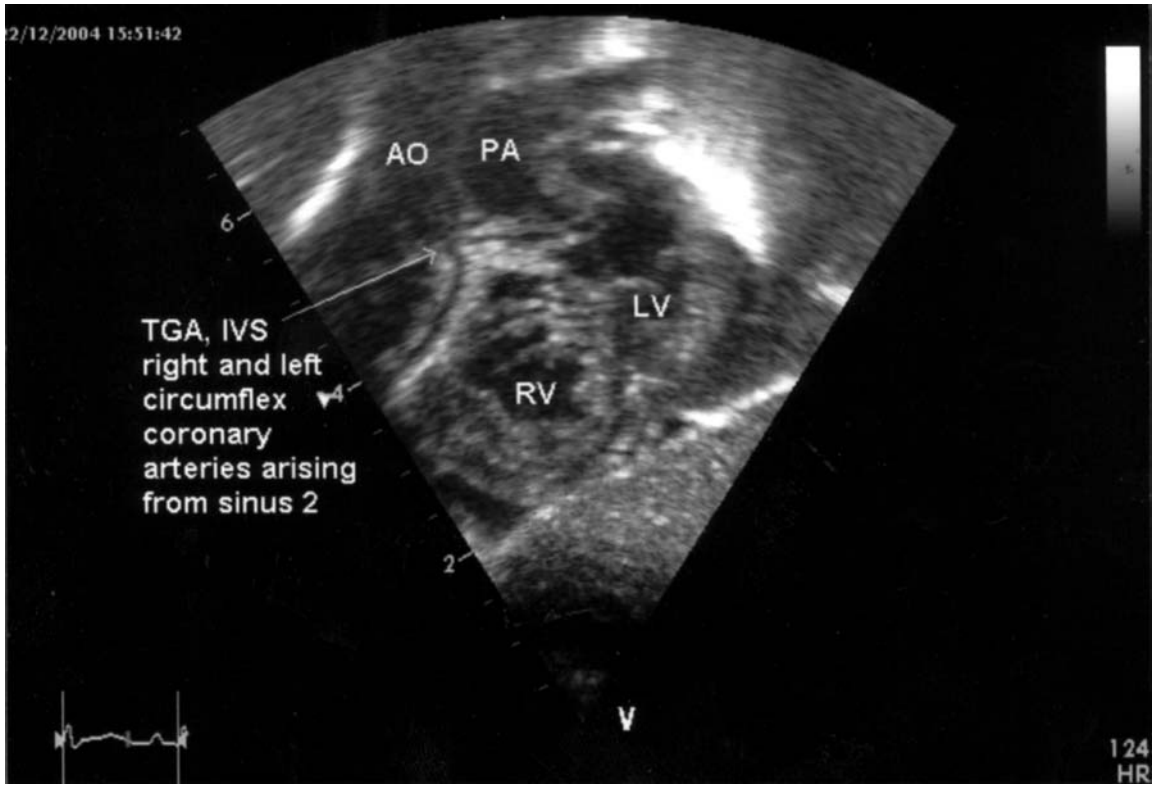


Figure 3.41

Consequently, when there is doubt about the significance of residual shunting detected intraoperatively by colour-flow mapping, the assessment should include the overall haemodynamic state of the patient, perhaps together with measurement of oxygen saturations or semi-quantitative assessment of the residual shunting by contrast echocardiography (Stumper *et al*., 1990a) in selected cases. Intraoperative echocardiography can be valuable in confirming satisfactory relief of systemic outflow tract obstruction, which may be difficult to localize after complex repairs (**Colour Plate 8**). The assessment of the severity of residual obstruction to flow, like the evaluation of residual atrio-ventricular valve regurgitation and residual shunting, must be interpreted in the context of the prevailing haemodynamic conditions (e.g. vasodilatation, impaired ventricular function) immediately after cardiopulmonary bypass. The assessment of left atrio-ventricular valve regurgitation, in particular, is more difficult than is sometimes supposed (Thomas, 2002). Routine intraoperative TOE is practised widely in the USA (Stevenson, 2003), justified on economic grounds (Siwik *et al*., 1999), whereas the usual approach in most European centres has been to apply the technique in selected cases.

POSTOPERATIVE ASSESSMENT

Bedside echocardiography in the intensive care unit provides key information for postoperative management. Surgical dressings, drains, pressure-monitoring catheters, pacing wires, prosthetic material inserted at operation, and hyperinflation of the lungs in ventilated patients may limit access and image quality. Infection is a potential concern, especially in neonates in whom the sternum may be splinted for a while postoperatively. Nevertheless, acceptable examinations can be obtained in most infants and young children, including patients in whom a surgical membrane has been used, but these factors may be indications for trans-oesophageal studies in selected cases. Ventricular function and regional wall motion can be assessed in a semi-quantitative manner as a guide to inotropic drug requirement, but abnormal loading conditions must be taken into account. An apparent reduction in ventricular contraction may be the result of excessively low preload or increased afterload. Ventricular septal motion is typically abnormal early after cardiopulmonary bypass, and this occurrence compromises some of the quantitative approaches to the echocardiographic assessment of ventricular function.

Imaging techniques must be modified in order to take account of surgically altered anatomy. For example, after the bifurcation of the pulmonary artery has been

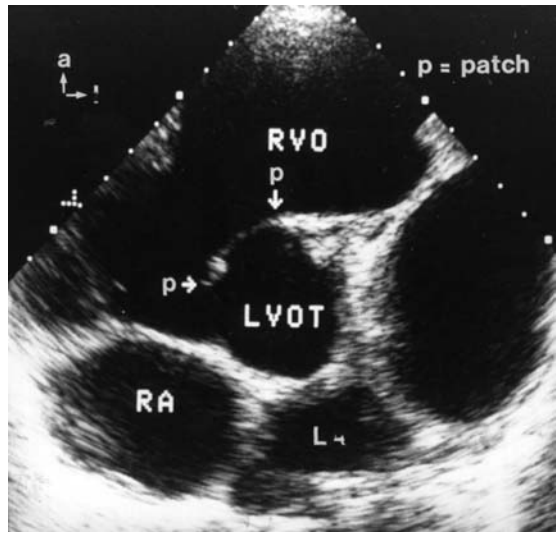


Figure 3.42

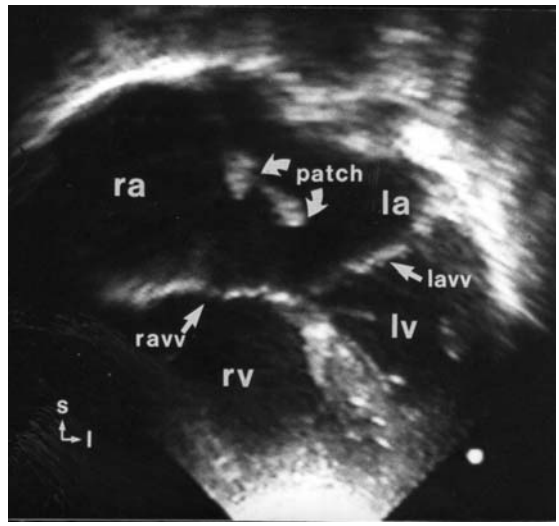
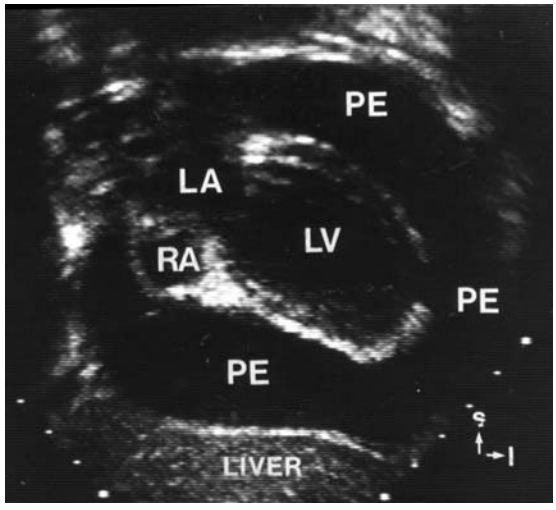


Figure 3.43

positioned anterior to the ascending aorta (Lecompte manoeuvre) as part of an arterial switch procedure, the spatial orientation of the great arteries is different from any naturally occurring relationship of these vessels (**Colour Plate 9**). Knowledge of residual structural lesions is crucial. Sometimes the significance is obvious, such as dehiscence of a ventricular (**Figure 3.42**) or atrial (**Figure 3.43**) patch or prolapse of a left atrio-ventricular valve, with a broad-based, high-intensity Doppler regurgitant signal into a enlarged, tense left atrium.

It is important not to consider the echocardiographic findings in isolation. As with intraoperative studies, colour-flow Doppler images demonstrate residual VSD flow or atrio-ventricular valve regurgitation in the early postoperative period in many patients with a completely satisfactory surgical outcome. Conversely an anatomically small residual VSD may be of more haemodynamic significance after repair of tetralogy of Fallot than after isolated VSD closure. Similarly, demonstration that a residual VSD is restrictive does not mean that it is insignificant, and the sensitivity of colour-flow mapping in confirming patency of a systemic-to-pulmonary artery shunt does not necessarily imply adequacy of volumetric shunt flow.

Extracardiac abnormalities may be identified. Echocardiography has been long recognized as the best technique for recognition of pericardial effusion (**Figure 3.44**). In the absence of a pericardial effusion, cardiac tamponade may be the result of mediastinal thrombus (**Figure 3.45**) or simply the result of tissue swelling after cardiopulmonary bypass. Ultrasound is helpful in the identification of pleural fluid (**Figure 3.46**) and is useful in guiding the optimal site for insertion of a pericardial or pleural drain. Infection of central venous lines may occur if postoperative recovery is prolonged (**Figure 3.47**). Paralysis of a hemi-diaphragm may occur after surgery for congenital heart defects. Confirmation of this complication at a patient's bedside can be reliably obtained by echocardiography (Balaji *et al*., 1991), which is important in infants because most need plication of the affected hemi-diaphragm before they can be weaned successfully from assisted ventilation.



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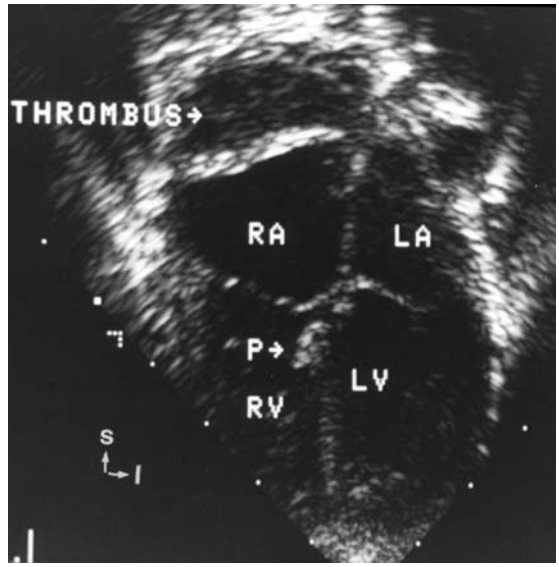

Figure 3.45

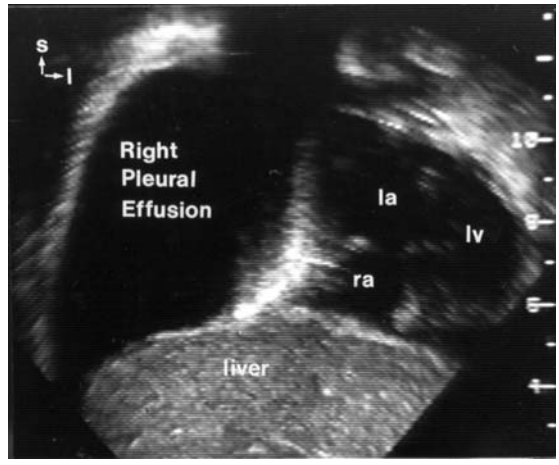

Figure 3.46

Echocardiography also plays a key role in late postoperative assessment. After palliative procedures, unexpected complications may occur, such as seroma formation after insertion of a modified Blalock–Taussig shunt. The usual aim at follow-up after palliative surgery is to determine the optimal timing of the next intervention. This might be influenced, for example, by evidence of a restrictive atrial septal defect through which there is obligatory flow (**Colour Plate 10**), impending encroachment of a pulmonary artery band onto right pulmonary artery, or an increasing Doppler gradient

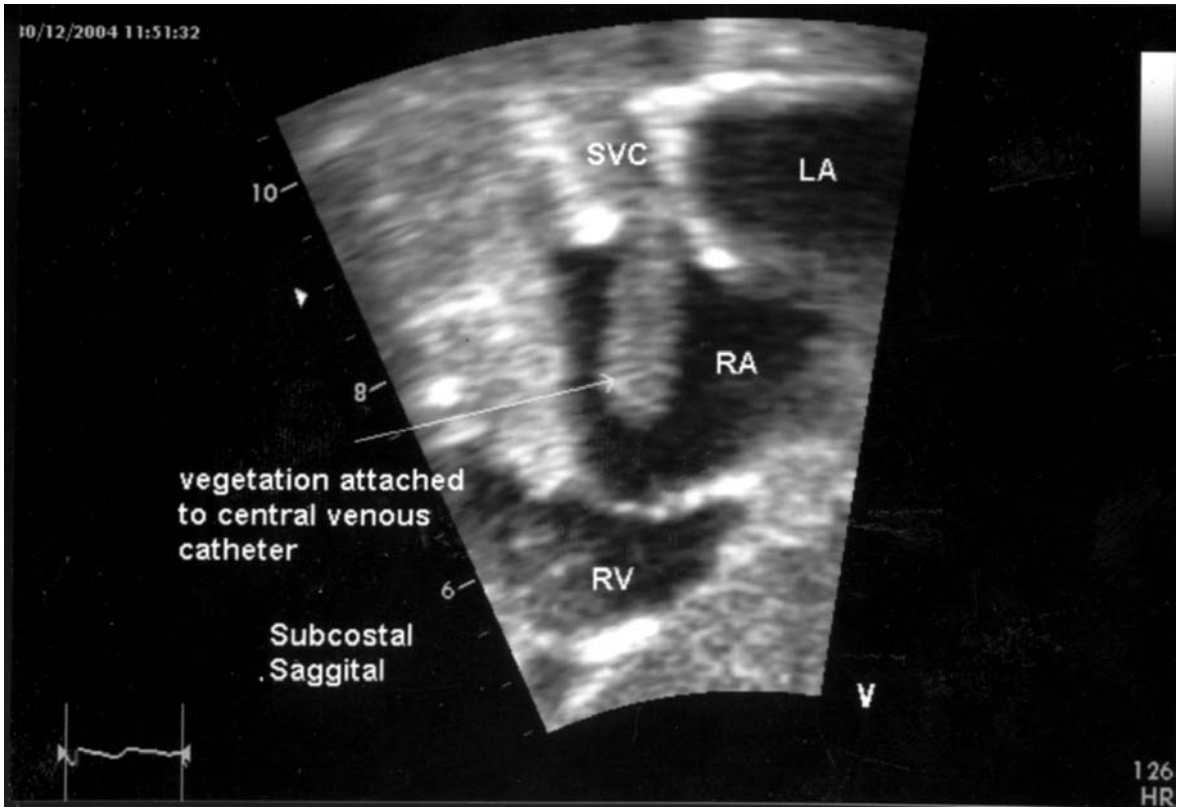

Figure 3.47

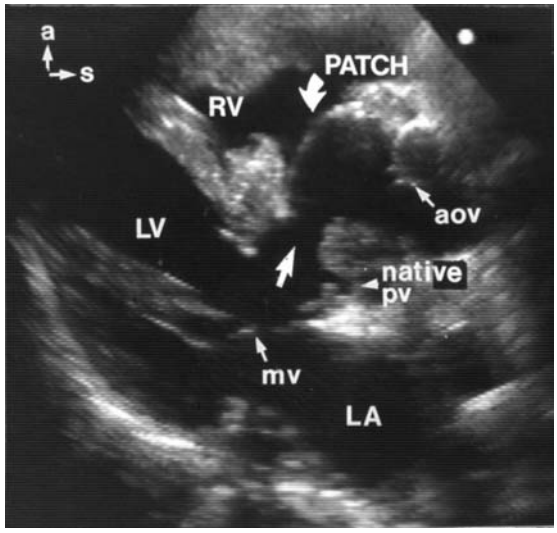

Figure 3.48

after aortic valvotomy. Echocardiographic surveillance is equally important after apparently definitive procedures. Progressive obstruction of an extracardiac valved conduit is almost inevitable and should be monitored. Other anticipated late complications might include recurrent aortic arch obstruction, pulmonary regurgitation after repair of tetralogy of Fallot (**Colour Plate 2**), venous pathway narrowing after atrial repairs for TGA, obstruction of pulmonary venous flow, either within the pulmonary veins or at the anastomosis between pulmonary vein confluence and left atrium after repair of TAPVD, recurrent left ventricular outflow tract obstruction after surgical relief of fixed subaortic stenosis (**Figure 3.30**) or at the site of the previous VSD after a Rastelli procedure (**Figure 3.48**), atrioventricular valve regurgitation after repair of atrioventricular septal defect, impaired ventricular function after many types of operation, or endocarditis. Such complications emphasize the need for sustained follow-up. Furthermore, an important role for echocardiography is as an aid in the audit of late functional results. The

continuing aim must be to acquire data either to reinforce a current surgical management strategy or to demonstrate the need for a different approach.

CONCLUSION

Two-dimensional and Doppler echocardiography enable comprehensive evaluation of structure and haemodynamics in congenital heart disease. Diagnostic, as opposed to therapeutic, cardiac catheterization and angiography are now rarely necessary. Continuing indications for invasive investigation might include angiographic assessment of multifocal pulmonary blood supply and detailed haemodynamic measurement when pertinent, such as when Fontan-type surgery or transplantation, is being considered. The trans-oesophageal approach has provided a semi-invasive technique extending the value of ultrasonography in the assessment of congenital heart disease, especially in the perioperative situation. Three-dimensional echocardiography (**Figure 3.3**) may have an increasing role as advances in computing make image reconstruction and manipulation easier, in a manner analogous to MR or CT image management. Intravascular and intracardiac ultrasound may have a role in selected patients.

Echocardiography remains the mainstay of clinical imaging and haemodynamic assessment in congenital heart defects, and will do so for the easily foreseeable future, but there has been increasing importance in using this easily accessible technique to select appropriate patients for more complex and expensive imaging, in particular MR imaging or multislice CT scanning. These techniques each have their specific strengths, but share the advantage over echocardiography of defining the heart and great vessels in relation to other thoracic structures. The aim should be to seek and interpret information required in a specific patient in a complementary fashion.

APPENDIX

Abbreviations in Figures Specific to This Chapter

A Ao, Asc Ao	ascending aorta
amvl	anterior mitral valve leaflet
ao	aorta
aoV	aortic valve
ASD	atrial septal defect
AVSD	atrio-ventricular septal defect
Az	azygos
CAVO	common atrio-ventricular valve orifice
CIRC	circumflex artery
coarct.	coarctation
conf.	confluence
CS	coronary sinus
cTGA	congenitally corrected transposition
D	diaphragm
DAo	descending aorta
dv	descending vein
ECG	electrocardiogram
Eust, EUS, eus	Eustachian
hv	hepatic vein
hpv	hepatic portal vein
ias	interatrial septum
ibl	inferior bridging leaflet
Inn., INN.	innominate
iv	innominate vein
IVC	inferior vena cava
ivs	interventricular septum
LA	left atrium
laa	left atrial appendage
LAD	left anterior descending
lavv	left atrio-ventricular valve
LAVVR	lavv regurgitation
LCA	left coronary artery
LIJ	left internal jugular
LMRA	left-sided morphological right atrium
LPA	left pulmonary artery
lpv	left pulmonary vein
LSA	left subclavian artery
L.SVC	left-sided superior vena cava
lu	left upper
LUPV	left upper pulmonary vein
LV	left ventricle
mb	moderator band
m s^{-1}	metres per second
MV	mitral valve
ncc	non-coronary cusp
p	patch
PA	pulmonary artery
PAB	pulmonary artery banding
PCG	phonocardiogram
PDA	persistent ductus arteriosus
pe	pericardial effusion
pmvl	posterior mitral valve leaflet
pv	pulmonary vein
pw	left ventricular posterior wall
ravv	right atrio-ventricular valve
RCA	right coronary artery
rcc	right coronary cusp
RIJ	right internal jugular
rl	right lower
RLPV	right lower pulmonary vein
RMRA	right-sided morphological right atrium

RPA	right pulmonary artery
rpv	right pulmonary vein
RSA	right subclavian artery
ru	right upper
RUPV	right upper pulmonary vein
RV	right ventricle
RVOT	right ventricular outflow tract
sa	subaortic
sbl	superior bridging leaflet
s-t	sinotubular
sub PS	subpulmonary stenosis
SVC	superior vena cava
tv	tricuspid valve
vs	ventricular septum
VSD	ventricular septal defect

Acknowledgements

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4

Foetal Echocardiography

L. D. Allan

The first detailed descriptions of the appearance of the normal human foetal heart as seen by cross-sectional ultrasound were published in 1980 (Lange *et al*., 1980; Kleinman *et al*., 1980; Allan *et al*., 1980). Since then, a complete spectrum of congenital heart disease (CHD) has been recognized in prenatal life (Allan *et al*., 1984, 1994). With experience and modern equipment, anomalies can now be reliably detected from as early as 12 weeks gestation, up to term (Huggon *et al*., 2002). The most easily obtained view of the foetal heart is the four-chamber view, and identifying the normality of this one section rules out many major defects. Nowadays, it is increasingly standard practice during the routine obstetric ultrasound examination to analyse not only the four-chamber view but also views of the great arteries. As the majority of pregnancies in the UK are now offered a routine anomaly scan, there is therefore the opportunity of screening all pregnancies for major forms of CHD (Fermont *et al*., 1986; Allan *et al*., 1986).

ORGANIZATION OF FOETAL ECHOCARDIOGRAPHY

Certain groups of pregnancies are at increased risk for CHD and these patients should be referred for a focused echocardiogram:

1. A family history of CHD is a predisposing factor. If one previous child has had CHD, the recurrence risk is about 1 in 50. When two children have been affected, the risk increases to about 1 in 10. When a parent is affected, the risk to the next generation is probably higher if the affected parent is the mother, and that risk is of the order of about 1 in 10.
2. Maternal diabetes is associated with a statistical risk of cardiac malformation of about 1 in 50. Good diabetes control in early pregnancy probably diminishes this risk.
3. Exposure to teratogens in early pregnancy, such as lithium or anticonvulsant drugs, is reported to be associated with a 1 in 50 risk of heart malformation.
4. The detection of an extracardiac foetal anomaly of almost any kind, on ultrasonography, should lead to a complete examination of the foetal heart because many types of abnormalities, e.g. exomphalos or a tracheo-oesophageal fistula, can be associated with CHD (Copel *et al*., 1986). Abnormalities in more than one system in the foetus should arouse the suspicion of a chromosomal defect underlying associated abnormalities. It may be appropriate to offer karyotyping by chorion villus sampling or amniocentesis, in order to give the most accurate prognosis to the mother.
5. Some foetal arrhythmias are associated with structural CHD, especially complete heart block, which produces a sustained bradycardia of less than 100 beats/minute. The normal foetal heart rate is around 140 beats/minute at 20 weeks gestation.
6. Non-immune foetal hydrops can be caused by CHD (Kleinman *et al*., 1982), and a foetal echocardiogram should be an essential part of the work-up of the affected foetus. Foetal hydrops may have a cardiac cause in up to 25% of cases.
7. An important high-risk group are those ‘‘normal’’ pregnancies where the ultrasonographer notices an abnormality of the heart during a routine scan. Nearly all pregnancies undergo scanning in the UK at the present time. The timing and intensity of the routine scan vary, but where a thorough ‘‘anomaly’’ scan takes place around 20 weeks gestation, severe cardiac anomalies can potentially be detected in up to 3/1000 studies. The majority of foetal cardiac anomalies seen in foetal cardiac referral units are suspected during the routine obstetric examination.
8. A more recently recognized high-risk group are those foetuses where the nuchal translucency measurement at 12 weeks gestation has been found to be increased (Hyett *et al*., 1995). This measurement

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of the skin-fold thickness at the back of the neck is becoming increasingly adopted as a screening tool for chromosomal defects (Snijders *et al*., 1998), particularly Down's syndrome, but an increased measurement is also associated with CHD. The incidence of CHD in this group will vary with the degree of increase in the measurement, but those with a measurement above the 95th centile are appropriate for referral for detailed foetal echocardiography. Up to 25% of major structural heart disease is associated with increased nuchal translucency and all types of CHD can present in this way.

In the hands of the specialist foetal echocardiographer, it is possible to visualize the cardiac connections in almost every patient. The ideal gestational age for specialist evaluation is between 14 and 20 weeks, depending on the experience of the examiner. Some minor defects, such as small ventricular septal defects and mild valve stenosis, are overlooked in the foetus and some lesions, such as cardiac tumours, develop in the latter half of pregnancy and therefore may not be detected if only one early scan is performed. Secundum atrial septal defects and, of course, persistence of the ductus arteriosus are not possible to predict prenatally. With the exception of coarctation of the aorta, which is a notoriously difficult diagnosis to make or exclude prenatally, false positive diagnosis of CHD by the specialist is rare.

NORMAL FOETAL CARDIAC ANATOMY

The Four-chamber View

The four-chamber view is achieved in a horizontal section of the foetal thorax just above the diaphragm (the following abbreviations are used in the illustrations: RV, right ventricle; LV, left ventricle; LA, left atrium; RA, right atrium; DAo, descending aorta; AAo ascending aorta; PA, pulmonary artery; IVC, inferior vena cava). In **Figure 4.1**, the four-chamber view is seen with the foetus in an ideal position with the apex of the heart close to the transducer. Whatever the foetal position, the same method of orientation is always used to identify the cardiac chambers. The first step is to locate the spine. Opposite the spine is the anterior chest wall or sternum, and below the sternum is the right ventricle. The descending aorta is seen as a circle in the mediastinum, lying anterior to the spine. Related to the aorta anteriorly is the left atrium. The remaining intracardiac chambers, the right atrium and the left ventricle, can then be identified. This view should be obtainable in every patient from 12 weeks gestation onward with generally available real-time equipment. The appearance of the four-chamber view varies according to the position of the foetus relative to the ultrasound beam, but no matter

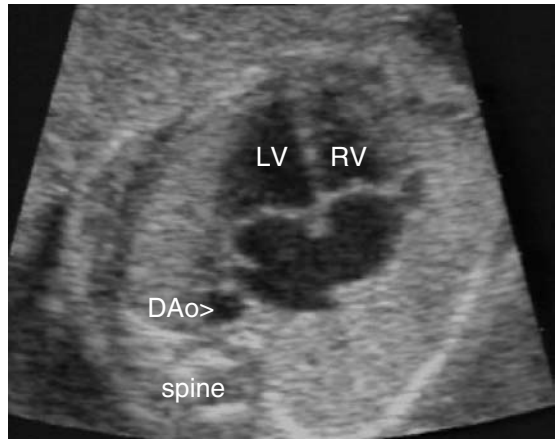


Figure 4.1

how the foetus is lying, the following important points can always be seen in the normal heart:

1. The heart occupies about one-third of the thorax.
2. The interventricular septum lies at an angle of about 45° to the midline.
3. There are two atria of approximately equal size.
4. There are two ventricles of approximately equal size and thickness. Both show equal contraction in the moving image.
5. The atrial and ventricular septa meet the two atrio-ventricular valves at the crux of the heart in an offset cross, with the tricuspid valve inserted a little lower into the septum than the mitral valve.
6. The ventricular septum appears intact whereas the foramen ovale defect occupies the middle third of the atrial septum.
7. Two opening atrio-ventricular valves are seen in the moving image.
8. There is forward flow across both AV valves and no regurgitation on colour-flow mapping.
9. The pulmonary veins enter the back of the left atrium on cross-sectional imaging and flow from at least one vein into the left atrium is confirmed on colour-flow mapping.

Additional features that can often be noted include the presence of the foramen ovale flap in the left atrium and the moderator band in the right ventricular apex.

Imaging the Great Arteries

Because neither the thoracic cage nor the lung fields obstruct the ultrasound beam prenatally, the great arteries can be imaged in a variety of projections (Allan, 1986).

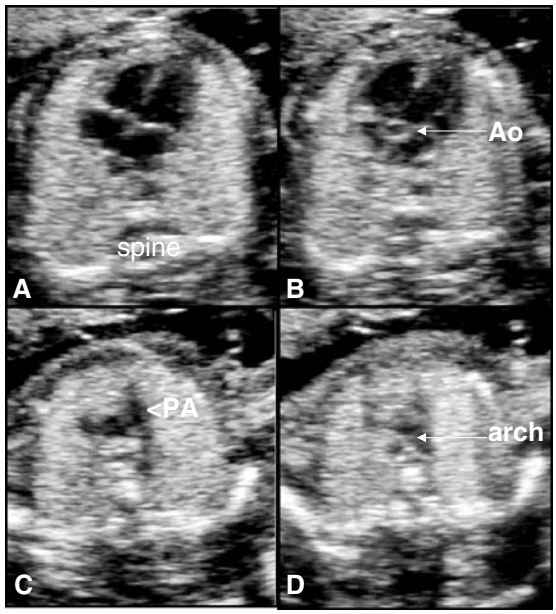


Figure 4.2

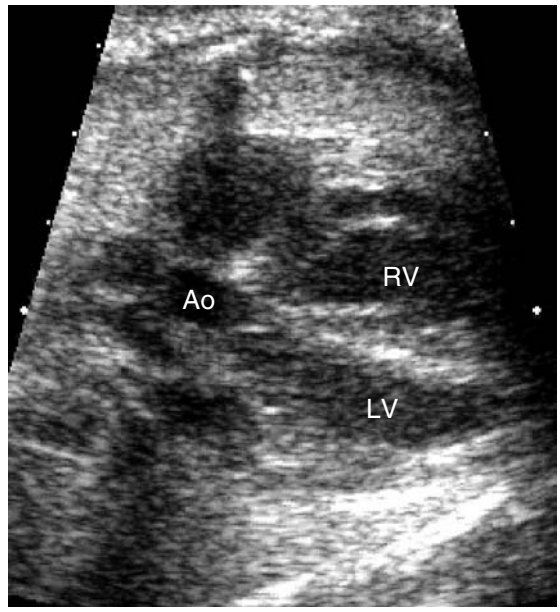


Figure 4.3

The easiest to obtain are those achieved in a transverse ''sweep'' from the four-chamber view, moving the ultrasound beam cranially to image the aortic origin from the left ventricle, the pulmonary artery from the right ventricle with the ductus arteriosus connecting to the descending aorta, and the aortic arch, sequentially (**Figure 4.2**). The longitudinal views are more familiar to examiners who are accustomed to postnatal cardiac imaging. When the transducer is angled from the four-chamber view towards the right shoulder, the aorta can be seen arising from the left ventricle (**Figure 4.3**) and sweeping out to the right of the thorax. In this long-axis view of the left ventricle, the mitral valve, between the left atrium and left ventricle, and the aorta arising from the left ventricle can be identified. A sagittal view of the foetus passing through the sternum enables the right heart connections to be seen (**Figure 4.4**). This section of the foetus demonstrates the inferior vena cava entering the right atrium, the tricuspid valve between the right atrium and ventricle, the origin of the pulmonary artery from the right ventricle, and its connection to the duct. This is a long-axis view of the duct. The aortic arch can also be imaged in a longitudinal section of the foetus, just to the right of the long-axis view of the duct (**Figure 4.5**). The aorta normally arises in the centre of the thorax and forms a tight hook shape with the vessels of the head and neck (arrows) arising from the crest of the arch. However the great arteries are imaged, the following important features should be noted:

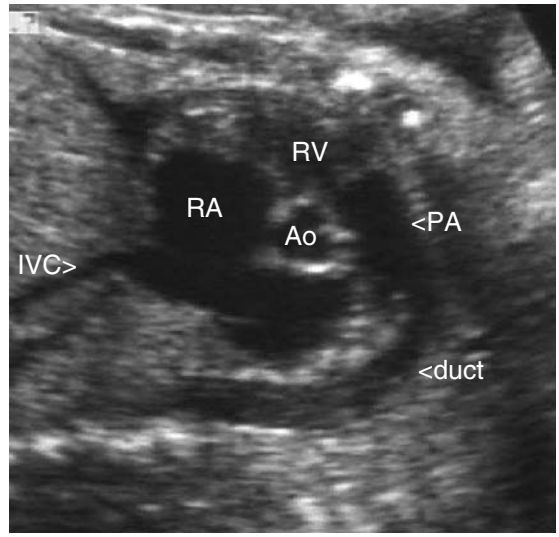


Figure 4.4

1. Two freely opening arterial valves are seen with laminar forward flow across them on colour-flow mapping.
2. The aorta arises wholly from the left ventricle.
3. The great arteries are similar in size, but on measurement the pulmonary artery at the valve ring is slightly bigger than the aorta.

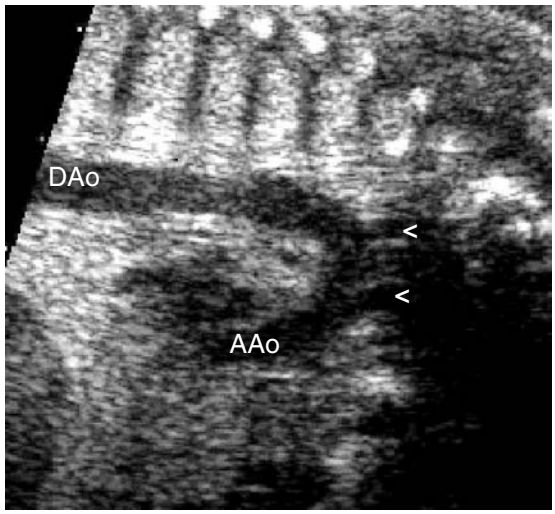


Figure 4.5

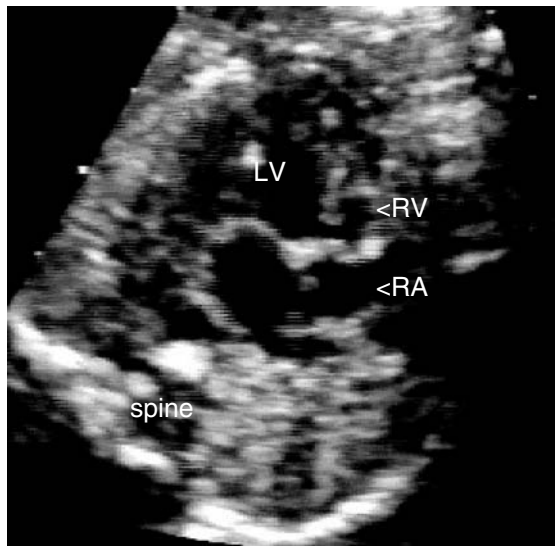


Figure 4.6

4. The pulmonary valve is anterior and cranial to the aortic valve.
5. At their origins, the great arteries lie at right angles to each other, with the pulmonary artery ''crossing over'' the aorta.
6. The size of the aortic arch is similar to that of the pulmonary artery and duct, and the arch is complete.
7. Flow in the arch and duct is in the same direction on pulsed Doppler or colour-flow mapping.

If all these normal features are seen, major anomalies of the great arteries can be ruled out.

CARDIAC MALFORMATIONS

Abnormalities of Connection

The echocardiographic method of diagnosing cardiac abnormalities prenatally generally follows the same rules as in postnatal life. For example, in *tricuspid atresia*, no patent valve is seen in the normal position between the right atrium and right ventricle. The associated right ventricular hypoplasia and ventricular septal defect are also appreciable (**Figure 4.6**). The connections of the great arteries in this condition must be identified because this will influence the prognosis. **Figure 4.7** shows a common atrio-ventricular valve, closed during systole, in a *complete atrio-ventricular septal defect*. This is one of the commonest forms of heart disease seen in prenatal life (Machado *et al*., 1988), representing almost 20% of foetal series, in comparison with the expected rate of around 5% found in postnatal life. This type of defect is found prenatally mainly in association with trisomy 21

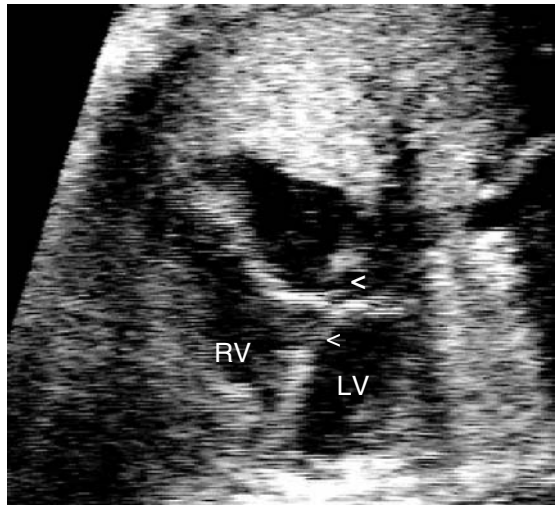


Figure 4.7

but also occurs with other complex cardiac anomalies and atrial isomerism. An isolated defect can occur in non-isomeric and chromosomally normal children, but it is uncommon. When the diagnosis of an atrio-ventricular septal defect with normal atrial situs is made, a foetal karyotype is recommended. Factors that influence the prognosis for corrective surgery, such as atrio-ventricular valve regurgitation or hypoplasia of either ventricle, can also be identified. In *pulmonary atresia*, the pulmonary root is not found in its usual position, or it is small

in relation to the aorta. Pulmonary atresia with intact ventricular septum, of the form most commonly seen postnatally, is characterized by a small and hypertrophied right ventricle. However, pulmonary atresia may also be found prenatally with a dilated right ventricle and severe tricuspid incompetence. In any form of pulmonary atresia, progression of the severity of valvar obstruction has been observed as pregnancy has advanced. Thus, a stenotic pulmonary valve with high-velocity flow may become completely atretic with no forward flow. In *aortic atresia*, the aorta is tiny and the left ventricle small. Prenatally, the left ventricular cavity can be impossible to find, or the left ventricular cavity can be small, thick-walled, and echogenic. An example is seen in **Figure 4.8**. In addition to the four-chamber view abnormality, the aorta is hypoplastic from its origin to the site of entry of the duct, where it becomes larger. Flow can be seen to be reversed on pulsed Doppler or colour flow-mapping of the transverse arch. It is the most common form of CHD recognized prenatally, representing about 20% of foetal abnormalities detected, which is more than twice the expected rate of detection of this defect in infants. The high detection rate of this obvious four-chamber view abnormality indicates the success of the four-chamber view screening programme. In *transposition of the great arteries*, the normal positional arrangement of the arteries is lost. The aorta arises anteriorly to the pulmonary artery and parallel to it (**Figure 4.9**),

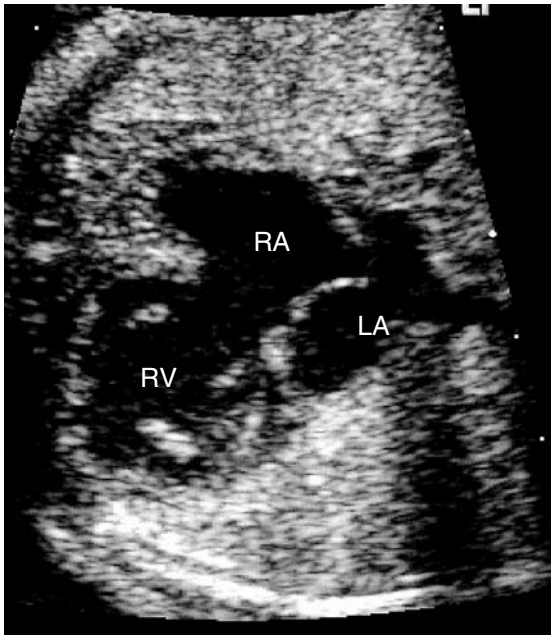


Figure 4.8

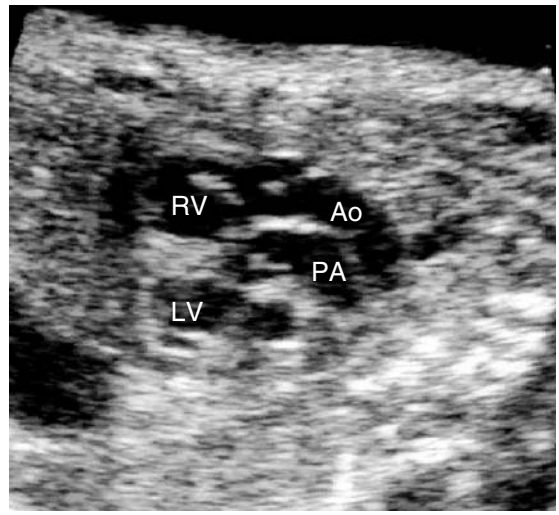
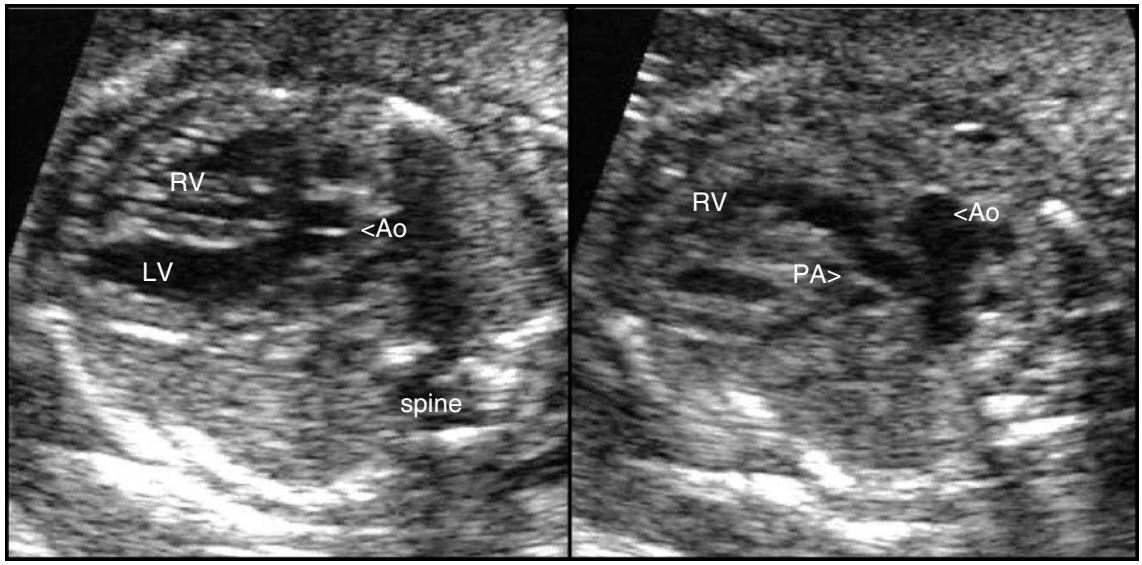


Figure 4.9

instead of being at right angles to the pulmonary artery at its origin in the normal foetus. The anterior vessel gives rise to the aortic arch, whereas the posterior vessel branches into the duct and pulmonary arteries. The aorta is connected to the right ventricle and the pulmonary artery to the left ventricle. Transposition is increasingly recognized during routine scanning as the ultrasonographer becomes more skilled at great artery evaluation in addition to four-chamber imaging. Aortic override can be recognized prenatally by the anterior displacement of the aorta and the presence of an outlet ventricular septal defect. The possible diagnoses, when aortic override is identified, of *Fallot's tetralogy* and *truncus arteriosus*, are differentiated by examination of the pulmonary outflow tract. In **Figure 4.10**, a great artery is shown arising astride the ventricular septum; just cranial to this view, the main pulmonary artery is seen to be smaller than the aorta. The size and confluence of the pulmonary arteries can be examined in order to predict prognosis in tetraology, as can the morphological features and competence of a truncal valve. In prenatal life, tetralogy of Fallot is frequently associated with extracardiac, particularly chromosomal, anomalies, much more than is anticipated from postnatal practice. In *double-outlet right ventricle*, both great arteries can be seen arising from the right ventricle anterior to the ventricular septum. An example is illustrated in **Figure 4.11**. In this case, the anterior artery proved to be the aorta. The small size of the aorta relative to the pulmonary artery raises the suspicion of coarctation as an additional lesion.



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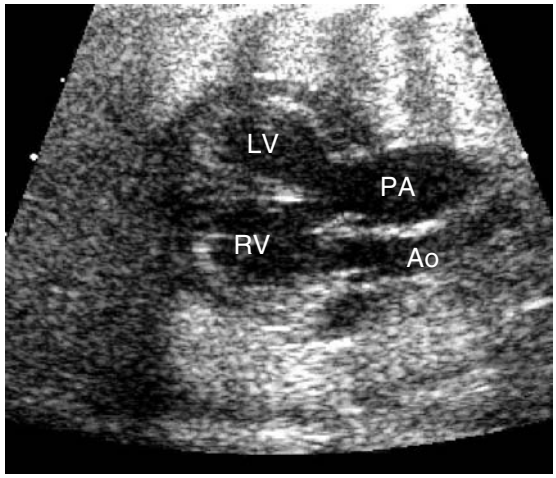

Figure 4.11

Additional Abnormalities

Many other cardiac malformations that do not involve the connections of the heart can be recognized prenatally. They include:

1. Valve stenosis.
2. Ventricular septal defect.
3. Valvar dysplasia or displacement.
4. Cardiac tumour.
5. Aortic arch abnormalities.

In *aortic or pulmonary valve stenosis*, the respective ventricular chamber and the valve itself may appear thickened. The affected artery is often disproportionately small in relation to the other artery. The Doppler sample volume placed in the stenosed artery sometimes shows a velocity of blood flow above the normal range, but this is not such a consistent feature as that produced by a valve stenosis postnatally. In critical aortic stenosis, the left ventricle is characteristically globular and echogenic (**Figure 4.12**) In the moving image, the poor contraction can be readily appreciated. *Ventricular septal defects*, when recognized prenatally, are frequently large and associated with chromosomal anomalies. A large mid-muscular defect is seen in **Figure 4.13**. *Tricuspid valve dysplasia*, with or without *Ebstein's anomaly*, is a common abnormality recognized prenatally. Both tricuspid valve dysplasia and Ebstein's anomaly result in incompetence and right atrial dilatation. This dilatation may be extreme and lead to secondary lung compression and hypoplasia. An example of relatively mild Ebstein's malformation is seen in **Figure 4.14**. There is mild right atrial dilatation and the displacement of the septal leaflet of the tricuspid valve into the right ventricle is more exaggerated than normal. Although both tricuspid dysplasia and Ebstein's malformation are relatively uncommon in paediatric practice, they are more commonly seen in prenatal series. This is probably because secondary lung hypoplasia results in early postnatal death in the most severe cases. There are few survivors in cases where the right atrium is significantly dilated during early gestation.

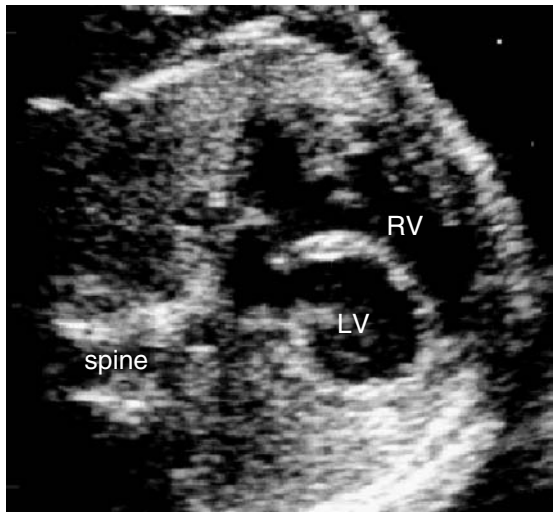

Figure 4.12

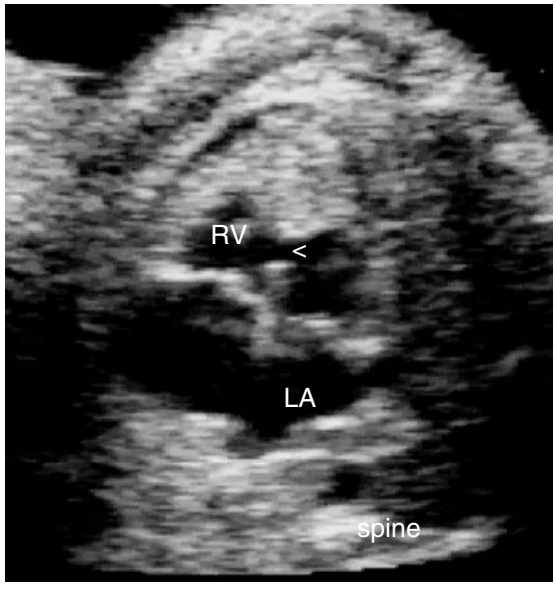

Figure 4.13

Cardiac tumours are not infrequently seen in prenatal life. They carry a high risk of blood flow obstruction, foetal hydrops and intrauterine death. Cardiac teratomas have been identified, but the majority of tumours are rhabdomyomas histologically and are associated with tuberous sclerosis. *Coarctation of the aorta* is a common form of heart disease, but the cases recognized prenatally constitute the more severe end of the spectrum of this disease. The association of cystic hygroma, foetal hydrops, coarctation and Turner's syndrome is

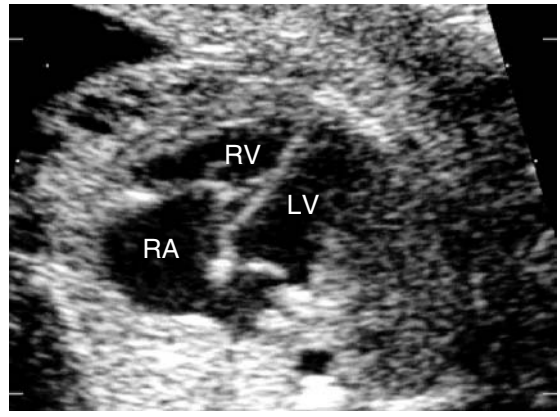

Figure 4.14

commonly recognized in early foetal life. Clues to the diagnosis of arch anomalies include the recognition of enlargement of the right ventricle and pulmonary artery in relation to the left ventricle and aorta respectively, in early pregnancy. Echocardiographic examination of the arch of the aorta reveals varying degrees of narrowing in coarctation, or the arch is incomplete in *interruption of the aorta*. Progressive arch hypoplasia has been observed during pregnancy, resulting in more severe aortic arch narrowing at term than was expected earlier in foetal life.

IMPLICATIONS OF FOETAL ECHOCARDIOGRAPHY

The spectrum of disease seen in prenatal life is different from that observed in infants (Allan *et al*., 1984b). Malformations detected tend to be the most severe forms of heart disease, and defects that are not commonly seen postnatally, such as tricuspid dysplasia and cardiac tumours, are disproportionately frequently recognized prenatally. Some affected foetuses do not survive intrauterine life, which accounts partly for the discrepancy between prenatal and postnatal incidences of severe CHD. A higher proportion (12–20%) of cardiac abnormalities detected in prenatal series are associated with chromosomal defects than the proportion found postnatally. The possibility of chromosomal defects must be included in the counselling of the parents, and karyotyping must be discussed with the mother when appropriate. The presence of multiple congenital anomalies contributes to the high mortality rates found in prenatally detected heart disease. The severity of the spectrum of disease seen is reflected in the outcome of pregnancies in which foetal heart disease has been detected. A recent personal series is illustrated in

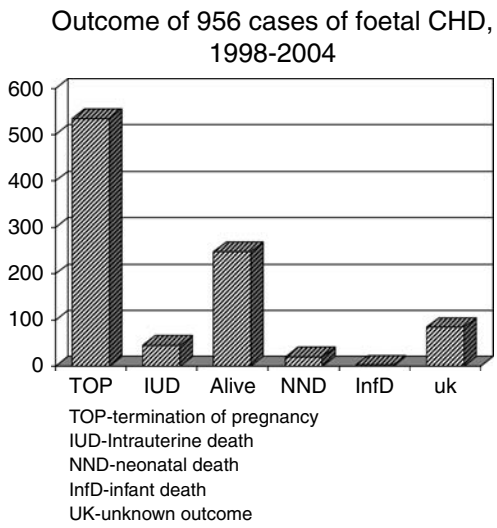

Figure 4.15

Figure 4.15. It can be seen that a high proportion (56%) of parents elected termination of pregnancy, but this is a higher rate than other centres due to the nuchal screening programme, which yields high incidence of chromosomal anomalies (nearly 50%) in our series. The decision concerning termination will vary with social class and religious beliefs, as well as the gestational age at diagnosis and the type of CHD found. It also varies internationally, with a reported incidence of 26% in Italy (Fesslova *et al*., 2003), 24% in the USA (Brick, 2002) and 52% in France (Mirlesse *et al*., 2001). However, the lower rates reflect a lower standard and later gestation of detection of CHD during obstetric screening. Termination of pregnancy was 65% when the diagnosis of CHD was made before 25 weeks in another Italian series (Paladini *et al*., 2002). Parents make their own decision concerning termination or continuation of the pregnancy, on the basis of the information and prognosis given by the paediatric cardiologist. It may also be helpful for the parents to talk to the paediatric cardiac surgeon. If there are associated extracardiac anomalies, or if the long-term outlook for complex forms of CHD is poor, the majority of parents will opt for termination, especially as the risk of recurrence of CHD in a subsequent pregnancy is low. In the future, therefore, some forms of CHD will become less frequent. For example, the prevalence of pulmonary atresia with intact ventricular septum fell in England from 5.6/100 000 to 4.1/100 000 live births, as a result of prenatal diagnosis and termination of pregnancy, in the study reported by Daubeney *et al*. (1998). One of the most important aspects of prenatal diagnosis of CHD is to deliver the newborn in ideal circumstances, such that haemodynamic stability can be established and maintained prior to surgical treatment. Foetuses with duct-dependent lesions would therefore be expected to show a benefit from prenatal diagnosis. This has proved difficult to demonstrate in practice because of the necessity of grouping sufficient numbers of cases by diagnostic category, and also due to the failure to ascertain cases who die without reaching the referral centre (Kumar *et al*., 1999). However, a large population-based study from Paris showed a clear benefit in mortality in transposition of the great arteries when the prenatal and postnatally diagnosed cases were compared. There was a preoperative mortality of 6% and a postoperative mortality of 8.5% in the cases postnatally diagnosed, in contrast to no losses in the prenatally diagnosed group (Bonnet *et al*., 1999). It is likely that this benefit from prenatal diagnosis will be demonstrable also in other categories of CHD in the future.

CONCLUSIONS

Foetal echocardiography can accurately detect structural heart malformations from as early as 12 weeks gestation. Almost all forms of CHD have been recognized prenatally. Once an ultrasonographer performing routine obstetrical scanning has learned to recognize the normal cardiac anatomy, deviations from the normal appearance or failure to find the standard views indicates the need for specialized help from the foetal or paediatric cardiac echocardiographer. An unusual and more severe spectrum of heart disease, frequently occurring in association with extracardiac anomalies, has been detected in foetal life. Many of the features of an individual case of CHD that are known to affect prognosis can be recognized prenatally. However, some defects have been observed to progress in severity between early diagnosis and delivery, in such a way that the surgical mortality has proved higher than expected.

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5

Magnetic Resonance Imaging and Computed Tomography in the Diagnosis of Congenital Heart Defects

V. Mutharangu, A. M. Taylor and R. Razavi

MAGNETIC RESONANCE (MR) IMAGING

Management of patients with congenital heart defects relies heavily on imaging, with echocardiography remaining the most common tool for diagnosis and follow-up. However, echocardiography is limited by acoustic windows, provides poor images of the distal vasculature, and is user-dependent. Thus, for more detailed assessment, X-ray guided cardiac catheterization remains important. Cardiac catheterization is not without risks, due to the invasive nature of the procedure. In addition, exposure to medical radiation is increasingly being recognized as a significant health risk for both patients and health care professionals. Furthermore, X-ray fluoroscopy provides only a projection image with limited three-dimensional (3D) capabilities. Cardiac magnetic resonance (MR) imaging is now an important tool for diagnosis and follow-up of adolescent and adult patients (grown up congenital heart disease, GUCH) with congenital heart disease (Higgins *et al*., 1984), and has become more widely used in neonates and younger children (Razavi *et al*., 2003 (Laucet & Cardioly only)). Cardiac MR provides the capability of imaging in any plane and the development of 3D MR techniques opens up the possibility of improved visualization of cardiac anatomy. In this chapter, we discuss the use of MR in congenital heart defects, its use in planning of surgical procedures and follow-up of patients following surgery.

BASICS

Magnetic resonance (MR) imaging relies on the property of certain nuclei (in cardiac MR almost exclusively hydrogen nuclei) to produce an MR signal when a radiofrequency energy (RF) pulse is applied to the body. The hydrogen nuclei can be thought of as a small bar magnet spinning on its own axis. When a patient enters the static magnetic field of the MR scanner, the hydrogen nuclei (mainly in water) line up in parallel, with the static magnetic field creating a spinning net magnetic vector. Application of a RF pulse flips the net magnetic vector more into the transverse plane, and it is the rotating transverse component of the net magnetic vector that induces the MR signal. Production of images from the MR signal requires spatial encoding, which is performed using magnetic gradients. In order for spatial encoding to be performed, an MR signal 'echo' must be produced sometime after the original RF pulse is applied. There are two main ways of producing a signal echo. The first is to apply a second RF pulse (spin echo); this method produces high signal but is slow, due to the need to apply a second RF pulse. It is therefore more suited to static imaging. The second is to produce an echo using the magnetic gradients (gradient echo); this method is faster than spin echo but produces less signal. It is therefore well suited to dynamic imaging. It is beyond the scope of this chapter to describe in full the basics of MR physics in cardiac MR; however, there are many texts available for further reading (e.g. Bogaert *et al*. 2005).

TECHNIQUES

Spin-echo Imaging

Initially, MR assessment of congenital heart defects was performed using spin-echo ''black-blood'' sequences (Higgins *et al*., 1984). Newer sequences, as discussed below, are becoming more important in cardiac MR; however, spin-echo black-blood sequences are still useful, particularly for accurate delineation of great vessel anatomy. In spin-echo sequences, blood appears black allowing excellent delineation of the vessel wall

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and myocardium. The other main benefit of spin-echo black blood sequences is that contrast improves when the blood flow velocity is high. Thus, it is a useful technique when imaging vascular obstructions and valvular stenosis, as, unlike gradient echo and angiographic sequences, it performs better in these situations. It is also useful when imaging vascular and respiratory structures. These sequences also appear to be much less affected by artefacts caused by metallic objects (such as surgical clips or stents).

Gradient-echo Imaging

Gradient-echo imaging is becoming increasingly important in cardiac MR. Gradient-echo sequences have high temporal resolution, enabling acquisition of multiple phases of the cardiac cycle. These images can be reconstructed into a cine MR representing one full cardiac cycle. Recently, newer gradient-echo sequences have become available which have the benefits of high signal noise ratio, excellent blood pool myocardial contrast and high spatiotemporal resolution. Gradient-echo sequences have various roles in cardiac imaging, as set out below. The most widely used gradient-echo sequence is steady-state free precession (SSFP), which is fast and has excellent signal from the blood pool.

Two-dimensional Gradient-echo Imaging

One advantage of newer gradient-echo sequences is that they are partially flow-compensated and thus the blood pool signal is homogenous. However, high velocity blood flow will lead to signal loss, which allows qualitative analysis of blood flow jets. Thus, single-slice imaging allows qualitative assessment valvular dysfunction and dynamic vascular anatomy.

Multi-slice two-dimensional (2D) gradient-echo imaging represents the gold standard for ventricular volumetry (Bellenger *et al*., 2002), as it is not reliant on complex geometrical models. Although gradient-echo sequences allow high spatiotemporal resolution imaging of the ventricles, they do require multiple breath holds. This may not be tolerated in patients, particularly children, with cardiac disease. In such cases the use of respiratory navigators may obviate the need for breath holds, although acquisition times increase significantly. In children less than 8 years of age, general anaesthesia can be used to provide multiple reproducible breath-holds.

Three-dimensional Gradient-echo Imaging

Recently, 3D gradient-echo techniques have been developed that allow fast imaging of the entire cardiac volume (Razavi *et al*., 2003). This technique offers high spatial resolution images with high contrast, allowing simpler volume rendering based on thresholding techniques. In addition, as reconstructed voxel size is isotropic, accurate multi-planar reformatting is possible (Razavi *et al*., 2003). This is particularly useful in the assessment of complex congenital heart defects.

Velocity-encoded Phase Contrast MR

Quantification of blood flow and velocity is an important tool in the management of congenital heart defects. Quantification of the pulmonary to systemic blood flow ratio (Qp:Qs), valvular regurgitation fraction or vascular stenosis is used to assess disease progression, suitability for surgery and timing of surgery. Velocity-encoded phase contrast MR enables non-invasive quantification of blood flow and velocity in major vessels. Cardiac output and Qp:Qs measured using this technique compare favourably with invasive oximetry (Beerbaum *et al*., 2001). In addition, phase-contrast MR has been validated in numerous phantom experiments (Hundely *et al*., 1995). The quantification of valvular regurgitation is also an important use of this technique (Rebergen *et al*., 1993). This technique has been internally validated against ventricular volumetry (Rebergen *et al*., 1993) and is superior to echocardiography for quantification of regurgitant volume.

Velocity-encoded phase-contrast MR also allows an estimation of pressure gradients across stenosis and have been shown to compare well with Doppler echocardiography and catheter data.

Gadolinium Contrast-enhanced MR Angiography

Gadolinium-enhanced MR angiography (Gd-MRA) relies on the signal enhancing properties of dilute gadolinium chelates. This technique has been shown to be useful in delineating thoracic vascular anatomy and identifying stenotic vessels, and the results compare well with X-ray angiography and surgical findings. The major drawback of Gd-MRA is image blurring, as this technique is not cardiac-gated (Razavi *et al*., 2003). This affects the ability of this technique to accurately assess intracardiac anatomy and may affect the accuracy of proximal vessel size measurements. In addition, in the presence of fast-moving blood signal drop-out occurs in Gd-MRA images, which can lead to overestimation of a stenosis.

ACYANOTIC HEART DEFECTS

Atrial Septal Defect

Atrial septal defects (ASDs) are an anatomically heterogeneous group of lesions whose exact nature

has an effect on the natural history and management of this disease. Management of ASDs has changed in recent years, particularly with the increasing use of trans-catheter ASD closure devices. However, trans-catheter techniques are only viable in patients with small to medium-sized ostium secundum defects (Fischer *et al*., 2003) Patients with large ostium secundum defects, sinus venosus lesions or deficiencies in the anterior or posterior inferior rim of the defect require operative repair. Evaluation of ASDs requires definition of type and location of the defect, quantification of the shunt, detection of any intra-atrial thrombus, assessment of RV function and visualization of the pulmonary venous anatomy. Although most patients with ASDs are evaluated by echocardiography, there is a subgroup of patients, in particular where there are concerns about the possible abnormality of the pulmonary venous return, in whom cardiac MR has a significant role to play.

2D spin-echo black-blood sequences and gradient-echo sequences can be used to accurately define ASD anatomy (Taylor *et al*., 1999) and the results compare well with operative findings (Lange *et al*., 1997). 3D balanced-SSFP techniques with isotropic resolution allow accurate multiplanar reformatted images to be produced with no loss of resolution, as well as 3D rendering of the ASD (**Figure 5.1**).

This is particularly useful for assessment of ASD anatomy when deciding to proceed to either catheter or surgical intervention. Indeed, it has been demonstrated that MR is a more powerful predictor of successful catheter closure than trans-thoracic or oesophageal echocardiography (Durongpisitkul *et al*., 2004). Sinus venosus defects are often associated with pulmonary venous abnormalities. Gd-MRA is a sensitive method for detecting pulmonary venous abnormalities (Puvaneswary *et al*., 2003) and can be useful prior to surgical intervention for sinus venosus defects.

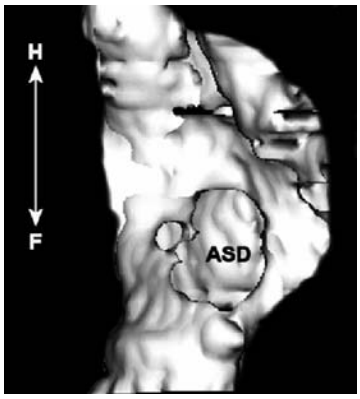


Figure 5.1

Haemodynamic assessment is also an important part of the evaluation of ASDs. Invasive catheterization has previously been used to accurately quantify left-to-right (L–R) shunts (Beerbaum *et al*., 2001). Quantification of L–R shunts using velocity-encoded phase-contrast MR compares well to invasive catheterization results. It has the benefits of being non-invasive, not requiring exposure to ionizing radiation and in certain situations being more accurate than invasive catheterization (Muthurangu *et al*., 2004). Ventricular overload can also be accurately assessed using multi-slice SSFP short-axis imaging (Fogel and Rychik, 1998) and can give important information regarding the timing of invasive intervention.

Ventricular Septal Defect

Ventricular septal defects (VSDs) are a heterogeneous group of lesions that have a common physiological result of shunting at the ventricular level. Cardiac MR can play an important role in the haemodynamic assessment of VSDs. Quantification of L–R shunts using velocity-encoded phase-contrast MR compares well to invasive catheterization results (Beerbaum *et al*., 2001) and has the benefit of being non-invasive. There is also some evidence that in certain situations combining phase-contrast MR and invasive pressure measurements is a more accurate method of calculating pulmonary vascular resistance than traditional methods that rely on the Fick principle (Muthurangu *et al*., 2004).

Operative closure is the accepted treatment for significant VSDs and is associated with low mortality and low residual shunting. However, trans-catheter techniques are increasingly being used in patients with suitable defects. Trans-thoracic echocardiography (TTE) can be used to assess suitability for operative or trans-catheter closure; however, TTE is associated with interoperator variability and difficulties assessing the 3D structure of the defect (Acar *et al*. 2002). Cardiac MR can provide accurate 2D (**Figure 5.2**) and 3D images of the defect. 3D balanced-SSFP techniques with isotropic resolution allow multiplanar reformat of the data to produce image planes that are useful in planning surgical or interventional procedures (Razavi *et al*., 2003). In addition, 3D volume rendering of the lesion can give better insight into the anatomy of the VSD and its relationship with valvular structures.

Optimum treatment (either operative or trans-catheter) of muscular defects depends on accurate demonstration of VSD anatomy. Muscular defects often have complex 3D structure with multiple defects, which can be difficult

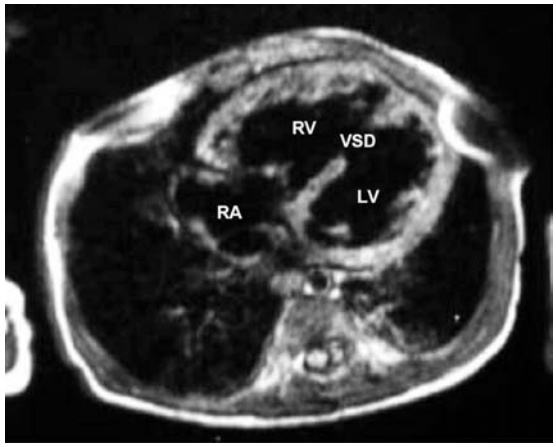


Figure 5.2

to assess with TTE. As explained above, cardiac MR can give accurate representations of 3D anatomy and this information may be particularly useful when planning treatment.

The management of multiple VSDs can be difficult, as both operative and trans-catheter closure can leave residual defects. In such situations palliation with pulmonary artery banding may be indicated. Assessment of pulmonary artery band adequacy relies on demonstration of a significant pressure gradient across the band. Gradient-echo cine MR has been used to assess band adequacy (Simpson *et al*., 1993) and has been shown to compare well with intraoperative and catheter findings. Furthermore, accurate assessment of the Qp:Qs ratio can be obtained, which represents the best way to assess band adequacy.

Coarctation of the Aorta

Coarctation is an area of narrowing, often situated at the site of ductal insertion. Although coarctation can be corrected surgically, up to one-third of patients are hypertensive postoperatively. X-ray angiography has been previously considered the gold standard for diagnosis of coarctation. However, cardiac MR, which is non-invasive, has been shown to be a powerful tool in the pre- and postoperative evaluation of aortic coarctation (Gomes *et al*., 1987). Prior to a surgical (or interventional) procedure, evaluation of the anatomy of the coarctation, as well as the anatomy of the rest of the aorta and head and neck vessels, is vital. Gd-MRA can be used to assess the 3D anatomy of the aorta (**Figures 5.3** and **5.4**), as well as accurately assess the relationship between the coarctation and the head and neck vessels (Debatin and Hany, 1998). However, high-velocity blood

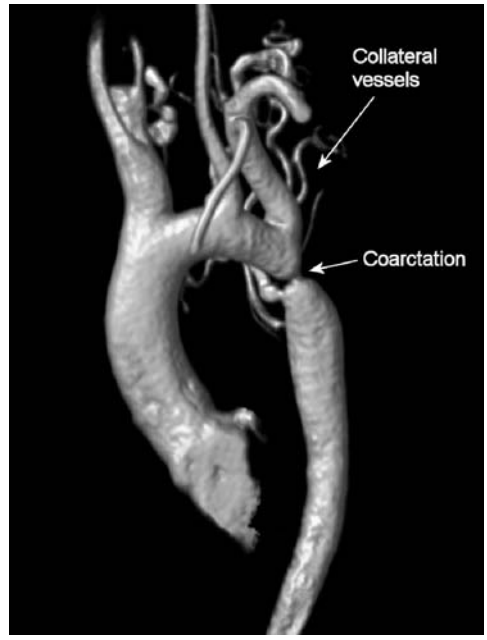


Figure 5.3

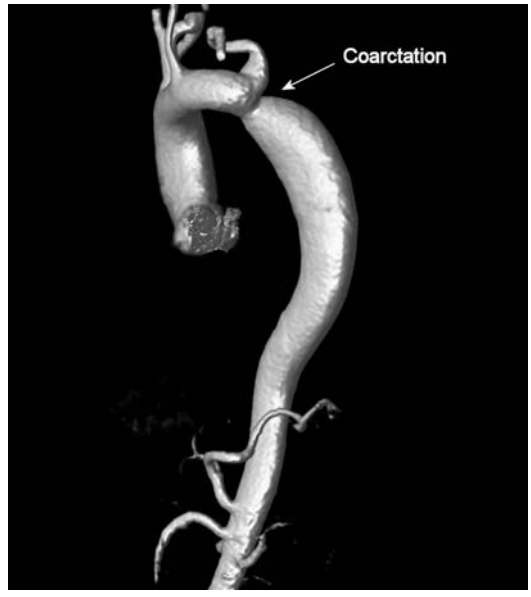


Figure 5.4

flow at the site of coarctation may lead to signal loss and thus overestimation of the narrowing. Spin-echo images (**Figure 5.5**) provide more accurate delineation of the vessel wall and thus more accurate measurement of

Figure 5.5

the degree of stenosis (Holmqvist *et al*., 2002). Velocity-encoded phase-contrast MR can be used to measure peak velocities, and therefore pressure gradients, at the site of coarctation (Mohiaddin *et al*., 1993). Velocity-encoded phase-contrast MR can also be used to measure collateral flow by measuring flow in the proximal and descending aorta (Szolar *et al*., 1996). Postintervention reassessment of collateral flow can be used to assess the success of the treatment (Araoz *et al*., 2003). Cardiac MR can also be used to assess secondary pathology in patients with coarctation, including assessment of the aortic root for dilatation secondary to a bicuspid aortic valve assessment of aortic incompetence and aortic stenosis, and an assessment of ventricular function and LV mass (an indirect indicator of increased LV afterload secondary to either the coarctation obstruction itself or associated hypertension). MRI is very useful for non-invasive follow-up after surgical repair (Bogaert *et al*., 2006), particularly in the evaluation of recoarctation, residual stenosis, persistence of aortic arch hypoplasia, and false aneurysm at the level of repair (Bogaert *et al*., 1995).

CYANOTIC HEART DEFECTS

Tetralogy of Fallot

Tetralogy of Fallot (ToF) is caused by malalignment of the infundibular septum, which leads to right ventricular outflow tract (RVOT) obstruction, a subaortic VSD with aortic override (**Figure 5.6**) and right ventricular hypertrophy. Current management consists of early

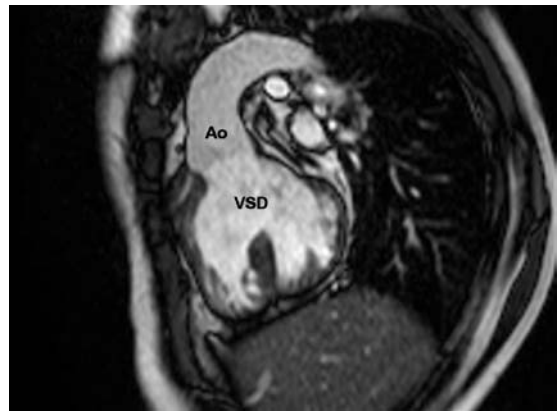


Figure 5.6

single-stage reconstructive surgery, which has the benefit of leaving the patient acyanotic and has good survival rates. However, staged reconstruction may still be required under special circumstances (see Chapter 29).

MR imaging can play a role in deciding between single- and multi-stage reconstructive surgery through delineation of the central pulmonary arteries. It has been demonstrated that Gd-MRAs of the pulmonary artery compare well with both operative and X-ray angiographic findings. In fact, MRI may be more powerful than X-ray angiography in defining the central pulmonary artery structure (Beekman *et al*., 1997).

The main role of MRI in patients with ToF is assessment of postoperative complications. Operative repair of ToF consists of resection of the infundibular stenosis, if necessary enlargement of the pulmonary valve/annulus and closure of the VSD (**Figure 5.7**). The most common late postoperative complication is pulmonary incompetence (Nollert *et al*., 1997), leading to right ventricular dysfunction (Helbing *et al*., 1996). Surgical valve replacement is the current method used in management of patients with severe pulmonary regurgitation. Accurate quantification of regurgitation fraction and definition of RVOT/main pulmonary artery anatomy is important in deciding the type and timing of procedures. Velocity-encoded phase-contrast MR has been shown to accurately quantify pulmonary regurgitation and has been internally validated against MR ventricular volumetry in patients with ToF (Rebergen *et al*., 1993). Although regurgitation can be qualitatively assessed using trans-thoracic or trans-oesophageal echocardiography, the ability of MRI to quantify regurgitant volume makes it a superior technique.

Pulmonary incompetence is often associated with anatomical abnormalities of the RVOT and main

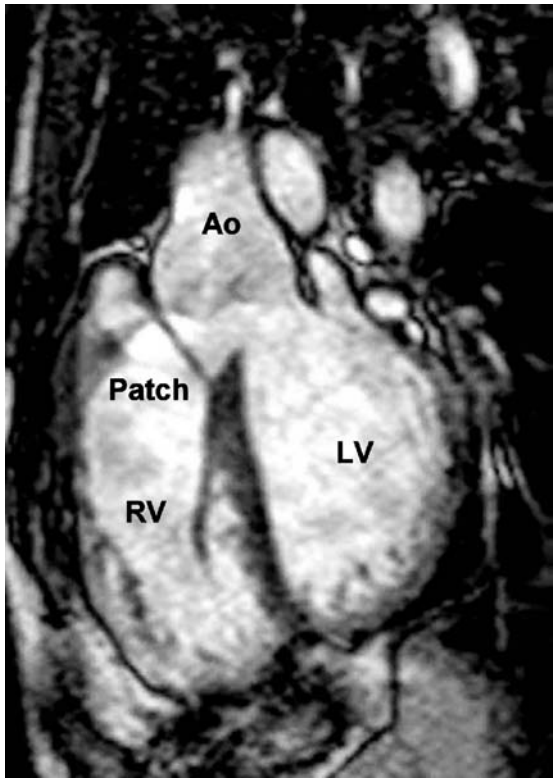


Figure 5.7

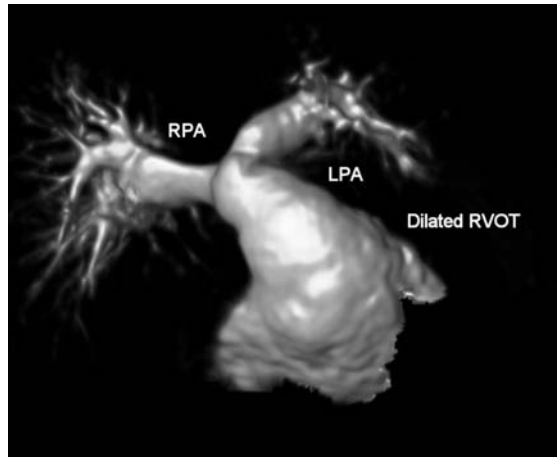


Figure 5.8

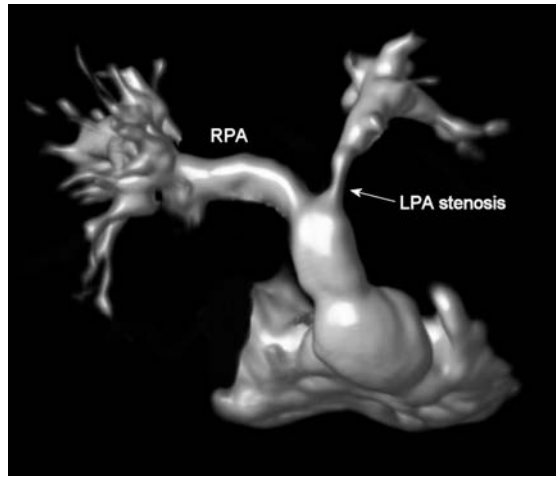


Figure 5.9

pulmonary arteries. Thus, delineation of RVOT anatomy is important for planning surgical or interventional valve replacement (Lutter *et al*., 2004). 2D spin-echo black blood and MRA can be used to delineate RVOT anatomy and quantitatively assess RVOT dilatation (**Figure 5.8**) or stenosis, and measurements made from MR data compare favourably with echocardiography and X-ray angiography (Greenberg *et al*., 1997). Furthermore, velocity-encoded phase-contrast MR can be used to calculate pressure gradients if RVOT obstruction is present, with MR findings comparing well with catheter pressure gradients (Holmvquist *et al*., 2001).

Branch pulmonary artery stenosis, which is often difficult to demonstrate echocardiographically, may also be present in this group of patients (**Figure 5.9**). It is important to detect any branch pulmonary artery obstruction, as such lesions should be repaired at the same time as valve replacement. It has been shown that MRA is more sensitive than echocardiography in anatomically detecting branch pulmonary artery stenoses, and MR findings compare well to operative findings (Greenberg *et al*., 1997).

The final role of MR is in evaluating right ventricular function. This is important for timing of invasive therapeutic measures. Measuring ventricular function is also important when evaluating the effect of any invasive procedure. Multislice short-axis balanced-SSFP imaging has been shown to be the most powerful method of ventricular function quantification (Bellinger *et al*., 2002).

Pulmonary Atresia

Pulmonary atresia is defined as a lack of continuity between the RVOT and the central pulmonary arteries, with a variable degree of hypoplasia of these structures.

Pulmonary atresia is separated into two groups; those with and those without a VSD. As the diagnosis and subsequent management of these two groups is different, it is useful to consider them separately.

Pulmonary atresia with a VSD is the most common variant and is considered by some to be a severe form of ToF, with a subaortic VSD, overriding aorta and often multiple aorta pulmonary collaterals (MAPCAs). Surgical therapy has evolved in recent years and there is now a move towards one-stage unifocalization of MAPCAs, establishment of RVOT to pulmonary artery continuity, with a valved conduit and repair of the VSD. Accurate delineation of the 'true' central pulmonary arteries and detection of all MAPCAs is vital for successful completion of the one-stage procedure. Gd-MRA has a role in preoperative assessment (**Figure 5.10**) and has been shown to be an excellent method of delineating central pulmonary arteries and MAPCAs (Geva *et al*., 2002).

As with ToF, the main role of MR in patients with pulmonary atresia and a VSD is assessment of postoperative complications. Homograft failure, usually with mixed stenosis and regurgitation, is not uncommon. Current management consists of either homograft replacement or some form of valved stent placement. As with ToF, velocity-encoded phase-contrast MR is useful in quantifying regurgitation and measuring peak velocities at the level of the conduit obstruction. Pressure gradients calculated from MR peak velocities compare well with catheter pressure gradients and give some indication of the functional significance of any stenosis (Holmqvist *et al*., 2001). Such functional data (including ventricular volumetry) is useful in deciding the timing of any invasive procedures. Due to the high velocity of blood through most stenosed conduits, spin-echo black-blood sequences are the most useful technique for accurate measurement of conduit obstruction, as they are less susceptible to flow artefacts (Martinez *et al*., 1992). However, Gd-MRA is also useful in assessment of homograft, as the complex 3D anatomy of the conduit is better appreciated in 3D (**Figure 5.11**).

One limitation of both these techniques is the inability to assess the dynamic anatomy of the conduit. 2D balanced-SSFP techniques can be used to assess the change in conduit architecture during the cardiac cycle. Other long-term complications are similar to those found in patients with ToF and are discussed in the previous section.

Pulmonary atresia with intact ventricular septum is the less common variant of pulmonary atresia, and is associated with a variable degree of RV hypoplasia. The presence of an RV infundibulum allows a biventricular repair. Thus, preoperative assessment of the RV cavity is vital. Spin-echo sequences, balanced-SSFP sequences (**Figure 5.12**) and MRA can all be used to assess the RV cavity. The complications of biventricular repair are similar to those in repair of pulmonary atresia and a ventricular septal defect. If the RV cavity is small, then single ventricular physiology is established. The MR assessment of such patients is considered in the section on hypoplastic left heart.

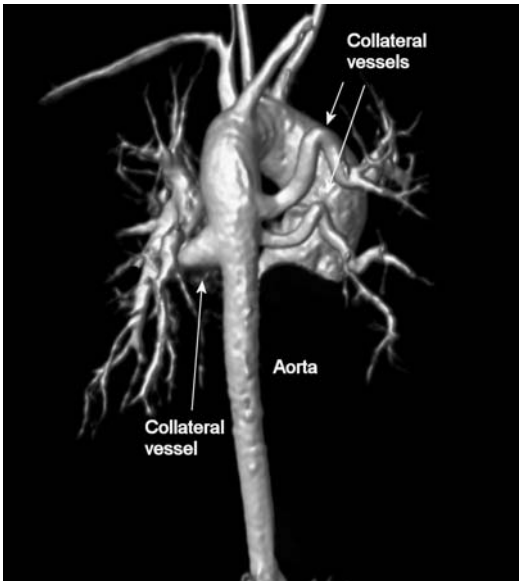
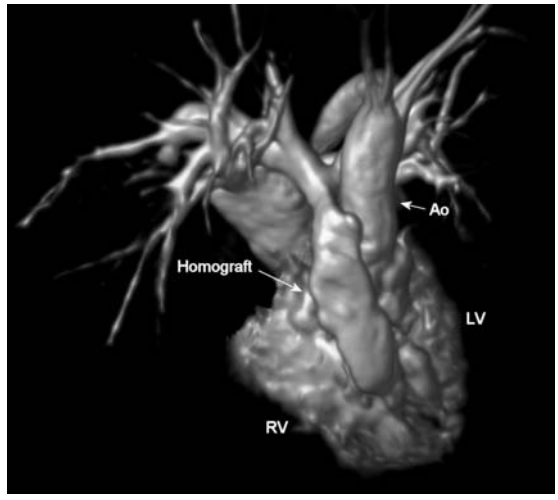


Figure 5.10



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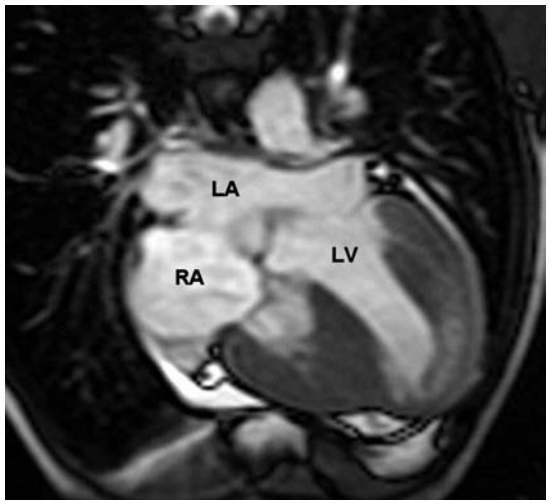

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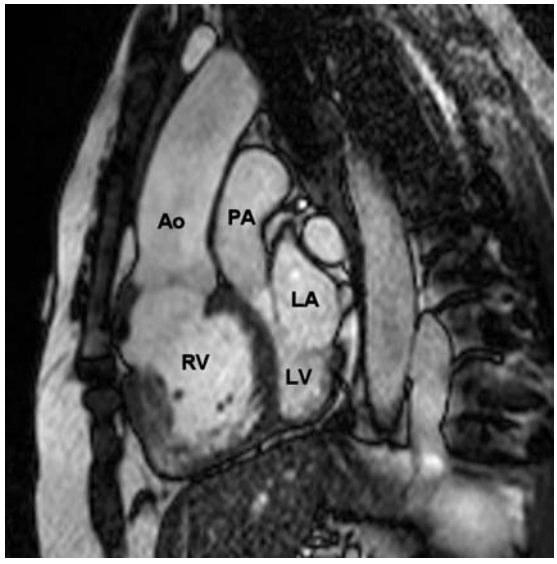

Figure 5.13

Transposition of the Great Arteries

Transposition of the great arteries (TGA) is defined as ventriculo-arterial discordance with an aorta arising from the RV (**Figure 5.13**), and the pulmonary artery arising from the LV. Surgical therapy for this condition was revolutionized with the introduction of intra-atrial switch procedures, which produce a physiologically normal circulation. The arterial switch operation produces both a physiological and anatomically normal circulation. In cases of TGA associated with a VSD and subpulmonary stenosis, the Rastelli procedure is preferred (see Chapters 35 and 36).

The main role of MR is in assessment of postoperative complications, such as RVOT or branch pulmonary artery obstruction (Gutberlet *et al*., 2000). Due to the unusual position of the pulmonary arteries immediately behind the sternum, trans-thoracic echocardiography is poor at detecting branch pulmonary stenosis in these patients. Cardiac MR is not constrained by the intrathoracic position of vessels and is an ideal imaging modality in this patient group. Gd-MRA can be used to visualize 3D pulmonary artery anatomy, particularly the relationship between the pulmonary arteries and the aorta i.e. the anterior–posterior relationship if the Lecompte manoeuvre has been performed (**Figure 5.14**), or side-by-side if the arterial switch was performed without the Lecompte manoeuvre (**Figure 5.15**). The degree of stenosis can be accurately assessed using spin-echo sequences. It has been shown that a combination of these techniques is superior to echocardiography in detecting such obstructive lesions (Hardy *et al*., 1994). In addition, MR compares favourably with X-ray angiography findings. RVOT/branch pulmonary artery narrowing, maybe associated with pulmonary regurgitation, and RV dysfunction. As already stated in the previous sections, MR represents the imaging modality of choice for quantifications of these parameters.

A less common complication of the arterial switch operation is coronary stenosis at the site of

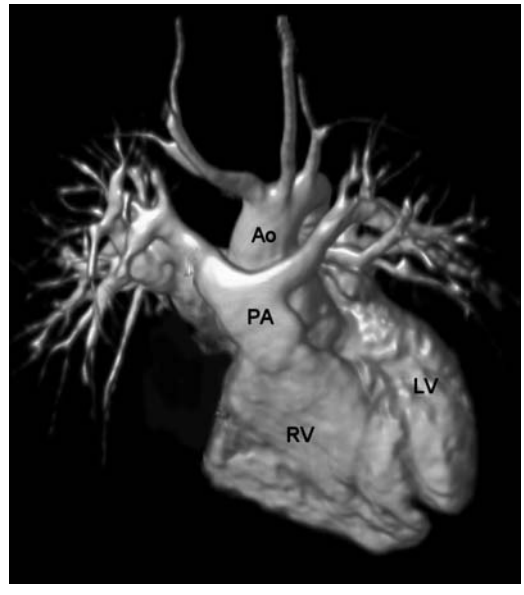

Figure 5.14

Figure 5.15

reimplantation. Although the majority of coronary complications cause early postoperative mortality, a subset of patients suffer from late coronary events. Coronary MR angiography is a useful non-invasive method for investigating coronary arteries, although X-ray coronary angiography probably represents the modality of choice.

Although intra-atrial repair has been superseded by the arterial switch operation, there is a sizeable population who have undergone either a Senning or Mustard operation. The most common complications of these operations are baffle obstruction or leak, arrhythmias and RV dysfunction (see Chapters 33 and 34). The venous pathways have a complex 3D structure and are difficult to accurately assess with trans-thoracic echocardiography. It has been shown that Gd-MRA can fully demonstrate the 3D anatomy of the Senning/Mustard anatomy and detect any luminal narrowing (Fogel *et al*., 2002). Unfortunately, this is a flow-sensitive non-gated sequence image and is susceptible to signal dropout. Spin-echo sequences are useful in accurately measuring the static baffle obstruction (Sampson *et al*., 1994), while 2D and 3D balanced-SSFP sequences are able to accurately delineate the dynamic anatomy of intra-atrial baffles and qualitatively assess flow (Sampson *et al*., 1994). A combination of all these MR techniques allows comprehensive assessment of intra-atrial baffles (**Figure 5.16**).

In cases of TGA with subpulmonary stenosis, the Rastelli procedure is usually preferred. This involves construction of an intracardiac tunnel through the VSD and placement of an RV to pulmonary artery conduit.

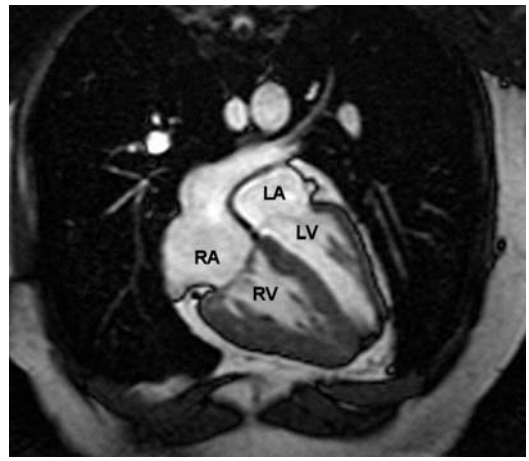

Figure 5.16

The most common complication is conduit stenosis, and the role of MR in assessment of this problem has already been discussed. LVOT obstruction can occur if the VSD becomes restrictive. Gd-MRA and 3D balanced-SSFP techniques can be used to assess the 3D structure of the tunnel.

Congenitally corrected transposition (ccTGA)

This rare disorder is characterized by atrio-ventricular and ventricle-arterial discordance (right atrium to left ventricle to pulmonary artery and left atrium to right ventricle to aorta). Congenitally corrected transposition may be asymptomatic and in some patients is an incidental finding (**Figure 5.17**).

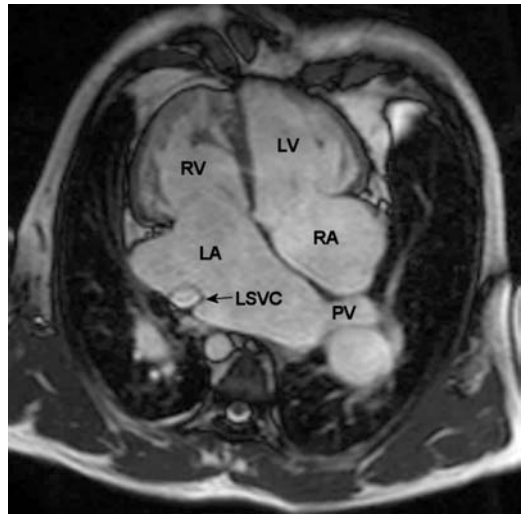

Figure 5.17

However, the majority of patients with ccTGA have associated cardiac lesions (Bjarke and Kidd, 1976), such as VSDs, pulmonary stenosis or tricuspid valve abnormalities. Even without associated abnormalities, the majority of patients with ccTGA develop systemic ventricular failure over time. The main role of MR is in evaluation of associated lesions, quantification of ventricular function and assessment of postoperative complications. Late failure of the systemic right ventricle has led to the increasing use of anatomical repair in ccTGA (either an atrial and arterial switch or a Rastelli procedure with an atrial switch). The complications of these operative procedures and the role of MR in investigating them have been described in the previously section.

Double-outlet Right Ventricle

In double-outlet right ventricle, both great arteries emerge from the right ventricle, with the left ventricle emptying through a VSD. The most common variant is normal arrangement of the great arteries and a subaortic VSD. This variant is often referred to as the Fallot's-type double-outlet ventricle, as it is often associated with pulmonary stenosis. Double-outlet right ventricle can also be associated with an anterior aorta and a subpulmonary VSD, known as the Taussig–Bing anomaly. The role of MR in the assessment of postoperative complications have been discussed in previous sections. However, MR can also play an important role in preoperative assessment in this group of patients. The anatomy of the VSD and the arrangement of the great vessels are particularly important when deciding the type of surgery. Spin-echo black blood imaging of the VSD has been shown to compare well with surgical findings and is able to predict the type of repair which will be finally done (Beekman *et al*., 2000). 3D balanced-SSFP and 2D balanced-SSFP cine techniques are also useful, as they give both anatomical and functional information (Kilner *et al*., 2002). Finally, MRA is useful in delineating great vessel arrangement and the 3D anatomy of the VSD.

Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) constitutes a spectrum of congenital heart defects, with hypoplasia or atresia of the left heart components, and normal relation of the great vessels (see Chapter 41). Surgical treatment was revolutionized by the introduction of the Norwood procedure in 1980 (Norwood *et al*., 1980), with subsequent staged conversion to a total cavopulmonary connection (TCPC). Prior to conversion to a TCPC circulation, preoperative invasive catheterization is performed to identify any increase in pulmonary vascular resistance and any complications that may require repair; branch pulmonary artery stenosis, aortic arch obstruction, tricuspid regurgitation and systemic/pulmonary shunt obstruction. Branch pulmonary artery stensosis, often associated with the BT shunt, is a common post-Norwood procedure. Gd-MRA can give excellent 3D images of any obstruction, although spin-echo images are better for accurate measurement of stenosis. Neo-aortic arch obstruction is also a recognized problem, detection of which is vital, as aortic reconstruction can be done at the same time as conversion of a bi-directional Glenn shunt. Gd-MRA is the modality of choice for imaging the often complex 3D arch obstructions present (**Figure 5.18**).

Obstruction of the BT shunt is another serious complication, with significant mortality. Blalock–Taussig shunt visualization can be difficult with echocardiography and routine invasive catheterization may fail to detect some stenoses. In addition, invasive selective catheterization of the shunt is associated with some risk. The use of spin-echo and MR angiographic sequences has allowed accurate non-invasive visualization of the shunt (**Figure 5.19**), and results compare well with X-ray angiography and operative findings (Ichida *et al*., 1992). Tricuspid regurgitation is often present in the patient's post-Norwood procedure; however, in a small proportion severe tricuspid regurgitation leads to RV failure. This requires annuloplasty, usually performed during

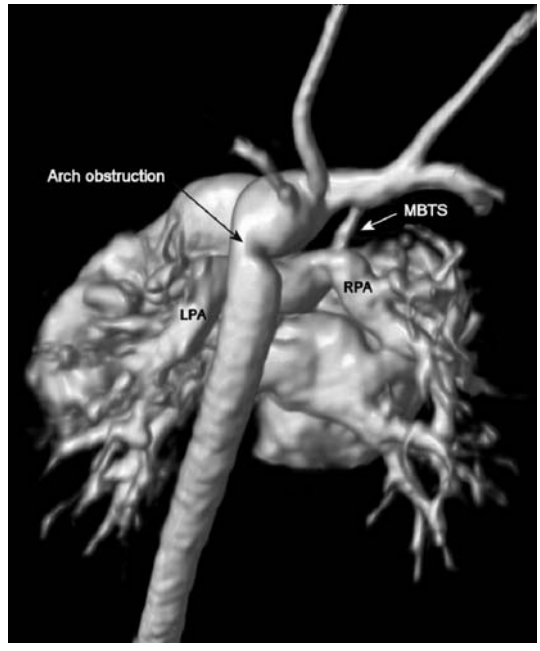

Figure 5.18

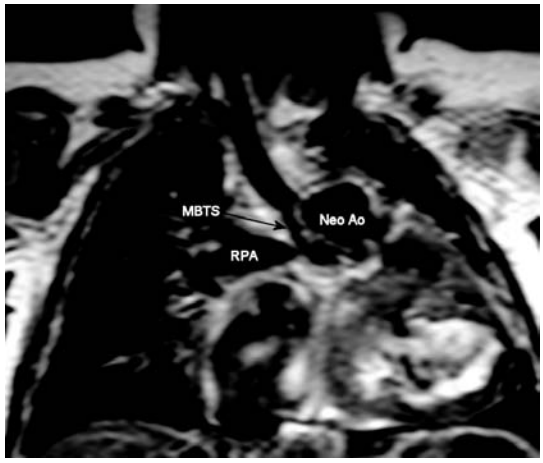

Figure 5.19

the stage II or III operation. Doppler echocardiography is able to detect valvular incompetence; however, with velocity-encoded phase-contrast MR the volume regurgitation and any associated RV dysfunction can be accurately quantified.

At 3–9 months, a stage II (hemi-Fontan, bidirectional Glenn) operation is performed. The main complication following stage II is branch pulmonary artery narrowing. Visualization of the Glenn shunt is difficult with echocardiography, and routine assessment with invasive X-ray catheterization is necessary. In addition, before stage III it is important to detect any pulmonary venous obstructions, as they may lead to failure of the Fontan circuit. MR can give excellent 2D (**Figure 5.20**) and 3D (**Figure 5.21**) images of the Glenn shunt and pulmonary venous anatomy and has been shown to compare well with X-ray angiography and operative findings (Julsrud *et al*., 1989).

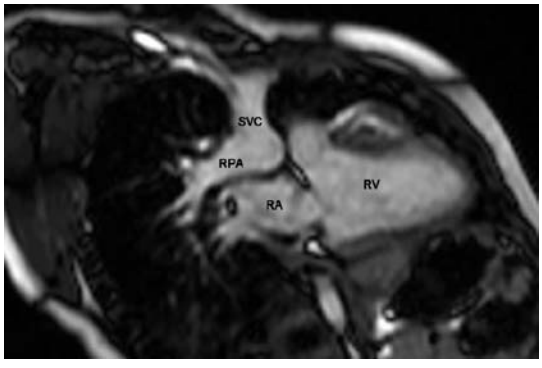

Figure 5.20

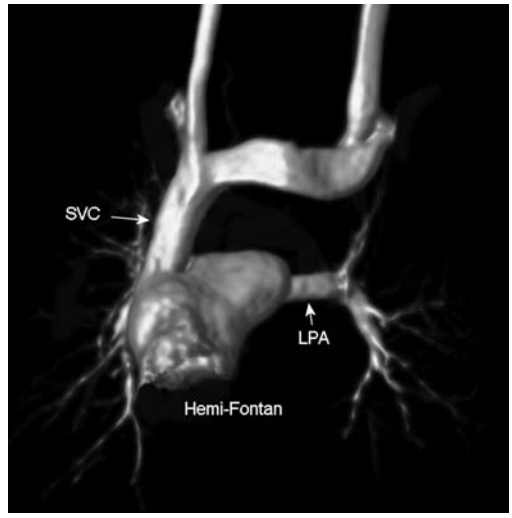

Figure 5.21

The final stage III (total cavopulmonary anatomosis, Fontan) consists of creation of a lateral or extracardiac tunnel between the inferior vena cava (IVC) and the right pulmonary artery or anastomosis of the right atrium to the pulmonary artery. Right atrial dilatation is the major complication of the Fontan procedure and, using MR (either MRA or 3D balanced-SSFP), the 3D anatomy of the atrium can be delineated. Right atrial dilatation can cause pulmonary vein compression and/or formation of atrial thrombi, which can lead to failure of the Fontan circulation. 3D MRA has been shown to be useful in assessing pulmonary vein obstruction (Pilleul *et al*., 2000). MR also has a role to play in the assessment of the modified Fontan procedures, particularly in assessment of the branch pulmonary arteries and tunnels (Fogel *et al*., 2001). Finally, MR can be used to accurately assess ventricular function. It has been demonstrated that ventricular function is lower in patients with Fontan circulation (Fogel *et al*., 1998). In some cases ventricular dysfunction may require cardiac transplantation. Multislice balanced-SSFP imaging of the ventricle is the most accurate method of assessing global ventricular function and is invaluable in this group of patients (Fogel *et al*., 1998).

CONCLUSION

Cardiac MR is becoming an important tool for diagnosis and follow-up of children and adult patients with congenital heart disease, as it provides an accurate, non-invasive method of imaging patients. It represents a viable alternative to X-ray angiography and in some

situations performs better than traditional angiography. Cardiac MR also represents the best available *in vivo* method for quantifications of ventricular function and vascular flow. The use of velocity-encoded phase-contrast MR allows accurate, non-invasive quantification of blood flow and pressure gradients. Finally, although less common in congenital heart disease patients, MR allows assessment of coronary anatomy, perfusion and scar. Thus MR provides a ''one-stop'' imaging modality, allowing assessment of anatomy, function and perfusion.

COMPUTED TOMOGRAPHY (CT)

As discussed in the previous section, MR imaging of the heart and great vessels is often used in the assessment of grown-up congenital heart disease (GUCH) and more recently in neonates, infants and younger children (Muthurangu *et al*., 2005). However, with its increasing availability and utility, multidetector computed tomography (MDCT) imaging is now becoming established as a further method of imaging congenital heart disease. Importantly, as both these cross-sectional imaging modalities place the heart back within the axes of the body (right/left, superior/inferior, medial/lateral), accurate description of cardiac and vascular anatomy in relation to the other structures of the chest is possible (Anderson *et al*., 2004).

The development of spiral (Westra *et al*., 1999; Kawano *et al*., 2000) and subsequently MDCT (Lawler and Fishman, 2001; Siegel, 2003) enables the acquisition of data during a single breath-hold and during the first pass of a contrast bolus, so that images can be reconstructed in any 2D plane or in 3D.

This section provides the reader with a brief overview of the technique of MDCT, and some of the indications for using MDCT in clinical practice.

MDCT Technique

'Slip-ring' technology allows the CT gantry to rotate smoothly, with many revolutions/second. When this is combined with motion of the CT table, the effect is to acquire a spiral of data of the body (somewhat analogous to an apple core). Increasing the number of detectors used increases the thickness of data that can be acquired per revolution of the CT gantry, reducing the total acquisition time without reducing the overall resolution of the image acquired.

For a 16-slice MDCT with 0.75 mm beam collimation and a table feed of 15 mm/rotation, a dataset through the thorax of a neonate can be acquired in 3–6 seconds. In order that the vascular structures are visualized, iodinated contrast must be administered—in effect all images are CT angiograms.

Triggering of the MDCT acquisition is crucial, and should coincide with the peak contrast enhancement in the vascular region of interest. MDCT can be triggered either automatically or by the CT operator. For both methods, an optimal axial slice is defined at the level of the ascending aorta for aortic pathology, pulmonary trunk for pulmonary artery pathology, or left atrium for pulmonary venous pathology.

To reduce radiation exposure, we use a tube current of 15–27 mAs at 100 kV for the majority of neonates and infants; also, the anatomical coverage is limited to the thorax (thoracic inlet to the diaphragm), and no precontrast images are obtained. For children older than 6 years of age, MDCT can usually be performed without sedation; feed and wrap techniques can be used for neonates; and sedation can be used for infants. We only resort to general anaesthesia after failure of sedation or in critically ill, intubated patients. In awake children and those imaged under general anaesthesia, imaging is performed during a breath-hold at end-expiration (Taylor *et al*., 1997).

Images are acquired with an axial spiral volume, and reformatted in sagittal, coronal and oblique planes. 3D volume-rendered images can be created for the majority of subjects.

Indications for Cardiovascular MDCT

In our own clinical practice, MR and MDCT are used as second-line investigations following echocardiography, and often as a replacement for a diagnostic cardiac catheterization.

Echocardiography remains the imaging technique of choice for the initial assessment of young subjects with suspected congenital heart disease, as it provides anatomical and functional information in real time. Echocardiography is non-invasive, portable, relatively inexpensive, and does not use ionizing radiation. Extra-cardiac structures, however, may not be easily visualized, due to the limited ultrasound window, especially in older subjects.

Cardiac catheterization with projectional angiography has been used to define anatomy not well shown by echocardiography and to assess haemodynamic status. However, imaging the great vessels of the thorax is often limited by the presence of overlapping adjacent vascular structures and the difficulty of demonstrating systemic and pulmonary vascular systems simultaneously. Furthermore, the procedure is invasive and relatively high doses of ionizing radiation and iodated contrast material may be required.

MDCT can overcome these limitations by providing high-resolution 3D images of extracardiac structures, without the limitation of echo window or orientation, and

without hindrance of overlying structures. In addition, MDCT provides diagnostic anatomical data that in many ways is superior to cardiac catheterization, in that it is 3D, allows assessment of the great vessels in relation to the other important structures of the thorax (airways, oesophagus and lungs) and is non-invasive. This 3D MDCT information is particularly useful as a road map for surgical/interventional procedures.

VASCULAR ANATOMY

Currently, we use cardiac CT for the following indications:

- Aortic pathology, in particular aortic arch vascular rings (plus airways imaging) (Siegel, 2003; Lee *et al*., 2004; Owens *et al*., 2005). **Figure 5.22** shows the axial reformat from a MDCT angiogram viewed from above, showing a double aortic ring (right arch, arrow; left arch, arrowhead). The compressed trachea (*) and oesophagus can be seen centrally.
- Pulmonary artery anatomy (only if no functional information is required) (Siegel *et al*., 2003; Lee *et al*., 2004; Owens *et al*., 2005). **Figure 5.23** shows a 3D volume-rendered MDCT angiogram viewed from the anterior. The patient was referred for assessment of the branch pulmonary arteries prior to cardiac transplantation. The patient was considered too unwell to undergo general anaesthesia for MR imaging or cardiac catheterization. A patent bidirectional Glenn circulation is shown, with widely patent branch pulmonary arteries.
- Postoperative systemic-to-pulmonary shunts—size and patency.

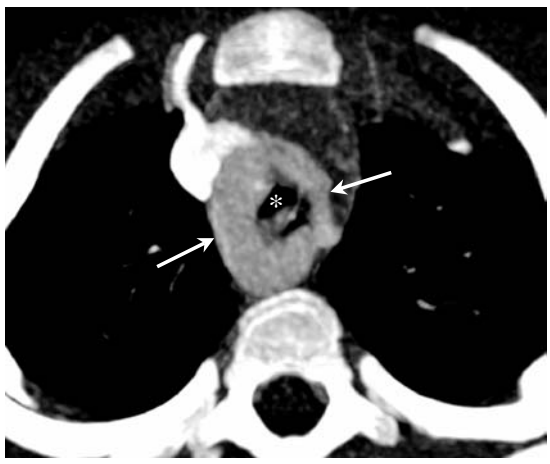

Figure 5.22

Figure 5.23

- Pulmonary venous anatomy—total and partial anomalous pulmonary venous drainage (Kim *et al*., 2000), pulmonary venous stenosis. **Figure 5.24**. shows a 3D volume-rendered MDCT angiogram viewed from the right posterior oblique. (A) Severe left lower pulmonary vein stenosis (PVst), and absent left upper pulmonary venous connection are shown (white arrow). (B) 3D volume-rendered MDCT angiogram viewed from left lateral, showing a supracardiac total anomalous pulmonary venous drainage. The pulmonary veins (black arrow) drain into an ascending vein (*) and then into the brachiocephalic vein (BCV).
- Vascular stent imaging.
- Contraindications to MR imaging—permanent pacemaker, cerebral aneurysm clips, etc.

CARDIAC ANATOMY

The accurate assessment of intracardiac anatomy requires good ECG gating and good temporal resolution. To date, this has only been possible at low heart rates (<60 beats/minute). The majority of neonates, infants and children have heart rates higher than this, and gated intracardiac MDCT imaging has not been performed. Furthermore, in the younger population, intracardiac anatomy is usually sufficiently well defined by echocardiography, and MDCT is usually indicated for the assessment of the great vessels. Despite these limitations, images of intracardiac anatomy are feasible and, with the advent of 64-slice scanners, imaging at faster heart rates should be technically feasible. **Figure 5.25** shows (A) a post-contrast MDCT oblique sagittal reformat through the outflow tracts, showing

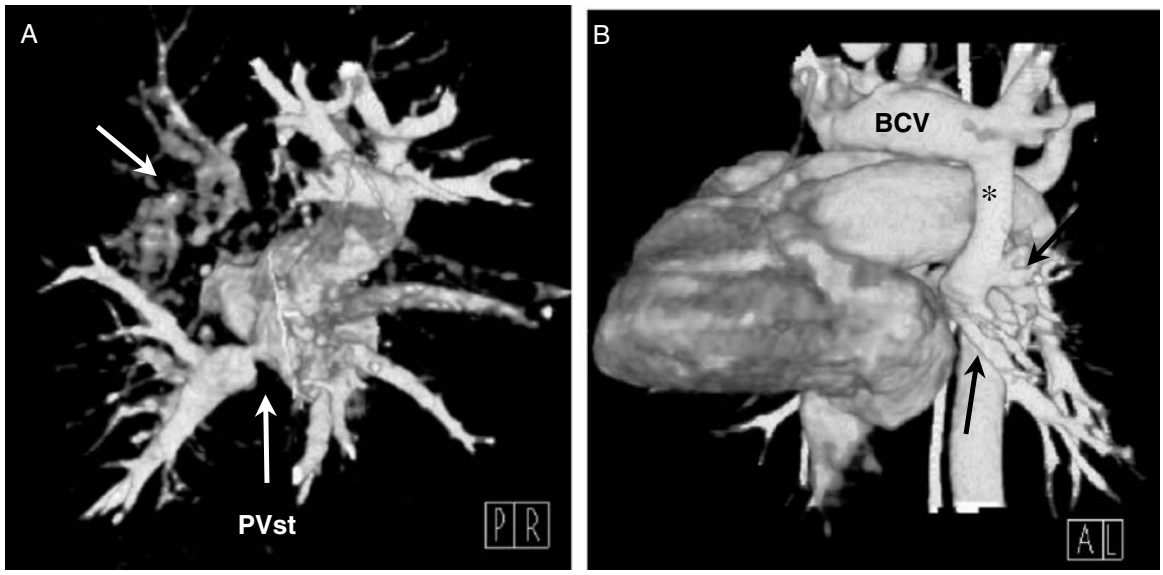

Figure 5.24

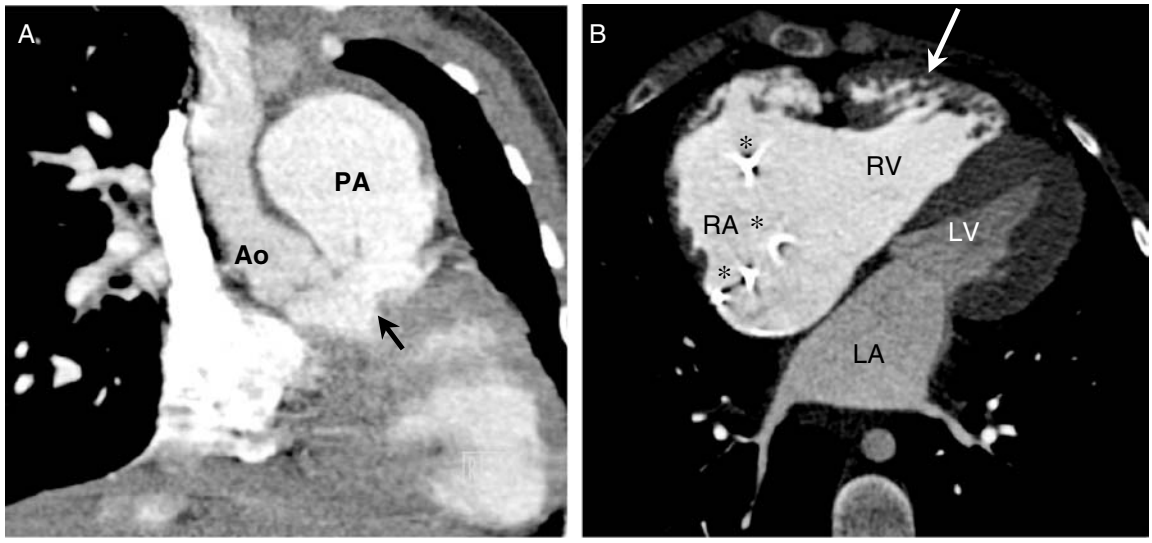

Figure 5.25

a doubly-committed ventricular septal defect (black arrow); Ao, aorta; PA, dilated pulmonary trunk; and (B) post-contrast MDCT four-chamber reformat through the heart in a subject with a restrictive cardiomyopathy. The right ventricle (RV) is small and hypertrophied (white arrow), whilst the right atrium (RA) is markedly dilated. Multiple linear regions of high attenuation in the right atrium (*) are secondary to an *in situ* venous long line. A disadvantage of ECG-gated MDCT is an increase in the radiation dose received (factor of 2–4), and this should be borne in mind.

MRI vs. MDCT

A major advantage of the recent advances in CT technology (16-slice and 64-slice MDCT) over MRI

is the very rapid acquisition time. Imaging can now be performed in a comfortable single breath-hold at the peak of the contrast bolus, reducing the need for general anaesthesia and sedation in young children. Currently, it is our policy to perform MR imaging in children less than 7 years of age under a general anaesthetic. Thus, in this age group, when anatomical information alone is needed, we perform an MDCT scan (unsedated or sedated). However, if quantification of ventricular function or vascular flow (e.g. pulmonary incompetence) is required, we perform an MR scan under general anaesthetic.

For subjects older than 7 years of age and with no contraindications, we perform MR imaging in the first instance. If there are contraindications to MRI (permanent pacemaker, cerebral aneurysm clips, etc.) or when stent imaging is necessary (metal artefact on MR imaging), we perform cardiovascular MDCT.

There are two main disadvantages of MDCT compared with MR imaging. The first is the use of ionizing radiation. As explained above, this can be kept to a minimum, by using low kV, low mAs acquisitions, with current modulation, and image acquisition over the minimal area of interest. Using such protocols, our mean dose for non-cardiac gated cardiovascular MDCT in children is 1.2 ± 0.57 mSv. This equates to approximately 60 chest radiographs (standard PA chest radiograph = 0.02 mSv) or 6 months of background radiation exposure (UK average background radiation = 2.2 mSv/year; Royal College of Radiologists, 2003; Owens *et al*., 2005). Furthermore, in critically ill patients the ability to perform a rapid examination without the need for general anaesthesia or even sedation may be more crucial, as the risk of prolonged sedation may be greater than that of radiation (Siegel, 2003). The second disadvantage of current MDCT techniques is that easy quantification of cardiac function (at high heart rates) and arterial flow are not possible compared with cardiac-gated MR imaging.

FUTURE PROSPECTS

MDCT scanning will become faster and faster, with installation of 64-slice scanners, now beginning, and the development of 256-slice scanners in the near future. This will enable more data to be acquired more rapidly, improving spatial and temporal resolution. This should allow ECG-gated images of cardiac anatomy to be feasible even at high heart rates. The assessment of cardiac function will then be routinely possible. Furthermore, such equipment may make routine, accurate assessment of coronary artery disease with a non-invasive method possible (Khositseth *et al*., 2005).

The recent improvement in non-invasive cross-sectional cardiovascular imaging modalities (MR and CT) has resulted in a change in our approach to the definition of anatomy and physiology in patients with congenital heart disease. The precise role of these imaging modalities in paediatric cardiology will evolve over time. Currently, our own clinical practice is to use cardiovascular MDCT in the initial diagnostic assessment of great vessel anatomy in young subjects, unless there is a very high degree of certainty after echocardiography. Cardiovascular MR imaging is used if further definition of intracardiac anatomy is required, or if ventricular/valvular function or blood flow needs quantifying. Finally, we use cardiac catheterization/angiography if haemodynamic information is required (pulmonary vascular resistance studies), or if there is a high suspicion of coronary artery stenoses. This approach can improve non-invasive diagnosis and reveal detailed anatomy that is important for both clinical decision-making and surgical planning.

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6

Invasive Investigation and Exercise Testing

G. Derrick and J. F. N. Taylor

INVASIVE INVESTIGATION

The main objectives for the detailed investigation of congenital heart disease are the assessment of the nature and severity of the haemodynamic disturbance and the anatomical basis for that disturbance. The predominant place historically taken by cardiac catheterization and angiography in defining these objectives has now been superseded by non-invasive methods, notably echocardiography, but supplemented by magnetic resonance imaging (MRI), nuclear medicine studies and, to a lesser extent, computerized X-ray tomography (CT). The practicability of echocardiography has resulted in this investigation becoming an extension of routine clinical examination, whilst also providing high-quality anatomical diagnosis and some haemodynamic information. If this is supplemented by blood gas analysis and oximetry, many of the objectives of an invasive investigation have been achieved, certainly for the common straightforward lesions.

What is now the place of the formal invasive study by cardiac catheterization and angiography? The current principal indications include:

1. Assessment of pulmonary vascular response.
2. Evaluation of complex pressure gradients.
3. Early postoperative investigation.
4. As an adjunct to interventional procedures.

Angiography remains unique amongst the imaging techniques in that it is not a tomographic modality and, although in a two-dimensional (2D) representation, it does provide information covering the third dimension. Digital subtraction angiography improves the visualization of vascular structures but does not alter the representation.

General Conduct of an Invasive Investigation

Investigation should be undertaken in an environment free from stress and with the infant or child in as near a stable haemodynamic state as feasible for the entire duration of the procedure. In practice, this is best achieved with general anaesthesia and controlled (mechanical) ventilation. The FiO_2 should be kept to the minimum necessary to maintain a normal pulmonary vein oxygen saturation and tension. A trans-cutaneous oxygen-monitoring system is useful in confirming stability. Ventilation is adjusted to maintain normal arterial pCO_2 levels. **Figure 6.1** shows the lowering effect on the pulmonary arterial pressure of correcting the arterial blood gas status by changes in ventilation.

Measures should be undertaken to ensure that the body temperature is maintained, particularly during an extended procedure, and the oesophageal or rectal temperature is monitored. Fluid balance should be maintained intravenously, and arterial blood gas and blood sugar measurements made at regular intervals. The ECG and arterial blood pressure will need to be monitored from both an anaesthetic and a cardiological viewpoint. Excessive blood loss is unlikely to be encountered during diagnostic procedures, although it may be necessary to replace losses during interventional procedures involving the use of large-diameter catheters, especially if unexpected and sudden.

Vascular Access

If cardiac catheterization is being undertaken for the first time, percutaneous vascular access rarely presents a problem. Difficulties arise after multiple approaches via the femoral vessels, particularly if one or more of those procedures have involved an interventional procedure (even as simple as balloon atrial septostomy). Following cardiac surgery, in particular in neonates, or

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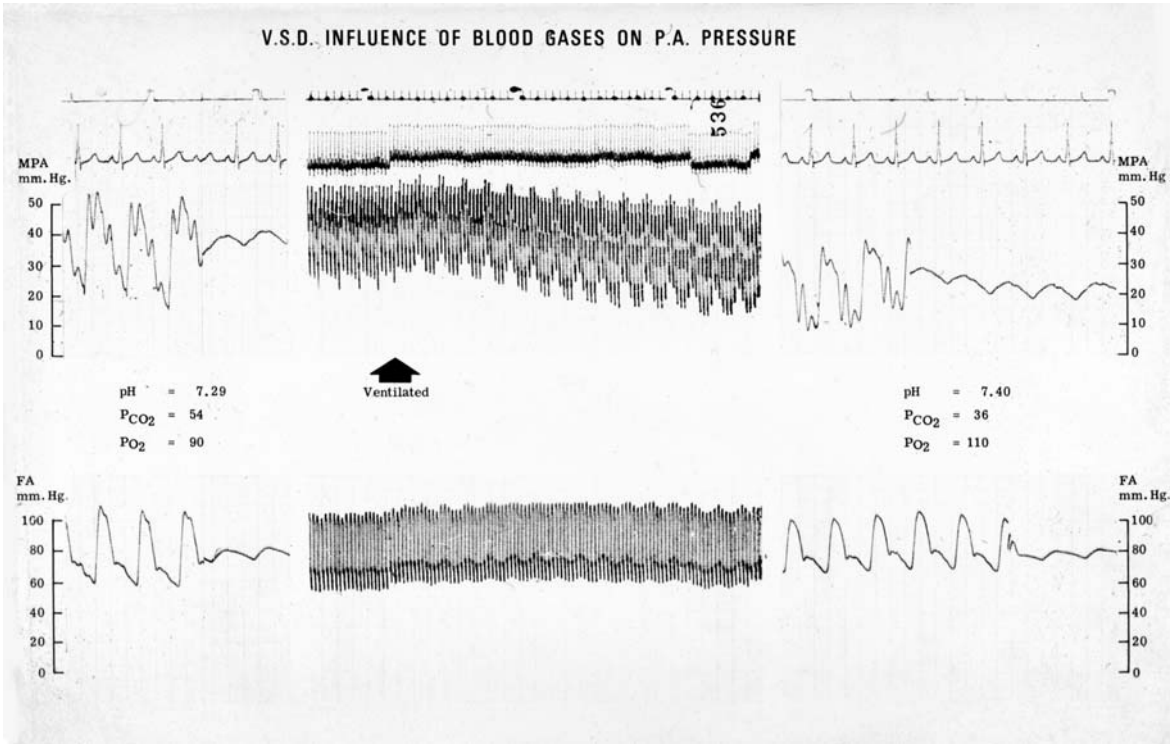

Figure 6.1

after a prolonged stay in an intensive care unit, various peripheral vessels may have been cannulated with long lines, and distortion or occlusion may have occurred to the subclavian, femoral or iliac venous systems. Whilst the preferred site of entry remains the femoral vessels, as access to all the cardiac chambers and great arteries is easier via the inferior vena cava. Another approach becomes necessary if the inferior vena cava can not be approached from below. This may be from the antecubital or (in small subjects) axillary approach; alternatively, in experienced hands, the subclavian route has much to offer. There may be occasions when even the jugular approach is not available to access the cardiac chambers (e.g. after a total superior cavopulmonary connection), but the transcutaneous trans-hepatic approach will allow access to the right heart and pulmonary arteries. The time taken to achieve vascular access if the procedure is not the initial one may be extended (Celermajer *et al*., 1993). **Figure 6.2** shows the relationship between the number of previous cardiac catheterizations and the percentage of patients with difficult vascular access.

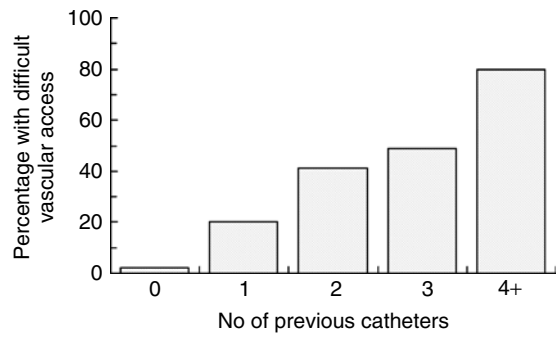

Figure 6.2

Ultrasonography may assist the percutaneous entry and is, of course, mandatory if the trans-hepatic approach is attempted. Direct exposure surgically (''cut-down'') of the femoral or axillary vessels provides little additional benefit, if any, as the vessels tend to be occluded central to the point of access. It is usual to catheterize both an artery and a vein simultaneously. Whilst it is convenient for these two catheters to be side by side, there may be instances when the venous and arterial approaches are from different sites; indeed, two venous sites may be necessary for the full investigation after a superior vena cavopulmonary connection has been undertaken. With multiple catheter sites, as with large diameter catheters

and sheaths, a watchful eye against disconnection and unnoticed blood loss is warranted.

Blood Oxygen Saturation Measurements

The objective is to enter all four chambers of the heart and both great arteries. One vena cava is traversed in order to enter the heart, and the other should be entered, together with one or more pulmonary veins, preferably one pulmonary vein from each lung (if there is full arterial saturation, pulmonary vein entry may not be necessary solely for oximetry). Reliance should not be placed on a single saturation measurement from one site, partly because of the effect of streaming (especially important in the main pulmonary artery and ascending aorta) and partly the accuracy of the oximeters. This does not mean that duplicate samples must be taken from all sites, as one sample may be used to verify the adjacent samples. It does mean that those samples to be used in the Fick calculation of total flow and shunt flow should be verified. The great arterial samples should be taken well away from the respective semilunar valve and care must be exercised in evaluating which samples are to represent the mixed venous saturation. Mention has already been made of the need to adjust the inspired oxygen concentration to achieve full pulmonary vein saturation without a major increase in the dissolved oxygen content.

Pressure Measurements

The accurate determination of the anatomical and physiological consequence of congenital heart disease requires the knowledge not only of the pressure relationship pertaining across each of the four valves in the heart but also of the relationship between the pulmonary and systemic circulations, and therefore the two ventricles and the two atria. It is therefore important that the pressure recordings obtained relate to each other, even though recorded over a time period of 20–30 minutes, or longer if there is an extended study, e.g. of the pulmonary vascular response. Basically this requires a continuous record to confirm haemodynamic stability with respect to time, and equally a contemporaneous record of the pressures representative of the pulmonary and systemic circulations.

The slow withdrawal of a catheter from one identifiable site to another may reveal multiple sites of pressure change as well as identifying the precise location of the most important change. Whilst this is important in the complex outflow abnormalities present after previous intervention, even in the unoperated it may reveal important preoperative detail. **Figure 6.3** shows a withdrawal trace from the pulmonary artery wedge position to the inferior vena cava in an infant with tetralogy of Fallot. There is a pressure gradient at the pulmonary arterial bifurcation, and in the infundibulum of the right ventricle, but not at the pulmonary valve level. The *x*-axis is, of course, time not distance.

Blood Gas Measurements

These measurements are routinely taken to demonstrate that the overall effectiveness of the systemic circulation is maintained and that alveolar ventilation is adequate. This is achieved by analysis of a systemic arterial sample, although the level of alveolar ventilation is better represented by a pulmonary vein or left atrial sample. In studies of the pulmonary vascular status, it is imperative to calculate the pulmonary venous oxygen content accurately and not assume it from an arterial sample; thus, the pulmonary vein sample is needed for both oximetry and pO_2 measurements.

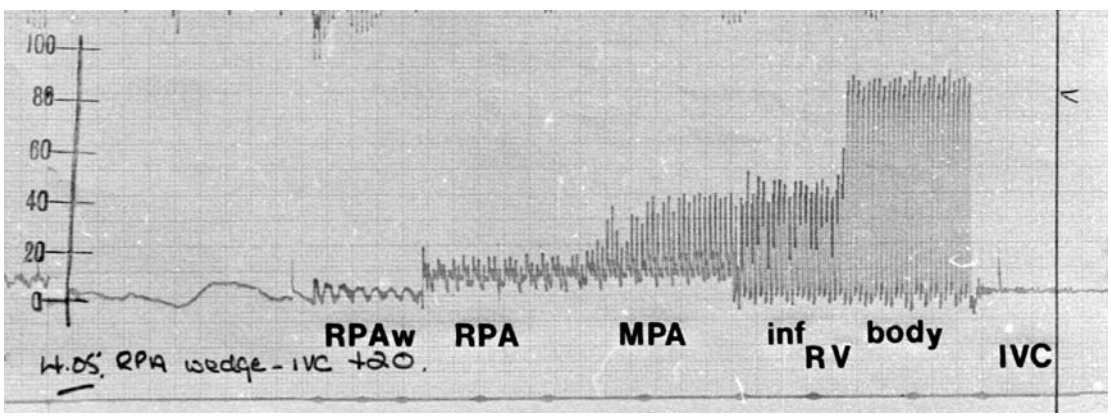

Figure 6.3

During a long procedure or a difficult intervention, regular estimates of the blood gas status will enable timely measures to prevent progressive acidaemia and its attendant haemodynamic instability.

Measurements of Pulmonary Vascular Resistance

Pulmonary vascular resistance is the dividend of the mean pressure drop across the pulmonary vascular bed and the mean pulmonary blood flow. It is an important measure in many congenital heart lesions, as the advisability and risk of intervention by surgical or interventional catheter techniques is determined by changes in the pulmonary vascular bed. It is relatively easy to measure the pulmonary artery pressure directly and, in many instances, the pulmonary vein/left atrial pressure; however, if the left atrium is inaccessible, the pulmonary artery ''wedge'' (pulmonary capillary) pressure may be used. This yields a damped representation of the pulmonary vein pressure waveform, but the mean pressure is an acceptable estimate of the true pressure. The more difficult task is to measure the pulmonary blood flow, which cannot be obtained directly in a clinical investigation. A number of indirect methods are available: the Fick principle, indicator dilution studies using indocyanine green, thermodilution studies or utilizing radioisotope indicators.

The Fick principle is the more widely used method. To calculate the pulmonary flow, it is necessary to measure the oxygen uptake through the lungs and the rise in blood oxygen content between the pulmonary artery and vein. In the past, a number of assumptions have been made in solving the Fick equation. First, the oxygen uptake has been assumed from a variety of tables and is almost certainly inaccurate when applied to an infant or child with congenital heart disease under sedation or anaesthesia. Second, the oxygen content has been calculated from the measured saturation, and the dissolved oxygen content either ignored or calculated from the pO_2. Using mass spectrograph and controlled ventilation, it is possible on a routine basis to measure the oxygen uptake on-line, and the oxygen content of the appropriate blood samples should be measured directly. This will give a measure of the pulmonary vascular resistance at the flows in the pulmonary and systemic beds at that time. More useful information can be obtained by altering the pulmonary flow mechanically and by specific pharmacological intervention on the pulmonary vascular bed. It is imperative that measures specific to the pulmonary bed are used, as any intervention that also alters the systemic circulation (after load, volume load, contractility, heart rate) will affect the calculation of the pulmonary vascular resistance by the effect on the left atrial performance independently of change in the pulmonary vascular bed. As these interventions are time consuming, and require a return to the base state if a series of pharmacological tests are being applied, it is essential that all other parameters associated with haemodynamic stability be maintained, viz. ventilation, blood gas status, fluid balance and body temperature. The relevant intravascular pressures will need to be measured during each phase of the investigation.

Testing the Pulmonary Vascular Response

Historically, the determination of resistance and reactivity of the pulmonary vasculature has implications for treatment and prognosis in patients with pulmonary vascular disease (Barst *et al*., 1999). Various inhaled or injected agents have been considered for reactivity testing, including oxygen, nitric oxide, calcium antagonists, adenosine and prostacycline. Many workers describe the construction of dose–response curves to multiple vasodilator agents, during long-catheter studies (Jolliet *et al*., 1997). Despite the various vasodilator agents available for testing, the maximal vasodilator response is seen with nitric oxide and oxygen (Turanlahti *et al*., 1998). Testing with additional agents serves only to reduce systemic vascular resistance and systemic blood pressure and prolong the investigation. In children, an accurate assessment of cardiac output for the determination of pulmonary vascular resistance is infeasible without general anaesthesia, but the induction and duration of anaesthesia in patients with severe pulmonary hypertension can be life-threatening. For these reasons, in our own unit, a streamlined protocol is employed to achieve the maximum yield of clinically applicable data in the shortest time possible. Pulmonary vascular resistance is calculated at baseline and during a single sequential stage combining both oxygen supplementation and inhaled nitric oxide. With experienced personnel delivering shorter anaesthetics, patient safety and morbidity is improved.

Catheter Placement and Haemodynamic Measurement

Resistance calculations require the measurement of cardiac output and mean trans-pulmonary pressure difference (pulmonary artery pressure – left atrial pressure). Of the methods of cardiac output measurement, there are known limitations. ***Thermodilution*** can be unreliable in small children when the proximal injectate port for the cold fluid injection may not lie in the right atrium, or where intracardiac shunts exist. ***Dye dilution*** is not commonly used for practical reasons. The ***Fick principle*** is easy to apply to a wide age and size range of patients and

pathologies, and is the method of choice in our unit. The principle requires the measurement of arteriovenous oxygen difference (bound and dissolved) and oxygen uptake in the lungs. Systematic error is introduced by assuming oxygen uptake from equations or tables (Li *et al*., 2003), so oxygen uptake should be measured directly with a medical mass spectrometer at each important stage of testing. Temporary balloon occlusion of aortopulmonary blood flow, such as an arterial duct or Blalock–Taussig shunt, is required in order to sample pure mixed venous blood. Systemic arterial blood is equivalent to pulmonary venous blood where intracardiac shunting is not present. Where right to left shunting does exist, pulmonary vein samples must be drawn from the left atrium or the pulmonary vein itself. If access to the left atrium or pulmonary vein cannot be gained by passing through an existing atrial septal communication, or retrogradely through the atrio-ventricular valve, atrial septal puncture can be performed. This latter technique can be hazardous in patients with severe pulmonary hypertension because the atrial septum bows to the left atrial free wall. Using the Fick principle, pulmonary blood flow is calculated and indexed to body surface area (litre/min/m^2).

Pulmonary vein or left atrial pressure can be assessed by pulmonary capillary wedge pressure, or direct measurement. Pulmonary vascular resistance (Wood units. 1 Wood unit = 80 dyne/s/cm^5) is calculated using Ohm's law and unindexed pulmonary blood flow. Pulmonary vascular resistance *index* is calculated using cardiac index, and units become Wood unit/m^2 (Wilkinson, 2001). Resistance measurements may be measured to help determine operability, feasibility of cardiac transplantation, and to assess the effects of pulmonary vascular disease modifying drug treatment.

Angiography

For a complete diagnosis of a congenital heart lesion, and to fulfil preoperative surgical requirements, every segment of the cardiovascular system from the great veins through to the major intrathoracic arteries (including the descending aorta) should be visualized. In most circumstances this is achieved by echocardiography. Angiocardiography and arteriography are necessary to delineate specific lesions and areas that are inaccessible acoustically, to display the totality, particularly of arteries that are represented only in section by the essentially tomographic representation afforded by echocardiography and MRI and to provide a better delineation of chamber size. This technique is also a prerequisite before, during and after a catheter intervention. Visualization of the lesion is enhanced by the use of digital subtraction angiography (DSA), which now has comparable resolution to classical photographically-based angiography, even at high frame rates (50 frames/s).

To achieve the best definition, two main principles should be followed. First, the contrast medium should be injected immediately upstream of the point of interest (even when using DSA), with a flow rate adjusted to deliver the contrast volume within 1.5 s. Second, the X-ray beam should be perpendicular to the lesion of interest. It may be helpful to have a simultaneous orthogonal projection. The anterior and lateral projections demonstrate the pulmonary vessels within the lung in detail, and of course these vessels cannot be imaged echocardiographically. If one of the X-ray beams is angled towards the long axial oblique projection (55–65° left oblique) and inclined 20° in a caudocranial direction, a left ventricular angiogram will clearly outline the perimembranous septum, aortic valve and ascending aorta (**Figure 6.4**). A steeper angulation to 40° left oblique, 20–40° cranial (hepatoclavicular or four-chamber view) will outline the trabecular part of the ventricular septum during a left ventricular angiogram (**Figure 6.5**), and the pulmonary artery bifurcation during the right ventriculogram (**Figure 6.6**). Other structures that are optimally displayed are the crux of the heart and its adjacent structures, the atrio-ventricular valves, the lower interatrial septum and the upper interventricular septum. This may be more conveniently achieved by using the lateral tube and intensifier. The foregoing descriptions are only illustrative of the principle that angulations to the beam from the traditional and overall most useful anterior and lateral projections will enhance the visualization of specific segments of the cardiovascular system. The objective of optimal

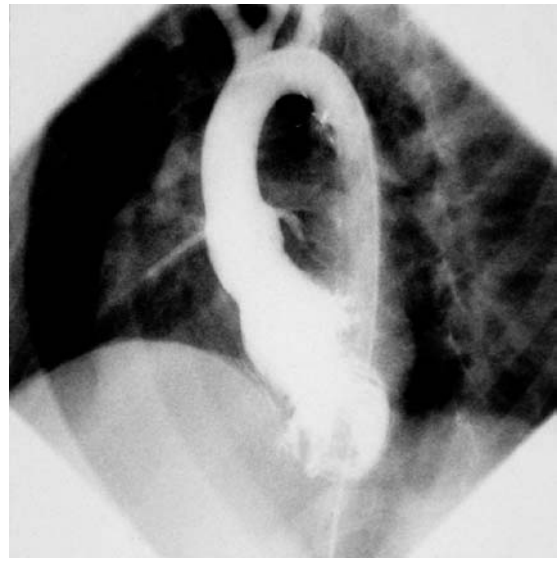

Figure 6.4

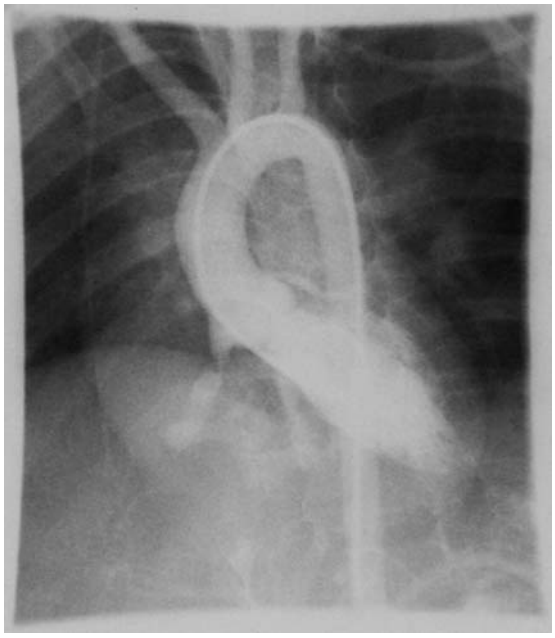

Figure 6.5

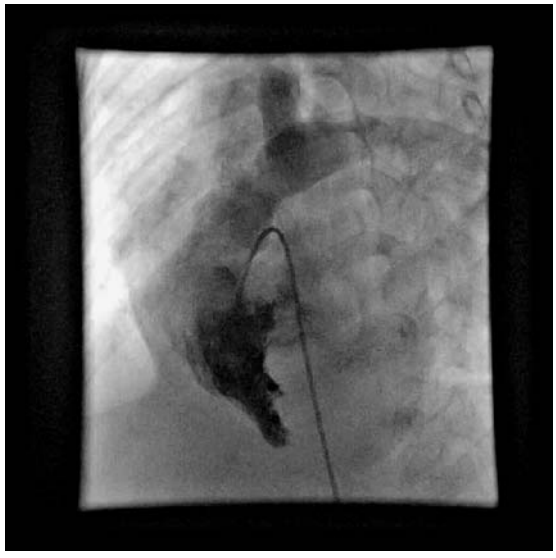

Figure 6.6

visualization of a specific lesion demands angulation tailored to the individual, and depends on the individual variation in position, size and hypertrophy or otherwise of component segments.

In reading an angiocardiogram, all the features displayed should be scrutinized, not just the feature of interest. An aberrant vessel or an unusual arrangement may not have been identified by other imaging techniques, and angiography is the only clinical technique that will display a tortuous vessel in continuity throughout its whole course. All the imaging modalities are capable of three-dimensional (3D) reconstruction, but these are expensive, not widely available at this date, and time consuming. Thus, angiocardiography is the single clinically applicable imaging modality that provides an image of cardiovascular structure covering three planes, albeit represented two-dimensionally.

Postoperative Investigation

Cardiac catheterization and angiocardiography have a significant role to play in early and late postoperative investigation, principally as the result of the degradation in the acoustic window which follows sternotomy. This limits both the spectral Doppler and imaging capabilities of echocardiography, particularly in the immediate (intensive care) phase of recovery. In complex lesions, accurate assessment of the magnitude of any residual shunt or pressure gradient and the contribution of a possible ventilation/perfusion mismatch will require detailed oximetric study, coupled with precise pressure measurement. Even a small residual lesion may reduce cardiac performance. The greater detail of anatomical information provided by angiocardiography as opposed to echocardiography in this period may outweigh the added difficulty in obtaining this information. As this group of patients is unstable haemodynamically, all the factors involved to maintain a steady state during the examination demand strict adherence (oxygenation, ventilation, acid–base balance, fluid balance, temperature control, inotropic support).

MRI is not affected by previous open access to cardiovascular structures and provides high-resolution images in multiple planes. As older children and adults will tolerate this procedure without general anaesthesia, it is valuable in later postoperative assessments. From the flow data, pressure relationships can be derived, although these do not always supplant the need for an absolute pressure measurement. Pressure changes derived from these studies as with those from Doppler-derived studies are not always adequately differentiated if the sites of obstruction are multiple, tortuous or tunnel-like. However, both these modalities can yield valuable performance and flow data from analysis of ventricular volume change in the normal resting state. The high-resolution images capable of 3D reconstruction, coupled with the increasingly sophisticated derived

haemodynamic parameters, make MRI the preferred investigation when cross-sectional and spectral Doppler echocardiography do not provide all the detail required for a full diagnostic preoperative diagnosis. This is more pertinent when reoperation is under consideration, as the problem of vascular access is avoided and the haemodynamic assessment is undertaken in a physiological, albeit resting, state. (i.e. it avoids sedation and anaesthesia). In the older child or adolescent, cardiovascular performance may be assessed at various levels of work by exercise testing (see below).

Risks of Invasive Investigation

The risks involved in invasive investigation revolve around two main factors. The first is the condition of the patient; the more ill, the more haemodynamically unstable, the greater the risk. The second is the mechanical complexity of the study undertaken. The stiffer and larger the diameter of sheaths, guide wires and catheters introduced, particularly into an artery, the more likely is there to be a problem. This will be a greater factor when an interventional procedure is undertaken, with the inherent reduced flexibility of the delivery systems and possibility of dislodgement.

General risks include:

1. Blood loss. If this exceeds 10% of the estimated volume from blood sampling or accidental loss, replacement with a blood substitute or blood should be considered.
2. Hypothermia becomes a problem in small infants, even if they are protected against heat loss, when the procedure exceeds 120 minutes.
3. Acidaemia is more frequently encountered after 90–120 minutes and is compounded by hypovolaemia and hypothermia.
4. Arrhythmias are not always predictable or avoidable but acidaemia and hypoxaemia are contributory. Many arrhythmic episodes are initiated mechanically; careful manipulation, particularly of the larger and stiffer catheters, etc. will reduce the likelihood.
5. Catheter displacement, particularly during angiocardiography, may induce an arrhythmia, intramyocardial extravasation, perforation and possible haemorrhagic pericardial effusion with tamponade. Cardiac chamber or vessel perforation during manipulation is associated more with stiffness, particularly of the tip, than with diameter.
6. All radio-opaque contrast media are toxic to some extent, and in excessive doses will reduce myocardial performance with a fall in cardiac output, and may induce both renal toxicity and changes in the coagulation properties of blood. Sensitivity reactions to the iodine content are rare. It is important to keep a strict record of all contrast volume injected as ''test doses'' as well as for the formal angiocardiograms, exercising caution when multiple injections are made.
7. The risks related to specific interventional procedures are discussed elsewhere.
8. Radiation dosage. With modern equipment this is not a relevant issue when only one or two diagnostic studies are undertaken in an individual patient. However, if multiply-staged surgical procedures are undertaken, each involving a detailed preoperative angiocardiographic assessment with multiple sites of injection (e.g. pulmonary atresia with multiple aortopulmonary communicating arteries), a significant radiological hazard can be attained over the child's growth period. A significant dose may also be achieved during prolonged interventional procedures and account must be taken of both anterior–posterior and lateral planes. A radiation record should be kept for all procedures; this should also include all the operators.

Certain circulatory states have an enhanced susceptibility to adverse reaction during invasive procedures:

1. States associated with pulmonary oedema and a low systemic blood flow.
2. Profound arterial hypoxaemia, whether caused by low pulmonary blood flow or additional severe pulmonary disease.
3. High pulmonary vascular resistance resulting from pulmonary vascular obstructive disease. Any fall in systemic resistance cannot be counteracted by a compensatory change in pulmonary blood flow characteristics, and a disastrous fall in cardiac output follows. The rate of adverse reaction to contrast media is high in this condition, due to the fall in systemic resistance induced, and angiocardiography may not be indicated when the pulmonary vascular resistance approaches or exceeds the systemic level.

A detailed account of all the risks involved in invasive investigations has been published elsewhere (Vitiello *et al*., 1998).

SUMMARY

Cardiac catheterization and associated angiocardiography is no longer the principal mode of investigation of heart disease. It is but one of a number of investigations into the form and function of the heart. It has its own particular strengths, but because of its invasive

nature carries an inherent risk. Thus, the importance of the specific data must be balanced with the risk of the procedure. The indications revolve around the need for precise haemodynamic measurements, particularly pressure, and visualization of structures either not at all or only poorly visualized by non-invasive imaging methods. It will uniquely provide an uninterrupted image of the complete course of a structure, however tortuous. It also provides the vehicle for testing pulmonary vascular function and interventional catheter procedures. To investigate the capability of the heart and circulation during exercise requires a distinctly different approach.

CARDIOPULMONARY EXERCISE TESTING IN CHILDREN AND ADOLESCENTS

In adult practice, exercise testing has largely focused on the evaluation of symptoms of ischaemic heart disease. Only a small subgroup of children have congenital or acquired reasons to suspect myocardial ischaemia (Kawasaki disease, cardiac surgery to the coronary arteries). Another subgroup of children may require investigation to assess inducible arrhythmia, including inherited arrhythmia syndromes such as Long QT and Brugada syndromes. There are numerous published protocols for exercise testing, dependent on the information required. Each laboratory should be accustomed to employing a variety of protocols and be aware of reasons for choice. For detection of inducible ischaemia, a rapid ramping maximal treadmill protocol, such as the Bruce protocol, is commonly employed. Arrhythmia provocation is best tested using the equivalent of shuttle runs—rapid escalation of exercise to maximum heart rate in a repetitive manner. The majority of children with congenital or acquired heart disease require investigation to determine their functional capacity as an estimate of their quality of life (Fleg *et al*., 2000).

The objective assessment of functional capacity has long been a ''Holy Grail''. A fundamental requirement for activities of daily living is the ability to perform aerobic exercise. An integrated approach for exercise testing (Wasserman, 1994) allows a unique opportunity to study simultaneously the cellular, respiratory and cardiovascular response to the metabolic stress of a controlled work load. The maximal capacity of an individual to perform physical work is defined by the maximal oxygen consumption ($VO_{2\,max}$), the product of cardiac output and arteriovenous oxygen difference (ΔaVO_2). This measurement is indexed to body weight or surface area to facilitate subject comparisons. Estimated oxygen consumption is sometimes defined in metabolic equivalents (MET), where one MET represents resting oxygen consumption (about 3.5 ml/kg); however, it is more appropriate to quote the absolute or indexed value for oxygen consumption.

In normal subjects, the maximal oxygen capacity is determined by delivery of oxygen to the exercising muscle by the cardiopulmonary system. This in turn is determined by the oxygen-carrying capacity of blood (haemoglobin), cardiac output (stroke volume and heart rate) and lung function. In patients with congenital or acquired heart disease, limitations of exercise capacity can be derived from any of these determinants. The use of maximal integrated cardiopulmonary exercise testing allows objective measurement of maximal exercise capacity and cardiopulmonary function. In this way, the evaluation of exercise intolerance allows differentiation into cardiac, respiratory, metabolic and motivational causes.

When maximal exercise testing is not possible or undesirable, cardiopulmonary exercise testing can still give useful data on submaximal function. Anaerobic threshold is a physiological event marking the change of aerobic to anaerobic respiration during exercise, and can be determined from respiratory gas exchange measurements. In adults, the anaerobic threshold has been found to be a good index of cardiopulmonary health, predicting all causes of surgical mortality (Older *et al*., 1999), but has not been extensively evaluated in children. Ventilatory equivalent (slope VE/VCO_2) correlates well with the severity of cardiac failure (Francis *et al*., 2000). Slope of oxygen uptake vs. exercise intensity may also be a useful submaximal index (Reybrouck *et al*., 2000).

Limitations

Although motor coordination and cooperation are important in the conduct of a successful exercise test in children, the major limitation is physical size. Specially designed cycle ergometers for smaller children exist or can be adapted from adult models; however, it is rare to be able to test a child less than 115 cm in height.

Potential Impact

Integrated cardiopulmonary exercise testing is a useful assessment of functional status, and allows disease states to be separated into causes derived from the respiratory, cardiovascular, metabolic or locomotor systems. The use of exercise testing allows a number of comparisons within congenital heart disease, e.g. the effects of a surgical intervention such as pulmonary valve replacement (Eyskens *et al*., 2000), catheter intervention such as atrial septal defect closure (Rhodes *et al*., 2002), drug treatment such as afterload reduction agents in Fontan circulation (Kouatli *et al*., 1997), and risk

stratification in patients with heart failure (Osada *et al*., 1998).

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7

Interventional Catheterization in Congenital Heart Defects

P. Bonhoeffer and Y. Boudjemline

Although the first attempts to treat congenital heart disease were surgical, several investigators have tried to develop trans-catheter techniques to replace surgical procedures. Numerous percutaneous techniques, viz. pulmonary and tricuspid valvotomy (Rubio-Alvarez *et al*., 1953), dilatation of atherosclerotic lesions (Dotter and Judkins, 1964), balloon atrial septostomy (Rashkind and Miller, 1966) and occlusion of patent ductus arteriosus (Porstmann *et al*., 1967), have been described before the more recent era of wider use of trans-catheter interventions. Interventional cardiology became a subspecialty with the report of percutaneous transluminal balloon angioplasty by Gruntzig and Hopff in 1974. Following animal experiments (Lock *et al*., 1981, 1982), the indications were subsequently extended to non-atherosclerotic stenotic valvar and vascular lesions in infants and children (Kan *et al*., 1982). To overcome problems of residual stenosis, secondary restenosis and undilatable vascular stenosis, balloon-expandable stents have been developed for intravascular use. After animal experiments (Mullins *et al*., 1988), such devices are now widely used with excellent results.

In parallel with these pioneering works, innovative approaches were being developed for non-surgical treatment of congenital cardiac defects. A number of devices have been described over the past 30 years for closure of patent ductus arteriosus (PDA), atrial septal defects (ASD) and ventricular septal defects (VSD) (King and Mills, 1974).

More recently, with the advances in stent technology and scaffolding, non-surgical valve replacement of the pulmonary valve has been performed enlarging the field of interventional cardiology (Bonhoeffer *et al*., 2000a,b). Currently a combined approach between the surgeon and the cardiac interventionist might improve patients' outcome in selected diseases.

INDICATIONS

Generally, indications for interventional catheterization can be divided into two major categories:

1. Opening of anatomical structures which are stenotic.
2. Closing of structures which are abnormally open.

For a number of lesions, treatment by interventional catheterization is now regarded as first-line treatment, surgery being discussed only if it fails. For some anomalies, both approaches are feasible and it becomes more difficult to decide from which approach the patient will benefit most. Certain new techniques, such as trans-catheter replacement of heart valves, Fontan completion and hybrid approaches, are still a field of clinical research and need further evaluation before they are fully introduced into clinical practice.

This chapter provides a comprehensive review of currently available interventional techniques, strategies and devices used in the management of congenital heart defects.

OPENING OF ABNORMALLY OR NORMALLY CLOSED STRUCTURES

Balloon Dilatation

Balloon dilatations are the most common procedures performed in a paediatric catheterization laboratory, involving valves, vessels, surgical pathways and intracardiac structures. Since the first balloon dilatation catheter, there has been a great improvement in materials, profiles and designs, allowing for balloon dilatation in small infants with low risk of injuries.

Valvuloplasty for Congenital Valvular Diseases

Pulmonic Stenosis

Since the first description of balloon dilatation of a valvar pulmonic stenosis by Kan *et al*. in 1982, this

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procedure is now considered to be first-line treatment. Pulmonary valve relief is usually indicated when the systolic gradient between the right ventricle and the pulmonary artery is higher than 50 mmHg. A routine right heart catheterization is first performed to assess the right ventricular pressure and right ventricular outflow tract anatomy. A right ventricular angiogram in lateral projection is then performed to visualize the doming valve leaflets and to locate the site of stenosis. Careful measurements of the annulus are then obtained. The diameter of the balloon catheter is chosen at 120–140% of the measured annulus diameter. For infants, young children, adolescents and adults, the length of the balloon catheter is 2 cm, 3 cm and 4 cm respectively. An end-hole catheter is advanced into a distal pulmonary artery and through it a 0.014–0.038 inch exchange wire is positioned. The chosen balloon catheter is then flushed using diluted contrast, inserted in the sheath, advanced over the guide wire to straddle the stenotic pulmonic valve and slowly inflated manually or using a hand deflator (**Figure 7.1**). The balloon catheter is inflated until the waist is obliterated. Thereafter, the balloon catheter is deflated and retrieved, leaving the wire in the distal pulmonary artery. If a waist is not visualized, the balloon catheter is repositioned more distally or proximally and the procedure is repeated until a waist is visualized. The pressure gradient is then remeasured, using an end-hole catheter or, more advantageously, a Multitrack catheter (NuMed Inc.) that allows precise assessment of the haemodynamic and angiographic results, leaving the wire in position. Dynamic infundibular stenosis is frequently seen after the procedure, making haemodynamic assessment difficult to distinguish from a residual valvar stenosis. Here again, the use of a Multitrack catheter is indicated to precisely locate the site of obstruction. The infundibular reaction usually disappears after a few hours spontaneously or after an oral beta-blocker administration. Right ventricular outflow tract angiogram is finally performed.

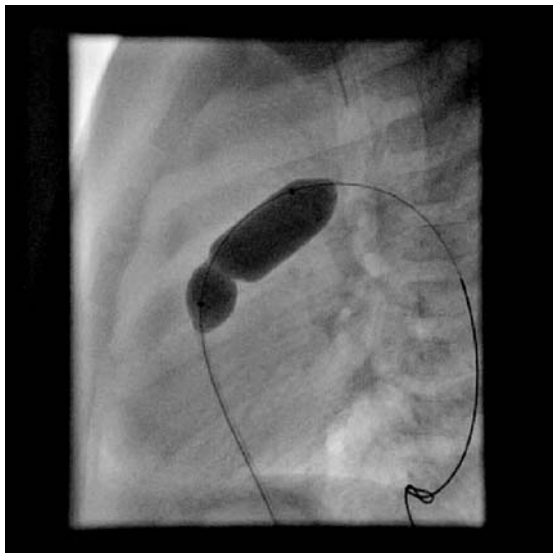

Figure 7.1

The efficacy of this technique in term of gradient relief has been well documented in a large number of series (McCrindle *et al*., 1994). Best results are obtained with the use of oversized balloon catheters. Patients with syndromic features, such as Noonan, usually exhibit poorer results (around 50% success) as compared with non-syndromic patients and might require surgical treatment to relief the obstruction. At follow-up, most patients usually have normal right ventricular pressure and no significant pulmonary regurgitation. A small number of patients (less than 10%) require a repeat balloon dilatation after a few years. The complication rate is low (0.4%,) and the mortality around 0.2% (Stanger *et al*., 1990).

Two particular groups should be discussed separately: newborns with critical pulmonary valve stenosis or pulmonary valve atresia with intact septum, and cyanotic patients with complex defects and pulmonary stenosis. Successful balloon dilatation has been reported in both groups.

Newborns with critical pulmonary stenosis or atresia are usually treated with continuous prostaglandin infusion to maintain the patency of the ductus arteriosus. A few days after birth, neonates with good-sized right ventricles are catheterized in order to enlarge or re-establish the continuity between the right ventricle and pulmonary artery. An end-hole catheter is placed in the infundibulum and a hand injection is obtained to look for an anterograde opacification of the pulmonary artery. In pulmonary atresia, an imperforate infundibulum is seen. To perforate the atretic valve, various techniques have been used since its first description (Latson *et al*., 1991), viz. stiff wires, laser and more recently radiofrequency. After the perforation, or directly in critical pulmonic valve stenosis where there is a tiny passage, a coronary wire is positioned in the distal pulmonary artery or in the descending aorta through the patent ductus arteriosus. In patients with atresia, careful assessment using various fluoroscopic projections is needed to ascertain that the wire is in the pulmonary artery or the descending aorta, and not in the pericardial cavity, before the balloon is advanced over it and inflated. A predilatation with a coronary balloon catheter is usually required in that population before final opening of the valve with a larger balloon catheter. In a full-term baby, the balloon catheter usually does not exceed 8 mm in diameter. Similarly in patients with pulmonary stenosis, the success rate is high

(85%) and the complication rate low (15%; Agnoletti *et al*., 2003). After the procedure, the prostaglandin infusion is discontinued after a few days or even weeks to allow the right ventricular compliance to recover. In some patients, a surgical shunt needs to be added later if oxygen saturation remains low.

The complex cyanotic patient with pulmonary stenosis and ventricular septal defect can benefit from a balloon dilatation. This is an unusual indication in difficult clinical situations. The procedure is performed to improve cyanosis by increasing pulmonary blood flow. Because full opening of the valve would lead to pulmonary hypertension, special attention should be given when choosing the diameter of the balloon catheter, to avoid exaggerated opening of the outflow tract that would expose the pulmonary artery bed to high pressures.

Aortic Valve Stenosis

Balloon dilatation valvuloplasty for congenital aortic valve stenosis was developed in the mid-1980s (Lababidi *et al*., 1984), and experience has been gained in most large centres. Dilatation is indicated in all patients with Doppler peak gradient greater than 70 mmHg, in patients with left ventricular strain on the ECG and peak gradient greater than 60 mmHg, and regardless of the gradient in patients with syncope, low cardiac output, severe left ventricular dysfunction or duct-dependent systemic circulation, as frequently seen in newborn babies. A femoral arterial sheath is placed. An aortography, using a pigtail catheter, is performed to measure the aortic annulus and to look for an aortic regurgitation. The left ventricle is usually entered using a retrograde approach. Pressures are measured either simultaneously, through the femoral sheath and the catheter positioned in the left ventricle, or after a pullback from the left ventricle into the ascending aorta. The use of a Multitrack catheter is useful in this particular setting, since there is no need to re-cross the aortic valve after the pullback. In bigger children, the catheter is exchanged for an 0.035 or 0.038 inch Amplatzer stiff wire with a broad hand-formed loop, in such a way that the wire loop is in the apex of the left ventricle away from the mitral valve. A balloon catheter is selected. The diameter of the balloon catheter is chosen at 90–100% of the measured annulus diameter, since the use of bigger balloons has been associated with the occurrence of a high incidence of aortic insufficiency. The balloon is flushed with diluted contrast, advanced over the wire in the area of the aortic annulus, inflated to its nominal diameter, and rapidly deflated after the disappearance of the waist that is usually seen during the balloon inflation. Postdilatation assessment (aortogram and haemodynamic measurements) is then performed. If gradient relief is inadequate, the dilatation frames are reviewed in order to measure ''real'' balloon diameter. A larger balloon may be used if the ratio of dilating balloon diameter to aortic valve annulus is less than 1.1. In neonates, alternative approaches, such as umbilical artery, carotid artery or femoral vein, have been used in order to limit the risk of secondary artery occlusion.

In comparison with pulmonic stenosis, aortic valvuloplasty raises additional issues:

1. Ventricular systole tends to eject the balloon catheter during inflation, making balloon position difficult to maintain.
2. The valve could be damaged after valvuloplasty, leading to significant aortic valve regurgitation. These two issues can be intricate, since the instability of the balloon during inflation can alter the aortic valve integrity, leading to tearing of the valve. Various techniques have been developed during the past few years to reduce this danger. Anterograde approach through a trans-septal puncture, right ventricular pacing or adenosine injection prior to balloon inflation have also been used to obtain a more stable position (**Figure 7.2**) Balloon dilatation has been applied to a wide range of patients, from neonates to adults, with various results. The technique, however, decreases the degree of aortic valve obstruction from severe to mild in a large majority of patients. Long-term follow-up studies of balloon dilatation valvuloplasty show that excellent relief of obstruction persists for several years. In a small number of the patients the balloon procedure will create severe aortic valve insufficiency.

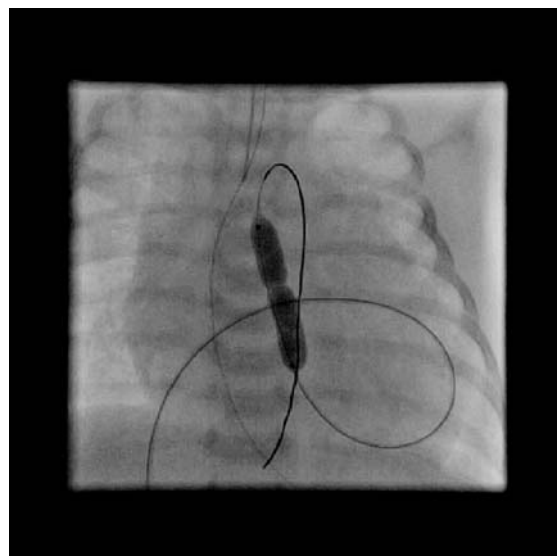

Figure 7.2

Like surgical valvotomy in the past (15–40% of patients require re-intervention 5–10 years after the initial surgical valvotomy), recurrent valve obstruction does occur in many children during the next 5–10 years, perhaps related to patients' growth and chronic valve changes. Many of these children will therefore require repeat balloon dilatation or surgical valve replacement procedures. Reich *et al*. (2004) and Balmer *et al*. (2004) reported their experience with balloon dilatation for valvar aortic stenosis in a combined series of 339 consecutive children of all ages. In both studies, early results were excellent with a significant decrease in pressure gradient from 78 ± 27.4 to 27.6 ± 16.2 mmHg ($p < 0.001$); and in systolic left ventricular pressure from 162.3 ± 37.1 to 124.3 ± 33.0 mmHg ($p < 0.001$). Similar changes were observed on Doppler measurements. The grade of aortic insufficiency increased from 0 to 1.0 ($p < 0.001$). For all age groups, the calculated mean actuarial probability of re-intervention-free survival at 14.4 years was 0.39 ± 0.07. This probability was worse in the neonatal subgroup, with 0.26 ± 0.08 free of re-intervention at 14.4 years. In patients less than 3 months, independent predictors for restenosis were young age and a small z score of the aortic annulus. Aortic regurgitation does progress. At the latest follow-up, the grade of aortic insufficiency increased from 1.0 to 2.0 (range 1.0–2.5) in Reich's series. In Balmer's study, 90% and 39% of patients had moderate to severe aortic incompetence after 1 month and 3 years, respectively. Independent predictors of aortic insufficiency were functionally bicuspid valve and a large z score of the aortic annulus. The causes for re-interventions by order of frequency were restenosis, significant regurgitation and restenosis with significant regurgitation. Procedure-related death occurred in 4.8%. Cardiac-related late mortality was 4.5%. Independent risk factors for death were a small z score of the aortic annulus, impairment of the left ventricular shortening fraction, and early date of procedure. The vascular access complication rate was 1.5–57%, with a higher incidence in the neonatal group. The great majority of these complications were transient and disappeared after fibrinolysis and/or heparin therapy.

To conclude, balloon valvuloplasty of congenital aortic stenosis has gradually become the treatment of choice. It is, however, a palliative procedure and re-intervention remains frequent. A mixture of restenosis associated or not with significant regurgitation are the indications for further procedures (surgery or balloon valvuloplasty). Comparable results in terms of immediate gradient relief, procedural mortality and longer-term survival are reported with balloon valvuloplasty and surgical valvotomy. Unfortunately, there are no randomized trials to allow for an accurate comparison of the two techniques. Most units favour balloon aortic valvuloplasty because of its avoidance of (a) a median sternotomy in patients who are likely to require further intervention and (b) cardiopulmonary bypass.

In neonates with critical aortic stenosis and left ventricular failure, trans-catheter dilatation is more challenging. The decision-making process between a single- vs. two-ventricle repair strategy is often complex, echocardiographic features of the left ventricle (size and aspect) being frequently impaired.

Angioplasty

Experiments establishing the basis for balloon angioplasty were performed in the early 1980s (Lock *et al*., 1981, 1982). In those experiments, the balloon dilatation of experimental pulmonary artery stenoses was responsible for medial and intimal tearing, leading to the increase of vessel size, which reflects the efficacy of the procedure. Since then, balloon catheters have been used to dilate various vascular lesions, either congenital or postoperative.

Coarctation of the Aorta

Balloon dilatation of aortic coarctation was first reported in 1982 (Singer *et al*., 1982). This technique is now widely accepted for the treatment of recoarctation but is more controversial for native coarctation. A sheath is first placed in the femoral artery. A catheter is then advanced across the coarctation site into the right subclavian artery or the ascending aorta and replaced by an exchange wire. After a pressure pullback, an angiogram is performed to locate and demonstrate the extension of the narrowing and its relationship with aortic branches. The diameter of the coarctation site as well as of the normal surrounding aorta is measured. The diameter of the balloon is no more than slightly longer than the diameter of the normal surrounding aorta. The chosen balloon catheter is advanced over the wire across the coarctation site and slowly inflated. The balloon is then deflated and removed. Because it is possible to perforate freshly dilated vessels when re-advancing wires or catheter through the dilatation site, pressure and angiographic controls are more safely performed using the Multitrack catheter over the previously placed wire.

The results of this procedure are good in terms of immediate gradient relief as well as long-term follow-up, with low incidence of complications and recoarctation. Complications after angioplasty for recoarctation, such as pulse loss, aortic disruption, aneurysm formation and re-intervention, are quite low. Those complications are much more frequent after ballooning of native coarctations (Rao *et al*., 1996) especially in neonates and infants. The aorta in postoperative coarctation is encased in scar tissue, which makes it less susceptible to disruption after ballooning. In Rao's report, during

a 6.5 year period, 51 neonates and infants (less than 3 months) with native coarctation underwent balloon angioplasty. Effective palliation was achieved in 92%. Surgical relief of aortic obstruction was required in four infants (8%) within the first month following the angioplasty. During intermediate-term follow-up, 22 infants (50%) developed recoarctation, requiring repeat balloon or surgical intervention 2–10 months (median, 3 months) after initial angioplasty. Decreased femoral pulses with cool lower extremity on the side used for angioplasty have been reported to be as high as 27% in that group of patients. However, with anticoagulant or thrombolytic therapy, permanent pulse loss or decrease has been rare (14%; Rao *et al*., 1996). The reported incidence of aneurysm varies (0–40%), depending on age group and investigators (Cooper *et al*., 1987). In a comparative group of children older than 3 months, Walhout *et al*. (2004) have reported similar outcomes in patients undergoing surgery or balloon angioplasty for native coarctation of the aorta; no statistical difference between surgery and angioplasty with respect to resultant pressure gradient decreases was found. There was no mortality or aneurysm formation. No statistical difference in freedom from re-intervention probabilities between the two groups of patients was noticed. Recoarctation occurred in one surgical patient (5.6%) and in two non-surgical patients (7%) (p = NS).

In summary, balloon dilatation is the treatment of choice for postoperative aortic coarctation or recoarctation. For native coarctation, balloon angioplasty is still controversial. Because of the high incidence of recoarctation and aneurysm formation, we do not use balloon dilatation in native neonatal coarctation.

Pulmonary Artery Stenosis

Surgical repair of proximal pulmonary branch stenosis is possible but has a high risk of restenosis, leading to re-interventions. The distal lesions are difficult to access surgically, making trans-catheter approach the technique of choice for treatment of this type of lesion. Indications for treatment are not clearly defined, but when some are treating aggressively every single pulmonary stenosis with the idea of recruiting vessels for lung growth, others have advocated treating only pulmonary artery stenosis with clinical manifestation or significant elevation of RV pressure. Everybody agrees to treat patients with univentricular circulation and cavopulmonary connection more aggressively. The technique is very similar to that described above. A right heart catheterization is performed. Haemodynamic assessment with pullback gradients in various pulmonary segments is carried out. Selective angiograms in multiple projections are then performed in order to locate, number and size the stenoses. An exchange wire is placed distally, its size depending on the balloon selected for the dilatation. The selected balloon is advanced over the wire and inflated with diluted contrast. When multiple stenoses are present, the most distal obstruction is dilated first, to avoid passing a wire through freshly dilated vessels. Angiography and haemodynamic assessment are repeated after each dilatation to assess the results and look for complications. Contrary to dilatation of proximal lesions, in distal lesions the effectiveness of the procedure cannot be judged on right ventricular pressure. Various criteria have been used to define success more adequately:

1. Increase of 50% or more in vessel size.
2. A lung perfusion scan showing a significant increase in flow to the affected lung segment.
3. A decrease in the pressure gradient across the stenosis.

The valvuloplasty and angioplasty registry reported the outcome from about 180 balloon angioplasties (Kan *et al*., 1990). Vessel diameter increased significantly from 5 ± 2 to 7 ± 3 mm ($p < 0.001$), with better results if the balloon diameter was > 3 times the diameter of the stenosis. The mean peak systolic pressure gradient across the stenosis decreased from 49 ± 25 to 37 ± 26 mmHg ($p < 0.001$) and the pressure proximal to the stenoses from 69 ± 25 to 63 ± 24 mmHg ($p < 0.001$). Complications were relatively frequent, occurring in 13% of patients. Vessel ruptures were encountered in five patients, with two deaths. Three more deaths occurred following one cardiac arrest, paradoxical embolism and low cardiac output. Various other studies have stressed the low rate of success, with nearly 50% of lesions considered as undilatable and 20–35% of recurrent stenoses (Lock *et al*., 1983).

No predictive factors were identified. Thus, predilatation parameters, technical aspects, nature of the lesions or immediate results of balloon dilatation were not predictive of restenosis. The use of high-pressure balloons (17–20 atm) has improved the success rate of this procedure. Gentles *et al*. (1993) applied this technique to 72 vessels in 52 patients. Of 36 vessels with unsuccessful low-pressure balloon dilatation, 63% were successfully treated with high-pressure balloons. Of the 36 remaining lesions, 81% had successful primary dilatation with a high-pressure balloon. Aneurysm occurred in 4.2%, which is similar to what is reported for a standard balloon; 13% had complications, including two vessel ruptures (one died). Despite the improvement in success rate, restenosis is still frequent. Studies are currently being undertaken to assess the efficacy of *cutting* balloons in relieving stenosis resistant to high- and low-pressure balloons and in decreasing the rate of secondary restenosis. These balloons have the advantage of creating controlled cuts on the stenosed vessels,

rather than uncontrolled tears. Rhodes *et al*., (2002) showed interesting early and mid-term results using these balloons. Longer follow-up studies are currently lacking.

Other Vascular Narrowing (Pulmonary Veins, Baffles, Surgical Pathway)

Balloon angioplasty has been applied to a variety of other congenital and postoperative vascular narrowings, including surgical pathways, bioprosthetic valves and pulmonary vein stenoses (Lock *et al*., 1984). The latter has been disappointing, giving only temporary results with rapid recurrence of haemodynamic impairments. Dilatation of postoperative baffle obstruction or bioprosthetic valve stenoses is, in contrast, more efficient, postponing the need for surgical intervention. In such conditions, the use of high-pressure balloons may be helpful to relieve calcified stenoses.

Atrial Septum

Excluding the transposition of the great arteries, where an atrioseptostomy, as first described by Rashkind and Miller in 1966, can benefit the patient, there are other situations in which an atrial communication is advantageous. Such situations are pulmonary hypertension with right heart failure and low cardiac output, failing Fontan circulation, or hypoplastic left heart with intact atrial septum or restrictive ASD. In such circumstances, the creation or the enlargement of an ASD has the advantage of improving clinical outcomes by decreasing the systemic venous or capillary pressure and congestion, and by increasing cardiac output. This can be achieved by different techniques. If the ASD is already present but restrictive, the hole can be progressively dilated using multiple standard static balloon catheters, increasing in size until the desired clinical result is obtained. However, sufficient enlargement can be difficult to obtain, particularly when the atrial septum is thick. Moreover, spontaneous closure of such an enlargement is usually observed after a few days or weeks. Redilatation is more difficult because of the fibrotic scar secondary to the first dilatation. In such cases, the use of cutting balloons can be helpful. If the atrial septum is intact, a hole can be created using a trans-septal needle, a blade or radiofrequency. It is then progressively dilated, as previously described (Thanopoulos *et al*., 1996).

Endovascular Stents

General Considerations

Successful balloon angioplasty of obstructive lesions associated with congenital heart diseases has been repeatedly reported. However, early failure may occur due to: (a) lesions that are too rigid to be dilated; (b) elastic recoil or external compression of the vessel wall; or (c) thrombosis or dissection of the vessel. Late failures have also been described with a large number of secondary restenoses (40%; Rothman *et al*., 1990). The development of stent technology has been one of the most exciting advances in the field of interventional cardiology. Although the concept was first introduced by Dotter and colleagues in 1969, first trials were carried out in late 1980s. These were done in adults with peripheral vascular and coronary artery obstructions. These studies have suggested that such devices may play an important role to deal with the failed balloon dilatation. In 1988, Mullins *et al*. reported the experimental results of implanting a Palmaz stent (Cordis, Warren, NJ) into pulmonary arteries and systemic veins in a canine model. Following this study, this stent was used to treat all vascular obstructions associated with congenital heart disease. These stents, however, have several limitations that have led to the development of other stents. The stent was designed for peripheral obstructions and not for congenital heart defects. It was rigid, inflexible and sharp-edged, making its delivery difficult in tortuous vessels and having the tendency to rupture the balloon during inflation and to traumatize the vessel after its expansion. Over-dilatation of the stent is limited because its strut design leads to a significant shortening. Several newer stent designs are currently available. These new stents are more flexible, making implantation in tortuous vessels more practical. Advantageously, Genesis stents are available premounted, allowing for the delivery without the use of a Mullins sheath. Self-expandable stents initially used for adults with iliac and femoral arterial stenosis have been introduced more recently and employed in the treatment of peripheral pulmonary stenosis and coarctation of the aorta. They require a shorter sheath for delivery and avoid the risk of balloon rupture with incomplete stent expansion seen occasionally with balloon expandable stents.

The technique for stent implantation is quite similar for all types of obstructive lesions. The procedure is usually performed under general anaesthesia to avoid adverse patient movement during stent inflation. Routine arterial and/or venous access is obtained. Anticoagulation is then given (50–100 IU heparin/kg body weight). A diagnostic cardiac catheterization with haemodynamic measurements and angiographic assessment in multiple projections is then performed in order to precisely locate the lesion to treat. Measurements of the vessel are performed at the site of the stenosis (diameter and length) and in the immediate adjacent areas to the stenosis, using inherent calibration systems if available

or a cardiomarker catheter as a calibration marker. The stent and balloon are then selected according to these measurements, taking care not to overstress the normal vessels. In certain circumstances, the diameter of stent dilatation may be deliberately chosen to be less than the diameter of the surrounding vessels, to limit the risk of rupturing the vessel. A stiff guide wire is positioned across the stenosis, with its soft end placed as distally as possible to obtain a stable position. A Mullins sheath 1–2 Fr larger than the diameter required for the balloon alone is then advanced over the wire across the stenosis. The dilator is retrieved with care, keeping the sheath and the wire position. The stent crimped over the selected balloon is then advanced within the sheath over the wire to the level of the stenosis. Once the sheath is withdrawn to expose the stent to the stenosis, angiography through the Mullins sheath is performed to confirm the position of the stent before balloon inflation. When an appropriate position is confirmed, the balloon is inflated, expanding the stent. A balloon in balloon catheter (BIB) can be advantageously used to limit the risk of stent flaring and reduce the risk of stent misplacement.

Haemodynamic and angiographic assessments are repeated following stent placement to (a) assess the luminal diameter of the stented vessel, the pressure gradient across the stent and the RV to aortic pressure ratio in case of PA stenting and (b) look for complications such as vessel rupture, intimal tears, dissection and occlusion of branch vessel. All sheaths and catheter are finally retrieved and haemostasis is achieved by manual compression. Antibiotic prophylaxis is given and the patients are generally discharged, the day after the procedure, on aspirin for 6 months.

Stent Implant for Peripheral Pulmonary Stenoses

Shaffer *et al*. reported in 1998 the results of prospectively collected data of intravascular stent placement in 152 patients with congenital and postoperative pulmonary artery stenoses (**Figures 7.3** and **7.4**.). The immediate results were encouraging, with a significant decrease in pressure gradients in both groups, from 46 ± 25 to 10 ± 13 mmHg in postoperative PA stenoses ($p < 0.001$); and from 71 ± 45 to 15 ± 21 mmHg in congenital PA stenoses ($p < 0.001$). Perfusion to a single affected lung increased significantly in both groups. At recatheterization 14 months after implantation, right ventricular pressure indexed to femoral artery pressure ratio increased in both groups, but remained low at 0.45 ± 0.01 in postoperative PA stenoses and increased from 0.55 ± 0.35 to 0.65 ± 0.3 in congenital PA stenoses. Repeat angioplasty of residual stent stenoses was safe and effective, allowing for further reduction of right ventricular pressure indexed to femoral artery pressure ratio. Complications included four stent migrations, with surgical removal in two and expansion in a benign position in the remaining four. Three patients had stent thrombosis and limited haemoptysis developed in four. One patient had an aneurysm of the stented area after redilatation. Two deaths occurred: one following a vessel rupture and the other secondary to pulmonary oedema with ventilation–perfusion mismatch. Neointimal proliferation occurs to some degree

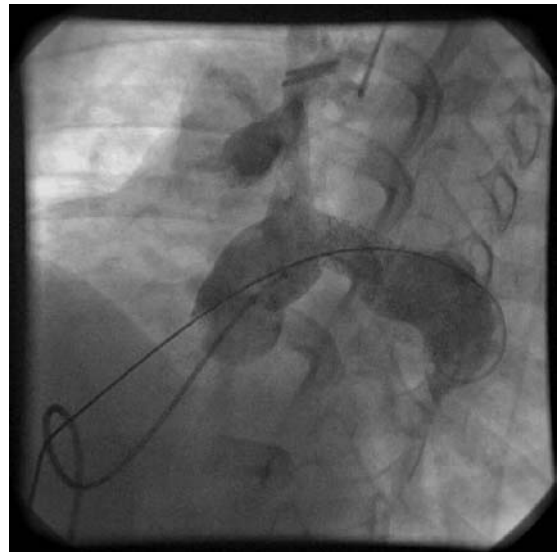

Figure 7.3

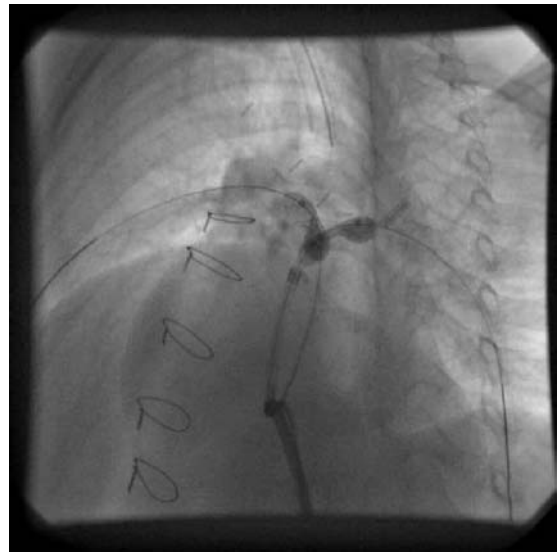

Figure 7.4

in all patients, with 1–2 mm decrease in the diameter of the lumen. Significant restenosis occurred in only three patients (2%) in this series.

Stent Implant for Right Ventricle-to-Pulmonary Artery Conduit Stenosis

RV–PA conduits have been widely used to establish the continuity between the RV and the PA. The lifespan of these conduits is, however, limited by the development of progressive but inevitable obstruction. Fibrotic intimal peel formation, calcification, kinking, external compression and inability to grow with the patient are all thought to contribute to the development of stenosis (Stark *et al*., 1998).

Surgical replacement of obstructed conduits is associated with increased morbidity, due to repeat sternotomy in patients who might already have had several previous operations and cardiotomies. Balloon angioplasty of those lesions had poor efficacy in delaying surgery. Balloon-expandable stents have been used to relieve conduit obstruction and delay surgical replacement.

Powell *et al*. (1995) reported a series of 44 patients with intermediate follow-up (1–48 months). Stent implantation resulted in significant immediate haemodynamic and angiographic improvement. The mean RV–PA systolic gradient fell from 61.0 ± 16.9 to 29.7 ± 11.9 mmHg ($p < 0.001$) and the RV:systemic pressure ratio from 0.92 ± 0.17 to 0.63 ± 0.20 ($p < 0.001$). During the follow-up period, 23 patients had re-interventions (redilatation in two, additional conduit stenting in seven and surgical conduit replacement in 14). Actuarial freedom from conduit re-operation was 65%, 30 months following stent placement. In a multivariate analysis, the strongest predictors of early surgical replacement were a greater postprocedure RV:aortic pressure ratio and a narrower post-stent diameter.

Stent Implant for Coarctation of the Aorta

Since the first series of Suarez de Lezo *et al*. in 1995, stents have been increasingly used to effectively treat various degrees (from mild to severe) of native aortic coarctation and re-coarctation. The use of stents in preference to simple balloon angioplasty is now accepted, since stents provide greater vessel support, maintaining the increased diameter, and limiting vessel recoil, restenosis and acute vessel rupture or aneurysmal formation (**Figure 7.5**). The stent diameter is usually chosen to equal the size of the descending aorta at the level of the diaphragm. However, depending on the nature and diameter of the coarctation site, a smaller balloon (12–14 mm) may be chosen to limit the risk of disruption. After 6 months, the stent can be fully expanded to its final diameter. As previously mentioned, the use of a BIB catheter is of value for precise stent placement and limitation of stent migration. Several teams reported very encouraging early and mid-term results (Suarez de Lezo *et al*., 1999).

Figure 7.5

Pressure gradient across the coarctation site is usually significantly decreased to less than 10 mmHg. This drop persists during the follow-up, with a low incidence of restenosis in patients older than 3 years of age at the time of stent implantation. In infants, the incidence of restenosis is greater (Suarez de Lezo *et al*., 1999). Similarly, the rate of re-stenosis is inversely related to aortic diameters before and after stent implantation and directly related to length of the lesion and the growth of the patient since the implantation. Complications, including stent migration, stent fracture, thromboembolic events and death, have been described. Complications related to vascular access were 5–19% (aneurysms in 0–7% of patients).

Stent implantation has become a worldwide practice to treat obstructive lesions of the aortic isthmus. From a haemodynamic and an anatomical point of view, the results, as shown previously, are impressive. However, these results appear to be disappointing when looking at persistent systemic hypertension, which has been reported at 0–28%, with a great proportion of patients being adults who presented with hypertension before stent placement. This is most probably related to the underlying damage to the vascular tree. Moreover, trans-catheter treatment exchanges an anatomical abnormality

for a ''stiff'' stent, which is unlikely to correct the abnormality of reflecting pressure waves. Long-term studies are obviously needed to assess the cardiovascular effect of the increase of the aortic wall rigidity, in particular in terms of affecting pulse pressure. Future studies will need to look at the management of coarctation.

Stent Implanted to Relieve Obstructions to Other Vessels

Stent implantation has been applied to a variety of other congenital and postoperative vascular narrowings, including surgical pathways (Abdulhamed *et al*., 1996) and systemic vein and pulmonary vein stenosis (Tomita *et al*., 2003), but also to natural vessels or heart lesions such as PDA and ASD, in order to maintain their patency (Gewillig *et al*., 2002).

As mentioned for balloon angioplasty, stent implantation in pulmonary vein stenosis has been disappointing, giving temporary results with rapid restenosis and recurrence of haemodynamic impairments secondary to severe intimal proliferation. General and local (using drug-eluding stents) immunosuppressive therapies are under investigation in some centres to try to improve the outcome of this terrible disease. Stent implantation of postoperative pathway obstructions, e.g. Blalock shunts, atrial baffles and Fontans, may in contrast be more efficient in postponing the need for reoperation.

Limitations and General Conclusions on Stent Technology

Despite encouraging results reported above, some concerns should be emphasized regarding the use of stent technology. First, endovascular stent implantation for pulmonary artery stenoses requires the use of a long and large vascular sheath to insure precise implantation and avoid embolization or malposition of the stent. A long vascular sheath may be difficult to position and usage may be associated with vascular damage and/or haemodynamic disturbance, especially in infants and small children. A large sheath requires a large vascular access to deliver the stent. That size may be too large to allow its use in small children. The possibility of redilating stents to follow children's growth is currently limited. For all these reasons, experience of stent placement is limited in that particular population. The recent availability of premounted stents (Palmaz, Corinthian and Genesis stents) that do not require the use of a Mullins sheath has permitted stent implantation in small children using a 6–7 Fr short sheath (Pass *et al*., 2002). However, because redilatation capability is limited, these stents cannot accommodate somatic growth until adulthood. They should therefore not be placed as a definitive procedure for peripheral lesions (< 12 mm in diameter) or elsewhere when patients are expected to undergo future open heart surgery. Research on stent design, biodegradable or breakable stents will, in future, hopefully permit stenting obstructive lesions in small children with the possibility of redilating the stent sequentially to accommodate somatic growth (Su *et al*., 2003).

Second, prospective and collaborative studies will be necessary to precisely delineate the place of each stent in the whole spectrum of lesions. It is imaginable that we will need different stents for various lesions: rigid stents with high radial strength for severely calcified lesions, and flexible stents for peripheral obstruction.

Third, the question of the superiority of direct stenting vs. stent implantation following balloon dilatation remains unanswered. Similarly, the place of covered stent should be clarified. Despite the need for a larger sheath, its use in native lesions has been advocated because of the increased risk of vascular rupture after stent placement. But the demonstration of efficacy, safety and superiority has not been made. Moreover, stent embolization is more dangerous, since the covering can occlude critical branches in proximity of the stented vessel. Finally, long-term data are currently missing.

Fourth, the risk of restenosis is of concern. It has been reported to be low (around 2–5%) in a number of series but some recent series have shown a percentage of restenosis as high as 28% (Ing *et al*., 1995). This discrepancy between studies might be related to different definitions used to qualify restenosis, but might also largely vary with types of vessels (location, native/postoperative and diameter) and the stents used. The progression of initial vascular trauma might explain the severity of intimal proliferation seen in some patients. Thus, it would be preferable to perform staged serial dilatation of stents, rather than attempting to gain maximum vessel diameter at initial stent implant. The development of drug-eluting stents may limit the intimal proliferation.

CLOSURE OF ABNORMALLY OPEN STRUCTURES

Patent Ductus Arteriosus (PDA)

In 1967, Porstmann *et al*. reported the use of an Ivalon plug to close PDA. Since then, several devices have been used, including the Rashkind PDA occluder (Rashkind *et al*., 1987), Gianturco or duct-occluder coils, detachable coils, Gianturco–Grifka sac, buttoned device, CardioSEAL and other umbrella devices and the Amplatzer PDA occluder. Using a technique described by Cambier *et al*. in 1992, the great majority of PDAs of small sizes are closed using vascular coils, which are usually delivered via arterial catheters of small sizes

Figure 7.6

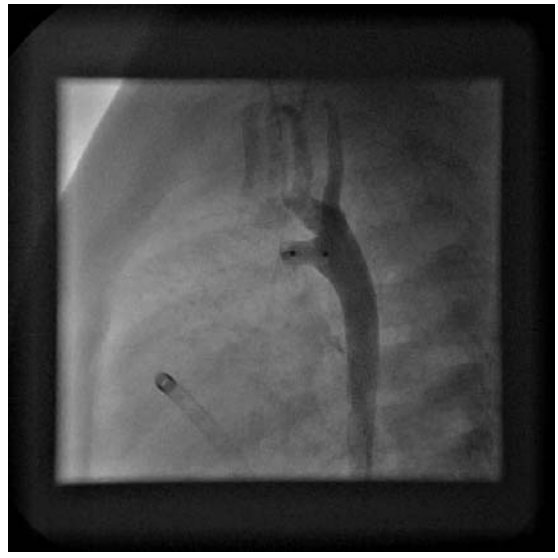

Figure 7.7

(4–5 Fr). A spiral of the coil is usually delivered in the pulmonary artery and the remainder of the spirals are positioned in the aortic ampulla (**Figure 7.6**), inducing an aggregation of platelets, an obstruction of blood flow and eventually endothelialization of the coil. Closure rate at 6 months is around 95% (Podnar *et al*., 1997).

The incidence of the complications (embolizations, residual shunt, haemolysis, protrusions into the left pulmonary artery with or without stenosis) is low (close to 5%). The incidence of the complications is more frequent when the PDA is large. Although closure of PDA of more than 4 mm in diameter is possible by inserting several coils simultaneously, the use of a plug seems preferable (**Figure 7.7**). The Amplatzer Duct Occluder is self-expandable, self-centering and retrievable, thanks to a screw attached to the body of the device. Its great flexibility allows its insertion in catheters of small sizes (5–7 Fr). It is advanced from the femoral vein into the descending aorta through the PDA before being released. Attached to a delivery cable via the screw, it is easily reintroduced into the sheath and repositioned if necessary. In the shape of a champagne cork, with its top in the area of the ampulla, it can close PDA up to 14 mm and, in most cases, perfectly matches the shape of the PDA, giving a rate of occlusion close to 100% up to 6 months (Waight *et al*., 2001). However, the use of such prostheses remains delicate in children weighing less than 6 kg because of the size of the introducer required and the risk of obstruction of aortic and/or pulmonary artery flows (Fisher *et al*., 2001).

The data comparing devices is not available. However, implantation feasibility is high for most devices (91–100%; Masura *et al*., 1998).

Device dislodgement varies (0–9%). Most of these embolized devices can be retrieved using specially designed trans-catheter equipment (snares, biotomes, etc.). However, in some instances surgical retrieval is necessary. Careful selection of the device according to the size and shape of the PDA may help to reduce the rate of embolization. The prevalence of residual shunting varies greatly (0–42%) with the type of the device used and the timing of evaluation within 24 hours of implantation to 0–16% during the follow-up. Cost is an important factor, particularly in developing countries. Coils cost about $50 US, whereas devices cost 10–20 times more.

Atrial Septal Defect

Since the first description by King *et al*. in 1974, catheter closure of secundum atrial septal defect (ASD) has been performed using various devices. The most-used devices are Amplatzer Septal Occluder (ASO) (**Figure 7.8**.), Clamshell (CardioSeal©) (Carminati *et al*., 2001), and the Helex (**Figures 7.9** and **7.10**). (Gore and associates©), which have been introduced more recently (Zahn *et al*., 2001). Amplatzer Septal Occluder, the most commonly used device since 1996 and the only device to have FDA approval, has a 4 mm long connection between the two disks, allowing the centring of the prosthesis on the ASD without requiring the ''oversizing'' of the

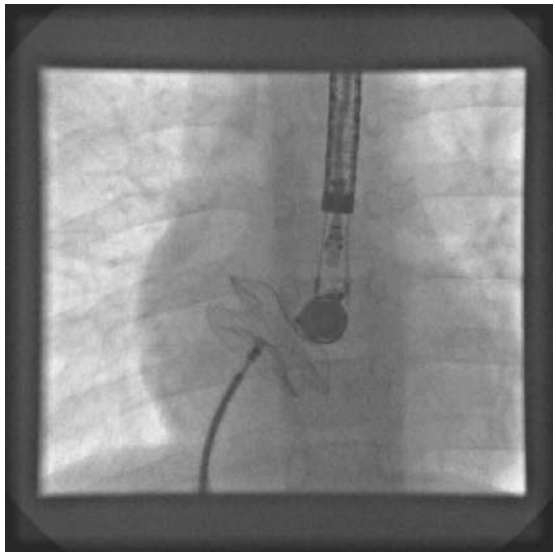

Figure 7.8

Figure 7.10

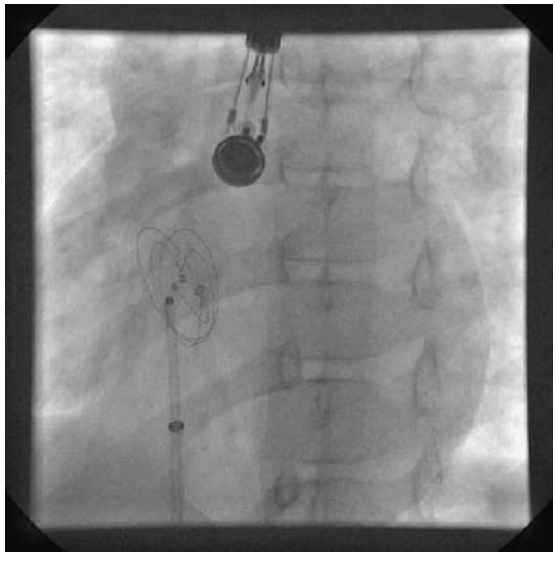

Figure 7.9

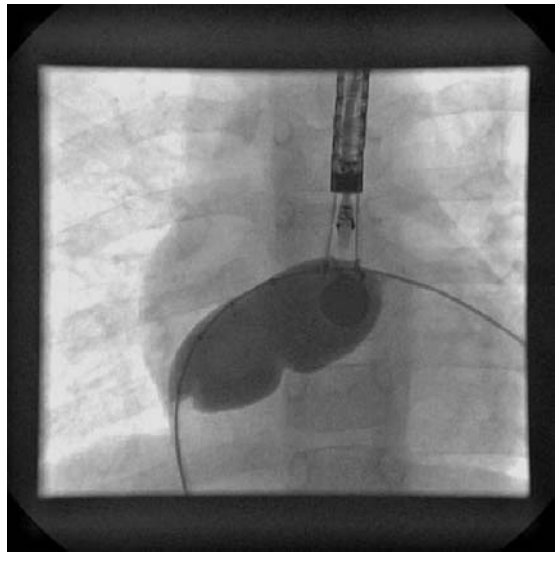

Figure 7.11

device. The device size is determined by the diameter of the waist (range 4–40 mm) (**Figure 7.11**). The two flat disks extend perpendicularly to the waist and have different diameters, the left disk being 12–14 mm larger then the waist and the right disk 8–10 mm larger than the connecting waist. The properties of the material (tightly woven Nitinol wire mesh) allow the device to be stretched, placed inside a small sheath (6–14 Fr) and then reform to its original configuration when not constrained by the sheath. Sealing is assured by three Dacron polyester patches sewn into each part of the device. A stainless steel sleeve is welded to the right disk, allowing for screwing the device to the delivery cable. The device therefore has the advantage of being deployable and repositionable until a perfect position is obtained. Effective occlusion is obtained in 68–99%

of patients immediately after the closure (Dhillon *et al*., 1999; Walsh *et al*., 2000).

This rate gradually increases during follow-up, and disappearance of residual shunt has been observed. Various complications have rarely been described, namely embolization (Peuster *et al*., 2003), arrhythmia or atrio-ventricular block, and aortic or pericardial perforation (Preventza *et al*., 2004). Other issues, such as incompleteness of endothelialization associated with theoretical risks of endocarditis and thrombus formation (Krumsdorf *et al*., 2004), and incidences of late arrhythmias and atrio-ventricular valve dysfunction remain uncertain. These issues have pushed Gore and associates to develop the more flexible Helex device. Its use in humans is increasing, but the implantation of this device is more complex, requiring a longer learning curve, and because of the absence of a centring system, is presently limited to small ASDs.

Muscular Ventricular Septal Defects

The success with occlusion devices for the closure of ASD and PDA prompted the trans-catheter closure of muscular VSD. The procedure for VSD was first attempted by Lock *et al*. in 1988 with devices originally designed for the closure of other intracardiac defects. These devices were used with a variable success rate and residual shunt. In 1997, AGA medical designed an occluder especially for trans-catheter closure of the muscular VSD (Amplatzer muscular VSD occluder, AMVSDO). It is made from a memory-shape alloy wire mesh, tightly woven, forming two flat discs separated by a connecting waist which has a length of 7 mm, corresponding roughly to the thickness of the muscular ventricular septum. A fabric of Dacron is bent into the two discs and the connecting waist to optimize the sealing of the device. The left and right retention disks are respectively 4 and 3 mm larger than the waist (**Figure 7.12**). A stainless steel sleeve with a female head is used to screw the delivery cable to the right disk of the device. It is therefore re-positionable after deployment and has a low profile (6–9 Fr), allowing its insertion in children. Devices are available in sizes of 6–18 mm. The results of the published studies indicate that percutaneous closing is feasible, safe and effective with a rate of occlusion very close to 95% (Arora *et al*., 2003). However, device closure of VSD remains challenging and controversial and should probably be reserved for a small group of children with defects that are difficult to close surgically and/or involve higher risk. Cases are selected by detailed trans-thoracic and/or trans-oesophageal echocardiography. Precise location of the defect, associated anomalies and the distance between the VSD and the surrounding structures (cardiac valves,

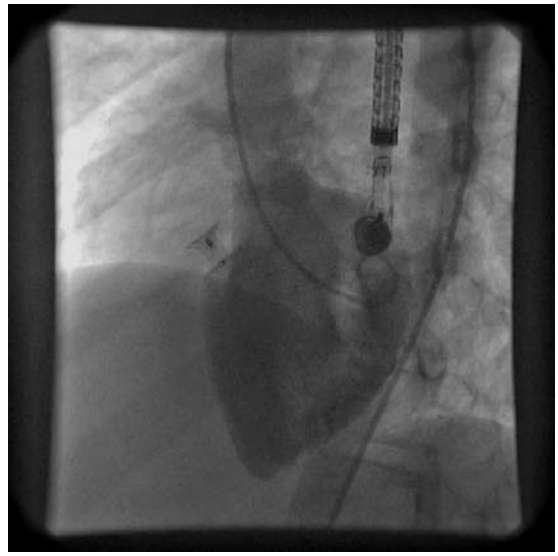

Figure 7.12

papillary muscles, chordae) should be identified. The procedure is performed under general anaesthesia. The VSD is usually crossed from the femoral artery but can be alternatively crossed from the right side. An exchange Amplatzer guide wire is then placed into the pulmonary artery and caught with a gooseneck snare to create a loop between the venous and the arterial accesses. The delivery system is then advanced over the wire into the left ventricle. To avoid kinking of the delivery system, a fine guide wire is left in its lumen while advancing the device attached to its delivery cable. Once the device reaches the tip of the delivery system, the glide wire is removed. To limit the sheath size in the artery, the device is usually deployed from the venous side (jugular or femoral vein). This is done under TOE and fluoroscopic guidance to properly position the device, and under continuous electrocardiographic monitoring to look for any arrhythmia/conduction problems that could be encountered during the procedure. The patient is usually discharged the day after the procedure. The patient is heparinized during the procedure and heparin is continued for 12 hours; 3–5 mg/kg aspirin, associated or not with clopidogrel, is given for 6 months. Current results were recently reviewed by Holzer *et al*. (2004). Sixty-nine patients were enrolled in the study and underwent a total of 77 procedures. Multiple VSDs were present in nearly 40% of patients. The procedure was successful in 66/77 procedures (85.7%). Repeated percutaneous procedures for unsuccessful closure or residual VSDs were required in 10% of patients. Seventeen patients (6.6%) received more than one

device. The median fluoroscopy time was 54 minutes. The authors observed adverse events in nearly half of the patients studied; 10% of them were major complications. Two deaths were procedure-related. One patient died of severe cerebral damage following the management of a cardiac perforation, the device being deployed in the pericardium. Another patient had a cerebral insult as a result of cardiac arrest secondary to significant blood loss during attempts to retrieve an embolized device. Two devices embolized and were subsequently retrieved either percutaneously or surgically. Hypotensive episodes and/or cardiac arrest occurred in about 13% of patients. Five patients required blood transfusion. Transient arrhythmias or conduction abnormalities were seen in nearly 20% of patients. Two patients developed permanent right bundle branch block. One patient had a transient ischaemic attack 24 hours after the procedure. After 24 hours, residual shunt was observed in more than 50% of patients. Most of them were trivial (93%). The closure rate improved with time, being around 93% at 12 months. Patients with body weight of less than 5 kg had longer procedures, a greater proportion of residual shunts and a higher rate of adverse effects and unsuccessful closure.

The current data suggest that trans-catheter closure of muscular VSD is efficacious, and that the technique can be considered as an alternative to often difficult surgery. However, this technique remains challenging and controversial in small children, particularly when weight is less than 6 kg.

Perimembranous VSD

Before the availability of the new perimembranous Amplatzer device, several authors have tried to close perimembranous VSDs using devices originally designed for ASD or PDA closure. The rates of complications (i.e. embolization, aortic insufficiency) and residual shunt were high. Following animal experiments, Hijazi *et al*. (2002) and Bass *et al*. (2003) recently reported their human experience with perimembranous VSD closure, using a specifically designed device, the Amplatzer perimembranous septal occluder (AGA, Golden Valley, MN) (**Figure 7.13**). The important feature is the asymmetrical left and right ventricular discs to accommodate the close proximity of the aortic valve.

Devices are available in sizes of 4–18 mm, in 2 mm increments. The delivery system is braided with a tight curve (180°) to prevent kinking and allow positioning into the left ventricular apex. It has a low profile (6–9 Fr), allowing its insertion in children. The technique of implantation is very similar to muscular VSD closure. Since, in animal studies, withdrawal of the expanded

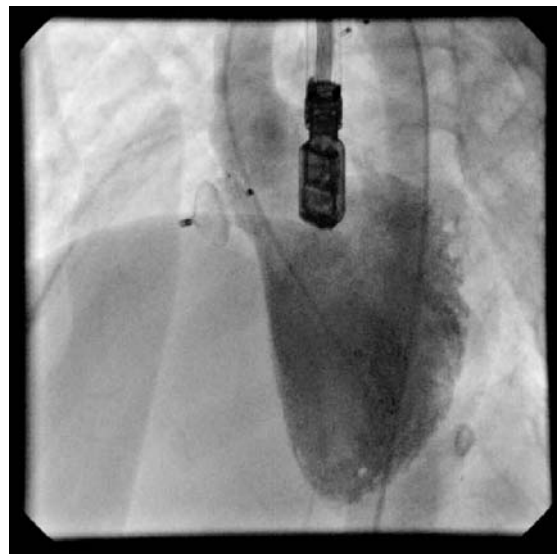

Figure 7.13

left ventricular disk through the mitral valve apparatus resulted in damage to the mitral valve, this disk should be opened in the left ventricular outflow tract rather than in the apex. As with all long sheaths positioned in the left side of the heart, full heparinization and careful flushing are mandatory to avoid air or thrombi that could embolize into the systemic circulation. Prophylactic antibiotic use is usually given during the procedure and the day after to reduce the risk of device-related infection. Bacterial endocarditis prophylaxis should be given during 6 months after placement and during lifetime if the shunt persists; it is discontinued if there is no residual shunt. Aspirin (3–5 mg/kg/day) may also be given during this 6 month period.

Since the first human reports (Thanopoulos *et al*., 2003), 96 patients with mean age and weight of 9 years and 28 kg, respectively, have received this implant worldwide. The mean pulmonary:systemic blood flow ratio was 1.6, range 0.9–10. Failed implantation was rare and was caused by device-related aortic insufficiency and atrio-ventricular (AV) block and usually resolved with removal of the device. Other technical failures included the inability to position the delivery sheath in the left side of the heart. The occluder was effective in eliminating the left-to-right shunt immediately or early during follow-up.

No mortality related to the procedure occurred. Eleven patients had adverse effects (11.5%), including aortic or tricuspid regurgitation in 5 and 7 patients, respectively, and arrhythmias or conduction abnormalities consisting of right and left bundle branch block, complete heart

block ($n = 3$) and ventricular tachycardia in 10 patients. Most of these electric abnormalities were temporary ($n = 8$). Two patients had AV block 48–72 hours after the implantation, which were revealed by syncope and required permanent pacing. One patient returned to sinus rhythm 3 weeks after VSD closure and pacemaker insertion. These electric abnormalities probably result from trauma of the device/delivery system/catheter against the conduction system, which runs on the margins of the defect. Device embolization occurred in one patient. The device was retrieved percutaneously and a bigger device was successfully implanted without any further adverse events. As for muscular VSD closure, patients with a lower body weight (less than 10 kg) had longer procedures, a greater proportion of adverse effects (41.7% vs. 16.3%) and unsuccessful closure (25% vs. 4.8%). The extension of the VSD into the inlet septum and the existence of an aortic valve prolapse were also predictive of worse outcome.

To conclude, despite interesting early results, this technique is still investigational. Before its general use, longer follow-up is necessary to determine the risk of bacterial endocarditis, and to ascertain the safety of the procedure with regard to trauma to the conduction system and aortic and tricuspid valves. The occurrence of a significant valvular insufficiency or a heart block before the release of the device should, in our opinion, lead to its removal. However, the absence of such complications in the short term does not preclude their occurrence in the mid- and long-term follow-up. Indeed, it is possible that long-term distortion of the left ventricular outflow tract and interference with the conduction system occur and may result in late complications. With regard to the management of device-related heart block, careful monitoring during the procedure and during the following days is mandatory, the risk seeming to be high during the first 96 hours after the procedure. Finally, since patients included in this registry were larger and older than many in whom closure of a membranous VSD is required, the utility of the Amplatzer membranous ventricular septal occluder in a younger population remains to be determined.

Other Vascular Abnormalities and Surgically Created Structures

Devices originally designed for the closure of specific intracardiac defects have permitted the closure of various abnormal vessels, such as coronary artery fistulae (Armsby *et al*., 2002), veno-venous or veno-arterial fistulae (Abushaban *et al*., 2004), major aortopulmonary collateral arteries (Perry *et al*., 1989) or vessels in pulmonary sequestration (Gao *et al*., 1993). Using the same devices, surgically created structures, viz. modified Blalock–Taussig shunt (Burrows *et al*., 1993), fenestration of the total cavopulmonary connection (Rueda *et al*., 2001) and paravalvular leaks (Piéchaud, 2003) have been closed by trans-catheter techniques. The success rate of these procedures varies and depends on the devices used and the lesions closed. Risks are low and include embolization, haemolysis, endocarditis and recanalization. Catheter techniques are relatively similar to those described previously.

TRANS-CATHETER PULMONARY VALVE INSERTION

A number of complex congenital heart defects involving the right ventricular outflow tract require a surgical repair in the early childhood. Surgery consists in repairing the intracardiac anomalies and interposing a prosthetic conduit or a homograft between the right ventricle and the pulmonary artery. Certain malformations of the aortic valve may also require pulmonary valve replacement as a part of the Ross procedure. The life span of conduits is limited due to somatic growth, calcification and/or accumulation of fibrous tissue on their walls. This commits the patients to multiple surgical reoperations to replace degenerated conduits. As shown previously, stent implantation in obstructed conduits delays the surgical replacement of failing conduits. This relieves obstruction but, by applying the valve of the conduit against its wall, creates a pulmonary insufficiency that in the long term is deleterious, or influences right ventricular function (Gatzoulis *et al*., 2000).

To overcome this drawback, we had the idea of mounting a bovine jugular venous valve (Medtronic Inc.) into a balloon-expandable stent (CP stent, Numed Inc.) (Bonhoeffer *et al*., 2000a,b). The valve–stent assembly was deployed using a custom-made delivery system. After *in vitro* testing and a series of successful animal experiments, human application began in September 2000. After a pilot study, in which only patients with obstructed conduits, with or without pulmonary regurgitation, were included, patients with pure pulmonary insufficiency were also eligible. The patient selection is crucial to the success of the technique, since we are currently limited by the availability of valves. Also, valved stents require a large sheath and can only be proposed for patients weighing more than 20 kg. The bovine jugular venous valves are available in diameters of 12–22 mm. This makes percutaneous valve insertion impossible in patients with a right ventricular outflow tract (RVOT) larger than 22 mm. Analysis of previous surgery and non-invasive assessment of RVOT anatomy are important for patient selection. In particular, MRI with 3D reconstruction of the RVOT is particularly helpful in searching for a stenotic component for stent

anchoring, but also for assessing RV function and quantifying pulmonary incompetence. Fifty-five patients in need of surgical replacement of the pulmonic valve and with adequate RVOT anatomy have been included in this study so far. After selection of patients, a right cardiac catheterization via the right femoral vein ($n = 54$) or the right jugular vein ($n = 1$) was performed under general anaesthesia. Haemodynamic evaluation and angiograms in multiple projections were done in order to confirm the indication for the procedure and to define the precise anatomy of the RVOT (**Figure 7.14**). Heparin (100 units/kg) and antibiotics (cephalosporins) were given intravenously at the beginning of the procedure, according to the standard protocol. After placement of a right coronary catheter in a distal branch pulmonary artery, a super-stiff exchange guide wire (Amplatz, Meditech) was positioned in the distal pulmonary artery. This was followed by an inflation of a balloon (Zmed II 18* 4 or Mullins 18*4; Numed Inc.) in the RVOT, in order to determine whether there was sufficient room to implant a pulmonary valve without creating additional obstruction, and to see that the stent can be dilated to its maximum required diameter, especially in those patients who already had some stenosis of the previously surgically implanted valve. An 18 mm bovine jugular venous valve (Medtronic Inc.) sutured into a platinum stent (Numed Inc.) is manually crimped onto a specially designed delivery system. The sheath can freely slide over the stent. At the tip of the catheter, a 1 cm long dilator has been glued to allow a smooth transition between the tip and the sheath, thus limiting the risk of traumatizing the vessel during skin insertion. Before insertion of the delivery system, a 22 Fr dilator is temporarily placed into the vein. After covering of the valved stent, the whole assembly is then passed over the guide wire and advanced into the pulmonary artery. Once in the desired position, the valved stent is uncovered. The inner balloon is initially inflated, followed by the outer balloon, thus deploying the valved stent. The balloons are then deflated and the delivery system is removed from the patient, leaving the valve in place and the guidewire in position. Angiographic and haemodynamic evaluation is repeated before finally removing the guidewire (**Figure 7.15**). Age at implant was 9–42 years (mean 18.1 years). Nearly 50% of patients were in NYHA classes III or IV, 40% in class II and 10% in class I. Homografts, biological valved conduits and native outflow tract represented 70%, 22.2% and 7.4%, respectively.

Percutaneous trans-catheter valve implantation was attempted 56 times in 55 patients. The technique was successful in 54 patients (96%). In the two failed procedures, the valved stent could not be manoeuvred into the RVOT despite prolonged manipulation, due to the presence of heavily calcified extra-anatomic conduits. The delivery system was subsequently modified; one of these two patients was subsequently successfully valved and the other is awaiting a new attempt. The median procedure time was 59 minutes and the median screening time was 25.5 minutes. After the procedure, the right ventricular systolic pressure and right ventricular outflow gradient fell significantly from 64.4 ± 18.7 to 50.8 ± 13.4 mmHg ($p = 0.0004$) and

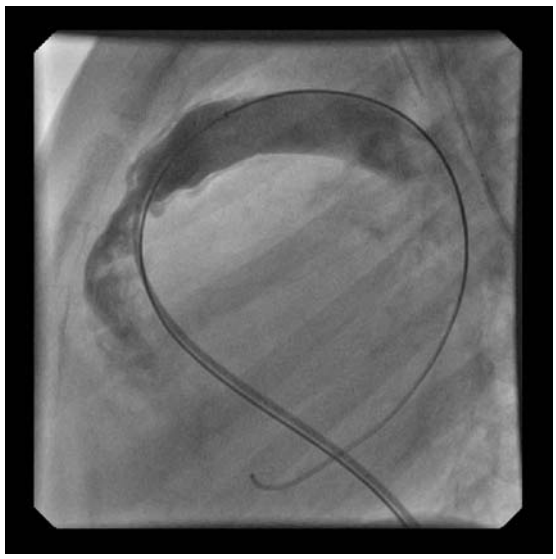

Figure 7.14

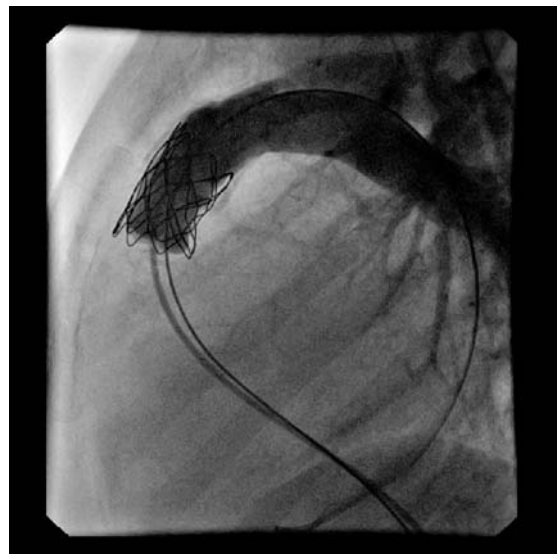

Figure 7.15

from 40.2 ± 21.9 to 25.0 ± 16.3 mmHg ($p = 0.0005$), respectively, without significant change in the systemic arterial pressure. There was a significant increase in the pulmonary artery diastolic pressure (9.5 ± 3.1 to 13.1 ± 3.6 mmHg, $p < 0.001$) after the procedure. Angiography showed an improvement in pulmonary valve function with only mild regurgitation, despite the presence of a guide wire and catheter across the valve. Echocardiography performed 24 hours later confirmed these data, showing a significant reduction in the tricuspid regurgitation velocity, in the right ventricular outflow tract velocity and in the grade of pulmonary regurgitation, with all patients having $\geq$ grade 2 PR before, and none with more than grade 2 PR after the procedure ($p < 0.001$). NYHA functional class, assessed during the latest follow-up, improved after PTCVI. There was no procedural mortality and there has been none up to nearly 4 years from the first implantation. Four patients, however, required urgent surgery: two because of acute device embolization or stent instability, one because of homograft rupture with haemothorax, and one because of unexpected device embolization 8 months after implantation. The device was explanted in three of these four patients. One patient had late explantation (9 months) for endocarditis after unprotected dental extraction. Two had detachment of the distal portion of the delivery system during removal of the system from the femoral vein. Both were retrieved successfully using a snare. The delivery systems were subsequently modified to prevent this complication. In-stent stenosis was apparent in 7 patients soon after implantation. Angiography showed the venous wall of the biological valve encroaching into the lumen of the stent, creating a ''hammock'' effect. This was the result of suturing only the ends of the venous wall to the ends of the stent. This was treated by implantation of an additional valved stent inside the first one ($n = 5$) and by surgical removal of the device ($n = 2$). The design of the device was modified to solve this problem by suturing the whole length of the venous segment to the entire length of the stent. So far, no further in-stent stenosis was encountered. All patients except those needing urgent surgery were ambulatory within 24 hours and were discharged within 48 hours.

To conclude, trans-catheter implantation of pulmonary valves has been the first clinical experience for non-surgical management of valvular regurgitation. Our group has shown the feasibility, safety and efficacy of pulmonary valve implantation in humans. There are still some unanswered questions about the durability of this valve in the long term and the benefits of this technique in the history of the disease. However, since all included patients who had a formal indication for surgery, this was delayed in all, making the present strategy of value. In the subgroup of obstructive lesions, larger multicentre studies are necessary to demonstrate the superiority of this technique over the standard approach, i.e. the ''non-valved'' stenting of the RVOT. At present, surgery remains the only option for patients weighing less than 20 kg and with small or too large RVOT (Coats *et al*., 2005).

We are in the process of developing devices and approaches for extending present indications to patients with large RVOT, who represent the large majority of patients requiring pulmonary valve replacement (Boudjemline *et al*., 2004).

HYBRID APPROACHES

Interventional cardiology has been generally considered as competitive to surgery, since some lesions are now ''curable'' without the need for surgery. Elsewhere, catheter treatments have been adjuvant to surgical treatment. However, a collaborative approach between surgeons and interventional cardiologists might be more appropriate in order to decrease cumulative morbidity and mortality and improve outcome of certain congenital heart defects. Various collaborations could be beneficial for the patient. For example, in small infants, vascular access for interventional treatment, i.e. balloon dilatation, or stent implantation for vascular or valvular stenosis, might be difficult. Better access can be provided by surgeons (Robinson *et al*., 2000). Using these approaches, interventional procedures can be performed with very low risk of complications. Advantages include avoidance of large catheters in the femoral vessels and reduction of haemodynamic compromise secondary to the passage of catheters across valves. Reciprocally, it is sometimes difficult for surgeons to perform a repair adequately (i.e. some VSDs, and distal pulmonary arteries stenoses). These situations can lead to:

1. Difficult weaning from cardiopulmonary bypass.
2. Early or late reoperations for residual shunting, or incomplete relief of obstruction.
3. Prolonged stay in cardiac intensive care.
4. Increased morbidity and mortality.

Recent advances in interventional techniques and the availability of new devices and stents have enabled interventional cardiologists to manage these types of lesions intraoperatively (Levy *et al*., 2003). Trans-ventricular closures of apical VSD in infants (Okubo *et al*., 2001) or balloon dilatation/stenting to relieve obstructions in the Fontan pathway or in pulmonary arteries (Hjortdal *et al*., 2002) have been attempted recently under direct vision, or fluoroscopic or echocardiographic guidance.

Avoiding the cardiopulmonary bypass is another aim of this approach. We and others have performed

trans-ventricular closure of VSD in infants (Fishberger *et al*., 1993). and trans-ventricular valve replacement experimentally. In such situations, the ability to convert this procedure to a bypass operation increases the security for the patient.

From a technical point of view, the surgeon places a purse-string suture. A guide wire is then passed via a needle (''18 gauge'') puncture at the site of the purse string. The delivery sheath is positioned and the intended intervention, ''bare or valved stent placement'' or ''device closure'' of a defect is performed.

Combined approaches have also been developed for the treatment of complex heart defects, such as hypoplastic left heart syndrome. Some groups are investigating a combined approach where the first procedure is a bilateral pulmonary artery banding, followed by or preceded by a PDA stenting through the main pulmonary artery (Gibbs *et al*., 1993).

The second stage is the creation of a partial cavo–pulmonary connection with some features of the Norwood stage 1 operation (Norwood stage I/II procedure). In some institutions with a programme of neonatal transplantation, this approach is considered to be a bridge to orthotopic transplant, because poor availability of neonatal heart donors was responsible for a significant number of deaths of patients on the waiting list.

The last subgroup of hybrid approaches is the staged procedure. The aim of this approach is to prepare the patient for further interventional procedures to avoid secondary inevitable surgery. A good example of this is the completion of partial cavo–pulmonary connection. This operation is usually performed surgically around 3 years of age. Hausdorf *et al*. (1996) reported an approach in which the surgeons would prepare the patient for trans-catheter completion during the PCPC. This involves a bidirectional Glenn shunt with simultaneous anastomosis of both sides of the superior vena cava with the right pulmonary artery, the closure or banding of the right atrium–SVC–PA connection, and the placement of a cuff around the IVC on the IVC–RA connection. Three years later, if the patient is able to tolerate the Fontan physiology, a trans-septal needle is used to perforate the occluded connection between the inferior part of the SVC and the right PA and a covered stent is inflated from the IVC to the PA. This diverts the blood from the IVC directly to the PA, excluding the passage into the univentricular heart. These techniques are currently being experimented upon by a number of groups, in collaboration between cardiologists and cardiac surgeons. Their effectiveness will be seen in future publications.

In the same manner, this approach could be applied to other defects, such as valve replacement. At present we are experimentally investigating a staged approach to make valve insertion amenable to trans-catheter technique. Atrio-ventricular and aortic valves could potentially benefit from such an approach. During surgery, the surgeon would replace the malfunctioning valve. Once the new valve has degenerated, percutaneous replacements would then become possible, avoiding the need for cardiopulmonary bypass and multiple reoperations.

The preparation for trans-catheter valve insertion implies the use of non-mechanical valve, resection of chordae and location of ''dangerous surrounding structures'', such as the coronary arteries, and of the inserted bioprosthesis, using radio-opaque markers. Using this approach in lambs, we were able to replace mitral and large pulmonary valves without cardiopulmonary bypass.

To conclude, collaborative approaches of physicians, industry, surgeons and interventional cardiologists may decrease cumulative morbidity and mortality and improve outcome of certain congenital heart defects. Various lesions and diseases are, to date, amenable to such approaches but, if superiority of these approaches is demonstrated, the number of patients treated with these approaches will increase in future. Hospitals will have to develop ''hybrid rooms'', including both fluoroscopic and surgical facilities. However, this new subspecialty at the frontier of both interventional cardiology and cardiac surgery will need trained personnel.

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8

The Anatomy of Ventricular Septal Defects and Their Conduction Tissues*

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The purpose in this chapter is to show the anatomical variation that can exist in communications between ventricles, and to show how this variation relates to the atrio-ventricular and ventriculo-arterial connections that are present. In each example, we will describe the surgical anatomy of the conduction tissues. The key for various parts of the conduction tissues as they are superimposed on photographs is illustrated in **Figure 8.1**. Note the different conventions used for the right and left bundle branches when they are seen on the surface of the septum, as opposed to their projected position as might be viewed through the septum. This code is used throughout the chapter.

BASIC RULES REGARDING DISPOSITION OF THE CONDUCTION SYSTEM

In any heart with a communication between two ventricles, application of a few basic rules enables fairly accurate prediction of the course of the conduction system. First, the regular atrio-ventricular node is always located at the apex of the triangle of Koch. Second, the branching atrio-ventricular bundle is positioned astride the crest of the muscular ventricular septum, regardless of whether that septum is aligned or malaligned relative to the atrial and muscular outlet septal structures. Third, with normal alignment of the atrial septum and the muscular ventricular septum, the apex of the triangle of Koch is itself adjacent to the crest of the muscular septum. This creates the situation for normal formation of the atrio-ventricular conduction axis. This is shown in the dissection in **Figure 8.2**, in which the course of the axis can be seen running from the atrial septum into the aortic outflow tract, where it is carried on the muscular ventricular septum. Malalignment between these septal structures prevents the normal atrio-ventricular node from making contact with the branching bundle carried on the ventricular septum. In this circumstance, as seen, for example, in congenitally corrected transposition, an anomalous anterior node gives rise to the penetrating atrio-ventricular conduction axis. The fourth rule is that this anomalous node is a remnant of a ring of conduction tissue that, during development, surrounds the morphologically right atrial vestibule (Anderson *et al*., 1974b). The node, therefore, may be formed at any point round the right atrio-ventricular orifice, but is usually an anterior structure. The fifth rule is that, when the ventricular septum is abnormally formed, as in hearts with univentricular atrio-ventricular connection, a regular atrio-ventricular node can make contact with the atrio-ventricular conduction axis only when the remnant of the muscular ventricular septum extends to the crux of the heart, as in double-inlet right ventricle. In all other circumstances, an abnormally located node is formed. Usually it is in an anterior position, but it can be located at any point around the right atrio-ventricular orifice. The point of its formation is determined by where the muscular ventricular septum joins the junction, as shown in **Figure 8.3**. Thus, when the septum is anterior to both atrio-ventricular valves, as in double-inlet left ventricle, the connecting node is also an anterior structure. When the right atrio-ventricular valve straddles the postero-inferior part of the ventricular septum, the septum does not reach the crux. The anomalous connecting node is then formed at the point where the ventricular septum joins the atrio-ventricular junctions, so that the anomalous node may well be posterior in relation to the regular node, but it is not located in the atrial septum. These basic rules are elaborated upon in the subsequent sections, but they provide the foundation

* Editors' note: The editors have decided to use Latin anatomical terminology to maintain uniformity throughout this book. This was done with the agreement of Professors Anderson and Becker. They wish to emphasize, however, their preference for usage of English rather than Latin words.

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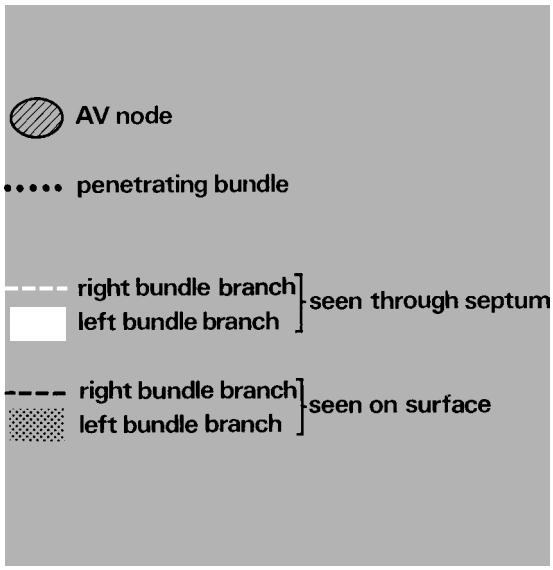

Figure 8.1

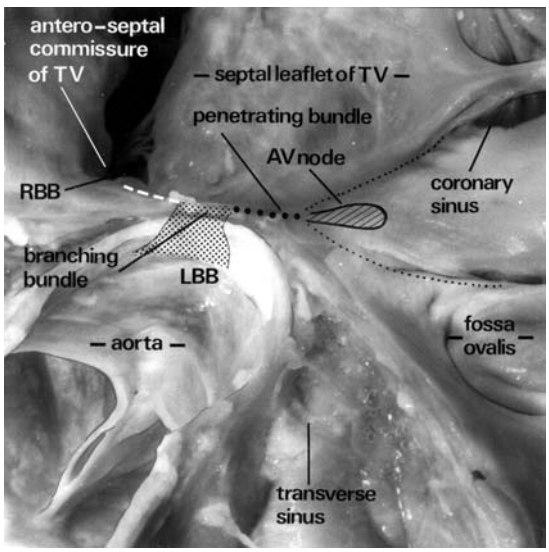

Figure 8.2

for understanding unusual dispositions of conduction tissues. In the subsequent sections, the morphological features and conduction tissues of hearts with the following defects are considered:

- Isolated ventricular septal defects.
- Tetralogy of Fallot.

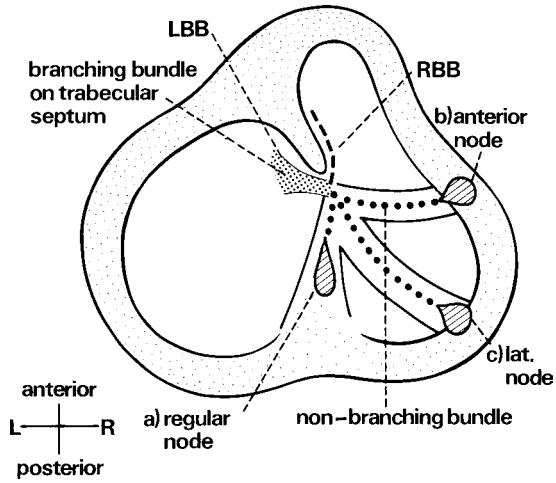

Figure 8.3

- Concordant atrio-ventricular and discordant ventriculo-arterial connections (transposition).
- Double-outlet ventricles.
- Single outlet of the heart.
- Discordant atrio-ventricular connections.
- Straddling atrio-ventricular valves.

ANATOMY OF ISOLATED VENTRICULAR SEPTAL DEFECTS

We use the term ''isolated'' ventricular septal defect to describe a hole between the ventricles in a heart with concordant atrio-ventricular and ventriculo-arterial connections, and normally positioned chambers and arteries, in other words otherwise normal hearts, with no other major associated lesions. The defect itself is considered to represent the margin around which the surgeon must place a patch so as to separate the pulmonary and systemic circulations, which before correction would have mixed across the defect. Understanding of the anatomy of such defects is greatly facilitated if the ventricular septum is thought of as possessing a large muscular component together with the fibrous membranous septum (**Figure 8.4**) (Moulaert, 1978). Previous classifications of defects have either been exceedingly simple, such as ''high'' and ''low'' defects, excessively complicated, as evidenced by the classification of Goor *et al*. (1971), or have been wedded to that enigmatic structure, the crista supraventricularis (supraventricular crest), as in ''supracristal'' and ''infracristal''. We suggest that, in simple terms of value to the surgeon, isolated ventricular septal defects can best be divided (**Figure 8.5**) into those in which an area of fibrous continuity between the leaflets of the aortic

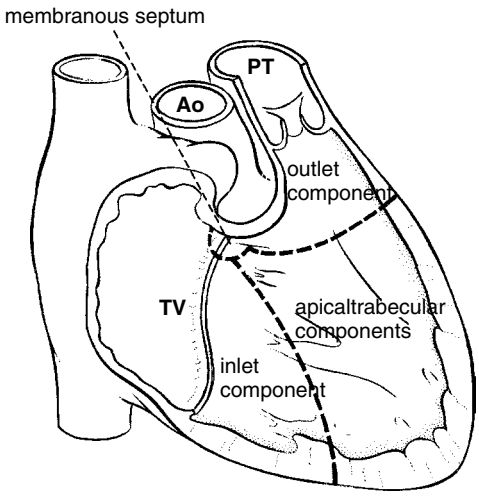

Figure 8.4

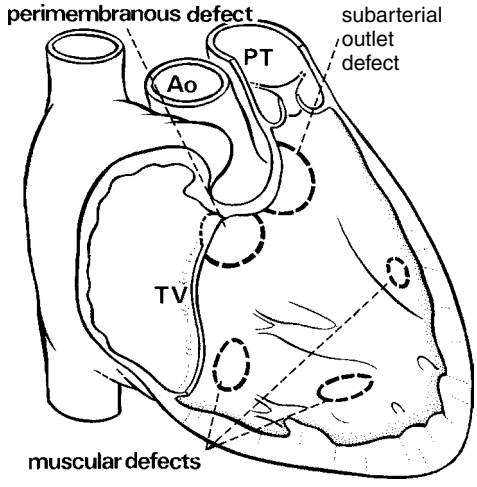

Figure 8.5

and tricuspid valves forms a margin of the defect, those that are roofed by the conjoined leaflets of the arterial valves, and those with entirely muscular rims. Each of these anatomical patterns, and the variation within each phenotype, are considered separately.

Defects with Fibrous Continuity Between the Leaflets of the Aortic and Tricuspid Valves

These defects have conventionally been termed ''membranous defects'', but this can be a misleading term. As Becu *et al*. (1956) indicated, the defect is rarely caused by a deficiency of the interventricular component of the membranous septum. Indeed, during the development of the heart, the interventricular component is frequently not formed until after birth (Allwork and Anderson, 1979). The defect with fibrous continuity between the leaflets of the aortic and tricuspid valves results instead from deficiency of the muscular septum forming the circumference of the membranous area. As such, the deficiency can extend to open toward the inlet, apical trabecular or outlet components of the right ventricle. Because all these defects extend around the persisting components of the membranous septum (**Figure 8.6**), we term them *perimembranous* defects. The direction of extension of the defects, although not altering their basic morphology, does significantly affect their relationship to important structures, such as the atrio-ventricular valves or the medial papillary muscle. The general relationship of a perimembranous defect to the atrio-ventricular conduction axis, nonetheless, is always the same, as demonstrated in **Figure 8.6**.

Perimembranous Defects Opening Centrally

These are the typical ''membranous'' defects. The defect opens to the right ventricle adjacent to the area of the central fibrous body, and the long axis of the defect is orientated toward the cardiac apex (**Figure 8.6**). When viewed from the right atrium (**Figure 8.7**), the zone of apposition between the septal and anterosuperior leaflets of the tricuspid valve, attached to the medial papillary muscle, is above and to the left of the defect. A cleft in the septal leaflet of the tricuspid valve extends frequently to the central fibrous body at the site of the defect, as shown in **Figure 8.7a**. Beneath the superior component of the septal leaflet thus formed, the area of contiguity of the tricuspid valve, the central fibrous body and the aortic valve forms the atrial margin of the defect as seen by the surgeon, frequently with a remnant of the interventricular membranous septum buttressing the right margin of the defect beneath the leaflet of the tricuspid valve. This membranous septal remnant must be distinguished from the leaflet of the tricuspid valve because the remnant may contain and cover conduction tissue. The leaflet, in contrast, only very rarely contains or covers conduction tissue (Ueda and Becker, 1985). This relationship is shown diagrammatically in **Figure 8.8**. If the remnant of the membranous septum is substantial, it can be safely used as an anchorage for sutures. Indeed, the remnant becomes aneurysmal in many hearts in an attempt to close the defect. It frequently forms a hammock-like structure attached by individual chordae to the edge of the defect. In other hearts with clefts in the septal leaflets of the valve, the leaflets of the tricuspid valve can fuse to the ventricular septum at

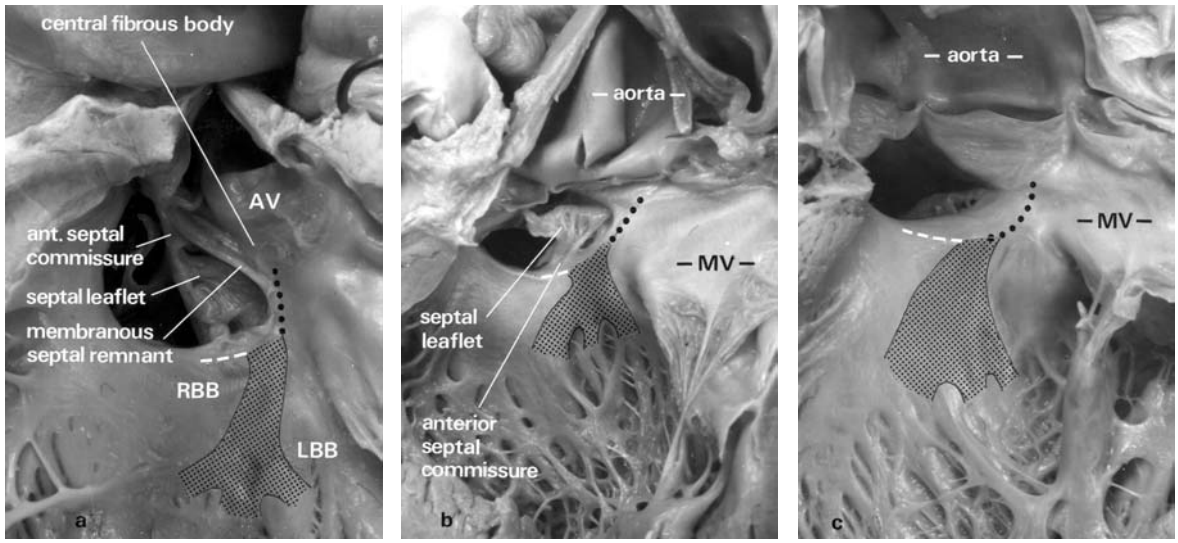

Figure 8.6

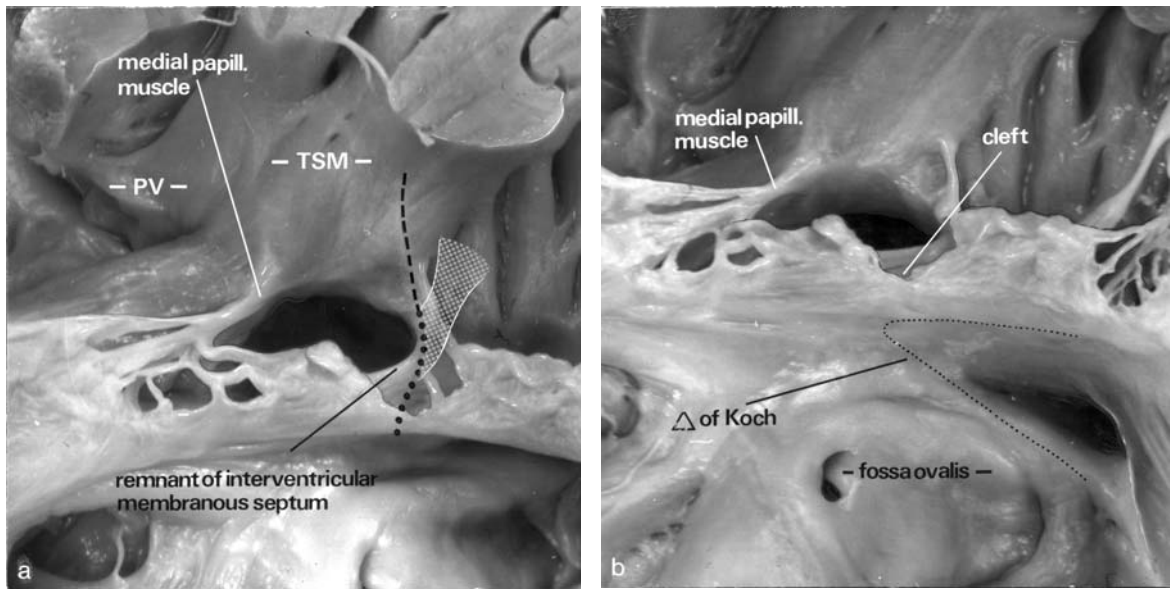

Figure 8.7

the apex of the cleft. The defect then appears to be a communication between the right atrium and the left ventricle. Such defects are often considered to exist in the atrio-ventricular component of the membranous septum. True defects of the atrio-ventricular membranous septum are rare (McKay *et al*., 1989). In most hearts with left ventricular to right atrial shunting through a ventricular septal defect, the atrio-ventricular component of the membranous septum is intact, with the attachment of the leaflets of the tricuspid valve on the atrial side of the defect, as shown diagrammatically in **Figure 8.9b**. If the defects were truly in the atrio-ventricular septum, the hinge of the septal leaflet would be on the ventricular aspect of the defect, as depicted in **Figure 8.9a**. This

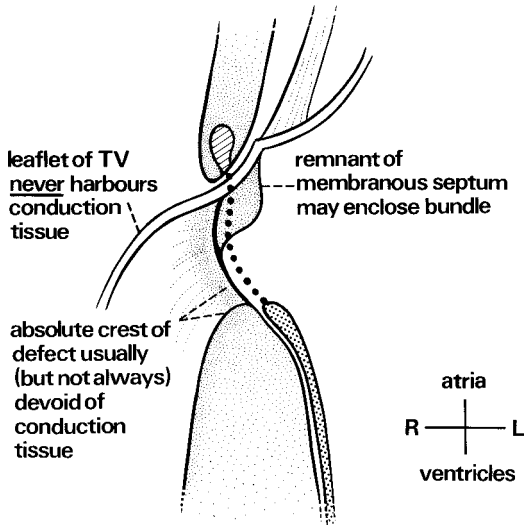

Figure 8.8

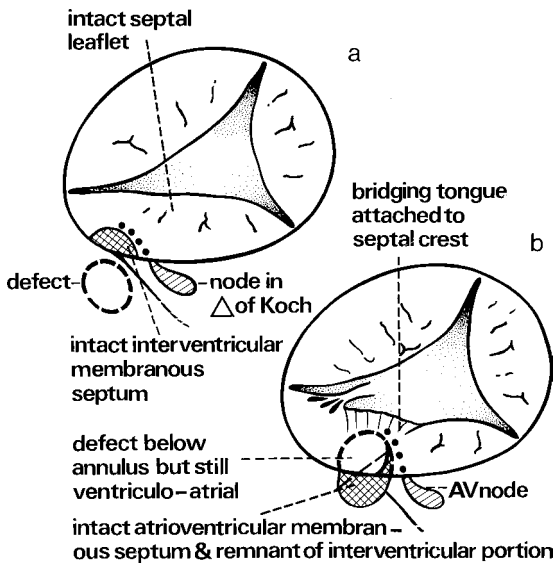

Figure 8.9

latter arrangement is exemplified by the case reported by McKay *et al*. (1989). On the ventricular aspect, the rim of the defect is muscular (see **Figure 8.6b**). When viewed from the right ventricle, the defect is cradled above the postero-inferior limb of the trabecula septomarginalis, and the medial papillary muscle is usually above and to the left of the defect, although it may be attached toward the apex. From the standpoint of the conduction tissues, the apex of the triangle of Koch is always an excellent guide to the position of the atrio-ventricular node. As indicated elsewhere, the triangle of Koch is brought into prominence by tension placed on the Eustachian valve. In the defects characterized by aortic-tricuspid valvar continuity, or perimembranous defects, the apex of the triangle is always on the inlet aspect of the defect, so that it points towards the coronary sinus and the crux of the heart. The most vulnerable area of the defect is the point at which the axis of conduction tissue passes from the right atrium to the ventricles, this being the site of penetration of the atrio-ventricular bundle. This is always in the central fibrous body and, in this region, the bundle is enclosed in ''white tissue''. A suture placed in this fibrous tissue may produce complete heart block. Our studies militate against the suggestion of Goor and Lillehei (1975) that white tissue is always safe from the standpoint of serving as an anchorage for sutures. The area of penetration, as indicated, is on the inlet side of the defect, and is the potential area of danger. Only one case that we have studied has a branch of the bundle penetrated into the leaflet of the tricuspid valve (Ueda and Becker, 1985). In almost all instances, therefore, if sutures in this area are passed through the leaflet rather than its attached margin, they should not damage the axis of conduction tissue (**Figure 8.8**), albeit that, as discussed above, if the remnant of the membranous septum is substantial, it too might be used as a safe anchorage for sutures. It should be remembered, nonetheless, that during penetration the bundle may be directly related to the margin of a perimembranous defect and may be enclosed within the remnant of the interventricular membranous septum. Once it has penetrated, the bundle tends to veer to the left ventricular side of the defect, where the fascicles of the left bundle branch cascade down into the apical trabecular region of the left ventricle. Thus, in most hearts, the branching bundle does not occupy the crest of the ventricular septum. In other hearts, however, the conduction tissues are more closely related to the rim of the defect. Thus far we are unable to distinguish these hearts from the safer variety simply by inspection from the right ventricle. In most of the unsafe hearts, the right bundle usually passes intramyocardially at the apex of the defect, and then descends as a thin cord within the substance of the trabecula septomarginalis. As a general rule, the muscular rim of the defect on the outlet side of the medial papillary muscle is unrelated to conduction tissues.

Perimembranous Defects Opening to the Inlet of the Right Ventricle

These defects have the same basic morphology as the typical perimembranous defect, except that instead of extending toward the apex, they extend so as to open

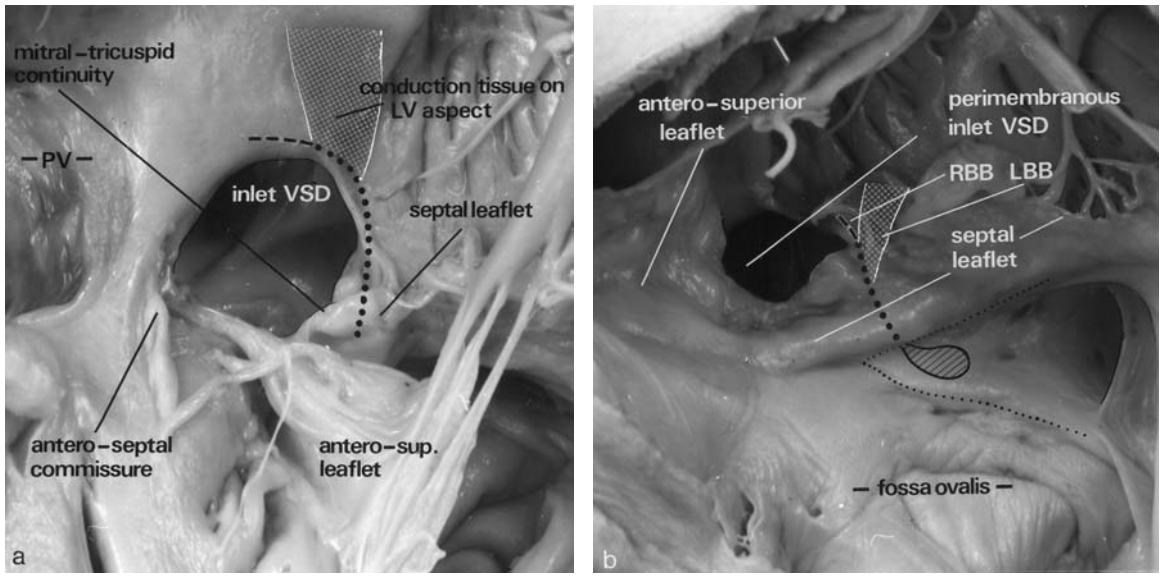

Figure 8.10

into the inlet of the right ventricle. Because of this, they occupy a position beneath the septal leaflet of the tricuspid valve. In addition to the central fibrous body, the contiguous leaflets of the mitral and tricuspid valves form the atrial margin of the defect, which is composed of fibrous tissue. This is shown in **Figure 8.10a**, in which the leaflets of the tricuspid valve have been retracted to show the atrial margin of the defect, as would be seen from the atrium. Defects with minimal excavation towards the inlet differ only in their position. When excavation is extreme, the septum may be deficient back to the crux of the heart. This defect, in the past, was often referred to as ''atrio-ventricular canal'' type (Neufeld *et al*., 1961). The essence of the atrio-ventricular canal, nonetheless, is a common atrio-ventricular junction. Most of the defects described in this fashion are no more than perimembranous defects with extensive communication to the inlet of the right ventricle. They have separate right and left atrio-ventricular junctions. Only when there is a common atrio-ventricular junction should they be classified as ''atrio-ventricular canal'' defects. The medial papillary muscle is to the left of the defects that open to the ventricular inlet, and, as with all perimembranous defects, the conduction axis is on the inlet side of the defect.

Perimembranous Defects Opening to the Outlet of the Right Ventricle

These defects also have the basic morphology of a perimembranous defect, but the deficiency of the muscular septum is toward the outlet of the right ventricle, with malalignment of the muscular outlet septum. Because of this, the aortic valve is often able to override the cavity of the right ventricle, and the medial papillary muscle is deviated more to the inlet margin of the defect. An approximate surgical view of such a defect from the right atrium is shown in **Figure 8.11**,

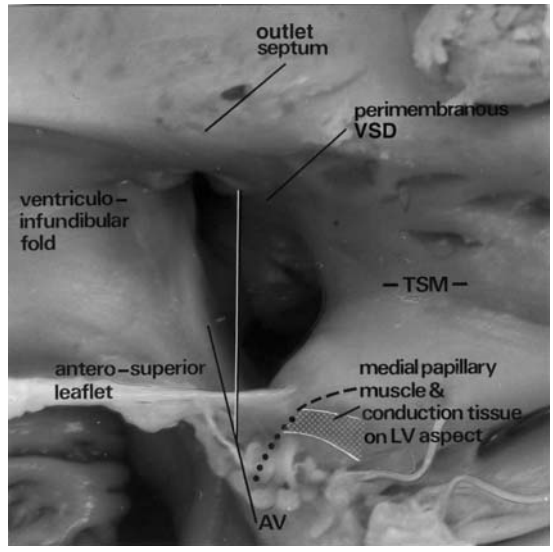

Figure 8.11

with the site of conduction tissue marked. The medial muscle tends to be to the right hand of the surgeon as the defect is viewed from the right ventricle. In addition, rather than being related to the septal leaflet of the tricuspid valve, the anterosuperior leaflet shields the defect. When aortic override is present, the supporting ventriculo-infundibular fold forms the left margin, and the crest of the trabecula septomarginalis reinforces the ventricular rim. These relationships are well seen in **Figure 8.11**. Because the excavation is toward the subpulmonary outlet, the conduction tissues tend to be more remote from the septal crest and the left-hand border, being carried on the left ventricular aspect of the septum. The same basic guides for perimembranous defects still apply.

Defects with Aortic to Pulmonary Fibrous Continuity

The characteristic feature of these defects is the formation of the roof by fibrous contiguity between the leaflets of the aortic and pulmonary valves. The defects are both subaortic and subpulmonary. Hence, they are called doubly-committed and juxta-arterial defects. As in muscular outlet defects (to be described), the membranous septum can be intact. When intact, it is separated from the margin of the defect by a muscular bar composed of the right margin of the ventriculo-infundibular fold and the postero-inferior limb of the trabecula septomarginalis. This is shown in **Figure 8.12**, in which a juxta-arterial defect is viewed from the apex of the right ventricle. The outlet septum is entirely deficient in these hearts, as is the septal component of the subpulmonary infundibulum. Because of this, the defect is located between the ventricular outflow tracts, as seen in **Figure 8.12b**, in which the defect is viewed from the left ventricle. This type of defect is frequently termed a supracristal defect but, because of the confusion surrounding the term ''crista'' (Anderson *et al*., 1977), we believe that the adjectives ''doubly-committed'' and ''juxta-arterial'' provide more accurate and less confusing appellations. For complete precision, nonetheless, the defects are best described on the basis of continuity between the leaflets of the aortic and pulmonary valves. In these defects, when the membranous septum is intact and is buttressed cephalically by the trabecula septomarginalis and the ventriculo-infundibular fold, the conduction tissues are distant from the edge of the defect, and are not at risk during repair. Because of the absence of the muscular outlet septum, the facing leaflets of the aortic valve are relatively unsupported. Prolapse of the aortic valve, with accompanying incompetence, is consequently a common feature. In other examples, however, the margins of doubly-committed and juxta-arterial defects can extend to be formed by fibrous continuity with the tricuspid valve. In this setting, the defects are also perimembranous. The leaflets of the aortic valve are still liable to prolapse with this combination, but the conduction tissue is at greater risk surgically because it

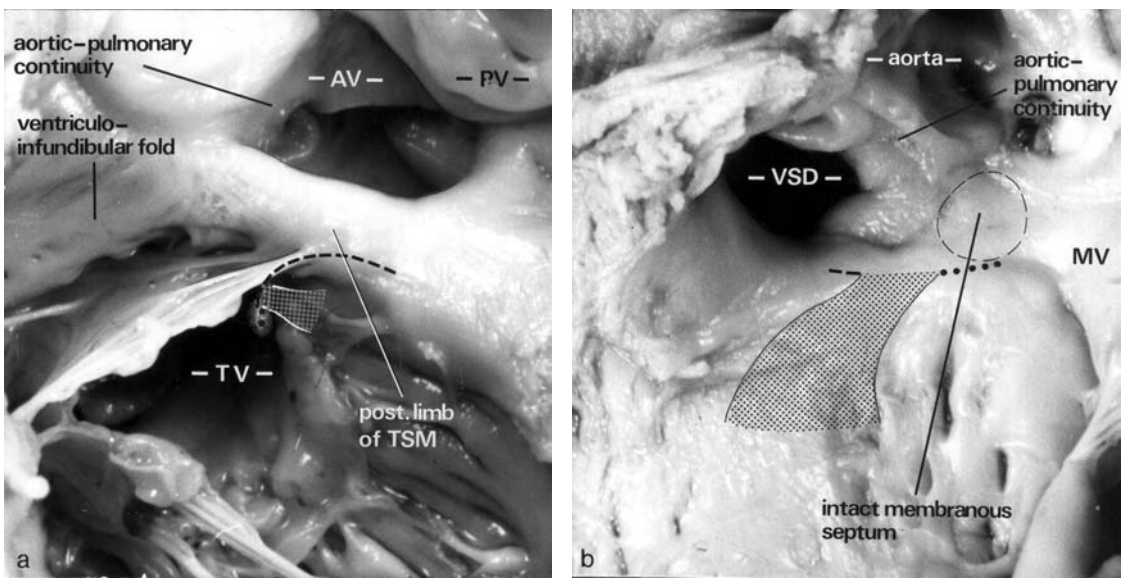

Figure 8.12

is unprotected by the buttressing bar. The rules are as described earlier for perimembranous defects.

Defects with Entirely Muscular Rims

Muscular defects can exist anywhere within the muscular septum, are frequently multiple and can coexist with defects whose rims are formed in part by the fibrous tissue of valvar leaflets or the central fibrous body. Their classification and morphology are simplified by considering their opening into the inlet, apical trabecular or outlet components of the right ventricle.

Muscular Defects Opening to the Inlet of the Right Ventricle

Muscular defects have different relationships to the atrio-ventricular conduction axis. The key to differentiation of the muscular inlet defect is that a rim of muscular tissue on its atrial aspect separates the margin of the defect from the attachments of the leaflets of the tricuspid, mitral and aortic valves. This is shown, as seen from the left ventricle, in **Figure 8.13b**. The defect is overlaid on its right ventricular aspect by the septal leaflet of the tricuspid valve, as shown in **Figure 8.13a**, which is viewed from the right atrium. The precise degree of overhang of the tricuspid valve depends on the position of the defect in relation to the inlet of the right ventricle. Because the postero-inferior limb of the trabecula septomarginalis still reaches to the apex of the triangle of Koch above the defect, the atrio-ventricular conduction axis penetrates into the ventricles on the outlet aspect of the defect. Thus, when viewed in surgical orientation from the right atrium, the conduction axis is seen to the left hand of the surgeon. This is in contrast to its constant position to the right hand in all types of perimembranous defects (**Figure 8.14**). Defects with entirely muscular rims that open to the inlet of the right ventricle, therefore, must be identified and distinguished from perimembranous defects that similarly open to the inlet of the right ventricle.

Muscular Defects Opening to the Apical Trabecular Component

These defects can exist at any place within the apical trabecular septum, are often well out toward the apex, but are usually positioned to one or other side of the trabecula septomarginalis. Smaller defects within the apical trabecular septum are frequently multiple and, when many are present, they produce the so-called ''Swiss-cheese septum''. The orifices of such defects, and indeed of most muscular defects within the apical trabecular septum, are better visualized from their left ventricular aspects. Indeed, it is often the case that visualization from the left side reveals a solitary opening, whereas it seems there are multiple holes as viewed from the right because the defect is crossed by right ventricular trabeculations. It has been suggested that these defects open to the infundibulum of the right ventricle (Kumar *et al*., 1997), but this is a confusing suggestion, since the defects are unequivocally between the apical components

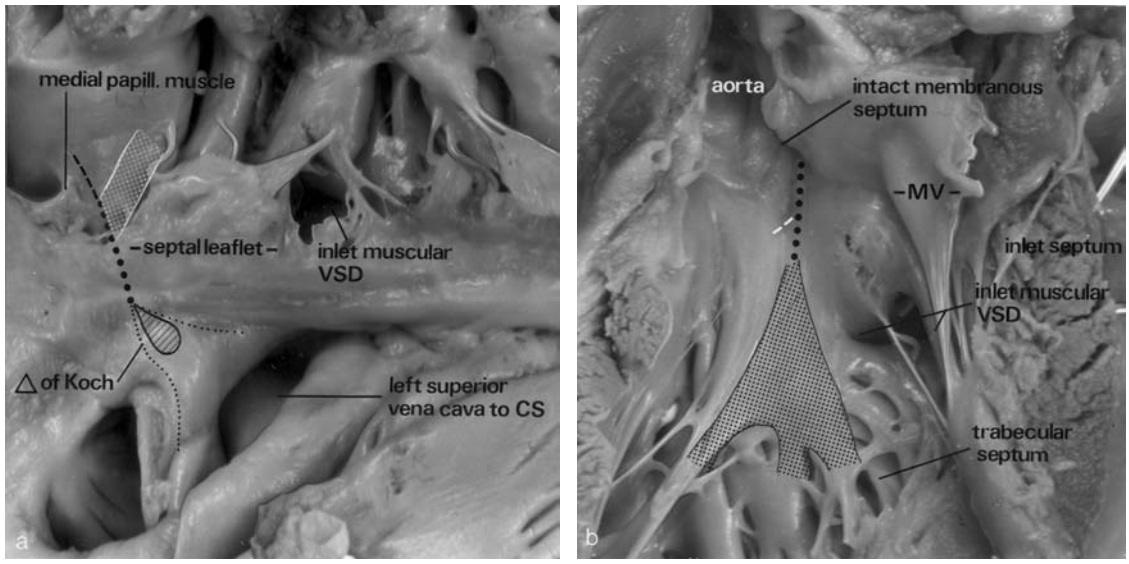

Figure 8.13

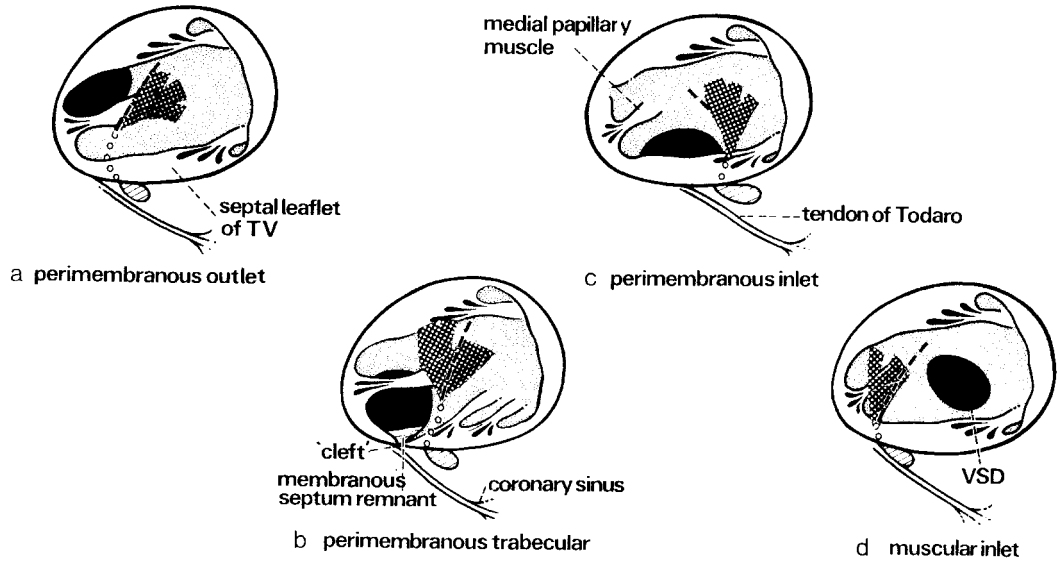

Figure 8.14

of the two ventricles (Tsang *et al*., 2002). Because the defects are beneath the crest of the muscular septum, they have no direct relationship to the branching or penetrating segments of the atrio-ventricular conduction axis. The fascicles of the bundle branches, in contrast, are frequently related to the margins and may form new branching portions on the septal rim (Latham and Anderson, 1972).

Muscular Defects Opening to the Outlet of the Right Ventricle

These defects have the same basic morphology as the doubly committed juxta-arterial type of defect with a muscular postero-inferior rim, except that the arterial valves are separated from each other in the roof of the defect by a remnant of the muscular outlet septum (**Figure 8.15**). The conduction tissue is protected from

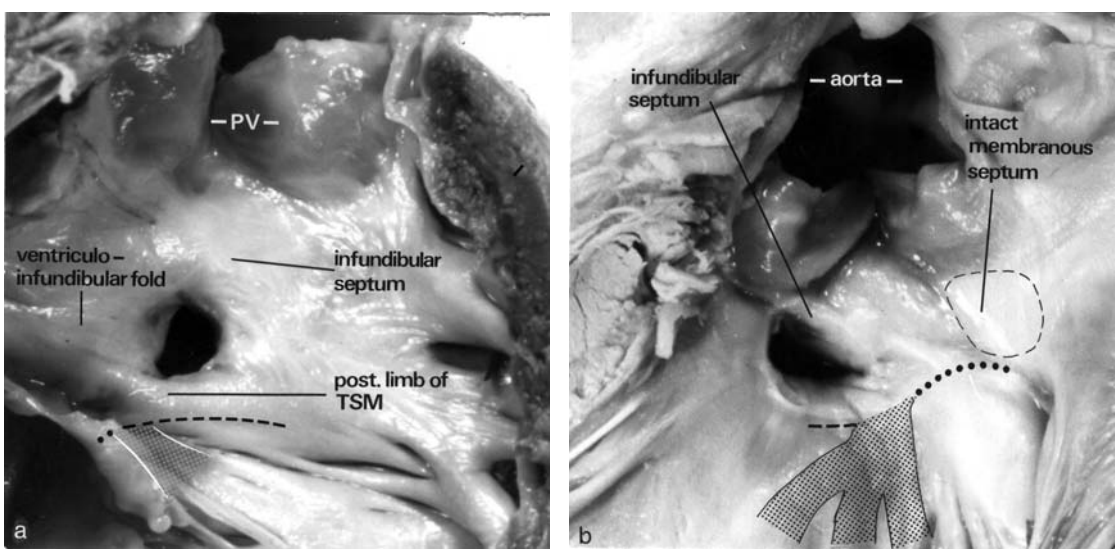

Figure 8.15

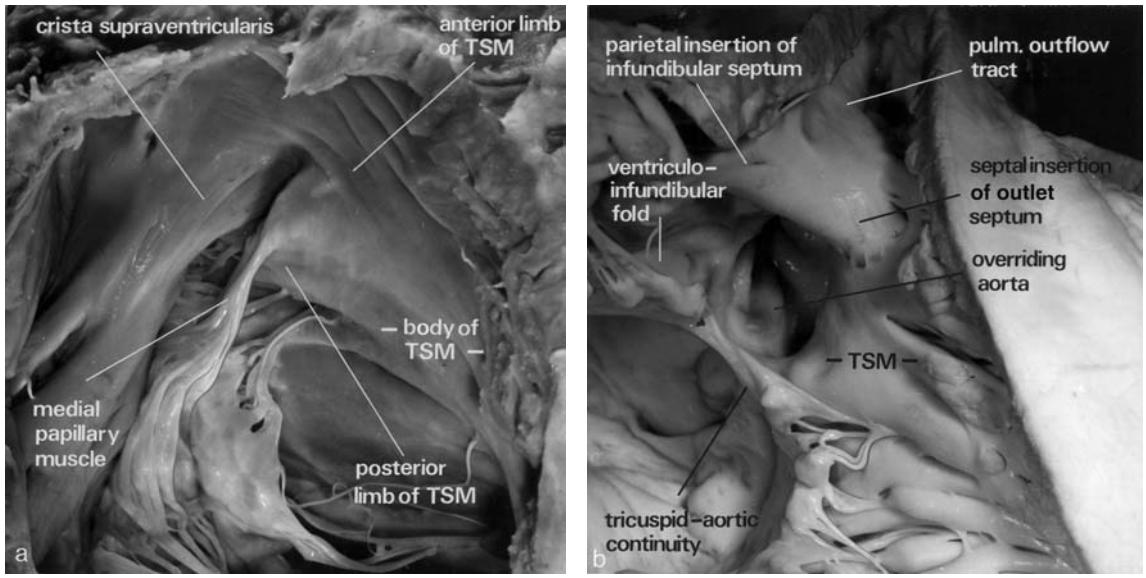

Figure 8.16

the edge of the defect by the muscle bar formed by fusion of the ventriculo-infundibular fold and the postero-inferior limb of the trabecula septomarginalis.

ANATOMY OF THE VENTRICULAR SEPTAL DEFECT IN TETRALOGY OF FALLOT

Although the various types of ventricular septal defects found in tetralogy are comparable with isolated perimembranous, muscular or juxta-arterial defects that open to the outlet of the right ventricle, tetralogy is such a well-defined entity that the defects warrant separate consideration. We define ''tetralogy'' as the combination of a ventricular septal defect, subpulmonary infundibular stenosis, aortic overriding and right ventricular hypertrophy. The essence of the anomaly is cephalad insertion of the septal insertion of the outlet, or infundibular, septum in relation to the trabecula septomarginalis, combined with hypertrophy of the septoparietal trabeculations. These two lesions combine to produce a ''squeeze'' at the mouth of the subpulmonary infundibulum. The muscular outlet septum is barely formed in the normal heart, being wedged between the most septal component of the supraventricular crest and the limbs of the trabecula septomarginalis. In tetralogy, it is a more extensive structure, and is inserted cephalad to the anterocephalad limb of the trabeculation. At one stroke, this cephalad insertion produces the ventricular septal defect, narrows the subpulmonary outflow tract, and brings the aortic valve above the right ventricle. This deviation in anatomy is demonstrated in **Figure 8.16b**, as viewed from the apex of the right ventricle. **Figure 8.16a** shows the same area in the normal heart. In most cases of tetralogy, part of the margin of the ventricular septal defect is formed by fibrous continuity between the leaflets of the aortic and tricuspid valves. Because of this feature, the defect is appropriately considered to be perimembranous. The anatomy as viewed by the surgeon through an infundibulotomy is illustrated in **Figure 8.17**. As with other perimembranous defects opening to the outlet

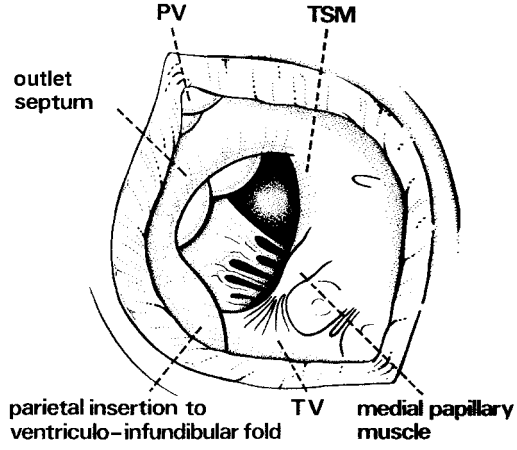

Figure 8.17

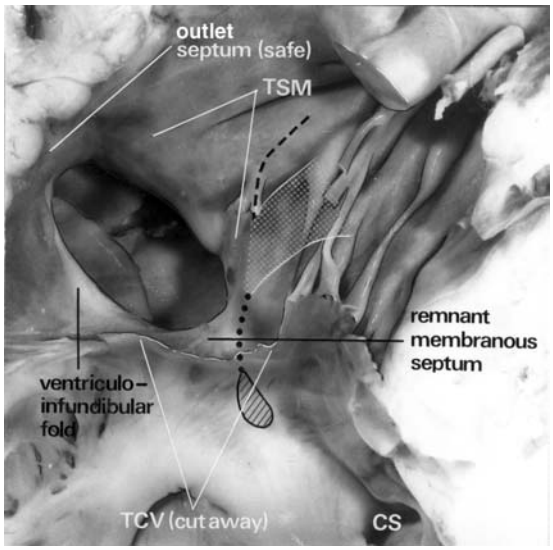

Figure 8.18

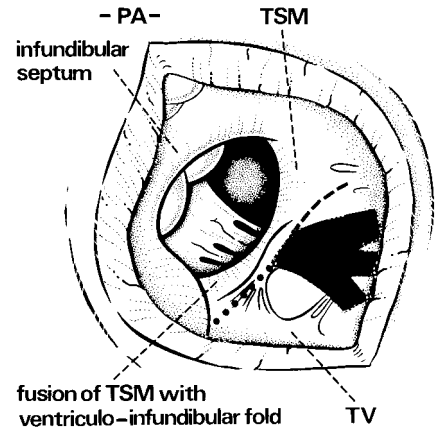

Figure 8.19

of the right ventricle, the conduction axis is usually disposed some distance away from the crest of the septum, being carried on its left ventricular aspect. The major danger area in repair of this type of defect is the area of continuity between the leaflets of the aortic and tricuspid valves hidden beneath the septal leaflets. This region is demonstrated in **Figure 8.18**. In the heart shown, the septal leaflet of the tricuspid valve has been cut back to its origin from the atrio-ventricular junction, showing the relationships of the defect to the right ventricle and the right atrium. The positions of the conduction tissues are marked, showing how the ventriculo-infundibular fold, the outlet septum, and the crest of the trabecular septum are all safe areas. The ''danger area'' is the right-hand corner when viewed from the right atrium. This is where the penetrating atrio-ventricular bundle passes from the atrial septum at the apex of the triangle of Koch to the subaortic aspect of the ventricular septum. There is frequently a remnant of the interventricular component of the membranous septum in this vulnerable corner. Again, the rule is that the septal remnant may conceal the conduction tissue, but the septal leaflet of the tricuspid valve never does. In about one-fifth of patients with tetralogy, the postero-inferior limb of the trabecula septomarginalis fuses with the ventriculo-infundibular fold across the atrial margin of the defect, producing a completely muscular rim to the defect. As with isolated muscular defects opening to the outlet of the right ventricle, this fusion of the trabecula septomarginalis with the ventriculo-infundibular fold buttresses the conduction tissue from the margin of the defects, making the entire muscular border a safe structure (**Figure 8.19**). In occasional hearts with aortic overriding and subpulmonary stenosis, the outlet septum may be lacking, so that the ventricular septal defect extends to become doubly-committed and juxta-arterial. With such a defect, the postero-inferior limb of the trabecula septomarginalis may again fuse with the ventriculo-infundibular fold, producing a completely muscular postero-inferior margin, or the defect may extend to come into contact with the continuous leaflets of the aortic and tricuspid valves. These defects are variants of tetralogy, and are common in the Far East (Ando, 1974) and South America (Neirotti *et al*., 1978). The position of the conduction tissue depends on whether the defect is bordered by aortic–tricuspid continuity, or whether the ventriculo-infundibular fold and trabecula septomarginalis fuse to protect the penetrating atrio-ventricular conduction axis.

VENTRICULAR SEPTAL DEFECTS IN HEARTS WITH CONCORDANT ATRIO-VENTRICULAR AND DISCORDANT VENTRICULO-ARTERIAL CONNECTIONS

The morphological features of septal defects in transposition are, broadly speaking, comparable with those of the isolated defects seen in hearts with concordant connections. Thus, various forms of perimembranous defects, including the defect extending toward the crux and opening to the inlet of the right ventricle, and muscular defects opening to inlet and trabecular components of the right ventricle, are all found in hearts with transposition (**Figure 8.20**). The significant difference from isolated defects is that, in transposition, because of the discordant ventriculo-arterial connections, it is the fibrous continuity between the leaflets of

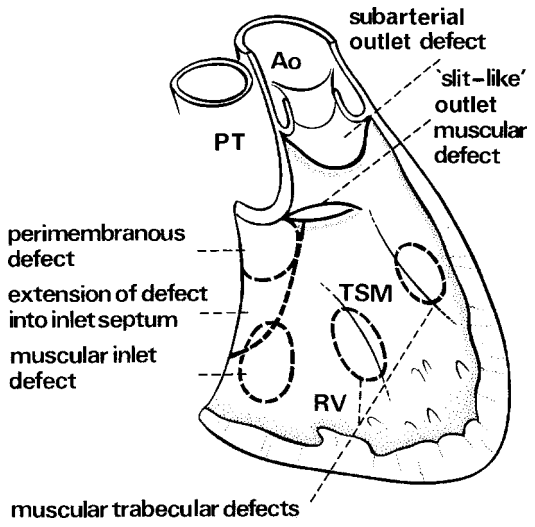

Figure 8.20

the tricuspid and pulmonary valves that forms part of the margin of the defect. The diagram shows the various defects superimposed on an outline of the right ventricle. Both muscular and doubly-committed juxta-arterial defects are shown opening to the right ventricular outlet, although it is highly unlikely that these defects could coexist. The conduction tissues are related to these various holes as they are to similar holes in hearts with concordant atrio-ventricular and ventriculo-arterial connections, these relationships having been described already in the section entitled ''Basic Rules Regarding Disposition of the Conduction System''. The important difference in the morphology of the communications is the high incidence of defects opening to the outlet of the right ventricle. As shown in **Figures 8.21a**, which is viewed from the right ventricular apex, and **8.21b**, seen from the left ventricle, these communications result from gross malalignment between the outlet septum and the muscular part of the ventricular septum, the latter reinforced by the trabecula septomarginalis. Indeed, in these defects, as with tetralogy of Fallot, the outlet septum is exclusively a right ventricular structure. In many defects of this type, the outlet and muscular ventricular septa are fused toward the atria and, in some hearts, an intact membranous septum is to be found posterior to this muscular bar. In other hearts, however, the defect may extend to an area of fibrous continuity between the pulmonary, mitral and tricuspid valves. This means that the defects have become perimembranous (**Figure 8.22**). Regardless of whether the defect is perimembranous or not, the important feature of these defects characterized by septal malalignment is that their margins are contiguous posteriorly, but diverge towards the anterior surface of the heart. Thus, the gap between the septal structures increases toward the parietal aspect of the subaortic infundibulum. Another important feature of these defects is that the tension apparatus of the tricuspid valve frequently crosses the right ventricular aspect of the defect to attach anomalously to the

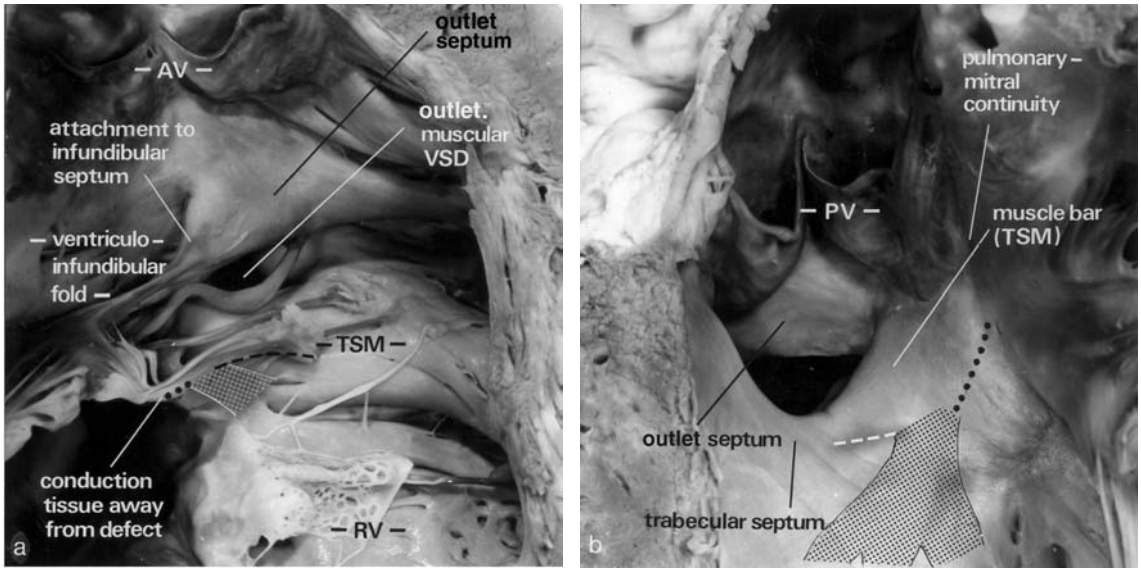

Figure 8.21

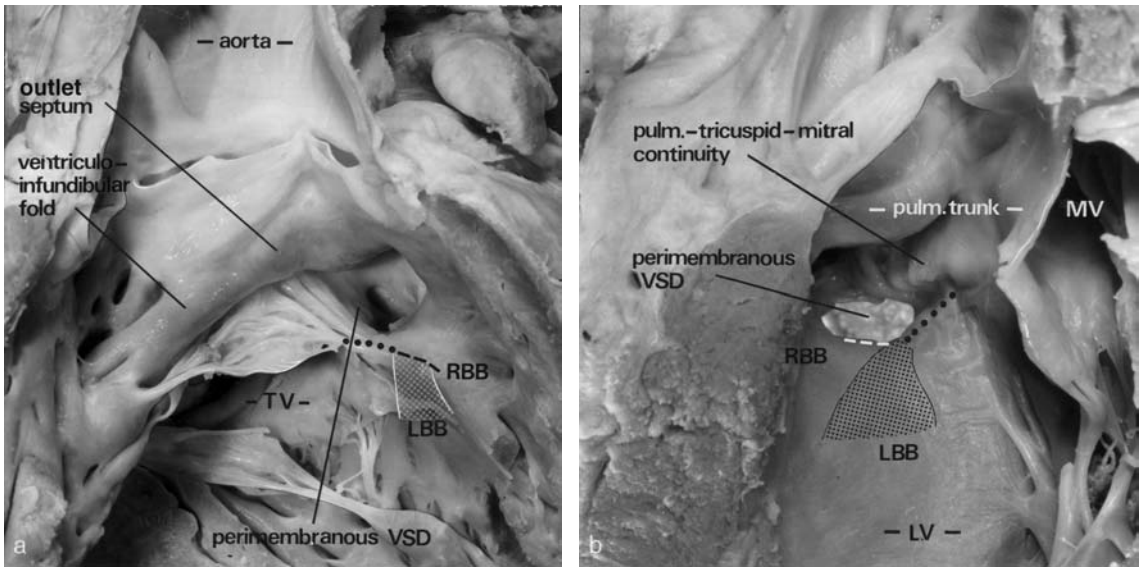

Figure 8.22

muscular outlet septum (**Figure 8.21**). These features are of major surgical significance, because failure to close the diverging antero-superior aspect may result in residual defects, and damage to the tension apparatus of the tricuspid valve can produce tricuspid regurgitation. From the standpoint of the atrio-ventricular conduction axis, the presence of the muscular rim in the postero-inferior position keeps the branching and non-branching segments of the axis away from the septal crest. This is shown in **Figure 8.21b**, in which the approximate position is marked on the left ventricular aspect of the septum. Before assuming the conduction tissue to be safe, it is important to ascertain that the postero-inferior rim of a defect with malalignment of the outlet septum is indeed a muscular structure. When the defect is perimembranous, it is possible for the penetrating and branching components of the bundle to be directly related to its postero-inferior edge (**Figure 8.22**). In the defects with malalignment of the outlet septum, be they perimembranous or muscular, the outlet septum is usually inserted anteriorly into the right ventricle. If it is inserted into the subpulmonary outflow tract from the left ventricle, then there is pulmonary obstruction.

THE INTERVENTRICULAR COMMUNICATION IN DOUBLE-OUTLET VENTRICLE

In hearts with double-outlet ventriculo-arterial connection, the hole between the ventricles forms the only egress for one of them. This hole, however, is not the space around which the surgeon secures a patch. It is important to appreciate this fundamental difference between interventricular communications in hearts in which each ventricle gives rise to a separate arterial trunk, and those in which there is a double-outlet ventriculo-arterial connection. The crucial feature in surgical repair is whether it is possible to construct a patch so as to connect the interventricular communication in unobstructed manner to the outflow tract of one or other great artery. It is the margins to which this patch must be secured that we emphasize in this section. When describing the position of the communication, nonetheless, it is convenient and useful for surgeons to describe the hole in relation to the great arteries, remembering that the relationships of the great arteries themselves can vary considerably. Thus, in simple terms, the interventricular communication can be considered related to the posterior, or right-sided, great artery, to the anterior, or left-sided, great artery, to both great arteries, making it doubly-committed, or to neither great artery, leaving it non-committed. This concept was promulgated by Lev *et al*. (1972) for the most common relationship of the great arteries, i.e. with the aorta being to the right of, and usually posterior to, the pulmonary trunk. It holds equally well for hearts in which the aorta is to the left of and anterior to the pulmonary trunk. In these hearts, the morphology of the subaortic defect is virtually identical to that of the subpulmonary defect found in double-outlet right ventricle with more usual relationships, the latter usually being called the Taussig–Bing heart. When this concept is combined with

considerations of whether the margins of the defect to be closed are perimembranous or muscular, a simple nosology is provided, which also gives important clues to the disposition of the conduction tissues. In hearts with the interventricular communication beneath the posterior and right-sided artery, the cephalad area of the margin to which the patch must be secured is formed by the outlet septum, which fuses with the rest of the muscular septum above the anterior limb of the trabecula septomarginalis. As with tetralogy, the outlet septum in hearts with double-outlet right ventricle is exclusively a right ventricular structure. The trabecula septomarginalis forms the crest of the septum. When it fuses with the ventriculo-infundibular fold to produce a muscular postero-inferior margin, then the bilateral infundibulum is usually also present. This morphological pattern is shown, as seen from the apex of the right ventricle, in **Figure 8.23a**. The atrio-ventricular conduction axis is buttressed from the edge of the defect, which consequently is a safe area. It must be remembered, however, that the presence of bilateral infundibula is not uniformly found in hearts with double-outlet ventriculo-arterial connection. Both arteries may arise in their entirety from the right ventricle, with fibrous continuity present between the posterior arterial valve and the leaflets of either the mitral or the tricuspid valve. In these circumstances, the defect is perimembranous, as illustrated in **Figure 8.24a,b**. The penetrating conduction axis may then be directly related to the fibrous tissue, as in other defects that are perimembranous. When the defect is beneath the anterior, or left-sided, great artery, be it the pulmonary trunk or the aorta, the outlet septum is usually fused with the ventricular septum in relation to the postero-inferior limb of the trabecula septomarginalis. In the presence of bilateral infundibula, both these structures fuse with the ventriculo-infundibular fold. The comparison between this morphology and that in a heart with a subaortic defect is seen in **Figure 8.25a,b**. The conduction tissues are usually remote from the edge of the defect, being most closely related to its postero-inferior rim (Bharati and Lev, 1976). In some instances, they may be directly related to the crest of the defect (Lincoln *et al*., 1975). When the defect is doubly-committed, the outlet septum is usually either very short or unattached to the trabecula septomarginalis, or else totally deficient. Because of this, the great arteries are free to override the septum. Such an arrangement is shown in **Figure 8.26**, in which the right ventricle has been opened through the pulmonary valve, and the aortic valvar orifice is seen posterior to an area of fibrous continuity between the leaflets of the aortic and pulmonary valves. Hearts with doubly-committed defects are frequently intermediate between double-outlet right and left ventricles, having been termed ‘‘double-outlet both ventricles’’ by Brandt *et al*. (1976). To the best of our knowledge, the conduction tissues in this anomaly have not been studied, but it is highly likely that the penetrating bundle is related to the postero-inferior rim and is probably not directly on the septal crest. The non-committed defects

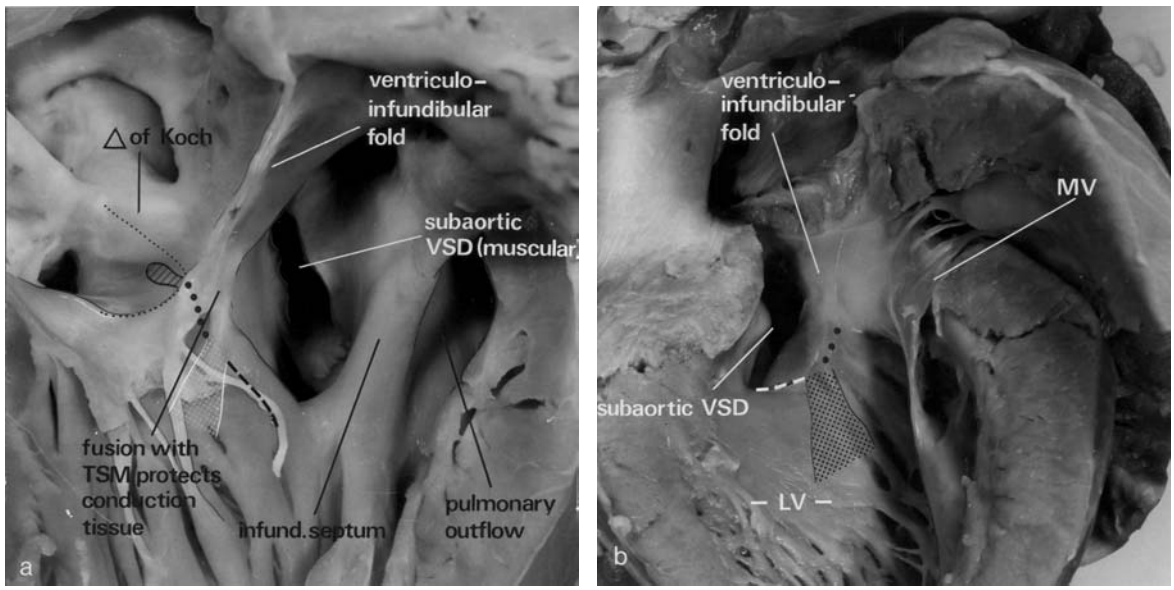

Figure 8.23

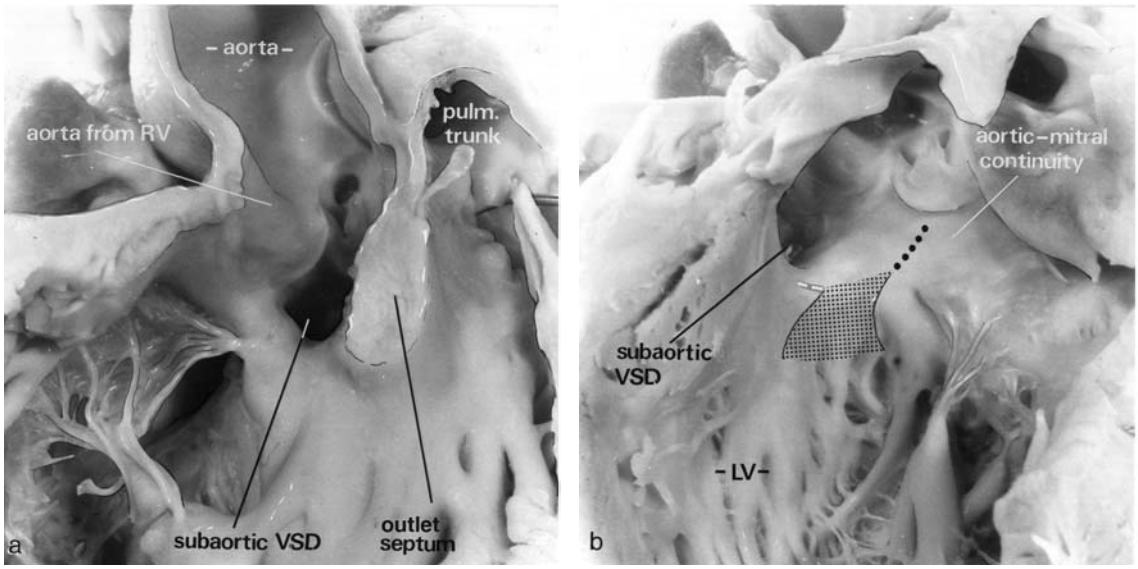

Figure 8.24

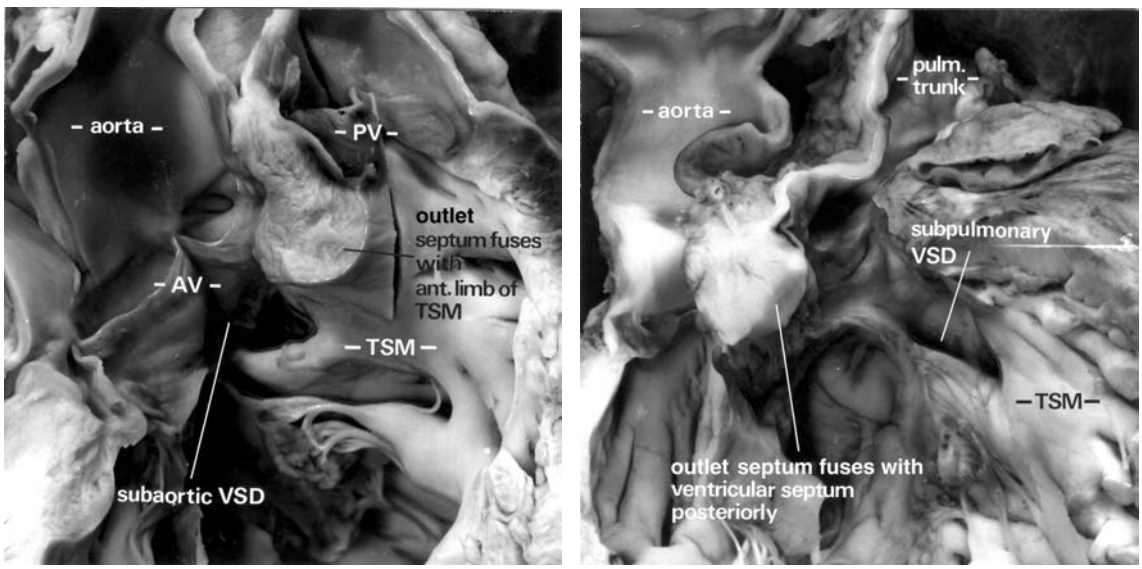

Figure 8.25

in double-outlet right ventricle can have variable morphology. Muscular defects opening to either the inlet or the apical trabecular components of the right ventricle would produce a communication distant from either great artery, and the conduction tissues would not be closely related. The precise orientation is like that found with isolated muscular defects. Perimembranous defects beneath the posterior great artery can also extend so far into the inlet component of the right ventricle that they are in a non-committed position. In this circumstance, the atrio-ventricular conduction axis is likely to be directly related to the postero-inferior rim of the defect. As more and more hearts with double outlet from the left ventricle are recognized, it becomes evident that the position of

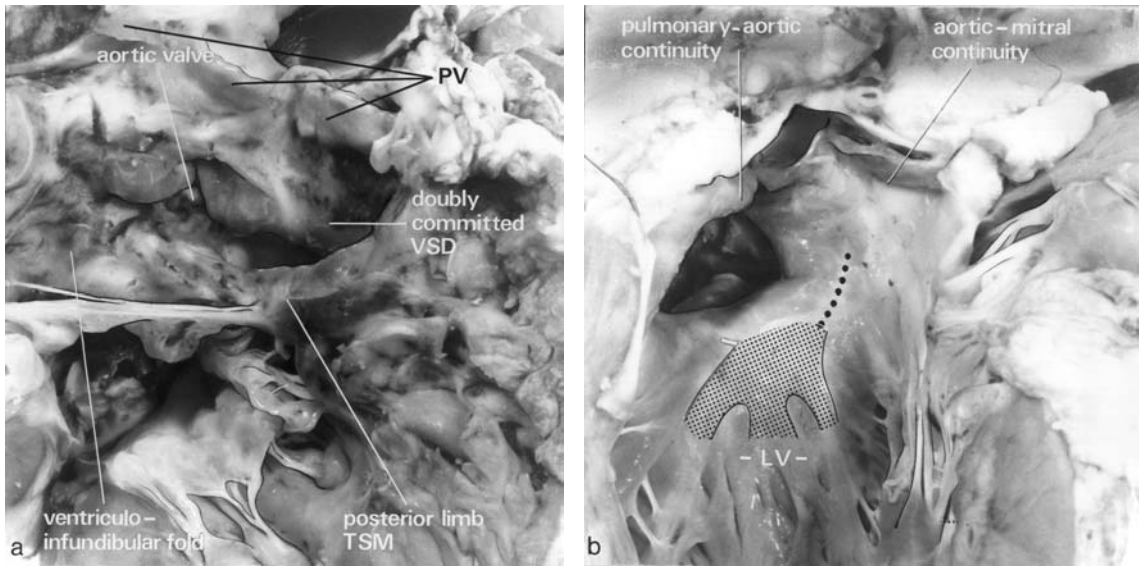

Figure 8.26

the interventricular communication and arterial relationships are just as variable as for double outlet from the right ventricle. It seems sensible, therefore, to use the same categorization as in double-outlet right ventricle. The guidelines for disposition of the conduction tissue are also the same.

VENTRICULAR SEPTAL DEFECTS IN SINGLE OUTLET OF THE HEART

Hearts with a single arterial outlet usually possess a ventricular septal defect. Its morphology varies, depending on the nature of the arterial vessel. Most usually this is a common arterial trunk. It is important to remember that, although truncal valves usually override the ventricular septum, as seen in **Figure 8.27**, the trunk may arise exclusively from either the left or the right ventricle. Enlargement of the ventricular septal defect, if needed, can be performed safely on the septal aspect adjacent to the anterosuperior ventricular wall. It would be dangerous to resect the cephalic aspect because this is the ventriculo-infundibular fold and, therefore, part of the wall of the heart. The conduction tissue is related to the inlet aspect of the defect, and is nearly always safe from damage, being buttressed by the continuity between the ventriculo-infundibular fold and trabecula septomarginalis, as in muscular or subarterial defects opening to the outlet of the right ventricle. This pattern is the most common variant with common arterial trunk. On occasion, nonetheless, the defect extends into perimembranous position, and then the conduction tissue may be closer to the edge of the defect, but it is usually on its left ventricular aspect, as in tetralogy of Fallot. When the single outlet from the heart is through the aorta with pulmonary atresia, the morphology of the ventricular septal defect varies, depending on the origin of the aorta. If the aorta is exclusively from the right ventricle, with the atretic pulmonary trunk in posterior position, the defect resembles those seen in transposition. In contrast, when the aorta straddles the septum, with the atretic pulmonary trunk positioned anteriorly and cephalad, the defect resembles that found in tetralogy of Fallot, usually being perimembranous but occasionally muscular. In the presence of single outlets that are due to aortic atresia with a patent pulmonary trunk, the ventricular septum is most frequently intact, this being the classical variant associated with hypoplasia of the left heart.

VENTRICULAR SEPTAL DEFECTS WITH DISCORDANT ATRIO-VENTRICULAR CONNECTIONS

The presence of discordant atrio-ventricular connections has a profound effect not only on the morphological characteristics of defects of the ventricular septum, but also particularly on the disposition of the conduction tissues. Discordant atrio-ventricular connections are most usually also associated with discordant connections at the ventriculo-arterial junctions. This combination is known as congenitally corrected transposition. The discordant atrio-ventricular connections, nonetheless,

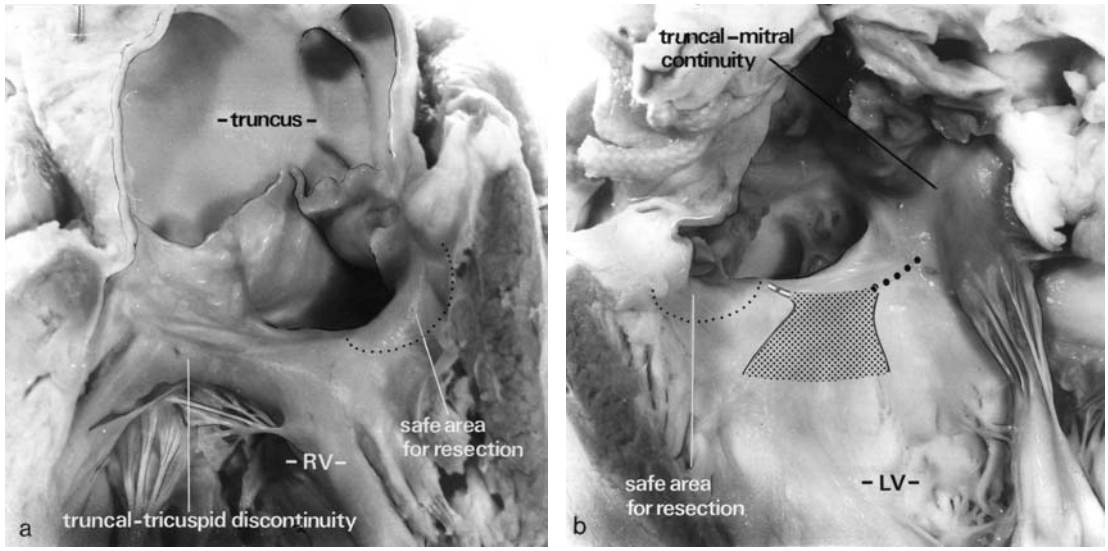

Figure 8.27

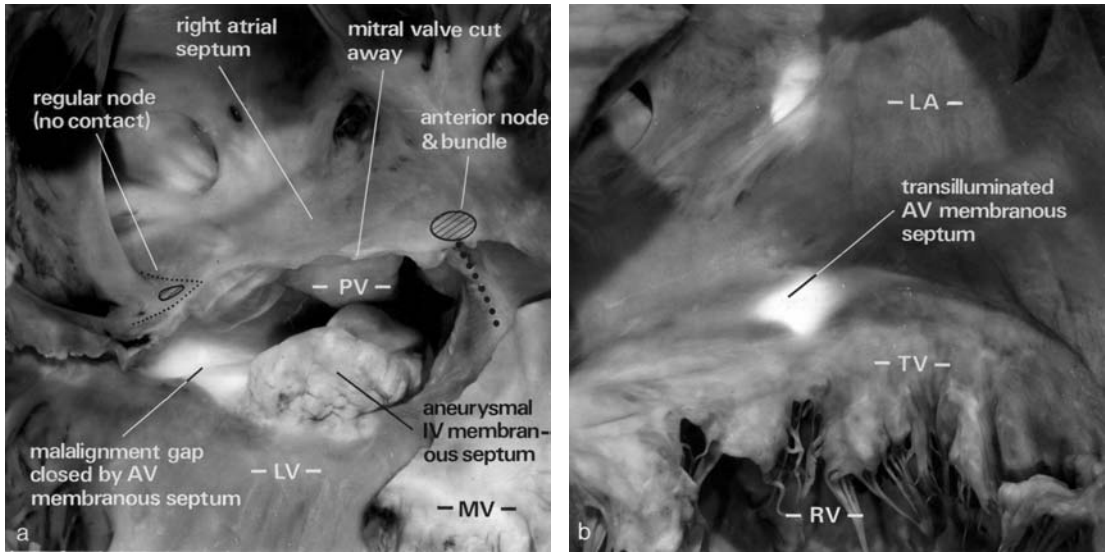

Figure 8.28

can also occur with double outlet from either the right or left ventricle, with single outlet of the heart or, in rare instances, with concordant ventriculo-arterial connections. Congenitally corrected transposition itself exists in two discrete forms, specifically with usual and mirror-imaged atrial situs. There are important differences between these anomalies. More than half the instances of congenitally corrected transposition with usual atrial situs exhibit defects of the ventricular septum (Allwork *et al*., 1976). The majority of these defects are wedged beneath the pulmonary outflow tract in a perimembranous position. The membranous septum itself, however, occupies a different position in hearts with discordant atrio-ventricular connections than in normal hearts. The major feature of hearts with discordant atrio-ventricular connections is malalignment between the atrial septum and the ventricular septum (**Figure 8.28a,b**). The two septal structures are contiguous at the crux but diverge as they are traced forward, the atrial septum veering

right-ward and the ventricular septum left-ward as viewed from the atrium. The pulmonary outflow tract is wedged into this gap (Becker and Anderson, 1978a, 1978b). The mitral valve forms its right border, and the membranous septum, when intact, forms its left border. The attachment of the tricuspid valve, a left-sided structure in corrected transposition, divides the intact membranous septum into an atrio-ventricular component between the left atrium and the pulmonary outflow tract from the morphologically left ventricle, and an interventricular component (**Figure 8.28**). Because of the malalignment, the interventricular component of the membranous septum is required to fill an exceptionally large area. When intact, it is frequently aneurysmal (**Figure 8.28a**). More usually, the membranous tissue fails to fill the gap, and the result is a perimembranous defect that excavates backwards so as to open between the ventricular inlets. Its roof is the remnant of the membranous septum, which is continuous with the central fibrous body and provides fibrous continuity between the leaflets of the pulmonary, mitral and tricuspid valves. The membranous septal remnant itself frequently becomes aneurysmal and contributes to subpulmonary obstruction (Anderson *et al*., 1974a), or else forms a diaphragm across the outflow tract, again resulting in subpulmonary obstruction. The latter arrangement is illustrated in **Figure 8.29**, which shows the diaphragmatic obstruction to the pulmonary outflow tract, as seen from the apex of the morphologically left ventricle. The position of the conduction tissues cephalad to the pulmonary outflow tract is marked.

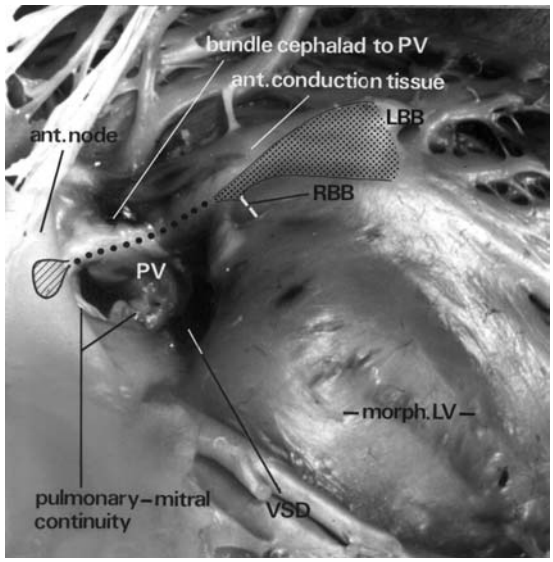

Figure 8.29

Because of its wedged position, the pulmonary outflow tract itself usually overrides the septum to some degree in the presence of a perimembranous defect. The major feature of this malalignment from the surgical standpoint is that it prevents establishment of a normal conduction system. Thus, the regular atrio-ventricular node in the atrial septum overlies the area filled by the fibrous membranous septum and is unable to make contact with the ventricular conduction tissues (Anderson *et al*., 1973; Becker and Anderson, 1978a, 1978b). Because the left ventricle is to the right of the right ventricle in the setting of left-hand ventricular topology, the bundle branches are arranged in a mirror-imaged manner. The atrio-ventricular bundle descends anteriorly down the muscular ventricular septum from the atrio-ventricular junction. The position taken by the conduction tissues has been superimposed on **Figures 8.28a** and **8.29**, and the gap between atrial and ventricular septal structures is well seen in these photographs. The anterior position of the non-branching bundle in relation to the pulmonary outflow tract is illustrated in **Figure 8.30**, and its position anterior to a small septal defect, as well as its relationship to a subpulmonary diaphragm, is shown in **Figure 8.29**. The view of this arrangement, as seen by the surgeon through a ventriculotomy in the morphologically left ventricle, is illustrated in **Figure 8.30**. The important point is that the descending course of the non-branching bundle from the anterior node takes it cephalad and anterior in relation to the pulmonary outflow tract. It reaches the morphologically left ventricle by penetrating through the lateral side of the area of continuity between the leaflets of the pulmonary and mitral valves, as seen in **Figure 8.28**. Thus when viewed from the morphologically left ventricle, the bundle descends

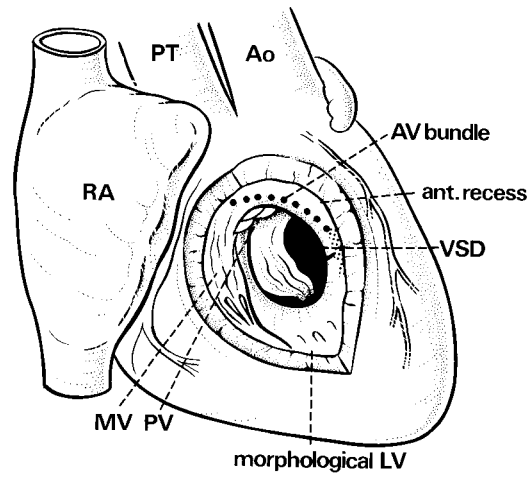

Figure 8.30

anteriorly and laterally to the pulmonary outflow tract (**Figure 8.30**). When a perimembranous defect is present, therefore, the bundle descends from in front of the pulmonary valve onto the anterior or cephalad edge of the defect. The branching bundle, and the left bundle branch, tend to occupy the morphologically left ventricular side of the septum, but the non-branching bundle, a long structure in congenitally corrected transposition, can occupy a subendocardial position on the cephalad edge of the defect. Both the anterior and posterior nodes have formed penetrating bundles in some hearts that we have studied. In these hearts, a sling of conduction tissue is formed around the edge of the ventricular septal defect (Symons *et al*., 1977). A clue to the presence of a sling is better alignment between the atrial and ventricular septal structures, a prerequisite for formation of a normal conduction system. It is easy to see the conduction tissues in fixed specimens. Whether this is so in the operating room remains to be established. Perimembranous defects are also to be found when congenitally corrected transposition coexists with mirror-imaged atrial situs. The pathologist less frequently sees these defects than those with congenitally corrected transposition and usual atrial situs. In most of those studied thus far to elucidate the course of the conduction tissues, these have been shown to arise from a normally situated regular node (Thiene *et al*., 1977; Dick *et al*., 1977; Wilkinson *et al*., 1978). It has recently been suggested that this is because all the studied cases have either pulmonary atresia or severe pulmonary stenosis, these associated lesions removing the malalignment gap and therefore promoting better septal alignment (Hosseinpour *et al*., 2004). In the presence of malalignment, therefore, the possibility still exists for formation of an anterior connection in the setting of congenitally corrected transposition with mirror-imaged atrial situs. The second most common type of defect in congenitally corrected transposition in either type of situs is the juxta-arterial and doubly-committed defect. As with doubly-committed defects in hearts with concordant atrio-ventricular connections, the defects seem more common in the Far East than in Europe and America. They constituted a large proportion of the cases described by Okamura and Konno (1973), and yet were rare in the series that we studied (Allwork *et al*., 1976). The defects are comparable with isolated juxta-arterial and doubly-committed defects in that the conjoined leaflets of the arterial valves roof them. The difference from the isolated defect is that, in the juxta-arterial defect with discordant atrio-ventricular connections, the trabecula septomarginalis is to the left of the septum when there is usual atrial situs, since it is a morphologically right ventricular structure. There is no reason why muscular defects of any type should not be found in congenitally corrected transposition, but they are certainly much rarer than the perimembranous and juxta-arterial and doubly-committed defects described earlier. Because of the likely anterior position of the conduction tissues, muscular defects are likely to be distant from the non-branching bundle, although they may well be related to the more distal bundle branches. After congenitally corrected transposition, and except for pulmonary atresia in which the defects are very much as in the heart with discordant ventriculo-arterial connections apart from the fact that the subpulmonary outflow is atretic, the next most common ventriculo-arterial connection with discordant atrio-ventricular connections is double outlet from the morphologically right ventricle. As in double-outlet right ventricle with concordant atrio-ventricular connections, any arterial interrelationship is possible in this segmental combination. The interventricular communication, forming the only egress from the right-sided morphologically left ventricle, may be subaortic, subpulmonary, doubly-committed or non-committed. In practice, this is a rare malformation, and most patients with usual atrial situs exhibit an anterior left-sided aorta and a subpulmonary ventricular septal defect. Many such hearts also exhibit subpulmonary stenosis and are in the right side of the chest. This constellation of anomalies has been termed the ''Mayo syndrome''. Monckeberg described such a case in 1913 and also had the prescience to study the conduction system. He demonstrated presence of anterior and posterior nodes and a sling of conduction tissue encircling the subpulmonary defect, again reflecting the better septal alignment because of the double outlet from the morphologically right ventricle. Establishment of the presence or absence of a sling is important because its presence effectively precludes enlargement of the septal defect. Double outlet from the left ventricle, or concordant ventriculo-arterial connections, can also accompany discordant atrio-ventricular connections (Brandt *et al*., 1976; de Leval *et al*., 1979). They are rare anomalies and, as far as we know, the conduction tissues have not been studied in such examples.

HEARTS WITH STRADDLING ATRIO-VENTRICULAR VALVES

Because it has both tension apparatus and an annulus, an atrio-ventricular valve can either straddle, when the tension apparatus is attached to both sides of the muscular ventricular septum, or override, when the atrio-ventricular junction is connected within both ventricles. Usually, valves both straddle and override, but on occasion a valve can override without straddling, or straddle without overriding. Overriding in isolation is not a major surgical problem, but straddling, with or without

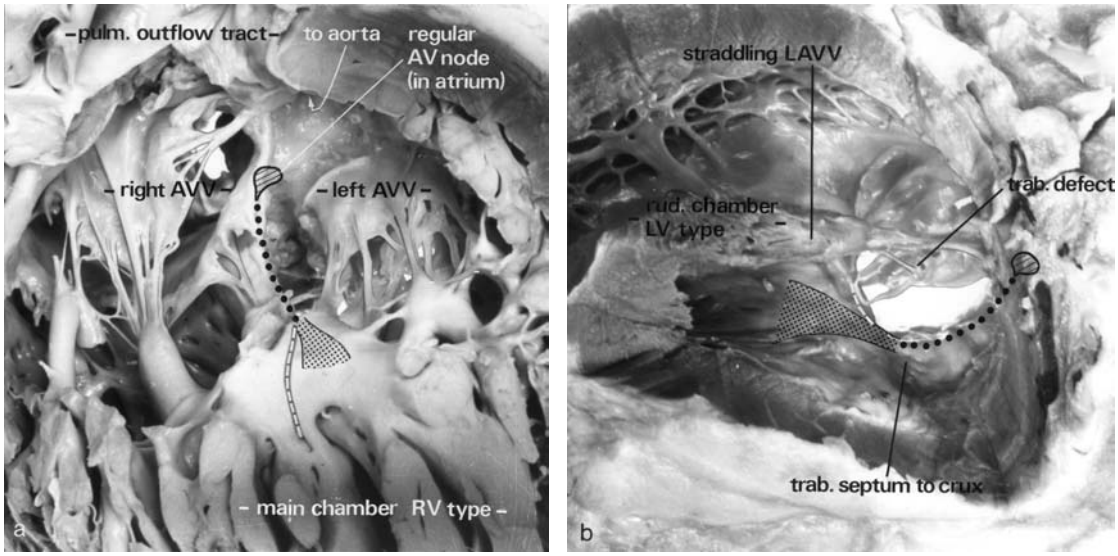

Figure 8.31

overriding, certainly is (Tabry *et al*., 1979). We conclude our reviews of ventricular septal defects, therefore, by considering the morphology of defects associated with straddling atrio-ventricular valves.

Straddling Left Atrio-ventricular Valves

A straddling left atrio-ventricular valve can be found in two distinct situations. The first is when the valve is of mitral structure, and is connected to a left-sided morphologically left ventricle and also to a right-sided morphologically right ventricle. This arrangement is basically that of concordant atrio-ventricular connections, but when the straddling valve is committed mostly to the ventricle receiving the other valve, then there is effective double-inlet right ventricle with left-sided rudimentary left ventricle, as illustrated in **Figure 8.31**. The second situation is when the valve is of tricuspid morphology, and is connected to a left-sided morphologically right ventricle and also to a right-sided morphologically left ventricle. This arrangement is basically that of discordant atrio-ventricular connections, and the conduction tissue of such a heart is illustrated in **Figure 8.32**. When the valve is committed mainly to the morphologically left ventricle, the arrangement is more akin to double-inlet left ventricle with left-sided rudimentary right ventricle (Milo *et al*., 1979). The important difference between the two types is that, in the hearts with the basic arrangement of concordant atrio-ventricular connections, the mitral valve straddles the anterior part of the septum. This is seen most frequently with discordant or double-outlet ventriculo-arterial connections with subpulmonary

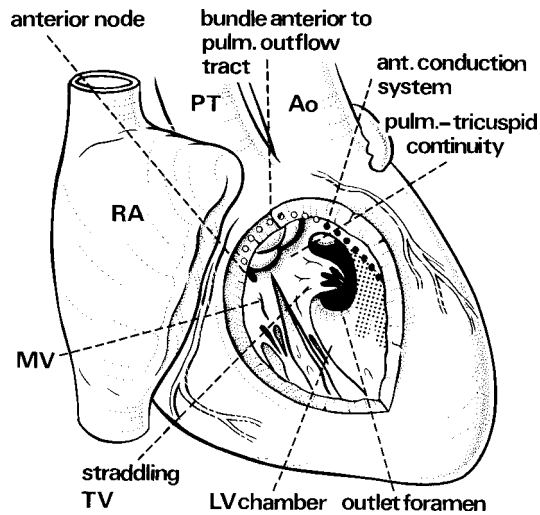

Figure 8.32

defect, the so-called ''Taussig–Bing malformation''. The association was first emphasized by Kitamura *et al*. (1974), and confirmed by Muster *et al*. (1979). The conduction tissues are in their usual position (Milo *et al*., 1979), as described in this chapter for transposition and double-outlet right ventricle. In the hearts with the basic arrangement of discordant atrio-ventricular connections, in contrast, the tricuspid valve straddles the posterior part of a septum that does not reach the crux. Because of this, the conduction tissue originates from an anomalous anterior node. The distribution is then as

would be found in congenitally corrected transposition (**Figure 8.30**), or as in double-inlet left ventricle with left-sided rudimentary right ventricle (Anderson *et al*., 1974b).

Straddling Right Atrio-ventricular Valve

As with straddling of the left valve, this condition can also exist in two basic forms, each of which may produce basically univentricular or biventricular atrio-ventricular connections, depending on the degree of override of the atrio-ventricular junction. The first represents a spectrum between concordant atrio-ventricular connections with straddling tricuspid valve, and double-inlet left ventricle with right-sided rudimentary right ventricle and straddling right valve. The straddling valve overrides the posterior part of a septum that does not extend to the crux. Because of this, the conduction tissue cannot originate from the regular node. Instead, an anomalous node is formed posterolaterally at the point where the septum reaches the atrio-ventricular junction. **Figure 8.33a** shows the view from the dominant left ventricle. There is, basically, a double inlet. Because of this, the muscular ventricular septum carrying the ventricular conduction tissues is separated from the atrial septum carrying the regular node. The view of the straddling valve as seen from the atrium is illustrated in **Figure 8.33b**. Most of the atrio-ventricular junction is committed to the dominant left ventricle. Also, the septum reaches the atrio-ventricular junction posteriorly, but a considerable distance from the crux. This is not shown in the figure, but its position is illustrated by the arrow and the star. The connecting node, therefore, is not the regular node but an anomalous posterior node. The principle underlying formation of this node is illustrated diagrammatically in **Figure 8.3**. Because of this unusual position of the node, a different area of the junction is at risk for damage during repair of hearts with straddling tricuspid valves. It is also fallacious to describe the defect in these hearts as an ''atrio-ventricular canal defect'', first because the hearts do not possess a common atrio-ventricular junction, and second because the conduction axis is not disposed as in atrio-ventricular septal defects, except when there is malalignment between the atrial septum and the muscular ventricular septum (Ho *et al*., 1992). The second situation, in which a straddling right valve is found, is when the valve straddles the anterior part of a septum between a right-sided morphologically left ventricle and a left-sided morphologically right ventricle. The affected hearts form a spectrum between discordant atrio-ventricular connection with straddling mitral valve, and double-inlet right ventricle with right-sided rudimentary left ventricle and straddling right valve. Either the conduction tissues arise from an anterior node, or there is a sling around the margins of the septal defect (Essed *et al*., 1980).

Acknowledgements

Figure 8.33 was photographed by Dr L. H. S. Van Mierop of Gainesville, FL, USA, and is reproduced with

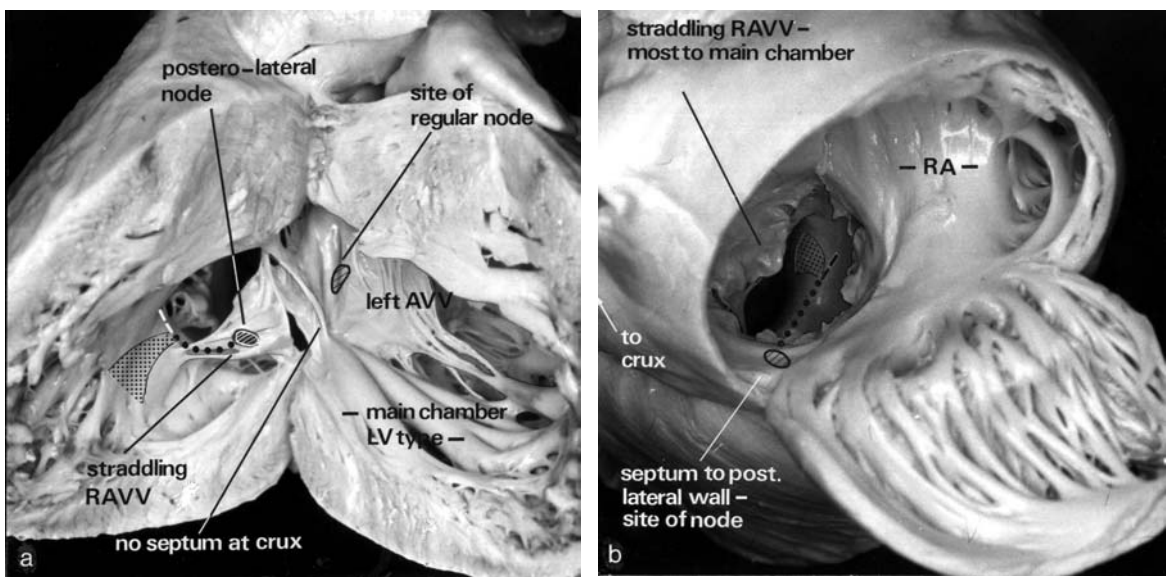

Figure 8.33

his permission. We are indebted to all our friends who have collaborated and contributed to the formulation of the concepts expressed herein, particularly James L. Wilkinson, Melbourne, Australia; the late Andre Moulaert, Utrecht, The Netherlands; Benigno Soto, Birmingham, AL, USA; Simcha Milo, Tel Aviv, Israel; Bob Van Mierop, Gainesville, FL, USA; and James R. Zuberbuhler, Pittsburgh, PA, USA. We also thank our other associates and friends who have worked with us on many of the studies quoted here. We are particularly grateful to Siew Yen Ho and Christine Anderson for their technical, photographic and artistic help in London, and to Ruud Verhoeven, Wilfried Meun, Rinus Klaver and Marscha Schenker for similar help in The Netherlands.

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9

Use of Pacemakers and Defibrillators in Children

P. G. Rees

In the late 1950s, C.W. Lillehei and colleagues devised a system of leaving at operation a wire attached to the ventricular epicardium and used it to pace the heart with a small external pacemaker system developed by Bakken (Weirich *et al*., 1958). In 1959, Elmquist and Senning described the first completely implantable pacing system involving the use of epicardial electrodes. At about that time, Furman and Robinson (1958) used endocardial leads introduced trans-venously. During the next few years these methods became widely used, although the systems paced the ventricle independent of the heart's cardiac action and were term asynchronous or fixed rate. The introduction of demand pacing with a sensing circuit allowed pacing only when there was no spontaneous heart action within a set time.

The development of the lithium iodide cell was a major advance. In addition, the circuitry and capacitors improved, becoming more reliable and complex, and were later replaced by microprocessors. This allowed the development of dual-chamber sensing and pacing in the early 1980s to maintain atrio-ventricular synchrony. In the mid-1980s sensor-driven rate-response pacing was introduced to allow increasing heart rate on exercise or emotional demand. Sensors include piezo-electric crystals, accelerometers, QT interval and impedance-derived minute volume, or a combination of two of these (**Figure 9.1**). This has been a significant advance for patients with sinus node disease, as the increased cardiac output on exercise very largely comes from increasing heart rate, rather than stroke volume. Numerous studies have confirmed that restoration of normal rate response on exercise is more important for improving exercise capacity than atrio-ventricular synchrony.

The increasingly complex circuitry allowed the pacemakers to be programmed externally, allowing alterations in many of their functions. Just a few examples are: filters to protect the pacemakers from external electrical interference; diagnostic features of monitoring the use of the pacemaker over time; arrhythmic events; on-line update assessment of pacing thresholds, which allows reduction in pacing output and therefore prolonged battery life. In the late 1990s the advent of biventricular pacing entered adult practice for the treatment of congestive cardiac failure by synchronization of left ventricular wall motion.

Alongside these advances there have been significant improvements in the technology of the leads that both deliver the output impulse from the generator to the myocardium and transfer the intracardiac electrograms from the myocardium to the sensing circuit. The leads have become thinner, with greater flexibility; the connector pins have reduced in size and are now standard at 3.2 mm; there have been improvements in the insulation with improved types of polyurethane; the small macro-surface electrodes allow for a lower threshold but with high micro-surface area, such as porous tip leads allows for better sensing. The development of steroid eluting leads, where a small amount of steroid is built into the tip, reduces the inflammatory response and keeps thresholds low and avoids exit block. Leads with a high pacing impedance have been produced, which will help battery life conservation.

Most implanted leads have a method of fixation. The passive type employ tines which catch under the trabeculae of the myocardium; the active type has a screw, often retractable, which is inserted into the myocardium (**Figure 9.2**). A new fine 4 French non-retractable screw electrode delivered with a long sheath has recently been introduced and may well be a significant advance on current bulkier leads. Temporary pacing leads and coronary sinus leads have no active fixation.

The lead may be *unipolar*, in which the tip is the cathode and the pacing box can is the anode, or *bipolar*, in which the anode is an additional electrode just proximal to the tip. Unipolar leads are smaller and more flexible and can be repaired; the stimulus

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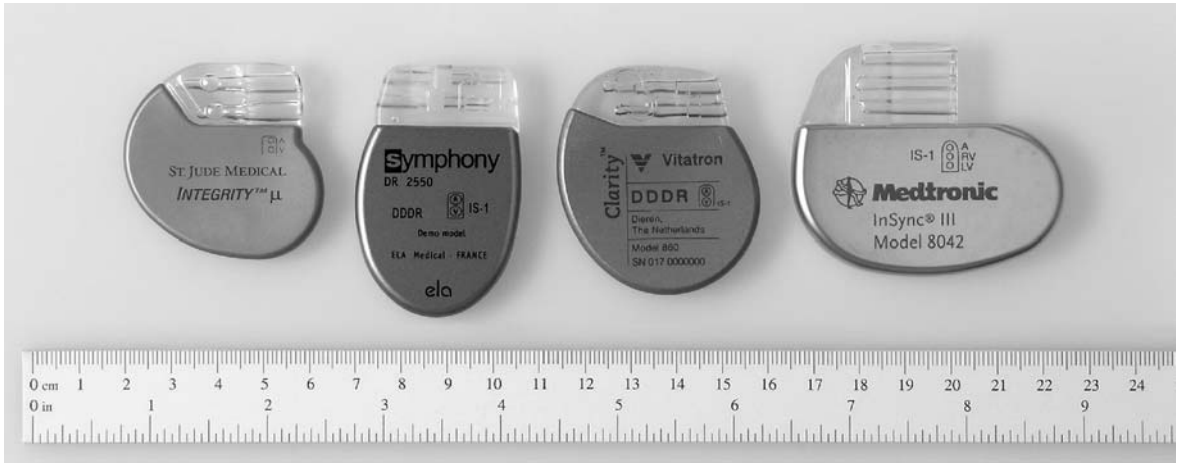

Figure 9.1

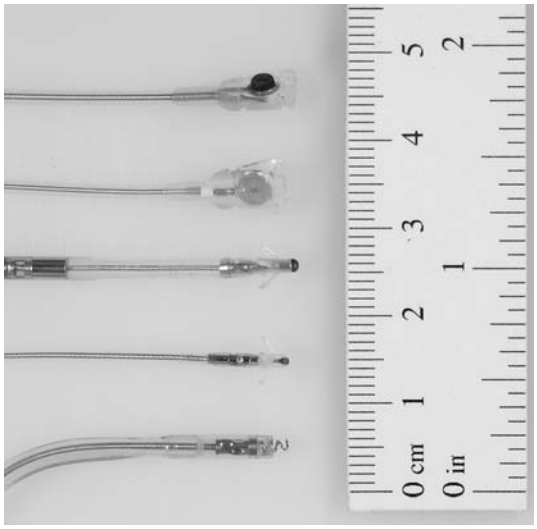

Figure 9.2

artifact is easier to see and may be more reliable. Bipolar leads have significantly better sensing potentials, particularly far-field sensing, and avoid local skeletal muscle twitch and skeletal myopotential over sensing and allow submuscular implantations of generators. Most leads implanted are endocardial but there have been significant advances, particularly with the steroid leads in epicardial leads. Finally, different length leads are now available, which reduces the amount of excessive lead that has to be hidden behind the pacing generator. Although the majority of these technological advances have been driven through adult cardiology, they have had the significant knock-on effect on the size and reliability of pacemakers for use in children.

PACEMAKER TERMINOLOGY

The nomenclature for pacemaker modes was introduced in 1987 by the North American Society of Pacing and Electrophysiology and the British Pacing and Electrophysiological Group (Bernstein *et al*., 1987). It is summarized in **Table 9.1**. For example, a VVIR mode is a ventricular-paced, ventricular-sensed mode that is inhibited by spontaneous activity and in addition has a rate response sensor; DDD means that both atria and ventricles are paced and sensed and both inhibited and triggered. The fifth letter relates to a pacemaker that has antitachycardia functions.

INDICATIONS FOR PACEMAKER IMPLANTATION IN CHILDREN

The American College of Cardiology, the American Heart Association and the North American Society of Pacing and Electrophysiology have produced a series of practice guidelines over the years, the latest update being that of Gregoratos *et al*. (2002), the section 1-G being relevant for children, adolescents and patients with congenital heart disease. The indications are divided into:

Class I:	Evidence and/or general agreement that pacing is indicated.
Class II:	Conditions in which there is conflicting evidence and/or divergence of opinion.
Class IIA:	The weight of evidence is in favour of therapy.

Class IIB: Therapy is less well established by evidence of opinion.
Class III: General agreement that pacemakers were not necessary.

The weight of the evidence was ranked highest at A, if the data was derived from multiple randomized clinical trials which involved large numbers of patients; intermediate, B, if the data was derived from a limited number of randomized trials that involved small numbers of patients or careful analysis of non-randomized studies or observational registries; a lower rank, C, was given when expert consensus was the primary basis for the recommendation. Indications are summarized in **Table 9.2**.

Our commonest indication for pacing used to be complete atrio-ventricular block following complex cardiac surgery that did not resolve within 2 weeks. However, with the advances in surgery this is now much rarer and currently our most frequent indication is

Table 9.1 NASPE/BPEG Generic Pacemaker Code.

Position	I	II	III	IV	V
Category	Chamber(s) paced	Chamber(s) sensed	Response to sensing	Programmability rate, modulation	Antitachyarrhythmia function(s)
	0 = None	0 = None	0 = None	0 = None	0 = None
	A = Atrium	A = Atrium	T = Triggered	S = Simple programmable	P = pacing (anti-tachyarrhythmia)
	V = Ventricle	V = ventricle	I = Inhibited	M = Multi-programmable	S = Shock
	D = Dual (A + V)	D = Dual (A + V)	D = Dual (T + 1)	C = Communicating	D = Dual (P + S)
				R = Rate modulation	

Table 9.2 Recommendations for permanent pacing in children, adolescents and patients with congenital heart disease.

Class I
1. Symptomatic advanced second- or third-degree AV block
2. Symptomatic sinus node dysfunction with correlation of symptoms
3. Postoperative advanced second- or third-degree AV block > 7 days
4. Congenital third degree AV block with wide QRS escape rhythm, complex ventricular ectopy or ventricular dysfunction
5. Congenital third degree AV block in the infant with a ventricular rate < 50/55 bpm or with CHD and a ventricular rate < 70
6. Sustained pause-dependent VT

Class IIa
1. Bradycardia-tachycardia syndrome requiring anti-arrhythmic therapy
2. Congenital third-degree AV block with an average heart rate < 50 bpm or abrupt pauses
3. Long QT syndrome with second- or third-degree AV block
4. Asymptomatic sinus bradycardia with congenital heart disease with low resting heart rate
5. Congenital heart disease with reduced function due to sinus bradycardia

Class IIb
1. Neuromuscular disease with any degree of AV block
2. Asymptomatic sinus bradycardia < 40 bpm with congenital heart disease
3. Asymptomatic congenital third-degree block with an acceptable rate and narrow QRS complex

isolated congenital complete A-V block, with or without symptoms, the latter situation becoming increasingly frequent with Holter monitoring, in accordance with the above recommendations and those of Dewey *et al*. (1987). We have not paced prophylactically asymptomatic, third-degree A-V block with acceptable rate, narrow QRS complex and normal ventricular function. Spontaneous sinus node dysfunction, or that occurring after atrial operations for transposition or Fontan type procedures, are the next most frequent indications. There is evidence that maintaining an adequate atrial rate reduces the frequency of atrial arrhythmias in this population of patients. The need to control tachyarythmias by pacing has been significantly reduced, as most patients can be helped by radiofrequency catheter ablation techniques or occasionally by surgical procedures, but in some of the complex Fontan type procedures pacemakers with overdrive pacing functions may well be used.

CHOICE OF SYSTEM TO BE IMPLANTED

The recommendations for pacemaker prescription for symptomatic bradycardia were introduced by a working party of the British Pacing and Electrophysiology Group (Clark *et al*., 1991) referring primarily to adult implantations (**Table 9.3**). These recommendations entailed a significant change in pacemaker practice, as they indicate that VVI pacing, even with rate response, although still a common type of procedure undertaken, is outdated.

In children, compromises are frequently required in order to obtain a reasonably good system that will produce a satisfactory long-term result with minimal battery and lead replacements. The more complex dual-chamber generators have generally a shorter lifespan than similarly sized single-chamber generators. Bipolar leads, which are most suitable for atrial sensing and pacing, are larger and stiffer than unipolar leads and two leads may be difficult to be put into a small vein. In addition, the pacemaker syndrome, which occurs with retrograde ventricular–atrial conduction and causes reflex hypotension in adults with ventricular pacing, is very rare in children.

Table 9.3 General principles of pacemaker choice.

1. The ventricle should be paced if there is actual or threatened atrio-ventricular block.
2. The atrium should be paced/sensed unless contraindicated.
3. Rate response is not essential if the patient is inactive or has a normal chronotropic response.
4. Rate hysteresis may be useful if the bradycardia is intermittent.

As described above, all systems currently implanted are multiprogrammable, i.e. alterations in many pacing parameters (e.g. pulse width, pulse amplitude, sensitivity, hysteresis and heart rate) can be made after implantation. Dual-chamber systems have additional features for altering the refractory periods of the atria and the ventricle, blanking period, and atrio-ventricular interval. It is important to have a fast enough maximal heart rate with appropriate sensors in young children. It is also very helpful to have pacemakers that can generate relatively high outputs (up to 8 V) to cope with long-term high thresholds, particularly on epicardial leads.

Small pacing generators are available for very small children; however, the disadvantage is a reduced lifespan. The new adult-sized generators are usually small enough for use in the majority of children, except for neonates and very small infants, particularly if they are implanted deep to the rectus muscle or in the suprarenal area.

Seventy-five percent of the increase in cardiac output during exercise occurs as a result of increased heart rate and the remainder occurs with an increase in stroke volume. Thus, the ability to increase pacing rate on exercise is extremely useful in children. Atrio-ventricular synchrony adds 20% to cardiac output at rest and is more important at slow heart rates.

We have therefore been somewhat more conservative in our choice of pacemakers and in small children have predominantly used single-chamber pacing and sensing and inhibitory systems with rate responsiveness. We have reserved endocardial dual-chamber systems for children above 8 years of age and those with poor ventricular function. If an epicardial system is used, then we would try to ensure both atrial and ventricular pacing.

WIRE IMPLANTATION

Our indications for epicardial implantation are summarized in **Table 9.4**. The advantages are that leads do not become displaced, but the disadvantages are the need for an operative procedure (often a thoracotomy), somewhat higher initial voltage threshold, and a higher incidence of lead fracture and late threshold rise and sensing problems (exit block) related to fibrosis at the junction between the electrode and the myocardium. These difficulties have been significantly reduced with the introduction of a steroid eluting epicardial lead.

Endocardial implantation is thus our usual approach because the initial and long-term thresholds are lower. With fixation at the distal end, either with tines or screws, an adequate loop can be created in the atria to allow for growth. Till *et al*. (1990) took this approach in infants

Table 9.4 Indications of permanent epicardial pacing system in children.

Small size (< 8 kg)
Right-to-left shunt
Venous access difficulties
Postoperative Glenn procedure
Superior vena caval obstruction
Ventricular access difficulties
Mechanical tricuspid valve replacement
Fontan-type operation
Failed transvenous system

with a body weight as low as 2.8 kg. In small children the ventricular pacing lead is usually unipolar because it has a smaller diameter and is more flexible. In the older child, bipolar leads work very well. For atrial pacing and sensing the bipolar leads are more appropriate.

IMPLANTATION TECHNIQUES

Temporary Systems

Transvenous

A temporary bipolar system can be inserted transvenously from any suitable venous access, the optimum being the subclavian vein, from which the electrode is less likely to displace and the site can be easily kept clean. Alternatives are the femoral vein, although this has a somewhat increased risk of sepsis and lead displacement, or the internal jugular vein. The wire is fixed to the skin and attached to a temporary external box, and the voltage threshold can be checked daily to ensure an adequate safety margin of $\times 2$.

Epicardial

At the end of most open-heart procedures, two atrial and two ventricular wires are sewn on. This allows atrio-ventricular sequential pacing, which is particularly important in patients whose cardiac output is borderline initially after operation. Junctional ectopic tachycardia (His bundle tachycardia) is a serious arrhythmia related to oedema around the His bundle. This is associated with significant morbidity and some mortality. Our current therapy involves cooling the patient, which decreases the rate of firing of the automatic focus, loading with Amiodarone, and atrio-ventricular sequential pacing at a slightly faster rate. This is usually a temporary requirement, with sinus rhythm returning over the next few days, when therapy can be stopped.

In patients with postoperative atrio-ventricular block, the voltage threshold can be checked daily to ensure an adequate safety margin (e.g. less than 75% of the maximal output of the external box). If sinus rhythm does not return within a reasonable time (7 days in recent guidelines, 10 days from our previous working practice), a permanent pacing system is implanted. If there are no worries concerning A-V block, the wires can be pulled out through the skin when no longer necessary and we now routinely remove these on day 1 or day 2 post-operatively, except in those patients where they have been used, and following a Fontan procedure or cardiac transplant.

Permanent Systems

Endocardial Implantation

The procedure requires X-ray fluoroscopy with a C arm and is performed in a catheter laboratory or a pacing theatre. Sterility is paramount and it is important to empower all members of the multidisciplinary team to point out areas of concern. We perform our procedures under general anaesthesia with a 24 hour course of anti-staphylococcal antibiotics starting at induction. Adult centres use basal sedation and local anaesthesia. Use of antibiotic cover is not universal but is very common. Meticulous preparation of the operative site with sterilizing fluid, draping widely with sterile towels and use of adhesive plastic drape over the operative area are all important.

The subclavian vein is our approach of choice, with modification of the Seldinger technique as described by

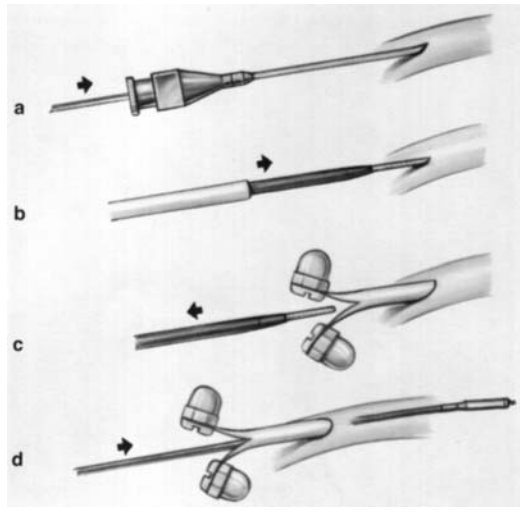

Figure 9.3

Vellani *et al*. (1969) (**Figure 9.3**). The patient is positioned supine, arms by the sides, with a bag of saline or a pad between the shoulders and the head turned to the opposite side. A venogram performed by a peripheral injection of contrast and screening over the shoulder area is often performed and is particularly useful if there has been vascular access from that side (following previous surgery, or in ex-premature babies, there may have been long lines from the left arm for parental nutrition early in life).

A small 3 cm incision is made 3 cm below the junction of the lateral one-third and medial two-thirds of the clavicle, where the clavicle bends posteriorly. The subcutaneous tissue is dissected down onto the pectoral fascia. The end of the needle is bent at 30° to keep it just under the clavicle and away from the subclavian artery. It is inserted in the lateral margin of the incision at the junction of the lateral one-third and medial two-thirds of the clavicle. It is directed towards the suprasternal notch or slightly more cephalad and is kept as superficial as possible. In small children we often use initially a fine 19-gauge needle with a 0.025 inch wire to enter the vein, followed by a fine dilator and sheath. Subsequently we replace these with a thicker wire and an appropriate peel-away sheath. In older children the puncture is performed with a larger needle and a 0.035 inch wire, over which in due course the appropriate dilator and peel-away sheath are directly inserted. We usually use an additional subclavian puncture for a separate lead, but that is not always necessary or possible. The dilator is curved to facilitate following the wire around the relatively tight venous bends of small children. The dilator is removed and the pacing electrode, which is an active or passive fixation type, is inserted with a curved stilette. Alongside the electrode, a fine straight wire is placed to allow subsequent venous access if necessary or the use of an additional lead. The sheath may be left in, particularly if it has a snug fit to the electrode, and this allows easier manipulation of the wires under the clavicle. However, if there is excessive blood loss, particularly with the tined leads, the sheath should be withdrawn and pressure applied.

Ventricular Pacing

The electrode with its curved stilette is passed to the right atrium, often a loop is formed with the stilette pulled back, and it passes across the tricuspid valve and out into the right ventricular outflow tract and pulmonary artery. In small children the curve of the stilette needs to be within 2 cm of the end. The entry into the pulmonary artery may be difficult, particularly in corrected transposition of the great arteries or in transposition after atrial surgery. In these cases the use of lateral screening or the presence of ventricular ectopics confirms that the ventricle rather than the coronary sinus has been entered (**Figure 9.4**).

The curved stilette is exchanged for a straight one, which is inserted to within 2 cm of the tip of the electrode, which is gradually withdrawn. As the electrode becomes more horizontal, the stilette is advanced to get the tip to the apex of the ventricle as far lateral as possible

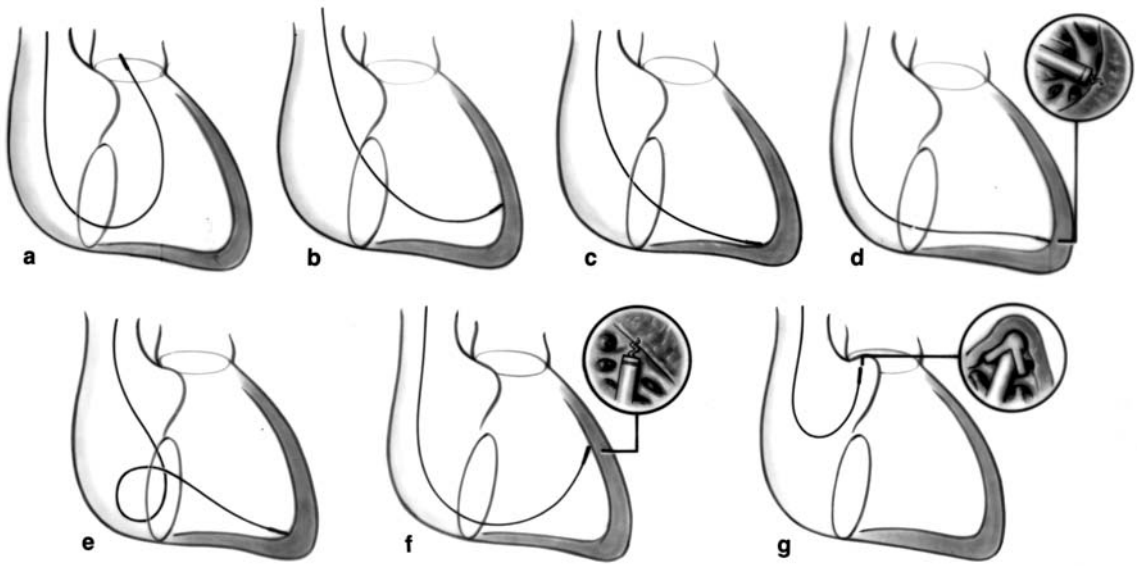

Figure 9.4

with the tip pointing downwards. A separate bobbing action may be visible when the tip of the electrode is caught under trabeculae. The lead is then fixed either by pushing gently with the tined lead or by turning the screw mechanism on the electrode, which delivers the screw, and this can be visualized by a change in shape near the electrode tip. The stilette is pulled gently backwards into the atrium and gentle traction is applied to see whether it has indeed caught in the myocardium. Other operators keep the stilette back from the tip during electrode movement and positioning in the apex of the ventricle to reduce the chance of perforation.

If the lead is a unipolar lead, an indifferent lead is inserted into the incision below the clavicle or into the box pocket. If the lead is a bipolar lead, the central core is the active lead and the ring electrode the indifferent. Electrical measurements are made by connecting with the sterile leads to a pacing system analyser (PSA). These are the intracardiac electrograms, intracardiac R wave, slew rate, pacing threshold, stability with hyperventilation, maximum output to check for phrenic stimulation (**Table 9.5**). An adequate atrial loop can usually be created for growth and the lead fixed to muscle below the clavicle, using the sleeve and non-absorbable sutures (the sleeve can be reduced in size, particularly in thin children).

There has been concern about the detrimental effect of right ventricular apical pacing with wide left bundle branch block type QRS pattern affecting cardiac function, and the increasing risks of atrial arrhythmias. The alternative sites of right ventricular septal or free wall have been suggested as a way to reduce this problem. However, current studies (Bourke *et al*., 2002; Schwaab *et al*., 1999) have shown no difference at early follow-up.

Atrial Lead Insertion

Bipolar pacing leads are used: either a tined lead for the atrial appendage or screw-in leads for both the atrial appendage or the free atrial wall. The leads may be a preformed J electrode or a J-shaped stilette (which in small children needs to be made even tighter) can be used with a straight electrode. The electrode with its shaped stilette placed within the atrium is withdrawn with the tip pointing leftwards and superiorly. In large atria it is sometimes helpful to open the curve a little by withdrawing the stilette somewhat. When the lead enters the right atrial appendage the electrode tip moves from side to side, a windscreen wiper motion. The activation screw is then deployed, the stilette gently withdrawn, an adequate loop created and appropriate electrical measurements recorded. Again, hyperventilation is used to check stability and output is increased to maximum to ensure that the phrenic nerve has not been stimulated. If the free atrial wall is used, a screw-in lead is essential. Again, the lead is fixed to the pectoral fascia, using the sleeve and non-absorbable sutures (**Table 9.5**).

Table 9.5 Optimal guideline measurements at lead implantation.

Parameter	Atrial	Ventricular
P–R amplitude (mV)	> 1.5	> 4.0
Slew rate (V/s)	> 0.5	> 0.5
Threshold (V) at 0.5 ms	< 1.2	< 1.0
Pacing impedance (Ohms)	> 500	> 500

Dual-chamber Pacing

It is usual practice to place the ventricular lead first followed by the atrial lead. The ventricular lead is initially left lying on the floor of the atrium until the atrial lead is in a stable position and then its atrial loop (to allow for growth) can be created. This makes it less likely that the leads will catch around each other and interfere (**Figure 9.5**). The concern regarding venous thrombosis in the relatively small veins in childhood has proved to be unfounded (Gillette *et al*., 1989). Screening just prior to fixation of sleeves is a useful check that the appropriate atrial loops are still in position.

Practical Difficulties with the Wire Position

1. A left superior vena cava to the coronary sinus can be used for both atrial and ventricular pacing, with

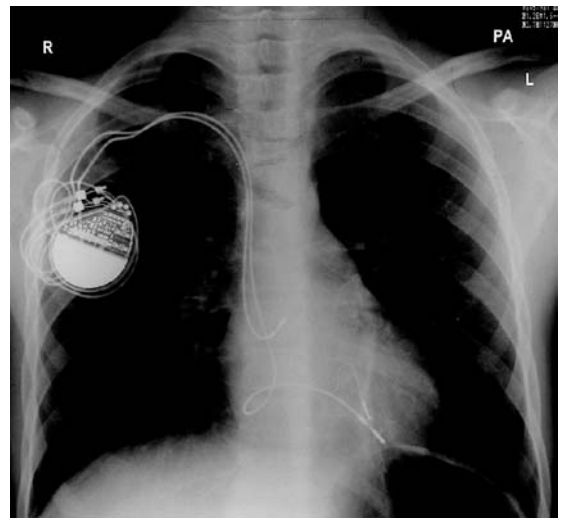

Figure 9.5

either a tined lead or a screw-in lead. It is relatively easy to enter the tricuspid valve on an atrial loop.

2. In a small heart, the bend between the right atrium and the pulmonary artery is too tight to take a straight stilette. In this case, the lead is withdrawn, leaving the stilette within the atrium, and the stilette is only advanced when the electrode has become almost horizontal. Occasionally a gentle curved stilette needs to be used for this manoeuvre. It is more of an issue with the fine unipolar leads than with the heavier bipolar leads.
3. In atrio-ventricular and ventriculo-arterial discordance (corrected transposition), the right atrio-ventricular valve is in a higher plane and has a different orientation than normal, which makes it more difficult to cross.
4. If the cardiac anatomy is unusual, the subclavian vein on the side of the innominate vein is used to reduce one of the tight bends and therefore make wire manipulation somewhat easier.
5. The atrial loop may slip into the ventricle (**Figure 9.6**). There has been some concern that this may cause A-V valve regurgitation or arrhythmias, but we have not seen this in practice.
6. If the ventricle to be paced has a left ventricular morphology, then a screw-in lead is usually chosen.
7. Other sites within the right ventricle may be used other than the apex, and this may be important in reducing ventricular dyssynchrony (**Figure 9.7**). Outflow tract or the interventricular septal sites, which are possible with active fixation leads, may be the optimum pacing sites in the future.
8. If the right atrium or right ventricle are large (related to significant tricuspid regurgitation), active fixation bipolar leads are used. These are heavier and more stable.

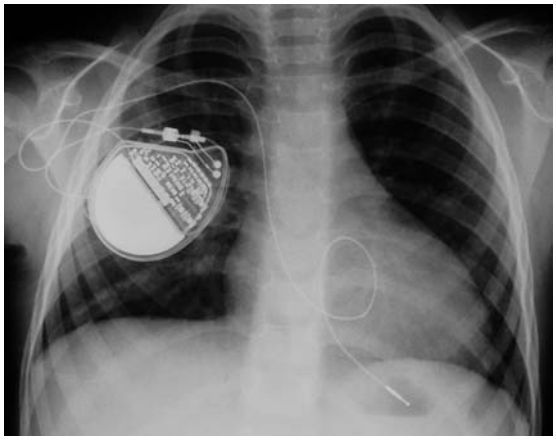

Figure 9.6

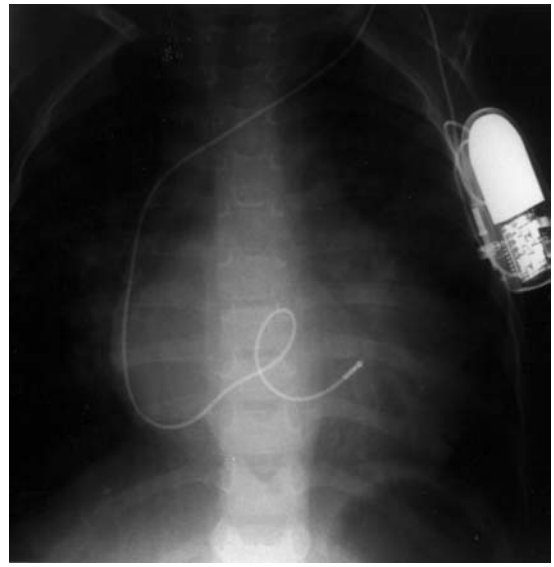

Figure 9.7

Alternative Venous Approach

Cephalic Vein

The Cephalic vein is often too small in younger children but may well be large enough in adolescent patients to take one or sometimes even two leads. The vein runs deep in the deltopectoral groove, before making an inferior bend to join the subclavian vein. The distal vein is ligated, the vein incised with a pair of scissors, and the orifice held open with a vein lifter. A guide wire, an introducer and sheath are frequently used or the lead may be inserted directly. Once an adequate position has been obtained, the lead is fixed and the box pocket created.

Internal/External Jugular Vein

The internal jugular vein can be used with a Seldinger technique. An additional incision is created to bury the wire. A cut-down procedure is used for the external jugular vein and that vein is ligated. The pacing wire should be brought down behind the medial part of the clavicle, so that erosion through the skin does not occur. This approach is currently only rarely used.

Trans-hepatic Approach

Venous access issues can be of major concern following operations for complex congenital heart disease, e.g. the Glenn procedure precludes approach from the

upper veins. Trans-hepatic venous access has become established for long-term intravascular feeding and the technique has been used occasionally for endocardial pacing. Under ultrasound guidance the hepatic vein is entered using the Seldinger technique; an appropriately sized sheath(s) can be inserted and active fixation leads inserted to atria and/or ventricle. With active fixation leads the diameter of the lead is similar to the sheath, reducing the chance of subcapsular haemorrhage. The leads can be fixed and a suitable box pocket created. This has occasionally been used when epicardial pacing is not possible.

Generator Pocket

This pocket can be constructed in the prepectoral or subpectoral area, or in the axilla. If the former is chosen, the box pocket can be made before the wire is placed with an incision 3 cm below the clavicle, and the pocket created by a blunt dissection downward and medially onto the prepectoral fascia. The needle can then be inserted into the subclavian vein through the top of this incision and thus no tunnelling is required.

If the axilla is chosen, an incision is made horizontally in the middle of the axilla, starting at the edge of the pectoris major, running posteriorly. Blunt dissection downward, keeping relatively anteriorly down onto the fascia on the ribs, will produce a box pocket of appropriate size. It is important to avoid extending the dissection too posteriorly, otherwise local muscle stimulation occurs if a unipolar system is used.

The box pocket should be dry; the wire is tunnelled down from the subclavicular region, cleaned, and connected to the generator, excess wire being positioned behind the box.

The subpectoral pocket has become much more frequently used, with the familiarity that has come from inserting defibrillator devices. This produces an extremely good cosmetic result and has reduced the chance of infection.

The connector pin of the leads is inserted into the correct (A or V) port of the pacemaker head of block. There is usually a diagram provided on the pacemaker can. The pin of the lead must be checked to be beyond the set screws, which are then tightened with a screwdriver that has a torque limiter. The redundant lead length is coiled underneath the generator in the pre-formed pocket. It is important that this pocket is large enough to accommodate the generator and that it and the redundant lead is away from the incision, to reduce risk of wound breakdown. The muscle or subcutaneous tissues are closed by loose absorbable sutures and the skin can be closed with either subcuticular absorbable sutures, such as Dexon, or a subcuticular suture that needs removal.

Epicardial Implantation

Permanent wires can be positioned on the epicardial surface through a left thoracotomy, also by a subxiphoid or mediasternotomy approach. Again, anti-staphylococcal cover is used for 24 hours.

Through a left-sided anterolateral thoracotomy in the fifth intercostal space, the pericardium is opened longitudinally away from the phrenic nerve. Sites free from coronary arteries, on both the left atrium and the left ventricle myocardium, are chosen. The area may be checked with a temporary probe before the permanent leads are attached (**Figure 9.8**).

The advantage of the thoracotomy is that the lateral wall of the left ventricle is paced, which reduces left ventricular dysynchrony (see later).

The subxiphoid or sternotomy approach has been used, with a small incision from the xiphoid process when both a portion of the right atrium and right ventricle can be visualized. The position of electrodes inserted through this approach is shown in (**Figure 9.9**).

The thresholds can be measured with a fine probe before the permanent leads are positioned.

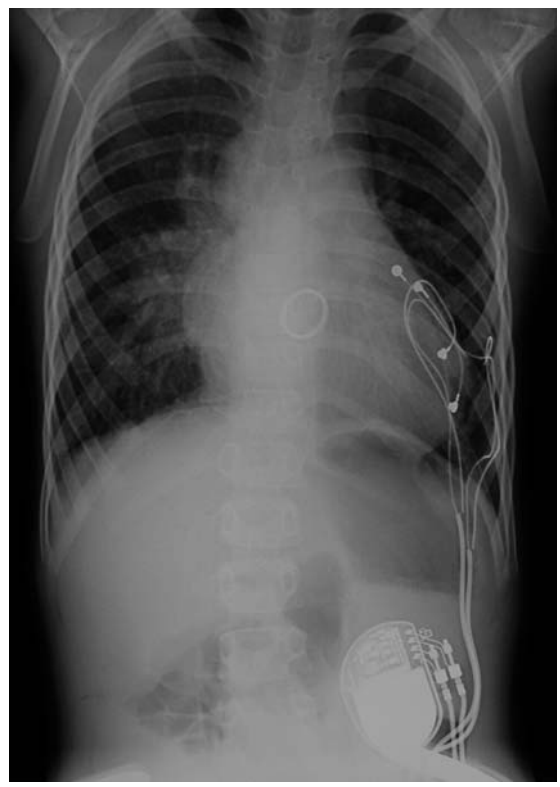

Figure 9.8

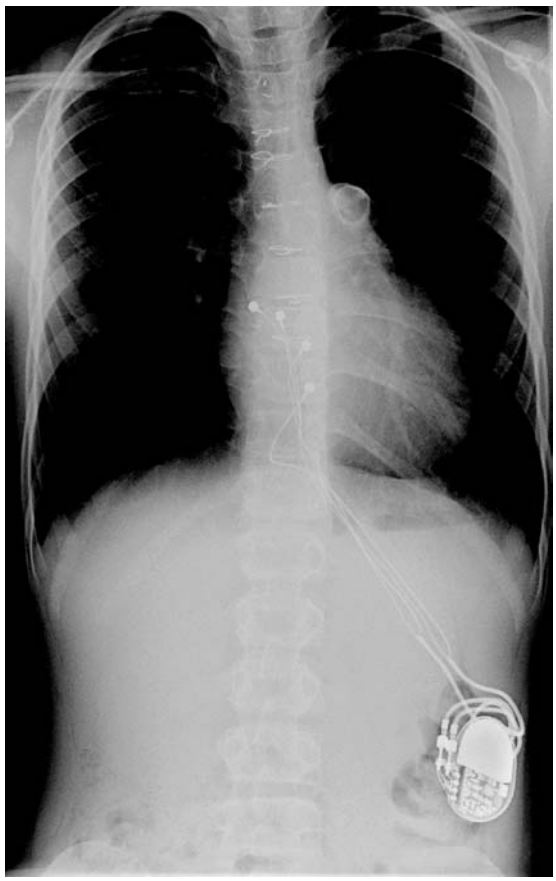

Figure 9.9

The electrodes are fixed with a gentle curve to minimize torque. The box pocket may well have been created prior to lead position. Initial thresholds that are acceptable usually are somewhat higher than on endocardial leads. Steroid eluting sew-on leads have now replaced the screw-in and fish hook leads and the long-term results have been really encouraging.

Generator Placement

Suprarenal Approach

This approach is frequently used in small infants and thin children because the pacemaker can be buried very deeply (**Figure 9.10**). An incision is made below and lateral to the 12th rib and dissection is carried down onto the muscle, which is split to reveal the extraperitoneal space above the kidney. The wire is tunnelled subcutaneously and connected to the box, the muscle edges are brought together with Dexon and the wound closed in layers.

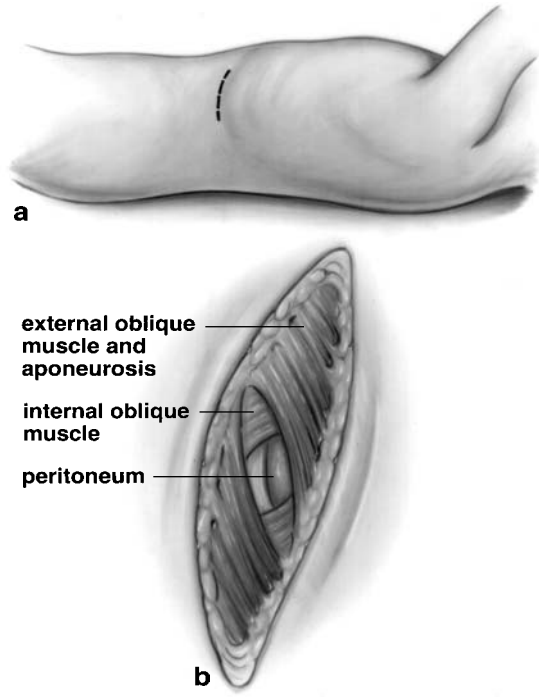

Figure 9.10

Sub-xiphoid Approach

A vertical incision is created down onto the aponeurosis of the rectus muscle, which is then incised close to the midline and mobilized. A space is created behind it (**Figure 9.11**). The wire is tunnelled subcutaneously down into this pocket and connected to the generator. The window (writing) of the pacemaker is turned away from the muscle to avoid twitching with the unipolar systems. It is helpful in these abdominal positions to fix the pacing box with a non-absorbable suture, so that it does not migrate.

Biventricular Pacing (Cardiac Resynchronization Therapy)

In association with heart failure, there are electromechanical abnormalities of prolonged atrio-ventricular, interventricular and intraventricular delay. This combination allows for poor ventricular filling, diastolic mitral regurgitation, delayed activation of the free wall of the left ventricle in comparison to the right ventricle, and finally dysynchrony between different segments of the left ventricle, particularly the septum and the free wall, all of which produce inefficient cardiac function with a high metabolic demand.

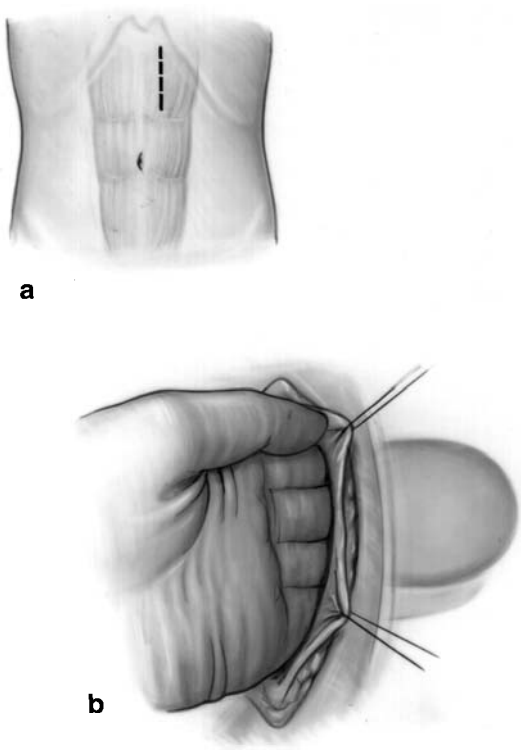

Figure 9.11

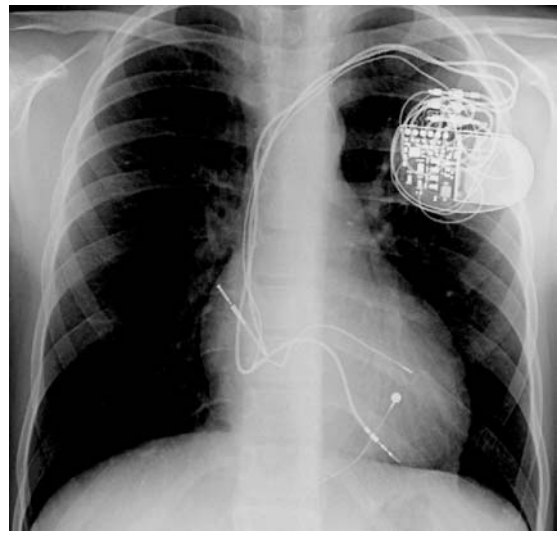

Figure 9.12

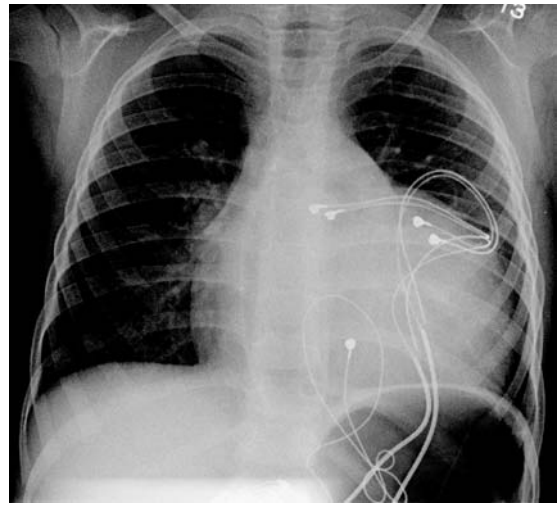

Figure 9.13

Cazeau *et al*. (1994) first used the term when reporting improved cardiac function in patients with dilated cardiomyopathy with epicardial pacing. The left ventricle can now be paced either epicardially or, more frequently in adults, endocardially through the coronary sinus to a suitable vein on the lateral free wall. There are now many studies confirming improvement in cardiac function in the short and medium term with this form of therapy in adults (Linde *et al*., 2002; Abraham *et al*., 2002).

In children the indications are evolving. Some patients with primary dilated cardiomyopathy with left bundle branch block pattern can be improved, unless in NYHA Class 4. Reversal of ventricular dysfunction secondary to prolonged right ventricular apical pacing is a very logical indication.

With biventricular pacing, additional right atrial and right ventricular leads are usually implanted first via the subclavian and/or cephalic approach. The coronary sinus is entered with a specially designed sheath, currently an 8 French. The necessity for three leads within the venous system in children makes it suitable only for older children (**Figure 9.12**).

In smaller children an epicardial approach has been used, with a combination of a left lateral thoracotomy and a low sternal approach, which allows placement of wires on the right atrium, right ventricle and lateral wall of the left ventricle. **Figure 9.13** shows the X-ray at implantation of the system and (**Figure 9.14**) 6 months later. Sometimes left atrial and left ventricular pacing via a thoracotomy can produce significant clinical long-term improvement. **Figure 9.15** shows the X-ray before and **Figure 9.16** again 6 months after the placement of a pacing system. During follow-up, Doppler assessments of mitral inflow velocity and echo assessments of

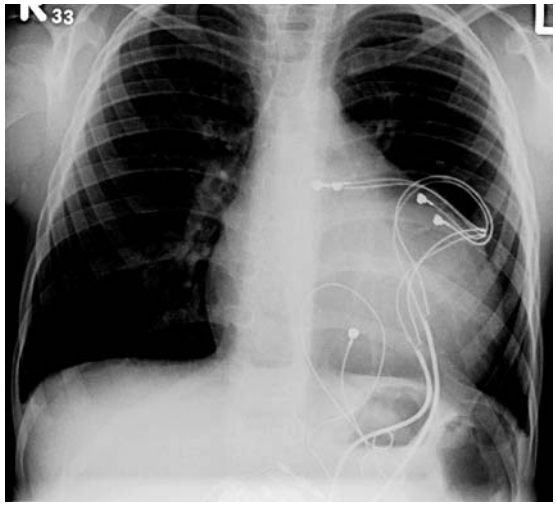

Figure 9.14

Figure 9.15

left ventricular function with tissue Doppler imaging, optimal AV delay and interventricular timings can be achieved. Further assessment of this form of pacing will be needed in future.

In congenital heart disease, right ventricular dysfunction is a major issue, particularly following repair to tetralogy of Fallot in association with right bundle branch block. Whether right ventricular septal or outflow tract pacing in this group of patients will improve electromechanical efficiency is as yet unknown and it may be an exciting development for the future.

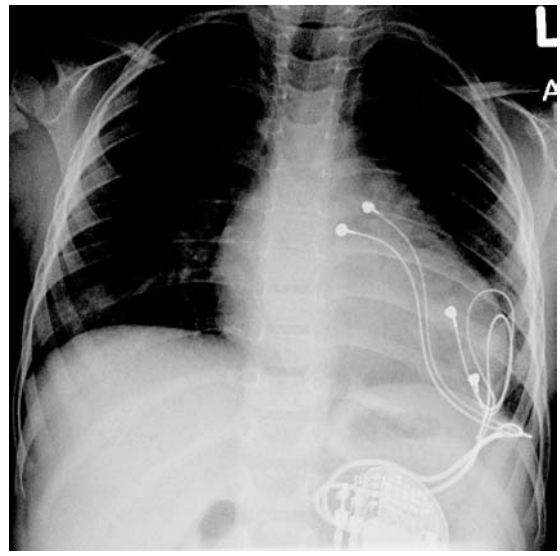

Figure 9.16

PACEMAKER PROBLEMS

Electrode Dislodgement

Electrode dislodgement may occur with endocardial leads, although it has become significantly rarer since active fixation leads have been routinely used. The patient may require a temporary trans-venous system to main cardiovascular stability while the permanent system is revised. The box pocket is opened and the pacing wire disconnected from the generator. The anchoring suture on the sleeve is removed; if necessary, a subclavicular incision would have to be opened. A new stilette is advanced into the electrode and the system is repositioned. We undertake this procedure under general anaesthesia and cover the procedure with a further course of anti-staphyloccal antibiotic for 24 hours.

High Pacing Threshold

This problem can occur with either epicardial or endocardial leads. It may require the use of a higher-output generator, accepting that this will buy time. This is particularly important when pacing is going to be used for life. If this is not satisfactory, a new lead would have to be inserted.

Lead Fracture

This may occur with crushing between the head of the first rib and the clavicle if the subclavian approach is too medial. It may also occur if the anchoring suture is

tied directly onto the insulation of the electrode, rather than using the anchoring sleeve. Occasionally a lead may sense adequately but not pace.

Muscle Twitching

This may occur with a unipolar system. It can be frequently abolished by reducing the voltage output and, if necessary, increasing the pulse width. If these procedures are not successful, the generator would need to be moved or occasionally a Gore-Tex sleeve placed around it. If the problem persists, the whole system would need to be replaced with a bipolar one.

Erosion

Occasionally the box pocket may be too tight and the skin becomes thin and shiny. If this problem is detected early enough, the generator can be moved and repositioned in a better area, perhaps deep into the pectoralis muscle. If it is left too long in the same place, infection can occur when the system erodes through the skin and then the whole unit would need to be replaced.

Left Ventricular Lead Malposition

Occasionally the lead can pass through a patent foramen ovale into the left side of the heart. If the position on AP screening looks unremarkable, the lateral screening would indicate a relatively posterior position. Pacing from this site produces a right bundle branch block pattern, which is more clearly seen if the monitoring leads include V_1, which may not be a routine. If this is detected, the lead should be repositioned early because of the danger of thrombus formation and embolization and potential damage to the mitral valve at later removal.

Infection

Infection is the major concern in pacemaker insertion. Very occasionally, long-term antibiotics may eradicate a local box infection but usually the whole system needs complete revision and replacement.

Systemic antibiotics are given and local skin care is rendered. If the patient is not pacemaker-dependent, the old system can be explanted, time allowed for the area to heal and a new system implanted in a new site. If the patient is pacemaker-dependent, a temporary lead can be inserted to cover the procedure to allow healing of the infected site. On occasion this is not possible; in those cases when the infection has been treated, a new system can be inserted into a clean site during the same procedure as the infected one is removed. If both systems are endocardial, the new wires are inserted into an adequate position and a separate operator explants the infected system and removes the old wire. After this, the new system is connected to its pacing generator and the wound closed.

Late infection usually requires lead and generator removal (see section).

Non-functioning Wire

Epicardial leads can be left in place; however, there is considerable debate concerning endocardial leads. They have become extravacular by being surrounded by a tube of endothelium and some centres leave them in position, others will remove them (see following section).

Endocardial Lead Extraction

Occasionally, endocardial leads need to be removed and the North American Society of Pacing and Electrophysiology have published recommendations for the indication, facilities and training required (Love *et al*. 2000).

The *Class I* (mandatory) ones include situations when the lead is involved in deep-seated infection/endocarditis, significant arrhythmias, thrombo-embolic events, interfering with another device, e.g. an ICD, or there is occlusion of all other usable veins and venous access is paramount.

The *Class II* indications are: localized box pocket infection not involving the lead; occult infection where the pacing system might be the culprit; chronic pain; leads preventing venous access for a newly required device; and non-functioning leads in young patients.

Leads should not be removed where the risk is higher than the benefit, particularly in those with a clotting abnormality. The lead is frequently stuck at several sites, e.g. entry to the subclavian vein, at the junction of the innominate vein and SVC, the tricuspid valve, and on the floor of the right ventricle as well as at the apex. The details of wire extraction are beyond the scope of this chapter but can be found in Ellenbogen and Wood (2005).

We have a very limited experience, removing only eight leads over the past 10 years. It is a procedure that needs to be planned and be performed either by or in conjunction with a colleague, often an adult cardiologist, who has significant experience, and with a surgical team on standby, in case major haemodynamic collapse occurs. Several large series with complete extraction rates around 90%, a complication rate of 3% and a mortality rate of $< 1\%$ have been published (Smith *et al*., 1994; Byrd *et al*., 2002).

Elective Generator Replacement

The timing of generator replacements can now be judged to the optimum (see follow-up). Different pacemakers have different end-of-life characteristics, a drop in cell voltage and/or reduction in pacing rate when a magnet is applied. It will be known whether the patient has a reliable underlying rhythm from the follow-up information. It is important to check the implant details of site of the box, whether subpectoral or prepectoral. The excess wire is usually placed beneath the pacing box. The skin is incised, the thick fibrous capsule incised and the box removed. If it is a unipolar system without reliable underlying rhythm, then a different electrode with a clip is attached to the casing of the generator and inserted into the pocket. This allows for continued pacing when the generator is removed from the patient. The generator is disconnected from the lead, which is then attached temporarily to an external pacing source. The voltage thresholds are again measured, intrinsic cardiac action is recorded and a new generator connected. The wound is closed in layers and the procedure covered with anti-staphylococcal antibiotics for 24 hours. Generator replacements are now frequently undertaken as day case procedures.

PACEMAKER FOLLOW-UP

The British Pacing and Electrophysiology Group have published guidelines for pacemaker follow-up and its aims, the equipment necessary and the training and support required (Sutton, 1996). The aims can be summarized as ensuring that the pacing system is optimized for individual needs, with safe optimization of battery life; identification of abnormalities and complications; prediction of end-of-life generator; accumulation of accurate data; patient support and education and training opportunities for medical and technical staff. Each patient will have a European pacemaker registration card. In the UK currently, Group I drivers can drive after a week of a pacemaker implant, whereas with Group II drivers, where driving anything larger than a minibus is involved, 6 weeks following pacemaker implantation is required before the patient is considered fit to drive. These rules are regularly updated.

Avoiding direct trauma to the pacing box is important, therefore contact sports such as rugby and boxing should be avoided. Very strong electrical fields, such as fairground dodgem cars, can also interfere with pacemakers. With regard to occupations, the electrical fields of arc welding and dynamos can affect systems but most patients are able to work well within factories. With surgical procedures using diathermy, the pacemaker should be programmed to a fixed rate beforehand to avoid interference if the patient is pacemaker-dependent. Patients with pacemakers cannot be exposed to MRI.

CONCLUSION

Pacing systems have become significantly more sophisticated and complex but more reliable and can now be tailored to the individual child's electrical and haemodynamic needs. With careful techniques during implantation, complications are very low.

IMPLANTABLE DEFIBRILLATORS

Implantable defibrillators, like pacemakers, have changed enormously over the years. Although the early work of Mirowski *et al*. (1973) used the endocardial approach, the first commercial systems involved epicardial patches and a large defibrillator box, which needed to be placed in the abdomen. The battery life was short and the system was unsuitable for paediatric or adolescent patients.

With improving technology the boxes are smaller and the systems are now endocardial. The lead may have a distal single coil in the right ventricle or a dual coil (**Figure 9.17**), where there is an additional coil usually positioned in the superior vena cava. The defibrillator can is now active and part of the electrical circuit, the shock being delivered jointly from the can and the proximal coil to the distal coil. The polarity may be reversed if necessary. The energy wave form is biphasic.

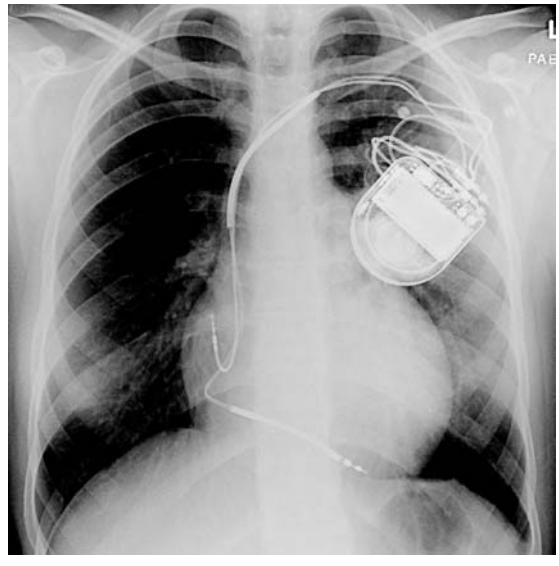

Figure 9.17

The circuitry has become very sophisticated, with continuous sensing of intracardiac electrograms, the ability to detect ventricular tachycardia and apply appropriate overdrive pacing and to detect ventricular fibrillation and institute defibrillation. There is standard post-shock ventricular pacing for a set time.

The ACC/AHA/NASPG 2002 guidelines (Gregoratos *et al*., 2002) give recommendations which are primarily for adults. The indications for implantation of defibrillators in the young are evolving, with improvement in battery life and sensing qualities, which differentiate ventricular arrhythmias from fast sinus tachycardia and atrial arrhythmias. These indications have broadened and are now considered for patients felt to be at high risk of sudden cardiac death, as well as those who have had a previous episode of ventricular tachycardia/ventricular fibrillation. (Elliott & McKenna 2004, Maron *et al*., 2000).

Currently, our most frequent indications for primary prevention are:

1. High-risk hypertrophic cardiomyopathy (two risk factors out of the following: a bad family history of syncope; severe myocardial hypertrophy; non-sustained ventricular tachycardia on Holter; and possibly a poor blood pressure response to exercise in adolescence).
2. Brugada syndrome.
3. Coumel, catacholamine-sensitive tachycardia.
4. Long QT syndrome, with either no response to beta-blockers or concern with regard to compliance.

We have not followed the adult practice of inserting defibrillators in dilated cardiomyopathy patients, despite having a large heart failure and transplant programme on site.

TECHNIQUE

The technique of insertion is very similar to a pacemaker insertion, performed under general anaesthesia with antibiotic cover and with additional external defibrillator leads attached. A subpectoral pocket for the box generator is ideal because of the size of the defibrillator can. The defibrillator lead, usually an active fixation type, is placed as far laterally in the right ventricular apex as possible, aiming to keep the distal coil completely within the right ventricular cavity.

Before leaving the hospital, the patient and the family are given further explanations of their ongoing care and in particular of what happens when the defibrillator fires. They should know that they should return for the system to be checked, to see if the therapy was appropriate. Changes may be necessary in the sensitivity of the system. If a series of inappropriate shocks occur, the system can be inhibited by a magnet placed over the box site in a local accident and emergency department. Group 1 drivers can drive 6 months after a device has been implanted, if it has not fired. A Group 2 licence cannot be held. MRIs have to be avoided and advice against contact sports is given. Ongoing specialist nursing/psychological support as well as medical care is essential to these patients and their families.

Acknowledgement

Figures 9.3, 9.4, 9.10 and **9.11** are reproduced from *Re-operations in Cardiac Surgery*, Stark J, Pacifico AD (eds). Springer-Verlag: New York, 1989.

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10

Catheter Ablation of Arrhythmias

M. Lowe

This chapter should be read in conjunction with Chapter 47 (Surgery for Arrhythmias).

The improved survival rates and clinical outcomes of patients with congenital heart disease (CHD) has led to an increase in the number of those presenting with arrhythmias in adolescence and adulthood. As a consequence, arrhythmias are the commonest cause of hospitalization in this age group and lead to significant morbidity, and even mortality. Electrophysiological evaluation and treatment of CHD patients with catheter ablation is becoming an increasingly important focus of management (Triedman, 2002).

Patients with CHD and arrhythmias represent a diverse group. Some arrhythmias are similar to those found in the general population, such as atrio-ventricular nodal re-entry tachycardia (AVNRT) and atrio-ventricular re-entry tachycardia (AVRT) mediated via an accessory pathway, the latter particularly associated with Ebstein's anomaly of the tricuspid valve. More commonly, however, the substrate for arrhythmias is acquired through surgical intervention. Here, the anatomical barriers for re-entry are incisions and patches in the atria or ventricles (**Figure 10.1**), compounded in many cases by chronic haemodynamic overload leading to conduction delay (Kalman *et al*., 1996). As a consequence, atrial flutters are the most commonly seen atrial arrhythmias, and multiple flutter circuits are often seen in individual patients following instrumentation of the right atrium (Nakagawa *et al*., 2001; Triedman, 2002). Repeated episodes of atrial flutter may lead to the development of atrial fibrillation with time, but this may also arise *de novo* in patients with left atrial dilatation and fibrosis (Kirsh *et al*., 2002).

CONDITIONS

Atrial Septal Defect (ASD)

Supraventricular arrhythmias, particularly atrial flutter, are very common in patients with an ASD. In those who have not had surgical correction, arrhythmias are seen in 30–40% of patients by the age of 35 years. Atrial flutter in these patients is often ''typical'', i.e. dependent on conduction through a critical isthmus between the tricuspid annulus and inferior vena cava, and the activation wavefront may circle in either a counterclockwise or clockwise direction (Chan *et al*., 2000).

In the postoperative patient, intra-atrial re-entry tachycardia (IART), also known as scar-related flutter or atypical flutter, may occur around a right atriotomy scar, cannulation site or, less commonly, an ASD patch (Kalman *et al*., 1996). With time, atrial fibrillation is seen in association with flutter, and may become the dominant rhythm during long-term follow-up.

Tetralogy of Fallot (TOF)

Most patients with TOF survive into adulthood with palliative or corrective surgery (Murphy *et al*., 1993). Patients with repaired TOF are at risk for both atrial and ventricular arrhythmias (Roos-Hesselink *et al*., 1995; Lucron *et al*., 1999). Following surgery, patients may have scarring of the right atrium from an atriotomy, and right ventricular scar from a ventriculotomy or ventricular outflow tract patch. In addition, residual haemodynamic lesions, such as RV dilatation caused by chronic pulmonary regurgitation, or RV hypertrophy secondary to residual pulmonary stenosis, may contribute to arrhythmogenesis.

Atrial arrhythmias are more prevalent, occurring in 12–35% of patients with repaired TOF, and in 50% of symptomatic patients (Triedman, 2002). The type of atrial arrhythmia documented is predominantly typical flutter or IART, but also includes atrial tachycardia and atrial fibrillation (AF). Atrial arrhythmias in this patient group are associated with older age at surgical repair, longer duration of follow-up and the presence of haemodynamic abnormalities.

The majority of patients with postoperative atrial flutter and TOF have circuits involving the typical flutter isthmus in either a counterclockwise or clockwise

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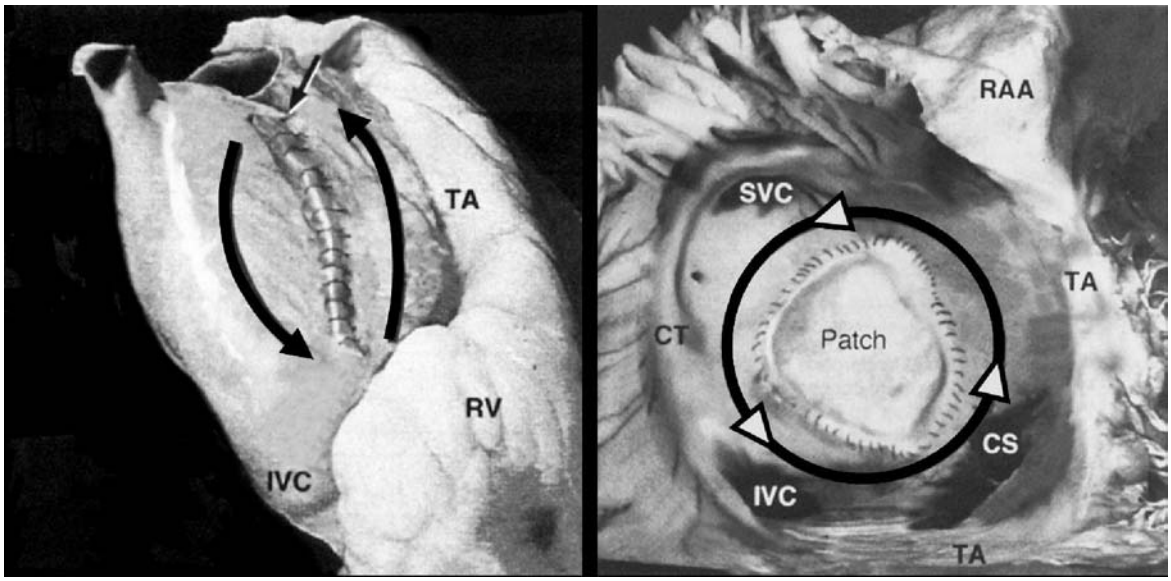

Figure 10.1

direction. Isthmuses responsible for IART include atriotomy scars and areas of fibrosis in the right atrium.

Sustained ventricular arrhythmias are less common, occurring in 0.5–6% of patients (Lucron *et al*., 1999). The incidence of sudden death in patients with TOF has been reported as 1–2% and increases with age. The substrate for ventricular tachycardia (VT) in this population is thought to be a macro-re-entrant circuit around the RV outflow tract patch or outlet septum. The occurrence of VT or sudden death is related to age at repair, duration of follow-up, surgical approach and chronic pulmonary regurgitation. Invasive electrophysiological evaluation may be useful for risk stratification.

Transposition of the Great Arteries

Atrial arrhythmias in patients with Mustard or Senning procedures are primarily IART secondary to a macro-re-entrant circuit developing around areas of scarring (Hayes and Gersony, 1986; Kanter *et al*., 2000). IART has been seen in around 10% at 5 years and 30% at 20 years of age, and often occurs in association with sinus node disease (Deanfield *et al*., 1988). Risk factors for flutter and IART include the occurrence of perioperative bradyarrhythmia, need for reoperation and sinus node dysfunction during follow-up.

The suture lines in Mustard and Senning patients are often complex. Conduction delay occurs in parts of the atria, inevitably giving rise to the substrate for re-entry. The many variations in the Mustard/Senning operation make delineation of the arrhythmogenic substrate difficult. The site of drainage of the coronary sinus, whether into the systemic venous atrium or pulmonary venous atrium, is critically important, as most Mustard and Senning IART appears to use the typical flutter isthmus (Kanter *et al*., 2000).

Ventricular arrhythmias have been occasionally reported but are much less common. Late sudden deaths have been seen in this patient population, but are more likely to occur in rapidly conducted atrial arrhythmias.

Functional Single Ventricle and Fontan Operation

The Fontan procedure offers surgical palliation to patients with a functional single ventricle. The primary arrhythmia following the atriopulmonary Fontan operation is IART, with atrial fibrillation also seen, but less frequently (Triedman, 2002). Retrospective studies from large cardiac surgical centres indicate that 25–50% of patients who have undergone the Fontan procedure will have clinical documentation of IART by 10 year follow-up. Frequently identified risk factors for IART include older age at operation and longer follow-up (Gelatt *et al*., 1994; Fishberger *et al*., 1997).

The substrate for atrial arrhythmias in these patients is thought to arise as a consequence of atrial hypertrophy and dilatation, surgical scarring and areas of fibrosis within the right atrium. This combination of factors leads to intra-atrial conduction block or delay, and the potential

for re-entry. Patients with cavopulmonary connections and lateral tunnel constructions appear to be at lower risk, because of the avoidance of atrial suture lines, although arrhythmias have also been documented in this population (Gandhi *et al*., 1996). As in patients with TGA who have undergone the atrial switch procedure, the development of atrial arrhythmias is often seen in conjunction with sinus node dysfunction.

In patients following the Fontan operation, re-entry barriers include atriotomy scars, the pulmonary artery/right atrial anastomotic zone, intra-atrial suture lines, AV valve rings, and residual AV valve tissue in those with tricuspid atresia. The multitude of barriers to conduction leads to many potential IART circuits, stressing the importance of rigorous electrophysiological evaluation in individual patients.

Other Defects

A variety of other congenital heart defects may be associated with late onset arrhythmia. Patients with Ebstein's anomaly of the tricuspid valve may have accessory pathway-mediated tachycardias, focal atrial tachycardias, atrial flutter and fibrillation. Those with corrected transposition are also at risk for accessory pathway-mediated tachycardias, AVNRT and atrial arrhythmias. Patients with mitral valve disease and a dilated left atrium are primarily at risk for atrial fibrillation. Ventricular ectopy may be seen in those with ventricular septal defects, and occasional patients develop sustained ventricular tachycardia.

SURGICAL ANATOMY

The surgical anatomy is described in Chapter 47.

CASE SELECTION AND PREPARATION

Knowledge of individual anatomy is essential when planning electrophysiological intervention. Review of the surgical case notes will allow assessment of the potential anatomical barriers for arrhythmia circuits and central obstacles for re-entry, such as atriotomy scars and patches. Documentation of clinical tachycardias via 12 lead electrocardiograms or Holter recordings is required to determine arrhythmia mechanisms and allow effective targeting of the arrhythmia substrate. Imaging modalities, including ultrasound, computed tomography and magnetic resonance imaging, are useful to delineate current anatomy, demonstrate potential isthmuses for ablation and detect thrombus in the chamber of interest.

In patients with CHD, arrhythmias are commonly seen secondary to haemodynamic lesions causing either volume or pressure overload. Adequate assessment of current haemodynamics is required in any patient presenting with an arrhythmia, and surgical correction of abnormal haemodynamics should be considered prior to ablation. For some patients with grossly enlarged atria or ventricles, surgical ablation may offer greater long-term arrhythmia-free survival, as well as the opportunity for chamber reduction surgery.

ELECTROPHYSIOLOGICAL STUDY

Equipment

Specialized equipment is required to perform an electrophysiological study and catheter ablation (Josephson, 2003).

Standard sites for electrogram recordings include the high right atrium, the superior tricuspid annulus (TA) in the region of the bundle of His, coronary sinus (CS, allowing simultaneous recordings of left atrial and left ventricular signals) and right ventricular apex (RVA). Multipolar catheters allow simultaneous recordings in the atria, coronary sinus, or ventricles (**Figure 10.2**). Standard tip ablation catheters (4 mm) have largely been superseded by 8 mm or irrigated tip catheters for complex ablations involving multiple applications of radiofrequency frequency (RF) energy.

Conventional Contact Mapping

Following characterization of arrhythmia subtype, mapping is performed to target an accessory connection, ectopic focus or critical isthmus. In patients with accessory or slow pathways, careful point-by-point mapping establishes the optimal site for energy delivery.

The ''typical'' atrial flutter circuit has as a critical isthmus the inferior portion of the right atrium between

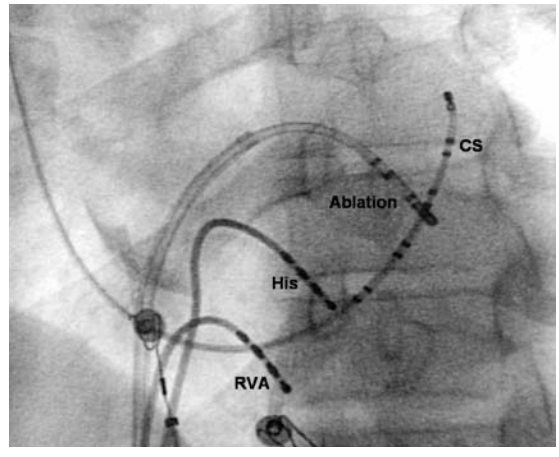

Figure 10.2

the tricuspid valve annulus and inferior vena cava (IVC). Mapping studies in CHD patients have demonstrated that, in addition to this isthmus, IART circuits may circle around non-conductive atriotomy scars or prosthetic material resulting from previous cardiac surgery. With these more complex circuits, traditional mapping techniques and programmed stimulation to assess the proximity of a mapping site to the re-entrant circuit have been utilized, but the multiplicity of potential circuits renders this approach difficult.

Traditional mapping techniques for assessing IART circuits in Fontan patients are particularly limited by the large size of the right atrium in those who have an atriopulmonary anastomosis. In the dilated right atrium, activation mapping during tachycardia remains problematic, despite the use of multiple-electrode catheters. This is compounded by the inability to reliably identify each catheter position in relation to an anatomical or surgical barrier. As a consequence, the primary success rates in ablating IART using conventional mapping techniques have been significantly lower than those achieved in patients with accessory pathways or AVNRT.

Advanced Mapping Techniques

The use of advanced mapping systems allows the correlation of electrical activity at any myocardial location with respect to underlying surgical anatomy. Each arrhythmia circuit can be plotted in detail and the optimal site established for ablation (Zipes and Jalife, 2004).

ABLATION STRATEGIES

Atrial Flutter and IART

Atrial Septal Defect

If the typical flutter isthmus is involved in the re-entry circuit, the area between the tricuspid valve (TV) and IVC is targeted for ablation (**Figure 10.3**). It is relatively narrow and in most patients good ablation catheter–tissue contact is achievable, although furrows and ridges may reduce tissue penetration with RF energy. Complete bidirectional conduction block across the flutter isthmus is the end point for ablation. IART may be re-entrant around a right atriotomy scar or, less usually, an ASD patch (Nakagawa *et al*., 2001).

Tetralogy of Fallot

The majority of patients with postoperative atrial flutter and TOF have circuits involving the typical flutter isthmus (Chan *et al*., 2000). Other isthmuses responsible for atrial flutter in postoperative TOF relate to atriotomy scars, breaks in the crista terminalis or areas of fibrosis. Ablation in relation to these circuits is performed by joining fixed anatomical structures or areas of scar that

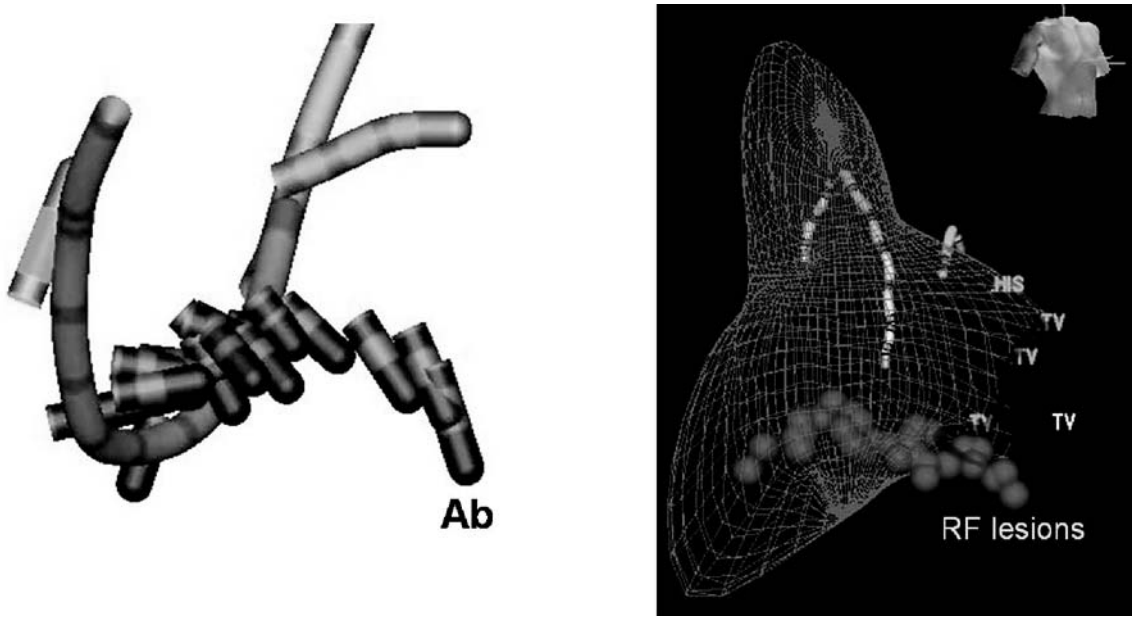

Figure 10.3

lie at either side of the critical isthmus (Nakagawa *et al*., 2001).

Transposition of the Great Arteries

In patients following the Mustard procedure, barriers to atrial conduction include the caval veins, atrioventricular valves, baffle suture lines and right atriotomy scar. In Senning patients the anatomy is more complex, with several potential lines of conduction block. These include, in addition to the above, multiple suture lines and the edge of the atrial septal remnant. In Mustard patients the most frequent site for successful ablation is in the region of the postero-inferior atrium near the coronary sinus ostium (Kanter *et al*., 2000).

Fontan

There are multiple potential re-entry circuits in the grossly dilated right atrium of the atriopulmonary Fontan patient. Central obstacles include surgical scars located on the free wall of the right atrium, atrial septal defects and the right AV valve, when present. In addition, areas of scarring and abnormal conduction at the lower border of crista terminalis may lead to periannular re-entry around the IVC. Ablation is directed to isthmuses, or ''channels'', between anatomical boundaries and areas of scarring, to produce conduction block (Nakagawa *et al*., 2001).

The use of complex mapping techniques, including both electroanatomical and non-contact mapping, and the use of irrigated tip ablation catheters, has improved ablation success rates. Procedure times remain long, however, and recurrence of IART may occur in at least one-third of patients. Surgical Fontan revision may be more appropriate in some patients with abnormal haemodynamics (Marcelletti *et al*., 2000) and surgical right atrial MAZE procedures with intraoperative cryoablation may be preferable in those with a very dilated and hypertrophied right atrium (Mavroudis *et al*., 1998).

Atrial Fibrillation

Atrial fibrillation may occur in any CHD patient with repetitive episodes of atrial flutter or IART, or may occur *de novo*. Patients predisposed to developing this arrhythmia more commonly have unrepaired defects or residual left-sided obstruction. The recognition that, in patients with structurally normal hearts, rapidly firing triggers, usually in the pulmonary veins (Haissaguerre *et al*., 1998), initiate and drive atrial fibrillation has led to similar efforts to identify the source of fibrillation in the CHD population. Ablation targeting a single source of fibrillation, such as a rapidly firing pulmonary vein, may result in AF termination, but recurrence rates with this strategy are high. More recently, a catheter-based MAZE procedure has evolved, with wide encircling RF applications around each pulmonary vein and additional linear ablation in the left atrium (Oral *et al*., 2003). This strategy has improved primary success rates and reduces the risk of medium-term AF recurrence.

Ventricular Tachycardia

Patients with ventricular dilatation, fibrosis or previous operative intervention are at risk of developing ventricular tachycardia. This is particularly seen in the patients with TOF who have undergone patch reconstruction of the right ventricular outflow tract (Lucron *et al*., 1999). Advanced mapping techniques are used to identify re-entry loops and corresponding channels between areas of scarring. Complex circuits may require ablation at several sites around the re-entrant zone to block all possible exit sites.

COMPLICATIONS OF CATHETER ABLATION

Catheter ablation in most patients is associated with low morbidity and a very low mortality risk. Serious complications relate to the risk of AV block in those with perinodal accessory or slow pathways, ectopic foci near the AV node, or flutter circuits that incorporate the interatrial septum adjacent to either AV valve. Damage to major vessels is rare, but left atrial ablation procedures may be complicated by pulmonary vein stenosis, even when energy delivery is directed to a site apparently remote from the pulmonary vein–left atrial junction.

Tamponade may occur with high-powered energy delivery in patients with thin atrial or ventricular walls. The risk of thrombus or char formation at the catheter tip has decreased significantly with the introduction of the irrigated tip catheter.

Acknowledgement

Figures 10.1 Modified by kind permission of author (Kalman *et al*.)

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11

Perfusion Techniques

T. Jones and M. J. Elliott

Cardiopulmonary bypass (CPB) is a form of extracorporeal circulation consisting primarily of a pump, a reservoir and an oxygenator. A wider review of cardiopulmonary bypass in neonates, infants and children may be obtained from the book edited by Jonas and Elliott (1994).

The first successful operation using total CPB was undertaken in 1953 (Gibbon, 1954), following the pioneering work of John Gibbon in the 1930s and 1940s. The subsequent development of the technique was driven by the need to undertake intracardiac operations in older children and young adults. Improved understanding, equipment and technique has enabled CPB to be undertaken routinely in all age groups, including neonates. While there are fundamental differences between adults and children undergoing CPB, the overall principles and objectives are the same—to provide a bloodless operative field enabling good surgical exposure with preservation of end organ function.

Prior to the advent of extracorporeal circulation, surgical correction of simple congenital heart defects was successfully being undertaken using total body (topical) hypothermia and cardiac inflow stasis. Working on ischaemic thresholds and inflow occlusion, Lillehei and Cohen discovered that by occluding all the venous inflow to the heart except the azygous vein, the resultant small cardiac output was enough to support all vital organs for a period of 30 minutes at normothermia (Cohen and Lillehei 1954). Prior to this discovery, attempts at total extracorporeal circulation had been using high flow rates intended to replicate cardiac output. Following further successes in the animal laboratory, Lillehei and colleagues (1986) undertook a series of human operations using ''controlled cross-circulation'', whereby intracardiac surgery was undertaken on a child using an extracorporeal circulation connected to the femoral artery and long saphenous vein of the child's mother or father to act as an oxygenator. A pump in the circuit regulated blood flow and volume. In essence, this ''cross-circulation'' resembled intrauterine circulation, with the parent's circulation correcting and maintaining that of the child. The technique was undertaken in 45 patients with acceptable short- and long-term results (Lillehei *et al*., 1986) and the principles that were established facilitated the modern development of CPB.

Following the successful application of ''cross-circulation'', interest was renewed in the search for a non-human oxygenator. Initially, canine and monkey lungs were used, but with poor results. The first series of eight patients undergoing intracardiac surgery at the Mayo Clinic using a pump oxygenator was published in 1955 (Kirklin *et al*., 1955). Also in 1955, despite concerns regarding the potential for gas embolism, DeWall and Lillehei developed (and used clinically) a disposable bubble oxygenator, with excellent results (Lillehei *et al*., 1956). The bubble oxygenator was further refined and remained in widespread use worldwide until the development of microporous membrane oxygenators, which have now become the standard in paediatric practice. There have also been important developments in materials science and circuit design, including reducing the size of the components which, allied with advances in surgical and postoperative care, have made complex neonatal surgery both achievable and successful.

PATHOPHYSIOLOGY

During total CPB, deoxygenated venous blood returning from the body to the heart is drained out of the circulation via venous cannulae into the artificial extracorporeal circuit. Typically, the blood collects in a reservoir, from which it is pumped through an oxygenator and heat exchanger before being returned to the patient, usually via an arterial cannula placed in the ascending aorta, thereby eliminating the need for blood to flow through the heart or lungs. This technique of CPB, in association with methods of myocardial preservation, enables the heart and lungs to be stopped and effectively emptied of blood for sufficient periods of time to enable surgical correction or intervention. Active cooling of the fluids in

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the circuit results in core cooling of the patient and thus a reduction in metabolic rate, allowing for a reduction in pump flow rate (cardiac output) and during periods of deep hypothermia (18°C) complete circulatory arrest.

Despite significant improvements in technique, CPB remains a non-physiological procedure. The effects of hypothermia, altered perfusion, haemodilution, acid–base management, embolization and the systemic inflammatory response need to be understood, and may be particularly marked in children and neonates. The specific challenges posed by neonates and infants are primarily related to their smaller circulatory volume and the relative immaturity of most organ systems at birth.

The normal changes in physiology in the first weeks of life, including the reactivity of the pulmonary vasculature, influence both technique and outcome of surgical repair. Pulmonary hypertension, present at birth, rapidly decreases within the first few months of life. The neonatal myocardium is relatively resistant to ischaemia but has limited functional reserve, being susceptible to increased afterload (systemic vascular resistance). Neonates have a higher metabolic rate, requiring higher pump flow rates and an impaired thermoregulatory response with greater dependence upon environmental temperature. Variability in heparin and protamine pharmacokinetics and immaturity of the liver may result in a coagulopathy that will be worsened by the presence of cyanosis. Renal perfusion and glomerular filtration is decreased due to increased renal vascular resistance, resulting in impaired sodium, water and acid–base regulation. While humoral and cellular immune factors are present, the immune response is impaired and either subnormal or, at the least, different. The underlying cardiac pathology may result in the presence of large intra- and extra-cardiac shunts that may need controlling during CPB in order to maintain adequate systemic perfusion and enable visualization of intracardiac defects. The development of collateral blood flow secondary to long-standing vascular obstruction or cyanosis may result in significant blood loss during surgical mobilization and poor visibility during surgery.

The combination of the above factors makes paediatric bypass a particular challenge. Patient age, size, underlying anatomy and surgical strategy influence the perfusion techniques and the construction of the CPB circuit. The bypass strategy is determined by the patient's needs. An understanding of the CPB pathophysiology is therefore required to make the correct choices and to optimize patient outcome.

HYPOTHERMIA

In non-cardiac surgical patients, the deleterious effects of intraoperative hypothermia with regard to increased infection, morbid cardiac events, blood loss, protein loss and length of hospital stay have been well documented. In contrast, during cardiac surgery, hypothermia has been used to reduce metabolic activity while preserving high-energy phosphate stores and increase tolerance to reductions in pump flow rates (cardiac output). It has also been used as a kind of insurance policy, potentially protecting the brain of the child against profound damage if there is mechanical failure of CPB components, now mercifully rare. At the extreme, deep hypothermia to 18°C enables the body to recover from periods of up to 45 minutes total circulatory arrest (Greeley *et al*., 1991), used by some for optimal surgical field exposure. The reduction in metabolic activity for every 10°C reduction in temperature is termed the Q_{10}. The effect is non-linear, such that a reduction in temperature from 37°C to 27°C is associated with a Q_{10} of 2.23 ml/100 g/min. The effect is greater at colder temperatures with a Q_{10} of 4.53 ml/100 g/min at 27–14°C (Michenfelder and Milde, 1991). There is evidence that the Q_{10} is higher for infants (Greeley *et al*., 1991), possibly secondary to an increased basal metabolic rate but implying that they may be able to tolerate longer periods of hypothermic circulatory arrest.

Mild to moderate hypothermia forms the basis of many myocardial and neuroprotective strategies during CPB. In the brain, the most important determinant of cerebral blood flow (CBF) is cerebral metabolic activity ($CMRO_2$) with regional and total CBF increasing or decreasing to match fluctuations in the metabolic demands of a region. This direct relationship is termed ''flow metabolism coupling''. General anaesthesia and hypothermia homogenizes metabolism and CBF, so that there is little difference between central areas or between grey and white matter. Cerebral metabolism is reduced 5–7% for each degree centigrade reduction in temperature, with a smaller but corresponding reduction in CBF (Stump *et al*., 1999). In health, CBF remains relatively constant over a wide range of cardiac output and mean arterial pressure via processes of ''perfusion-pressure autoregulation''. This effect is preserved during mild to moderate hypothermic CPB (> 22°C) with non-temperature-corrected blood gas management (alpha stat). Such processes help match oxygen supply to demand. The kidney is another organ that is capable of perfusion-pressure autoregulation, but this effect is disrupted during hypothermic and normothermic CPB, with renal blood flow being primarily determined by pump flow rate and secondarily by mean arterial pressure (Jones, 2005).

Laboratory studies have demonstrated the neuroprotective effects of hypothermia, with the most noticeable benefit occurring at 34–36°C (Busto *et al*., 1987). In contrast, hyperthermia (> 37°C) increases the extent and

severity of brain injury, with postischaemic hyperthermia occurring up to 24 hours after injury, worsening brain injury and outcome (Ginsberg and Busto, 1998).

ALTERED PERFUSION

Altered perfusion states during CPB are manifested by reduction in pump flow rate (cardiac output), mean arterial pressure, and the use of non-pulsatile perfusion during CPB. All have been implicated as causes of postoperative organ dysfunction. However, every type of artificial blood pump is associated with trauma to circulating blood components. In an attempt to limit this effect, to avoid large pressure gradients and to maintain a surgically dry field, a reduced blood flow rate continues to be used during CPB. In paediatrics the pump flow rate is determined by patient weight (**Table 11.1**).

Mechanisms of cerebral autoregulation maintain cerebral blood flow relatively constant over a range of arterial blood pressures and are well preserved during CPB (Murkin *et al*., 1987). Thus, whether it is more important to maintain pump flow as opposed to arterial pressure has been the focus of much research. Provided that mean arterial pressure remains in the autoregulatory range, fluctuations in pump flow do not result in significant changes in CBF (Schwartz *et al*., 1995). However, it is difficult to estimate the relevance of such studies because, in practice, pump flow and arterial pressure are inextricably linked. The exact limits of the autoregulatory range during paediatric CPB remain to be established.

The importance of pulsatility during CPB is also controversial, yet there are few data relating to children, largely because perfusion system manufacturers have little market stimulus to develop appropriate small pulsatile systems. In a porcine model of paediatric CPB, Lodge *et al*. (1997) demonstrated no difference in cerebral blood flow between pulsatile and non-pulsatile perfusion, but pulsatile flow was associated with an increase in renal and myocardial blood flow. Few of the pulse-generating systems available on the market produce a stroke volume small enough to be applicable to the paediatric population, and it remains unclear whether the pulse generated at the pump is transmitted to arterioles. To date there is no established clinical benefit to either technique.

Table 11.1 Pump flow rates during cardiopulmonary bypass.

Patient group	Estimated pump flow
Neonates	120–200 ml/kg/min
Infants (up to 10 kg)	100–150 ml/kg/min
Children	80–120 ml/kg/min
Adults	2.4 l/min/m^2

HAEMODILUTION

Before the extracorporeal circulation can be connected to the patient's circulation, it must be primed with fluid and de-aired to reduce the possibility of gaseous macro- and microemboli being delivered to the patient. Initiation of CPB therefore results in haemodilution, with a reduction in the plasma colloidal oncotic pressure that may result in a movement of fluid from the intravascular space into the inter- and intracellular spaces, causing tissue oedema. An increase in interstitial fluid was noted as early as 1970 (Pacifico *et al*., 1970) and this was subsequently noted to be more common in neonates (Mavroudis and Ebert, 1978). Maehara *et al*. (1991), using bio-electrical impedance methods, were able to demonstrate an elevation in total body water after CPB in children, with low body weight, small size, long duration of bypass, low temperature and low haematocrit being risk factors for the increased fluid accumulation.

The rationale for haemodilution is based on the need for hypothermia during bypass to permit reduction of cerebral and other organ metabolic rates. This enables the use of lower flows during bypass, thus reducing blood cell trauma and the amount of venous return to the heart, permitting good visualization of the inside of the heart. Haemodilution decreases the viscosity of blood, resulting in an increase in blood flow and an increase in tissue perfusion during CPB (Cooper and Giesecke, 2000). Hypothermia induces direct vasoconstriction and increases the apparent viscosity of blood and this effect is partly reduced by haemodilution. The average blood volume of a neonate is around 85 ml/kg; thus, a 3 kg child has a blood volume of approximately 255 ml. At Great Ormond Street, CPB prime volume for a 3 kg child is approximately 350 ml. Despite a lower ratio of 1:1, additional donor blood is still required in most cases in order to maintain an adequate haematocrit (> 30%). However, there is an increasing belief that donor blood should be avoided when possible. The risks of transmitted infection and the metabolic load (Ratcliffe *et al*., 1986), together with cost, availability and the objections of the patients' families, all argue against the use of donor blood.

Donor blood also has important metabolic consequences when added to the prime. There is a high sodium, potassium, glucose and lactate load associated with citrate phosphatedextrose (CPD)-stored blood (Ratcliffe *et al*., 1986), and this forms the dominant component of the substrate load provided to the patient at the onset of bypass. This load can be avoided by washing the prime

through the haemofilter, using the technique described by Ridley *et al*. (1990) at the onset of bypass. Recent work suggests that a relatively high haematocrit may provide optimal perfusion in both the brain and the rest of the body, contrary to the assertions of risk proposed above (Jonas *et al*., 2003). To date despite pressure to avoid donor blood and the apparent success of operating on Jehovah's Witness patients without blood (Tsang *et al*., 1994) there has been no systematic assessment of the safety and efficacy of blood vs. blood-free cardiac surgery in children. Such a study would need to be large and multicentred, but offers a real chance to quantify risk and develop sensible protocols.

INFLAMMATORY RESPONSE

At the initiation of CPB, blood is drained from the patient into the extracorporeal circuit. The circuit is predominantly made from polyvinyl chloride, silicone rubber and latex. Despite attempts to improve the biocompatibility of the blood–circuit interface, with processes of heparin-bonding or surface-modifying agents, the circuit is not covered by endothelium and therefore is recognized as ''foreign'' by the circulating blood components. This results in an inflammatory response, which occurs in all patients undergoing cardiac surgery to greater or lesser degrees. In its most severe format the systemic inflammatory response syndrome (SIRS) is characterized by a hyperdynamic circulatory state and increased capillary permeability, which may be accompanied by coagulopathy, fever, tachypnoea, leucocytosis or leucopenia.

The inflammatory response is complex and a complete review is outside the scope of this chapter. In general, activation occurs due to blood contact with non-endothelial surfaces and air within the extracorporeal circuit. In addition, the blood pump, membrane oxygenator, arterial cannula, cardiotomy suction and vent circuits generate shear stresses which damage red blood cells, leukocytes and platelets, leading to further activation of the inflammatory response. The response is mediated by cellular and non-cellular or humoral components of the blood. The cellular components are red blood cells, platelets and leukocytes, with the most significant mediator being the neutrophil. The key sequence in the non-cellular or humoral response is the activation of factor XII to factor XIIa, resulting in the activation of the kallikrein, intrinsic coagulation, fibrinolytic and complement pathways. The whole array of cytokines is turned on, with each child having a different response, almost certainly based on its genotype. We now know, for example, that genotypic differences in the pathways controlling IL-6 expression are crucial to postoperative recovery after cardiac surgery. Patients predestined to have a high plasma IL-6 response usually stay longer in the ICU after cardiac surgery. Similarly, failure of one's monocytes to express HLA-DR dramatically increases the risk of developing sepsis postoperatively (Allen *et al*., 2002). These observations have stimulated a wide body of work around the world, and genotype-targeted therapy will undoubtedly become part of our practice in the next decade.

Heparin is administered prior to CPB as an anticoagulant. It binds to and activates antithrombin III but it does not stop the activation of circulating factor XII to activated factor XIIa. Further, despite adequate heparinization, subclinical coagulation occurs due to the formation of fibrin, which in association with activated white cells and platelets may lead to the formation of microemboli consisting of fibrin, cellular aggregates and denatured proteins and lipoproteins, in addition to the release of endotoxins, vasoactive, cytotoxic, procoagulant and anticoagulant mediators.

An awareness of the inflammatory response has led to the development of strategies aimed at reducing its effect. The use of heparin-coating and surface-modifying agents to improve the biocompatibility of the circuit blood interface may reduce the response. Serine protease inhibitors, such as aprotinin, interact with the inflammatory response at many levels, including inhibiting the extravasation or transmigration of leukocytes from the microcirculation into surrounding tissue (Asimakopoulos *et al*., 2000). The technique of modified ultrafiltration at the termination of CPB is aimed at improving organ function by removal of excess total body water that may have accumulated in the tissues secondary to capillary leak.

The evidence for the use of corticosteroids during CPB is inconsistent. Steroids suppress inflammation in a number of ways. There is research demonstrating decreased levels of tumour necrosis factor and interleukin during CPB following the administration of dexamethosone (Jansen *et al*., 1991; Engelman *et al*., 1995), but other work suggests an impairment of host defences and elevated levels of endotoxins following treatment (Andersen *et al*., 1989). The difference in outcomes may be explained by the timing of steroid administration, with maximal beneficial effects following high-dose steroids given several hours prior to CPB (Lodge *et al*., 1999).

All of these interventions are likely to come under renewed focus once the role of the genotype in various aspects of the inflammatory response is understood. Monotherapy, such as blocking a complement receptor, makes little sense if it is only going to work in a limited number of patients.

Emboli

Intravascular emboli occur during all types of extracorporeal circulation. In adults, the number of emboli detected intraoperatively has been demonstrated to be associated with postoperative neurobehavioural deficits. The adult atherosclerotic aorta has been identified as a significant source of intraoperative emboli. This is obviously less of a problem in paediatric surgery, but gaseous and other types of particulate emboli can and do occur. They can also be more significant if there are intracardiac communications, such as a VSD or ASD, since venous emboli can pass directly to the systemic circulation.

Emboli may be classified according to their composition or source, with emboli being either biological (blood-borne), non-biological (foreign to host) or gaseous. Biological emboli may consist of debris from disruption of calcific plaques, remnants of damaged cells, platelets, neutrophils and cellular aggregates, fibrin, chylomicrons and lipids from the operative field. Non-biological emboli may comprise plastic fragments generated by the pump or flushed from the circuit components, aluminium, silicone antifoam and glove powder. Gaseous emboli may be inadvertently introduced into the circulation by the surgeon or perfusionist (Taylor *et al*., 1999) or they may be generated by the components of the circuit through entrainment, cavitation or temperature and pressure changes (Mitchell *et al*., 1997). There is significant variability between individual products in the ability of the circuit components to remove entrained gaseous microemboli (Jones *et al*., 2002a).

THE EXTRACORPOREAL CIRCUIT

Prime

The composition of the prime remains controversial. The recipe for the make-up of the CPB prime depends on a number of factors: (a) the desired haematocrit on bypass; (b) the perceived requirement for colloid osmotic pressure maintenance during bypass; (c) the need for such additives as mannitol, bicarbonate, steroids or heparin; and (d) the temperature at which bypass is performed. Current composition of prime used at Great Ormond Street is shown in **Table 11.2**. The overall volume will vary slightly, depending on the patient's size and placement of the reservoir level sensor.

Acceptable levels of haematocrit are not yet clearly defined and they vary between 20% and 30% for neonatal deep hypothermic bypass (Elliott, 1993a). Haemodilution increases cerebral blood flow velocity (Gruber *et al*., 1999) and at extremes (~10%) it is associated with impaired oxygen delivery and delayed cerebral recovery following hypothermic circulatory arrest (Shin'oka *et al*., 1996). The use of a haematocrit of 30% is likely to require the addition of more donor blood than that of a haematocrit of 20%. We currently maintain a haematocrit of > 30% for deep hypothermia (under 25°C), 30–35% for moderate hypothermia (25–28°C) and > 35% for modest hypothermia (approximately 32°C). We have pointed out earlier the absence of comparative data with regard to the risk–benefit of blood transfusion. Advances in the use of ultrafiltration argue in favour of the acceptance of lower haematocrits during CPB and predict the potential avoidance of donor blood altogether. However, in a recent study (Jonas *et al*., 2003), 147 infants less than 9 months of age were randomized to CPB with either low (20%) or high haematocrit (30%). The low haematocrit group had reduced cardiac index and an increased serum lactate and total body water content postoperatively. At 1 year of age they had worse psychomotor development. There was no standardization of intraoperative temperature, but the group reported there was no interaction between haematocrit, temperature and early and 1 year outcomes. More work needs to be done in this area and we need to know the relative long-term risks of blood donor exposure in our patients.

Table 11.2 Pump prime for a circuit up to 800 ml/min flow (3.1 kg patient).

Plasmalyte A	120 ml
Gelofusin	120 ml
Packed red cells	100 ml
Mannitol 20%	11 ml (2.5 ml/kg)
Heparin	2000 IU
Sodium bicarbonate 8.4%	10 ml
Calcium chloride	0.3 ml
Total	361 ml

Surprisingly little is known about the appropriate colloidal solutions to be added to the bypass circuit; however, a variety of substances are used in routine clinical practice, viz. human albumin (5% or 20%), fresh frozen plasma and the synthetic colloids, such as hetastarch, dextran and gelofusin. In addition, mannitol and bicarbonate are added by most units. PlasmaLyte 148 and PlasmaLyte A are probably superior to Hartman's solution because they avoid an unnecessary lactate load. Glucose-containing fluids should be avoided in order to prevent massive osmotic shifts at the beginning of bypass and to avoid hyperglycaemia (which may be induced by cold or stress-induced insulin intolerance) during times of potential neurologic damage. Hyperglycaemia appears to inhibit recovery from neurologic insult. If glucose, sodium and potassium concentrations in the prime are

high before the onset of CPB, the prime should be washed (Ridley *et al*., 1990). This strategy is particularly appropriate for the very small sick neonate, who may be very intolerant of electrolyte and osmotic shifts.

The priming volume should be kept as small as possible to reduce haemodilution and avoid blood usage. This can be achieved by the utilization of appropriately sized equipment with smaller priming volumes. Correct positioning of the CPB apparatus as close to the patient as possible avoids unnecessary lengths of tubing. One metre of 1/4 inch tubing contains 32 ml blood. Thus, if two or three suckers are used in a bypass circuit in addition to the arterial and venous line, the tubing alone can accommodate almost the entire blood volume of the patient. A reduction in prime volume and donor blood usage can be achieved simply by attention to these details. For example, we now use 3/16 inch tubing in neonates. Many surgeons and perfusionists have demanded smaller circuits from the manufacturers over the years, but market forces (too few unit sales) prevent them from undertaking the required development work.

Venous Reservoirs and Venous Drainage

Blood drains from the patient via venous cannula into a venous reservoir before being pumped through the circuit. The reservoir acts as a holding tank, allowing fluctuations in the patient's circulatory volume to be tolerated. **Figure 11.1** shows a diagram of the cardiopulmonary bypass circuit used at our department. Line 1 is the venous line from the right atrium or superior vena cava (SVC) to the venous/cardiotomy reservoir (A). Line 2 is the venous line from the reservoir to the arterial pump (B). Line 3 takes the blood from the arterial pump (B) to the oxygenator/heat exchanger (C). Line 4 takes the blood from the oxygenator via an in

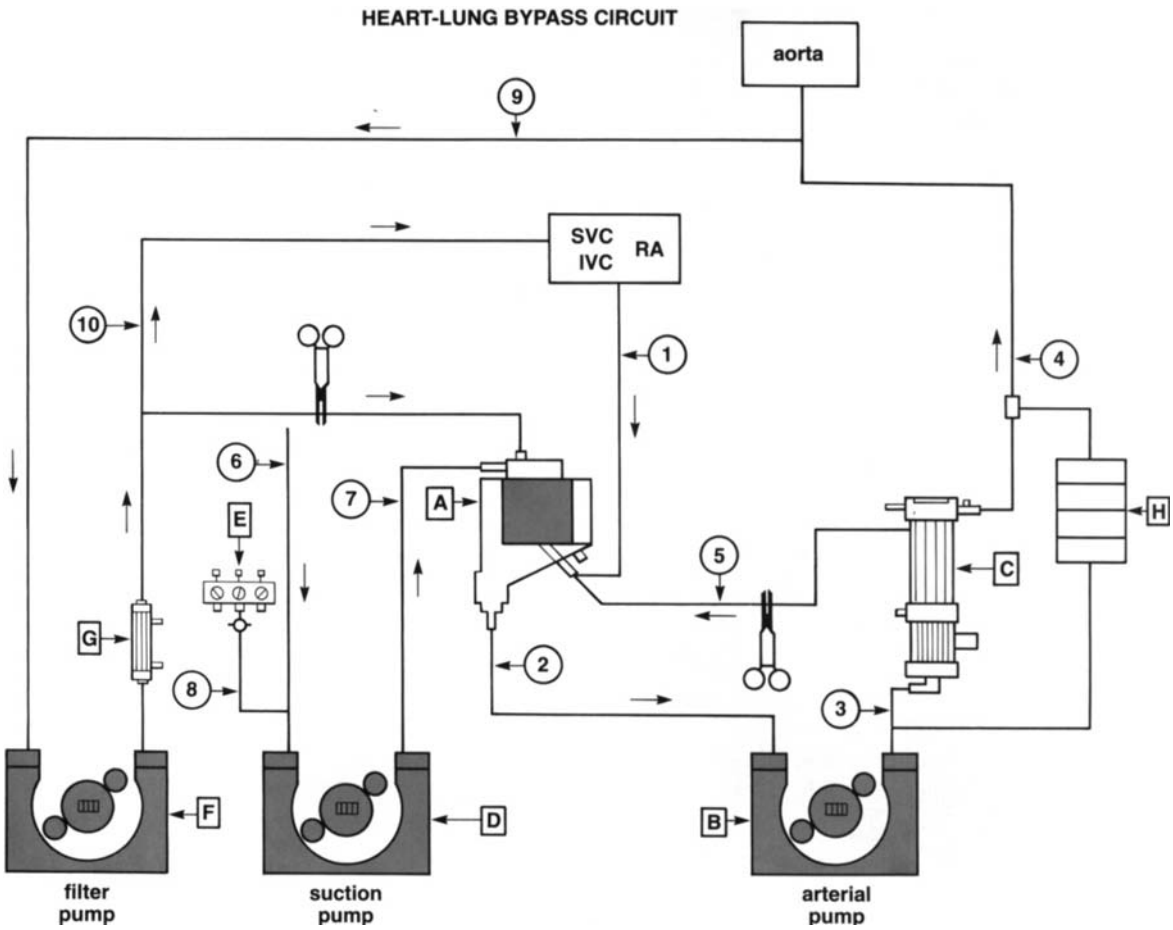

Figure 11.1

line arterial filter (40 μm) to the aorta. Line 5 is a shunt from the oxygenator (C) to the venous/cardiotomy reservoir (A). This shunt is clamped during bypass. Line 6 represents a suction line from the patient to the suction pump (D), from which line 7 carries the blood to the combined venous/cardiotomy reservoir (A). Line 8 is a suction bleed line to a pressure regulator (E). Line 9 shows the inlet line to an ultrafilter (G) via a filter pump (F) (described later in the section on ultrafiltration). This inlet line comes from a three-way connector inserted into the arterial line close to the arterial cannula from the filter (G); line 10 passes to its point of insertion in the right atrial appendage. To permit priming and prebypass ultrafiltration, line 11 provides shunting to the venous/cardiotomy reservoir (A). Block (H) represents monitoring equipment (e.g. reservoir level, line pressure, bubble activity and oxygen pressure).

During periods of low or no flow, the patient can be exsanguinated into a high-capacitance reservoir. Blood, drugs and fluid can be added to the reservoir via filtered ports. There are two types of venous reservoir in general use: a hard shell and a soft shell. The hard shell is made from a rigid plastic container. It also referred to as ''*open system*'' because there is an air–blood interface present. In contrast, the soft shell is made from a distensible plastic bag and, as such, there is no air–blood interface (''*closed system*''). The hard shell reservoir handles entrained air more efficiently and can be used with vacuum-assisted venous drainage. The soft shell reservoir has been described as associated with less bleeding and a less marked inflammatory response, possibly secondary to exclusion of the air–blood interface (Schonberger *et al*., 1995).

As CPB starts, blood is siphoned from the patient into the reservoir by gravity. The use of small lumen venous cannulae and cannulation remote to the heart may prevent adequate venous drainage. This has led to a renewed interest in methods of assisted venous drainage, using either a centrifugal or roller pump in the venous line (*kinetic-assisted venous drainage*; KAVD) or by applying a vacuum to the top of the hard shell venous reservoir (*vacuum-assisted venous drainage*; VAVD). Both systems have the potential to aspirate air into the circulation, with the potential to develop an air lock when using KAVD (LaPietra *et al*., 2000). Micro-air embolism remains a major concern (Walther *et al*., 2002). In contrast, VAVD at low levels of suction ($\leq$ 40 mmHg) does not appear to influence the ability of the circuit to deal with entrained air (Jones *et al*., 2002b).

Blood Pumps

There are currently two types of blood pumps in use. The *roller pump* consists of a rotating arm which compresses silastic tubing between the parallel wall of the roller and a backing plate, causing positive forward displacement of blood in a continuous flow. It is therefore occlusive. Rate of flow is dependent upon the speed of rotation of the rotor arm. Careful adjustment of the rotating arm is required to provide forward blood flow and yet minimize damage to the cellular components of the blood. *Centrifugal blood pumps* utilize a rapidly rotating impeller or inverted cone enclosed within a rigid plastic housing, from which blood is propelled. The impellers or cones are magnetically coupled to an electric motor and, when rotated, rapidly generate a pressure differential that causes the movement of blood. Unlike roller pumps they are non-occlusive, resulting in less damage to blood cells and platelets. They are afterload-dependent, meaning that an increase in downstream resistance secondary to a line obstruction or occlusion will decrease forward flow. For these reasons they tend to be favoured for more prolonged usage, such as during extracorporeal membrane oxygenation (ECMO). There is as yet no proven clinical advantage for either type of pump when used for routine CPB.

Oxygenators

There is currently no role for the bubble oxygenator in paediatric practice, unless there are strong economic reasons for its use. In comparison to membrane oxygenators, they are associated with increased production of gaseous microemboli, protein denaturation, increased haemolysis and complement activation (van Oeveren *et al*., 1985). In addition to gaseous exchange, membrane oxygenators also remove gaseous microemboli from the circuit (Jones *et al*., 2002a).

Of the membrane oxygenators available, three types dominate the market: (a) a hollow fibre oxygenator, which is really a microporous membrane (e.g. Dideco 701 or Terumo); (b) a variable prime flat sheet membrane oxygenator, also microporous (e.g. Cobe VPCML); (c) the Silastic oxygenator, which is a truly semi-permeable membrane oxygenator (e.g. Scimed). These have been used predominantly for longer-term bypass, as in extracorporeal membrane oxygenation (ECMO), but a small number of units use them primarily for bypass in children. Many of these oxygenators come with integral or associated venous and cardiotomy reservoirs and an integral heat exchanger. All membrane oxygenators have relatively large priming volumes but the hollow-fibre products currently have the lowest prime requirements.

Heat Exchanger

The heat exchanger, integral to modern CPB circuits, is capable of rapidly changing the temperature of

blood returning to the patient over a range of temperatures (8–41°C). Such is their heat transfer capacity that modern heat exchangers easily override normal thermoregulatory mechanisms. Consequently, the temperatures of the blood, tissues and organs become dependent upon the perfusionist. Temperature should be monitored in at least two places, viz. close to the most vulnerable organ, the brain (nasopharynx or tympanic membrane), and peripherally, to judge the effectiveness of peripheral perfusion and rewarming. In addition to reducing metabolic rate and increasing the apparent viscosity of blood, hypothermia increases the solubility of gases in biological fluids. This results, in association with a direct reduction in the dissociation constant of water, in significant disruption of the normal acid–base balance relationships.

CONDUCT OF CPB

Heparin Dosage and Heparin Neutralization

Heparin is given prior to cannulation at a dose of 3 mg/kg of body weight. More heparin may be added, depending on the activated clotting time (ACT), which is monitored at frequent intervals during bypass to maintain an ACT of more than 450 seconds. A baseline ACT measurement is performed before heparinization and a second measurement before the institution of bypass, in order to ensure adequate anticoagulation. Heparin binds to and activates antithrombin III. It is not a uniform substance, and its activity varies. Thus, monitoring of ACT is crucial for maintaining adequate anticoagulation. After cessation of CPB and confirmation that bleeding is not severe and that bypass will not be required again, the heparin is reversed with the use of protamine, in the ratio of approximately 1.5 mg protamine to 1 mg circulating heparin. This usually represents an underestimate, and more protamine is added as needed, on the basis of ACT measurements. Recently, many groups have been experimenting with thrombo-elastography as a means of monitoring the management of postoperative bleeding, but more work needs to be done to routinely recommend it.

The use of aprotinin, which is being used increasingly in reoperations and complex neonatal surgery, can artificially prolong the celite ACT (Wang *et al*., 1992), leading to inadequate heparinization. This can be overcome either by maintaining the ACT in excess of 750 seconds when using aprotinin or by using a kaolin ACT, which is less affected by aprotinin.

SURGICAL ASPECTS

After suspension of the pericardium to the wound margins, the heart is inspected. The pericardial reflection

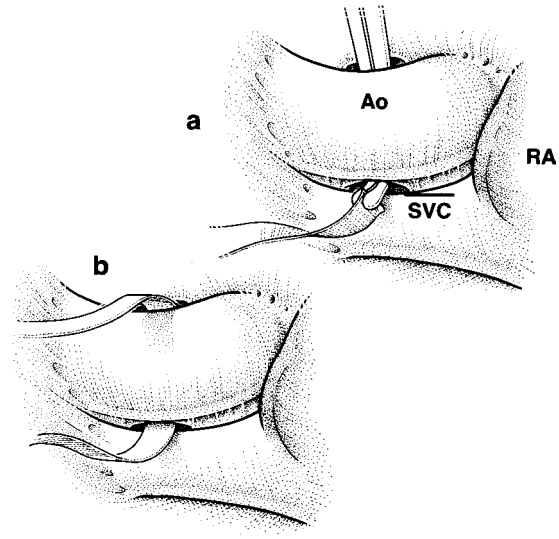

Figure 11.2

between the ascending aorta and the main pulmonary artery is routinely exposed and divided with diathermy. A right-angled dissector can then be passed through the incision and around behind the aorta, anterior to the right pulmonary artery. A nylon umbilical tape can then be passed around the aorta and secured with a small clamp (**Figure 11.2**). This tape aids in subsequent aortic cross-clamping and can assist in mobilization of branch pulmonary arteries by suitable retraction. The visceral pericardium overlying the right pulmonary artery is then divided just to the left of the SVC, and a right-angled dissector is then passed from the right aspect of the SVC and under the SVC, to emerge in the divided visceral pericardium to its left. A tape (or, in very small children, a thick silk ligature) can then be passed around the SVC in preparation for snaring that vessel (**Figure 11.3**). Remaining snares and tapes can be positioned when on bypass during the early stages of cooling. The inferior vena cava (IVC) is then encircled with a tape, with the use of a large, fully curved dissector (**Figure 11.4**). Close proximity of the right phrenic nerve should be noted.

Cannulation

The aim of cannulation is to provide adequate flow, unobstructed venous drainage, unobstructed perfusion of all organs and an unobstructed operating field.

Arterial Cannulation

The site of arterial cannulation will vary, depending on the requirements of the individual operation and

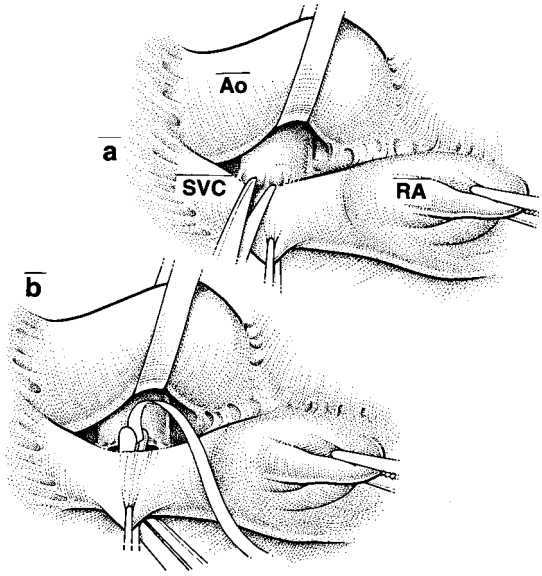

Figure 11.3

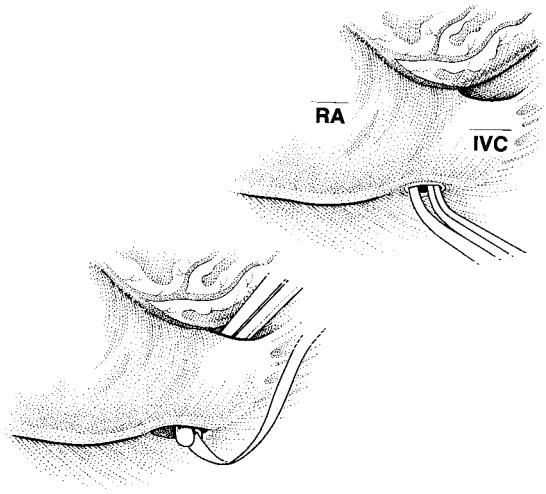

Figure 11.4

the morphology of the circulation. Some situations are obvious, e.g. if the surgeon is to operate on the aortic root, the cannula should be positioned as distally as possible. However, more subtle positioning is also helpful. In an operation for tetralogy of Fallot, most of the surgical procedures take place on the right ventricular outflow tract or the pulmonary arteries, to the left of the aorta, and thus it is helpful to position the aortic cannula slightly to the right. The opposite is true for a Fontan or total cavopulmonary connection (TCPC) operation, in which the aortic cannula is better positioned slightly to the left.

The placement of the purse string suture is important. The aorta may be very small in relation to the cannula, and an incorrectly placed or oversized purse-string suture may result in an iatrogenic stenosis. Thus, it is wise to position the purse-string suture in a longitudinal orientation and to limit its size to just greater than the chosen cannula. A horizontally orientated purse-string suture may result in a stenosis at the end of the procedure, with exaggerated obstruction to cardiac ejection and in distal hypotension. The choice of cannula is largely one of personal preference, although its size can be determined from published tables relating cannula size to flow rate. Our own preference is for the DLP arterial cannula (Medtronic).

Several methods for inserting the cannula have been described. Direct cannulation through a stab incision is a controlled and safe technique (**Figure 11.5**). The adventitia within the purse-string suture is incised transversely or vertically with the use of either very fine pointed scissors, a No. 11 or a Beaver scalpel blade. Once this incision has been made, the adventitia can be gripped above or to one side of the incision by a pair of forceps held in the left hand, after which a No. 11 scalpel blade, or equivalent, can be advanced slowly through the aortic wall, within the purse-string suture; the flap of adventitia is used to control any haemorrhage. As the knife is withdrawn, the hole can be closed by the adventitial flap. The aortic cannula can then be taken in the right hand and slid into the incision by simultaneously

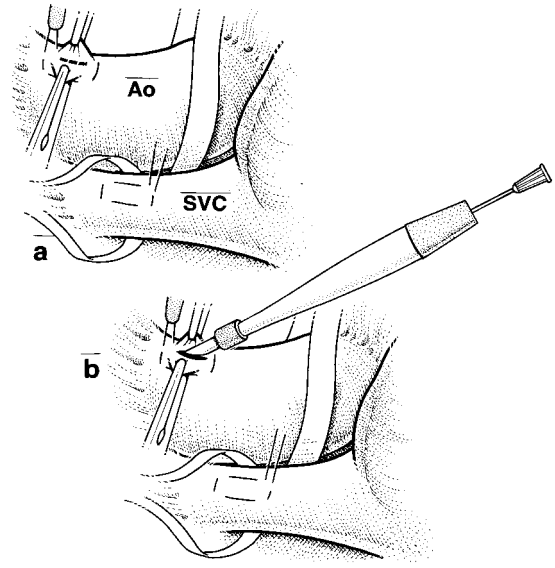

Figure 11.5

moving aside the adventitial flap. The purse-string suture can be snugged and the cannula tied to the rubber of the purse string snugger to prevent displacement. Any air bubbles must be tapped out of the cannula before it is connected to the arterial line, and it is helpful if the perfusionist slowly advances blood through the arterial line, in order to de-air the line at the same time that the aortic cannula is de-aired, by gradually removing the clamp. Fixing the cannula to the edge of the wound using a strong silk suture avoids movement and keeps the cannula away from the operating field.

Retrograde cannulation from an iliac or a femoral artery is indicated either when the ascending aorta is severely affected by the congenital anomaly (leaving insufficient room for cannulation) or when resternotomy is considered dangerous (e.g. if there is an aneurysm of the aorta or the right ventricular outflow tract or an adherent conduit). The iliac vessels are larger than the femoral vessels and, if required, can be approached via a muscle-splitting incision in a position similar to that used for appendectomy and through an extraperitoneal approach to the vessels as they overlie the rim of the pelvis. Once the vessels are exposed, umbilical tapes can be passed around them and snuggers prepared. A transverse incision should be made in the anterior surface of the iliac artery before cannulating the vessel with either an aortic cannula, pushed in as far as it will go in a cephalad direction, or with a specifically designed DLP femoral arterial cannula, which has an excellent orifice: overall diameter ratio. Again, the snuggers can be tied to the cannula and the cannula fixed to the skin edge to prevent displacement. The incision in the vessel is repaired by transverse closure with 7-0 Prolene at the end of the operation. Snugger-induced damage to the artery and the vein can be minimized by using a small piece of rubber tube or a Silastic vessel loop within the snare. Through the same skin incision, the iliac vein or, often, the IVC can be cannulated, should venous cannulation by this route also be preferred. Reinforced DLP femoral venous cannulae can be advanced high up the IVC toward, or even into, the right atrium. Full flow can often be maintained via these cannulae. Cannulation via this route can be very rapid and is a useful escape route, should problems be encountered during median sternotomy.

Venous Cannulation

Single Venous Cannulation

This is usually the method of choice for operations certain to involve circulatory arrest, or those in which the left side of the heart requires attention in the absence of intracardiac shunts. However, it is also preferred by some teams when low-flow bypass is required or when surgery on the inside of the heart is minimal (e.g. the situation in the arterial switch procedure with intact ventricular septum). The choice of cannula is important for facilitating positioning within the right atrium away from the field of surgery and for maintaining adequate venous drainage. For example, in the arterial switch operation, some surgeons prefer to use a short right-angled Rygg cannula, positioned via the right atrial appendage (**Figure 11.6**). A purse-string suture is usually placed around the atrial appendage, with the use of three or four bites of tissue. It is wise to position the snugger away from the field of surgery. After the snugger has been tied to the cannula, it is sometimes helpful to leave the ends of this tie long and clip those to the surrounding towels in order to make sure that the venous cannula is positioned out of the surgeon's way. Cannulation of the right atrial appendage for single venous cannulation can be done in many ways, e.g. with the use of a side biting clamp, such as a Cooley or Castenada clamp, or more simply by placing DeBakey forceps across the base of the appendage and then cutting off the tip of the appendage before positioning the cannula and releasing the DeBakey forceps, at the same time that the purse-string suture is snugged down. The cannula can also be positioned by making a stab wound in the centre of the purse-string suture and then enlarging this stab wound with a pair of scissors or artery forceps.

Bicaval Cannulation

Bicaval cannulation is the most frequently used technique in paediatric cardiac surgery because it provides complete venous return and permits exposure of the inside of the heart. Bicaval cannulation may be either indirect or direct.

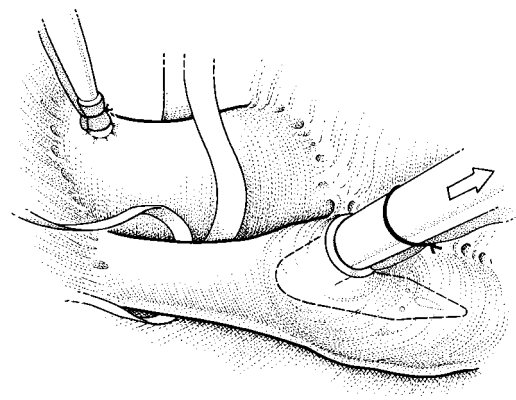

Figure 11.6

For *indirect bicaval cannulation*, purse-string sutures are placed in the right atrial appendage for the SVC cannula and in the low right atrium close to the mouth of the IVC for the IVC cannula. The cannula for the SVC should be long enough to pass beneath the ridge of the crista terminalis and well up into the SVC. The cannula for the IVC, however, should be shorter and designed in such a way that its tip lies just below the level of the diaphragm, so as to avoid obstructing venous return from the liver during bypass. The SVC cannula is usually inserted first (**Figure 11.7a, b**) and must pass beneath the ridge of the crista terminalis of the atrium to pass into the SVC. Care must be taken to avoid pushing the tip of the cannula through the atrial wall or through an atrial septal defect into the left side. Bypass can then be established on one cannula before either inserting the purse-string suture for the IVC or cannulating the IVC (**Figure 11.8a, b, c**). Once bypass is established, full flow can usually be obtained through this single cannula and ventilation may be stopped. This makes it much easier to place a purse-string suture low in the right atrium and to cannulate the IVC. The surgeon must avoid catching the cannula in the Eustachian valve, which guards the entrance to the IVC. Once cannulation has been achieved and the clamps have been removed, the cannulae are fixed in the usual way and a second umbilical tape or a thick silk ligature can be passed around the IVC.

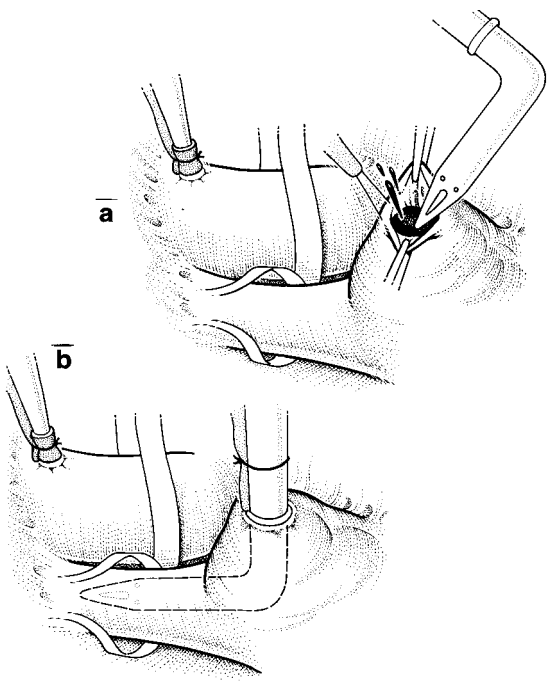

Figure 11.7

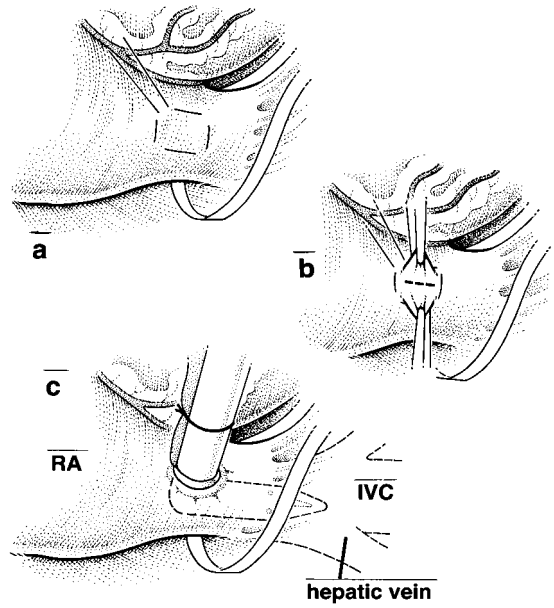

Figure 11.8

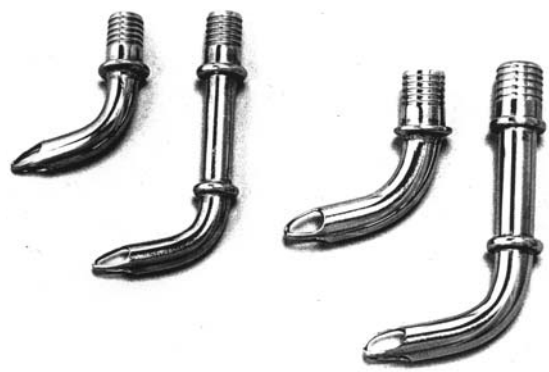

Figure 11.9

Since the advent of fine sutures and thin-walled metal cannulae, the use of *direct bicaval cannulation* has become standard for most intracardiac surgery performed by those who prefer continuous flow CPB. **Figure 11.9** shows our own specially designed venous metal cannulae, which have a caged tip and are relatively short. They are different from the DLP metal cannulae which have a sharp end opening. The purse-string suture should be positioned in the anterior aspect of the SVC, about 0.5 cm above the junction with the right atrium. The orientation of the purse-string suture can be critical, because SVC narrowing is a serious

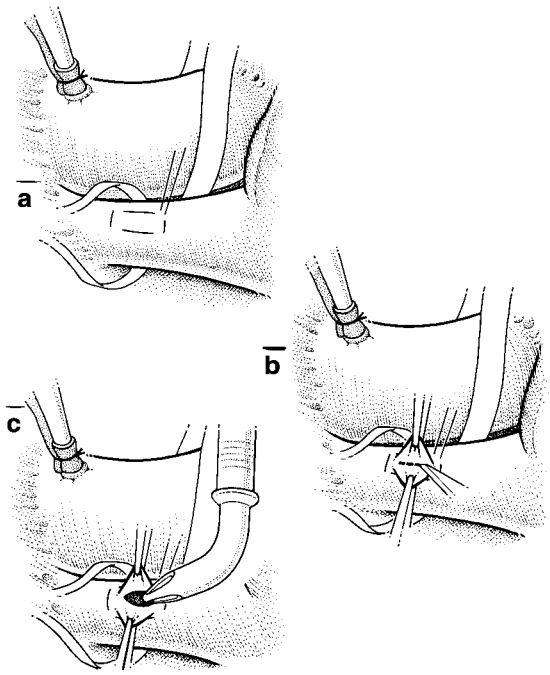

Figure 11.10

potential complication after cannulation. Thus, the purse-string suture should be orientated longitudinally and not be too large (**Figure 11.10**). Cannulation through this suture requires the placement of either a fine-tipped mosquito forceps, thereby distracting the vessel; a No. 11 scalpel blade is placed through the centre of the purse-string suture, and the mosquito forceps are used to control haemorrhage. A small mosquito forceps can then be used to dilate the incision, if necessary, before the cannula is passed through the incision into the SVC. The snuggers are tightened, and the cannula tied to the rubber of the snugger. At the end of bypass, the SVC cannulation site can usually be repaired by simply tying the appropriately sized purse-string suture or, occasionally, by fine transverse sewing of the longitudinal incision within the purse-string suture.

For the IVC cannulae, it is better to release the pericardial reflection, anterior to the IVC, in order to expose the immediate subpericardial IVC down to as far as the first hepatic branch. A purse-string suture can then be inserted into the IVC, below the pericardial reflection, during CPB. This will avoid lowering the cardiac output by compressing the right atrium. A direct stab incision into the centre of the purse-string suture can be created and the cannula positioned through the incision before fixing it in place.

Cannulation can also be carried out in a different sequence. This requires the placement of an additional purse-string suture in the right atrial appendage. The venous cannulae can be connected to a Y connector, and the first cannulation can be made by using the IVC cannula via the right atrial appendage purse-string suture to establish bypass. Once bypass is established via this route, the SVC cannula can be inserted, after which the IVC cannula can be clamped and a cardiotomy suction catheter placed into the right atrial appendage to maintain the flow, while the IVC purse-string suture is inserted and the IVC cannula positioned within the IVC, unclamping the line. This is the least haemodynamically disturbing form of cannulation.

Air in the venous line results in gaseous microemboli being delivered to the patient (Jones *et al*., 2002b). Prior to commencement of CPB, the venous line should be de-aired, either by filling the line with saline or by applying a little pressure on the abdomen to elevate venous pressure and allowing blood to pass back up in the cannula before connection.

Venting

There may be considerable venous return to the left atrium during bypass, particularly if cyanosis has resulted in excessive bronchial collateral flow. An inadequately controlled shunt or an unsuspected patent ductus arteriosus may also result in increased left atrial return, which, uncontrolled, would cause left ventricular distension and damage the myocardium. It is very important to remember that cannulation of the left side of the heart with a vent carries some risk, particularly if the heart is beating and the aorta is unclamped. Diastole is an active phase of the cardiac cyle and an open vent site can thus suck air into the left side of the heart, ready to be ejected through the aortic valve and be sent on its way to the brain. Many lawsuits testify to this over the years. For this reason, whenever possible, the vent should be inserted after the aorta is clamped, unless the heart is fibrillating or demonstrably distended.

There are three approaches to venting the heart:

1. Left atrium (direct or indirect).
2. Pulmonary artery.
3. Left ventricle.

If the right atrium is open and if there is a communication between the two atria, an indirect approach to the left atrium by placing a vent across the atrial septum is usually adequate. If the septum is closed, the vent is positioned either through an incision in the fossa ovalis or, more often, directly into the left atrium via the junction with the right superior pulmonary

vein. A useful manoeuvre is to place a right-angled instrument through the patent foramen ovale and back into the right superior pulmonary vein, withdrawing it slightly to the junction with the left atrium. At this point, the tips of the instrument can be pushed out to the edge of the atrial wall so that they can be seen externally, and a No. 11 scalpel blade can be passed between the tips of the instrument, thereby ensuring that the knife does not pass either directly across the superior pulmonary vein or out through the back of the left atrium (**Figure 11.11a.**). Once this incision has been made, a vent can be positioned through it and a purse-string suture can be placed around it. This method produces a very secure and accurate placement of the vent (**Figure 11.11b**). The vent itself should be designed so that it works efficiently at a low flow rate without sucking in the atrial or ventricular tissues. The one commonly used is the left heart vent catheter by DLP.

In larger children, or if the heart begins to distend before the right atrium is opened, a direct external stab is quite safe. Vents can also be placed via purse-string sutures at the left atrial appendage, in the left atrial wall between the superior and inferior pulmonary veins on the right, or superiorly below the right pulmonary artery, directly to the left atrium. Should both left and right sides of the heart be too full at any stage of the procedure, it may be helpful to make an incision in the superior surface of the main pulmonary artery and insert a vent into the pulmonary artery. This procedure has the effect of both decompressing antegrade flow into the pulmonary arteries and, because the pulmonary circuit is valveless, decompressing the left atrium as well. This technique is particularly useful in obstructed TAPVC.

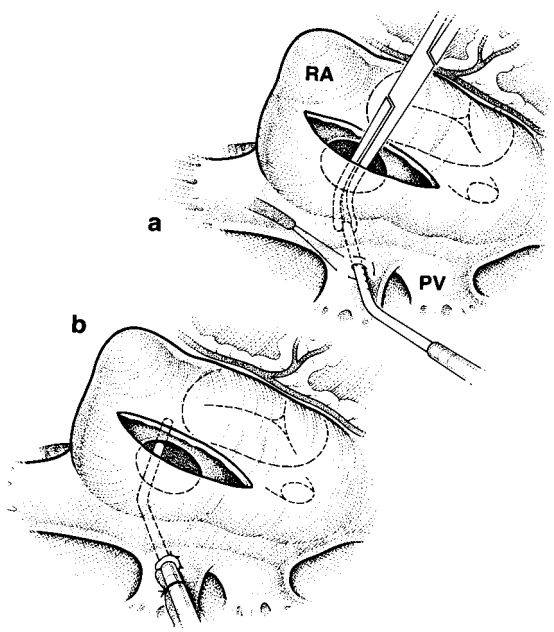

Figure 11.11

Direct venting of the left ventricular apex is used less frequently in children. Left ventricular apical venting can undoubtedly cause problems ranging from persistent haemorrhage to ventricular rupture, but perhaps of more importance, especially in small infants, are the potential haemodynamic consequences of displacing the apex of the heart toward the end of the procedure in order to close the vent site.

Intracardiac Suction

Typically, three suction heads are used to provide three suction lines to the patient. One of these acts as a vent, and the other two are available for intracardiac or pericardial suction. A sump sucker is connected to one of these, and a hand-held metal-tipped sucker is connected to the other. The tubing to these suction devices should be as short as possible. For neonates, we use only two suction lines to reduce such prime loss. It is important to use low-pressure suction to avoid haemolysis.

BLOOD GAS MANAGEMENT

Alpha Stat or pH Stat Mode

The optimal management of acid–base and arterial carbon dioxide tensions during CPB has been the focus of much research and debate. The solubility of carbon dioxide is influenced by temperature. Decreasing core body temperature increases carbon dioxide solubility, resulting in a reduction of blood $PaCO_2$ despite no change in total carbon dioxide content. Acid–base balance is defined as maintaining a pH of 7.4 and an arterial $PaCO_2$ of 40 mmHg at *normothermia* (37°C). During ''pH stat'' management, the *temperature-corrected* $PaCO_2$ is maintained at 40 mmHg by the addition of CO_2. During ''alpha stat'' management, *non-temperature-corrected* blood gas management is employed. At 28°C the difference in $PaCO_2$ between the two techniques is approximately 15 mmHg $PaCO_2$.

During alpha stat management, intracellular pH, enzymatic activity and perfusion-pressure autoregulation is preserved (Murkin *et al*., 1987). In contrast, during ''pH stat'' management, autoregulation is lost and cerebral perfusion is in excess of metabolic demands. The increased cerebral blood flow improves brain cooling, with redistribution of blood flow to deep brain structures (Aoki *et al*., 1993). pH stat results in lower intracellular pH, which potentially increases cerebral

tissue oxygenation by suppressing cellular function (Hindman *et al*., 1995) and displacing the oxygen dissociation curve to the right, thereby liberating more oxygen to the tissues. There are concerns that suppression of cellular function with pH stat results in delayed cerebral metabolic recovery following hypothermic circulatory arrest (Skaryak *et al*., 1995). This has led some groups to use a strategy of pH stat during cooling with conversion to alpha stat prior to circulatory arrest and subsequent rewarming, in order to enhance cerebral cooling and subsequently improve metabolic recovery (Skaryak *et al*., 1995). The optimal clinical strategy remains unclear because most of the data comes from animal work. In a prospective clinical trial undertaken by the Boston group, the use of a pH-stat strategy throughout in infants undergoing deep hypothermic CPB was associated with lower postoperative morbidity and a shorter time to first EEG activity, supporting the clinical advantages of pH management (du Plessis *et al*., 1997). In a follow-up study, the group did not find a significant difference in development or neurological outcome at 1, 2 or 4 years between pH stat or alpha stat groups (Bellinger *et al*., 2001). The jury is still out.

HYPOTHERMIC CIRCULATORY ARREST AND LOW-FLOW CPB

Each individual operation imposes its own constraints on the surgeon and the perfusion practice. The use of circulatory arrest, low flow, full flow, and high or low perfusion pressures is based on a combination of patient size, anatomy, surgical procedure, collateral blood flow and personal (surgeon) preference, as opposed to predetermined protocols. There is a trend towards less use of deep hypothermia and circulatory arrest, with a move towards more physiological parameters. Currently, the majority of CPB users employ full-flow bypass. Recommended flows in various age groups are shown on **Table 11.1**. Good venous cannulation with snaring of the venae cavae can produce a near-bloodless operating field and yet maintain full flow throughout the systemic circuit. In this way, slightly higher temperatures can be maintained on bypass, with a higher haematocrit and probably with less potential for capillary leakage. Even within this policy, there is space for short periods of circulatory arrest as required, e.g. for closure of an atrial septal defect during the arterial switch operation.

If deep hypothermic circulatory arrest (DHCA) is required, single venous cannulation is usually employed, a lower haematocrit predetermined, and the temperature of the prime adjusted accordingly. Bypass is established at full flow and continued until the nasopharyngeal or tympanic membrane temperature has reached the predetermined level. Bypass should be continued for at least 20 minutes before the circulation is arrested (Bellinger *et al*., 1991). This results in even distribution of blood flow to the brain with adequate cerebral cooling.

The optimal temperature for circulatory arrest is thought to be 14–20°C (Gillinov *et al*., 1993; Mezrow *et al*., 1994), with a reported range in practice of 12–22°C (Elliott, 1993a). Cyanotic patients with large aortopulmonary collaterals have been identified as a group with increased risk of neurological injury associated with circulatory arrest. Evidence from animal work suggests that the period of cooling should be extended in this group, due to delayed brain cooling secondary to reduced cerebral blood flow (Kirshbom *et al*., 1995). The maximum duration of ''safe'' DHCA is unknown. It is logical that the period should be as short as possible but long enough to accomplish satisfactory surgical repair. Increased periods of DHCA are associated with increased brain injury (Mault *et al*., 1993).

There is considerable debate about whether circulatory arrest is really necessary at all for most of the operations performed in the neonatal and infant period. The reduction in cerebral metabolic activity during cooling is greater than the reduction in cerebral blood flow and oxygen delivery. This has led to the development of low-flow, low-temperature CPB. In a trial of DHCA vs. low-flow CPB in children undergoing arterial switch procedure, the DHCA group had a higher incidence of seizures and delayed EEG recovery (Newburger *et al*., 1993). In subsequent reports, at 1 year of age the DHCA group had increased abnormal neurology and worse motor function (Bellinger *et al*., 1995), with impaired language skills reported by parents at 2.5 years of age (Bellinger *et al*., 1997). By 4 years of age, the DHCA group had worse motor function and more severe speech abnormalities but there was no difference in IQ between the groups (Bellinger *et al*., 1999). By 8 years of age there were no differences in neurologic status, IQ, academic achievement and problem solving between the groups, but overall neurodevelopmental status for both groups was below expectations. The DHCA group continued to demonstrate lower test scores with regard to motor function and speech (Bellinger *et al*., 2003).

Circulatory arrest may still be required when access is otherwise impossible or if venous return is excessive. In such circumstances, intermittent flow during periods of DHCA produces considerably improved cerebral metabolic recovery (Langley *et al*., 1999). Thus, a period of 60 minutes of DHCA is well tolerated, with complete cerebral recovery, if divided into 15–30 minute intervals interrupted by short periods of perfusion, even at low flow.

An alternative to DCHA, particularly when working on the aortic arch, is selective antegrade cerebral perfusion with direct cannulation of the carotid artery

and low-flow CPB. There is increasing experience of this technique in adults undergoing aortic arch surgery (Bachet *et al*., 1999). The technique is readily applicable to paediatric surgery. Following a brief period of circulatory arrest, the aortic arch is opened and the aortic cannula can be advanced directly into the innominate artery and held in place with a snugger (Pigula *et al*., 2000). Despite widespread acceptance in recent years, techniques similar to this have never been tested against other techniques, such as intermittent perfusion. Once again, the data are not clear and readers are advised to review the available literature with caution. However, it makes good sense to reduce periods of low or absent flow to a minimum and, if one has to use them, to take all necessary precautions to protect the brain, including appropriate cooling and the avoidance of late hyperthermia or low cardiac output. Most brain damage is the result of several concatenating elements.

MYOCARDIAL PRESERVATION

Since the publication of the first edition of this book, a wealth of literature has been generated concerning intraoperative protection of the paediatric myocardium. A majority of the work reported in this literature is experimental and includes various modifications of crystalloid cardioplegic solutions, blood cardioplegia and, more recently, reperfusion solutions. Despite extensive literature, there remains no generally accepted strategy for the intraoperative protection of the paediatric myocardium. This is partly because the myocardium of neonates and infants is not the same as that of adult patients or animal models. The large number of different types of operations for congenital heart disease also makes comparison of different types of myocardial protection difficult. However, the principles of myocardial protection (i.e. diastolic arrest using high potassium concentrations, with ventricular decompression and hypothermia to reduce metabolic demand) remain unchanged.

There are major metabolic, functional and biochemical differences between the heart of a newborn and that of an older child (Coles *et al*., 1987). The neonatal heart uses glucose as its principle energy source. Experimental evidence suggests that neonates are better able to tolerate hypoxia than are children, but are less tolerant of the increase in myocardial water associated with repeated doses of crystalloid cardioplegic solution (Grice *et al*., 1987; Yano *et al*., 1987). Thus, when using crystalloid cardioplegia, many surgeons use only a single dose for operations in newborns, extending up to about 80 minutes of aortic cross-clamping time. Others repeat cardioplegia every 20–30 minutes, as in older children. The myocardium is also cooled topically with the use of repeated applications of ice slush during the period of cross-clamping, so as to reduce the temperature gradient between the heart and the surrounding tissues.

Typically, following application of the aortic cross-clamp, an initial dose of cardioplegia of 20–30 ml/kg is given, and supplemental doses of 10 ml/kg are administered every 20–30 minutes. Cardioplegic solutions are usually delivered in a prograde manner into the aortic root, via a cardioplegic cannula placed through a purse-string suture. In the presence of aortic (truncal valve) incompetence, the cardioplegic solution is infused through a coronary artery cannula directly into the coronary arteries after opening of the aorta. Direct infusion into the coronary arteries is also used in all situations in which the aorta is opened (repair of truncus arteriosus, arterial switch operation). In experimental studies, various additives to the cardioplegic solution have been found to offer better protection of the myocardium. These include L-aspartate, L-glutamate, adenosine, calcium channel blockers, deferoxamine, co-enzyme Q10 and free radical scavengers. However, their precise role in clinical practice has not, as yet, been properly defined. It has been demonstrated experimentally (Rosenkranz *et al*., 1984) that the initial warm cardioplegic solution may optimize the conditions for myocardial recovery in the post-ischaemic period. Factors other than cardioplegia often contribute to the adequacy of myocardial preservation and must be borne in mind. These include preoperative glycogen reserves, anaesthetic techniques, total body (perfusate) hypothermia, topical cooling, avoidance of coronary air embolization and avoidance of distension of the heart. Gentle handling of the heart and the completeness of the surgical repair are also extremely important.

Several experimental and some clinical studies have shown the superiority of providing oxygen in the cardioplegic solution. The vehicle for providing oxygen may be blood, perfluorocarbons, stroma-free haemoglobin, or oxygen dissolved in crystalloid cardioplegic solution. Of these different agents, blood is the only vehicle that is used clinically. The advantages of blood in cardioplegia include: (a) keeping the heart oxygenated while it is being arrested; (b) improved buffering and rheology; and (c) reducing the risk of reperfusion damage. Unlike in adult surgery, the advantages of blood cardioplegia in paediatric myocardial protection are less well established. There is evidence to support its use over crystalloid cardioplegia during periods of prolonged myocardial ischaemia (Corno *et al*., 1987).

Induced ventricular fibrillation without aortic cross-clamping is a useful technique for less complicated lesions, in which the intracardiac procedure is short. It is usually combined with mild hypothermia and can be used in the majority of patients with secundum atrial septal

defects (Rosengart and Stark, 1993). This technique may also be used in operations involving the right ventricle, the right ventricular outflow tract, and the pulmonary arteries. The technique is also useful whenever it is impossible to cross-clamp the aorta safely (e.g. in the presence of severe calcification). If induced fibrillation is used, great care should be taken not to distend the heart because this would seriously impair myocardial blood flow and cause subendocardial ischaemia.

MANAGEMENT OF VENTILATION

Mechanical ventilation is usually discontinued during bypass. A small constant positive pressure of 5 cmH_2O does keep the lungs partially inflated and minimizes micro-atelectasis. However, the capillary bed is static, and there is evidence of sequestration of neutrophils within this capillary bed. These neutrophils are activated, increasingly adherent, and therefore capable of generating massive endothelial damage (Finn *et al*., 1993). Thus, many workers have suggested increasing the ventilation or ventilating the child as early as possible in the course of bypass. A solution is to ventilate the child as soon as practicable during rewarming, once the cardiac chambers have been closed. This gives plenty of time to treat atelectasis, to lower pulmonary vascular resistance and, it is hoped, to allow the pulmonary lymphatic channels to deal with any accumulated extra vascular fluid. Recently, continuous direct perfusion of the pulmonary arteries has been suggested, and is reported to improve pulmonary compliance and lower pulmonary vascular resistance postoperatively. This work needs formal testing.

SPECIAL CIRCUMSTANCES

Left Superior Vena Cava

The special circumstances of cannulation for the various procedures are covered in the individual operations described in this book; however, the management of a left SVC is common to many conditions and is considered separately here. It can be difficult to determine in advance whether it is necessary to cannulate a left SVC. However, if pressure-monitoring cannulae have been inserted into the neck above both right and left SVCs, the decision can be made during CPB. When full flow is established with the use of right SVC and IVC cannulae, the base of the left SVC can be clamped or snared and the pressure above the clamp observed. If the venous pressure rises to > 18 mmHg, the left SVC should be drained. This is often the situation if the innominate vein is absent. Occasionally, however, even in the absence of a ''bridging'' vein, the left SVC pressure does not rise on clamping, and cannulation of the left SVC is not necessary.

Direct Cannulation

The left SVC is dissected (on bypass) above the left atrium and mobilized slightly. The left SVC should be dissected carefully in order to avoid the phrenic nerve, which lies on its anterolateral surface. A purse-string suture similar to that used in the right SVC can be positioned anteriorly, and a small, right-angled, metal-tipped cannula is positioned. This cannula can be connected to a triple venous connector to join with the right SVC and IVC cannulae and thereby permit complete venous drainage. The left SVC can then be snared. However, access to this area is sometimes difficult, particularly in a reoperation, and under these circumstances an alternative, indirect method can be useful.

Indirect Cannulation

An open heart sump sucker placed into the coronary sinus orifice of the LSVC usually provides adequate drainage. Alternatively, a third venous cannula or a balloon-tipped Foley catheter can be advanced up the coronary sinus with the balloon inflated in the left SVC (not within the heart). The central pipe of the catheter can then be connected to the bypass circuits. Direct left SVC cannulation is preferable to indirect cannulation in most circumstances.

No Cannulation

In small infants with a left SVC or in patients with more complex anomalies of systemic venous return, it may be preferable to use single atrial cannulation and operate under deep hypothermic circulatory arrest.

Other Cannulation Techniques

There are other situations, especially during reoperations, that may require unorthodox cannulation. For example, in the redo replacement of the systemic atrio-ventricular valve in corrected transposition of the great arteries, or placement of a left ventricle to pulmonary artery conduit in a patient who has previously undergone a Mustard or Senning procedure, the operation is accomplished through the left thoracotomy; the descending aorta is cannulated for arterial return, and the main pulmonary artery or the left (functionally right) atrium is cannulated for venous return. Patients undergoing reoperation for failed Fontan procedure may have a grossly dilated right atrium and be in a state of low cardiac

output. Iliac artery/iliac venous bypass is ideal for such patients. If the iliac veins are thrombosed after previous catheterizations and interventions, it may occasionally be prudent to cannulate the artery and then perform a mini-thoracotomy to cannulate the right atrium before proceeding with sternotomy and formal cannulation.

Control of Shunts

Congenital, or surgically created, systemic-to-pulmonary-artery connections must be dissected out and controlled but not occluded before the onset of CPB. Failure to control shunts can result in massive run-off into the pulmonary circulation once CPB is commenced, with a corresponding fall in diastolic pressure. Thus, myocardial and cerebral perfusion may be severely affected, particularly in watershed areas. This is true of large patent ductus arteriosus in patients with significant left-to-right shunts with pulmonary blood flows dependent on such a duct. It is also important to control or occlude the branch pulmonary arteries in repair of truncus arteriosus. Similarly, if a patient has Blalock–Taussig shunts or aorto-pulmonary communications that have been surgically created, the onset of bypass is associated with massive run-off into the pulmonary circulation with a low arterial pressure, which can result in severe cerebral damage, poor diastolic flow down the coronary arteries, or damage to the lungs. Thus, these shunts must be controlled. Once the vessels or shunts have been dissected and ligatures passed around them, they are closed immediately after the onset of bypass. If a shunt is constructed on CPB in the presence of a patent, previously constructed shunt, an alternative strategy may have to be used. Ligature or clamp may damage the shunt, which may occlude soon afterwards. Under such circumstances, control of the shunt with a Foley catheter placed from the pulmonary artery may be advantageous. Alternatively, a hybrid approach, placing a balloon in the catheterization laboratory and inflating at surgery, may be used. The same constraints apply also to major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect. Detailed individual preoperative strategies for their control must be worked out by the multidisciplinary team in advance.

ULTRAFILTRATION (UF)

The increasing use of CPB in the neonatal period with haemodilution has exposed the tendency toward massive capillary leakage in these patients. Such capillary leakage can result in organ oedema and dysfunction and potentially fatal results. This problem reawakened interest in ultrafiltration, which was used in adult cardiac surgery for some time for removal of water from patients in chronic heart failure or for haemoconcentration towards the end of bypass. In conventional ultrafiltration (CUF), an ultrafilter within the bypass circuit is used to shunt some blood from the arterial to the venous sides of the circuit, either throughout CPB or during rewarming toward the end of bypass (usually with temperatures higher than 28°C), to remove excess water from the combined circuit. We employed this method in our unit for some time and were dissatisfied with the results (Elliott, 1993b). By changing the technique and timing of ultrafiltration, in such a way that the modified ultrafiltration (MUF) was started after bypass for 10–15 minute. MUF was able to reduce significantly the accumulation of total body water observed in association with cardiopulmonary bypass. In order to use ultrafiltration after termination of bypass, the arterial cannula is left in place and venous cannulae are removed. The (heated) line from the ultrafilter is placed through the venous cannulation site into the right atrial appendage. The inlet tube to the ultrafilter can be repositioned into one of the suction roller pump heads of the bypass machine. Low-pressure suction is applied to the outlet of the ultrafilter at approximately 100 cmH_2O. The venous blood can be drained back into the bypass circuit and followed by saline to leave the circuit primed and, while the left atrial pressure is monitored, ultrafiltration is begun. Blood flows up the aortic cannula to the ultrafilter inlet, to be pumped through the filter at 100–150 ml/minute. A circuit diagram for the technique of modified ultrafiltration is shown in **Figure 11.12**. The haemoconcentrated blood from the haemofilter passes to the right atrium. If the left atrial pressure falls,

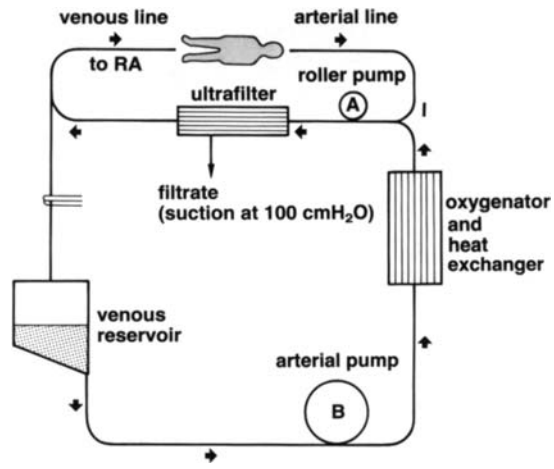

Figure 11.12

diluted blood in the venous reservoir can be pumped through the oxygenator/heat exchanger to the inlet to the ultrafilter. This blood is thus haemoconcentrated as it flows to the right atrial appendage. Ultrafiltration continues until the patient's haemotocrit is elevated to the desired level, which varies with age and diagnosis. Neonates and patients with elective cyanosis at the end of the procedure require a higher haematocrit. This process usually takes 10–15 minutes. Custom-designed ultrafiltration circuits are now available from many manufacturers. Conventional ultrafiltration was successful in some patients, but in others it was clearly unsuccessful; indeed, there was no significant difference between patients who underwent conventional ultrafiltration and control patients in terms of removing total body water. Encouraged by the results of the pilot study, we conducted a prospective randomized assessment of the use of ultrafiltration during bypass and were able to demonstrate a significant reduction in the rise of total body water, a reduction in blood loss, and a reduction in the need for blood transfusion during the early postoperative phase (Elliott, 1999). In small infants undergoing long, low-temperature perfusion bypasses (patients who had the greatest tendency for accumulation of total body water), there was a suggestion that ventilation times and hospital stay were considerably reduced. Most surprising, however, was that all the patients who underwent ultrafiltration showed a highly significant rise in systemic blood pressure during the 10–15 minutes of ultrafiltration in comparison with the controlled patients, who showed no change. The observation was subsequently investigated and it was found that modified ultrafiltration, by the method that we used, was associated with a marked rise of about 40% in cardiac index, no significant change in systemic vascular resistance, and a 40% fall in pulmonary vascular resistance over the 10 minute period of ultrafiltration, which routinely elevated haematocrit from 20 on bypass to 35. In other words, this technique has produced very favourable haemodynamic changes at the same time as elevating the haematocrit to predetermined levels, without the need for donor blood. The technique offers the advantages of a non-pharmacological control of blood pressure and cardiac index with the ability to improve oxygen delivery to the tissues by elevating haematocrit. Ultrafiltration permits the use of lower haematocrits during CPB and potentially obviates the need for donor blood, both during and after the operation, and thus has become an integral part of blood-sparing strategies and for the management of Jehovah's Witnesses.

The results we obtained initially have now been reproduced by many other workers in several centres around the world. However, Shin'oka *et al*. (1998) using magnetic resonance spectroscopy, demonstrated, in pigs undergoing CPB, that a high haematocrit CPB strategy had an equal or greater impact on recovery from DHCA than MUF.

In the current era, MUF is widely used in combination with CUF to optimize fluid balance and oxygen delivery. The benefits in terms of reduction in blood loss and transfusion requirement and the improvement in pulmonary vascular resistance and compliance have led to its increasing use in adult practice. In paediatric cardiac surgery, these benefits are of particular value after Fontan-type procedures and the Norwood operations.

Ultrafiltration helps manage the consequences of CPB. It does nothing to prevent the injury. While we firmly believe that ultrafiltration beneficially affects many physiological parameters and consequent morbidity, no data exist to suggest that it reduces mortality. It would be far better to prevent the injury, but until we can do that ultrafiltration is useful in reducing its consequences.

CONCLUSION

Paediatric CPB is generally a surprisingly well-tolerated procedure. Significant advances in understanding and technique have enabled complex neonatal cardiac surgery to be undertaken. The advances have been largely iterative and to some extent intuitive. Conventional Cochrane-style measures of the quality of evidence reveal that we have little scientific basis for most of our practice (Bartels *et al*., 2002). The large variability in children, cardiac pathology and surgical techniques has made effective randomized, clinical trials hard to conduct and difficult to analyse. Consequently, much practice has evolved from the application of animal data and shared experience. In order to continue to improve surgical outcomes, we must continue to ask the correct questions and attempt to answer them with the appropriate studies. The design of such studies will be complex, but should not be ignored because of that.

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12

Anaesthesia

E. Sumner

The anaesthetist is a vital member of the surgical team and is involved in patient management from admission to recovery. The role with this type of surgery is considerably wider than involvement only in the operating theatre.

Good practice in anaesthesia for congenital heart surgery in children, whether for palliative or corrective procedures, requires an extensive knowledge of the pathophysiology of the particular disease process, the effect of and reaction to the drugs and processes of anaesthesia and the likely outcome of surgery.

The basic principles of anaesthesia for children with congenital heart disease have not greatly changed since the last edition of this book and involve:

1. Adequate anaesthesia and analgesia, so that endogenous catecholamine levels are not excessive at any stage. High levels of endogenous catecholamines increase the risk of dysrhythmia, ventricular infundibular spasm in patients who have hypertrophied ventricular outflow tract, and peripheral vasoconstriction.
2. Full muscle relaxation, so that sudden contraction of the diaphragm or inspiratory gasping cannot take place, thereby reducing the risks of tearing vessels held within a clamp or entrainment of air if the heart is open.
3. Life support in its various aspects. This involves maintenance of normal alveolar ventilation, circulatory volume, a satisfactory peripheral circulation, acid–base balance and electrolyte homeostasis. In order to achieve this, extensive monitoring is essential.
4. Avoidance of cerebral air embolism, an ever-present danger in most patients with a congenital heart defect.

Problems facing the anaesthetist relate to the physiological differences between the infant and the older child or adult and the type of cardiac disorder. In infancy all physiological processes are immature and develop at different rates, e.g. at birth hepatic function is immature and the metabolism of drugs, such as morphine, will be greatly slowed, so that even a single dose can have a prolonged and profound effect, but by 4 months of age, the processes have matured; respiratory function does not fully mature for several years.

Infant haemodynamics are characterized by greater interventricular dependency, a lower myocardial contractile mass (30% of the adult), a cardiac output which is largely rate-dependent, and a high cardiac output, which is optimal with a low systemic vascular resistance and a heart functioning close to the top of the Starling curve. At the cellular level, the sarcoplasmic reticulum is immature with immature Ca^{++} storage and exchange mechanisms, which make the infant myocardium particularly sensitive to the inhalational anaesthetic agents for at least the first 6 months (Friesen *et al*., 2000).

Sympathetic innervation is underdeveloped, with lower catecholamine stores and catecholamine receptors are easily ‘‘downregulated’’, both of which have implications for inotropic therapy. Pulmonary vascular resistance is high at birth and may be labile, especially where L–R shunts cause failure of the regression of muscularity in the pulmonary arterioles, failure of development of the intra-acinar vessels, with implications for intra- and postoperative management.

Because of an infant’s high oxygen requirement, the minute alveolar ventilation is, on a weight basis, twice that of an adult. This is attained by an increase in respiratory rate rather than in tidal volume. Tidal volume is relatively fixed because it is achieved by diaphragmatic function only. Anything that interferes with the function of the diaphragm, such as abdominal distension or phrenic nerve palsy, is likely to precipitate respiratory failure in an infant.

Airways resistance and the work of breathing are high during the first years of life because of the small bore of even major airways. In infants, as in the elderly,

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there is a tendency to peripheral airway closure within normal tidal breathing, leading to ventilation/perfusion inequalities and an early increased need for supplemental oxygen, e.g. with the increased lung water commonly seen after cardiopulmonary bypass.

Respiratory support is more likely to be required for a sick infant and may be beneficial preoperatively to promote oxygenation and to provide cardiac support by removing the work of breathing.

An infant has a large surface:weight ratio and, in the early stages of life, poor peripheral insulation, which can result in extensive heat loss in the thermally unfavourable environments of the catheterization suite, induction and operating rooms and the intensive care unit. Energy is expended in attempting to maintain normothermia and oxygen demand may outstrip availability in patients with heart disease, leading to hypothermia and respiratory failure. The aim is always to maintain a neutral body temperature with a minimal expenditure of energy and oxygen consumption. Postoperatively, central temperature may rise, with marked peripheral vasoconstriction as a result of low cardiac output or excessive adrenergic activity, which require treatment.

CARDIAC DEFECTS

The common classification of congenital cardiac disease is based on a sequential, segmental approach, which is unambiguous and is useful for cardiologists and surgeons. Anaesthetists are more concerned with applied pharmacology and physiology and a functional approach may be clinically more useful. Four basic functional groups can be identified.

Many patients with congenital heart disease have abnormal communications between pulmonary and systemic circulations at either the atrial, ventricular or great arteries level, resulting in shunting of blood:

- An *obligatory* shunt is from a high-pressure chamber to a low-pressure chamber and is therefore continuous, although the quantity of flow may change.
- A *dependent* shunt occurs between chambers of equal pressure; the shunt flow is determined largely by the shunt orifice and the outflow resistances. When the shunt orifice is not restrictive, the direction and the magnitude of the shunt are dependent mainly on the relative resistances (PVR and SVR) of the pulmonary and systemic vascular beds.
- *Complex* shunt lesions are associated with ventricular outflow tract obstruction at subvalvar, valvar, supravalvar or major vessel level. In these cases, the outflow obstruction may be a much greater determinant of the shunt than the resistance of the peripheral vascular beds. Patients who have complete valvar obstruction have an obligatory and sometimes a nearly total shunt proximal to the obstruction, and this is always associated with a second shunt downstream to provide flow to the obstructed side of the circulation. This second shunt is usually a persistent ductus arteriosus, and flow can be in either direction, depending on the side of the obstruction.

In a neonate with outflow atresia, it is crucial to maintain ductal patency preoperatively, and this is achieved by an infusion of prostaglandin E, the usual dose of which is 0.05–0.1 μg/kg/min intravenously.

Defects with High Pulmonary Blood Flow

Defects in which left-to-right shunting is continuous or predominant are associated with high pulmonary blood flow, and affected patients may eventually develop irreversible pulmonary vascular disease. The pressure and volume of flow, together with low oxygen saturations, are important determinants of structural changes in the lung vessels.

These defects include persistent ductus arteriosus, aortopulmonary window, atrial and ventricular septal defects, atrio-ventricular septal defects, persistent truncus arteriosus and total anomalous pulmonary venous connection.

Immediately after birth, PVR is high but falls soon after, so that most infants have a low PVR and a high pulmonary blood flow, which causes actual or incipient pulmonary oedema, increased work of breathing with tachypnea and varying degrees of cardiorespiratory failure. These conditions may also cause repeated chest infections and failure to thrive, with feeding difficulties, in spite of diuretic therapy.

Prolonged intensive antifailure therapy may result in a contraction of circulating blood volume with peripheral vasoconstriction, making venepuncture and the placement of central venous catheters more difficult. Even with supplements, deficits of total body potassium and magnesium may occur and should be assumed.

In the early stages, systemic vasodilatation and mild hypotension are well tolerated.

Irreversible pulmonary vascular disease is often the limiting factor for corrective cardiac surgery and even small increases may prevent the formation of a Fontan type of univentricular circulation. If pulmonary vascular disease is advanced, any reduction of systemic vascular resistance may cause a reversal of the shunt, with sudden cyanosis and haemodynamic collapse.

Induction of anaesthesia by inhalation or intravenously will be normal.

Defects with Low Pulmonary Blood Flow

Right ventricular outflow tract obstruction with right-to-left shunting results in low pulmonary blood flow and cyanosis. This group of defects includes tetralogy of Fallot, pulmonary stenosis with atrial or ventricular septal defect or both, pulmonary atresia and tricuspid atresia. Polycythaemia is the result of the chronic hypoxaemia and may be severe. Dehydration will increase the risk of cerebral thrombotic episodes in children with a very high haematocrit. These patients may also be expected to have a compensatory coagulopathy with reduced prothrombin and poor platelet function, which may cause problems with haemostasis after cardiopulmonary bypass.

Inhalation induction of anaesthesia is slow with insoluble anaesthetic agents, but with shunts greater than 70% it is slow with all agents, but usually acceptable in small infants (Tanner *et al*., 1985). Older children should always undergo intravenous induction, because inhalation induction may be extremely prolonged and associated with poor airway maintenance and laryngospasm. For intravenous induction, the rate of injection should be slow, as the R–L shunt allows immediate access to the arterial circulation of the brain and heart.

These patients are at particular risk from air embolism, which has direct access from the venous to the arterial circulation, and meticulous care must be taken to prevent this. Stopcocks may be a source of air, so the line should be aspirated before injection and the last of the syringe contents should not be injected.

Overventilation occurs easily because the lungs are highly compliant, and a high mean inflation pressure may cause a further reduction in pulmonary blood flow.

Patients with tetralogy of Fallot may suffer from a hypercyanotic attack from right ventricular infundibular spasm induced by an increase in circulating catecholamines, associated, for example, with induction of anaesthesia or with light anaesthesia, or from reduction in systemic resistance. Direct handling of the heart further increases the R–L shunting. Morphine reduces the severity of infundibular spasm, and systemic vascular resistance can be safely increased with a noradrenaline infusion (dilute 300 μg/kg in 50 ml 5% dextrose; start at 1 ml/h (= 0.1 μg/kg/min).

Systemic hypotension may also reduce flow through systemic-to-pulmonary collaterals or a Blalock–Taussig shunt, thus increasing hypoxaemia, and the combination of hypotension, hypoxaemia and metabolic acidosis may be rapidly fatal without treatment.

If hypotension occurs, blood pressure should be returned to normal levels by transfusion, correction of metabolic acidosis, and the use of noradrenaline. If serious arterial hypoxaemia persists [arterial oxygen pressure (PaO_2) < 30 mmHg (4 kPa)] with a systolic pressure higher than 100 mmHg, a small dose of a β-adrenergic blocking agent, such as propranolol, should be used to relax the infundibulum of the right ventricle.

Transposition of the Great Arteries

Patients with this defect have a combination of severe cyanosis and increased pulmonary blood flow. In patients with simple transposition, a Rashkind's balloon septostomy is performed as soon after birth as possible to increase mixing at atrial level, although severe desaturation may persist. If a small ventricular septal defect or persistent ductus arteriosus is present, systemic oxygenation is usually higher. In most cases an arterial switch operation is performed in the first 2 weeks of life before the pulmonary vascular resistance falls, after which the left ventricle weakens and may not support the systemic circulation postoperatively (see Chapter 36).

Anaesthesia must take into account the physiological limitations of the newborn and systemic desaturation with metabolic acidosis may occur. Oxygenation depends on interatrial blood mixing, which is easily disturbed by surgical manipulation or overtransfusion.

Left Ventricular Outflow Tract Obstruction

Infants with aortic obstruction or coarctation of the aorta may have severe left ventricular hypertrophy with myocardial ischaemia and fibrosis, which may result in very poor myocardial function. Patients with the supravalvar aortic stenosis with Williams' syndrome are at particular risk from reduction in systemic pressure, as the myocardium is used to a high perfusion pressure.

Preoperative support with mechanical ventilation, inotropes, prostaglandin and sometimes peritoneal dialysis improves the preoperative condition enough to enable surgery to take place. Transfusion may precipitate pulmonary oedema, and hypotension may further reduce myocardial oxygenation and cause ventricular dysrhythmias or even cardiac arrest.

SYSTEMIC AND PULMONARY VASCULAR RESISTANCES

The anaesthetist is in a position of control of both SVR and PVR (**Table 12.1**) by manipulating CO_2 and O_2 with ventilation strategies and the use of drugs. This is particularly useful in the pre- and postoperative management of the Norwood procedure.

Pulmonary Vascular Resistance

At birth, the media of the peripheral pulmonary arterioles have copious smooth muscle that regresses

Table 12.1 Anaesthetic implications of cardiac pathophysiology. Perianaesthetic factors influencing cardiovascular flow and resistance.

Flow (cardiac output)		
Systemic and pulmonary circulations		
	Increase	Volume loading
		Chronotropic agents
		Inotropic agents
		Vasodilators (with adequate volume)
		Inhalation anaesthetics
		β-Adrenergic antagonists
	Decrease	Inhalation anaesthetics
		Hypovolaemia
		Vasodilators (with inadequate volume)
		Arrhythmias
		Ischaemia
		Calcium slow-channel blocking agents
		High mean airway pressure (with inadequate volume)
Resistance		
Systemic circulation		
	Increase	Sympathetic stimulation
		α-Adrenergic agonists
	Decrease	Anaesthetics[a]
		Vasodilators
		α-Adrenergic antagonists
		β-Adrenergic agonists
		Calcium slow-channel blocking agents
Pulmonary circulation		
	Increase	Hypoxia
		Acidosis
		High mean airway pressure
		Sympathetic stimulation
		α-Adrenergic agonists
		Hypervolaemia
		Anaesthetics[a]
	Decrease	Oxygen
		Hypocarbia
		Alkalosis
		Prostaglandin E_1/prostacyclin
		α-Adrenergic antagonists
		Vasodilators
		Anaesthetics[a]
		Nitric oxide

[a] Effects of non-inhalational agents may be unpredictable.

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gradually; during this time, the vascular bed remains reactive. Lesions resulting in high pulmonary artery pressure are initially associated with high pulmonary blood flow, but obstructive vascular disease eventually develops. Small intrapulmonary vessels rapidly become muscularized, and intra-acinar vessels fail to develop from birth. Pulmonary vascular disease is a serious risk if the PA pressure is greater than half the systemic and the pulmonary:systemic flow ratio is greater than 3:1. This eventually causes fixed pulmonary vascular obstruction, but in the early stages there is a reactive and reversible component (Hall and Haworth, 1992). PVR is increased by hypoxia and acidosis, sympathetic stimulation and surgical retraction. PVR also increases with hyperinflation (high lung volumes) and atelectasis (low lung volumes).

High levels of positive end-expiratory pressure (PEEP) increase PVR, but if modest levels of PEEP reverse pulmonary oedema and atelectasis, PVR is lowered. Modest hyperventilation, raising the pH to 7.5, always lowers PVR in infants.

Some anaesthetic agents may affect PVR, although responses may be variable. Ketamine was initially reported to increase PVR but does not appear to do so in infants if ventilation is normal (Hickey *et al*., 1985). High doses of fentanyl suppress sympathetic stimulation in infants during anaesthesia and thus reduce pulmonary vascular responses to stimuli.

Systemic Vascular Resistance

Most agents used for anaesthesia cause some reduction in SVR, although this may be dose-related. Ketamine causes a modest increase in systemic vascular resistance.

Hypotension resulting from a reduction in SVR may have serious consequences in patients in whom oxygenation is shunt-dependent, either by a surgically created shunt or from systemic–pulmonary collaterals. Patients with tetralogy of Fallot may exhibit an increase in right-to-left shunting, which can cause life-threatening hypoxaemia and the development of serious metabolic acidosis.

Patients with left ventricular failure, as in aortic stenosis or coarctation, require high coronary perfusion pressure in order to adequately oxygenate the myocardium; those with supravalvar aortic stenosis are particularly at risk.

Minor degrees of hypotension may be tolerated, but if oxygenation is falling, perfusion pressure should be increased by the use of an α-adrenergic agonist, such as noradrenaline or phenylephrine.

PREOPERATIVE MANAGEMENT

The anxiety involved for a family coming into hospital for a child to have cardiac surgery cannot be underestimated. Patients and parents may already be familiar with the hospital from previous admissions for investigation, medical treatment or surgery. However, despite the greatly improved operative and long-term outlook for children with congenital heart disease, with mortality approaching zero for many lesions, the operation remains the major episode, and parental anxiety is often considerable. This anxiety is rapidly transmitted to the child.

The most satisfactory way of avoiding parental overanxiety is by regular contact between medical and nursing staff and the parents. Explanations should be simple, factual and honest, and at all times parents must be aware of the reasons for, and timing of, different aspects of care. All aspects of the anaesthesia and surgery are fully discussed, which includes the known risks of cardiac, renal and neurological complications, which should be quantified where possible. *Most* units also provide booklets or videos and arrange for preoperative hospital visiting. It is usual for the parents to be shown the intensive care unit preoperatively, to lessen the shock of seeing their own child in that environment.

The anaesthetist's preoperative visit is also an opportunity to find out about the lesion and what the surgeon intends to do, as well as an assessment of the clinical condition, the exercise tolerance, degree of cyanosis, the ability to feed and the presence of other congenital abnormalities, e.g. one that might cause intubation problems. Each patient requires very careful individual assessment. The anaesthetist is usually involved in the preoperative joint cardiac conference between the cardiologists, surgeons and intensivists, where management decisions are made.

Although starvation protocol insists on no solid food or formula milk for 6 hours, or breast milk for 4 hours, clear fluids such as water or clear juice can be taken up to 2 hours preoperatively. Polycythaemic children should not be without fluids for too long and if the start time of surgery is uncertain, then fluids should be given intravenously.

In addition to establishing a rapport with the patient and parents, most anaesthetists favour some form of anxiolytic premedication. Midazolam, which is also an amnesic, may be given orally in a dose of 0.5–1 mg/kg. The larger doses are well tolerated and do not cause haemodynamic instability (Masue *et al*., 2003). Even very small babies, such as those for the arterial switch procedure, may benefit from sedative premedication with chloral hydrate 30–50 mg/kg orally 40–60 minutes preoperatively.

Local anaesthesia cream, such as EMLA or Ametop, is used routinely, as this allows painless venepuncture, either for an intravenous induction of anaesthesia or earlier during an inhalational induction.

Table 12.2 Antibiotic prophylaxis for cardiac surgery.

Age	Drug	Dose	Comments
All cases except for reoperations and patients allergic to penicillins			
0–4 weeks	Amikacin	10 mg/kg, stat dose	
> 4 weeks	Amikacin	10 mg/kg, 12 hourly	
0–4 weeks	Flucloxacillin	25 mg/kg, 8 hourly	Repeat 25 mg/kg, when stable off bypass
> 4 weeks	Flucloxacillin	25 mg/kg, 6 hourly	Repeat 25 mg/kg, when stable off bypass
Re-operations during same hospital admission or patients allergic to penicillins			
0–4 weeks	Amikacin	10 mg/kg, stat dose	
5–8 weeks	Amikacin	10 mg/kg, 12 hourly	
8 weeks	Amikacin	10 mg/kg, 12 hourly	
0–4 weeks	Teicoplanin	16 mg/kg, stat dose	
5–8 weeks	Teicoplanin	15 mg/kg, stat dose	
> 8 weeks	Teicoplanin	10 mg/kg, 12 hourly	

Duration of prophylactic cover should be 24 h. First dose, on induction of anaesthesia; bypass cases, additional flucloxacillin dose off bypass.
Subsequent doses: to complete 24 h cover from induction.
Antibiotic levels are not indicated as prophylaxis is for 24 h only.
Preterm infants and special situations, e.g. renal failure, long-standing and complex cases. Discuss regime with consultant microbiologist.
Teicoplanin administration: patients > 8 weeks, intravenous bolus; neonates 8 weeks, < 30 min infusion.

Antibiotic prophylaxis is routine, to prevent endocarditis from a transient bacteraemia (Dajani *et al*., 1997; **Table 12.2**).

INDUCTION

All anaesthetic agents and opioids affect haemodynamics and if myocardial function is poor or the patient is hypovolaemic, the effects may be profound. Induction should be performed with care and full non-invasive monitoring (ECG, pulse oximetry and non-invasive blood pressure monitoring, as well as a precordial stethoscope). Peripheral vascular access is achieved as soon as possible, if not already present.

Sevoflurane has become the agent of choice for inhalational induction (Mori and Suzuki, 1996). It is non-pungent, fast-acting and well accepted by children. The cardiac output is well maintained and even though there is direct myocardial depression, especially in infants, a moderate rise in pulse rate compensates for this. This agent is less dyrrhythmogenic than other inhalational agents (Ebert *et al*., 1995). In infancy, even sevoflurane decreases mean blood pressure from an awake level to a similar degree to isoflurane (and halothane), with a significant reduction in cardiac index at 1.25 minimum alveolar concentration (MAC). Isoflurane is an unsatisfactory induction agent, as it tends to be an irritant, but is an ideal agent for maintenance of anaesthesia if used in low doses (up to 1%) together with an opioid analgesic (Housmans and Murat, 1998). After a fluid load, the ejection fraction will increase with this agent. It is a peripheral vasodilator, which may be beneficial but will also dilate small coronary vessels and can cause ''steal'', by diverting flow from areas of borderline perfusion to areas already well perfused, producing ischaemia. This could be significant in children whose coronary blood flow is marginal.

Ketamine is the agent of choice for intravenous induction in a dose of 2 mg/kg (or 6–8 mg/kg if given i.m.) and extensive clinical practice has confirmed its efficacy and safety. It produces rapid hypnosis and profound analgesia without cardiovascular or respiratory depression. Although it is a direct-acting negative inotrope, it prevents neuronal uptake of catecholamines, causing a centrally mediated sympathetic stimulus, which usually predominates. There is a non-dose-related rise in SVR and PVR with increased myocardial oxygen consumption. The effect on PVR will be trivial if the respiration has been controlled. Blood pressure will fall, however, if catecholamine levels are depleted after prolonged infusions of dobutamine, for example, and catecholamine receptors have been downregulated. Increased cardiac output and increased myocardial oxygen consumption, with tachycardia, may be undesirable in the presence of severe valvar obstruction and ventricular strain. All undesirable side-effects of ketamine are reversed by

small doses of midazolam. Its action is that of an NMDA receptor antagonist, which may possibly have a cerebral protective effect during deep hypothermic circulatory arrest (Orser *et al*., 1997).

Etomidate is also an older drug for induction of anaesthesia that has regained popularity, especially for use in very high-risk patients, such as those with a very poor ejection fraction. This drug has GABA-mimetic effects in doses of 0.2–0.3 mg/kg and has minimal cardiovascular side-effects. It causes a rise in heart rate and cardiac index and a rise in myocardial blood flow without a rise in myocardial oxygen uptake (Donmez *et al*., 1998). Injection is painful and causes involuntary limb movements, which can be minimized by giving 1 μg/kg fentanyl first.

Thiopentone and propofol, extensively used in fit children, reduce sympathetic tone and availability of calcium to cardiac muscle and are not recommended for bolus dose in sick children for induction.

After a short induction with minimal doses of agents, which may depress the myocardium, tracheal intubation and controlled ventilation are quickly achieved after using a cardiostable muscle relaxant, such as pancuronium or vecuronium.

In infants, the nasal route for intubation is preferred for postoperative comfort and stability, especially for smaller children. The tube should be of a size that allows a small air leak through the cricoid at 25 cmH_2O pressure; otherwise there is a risk of mucosal damage with oedema on extubation and possible long-term tracheal stricture. The length should be such that the tip of the tube is midtracheal and can be seen lying between the clavicles on the postoperative chest X-ray. Some tubes, such as the Portex, have a mark that suggests the level, which should lie at the vocal cords. After intubation, careful auscultation of the chest should reveal bilateral breath sounds and there should be a square end-tidal CO_2 trace on the capnograph. Many centres nowadays use cuffed tracheal tubes, even for infants, who have the advantage of sealing the trachea, thus allowing the use of sophisticated pulmonary function analysis, and with care these should not cause laryngeal damage.

Ventilation, usually with an air/oxygen mixture, is adjusted to produce an arterial carbon dioxide pressure [$PaCO_2$ of about 35 mmHg (4.6 kPa)] and an appropriately sized heat and moisture exchanger is put in the circuit close to the patient. Patients with L–R shunts have non-compliant lungs, whereas those with low pulmonary blood flow have compliant lungs and overventilation will further reduce pulmonary blood flow.

Vascular Access Lines

It is usual practice to insert a multi-lumen catheter, using the Seldinger technique. The internal jugular, subclavian and femoral veins are possible sites, of which the right internal jugular is the most common. However, for patients with cavo–pulmonary connections, the femoral route may be preferred. At least three lumens are required, one for CVP measurement, one for continuous infusion of drugs, such as inotropes, and the third for maintenance, blood sampling and colloid infusions.

The patient is placed with a small sandbag under the shoulders, so that the neck is extended with the face to the left. Extension and rotation should be decreased in older children. The anterior border of the sternomastoid muscle is identified, as is the apex of the triangle formed by the sternal and clavicular heads of the muscle. The internal jugular vein passes behind the clavicle at a point behind the medial edge of the clavicular head of the muscle. It may be punctured just above the clavicle or, preferably, through the muscle body at a point just below the apex of the triangle formed by the two heads of the sternomastoid muscle. The position of the skin puncture varies slightly, depending on the degree of extension of the neck. Firm pressure over the liver and a head-down tilt causes the vein to fill, so that the line of the vein can be identified. The higher approach is preferable because it is associated with a lower incidence of complications. It is important to remember that the internal jugular vein is relatively superficial in a child and should be entered within 1–2 cm. Failure to observe this basic rule may lead to puncture of the cervical pleura and lung or one of the major vessels in the root of the neck, which may be abnormal in both size and position in patients with congenital heart defects. In very small infants, particularly those who are hypovolaemic, it is usual to transfix the vein and enter the lumen only during withdrawal of the cannula. The venous pressure must be checked to ascertain correct placement of the cannula because the colour of the blood is unreliable.

For those less experienced and for training purposes, it is recommended that ultrasonic guidance be used to locate the internal jugular vein (Grebenik *et al*., 2004). At all times, there must be close attention to detail, the avoidance of even minute air bubbles, and a full aseptic technique in order to avoid line colonization with *Staphylococcus epidermidis*.

A 22- or 24-gauge Teflon cannula is inserted percutaneously into the radial or ulnar artery and connected to a short length of manometer line and three-way tap. Sampling therefore takes place a short distance from the cannula, and so the cannula can be securely strapped in position. Children over 2 years of age usually have easily identifiable radial arteries, and puncture of only the anterior arterial wall enables the cannula to be inserted easily. In smaller infants, success is more readily achieved if the artery is transfixed and the cannula withdrawn until blood appears when the cannula

is then inserted up the lumen of the vessel. Imprecise arterial puncture results in a subintimal haematoma, which makes further attempts at cannulation unlikely to succeed. Alternative sites are the brachial, axillary and femoral arteries. The smaller the cannula inserted, the longer the line tends to last postoperatively. Most satisfactory results are obtained with a fine-gauge, non-tapered cannula. Also, the life of a near-occlusive cannula is usually less than 24 hours because of thrombosis of the artery.

The system must have a continuous slow flush of heparinized saline (1 U/ml) using an intraflow device at 3 ml/h, except in small infants, when a syringe pump set at 1 ml/h is used to avoid excess flush fluid. With care, complications of percutaneous lines are very few in children.

Maintanance of Anaesthesia

A nasogastric tube, a urinary catheter, and nasopharyngeal and peripheral skin temperature probes are placed, and the patient is transferred to the operating table on a warming mattress, switched off at this stage. Arterial blood is taken at the earliest opportunity for baseline blood gases, electrolytes, sugar and the activated clotting time (ACT) measurements.

After connection of the monitoring lines, anaesthesia is continued with, for example, fentanyl by small boluses of 1–2 μg/kg up to 10–20 μg/kg just before the incision is made and repeated before bypass, together with a full dose of relaxant to prevent respiratory movements (Duncan *et al*., 2000). The combination of pancuronium, which causes tachycardia, and fentanyl, which produces bradycardia, usually achieves very stable haemodynamics and has been in use since the 1970s.

Remifentanyl, the ultra-short-acting opioid, can be used as a continuous infusion throughout the procedure in children of all ages at a dose of 0.5–1.0 μg/kg/h (Weale *et al*., 2004). Other forms of analgesia are given before the infusion is stopped, as the activity of remifentanyl is so short. The risk of awareness is reduced by the addition of up to 1% isoflurane or boluses of 0.2 mg/kg midazolam.

The use of the anaesthetic agent, propofol, as a continuous infusion of 6–8 mg/kg/h is increasingly popular for the older child (not licensed for use in children below 3 years of age). The infusion is continued in the early postoperative period at a lower rate of 2–3 mg/kg/h and allows transfer from theatre to the ICU with stable haemodynamics. When the infusion is stopped, the patient rapidly wakes up and can then be extubated.

Epidural and spinal analgesia are increasingly used in some centres and have the advantage of greatly reducing the stress response of the surgery without large doses of intravenous opioids (Hammer *et al*., 2000). However, the risk of haematoma in the heparinized patient remains an anxiety (Bosenberg, 2003).

The inspired oxygen concentration should be increased if the cardiac output falls in the pre-bypass period. Blood gas analysis is repeated as increasing acidosis is common and is corrected with 8.4% sodium bicarbonate if the base deficit is greater than 5 mmol/l.

Rapid transfusion may be necessary during sternotomy in cyanotic children or those having reoperations, as bleeding from collaterals or dense adhesions may be profuse. Surgical manipulation always interferes with cardiac output, although it may be useful to begin bypass with only one venous cannula in the right atrial appendage, which causes minimal disturbance, and to put the second cannula in the IVC after partial bypass is established. If pressure is low after manipulations cease, then a bolus of 10% $CaCl_2$ is a powerful inotropic stimulus, but there is a risk of severe bradycardia or even asystole if K^+ is low or myocardial oxygenation is borderline. A very small dose of 1:100 000 adrenaline is the alternative.

A 300 U/kg dose of heparin is administered at least 5 minutes before arterial cannulation, and the adequacy of heparinization is estimated after 3 minutes using the activated clotting time, which should be prolonged to more than 400 seconds and maintained above that level throughout cardiopulmonary bypass. Aprotinin (Trasylol) is a proteinase inhibitor, preventing fibrinolysis, and enhances platelet function by preserving α2-antiplasmin levels. It is commonly used to reduce the need for blood products in children having complex surgery, reoperations or any surgery where excessive bleeding is anticipated (Miller *et al*., 1998). The common dosage regime is a slow bolus of a test dose of 1–2 ml, then a slow bolus dose to the patient of 1–2 ml/kg, followed by a continuous infusion of 1–2 ml/kg/h until bleeding stops. The same bolus dose is put into the pump prime after it has been demonstrated that the patient is not sensitive to the drug. Hypersensitivity is, however, rare in children, even after multiple exposures.

CARDIOPULMONARY BYPASS

Once bypass is fully established and venous return blood is well saturated, ventilation is discontinued and the lungs are allowed to deflate. No apparent benefit is derived from the application of 2–3 cmH_2O of pressure during bypass.

Any systemic-to-pulmonary shunt, such as a persistent ductus arteriosus or a Blalock–Taussig shunt, is occluded as soon as perfusion is started, so that the lungs are not flooded. The left side of the heart is vented, and cooling is commenced with a full flow of 2.4 l/min/m^2. A perfusion

pressure lower than 30 mmHg (4 kPa) is usually caused by flow through systemic pulmonary collateral vessels, and the flow should be temporarily increased above the calculated level. Vasoconstrictors, such as methoxamine, should be avoided where possible. As cooling progresses, a perfusion pressure of 40–60 mmHg is usual during periods of full flow. If the pressure exceeds these levels, either isoflurane can be introduced from the pump or an infusion of glyceryl trinitrate (2–10 μg/kg/min) will be effective. Haemodilution to a haematocrit of 20–25% promotes peripheral blood flow and optimizes tissue oxygenation. For stabilizing membranes and to reduce the inflammatory response, 30 mg/kg methylprednisolone is used in some centres before circulatory arrest, but there is no convincing evidence that it alters outcome (Lodge *et al*., 1999).

At low temperatures cerebral metabolism and oxygen consumption are greatly reduced, so that at core temperature of 18°C they are approximately 17% of basal levels. Periods of deep hypothermic circulatory arrest are kept to a minimum, but 30 minutes is usually considered reasonable at core temperatures of 18°C (Swain *et al*., 1991a). This technique is often used for small babies where the usual venous cannulas would obstruct the surgical field. Cessation of cerebral electrical activity and maximal jugular venous bulb oxygen saturations are a guide to optimal cerebral cooling, which should be even and adequate. To aid complete cerebral cooling and to ensure that rewarming does not occur during the arrest period, the head is surrounded by ice packs. Topical cooling and modified ultrafiltration help cerebral function to normalize very quickly after rewarming.

The alternative is to maintain a low flow with the venous cannulas left in place. Flow rates as low as 0.5 l/min (50 ml/kg/min) can be used, which provide a reasonably blood-free surgical field and therefore require only moderate hypothermia (22–25°C) (Swain *et al*., 1991b).

During hypothermia, pH and $PaCO_2$ are maintained within the normal range without correction for temperature, the alpha-stat mode of regulation that maintains electrochemical neutrality. This results in a high intracellular pH during hypothermic ischaemia and helps in the preservation of high-energy phosphates, functional enzymes, ion concentrations, and cell volumes necessary for the regeneration of depleted high-energy phosphates at the time of reperfusion. Myocardial function may be better preserved.

Potassium supplementation of 1 mmol/kg is always required on bypass to compensate for preoperative deficits and losses into the cells, urine and bypass fluid and can be given slowly during the course of the bypass. Serum potassium should be kept in the high normal range (above 4 mmol/l). Serum Mg^{++} is also low after diuretic therapy and is further reduced by bypass, and is not reflected in serum levels. The administration of Mg^{++} (0.25 mmol/kg) prevents coronary spasm, reduces the incidences of dysrhythmia, reduces SVR and promotes cardiac output. It may also minimize deleterious effects of myocardial reperfusion. Lactate levels are usually low, even after long periods of arrest.

Before the commencement of the intracardiac repair, the aorta is cross-clamped and blood cardioplegia is used to produce asystole and further cool the myocardium. A reduced dose of cardioplegic solution is used at half-hourly intervals. Perfusion techniques and cardioplegia are discussed in detail in Chapter 11.

Catecholamine levels are enormously elevated after deep hypothermic circulatory arrest and cause peripheral vasoconstriction, which is occasionally severe. The increase occurs largely at the time of reperfusion and involves adrenaline and noradrenaline. Rewarming should be slow, avoiding gradients in excess of 10°C between perfusate and body core temperature. The heated mattress can be turned on at this stage. During rewarming, the intense vasoconstriction requires the use of a vasodilator, such as glyceryl trinitrate (GTN) or the ino-dilator milrinone. The perfusion pressure may be maintained at approximately 40 mmHg, but towards the end of rewarming it is essential that the pressure rises, to allow a period of adequate myocardial perfusion before bypass is terminated, and this may require the addition of an inotropic agent. Before the aortic cross-clamp is removed, every trace of air must be removed from the heart, particularly from the left side, and the aortic root is vented with low-level continuous suction until bypass is discontinued. To aid the de-airing process, the patient is put in a head-down position and the lungs distended to expel blood into the left side of the heart, displacing any air, which can be removed from the aortic root. The venous pressure is raised to 5–6 mmHg in order to achieve a pulsatile flow with some cardiac output and to prevent entry of air through an atrial suture line. Direct intracardiac monitoring lines are placed; left atrial pressure measurement is essential when left ventricular function is poor, when mitral valve surgery has taken place or disease is present, or when a Fontan-type procedure has been performed. Direct pulmonary artery pressure measurement is helpful when a reactive pulmonary vasculature is suspected. The incidence of complications is low, although bleeding may occur on removal or the line may fracture, thus requiring surgical removal.

Ventilation of the lungs is recommenced, initially with 100% oxygen, tracheal suctioning undertaken, and areas of atelectasis re-expanded so that bypass can be discontinued when the nasopharyngeal temperature has

been at 35–36°C for several minutes and the muscle mass and extremities are warm.

POST-BYPASS

After venous drainage has stopped, the patient is transfused via the aortic cannula to a systolic pressure of 80 mmHg and SVC pressure of 10–12 mmHg. Where there is anxiety about LV function, LA pressure is also measured and used as a guide to ventricular filling. It is difficult to maintain peripheral vascular dilatation in a hypovolaemic patient, and so the atrial pressure is raised to 10–12 mmHg unless there is a specific reason to use a lower filling pressure. Overtransfusion from the bypass must be avoided because it is easy to push volume beyond the optimal point on the Starling curve, and the distensible liver has a high venous capacitance in the infant. Caval cannulae are removed quickly because of the danger of caval obstruction.

Occasionally cardiopulmonary bypass may produce lung and tissue damage, with severe extravasation of plasma and water into all tissues and subsequent organ dysfunction. This is a problem particularly in small infants, in whom the ability to quickly excrete large volumes of crystalloid fluid may be limited and increased lung water results in difficulty in weaning from respiratory support. Complement activation is assumed to play a role, with sequestration of granulocytes in the lungs. Many of these deleterious effects may be reversed by modified ultrafiltration, using the technique described by Naik *et al*. (1991). As ultrafiltration proceeds, the haematocrit rises, pulmonary artery pressure falls and cardiac output improves.

If systolic pressure remains low with high filling pressures, then inotropic support is necessary (Booker, 1998). Infants tend to have a rate-dependent cardiac output, so that if the rate is low or there is junctional rhythm, an optimal cardiac rate is achieved either by external pacing or the use of isoprenaline. Isoprenaline is seldom used alone, as it may cause tachycardia and should be avoided if there is residual muscular outflow tract obstruction, as this may increase. Dopamine in a dose of 5–10 μg/kg/min may be effective, or dobutamine in the same dosage if there is anxiety about reactive pulmonary vessels. Dopamine is often used in a lower dose to improve renal perfusion, although this effect on mesenteric flow may be the result of improved myocardial activity. Neonates have a variable response to dopamine and may require a higher dose, up to 15 μg/kg/min or more, to achieve the same clinical response. Higher doses than 10 μg/kg/min cause vasoconstriction and increased ventricular afterload, which should be offset by an infusion of GTN 1–2 μg/kg/min.

An effective combination (nowadays becoming the first choice) is an infusion of milrinone, a phosphodiesterase inhibitor, at a rate of 0.5–0.75 μg/kg/min, after a slow loading dose, together with adrenaline, (300 μg/kg in 50 ml 5% dextrose, starting at 1 ml/h = 0.1 μg/kg), the infusion titrated to achieve the desired haemodynamics (Bailey *et al*., 1999; Hoffman *et al*., 2003). Afterload reduction is particularly important in patients with mitral valve regurgitation.

After ultrafiltration, when the haematocrit has been raised to approx 40, an infusion of protamine is started with a very small test dose, and the amount is slowly increased to a dose of 6 mg/kg under ACT control. Severe hypotensive episodes have been described, with pulmonary vasoconstriction associated with the use of protamine, and are minimized by very slow infusion into a peripheral rather than a central line (Ralley, 1999).

The aortic cannula is removed as soon as major bleeding is controlled. In newborns, the aortic cannula may be partially obstructive and may have to be removed shortly after termination of bypass. Blood or fresh-frozen plasma is given so as to maintain atrial pressure at a preselected level, usually about 12 mmHg, although this may be increased to 15 mmHg if the systemic pressure is less than 80% of normal. LA pressure should not exceed 15 mmHg.

Bleeding after cardiopulmonary bypass is common and is multifactorial. There is thrombocytopenia with abnormalities of platelet function and a reduction in fibrinogen and the activity of coagulation factors, complement activation and fibrinolysis. Small babies are particularly affected, because below 6 months of age they have reduced levels of coagulation factors and below 3 months a prolonged partial thromboplastin time, together with an increased inflammatory response to CPB. Blood products, such as fresh frozen plasma (FFP), cryoprecipitate or platelets, drugs such as aprotinin, and recombinant coagulation products may all be required (Miller *et al*., 1997). Topical agents, such as ''FlowSeal'', have been shown to be very useful. The thromboelastograph, which measures whole blood coagulation, is increasingly used after cardiopulmonary bypass. It correlates well with the platelet count, shows fibrinolysis and is useful in assessing the effectiveness of treatments (Shore-Lesserson *et al*., 1999).

Temporary pacemaker wires are inserted routinely, usually to allow sequential atrio-ventricular pacing if necessary. Significant dysrhythmia, such as junctional tachycardia, although not common in children, does occur, causing a fall in cardiac output when the myocardium is stressed. Fast dysrhythmia with a marked fall in cardiac output can be treated with D.C. shock or 50 μg/kg increments of adenosine to a maximum of 250 μg/kg, otherwise amiodarone 5 mg/kg, followed by

infusion to a maximum of 20 mg/kg/day, is the treatment of choice.

Urine output should be at least 1 ml/kg/h after bypass, and if this is not achieved, 0.2–0.3 mg/kg furosemide should be given and cardiac output optimized. Diuretics should also be given if haemoglobinuria is observed. Potassium levels require regular checking and the serum level should be maintained above 4 mmol/l by a slow, controlled infusion of dilute potassium.

Analgesia is achieved with an infusion of 1.0 mg/kg morphine in 50 ml 5% dextrose running at 1–2 ml/h, which yields 20–40 μg/kg/h. Neonates may be given 5–10 μg/kg/h because excretion of morphine is significantly reduced in this age group. Morphine given by the caudal route is popular in some centres (Rosen and Rosen, 1997).

Fast-tracking

Many units have evolved clinical care pathways for the management of many lesions, so that deviations from the normal progress of, for example, a repair of tetralogy of Fallot, can be easily and quickly realized and corrected (Price *et al*., 1999). This has led to the concept of fast-tracking for many children with even complex lesions (Lake, 2000). Short-acting anaesthesia drugs, such as propofol and remifentanyl, together with efficient postoperative analgesia, allow many children to be extubated very soon after surgery and to pass through the intensive care unit in the shortest possible time. Early extubation reduces the time in the ICU and saves costs (Heinle *et al*., 1997). Criteria for early extubation include return of reflexes, good pain relief, good gas exchange on a low FiO_2 and low respiratory rate and stable haemodynamics, with reasonable cardiac support and no bleeding.

PULMONARY HYPERTENSION

Pulmonary hypertension is a common cause of problems post-bypass in children with pre-existing pulmonary hypertension and is worsened by pulmonary endothelial dysfunction caused by the bypass. The subject is well reviewed by McEwan (2005). With L–R shunts and with pulmonary venous obstruction (TAPVC), mitral stenosis, duct-dependent circulations and TGA, the smooth muscle of the media of the pulmonary arterioles does not regress at birth but increases, so that these vessels become very contractile. This causes lability in the pulmonary circulation, as the vessels readily constrict with stimuli such as hypoxia and acidosis via an adrenergic mechanism (Morray *et al*., 1988). If the foetal channels (DA and PFO) are open, the increased PVR causes a reverse to a R–L shunt with profound hypoxaemia. This is the situation with persistent pulmonary hypertension of the newborn.

If the channels are closed, as they are after cardiac surgery, a rise in PVR causes right heart failure, followed by failure of the left ventricle, since in infancy there is marked interventricular dependency. The pulmonary vascular changes eventually become irreversible with the onset of pulmonary vascular disease.

In high-risk cases, the PA pressure is usually monitored after bypass via a direct line into the PA and the effect of pulmonary artery vasodilators can be assessed. A fall in PVR is usually seen during modified ultrafiltration by a mechanism involving reduction in endothelin-1 levels. The phosphodiesterase inhibitors, such as milrinone, improve ventricular function and also promote pulmonary vasodilatation and are the first-line treatment of choice. Other drugs previously used in the treatment of sustained pulmonary hypertension post-bypass included phenoxybenzamine, nitroglycerine and prostacycline, but have been largely superseded by inhaled nitric oxide (NO) (Frostell *et al*., 1991). The main effect of inhaled NO is the activation of guanylate cyclase, which promotes cyclic GMP, a vasorelaxant. Nitric oxide is occasionally required to wean patients from bypass where other measures have failed (Kadosaki *et al*., 2002).

RE-DO SURGERY

Many patients undergo reoperation at some stage in their lives. Those whose lesions required the use of a homograft conduit, such as truncus arteriosus, Rastelli procedure or correction of pulmonary atresia, will need conduit replacement, either because they have grown out of the size originally used or because the conduit has become progressively narrowed with calcification. The conduit may lie just beneath the sternum or even attached to it. The greatest care is required during the opening of the sternum and usually one iliac region is prepared for immediate bypass should excessive bleeding be encountered. Occasionally femoro-femoral bypass is started from the beginning. Extra venous access, with as wide-bore a cannula as possible, is advisable, and also having blood and colloid ready, together with a rapid infusion device, in case urgent transfusion is suddenly required. Aprotinin is usually used in redo cases to reduce bleeding and to minimize the use of blood products. External defibrillator pads are placed beneath the chest before the start of surgery.

SPECIAL CONSIDERATIONS

Norwood Operation

The Norwood procedure is undertaken for the definitive palliation of hypoplastic left heart syndrome, which is a spectrum of disease of left ventricular and aortic

hypoplasia. The surgery is performed in three stages and is discussed in Chapter 41. Stage I consists of reconstruction of the aortic arch and a central shunt to supply the pulmonary blood. After this, the pulmonary and systemic blood flow depends on the relative resistances, the aim being to achieve a Qp:Qs of 0.8:1.0. These infants are usually sick preoperatively and require careful management and ventilation at all stages with a low FiO_2 to keep the pulmonary resistance high, thus minimizing pulmonary blood flow at the expense of the systemic circulation. The infusion of prostaglandin to maintain ductal patency, which is crucial for systemic blood flow, is continued until the start of bypass. Arterial oxygen saturations of approximately 80% are ideal. Strategies to change the PVR may be necessary postoperatively and calculation of the Qp:Qs can be made from the mixed venous (SVC as a close approximation) saturation, which provides mixed venous values. Function of the right ventricle is maximized with the use of inotropic agents postoperatively. The Sano modification (RV–PA conduit, rather then a B–T shunt) has decreased operative mortality, possibly by improving ventricular diastolic function, and has made postoperative management of the first stage more straightforward. As the reconstruction is in three stages, all of which require bypass, sites of venous and arterial vascular access must be preserved where possible. The femoral route is favoured to avoid thrombo-embolic problems in the SVC–PA anastomosis during stages II and III.

Univentricular Heart

Surgical management involves the Fontan procedure or one of its variations, such as the total cavopulmonary connection (TCPC). For success, the procedure, which is classified as definitive palliation and which involves passive pulmonary blood flow, relies on good function of the sole ventricle and low/normal pulmonary vascular resistance. The procedure may be staged, starting with a Glenn shunt (SVC to right PA) followed by a completion of TCPC at a later date. Sometimes a fenestration between the pulmonary and systemic venous atrium or the IVC/PA conduit is created. The details are discussed in the Fontan chapter. After bypass it is mandatory to continuously monitor both left atrial and central venous (PA) pressures. This gives a guide to the trans-pulmonary blood flow. If the gradient is greater than 12–15 mmHg, steps must be taken to reduce pulmonary vascular resistance if there is thought to be a reversible component. Modified ultrafiltration and the phosphodiesterase inodilator, milrinone, may be sufficient, but inhaled NO may also be needed if the Fontan circulation is to succeed (Miller *et al*., 2000).

It is advisable to have central venous access to the SVC and IVC. The SVC lines are removed in the early postoperative period to avoid thrombo-embolism in the SVC–PA pathway. Continuing access is then via the IVC route.

Transplantation

The management of anaesthesia and perfusion are similar to those for any patient undergoing open-heart surgery, but there are additional problems in a number of areas. The topic is reviewed by Schindler *et al*. (2004).

Most paediatric transplant recipients are not in hospital at the time when a donor organ becomes available and preparation time is usually at a minimum. An adequate fasting period is unlikely to be available. Induction and pre-bypass anaesthesia is unlikely to present severe problems in heart–lung recipients, even if respiratory failure is advanced, and these patients usually benefit from mechanical ventilation and tracheal toilet. Patients with progressive cardiomyopathy may deteriorate rapidly and irreversibly, and pre-bypass anaesthesia must be administered with the greatest care. Ventricular stroke volume is reduced with a fixed upper limit, and cardiac output is dependent on both heart rate and preload. Drug-induced myocardial depression, heart rate reduction, and small changes in preload and afterload may all cause an irreversible decline in blood pressure and cardiac output. This condition may not respond well to β-adrenergic agonists, because of receptor downregulation caused by a reduced receptor density in the failing heart. All anaesthetic agents should be used cautiously by titration against clinical effect, but once even modest hypotension occurs, only the institution of cardiopulmonary bypass may break a downward path. Etomidate or sevoflurane, pancuronium and fentanyl form the basis of a satisfactory anaesthetic technique. Ketamine, which is such a useful induction agent in most children with cardiac disease, should be used with caution in end-stage cardiac disease, where endogenous catechoalamine levels are likely to be depleted. A rapid sequence induction should be used where there is a possibility of significant gastric contents. Insertion of vascular access catheters must be undertaken quickly, with a careful aseptic technique, because infection is a major cause of morbidity. An additional catheter for administering immunosuppressant drugs is helpful. Right internal jugular access should be avoided in order to facilitate postoperative myocardial biopsies; however, if vascular access is difficult, this route should be used and an alternative found postoperatively. During cardiopulmonary bypass, attempts should be made to correct pre-existing electrolyte and acid–base abnormalities that may be severe and to improve the peripheral circulation by the use of vasodilators.

The implanted donor heart is denervated and responds only to agents that activate receptors directly. Isoprenaline can be used at the termination of bypass because of its combined chronotropic and inotropic effects, but the combination of milrinone and adrenaline is a successful one. The transplanted heart requires adequate preload, and higher-than-usual right atrial pressures may be required. Right ventricular failure is not an uncommon problem and assist devices can be used postoperatively until recovery.

The transplanted lung usually requires a period of PEEP at 8–10 cmH_2O before systemic oxygenation is satisfactory, presumably in order to reverse distal airway closure. Excessive levels of PEEP should be avoided if the lungs are small for the recipient chest cage, because lung rupture may occur. Postoperative and long-term rejection remains a problem after paediatric heart–lung transplantation and regular trans-bronchial lung biopsies may be required in order to monitor rejection and the development of fibrosing alveolitis.

CLOSED CARDIAC OPERATIONS

Approximately 25% of cardiac surgery for congenital disease is performed without bypass, usually via a thoracotomy for procedures such as ligation of persistent ductus arteriosus, relief of coarctation of the aorta, formation of Blalock–Taussig shunts and banding of the pulmonary artery. Many of the patients are very cyanosed and/or in severe heart failure. Some may require preoperative intensive care with respiratory and cardiac support. Levels of monitoring, vascular access and induction and maintenance of anaesthesia are usually the same as for open-heart surgery. A nasogastric tube is required for all patients, as ileus and gastric stasis are common after thoracotomy. Small, sick infants benefit from a period of respiratory support postoperatively.

Patent Ductus Arteriosus (PDA)

All premature babies are at risk for PDA, as the mechanisms that cause the duct to close at birth are also immature (Friedman and Fahey, 1993). Fluid overload in the neonatal period will also cause the duct to reopen. A PDA is usually a single chance finding, but is also associated with other congenital cardiac conditions, some of which require ductal patency for continuing survival. Approximately 25% of very low birth weight babies with hyaline membrane disease have a PDA. The increased pulmonary blood flow and reduced pulmonary compliance contribute to the development of chronic lung disease, so that any ventilator-dependent baby with a PDA should have this occluded. In the absence of a murmur, the diagnosis is made on routine echocardiography. As the babies are small, sick and often oxygen/ventilator-dependent, they require very careful monitoring, with the operation sometimes being performed in the neonatal intensive care unit. An anaesthesia technique with relaxant, ventilation with an air/oxygen mixture and up to 10 μg/kg fentanyl is a suitable technique for those infants not expected to breathe afterwards. If the infant is to be extubated, then a lower dose of opioid should be given together with a regional technique, such as intercostal nerve blocks with bupivacaine 0.25% 0.5–1 ml per space. Spinal anaesthesia has been used successfully for this procedure (Williams and Abajian, 1997).

Attention to temperature maintenance, blood glucose level and blood calcium level is required. Rapid changes in arterial and venous pressure from either hypoxaemia or rapid blood volume changes must be avoided because of the danger of intraventricular haemorrhage from rupture of fragile cerebral capillaries. Careful monitoring of arterial and inspired oxygen levels is required and development of retinopathy of prematurity is minimized if the oxygen saturations are maintained at 90–92% during the procedure. Blood should be ready to transfuse in the rare event of rupture of the duct, otherwise transfusion is rarely necessary.

Weaning from respiratory support by means of intermittent mandatory ventilation and constant positive airway pressure may take several days, during which time chest physiotherapy is essential.

In the older child a PDA may be a chance finding or may present with heart failure and shortness of breath. Most of these are occluded by interventional cardiology, an occlusion device being introduced via a cardiac catheter. This requires a straightforward anaesthetic. Occasionally a very large duct requires surgical treatment, and during the ligation a short period of induced hypotension with isoflurane or sodium nitroprusside will make the surgery safer. An alternative technique is video-assisted minimal invasive closure. This usually requires the use of an endobronchial tube or bronchial blocker, so that the left lung can be collapsed during surgery.

Coarctation of the Aorta

Neonates These babies are often very sick because the coarctation is usually severe and the anastomotic circulation may be minimal at this stage. Clinical diagnosis is confirmed by echocardiography, which will also pick up any other cardiac pathology. The left ventricle may be hypertrophied with poor function and patchy ischaemic necrosis; pulmonary oedema and renal failure are common. Prostaglandin infusion to maintain patency of the ductus arteriosus is necessary,

but respiratory and inotropic support may also be required preoperatively. It may be beneficial for all the monitoring lines to be inserted before the baby comes to the operating theatre. It is preferable if the arterial line and the pulse oximeter probe are in the right arm, as the left subclavian artery is clamped during the surgery. Anaesthesia, which provides the greatest cardiovascular stability, is required and a technique involving relaxant, oxygen/air and analgesic, such as fentanyl 10–20 μg/kg, given slowly, is very suitable. The hypertrophied ventricle does not usually tolerate inhalational agents. Serum potassium may be low and will contribute to cardiac instability.

Careful attention to intraoperative ventilation, correction of acidosis and blood replacement is required. Unclamping the aorta after the repair is the time of greatest risk and the patient may become hypotensive, which is not tolerated by the myocardium, used to a high coronary perfusion pressure, and ventricular fibrillation may occur. It is advisable to have a syringe of colloid ready to transfuse at this stage. The aortic clamp can be reapplied for a brief period to allow adequate coronary perfusion and replacement of blood loss, although care should be taken to avoid over-transfusion. The peripheral vasculature is reactive and an infusion of sodium nitroprusside may be necessary later to maintain systolic pressure around 80 mmHg. A period of respiratory support postoperatively is advisable, although recovery from simple coarctation is usually rapid.

Older Children These patients are usually hypertensive, but otherwise fit and have developed collaterals through the intercostal arteries. During thoracotomy, mild induced hypotension without tachycardia is useful to minimize bleeding from the collaterals in the chest wall, which may be profuse. This is achieved by using a combination of labetalol 0.05–1.0 mg/kg and isoflurane to obtain the desired level of pressure. Arterial monitoring is from the right arm. If the arterial pressure rises dramatically with clamping of the aorta, this indicates poor collateral flow and is an indication for further induced hypotension, e.g. with sodium nitroprusside. In the absence of adequate collaterals, reducing the pressure may be dangerous and knowledge of distal pressure is important. The clamping time should not exceed 25–30 minutes, as ischaemia of the spinal cord is a risk the longer the clamping time. More complicated cases may require the use of heparin-bonded shunts or a form of left heart bypass. Sodium nitroprusside or labetalol infusion is usually required postoperatively to maintain the blood pressure below 100 mmHg. The patients are extubated in theatre and require full analgesia and sedation. Continuous thoracic epidural infusions have been used for intra- and postoperative analgesia in these cases (Bennett *et al*., 1995). It is, however, not practised in our institution.

Pulmonary Artery Banding

Banding of the pulmonary artery is undertaken in those infants with a large L–R shunt, not amenable to early corrective surgery. The banding reduces the extent of the shunt, reduces the likelihood of permanent pulmonary vascular disease and reverses the deleterious effects of high pulmonary blood flow. The work of the left ventricle is reduced, lung compliance improves and the work of breathing is lessened. Currently this operation is performed infrequently.

These patients may be very sick, ventilator-dependent and/or in unchecked cardiac failure. The surgical approach is via a left thoracotomy or sternotomy and requires full cardiovascular monitoring. The limiting factor may be systemic desaturation, with a reversal of the shunt which, if severe, causes bradycardia and fall in cardiac output. Severe desaturation is also a limiting factor when banding cyanotic patients (TGA or DORV + VSD). Pulse oximetry is very useful to check progress during the banding process and final oxygen saturation is not usually below 80% (Casthely *et al*., 1987). Occasionally pre-existing lung disease is severe and a marked oxygen gradient may make difficult a final level of banding. Postoperative ventilation is usually required for a period of time to allow recovery of lung function.

Systemic–Pulmonary Artery Shunt

An anastomosis between the subclavian and pulmonary arteries of the same side is used to improve pulmonary blood flow and oxygenation in patients in whom pulmonary blood flow is reduced as a result of severe pulmonary stenosis or atresia, in patients with tricuspid atresia, and in some patients with tetralogy of Fallot or complex transposition. The commonly performed shunt in infants is a modified Blalock–Taussig shunt, with a 4 mm Gore-Tex tube end-to-side between the two vessels. Details of the Gore-Tex shunt and alternative types of shunts are described in detail in Chapter 16. The main problems are polycythaemia, hypoxaemia and metabolic acidosis. Venous and central venous access is required for transfusion, central venous pressure measurements and inotropic agents if necessary. If the internal jugular vein on the side of the shunt is to be used, the greatest of care must be taken not to damage the subclavian artery. Direct arterial measurements should avoid subclavian territory to be shunted. The other radial or a femoral artery is suitable. Excessive perioperative fluid deprivation associated with polycythaemia may lead to cerebral thrombosis or thrombosis of the

shunt, although the latter is more likely to arise from hypotension or technical problems associated with the vascular anastomosis. In older children, considerable bleeding from small anastomotic vessels may occur during dissection of the lung hilum to isolate the pulmonary artery. Such bleeding may be aggravated by a mild coagulopathy caused by prolonged severe polycythaemia. Volume replacement is by fresh-frozen plasma or plasma protein fraction. While the vascular anastomosis is performed, the shunted pulmonary artery is clamped, and great care must be taken to ensure that there is no interference with the blood flow to the opposite lung. A high FiO_2 is used, full muscle relaxation maintained, and 0.5–1.0 mmol/kg sodium bicarbonate given if the patient is severely hypoxaemic, in order to avoid an increase of acidosis during the procedure. Heparin (1 mg/kg) is given before the clamps are applied on the pulmonary artery, so as to prevent thrombosis along the shunt suture line during the first few hours. It is also important to maintain an adequate blood pressure because any period of hypotension predisposes to shunt occlusion and episodes of desaturation.

With a large shunt, pulmonary blood flow may be excessive, with very high oxygen saturations (> 90%). In these cases, inspired oxygen should be minimized to promote pulmonary vasoconstriction and limitation of lung blood flow.

Vascular Ring

A vascular ring around the trachea and oesophagus is usually based on a double aortic arch, with several variations (see Chapter 22). Less commonly it is caused by a pulmonary artery sling when the left pulmonary artery rises from the right pulmonary artery. Patients with pulmonary sling may have a hypoplastic trachea with complete cartilaginous rings, so-called ''stovepipe trachea''. This should be excluded preoperatively, as an extensive tracheal reconstruction may be required as well as treating the sling. The common presenting symptom is stridor, and some degree of oesophageal narrowing can usually be demonstrated on preoperative oesophageal swallow studies and confirmed by echocardiography. The degree of tracheal obstruction is variable, and ranges from minimal with little or no stridor to extensive with severe stridor, with considerable compression and softening of the tracheal wall and in some cases severe tracheal stenosis. Vascular ring is a cause of respiratory arrest and sudden infant death.

The surgical approach in the majority of patients is via a left thoracotomy and may cause considerable retraction on the trachea, which may result in intermittent airway obstruction. Care should be taken with intubation to have the tip of the tube at least in the mid-trachea. Hand ventilation allows early warning of tracheal obstruction during the dissection. Tracheal suction may be required, as peripheral secretions may become troublesome with lung handling and retraction. Most patients can be extubated after reversal of muscle relaxant and nursed in a humidified head box with added oxygen. Stridor is often still obvious postoperatively and intravenous dexamethasone (0.25 mg/kg) can be given in order to prevent any additional problems from acute oedema. Nebulized adrenaline is also useful.

A small number of patients develop severe stridor and require nasotracheal intubation for several days. Airway distending pressure, which can be continued after extubation with nasal cannulas, may be necessary because tracheomalacia is usually present.

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Further Reading

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13

Intensive Care Management of the Postoperative Patient

D. Macrae and J. Larovere

Successful postoperative recovery from cardiac surgery is best assured by efficient preoperative diagnosis and management, timely presentation for surgery, and excellent perioperative teamwork. Cardiologists, surgeons, intensivists, anaesthetists and nurses must understand the anatomical diagnosis and associated physiology, the treatment plan and their role in its achievement. Preoperative diagnosis must be complete if intraoperative or postoperative problems related to incorrect or unsuspected problems are to be prevented. Intensive care management should be patient and lesion-specific and planned to anticipate predictable and deal with possible postoperative events. Successful intensive care management is reliant on cohesive teamwork between intensivist, surgeon, cardiologist and nurses, with good communication at all levels being central to good clinical outcomes. A firm base for successful intensive care is to have a clear structure to guide the process of admission of the child following surgery (**Table 13.1**).

Table 13.1 Immediate management on ICU admission.

Transfer patient to ICU ventilator and ICU monitors
Obtain a detailed handover from surgeon and anaesthetist (procedure, intraoperative events, current status)
Rapid clinical examination
Chest X-ray (to determine tube, line and drain positions; lung fields clear; effusions; heart size)
12-Lead electrocardiogram
Baseline laboratory profiles, including full blood count, coagulation screen and plasma electrolytes, arterial blood gases and lactate
Mixed venous saturation measurement to calculate arterial–venous O_2 difference
Echocardiogram (if not performed intraoperatively) to assess surgical result and myocardial function
Discussion with parents to explain initial status and plans for immediate postoperative management

CARDIOVASCULAR SYSTEM

Monitoring

Minimal monitoring standards for children in a paediatric cardiac intensive care unit include ECG, arterial and central venous pressure monitoring, central and peripheral temperature, and pulse oximetry. End-tidal CO_2 monitoring is desirable in all ventilated children. In selected cases, left atrial or pulmonary artery lines may be placed by the surgeon to assist in postoperative management of children at risk of significant left ventricular dysfunction or pulmonary hypertension. Immediately following surgery most children require a 1:1 nurse:patient ratio, although some complex cases, such as those requiring ECMO support or haemofiltration, may require 2:1 nursing. Once a child is more stable, especially when no longer requiring mechanical ventilation or additional complex care, they may make the transition through a short period of ''high-dependency'' care, during which nursing care of two patients is shared by one nurse, before transfer to a paediatric ward. Knowledgeable bedside nurses are a key element in the safe delivery of intensive care to a postoperative child. Their roles include making and interpreting clinical observations, administering drugs and fluids, reassuring the child, supporting parents and, very importantly, acting as an advocate for the child and as a focus for the child's clinical care.

Even relatively simple monitoring of postoperative modalities can provide useful pathophysiological insights which will guide management. Increasing tachycardia may coincide with rising central temperature,

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which, following cardiopulmonary bypass, can be an indication of a significant inflammatory response, may be associated with onset of low cardiac output, arise from an accelerating abnormal rhythm such as junctional ectopic tachycardia, or simply be an indication of pain or discomfort.

Vascular Access

Arterial cannulae to facilitate continuous monitoring of arterial blood pressure and intermittent blood gas sampling are placed either preoperatively in the intensive care unit in an unstable patient or at induction of anaesthesia for surgery, generally in the radial or femoral arteries. Alternative sites include axillary, brachial and pedal arteries. The ulnar artery should be avoided, as the ipsilateral radial artery may have been previously cannulated or will be in future. The brachial artery territory has little or no collateral supply, therefore lines in this site should be used infrequently and removed as soon as alternative access is achieved. A small term neonate requires a 24G cannula in the radial position, or a 22G in the femoral or axillary artery. In children undergoing coarctation or more extensive aortic arch repair, arterial lines may be placed in both the right radial (precoarctation) and a femoral artery (postcoarctation).

In the early neonatal period, an umbilical arterial catheter can be utilized. The umbilical stump is prepared and one of the arteries is identified, with sterile dissection in the 4 or 8 o'clock position. An umbilical arterial catheter is inserted and advanced. The position of the tip should lie above the diaphragm but below the left subclavian artery, usually in the aorta above the ductus arteriosus. In a child with no other access, resuscitation fluids and drugs may be given into a properly placed umbilical arterial catheter, provided that no air bubbles are introduced.

Central venous cannulae are vital for both intra- and postoperative patients to provide secure vascular access for fluids and infusions as well as access for oxygen saturation measurement. Small diameter multilumen catheters (4 or 5.5 Fr) are available in a range of lengths suitable for percutaneous placement, even in neonates. Choice of site for percutaneous cannulation depends mainly on the preference of the operator, with internal jugular, subclavian and femoral veins commonly used. To prevent damage to cardiac structures, the tips of central venous catheters must not encroach into the right atrium. Some units insert single or multilumen catheters at the time of surgery, which can be removed percutaneously several days postoperatively, aiming to preserve percutaneous sites for later procedures or investigations. Insertion-related risks from percutaneous central venous cannulation include bleeding, accidental arterial cannulation and damage to adjacent structures, the occurrence of which can be minimized by training. Two particular complications of central venous lines relevant to the intensivist are infection (O'Grady *et al*., 2002; Polderman and Girbes, 2002) and line-related venous thrombosis. Lines must always be placed and manipulated aseptically, and removed as soon as possible in the postoperative period. Heparin-bonded catheters may reduce the incidence of catheter-related thrombosis, and antibiotic-coated catheters may reduce the incidence of infective and thrombotic complications (Randolph *et al*., 1998; Pierce *et al*., 2000).

Additional monitoring may be required in selected patients. In patients with poor left ventricular function, following mitral valve repair or Fontan surgery, a left (systemic) atrial pressure line is often inserted surgically to permit direct assessment of filling conditions of the systemic ventricle. For those patients at significant risk of severe pulmonary hypertension in the postoperative period, a catheter can be placed in the pulmonary artery by the surgeon. Trends in pulmonary artery pressure give early warning of the onset of pulmonary hypertension, which may otherwise only be detected with the onset of acute right ventricular failure.

Trans-cutaneous intracardiac lines should be removed when no longer required. Complications occurring at removal include retention (inability to remove all or part of the catheter) and bleeding (Gold *et al*., 1986). It is essential to check that coagulation times and platelet count are normal, that a unit of cross-matched blood is available and that a surgeon is available within the hospital for the rare occasion that bleeding into the pericardium results in haemodynamically compromising pericardial tamponade. An echocardiogram should be obtained soon after line removal to look for pericardial effusion, particularly in those patients in whom their mediastinal drains have already been removed. Cardiac tamponade must be ruled out if haemodynamic deterioration occurs soon after removal of a transthoracic device, such as a pacing wire or monitoring line.

Low Cardiac Output

The heart must pump blood around the circulation at a sufficient flow rate (cardiac output) to meet the body's metabolic needs. If cardiac function is sufficiently impaired (a low cardiac output state), delivery of oxygen and other substrates will be compromised. A sustained period of low cardiac output is associated with a high risk of multiorgan dysfunction, often encapsulated by the term *low cardiac output syndrome* (LCOS). The occurrence of LCOS is predictable in many situations, and its management must be proactive to ameliorate the potentially serious consequences of multiorgan

dysfunction, which commonly include impairment of renal, hepatic and/or cerebral function.

Objectively, if somewhat arbitrarily, low cardiac output (LCO) is said to exist when measured cardiac index falls below 2 l/min/m^2. Cardiac indices below this level have been shown to be associated with high mortality (Parr *et al*., 1975). The precise incidence of LCO meeting this definition is unknown in the current era, as relatively few centres measure cardiac output routinely. However, it is known that haemodynamics immediately after complex surgery, such as the arterial switch operation, are often impaired (Wernovsky *et al*., 1995) detected a fall in cardiac index following arterial switch operations, with a median maximal decrease of 32% occurring 6–12 hours after separation from bypass, with gradual return to the prebypass cardiac index at 24 hours after the procedure (Wernovsky *et al*., 1995). This fall in cardiac index was associated with elevation of both systemic and pulmonary vascular resistance.

Clinical assessment of the child should be performed sequentially throughout the postoperative period, as interpretation of trends is more useful than a single set of observations. The clinical manifestations of LCO include slow capillary refill time, large core–peripheral temperature gradient and oliguria. Hypotension may be present, but LCO can exist even in the absence of hypotension, as systemic vascular resistance may be elevated. Reduced cerebral perfusion may manifest as irritability in neonates and infants or confusion in older children.

Metabolic indicators associated with LCO include high or rising plasma lactate, and low mixed venous oxygen saturations. Lactate is used as an energy-producing substrate in tissues, including skeletal and cardiac muscle, when mitochondrial oxidation is not restricted by lack of oxygen. As tissue metabolism shifts from aerobic to anaerobic, there is both an increase in lactate production and a reduction in lactate utilization, which results in increased plasma lactate. In the postoperative child hyperlactaemia with clinical signs of low cardiac output should be assumed to be the result of tissue hypoxia and treated accordingly. Normal plasma lactate concentrations are < 2 mmol/l. A high plasma lactate which falls postoperatively at a rate of 1–2 mmol/l/hour is an indication of previous tissue hypoxaemia, whereas a persisting high or rising lactate indicates probable concurrent tissue hypoxaemia. Also of value in assessing circulatory sufficiency is the arteriovenous (A-V) oxygen difference, derived from the Fick principle relating cardiac index to oxygen consumption:

$$\text{Cardiac index} = \frac{\text{Oxygen consumption}}{\text{Arteriovenous oxygen difference}}$$

Therefore, if oxygen consumption and oxygen carrying capacity of the blood remain constant, then a fall in mixed venous oxygen saturation indicates increased tissue extraction and implies a fall in cardiac index. The normal mixed venous oxygen saturation is 75%, which, if the systemic arterial saturation is 100%, gives an A-V oxygen difference of 25%. In low output states, as A-V extraction increases, the mixed venous saturation falls and the A-V difference rises. In the postoperative patient a mixed venous saturation below 65% strongly suggests high tissue oxygen extraction, due to low cardiac output. Measurement of the true venous saturation can only be achieved via a surgically placed pulmonary artery catheter. In a child with no residual intracardiac shunt values, values recorded from the superior or inferior vena cava are commonly used to reflect the mixed venous saturation. Such samples reflect only upper or lower body oxygen consumption, respectively, but can be expected to reflect significant improvement or deterioration in cardiac output.

Pharmacological Management of Low Cardiac Output (LCO)

The measures necessary for the management of LCO depend on its precise cause (Wessel, 2001), the main determinants of which are listed (**Table 13.2**).

Inotropic Drugs

In clinical practice, adrenergic agonists delivered by continuous infusion and titrated to haemodynamic effect are the mainstay of pharmacological management of LCO (Booker, 2002). When systolic ventricular function is impaired, dopamine, dobutamine or low-dose adrenaline are used as ''first-line'' inotropes. Additional agents should be administered according to assessment of response judged clinically and from available haemodynamic monitoring. Higher-dose adrenaline is useful when more potent inotropic stimulation is required. Noradrenaline or vasopressin can be used if refractory vasodilatation is present, such as occurs rarely after cardiopulmonary bypass in children and in septic shock. Isoprenaline is a non-specific β-adrenergic agonist whose principal cardiovascular effects are vasodilatation and increasing heart rate effects. The drug is rarely used in intensive care except to increase heart rate where atrial pacing is not feasible. Caution is needed when higher dose catecholamine support is used, as the agents may reduce ventricular compliance (worsen diastolic function) (**Table 13.3**).

Historically, low-dose dopamine infusions have been used for renal protection. Whilst dopamine augments renal blood flow and increases urine volume and sodium excretion in animals and healthy humans, there is

Table 13.2 Factors contributing to postoperative low cardiac output.

Factor	Comment	Action
Heart rate		
Slow heart rate	Neonates and infants have a rate-dependent cardiac output	Consider atrial (AAI) pacing or infusion of isoprenaline
Fast heart rate	May impair ventricular filling by shortening diastole	Treat pyrexia; ensure sedation/analgesia, hypovolaemia
		ECG to diagnose possible tachyarrhythmia
		Treat arrhythmia if present
Preload	Inadequate preload will impair ventricular filling	Assess (clinical, CVP, LAP).
		Fluid challenge 5–10 ml/kg and assess response
Afterload		
Low afterload	Causes include excessive vasodilatation and run-off through systemic–pulmonary artery shunts or a patent ductus arteriosus	Discontinue vasodilator drugs. Consider vasoconstriction with noradrenaline or vasopressin
	Danger of myocardial ischaemia if diastolic pressure too low	Shunting through PDA/surgical shunts may be reduced by factors which cause pulmonary vasoconstriction (high CO_2, low O_2, etc.)
High afterload (SVR and PVR)	Common consequence of cardiac surgery and cardiopulmonary bypass	Consider surgical revision of shunt/duct closure
Pulmonary hypertension	Predominant pulmonary hypertension: particular risk following repair of anomalous pulmonary venous drainage; truncus arteriosus; and patients with long-standing pulmonary recirculatory (L–R) shunts or high pulmonary venous pressures	Anticipate. Use milrinone or a similar inodilator as part of the pharmacological support postoperatively
		General measures to lower PVR include optimizing oxygenation; inducing mild alkalosis; and optimizing cardiac output
		Consider pulmonary vasodilatation with inhaled nitric oxide
Contractility	Impairment of ventricular function following surgery may be due to direct myocardial (surgical) injury; ischaemia–reperfusion; the systemic inflammatory response to cardiopulmonary bypass and other factors, including metabolic derangements	Assess function with echocardiogram
		Predominant *systolic dysfunction* ≫ inotropic support
		Predominant *diastolic* dysfunction ≫ maintain preload and consider phosphodiesterase inhibitor
	Preoperative steroid; careful surgical technique; effective use of cardioplegia and modified ultrafiltration may prevent postoperative cardiac dysfunction	
Cardiac tamponade	Haemodynamically significant haemorrhagic cardiac tamponade	Surgical drainage/haemostasis required
		Re-open sternal wound or delay sternal closure
Residual lesions	''Tissue tamponade'' may result from oedema of intrathoracic tissues. Always seek to identify, and if necessary correct, residual or previously undiagnosed lesions	Echocardiogram
		Cardiac catheterization and angiography should be undertaken if postoperative progress is unexpectedly slow
Metabolic milieu	Severe acidaemia may produce significant myocardial depression	Correct acidaemia
	Low ionized calcium impairs contractility	Correct calcium to normal concentrations
	Hyperthermia increases tissue oxygen demand and is undesirable in low cardiac output states	Aggressive cooling to prevent hyperthermia
		Induced mild hypothermia (34–36°C) reduces metabolic oxygen demands and may assist in maintaining oxygen balance in LCO states

Table 13.3 Vasoactive infusions.

Vasoactive amines	Infusion rate	Receptor pharmacology				Comments
		DA	β^1	β^2	α	
Dopamine	1–5 μg/kg/min	++	+	−	−	First-line inotropic drug
	5–10 μg/kg/min	++	++	−	+	If > 10 μg/kg/min, change to adrenaline
	10–20 μg/kg/min	++	++	−	++	
Dobutamine	1–20 μg/kg/min	−	++	+	−	Alternative first-line inotropic drug
Isoprenaline	0.05–2 μg/kg/min	−	+++	+++	−	Inotropic, chronotropic and vasodilating actions Used to treat bradycardia
Adrenaline (USA, epinephrine)	0.01–0.1 μg/kg/min	−	++	++	+	First-line inotrope if used at low dose
	0.2–0.5 μg/kg/min	−	++	+++	+++	Potent inotropic and dose-related vasoconstriction at doses > 0.1 μg/kg/min
Noradrenaline (USA, norepinephrine)	0.01–0.5 μg/kg/min	−	++	+	++++	Predominantly vasoconstrictive action Use to treat hypotension due to vasodilatation
Inodilators						
Milrinone	50–75 μg/kg loading 0.25–0.75 μg/kg/min	Acts by inhibition of cAMP breakdown by phosphodiesterase				Administer loading dose over 20–30 minutes to avoid hypotension Systemic and pulmonary vasodilatation; mild inotropic and lusiotropic action; synergistic effect with β-adrenergic inotropes
Vasodilators						
Glyceryl trinitrate (USA, nitroglycerine)	0.5–5 μg/kg/min	Dilates smooth muscle by drug metabolism, leading to release of nitric oxide				Mild reduction in systemic vascular resistance Systemic venodilatation and pulmonary dilatation
Sodium nitroprusside	0.5–5 μg/kg/min	Similar mechanism of action to glyceryl trinitrate				Potent arteriolar dilator Easily titrated due to short half-life Danger of cyanide toxicity with prolonged or high-dose infusion
Prostaglandin E1 or E2	5–30 ng/kg/min	Direct action on tissue of ductus arteriosus or PDA				Used to maintain patency of the PDA in neonates with duct-dependent circulations
Prostocyclin (epoprostenol)	1–20 ng/kg/min	Acts via cyclic AMP pathways				Potent vasodilator of both pulmonary and systemic circulations Inhibits platelet aggregation and may cause bleeding

now compelling evidence from clinical trials which proves that low-dose dopamine does not prevent the onset or alter the course of acute renal failure in critically ill humans. Finally, there is some evidence that dopamine is harmful to the gastrointestinal, endocrine, immunological and respiratory systems in patients with critical illness (Debaveye and Van den Berghe, 2004).

Phosphodiesterase inhibitors have positive inotropic, vasodilitory and lusiotropic effects. Milrinone has been investigated extensively in adult and paediatric practice and its use in the immediate postoperative period is now common practice particularly in combination with low dose β-adrenergic drugs such as dopamine, dobutamine and adrenaline. It is also commonly started prophylactically, either coming off bypass or shortly after arrival in PICU, in those patients deemed at significant risk of developing LCOS. Its use in this manner is supported by the findings of the PRIMACORP trial, which reported a significant reduction in a clinical diagnosis of LCOS in patients receiving prophylactic infusion of milrinone compared to the control group. It is interesting to note that patients who developed LCOS had a significantly longer cumulative duration of mechanical ventilation (1.7 days, $p = 0.001$) and hospital stay (2.4 days, $p = 0.016$) when compared to those who did not develop LCOS, suggestive of a positive benefit to therapy which avoids LCO (Hoffman *et al*., 2003). Phosphodiesterase inhibitors are therefore indicated in situations where high SVR is poorly tolerated by the myocardium and impairs cardiac output, as in postoperative neonates and young children.

Ionized hypocalcaemia has a negative inotropic influence on the myocardium, especially in neonate, where mechanisms for the storage and handling of calcium are immature. In neonates with a clinical picture of LCO, ionized hypocalcaemia should be treated with intravenous calcium replacement and a daily maintenance allowance of calcium administered via i.v. fluids or milk feeds.

Systemic Vasodilators

Vasodilators may be employed in the management of systemic hypertension, which can occur in children following repair of aortic coarctation or relief of left ventricular outflow obstructive lesions, and in the management of cardiac failure. Systemic vasodilators should be used with extreme caution in patients with systemic hypotension and those with left ventricular outflow obstruction who are at risk of uncompensated severe systemic hypotension and myocardial ischaemia. In children sodium nitroprusside is the most commonly used agent in the ICU because of its powerful arteriolar dilating effect and short half-life, which render it both effective and highly titratable. Glyceryl trinitrate (nitroglycerine) is an alternative short-acting drug which acts only as a weak arteriolar dilator at high doses, but is an effective venodilator and is used in treatment of severe congestive heart failure and in the management of coronary ischaemia. Phenoxybenzamine, a long-acting α-adrenergic blocker, may reduce variations in systemic vascular resistance and maximize systemic blood flow in, for instance, the post-Norwood Stage 1 circulation (Tweddell *et al*., 1999). Phenoxybenzamine bonds covalently to α-adrenergic receptor, has a very long duration of effect and is difficult to titrate. If hypotension occurs in a patient receiving phenoxybenzamine, noradrenaline or vasopressin will be required to counteract the effects of the drug. Alternative vasodilatory strategies are believed to achieve similar effects, although a recent study concluded that phenoxybenzamine was superior to sodium nitroprusside when used intra-operatively (Motta *et al*., 2005).

Longer-term vasodilator therapy using angiotensin-converting enzyme (ACE) inhibitors, (in children able to absorb enterally) such as captopril and enalapril, have been found to have haemodynamic benefit in heart failure caused by left-to-right (L–R) shunts and systolic dysfunction of the systemic ventricle. Prolonged treatment with ACE inhibitors has been shown to be effective in reducing not only LV volume overload but also LV hypertrophy in the hearts of growing children with chronic LV volume overload (Mori *et al*., 2000).

Recently, preliminary data have suggested that levosimendan (Kivikko *et al*., 2003), a newly available inotropic drug which acts by calcium-sensitizing mechanisms, may have a role in the management of ventricular failure in the perioperative child.

Non-pharmacological Strategies in the Management of Postoperative Low Cardiac Output

Core cooling may be used as a temporary strategy in children with LCOS, aiming to lower tissue oxygen demand and therefore improve tissue oxygen balance. Cooling to 34–35°C requires that the patient be sedated and paralysed with a neuromuscular blocker to inhibit shivering. A vasodilator such as sodium nitroprusside may be used to facilitate cooling, provided systemic blood pressure is adequate.

Ventilatory strategies can be used to influence cardiac output in some situations. Positive airway pressure improves cardiac output in left ventricular failure, whereas it has the opposite effect on right ventricular function and cardiac output in the Fontan circulation. Negative pressure ventilation has been shown to improve cardiac output in children with severe right ventricular dysfunction following repair of Fallot's tetralogy and in the Fontan circulation

(Shekerdemian, 2001). Clinical use of negative-pressure ventilation in the early postoperative period, requires a cuirasse to surround the chest, which can be complicated by the presence of wires, lines, wound dressings and drains. Ultimately, if adequate systemic perfusion cannot be established, the use of mechanical circulatory support (ECMO or VAD) should be considered sufficiently early to avoid secondary organ damage (see Chapter 14).

Other Cardiovascular Drugs

Diuretics such as frusemide (often given i.v.), are widely used in heart failure treatment in the perioperative period (Singh *et al*., 1992) to optimize fluid balance, and are crucial in the face of potentially impaired renal function. Continuous infusions of frusemide produces a smoother control of fluid and electrolyte shifts than intermittent i.v. bolus administration (Klinge, 2001). If tolerance to frusemide develops, concomitant use of thiazide diuretics such as chlorthiazide or metolazone, which act synergistically with loop diuretics, may improve diuresis. Care must be taken in all patients receiving diuretics to maintain circulating volume and normal electrolyte concentrations, including potassium, calcium and magnesium.

β-Blockers such as propranolol or atenolol are primarily used in the management of systemic hypertension following coarctation repair, although they may also have a place in the management of dynamic right ventricular outflow tract obstruction secondary to muscular obstruction. Initial β-blockade may be achieved via a continuous esmolol or labetolol infusion, titrating from a low dose until the desired end-point is achieved. Intermittent propranolol doses may also be used.

Pulmonary Hypertension

Children with L–R shunts leading to pulmonary overcirculation or obstruction to pulmonary venous drainage (pulmonary venous obstruction, mitral valve stenosis) are at risk of developing pulmonary hypertension (PHT) (Wessel, 2001). PHT increases right ventricular afterload, potentially resulting in acute right ventricular failure, and may occur despite technically successful cardiac surgery. Pulmonary vascular reactivity has been related not only to the presence of preoperative pulmonary hypertension but also to longer cardiopulmonary bypass times and the associated inflammatory response. A number of strategies for the treatment of pulmonary hypertension are listed in **Table 13.4**.

Management of pulmonary hypertension following surgery should be directed at the cause. In addition to vascular reactivity, elevation of left atrial pressure, pulmonary artery stenosis and residual L–R shunts

Table 13.4 Pulmonary hypertension.

Factor	Influence	Management strategy
Oxygen	Lowers pulmonary vascular resistance (PVR)	Maintain good oxygenation
pH	PVR rises as pH falls	Avoid respiratory acidosis Avoid metabolic acidosis Consider inducing moderate alkalosis
Lung volume	PVR elevated at low and at high lung volumes	Avoid atelectasis Avoid overdistension
Cardiac output	Low output impairs coronary perfusion and results in acidosis	Optimize cardiac output Consider inotropic support
		Treat pain and agitation with appropriate analgesics and sedatives and, if required, neuromuscular blockade
Sedation/analgesia	Associated with lower PVR, less reactivity	Most intravenous vasodilators lower both PVR and systemic vascular resistance (SVR)
Vasodilators	Lower PVR	Inhaled nitric oxide is a selective pulmonary vasodilator

may raise pulmonary artery pressure postoperatively. Vascular reactivity is managed initially by optimization of oxygenation, ventilation, cardiac output and analgesia, following which either a non-selective (milrinone) or selective (inhaled nitric oxide) pulmonary vasodilator may be added. Inhaled nitric oxide (iNO) is commonly commenced at 20 parts per million (ppm) which may be reduced to 5–10 ppm once a satisfactory response and clinical stability are achieved (Miller *et al*., 2000; Macrae *et al*., 2004). Other relatively selective pulmonary vasodilators include nebulized prostacyclin, oral or intravenous sildenafil although the clinical safety and efficacy of these agents in paediatric congenital heart disease remains unproven at present. Oral sildenafil has been shown to facilitate discontinuation of iNO in children with postoperative pulmonary hypertension and may act synergistically with iNO in management of PHT (Atz and Wessel, 1999; Atz *et al*., 2002).

Cardiac Tamponade

Cardiac tamponade occurs when fluid or intrathoracic structures compress the heart, restricting venous return and cardiac chamber filling, thereby reducing cardiac output and blood pressure. In the immediate postoperative period the two common causes of cardiac tamponade are the presence of blood within the pericardium and compression of the heart by adjacent mediastinal structures, so-called 'tissue tamponade'. Cardiac tamponade should be suspected if there is a sudden or gradual onset of hypotension associated with elevated heart rate, left atrial and central venous pressures, and reduction in cardiac output and hence pulse volume with inspiration (pulsus paradoxus).

A pericardial effusion may be evident on both chest X-ray and echocardiogram, the latter of which may demonstrate the presence of atrial diastolic collapse. It is important to stress that neither a chest X-ray nor an echocardiogram can completely exclude cardiac tamponade, so if haemodynamically compromising tamponade is suspected following cardiac surgery the chest should be reopened to facilitate evacuation of fluid or blood clot and control bleeding if necessary. In the case of ''tissue tamponade'', cardiac output improves if the chest is reopened and stented, creating extra space for mediastinal structures. Delayed sternal closure following surgery may offset potentially compromising haemodynamics in the postoperative period, a strategy which is associated with low rates of infectious complications despite the open wound (Iyer *et al*., 1997). Until cardiac tamponade is relieved, the maintenance of adequate preload (volume status) and afterload (avoidance of vasodilators) is essential to maximize cardiac output.

CARDIOPULMONARY RESUSCITATION

Cardiopulmonary resuscitation in the postsurgical child is fundamentally different to that in the community, and must be directed towards rapid restoration of the patient's cardiac and respiratory function, coupled with identification and treatment of the underlying cause.

In the non-ventilated child, rapid assessment of the conscious level, airway patency and work of breathing and circulation should be performed as per normal basic life support, prior to proceeding to intubation, vascular access and advanced life support. In the ventilated child, rapid assessment of the endotracheal tube and ventilatory circuit should be rapidly performed to exclude mechanical obstruction. Hand ventilation in 100% oxygen not only improves ventilation and oxygenation but also provides valuable information on airway compliance. The majority of children in intensive care will have central intravenous access, but if not this should be established immediately. Resuscitation drugs, including adrenaline, sodium bicarbonate and calcium, are administered as per normal protocol. The ratio of cardiac compressions to ventilated breaths varies according to age; 5:1 for infants and children under 8 years and 15:2 for children over 8 years. The cardiac compression rate is 100 compressions/minute for all age groups. The UK Resuscitation Council produces a wide range of guidance on resuscitation (http://www.resus.org.uk).

In infants and children with congenital cardiac disease, identifiable causes of cardiac arrest should be considered and may be coincident with recent events, such as cardiac tamponade following intracardiac line removal or tension pneumothorax following subclavian venous puncture. Once basic life support has been established, rapid progression to maximal cardiovascular and respiratory monitoring is mandatory, including ECG to assess cardiac rhythm, end-tidal CO_2 to quantify efficacy of cardiopulmonary resuscitation and arterial waveform to analyse cardiac output and arterial pressure, either with or without chest compressions. If central venous or arterial access is in place, a blood gas to assess gas exchange, electrolyte and metabolic status should be performed as soon as permissible.

Cardiopulmonary resuscitation in cardiac intensive care is only as effective as those providing it. Efficient teamwork and adequate, recurrent training of all concerned coupled with a comprehensive knowledge of potential causes of cardiac arrest helps ensure a positive

outcome. Rapid deployment ECMO has also shown to be effective in some resuscitation situations.

Postresuscitation Stabilization

Resuscitation does not end with return of spontaneous circulation, as efforts must then be directed at supporting and improving cardiac output, optimizing tissue reperfusion. It is usual for the post-arrest myocardium to require inotropic support as the effect of resuscitative epinephrine recedes. Ventilation of the lungs should be continued until cardiorespiratory stability is achieved, avoiding both hypercapnia (risk of raising intracranial pressure) and hypocapnia (risk of excessive restriction of cerebral blood flow). Metabolic normality should be obtained through gradual correction of acidosis, maintenance of normoglycaemia and avoidance of hyperthermia.

FLUID THERAPY AFTER CARDIAC SURGERY

Fluid shifts occur in response to the activation of neuroendocrine and inflammatory pathways induced by cardiopulmonary bypass, particularly increased vascular permeability, which in its most florid form results in generalized tissue oedema and multiorgan dysfunction, often termed ''capillary leak syndrome''. Cardiopulmonary bypass is associated with an increase in total body water, even in the absence of major organ dysfunction (Naik *et al*., 1991). Neonates and infants and those with preoperative activation of their immune system are at particular risk of developing clinically significant capillary leak syndrome (Seghaye, 2003). Strategies such as preoperative steroid administration, intraoperative high-dose aprotinin and the use of modified ultrafiltration (Gaynor, 2003) have been promoted as potentially useful means of modifying these adverse responses.

Volume Replacement

All blood products, including human albumin solution, carry a small risk of transmission of infective agents, including viruses and prions. For this reason all blood products must only be used for clearly defined indications. Alternative non-blood fluids should be used for routine red cell-free volume replacement. A target haematocrit should be decided on a patient-by-patient basis, based on their pathophysiological status. Children with cyanotic heart disease and patients in cardiac failure require higher haematocrits (40–45%) to optimize systemic oxygen delivery than do other 'well' postoperative children, in whom lower haematocrits (30–35%) can be tolerated and therefore transfusion avoided. The choice of *colloid* or *crystalloid* solutions for non-red cell volume replacement is controversial. Although slightly larger amounts of crystalloid solutions are required to sustain plasma volume expansion compared to colloids solutions, there is no evidence that the routine use of colloid solutions leads to substantially improved outcomes compared to crystalloids. All plasma volume expanders cause dilution of circulating clotting factors. We recommend two alternative strategies for plasma volume expansion: 0.9% sodium chloride or lactated Ringer's solution in children at low risk of developing ''capillary leak'', and polygeline solution (Gelofusin) in others, given as infusions of 5–10 ml/kg of the fluid over 30–60 minutes.

BLEEDING AND BLOOD TRANSFUSION

Bleeding is an ever-present risk after cardiac surgery, the result of inadequate surgical haemostasis or acquired clotting abnormalities. Factors commonly associated with postoperative coagulopathy include incomplete heparin reversal, dilution of clotting factors and destruction or inactivation of platelets. In addition to assuring surgical haemostasis, strategies to minimize postoperative blood loss include complete reversal of heparin with protamine, the intraoperative use of aprotinin and the use of modified ultrafiltration. Although the whole blood activated clotting time (ACT) is the mainstay of anticoagulation management during cardiopulmonary bypass, more sophisticated approaches available in the operating room and ICU include the use of combined standard and heparinase ACT to detect residual heparin activity and thromboelastography, which may reveal coagulation defects at an early stage and guide their management.

On admission to the ICU following surgery, a baseline coagulation screen and platelet count should be obtained. If ongoing bleeding occurs, administration of protamine and blood products should be guided by the results of admission and follow-up haematological tests (**Table 13.5**). Whilst some blood loss via chest drains is expected after cardiac surgery, drainage is usually minimal after 3–4 hours. Abnormal surgical bleeding may be sufficient to require re-exploration of the chest cavity, especially if blood loss exceeds 10% blood volume in 1 hour or 20% in 4 hours.

ARRHYTHMIAS AND PACING

Postoperative arrhythmias, a relatively common complication following cardiac surgery (Hoffman *et al*., 2002),

Table 13.5 Blood products.

Product	Indication for treatment—bleeding occurring *and*:	Haematological abnormality
Protamine (0.5–1 mg/kg)	Residual heparin activity	Prolonged ACT, normal heparinase ACT APTT > 2 × control, PT < 1.5 × control
Fresh frozen plasma (10–15 ml/kg)	Prolonged prothrombin time	PT > 1.5 × control
Cryoprecipitate (5 ml/kg)	Low fibrinogen	Fibrinogen < 1 g/l
Platelets (10–15 ml/kg)	Low platelets following CPB	Platelets < 100×10^9/l
	Low platelets following surgery without CPB	Platelets < 50×10^9/l
	Prophylaxis for extreme thrombocytopenia	Platelets < 20×10^9/l

APTT, activated partial thromboplastin time; ACT, activated clotting time; PT, prothrombin time.

result from a variety of causes, including injury or oedema to the cardiac conduction system, changes in myocardial electrophysiological properties, myocardial ischaemia, or acute changes in haemodynamics causing, for instance, atrial distention. Arrhythmia management (**Table 13.6**) should be directed towards restoration of a physiological heart rate and atrio-ventricular synchrony to maximize cardiac output, especially in the early postoperative period when arrhythmias may exacerbate low cardiac output syndrome. All patients should have a preoperative 12-lead ECG recorded and those with a complex arrhythmia history or at high risk for arrhythmias discussed preoperatively with an electrophysiologist. Further 12-lead ECGs should be performed on return from the operating theatre and in the event of sustained arrhythmia *prior* to any subsequent drug administration or intervention.

Table 13.6 Basic principles of arrhythmia management for the intensivist.

Ensure preoperative and immediate postoperative 12-lead ECGs have been recorded for later comparison
Discuss patients with complex arrhythmia histories with an electrophysiologist prior to surgery
Discuss any incidents of perioperative arrhythmias and precipitating factors with surgeon and anaesthetist
Ensure abnormalities of plasma electrolytes are corrected
Obtain a 12-lead ECG of arrhythmia prior to antiarrhythmic drug administration
Record ECG after any subsequent interventions including drugs and overdrive pacing
Aim to restore atrio-ventricular synchrony and physiological heart rate as quickly as possible
Broad complex tachycardias should be treated as VT until otherwise proved
Profound haemodynamic instability due to arrhythmia requires urgent DC cardioversion

All patients at risk of arrhythmias, and probably all children undergoing cardiac surgery, should have temporary epicardial pacing wires attached at the time of surgery to facilitate arrhythmia diagnosis and management in the early postoperative period. Two wires are placed on the anterior surface of both the right atrium and right ventricle, which when attached to a temporary pacing box will provide bipolar sensing and pacing. Temporary pacing allows restoration of cardiac excitation and rate to as near normal as possible, may act as a back-up in situations of sudden loss of cardiac excitation and facilitate the overdrive pacing of tachyarrhythmias.

Early Postoperative Tachyarrhythmias

Atrial Ectopic Tachycardia

Atrial ectopic tachycardia (Rosales *et al*., 2001) occurs when an area of atrial myocardium develops increased automaticity. This may be from a single or multiple foci within the atria. Atrial ectopic tachycardia is therefore classified as an automatic tachycardia, which cannot be terminated by direct current or pharmacological cardioversion or overdrive pacing On the surface ECG, changes in P wave morphology, axis and PR interval may all be apparent with a normal QRS complex as ventricular activation proceeds via the His–Purkinje system. Atrio-ventricular conduction is most commonly 1:1, although atrio-ventricular block may be apparent

if the atrial rate is sufficiently fast. If the diagnosis is uncertain, intravenous adenosine will increase the degree of atrio-ventricular block, revealing persisting discrete atrial activity with variable conduction to the ventricles. Management is directed at correction of potentiating factors (serum electrolyte abnormalities) and slowing the ectopic rate with either intravenous β-blockade (esmolol), amiodarone or class 1 agents, such as flecainide. Atrial ectopic tachycardia commonly settles after the immediate postoperative period, although treatment may need to be continued as an outpatient.

Junctional ectopic tachycardia (JET), also referred to as *His bundle tachycardia*, is the tachyarrhythmia which most commonly requires treatment in the postoperative period (Dodge-Khatami *et al*., 2002a, 2002b). It is a malignant arrhythmia associated with serious morbidity, including prolonged ventilation and intensive care stay. Its mechanism is most likely secondary to trauma or haemorrhagic infiltration of the His bundle during surgery. JET is also an automatic tachycardia driven by a focus within or immediately adjacent to the atrio-ventricular (A-V) junction and His bundle and, like AET, does not convert with programmed stimulation or cardioversion. JET displays a ''warm up'' and ''cool down'' phenomenon. In the immediate postoperative period the junctional focus will gradually ''warm up'' and the heart rate increase leading to sequential changes in the ECG, viz. a shortening of the PR interval followed by retrograde atrial activation (P waves after the QRS complex) and ultimately complete A-V dissociation, as the A-V node lying on the atrial aspect of the junctional focus cannot conduct every impulse back to the atria. If the diagnosis is not immediately apparent, administration of adenosine will demonstrate a narrow complex tachycardia independent of the atria, therefore originating from the His bundle. If the atrial complex is difficult to see on the surface ECG, an atrial ECG may also provide useful diagnostic information.

As atrial systole provides approximately 25% of cardiac output, the loss of A-V synchrony and an associated tachycardia, further reducing the time for passive ventricular filling, can prove catastrophic, especially in a patient who already has low cardiac output syndrome. Our aim in managing JET is to slow the ventricular rate sufficiently to permit atrial overdrive pacing, thereby restoring A-V synchrony. This may be achieved at first by surface cooling to 34–35°C and, if unsuccessful, by the administration of intravenous amiodarone (25 μg/kg/min for 4 hours, then a continuous infusion at 5–15 μg/kg/min). The surface temperature can often be allowed to normalize once the child has been sufficiently loaded with amiodarone. While JET usually resolves within 2–3 days of the restoration of normal sinus rhythm, it may be occasionally necessary to continue amiodarone orally for a further 5–10 days.

Atrio-ventricular Re-entry Tachycardia (AVRT)

Atrio-ventricular re-entry tachycardia (AVRT) mediated by accessory pathways may be seen for the first time following surgery or may be a pre-existing condition. As AVRT is dependent on the atrio-ventricular node, acute termination can be achieved by the administration of intravenous adenosine (0.2 mg/kg), thereby interrupting the re-entrant circuit. In patients dependent preoperatively on antiarrhythmic drugs, the same regimen should be introduced as soon as possible, always considering the potentially negative inotropic and chronotropic effects of such agents.

Ventricular Tachycardia (VT)

VT following cardiac surgery is rare, occurring most commonly in the setting of pre- or postoperative ventricular dysfunction. All broad complex tachycardias should be managed as VT until proved otherwise. Lesions classically associated with a risk of VT following surgery include complex left ventricular outflow tract obstruction, tetralogy of Fallot and anomalous coronary artery from the pulmonary artery (ALCAPA). ''Torsades des pointes'' is a polymorphic ventricular tachycardia, where the QRS complexes rotate around an imaginary baseline. It is an unstable rhythm which may rapidly degenerate to ventricular fibrillation. It may be caused by profound ventricular disease or electrolyte imbalance prolonging the baseline QT interval. DC cardioversion should be used wherever haemodynamic instability is present. Amiodarone, lignocaine or magnesium may be used if VT has not resulted in haemodynamic deterioration.

Atrial Flutter (AFL)

AFL is rarely seen after cardiac surgery. It may occur in the setting of right atrial dilatation, e.g. following repair of total anomalous pulmonary venous drainage or a large atrial septal defect. The 12-lead ECG shows a classic ''saw-tooth'' baseline due to continual atrial activation, with varying degrees of atrio-ventricular block. If the diagnosis is in doubt, intravenous adenosine (0.2 mg/kg) will unmask the flutter waves. Antiarrhythmic medication rarely terminates atrial flutter and in certain circumstances may slow the atrial conduction, permitting 1:1 atrio-ventricular conduction and causing severe haemodynamic compromise. DCC cardioversion or overdrive oesophageal pacing are the treatments of choice.

Postoperative Bradycardia and Pacing

Bradycardia following cardiac surgery may be caused by sinoatrial or atrio-ventricular nodal dysfunction, leading to reduced ventricular rate and hence lowering cardiac output.

Sinus Bradycardia and Sinus Node Dysfunction

As heart rate is intricately linked to cardiac output, disproportionate slowing of the sinus rate (due to excessive sedation or reduced central body temperature) may lead to haemodynamic compromise. This may be overcome by atrial pacing (AAI) at faster rates. Surgery at the junction of the right atrium and superior vena cava or right atrial roof may injure the sinus node itself or its arterial supply. Operations classically associated with this phenomenon are repair of sinus venosus atrial septal defect, especially if baffling of anomalous pulmonary veins to the left atrium is necessary, and either the Mustard or Senning procedures. As with sinus bradycardia, this can be managed with atrial pacing until sinus node function resumes. This is often a transient phenomenon which resolves, although if prolonged it may require implantation of a permanent pacing system.

Atrio-ventricular Node Dysfunction

Atrio-ventricular (A-V) block complicates 2–3% of congenital cardiac surgery. In a large prospective series Weindling and colleagues (1998) found the greatest risk for A-V block with surgery for left ventricular outflow tract obstruction, ventricular septal defect and tetralogy of Fallot, or in patients with congenitally corrected transposition of the great arteries. Two-thirds of patients recovered A-V conduction, > 95% of whom did so by the ninth postoperative day, suggesting that there is little benefit in delaying permanent pacemaker implantation beyond this point. In patients in whom concern has been raised intraoperatively about integrity of the A-V conduction system, careful evaluation, including a 12-lead electrocardiogram, should be made on return to the ICU, as simultaneous atrial and ventricular rates may often mask underlying A-V block. If uncertain, a diagnosis may be made by atrial pacing at a faster rate, which would fail to conduct to the ventricles in the presence of A-V node dysfunction. DDD pacing is the treatment of choice for postoperative A-V block, with daily evaluation for signs of recovery. Due to the sudden loss of A-V conduction in some patients, where normal conduction has apparently returned, all patients with documented postoperative block should remain on back-up ventricular pacing (VVI at 70–80 bpm) until normal AV node function has been confirmed by an electrophysiologist.

RESPIRATORY SYSTEM

The lungs may be directly affected during cardiac surgery by exposure to cardiopulmonary bypass, handling, retraction or as a result of indirect factors, such as cardiac failure, sepsis and blood transfusion. The need for ventilatory support following surgery varies. Respiratory drive is often suppressed in the early postoperative period by the residual effects of anaesthetic sedative, analgesic and muscle-relaxant drugs. After simple open or closed heart procedures extubation is possible at the end of surgery, and the only respiratory support needed is supplemental oxygen. In contrast, some sick patients, especially neonates undergoing complex surgery, will require full respiratory support for several days until haemodynamic stability is achieved and then ventilatory support is reduced more gradually.

Where postoperative ventilation is anticipated, nasotracheal intubation for surgery is preferred, as it provides greater comfort and security than oral tube fixation. The tip of the endotracheal tube must lie in the midtracheal position and must be confirmed on admission to the ICU and regularly thereafter. Inspired gases must be humidified and the tracheal tube aspirated periodically to prevent atelectasis or tube occlusion. In children, there is a danger of late subglottic stenosis if endotracheal tubes exert pressure on the cricoid ring. When uncuffed endotracheal tubes are used in infants and young children, the presence of a small leak audible at an airway pressure of 20–30 cmH_2O is an indication that the tube is of correct diameter to provide ventilation whilst preventing excessive pressure on the respiratory epithelium of the subglottic area.

Oxygen Therapy

Most children require supplemental oxygen in the postoperative period, whether ventilated or recently extubated. It is essential that the intensivist understands the child's circulation, the level of systemic oxygen saturation expected, and seeks to diagnose the cause of a lower or higher than expected SaO_2. Oxygen is a pulmonary vasodilator and its use in the presence of L–R shunts will increase pulmonary blood flow, potentially compromising systemic circulation. Mild to moderate degrees of ventilation perfusion mismatch commonly occur in postoperative patients, particularly those who remain ventilated. Augmenting inspired oxygen concentrations to 30–40%, together with lung recruitment ventilatory strategies (increased positive end expiratory pressure), are usually all that is required to manage the pulmonary contribution to hypoxaemia, although higher oxygen concentrations may be required in severe lung injury.

Positive Pressure Ventilation

Positive pressure ventilation via an endotracheal tube is the most commonly used ventilatory strategy in the immediate postoperative period (invasive ventilation). There have been many advances in ventilator technology over recent years. The current generation of intensive care ventilators have much improved monitoring technology, more flexible ventilatory and weaning modes, superior ventilator–user interfaces and may be configured to all age groups. Despite this, the skill of the user is a far more important factor in delivering high quality respiratory care than the availability of the latest technology. There is little evidence that sophisticated ventilators or particular weaning modes substantially impact postoperative recovery.

Postoperative Ventilation

Initial ventilator settings in postoperative patients are a matter of clinical judgement. Initial peak inspiratory and expiratory pressures of 20 and 5 cmH_2O to achieve tidal volumes of 6–8 ml/kg and a respiratory rate appropriate to age (neonate, 20–25 breaths/minute (bpm); infant, 15–20 bpm; and child, 12–15 bpm) are suitable parameters. An initial Fraction of inspired oxygen (FiO_2) of 1.0 may be chosen, this being rapidly titrated down to achieve the desired PaO_2, which in the absence of R–L shunting would be 12–15 kPa (85–115 mmHg). End-tidal CO_2 monitoring, if provided, will give an early indication of ventilatory adequacy, and oxygen saturation (SaO_2) monitoring will give beat-to-beat information of arterial oxygen saturation. The results of an arterial blood gas taken 10 minutes after connecting the ventilator should be used to fine-tune ventilator settings.

Occasionally severe pulmonary oedema secondary to cardiac failure, acute respiratory distress syndrome (ARDS) or severe pulmonary infection require more aggressive ventilatory strategies. Provided they are tolerated haemodynamically, higher end expiratory pressures increased gradually up to a maximum of 10–12 cmH_2O can be used maintain alveoli recruitment. So-called permissive ventilatory strategies, particularly allowing the $PaCO_2$ to rise provided that pH is 7.20 or above, have been shown to be associated with better outcomes in ARDS than using higher pressures to achieve normocapnia (Sevransky *et al*., 2004). Although this data arises from adult studies, the concept of treating the lung gently in the hope of avoiding secondary damage induced by high ventilatory pressures and volumes is logical. If there is severe loss of lung volume which has not been successfully managed with conventional measures, the use of high-frequency oscillatory ventilation (HFOV) should be considered. This mode of ventilation is highly effective in those patients with significant interstitial lung disease in whom oxygenation and CO_2 removal are difficult to achieve with a conventional ventilator despite high ventilatory pressures.

Non-invasive Positive Pressure Support

Soft-seal face masks or nasal cannulae or masks can be used to successfully deliver either a continuous distending pressure (continuous positive airway pressure, CPAP) or tidal ventilation (bilevel positive pressure ventilation). Nasal CPAP is easily delivered to neonates and small infants via nasal cannulae and may be started electively at the moment of tracheal extubation in infants believed to be at high risk of reduced lung volume and developing atelectasis following extubation. Older patients can similarly be supported with CPAP using a face mask, and in addition can receive tidal ventilation non-invasively if required. This may prevent intubation or reintubation in children exhibiting increasing signs of respiratory failure. Non-invasive CPAP or ventilation is particularly useful in patients who develop pulmonary oedema or are compromised by developing atelectasis. Non-invasive support given whilst dealing with the causes of respiratory insufficiency may well lead to success without resort to invasive ventilation.

Weaning and Extubation

Weaning from mechanical ventilation may be ''routine'', usually within hours of ICU admission, once haemodynamic stability is achieved and major bleeding subsided, and is typically rapid and uneventful. It is essential to assess levels of sedation prior to extubation, as child must be sufficiently alert to protect their airway and cough to clear secretions. If stridor occurs following extubation, nebulized adrenaline 1:1000 may act to vasoconstrict engorged mucosa and improve the situation, and dexamethasone 0.25 mg/kg i.v. may be given in addition. ''Complex'' weaning, on the other hand, describes the process of reducing ventilatory support in a child after a prolonged period of ventilation or the situation where initial attempted weaning has failed. In children following a difficult or protracted intensive care course where difficulty in weaning is anticipated, or those who have failed a previous attempt to wean, a number of factors should be addressed. Reduced myocardial dysfunction and residual or undiagnosed cardiac defects should be excluded and cardiorespiratory function optimized. Poor nutritional status alone is sufficient cause for weaning failure, particularly in very small infants and neonates. In such circumstances, it may be necessary to delay weaning while nutrition is optimized and a weight

gain target is achieved. Extrinsic compression of the trachea or bronchi (including tracheobronchomalacia) may be diagnosed by flexible bronchoscopy and/or bronchography. A bronchogram is particularly useful in demonstrating the extent of tracheobronchomalacia, which may require intraluminal stenting or long-term CPAP support via a tracheostomy. Unilateral phrenic nerve palsy is poorly tolerated in neonates and infants in whom intercostal inspiratory mechanisms are inefficient. Clinically, the abdominal contents visibly slew to the paralysed side during spontaneous inspiration. Diaphragmatic paralysis can be confirmed by screening the diaphragm using ultrasound or video fluoroscopy or, if available, transcutaneous phrenic nerve electromyography. Although the phrenic nerve may recover slowly if it has not been divided, surgical placation of the diaphragm greatly increases efficiency of the contralateral hemi-diaphragm and usually leads to successful weaning. Bilateral diaphragmatic paralysis is an uncommon and challenging problem, and although bilateral placation is probably beneficial, weaning thereafter is more problematic.

Ventilation in Special Situations

Parallel Circulations

In any children where the systemic and pulmonary circulations are in parallel, run-off can occur into the lower resistance pulmonary vascular bed, diverting flow away from the systemic circulation and resulting in a low systemic cardiac output. This may occur preoperatively in the newborn period through the ductus arteriosus or following surgery if the two circulations are connected by a systemic–pulmonary artery shunt. A similar pathophysiological situation occurs in the presence of a large VSD where a low PVR will result in increased pulmonary blood flow and congestive cardiac failure. The intensivist can assist in balancing pulmonary and systemic blood flow by avoiding factors which lower PVR (high inspired oxygen fraction, inducing alkalosis, inhaled nitric oxide or other specific pulmonary vasodilators), actively dilating the systemic circulation (phenoxybenzamine, sodium nitroprusside), and considering lowering pH (hypoventilation, adding CO_2 to inspired gas) or reducing further inspired oxygen fraction (adding nitrogen to inspired mixture). The alteration of gas inspired mixtures were frequently recommended in the perioperative management of Norwood Stage 1 operations for HLHS (see Chapter 41), but in the current era, the use of smaller diameter systemic–pulmonary artery shunts and the advent of RV–PA conduits have reduced the need for these complex measures in the management of such babies.

Ventilatory Manipulation of Cardiac Function

Right ventricular diastolic dysfunction immediately following repair of tetralogy of Fallot is manifested by a low cardiac output state as right ventricular filling is reduced. This ''restrictive physiology'' can be diagnosed echocardiographically by the presence of antergograde flow in the pulmonary artery during atrial systole, suggesting that the right ventricle acts as a stiff conduit rather than a compliant chamber. Pulmonary blood flow, and hence cardiac output, is therefore augmented by measures which increase systemic venous return (spontaneous negative pressure ventilation) and hindered by a further reduction in RV filling (positive pressure ventilation). The adverse effects of RV restriction can therefore be best managed by minimizing mean intrathoracic pressure during positive-pressure ventilation, encouraging spontaneous modes of ventilation and, if feasible, using negative-pressure respiratory support to increase cardiac output (Shekerdemian *et al*., 2000). Similar considerations apply to the Fontan circulation, in which pulmonary blood flow and hence systemic ventricular filling are secured by passive return of systemic venous blood across a low-resistance pulmonary vascular bed to reach the pulmonary venous atrium. Excessive intrathoracic pressure induced by injudicious use of positive-pressure ventilation will reduce cardiac output. Conversely, spontaneous or negative-pressure modes increase cardiac output by augmenting systemic venous return. Care must be taken by the intensivist in responding to pleas for ''early extubation'' in these situations. In our view, we should seek to extubate all of our patients as soon as they fulfil extubation criteria. Unthinking adherence to an over-aggressive policy of ''early'' extubation of the Fontan or restrictive Fallot child risks compromising pulmonary blood flow and cardiac output if PVR rises as a result of respiratory acidosis or hypoxaemia due to excessive sedation or atelectasis.

Analgesia and Sedation

All patients following surgery require effective analgesia and sedation, which must be optimized and titrated by continued assessment and re-evaluation. Analgesia also has beneficial effects in modifying reflex, endocrine metabolic and immune-mediated responses, which may be harmful to the patient. High-dose opioid analgesia during surgery is associated with decreased stress response and fewer postoperative complications and decreased mortality (Anand *et al*., 1987; Anand and Hickey, 1992), and has been shown to be effective in the prevention and treatment of episodic pulmonary hypertension in the at risk infant. The use of intravenous opioids in non-surgical, mechanically

ventilated neonates results in reduced physiological and behavioural measurements of pain and stress and is associated with fewer periods of hypoxaemia and improved neurological outcomes (Orsini *et al*., 1996; Guinsburg *et al*., 1998). However a balance of analgesia and sedation must be maintained which allows satisfactory pain control and alleviates distress, whilst at the same time promoting weaning of ventilation and extubation. Pharmacological analgesia and sedation should be supplemented with positive environmental factors, including lack of noise, parental presence, warmth, nutrition and friendly distraction to promote comfort for the ICU patient. These strategies are inexpensive and are without the risks associated with sedatives and analgesics, which include prolonged sedation and withdrawal phenomena.

Analgesic drugs commonly used in PICU include opioids (morphine and fentanyl), non-steroidal antiinflammatory drugs (ibuprofen, diclofenac and ketoralac) and the α-2 adrenergic agonists (clonidine and dexmedetomidine). *Sedative* drugs used include benzodiazepines (midazolam, lorazepam), phenothiazine-related drugs (trimeprazine, chlorpromazine, levomepromazine) and chloral hydrate-related preparations. In addition, the general *anaesthetic* drugs, including volatile agents (sevoflurane, isoflurane), propofol and barbiturates (thiopentone), may be used for specific indications or procedural anaesthesia.

Intravenous morphine is recommended as the first-line analgesic in the early postoperative period, administered as an intravenous infusion with additional intermittent bolus doses titrated to effect. The pharmacokinetics of morphine are highly age-dependent, requiring age-sensitive dosing regimes, especially in the neonatal period where opioid related respiratory depression may be profound. Fentanyl, a potent opioid, is effective in the prevention and treatment of pulmonary hypertensive crises in infants (Hickey *et al*., 1985). Due to its high lipid solubility, fentanyl has a more rapid onset of action than morphine; however, after frequent dosing or long-term infusions clinical recovery can be prolonged due to its redistribution from peripheral stores. Alfentanil is highly protein-bound and therefore has a smaller volume of distribution than fentanyl, and is also less fat-soluble, which results in a shorter context-sensitive half-life. Its rapid onset of action makes it particularly useful for providing short intense periods of analgesia. Remifentanyl is unique among opioids in that it is not metabolized by liver and kidney, but by plasma and tissue cholinesterases. Its place in the postoperative management of children undergoing cardiac surgery has yet to be fully explored, but it may become the opioid of choice in children with postoperative renal or hepatic dysfunction. Although the sedative effects of opiods are beneficial, it is common to use a benzodiazepine such as midazolam in addition, again titrated to effect. The potential cardiovascular depressant effects especially of the benzodiazepines should always be considered. Paracetamol and non-steroidal antiinflammatory agents may be administered by the oral or rectal routes, which reduce opioid requirements and facilitate weaning from opioids once acute pain is subsiding. Central α 2-agonists have significant analgesic and sedative properties and may facilitate weaning of benzodiazepines and opioids and in the treatment of drug withdrawal syndromes occurring during recovery from critical illness. The intravenous anaesthetic agent propofol has predictable pharmacokinetics and an excellent rapid recovery profile. It is extremely useful when used in combination with an infusion of morphine to provide a short period of postoperative anaesthesia for patients who will extubate quickly, e.g. within 6 hours (Cray *et al*., 2001). However, propofol should not be used for prolonged periods as a sedative in PICU patients because of the risk of propofol infusion syndrome (Bray, 1998). We limit its use to a maximum of 6 hours at a maximum infusion rate of 4 mg/kg/hour.

Indications for the use of muscle relaxants in postoperative intensive care are few, but continuous muscle paralysis may be beneficial in children with very unstable circulations, including reactive pulmonary hypertension, those in whom spontaneous respiration must be suppressed to lower pH or PaO_2, in patients with severe lung disease who cannot synchronize with the ventilator despite sedation/analgesia, and patients who require systemic cooling. Some units consider children with open chests to be candidates for continuous paralysis, whilst others manage such patients with analgesia–sedation only. Analgesia and sedation must be confirmed prior to the institution of muscle relaxation, and doses must be adjusted based on continuous or intermittent measurement of neuromuscular blockade or by implementing daily ''relaxant holidays,'' during which the relaxant infusion is temporarily discontinued.

GASTROINTESTINAL SYSTEM

Nutrition

The goals of metabolic and nutritional support in the paediatric cardiac ICU are first, to minimize the deleterious effects of the hypermetabolism and catabolism induced by the 'stress' of major surgery or critical illness, and second, to promote anabolism and growth (Irving *et al*., 2000). Assessment of a child's nutritional needs should be undertaken on ICU admission and a feeding plan established. Feeds should commence as soon as the child's haemodynamic status permits, with preference given to enteral feeding. Caloric requirements for

Table 13.7 Recommended daily intake (RDI) of calories.

Age (years)	Weight (kg)	Caloric requirement (kcal/kg)
Preterm		130–150
< 1	3–10	90–120
> 1–6	11–20	75–90
7–12	21–40	60–75
12–18	40–70	25–30

critically ill children are usually calculated from standard formulae. Pathological conditions, such as cardiac failure, increase the work of breathing and hence energy requirements; infants with chronic heart failure may require caloric intakes of 140–160 kcal/kg/day. Many babies have not achieved such intakes preoperatively and are therefore nutritionally impaired when admitted to the ICU following surgery. Allowance must therefore be made in planning postoperative feeding, to provide sufficient calories for ongoing needs and ''catch-up'' requirement (**Table 13.7**).

Enteral Feeding

Enteral feeding is absolutely contraindicated in cases of proved or suspected necrotizing enterocolitis (NEC) and for 24–48 hours after gastrointestinal surgery. Feeding is relatively contraindicated in infants and children with severe low cardiac output or reduced regional blood flow (e.g. aortic coarctation), due to low splanchnic perfusion and consequent risk of developing NEC. Unless a contraindication exists, enteral feeds should start within 12 hours of PICU admission.

Parenteral Nutrition

Parenteral nutrition (PN) should only be used if enteral feeding is absolutely contraindicated or in situations in which tolerance of enteral feeds cannot be maximised and fails to meet caloric requirements. Parenteral nutrition is more expensive, more complex to deliver, requires central venous access and it is associated with complications including catheter-related thrombosis and infection and specific physiological derangements including cholestasis, gut mucosal atrophy and induced biochemical abnormalities (Heine and Bines, 2002). In critically ill children, parenteral nutrition should be prescribed based on standard paediatric regimes modified to take into account the particular metabolic circumstances of the child (**Table 13.8**). Frequent monitoring of biochemical parameters is

Table 13.8 Modifications to caloric requirements in postoperative and critically ill children.

Age	RDI(%)	Comment
RDI	100	Recommended intakes for growth in healthy children undertaking *normal life activities*
Basal	55	Deep sedation, ebb phase injury, full mechanical ventilation
Maintenance	66	Mechanical ventilation, enteral feeds, lying quietly
Minor stress	76	Minor surgery, peritonitis, fever < 39°C
Major stress	98	Sepsis, major cardiac surgery

RDI, recommended daily intake.

essential. Disturbances of sodium, potassium, calcium, phosphate and other electrolytes as well as hyper- or hypoglycaemia are relatively commonly seen in children and especially in neonates receiving PN. Lipid tolerance may be impaired in metabolically stressed children and should be introduced incrementally with monitoring of serum triglyceride concentrations, aiming for plasma triglyceride levels of less than 1.5 mmol/l.

Central venous access for PN is a frequent source of complications, of which entry site infection, line-related blood stream infection and the occurrence of central venous thrombosis are prominent. The catheter entry site should be observed daily and kept clean and dry and strict aseptic techniques used when accessing lines. Infection of the line entry site is an indication for line removal and resiting. A 7–10 day course of antibiotics is required to treat established entry site infection or line-related blood stream sepsis. Fungal septicaemia is usually associated with indwelling lines.

Consistent weight gain is the best acute guide to the anabolic success of any feeding regime. Weight, length and head circumference referenced to appropriate centile charts provide longer-term benchmarks.

Gastroduodenal Paralysis

Often gastric emptying is impaired in children recovering from surgery and critical illness. Large residual feed volumes are present in the stomach. Management includes temporary reduction in feed volumes, introducing small volume frequent feeds or continuous

feeds, introducing a gastric promotilant drug, such as *erythromycin* or *domperidone*, or bypassing the pylorus by siting a nasojejuenal feeding tube.

Stress Ulceration

The incidence of stress-related bleeding is very low in the children and, given the lack of evidence of effect from adult studies and some evidence of possible harm, the routine use of stress ulcer prophylaxis cannot be recommended for children undergoing intensive care. However, prophylaxis with a proton pump inhibitor (Kearns and Winter, 2003) or H_2 receptor antagonist should be considered in known ''at-risk'' groups, such as critically ill children in whom anticoagulants are being administered, those receiving systemic steroid therapy and those with a history of upper gastrointestinal haemorrhage. Upper gastrointestinal bleeding is managed by volume replacement and supplementary measures, including discontinuing feeds, continuous gastric tube drainage or suppression of acidity with an H_2 receptor antagonist (ranitidine) or a proton pump inhibitor (omeprazole). If haemorrhage is not controlled, an urgent referral should be made to a paediatric gastroenterologist and/or surgeon.

Chylothorax

The most common cause of chylous pleural effusion is lymphatic hypertension arising from high venous pressure or occlusive thrombus in the superior vena cava (Sharoni *et al*., 2001). In addition, iatrogenic damage to the thoracic duct during surgery or during central venous cannulation occasionally occurs. Chyle may also collect in the pericardium, causing pericardial tamponade (Campbell *et al*., 2001). Chyle is a mixture of lymph and triglycerides derived from abdominal lymphatics with the addition of lymphocytes. In enterally fed patients, high levels of triglycerides (pleural fluid:plasma ratio > 1.0) are present in the effusate. Diagnosis of chylous effusion is confirmed by a differential leukocyte count showing $> 80\%$ lymphocytes.

Chylothorax can compromise immunity and nutritional status through uncompensated losses of lymphocytes, protein and fat. Initial management of chylothorax consists of draining the pleural fluid and dietary substitution of long-chain triglycerides with medium-chain triglycerides (Cormack *et al*., 2004), or complete replacement of enteral feeds with parenteral nutrition, completely abolishing the lipid load of the lymphatic system. If the thoracic duct has been damaged surgery may be required (Merrigan *et al*., 1997). Although surgical or chemical pleurodesis has its advocates in cases of diffuse chylous leakage, addressing any identifiable causes of the chylous effusion, such as venous thrombus or hypertension, are likely to be most effective.

Necrotizing Enterocolitis

Necrotizing enterocolitis (NEC) is the most common acquired intra-abdominal emergency in newborn infants and is associated with significant mortality (Lee and Polin, 2003). In congenital heart disease NEC is believed to multifactorial, relating to intestinal ischaemia, colonization of the gut by pathogenic bacteria, and excess protein substrate in the intestinal lumen, leading to ischaemic necrosis. Congenital heart disease in itself is a risk factor for the development of NEC, with truncus arteriosus and hypoplastic left heart syndrome demonstrated to be especially susceptible (McElhinney *et al*., 2000). Lesions where pulmonary blood flow is dependent on a systemic–pulmonary shunt have absent or reversed splanchnic blood flow, both at rest and after feeding, which may explain the particular predisposition of these infants (Cheung *et al*., 2003).

NEC may occur pre- or postoperatively, presenting with non-specific findings such as temperature instability, lethargy, apnoea, feed intolerance to frank symptoms of abdominal distension, bilious vomiting and bloody stools and ultimately peritonitis and cardiovascular collapse. Laboratory findings are those of sepsis and inflammation, including neutrophilia or absolute neutropaenia, coagulopathy, raised acute phase proteins and positive blood or peritoneal fluid cultures. On suspicion of NEC, enteral feeds should be withheld, nasogastric drainage commenced and ''enteral'' antibiotic cover, typically penicillin, gentamicin and metronidazole, initiated for a minimum period of 10 days, during which time parenteral nutrition may be necessary. Laparotomy is necessary if perforation or severely ischaemic bowel is suspected (Pierro and Hall, 2003).

RENAL SYSTEM AND FLUID THERAPY

Postoperative Fluid Management

Normal parenteral fluid requirements in children can be estimated according to patient's weight (**Table 13.9**). Fluid intake, which should include fluids from all sources, is restricted to 50% of these requirements in the immediate postoperative period to take account of intraoperative fluid gain (see above) and the stress-induced antidiuretic state. Parenteral fluids and those used for drug infusions are commonly 0.9% sodium chloride and/or 5% dextrose, adjusted according to the specific metabolic and electrolyte needs, which should be analysed regularly. Accurate measurement of blood and fluid losses must be charted postoperatively.

Table 13.9 Intravenous fluid requirements.

Body weight (kg)	Daily intravenous fluid requirement
< 10	100 ml/kg
10–20	1000 ml + 50 ml for each kg over 10 kg
> 20	1500 ml + 20 ml for each kg over 20 kg

Most children have normal or moderately raised blood glucose levels with this fluid regime. By convention, moderate hyperglycaemia, which usually resolves spontaneously within 12 hours of surgery, is not treated aggressively, although insulin infusions should be used (0.05–0.1 units/kg/hour) if blood glucose levels persist at > 12–14 mmol/l. Recent studies in adult patients have demonstrated an improved outcome with tighter glycaemic control (Van den Berghe, 2001). Additional glucose administration may be required in the sick neonate to maintain glucose above a safe level of 2.5–3 mmol/l. This may be achieved by permitting higher fluid infusion rates or increasing the concentration of glucose infused, typically by using a 10% glucose solution.

Subsequent fluid intake should be adjusted according to clinical, chest X-ray and laboratory evidence of the child's hydration status. In the absence of pulmonary or systemic oedema, fluid intake may be increased over 2–3 days to achieve normal daily maintenance, often with a transition from enteral to parenteral regimens as soon as possible. Increases in fluid intake should be mainly permitted to accommodate increases in nutritional intake, with the judicious use of diuretics increasing urinary output and preventing excessive fluid retention.

Electrolytes

Potassium

Plasma potassium levels may change rapidly in children following cardiac surgery in response to factors such as changes in cardiac output, renal perfusion, urine output and perioperative diuretic administration. Whilst extreme hypokalaemia ($K^+ < 2.0$ mmol/l) is associated with risk of serious arrhythmias, mild hypokalaemia (2.5–3.5 mmol/l) is usually well tolerated; 2–4 mmol/kg/day added to the intravenous maintenance fluid solution or titrated as a separate potassium infusion is required to maintain plasma levels in the range 3.5–4.5 mmol/l. Plasma potassium levels must be checked hourly during potassium infusions to avoid unanticipated hyperkalaemia ($K^+ > 5.5$ mmol/l), which is associated with a high risk of haemodynamically significant ventricular arrhythmias (for treatment of hyperkalaemia, see renal section, below).

Calcium

The normal requirement for calcium is 1.5 mmol/kg/day. In the immediate postoperative period, ionized calcium should be measured and maintained in the normal range (approximately 1–1.25 mmol/l). Hypocalcaemia manifested as tetany, seizures and myocardial dysfunction may be seen in neonates, those with hypoparathyroidism and children receiving high doses of diuretics or rapid transfusion of citrated blood. This can be successfully treated with a slow bolus of calcium. Patients at particular risk of hypocalcaemia are those with 22q11 deletion syndrome (variously termed DiGeorge, velocardiofacial or Sphrintzen syndrome) who display a constellation of features including a characteristic heart defect (interrupted aortic arch, truncus arteriosus, tetralogy of Fallot), hypocalcaemia secondary to hypoparathyroidism and immune deficiency as a result of thymic hypoplasia (Perez and Sullivan, 2002).

Magnesium

Hypomagnesaemia occurs as a result of urinary losses induced by diuretics or impaired intake in association with parenteral nutrition. Clinical problems are unlikely until depletion is severe with plasma concentrations of < 0.6 mmol/l. Usual daily requirement are 0.5 mmol/kg/day.

Capillary Leak Syndrome

Capillary leak syndrome is a transient phenomenon in the early postoperative period, manifested as severe systemic oedema secondary to fluid leak from the intravascular space into the extracellular tissues. It is associated with profound systemic inflammatory response to surgery and to cardiopulmonary bypass, and probably results from endothelial injury and subsequent increased permeability. Risk factors for include young age, long periods of cardiopulmonary bypass, perioperative low cardiac output, and the presence of preoperative inflammation. Despite the obvious presence of tissue oedema, patients are often intravascularly deplete and continue to require fluid resuscitation. The magnitude of capillary leak may be favourably influenced by the use of steroids preoperatively, low prime volume CPB circuits and the use of modified ultrafiltration following cardiopulmonary bypass (Gaynor, 2003). Small-volume peritoneal dialysis to remove inflammatory cytokines has also been proposed (Bokesch *et al*., 2000).

Perioperative Renal Dysfunction

By far the most common renal problem seen in children with heart disease is acute renal failure, the onset of

which is usually the result of an episode of low cardiac output or cardiac surgery (Mentser and Bunchman, 1998). Children may have pre-existing renal anomalies unrelated to their cardiac condition, often associated with genetic syndromes.

Acute Renal Failure (ARF)

The occurrence of ARF following cardiac surgery, which is associated with increased mortality, usually occurs as part of the spectrum of multi organ system failure (Giuffre *et al*., 1992; Filler, 2001). Clinical risk factors for the development of perioperative renal failure include low cardiac output, hypoxaemia, hypotension, congestive heart failure and the use of vasopressors. Other factors such as sepsis, disseminated intravascular coagulation and the use of nephrotoxic drugs, such as aminoglycosides, frusemide and amphotericin, may also be relevant. Acute renal failure complicating cardiac disease usually presents with oliguria (urine output of $< 300\ \text{ml/m}^2\text{/day}$ or $< 0.5\ \text{ml/kg/h}$), and is confirmed by rising serum urea and creatinine. Clinically the differentiation of the causes of renal failure into prerenal, renal and postrenal is important. Children with oliguria and ARF due to prerenal causes tend to have high urinary osmolality and low urinary sodium excretion, a homeostatic response to retain water and sodium to augment circulating volume and improve renal blood flow. In intrinsic renal failure renal cellular function is impaired and urine of low osmolality with high sodium concentrations.

Management of Acute Renal Failure

The aim is early diagnosis and treatment of reversible causes while supporting renal function. Prerenal causes, such as hypotension and intravascular volume depletion, should be urgently corrected. Any potentially nephrotoxic drugs should be withheld and reintroduced only if absolutely necessary.

Frusemide

There is no evidence that frusemide reliably prevents the onset of acute renal failure following cardiac surgery. Frusemide may convert oliguric renal failure to non-oliguric renal failure, making fluid management simpler and preventing the need for renal replacement therapy. In severe oliguria, bolus doses of up to 5 mg/kg may elicit a diuretic response but if little or no urine flow occurs, further administration is probably futile until kidney recovery occurs. Both in ARF and when used to achieve general diuresis, continuous intravenous infusion of frusemide (0.1–0.3 mg/kg/hour) reduces haemodynamic instability, providing a more controlled and predictable urine output.

Mannitol

Mannitol has no place in the therapy of established oliguric renal failure, although it may slow its onset if given early.

Dopamine

Although low-dose dopamine infusions have long been accepted to afford a degree of renal protection, recent studies in high-risk adult patients have failed to show that dopamine provides any benefit, has potential widespread adverse effects and therefore should not be used either as prophylaxis or therapy for ARF (Kellum JA and M Decker J, 2001).

Hyperkalaemia

Hyperkalemia is a recognized complication of ARF, and plasma levels should be monitored closely in patients with renal dysfunction. If hyperkalaemia occurs, urgent steps must be taken to stop potassium administration and lower plasma potassium to safe levels. Intravenous calcium chloride counteracts the physiological effects of hyperkalaemia, while sodium intravenous bicarbonate, nebulized salbutamol and insulin/glucose infusions (0.1 U/kg insulin i.v. and 2 ml/kg 50% glucose) all act to lower plasma potassium by promoting cellular uptake. Urgent haemodialysis or haemofiltration should be considered if the above measures are unsuccessful.

Renal Replacement Therapies

In ARF renal replacement therapy (RRT) may become necessary to remove endogenous and exogenous toxins and to maintain fluid, electrolyte and acid–base balance until renal function returns. RRT may be provided by peritoneal dialysis or haemofiltration. In neonates and infants, the choice of RRT modality for acute dialysis depends more on the availability of vascular or peritoneal access and familiarity of ICU personnel with the particular technique than with the proven superiority of either method. Peritoneal dialysis is a relatively simple technique which is particularly useful in the postoperative neonate or infant with acute renal failure, and prevents the complications of vascular access with large calibre catheters and extra-corporeal renal support (Reznik *et al*., 1991). A detailed description of haemofiltration/haemodialysis is beyond the scope of this chapter and readers are referred elsewhere for information (Bunchman, 2002).

SEPSIS

Hospital-acquired infections are a major cause of morbidity and mortality in children undergoing cardiac

Table 13.10 Suggested first-line antibiotic therapy for suspected infections in the postoperative cardiac child.

Infection site	Common causative organisms	Empirical antibiotic regime
Wound infection	*Staphylococcus*, Coliforms	Teicoplanin or vancomycin + gentamicin
Bacteraemia	Staphylococci (especially *S. epidermidis*), Coliforms	Teicoplanin or vancomycin + gentamicin
Pneumonia (acquired in ICU)	*Streptococcus pneumoniae, Haemophilus influenzae*, Coliforms including *Pseudomonas, Staphylococcus*	Piperacillin/tazobactam + gentamicin
Urinary tract infection	Coliforms	Cefuroxime + gentamicin

Therapy must be reviewed in the light of microbiological cultures, antibiotic sensitivities and clinical response of the child.

surgery. Risk factors include the presence of wounds, drains, lines, infusion of fluids and drugs and prolonged ICU stay. Preoperative poor nutritional status, pre-existing immunocompromise and neonatal age also confer added risk. The most common causes of postoperative infections include bacteraemia, wound infection, respiratory tract infections and urinary tract infections. **Table 13.10** lists the organisms most commonly associated with sites of infection and reasonable ''first-line'' antibiotics to be used when treatment is required. Antibiotic therapy is best guided by agreed local antibiotic policies administered jointly by intensivists, microbiologist and pharmacist. Antibiotic treatment should be started only when clear indications exist and must always be reviewed in the light of culture results and antibiotic sensitivity testing.

Prevention of infection is a huge issue in any intensive care unit. Adequate staffing and good intensive care unit design play some part in minimizing cross-infection risk. However, strict adherence to infection control measures, such as hand washing before and after every patient contact, however minor, and strict application of aseptic techniques during ICU-related interventional procedures and in the care of vascular catheters (Hodge and Puntis, 2002; Polderman and Girbes, 2002), is of the utmost importance.

Patients known to be colonized with antibiotic-resistant organisms, such as methicillin-resistant *Staphylococcus aureus* (MRSA), and those with infective secretions of fluids, e.g. infective diarrhoea, respiratory viral infections, should be isolated. Despite these measures, patients' skin, mouth and bowel flora provide a reservoir from which infection may arise. Prophylactic antibiotics are highly effective in reducing wound infections. Ideally, tissue concentrations of antibiotics need to reach effective concentrations by the time of skin incision and remain so until wound closure. Although some surgeons recommend continuing antibiotics until chest drains and/or lines are removed, this practice is not based on good prospective evidence, which only supports single-dose prophylactic regimes. Extended antibiotic use is associated with the generation of antibiotic-resistant organisms. The choice of antibiotic regime must depend to some extent on local patterns of antibiotic resistance in the population served.

Antibiotics for surgical prophylaxis should be targeted at *Staphylococcus aureus* and *Staphylococcus epidermidis* as well as aerobic Gram-negative bacteria. Many centres use a cephalosporin, such as cefazolin or cefuroxime; however, a combination of a penicillinase-resistant penicillin, such as flucloxacillin, in combination with gentamicin provides more effective anti-staphylococcal cover. A combination of teicoplanin or vancomycin and gentamicin is suitable for situations where the risk of infection with MRSA is high, and for those resistant to penicillin.

Children should be constantly observed for the onset of infective complications. Wound and catheter entry sites must be regularly inspected. Pyrexia, high or low leukocyte counts, elevated biochemical markers, such as procalcitonin or high C-reactive protein, may give an early indication of evolving sepsis and suggest the need for screening cultures and detailed clinical assessment.

NEUROLOGICAL DYSFUNCTION

Both acute and chronic neurological morbidity are significant concerns for children with congenital heart disease. Neurological impairment may be a preadmission feature of the child's overall condition or may result from an acute perioperative insult. Neurological injury may be caused by reduced cerebral perfusion during periods of low cardiac output, chronic hypoxia or thromboembolism (Fallon *et al*., 1995;

Bellinger *et al*., 1999; Mahle *et al*., 2002). Peripheral neurological injury to the recurrent laryngeal and phrenic nerve are also recognized complications of cardiac surgery.

Common acute manifestations of neurological damage associated with cardiac surgery and intensive care include alterations of consciousness, seizures, hemiparesis, choreo-athetoid movements, neuro-ophthalmic deficits, global hypoxic–ischaemic encephalopathy and intracranial bleeds. Although the incidence of acute neurological complications is low (Menache *et al*., 2002), their impact on both the child and family is substantial (Mahle *et al*., 2000).

Maintenance of adequate cerebral blood flow throughout the perioperative period, especially by optimizing cardiac output, is a key factor in the prevention of neurological morbidity. Modified ultrafiltration may improve cerebral recovery (Skaryak *et al*., 1995). Rapid increases in central temperature following CPB are associated with increased oxygen consumption, which should be treated by proactive and aggressive cooling strategies. Major metabolic disturbances of glucose and electrolyte balance may injure the central nervous system or provoke seizure activity. Sedative, analgesic and anticonvulsant drugs have the potential to accumulate if metabolism or excretion is impaired or inhibited by factors such as renal failure or low cardiac output, resulting in drug-induced narcosis and a range of withdrawal syndromes.

A structured approach must be adopted to the management and investigation of acute postoperative central neurological dysfunction (Tasker, 2000). Therapy is mainly supportive, with an emphasis on the maintenance of adequate cardiac output, cerebral perfusion and oxygen delivery. Seizures should be suppressed with anticonvulsants, and measures taken to prevent aggravation of cerebral oedema through appropriate use of positioning, ventilatory strategies to lower $PaCO_2$ and the administration of analgesia. Manifestations resulting from sedation–analgesia withdrawal range from mild symptoms such as tachycardia, irritability or diarrhoea to hallucinations and major motor or sensory deficits. Withdrawal effects can be reduced by slow staged scaling-down of sedative analgesics if used for > 48 h.

Spinal cord injury may occur during cardiac surgery and anaesthesia, especially in children undergoing repair of aortic coarctation, where spinal cord ischaemia and permanent damage may occur, especially if the cross-clamp time exceeds 30 minutes.

Positive confirmation of the return of normal lower limb motor function is therefore reassuring in the early postoperative period following coarctation repair (for details of spinal cord injury, see Chapter 20).

APPENDIX 1 (WITH J. STARK) TRACHEOSTOMY

Indications for tracheostomy after operation for congenital heart defects have greatly decreased since the early 1980s. Because it is infrequently undertaken, we believe that previous experience updated to reflect current practice is worth including in this edition of the text.

Tracheostomy is normally reserved for children requiring long-term ventilation due to chronic cardiorespiratory failure (Amin and Fitton, 2003) and children with obstructive airway anatomy. Tracheostomy enables easier feeding and stimulation of infants and young children on ventilatory support. Secretions are readily accessible, and weaning of children with chronic respiratory failure from ventilation, if facilitated by the ease with which ventilation can be recommenced. It is particularly important to learn the techniques of tracheostomy when dealing with infants and young children. In our view, the technique suggested and described in detail by (Aberdeen, 1965) is most suitable.

Surgical Technique

Tracheostomy should be performed in the operating room, under general anaesthesia. The child is positioned on the operating table with the neck extended over a plastic wedge (**Figure 13.1**). The anterior portion of the neck, chin and upper chest are prepared and draped. A short transverse incision is made just below the level of the cricoid cartilage. When the neck is extended, the thyroid and cricoid cartilages are easily palpable. The incision in an infant is usually less than 15 mm in length. Subcutaneous tissue is cut with diathermy to avoid bleeding. Two artery forceps grasp the subcutaneous tissue above and below to facilitate dissection (**Figure 13.2**). Sharp dissection with scissors and diathermy opens the fascia and enables visualization of the anterior trachea. It is important to limit dissection to the midline to avoid injury to the recurrent laryngeal nerve and major blood vessels. Dissection is facilitated by placement of two small, self-retaining retractors (**inset, Figure 13.3**). Once the trachea is visualized, tracheal rings are carefully identified. The first two rings should never be incised because of the possibility of subsequent subglottic stenosis. The third and fourth tracheal rings are opened with diathermy. The point of the diathermy must be used to avoid necrosis of the adjacent tissues. Alternatively, a knife is used, and any bleeding from the edges is controlled with diathermy. Cartilage *should never be excised* and a window should never be formed in a child's trachea.

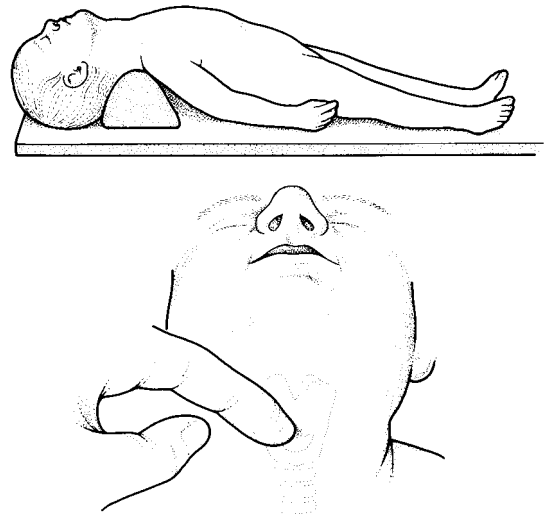

Figure 13.1

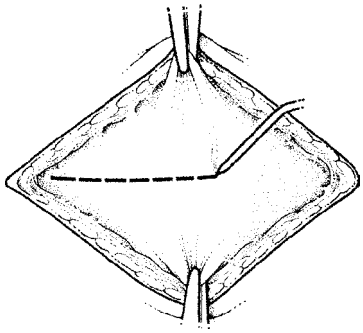

Figure 13.2

Typically, a slightly larger external diameter tracheostomy tube than the preceding endotracheal tube is required. Appropriate catheters for aspiration of secretions and for saline and a trap-sputum bottle should be available. The incision in the trachea is then opened with fine forceps or two stay sutures (**Figure 13.4**). At this point, the anaesthetist withdraws the endotracheal tube to allow the insertion of the tracheostomy tube (**Figures 13.5** and **13.6**). It is important not to remove the endotracheal tube from the larynx because it must be re-inserted immediately if there is any difficulty in placing the tracheostomy tube. Once the tracheostomy tube is safely inserted, it is connected to the gas supply and the child is ventilated. We check the size of the tube for leakages and ensure that subsequent IPPV is possible. It is important that the opening in the trachea

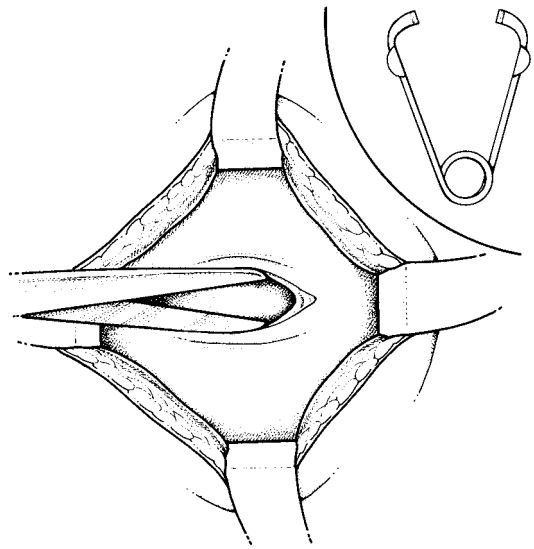

Figure 13.3

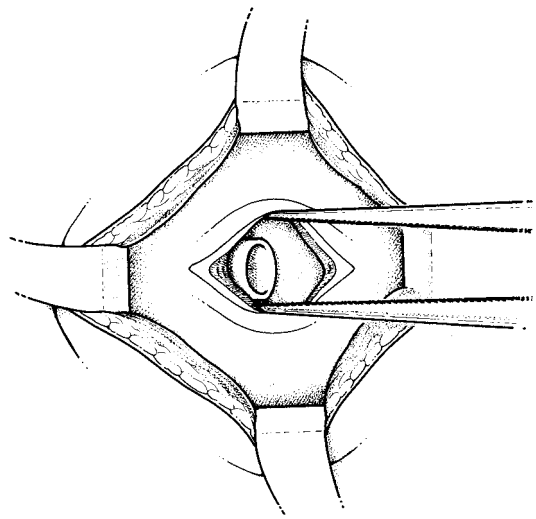

Figure 13.4

is not too long, in order to avoid excessive air leakages and possible dislodgement of the tube. It is equally important that the opening is not too small because the insertion of the tracheostomy tube may be difficult and pressure on the edges of the incision may cause necrosis. Subcutaneous tissue is approximated with a fine Dexon suture. The skin is not closed. We prefer to secure the tube with a piece of tape that is padded with plastic foam

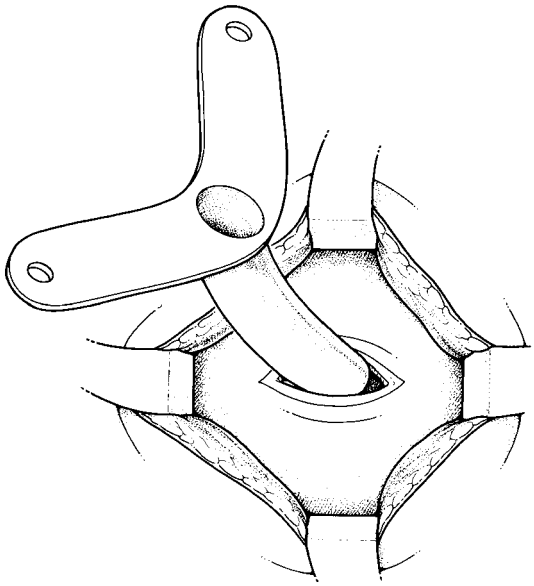

Figure 13.5

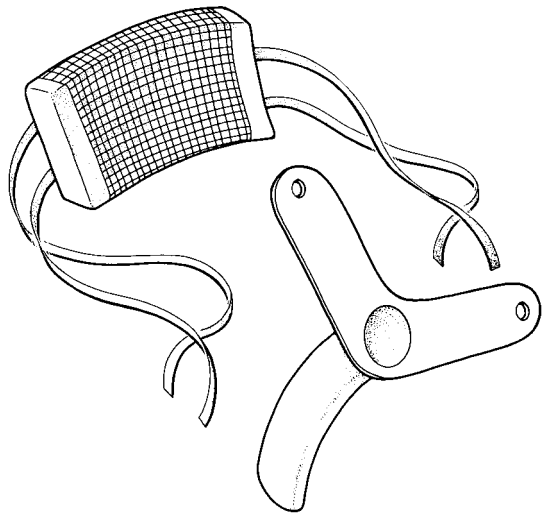

Figure 13.7

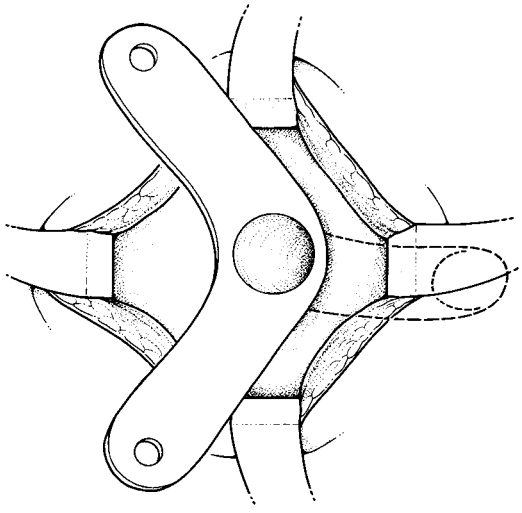

Figure 13.6

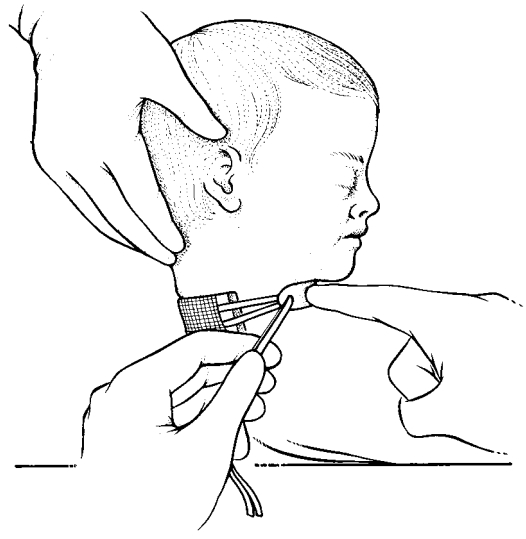

Figure 13.8

to protect the skin at the back of the neck (**Figure 13.7**). It is tied firmly while the neck is flexed. The neck of an infant is short, and if the tube is affixed while the neck is in extension, the ties may become loose when the infant flexes the neck (**Figure 13.8**).

Posttracheostomy Care

Secretions must be aspirated from tracheostomy tubes and gases humidified, as with endotracheal tubes, to prevent tube occlusion and atelectasis. The first tracheostomy tube change should take place after 7 days under medical supervision. This is usually uneventful. A safe technique is to pass a soft flexible bougie through the original tracheostomy tube into the trachea and use this to guide the new tube into place. After an uneventful

initial change of tube, subsequent changes are not usually problematic, as the tract matures rapidly.

Decannulation

This procedure does not usually present any special problems. When a child has been off the ventilator for 2–3 days, we start reducing the size of the tracheostomy tube. The size is reduced daily; this enables gradual closure of the stoma with an increasing flow of air through the larynx. If the tracheostomy tube has been in place for more than 1 month, tracheoscopy should be performed before decannulation. This would visualize any granuloma usually located above the stoma on the anterior tracheal wall. It can be brought into vision during tracheoscopy by a skin hook and excised by diathermy. When a child is ready for extubation, 0.2 mg/kg morphine is used for sedation. The tracheobronchial tree is aspirated and the tube is gently removed. It is preferable to perform extubation in the morning; spare tubes and laryngoscopes should be readily available for the next 24 hours. The stoma usually closes within the next 2–3 days.

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14

Paediatric Cardiac Mechanical Support

A. Karimova and A. Goldman

The history and evolution of paediatric cardiac mechanical support is integral to the history of cardiopulmonary bypass. In the 1960s it was demonstrated that extracorporeal devices could be used to support patients for several days outside the operating theatre. Hill *et al*. (1972) reported the first adult extracoporeal membrane oxygenation (ECMO) survivor in 1972. Bartlett *et al*. (1976) reported the first neonatal ECMO survivor, who was appropriately named ''Esperanza'' (''Hope''). Following this, there was a proliferation of the use of ECMO for a variety of conditions leading to refractory cardiorespiratory failure.

In 1989 over one hundred ECMO centres from around the world established the Extracorporeal Life Support Organization (ELSO) as a platform for communication and research in extracorporeal support. Today, there are over 30 000 patients on the registry with an overall survival to hospital discharge rate of 65% (ECLS Registry, 2005).

By far the largest and most successful single group of patients supported on ECMO are neonates with acute hypoxaemic respiratory failure with an overall survival to hospital discharge rate of 80%. It is also worth noting that this is the only group of ECMO patients who have been subjected to a rigorous randomized controlled trial (Field *et al*., 1996). This conclusively showed a survival and long-term morbidity benefit at 4 years follow-up in babies treated with ECMO (60% survival) compared to those who received conventional treatment (± 30% survival) (Bennett *et al*., 2001).

Currently the fastest-growing group of patients supported on mechanical assist devices are infants and children with intractable cardiac failure. This chapter will specifically focus on this group of patients with ECMO as the main mode of support, although a variety of other mechanical assistance devices available for this population will be discussed.

AIMS OF CARDIAC MECHANICAL SUPPORT

The primary aim of cardiac mechanical support is to restore adequate oxygen delivery to vital organs in situations where all appropriate conventional therapies have failed. The secondary aim is to provide time and the appropriate physiological milieu for myocardial recovery. In this sense, it is vital that the affected ventricle is off-loaded, with reduced ventricular wall stress, reduced myocardial oxygen consumption and adequate coronary perfusion.

In circumstances where there is failure of myocardial recovery over an expected time course of days to weeks, the aim of mechanical support may change from recovery to a bridge to heart transplantation. Mechanical support as a destination therapy is currently not available for children.

CHOICE OF MECHANICAL ASSISTANCE DEVICE

Selection of the appropriate mechanical assistance device is determined by the size of the patient, the indication for support, the pathophysiology of the underlying disease and the predicted length of time for which the support will be needed (bridge to recovery or transplantation) and the availability of different devices in various institutions. Unlike adult practice, where there are a wide variety of paracorporeal and implantable ventricular assistance devices (VADs) to support patients as a destination therapy or as a bridge to recovery and/or transplantation for prolonged periods of months or years, there is a paucity of devices in the paediatric population, particularly for neonates and infants.

Currently the two most common modes of short-term cardiac support used in infants and children are ECMO

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Table 14.1 Comparison between ECMO and extracorporeal centrifugal VAD.

	ECMO	VAD
Oxygenator	Yes	No
Circuit	Longer, with reservoir	Short, no reservoir
Anticoagulation	ACT 180–220 seconds	ACT 140–160 seconds
Risk of air embolus	Low (air trapped in the circuit)	Higher, particularly with L-VAD
Type of support	Cardiorespiratory	Cardiac
Ventricular decompression	May need atrial septostomy/LA vent	Not required
Length of support	Up to weeks	Up to weeks
Cannulation	Trans-thoracic or neck	Trans-thoracic

and centrifugal extracorporeal VADs (**Table 14.1**). It is encouraging that there are now also newer paracorporeal ventricular assistance devices available for smaller patients, which have been used with reasonable success in bridging children to transplantation for periods of up to several months. For completeness, although rarely used in paediatric practice, intra-aortic balloon pump will also be discussed.

EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO)

ECMO remains the mainstay form of mechanical cardiac support used in the paediatric population. There are two principal modes of ECMO support: *veno-venous ECMO* provides only respiratory support, whilst *veno-arterial ECMO* bypasses both the heart and lungs and is thus the appropriate type of ECMO for patients with circulatory failure. The great advantage of ECMO over other devices is that it can be used to support patients with biventricular failure as well as patients with associated respiratory failure and refractory pulmonary hypertension.

Survival-to-discharge rates of patients treated with ECMO for short-term cardiac support are in the range 40–80%, depending on the patient's diagnosis and underlying pathophysiology, as described previously (ECMO Registry, 2005). ECMO is, however, limited to support patients only for several weeks (in our experience, maximum 4–6 weeks), as the incidence of complications, particularly infection, increases with time (Chaturvedi *et al*., 2004). For patients requiring longer-term support, other types of ventricular assistance devices might be more suitable.

An ECMO circuit as used in our institution is shown in **Figure 14.1**. The circuit comprises the following: 1, mixed venous O_2 saturation probe; 2, blood sampling port; 3, heparin infusion/venous pressure monitoring port; 4, bladder reservoir; 5, roller pump; 6, membrane oxygenator with pre-postmembrane pressure monitoring ports and integrated heater; 7, gas blender; 8, postmembrane blood sampling port (also used for administration of clotting products and platelets); 9, bridge (allows recirculation when the patient is clamped off); 10, flow probe. A haemofilter may be added into the circuit and driven by the pressure gradient between port (8) and bladder (4).

VENTRICULAR ASSISTANCE DEVICES (VADs)

Extracorporeal support with non-pulsatile centrifugal VADs have been used to effectively support infants and children with ventricular failure after congenital heart surgery. This applies in particular to patients with left ventricular failure, following repair of anomalous origin of left coronary artery from the pulmonary artery or late arterial switch operation and right ventricular failure

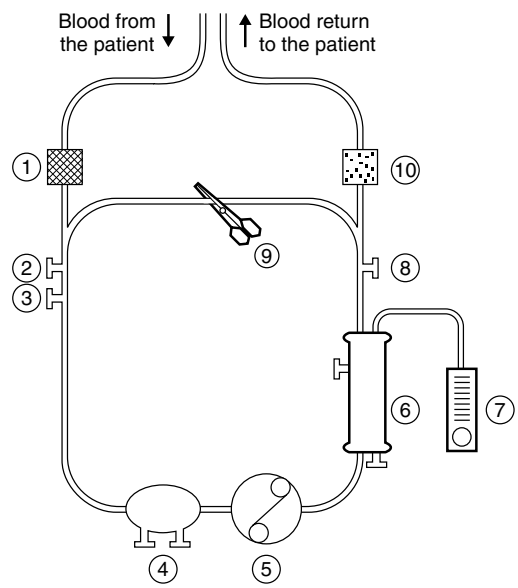

Figure 14.1

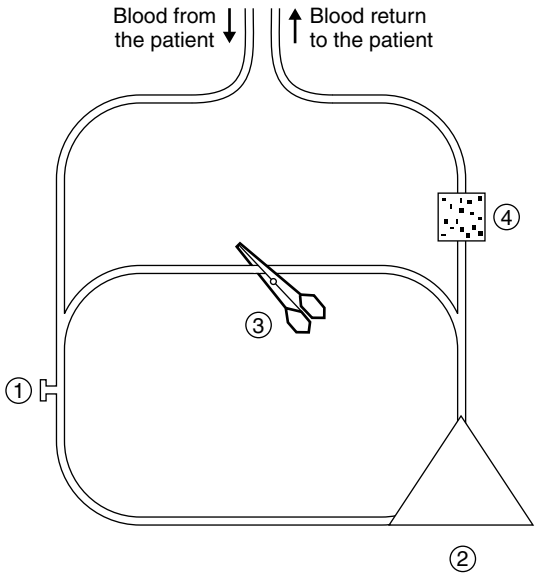

Figure 14.2

secondary to pulmonary hypertension following heart transplantation (del Nido *et al*., 1999).

There are two large reports on children supported perioperatively with centrifugal VADs, with survival-to-hospital-discharge rates similar to that of ECMO, at around 40% (Duncan *et al*., 1999; Thuys *et al*., 1998).

Figure 14.2 demonstrates our paracorporeal VAD circuit with a centrifugal pump. The VAD circuit comprises the following: 1, heparin infusion/venous pressure monitoring port; 2, centrifugal pump; 3, bridge (allows recirculation when the patient is clamped off); and 4, flow probe.

The great advantage of using centrifugal VADs over ECMO in this circumstance is the simplicity of the circuit, better decompression of the supported ventricle, no need for the oxygenator, lower level of anticoagulation required and probably also lower staff:patient ratios. The limitation of this device is that, although it can in theory be used for biventricular support, this is unrealistic in babies and infants, as it requires four cannulae and two pumps. Univentricular VADs are also unsuitable for patients with large intracardiac shunt, patients with pulmonary hypertension and those with refractory respiratory failure. The other limitation of this type of VAD is that, as with ECMO, it can only be used for short-term support up to several weeks. The main characteristics of ECMO and centrifugal VAD are shown in **Table 14.1**.

For long-term mechanical support there are a variety of devices, ranging from paracorporeal ventricular assistance devices to fully implantable impeller pumps. The two most common paracorporeal VADs used in infants and smaller children are the Excor from the Berlin Heart AG and the Medos HIA. These are pneumatically driven devices generating pulsatile flow. The pumps are available in a variety of sizes with stroke volumes down to 10 ml, which is suitable for term neonates (Merkle *et al*., 2003; Reinhartz *et al*., 2002). The Thoratec paracorporeal device has been used with great success in adults as well as children over 10 years of age (Reinhartz *et al*., 2001).

The mainstay use of the paracorporeal devices in children has been as a bridge to heart transplantation for patients with cardiomyopathy or myocarditis. Some of these patients have been successfully supported for up to 100 days, during which time some patients have been extubated and ambulatory within the hospital. The use of these devices in the postcardiotomy setting has been limited, with poor results. The major risk with these VADs has been either thrombo-embolic or haemorrhagic stroke; neurological events vary (10–30%) (Merkle *et al*., 2003; Reinhartz *et al*., 2001).

Other recent advances in long-term mechanical assistance devices centre around the use of impeller axial flow pumps, such as the DeBakey from Micromed, Jarvik, and Incor from Berlin Heart AG (Song *et al*., 2003). There is now a paediatric version of the DeBakey pump, but it is too early to comment on the success of these devices in children.

INTRA-AORTIC BALLOON PUMP (IABP)

The aim of the intra-aortic balloon pump (IABP) is to improve coronary perfusion and reduce left ventricular afterload in patients with left ventricular failure refractory to optimal medical therapy. The balloon is placed in the descending thoracic aorta. It inflates during diastole, enhancing coronary blood flow, and deflates at the beginning of systole, reducing left ventricular afterload, thus augmenting cardiac output. Although the balloons have been appropriately downsized for infants and small children, there is still a concern about peripheral vascular complications leading to mesenteric and limb ischaemia (Akomea-Agyin *et al*., 1999). Some reservations exist as to whether the more compliant aorta seen in children limits the effectiveness of balloon counter-pulsation. Faster heart rates in children also make synchronization of the balloon pump difficult.

The vast majority of experience with IABP remains in the adult population and these devices, therefore, cannot routinely be recommended for infants and small children. In the older children and adolescents, the IABP may have a role in treating patients with moderate left ventricular failure prior to inserting a different form of mechanical assistance or as an adjunct for weaning from ECMO.

INDICATIONS AND TIMING FOR MECHANICAL SUPPORT

The indications for paediatric cardiac mechanical support can be divided into two groups, (a) patients with cardiac failure following open heart surgery, and (b) medical patients with cardiac failure due to a spectrum of diseases, including myocarditis, cardiomyopathy, pulmonary hypertension, arrhythmias, events related to cardiac catheterization and infants who decompensate prior to surgery. Circulatory failure secondary to overwhelming sepsis will not be dealt with further in this chapter, other than to say that mechanical support can be used to salvage such patients, with an expected survival rate of around 50–60% (Goldman *et al*., 1997).

Postcardiotomy Circulatory Failure

Children with structural heart disease following cardiac surgery comprise by far the largest group of paediatric cardiac patients requiring mechanical circulatory support. The postcardiotomy myocardial dysfunction is usually related to myocardial stun, due to myocardial ischaemia–reperfusion from aortic cross-clamp, direct myocardial injury from surgery and the inflammatory response to cardiopulmonary bypass. It is vital to identify any potentially correctable residual anatomical lesion as the contributory cause of the patient's circulatory failure. Other, less common indications for mechanical support in postcardiotomy patients include refractory pulmonary hypertension and malignant arrhythmias.

When choosing the optimal time to initiate mechanical support, one has to balance the risks of waiting too long and the patient suffering severe end-organ injury vs. the risks inherent in an invasive treatment such as ECMO. Unfortunately, there is no easy formula for predicting severity of illness and patient's outcome as there is for respiratory ECMO using the oxygenation index. In this sense, practice varies widely between different institutions in terms of the optimal timing of starting mechanical support.

In most centres postoperative mechanical support is initiated in patients with worsening signs of inadequate oxygen delivery (rising lactate, falling mixed venous oxygen saturation, wide core–peripheral temperature gap, poor urine output and other signs of end-organ ischaemia) despite appropriate vasoactive medical management. Because of the concerns of myocardial damage associated with high doses of epinephrine and to avoid organ damage from periods of low cardiac output, there is a trend towards a pre-emptive, earlier institution of mechanical support in patients not responding to conventional treatment (Chaturvedi *et al*., 2004). Our policy is to consider mechanical support in patients who continue to deteriorate with signs of low cardiac output despite increasing inotropic support (epinephrine dose is approaching 0.3 μg/kg/min).

Some centres have proposed, with good effect (90% survival), the semi-elective use of ventricular assist devices in postcardiotomy patients who have a high probability of severe left ventricular myocardial dysfunction in the early postoperative period, e.g. following repair of anomalous left coronary artery from pulmonary artery and after a late arterial switch operation (del Nido *et al*., 1999; Kang *et al*., 2004).

Other centres advocate an elective use of ventricular assistance support for all patients after the Norwood stage 1 operation. This is based on the premise that ensuring adequate cardiac output and thus cerebral perfusion during the first 48 hours after surgery will protect these patients against the known long-term high risks of neurological morbidity. Preliminary reports have suggested an 80% survival to discharge in this group of patients, which is in fact comparable to centres not using mechanical support after the Norwood stage 1 operation. While there does not appear to be a short-term survival benefit using this strategy, the results of long-term neurological follow-up are eagerly awaited (Ungerleider *et al*., 2004).

Controversy still exists as to the prognostic significance of the need to place patients on ECMO directly from cardiopulmonary bypass, as opposed to the need to initiate ECMO in the intensive care. We have reported a survival benefit for those patients who went on ECMO directly from cardiopulmonary bypass. This may be related to the fact that these patients avoided the ill-effects of prolonged low cardiac output state prior to the institution of mechanical support (Chaturvedi *et al*., 2004). Others have reported increased mortality and morbidity (bleeding complications) in those patients who failed to separate from cardiopulmonary bypass (Duncan *et al*., 1999).

In terms of univentricular vs. biventricular anatomy as an indication for mechanical support, there is now increasing evidence that patient's outcome is related more to the potential reversibility of myocardial dysfunction than to the underlying cardiac anatomy. Thus, patients with single ventricle physiology and other complex heart lesions are now readily supported, provided the cardiac lesions are deemed operable and the principal indication for support is potentially reversible (Morris *et al*., 2004; Aharon *et al*., 2001).

Circulatory Failure Due to Myocarditis or Cardiomyopathy

The traditional belief has been that children who present with fulminant myocardial failure usually have a diagnosis of myocarditis, whilst those with more long-standing

cardiac failure have cardiomyopathy. It is now becoming increasingly apparent that the distinction between these two groups of patients is almost impossible at the time of presentation, and that even with the use of myocardial biopsy the diagnosis is not always clear. It is not unusual that patients thought to have myocarditis do not recover to wean from mechanical support, while patients with long-standing cardiomyopathy with acute deterioration do recover sufficiently to wean from support and need transplantation at a later stage. It therefore seems more appropriate that both these groups of patients are seen as a single entity of acute cardiac failure, and that the aim of support in the first instance should always be as a bridge to recovery.

When choosing the optimal time for initiation of mechanical support for these patients, the same principles apply as to postcardiotomy patients. Occurrence of arrhythmias, intolerance of enteral feeding and oliguric renal failure are often ominous signs and a pre-emptive, semi-elective initiation of mechanical support may be warranted.

In patients who fail to show improvement in ventricular function over the course of the first week the aim of support should be reviewed as a bridge to transplantation. At this time, changing to a device capable of long-term support (months) should be considered.

Rapid Deployment ECMO or Extracorporeal Cardiopulmonary Resuscitation (ECPR)

In view of the high risk of severe neurological injury in patients who suffer a prolonged cardiac arrest, many centres have opted to have extracorporeal cardiopulmonary resuscitation (ECPR) capability. There are now several reports in which the outcome of patients placed on ECMO during cardiac arrest are encouraging, with survival rates of around 40% (Duncan *et al*., 1998; Morris *et al*., 2004). Several of these patients have suffered prolonged arrests of up to 60 minutes, with apparently intact neurological survival. It is, however, vital that long-term follow-up studies of neurological outcome of these patients are carried out, so that a balanced view of the merits of this practice is forthcoming.

The following should be available for delivery of ECPR programme: staff trained and capable of rapidly priming and managing an ECMO circuit, surgeons trained in performing cannulation, theatre nurses and ECMO specialists. In reality, this infrastructure is only available out of working hours in a select number of large tertiary centres. An alternative approach is to have a clear primed ECMO circuit always available within the cardiac intensive care unit, together with surgical and ECMO expertise. In a recent study we have demonstrated (Karimova *et al*., 2005) that if ECMO circuits are primed with crystalloid solution and stored in the appropriate fashion, these circuits can be safely maintained for a period of up to 2 weeks. This may go some way to improve the availability of ECPR. However, it should be stressed that ECPR should only be used as a last resort for patients suffering an unexplained cardiac arrest. In most circumstances an anticipatory strategy to initiate ECMO support is preferable, as described earlier.

PATIENT SELECTION CRITERIA

The criteria for mechanical support have continued to evolve with the increasing ECMO experience and the development of new mechanical devices. It is therefore important to focus on the principles of patient selection, rather than absolute criteria:

- First, the patient is rapidly deteriorating despite maximum conventional treatment.
- Second, the underlying cardiac and/or respiratory pathophysiology is potentially reversible, unless the aim of support is as a bridge to heart transplantation.
- Third, any haemodynamicaly important, surgically correctable, residual lesions should be identified, as the presence of such unrecognized defects may exacerbate the haemodynamic compromise and minimize the chances of recovery. It is crucial that patients have detailed postoperative imaging, either by echocardiography, cardiac catheterization, computed tomography or magnetic resonance scan.
- Fourth, the patient should not have suffered any other complication for which intensive care support would otherwise be discontinued (such as severe neurological injury from hypoxic–ischaemic encephalopathy) and/or does not have an absolute contraindication to anticoagulation (such as a recent significant cerebral haemorrhage). From a practical point of view, it is often impossible in the emergency setting to adequately assess the patient's cerebral function. In this circumstance the patient is given the benefit of doubt and the neurological function tests are carried out as soon as feasible on mechanical support. The wishes and expectations of the patient's family should always be taken into consideration.

PRACTICAL ASPECTS OF MECHANICAL SUPPORT

Cannulation

Veno-arterial ECMO in children can either be instituted via cannulation of the neck vessels (right common carotid artery and right internal jugular vein ± an additional femoral venous cannula) or cannulation via a median sternotomy, usually through the operative site. The

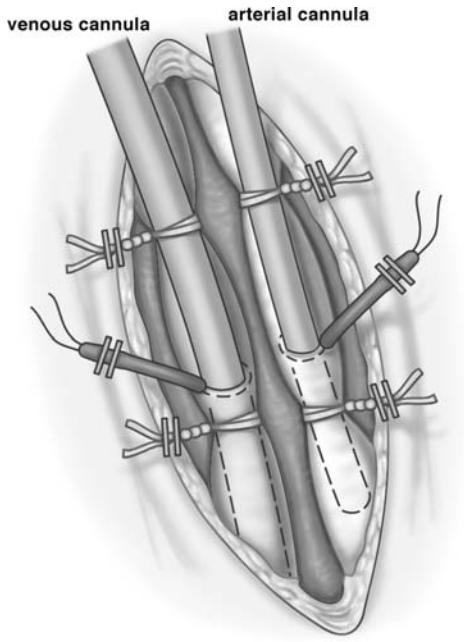

Figure 14.3

technique of infant ECMO cannulation (**Figure 14.3**), allowing preservation of the carotid and jugular vessels, was well described by Karl *et al.* (1990). The Silastic vessel–loop technique provides excellent haemostasis around the cannulae during ECMO and also allows reconstruction of the vessels during decannulation.

In patients who fail to wean from cardiopulmonary bypass and are placed directly on ECMO in theatre, cannulation is usually achieved using the bypass cannulae in the aorta and the right atrium, with most patients also requiring an additional left atrial vent for decompression. The same would apply for postcardiotomy patients requiring urgent cannulation in a cardiac arrest or near-arrest situation in the intensive care unit, as this would be the quickest way to deliver mechanical support. However, if time permits and there are no contraindications to neck cannulation, then this is probably the preferred site of cannulation, as it reduces the risk of bleeding and infection.

It is vital to decompress the left heart, particularly where there is marked left ventricular impairment and failure of aortic valve to open. In this circumstance the left atrial pressure is likely to be very high (above 30 mmHg), causing lung congestion. A high left ventricular end-diastolic pressure is also likely to increase ventricular wall stress and impair coronary perfusion. The decompression of the left heart can be achieved either by creating an atrial communication in the catheter laboratory or by directly placing a left atrial vent.

Cannulation for a paracorporeal device is done on cardiopulmonary bypass in the operating theatre. The drainage cannula for the LVAD can be inserted into either the left atrium or directly into the apex of the left ventricle, with the return cannula placed directly into the ascending aorta. Left ventricular cannulation is now thought to be the preferred site, as it is associated with improved drainage and decompression of the left ventricle, which may lead to lower rates of cerebral events (Merkle *et al.*, 2003; Reinhartz *et al.*, 2001).

Management of Bleeding

Excessive bleeding is one of the most common complications of postoperative cardiac ECMO, with important implications. This starts a vicious circle in which there is a need for massive blood transfusion, leading to worsening coagualopathy and further bleeding. The management of postcardiotomy bleeding in our practice involves:

1. Aggressive correction of any coagulopathy (maintaining platelet count > 100 000 with platelets transfusion) and fibrinogen > 2 g/l with cryoprecipitate.
2. Maintaining the activated clotting time (ACT) in the range 160–180 seconds.
3. Administration of antifibrinolytic agents, such as aprotinin.
4. Chest re-exploration for significant ongoing bleeding (> 10 ml/kg/hour) despite correction of the patient's coagulopathy and the above mentioned manoeuvres.
5. Conversion of chest to neck cannulation if applicable.
6. Using a blood cell-saver.
7. If the bleeding continues, running the circuit heparin-free for a period of 4–6 hours until the bleeding is controlled. In this situation a spare primed ECMO circuit should be readily available on stand-by in case of major clotting of the circuit.
8. More recently, the use of recombinant activated factor VII has been advocated for refractory bleeding in postcardiotomy patients (Pychynska-Pokorska *et al.*, 2004). This treatment cannot yet be recommended for routine use until its safety and efficacy has been tested in randomized clinical trials.

Determining Adequacy of ECMO

The usual markers of end-organ perfusion should be used to assess the adequacy of extracorporeal support. These include serial measurements of acid–base status, lactate and mixed venous oxygen saturation (the latter is of limited use in the setting of a left atrial vent

or atrial communication) as well as urine output. Although there is insufficient knowledge about optimal ECMO flows to facilitate ventricular recovery (usually 100–200 ml/kg/min), it seems logical that the lowest ECMO flow, that provides satisfactory oxygen delivery and ventricular decompression, should be used.

In circumstances where there is deemed to be inadequate systemic oxygen delivery, the haematocrit should be optimized to 40% and the blood leaving the oxygenator should be checked to be fully saturated. The use of ultrasonic measurements of blood flow on the arterial limb of the ECMO circuit distal to the bridge (**Figure 14.1**) serves as a useful guide to the actual blood flow. In a situation in which one does not have a device to measure the actual flow, it should be always checked that the roller pump occlusion is appropriate and that the circuit bridge is fully clamped. If the patient continues to have signs of inadequate end-organ perfusion despite the above manoeuvres, then the pump flow should be increased to meet the patient's metabolic demands. In the rare situation where very high flow, i.e. > 200 ml/kg/min, is not able to achieve adequate end-organ perfusion, reducing the patient's metabolic demand by cooling him/her to 34°C can be considered.

Generous use of vasodilators to optimize perfusion, control hypertension and reduce afterload is frequently necessary on ECMO. The afterload reduction and maintenance of optimal volume status (right atrial pressure > 5 mmHg) are of key importance when centrifugal pump is used.

Management of ECMO in Patients with Systemic–Pulmonary Shunt

In patients after Norwood stage I operation with Blalock–Taussig shunt (BT shunt), VAD support is preferable to ECMO, unless there is an evidence of associated lung injury (Ungerleider *et al*., 2004). The VAD flow should be maintained high (at around 200 ml/kg/min) to compensate for the patent shunt and to ensure adequate systemic perfusion. When ECMO is used for these patients, the BT shunts should not be fully occluded, as this can result in shunt thrombosis and infarction of the lungs (Jaggers *et al*., 2000).

In patients where the Sano modification of Norwood stage I procedure has been performed (see Chapter 41) and mechanical support is needed, ECMO (rather than VAD) is required, as there may be inadequate blood flow to the lungs in this physiology.

Management of Patients on Mechanical Support

Patients should be managed in the most physiological manner possible, i.e. enterally fed (unless contraindicated) and minimally sedated to keep them comfortable and safe; the parents should be encouraged to participate in their child's bedside cares. Ventilation on ECMO is usually maintained on ''resting'' settings, with tidal volumes of 5–10 ml/kg (maximum peak inspiratory pressure 25 cmH_2O), PEEP 7–10 cmH_2O and a ventilator rate of 15–20/min. In order to avoid pulmonary venous desaturation, the FiO_2 on the ventilator is typically set at 40%. In contrast to ECMO, full ventilation is mandatory in patients on VAD. Although antibiotic cover is not routinely necessary for patients on ECMO, it is common practice to perform daily blood cultures and treat any new infection aggressively.

Weaning from ECMO

One of the greatest challenges of cardiac ECMO is determining the point at which there has been adequate ventricular recovery to allow for a trial of weaning from mechanical support. Our practice is to rest the patient on full ECMO support for 48–72 hours. After this time, a ''stress test'' is performed on an infusion of inotropes (usually adrenaline or dopamine in combination with milrinone): the left atrial vent (if present) is clamped off and the ECMO flow is gradually reduced over a period of 1–2 hours until the lowest safe flow is achieved (usually around 100–300 ml/minute). The ECMO flows should not be reduced below this level because of the increased risk of clotting of the cannulae and the circuit. If flow reduction is successfully achieved without haemodynamic compromise, the patient is clamped off ECMO support and the pump left to recirculate via the bridge. The patient is usually tried off ECMO for a period of 1–3 hours before a decision about decannulation is made. The echocardiogram is useful in this setting to assess ventricular function as well as to check for any undiagnosed lesions, pulmonary hypertension, regional wall dyskinesis, pericardial effusions or any other complication.

If the patient is deemed to have failed the trial off ECMO, as indicated by unstable haemodynamics and/or signs of inadequate tissue oxygen delivery, then full ECMO support is recommenced. It is advisable to wait a further 48 hours before the next trial of weaning from ECMO to allow a decent period of time for further ventricular rest and recovery.

When to Stop Mechanical Support (How Long Is Long Enough?)

The duration of anticipated recovery on mechanical support depends on the underlying pathophysiology. In postcardiotomy patients, where the pathology is essentially postoperative myocardial stun, some improvement of ventricular function should be evident within 72–96 hours of support (Duncan *et al*., 1999; Aharon *et al*.,

2001). Duncan *et al*. (1999) proposed that if there are no signs of ventricular recovery by this time, then these patients should be considered for early listing for transplantation. The timing for listing will also depend on organ availability.

The issue of when to stop mechanical support in patients who fail to show signs of effective ventricular recovery, and who are not suitable for transplantation, is understandably emotive. Most survivors are on ECMO for periods of 5–10 days (Chaturvedi *et al*., 2004). Recovery is thought to be less likely beyond 10 days in this patient group, although this view has been recently challenged by a report of reasonable survival rates after 250 hours of mechanical support (Morris *et al*., 2004). It is our current policy to withdraw mechanical support if there are no signs of recovery of ventricular function after 14 days of support and the patient is not a candidate for heart transplantation.

One of the greatest challenges of cardiac ECMO remains identifying the markers of potential ventricular recovery. Unfortunately, at the present time, we still rely on crude assessment of ventricular function using traditional echocardiography and markers of adequacy of oxygen delivery when the support is weaned. There have been some encouraging reports recently that myocardial tissue Doppler may be a more sensitive predictor of ventricular recovery, as this provides a measure of myocardial function independent of loading conditions (Cheung *et al*., 2004).

In patients with myocarditis, one would anticipate adequate ventricular recovery, sufficient for a trial of weaning from mechanical support within a 10–14 day period. If weaning fails and the patient remains a candidate for heart transplantation, then mechanical support should be continued as a bridge to transplant for as long as the patient remains a transplant candidate (Duncan *et al*., 2001). However, availability of the appropriate mechanical support device for the anticipated long waiting time for a donor heart becomes a limiting factor.

It is interesting that some patients with cardiomyopathy have demonstrated an improvement in ventricular function sufficient to wean off ECMO over a 1–3 week period of support. These patients require very close follow-up and often remain listed for transplantation (McMahon *et al*., 2003).

COMPLICATIONS OF MECHANICAL SUPPORT

The four major complications of cardiac ECMO reported by ELSO (ECLS Registry, 2005) are:

1. Surgical site bleeding (incidence ± 30%).
2. Infection (incidence ± 10%; associated with increased length of time on the device).
3. Neurological events, i.e. haemorrhage and infarcts (incidence ± 6%; higher in neonates at ± 12%).
4. Oxygenator, pump or circuit failure (incidence ± 10%).

The major complications of the paracorporeal devices include cerebral events (haemorrhage and thromboembolism) and infection, as described earlier. The management of these complications has been addressed earlier in the chapter.

OUTCOMES

It is disappointing to note that over the past decade, despite the increased use of paediatric cardiac ECMO, survival to hospital discharge of patients requiring postcardiotomy mechanical support remains relatively unchanged at around 40% (ECMO Registry, 2005). Survival rates appear to be independent of the mode of mechanical support used and whether this was initiated semi-electively or in the setting of a cardiac arrest (Duncan *et al*., 1999; Morris *et al*., 2004). The late mortality noted after weaning from mechanical support appears to relate to a progressive low cardiac output state in some patients with multi-organ failure, as well as to neurological complications acquired prior to commencing of ECMO (Hintz *et al*., 2005).

The outcomes of patients with myocarditis or cardiomyopathy are more favourable. When mechanical support is used as a bridge to recovery for myocarditis patients, the survival to discharge is around 70% (Acker, 2001). Mechanical bridge to transplantation has been very successful in the paediatric population, with 70–80% of patients surviving to transplantation and around 70% surviving to hospital discharge (Goldman *et al*., 2003; Levi *et al*., 2002).

FUTURE DEVELOPMENTS

One of the greatest challenges for mechanical support teams for the paediatric population is the development of ventricular assistance devices capable of providing long-term support for infants and small children. Ideally, these devices should be implantable and allow patients to be ambulatory. Other, much needed advances in technology relate to the development of biocompatible tubing that does not require systemic anticoagulation. Additionally, better methods are needed for evaluation of ventricular function. These would potentially guide us in the decision-making process during the initiation and weaning from mechanical support.

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15

Surgical Approaches

J. Stark and V. T. Tsang

Several factors should be considered before a surgeon decides on the optimal approach to the heart. Adequate exposure is the most important part of any operation. The surgeon must have the best possible view of the part of the heart in which the lesion is located. In addition, an adequate approach to other parts of the heart and great vessels must be available in order to facilitate cannulation for cardiopulmonary bypass, to evacuate air and to apply electrodes for defibrillation. From the respiratory point of view, patients are more satisfactorily ventilated in a supine position than in a lateral position. A bilateral thoracotomy incision (clamshell incision), although it may interfere with respiratory function, is used routinely at our institution for heart–lung or sequential bilateral single lung transplantation because of superior surgical exposure. The cosmetic effects, although usually of secondary importance, should also be considered.

Midline sternotomy is a standard procedure for most operations performed on cardiopulmonary bypass. Our preference is to combine the cosmetic advantages of a limited skin incision and a complete median sternotomy. An alternative is a limited midline sternotomy leaving the manubrium intact, which has gained some popularity for repair of some simple congenital heart defects in children. There are, however, no firm data to suggest the superiority of this approach in terms of postoperative recovery and pain control (Laussen *et al*., 2000). In addition, median sternotomy can also be used for both central and lateral systemic–pulmonary artery shunts and pulmonary artery banding.

Right thoracotomy is used for right-sided systemic–pulmonary shunts and, rarely, for Glenn and Blalock–Hanlon atrial septectomy. Right thoracotomy also provides excellent exposure to atrial septal defects and the tricuspid and mitral valves. It is useful in reoperations after Mustard or Senning operation (Szarnicki *et al*., 1978).

Left thoracotomy is used for ligation of persistent ductus arteriosus, coarctation repair, repair of most forms of vascular rings, pulmonary artery banding, and left-sided systemic–pulmonary shunts. The increasing use of mechanical assistance devices for patients in severe heart failure may require the placement of the device from the apex of the left ventricle to the descending thoracic aorta.

Reoperations require special consideration. In some patients it may be possible to use an approach different from the one used in the original operation in order to avoid dissection of the troublesome adhesions. An example is the right thoracotomy for reoperations on the mitral valve or systemic or pulmonary venous obstruction after Mustard or Senning operation. Dissection of the right ventricle and possible damage to the coronary arteries is avoided; access for the cannulation of the superior vena cava (SVC) and the inferior vena cava (IVC) is excellent. If a repeated midline sternotomy is to be performed, careful assessment of the proximity of the important structures (aorta, conduit) to the sternum should be made. It may be prudent to prepare or even to cannulate the iliac or femoral vessels before performing the sternotomy, using thin-walled DLP or Biomedicus cannulae (these provide superior flow with smaller diameter). The patency status of the groin vessels should be part of the surgical planning process. Details of surgical approaches at reoperations have been described elsewhere (Stark and Pacifico, 1989).

MIDLINE STERNOTOMY

A midline sternotomy is the approach most commonly used in patients undergoing operation with cardiopulmonary bypass. A patient is placed on the operating table in the supine position with a towel underneath the shoulders (**Figure 15.1**). At the authors' institution, betadine is currently used for skin preparation. After the third painting, the solution is allowed to dry, and Steri-Drape is applied to the chest, neck and abdomen. It is important not to pull the skin while the Steri-Drape is applied and to position the head of the patient straight, otherwise the skin incision and the resulting scar may

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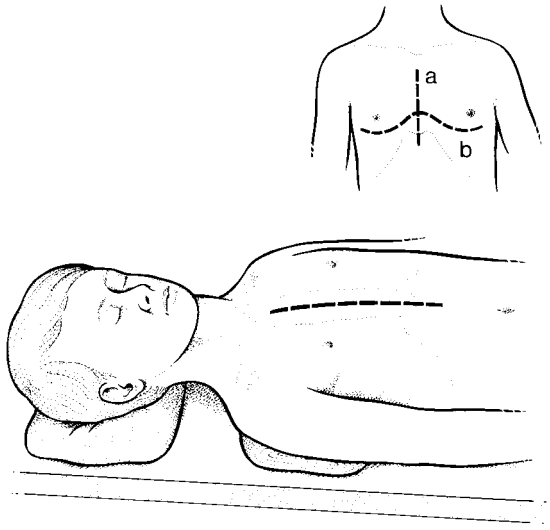


Figure 15.1

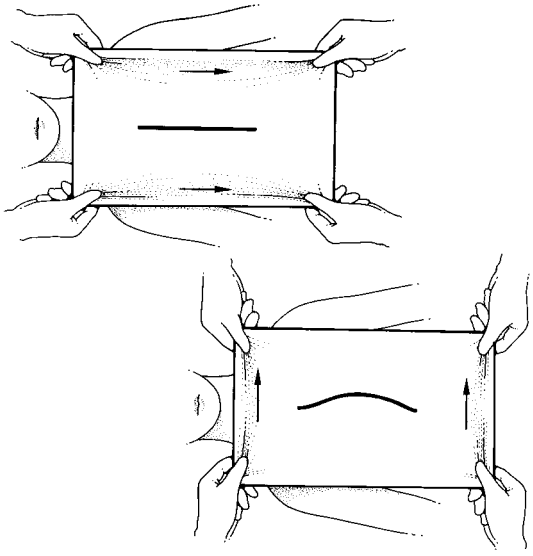


Figure 15.2

be asymmetrical (**Figure 15.2**). If the iliac vessels are to be used for cannulation, one side of the groin is also prepared.

The skin incision starts at the manubrio-sternal junction and stops just short of the xiphoid process (**Figure 15.1**). For cosmetic reasons, the superior part of the incision can be made slightly shorter in female patients. However, the superior end of the incision should be extended towards the sternal notch in operations involving the head vessels and the aortic arch.

A transverse submammary incision (Brutel de la Riviere *et al*., 1981) can also be used. This produces a better cosmetic result. Skin incision is illustrated in **Figure 15.1**, line b. The skin flap is raised superiorly and inferiorly and then a midline sternotomy is performed in the usual way. However, one should be aware of the possibility of distorting the development of the breast and/or denervation. The authors do not use this incision.

The skin is incised with a scalpel, and diathermy is used for all subsequent layers in order to minimize bleeding (**Figure 15.3**). The linea alba is opened, and the space underneath the xiphoid and sternum is dissected with a Denis Browne dissector (**Figure 15.3b**). The interclavicular ligament is dissected with a right-angled dissector (**Figure 15.4**). A Denis Browne dissector is very suitable for this dissection (**Figure 15.5**). The incision is then carried to the periostium and all bleeding points are carefully stopped. The sternum is then split from the xiphoid process to the top by an electric or air-driven saw (**Figure 15.6**). It is important to stay in the midline; a deviation can cause cutting through the cartilages of the ribs and make subsequent sternal closure difficult and unstable. During sawing, the anaesthetist stops the ventilation and releases pressure on the lungs, so that the pleura and soft tissues fall away from the sternum. Some surgeons prefer to open the sternum from the top. After the suprasternal ligament is cut, the space under the manubrium is developed with a right-angled dissector. The blade of a saw is introduced into this

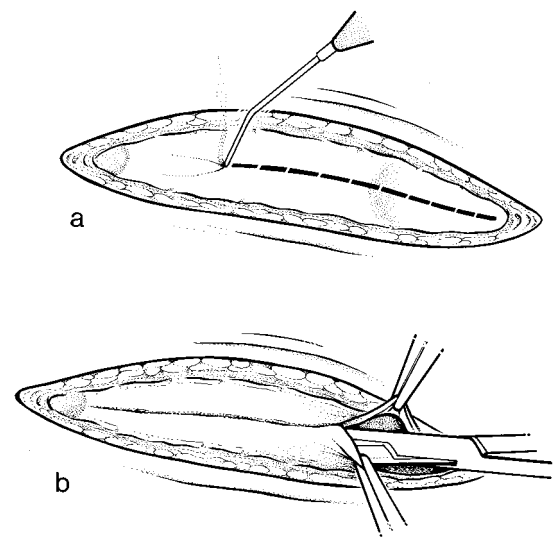


Figure 15.3

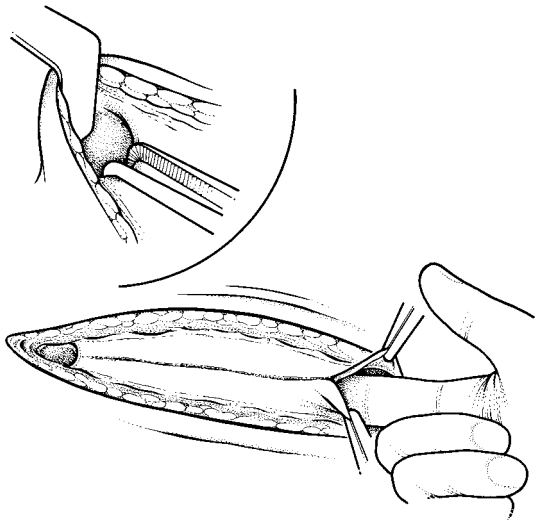


Figure 15.4

Figure 15.5

space, and the sternum is split. The saw tip should be kept elevated so that the saw toe presses firmly against the sternum. An oscillating saw is very useful for cutting the external table of the sternum during reoperations; it is also possible to cut the external table with a saw and then cut the internal table with Mayo scissors. In older children who are severely cyanotic, blood loss during sternotomy may be considerable. Bleeding occurs from multiple collateral vessels but also from the bone and periosteum. An adequate amount of blood should be available at this stage. Bleeding is controlled by diathermy and bone wax (**Figure 15.7**).

Using a gauze swab to protect surgeon's gloves from being caught by sharp bone edges can facilitate the application of bone wax. A minimal amount of bone wax should be used to avoid non-union of the sternum;

Figure 15.6

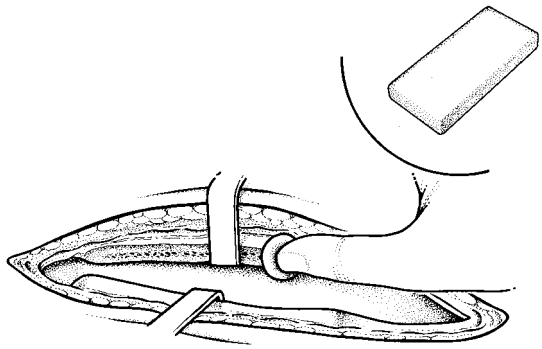


Figure 15.7

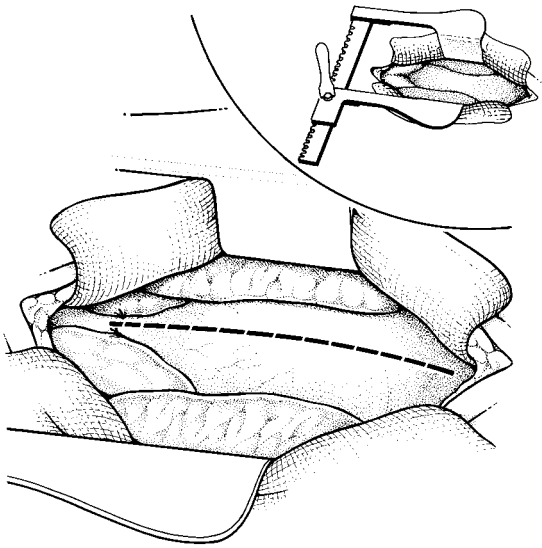


Figure 15.8

excessive use of bone wax is also thought to increase the risk of infection. The edges of the sternum are covered with small towels and a Finochietto retractor is inserted. The authors prefer to insert it with the horizontal bar placed superiorly (**Figure 15.8**). In this way, the lower part of the wound is free and the bar does not interfere with subsequent steps in the operation. The sternal retractor is opened gradually to avoid sternal fracture. The smallest opening of the sternal retractor that gives adequate exposure should be used. Mediastinal fat is dissected with sharp and blunt dissectors, and both pleurae are pushed to the sides.

The thymus is separated between its lobes and the thymic veins are ligated or occluded with diathermy. The proximity of the phrenic nerves must be borne in mind during dissection. If the thymus is very large or an external valved conduit is being inserted in an infant or a small child, subtotal thymectomy is performed. Tissue plane must be respected in the case of a subcapsular excision, because bleeding to the thymus may cause severe mediastinal compression or tamponade.

The pericardium is usually opened in the midline (**Figure 15.8**) and the edges are suspended to the edges of the wound and to the sternal retractor. If a pericardial patch is to be used for repair, the pericardium is opened on the right side.

Cannulation

The tissue plane between the right pulmonary artery and the aorta is opened with scissors, and then a right-angled Denis Browne dissector is used to encircle the aorta with a tape. Similarly, the SVC is dissected just above the right pulmonary artery. For IVC dissection, the tissue plane lateral to the low right atrium and the IVC is dissected with blunt dissection, and then a right-angled dissector is passed around from the medial aspect of the IVC. It is important to avoid damaging the right phrenic nerve, which runs very close to the lateral aspect of the IVC.

Purse-string sutures for cannulation are then placed (**Figure 15.9**). At the authors' institution, 5-0 Ti-cron cylindrical purse-string is used in infants under 6 months of age, and 4-0 Ti-cron ''diamond'' purse-string in older children. We routinely place two purse-strings on the aorta for better security during decannulation. If the aorta is particularly thin, purse-string sutures with felt pledgets are used.

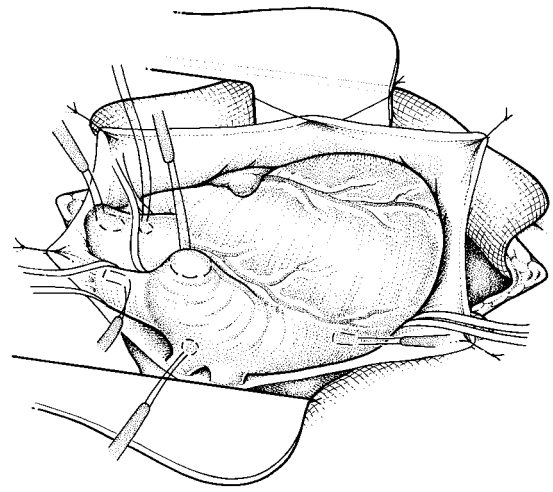


Figure 15.9

The aortic cannula is routinely placed at the base of the innominate artery, pointing towards the aortic arch. If necessary, the small arterial cannula can be repositioned into the innominate artery for regional low flow perfusion. Under special circumstances, such as in the arterial switch operation or truncus arteriosus repair, the cannula may have to be placed on the aortic arch. At the authors' institution, a DLP or Stockard-Shiley wire-reinforced cannula is routinely used. Another purse-string suture is placed on the ascending aorta for cardioplegia and subsequent de-airing.

Some sick children and especially infants do not tolerate retraction of the heart, which is required for placement of IVC purse-string sutures and IVC cannulation. For this reason, cardiopulmonary bypass is often begun with one cannula in the right atrial appendage or in the SVC. When using lower median sternotomy, the SVC is cannulated via the right atrium appendage, using the thin-walled Biomedicus straight cannula. This cannula provides superior flow dynamics. The IVC is then easily cannulated on partial bypass.

The SVC purse-string suture is placed either directly on the SVC or on the right atrial appendage. If direct SVC cannulation is planned, the purse-string suture should be oblong rather than circular to avoid SVC stenosis. The IVC purse-string suture is placed posteriorly on the lower right atrium or directly on the IVC. This provides better surgical exposure for heart transplantation, total cavopulmonary connexion or for Senning operation.

Venting the left side of the heart is important to avoid distension and to facilitate de-airing at the end of the procedure. The authors prefer to vent the left side through the right superior pulmonary vein (**Figure 15.9**). The purse-string is placed close to the interatrial groove, and at the end of the procedure, if indicated; a left atrial line is placed through this purse-string. Other methods of venting the left heart are through the patent foramen ovale or, if the atrial septum is intact, through a small incision made in the fossa ovalis. Pulmonary artery can also be used for venting. Venting the left heart via the left ventricular apex is effective, although rarely used today.

Closure of Sternotomy

When the intracardiac procedure is completed, all bleeding points are first controlled. All suture lines and purse-string sutures are checked. Areas of aortic, SVC and IVC dissection are inspected. Previous systemic–pulmonary shunts that have been dissected and ligated are often sources of oozing and should be carefully checked. Sometimes blood running down into the pericardial cavity makes it difficult to identify cardiac bleeding points. It may therefore be advantageous to check the edges of the sternum, the remnant of the thymus, the suprasternal notch and the edges of the pericardium first. Bleeding from the heart is usually controlled with fine adventitial polypropylene sutures. Troublesome surface bleeding after reoperations, bleeding from the area of dissection of the Blalock–Taussig shunt, or bleeding from the myocardium close to a major coronary artery can be controlled by application of collagen with fibrin seal (Tisseel) (Stark and de Leval, 1984). Other agents include Gelfoam/thrombin and more recently Flowseal (Oz *et al*., 2000). Considerable decreases in postoperative bleeding have been observed in patients who received aprotinin (Trasylol, Bayer) (van Oeveren *et al*., 1987; Elliot and Allen, 1990).

The pericardium is usually not closed. In patients in whom the chance of reoperation is considered high (aortic valvotomy in infants, conduits, valves), use of a thin-walled polytetrafluoroethylene (Gore-Tex) membrane to facilitate the safe reopening of the sternum has been advocated (Imai *et al*., 1978; Sakamoto *et al*., 1978). The membrane is sutured with a fine polypropylene suture to the left edge of the pericardium. On the right side, single stitches attach the membrane to the mediastinal substernal tissue. It is important to cut the membrane to the appropriate size and to partially close the sternal retractor, so that the membrane does not wrinkle. However, it has been noticed that at reoperation the membrane can be very adherent to the surface of the heart. This may make the identification of epicardial coronary arteries more difficult.

When haemostasis has been achieved, the sternum is closed with stainless steel wires (2-0 for neonates, size 1 for infants and size 4 for older children). Alternatively, Ethibond synthetic sutures can be used (**Figure 15.10**). The deep fascia is closed with a continuous suture of

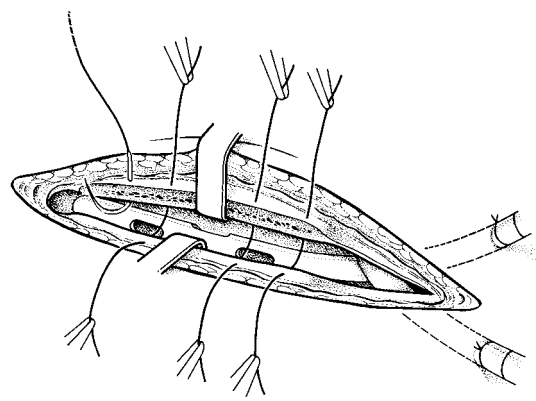


Figure 15.10

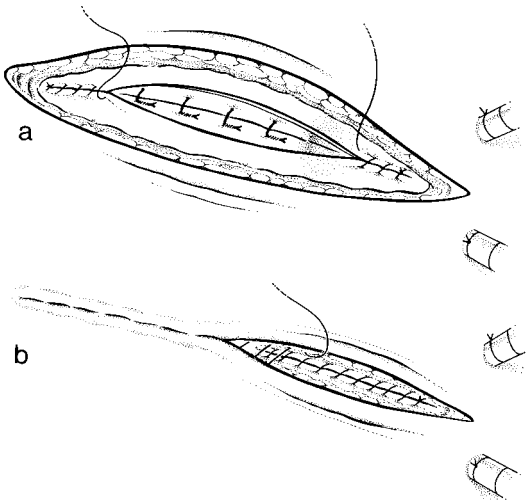


Figure 15.11

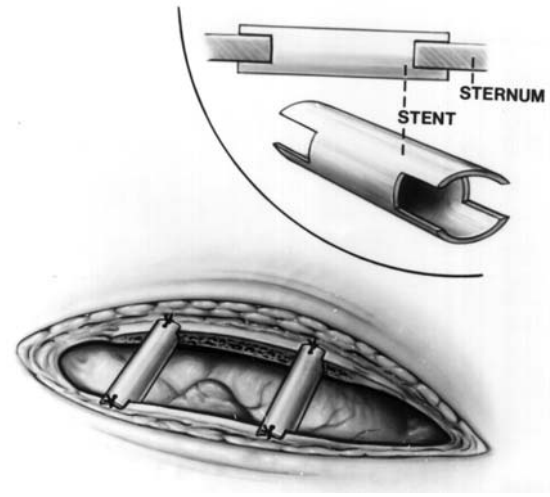


Figure 15.12

2-0 or 3-0 Dexon or Vicryl (**Figure 15.11a**). Continuous 3-0 or 4-0 Dexon or Vicryl is used for subcutaneous tissue. The skin is closed with subcuticular Dexon or Vicryl (**Figure 15.11b**). In reoperations, the authors may use nylon single stitches. The wound is painted with betadine, dried and covered with dressings for 24 hours.

On occasion, the sternal closure is not tolerated at the end of complex neonatal operations without causing major haemodynamic embarrassment. Also on occasion, the sternum cannot be closed safely, even in older children, after prolonged complex repairs. If there is a possibility that the sternal closure would not be tolerated, one or two stents tailored from polyvinyl chloride tubing are placed between the two halves of the sternum and sutured to the bone (**Figure 15.12**) (Shore *et al*., 1982). Polymeric silicone (Silastic) membrane can be sutured to the edges of the skin to provide watertight closure. Stenting of the sternum provides stability of the chest; physiotherapy can safely be given. After the patient improves, stents can be removed and the chest closed in the intensive therapy unit. This is usually possible after 1–4 days, when catecholamine requirements decrease, atrial pressures come down and blood gases remain stable.

Between January and June 2004 we used delayed sternal closure in 29 of 187 patients undergoing open heart surgery (15%). The majority of patients were neonates. Average time to closure was 3 days. There was no increased incidence of infection and delayed closure in ITU was performed without complications.

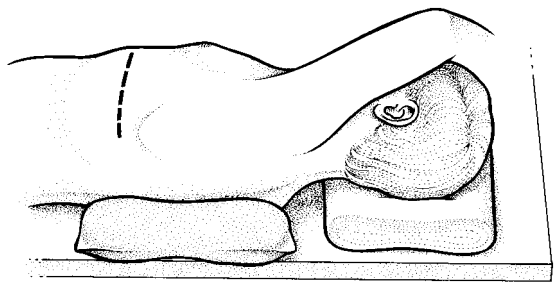
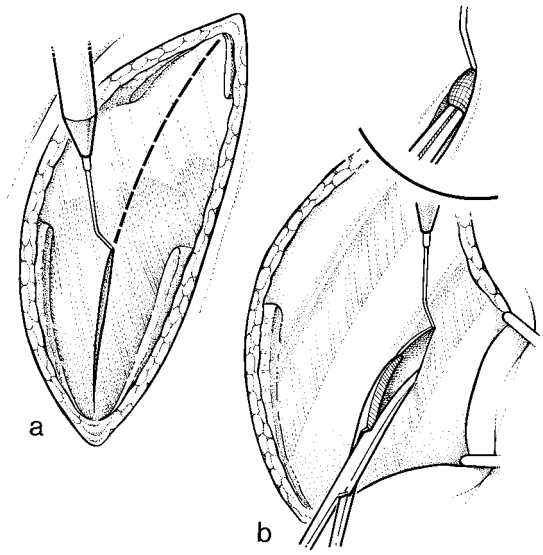


Figure 15.13

POSTEROLATERAL THORACOTOMY

The patient is secured in the lateral position, with midportion of the chest slightly elevated (**Figure 15.13**) and a tape over the hips. It is advantageous if the patient lies slightly across the operating table with shoulders slightly closer to the surgeon, because in this position the ribs run more horizontally. The length of the skin incision depends on the particular lesion; for example, it is shorter for persistent ductus arteriosus and longer for the full exposure needed for coarctation of the aorta or for systemic–pulmonary shunts. It is important to stay away from the nipple and the area of the breast because the future development of the breast may be compromised by the incision. Usually a straight incision is adequate (**Figure 15.13**). Curving the incision upwards above the angle of the scapula is not necessary in children. The scar grows with the



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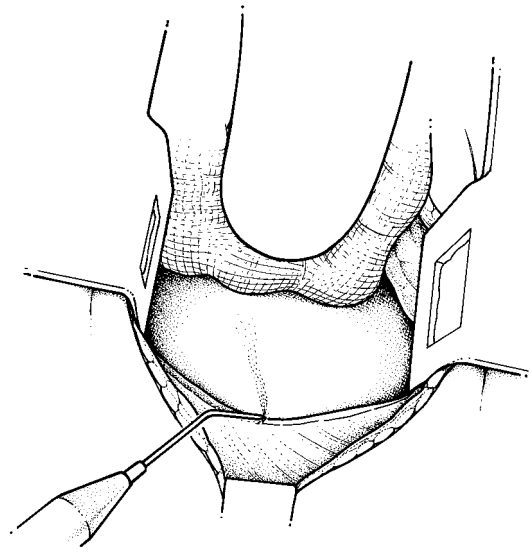


Figure 15.15

child, and the top extension becomes very evident in later years. The skin is incised with a knife, and subsequent layers are opened with the point of a diathermy (**Figure 15.14a**). The muscles are incised carefully in layers (pectoralis major, latissimus dorsi). The serratus is often preserved. With the appropriate current setting of the diathermy, the vessels can be coagulated without the need for ligation, except for coarctation of the aorta in older children. We do not use the subperiosteal approach in children; instead, we open the intercostal space between two ribs with a diathermy (**Figure 15.14b**), staying close to the upper border of the lower rib. Intercostal muscles and ribs are spread with artery forceps. When the pleura is entered, a small, moist swab on an artery forceps is inserted, and the rest of the intercostal muscles are opened with the tip of diathermy pressed against this swab (**inset, Figure 15.14**). Very quick, bloodless and efficient opening of the chest is thus achieved. Posterior extension toward the vertebral column is best achieved from inside after the Finochietto retractor is placed and the lung is held medially (**Figure 15.15**).

Closure of the thoracotomy is simple. After the procedure is completed and haemostasis is secured, the edges of the thoracotomy are again checked for bleeding. It is important to place the drain through a subcutaneous tunnel so as to avoid air leak through the thin chest wall after removal of the drain. Appropriate gauge pericostal stitches of Vicryl or Ethibond are then inserted (**Figure 15.16**). Usually three or four stitches are sufficient. Before the pericostal stitches are tied, a

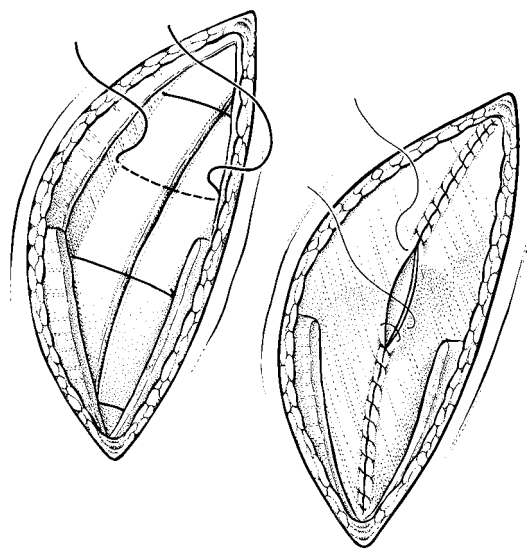


Figure 15.16

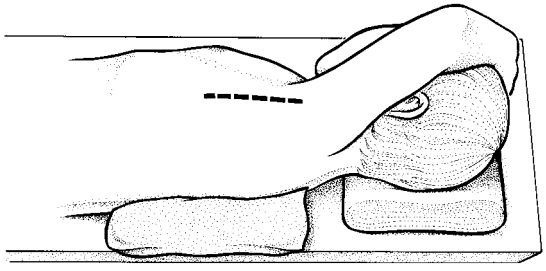
long-lasting local anaesthetic may be injected posteriorly into three or four pericostal spaces for postoperative analgesia. Both muscular layers are then closed with running stitches of Vicryl or Dexon (**Figure 15.16**). A running stitch is used for subcutaneous tissue. The skin is closed with subcuticular Dexon. In reoperations, the authors may use interrupted nylon stitches.

MUSCLE-SPARING INCISION

In neonates and small infants, an alternative incision can be used for thoracotomy (Browne, 1952). The position of the patient is the same as that chosen for a posterolateral thoracotomy, except that the arm on the side of the thoracotomy is placed over the head (**Figure 15.17**). A vertical skin incision from the middle of the axilla runs down parallel to the posterior border of the axilla. The edges of the pectoralis major and latissimus dorsi are identified and retracted. The serratus anterior is divided and then retracted backwards with a scapula. The pectoralis muscles are dissected from the chest wall and retracted, and the cartilages of the third and fourth ribs are thereby exposed. The ring retractor is then placed over the chest with one blade retracting the pectoralis muscle and the other the latissimus dorsi and serratus anterior (**Figure 15.18**). The chest is then opened through the 3rd–4th intercostal space and the ribs are spread with an adjustable screw retractor (**Figure 15.19**). Closure of this incision is simple. After the pericostal sutures are placed, the serratus anterior is reattached with Dexon, and the subcutaneous tissue and skin closed as with other incisions. This incision yields an excellent cosmetic result. The scar is less visible and is almost completely obscured by the arm in adduction.

BILATERAL THORACOTOMY

Bilateral anterolateral thoracotomy with transection of the sternum provides excellent exposure of the heart. Surgical access to both pleural spaces is enhanced. The patient is positioned on the operating table in the supine position and the clamshell incision is made (**Figure 15.20**). The pectoralis major and the first insertions of the serratus anterior are divided with diathermy. The right side of the chest is opened by means of the same technique as that outlined for posterolateral thoracotomy. The internal mammary artery and vein are doubly ligated and divided (**Figure 15.21**). The left side of the chest is then opened, and the sternum is transected with a saw or, in infants, with scissors. Two Finochietto retractors are then placed. Further steps, such as opening of the pericardium and dividing the two lobes of the thymus, are identical to those described for median sternotomy. When the bilateral thoracotomy is closed,



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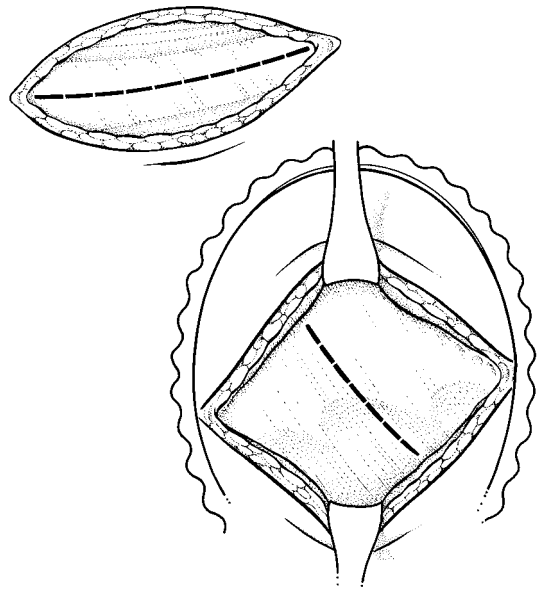

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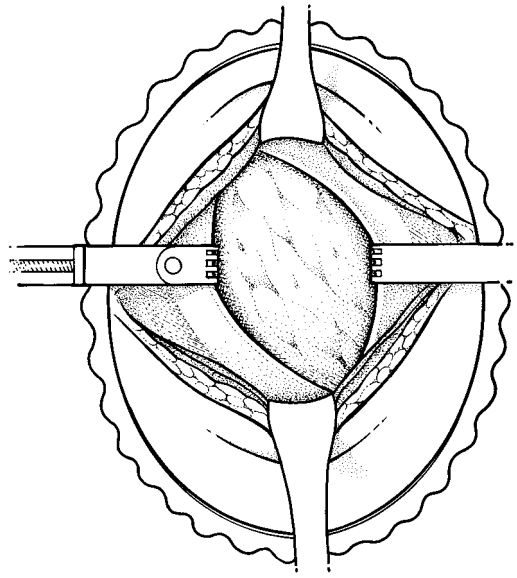

Figure 15.19

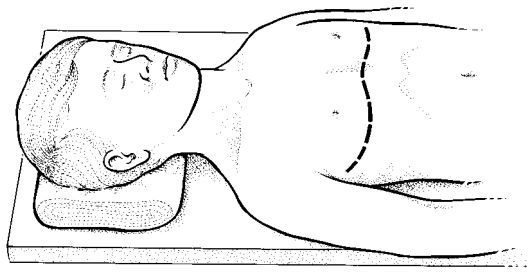

Figure 15.20

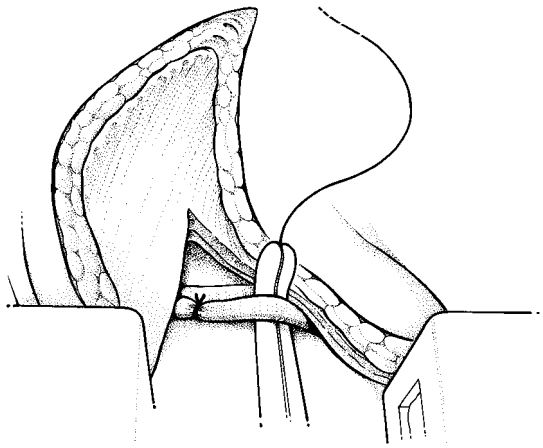

Figure 15.21

pericostal stitches are used for both sides of the chest. The sternum is approximated with sternal wires.

ENDOSCOPIC APPROACH

Today some procedures are performed using video-assisted approach. It is an alternative to surgery. The aim is reduction of invasiveness, improved postoperative recovery and shorter hospital stay. For persistent ductus arteriosus, see Chapter 19. Atrial septal defect closure and mitral valve repair (in adults) has been described from this approach (Morgan *et al*., 2004; Casselman *et al*., 2003). A learning curve may present a problem in view of the outstanding results achieved by conventional surgery.

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Part II
Surgical Procedures

16

Systemic-to-Pulmonary Artery Shunts

C. van Doorn and M. R. de Leval

The concept of improving systemic oxygen saturation in patients with cyanotic congenital heart disease and diminished pulmonary blood flow by creating a shunt between a systemic artery and the pulmonary artery was introduced clinically by Blalock and Taussig in 1945. This was followed, 1 year later, by the description of a direct anastomosis between the descending aorta and left pulmonary artery by Potts *et al*. (1946). In 1962, Waterston described a technique that consisted of an anastomosis between the ascending aorta and the right pulmonary artery. The use of prosthetic material for the construction of systemic–pulmonary artery shunts was reported by Redo and Ecker in 1963. Whereas these anastomoses are arterial in nature, Glenn and Patino (1954), following the work of Carlon and colleagues (1951), described an experimental veno-arterial shunt for certain cyanotic congenital heart defects.

INDICATIONS

The need for shunt operations has decreased with the increasing number of intracardiac repairs performed in infancy. The shunts are, however, still indicated when the outcome after primary repair is less successful than that after the two-stage approach, and for a number of complex cyanotic heart defects that are not correctable. To this end, advances in the surgical management of hypoplastic left heart syndrome have led to a renewed interest in systemic–pulmonary artery shunts (Norwood *et al*., 1983).

DISADVANTAGES

The Waterston anastomosis proved to provide satisfactory palliation in small infants. However, it is rarely used today because of the relatively high incidence of distortion of the right pulmonary artery and the risk of creating too big a shunt, which results in the development of congestive heart failure and, later, pulmonary vascular disease. The Potts shunt has been almost completely abandoned because of the same risks and because of difficulties in closing the anastomosis at the time of intracardiac repair. The classical Blalock–Taussig shunt on the opposite side of the aortic arch remains a good operation. However, the modified Blalock–Taussig shunt (MBT) has been our shunt of choice since the late 1970s (McKay *et al*., 1980). Although the MBT shunt continues to provide satisfactory early palliation, it is still associated with important shunt-related morbidity.

If the pulmonary arteries are small, peripheral shunts are likely to produce distortion and peripheral stenoses of the pulmonary arteries. In those circumstances, central shunts are preferable. The transsected small main pulmonary artery can sometimes be anastomosed onto the side of the ascending aorta (Watterson *et al*., 1991) or a prosthetic interposition shunt can be placed between the ascending aorta and main pulmonary artery. Those shunts are described in Chapter 30. Systemic–pulmonary artery shunts that form part of the first stage of palliation for hypoplastic left heart syndrome are detailed in Chapter 41.

SURGICAL TECHNIQUE

Classical Blalock–Taussig Shunt

When the aortic arch is on the left, the patient is positioned with the right side up, and a right lateral thoracotomy incision in the fourth intercostal space is made. The pulmonary artery is dissected first. To mobilize the pulmonary artery, the lung is retracted posteriorly and inferiorly, so that the artery is placed under a little tension. The periarterial plane of cleavage, which results in an easy anatomical dissection, is found close to the vessel wall. In severely cyanotic patients, numerous small collateral vessels must be cauterized, with care taken not to injure the phrenic nerve. It is important to identify the main branch of the right pulmonary artery rather than its upper lobe branch. It is easy to overlook the lower branch, which thereby causes

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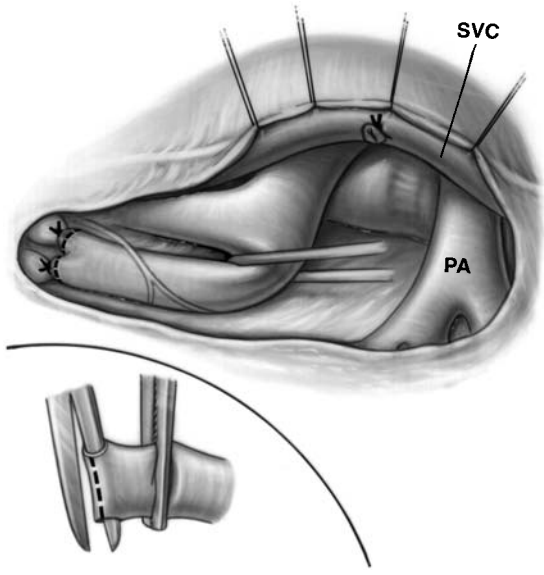

Figure 16.1

the shunt to be constructed into the upper lobe artery (Kirklin and Karp, 1970).

The dissection is then carried centrally well beneath the superior vena cava to free an adequate length of the pulmonary artery and peripherally to include the proximal branches (**Figure 16.1**). A vessel loop is passed around the upper and a further one around the lower branch of the pulmonary artery. They are passed by the subtraction technique, to make sure that all branches are encircled but the branches are not occluded until the pulmonary artery anastomosis is started. The mediastinal pleura is incised from the pulmonary artery up to and along the subclavian artery. The azygos vein and any venous branches crossing the subclavian artery (usually the superior intercostal vein) are divided between ligatures. Obviously, in a case of azygos continuation of the inferior vena cava, the azygos vein—which under these circumstances is large—is not divided. A groove is then created by blunt dissection between the trachea and the superior vena cava, taking care to cauterize the small blood and lymphatic vessels often present in that area.

After careful identification of the vagus nerve and its recurrent branch, the subclavian artery is dissected (**Figure 16.1**). Small sympathetic nerve fibres usually loop around the subclavian artery and may inadvertently be divided; this may cause Horner's syndrome. The most accessible portion of the vessel is dissected, and a vessel loop is placed around it for traction to expose the branches, which are doubly ligated and divided separately. The following branches often arise close together: the vertebral artery, the internal mammary artery, the thyrocervical trunk, the costocervical trunk and the dorsal scapular artery. The subclavian artery is tied distally, and a straight vascular clamp is placed across its midportion. The artery is divided just proximal to the distal ligatures. Right-angled forceps are then passed beneath the vagus and recurrent nerves to grasp the end of the subclavian artery. The clamp is released, and the artery is drawn through the loop of the recurrent nerve. The vascular clamp is then reapplied, and the lumen of the artery is flushed with heparinized saline. The subclavian artery is dissected to its origin, and the carotid and innominate arteries are freed in order to provide even more mobility. The subclavian artery is then turned down, and its distal end trimmed and cleaned.

With good mobilization of the innominate and carotid arteries and a deep groove between the superior vena cava and the trachea, it is possible in most instances to sever the subclavian artery at the level of its first branch, so that the end of the divided vessel can be spatulated (**inset, Figure 16.1**). This is particularly important in small infants, as an adequately sized stoma is needed for the anastomosis. Further mobility of the structures to be approximated can be gained by dividing the inferior pulmonary ligament. This is accomplished with diathermy. With traction on the main branch of the pulmonary artery, a straight vascular clamp is applied to the main pulmonary artery as medially as possible with the distal vessels snared. Alternatively, a C-clamp can be placed over the upper border of the pulmonary artery. Heparin 100 IU/kg body weight is given intravenously before the pulmonary artery is occluded. A longitudinal incision is then made on the superior aspect of the pulmonary artery (**Figure 16.2**).

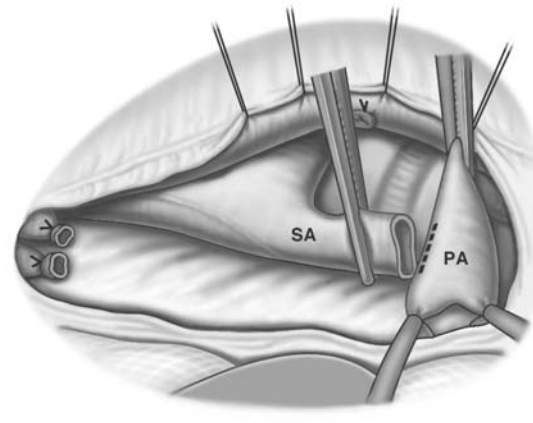

Figure 16.2

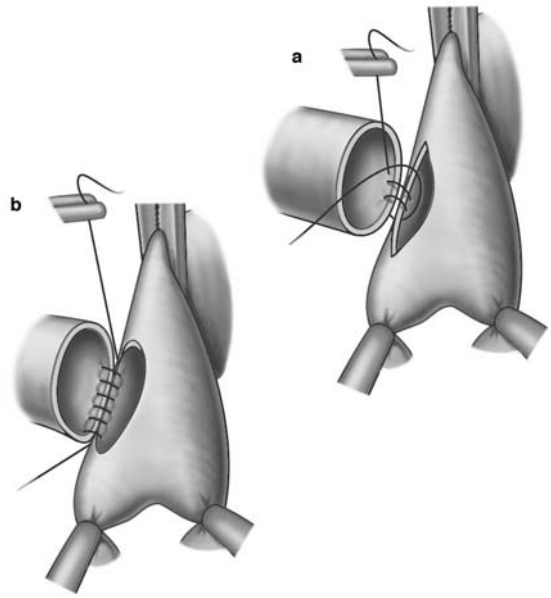

Figure 16.3

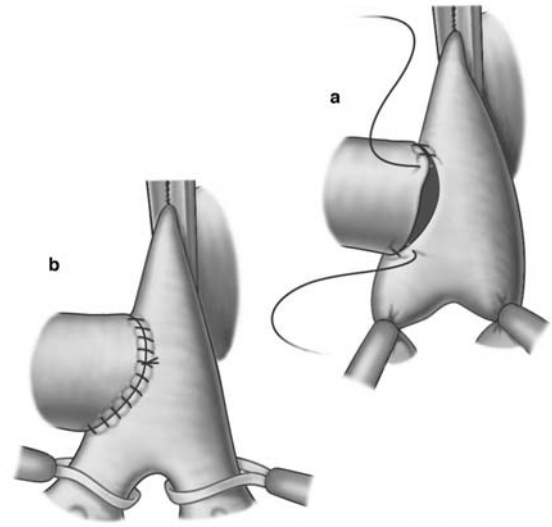

Figure 16.4

The anastomosis is constructed with a continuous 6-0 or 7-0 polypropylene (Prolene) suture, starting at the medial end of the incision and running the posterior suture line first (**Figure 16.3**). When the suture line is completed, the snares are released from the distal pulmonary arteries. The clamp on the proximal pulmonary artery is released, and finally the clamp on the subclavian artery is also released (**Figure 16.4**). A continuous thrill must be felt on the pulmonary artery when the shunt is completed. When heparin is used, it is not reversed with protamine. Systemic heparinization is used for a further 8–12 hours to reduce the risk of early shunt thrombosis.

Closure of Functioning Blalock–Taussig Shunts

Shunts are usually closed at the time when complete repair or further palliation is performed. After median sternotomy, the shunt is exposed. On the right side it is often safe to dissect the shunt before heparinization. On the left, dissection is often more difficult and it is usually safer to go on cardiopulmonary bypass with normothermia or moderate hypothermia to keep the heart beating until the shunt is controlled.

When there is a right-sided Blalock–Taussig shunt, the thrill is felt on palpation between the superior vena cava and the ascending aorta. The posterior pericardium is opened vertically about 1 cm above the right pulmonary artery to avoid dense scar tissue surrounding the site of the anastomosis. The right subclavian artery is

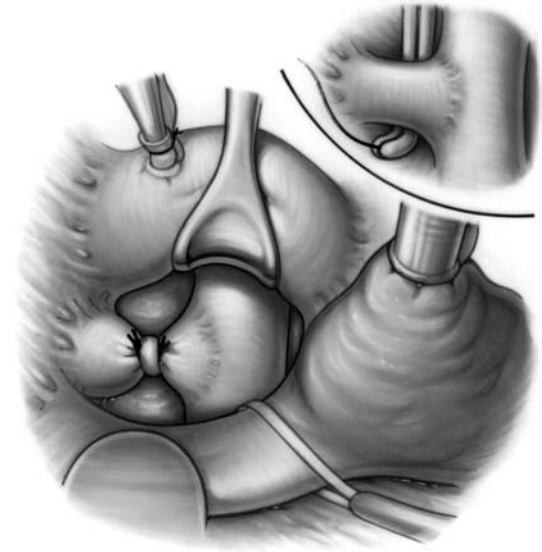

Figure 16.5

cleanly dissected from the mediastinal tissues, and right-angled forceps are passed beneath the artery to grasp and withdraw ligatures of heavy silk, which are tied after initiation of cardiopulmonary bypass (**Figure 16.5**). Alternatively, the shunt can be occluded with one or two Ligaclips or transsected and both ends oversewn.

A functioning left-sided subclavian–pulmonary artery anastomosis may be dealt with from three different

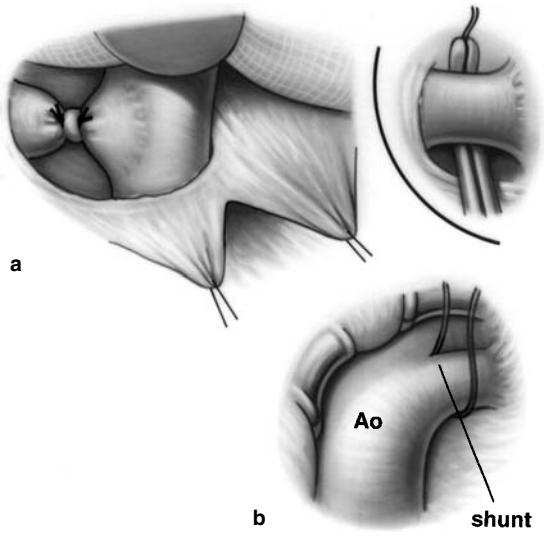

Figure 16.6

approaches. The first involves control of the shunt at the superior aspect of the left pulmonary artery. The other approach consists of dissecting the subclavian artery 1–2 cm above the left pulmonary artery. This requires an extrapericardial and often trans-pleural dissection of the subclavian artery, which has to be identified and encircled (**Figure 16.6a**). The proximity of the left phrenic nerve, which can easily be injured during dissection, and persistent haemorrhage from numerous small collateral vessels are potential hazards of this method. The third approach consists of dissecting and encircling the left subclavian artery immediately distal to its origin from the innominate artery (right aortic arch) or the aorta (left aortic arch). The aortic arch is exposed and dissected behind the innominate vein, which is retracted anteriorly and superiorly. The dissection is carried laterally along the appropriate vessel, giving way to the subclavian artery. When this artery is positively identified and distinguished from the carotid artery, a heavy ligature is passed around the subclavian artery, which is ligated (**Figure 16.6b**) or the shunt is occluded with Ligaclips once cardiopulmonary bypass is started.

Potts Shunt and Waterston Shunt

In the current era, we do not recommend the use of the Potts or the Waterston shunt. For the interested reader, a description of the operation can be found in the original papers (Potts *et al.*, 1946; Waterston, 1962).

The surgeon may, however, still be called upon to close a Waterston shunt. A detailed description of these operations is therefore included in this chapter. Catheter-based interventions may also be useful to control these shunts and the input of an interventional cardiologist should be considered, either at the time of surgery or as a separate procedure.

Closure of Waterston Shunt

Kinking, stenosis and obstruction of the right pulmonary artery at the level of the anastomosis are well documented complications of the Waterston shunt. Cardiac catheterization and selective pulmonary angiography or magnetic resonance imaging are required before complete repair is performed, in order to demonstrate the severity of the distortion of the right pulmonary artery, if present. Preparation for cardiopulmonary bypass proceeds as usual; care is taken to cannulate the ascending aorta near the origin of the head vessels, in order to retain enough length to place the aortic clamp between the cannulation site and the shunt. Femoral or iliac vessel cannulation is rarely necessary. Immediately after the institution of cardiopulmonary bypass, the shunt is controlled either by direct occlusion or by occlusion of the pulmonary arteries. When the desired level of hypothermia is reached, and with the shunt still occluded, the aorta is cross-clamped immediately proximal to the cannulation site, and cold cardioplegic solution is then infused into the root of the aorta.

In the absence of any distortion of the right pulmonary artery, a small transverse aortotomy is made over the stoma of the aorto–pulmonary anastomosis, which can be closed with running sutures securely fixed into the anastomosic junction (**Figure 16.7**).

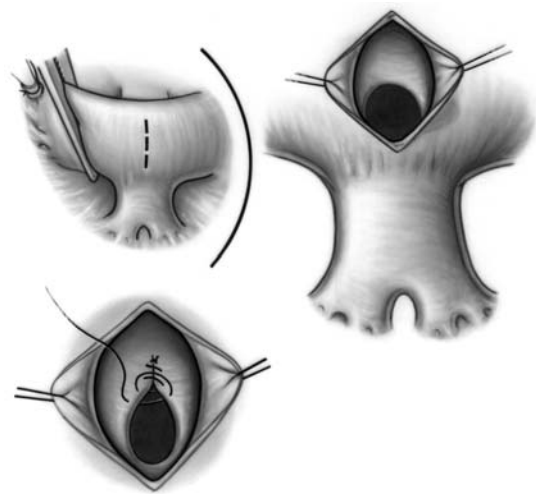

Figure 16.7

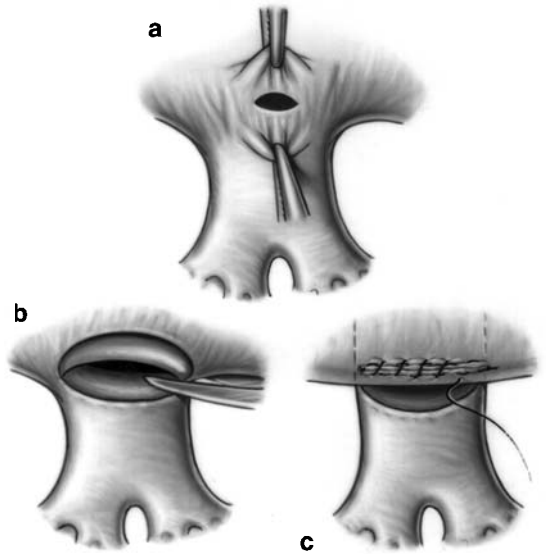

Figure 16.8

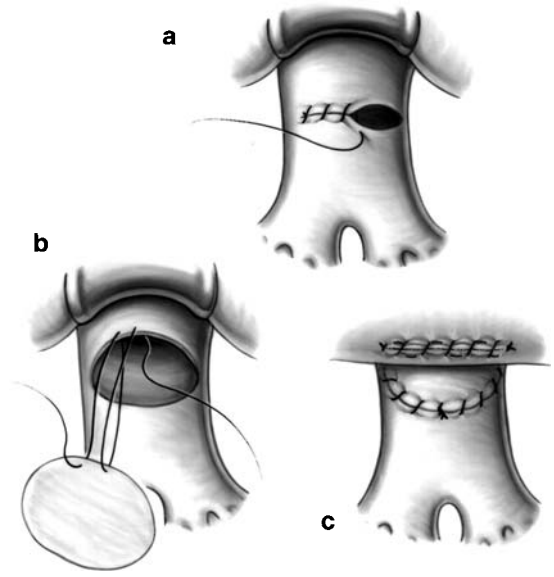

Figure 16.9

In patients in whom the right pulmonary artery is distorted or its proximal portion is partly occluded, the artery must be reconstructed. An incision is made in the aortic wall 1–2 mm above the suture line with the pulmonary artery (**Figure 16.8a**). The right pulmonary artery is then carefully dissected with scissors and completely detached from the aorta (**Figure 16.8b**). The defect in the aorta is either closed directly (**Figure 16.8c**) with a running everting mattress suture, reinforced with an over-and-over 4-0 or 5-0 monofilament suture, or with a small patch. The defect in the pulmonary artery can be closed transversely by direct suture (**Figure 16.9a**) or a small patch is frequently used to enlarge this portion of the pulmonary artery (**Figure 16.9b,c**). This patch is secured with a continuous monofilament suture. We currently recommend detachment of the pulmonary artery from the aorta in all patients with a previous Waterston shunt.

In a small number of patients, the blood from the Waterston shunt is directed almost exclusively to the right pulmonary artery; this may result in the development of a proximal hypoplastic segment, which can extend to the bifurcation of the pulmonary artery. For such cases, transection of the ascending aorta can greatly facilitate the exposure and reconstruction of the pulmonary artery (Yamamoto *et al*., 1976). The hypoplastic segment of the right pulmonary artery is incised (**Figure 16.10**) and then enlarged with a pericardial or a xenograft patch. When the reconstruction is completed and haemostasis is achieved, the aorta is re-anastomosed end-to-end (**Figure 16.11**). Because the

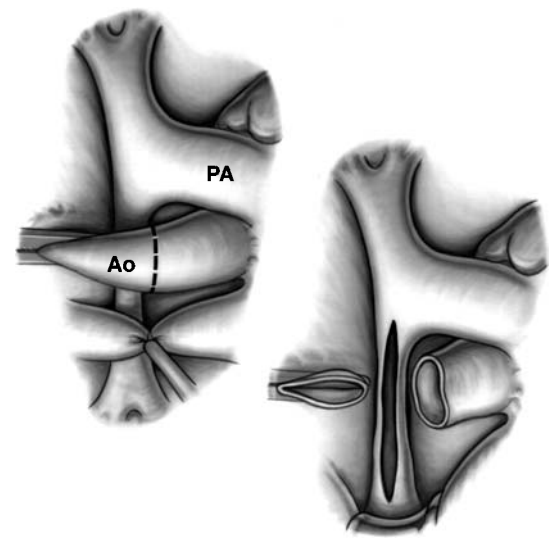

Figure 16.10

reconstruction of the aorta prolongs the cross-clamp time, we use this technique only in exceptional cases.

Prosthetic Systemic–Pulmonary Artery Shunt

Various shunts connecting the aorta with the main (central) or peripheral pulmonary arteries by using woven Teflon or Dacron prostheses have been

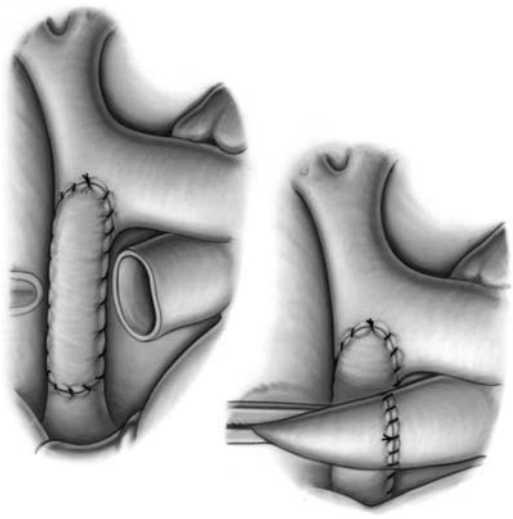

Figure 16.11

described since 1963 (Klinner *et al*., 1962; Redo and Ecker, 1963). In the past, their main disadvantage was the high incidence of thrombosis, particularly when small grafts were used. Advances in the production of arterial prostheses with long-term patency rates, even in those with small diameters, renewed the interest in prosthetic shunts (Gazzaniga *et al*., 1976). When an aorto–pulmonary shunt is required during infancy, a small graft must be used in order to avoid pulmonary oedema. This, of course, raises the question of inadequacy of these shunts as the child grows. The modified Blalock–Taussig shunt was developed with the assumption that a subclavian–pulmonary artery shunt offers adequate palliation for all ages (McKay *et al*., 1980; de Leval *et al*., 1981). Initially, a graft of expanded polytetrafluoroethylene (thin-walled Gore-Tex), was anastomosed end-to-side to the undivided subclavian artery and end-to-side to the pulmonary artery. This was with the assumption that the flow would be limited by the diameter of the subclavian artery and that the flow would increase with growth (de Leval *et al*., 1981). This, however, has never been demonstrated. Currently, shunts of 3.5–4 mm diameter are used for neonates.

Modified Blalock–Taussig Shunt via Lateral Thoracotomy

The shunt can be performed on either side of the aortic arch. We prefer to perform the shunt on the side of the innominate artery because access to the vascular structures is generally easier on the opposite side of the aortic arch. The added advantage is that a right Blalock–Taussig shunt is easier to take down, and there is less risk of injury to the phrenic nerve, compared with the left Blalock–Taussig shunt.

Although we prefer the right-sided shunt, here we describe the shunt on the left for a case of left aortic arch, as it is technically more demanding. A left thoracotomy in the fourth intercostal space is performed, and the left lung is retracted inferiorly and posteriorly. The pulmonary artery is identified at the upper border of the hilum. When the pulmonary artery is hypoplastic and very small, numerous collateral vessels may render dissection difficult. Care must be taken to avoid isolating the anteriorly located left upper pulmonary vein instead of the artery. If there is any doubt as to the identity of these vessels, the vessel is followed intrapericardially. The artery is freed medially to the pericardial reflection and laterally to its bifurcation. The upper and lower branches may be encircled with vessel loops, as described for classical Blalock–Taussig shunt. The superior intercostal vein is divided, and the mediastinal pleura is incised over the subclavian artery and its aortic origin (**Figure 16.12**). A vessel loop is passed around the subclavian artery, which is pulled anteriorly in order to isolate a segment of it with a C-type vascular clamp (**Figure 16.13**). A longitudinal incision is made on the lateral aspect of the subclavian artery, and an obliquely trimmed Gore-Tex graft is anastomosed end-to-side with a continuous 6-0 or 7-0 Prolene suture. A 3.5–4 mm thin-walled Gore-Tex graft is used in small infants and a 5–6 mm graft in bigger children. The length of the graft is then adjusted so that it lies straight along the mediastinum without traction on the pulmonary artery.

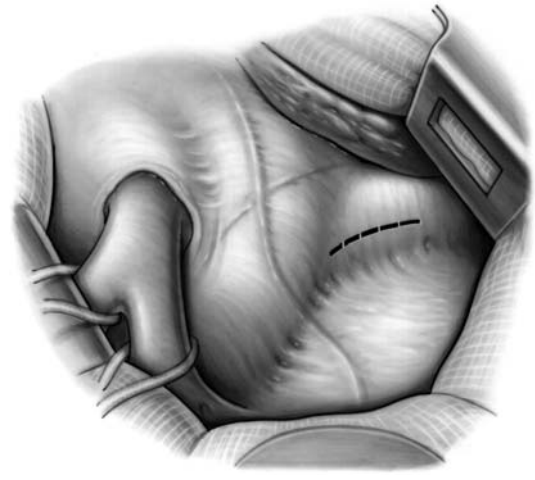

Figure 16.12

Figure 16.13

When the anastomosis between the Gore-Tex tube and the subclavian artery is completed, the end of the Gore-Tex tube is occluded with a pair of forceps and the vascular clamp temporarily released from the subclavian artery, to check patency of this anastomosis and any bleeding points. The clamp is then reapplied onto the subclavian artery. It is important not to place a vascular clamp on the Gore-Tex itself because this may damage the shunt and possibly lead to early thrombosis and occlusion of the shunt. The Gore-Tex tube is trimmed and flushed with heparinized saline to remove all blood clots. Heparin (100 IU/kg) is then given intravenously. The left pulmonary artery is clamped centrally with a vascular clamp and vessel loops are tightened around the peripheral branches. Alternatively, a C-clamp can be placed across the upper border of the pulmonary artery. A longitudinal incision is made over the upper margin of the pulmonary artery (**Figure 16.14**). The anastomosis is done with a continuous 6-0 or 7-0 Prolene suture. After completion of the shunt the vessel loops and pulmonary artery clamp are released, followed by the subclavian arterial clamp. An immediate rise in arterial oxygen saturation, with a drop in diastolic blood pressure and thrill over the shunt, are indicators of adequate shunt function. The completed shunt is shown in **Figure 16.15**.

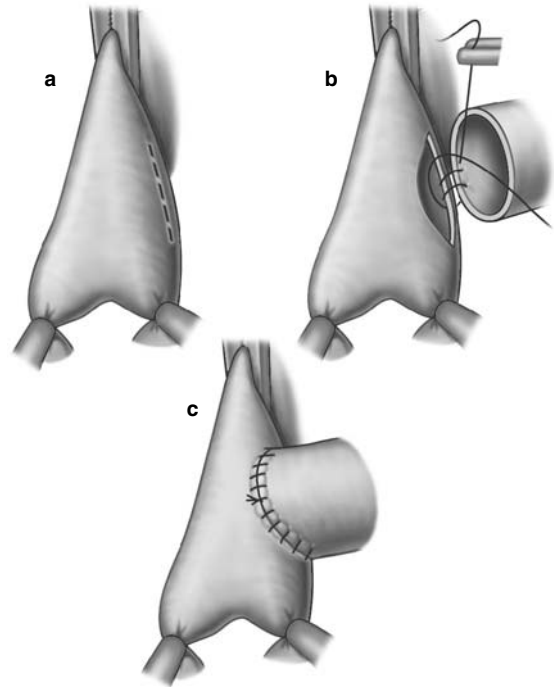

Figure 16.14

Figure 16.15

Modified Blalock–Taussig Shunt via Median Sternotomy

Some surgeons prefer to construct the shunt via a median sternotomy. This approach was originally introduced after the observation that patients with previous thoracotomies seem to form aortopulmonary collateral connections across chest wall adhesions, which may complicate subsequent Fontan operations (Odim, *et al*., 1995). This approach has the advantage that there is little distortion of peripheral pulmonary arteries, no

resultant chest deformity and in hypoxic patients the lungs are not interfered with. Additional advantages are that in selected patients a patent duct can be ligated after construction of a shunt, there is no risk of chest wall deformity, and in unstable patients cardiopulmonary bypass can be instituted quickly. A drawback to the central approach is the need for redo sternotomy at the time of future procedures.

A right-sided shunt is constructed as follows. The patient is positioned supine on the operating table. The sternum is opened and the right lobe of the thymus excised, taking care to spare the phrenic nerve. The upper half of the pericardium is opened. The right pulmonary artery is dissected between the aorta and superior vena cava. The innominate artery and its junction with the subclavian artery are dissected and encircled with slings. (**Figure 16.16**). The artery is retracted into the mediastinum and a C-clamp is placed from below across the innominate artery and rotated upwards so as to expose the undersurface of the artery. A vertical incision is made and an obliquely cut Gore-Tex tube (generally 3.5 mm for infants of 2.5–3.5 kg and 4 mm for larger infants) anastomosed end-to-side using a continuous 7-0 Prolene suture (**Figure 16.17**). The clamp is rotated back and temporarily released to check shunt patency. The shunt is washed out with heparinized saline, and heparin given as described before. A vascular clamp is then placed across the upper border of the right pulmonary artery, between the ascending aorta and superior vena cava, and the clamp gently turned downwards to improve exposure. One should be aware of the close proximity of the right coronary artery, which may be distorted by the clamp. A longitudinal incision is made over the upper border of the pulmonary artery and the distal end of the Gore-Tex tube anastomosed end-to-side (**Figure 16.18**).

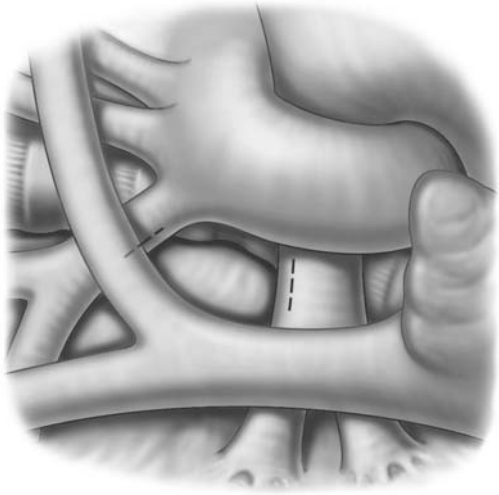

Figure 16.16

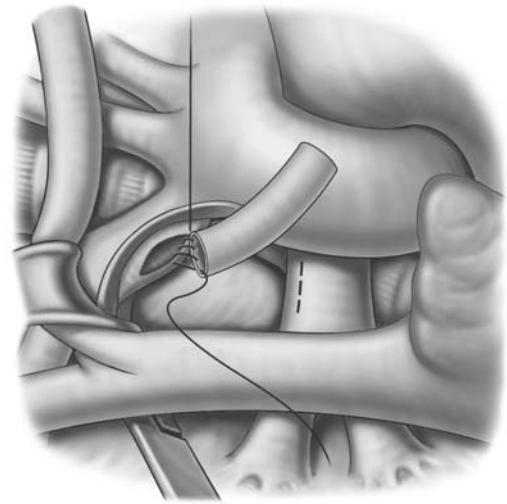

Figure 16.17

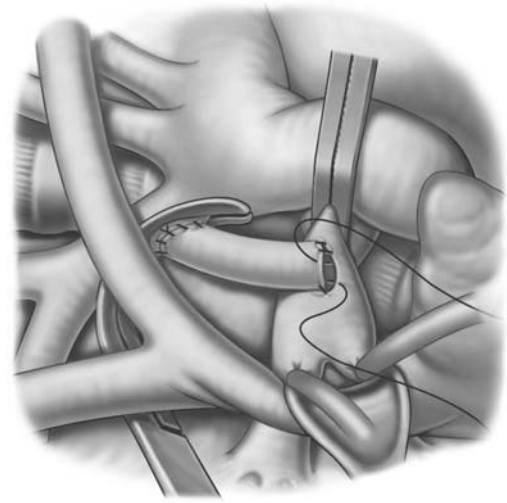

Figure 16.18

Closure of Gore-Tex Shunt

These shunts can be closed by ligation or with metal clips. Ideally, closure with division of the shunt should be performed to reduce the risk of pulmonary artery distortion in the growing child, but this may be difficult to achieve in the presence of adhesions. On the right side, the shunt can often be dissected safely through the posterior pericardium between the superior vena cava and the aorta before cardiopulmonary bypass is started. The shunt is occluded as soon as the cardiopulmonary bypass is established.

If a cavo-pulmonary anastomosis is to be performed, the shunt is disconnected from the right pulmonary artery and its distal end is oversewn, as indicated in Chapter 40. On the left side, we prefer not to dissect the interposition shunt before cardiopulmonary bypass is established. After normothermic bypass is instituted, the left pulmonary artery is then dissected distally, and the shunt is occluded just above the pulmonary anastomosis. It is obviously important to avoid ventricular fibrillation while the shunt is still open.

RESULTS

In view of long-term complications, such as distortion of the pulmonary artery and development of pulmonary vascular disease, direct aortopulmonary shunts (Waterston and Potts) are not used today. The mortality rate associated with the modified Blalock–Taussig shunt is low and does not differ from that reported for the classical Blalock–Taussig shunt. However, it is related to the severity of the underlying lesion (Mee, 1991).

In a series of 86 consecutive children, including 44 infants, the patency rate of a modified Blalock–Taussig shunt was $97.1 \pm 2\%$ at 6 months and $88.9 \pm 5\%$ at 2 years (McKay *et al*., 1980). There was a significant difference ($p < 0.05$) in shunt patency according to the shunt size. The patency rate was greater for 5 mm shunts than for 4 mm shunts. The beneficial effect of aspirin in maintaining shunt patency remains open to debate.

In a recent review of 92 infants who underwent an isolated modified Blalock–Taussig shunt between 1994 and 2003 at our Institution, hospital mortality was 3.3% (Vricella *et al*., 2004) and a further 3.3% required early shunt revision. Weight less then 2.5 kg was a predictor of acute shunt-related morbidity and late death.

Iatrogenic pulmonary artery problems remain common. In our recent review, 21% of hospital survivors developed significant shunt-related pulmonary stenosis. In patients with tetralogy of Fallot, angiography prior to definitive repair showed significantly smaller distal right pulmonary arteries in those who had received a right modified Blalock–Taussig shunt, compared to patients who had not been palliated (Gladman *et al*., 1997). Furthermore, in 33% there was angiographic evidence of pulmonary artery distortion, and shunt stenosis was common in those who had been palliated at a younger age.

An excessive serous leak through the Gore-Tex, leading to prolonged chest drainage or local seroma formation, is a recognized complication (LeBlanc *et al*., 1984), as is false aneurysm formation, which carries a risk of rupture (Sethia and Pollock, 1986).

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17

Pulmonary Artery Banding

V. T. Tsang and J. Stark

Pulmonary artery banding (PAB) was suggested by Muller and Dammann in 1952 as a palliative operation for children with a large left-to-right (L–R) shunt, such as large ventricular septal defect (VSD) or single ventricle. Since then, it has been used in the palliative treatment of congestive heart failure and also in the hope that it would prevent or arrest the further development of pulmonary vascular obstructive disease (Goldblatt *et al*., 1965; Stark *et al*., 1969).

CLASSICAL INDICATIONS

Originally, pulmonary artery banding was suggested for patients with ''functional single ventricle'', i.e. those with a large VSD or a single ventricle. With the development of open-heart surgery, symptomatic VSDs were more routinely closed during intracardiac operations. However, as the risk of VSD closure in sick infants with other associated defects remained high, pulmonary artery banding was commonly used as a palliative procedure to reduce pulmonary arterial pressure and blood flow in this age group. Nevertheless, the disadvantages of too tight a band and the potential distortion of the pulmonary bifurcation and the valve were well appreciated.

Other conditions causing increased pulmonary blood flow have also been palliated by banding: acyanotic lesions (atrio-ventricular septal defect, double-inlet ventricle, and some forms of double-outlet ventricle without pulmonary stenosis) and cyanotic lesions, including transposition of the great arteries (TGA) with VSD, some forms of tricuspid atresia, truncus arteriosus, congenitally corrected transposition with VSD, hypoplastic left heart syndrome and mitral atresia. Since early primary repair of VSD, TGA and several other lesions became the treatment of choice, pulmonary artery banding has been used less frequently. However, the latter trend was not immune to changes. Pulmonary artery banding has recently been employed to ''train'' the subpulmonary left ventricle post atrial switch with a view to an arterial switch conversion (see Chapter 36), and ''shift'' the septal configuration in the management of significant tricuspid regurgitation with failing systemic right ventricle in congentially corrected transposition (see Chapter 39). Acute PAB may lead rapidly to ventricular failure. A more gradual approach to PAB may be required to achieve a successful outcome.

CURRENT INDICATIONS

Ventricular Septal Defect

Most VSDs are submitted to rigorous medical treatment in the first instance of severe heart failure. If medical treatment fails, primary closure of such defects is our preference. There are, however, a few exceptions in which PAB is considered:

1. *Multiple VSDs* in early infancy. Surgery remains risky, although new approaches, such as double-patch sandwiching the septum (Brizard *et al*., 2004), have been reported, with encouraging results. Some of the muscular VSDs may close with muscular septal hypertrophy secondary to PAB. The authors would prefer to delay repair until late infancy with a view to undertaking closure of the ''remaining'' VSDs via the right atrium, with division of the moderator band if necessary and apical right ventricular approach if required. Left ventriculotomy is usually not necessary or advisable (Kitagawa *et al*., 1998). Intraoperative and postsurgical device closure of any important residual communications should be part of the management strategy (see Chapter 7).
2. *Coarctation of the aorta with a large VSD*. In these patients, coarctation of the aorta is repaired. If the VSD is large, three alternatives are available. First, the coarctation alone may be repaired via a left thoracotomy, and the patient continues to receive medical treatment; if the patient fails to be weaned from the ventilator, the VSD is

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closed. Second, the coarctation of the aorta is repaired, and the pulmonary artery is banded via left thoracotomy; VSD closure is then delayed for 3–6 months. Third, the VSD is closed and the coarctation is repaired simultaneously. Both procedures can be accomplished through midline sternotomy (Ungerleider and Ebert, 1987; Karl *et al*., 1992). This concomitant repair is currently our preferred approach if the VSD is technically closable and arch hypoplasia. Alternatively, coarctation is repaired from the left thoracotomy; the patient is then turned over, and the VSD is closed through midline sternotomy (Doty *et al*., 1975). The latter approach is rarely used today, but the conceptual advantage of avoiding circulatory arrest is still appreciated. Nowadays, the general opinion of completing the hypoplastic aortic arch and coarctation repair within 30 minutes at deep hypothermic circulatory arrest is considered ''safe''. Low-flow regional cerebral perfusion can also be employed to avoid circulatory arrest.

3. *VSD complicated by other congenital non-cardiac anomalies* (e.g. large omphalocoele, oesophageal atresia, diaphragmatic hernia, syndromal anomalies, premature baby with low birth weight, or VSD in severely undernourished babies and the presence of severe infection (sepsis, respiratory, skin) that cannot be brought under control because of severe heart failure). The rationale is that a non-bypass operation might alleviate the heart failure and therefore improve the general condition of the infant, so that a subsequent open-heart procedure would have a more favourable chance of success. Deciding whether to perform PAB or close the VSD may be very difficult in this group of patients. One important factor would be the experience of the team to perform open heart surgery in very young and ill infants.

Transposition of the Great Arteries

The indications for PAB for patients with TGA have changed considerably. Originally, PAB was used for patients with TGA and VSD (Carpena and Subramanian, 1974; Trusler and Mustard, 1972) and the Mustard or Senning operation and VSD closure were delayed. However, with increasing experience in neonatal surgery, the results of the arterial switch operation and VSD closure improved so much that, in several series, young age ceased to be a risk factor (Norwood *et al*., 1988; Brawn and Mee, 1988).

Therefore, at present, there is no strong indication for PAB in infants with TGA and VSD, except for occasional patients with multiple VSDs and additional severe non-cardiac malformations. However, PAB in an already cyanosed patient can be difficult, and an unrestrictive atrial communication is essential for optimal mixing. Also, care must be taken not to distort the pulmonary valve.

An interesting approach was developed by the Boston group (Jonas *et al*., 1989). Patients with simple TGA who present after the first 3–4 weeks of life undergo construction of a systemic–pulmonary artery shunt and PAB. They are kept for 7–10 days in the intensive care unit, and then an arterial switch operation is performed. The left ventricle should then be able to support the systemic circulation after the arterial switch operation is performed. In our institution, instead of PAB and staged arterial switch, we are extending the time limit (up to 2 months) by which to perform primary arterial switch, with postoperative mechanical circulatory support if required (Kang *et al*., 2004).

Another indication for PAB in TGA is severe right ventricular failure after a Mustard or Senning operation. The pulmonary artery is banded, unless the left ventricular pressure is already increased (such as in associated left ventricular outflow tract obstruction). The arterial switch operation is then performed about 12 months later, when the hypertrophied subpulmonary LV is considered adequate to sustain the systemic afterload (see Chapter 36) (Mee, 1986, 1989). However, important ventricular dysfunction post-PAB was noted in the adolescent group.

Double-inlet Ventricle and Tricuspid Atresia without Pulmonary Stenosis

In the absence of pulmonary stenosis, patients with double-inlet ventricle and tricuspid atresia present in heart failure. A Fontan-type procedure or total cavopulmonary connection (TCPC), is usually the only definitive solution. Staged approaches with a bidirectional superior cavo-pulmonary anastomosis (Glenn shunt) or a hemi-Fontan are currently not favoured for patients under the age of 3 months because the pulmonary vascular resistance is still too high. If the pulmonary blood flow is not restricted and the pulmonary arterial pressure not reduced, pulmonary resistance continues to increase. PAB is therefore indicated. It may be difficult to achieve adequate banding without causing an increase in cyanosis. These patients have major obligatory mixing of the pulmonary and systemic venous returns within the dominant ventricle. Incomplete mixing may, result in streaming, so that in some patients the amount of pulmonary blood flow may be difficult to assess from the degree of cyanosis. However, if the pulmonary artery is not constricted adequately, a Fontan-type procedure may not be possible later. For this reason, an alternative procedure may be performed.

The pulmonary artery is completely occluded and a modified Blalock–Taussig shunt is constructed. This will regulate the pulmonary blood supply and ensure that the pulmonary artery pressure is not excessive.

With concomitant transposed great arteries and the coexistence of aortic coarctation/hypoplastic aortic arch, we would favour a Norwood-type or Damus–Kaye–Stansel (DKS) procedure with a systemic–pulmonary shunt in the first instance. This is based on the potential development of a restrictive VSD with ''subaortic stenosis'' ascribed to PAB, and this may compromise subsequent Fontan strategy. However, Daenen *et al*. (2000) were unable to demonstrate these secondary changes of ventricular hypertrophy and dysplastic changes to the pulmonary valve from PAB to be a major disadvantage in the strategy of a DKS or a Fontan procedure.

Other Indications

PAB for double-outlet right ventricle, truncus arteriosus, and complete atrio-ventricular septal defect has been largely abandoned in favour of early complete repair. Under special circumstances (such as those outlined for VSD, late-presenting atrio-ventricular septal defect with pulmonary hypertension, straddling atrio-ventricular valves, and atrio-ventricular discordance), PAB may be considered.

Hypoplastic Left Heart (HLH)

The surgical concept, with bilateral PAB, preceded by endovascular stenting of the patent ductus arteriosus, is being very selectively applied as part of the Norwood strategy in the management of HLH. This new approach, together with atrial septectomy or septostomy, was first reported by Gibbs *et al*. (1993).

It has been used in very sick neonates with HLH to improve the haemodynamics before a delayed stage 1 Norwood procedure (Pizarro and Norwood, 2003). Another option being considered is to offer this approach as an alternative to stage 1 Norwood. The next combined stage would then involve a bidirectional Glenn shunt, major reconstruction of the pulmonary arteries and the aortic arch. Alternatively, this palliative approach allows the opportunity for heart transplantation after an extended waiting period because of poor availability of infant donor hearts (Akintuerk *et al*., 2002).

OPERATIVE TECHNIQUE

Approach

The approach depends on the type of underlying lesion. The authors usually use a left lateral thoracotomy through the third or fourth intercostal space. This approach enables closure of PDA and repair of coarctation of the aorta, if present. A midline sternotomy may be used for patients with complex transposition, some forms of double-outlet right ventricle and hypoplastic left heart syndrome, when banding may have to be performed, together with atrial septectomy.

We often start a dopamine infusion (5 μg/kg/min) before pulmonary artery banding if the systemic pressure is low. However, inotropes are used with caution, as they may alter the balance between the systemic and pulmonary vascular resistances. An arterial monitoring catheter is always inserted, because the changes in systemic pressure are used to evaluate the efficiency of banding together with oxygen saturation monitoring.

Technique

Left Thoracotomy

Usually, the third intercostal space is opened, and the ductus or ligamentum arteriosum is dissected and ligated (**Figure 17.1**). If coarctation of the aorta is present, it is repaired. The lung is retracted posteriorly, and the pericardium is opened in front of the phrenic nerve. It may be necessary to partly dissect the thymus off the pericardium in order to gain adequate access to both the aorta and the pulmonary artery. The exposure is further assisted by retracting the tip of the left atrial appendage out of the operating field with a fine silk ligature (**Figure 17.2**).

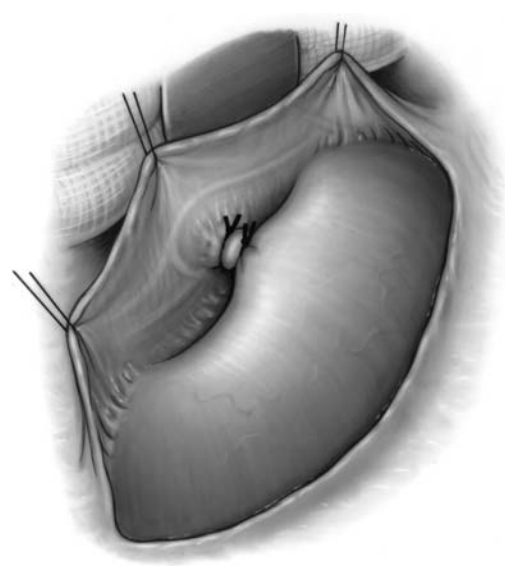

Figure 17.1

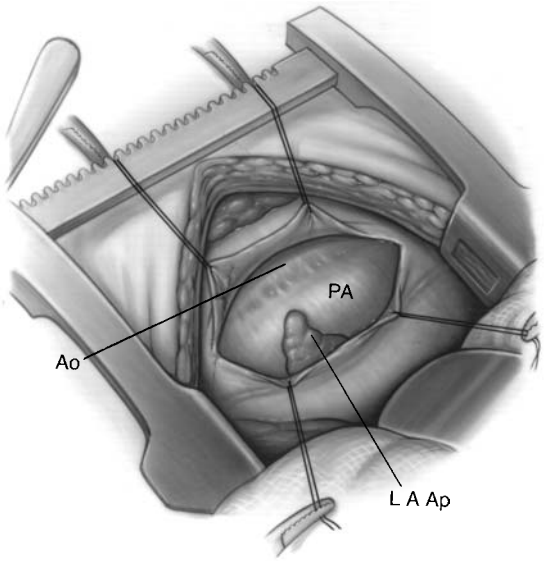

Figure 17.2

Figure 17.4

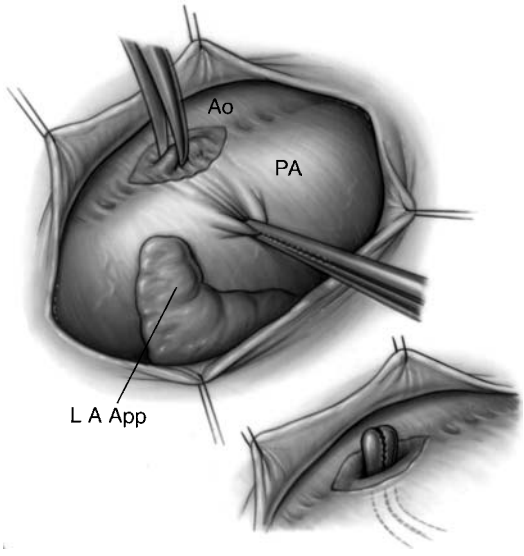

Figure 17.3

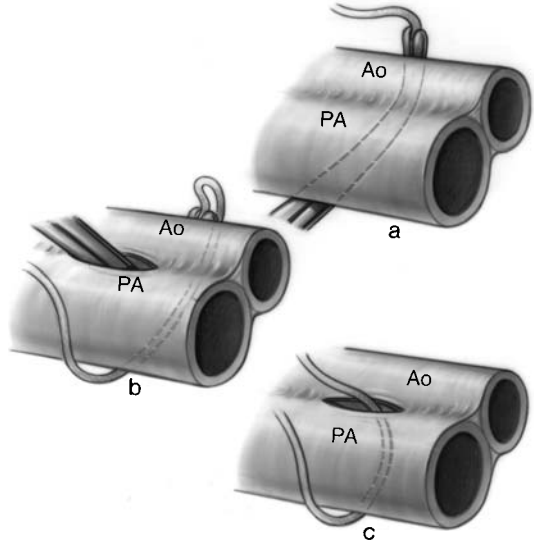

Figure 17.5

The space between the aorta and the pulmonary artery is dissected out. Care must be taken to avoid or to coagulate the small vein that runs between the two vessels. A ligature of thick (1.2 mm) plaited silk or 3 mm cotton umbilical tape is then passed around the pulmonary artery (**Figure 17.3**) with the use of a right-angled Denis Browne dissector (**Figure 17.4**). Should direct dissection of the pulmonary artery prove difficult, the ligature/tape may be passed more easily by the subtraction technique (**Figure 17.5**). A thick ''ductus'' silk ligature/tape is passed through the transverse sinus around both vessels and then brought underneath the aorta. The surgeon must carefully check to see that the ligature/tape has been passed around the main pulmonary artery and not just around the left pulmonary artery. The lungs are then released and re-expanded, and the patient

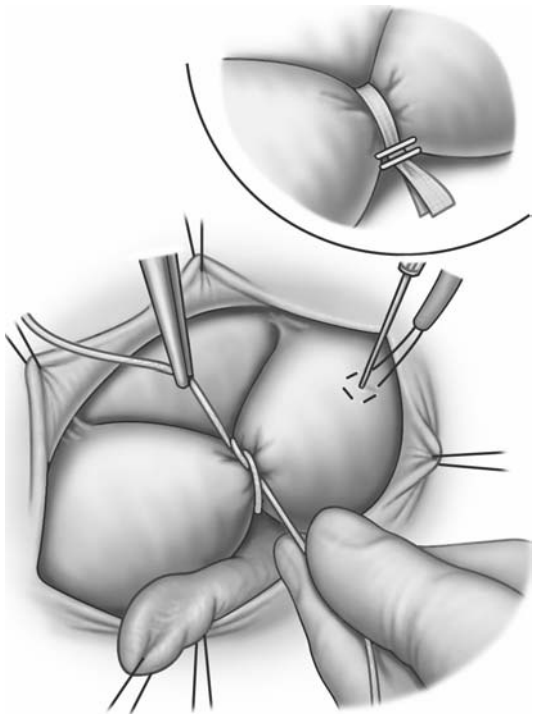

Figure 17.6

is hyperventilated for a few minutes. During the banding procedure, the patient is ventilated with 50% oxygen; blood gases are checked before and after banding, or the patient is monitored with the pulse oxymeter. A purse-string suture is placed on the distal main or left pulmonary artery. The lung is gently retracted without being compressed completely, the pressure-monitoring needle is inserted into the pulmonary artery, and the banding ligature is slowly tied (**Figure 17.6**) or the cotton tape is constricted in a step-wise fashion, using two ligaclips (**inset, Figure 17.6**).

Median Sternotomy

This technique has gained some popularity because of the advantage of ease of access, especially in the setting of complex great vessel arrangements, minimal interference with the ventilation at the time of banding, and one surgical incision for subsequent staged procedures. The superior two-thirds of the anterior pericardium is opened and the ductus or ligamentum arteriosum is always ligated. A word of caution: with the banding from the front, the band itself may be a lot closer to the right pulmonary artery than anticipated.

Anterior Extrapleural Approach

In premature infants and some small, undernourished infants, the author (J.S.) has used an anterior extrapleural approach through the third intercostal space. The infant is placed on the operating table in a supine position with a small towel underneath the left shoulder. A small incision is made just below the left clavicle (**Figure 17.7**) and the third intercostal space is opened. The ribs are spread with an edentulous gag and the pleura is pushed laterally. It is sometimes necessary to resect the left lobe of the thymus. The pericardium is then opened, and excellent access to the main pulmonary artery and the PDA is obtained (**Figure 17.8**). Banding is performed and the pericardium is then closed (**Figure 17.9**).

THE DEGREE OF PULMONARY ARTERY BANDING

The degree of constriction depends on the underlying cardiac lesion. The principle is to balance the systemic and pulmonary blood flow. Special consideration needs to be given to the unique pathophysiological features in the patient with a functionally univentricular circulation (Wessel, 1996). Based on the principles outlined by Barnea *et al*. (1998), assuming a pulmonary venous oxygen saturation of 96%, to achieve a mixed venous oxygen saturation of 70%, the appropriate aortic oxygen saturation for optimal systemic oxygen delivery is slightly greater than 80%. For banding in the context

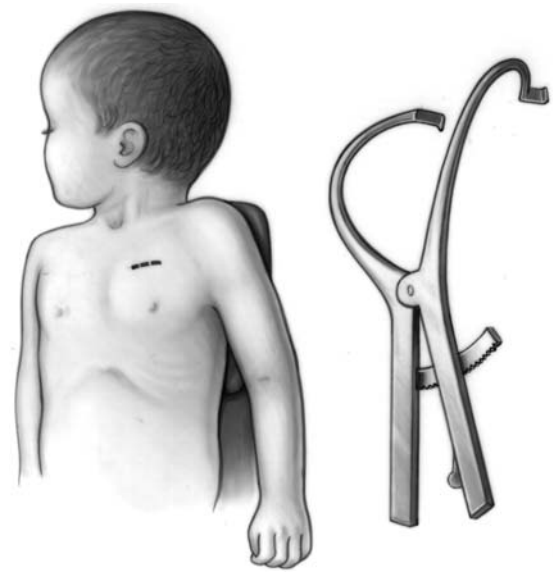

Figure 17.7

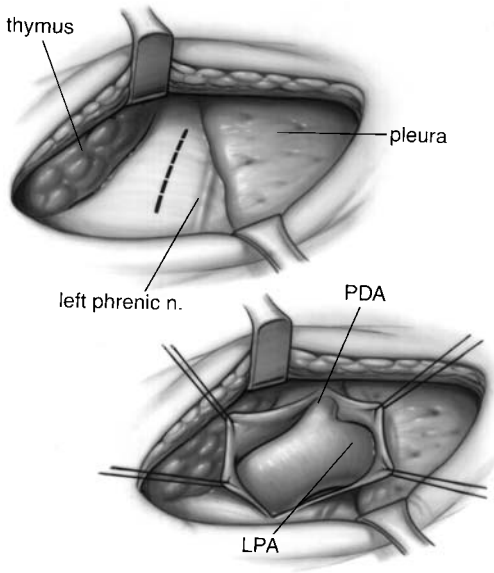

Figure 17.8

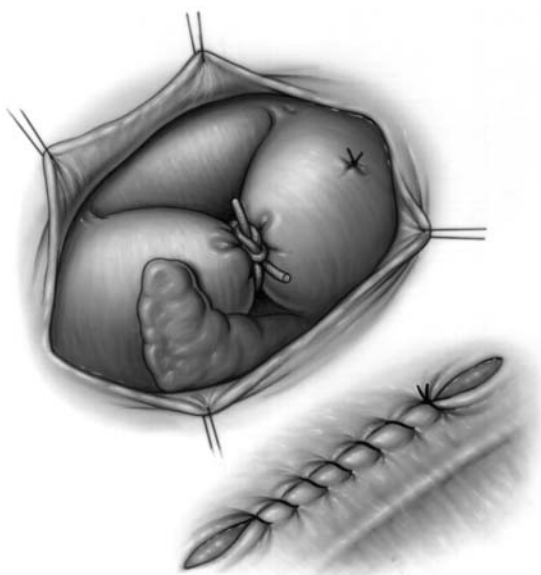

Figure 17.9

of a functional univentricular heart, refer to Special Considerations (see subsequent text).

For VSD, the aim of PAB is to reduce the pulmonary artery pressure distal to the band close to the normal systolic pressure of 30 mmHg. Another very good guide for assessing the degree of narrowing is to follow the changes in systemic pressure, pulse rate and oxygen saturation.

As the pulmonary artery is being constricted, the systemic pressure starts rising while the distal pulmonary artery pressure is falling. The optimal degree of constriction is achieved when the systemic pressure reaches a plateau, usually an increase of 10–20 mmHg without bradycardia and oxygen desaturation beyond the low 90s. Further constriction would be dangerous because it would put an extra load on the ventricles and subendocardial ischaemia may occur. Trusler and Mustard (1972) calculated the optimal length of the band from the infant's body weight by using a premarked 4 mm polytetrafluoroethylene (Teflon) band impregnated with polymeric silicone (Silastic). If the banding is done for biventricular heart, 20 mm + weight in kg is used. If done for complex cardiac malformations, it is 24 mm + weight in kg.

When the optimal degree of constriction is achieved, a second knot is placed on the ligature. A square knot on this rather thick material ensures that the ligature will not become loose and that the degree of stenosis can be maintained. Alternatively, one to two Ligaclips would secure the constriction on the cotton tape, together with a fine 6-0 Prolene mattress suture. The monitoring needle is removed from the pulmonary artery, the purse-string suture is tied, and all blood is carefully aspirated from the pericardial cavity. The band itself is secured to the proximal main pulmonary artery with two fine 6-0 Prolene sutures to prevent any band migration. The pericardium is closed with a running stitch, and a small opening, superiorly and inferiorly, is left for drainage (**Figure 17.9**).

In very small neonates and in some patients with cyanotic heart lesions, this degree of constriction may increase the cyanosis considerably. For this reason, a milder degree of banding is planned, with the systolic pressure in the distal pulmonary artery at about 40 mmHg, or about half of the systemic pressure. In cyanotic lesions, it is useful to check arterial oxygen pressure (pO_2) and saturation before and after banding. The aim is not to reduce arterial pO_2 below 30 mmHg (4 kPa).

When the arterial pO_2 is measured, it is important that the anaesthetist keeps the fraction of inspired oxygen constant (usually 0.5) and that the lungs are not excessively compressed or overventilated. If the pulmonary artery is banded in preparation for arterial switch operation in a patient with simple TGA, a 4 mm shunt is added (Jonas *et al*., 1989). However, this type of ''rapid two-stage approach'' to simple TGA presenting ''late'' is not a standard approach at our institution.

Material Used for Pulmonary Artery Banding

Since the original description by Muller and Dammann (1952), different materials have been suggested and used for banding. These include umbilical tape, knitted tubular Dacron, Teflon tape, Silastic tape and plaited silk ligature (Albert *et al*., 1961; Stark *et al*., 1969). Fishman and Roe (1971) suggested the use of an adjustable band. When selecting the material for banding, the surgeon should consider two factors: (a) the ease of its application and adjustment; and (b) the ease with which the band can be subsequently removed.

Stark *et al*. (1972) evaluated several materials used for banding (the plaited silk ligature, 3 mm and 7 mm polytetrafluoroethylene (Teflon) tape, nylon tape, and cotton umbilical tape) on young piglets. The damage to the wall of the pulmonary was related to the degree of stenosis but not to the materials used. A ligature of plaited silk, because of the ease of application, has been successfully used at our institution. Another option is the use of cotton tape with Ligaclips to adjust the tightness of the banding. The latter technique has been favoured by the authors recently because of the simplicity of banding and the ease with which the constriction is adjusted. In addition, debanding can be done by an interventional technique if indicated (see later).

Other approaches to PAB

There has been some recent interest in reducing the number of surgical interventions through the application of either an absorbable pulmonary trunk band, or a band that may be inflated at interventional cardiac catheterization.

Absorbable PAB

The concept is attractive but so far has had limited clinical impact. A successful case report of the use of an ''absorbable Polydioxanone ribbon'' as a band in tricuspid atresia to induce a restrictive VSD has been published (Bonnet *et al*., 2001). However, the integrity of the band, with the suture being absorbed or losing tensile strength prematurely, leading to high pulmonary blood flow, is always a risk.

Dilatable PAB

We have performed, in four patients, pulmonary artery constriction with cotton umbilical tape secured with Ligaclips and debanded by balloon angioplasty at the same catheter intervention as device closure of a muscular ventricular septal defect. We believe that this approach may have a useful role in the future in this carefully selected subgroup of patients, who are suitable for device VSD closure yet require initial surgical palliation (PAB).

Adjustable PAB

Fishman and Roe suggested the use of an adjustable band in 1971. Most recently, this attractive concept was radically rekindled with the application of telemetric adjustable PAB, operated with the help of an external control unit that transmits to the implanted FloWatch-PAB energy. The commands may further narrow or release the band, using a radiofrequency source (Corno *et al*., 2003). This new device may have a role in selected children who may require PAB for complex heart defects. The use of this technology may obviate the need for an early reoperation. Experience with this technique is limited.

SPECIAL CONSIDERATIONS

Transposition of the Great Arteries with Ventricular Septal Defect

It is very rare to undertake PAB for this condition. However, for completeness, the principle is described below. Before banding, the surgeon should ensure that there is an adequate interatrial communication. If there is not, it can be produced by balloon atrial septostomy. Banding in the absence of an adequate interatrial communication may increase cyanosis to an unacceptable level. For this reason, when banding cyanotic patients, the authors monitor arterial pO_2 and arterial saturation and try to avoid any reduction in PaO_2 below 30 mmHg (4 kPa).

Double-inlet Ventricle and Tricuspid Atresia

Currently we prefer to defer a Fontan-type procedure for these lesions until patients reach the age of 2–4 years. In symptomatic infants, we try to improve congestive heart failure and prevent the development of pulmonary vascular obstructive disease. A careful balance between excessive and inadequate banding may be difficult. In our practice, it is preferable to err on the side of tight banding. If the band has become too tight with growth, a bidirectional Glenn shunt or systemic–pulmonary shunt can be created before the optimal age for a Fontan-type procedure is reached. If, however, the band is left too loose, a certain degree of pulmonary hypertension may persist and may lead to the development of pulmonary vascular obstructive disease. As explained in Chapter 40, even mildly elevated pulmonary arteriolar resistance may preclude success of the Fontan operation.

Some of these babies with increased pulmonary blood flow may have coexisting systemic outflow obstruction

at the subaortic level (e.g. restriction at the VSD or within the rudimentary right ventricle in double-inlet left ventricle and discordant ventriculoarterial connections). Under such circumstances, the subaortic stenosis may be exaggerated by the PAB.

Absent Left Atrio-ventricular Connection

If PAB is considered, atrial septectomy is indicated (see Chapter 18). Balloon atrial septostomy is often not adequate in the context of an obligatory L–R shunt. Determining the optimal PAB can be difficult after cardiopulmonary bypass. One should aim for oxygen saturation in the low 70s. The lungs would improve post-bypass and the PaO_2 will rise. However, redo PAB may become necessary.

Banding in Very Small/Young Infants

Very small undernourished infants require special consideration. If banding is successful and heart failure is alleviated, the child may start feeding well and grow rather rapidly. Under these circumstances, the band, which was acceptable at the time of operation, may become too tight. It may be necessary to proceed with an early repair and debanding.

COMPLICATIONS

Various complications of PAB have been described. Some are potentially avoidable with attention to surgical details. The band may cut through the pulmonary artery (Hunt *et al*., 1971; Stark *et al*., 1969). This is rare in neonates and small infants. If such cutting is acute, fatal bleeding may occur; if it is more gradual with prolonged PAB left *in situ*, the degree of pulmonary constriction may become inadequate, due to the progressively tight band eroding through the pulmonary arterial wall, and pulmonary vascular obstructive disease may develop as a result of unsuspected high pulmonary blood flow. Infection at the site of the band is another possible, serious complication.

Changes in the wall of the pulmonary artery (Berry, 1969) and the pulmonary valve (Hunt *et al*., 1971) have been reported. Freed *et al*. (1973) observed the development of left ventricular outflow tract obstruction after banding. Spontaneous closure of VSD after banding (Edgett *et al*., 1968; Stark *et al*., 1970), and extensive destruction (thrombosis) of the pulmonary artery (Osborn *et al*., 1966) have also been reported. Migration of the band toward the bifurcation of the pulmonary artery may lead to distortion and narrowing of one or both pulmonary arteries, necessitating major reconstruction at the time of intracardiac repair. Infection, possibly with an aneurysm at the site of the banding, is another possible, serious complication. Airway compression may also coexist.

PULMONARY ARTERY RECONSTRUCTION AFTER BANDING (DEBANDING)

It has been our experience that the band can be removed and the pulmonary artery dilated at the time of total repair if the VSD repair is undertaken within 2–3 months of PAB. After a longer interval, we have found it necessary to reconstruct the pulmonary artery in order to avoid residual gradients. We have used three techniques:

1. Removal of the band plus angioplasty (**Figure 17.10a**).
2. Removal of the band plus patch (**Figure 17.10b**).
3. Resection of the segment of the pulmonary artery with the band and end-to-end anastomosis (our preferred technique) (**Figure 17.10c**).

At the time of corrective operation, we dissect the pulmonary artery after establishing cardiopulmonary bypass. The pulmonary artery is opened across the band and, if possible, the band is partially or totally removed. A shelf caused by the band on the posterior wall of the pulmonary artery may have to be excised and the raw area covered with a running stitch of fine, monofilament material. In some patients in whom the pulmonary artery is large, a longitudinal suture may be closed transversely (**Figure 17.10a**).

If angioplasty does not adequately relieve the stenosis, the shelf is excised, and a patch of pericardium or Gore-Tex is sewn in place (**Figure 17.10b**). This procedure usually provides satisfactory relief. Because of the possibility of residual turbulence and gradients across the site of banding, we prefer a technique of resection and end-to-end anastomosis. The pulmonary artery is dissected and the short segment of the artery with the band is resected (**Figure 17.11a**). An end-to-end anastomosis is then performed with the use of a running stitch of 5-0 or 6-0 Prolene sutures (**Figure 17.11b**). Because the size of the pulmonary artery at the time of total repair is usually adequate, a running stitch without any interruption could be used. Excellent reconstruction of the pulmonary artery is achieved.

RESULTS

In earlier years, PAB carried a risk of 13–53%. The main determinant factor for operative mortality was the underlying cardiac defect: the mortality rate was lowest for VSD and highest for truncus arteriosus. Between

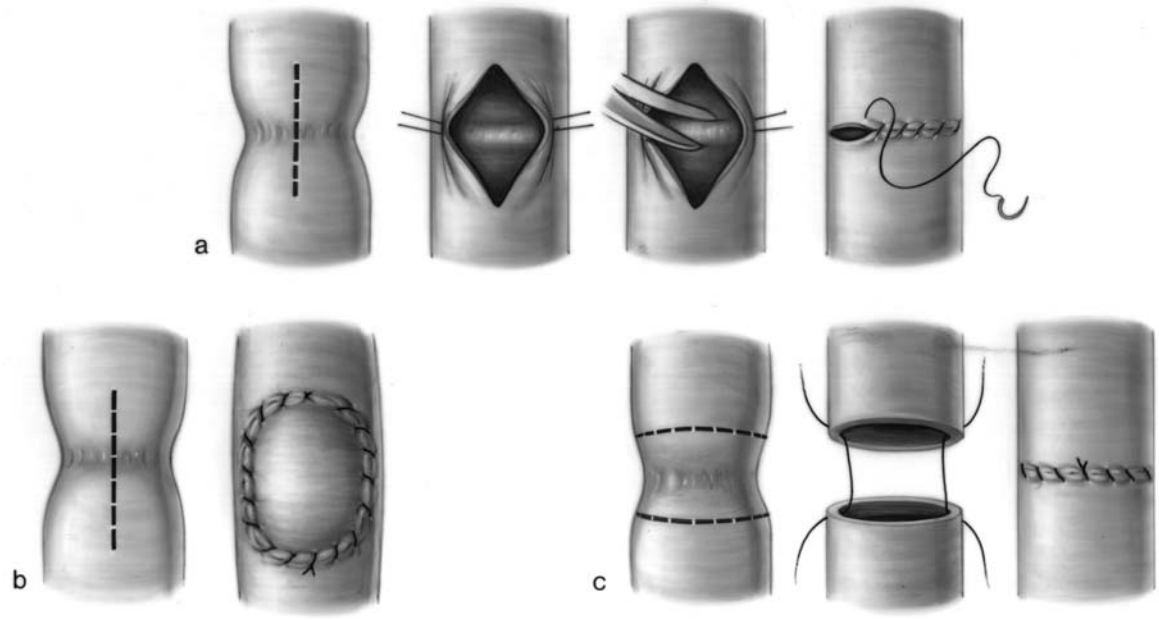

Figure 17.10

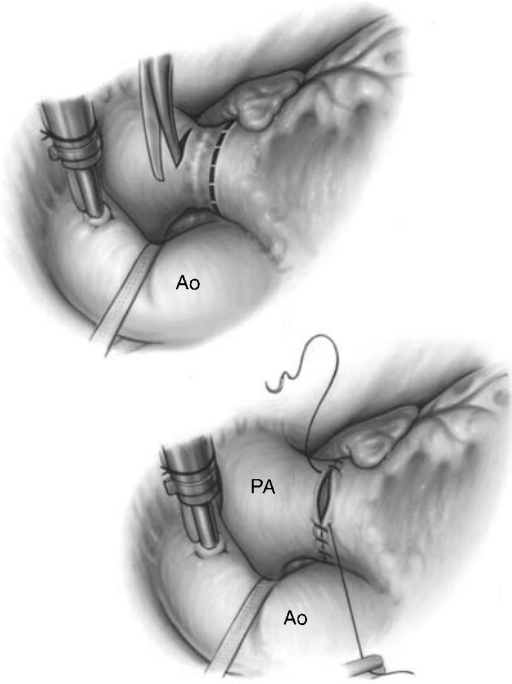

Figure 17.11

1985 and 1991, the authors and associates performed PAB in 158 patients; 145 (92%) survived the operation.

Currently, PAB is used almost exclusively for very complex lesions or for sick neonates with additional cardiac and non-cardiac lesions, as outlined in the section on Indications. During 2000–2004, 102 patients underwent PAB at our unit, with five hospital deaths. There was no early mortality in 24 patients with multiple VSDs, but two deaths (premature babies with low birth weight) in 26 infants with coarctation and large VSD. However, there were 3 deaths out of 27 operated for complex cardiac lesions such as DORV, AVSD complicated by late presentation or low birth weight with coarctation, straddling atrio-ventricular valve, or atrio-ventricular discordance. Decision making in some of these complex cases can be difficult, with many variables involved.

Takayama *et al*. (2002) reported their early mortality of PAB (1990–2001) of 13.8% (12/87), little different from the previous era (1980–1989) of 13.5% (15/111). The authors suggested that, because of the continuing high mortality of PAB, early repair should be considered whenever possible.

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18

Atrial Septectomy

V. T. Tsang and J. Stark

Surgical creation of an atrial septal defect was one of the milestones in the treatment of patients with transposition of the great arteries (TGA). After extensive experimental work, Blalock and Hanlon (1948) reported the first clinical success in 1950. Balloon atrial septostomy (BAS) was introduced by Rashkind and Miller in 1966 and almost completely replaced surgical septectomy in young infants. Currently, Blalock–Hanlon septectomy is performed only rarely; when surgical septectomy is needed, it is usually done on cardiopulmonary bypass as an open procedure, or under circulatory arrest as part of the stage 1 Norwood repair.

INDICATIONS

TGA

Balloon atrial septostomy (BAS) is performed prior to arterial switch operations, but opinions about the use of BAS differ. Some units perform BAS electively in all patients; others reserve BAS only for patients admitted to the hospital in critical condition or for patients who deteriorate before surgical repair (for details, see also Chapters 33–36).

We perform BAS routinely with the aim of improving mixing at atrial level and decompressing the left atrium, which in turn improves the effective pulmonary blood flow. In the case of TGA and VSD, the mixing at atrial level is not so critical, but decompressing the left atrium is still useful. Children with TGA, ventricular septal defect, and left ventricular outflow tract obstruction are usually treated with BAS and a shunt operation. Surgical septectomy is performed if the patient is referred late, usually after 2 months of age, or if BAS becomes ineffective and child is considered too small for a conduit-type operation. A similar situation may arise in some patients with double-outlet right ventricle with transposition physiology.

Univentricular Heart

In neonates with either AV valve atresia or stenosis, the interatrial communication may become progressively obstructive beyond the neonatal period. Surgical septectomy is used mostly in infants with univentricular heart in whom a BAS was ineffective. It should be performed as an integral part of any initial palliation to protect the pulmonary vascular bed.

In some conditions that are not amenable to total repair, decompression of the left (pulmonary venous) atrium may achieve considerable improvement and/or prepare grounds for a subsequent, more permanent palliation (e.g. total cavo-pulmonary connection). This category includes mitral atresia, tricuspid atresia, some forms of double-inlet ventricle with pulmonary stenosis, and double-outlet ventricle with hypoplasia of one of the ventricles.

SURGICAL TECHNIQUE

Various techniques have been suggested and described for the creation of an atrial septal defect. The original Blalock–Hanlon procedure has been probably used most frequently. Alternative techniques include septectomy under inflow occlusion (Lindesmith *et al*., 1965) and closed septectomy with special instruments (Rastan and Koncz, 1971; Kratz *et al*., 1977).

Currently, atrial septectomy is performed on *cardiopulmonary bypass* as an open procedure. As it is quick, normothermia or mild hypothermia can be used. Atrial septectomy during a stage 1 procedure (Norwood) for hypoplastic left heart syndrome is usually performed under *circulatory arrest* in deep hypothermia.

Atrial septectomy can also be performed using the *inflow occlusion technique*. The approach may be through either a right thoracotomy or a midline sternotomy. The aorta, pulmonary artery, SVC and IVC are dissected and encircled with tape. The patient is then ventilated for 2–3 minutes with 100% oxygen, and the SVC and IVC are snared. After a few heartbeats, the aorta

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is cross-clamped. The pulmonary artery may be clamped separately, or both vessels may be occluded with the same clamp (**Figure 18.1**). Ventilation is stopped, the right atrium is opened, the fossa ovalis is grasped with forceps, and an incision made with a pointed blade (**Figure 18.2**). A large segment of septum is then excised with scissors. Caval snares are released, and the atrium is filled with blood. A partial occlusion clamp is placed on the atrial incision (**inset, Figure 18.2**) and aortic and pulmonary artery clamps are released. The procedure usually takes 2–3 minutes and is well tolerated.

As the *Blalock–Hanlon procedure* may still be used in some countries and institutions, we briefly describe the surgical technique (Aberdeen, 1968) here. A right lateral thoracotomy is performed through the fifth intercostal space. The pericardium is incised behind the phrenic nerve from the superior vena cava (SVC) towards the inferior vena cava (IVC). Using this approach and closure of the pericardium, the formation of adhesions is usually minimal (**Figure 18.3**). A thick ligature is passed around the upper and lower pulmonary veins and placed into snares. It is preferable to pass the first ligature by the substraction technique (**inset, Figure 18.3**) to ensure that all pulmonary veins are encircled. The right pulmonary artery is then encircled. A partial occlusion clamp is placed with one jaw behind the pulmonary veins and the left atrium and the other jaw anterior to the right atrium, so that when the clamp is closed, portions of the left and right atria are grasped (**Figure 18.4**). In some older children with a large and tense left atrium, it may be possible to place a large partial occlusion clamp anteriorly across the interatrial groove and part of the left and right atria.

The pulmonary artery is snared and an incision is then made, first on the right side of the septum, using a scimitar blade. A similar incision is then made on the left side of the septum (**Figure 18.5**). Care must be taken not to carry this incision into the right superior pulmonary

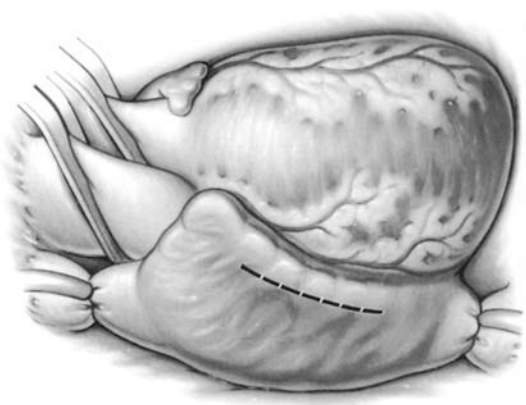


Figure 18.1

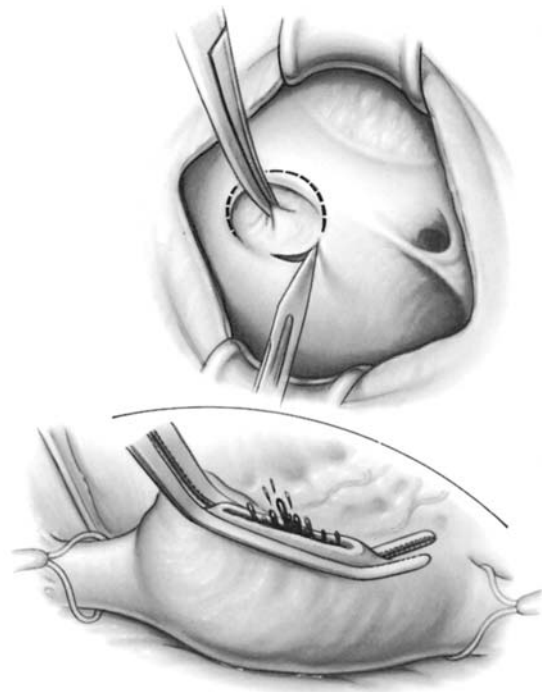
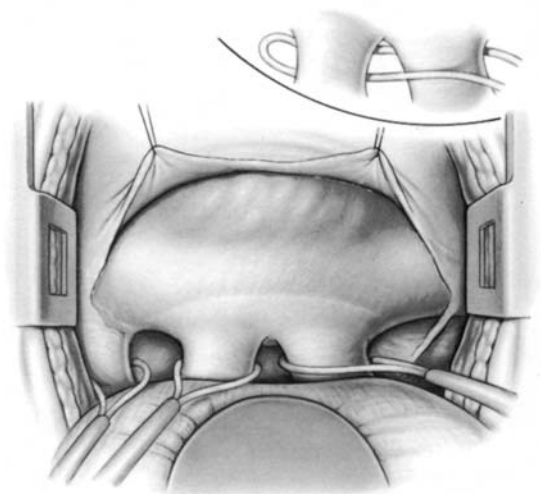


Figure 18.2



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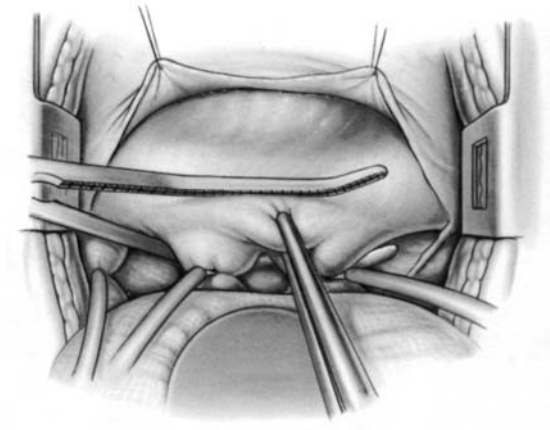

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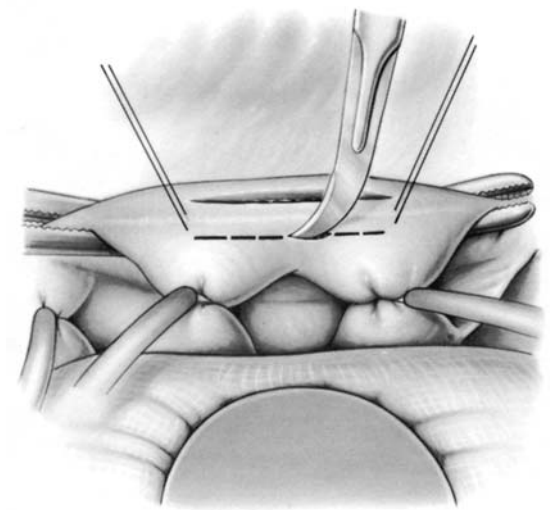

Figure 18.5

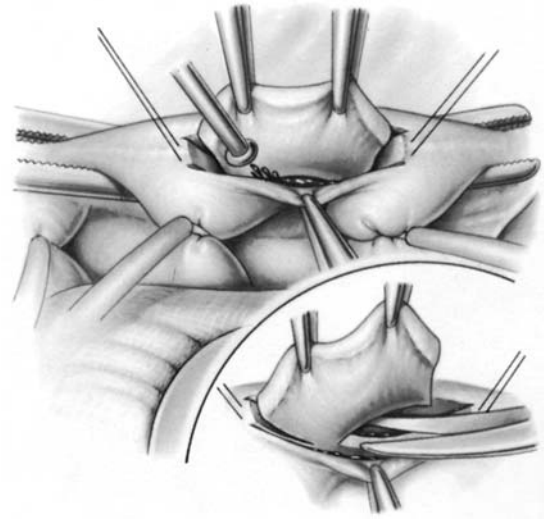

Figure 18.6

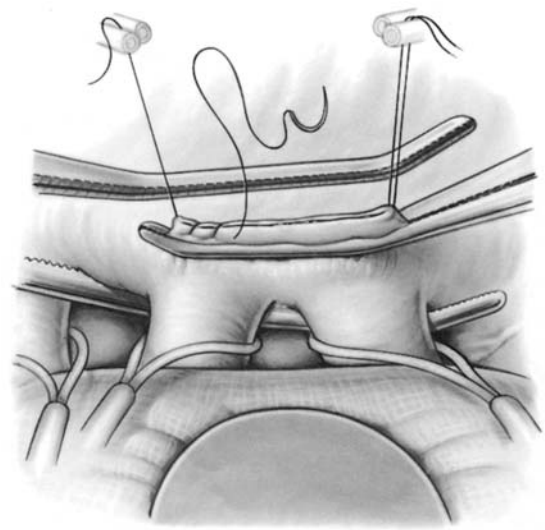

Figure 18.7

vein because it may lead to subsequent narrowing or even occlusion of this vessel. The septum is then grasped with two straight artery forceps or with Babcock forceps. An incision is made superiorly and inferiorly and the clamp is then released slightly, while traction is placed on the two artery forceps. The septum is pulled out into the operating field (**Figure 18.6**). The fossa ovalis is usually visualized, and a large piece of the septum is then excised (**inset, Figure 18.6**). After the septum has been excised, the clamp is again temporarily released to allow the rest of the septum to drop down.

If the atrium is large enough, another vascular clamp is applied to catch the edges of the incision. The atriotomy is closed. The pulmonary veins, pulmonary artery and the first clamp can then be released (**Figure 18.7**). With some practice, the procedure is quick, and the pulmonary veins are occluded for 2 minutes or less before mixing through a newly created atrial septal defect starts (**Figure 18.8**).

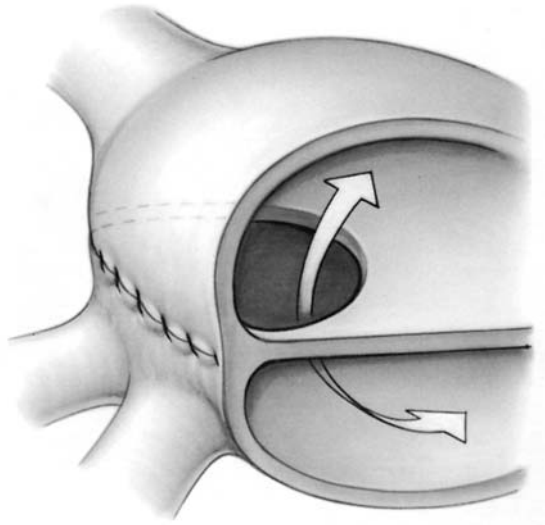

Figure 18.8

RESULT

BAS has greatly lowered the high mortality of surgical septectomy. Blalock–Hanlon septectomy used to carry a considerable risk; mortality rates between 30% and 60% were reported (Ochsner *et al*., 1961; Deverall *et al*., 1969). More recent results are better, although evaluation is difficult: small numbers of patients undergo this operation, and reports of outcomes in the literature are rare. We performed 34 atrial septectomies between 1985 and 1991 for the following diagnoses: TGA, double-outlet right ventricle, mitral atresia, and pulmonary atresia with intact ventricular septum with double-inlet ventricle. Thirty-one patients (91%) also received a modified Blalock–Taussig shunt. Three patients had concomitant banding of the pulmonary artery. There were three hospital death (9%).

Unsatisfactory results were reported with balloon and/or blade septostomy. Even with surgical septectomy, recurrent stenosis across the atrial communication was noted. Perry *et al*. (1986) reported retrospectively the incidence of early and late failures of procedures to enlarge an interatrial communication in patients with left atrio-ventricular valvar atresia and stenosis. BAS was unsuccessful, and blade septostomies were inadequate. Of 63 surgical septectomies, 17% were inadequate, partly related to restenosis of the atrial defect. The importance of maintaining an adequate atrial communication cannot be overemphasized.

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19

Persistent Ductus Arteriosus

J. Stark and V. Hjortdal

The ductus arteriosus connects the main pulmonary trunk with the descending aorta, distal to the origin of the left subclavian artery. In most patients, it is on the left side. If a right aortic arch is present, the ductus may be either on the right or on the left; very rarely is it bilateral. It varies in both length and diameter. The media of the ductus consists mainly of smooth muscle that is arranged spirally, and the intima is much thicker than that of the aorta. Contraction of the medial muscle after birth causes shortening of the ductus and its functional closure. Folding of the endothelium and proliferation of the subintimal layers causes a permanent closure, usually during the first 2–3 weeks of life. Prostaglandins E1, E2 and prostacyclin delay the constriction of the ductus arteriosus at hypoxaemia. Delayed closure of the ductus in premature infants is recognized. It is related to gestational age and low birth weight; 80% of premature babies weighing less than 1200 g will present with a PDA; 45% of those weighing less than 1750 g will have a PDA (Mitchell *et al*., 1971).

DIAGNOSIS

In premature infants, the murmur of persistent ductus arteriosus (PDA) is usually systolic. With increasing size of the shunt, the murmur becomes louder and more prolonged, extending into early diastole. The classical continuous machinery-like murmur described in older children with PDA (Gibson, 1900) is not usual in premature infants. Many children with PDA are asymptomatic, and the clinical features depend on the magnitude of a left-to-right (L–R) shunt. The clinical diagnosis is based on increased or bounding peripheral pulses and the classical crescendo–decrescendo murmur in the second and third intercostal spaces. The murmur is truly continuous only in patients with very large shunts. Electrocardiogram is normal in small PDAs; with larger shunts, mild to moderate or even severe left ventricular hypertrophy is noted. Chest X-ray findings range from normal to moderate cardiomegaly and pulmonary plethora. Two-dimensional echocardiography and Doppler echocardiography is the gold standard for demonstrating patency of the ductus and enlarged left-sided cardiac chambers. Associated cardiac lesions should be carefully ruled out in the presence of any atypical features, such as slight cyanosis, atypical murmur, ejection click, evidence of severe hypertrophy over one or both ventricles on electrocardiograms, and gross cardiac enlargement with evidence of pulmonary oedema on X-ray. In small infants, the high incidence of associated cardiac lesions makes the detailed assessment mandatory. Cardiac catheterization and angiocardiography is used in exceptional, complex cases. Most patients can be assessed accurately by an expert echocardiographer.

INDICATIONS

Congestive heart failure is an indication for PDA closure in infancy. If antifailure treatment is not successful, it is urgent that the PDA be closed.

In premature infants and/or low birth weight infants, indomethacin and ibuprofen are effective in closure of PDA (Heymann *et al*., 1976; Friedman *et al*., 1976) and the two drugs are equally effective (Ohlsson *et al*., 2003). Even in asymptomatic preterm infants with PDA, indomethacin significantly reduces the incidence of developing symptoms, the need for duct ligation, and the duration of supplemental oxygen and is therefore recommended (Fowlie and Davis, 2003).

If the medical therapy fails, operation is considered for all symptomatic children. Cassady *et al*. (1989) conducted a randomized controlled trial of early (day 1) prophylactic ligation of the PDA in infants who weighed 1000 g or less at birth. The control group underwent PDA ligation only if the shunt was haemodynamically important. Cassady *et al*. (1989) concluded that early surgical closure reduced the risk of necrotizing enterocolitis in these infants of very low birth weight.

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In children with mild or no congestive heart failure, PDA closure can be planned electively. We believe that all PDAs should be closed because of a small but definite risk of subacute bacterial endocarditis. In children with elevated pulmonary arteriolar resistance ($> 8\,U/m^2$), a severe degree of pulmonary vascular obstructive disease has probably developed, and their PDAs should not be closed.

METHODS FOR CLOSURE OF PERSISTENT DUCTUS ARTERIOSUS

Medical

Closure of a PDA can be stimulated by the administration of prostaglandin synthesis inhibitors, such as indomethacin or ibuprofen. These agents have been shown to be effective in premature infants (Heymann *et al*., 1976; Friedman *et al*., 1976, Fowlie and Davis, 2003; Ohlsson *et al*., 2003). Indomethacin (0.2 mg/kg) is given by intravenous infusion over 20 minutes as an initial dose; this is followed by two more doses after 12 and 24 hours. These doses are 0.1 mg/kg in infants weighing less than 1250 g and 0.2 mg/kg in larger infants and those more than 7 days old. With this regime, the number of surgical ligations was reduced from 27% to 5% of haemodynamically significant PDAs (Clyman, 1990).

Surgical Technique

The technique of duct ligation was described by Munro in 1907. The first successful operation was performed in 1939 by Gross and Hubbard (1939). The PDA is ligated or divided. Under special circumstances, such as a calcified PDA or the presence of dense adhesions after previous intrapericardial operations, it is closed on cardiopulmonary bypass by a suture from inside the pulmonary artery. The technique is similar to that used in the closure of a Potts shunt. Alternatively, the orifice of the PDA can be closed with a patch from an aortotomy. The usual approach to a PDA is through the third or fourth intercostal space, using a left posterolateral thoracotomy. In small infants, this can be performed through the vertical skin incision (Browne, 1952), as described in Chapter 15. Some surgeons prefer an anterolateral thoracotomy. More recently, video-assisted thoracoscopic approach (Laborde *et al*., 1993) or even robotic surgery (Le Bret *et al*., 2002) has been suggested.

Ligation

A standard left thoracotomy through the fourth intercostal space is performed. The lung is gently retracted medially over a wet swab, and the posterior pleura is incised (**Figure 19.1**). The left superior intercostal vein is ligated and divided. The edges of the posterior pleura are cauterized, care being taken not to injure the vagus nerve and the thoracic duct. Stay sutures are then placed on the anterior edge of the pleura (**Figure 19.2**). Sharp dissection is continued above and below the level of the ductus. Dissection is easiest in the plane close to the aortic wall, which is avascular. The recurrent laryngeal nerve is left on the pleural flap, which

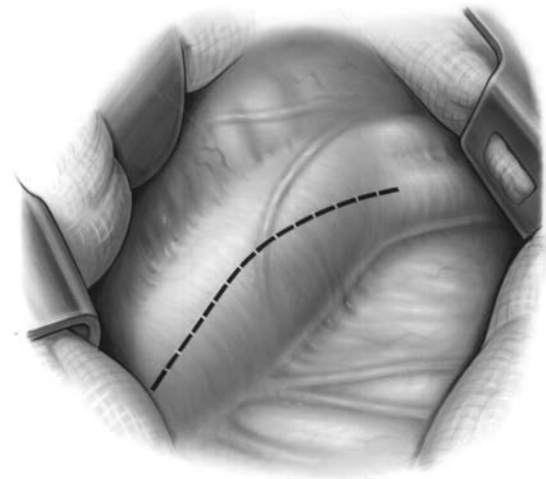

Figure 19.1

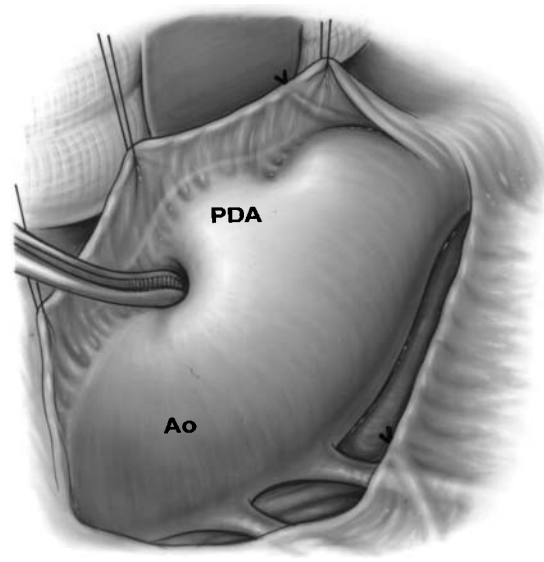

Figure 19.2

Figure 19.3

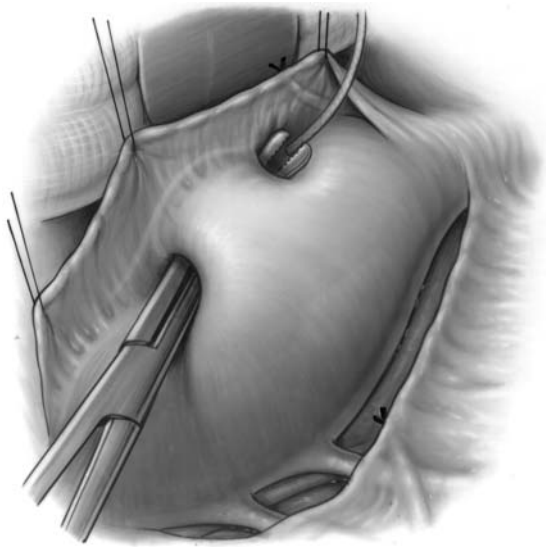

Figure 19.4

is retracted medially (**Figure 19.2**). For dissection of the PDA, we use a Denis Browne dissector (**Figure 19.3**) with a blunt end. First, a sharp dissection above and below the PDA opens access into the right tissue plane. A curved dissector is then used, the branches being opened parallel to the ductus to avoid any injury. An attempt to come around the ductus with a fully curved dissector is made only when the dissection is deep enough both above and below the ductus (**Figure 19.4**).

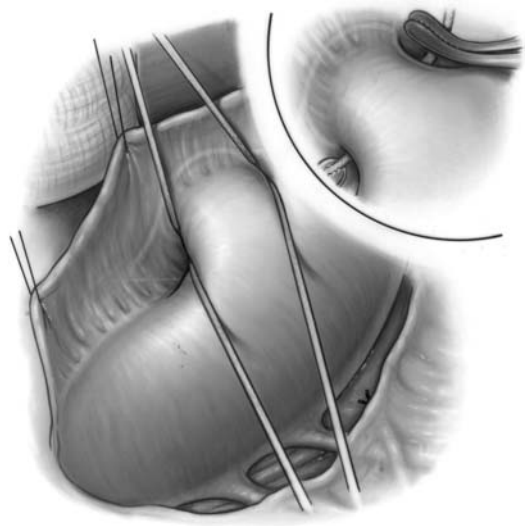

Figure 19.5

It must be remembered that the ductal tissue is often fragile and, therefore, dissection should be performed carefully. When the instrument passes freely behind the PDA, a ligature is passed around it. The authors and associates prefer to use a thick ligature of plaited silk (**Figure 19.5**); a finer ligature may cut through the ductal tissue and cause dangerous bleeding. Care is taken not to pull the ligature too fast because it may act like a Gigli saw and damage the posterior wall of the ductus. A right-angled dissector keeps the ligature deep down while it is being pulled around the PDA in order to avoid damage to the posterior wall (**inset, Figure 19.5**).

Trial occlusion for 30–60 seconds, either by pressure against the dissector or by temporary clamping with the dissector, should abolish the thrill. Development of bradycardia or hypotension is a warning sign that the ductus may be providing the sole blood supply to the pulmonary artery (as in pulmonary atresia) and the ligation must therefore be avoided. In patients with a large L–R shunt through the ductus, a rise in the systemic pressure may be seen during trial occlusion.

Before ligation of the PDA, we always check for the following items:

1. The blood supply is available in the operating room.
2. Vascular clamps are selected and available should any bleeding occur.
3. Two independent suction units are functioning.

The ligature close to the pulmonary artery is first tied. Both index fingers should be deep in the chest in the plane of the PDA (**Figure 19.6**). Occasionally, double

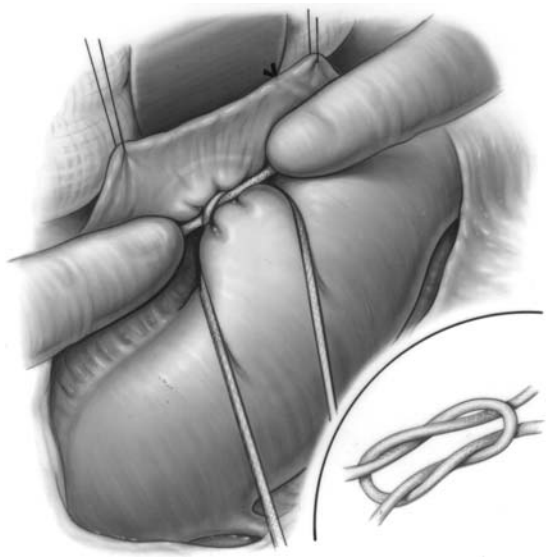

Figure 19.6

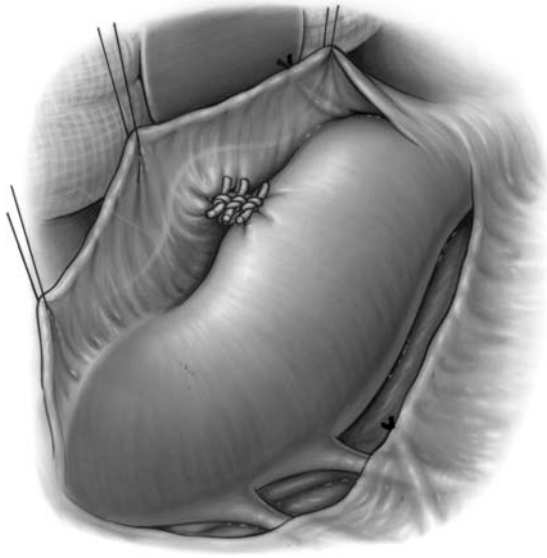

Figure 19.7

ligature is not adequate and three or four ligatures may have to be placed (**Figure 19.7**).

In patients with pulmonary hypertension, the aorta is cross-clamped above and below the PDA before the first knot is tied. The dissection is then slightly more extensive and the two rubber slings are passed around the aorta above and below the PDA. Two straight vascular clamps

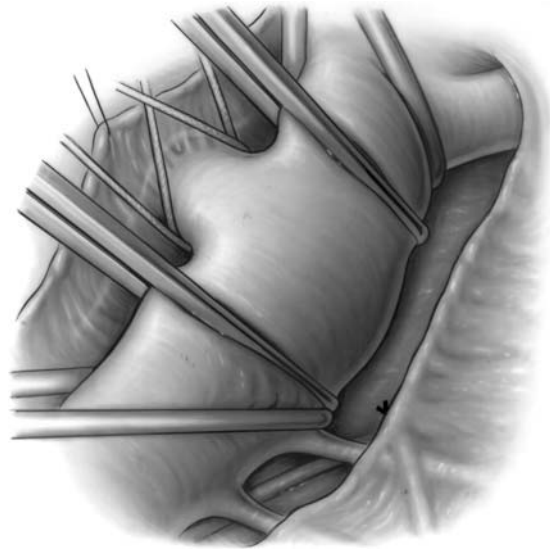

Figure 19.8

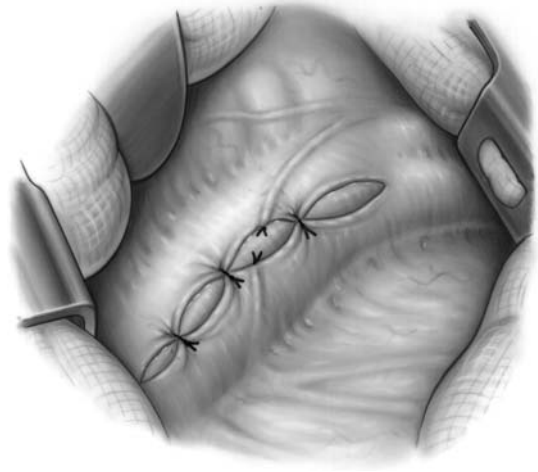

Figure 19.9

are applied (**Figure 19.8**), the PDA is ligated, and the clamps are immediately released. Additional ligatures are placed later. In this technique, the time of cross-clamping is very short (only 10–20 seconds). Ligation by two purse-string sutures in the middle was suggested and performed by Blalock (1946). When the PDA is safely ligated, we check that the thrill has disappeared. All bleeding points are carefully stopped, and the posterior pleura may be approximated with two or three single stitches (**Figure 19.9**). The chest is then closed in a routine manner with an intercostal drain in place.

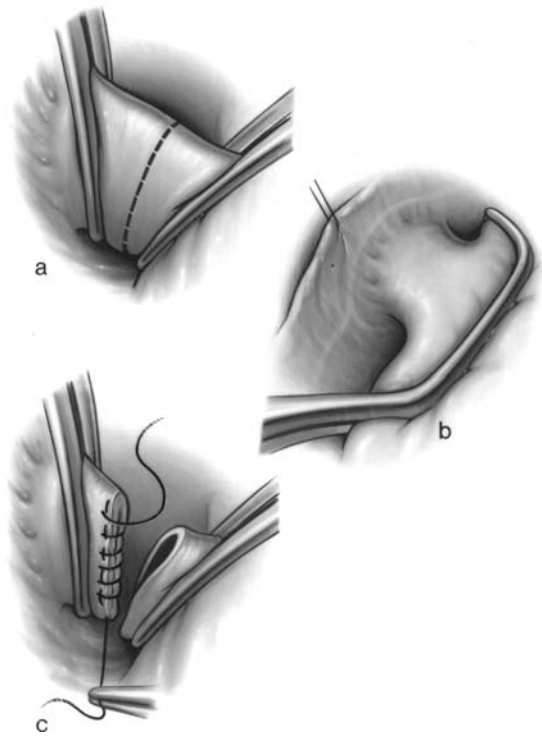

Figure 19.10

Division

The approach is the same as that described earlier: through the left lateral thoracotomy, through the fourth intercostal space. When the PDA is dissected, fine vascular clamps are applied, and the PDA is divided (**Figure 19.10a**). Alternatively, a partial occlusion clamp may be used at the aortic end to obtain more length for division (**Figure 19.10b**). Care must be taken not to crush or tear the PDA tissue with clamps. Both ends are oversewn, usually with a running mattress suture in one layer and then a continuous over-and-over stitch in the second layer (**Figure 19.10c**).

Closure from a Midline Sternotomy

In earlier years, a separate left thoracotomy was performed for patients with intracardiac lesions complicated by a PDA. Kirklin and Silver (1958) recommended an anterior approach to PDA in 1958. The PDA is dissected before administration of heparin and before cardiopulmonary bypass is started. We place a mosquito clamp on the adventitia of the ascending aorta and apply gentle traction to the right and toward the inferior vena cava. The main pulmonary artery is pushed downwards and to the left by a swab or the surgeon's finger (**Figure 19.11**). The left pulmonary artery and PDA are then dissected with sharp and blunt dissectors. The PDA is encircled with ligatures of plaited silk and ligated just before the institution of cardiopulmonary bypass (**Figure 19.12**). If the PDA is short, it may be excised from the pulmonary artery; the aortic end is oversewn on the clamp, and the opening in the pulmonary artery is sutured when cardiopulmonary

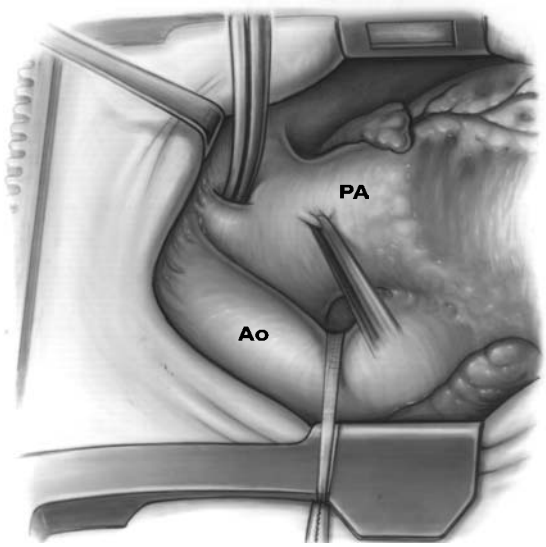

Figure 19.11

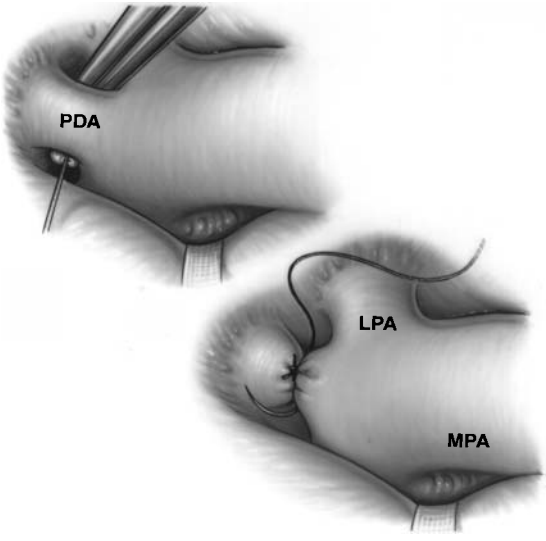

Figure 19.12

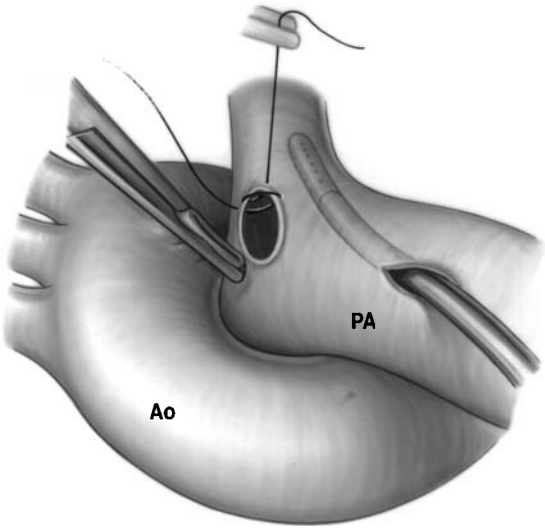

Figure 19.13

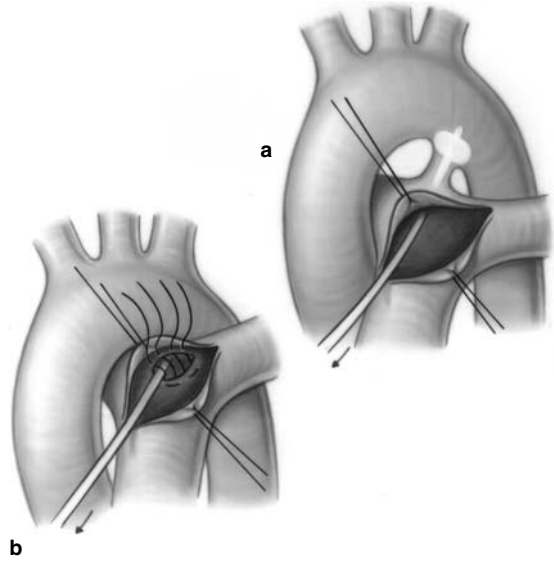

Figure 19.14

bypass is instituted. A sump in the pulmonary artery minimizes blood loss (**Figure 19.13**) (McGoon, 1964). In patients in whom the PDA provides the sole blood supply to the pulmonary artery, ligation is deferred until cardiopulmonary bypass is begun.

In a patient with a large and tense PDA, it may be occluded temporarily and the patient may be cooled on cardiopulmonary bypass. Once the core temperature is lowered, the perfusion flow is reduced and the PDA safely ligated. As the intraluminal pressure is decreased, the chances that the ligature will cut through are minimized. During some procedures when full mobilization of both pulmonary arteries is required (arterial switch operation, Rastelli operation), the PDA should be divided. In some patients, ligation of the PDA or ligamentum arteriosum can cause some distortion of the bifurcation of the pulmonary artery.

Closure from Inside the Pulmonary Artery

If the PDA is not easily accessible, either because of the adhesions after previous intrapericardial operations or because of its position, it can be sutured from inside the pulmonary artery. The patient is cooled on cardiopulmonary bypass while the PDA is occluded by the surgeon's finger from outside. When the nasopharyngeal temperature reaches 28°C, the rate of perfusion is temporarily reduced to 0.5 L/min/m^2. The pulmonary artery is opened, and the PDA firmly occluded by the surgeon's finger or a balloon catheter (Bhati *et al*., 1972) (**Figure 19.14a**). The incision is extended towards the left pulmonary artery, and the orifice of the PDA is identified and oversewn (**Figure 19.14b**). As a precaution against possible air embolization, the child is placed in the head-down position. When the sutures are being placed, care must be taken not to occlude a branch of the left pulmonary artery instead of the PDA and also to prevent air from entering the aorta. The intracardiac operation then proceeds in the usual manner.

Closure in the Presence of Acute Endocarditis

A patient with active endocarditis on the PDA is usually treated with massive doses of antibiotics. However, if the infection is not brought under control or if embolization to the lungs occurs, operation during the acute phase of endocarditis may be undertaken on cardiopulmonary bypass.

Patch Closure

It has been suggested that the PDA may have to be closed by a patch from inside the aorta (Morrow and Clark, 1966). However, today, severely calcified PDA in an adult or a recanalized PDA would be treated by Device closure in the catheterization laboratory.

Ligaclip Occlusion in Premature Infants

In premature infants, we prefer occlusion of the PDA with a Ligaclip. The posterior pleura is opened over the descending aorta, and dissection is made above and below the PDA. When the dissection is deep enough, a Ligaclip is carefully applied (**Figure 19.15**). The

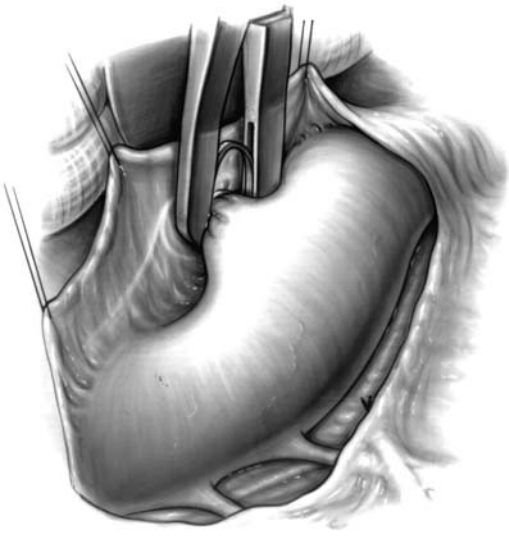

Figure 19.15

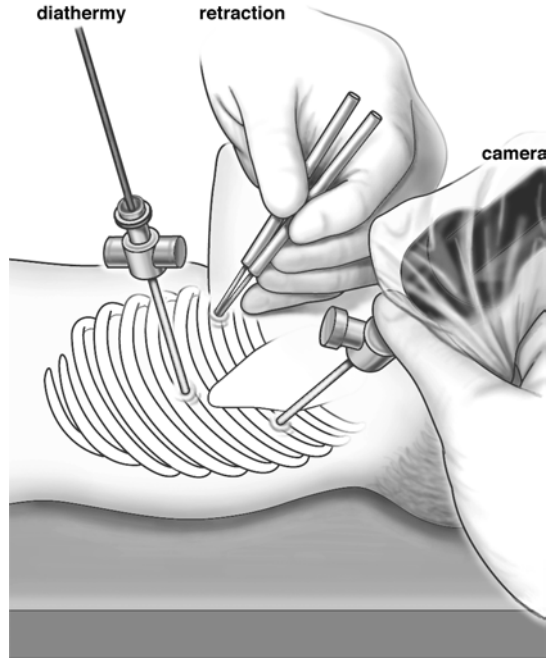

Figure 19.16

applicator of the clip must be opened slowly because a clip sticking to the applicator may possibly tear the fine ductal tissue. A second Ligaclip may be applied if necessary. The advantage of this approach is a limited dissection. The friable duct does not have to be encircled, and this diminishes the chances of tear and bleeding. Safe and fast occlusion is achieved.

Transcatheter Closure of PDA

In older infants and children, most PDAs can be closed without thoracotomy during cardiac catheterization. Porstmann *et al*. (1971) and Rashkind and Cuaso (1979) described the two alternative techniques. Currently, the coils are used with excellent results. This is currently used for most children who weigh 6 kg or more. Details of interventional cardiology techniques and the results are described in Chapter 7.

Video-assisted Thoracoscopic Surgery (VATS)

Video-assisted thoracoscopic surgery (VATS) approach to PDA ligation was reported by Laborde *et al*. (1993, 1995). Burke (1995) published results from Boston. Under general anaesthesia, the patient is placed in the right lateral position with the left arm in a 90° angle with the chest wall (**Figure 19.16**). Three to four small 4–7 mm incisions are made around the tip of the scapula for positioning of a videoscopic camera, one to three lung retractors, an L-shaped diathermy dissector and the endoscopic clip applicator. The camera and the diathermy dissector can be easily manipulated in placed through 4–5 mm trocars. No chest wall muscles are cut and the ribs are not retracted.

Depending on the size of the child, the videoscope size is 2.7–4 mm. Cotton-tip applicators are used as lung retractors for the premature infants and 60–90° angled hooks or fan-shaped retractors for infants and older children.

We prefer the assistant to hold the camera. The same approach is used by Laborde *et al*. (1993), but the camera can also be held by a mechanical arm (Jacobs *et al*., 2003).The lung is retracted inferomedially. Using the diathermy, the parietal pleura overlying the aorta is incised from 1 cm below the PDA up to the left subclavian artery while pulling the pleura away from the aorta and the PDA. The hemiazygos vein crossing the aorta is divided, using cautery. A 90° hook is introduced through the same hole as the other hooks to retract the pleura up against the heart and towards the sternum.

Dissection on the cranial side of the PDA is usually performed using diathermy, while the dissection on the caudal side of the PDA is done using the diathermy hook without the current applied. The PDA is dissected on both the cranial and caudal side so that the two arms of the Ligaclip can be applied.

The right side of the PDA is usually not dissected. When dissection has been completed the diathermy hook

and the trocar are removed. The hole below the lower tip of scapula is slightly enlarged using blunt dissection. The clip applicator with the clip mounted is introduced through the hole between the legs of a long slim artery forceps. The clip is gently placed with an arm on each side of the PDA, using soft wriggling movements and avoiding tissue slipping in between the clip and the PDA. The first clip is placed close to the pulmonary artery; the second clip is placed on the aortic side of the PDA.

A large PDA may be reduced in size with an endoscopically applied ligature after complete circumferential dissection and then completely occluded with a clip. The positioning of the clip is controlled using the diathermy hook to push tissue aside, so that the clip can be visualized from both sides. Air is evacuated from the pleural cavity using a small drain, which can be removed immediately or after a few hours. The three port holes are closed with absorbable sutures in the fascia and Steristrips on the skin. When performing a VATS procedure, we have the instruments for thoracotomy and vascular clamps available in the theatre.

Robotically assisted PDA closure has recently been introduced. It seems to be equally safe but is more time consuming (Le Bret *et al*., 2002)

COMPLICATIONS

Bleeding at the time of operation is a major complication that can occur when the PDA is ligated or divided. The risk of haemorrhage is higher in the presence of an aneurysm of the PDA. This may be present before surgery. It was described after both ligation and division (Ross *et al*., 1961). Recanalization of the PDA is rarely observed and is presumed to be the result of incomplete ligation rather than recanalization. In our series, it occurred in four of 936 patients. Injury to the recurrent laryngeal nerve is rare. On occasion, chylothorax may occur with the dissection and damage to the small lymphatic vessels overlying aorta and the ductus.

In premature infants with PDA, operative mortality may be related to severe associated problems: retrolental fibroplasia with possible blindness, bronchopulmonary dysplasia and cerebral palsy. All these are probably related to long-term ventilation, oxygen toxicity and prematurity (Brandt *et al*., 1981).

After VATS procedures, recurrent laryngeal nerve injury has been noted in 3% (13.6% in low birth weight infants) but only 0.4% patients have permanent dysfunction. Chylothorax (0.6%), conversion to thoracotomy (1%), transfusion for bleeding (0.1%) and pneumothorax (1.3%) were also described. They were noted more often in low birth weight infants (Villa *et al*., 2004).

Residual patency of PDA has been detected immediately after the procedure in 1.4% of operated patients, leading to additional surgery. The incidence of residual patency at follow-up and conversion to thoracotomy have been reported to be 0.6% and 1%, respectively, by Villa *et al*. (2004). However, higher incidence (16% and 12%) was observed in the series of Kennedy *et al*. (1998). In the last-mentioned paper, no benefits were identified in terms of hospital stay, length of operation or morbidity when compared to a muscle-sparing thoracotomy.

Complications related to trans-catheter closure of PDA are mentioned in Chapter 7.

RESULTS

In the earlier years, the risks of operation for children with PDA as a single lesion were less than 0.5% (Panagopolous *et al*., 1971). A high risk has been reported in the presence of associated lesions and increased pulmonary arteriolar resistance and, in adults, when the ductus becomes calcified or even aneurysmal.

Perioperative mortality for PDA occlusion in premature infants used to be high. Clark *et al*. (1976) and Nelson *et al*. (1976) reported mortality rates of 20–40%. Even in the current era, the overall hospital mortality rate for this group of patients is around 10% (Niinikoski *et al*., 2001). Only 50% of hospital survivors of PDA closure in premature infants in one series were alive and well 5 years later (Brandt *et al*., 1981). The poor prognosis is reflected by the fate of babies with extremely low gestational age and birthweight in general (Kamper *et al*., 2004).

The largest series of more than 700 cases of VATS occlusions of PDA has been reported by Laborde's group in Paris (Villa *et al*., 2004). There was no operative mortality. Even low birth weight infants can be treated by VATS without mortality (Hines *et al*., 2003).

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20

Coarctation of the Aorta

V. T. Tsang and J. Stark

Coarctation of the aorta is a congenital narrowing of the aorta, usually situated between the origin of the left subclavian artery proximally and the junction of the aorta and the ductus arteriosus distally. In occasional patients, however, the constriction may be between the left carotid and the left subclavian arteries. Although it has long been recognized that the clinical manifestations of this disorder are strikingly different in infants in comparison with older children, the mechanism of this difference remains unknown. Bonnett (1903) attempted to establish a clinical pathological classification, dividing the coarctation into an ''infantile'' type, characterized by an elongated and diffuse narrowing of the aorta proximal to the ductus arteriosus, and an ''adult'' type, consisting of a sharp constriction in the area of aortic insertion of the ductus. Johnson *et al*. (1951) similarly based their classification on the position of the ductus and its relation to the coarctation, recognizing the preductal and postductal coarctations. These classifications have proved to be unsatisfactory because the ''infantile'' or preductal type is encountered in older children and the ''adult'' or postductal type is found in infants. Occasionally, in neonates, if the coarctation is postductal, prostaglandin infusion would not be effective. For practical purposes, the clinician deals with two groups of patients: symptomatic infants and asymptomatic older children or, occasionally, adults.

DIAGNOSIS AND MEDICAL MANAGEMENT

Symptomatic Infants

Isolated Coarctation

These patients, many of whom are neonates, are often admitted to the hospital with severe congestive heart failure. They are often in critical condition. Typically, the radial pulses are easily palpable and prominent; the femoral pulses, in contrast, are weak or cannot be palpated. These findings may change, especially if an infant is treated with prostaglandin, which may reopen the ductus arteriosus. Thus, the situation in which one observer feels strong femoral pulses whereas another finds them absent is not infrequent. Auscultatory findings are influenced by associated anomalies, which are present in 75% of the cases in the neonatal period (Tawes *et al*., 1969). This emphasizes the fact that coarctation is rarely a simple lesion. Besides persistent ductus arteriosus, the most commonly associated cardiac anomalies are bicuspid aortic valve, ventricular septal defect (VSD), and mitral valve anomalies. The association of coarctation of the aorta, parachute mitral valve, supravalvar ring of the left atrium and subaortic stenosis is known as Shone's syndrome (Shone *et al*., 1963). On chest X-ray, the heart is usually markedly enlarged, and the lung fields show congestion, increased pulmonary vascularity and sometimes frank pulmonary oedema. Electrocardiograms invariably show right ventricular hypertrophy with or without left ventricular hypertrophy. At our institution, neonates with coarctation of the aorta are currently managed as follows. If coarctation is suspected in a critically ill neonate, resuscitative measures are initiated first. These include placement of central venous and arterial cannulae and starting prostaglandin E_1 infusion (5 ng/kg/minute). In many patients, an infusion of dopamine is started as well, and diuretics are given. The sickest infants are intubated and ventilated. Patients in established renal failure may have to undergo dialysis even before surgery. The diagnosis is then established by two-dimensional and Doppler echocardiography. The aortic arch anatomy is demonstrated, and the coarctation shelf is usually visible just distal to the left subclavian artery. Continuous-wave Doppler echocardiography enables measurement of the maximal postcoarctation jet velocity, from which the gradient across the defect can be estimated using the modified Bernoulli equation. The results are not always accurate in neonates (Marx and Allen, 1986). The gradients may be underestimated in the presence of the collateral flow. Colour-flow mapping techniques

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have improved the accuracy of diagnosis because the presence of diastolic flow tail in the descending aorta is diagnostic. In the majority of patients, this approach is adequate for assessing both the coarctation and the associated cardiac lesions. If the patient responds well to the aforementioned measures, repair of the coarctation is performed within 24 hours. If, however, the child does not respond adequately and continues to be acidotic or anuric or both, the operation is performed without delay. For infants who are not critically ill or are asymptomatic but have persistent cardiomegaly or severe hypertension, early elective repair is planned. The risk of anastomotic stricture is lower in older infants and hopefully the risk of persistent hypertension does not increase if the operation is performed within 1 year of age (Brouwer *et al*., 1994).

The use of balloon angioplasty for native coarctation remains controversial. There is an important risk of the recurrence of coarctation and risk of vascular injury to femoral vessels, especially in neonates (Rao *et al*., 1996). We consider these risks unacceptable and do not perform balloon dilatation in these patients. The angioplasty data is less clear-cut in older children. The major concern is the rupture of the aorta at the time of the procedure and aneurysm formation at the site of the intimal and medial tear of the aorta (Hijazi *et al*., 1997).

Coarctation and Ventricular Septal Defect

In the past, we used to assess the size of the VSD angiocardiographically and determine the probability of survival without banding (Leanage *et al*., 1981). Nowadays, echocardiography and Doppler assessment should suffice. The conventional approach is banding of the pulmonary artery for a moderate or large VSD, performed at a time of coarctation repair. For patients who do not undergo banding and fail to be weaned off the ventilator rapidly, early closure of the VSD is considered. Alternatively, coarctation can be repaired and VSD closed while a patient is on cardiopulmonary bypass from a midline sternotomy (Ungerleider and Ebert, 1987). The preoperative neurological status and the complexity of the VSD and arch hypoplasia would influence surgical strategy.

Older Children

Currently, the majority of patients with coarctation of the aorta are diagnosed and treated in infancy. In older children, aortic coarctation rarely produces symptoms and is often first recognized during routine physical examination at school, which reveals weak, delayed or absent femoral pulses and a heart murmur or the presence of upper extremity systemic hypertension. The chest X-ray may be normal or may show cardiomegaly. Rib notching is rarely seen in patients under 8 years of age. The electrocardiogram may be normal or may show left ventricular hypertrophy. Echocardiogram may not provide an adequate assessment. Cardiac catheterization and angiocardiography or MRI are performed if the clinical diagnosis is not sufficiently clear or typical and if associated cardiac anomalies are suspected. Associated malformations are much less common in asymptomatic children than in symptomatic infants (Tawes *et al*., 1969). Despite the lack of symptoms, surgical treatment is usually recommended as soon as the diagnosis of significant narrowing is made. Late hypertension after successful surgery remains an ongoing concern.

SURGICAL TECHNIQUE

If not in place preoperatively, an arterial catheter is inserted, in all patients, into the radial artery on the side opposite to the coarctation (usually the right side) to monitor the arterial blood pressure in the upper body and the blood gases. It is very important not to reduce the upper body blood pressure below normal and to maintain normal acid–base status, as spinal cord injury could occur. Of equal importance is adequate venous access for transfusion. The patient is positioned on the table with the left side up.

A lateral thoracotomy incision is made through the third or fourth intercostal space. The left lung is retracted anteriorly and inferiorly by a moist sponge. A longitudinal incision is made in the mediastinal pleura over the upper descending aorta, posterior to the vagus nerve, and is extended superiorly over the left subclavian artery, ligating and dividing the superior intercostal vein in the process (**Figure 20.1**). Stay sutures are then placed on the anterior edge of the pleural incision; care must be

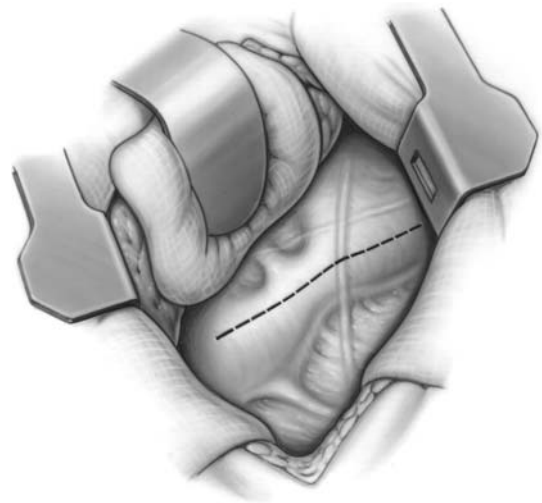

Figure 20.1

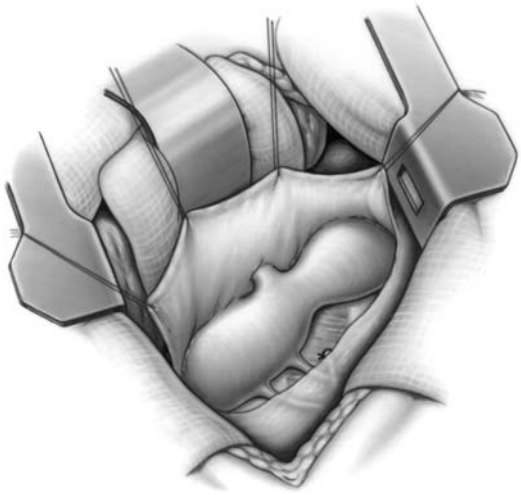

Figure 20.2

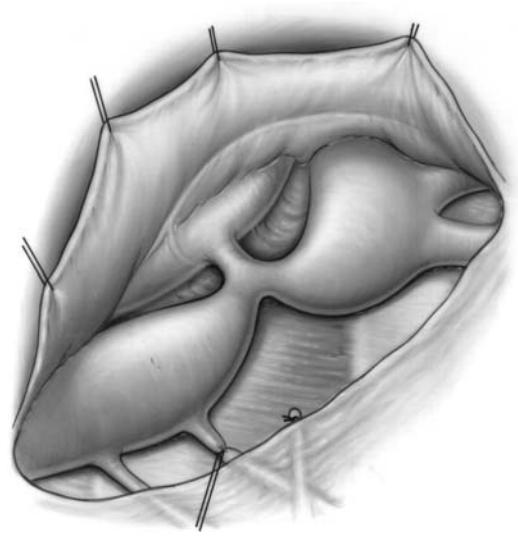

Figure 20.3

taken not to damage the vagus nerve. The stitches are anchored to the thoracotomy retractors (**Figure 20.2**). The ductus arteriosus is often patent; some degree of tubular hypoplasia of the aortic isthmus is almost a constant feature in these infants (Ho and Anderson, 1979). The ductus arteriosus is then dissected. Care must be taken not to damage the recurrent laryngeal nerve. The areolar tissue is separated from the lateral and inferior surfaces of the ductus by sharp dissection. The dissection on its medial aspect is facilitated by the use of a blunt tissue dissector to open the fibrous layer and pass a braided silk ligature around the ductus, which is then securely tied just before the vascular clamps are applied. The aorta is mobilized proximally and distally. Sometimes the first few pairs of intercostal arteries are looped (**Figure 20.3**).

Several techniques for repairing coarctation of the aorta in infancy have been described. There is no consensus about the preferred technique. We believe that optimally the diseased segment of the aorta should be removed, and therefore resection and end-to-end anastomosis is our technique of choice. However, because the anatomical situations vary, the technique should be suited to each patient. With a long hypoplastic distal segment, the subclavian flap technique may be preferred. If, in addition to the coarctation, the segment of the aorta between the left carotid and left subclavian arteries is hypoplastic, reversed subclavian flap may be considered (Hart and Waldhausen, 1983). In a relatively rare form of hypoplasia of the entire aortic arch, extended resection and anastomosis of the descending aorta onto the undersurface of the aortic arch is an appealing technique. Aortoplasty with a prosthetic patch is a quick and simple procedure; unfortunately, a considerable number of late aneurysms have been reported with the use of this technique. Because these aneurysms may develop 15–20 years later, we prefer not to use a patch aortoplasty and reserve it for special situations, such as during some reoperations.

Resection and End-to-end Anastomosis

Resection and direct end-to-end anastomosis, originally described in 1945 by Crafoord and Nylin, should be preferred to any other technique. When the dissection of the aorta and the ductus is completed, the ductus arteriosus is doubly ligated. The aorta is then occluded above the coarctation; if the pressure in the distal aorta seems to be low, it should be measured. However, the level of acceptable pressure is still controversial.

Paraplegia is very rare but a serious complication after repair of coarctation of the aorta. Blood supply to the spinal cord (anterior spinal artery) is variable. Brewer *et al*. (1972) reported an incidence of 0.41% of paraplegia in a collective series of 12 532 operated coarctations and re-coarctations. There is no general agreement as to what the ''safe'' signs of adequate collateral circulation in children with coarctation are; pressures higher than 40 mmHg below the clamp have been quoted as safe (Stark and Pacifico, 1989). Even the presence of anomalous right subclavian artery does not seem to increase the risk of paraplegia in neonates (Hjortdal *et al*., 2003).

In neonates, measures to avoid paraplegia are shortest possible clamping time (nowadays rarely exceeding

20 minutes), avoidance of haemorrhage with hypotension, low proximal aortic pressure and avoidance of acidosis. Mild hypothermia (surface) and pleural lavage with cold saline may possibly provide some protection. Other measures, such as steroids, local cooling of the spinal cord, and intrathecal injection of papaverine, are not currently used. In children another form of spinal cord protection would be perfusion of the descending aorta at the time of aortic cross-clamping. It can be achieved by femoral–femoral or left atrium–descending aorta bypass, or by using a temporary shunt (Gott) between the subclavian artery or ascending aorta and the descending aorta.

In older children the soft tissues are abundantly supplied with collateral vessels, most of which require ligation. Blood loss can be minimized by manually compressing the muscles against the ribs. To avoid damage to the large and often tortuous intercostal vessels, the chest may be entered through the bed of the fourth rib rather than through the intercostal space. The retractor is inserted and opened slowly. Dissection and mobilization of the aorta is performed, with special attention to the well-developed collateral circulation. The easier dissection is performed first; the left subclavian artery, the adjacent aorta, the coarctation, and the ligamentum arteriosum or ductus arteriosus are mobilized. The proximal dissection is carried up to and sometimes beyond the left carotid artery, in order to provide adequate mobilization of the arch. Special care must be taken in the posterior aspect, where small branches to the viscera of the posterior mediastinum (bronchial and oesophageal arteries) may be found and, if severed, can cause distressing haemorrhage. An anomalous artery may arise from the back of the aorta immediately above the coarctation and course medially beneath the aortic arch or carotid artery. That artery, originally described by Hamilton and Abbott in 1928, was named Abbott's artery by Schuster and Gross in 1962. It may be aneurysmal.

The post-stenotic segment is then mobilized; great care is taken not to injure the intercostal vessels, which are dilated and thin-walled and bleed briskly if injured. However, these vessels should be preserved whenever possible. The distal dissection is carried to the level of the third or fourth pair of intercostal vessels, and beyond if necessary, to provide adequate mobilization.

If the collateral circulation is considered adequate, vascular clamps are selected and tried. If the clamps seem suitable, the lungs are temporarily ventilated for 1–2 minutes. This is beneficial, especially in sick infants with associated cardiac lesions and poor left ventricular function. The left lung is then retracted again, and clamps are applied on the aorta from the surgeon's side (**Figure 20.4**). We prefer, if possible,

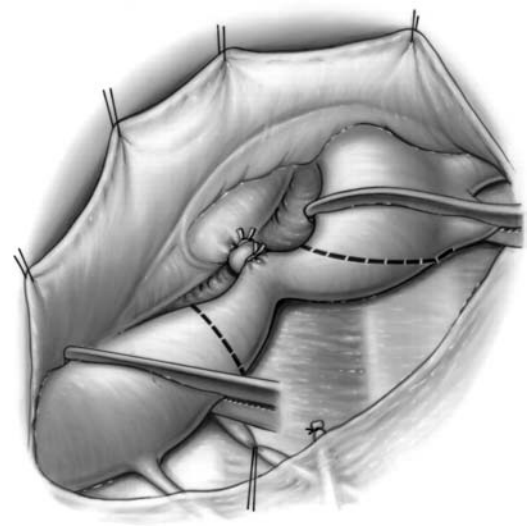

Figure 20.4

not to occlude or to only partially occlude the left subclavian artery in order to allow some collateral flow to the lower part of the body. The coarcted segment is then resected. It is very important to excise all the ductal tissue to minimize the risk of recoarctation. It may be necessary to enlarge the proximal aortic orifice by a cutback toward or into the origin of the left subclavian artery (**Figure 20.5**). The second assistant standing to the right of the surgeon holds the two clamps

Figure 20.5

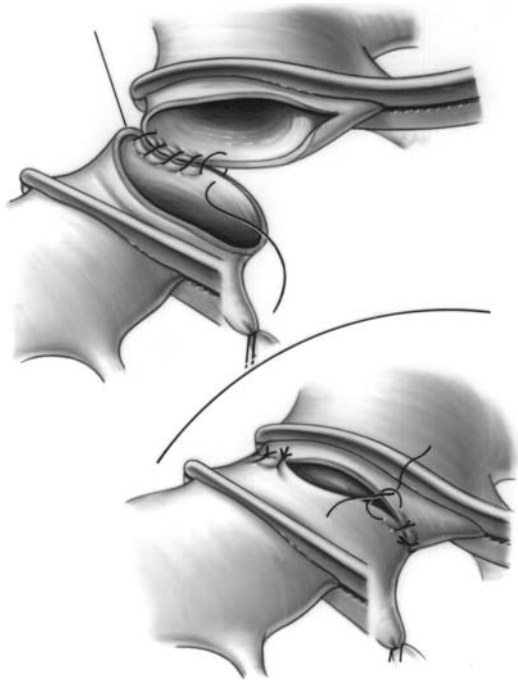

Figure 20.6

about 2 cm apart. Various types of suture techniques and suture materials have been described. The authors currently use an over-and-over running stitch with 7-0 Prolene sutures in infants and 5-0 in children. The first suture is placed from inside into the medial aspect of the distal aorta and is anchored with a rubber-shod clamp. The other needle is then passed from inside to the outside of the proximal aorta. Part of the posterior suture line is performed before the clamps are approximated and the two ends pulled together (**Figure 20.6**). Three or four interrupted sutures may be used anteriorly (**inset, Figure 20.6**). However, the continuous circumferential suture is more haemostatic and the growth of the anastomosis does not seem to be compromised.

Subclavian Flap Technique

This technique was described by Waldhausen and Nahrwold in 1966. The incision in the mediastinal pleura is extended superiorly to the origin of the vertebral artery. Some lymphatic vessels may be oversewn. After mobilization of the aorta and ligation of the persistent ductus arteriosus, a fine braided ligature is placed distally on the subclavian artery. Some authors have suggested ligating the vertebral artery at its origin in order to prevent subsequent steal from the cerebral circulation

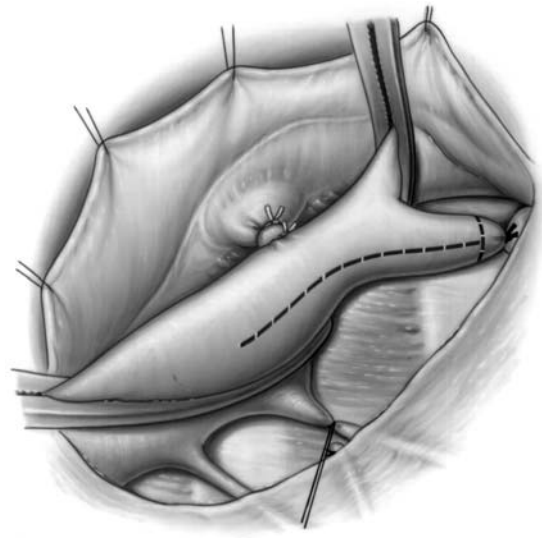

Figure 20.7

to the left upper limb (Hamilton *et al*., 1978). Vascular clamps are then applied across the transverse aortic arch between the left carotid and left subclavian arteries and the descending thoracic aorta, well beyond the level of the coarctation. The lower clamp, usually spoon-shaped, is placed so as to occlude the intercostal arteries arising near the coarctation (**Figure 20.7**).

Alternatively, one curved vascular clamp, such as a Castaneda clamp, can be used to occlude the aorta both distally and proximally. The aorta is then incised from well below the coarctation, across the narrow segment through the isthmus and along the lateral border of the subclavian artery, which is transected proximal to the ligature. In many patients, there is an intimal shelf of tissue at the site of the coarctation. Excision of this shelf is a subject of controversy (**Figure 20.8**)—too radical an excision is blamed for the development of late aneurysms, and failure to excise it is blamed for recurrent coarctation. Currently, the authors do not excise the shelf. The incision in the descending aorta should extend well below (at least 1 cm) the coarcted segment in order to avoid residual coarctation. The distal end of the subclavian artery and the lower part of the aortotomy are then approximated with a fine suture and anchored with a rubber-shod clamp. When this suture is pulled down, the flap is positioned so as to lie within the margins of the aortic incision and is inserted with two longitudinal sutures of 7-0 polypropylene (Prolene), starting at the proximal end. The tip of the flap should be left untrimmed, so as to allow all the tissue of the subclavian artery to enlarge the

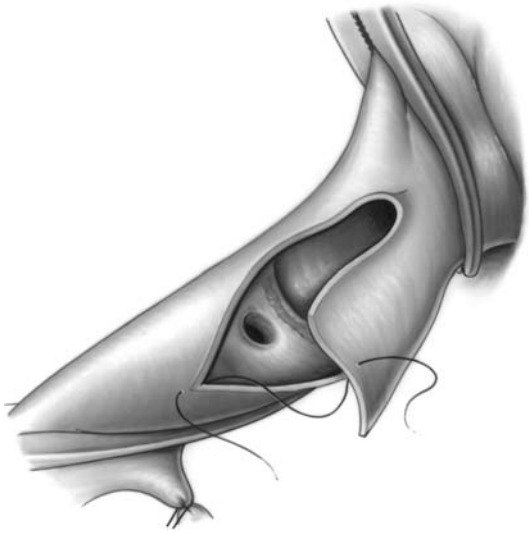

Figure 20.8

narrow segment (**inset, Figure 20.9**) (Hamilton *et al*., 1978). After completion of the anastomosis, the distal aortic clamp is removed to allow the excluded segment to fill with blood and to expel air. The proximal aortic clamp is then slowly removed, and the systemic pressure is carefully watched. Transfusion of blood or crystalloids is often necessary at this time to maintain the blood pressure. The clamp must be re-applied if the blood pressure continues to fall in spite of the transfusion. An infusion of catecholamines is occasionally required. The mediastinal pleura is not routinely closed, and a single pleural drain is inserted before closure of the chest.

Reversed Subclavian Flap

Infants with hypoplasia of the aortic arch between the left carotid and left subclavian arteries can be treated by reversed subclavian flap angioplasty, as suggested by Hart and Waldhausen (1983). The dissection has to be somewhat more extensive, mobilizing part of the left carotid artery and the arch proximal to its origin. The left subclavian is ligated, as in the subclavian flap operation. One vascular clamp is placed across the left carotid artery and the aortic arch proximal to its origin, and the other clamp is placed on the descending aorta (**Figure 20.10a**). The subclavian artery is then transected near to the ligature and opened down toward the aorta and the aortic arch and into the base of the left carotid artery (**Figure 20.10b**). The flap thus created is then sutured in place with 7-0 polypropylene sutures (**Figure 20.11**). The reversed flap for arch hypoplasia can also be done in conjunction

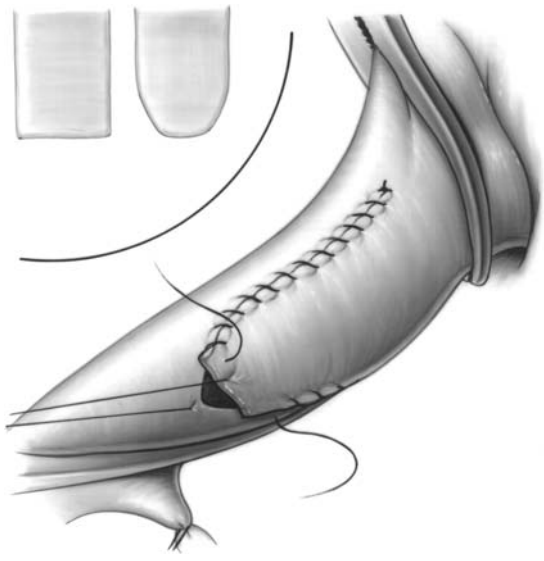

Figure 20.9

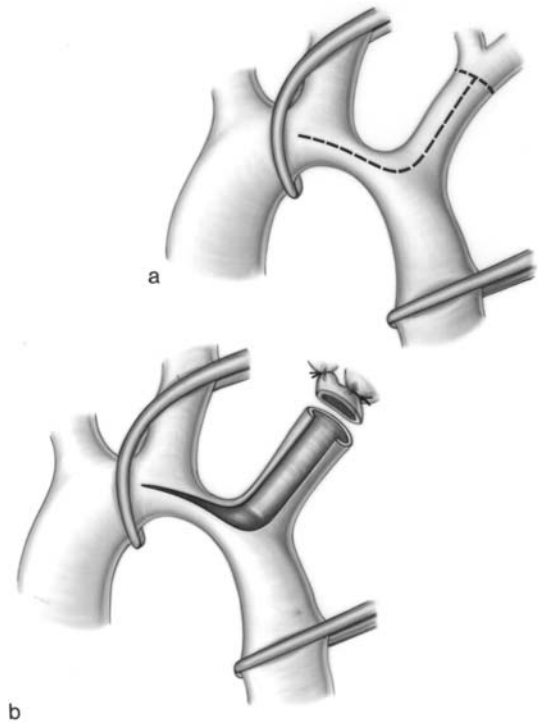

Figure 20.10

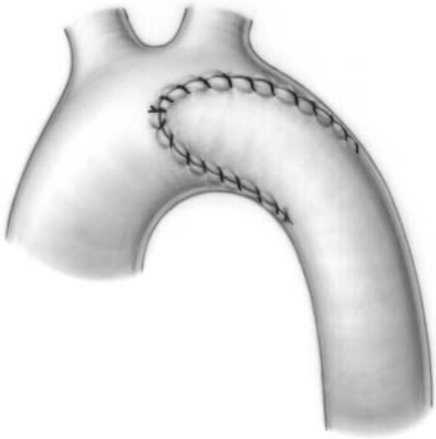

Figure 20.11

with a resection and an end-to-end anastomosis for a juxtaductal coarctation.

Extended Resection and Anastomosis of the Distal Aorta to the Undersurface of the Aortic Arch

If the aortic arch is hypoplastic, resection or repair of the coarctation alone may leave a residual gradient. The definition of aortic arch hypoplasia is difficult. In normal neonates, the segment of the aorta between the left carotid and the left subclavian arteries is always the narrowest part of the arch. Some surgeons (Karl *et al*., 1992) use a formula to define normal size of the arch between the left carotid and left subclavian artery. The dimension (mm) should be body weight + 1 mm. For example in a 3 kg baby, it would be 3 + 1 = 4 mm. Opinions differ: some surgeons consider the use of the extended repair in a high proportion of patients (Trinquet *et al*., 1988); others believe that repair of coarctation alone encourages the growth of the narrow arch and reserve the extended repair for only a very few patients (Jahangiri *et al*., 2000). The first description of the extended repair was published by Amato *et al*. (1977). The descending aorta is first dissected and mobilized towards the diaphragm. All the tissue between the intercostal arteries is dissected free. Proximally, the ductus arteriosus or ligamentum arteriosum, the left subclavian artery, the left carotid artery and the aortic arch are dissected. After the ductus arteriosus is ligated, a curved vascular clamp (Elliott, 1987) is placed across the origin of the left subclavian artery and the left carotid artery and catching part of the distal ascending aorta (**Figure 20.12**). Care must be taken to allow adequate flow into the innominate artery. The distal clamp is placed across the descending aorta, and the coarcted segment with the ductal tissue is resected (**Figure 20.13**). The incision is extended to the undersurface of the aortic arch and onto the distal aorta. A posterior suture line is then placed in an open manner. Clamps are approximated, and the anterior suture line is completed (**Figure 20.14**). This technique achieves excellent reconstruction of the arch but requires more extensive dissection and more time.

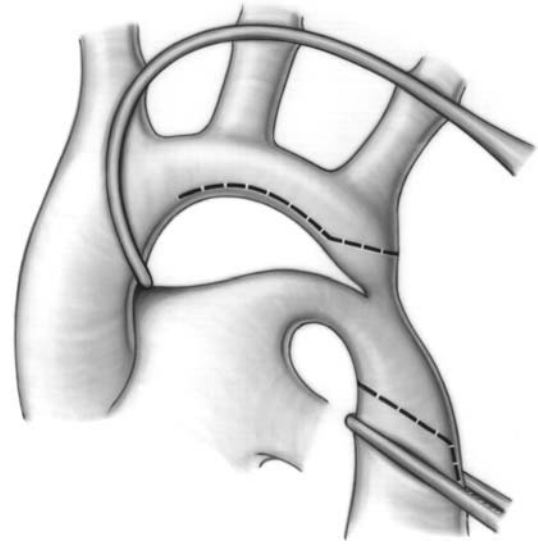

Figure 20.12

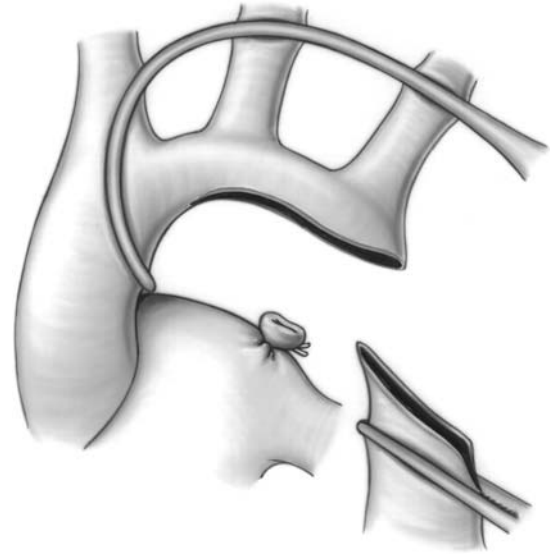

Figure 20.13

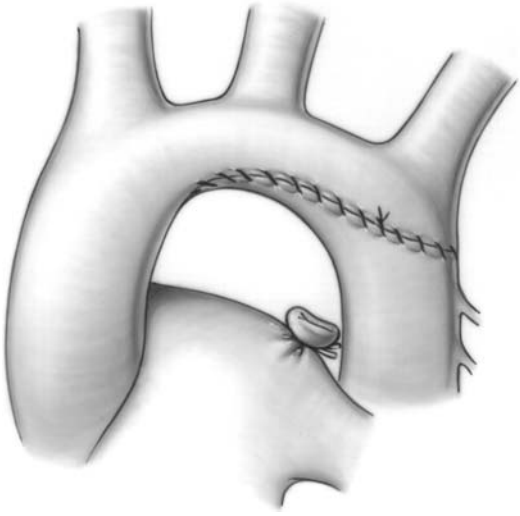

Figure 20.14

Patch Graft Aortoplasty

Direct and indirect isthmoplasty has been described by Bernhard (1949) and Vosschulte (1957). It is an easy operation that does not require such an extensive dissection as do some of the other techniques. The ductus arteriosus or ligamentum arteriosum is ligated and divided, and vascular clamps are placed with minimal interference with the collaterals. The aorta is opened longitudinally; some surgeons recommend careful resection of the shelf. The authors do not resect it. A generous Gore-Tex patch is sutured in place to widen the aorta (**Figure 20.15**). The patch must extend from well above to well below the coarctation. The aortic patch angioplasty has been favoured by several authors (Reul *et al*., 1974; del Nido *et al*., 1986). The technique is easy, but the possibility of late aneurysm formation is a major setback (Ala-Kulju *et al*., 1983; Clarkson *et al*., 1985). Backer *et al*. (1995) recommend patch aortoplasty using a Gore-Tex patch in patients older than 2 years and younger than 16 years. They have observed only one false aneurysm.

Patch aortoplasty may be used occasionally in conjunction with resection and end-to-end anastomosis or with the subclavian flap technique. Once one of these operations has been completed and the residual gradient measured, the clamps are re-applied, an incision is made anteriorly across the narrow area, and a patch is sutured in place. Barratt-Boyes used this modification in 12 patients after resection and in two after the subclavian flap technique; interestingly, aneurysms developed in none of these patients in whom the Dacron patch was used in conjunction with other techniques (Clarkson *et al*., 1985).

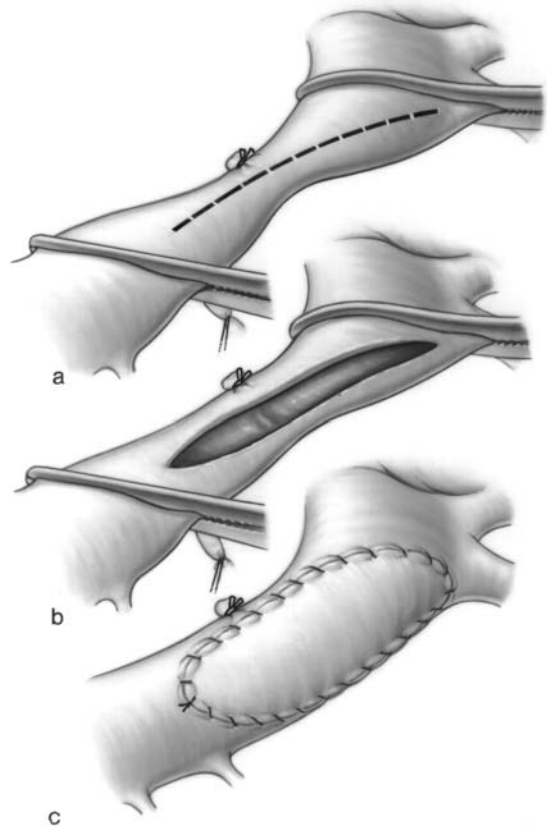

Figure 20.15

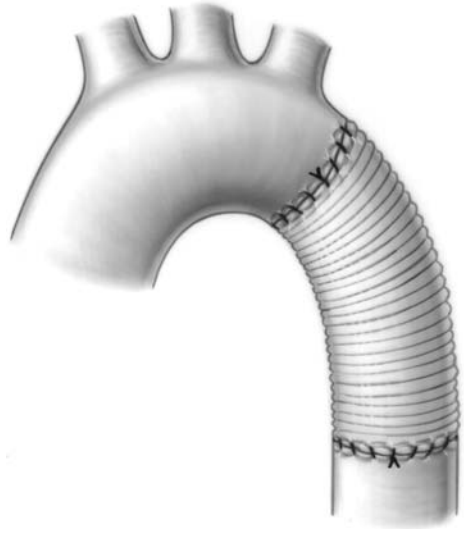

Figure 20.16

In a grown-up with a very long narrow segment, full interposition prosthetic tube can be used (**Figure 20.16**).

Simultaneous Repair of Coarctation of the Aorta and Intracardiac Defects

If intracardiac defects such as VSDs are associated with coarctation of the aorta, the coarctation can be repaired and the pulmonary artery banded, and repair of the intracardiac defect delayed. This approach requires no circulatory arrest, but two operations. Ungerleider and Ebert (1987) performed simultaneous repair of the defects through a midline sternotomy. They used cardiopulmonary bypass with deep hypothermia and either total circulatory arrest or low-flow perfusion. The coarctation was repaired with a patch of pericardium in infants and with Gore-Tex or Dacron in older children. Karl *et al*. (1992) reported their experience, adding isolated myocardial perfusion during arch repair.

The strategy would depend on the confidence of the surgical and the ITU team to deal with neonatal/infant open heart surgery. At our unit the decision for one-stage repair is based on the preoperative neurological status, severity of the arch hypoplasia and the complexity of the VSD. In the usual setting of a moderately large malaligned VSD, the VSD would be closed transatrially during cooling, and the hypoplastic arch/coarctation would be repaired under a brief period of circulatory arrest at 18°C, using either direct anastomosis or a pulmonary homograft patch with its inner curve. The proximal end of the patch should be tailored not to impinge on the ascending aorta (**Figure 20.17**).

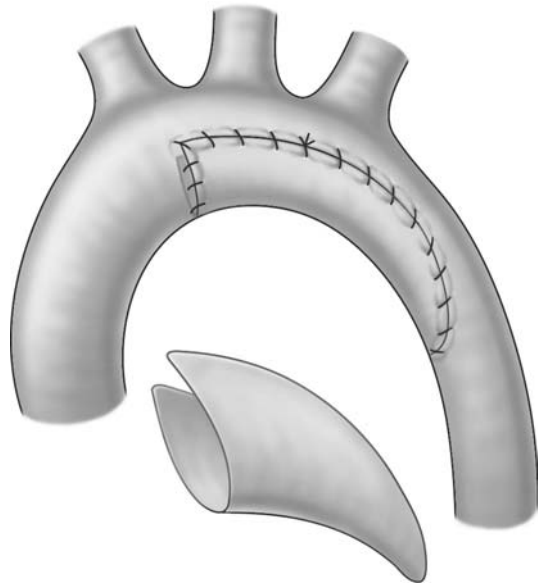

Figure 20.17

EARLY COMPLICATIONS

Bleeding

Bleeding after coarctation repair is not an uncommon complication, particularly in older children with many collateral vessels. The usual principles for preventing and treating the haemorrhage apply.

Paradoxical or Rebound Hypertension

Paradoxical or rebound, hypertension is a common complication, occurring in up to 30% of all patients (Pennington *et al*., 1979). It is less common in infants. Perioperative catecholamine surge and renin production have been implicated (Sealy, 1990). Mild forms are treated by sedation; for severe hypertension, intravenous labetalol (α- and β-blockade) or sodium nitroprusside is given in the immediate postoperative period and is usually followed by oral propranolol.

Paraplegia

Paraplegia is a rare but extremely serious complication of coarctation repair and re-coarctation repair. The estimated incidence was reported as 0.41% (Brewer *et al*., 1972). Lerberg *et al*. (1982) observed paraplegia in 5/334 coarctation repairs (incidence, 1.5%). There was correlation between the length of aortic cross-clamping and paraplegia (mean cross-clamp time in patients who developed paraplegia was 49 minutes). The measures for avoiding paraplegia have been described earlier in this chapter, such as the shortest possible cross-clamp time, avoidance of acidosis and preservation of collateral vessels. In older children, distal aortic pressure should be measured and, if deemed necessary, perfusion of lower body could be indicated. Careful technique of anastomosis and avoidance of haemorrhage should obviate the need for reapplication of the cross-clamp. Moderate hypothermia to 34–35°C may be also helpful.

Renal Failure

Renal failure may occur in severely ill infants. It is a consequence of poor perfusion of the lower part of the body; it may be present before operation and may have to be treated by peritoneal dialysis.

Abdominal Pain

Abdominal pain with the ileus occasionally occurs on the second or third postoperative day. It is probably related

to mesenteric vasculitis, which in rare cases becomes necrotizing and necessitates surgical intervention. Aggressive control of postoperative hypertension has reduced the incidence of this complication (Sealy, 1990).

Chylothorax

The thoracic duct can rarely be injured during dissection. Chylothorax is more likely due to the incision of small lymphatic vessels near the subclavian artery. These lymphatic vessels, if seen, should be oversewn rather then electrocoagulated. If a milky drainage starts appearing from the chest drains, a conservative treatment consisting of a medium-chain triglyceride diet is started. If, however, the drainage is copious, reoperation is indicated. The site of the chylous leak is oversewn.

LATE COMPLICATIONS

Re-coarctation

Re-coarctation occurs in a number of patients, regardless of the method of surgical repair. The incidence of re-coarctation is highest in patients who have undergone operation early in infancy (Hopkins *et al*., 1988; Cobanoglu *et al*., 1985; Kirklin and Barratt-Boyes, 1986), although in some smaller series no incidence of re-coarctation was reported (Hamilton *et al*., 1978; Waldhausen *et al*., 1986). In general, reported incidence is 11–42% (Beekman *et al*., 1981; Hesslein *et al*., 1981; Penkoske *et al*., 1984; Hopkins *et al*., 1988; Kappetein *et al*., 1994). These differences may be to some extent explained by different criteria used by different authors to define re-coarctation.

Williams *et al*. (1980) observed gradients in 54% of survivors after the operation, but only 11 underwent reoperation. It is not easy to distinguish between residual and recurrent coarctation, unless pressures are measured routinely at the end of the coarctation repair. We therefore recommend measuring and recording pressures above and below the site of repair at the conclusion of each primary operation A residual coarctation may result from a technically inadequate repair (**Figure 20.18a**), from hypoplasia of the arch above the coarctation (**Figure 20.18b**) or from the short subclavian flap used for repair (**Figure 20.18c**). Tension on the anastomosis also predisposes to the development of re-coarctation. Persistent systemic hypertension in the upper body, with delayed or even absent femoral pulses, left ventricular hypertrophy, or strain pattern on electrocardiograms, is suggestive of recurrent or residual narrowing. The gradient is assessed by measurement of blood pressure with an appropriately sized cuff in the upper and lower extremities. Doppler echocardiography may obtain an accurate gradient; nowadays, MRI is the investigation of

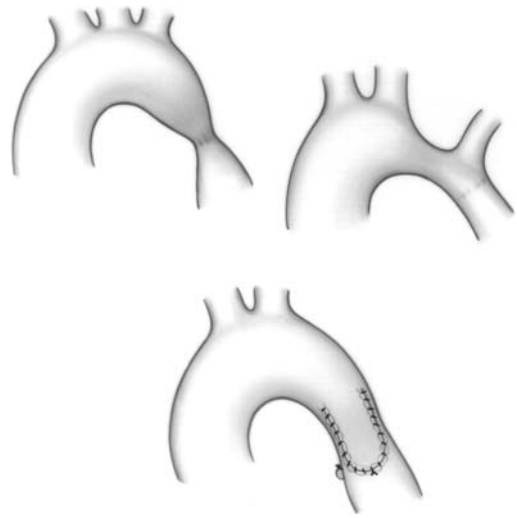

Figure 20.18

choice (Chapter 5). Cardiac catheterization and angiography provide an exact measurement and elucidate the length and degree of the narrowing, as well as a platform for interventional angioplasty.

In general, intervention is indicated when a resting gradient of 20–30 mmHg is found in the presence of significant narrowing on imaging. With the development of interventional cardiology, balloon dilatation of re-coarctation (Lababidi *et al*., 1984) has become a technique of choice for most patients with residual/recurrent coarctation. The freedom from reintervention 12 years after successful dilatation was over 70% (Yetman *et al*., 1997). Stenting in conjunction with dilatation has been used, but the long-term outcome remains uncertain. The details of the balloon dilatation technique, with or without stenting, are described in Chapter 7.

REPAIR OF RE-COARCTATION

If balloon dilatation/stenting is not considered appropriate, surgery may be required. Several surgical techniques have been used. The left side of the chest can be reopened, or a sternotomy performed. In some special cases, a right thoracotomy may be useful for some bypass operations. The criteria for spinal cord protection are similar to those outlined earlier for the repair of primary coarctation, but more care has to be exercised in the absence of significant collaterals. To avoid dissection of the adhesions at the former suture line, the coarctation may be bypassed with a woven Dacron or Gore-Tex graft placed from the base of the left subclavian artery to the distal aorta (**Figure 20.19**) or on the aorta from above to below the

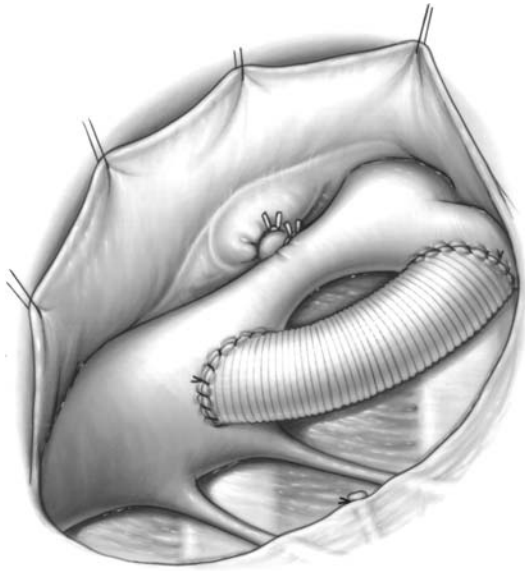

Figure 20.19

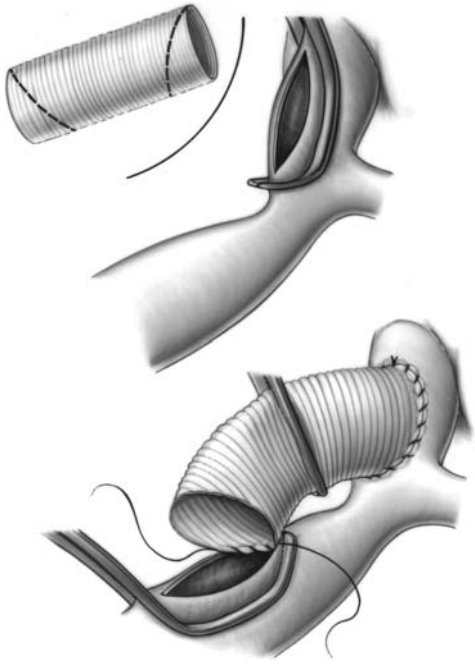

Figure 20.20

narrowing (**Figure 20.20**). If the narrow segment is very long or if the adhesions are particularly difficult to dissect, a long graft may be placed from the ascending to the descending aorta. Another alternative is to place a tube graft through a midline sternotomy, on cardiopulmonary

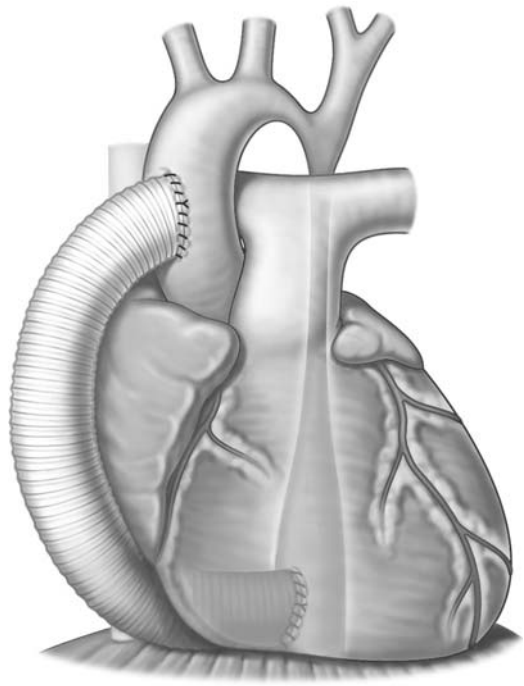

Figure 20.21

bypass and moderate hypothermia, from the ascending aorta to the aorta just above the diaphragm via posterior pericardium opening behind the heart (Kanter *et al*., 2000) (**Figure 20.21**).

RESULTS

During the 1980s the mortality rates in infancy were closely related to the presence or absence of associated lesions and to the age of the patient. In a review of 179 infants undergoing operation for coarctation of the aorta (Hopkins *et al*., 1988), the overall mortality rate was 15%. There was no difference between the types of operative repair used. Only 20 patients had simple coarctation; one died. Of 159 patients with complex coarctations, 26 (16%) died. The mortality was highest in neonates. Trinquet *et al*. (1988) analysed their series of 178 infants younger than 3 months. Of the 63 with simple coarctation, 8% died; of the 47 with associated VSD, 11% died; and of the 68 with complex associated lesions, 37% died. Ziemer *et al*. (1986) reported a 12-year experience from Boston Children's Hospital with 100 neonates: of 29 with simple coarctation, one (3.4%) died; of 32 with associated VSD, four (12.5%) died; and of 39 with complex associated lesions, 12 (30.7%) died.

The advantages of any particular surgical technique remain controversial. A review of 151 infants under age

3 months underwent repair between 1985 and 1990 (Van Heurn *et al*., 1994). Early mortality (8%) was related to the presence of associated major intracardiac defects, preoperative resuscitation and postoperative gradient across the repair. Freedom from re-coarctation was 54% after subclavian flap, 77% after end-to-end anastomosis and 83% after the extended repair. Quaegebeur *et al*. (1994) reported results in 322 neonates from a multi-institutional study; 20 (14%) of 139 neonates died after resection and anastomosis; nine (8%) of 112 died after subclavian flap. In those with single moderate or large associated VSD, the repair of coarctation, pulmonary artery banding and subsequent repair of VSD were associated with the highest 2 year survival of 97%.

We operated on 141 patients for coarctation of the aorta during 2000–2004. End-to-end anastomosis was performed in 116; 22 of those had concomitant PA band. There were two hospital deaths, Shone syndrome in one and very poor preoperative LV function in the other. 25 had subclavian flap and nine had concomitant PA band. All survived. 14 patients had one-stage repair of coarctation and VSD closure. One patient died due to severe hypernatraemia. Among patients over the age of 1 year, mortality rates under 2% have been reported (Koller *et al*., 1987; Kirklin and Barratt-Boyes, 1986).

LONG-TERM FOLLOW-UP

Although coarctation of the aorta seems to be a relatively simple technical problem, the long-term results are rather poor. Clarkson *et al*. (1983) showed that only 20% of patients were alive without a complication and with a normal blood pressure 25 years after coarctation repair. Of Olley's patients, 75% manifested some complication during the long-term follow up (Olley, 1979).

Hypertension

The incidence of persistent postoperative hypertension seems to be related to the age at operation. It is less common among patients undergoing operation before the age of 5 years (Liberthson *et al*., 1979). Recent data (Brouwer *et al*., 1994) suggested that the risk of persistent hypertension does not increase if the operation is performed during the first year of life. Among patients observed for a long period, the incidence of hypertension is higher (Clarkson *et al*., 1983). Even with a successful repair performed in late childhood, the life expectancy is reportedly reduced. It is related to secondary cardiovascular complications (Presbitero *et al*., 1987). Gardiner *et al*. (1994) demonstrated abnormal endothelial responses in normotensive young adults following ''successful'' surgery for coarctation performed in young age.

Aneurysm

An aneurysm at the site of the coarctation repair can develop as late as 20–25 years after surgery. The etiology remains unclear. Differences in compliance of the patch and the aortic wall have been implicated. The mortality rate, both with and without surgery, is considerable (Stark and Pacifico, 1989). Hehrlein *et al*. (1986) reported 18 aneurysms in 317 patients treated by patch aortoplasty. Because almost all the aneurysms described in the literature have occurred in patients undergoing patch aortoplasty (Ala-Kulju *et al*., 1983; Bromberg *et al*., 1989), we do not use and do not recommend this technique, except for special cases.

Aneurysm formation after balloon dilatation at the site of coarctation is of particular concern. Careful follow-up is essential. If the diameter of the aorta continues to increase, urgent attention (intraluminal stenting or surgery) is needed. If surgery is required, the approach would be similar to what was described earlier. Resection of aneurysm on cardiopulmonary bypass with deep circulatory arrest and the use of interposition graft is our technique of choice.

Acknowledgements

Figures 9, 10, 11, 17, 19, 20 and 21 are modified from illustrations (**Figures 9.1, 9.14, 9.15, 9.16, 9.17 and 9.20**) in *Reoperations in Cardiac Surgery*, Stark J, Pacifico AD (eds). Springer-Verlag: London, 1989, with permission of Springer-Verlag.

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21

Interruption of the Aortic Arch

J. L. Monro

Interruption of the aortic arch (IAA) is a rare condition, accounting for about 1% of infants with critical congenital heart disease (Collins-Nakai *et al*., 1976). The natural history is very poor; 80% of children die within the first month if untreated (Van Praagh *et al*., 1971). Since the introduction of prostaglandin (Olley *et al*., 1976) and the improvement in the management of complex neonatal cardiac lesions, the outcome following surgery has improved considerably (Fulton *et al*., 1999; Rousin *et al*., 2002).

ANATOMY

Figure 21.1 shows the three types of interruption (Celoria and Patton, 1959). The interruption may be distal to the left subclavian artery (type A), between the left carotid and the subclavian arteries (type B) or between the innominate and left carotid arteries (type C). Type B is the commonest and type C is very rare. Associated defects usually accompany such interruptions; a ventricular septal defect (VSD), which is virtually always present, as is a persistent ductus arteriosus, through which the blood flows to the descending aorta. Truncus arteriosus and aortopulmonary window and a number of other lesions, such as univentricular heart, may also be associated. The VSD is commonly infundibular (subarterial, muscular or perimembranous) and is often associated with left ventricular outflow tract obstruction LVOTO (Van Praagh *et al*., 1971; Freedom *et al*., 1977). The obstruction has been attributed to a leftward displacement of the infundibular septum (Van Praagh *et al*., 1971). Occasionally there are multiple VSDs and LVOTO may be present initially or develop later. In addition, the aortic valve is bicuspid in approximately 30% of affected patients, the aortic ring may be small, and the ascending aorta is usually small. The aorta is occasionally right-sided, and an aberrant right subclavian artery originating from the descending aorta is common in type B but also occurs in type A.

In addition, 22q11 deletion and DiGeorge's syndrome with absent thymus is quite frequently associated with type B interruption, resulting in hypocalcaemia, immunological problems and developmental delay. The use of irradiated blood is recommended.

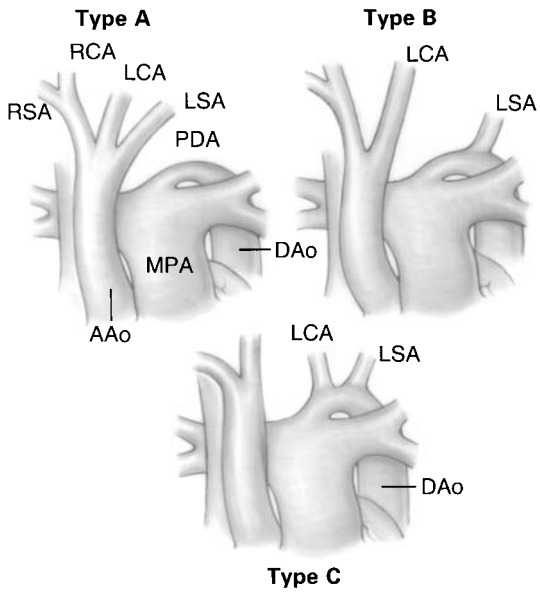

Figure 21.1

DIAGNOSIS AND MEDICAL MANAGEMENT

Soon after birth, the ductus arteriosus begins to close and severe congestive heart failure, metabolic acidosis and anuria develop. Femoral pulses, which may have been present before this, disappear with the clinical deterioration. The baby should be admitted to a neonatal unit as soon as possible and prostaglandin should be given at this stage to reopen the duct. This, together with other resuscitative measures, including ventilation

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if necessary, should greatly improve the metabolic state and femoral pulses reappear. Before 1976 when prostaglandin became routinely used, these neonates were extremely ill and required urgent operation; even so, the outcome was often fatal. However, since the introduction of prostaglandin, the management has been dramatically improved and surgery can be delayed for a day or two if necessary. Echocardiography is used to confirm the diagnosis, and the site and length of the interruption. The position and size of the VSD should be noted, or whether there is a univentricular heart. Any posterior displacement of the infundibular septum should be seen and measurements of the LVOT made, as this has a bearing on the type of repair and whether relief of the outflow tract obstruction is necessary. Measurements of the LVOT also give the best indication of whether postoperative obstruction is likely (Apfel *et al*., 1998). A Z-score of less than −5 is very predictive of severe postoperative obstruction problems (Salem *et al*., 2000). Because echocardiography gives such a good idea of the anatomy, other investigations, such as cardiac catheterization, are rarely necessary. In some patients the diagnosis may have been made by echocardiography antenatally. In this case delivery should be arranged in a hospital with a neonatal unit and prostaglandin given promptly.

SURGICAL TECHNIQUE

The surgical approach depends on the presence of associated defects. In the majority of patients, only an isolated VSD and persistent ductus arteriosus are present. Such patients will have been resuscitated after prostaglandin infusion and are likely to be in the first month of life. Although many different methods of managing this typical situation have been advocated, most surgeons agree that a one-stage correction through an anterior approach is the method of choice. This is basically the method used by Trusler, who reported the first successful neonatal correction without the use of a graft in a patient with type B aortic arch interruption (Trusler and Izukawa, 1975). Barratt-Boyes had previously been the first to describe the successful correction of a type A interruption in an infant but used a Teflon graft in the repair (Barratt-Boyes *et al*., 1972). Murphy *et al*. (1973) reported successful correction of a type B aortic arch interruption in a 3 day-old infant, but they used a vein graft from the patient's father. However, although a two-stage approach has mostly produced worse outcomes, occasional good results have been achieved with the repair of the aorta through the left chest, together with pulmonary artery banding and later debanding and VSD closure (Mainwaring and Lamberti, 1997). Turning down the left carotid or the left subclavian artery can be a satisfactory way to obviate tension on the anastomosis. The important thing is to achieve survival of the patient. This approach is analogous to repair of coarctation of the aorta with VSD. Although most surgeons would still repair a coarctation through the left chest and band the pulmonary artery as necessary, there is an increasing trend to close a VSD and repair the coarctation from the front (Murakami and Kado, 2001; Uemura *et al*., 2001). This seems a somewhat illogical state of affairs, but the important thing is to have a live patient.

Midline Approach

Traditionally, profound hypothermia with total circulatory arrest has been used to provide optimal operating conditions. This can be used for the whole procedure or preferably just for the arch repair, and bypass is restarted for closure of the VSD. Recently more surgeons have used low-flow cerebral perfusion during the arch repair (Asou *et al*., 1996; Takeuchi *et al*., 1998; Korkola *et al*., 2002) and the descending aorta may be perfused as well (Uemura *et al*., 2001; Murakami and Kado, 2001). With the very small modern aortic cannulae and moderate cooling, perfusion of the innominate artery at reduced flow can provide safer operating conditions (Tchervenkov *et al*., 2001). **Figure 21.2a** shows how the small aortic cannula is inserted into the distal ascending aorta for the initial cooling bypass.

If surface cooling is used initially down to 28°C, it is only necessary to use one aortic cannula. Otherwise it is safer to place a second arterial cannula through the pulmonary artery and duct into the descending artery, with snaring of the duct when on bypass.

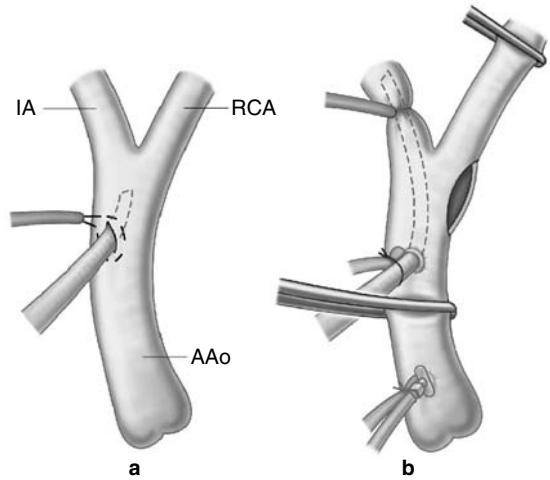

Figure 21.2

Once moderate hypothermia (e.g. 28°C) has been achieved, bypass is briefly stopped while the aortic cannula is advanced into the innominate artery and the artery snugged around the cannula (**Figure 21.2b**) Bypass is recommenced with a reduced flow (10 ml/kg/min has been recommended by (Takeuchi *et al*., 1998), but a flow of 30–40 ml/kg/min is probably safer and cooling to 20°C may result in better subsequent renal function. This should probably equate to a right radial artery pressure of 25–30 mmHg (Pigula *et al*., 2001). The ascending aorta is clamped proximal to the cannula with a reduced flow and cardioplegic fluid is then infused into the ascending aorta.

Good mobilization of the aorta and head vessels is essential, as tension must be avoided. This can mostly be achieved while the patient is being cooled. Once the innominate artery is perfused and the cardioplegia is in, the duct cannula is removed, the left carotid and left subclavian arteries are snugged and the descending aorta clamped as low as possible. The duct is divided and all duct tissue removed from the descending aorta, which is then brought up and anastomosed direct to the distal ascending aorta and possibly into the base of the left carotid artery (**Figure 21.3**). It may be necessary to sacrifice the left subclavian artery and care must be taken to preserve the recurrent laryngeal nerve. The anastomosis should not be too low or compression of the left main bronchus can occur, leading to bronchomalacia. This is particularly likely if the truncus arteriosus is associated (Sano *et al*., 1990).

There are several ways of closing the VSD. This is usually done through the right atrium. If the defect is high, it may be necessary to close it through a right ventriculotomy. It is also possible to close a high VSD through the large pulmonary artery. Some surgeons have used polydioxanone (PDS) absorbable sutures for the aortic anastomosis, in the hope that the anastomosis is more likely to grow, but probably 7-0 Prolene sutures are more satisfactory because they run more easily and break or unravel as the child grows.

Another method of repairing a type B aortic arch interruption through the front is to turn down the left carotid artery to reconstitute the aortic arch (Monro, 1981). If the left carotid artery is of an adequate size, this is a satisfactory method involving less mobilization of the descending aorta, and the distal end of the left carotid artery can be anastomosed to the right carotid artery in order to reconstitute circulation to the left side of the head, although this should probably only be done with deep hypothermia and circulatory arrest (**Figure 21.4**). Satisfactory growth is achieved by this method, but again it is important to remove all ductal tissue from the descending aorta; otherwise, anastomotic stenosis can occur (Monro *et al*., 1996). In a type A interruption, it is possible to similarly turn down the left subclavian artery to reconstitute the arch if this artery is of an adequate size (**Figure 21.5a, b**).

In older patients, the ductus arteriosus has been used successfully to reconstitute the aorta (Monro *et al*., 1977). Tissue from this duct can grow, but it may be

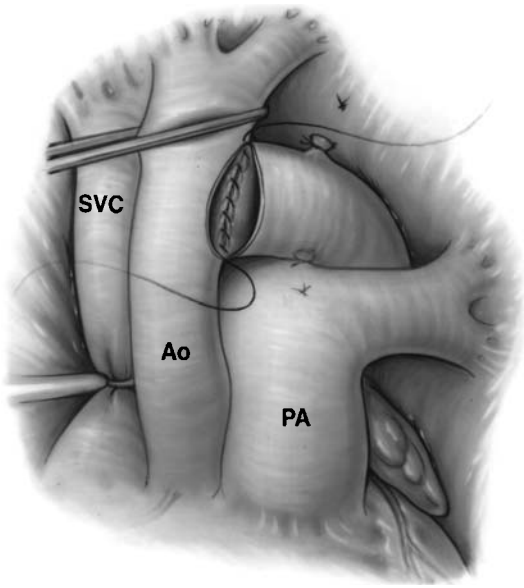

Figure 21.3

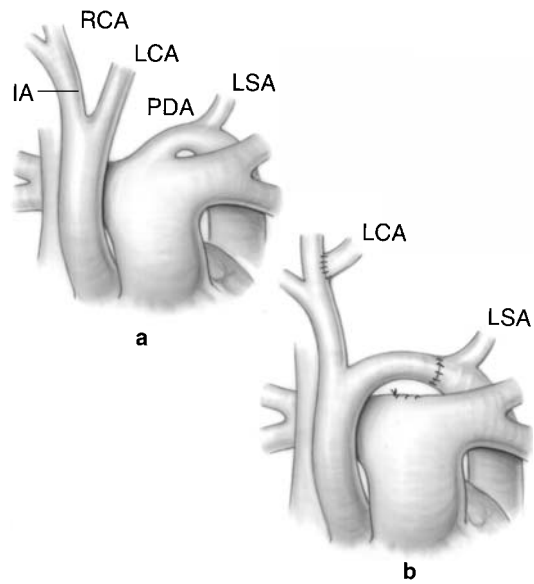

Figure 21.4

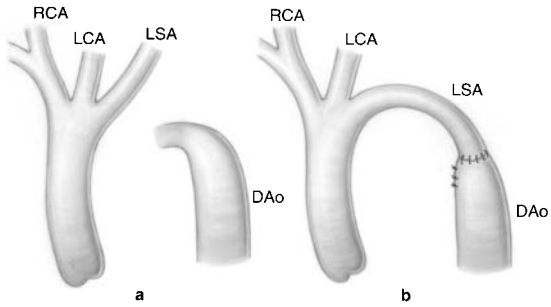

Figure 21.5

necessary to add to the ductal tissue at the time of initial repair with either a flap of the pulmonary artery or a pericardial patch. In infants under 3 months of age (who are, of course, the vast majority) and particularly in those under 1 month of age, the ductal tissue is likely to cause stenosis, and therefore this method is applicable only to the rare older patients. Some workers have advocated the insertion of prosthetic tube grafts, but this is unnecessary and inevitably leads to the need for reoperation.

Severe LVOTO is one of the major problems in correction of IAA. It should be detected and measured by ehocardiography preoperatively. Although resection of obstructing tissue is sometimes feasible, the best technique is to suture the VSD patch to the left side of the infundibular septum, so that it is pulled anteriorly out of the way in systole (Luciani *et al*., 1996; Watanbe *et al*., 1998). If the LVOTO is extremely severe, it may be better to perform a modified Norwood procedure (Jacobs *et al*., 1995).

Some severe associated defects may also be dealt with at the same time through an anterior incision. Truncus arteriosus and aortopulmonary window can certainly be corrected successfully at the same time through a midline approach, as can total anomalous pulmonary venous drainage. Transposition of the great arteries with associated aortic arch interruption probably should be dealt with by anatomical correction of the great arteries (switch procedure) and direct arch repair in one operation, and the Taussig–Bing type of double-outlet right ventricle should be dealt with similarly.

Repair of the Aortic Arch through the Left Chest

When the arch interruption is associated with a double-inlet ventricle, it is best to perform the arch repair through the left side of the chest, and if the pulmonary flow is increased, as is most commonly the case, the main pulmonary artery should be banded. The type of repair of the aortic arch again depends on the position of the interruption and the size of the head vessels, but the goal is to avoid the use of prosthetic grafts, because this inevitably necessitates an additional operation to increase the size of the aortic arch.

A type A interruption is actually somewhat similar to an exaggerated coarctation, and therefore the left subclavian artery can be turned down and anastomosed to the descending aorta (see **Figure 21.5b**); alternatively, the descending aorta can be brought up and anastomosed to the base of the left subclavian artery (**Figure 21.6**). In a type B interruption, the left subclavian artery, if large, can be divided and turned up to be anastomosed to the base of the left carotid artery, thus reconstituting the arch (**Figure 21.7a, b**), or if the left subclavian artery is too small, the left carotid artery can be turned down to the descending aorta (**Figure 21.8**). Again, it is very important to remove all ductal remnants. The theoretical disadvantage of this latter approach is that the blood flow to the left side of the head may be compromised, but

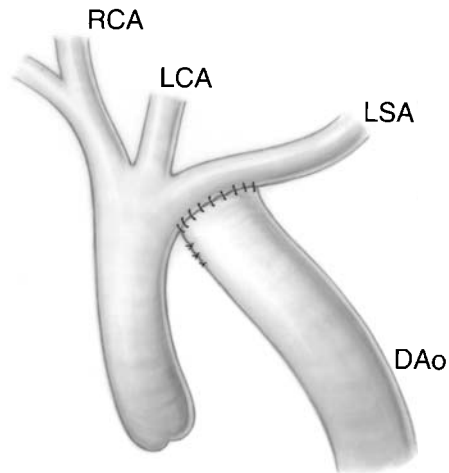

Figure 21.6

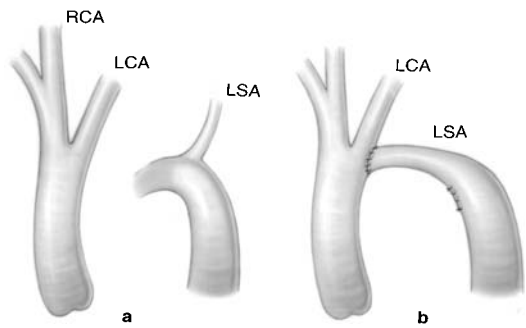

Figure 21.7

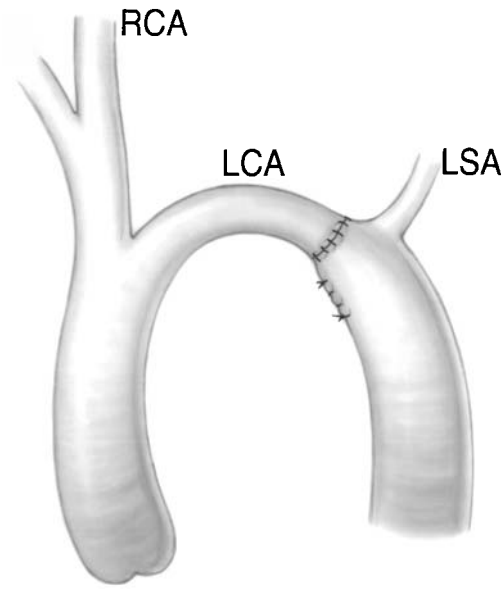

Figure 21.8

at this young age collateral flow is probably adequate; however, it is advisable to use moderate hypothermia to about 32°C (which can easily be achieved with surface cooling) in order to help protect the brain and spinal cord at the time of operation. In either the carotid turn-down or the subclavian turn-up procedures, the new arch, if inadequate in size, can be patched with a piece of pericardium, which still allows for growth. The pulmonary artery is banded during the same operation, but these patients must be carefully observed because they are particularly prone to development of subaortic stenosis, which must be relieved either before or at the time of the definitive Fontan procedure. Care also must be taken to ensure that the band is tight enough, so as to prevent pulmonary vascular disease, which would jeopardize a subsequent Fontan procedure.

RESULTS

The outcome following surgery for IAA must be divided into those undergoing surgery through the left chest and those undergoing complete repair through the midline.

The early survival for those having aortic repair and pulmonary artery banding through the left chest is reasonably good. However, there is a fall-off during the years before a Fontan type of procedure can be done.

For those undergoing a complete repair through the front, results have improved remarkably during the last 30 years. Prior to the use of prostaglandin a high mortality was inevitable, as many infants were moribund preoperatively. However, since 1976, when prostaglandin became available, surgeons have converted to the one-stage approach and the early mortality has dropped steadily (Serraf *et al*., 1996; Schreiber *et al*., 1997). Because of the rarity of the condition, there are few large series and most of them include patients from more than 10 years ago. In the mid-1990s, some large series were reporting an early mortality in the region of 20% (Jonas *et al*., 1994; Schreiber *et al*., 1997). Since then, and particularly with the use of minimal circulatory arrest and cerebral perfusion, the best reports have an early mortality of less than 10% (Fulton *et al*., 1999; Roussin *et al*., 2002).

The one-stage vs. two-stage approach for IAA + VSD + PDA has been debated for years. Twenty-five years ago there were only a few reports of successful one-stage repairs but, as with other conditions (e.g. Fallot's tetralogy), surgeons have increasingly used a one-stage approach. This has the obvious advantage of only having to undergo one operation rather than two. There is no evidence that a one-stage approach has a higher incidence of subsequent reoperation.

LATE FOLLOW-UP

Late survival obviously depends on early survival, and interrupted arch has in addition a high incidence of reoperation, which in turn carries a further mortality. Survival at 4 years has been quoted as 63% in a multi-institutional study of 174 patients (Jonas *et al*., 1994) but the patients were operated upon in 1987–1992. In a series of 79 patients during 1985–1995, the 5 year survival was 70% (Seraff *et al*., 1996).

In a large multicentre report from North America, a group of 472 neonates operated on between 1987 and 1997 had an overall survival of 59% at 16 years. This included those operated on from both an anterior and lateral approach; 25% had further arch intervention and late survival was less for those who had LVOTO (McCrindle *et al*., 2004). The early mortality among the 51 patients with associated truncus arteriosus was 67%. This seems remarkably high, but underlines how important it is to have these large multicentre studies, where one sees the true overall picture of mortality as compared with that reported by smaller groups with good results. Because only the best results tend to be reported, one gets an impression of what can be achieved, rather than the overall mean. Unfortunately, in this large series the approach has not been separated out, so one is unable to compare the outcomes in the commonest situation (IAA + VSD + PDA) repaired by a one-stage frontal approach, as opposed to a two-stage (lateral incision with repair of IAA and pulmonary artery banding and later debanding and VSD closure). It would need a

controlled trial of these two techniques on matched groups of patients suitable for an anterior approach to determine the best method.

Reoperation has been common after repair of IAA, due mainly to the occurrence of LVOTO and aortic arch obstruction, and less commonly for residual VSD and tracheal or bronchial compression. As many patients have bicuspid aortic valves, operations on the aortic valve for repair or eventual replacement are quite common.

In a series of 94 patients operated on in Munich between 1975 and 1999, the 10 year freedom from reoperation was 49% (Schreiber *et al*., 2000). Sell *et al*. (1988) had an incidence of 21% for late LVOTO and 24% for recurrent or persistent aortic arch obstruction.

Anastomotic Obstruction

Anastomotic obstruction has occurred quite frequently following IAA repair and is usually due to tension on the anastomosis, caused by inadequate mobilization of the head vessels and descending aorta, or to inadequate resection of all duct tissue. Since these two factors have been well reported and surgeons are taking care to avoid them, the incidence of reoperation for this cause has fallen. If it does occur, balloon angioplasty is the preferred treatment for late anastomotic obstruction.

Left Ventricular Outflow Tract Obstruction

The occurrence of postoperative LVOTO is mostly related to its presence preoperatively. Salem *et al*. (2000) identified an aortic valve diameter < 4.5 mm (Z-score < -5) to be predictive of subsequent LVOTO and it usually occurs within 1 month (Apfel *et al*., 1998). Hopefully, with better preoperative echocardiographical assessment and the suturing of the VSD patch to the left side of the infundibular septum (Luciani *et al*., 1996), this will become less common.

An alternative method of overcoming the problem of extreme LVOTO is to patch the VSD into the pulmonary artery, which in turn is divided and the proximal end anastomosed to the ascending aorta. Right ventricular-to-pulmonary artery continuity is restored with a valved conduit (Yasui *et al*., 1987).

SUMMARY

Most neonates presenting with this rare condition are initially stabilized in the first week or so of life with prostaglandin and then go on to one-stage repair of the aorta and VSD through a midline incision. Improved bypass techniques allow for continued cerebral perfusion with minimal circulatory arrest. Good mobilization of the aorta and its branches with removal of all duct tissue allows for a satisfactory anastomosis of the descending aorta to the ascending aorta, with much less frequent recurrence of arch obstruction. LVOTO remains a serious complication and suturing the VSD patch to the left side of the septum may overcome this.

Patients with complex associated lesions, such as univentricular heart, could have their aortic interruption repaired through the left chest with concomittant pulmonary artery banding, and will require further surgery.

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22

Vascular Rings

M. R. de Leval and M. J. Elliott

Malformation of the aortic arch, which may cause compression of the trachea, oesophagus or both, is considered in this chapter even though some aortic arch malformations do not constitute a complete ring. Edwards (1948) proposed a classification for these anomalies that was derived primarily from a developmental viewpoint of the aortic arch system. Formation of the usual left aortic arch system is based on regression of the eighth segment of the right dorsal aortic root. Most vascular rings result from either lack of regression of this segment or an abnormal regression of other segments or from both. They arise more commonly at four locations of a hypothetical double aortic arch and bilateral ductus arteriosus (Edwards, 1948), as illustrated in **Figure 22.1**. Associated tracheobronchial stenosis/malacia is common. It will be discussed in the latter part of this chapter.

SURGICAL ANATOMY

Double Aortic Arch

Double aortic arches are the most common types of vascular rings. They represent a persistence of both right and left dorsal aortic arches. They arise from the ascending aorta anterior to the trachea, cross dorsally on both sides of the trachea and oesophagus, and join posteriorly to form the descending aorta. Each arch gives rise to its common carotid and subclavian arteries, which usually arise as separate branches without an innominate artery. The ductus arteriosus may be present on the left or right side, or it may be bilateral. The relative size of the lumen, the patency of each arch, and the position of the upper descending aorta are all variable. If the descending aorta is on the left, there is a left anterior arch and a right posterior arch. A major posterior arch is more frequently encountered when the descending aorta is on the left (**Figure 22.2**). In most cases of right-sided upper descending aorta, the right anterior arch is the major arch. The relative sizes of the arches are important because at operation the surgeon divides the smaller one.

Atresia of one segment of the arch is also sometimes seen. The ductus arteriosus is most often left-sided. If short, it pulls the pulmonary artery backwards and compresses the trachea from the front, thus contributing to the ring.

Vascular Compression Associated with a Left-sided Aortic Arch

Anomalous Innominate Artery

Compression of the trachea by the innominate artery was recognized by Gross and Neuhauser in 1948. The anatomical substrate of the compression of the trachea is still controversial. In some patients, the innominate artery arises more posteriorly from the aorta than normally; in others, a congenitally short artery may give rise to tracheal compression. In many instances, the anatomy of the great arteries is normal and the compression is located in the thoracic inlet.

Anomalous Left Carotid Artery

This anomaly is similar to the one just described. It occurs when the common carotid artery arises farther to the right than usual.

Aberrant Right Subclavian Artery

Aberrant right subclavian artery is the most common type of vascular anomaly of the aorta and occurs in approximately 0.5% of the population. This malformation results from an embryological interruption of the right fourth arch between the right common carotid and the right subclavian arteries. In this anomaly, the right subclavian artery arises as the last branch of the aortic arch and passes obliquely upward from left to right to reach the right arm. The aberrant vessel most commonly passes behind the oesophagus and the trachea; in rare instances, it passes in front of the trachea.

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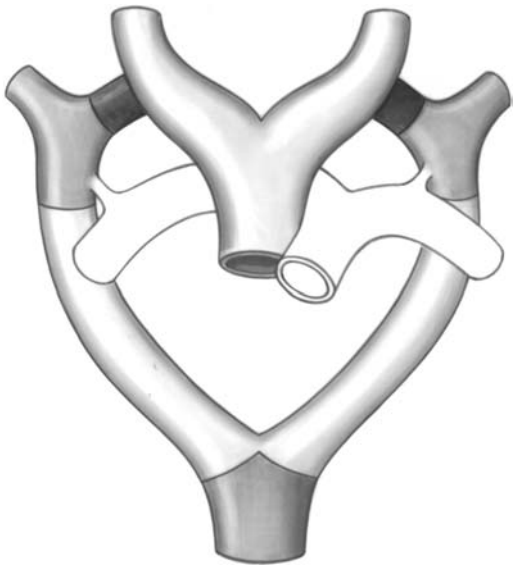

Figure 22.1

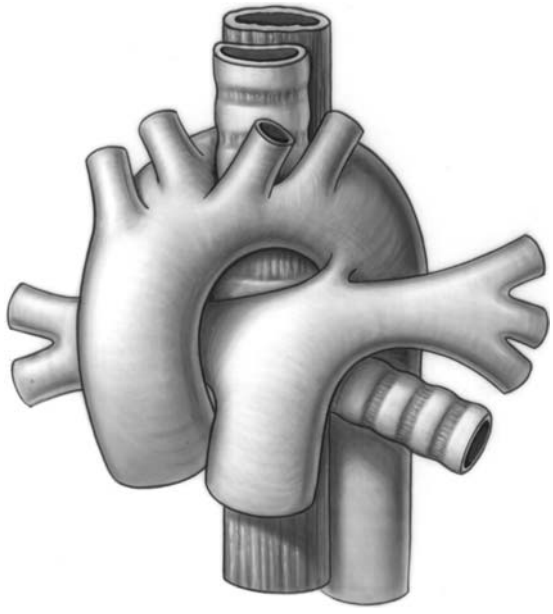

Figure 22.2

Vascular Compression Associated with Left-sided Aortic Arch and Right-sided Upper Descending Aorta

This is a rare anomaly, in which the ascending aorta passes upwards and arches to the left and around the trachea, proceeds backwards and crosses to the right behind the oesophagus, and continues to a right-sided upper descending aorta. The ductus arteriosus may connect the right pulmonary artery to the right-sided upper descending aorta or to the origin of the aberrant right subclavian artery, thus forming a complete ring around the oesophagus and the trachea (**Figure 22.3**). This anomaly results from interruption of the right aortic arch between the right subclavian artery and the right ductus arteriosus.

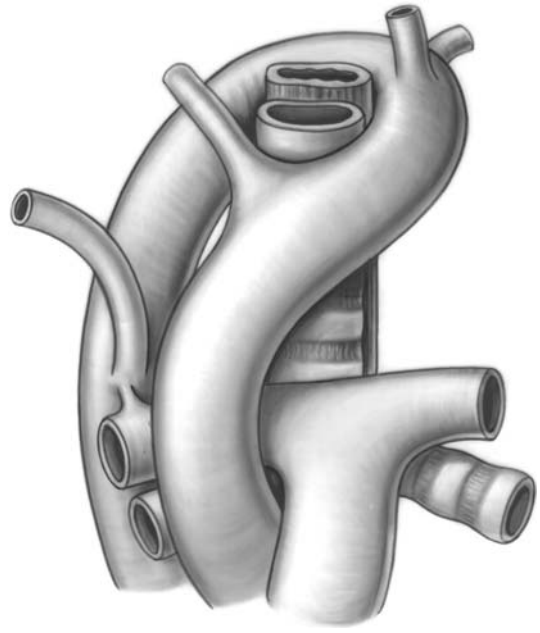

Figure 22.3

Vascular Compression Associated with Right-sided Aortic Arch

The right-sided aortic arch passes backwards to the right of the trachea and oesophagus to join the descending aorta. This malformation results from regression of the left dorsal aortic root. Regression between the left ductus arteriosus and the descending aorta produces a mirror-image branching of the aorta with the left innominate, right carotid, and right subclavian arteries arising in that order (**Figure 22.4**). A left-sided ductus arteriosus may connect the left pulmonary artery to the subclavian portion of the innominate artery, or it may connect the left pulmonary artery to the upper descending aorta. In the latter case, a complete vascular ring exists. Regression of the left fourth arch between the left common carotid and left subclavian arteries results in a right aortic arch with aberrant left subclavian artery (**Figure 22.5**). In

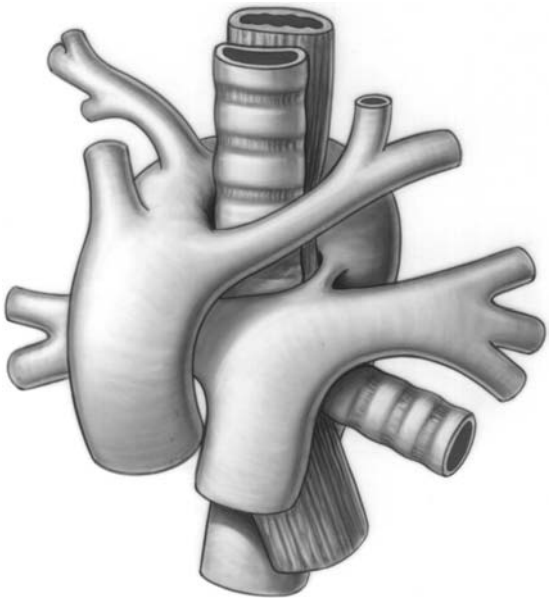

Figure 22.4

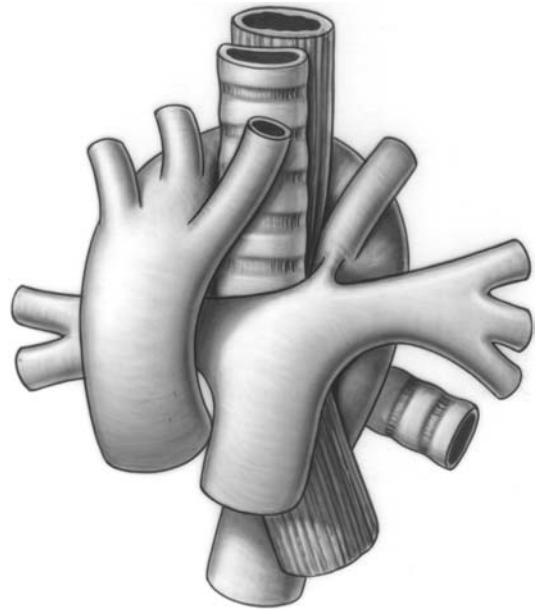

Figure 22.5

this arrangement, the left common carotid artery is the first branch to rise from the aortic arch, followed by the right common carotid and right subclavian arteries, and then followed by the left subclavian artery as the last branch passing to the left behind the oesophagus. A right-sided aortic arch, an aberrant left subclavian artery and a left-sided ductus arteriosus also form a complete ring that consists of the ascending aorta and pulmonary artery anteriorly with the aortic arch on the right and the ductus arteriosus and the left subclavian artery on the left. Aberrant subclavian arteries may exhibit a bulbous malformation at their origin that produces a large, concave retro-oesophageal indentation (Kommerell's diverticulum; Kommerell, 1936). Associated congenital heart anomalies are very common among cases of mirror-image branching but are fairly uncommon in the presence of right-sided aortic arch with aberrant left subclavian artery. In comparison with a double aortic arch, the ring formed by a right-sided aortic arch with aberrant left subclavian artery and left-sided ductus arteriosus is loose in most instances.

Cervical Aorta

This is a rare anomaly in which the aorta ascends into the neck on the right or left side, forming a pulsatile mass in the supraclavicular region. Several morphological types have been described according to the side of the aortic arch (contralateral or ipsilateral) and the origin of the head vessels.

Pulmonary Artery Sling

In this anomaly, an aberrant left pulmonary artery arises from the right pulmonary artery, passes over the right main bronchus posteriorly and to the left, and traverses the mediastinum between the trachea and the oesophagus to enter the hilus of the left lung (**Figure 22.6**). The sling thus formed compresses the proximal portion of the right main bronchus and the distal end of the trachea. Hypoplasia of the distal trachea with complete cartilaginous rings is frequently associated with pulmonary artery sling (Sade *et al*., 1975). Complete tracheal rings are present in 50–65% of patients with pulmonary artery sling (Fiore *et al*., 2005).

DIAGNOSIS

A vascular ring may be asymptomatic and thus may be discovered incidentally during chest radiography, arteriography or a gastrointestinal series or at post mortem examination. When symptoms are present, they are caused by compression of either the trachea or the oesophagus or both. The severity of the symptoms and the age at which onset occurs tend to reflect the degree of compression, which is in turn somewhat characteristic of the particular vascular ring. In most cases, the patient is typically a small infant, is poorly developed and has a history of respiratory difficulties and repeated chest infections dating from birth. Of 51 patients with vascular rings operated on at the Hospital for Sick Children, Great

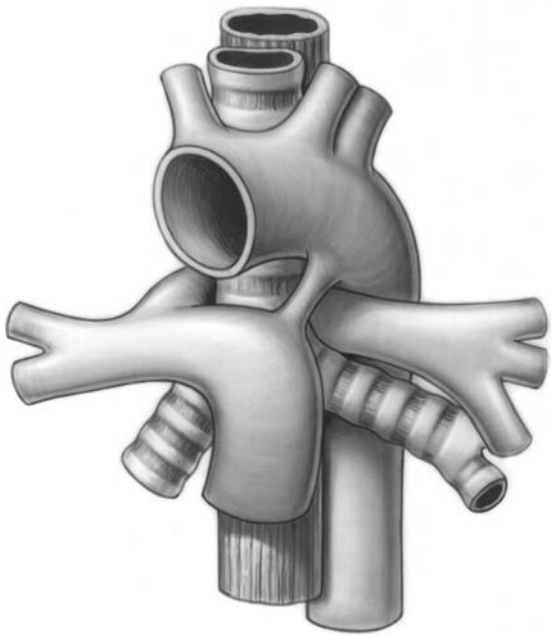

Figure 22.6

Ormond Street, in London between 1968 and 1980, 25 were symptomatic at birth and all but two exhibited symptoms before the age of 6 months (Roesler *et al*., 1983). The term ''reflex apnoea'' was introduced by Fearon and Shortreed (1963) to describe episodes of respiratory arrest initiated by irritation of the area from tracheal compression. Respiratory symptoms are often exacerbated by flexion of the neck and are relieved when the head is extended. Feeding can exaggerate the stridor and the wheezing, and this may cause cyanosis. Feeding difficulties tend to become more apparent when patients begin eating solid foods.

Plain chest X-ray and barium oesophagogram are the most important classic investigations in a patient suspected of having a vascular ring. The chest X-ray shows the laterality of the aortic arch in most instances, and tracheal compression can often be seen on penetrated films. Obstructive emphysema and atelectasis can also be related to bronchotracheal compression.

Anteroposterior and lateral oesophagograms show indentations or compression produced by the vascular anomaly. In double aortic arches, the barium swallow study reveals indentations on both sides of the oesophagus in the anteroposterior view. The larger arch usually produces the larger indentation. The anteroposterior view shows an oblique linear defect (**Figure 22.7a**). In the lateral view, the posterior arch is shown to produce a posterior indentation (**Figure 22.7b**).

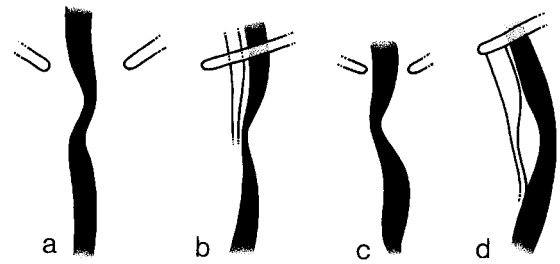

Figure 22.7

In the presence of aberrant right subclavian artery, there is also a posterior impression on the lateral projection; **Figure 22.7c** illustrates the findings in patients with left-sided aortic arch and right-sided persistent ductus arteriosus. Anterior indentation of the oesophagus is, however, typical of a pulmonary sling (**Figure 22.7d**).

Whereas the barium oesophagogram used to be the single most important study to evaluate vascular rings, it has now been supplanted by computed tomography (CT) and magnetic resonance imaging (MRI). With high-speed high-resolution CT scanning, general anaesthesia is usually not necessary, which is not the case for MRI. Three-dimensional (3D) reconstructions of CT scans are particularly useful (Backer *et al*., 2005).

Two-dimensional (2D) echocardiography with Doppler colour-flow mapping has proved to be of limited value in the diagnosis of vascular rings because of poor windows available and because vascular ring segments without lumens cannot be displayed. 2D echocardiography, however, remains a very useful tool to establish the diagnosis of pulmonary artery slings. All patients with vascular rings should have a preoperative echocardiogram to rule out or diagnose associated intracardiac anomalies, which are present in more than 10% of cases (Backer *et al*., 2005).

SURGICAL TECHNIQUE

Double Aortic Arch

The operation is performed through a left-sided lateral thoracotomy in the fourth intercostal space. The two arches are dissected anteriorly and posteriorly, and the ductus arteriosus or ligamentum arteriosum is divided in all patients to provide more mobility of the great vessels. In case of an atretic segment, the ring is divided at the level of that segment. In other cases, the smaller ring is most commonly divided distal to the subclavian artery. If the two arches are of equal size, it is preferable to divide the right arch in order to restore normal anatomy with ample relief of compression. **Figure 22.8** shows a

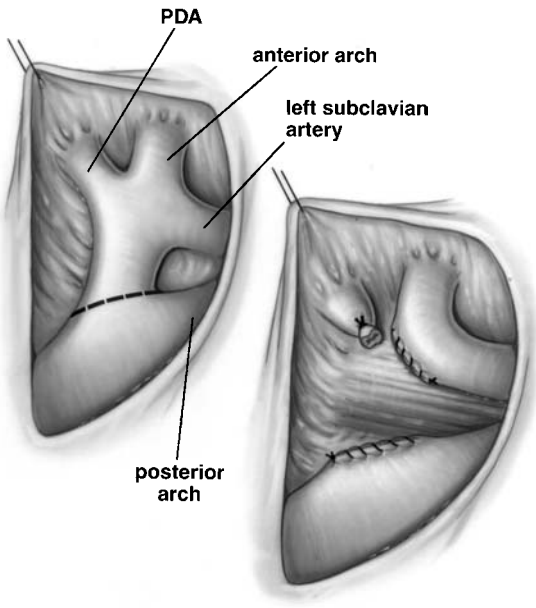

Figure 22.8

double aortic arch with a major posterior arch and a left-sided ductus arteriosus, after division of the ring. The dotted line indicates the site of division of the smaller arch distal to the origin of the left subclavian artery. The ductus arteriosus has been ligated and divided, the anterior arch has been divided between vascular clamps distal to the origin of the left subclavian artery, and the ends are oversewn. The oesophagus and trachea are then dissected thoroughly, and all fibrous bands between these two structures are divided. The left recurrent laryngeal nerve is carefully identified and protected from injury.

Vascular Compression Associated with Left-sided Aortic Arch

Anomalous Innominate Artery

The surgical treatment of this anomaly consists of suspending the innominate artery from the sternum. This procedure can be performed from either side. We prefer the left-sided thoracotomy approach. The pericardium is incised anterior to the left phrenic nerve, and two or three interrupted sutures of heavy monofilament are used to approximate the adventitia of the aorta or innominate artery to the posterior aspect of the manubrium. When it is obvious from the arteriogram that the origin of the innominate artery is abnormal, a midline sternotomy incision can be performed to reimplant the innominate artery anteriorly to the aorta (Binet and Langlois, 1977). In our experience the need for this operation has been very exceptional.

Aberrant Right Subclavian Artery

This is usually asymptomatic. When responsible for symptoms, aberrant right subclavian artery can be divided through a left-sided thoracotomy. As this may lead to late development of a subclavian steal syndrome, it has been suggested that the subclavian artery be reimplanted (Siderys, 1969).

Left Aortic Arch with Right-sided Upper Descending Aorta

In symptomatic patients, the surgical treatment consists of dividing the ductus arteriosus or ligamentum arteriosum through a right-sided lateral or posterolateral thoracotomy incision.

Vascular Compression Associated with Right-sided Aortic Arch

Surgical treatment of this condition also consists of dividing the ductus arteriosus or the ligamentum arteriosum. Division of the left subclavian artery is not generally necessary for relieving tracheal compression. Patients with an anomalous left subclavian artery and Kommerell's diverticulum are at risk of developing severe tracheal compression. Backer *et al*. (2002, 2005) have recommended resecting the diverticulum and reimplanting the left subclavian artery to the left carotid artery as a primary operation. The operation is performed through a left lateral thoracotomy. After dissection of the vascular structures, the patient is anticoagulated (1 mg/kg heparin). The base of Kommerell's diverticulum is taken in a side-biting clamp and the distal subclavian artery is occluded with a vascular clamp prior to its division. The diverticulum is resected and its base is oversewn. The left subclavian artery is then anastomosed to the origin of the left carotid artery.

Cervical Aorta

Tracheo-oesophageal compression, when present, is caused by a ring formed by the retro-oesophageal aorta, the ductus arteriosus and the main pulmonary artery. Division of the ductus arteriosus alone has been reported to relieve the symptoms in several cases.

Pulmonary Artery Sling

The most common procedure is the division and reimplantation of the left pulmonary artery. This was

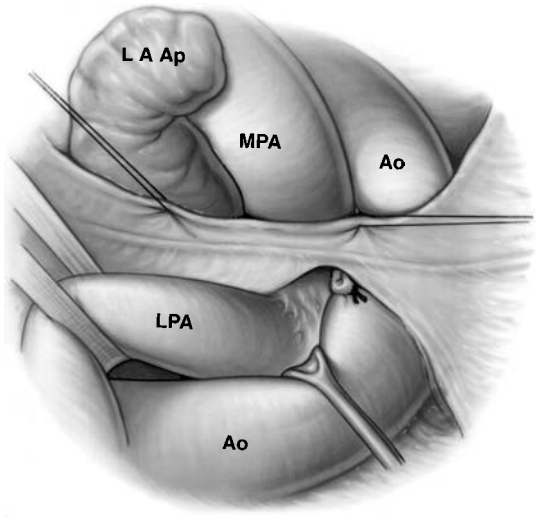

Figure 22.9

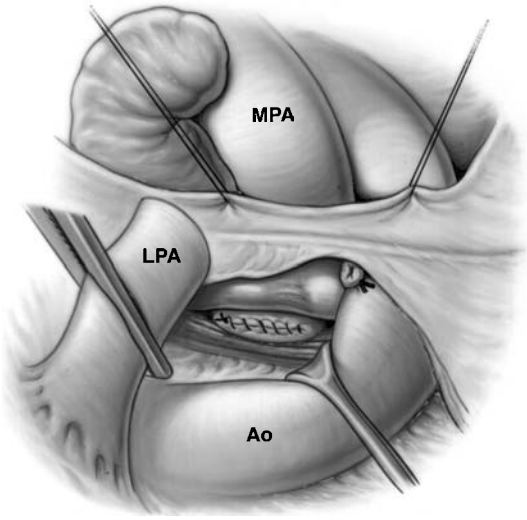

Figure 22.10

originally described by Potts *et al*. (1954). Through a left-sided thoracotomy in the fourth intercostal space, the mediastinal pleura is incised, the ligamentum arteriosum or persistent ductus arteriosus is divided, and the left pulmonary artery is dissected between the trachea and the oesophagus (**Figure 22.9**). After systemic heparinization (1 mg/kg body weight), the left pulmonary artery is divided between two vascular clamps as close to the right pulmonary artery as possible. Care must be taken not to obstruct the trachea with the vascular clamp. The proximal stump is oversewn (**Figure 22.10**) and the main pulmonary artery is then dissected through an opening in

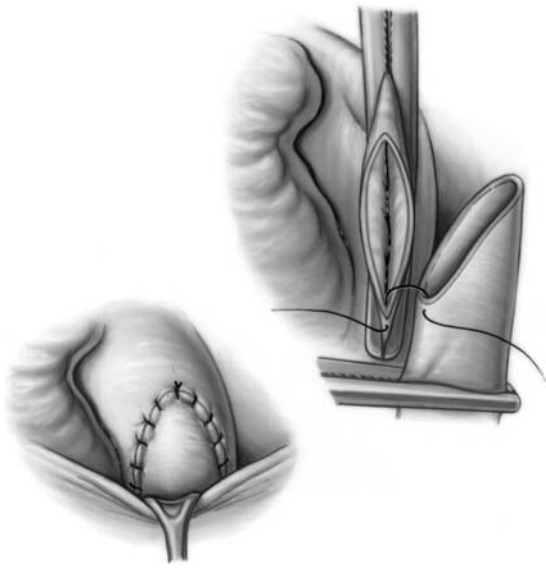

Figure 22.11

the pericardium in front of the left phrenic nerve. A partial occlusion clamp isolates a portion of the main pulmonary artery, which is then incised. The left pulmonary artery is trimmed obliquely and brought through a pericardial incision posterior to the phrenic nerve. An end-to-side anastomosis between the left pulmonary artery and the main pulmonary artery is then created (**Figure 22.11**), using fine monofilament suture (Dunn *et al*., 1979). The left thoracotomy approach remains a satisfactory option for patients who do not require tracheal and/or intracardiac surgery. However, some authors (Fiore *et al*., 2005) have recommended repair of the vascular sling through a median sternotomy incision on cardiopulmonary bypass even for those patients.

TRACHEAL STENOSIS AND MALACIA ASSOCIATED WITH VASCULAR RINGS

The anatomical descriptions of the various rings are described earlier in this chapter, and this section is devoted solely to the diagnosis and management of the airway component of the defect. By definition, vascular rings compress the major airways. Respiratory signs and symptoms, usually in the form of biphasic stridor, represent the most common mode of presentation. Often the compression is relatively mild at birth, but as the child grows, its airway and the structures comprising the vascular ring grow, then the space for the airway becomes less and symptoms develop. This means that many patients present between the ages of 6 and 18 months.

It is important to realize that airway compression alone is not the complete story. There is an important association between vascular ring abnormalities and the presence of complete tracheal rings with, usually, long-segment congenital tracheal stenosis (LSCTS), particularly with pulmonary artery sling. Thus, the airway compression may be in the form of tracheobronchomalacia, in which continuous vascular compression is associated with soft cartilage support in the airway or true tracheal stenosis. The correct management depends on the thorough assessment of the airway and a rational decision tree. Having said that, it is possible to use common sense, based on the frequency of the associations of severe airway obstruction with particular types of vascular ring. Thus, most forms of double aortic arch are rarely associated with complete tracheal rings and true stenosis and so one can proceed to division of the ring and careful follow-up to assess the progress of airway symptoms. However, in the presence of pulmonary artery sling, LSCTS is frequent (60% in our practice) and formal airway assessment should be mandatory. We are now firmly convinced that the management of the airway is a complex, specialized and multidisciplinary task. In our view such care should take place in specialized units where the complex postoperative care can be delivered by those with both appropriate skills and time.

Assessment of the Airway

Clinical examination is of little value in defining the morphology of the airway problem. Echocardiography can define the vascular relationships and may suggest a narrow airway, but cannot tell you what is the nature of the narrowing. Thus, in any child in whom significant airway obstruction is suspected we (Elliott *et al*., 2003) recommend the following strategy of investigations.

High-resolution CT Scan

Modern scanners are capable of outstanding resolution of detail and, in the context of this diagnosis, have the supreme advantage of demonstrating perfectly the spatial relationships of vascular and airway structures. In our view and those of others (Chen *et al*., 2003), it is vital to use no greater than 0.5 mm slices and post-hoc 3D reconstruction is very valuable in choosing approach and assessing risk. Virtual bronchoscopy can also be created from the 3D dataset and this has some value in follow-up. It cannot yet demonstrate complete tracheal rings. Further, as with all digital data interpretation, one must not forget the degree of operator dependence and remain vigilant to data from supportive investigations.

Bronchoscopy and Bronchography

These are combined together for a purpose. In children with severe tracheal stenosis, the airway is often critically narrow, perhaps down to 1 mm in transverse diameter, thus bronchoscopy can often be performed safely only using a small (currently 2.2 mm) fibre-optic bronchoscope. The bronchoscope cannot usually be passed beyond the stenosis in severe cases, and to demonstrate the distal anatomy better it is necessary to perform a bronchogram, details of which we have published elsewhere (Mok *et al*., 2005). Bronchography demonstrates not only the morphology of the airway and permits its classification (Kocyildirim *et al*., 2004b), but also give some clue as to the dynamics of the airway and thus the presence or absence of severe distal malacia, one of the key risk factors in outcome (Kocyildirim *et al*., 2004a). Recently, we have been developing the technique of endobronchial ultrasound for this purpose, and preliminary data are encouraging.

Pressure-flow and Pressure–Volume Loops

In ventilated children, detailed airflow dynamics are helpful in judeging severity and for providing baseline data for postoperative assessment in the ICU. The resultant data should be discussed in a multi-disciplinary setting, since the relevant postoperative care may be complex and involve several disciplines. We have demonstrated that cross-skilling between team members greatly improves both the quality and efficiency of care (Kocyildirim *et al*., 2004a).

Management of the Major Airways in Association with PA Sling

The greatest likelihood for tracheal surgery at the time of vascular ring repair is with a diagnosis of pulmonary artery sling. Video clips of the procedure can be seen at www.congenitalheartsurgery.com The key prognostic information required to plan the management of long-segment tracheal stenosis seen in PA sling is the presence or absence of complete tracheal rings, together with the severity and length of the stenosis. As Backer *et al*. (1999) have pointed out, there is often an increased number of complete rings in this setting and the trachea is, as a result, unduly long. Only recently has a categorization system been devised (Kocyildirim *et al*., 2004b), so there are no data indicating exactly which anatomic variants warrant primary tracheal repair. We take the view that if the stenosis is severe enough to give symptoms or require ventilation, and if there are complete tracheal rings, then concomitant repair of the trachea with the PA sling is the correct management. If symptoms are not severe, and there are no tracheal rings,

then a lateral approach to the PA sling alone is likely to suffice.

Let us assume that we are fixing both lesions simultaneously. In our view, the correct strategy for repair of the PA sling is to perform division and reimplantation via a median sternotomy as Backer *et al*. (1999) described. We do not support the procedure originally described by Jonas *et al*. (1989), in which the trachea is divided and repaired posterior to the repositioned LPA, which is left undisturbed. We have seen both severe LPA stenosis and tracheal stenosis late after this procedure.

Implantation of the Left Pulmonary Artery

The reimplantation operation is performed on cardiopulmonary bypass (single venous cannulation is sufficient unless there are other intracardiac lesions, which should also be repaired), usually normothermic perfusate and without aortic cross-clamping. The pulmonary artery is identified to the right of the aorta and careful dissection continues until the origin of the LPA from the MPA is identified. The LPA passes posteriorly around the back of the trachea in close proximity to the right main bronchus. During the dissection, the recurrent laryngeal nerve and blood supply to the right main bronchus are vulnerable. Once the structures (MPA, RPA, LPA and trachea) have all been identified, vascular slings can be passed around the blood vessels. Sometimes the origin of the R upper lobe artery is in close proximity to the LPA origin and this requires a little extra care to avoid damage. Using the sling on the LPA as traction towards the surgeon, the areolar tissue around the LPA between the trachea and oesophagus can be mobilized until the LPA itself is freely mobile. The LPA is then ligated a little way from its origin. We find it an advantage to leave the tails of this ligature long, for reasons which will become clear. If the RPA is empty of blood, then the LPA can simply by cut away from the RPA, leaving a short stump to suture, otherwise clamping the vessel using a side-biting Castaneda clamp is helpful. Either way, the defect in the RPA should be repaired by two-layer closure, using a fine (7-0) suture, preferably aligned transversely to prevent any narrowing of the RPA.

Next, the left pleura should be opened widely, and the LPA in the hilum identified and mobilized. Care has to be taken to ensure that the main LPA rather than its branches is identified. We place a right-angled instrument behind the LPA at this site and pull the vessel up. The previously applied ligature will shortly come into view and it is preferable to use this loop as the major source of traction to avoid further damage to the intrapulmonary vessels. A small mosquito clip can then be applied to the ligature to maintain the correct orientation of the LPA for subsequent reimplantation. The LPA needs to be reimplanted into the MPA. We have found that this is made much easier if any left-sided ductal ligament is divided between ligatures, as the MPA becomes more accessible to a side-biting clamp. It is then necessary to decide whether the LPA is delivered to the MPA behind or anterior to the left phrenic nerve. In our experience, this decision is best individualized, since the phrenic nerve position on the left pericardium is remarkably variable. The nerve must not be damaged and the LPA must not be obstructed. If the decision is to deliver the LPA behind the phrenic nerve, then an opening must be made in the pericardium behind the nerve. If anterior, then the pericardium can simply be divided down to the position of the nerve and the LPA positioned directly onto the MPA.

Next the LPA can be clamped with a Castaneda clamp close to the hilum, and a side-biting clamp positioned obliquely on the MPA. An incision is made in the MPA and the LPA is transected and spatulated to maximize the anastomotic area. A continuous 7-0 prolene suture is used for the anastomosis.

Tracheal Repair

Once the LPA has been reconnected, it is time to repair the trachea. Over recent years, many different types of repair have been undertaken, ranging from simple end-to-end repair, various forms of patch tracheoplasty, tracheal homograft implantation to slide tracheoplasty. A review of these methods is beyond the scope of this chapter and the reader is referred to more comprehensive reviews elsewhere (Backer *et al*., 2001; Elliott *et al*., 2003). Over the last decade, it has become clear that the slide tracheoplasty (Tsang *et al*., 1989) is probably the best option for repair. It can be performed with low mortality, lower morbidity than other techniques and seems to result in good medium-term quality of life with growth of the trachea. It is now our procedure of first choice in primary tracheal stenosis (Kocyildirim *et al*., 2004a).

Slide tracheoplasty

Is possible only with extensive mobilization of the trachea, and is easiest in small children whose tissues are anyway highly mobile. The operation should be performed on cardiopulmonary bypass in almost all children, and careful preparation of the operative field is helpful. The innominate artery, innominate vein, superior vena cava, aorta, and right pulmonary vein should be mobilized and vascular silastic slings passed around them. The trachea is most easily found via a median sternotomy at the superior end of the incision between the innominate and left carotid arteries. It

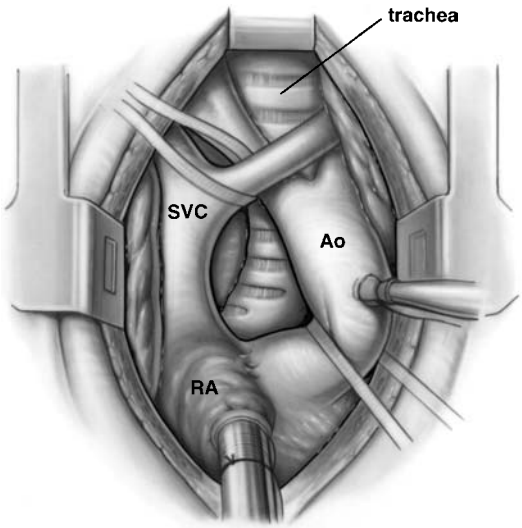

Figure 22.12

can also be readily located above the RPA between the aorta and superior vena cava (**Figure 22.12**). All approaches to the trachea must be strictly over its anterior midline to avoid any damage to the recurrent laryngeal nerves, which lie closely applied in the groove between trachea and oesophagus. By taking this midline approach, it is easy to divide the pretracheal fascia directly onto the trachea and gradually mark out its entire length, externally identifying the extent of the stenosis. Extensive mobilization of the trachea is often needed. It is extremely important to avoid damage to the recurrent laryngeal nerves during mobilization. This can be achieved by making the dissection extremely close to the trachea, whilst preserving as much areolar tissue as possible. A combination of sharp and blunt dissection suffices, and limited use of diathermy. Once the trachea has been mobilized an assessment must be made, as to the length of the slide necessary to correct the stenosis, and a decision made as to whether the slide should extend to one or other bronchus.

Details of the many technical variations of slide tracheoplasty are published elsewhere (Beierlein and Elliott, 2006). The trachea is then transected across the mid-point of the stenosis and an anterior incision is made along the distal segment until normal trachea or bronchus is identified (**Figure 22.13a**). This must be beyond the end of the complete tracheal or bronchial rings and is often 1 or 2 rings into the bronchus. A similar incision is made posteriorly on the upper segment (**Figure 22.13b**),

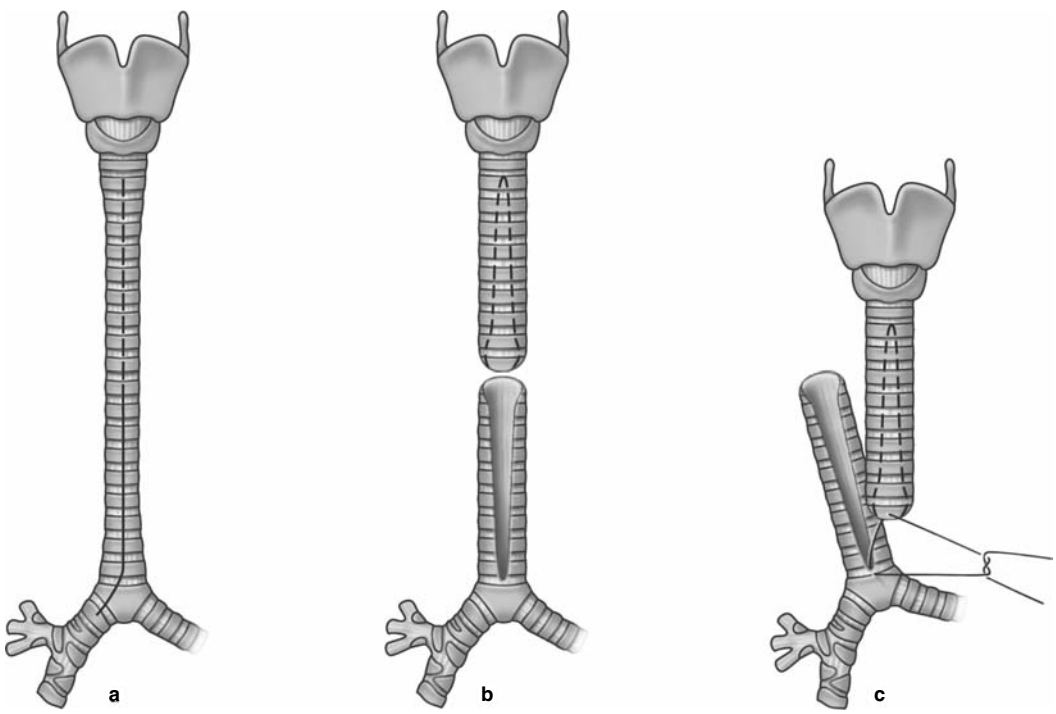

Figure 22.13

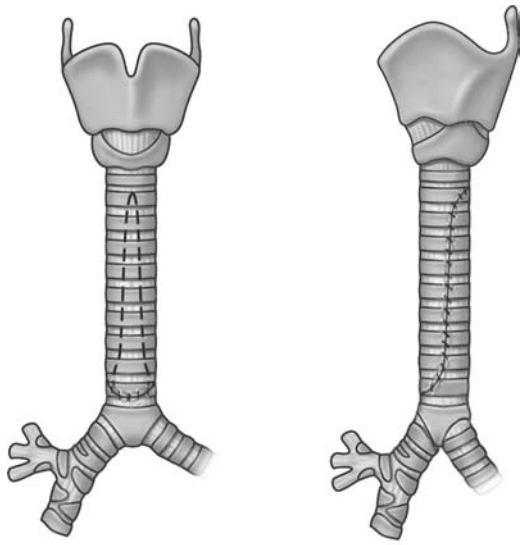

Figure 22.14

again extending right up to normal trachea, occasionally right up to the cricoid cartilage. Using sutures applied to each segment a temporary slide should be performed to ensure adequate mobilization of the trachea (**Figure 22.13c**). If there is excess tension, then the bronchi, posterior pericardial reflection, carinal nodal tissue and hilar reflections of the pleura may all need to be mobilized, and this can produce up to a centimetre of additional length. Superiorly, a hyoid release of the larynx may also provide extra length for the slide.

The anastomosis is completed using slightly everting horizontal mattress sutures of 5-0 or 4-0 PDS sutures, attempting to lay the sutures in a submucosal plane (**Figure 22.14**). It is best to place three or four sutures before tying them down, to distribute the tension more evenly. The anastomosis can be sealed with Tisseal fibrin glue after testing for air leak under a pool of saline. A check fibre-optic bronchoscopy (video-recorded) to confirm integrity and the quality of the repair is mandatory, and is easiest done whilst still on CPB. After completion of the procedure, it is advisable to leave the closed chest irrigated with weak Betadine for 48 hours to reduce the considerable chances of mediastinitis.

Other Options for Repair

These include pericardial patch (Idriss *et al*., 1984) and homograft techniques (Jacobs *et al*., 1999). We have abandoned these after years of experience because of the high complication rate. Recurrent stenosis can be dealt with by balloon dilatation in the first instance, stenting if that fails and, for serious recurrence, tracheal homograft. Tissue engineering and stem cell research will hopefully result in suitable patch material being developed and the more complex patients will benefit from the development of absorbable stents.

POSTOPERATIVE SURVEILLANCE

Postoperative surveillance is a multidisciplinary task. We perform regular fibre-optic examinations of the airway in the ICU during the first 5 days of elective ventilation. At 5 days, a bronchoscopy and bronchogram are performed to confirm the safety of extubation, which is done under bronchoscopic control. The risk of granulation tissue obstructing the airway is high for a while until the relatively ischaemic anastomosed trachea re-establishes a stable blood supply. During this phase, regular balloon dilatations (such as angioplasty) may be required to compress the granulations and prevent ischaemic stricture. Long-term follow-up is necessary and should comprise bronchoscopy, bronchography and respiratory function monitoring. Little is known about the very long-term results and quality of life after this procedure.

MANAGEMENT OF TRACHEOBRONCHOMALACIA

The management of malacia associated with vascular rings and slings is more controversial and associated with both institutional and patient-related variation in practice. As indicated above, malacia is usually relieved by removing the vascular compression. This must therefore always be the first step. If malacia persists, then several options should be considered:

1. ***Tracheostomy and long-term ventilation*** followed by non-invasive ventilatory home support (BIPAP). Malacia usually resolves with time in small immature babies. We have found this to be less reliable after vascular compression and thus rarely use this approach.
2. ***External splinting/support.*** The malacic segment can be wrapped in ringed Gore-Tex tubing and supported by sutures through the Gore-Tex, as suggested by Hagl *et al*. (1997). We have found this method useful, but the gap between the airway and the Gore-Tex fills with amyloid material over time and the long-term results are not known.
3. ***Internal stenting.*** We use internal stenting with Palmaz, silastic or self-expanding stents from time to time, but never if there is residual vascular compression, since erosion and haemorrhage will

surely follow. The results are highly variable and beyond the scope of this chapter.

In summary, airway problems in vascular rings may be severe. If long-segment tracheal stenosis with complete tracheal rings is present, repair with slide tracheoplasty at a specialized unit is recommended. Postoperative care is complex and skilled. The management of malacia should be individualized, with conservative treatment preferred.

RESULTS

Early diagnosis, prompt surgical treatment, avoidance of interoperative damage to the trachea, and proper postoperative care should result in very low mortality rates after division of vascular rings. Between 1968 and 1980, we operated on 51 children with various types of vascular rings. The anatomical features of the vascular ring are summarized in **Table 22.1**. Of the 51 patients, 75% were under 1 year of age. Two patients died, but the majority of patients (78%) underwent extubation in the operating theatre at the end of the procedure and did well. Three patients required tracheostomy, one of them for 2 years because of tracheomalacia. Long-term results are shown in **Table 22.2**.

Backer *et al*. (2005) reviewed the overall experience of the Children's Memorial Hospital in Chicago of 209 cases of vascular rings operated between 1946 and 2003. There were no hospital deaths since 1959. The mean and median hospital stays have decreased over time. Prolonged hospital stays were mainly related to airway problems. Four patients (2% of the total series) underwent an aortoplasty 2 months to 4 years after the initial vascular ring division.

The results in patients with pulmonary sling are also excellent (Pawade *et al*., 1992). Backer *et al*. (1999) reported their current practice of correcting pulmonary sling through midline sternotomy using cardiopulmonary bypass. There was no operative mortality in 16 operated infants; 12 of these 16 had complete tracheal rings, seven underwent tracheal repair with pericardial patch, four had tracheal autograft inserted and one had distal tracheal resection. One patient died late after pericardial tracheoplasty. We currently prefer sliding tracheoplasty in this group of patients (Beierlein and Elliott, 2006). Insufficient data are available for evaluating the results in patients with pulmonary sling and associated tracheal stenosis.

Table 22.1 Anatomy.

Malformation	No. of Patients
Double aortic arch	27*
Right aortic arch and left ligamentum arteriosum	8
Anomalous subclavian artery	6†
Pulmonary artery sling	6
Anomalous innominate artery	3
Left aortic arch, right descending aorta and right patent ductus arteriosus	1
Total	51

* One patient died during operation.
† One patient with associated Fallot's tetralogy died after operation.

Table 22.2 Long-term results.

Symptoms and signs	No. of patients	(%)
Asymptomatic	35	(76)
Minimal stridor	3	(7)
Moderate stridor	3	(7)
Recurrent chest infections	3	(7)
Tracheal stenosis and recurrent pneumonia	1	(2)
Dysphagia	1	(2)
Total	46*	

* Three patients were lost to follow-up.

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23

Anomalies of the Systemic Venous Return

C. van Doorn and M. R. de Leval

Anomalies of systemic venous return are frequently associated with other congenital cardiac defects and with atrial isomerism. In the latter condition, the right-sided and left-sided atria are morphologically similar and have either right or left atrial configuration. What determines configuration is the structure of the atrial appendages and their junction with the atrial chamber (Macartney *et al*., 1980). The morphological right atrial appendage has a broad triangular shape and an extensive junction with the atrium, accompanied by a pronounced crista terminalis. The morphological left appendage is long and tube-like and has a narrow and constricted junction with the atrial chamber and no identifiable crista terminalis. Rarely there is mixed right and left atrial morphology (situs ambiguus). Cases of atrial isomerism have long been associated with left- or right-sidedness of the abdominal viscera. Patients with the so-called ''asplenia syndrome'' are likely to have right-sided isomerism, whereas patients with the polysplenia syndrome are likely to have left-sided isomerism. The concordance between the splenic status and the atrial isomerism is, however, rather weak. Some patients with right atrial isomerism have a spleen, whereas some patients with left isomerism do not have multiple spleens. In contrast, there is a stronger concordance between the arrangement of the lungs and bronchi and the morphology of the atrial appendages. The bronchial arrangement is usually asymmetrical in patients with lateralized arrangement but symmetrical in patients with isomerism (Van Mierop *et al*., 1970). Bilateral long and hyparterial bronchi are indicative of left-sided isomerism, and bilateral short eparterial bronchi are indicative of right-sided isomerism. However, discordance between thoracic organs may occasionally be present. Because of these inconsistencies, Anderson suggested the use of the term ''heterotaxy'' to refer to any defects in lateralization, and then to describe the specific arrangement of the organs within each individual (Anderson *et al*., 1998).

Penetrated chest radiographs or specially prepared filter films are useful for evaluating patients suspected of having atrial isomerism (Deanfield *et al*., 1980). The echocardiographic demonstration of the relationship between the inferior vena cava and the descending aorta has proved to be a valuable tool in diagnosing isomerisms (Huhta *et al*., 1982a, 1982b). In right-sided isomerism there is juxtaposition between the abdominal aorta and the inferior vena cava, the vein being in anterior position. In at least two-thirds of cases of left atrial isomerism there is interruption of the inferior vena cava, which is continued via the azygos vein (''azygos continuation''). The azygos vein is also juxtaposed with the aorta but is in a posterior position. Magnetic resonance imaging is also increasingly used to define anatomic relationships and cardiovascular shunts in patients with complex congenital heart disease (Rebergen *et al*., 1996; see Chapter 5).

In right atrial isomerism there is a sinus node on either side lying within the sulcus terminalis. There is often duplication of the superior vena cava, which on both sides connects to the atrial roof in relation to the appropriate crista terminalis. Then the coronary sinus is absent and the cardiac venous return drains directly to the atrial chamber via the Thebesian veins. The inferior vena cava is connected to the atria, but the connection can be left- or right-sided. Anomalies of the pulmonary venous return exist in almost all cases of right isomerism. In the majority of cases the pulmonary venous connection is extracardiac. If the veins return to the atrium, it is usually to a common collector in the roof of a common atrium.

In left atrial isomerism there is no sulcus terminalis and no obvious site for a sinus node (Dickinson *et al*., 1979). The most characteristic feature of left atrial isomerism is the course of the inferior vena cava (IVC), which is interrupted in the abdomen above the renal veins. The abdominal vein continues through either the azygos or the hemi-azygos system to drain to either the right- or left-sided superior vena cava. Most commonly, the hepatic veins are connected bilaterally with the atrial chambers. Bilateral superior venae cavae (SVCs) are present in the majority of cases of left atrial isomerism, in which case either each vein joins individually the

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atrial roof between the appendage and the pulmonary veins or with one of the veins draining via the coronary sinus. It is present in approximately half the cases and occasionally is partially unroofed along its course in the atrio-ventricular groove (Uemura, 1995).

In this chapter, the anomalies of the systemic venous return are classified according to the surgical anatomy and the functional disturbance that they create rather than to embryological features. Four groups have been distinguished:

1. Anomalies of systemic venous return to the systemic venous atrium.
2. Anomalies of systemic venous return to the pulmonary venous atrium.
3. Anomalies of systemic venous return to both atria.
4. Combination of anomalies of systemic and pulmonary venous return.

Other anomalies, such as a retroaortic course of the bridging (innominate) vein, generally have little consequences for surgical repair.

ANOMALOUS VENOUS RETURN TO THE SYSTEMIC VENOUS ATRIUM

In this group of patients, the anomalous return does not produce any functional disturbance. The surgical importance lies in the techniques of venous cannulation for the extracorporeal circulation and, for some cases, in the route for administration of cardioplegia and the intracardiac repair of the associated anomalies.

Anatomy

Drainage of the Left Superior Vena Cava to the Coronary Sinus

This is the most common anomaly of systemic venous return, occurring in 2–4% of all congenital cardiac defects (Campbell and Deuchar, 1954). A right-sided superior vena cava is usually but not always present. In approximately 75% of cases there is hypoplasia or agenesis of the innominate vein (Winter, 1954).

The left-sided superior vena cava originates at the confluence of the left internal jugular and subclavian veins and descends anterior to the aortic arch (if the arch is left-sided) and the hilus of the left lung; it then pierces the pericardium and runs between the left-sided atrial appendage and the left pulmonary veins to become continuous with the coronary sinus, which is typically enlarged (**Figure 23.1a**).

Interrupted Inferior Vena Cava

Azygos continuation is seen in 0.6% of patients with congenital heart disease (Anderson *et al*., 1961). It is frequently associated, as indicated earlier, with left atrial isomerism. The hepatic veins continue to drain into the right atrium as a single trunk or as separate vessels.

Exceptional Anomalies

Other anomalies, such as hepatic veins draining into the coronary sinus (Nabarro, 1903), coronary sinus orifice atresia with persistent left-sided superior vena cava (Gerlis *et al*., 1984) or absence of bilateral venae cavae with drainage of an upper body venous confluence into the inferior vena cava (Krasemann *et al*., 2003), have been the subject of isolated case reports.

Surgical Technique

Cannulation for Cardiopulmonary Bypass

If there is a small left-sided superior vena cava to the coronary sinus or an adequate size bridging vein is present, the left SVC can be temporarily snared during the intracardiac repair. In the other cases, adequate drainage of the left-sided superior vena cava blood must be ensured. This can be achieved by cannulation of the vein, either directly or from within the right atrium through the coronary sinus.

Direct cannulation of the left superior vena cava, because it requires retraction of the heart, is best performed on cardiopulmonary bypass, after the more accessible vein or veins or the right atrium has been cannulated. In cases of azygos or hemi-azygos continuation of the inferior vena cava, the superior vena cava cannula must be large enough to allow for the additional lower body venous drainage, and its tip must be proximal to the entrance of the azygos or hemi-azygos vein into the superior vena cava. Cannulation of the coronary sinus can be done under direct vision when the right atrium is opened. Alternatively, a small sump suction catheter can be placed in the coronary sinus during the intracardiac repair to aspirate the blood return from the left-sided superior vena cava. This is currently our technique of choice, particularly in small infants, under deep hypothermia and low flow. Total circulatory arrest can also obviate the need for individual caval cannulation.

All systemic veins are routinely encircled with snares near their connections to the cardiac chambers. To encircle a left-sided superior vena cava, the heart is retracted slightly to the right and the left atrial appendage is retracted caudally, so as to expose the anomalous vein. A short incision is made into the posterior pericardium on each side of the vessel, and the snare is passed around it with a right-angled dissector. Care must be taken not to damage the phrenic nerve, which usually lies along the left-sided superior vena cava.

When the hepatic veins drain as a single vessel into the right atrium, the common trunk is cannulated through the adjacent atrial wall with a very short cannula, the tip of which remains above the bifurcation of the hepatic veins. If the hepatic veins drain as two vessels, it might be necessary to use two cannulae, or if one of the veins is small; the blood return can be controlled with an intracardiac suction tip, thus obviating the need for direct cannulation. When the exact anatomy of the systemic venous return is not apparent or when adhesions limit access to the heart, the right atrium is cannulated with a single catheter, the patient's temperature is lowered on cardiopulmonary bypass, temporary circulatory arrest or low flow is instituted, and the anatomy of the systemic venous drainage is assessed under direct vision. Then, depending on the cardiac defect, the systemic veins can be cannulated separately to establish conventional bypass or, particularly for small infants, the intracardiac repair may be performed during total circulatory arrest.

If the surgeon wishes to deliver cardioplegic solution retrograde via the coronary sinus, it is mandatory that the persistent left superior vena cava is occluded close to the heart to prevent back-flow of cardioplegia into the systemic veins (Shahian, 1992). It is also important to ensure that the coronary sinus pathway is intact, because if fenestrations are present (unroofed coronary sinus, see below) cardioplegia will leak into the left atrium rather than perfuse the myocardium, with resultant inadequate myocardial protection (Ruengsakulrach, 2001).

SPECIFIC SURGICAL SITUATIONS

Besides their importance in relation to the cannulation for cardiopulmonary bypass and cardioplegia delivery, anomalies of systemic venous return to the right atrium may influence the surgical procedure.

Atrial Switch (Mustard or Senning)

For intra-atrial repair of transposition of the great arteries, two basic options are available. If small, the left-sided superior vena cava may be ligated, and the intra-atrial repair (Mustard or Senning) may be performed in such a manner that the coronary sinus drains into the newly created pulmonary venous (''left'') atrium. If the left-sided superior vena cava is large, and if the left SVC pressure is higher than 15–20 mmHg after occlusion of the SVC, the SVC is cannulated directly and the coronary sinus remains draining into the physiological right atrium.

A clamp or forceps is introduced into the enlarged coronary sinus as a guide to make a deep incision in its roof. For the Mustard repair, the suture line starting on the remnant of the atrial septum is carried along the cut edge of coronary sinus and then within the mouth of the sinus, so as to stay away from the conduction system. This directs the left-sided superior vena cava and coronary sinus blood into the systemic venous atrium (see Chapter 33). For the Senning operation, the posterior flap created by the incision of the coronary sinus can be sewn on the caudal aspect of the atrial septal flap to create the floor of the systemic venous pathway (see Chapter 34).

Atrio-ventricular Septal Defect

Similarly, for patients with atrio-ventricular septal defects, the authors prefer placing the ostium primum patch superior and around the coronary sinus, directing its blood flow to the left atrium. This is possible if the left-sided superior vena cava can be ligated; if not, the coronary sinus orifice has to remain in the right atrium to avoid creating a large right-to-left (R–L) shunt, resulting in cyanosis. In this situation the suture line for the atrial patch should be placed at the base of the left inferior bridging leaflet and continue downwards on the outer margin of this leaflet until the free edge of the atrial septum is reached, leaving the coronary sinus rightward.

Fontan Operation

Anomalies of the systemic venous return also have practical implications in right heart bypass operations. In the presence of bilateral superior venae cavae, cavopulmonary anastomoses must be created on both sides *(bilateral, bidirectional cavopulmonary anastomoses)*; otherwise, the blood from the high-pressure system flows to the lower-pressure vein, which has remained connected to the atrium, through either natural or acquired communicating and collateral veins. Similarly, if a *total cavopulmonary connection* is created in the presence of bilateral superior venae cavae, both veins must be connected to the pulmonary arteries. However, if an *atriopulmonary connection* is constructed, a left-sided superior vena cava draining into the coronary sinus does not have to be connected to the pulmonary artery. In the presence of azygos or hemi-azygos continuation, a cavopulmonary anastomosis can present some technical difficulties because the length of the superior vena cava downstream to the azygos connection can be so short that flow from the azygos vein may be disturbed during the construction of the anastomosis. We therefore prefer to occlude the SVC downstream to the azygos connection with a vascular clamp rather than a tourniquet. In addition, the SVC can be divided 2–3 mm below its connection to the right atrium, so as to amputate the top of the cavo-atrial junction; care must be taken to perform this division above the expected area of the sinus node, so as to provide a few millimetres of additional length to the SVC.

A left-sided or bilateral hepatic veins draining to the systemic venous atrium may present difficulties for

completion of the total cavopulmonary connection. After careful consideration of the anatomy, connection of the hepatic veins to the pulmonary artery is usually possible. This may be achieved using intra- or extracardiac baffling of the veins, either individually or together, with a conduit or lateral tunnel technique or a combination of both. Obviously, care has to be taken not to obstruct the pulmonary venous return or the atrio-ventricular valve. If any intracardiac opening of the hepatic veins remains in the atrium after completion of a total cavopulmonary connection, there will be a R–L shunt and cyanosis. Cyanosis tends to increase over time because of the formation of intrahepatic collateral vessels that allow shunting of desaturated hepatic venous blood into the lower pressure pulmonary venous atrium.

However, we have observed at the subsequent intervention that, in the presence of well-established collaterals, it is safe to occlude the atrial orifice of the hepatic vein, thus directing all hepatic venous return to the lungs.

Cardiac Transplantation

For cardiac transplantation, abnormalities of systemic venous drainage—either of the recipient or the donor heart—will require modification of the surgical technique. Systemic venous rerouting using extra- or intracardiac baffles is usually possible. For a persistent left superior vena cava in the recipient, retrieval of the bridging vein *en bloc* with the donor heart will provide a natural conduit to connect the left-sided SVC. A persistent left SVC in the donor heart, particularly if small, can usually be ligated and the right superior vena cava anastomosed only. A dominant donor heart left SVC can be anastomosed to the recipient innominate vein or, alternatively, be ligated and the recipient right SVC anastomosed directly to the right atrial appendage.

ANOMALOUS VENOUS RETURN TO THE PULMONARY VENOUS ATRIUM

Anatomy

The most common malformation of this group of disorders is a left-sided superior vena cava draining into the left atrium. The orifice of the left-sided superior vena cava is situated in the left upper corner of the left atrium, between the base of the left atrial appendage anteriorly and the left pulmonary veins posteriorly.

In most such cases, there is a right-sided superior vena cava draining to the right atrium, and the innominate vein is often absent. In exceptional cases, there is no right-sided superior vena cava (Sherafat *et al*., 1971) and all the systemic venous blood of the upper part of the body returns to the left atrium. The coronary sinus is typically absent, and there may be an atrial septal defect in the postero-inferior aspect of the secundum atrial septum in the area normally occupied by the orifice of the coronary sinus (coronary sinus atrial septal defect) (**Figure 23.1c**). The defect is separated from the atrio-ventricular valves by a rim of septal tissue. Raghib *et al*. (1965) described this as a ''developmental complex''. The coronary veins drain individually into the atria. An additional patent foramen ovale or secundum atrial septal defect may be present. The atrial septal defect is sometimes larger, resulting from a confluence of a coronary sinus type of atrial septal defect and an ostium secundum or, in other cases, there is an ostium primum or a common atrium with the features of a partial atrio-ventricular septal defect. These anomalies have been interpreted embryologically as the result of a complete absence of partition between the coronary sinus and the left atrium and would therefore be a form of unroofed coronary sinus (Quaegebeur *et al*., 1979; Lee and Sade, 1979).

Figure 23.1 illustrates various types of unroofed coronary sinus with left-sided superior vena cava. **Figure 23.1a** shows a persistent left-sided superior vena cava draining into the coronary sinus. The dotted lines indicate the roof of the sinus. A fenestration in the midportion of the roof is represented in **Figure 23.1b**. **Figure 23.1c** shows complete absence of the roof, with the left-sided superior vena cava emptying directly into the upper corner of the left atrium, with a coronary sinus atrial septal defect. In **Figure 23.1d** there is complete absence of the roof, absence of coronary sinus and drainage of the left SVC into a common atrium.

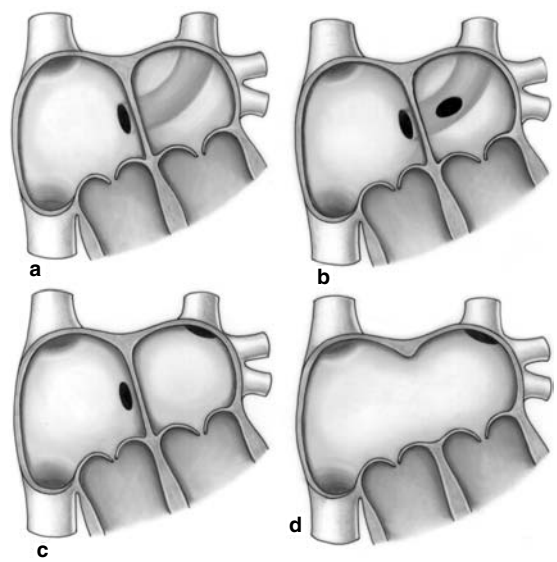

Figure 23.1

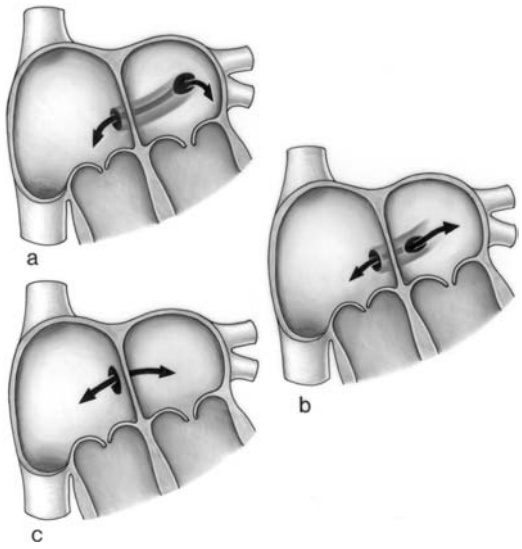

Figure 23.2

Other types of unroofed coronary sinus without left-sided superior vena cava include an unroofed upstream portion of the coronary sinus (**Figure 23.2a**), a fenestration in its midportion (**Figure 23.2b**) and a complete absence of the roof (**Figure 23.2c**) (Quaegebeur *et al*., 1979; Mantini *et al*., 1966; de Leval *et al*., 1975). In the presence of a left-sided SVC, these anomalies result in a R–L shunt at the atrial level, and a history of brain abscess or cerebral embolism, resulting from the draining of systemic venous blood into the left atrium, is fairly common. Without a left-sided SVC, a L–R or R–L shunt, depending on the atrial pressure relations, can exist at atrial level. Unroofed coronary sinus can exist as an isolated defect or in association with other cardiac anomalies. When complex, this anomaly often includes situs ambiguus, common atrium and a complete atrio-ventricular septal defect.

Other less common types of anomalies of systemic venous return to the left atrium include drainage of the IVC to the left atrium (Gallaher *et al*., 1963), drainage of hepatic veins to the left atrium, and hemi-azygos continuation of the left-sided inferior vena cava to the left-sided superior vena cava, which drains into the left atrium. Several cases of total anomalous systemic venous return have also been reported (de Leval *et al*., 1975).

SURGICAL TECHNIQUE

Drainage of the Left-sided Superior Vena Cava to the Left Atrium

Three surgical procedures have been used: ligation of the left-sided superior vena cava; reimplantation of the vena cava into the right atrium or pulmonary artery; and intra-atrial redirection of flow from the vena cava.

Ligation of Left-sided Superior Vena Cava

This procedure can be performed in the presence of an adequate-sized right-sided superior vena cava. Ligation is sometimes possible even in the absence of a bridging vein but it cannot, of course, be performed in the exceptional cases in which the left-sided superior vena cava carries the coronary sinus blood or the pulmonary venous blood. The first case can result from atresia of the right atrial ostium of the coronary sinus and retrograde drainage of the coronary sinus blood into the innominate vein through the left-sided superior vena cava (Mantini *et al*., 1966). The second case, reported by Edwards and DuShane (1950), consists of mitral atresia with intact atrial septum, and drainage from the left-sided superior vena cava to the left atrium is the only route of exit for the blood from the left atrium.

Re-implantation of the Left Superior Vena Cava

Extracardiac redirection of left superior vena cava blood flow is preferred if there is concern that an intra-atrial baffle may obstruct systemic or pulmonary blood flow. Division and reimplantation of the left-sided superior vena cava into the right atrium (Shumacker *et al*., 1967; Reddy *et al*., 1997), right-sided superior vena cava (Reddy *et al*., 1997) or into the pulmonary artery (Foster *et al*., 1978) have been successfully performed. The long-term patency of these venous anastomoses has generally been encouraging. After cavopulmonary anastomosis, a high pulmonary artery pressure may well reverse the flow into the left-sided superior vena cava. It has also been associated with formation of venous collaterals and desaturation in patients with single ventricle physiology (McElhinney *et al*., 1997). In case of a short SVC, an interposition conduit may be necessary to reach the appropriate reimplantation site.

Intra-atrial Redirection

Redirection of left-sided superior vena caval blood is the procedure of choice. Three techniques can be used:

1. ***Roofing procedure***. This procedure is recommended for partial absence of the roof, as illustrated in **Figure 23.3**. In small infants, the repair can be performed with a single venous cannula, profound hypothermia and total circulatory arrest. In older patients, the right-sided superior vena cava and the right-sided inferior vena cava are cannulated directly or through stab wounds in the adjacent right atrial

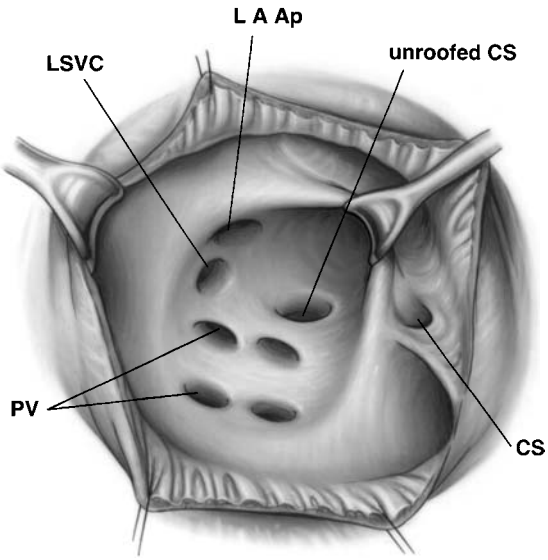

Figure 23.3

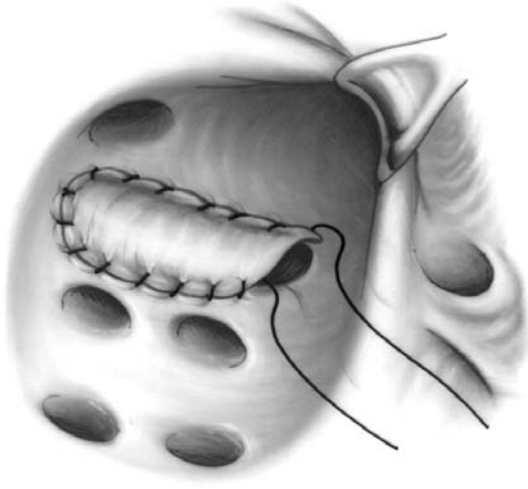

Figure 23.4

wall. The roofing can be accomplished by using a patch (**Figure 23.4**.)

The repair is started at the point of entrance of the superior vena cava into the left atrium, that is between the left pulmonary veins posteriorly and the base of the left atrial appendage anteriorly. The newly created roof runs along the postero-inferior wall of the left atrium anterior to the left pulmonary veins and behind the mitral valve and terminates in the right atrium at the ostium of the coronary sinus. When only a coronary sinus atrial septal defect is present, an incision into the adjacent fossa ovalis is usually necessary for providing adequate exposure into the left atrium. When a large atrial septal defect or a common atrium is present, a patch is used for the roofing, and its septal end can be trimmed so as to serve for the closure of the septal baffle.

2. ***Intra-atrial baffle***. Because of the risk in the roofing procedure of obstructing the left pulmonary veins or the mitral valve, and also the risk of creating too small a tunnel, an intra-atrial baffle to direct blood from the left-sided superior vena cava to the right atrium can be used (de Leval *et al*., 1975). In small infants, the repair is performed with a single cannula and total circulatory arrest. In larger patients, the left-sided superior vena cava is cannulated directly. If the atrial septal defect is small, it is first enlarged anteriorly by excising part of the atrial septum. A patch of autologous or heterologous pericardium or prosthetic material is then inserted between the two atrio-ventricular valves on the rim of the atrial septum, if there is a secundum atrial septal defect, or along the annulus of one atrio-ventricular valve if a primum atrial septal defect is present. The suture line is then carried down toward the postero-inferior corner of the left atrium so as to go around the left-sided SVC and the base of the left atrial appendage if the vena cava was cannulated through it, so as to redirect the vena caval blood in front of the patch (**Figure 23.5**). The suture line then runs on the roof of the left atrium to join the rim of the atrial septum laterally, thus separating completely the pulmonary and systemic venous return.

   If a common atrium is present, the coronary sinus is absent, and thus the usual landmark of the conduction system is not available to the surgeon. The sutures are placed on the tricuspid valve posteriorly between the two atrio-ventricular valves, and they are carried out onto the right atrial wall beyond the tricuspid valve annulus to include the whole area of conduction system in the left atrium, as shown in **Figure 23.5**.
3. ***Intra-atrial prosthetic tube***. This may be an option in an older child or adolescent.

Unroofed Coronary Sinus without Left-sided Superior Vena Cava

The treatment consists of closing the coronary sinus atrial septal defect directly or with a patch, taking superficial bites on the margin of the defect in order to avoid the conduction system.

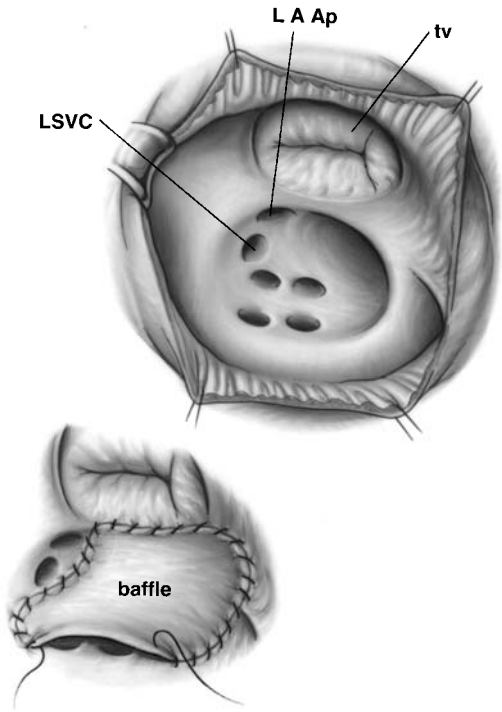

Figure 23.5

Drainage of Inferior Vena Cava or Hepatic Veins to the Left Atrium

An inferior vena cava or hepatic veins that drain into the left atrium are cannulated directly, or the blood return is aspirated from within the left atrium. The repair consists of constructing an intra-atrial baffle for directing the systemic veins anteriorly and the pulmonary veins posteriorly in cases of ventriculo-arterial concordance. In cases of ventriculo-arterial discordance (transposition of the great arteries), the baffle is of course built so as to direct the systemic venous blood into the right ventricle if an arterial switch operation is not performed. The same techniques are applied in cases of total anomalous systemic venous return. For patients with single-ventricle physiology who undergo completion of total cavopulmonary connection, the inferior vena cava or hepatic veins are connected to the pulmonary artery, with the use of either an intra-atrial baffle or an extracardiac conduit. Care has to be taken not to cause obstruction to either the pulmonary venous return or systemic atrio-ventricular valve.

ANOMALOUS VENOUS RETURN TO BOTH ATRIA

This group of conditions includes combinations of anomalies described in the two previous sections. An azygos continuation of the inferior vena cava to the right-sided SVC and of a left-sided SVC to the left atrium is one example. These anomalies are dealt with by following the principles described earlier (Jacob *et al*., 1977).

COMBINED ANOMALOUS SYSTEMIC AND PULMONARY VENOUS RETURN

Numerous variants are possible. **Table 23.1** is a list of such anomalies reviewed in an article from the Mayo Clinic (de Leval *et al*., 1975). These anomalies are often associated with complex cardiac malformations. Principles of repair include division of any significant extracardiac communication between the pulmonary and the systemic veins and intracardiac separation of the venous return, so that pulmonary venous blood drains into the systemic ventricle and systemic venous blood drains into the pulmonary ventricle. Intra-atrial baffles are usually the most satisfactory means of separating the circulations. The drainage of one pulmonary vein into a systemic

Table 23.1 Combined anomalies of systemic and pulmonary venous connections.

Anomalous systemic and pulmonary venous connections to the right atrium
Left-sided superior vena cava to coronary sinus; right upper-lobe pulmonary vein to right atrium
Azygos continuation of inferior vena cava; right pulmonary vein to right atrium
Left-sided superior vena cava to coronary sinus; left upper-lobe pulmonary vein to left-sided superior vena cava
Left-sided superior vena cava to coronary sinus; total anomalous pulmonary venous connexion to coronary sinus
Left-sided inferior vena cava, left-sided superior vena cava, and left pulmonary veins to coronary sinus
Anomalous systemic venous connection to left atrium and anomalous pulmonary venous connexion to right atrium
Left-sided superior vena cava to left atrium; right pulmonary veins to right atrium
Left-sided inferior vena cava to left atrium; total anomalous pulmonary venous connection to left vertical vein
Left-sided inferior vena cava and right-sided superior vena cava to coronary sinus; hepatic veins to left atrium; left pulmonary veins to right atrium
Left-sided inferior vena cava and left-sided superior vena cava to left atrium; total anomalous pulmonary venous connection to right atrium

vein produces little haemodynamic disturbance and can be left uncorrected. Although never seen at the authors' institution, total anomalous pulmonary venous drainage to the coronary sinus in the absence of a right-sided superior vena cava would present an interesting surgical challenge. Inability to separate pulmonary and systemic venous blood within the coronary sinus might be an indication for direct implantation of the left-sided superior vena cava to the left atrium and the intra-atrial baffle.

RESULTS

Results of surgical repair of anomalies of systemic venous return are closely related to the complexity of the associated cardiac anomalies. Most of these anomalous venous connections can be diagnosed preoperatively. They should be suspected in any patient with abnormal situs or a positional abnormality of the heart. In view of the high incidence of this anomaly, it is advisable to routinely inspect the heart and search for a left-sided superior vena cava before cannulation. Failure to detect anomalies of systemic venous return preoperatively can result in excessive blood return to the heart, inadequate venous return to the pump oxygenator, and residual intracardiac shunts.

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24

Total Anomalous Pulmonary Venous Return and Cor Triatriatum

V. T. Tsang and J. Stark

TOTAL ANOMALOUS PULMONARY VENOUS RETURN

Total anomalous pulmonary venous return (TAPVR) is a rare congenital cardiac anomaly. It occurs in only 1.5% of children born with congenital heart defects (Keith *et al*., 1978). However, in specialized centres caring for large numbers of infants, it is one of the few lesions necessitating urgent intracardiac operation in the neonatal period (Stark *et al*., 1980).

Several anatomical types of TAPVR are recognized (**Figure 24.1**):

1. Supracardiac (the commonest), in which the pulmonary veins drain either via the ascending vertical vein to the innominate vein (**Figure 24.1a**) or directly to the superior vena cava (SVC), the orifice usually being close to the orifice of the azygos vein (**Figure 24.1b**).
2. Cardiac, in which drainage is directly into the right atrium or into the coronary sinus (**Figure 24.1c**).
3. Infracardiac, in which drainage occurs via the descending vertical vein into the portal vein or very rarely into the inferior vena cava (IVC) (**Figure 24.1d**). Most are obstructed, as the pulmonary venous return has to pass through the hepatic capillary system.
4. Mixed (the least common type) consists of a combination of one or more of the three types just described.

Partial anomalous pulmonary venous return (PAPVR) is an incomplete form of the anomaly and is discussed in Chapter 25 on atrial septal defects.

DIAGNOSIS

Many patients present in the first days of life with severe congestive heart failure, which is often accompanied by cyanosis. The severity of the symptoms depends on several factors, but the obstruction to the pulmonary venous drainage is probably the most important one.

A systolic murmur is usually present. Chest X-rays show pulmonary plethora and some times cardiomegaly. Pulmonary oedema in the presence of a small heart on chest X-ray is an almost diagnostic feature of obstructive forms of TAPVR. The wide upper mediastinum (snowman sign) is seen more frequently in older infants and children than in neonates. Electrocardiograms usually show right ventricular hypertrophy and right atrial hypertrophy.

TAPVR is most often an isolated lesion, but can be accompanied by other complex congenital heart defects and is frequently associated with asplenia–polysplenia syndrome (heterotaxy syndrome) with a univentricular heart (Sadiq *et al*., 1996; Caldarone *et al*., 1998). Diagnosis is established by two-dimensional echocardiography and colour Doppler (Sahn *et al*., 1979; Smallhorn *et al*., 1981). Squashed left heart secondary to enlarged, hypertensive right ventricle is noted. The size of the left ventricle is of concern. However, Haworth and Reid (1977) measured the dimensions of the left ventricle; almost all proved to be normal. Only in exceptional circumstances are cardiac catheterization and angiocardiography indicated in neonates and infants. In children, invasive investigation may be necessary to evaluate the complex morphology of TAPVR, the site of obstruction and the pulmonary arteriolar resistance.

Recent advances made CT and MRI very promising modalities for three-dimensional reconstruction of the pulmonary veins. This may be particularly valuable after the operation, when pulmonary venous obstruction is suspected.

INDICATIONS

Infants with TAPVR often present very early and are in critical condition. Those with obstruction of

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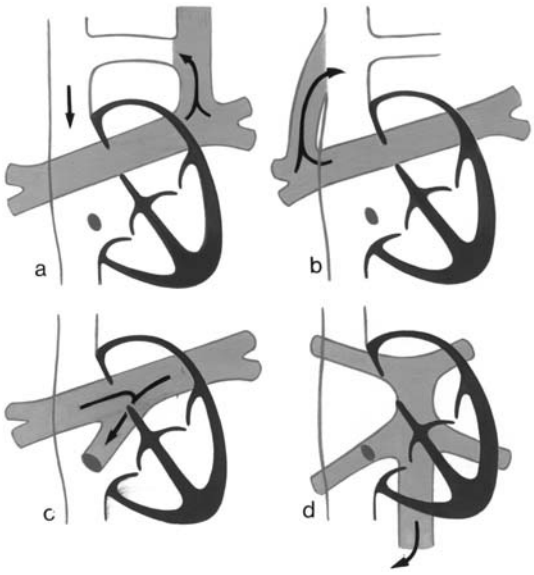


Figure 24.1

pulmonary venous drainage have elevated pulmonary artery pressure, often at the suprasystemic level. Histological lung specimens of these patients show changes not only in the wall of the pulmonary arteries but also in the pulmonary veins (Haworth and Reid, 1977). Most patients in whom the diagnosis of TAPVR is established early in life have severe symptoms. In the few who do not, the pulmonary venous drainage is usually not obstructed. We believe that in such patients the operation should not be unnecesarily delayed. It may be argued that such patients are very good candidates for surgery, and in the absence of obstructed pulmonary venous return, the results of surgery are likely to be favourable.

Neonates and infants who are admitted with severe failure or cyanosis or both are treated urgently. Sometimes, a short period of optimization (around 24 hours), with correction of fluid and electrolyte imbalances and with positive-pressure ventilation, may improve the preoperative condition considerably and, in our opinion, improve the chances of a successful outcome. Palliation of TAPVR through the use of balloon atrial septostomy was suggested by Serrato *et al*. (1968). However, the medical management of such patients in the presence of pulmonary venous obstruction is not satisfactory. Corrective surgery should be promptly performed to avoid further deterioration.

SURGICAL TECHNIQUE

Muller attempted in 1951 to treat this condition by an anastomosis between the common pulmonary vein and the left atrium. In 1956, Burroughs and Kirklin reported the ''atrial well'' technique, and in 1956 Lewis *et al*. successfully corrected TAPVR in a 5-year-old child by using hypothermia and inflow occlusion. The first successful repair with the use of cardiopulmonary bypass was performed by Cooley and Ochsner in 1957. Currently, repair is accomplished on cardiopulmonary bypass, using bicaval cannulation and moderate hypothermia, or using deep hypothermic circulatory arrest (DHCA). The negative long-term effects of prolonged DHCA have been demonstrated (Bellinger *et al*. 1995); however, the advantage of achieving a clear bloodless field during the brief period (15–20 minutes) of DHCA should be considered.

In this chapter, for the purpose of completeness, both the past and present techniques used in the authors' unit are described. Monitoring does not differ from other cardiac operations performed on sick neonates. An arterial catheter and central venous catheters are inserted percutaneously.

A median sternotomy is performed, and the ductus arteriosus or the ligamentum arteriosum is dissected. If the patient is not stable, dissection of the ductus arteriosus can be delayed until the beginning of perfusion. We believe that it is important to dissect and ligate the ductus arteriosus in all patients with TAPVR. With systemic pressure in the pulmonary artery, ductal flow may not be visualized before the operation. Failure to occlude the ductus arteriosus may result not only in overperfusion of the lungs but also in possible air embolism into the systemic circulation during the circulatory arrest.

Because of the risk of pulmonary hypertensive crises, some preventive measures are recommended to minimize reactive response of the pulmonary vasculature. We prefer the judicious perioperative use of milrinone or sodium nitroprusside infusion. Short-acting α-blockade (such as phentolamine) can also be used. Some surgeons prefer the longer-acting phenoxybenzamine (Sano *et al*., 1989).

Heparin is given, the aorta and right atrium are cannulated. Perfusion is then started, and the ductus arteriosus is ligated. On bypass, the patient's nasopharyngeal temperature is lowered to 18°C. During the cooling process, the main pulmonary artery is vented to prevent pulmonary congestion when the heart beat becomes ineffective during the later phase of cooling. The vent site would subsequently be used for the placement of a pulmonary artery catheter for postoperative monitoring. We prefer deep hypothermic circulatory arrest for the infra- and supra-diaphragmatic TAPVC. When temperature of 18°C is reached, the aorta is cross-clamped, and cold blood cardioplegic solution is infused into the root of the aorta. The perfusion is stopped. Blood is drained into the oxygenator and the right atrial cannula is removed.

Vertical Vein

Opinions still differ as to whether the ascending or descending vertical vein should be ligated (Cope *et al*., 1997). It may provide a useful ''vent'' for the non-compliant left heart in the early postoperative period. One approach is to measure the left atrial pressure after discontinuation of bypass; the vertical vein is then snared. If the left atrial pressure does not rise and the systemic pressure does not fall, the vein is ligated; otherwise it is left open. The latter situation is most often the case in patients with infra-diaphragmatic TAPVR. It provides a useful vent for the left atrium or left ventricle for the first hours or days after the repair. Only rarely does the vertical vein not close spontaneously in subsequent weeks/months, and may have to be either occluded by coils or balloons during catheterization or ligated surgically to treat the left-to-right shunt.

Not infrequently (mainly in infra-diaphragmatic type) the distance between the confluence of the pulmonary veins and the left atrium would not permit the construction of a tension-free anastomosis. Under such circumstances, the vein is doubly ligated and transected (Stark *et al*., 1977). The transsected vertical vein can be then incorporated in the anastomosis.

If the ascending vertical vein is to be ligated, care must be taken not to injure the left phrenic nerve, which lies on the anterolateral surface of the vein. Care must also be taken to place the ligature close to the innominate vein, so as to avoid misdirection of the left upper lobe pulmonary vein to the innominate vein. Similarly, the descending vein should only be ligated when the lower pulmonary veins are identified.

Supracardiac Type

In small infants the aorta and the right atrium are cannulated, and a short period of DHCA at 18°C is used. For older children, bicaval cannulation is used. The operation is performed with the use of hypothermic perfusion, aortic cross-clamping, and cold blood cardioplegia, which is repeated every 20 minutes if necessary.

Posterior Approach

This approach was described by Williams *et al*. (1964). The apex of the heart is elevated out of the pericardial cavity towards the right side, providing exposure to the posterior left atrium, the posterior pericardium and the confluence of the pulmonary veins. The elevated apex is best supported with a pledgeted suture on gentle traction to the patient's right. The pulmonary venous confluence is identified through the posterior pericardium. An incision is made in the pulmonary

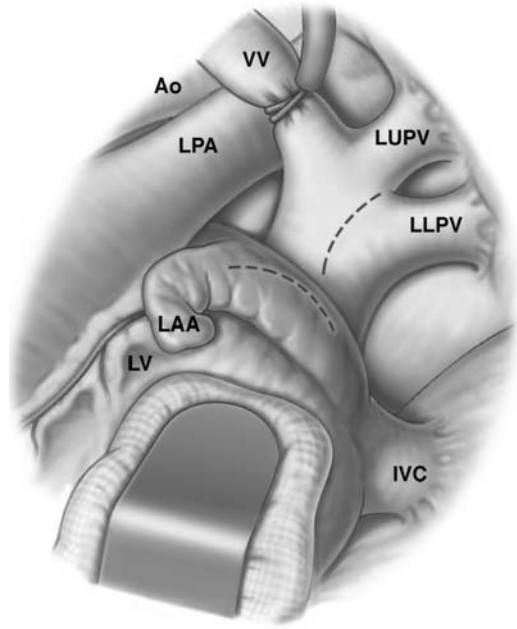


Figure 24.2

confluence from the left to the right pulmonary veins. The incision in the left atrium is slightly oblique, to match the incision in the confluence of the pulmonary veins, when the heart is eventually replaced into the pericardial cavity (**Figure 24.2**). The anstomosis between the left atrium and the common pulmonary confluence is made as large as possible to minimize the risk of anastomotic stenosis. The common pulmonary vein incision can be extended into the lobar branches or the ligated and divided vertical vein. The left atrial incision is anastomosed to the opening in the pulmonary venous confluence, using continuous 7-0 prolene without purse-stringing of the suture line. The patent foramen ovale or ASD can be closed either from the left side or through a small incision in the right atrium. The left atrium and left ventricle are filled with cold saline, usually from the right atrium, before the PFO/ASD is closed. Then the transfusion is slowly started from the aortic cannula. Bypass is then established, and while the aortic cross-clamp remains on, de-airing is achieved via the aortic needle vent, coupled with lung inflations by the anaesthetist. The patient is rewarmed to 36°C. When the aortic clamp is removed and the heart regains its tone, de-airing is carefully repeated via the ascending aortic needle vent. The ascending aorta is usually quite small in these patients, and in occasional cases the aortic cannulation may be obstructive immediately after bypass is discontinued. Its immediate removal may be

required, and this is usually followed by improvement in haemodynamics. After bypass is discontinued, modified ultrafiltration is helpful to improve pulmonary vascular compliance and the ventricular function (see Chapter 11). A pulmonary artery line is routinely placed for postoperative monitoring, because pulmonary hypertension is common in these patients.

Approach from the Right Side

The confluence of the pulmonary veins is dissected free from the right and left atria. When the atrial cannula is removed and circulatory arrest is established, the right atrium is retracted to the left, exposing the left atrium. The left atrium is opened behind the interatrial groove (**Figure 24.3**). The confluence of the pulmonary veins is then opened and anastomosed to the left atrium with fine continuous polypropylene sutures (**Figure 24.4**). Absorbable sutures may be used as an alternative. Because the two structures to be anastomosed lie in close apposition, a large, tension-free anastomosis can be constructed.

Superior Approach

This was described by Tucker *et al*. (1976). This approach is particularly suited to patients in whom the pulmonary venous confluence drains to the SVC–RA junction. Dissection is conducted between the aorta, the right pulmonary artery and the roof of the left atrium.

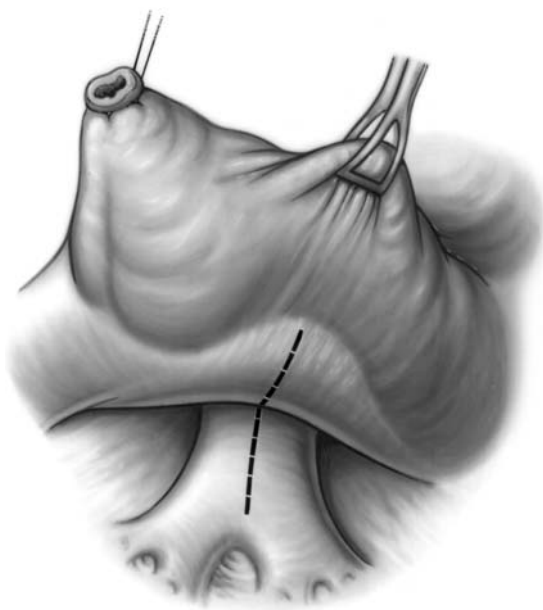


Figure 24.3

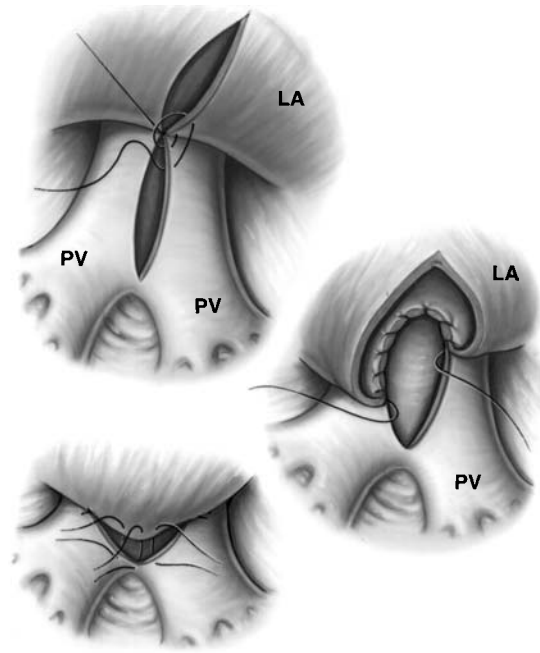


Figure 24.4

Bi-atrial Approach

An alternative approach is through a bi-atrial incision, as suggested by Shumacker and King (1961). The incision is directed toward the interatrial groove (**Figure 24.5**). It is then continued posteriorly across the atrial septum into the left atrium. The incision on the posterior wall of the left atrium is directed toward the orifice of the left atrial appendage but not into it. The incision thus opens both right and left atria and provides another approach to the anastomosis. The confluence of pulmonary veins behind the heart is then opened (**Figure 24.6**). The anastomosis between the confluence of the pulmonary veins and the posterior wall of the left atrium is started at the far corner (**Figure 24.7**), close to the orifice of the left atrial appendage; fine double-armed monofilament suture material (usually 7-0 Prolene) is used. The suture line must be performed very meticulously because any bleeding from the anastomosis is difficult to control after cardiopulmonary bypass is discontinued. The left atrial appendage sometimes becomes inverted and obscures the beginning of the anastomosis; it is useful to place a ligature on its tip. Gentle leftward traction on the ligature via the transverse sinus facilitates the exposure.

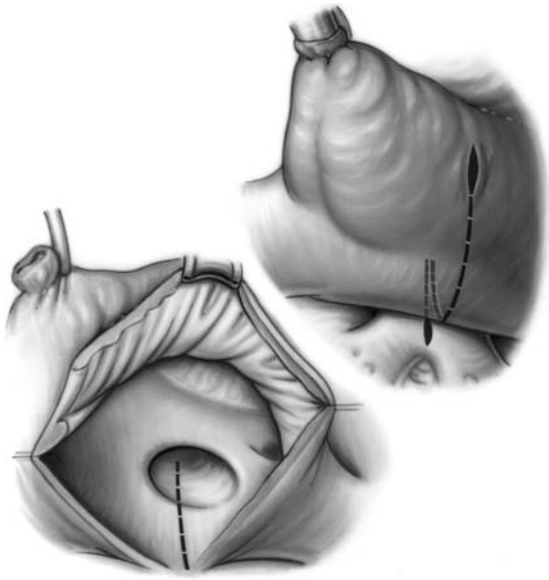


Figure 24.5

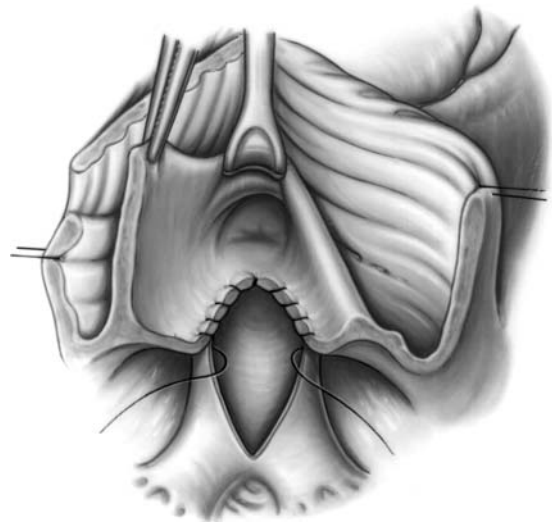


Figure 24.7

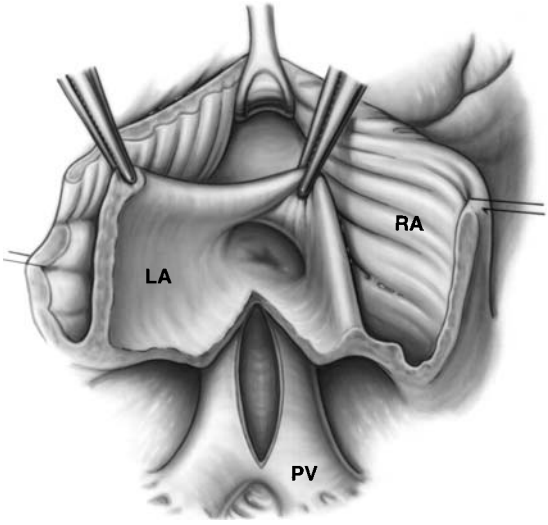


Figure 24.6

Figure 24.8

It is important to make sure that the right end of the anastomosis of the pulmonary veins to the left atrium is not compromised by the suture of the atrial septum (**Figure 24.8**). For this reason alone, the septum is preferably closed with a small pericardial patch (**Figure 24.9**). When the right atrium is closed, the venous cannula is replaced into the right atrial appendage (**Figure 24.10**), bypass is restarted and the patient is rewarmed. All air must be carefully evacuated, as described earlier.

Cardiac Type

Cardiac type of TAPVR can be corrected either on cardiopulmonary bypass or with the use of hypothermic circulatory arrest. The pulmonary veins drain either directly into the right atrium or into the coronary sinus.

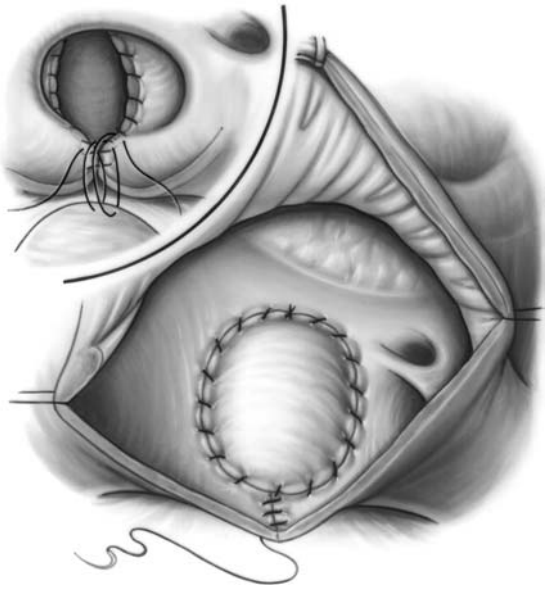


Figure 24.9

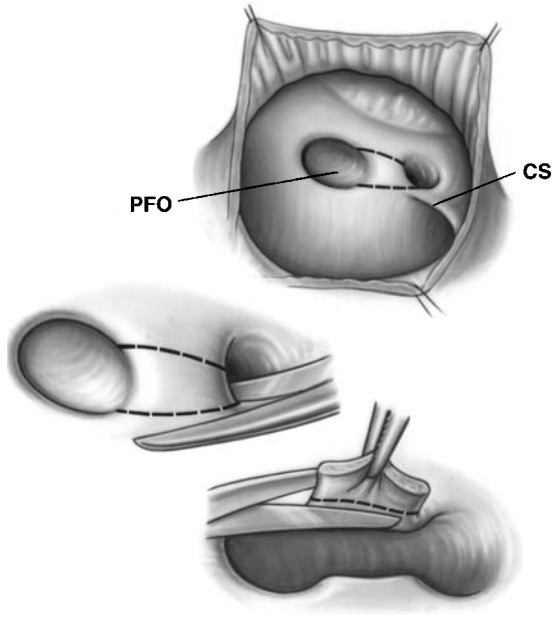


Figure 24.11

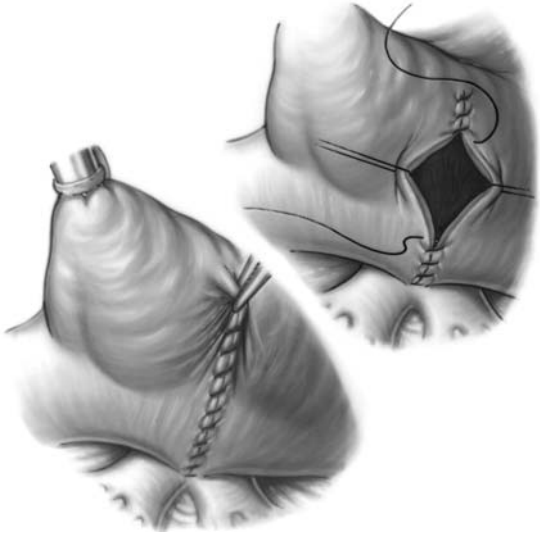


Figure 24.10

The principle of correction is the same in both instances. The ASD must be large enough to accept all pulmonary venous return without any obstruction. This often means resection of part of the atrial septum. The patch is then sutured around the defect and around the orifice of the pulmonary veins, so that a wide channel is created for pulmonary venous blood. If the pulmonary veins drain into the coronary sinus, a cutback is made from the roof of the coronary sinus into the foramen ovale or ASD (**Figure 24.11**). Part of the lower atrial septum may have to be excised. The autologous pericardial patch is then sutured in place. Care is taken to avoid the area anterior to the coronary sinus because it contains the atrio-ventricular node. Therefore, the suture line (depicted by dotted line in **Figure 24.12**) moves away from the edge of the septum into the floor of the coronary sinus.

An alternative technique, described by Van Praagh *et al*. (1972), is illustrated in **Figures 24.13 and 24.14**. An incision is made in the fossa ovalis (**Figure 24.13a**). A curved instrument is then passed through the coronary sinus in order to tent the roof of the coronary sinus, which is excised (**Figure 24.13b**). This ensures a wide communication between the coronary sinus and pulmonary veins and the left atrium. The orifice between the coronary sinus and the right atrium is then closed with fine sutures, and repair is completed by closing the patent foramen ovale (**Figure 24.14**).

Infracardiac Type

For a number of years, infracardiac TAPVR presented one of the most challenging surgical problems (Bove *et al*., 1981). Most patients present in the first few days of life with severely obstructed pulmonary venous return. Many such patients are moribund on admission.

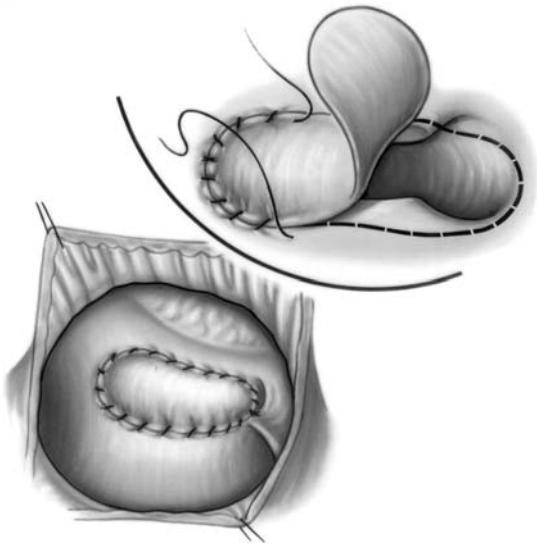


Figure 24.12

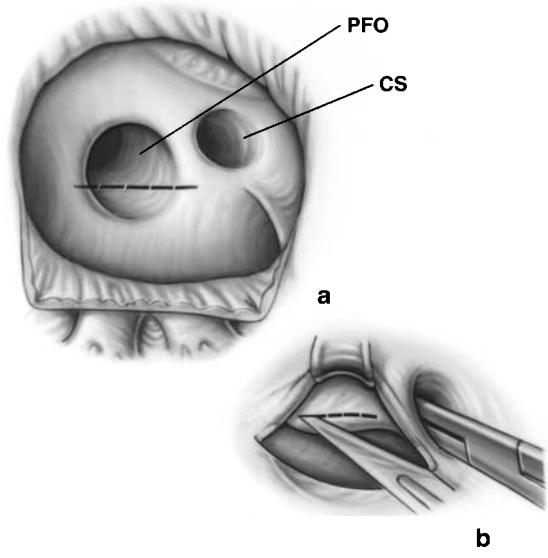


Figure 24.13

Preoperative considerations are identical to those described for supracardiac TAPVR and for operations in other severely ill neonates.

The posterior approach, as described earlier, is used for this anomaly. A midline sternotomy is performed, and the aorta and right atrial appendage are cannulated. The ductus arteriosus or ligamentum arteriosum is dissected and ligated at the beginning of cardiopulmonary bypass. During cooling, the heart is gently lifted and the

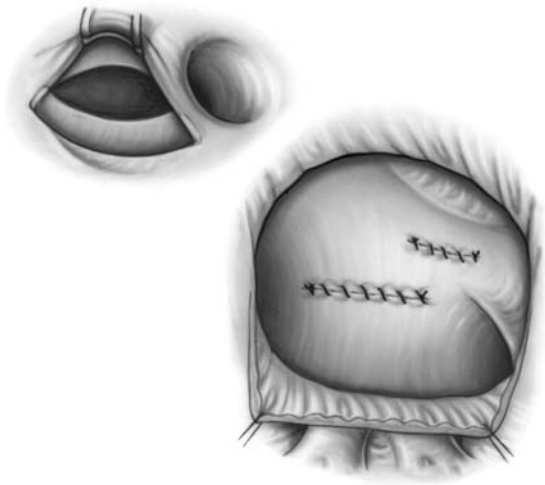


Figure 24.14

confluence of the pulmonary veins is identified. The pulmonary artery is vented or the confluence is opened with a small incision, and a small sump sucker is inserted into this opening to decompress the pulmonary veins. We prefer correction using a brief period of DHCA for all infants with infradiaphragmatic TAPVR.

When the nasopharyngeal temperature reaches 18°C, the aorta is cross-clamped, cold blood cardioplegic solution is infused to the root of the aorta, perfusion is stopped, and the blood is drained into the oxygenator. The heart is then gently lifted to the right with a moist swab, and all pulmonary veins, their confluence, and the descending vertical vein are identified and dissected.

The configuration of the pulmonary veins often differs from that in supracardiac TAPVR. It has been described as tree-like in appearance, with the largest and longest parts of the confluence orientated vertically (Kawashima *et al*., 1977). We have long recognized this fact and recommended that the incision in the pulmonary venous confluence be orientated vertically (Stark *et al*., 1977). The atrial incision should match the vertical incision in the pulmonary veins (**Figure 24.15**) once the heart is placed back in the pericardial sac. This, in our opinion, is the only difficult part of the operation performed from this approach. It is rarely necessary to extend the incision into the orifice of each pulmonary vein with corresponding side incisions on the left atrium (Clarke *et al*., 1977) (**inset, Figure 24.16**). In rare cases, the confluence of the pulmonary veins may be rather low (with the left lower pulmonary vein close to the diaphragm) and therefore the anastomosis to the left atrium may be under tension. In such circumstances, double ligation and division of the descending vein may be helpful (**Figure 24.17**) Nowadays we transsect the descending vertical vein

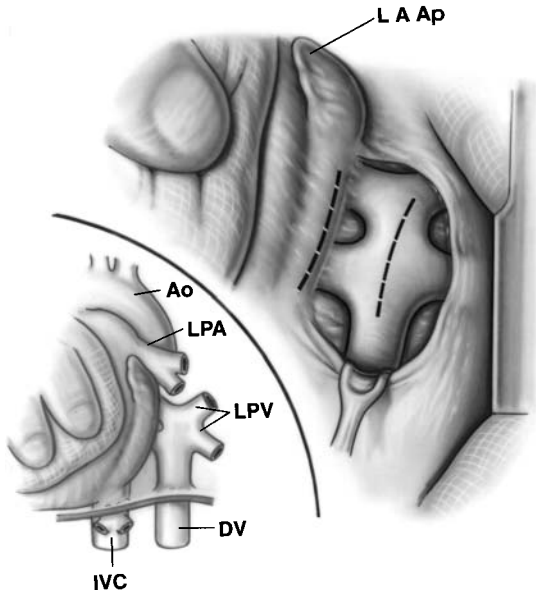
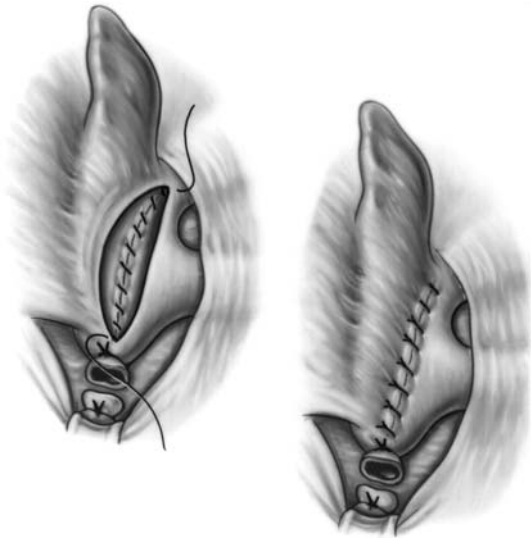


Figure 24.15



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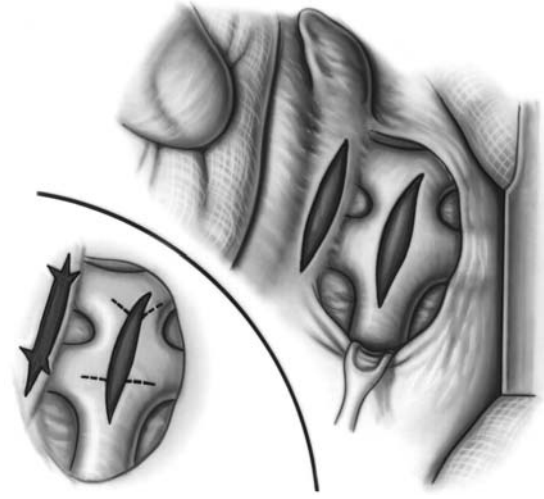


Figure 24.16

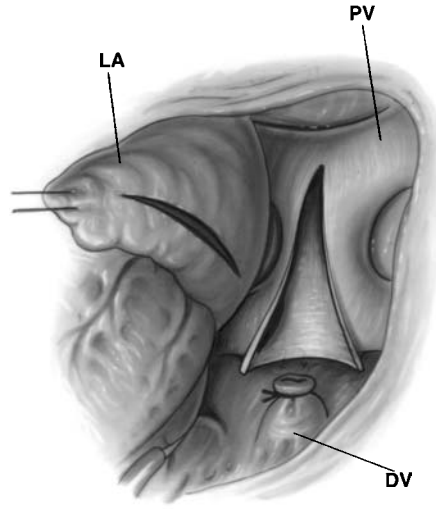


Figure 24.18

routinely and incorporated it in the anastomosis to achieve unrestricted flow (**Figures 24.18 and 24.19**).

Mixed Type

The operative technique depends on the exact anatomy (de Leval *et al*., 1973; Delius and Stark, 1996). Most often, some pulmonary veins drain into the coronary sinus and the others drain directly into the right atrium. When the left upper pulmonary vein drains to the vertical vein and all other veins to the coronary sinus, a separate anastomosis of the left upper pulmonary vein to the amputated left atrial appendage can be made (**Figure 24.20**). When only one pulmonary vein drains anomalously (PAPVR), it may be left uncorrected without significant physiological consequences; the advantage is that a complex or hazardous repair is avoided. This is particularly true with regard to isolated return of one pulmonary vein high into the SVC, because

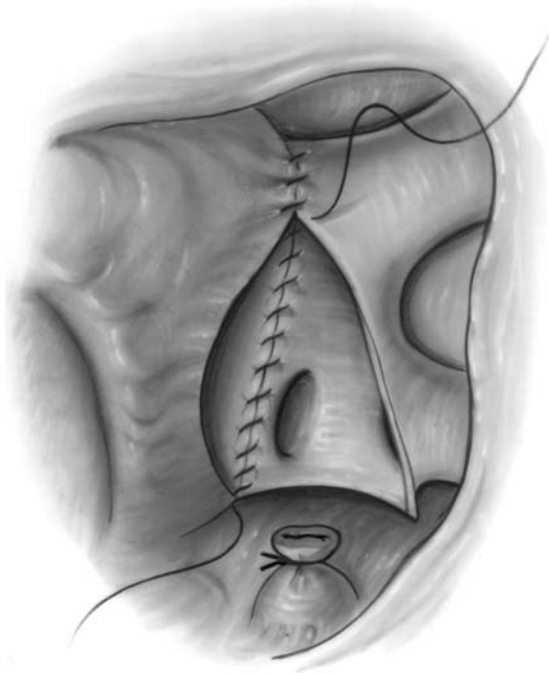


Figure 24.19

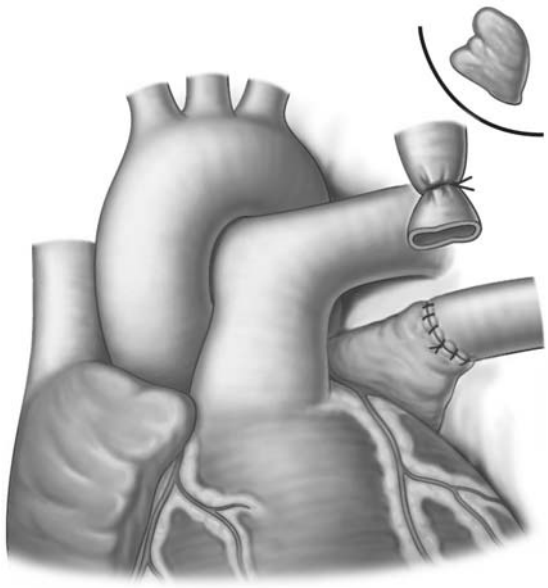


Figure 24.20

the repair might compromise SVC, the sinus node or the sinus node artery.

EARLY COMPLICATIONS

Pulmonary Oedema

Many infants with TAPVR have pulmonary oedema before operation. It is therefore not surprising that many patients show signs of pulmonary oedema after surgery. The left atrial pressure is usually higher than the right atrial pressure in these patients. It is important to monitor these pressures and to try to keep the left atrial pressure as low as is compatible with adequate cardiac output. A high left atrial filling pressure may trigger pulmonary arteriolar vasoconstriction. Diuretics are usually required for several days after the operation to minimize pulmonary oedema.

The left atrium and the left ventricle were reported to be small in patients with TAPVR (Burroughs and Edwards, 1960; Matthew *et al*., 1977). It was not clear whether the cavity of the left atrium should be enlarged with a pericardial patch at the time of repair. The experience of Katz *et al*. (1978) failed to show any difference in survival between patients who did and patients who did not undergo left atrial enlargement.

Enlargement of the left atrium has not been considered necessary at our unit. Similar experience has been reported by Sano *et al*. (1989). The non-compliant left heart may be responsible for the marginal early postoperative haemodynamics and inotropic agents may be required to maintain adequate cardiac output.

Pulmonary Hypertensive Crises

Many infants with TAPVR may have an increase of muscle in the walls of the small pulmonary vessels (Haworth and Reid, 1977). This may lead to pulmonary vascular constriction and, in some patients, may cause pulmonary hypertensive crises. The perioperative administration of potent vasodilators may minimize the development of such crises, but the systemic hypotension would be a limiting factor. Detailed attention to postoperative acid–base balance and ventilatory support with paralysis is required. Endotracheal tube suctioning should be done with extreme care. The monitoring of the PA pressure is very useful to assess the effectiveness of the treatment as well as the lowering of the pulmonary vascular resistance. If the crisis occurs, hyperventilation with 100% oxygen and inhaled nitric oxide is the treatment of choice (Pepke-Zaba *et al*., 1991). Pulmonary vasodilators, such as infusion of prostacyclin, may also be useful (Hopkins *et al*., 1991). Rebound pulmonary hypertension following withdrawal from inhaled nitric oxide has been described (Miller *et al*., 1995).

Today, treatment with inhaled nitric oxide is very effective. Sildenafil may be added in some patients with persistent pulmonary hypertension and during

weaning from nitric oxide (Atz and Wessel, 1999). For further details about pulmonary hypertensive crises, see Chapter 13. Patients with elevated right ventricular pressure may show signs of right ventricular dysfunction. Delayed sternal closure may be beneficial in such patients (Serraf *et al*., 1991). The authors also use this technique whenever it is deemed necessary.

Phrenic Nerve Damage

The phrenic nerve may be damaged by dissection of the ascending common vein, by traction or by ice slush, if it is used for topical cooling. Early diagnosis by diaphragmatic screening using echocardiography or fluoroscopy and early diaphragmatic plication shorten the postoperative recovery (Hamilton *et al*., 1990).

LATE COMPLICATIONS

Pulmonary Venous Obstruction

This formidable complication occurs in around 10–20% of patients operated for TAPVC, usually because of anastomotic obstruction or pulmonary venous stenosis (Hamilton and Van de Wal, 1989; Lacour-Gayet *et al*., 1999). It usually develops within the first few months after initial repair, although rarely it may manifest 1 or more years later. Early presentation and postoperative pulmonary hypertension have the greatest impact on adverse outcomes (Ricci *et al*., 2003) Patients represent with tachypnoea and pulmonary venous congestion. Echocardiogram identifies accelerated turbulent flow in the pulmonary veins, usually at the site of the anastomosis. Although anastomotic technical failure may be implicated, the pulmonary venous stenosis is more often related to a poorly understood process of neo-intimal hyperplasia, and has been associated with all surgical techniques used for primary TAPVC repair. A sclerosing vasculopathy, similar to congenital pulmonary vein stenosis, develops, which can extend beyond the anastomotic site and progress inexorably into the pulmonary veins (Sadr *et al*., 2000).

Anastomotic Stricture

Hamilton and Van de Wal (1989) found reports of 249 cases of TAPVR in 10 series. Twelve late anastomotic strictures (4.8%) occurred among these 249 patients. The incidence of anastomotic strictures used to be higher when vascular clamps were used for the anastomosis; it has decreased since large anastomoses have been constructed under deep hypothermic circulatory arrest. Obstruction can occur even after repair of TAPVR to coronary sinus. Jonas *et al*. (1987) reported six obstructions in 27 operated cases of coronary sinus TAPVR.

Pulmonary Venous Stenosis

Strictures caused by intimal hyperplasia may develop within individual pulmonary veins after the TAPVC operation (Haworth and Reid, 1977). The mechanism for this TAPVC disease complex and stenosis remains unclear, regardless of the type of drainage. Hamilton and Van de Wal (1989) encountered ostial stenosis of pulmonary veins in six of 82 patients who underwent operations for TAPVR. More recently Lacour-Gayet *et al*. (1999) reported incidence of 9% of progressive pulmonary venous obstruction in a series of 178 patients. Small-sized pulmonary veins at the time of presentation of TAPVC may predict the progression of pulmonary venous obstruction and survival (Jenkins *et al*., 1993).

REPAIR OF PULMONARY VENOUS OBSTRUCTION (PVO)

This dreaded complication involving the veno–atrial junction has led to a number of innovative techniques for reoperation. The so-called ''sutureless'' technique of repair was described by Lacour-Gayet *et al*. (1999), Caldarone *et al*. (1998) and Ricci *et al*. (2003). Stenosis of the right pulmonary veins is relieved by an incision; sometimes excision of the fibrous tissue is required. Left pulmonary veins are also incised from inside the lumen, as shown by a dotted line in **Figure 24.21a**. An atrial well is then created by anastomosing pedicled pericardium to the fibrous tisues surrounding the right pulmonary veins, rather than to the endothelium itself (**Figure 24.21b**). This concept was derived from modifications of the Senning operation (Turina *et al*., 1988), in which the *in situ* pericardium was used to augment the pulmonary venous pathway. The main purpose of the sutureless technique is to provide a wide open connection with little geometric distortion and turbulence. Some surgeons have advocated this principle of repair at the primary operation for TAPVC, as a means of avoiding development of this very serious complication (Yun *et al*., 2005).

Other techniques for re-operation (Ricci *et al*., 2003) include patching the right-sided stenosed veins with a free pericardium or *in situ* left atrial appendage. Another endocardial approach with resection of the fibrosis and disobliteration was reported by Louhimo and Tuuteri (1983). All methods have been associated with significant failure rates, further supporting the notion that the disease process is intrinsic to the veins themselves, rather than being attributable to the surgical technique.

Congenital Stenosis of Pulmonary Veins

This is a very rare condition with a poor prognosis (Sadr *et al*., 2000). It may be present even in the absence of TAPVD. The progressive myofibroplastic proliferation

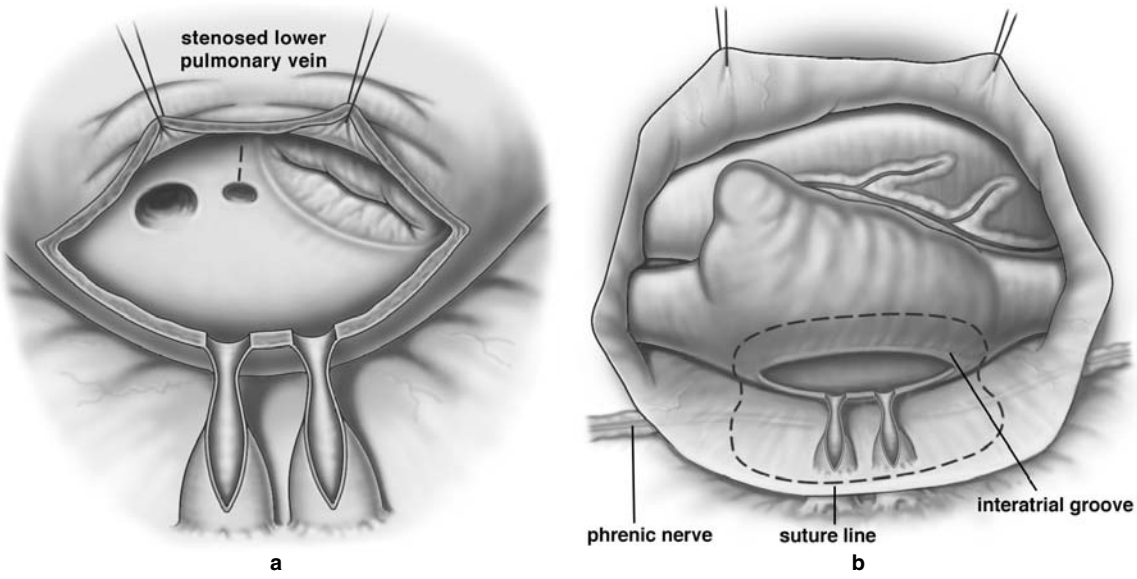


Figure 24.21

at the veno-atrial junction explains the disappointing surgical and interventional stenting approaches (Hyde *et al*., 1999). Steroids (Sands *et al*., 1998) and cytotoxic drugs do not appear to slow the relentless progress in these cases.

Heterotaxy

Patients with isolated TAPVC who survive surgery and who do not develop pulmonary vein stenosis can be expected to grow normally and are usually asymptomatic in NYHA functional class 1 at late follow-up (Bando *et al*., 1996). In atrial isomerism, the long-term prognosis must be much more guarded and is dictated by the associated complex cardiac and extracardiac lesions. Several small series report long-term survival of less than 40% in this group of patients, with survival of 5–13% in the subgroup who require neonatal surgical intervention (Hashmi *et al*., 1998; Sadiq *et al*., 1996; Caldarone *et al*., 1998). Whereas operative mortality for ''simple'' TAPVC has improved over the years, the group of patients with isomerism (or other complex associated lesions) has remained poor. Part of the explanation for this situation lies in the difficulty in regulating pulmonary blood flow in univentricular heart with labile parallel circulations. The management is further complicated by the need for either a systemic to pulmonary shunt or pulmonary artery banding, depending on the exact anatomy.

RESULTS

After Primary Repair

In the late 1960s, mortality rates of 65–88% were reported (Mustard *et al*., 1968; Leachman *et al*., 1969). In more recent years, the risk decreased considerably. Between 1979 and 1987, Sano *et al*. (1989) operated on 44 patients (median age, 15 days), 33 (50%) of whom had obstructed pulmonary drainage. There was one hospital death (2.3%). Four patients required reoperation; two (4.6%) died. Serraf *et al*. (1991) reported a series of 57 patients with TAPVR and severe pulmonary venous obstruction. The mortality rate decreased from 59% in 1980–1984 to 13.3% in 1985–1989.

Our results of the operations also showed considerable improvement in comparison with earlier years (Stark *et al*., 1977). During the 5 year period 1986–1991, we operated on 44 infants with TAPVR. There were five early deaths (11.3%) and one late death (2.3%). Two of the intraoperative deaths were in neonates who had required preoperative resuscitation; after the repair, they had suprasystemic right ventricular pressure. Of the three other early deaths, one was in a neonate with mixed-type TAPVR. Left pulmonary veins were grossly underdeveloped and were not correctable; repair therefore involved right pulmonary veins only. The patient died on day 23 in sepsis. The other two patients died in uncontrollable pulmonary hypertensive crisis on days 1 and 3, respectively.

Our most recent experience (2000–2004) demonstrates the continuing improvement in survival rates. 35 patients underwent isolated TAPVC repair with 1 hospital death (3% mortality). The patient was a premature baby (34 weeks gestation), weight 1.8 kg with obstructed infracardiac TAPVC, complicated by hyaline membrane disease.

Bando reported in 1996 a group of 105 patients operated on during 1966–1995 with a 5% mortality rate. In Birmingham, UK, 85 patients underwent surgery for TAPVR in 1988–1997 with a 7% mortality (Hyde *et al*., 1999).

Results after Reoperations for Pulmonary Venous Obstruction (PVO)

In 1996, Bando *et al*. reported eight patients who developed PVO after TAPVC repair and were managed surgically. Five survived the operation, while three succumbed to pulmonary hypertensive crises (mortality 37%). Lacour-Gayet *et al*. (1999) reported 16 patients (9%) out of 178 patients who required reoperation for progressive pulmonary venous stenosis after repair of TAPVC. Four died (25%). The only significant risk factor in their series was the presence of bilateral stenosis. *In situ* pericardial technique was used in seven patients, with five survivors. Pulmonary artery pressure was normalizing at follow-up.

In our own experience (Ricci *et al*., 2003), 20 patients were reoperated between 1992 and 2002. Five patients died (25%). The risk factors for mortality were early presentation (under 6 months) and persistent pulmonary hypertension after reoperation without any residual pulmonary venous obstruction.

COR TRIATRIATUM

Cor triatriatum is a rare congenital cardiac anomaly, in which the pulmonary veins drain into a common atrial chamber behind the heart. This chamber, which is usually behind and above the true left atrium, is separated from the left atrium by a diaphragm (**Figure 24.22**). We prefer to identify the upper and lower chambers as proximal and distal chambers, based on the direction of the blood flow. The proximal chamber receives the pulmonary veins; the distal one (true left atrium) contains the left atrial appendage and the mitral valve. Van Praagh and Corsini (1969) defined cor triatriatum as a malformation in which the left atrium is subdivided into the dorsal and ventral chambers by a fibromuscular diaphragm. The communication between the two chambers may be small, large or even absent. The right atrium may

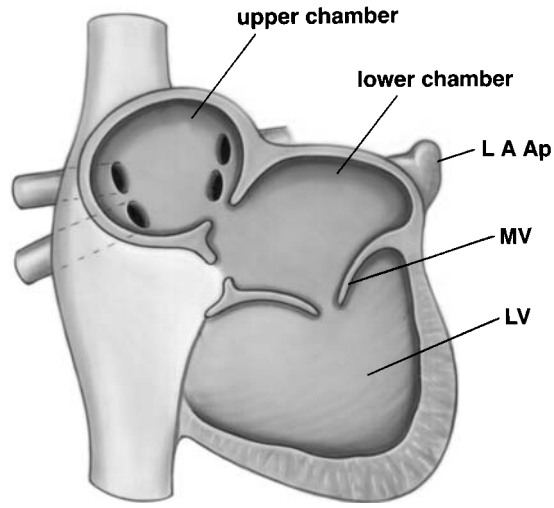


Figure 24.22

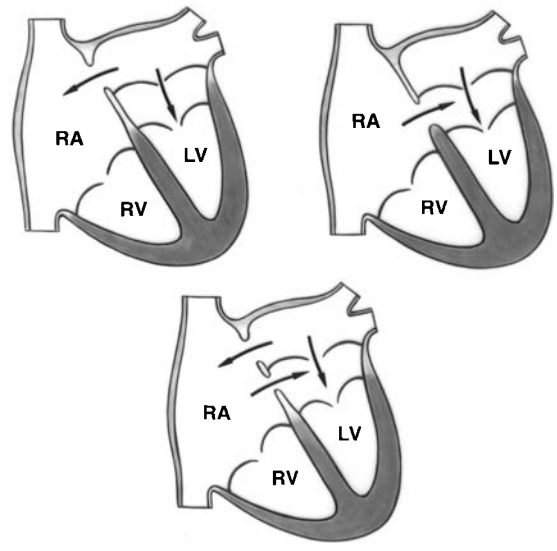


Figure 24.23

communicate with either chamber or even with both (**Figure 24.23**).

DIAGNOSIS

The symptoms and signs depend on the degree of pulmonary venous obstruction, the degree of pulmonary venous hypertension, and the presence or absence of associated shunting (Brickman *et al*., 1970). There is a systolic murmur along the left sternal border. In the

presence of pulmonary venous obstruction, the second pulmonary sound is loud. The electrocardiogram shows right axis deviation, right atrial enlargement and right ventricular hypertrophy. The chest X-ray usually shows pulmonary venous congestion. An X-ray picture of a small heart and pulmonary oedema is typical for two conditions only, cor triatriatum and obstructed TAPVR. Cardiac catheterization and angiocardiography used to be the standard diagnostic techniques. Currently, two-dimensional echocardiography with colour Doppler imaging can diagnose this condition and other associate lesions accurately. Cor triatriatum must be distinguished from supramitral membrane, which is usually closely related to the mitral valve (Sethia *et al*., 1988). Both these lesions may be missed by our current techniques. Cardiac MRI provides excellent imaging of pulmonary venous system.

INDICATIONS

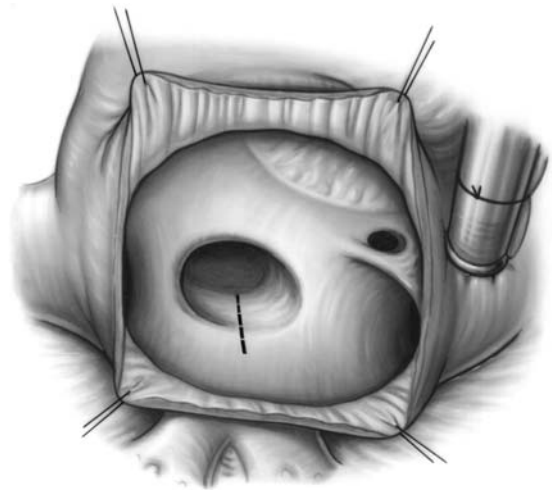
When cor triatriatum manifests as an obstructive lesion, severe pulmonary hypertension make early operative intervention mandatory. If there is a good communication between the two chambers within the left atrium, or if the chambers communicate widely with the right atrium, the symptoms are those of a large ASD and surgery may be performed electively.

SURGICAL TECHNIQUE

The first successful repair was achieved by Lewis *et al*. (1956), using inflow occlusion with hypothermia. Today, the operation is performed on cardiopulmonary bypass with moderate hypothermia, cardioplegia and aortic cross-clamping. In very small infants, it may be advantageous to use deep hypothermia and have the benefit of a bloodless field under very brief circulatory arrest. The right atrium is opened and the anatomy visualized. The patent foramen ovale or ASD is enlarged in order to gain access to the left atrium (**Figure 24.24**). The dividing membrane is then carefully visualized. An eyelid retractor is placed under the edge of the septum, and the membrane is resected (**Figure 24.25**). Care must be taken not to cut outside the heart and not to damage any adjacent structures. **Figure 24.26** shows the posterior wall of the left atrium after the membrane has been resected. The atrial septum may be closed with a direct suture or with a patch (**inset Figure 24.26**). The patient is rewarmed, and perfusion is terminated in the usual manner.

RESULTS

Because cor triatriatum accounts for only about 0.1% of all congenital heart defects (Keith *et al*., 1978), the number of reports on surgical treatment is limited. Richardson *et al*. (1981) reported 21 patients, of whom 13 underwent surgical correction; eight survived the operation (62%), and there was only one late death. Rodefeld *et al*. (1990) reported 12 patients with overall mortality rate of 25%. The risk appeared to be higher when complicated by concomitant cardiac anomalies. We have operated on 16 patients, three of whom have



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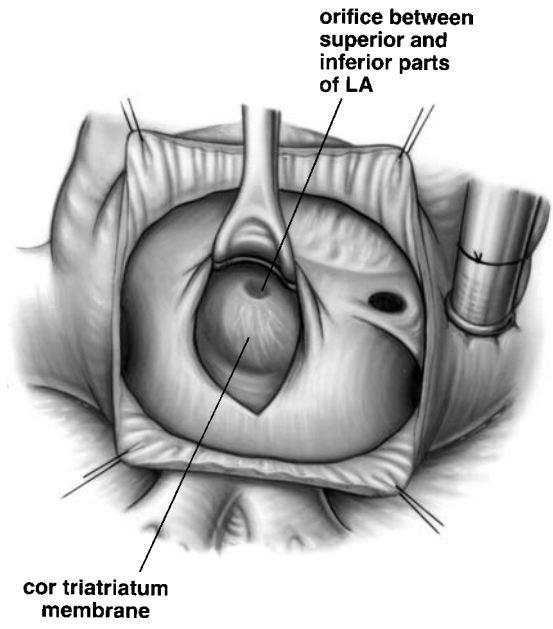

Figure 24.25

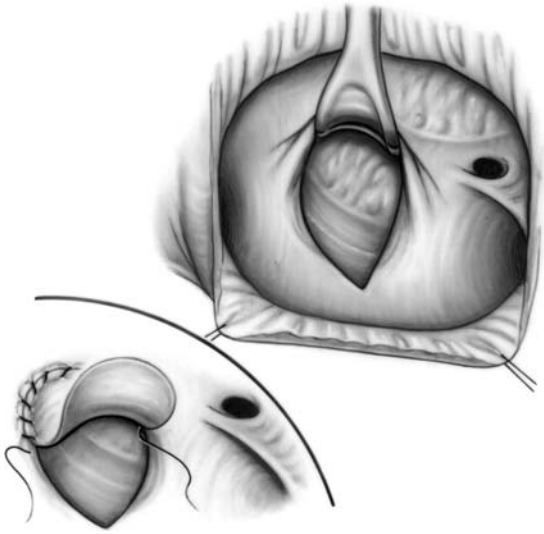

Figure 24.26

been reported previously (Somerville, 1966; Oelert *et al*., 1973). One 4-month-old infant with associated complete atrio-ventricular septal defect and diaphragmatic hernia died. All 15 survivors are well.

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25

Secundum Atrial Septal Defect and Partial Anomalous Pulmonary Venous Return

J. Stark and V. T. Tsang

In this chapter we discuss defects in the septum secundum, together with partial anomalous pulmonary venous return. Defects in the septum primum are discussed in Chapter 27 (atrio-ventricular septal defects). The communications between the left ventricle and the right atrium are described in Chapter 26 (ventricular septal defects).

Secundum atrial septal defect (ASD) is one of the most common congenital heart defects (Keith, 1978). ASD was among the first anomalies to be corrected by operative treatment. Before the advent of cardiopulmonary bypass, several ingenious techniques were developed and used. In 1953, Gross *et al*. used an atrial well technique, Lewis and Taufic (1953) closed an ASD successfully by using hypothermia and inflow occlusion, and Sondergaard (1954) described his method of purse-string suture closure. Limited time and poor visualization sometimes led to incomplete closure or the overlooking of some additional cardiac malformations. Since the development of the pump oxygenator, most centres preferred closure of ASD during cardiopulmonary bypass. In the mid-1970s, attempts were made to close ASDs during cardiac catheterization with the use of various types of umbrella or balloon devices (King and Mills, 1974; Rashkind, 1975) and recently, ''clamshell'' and Amplatzer devices. The details about the current state of art in these techniques are discussed in Chapter 7. The surgical approach has also evolved with the introduction of minimal invasive surgery.

DIAGNOSIS

The majority of children with secundum ASD are asymptomatic. The diagnosis is usually made after the discovery of a murmur during routine school examination. Typical findings include a soft ejection systolic murmur with fixed split-second sound over the base of the heart and a mid-diastolic inspiratory rumble at the left sternal edge. The chest X-ray shows cardiac enlargement and pulmonary plethora if the left-to-right (L–R) shunt is significant. The haemodynamic changes are reflected by the echocardiographic appearance of right ventricular volume overload. Cardiac catheterization, with or without angiocardiography, used to be performed to confirm the diagnosis. At present, careful clinical examination combined with two-dimensional echocardiography and colour Doppler imaging provide an adequate base for determining indications for surgery in children. If the clinical signs suggest pulmonary hypertension, catheterization may be required for establishing the degree of pulmonary arteriolar resistance. This is especially important in adolescents and adults. In very rare instances, ASD manifests with congestive heart failure in infancy (Bull *et al*., 1981). Careful evaluation is necessary in such infants, so that additional cardiac lesions that may be responsible for heart failure are not overlooked.

INDICATIONS

The natural history suggests a slow development of symptoms and rising pulmonary artery pressure. Craig and Selzer (1968) evaluated 128 consecutive patients over the age of 18 years who were catheterized; 75% were symptomatic, 22% had significant pulmonary hypertension and 15% had high pulmonary arteriolar resistance. When the pulmonary resistance rises and a right-to-left (R–L) shunt appears, the patient starts to deteriorate more rapidly (Eisenmenger syndrome). For these reasons, it is accepted that significant secundum ASD should be closed in children. We prefer closure before patients reach school age (i.e. at about 3–4 years), or earlier if there is a significant RV volume overload. Only rarely does secundum ASD cause severe heart failure in infancy needing closure (Bull *et al*., 1981). If

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the pulmonary arteriolar resistance is severely raised, the condition is considered inoperable.

TYPES OF ATRIAL SEPTAL DEFECT (ASD)

Various types of ASD are illustrated in **Figure 25.1**. In all figures the ASD is viewed from the right atrium. The patent foramen ovale is shown in **Figure 25.1a**; in the ASD of fossa ovalis type (or ostium secundum ASD), there may be a complete septal rim (**Figure 25.1b**), or the lateral rim may be missing (**Figure 25.1c**). As a variant, a fossa ovalis type of ASD may present with multiple fenestrations of the fossa (**Figure 25.1d**). A large fossa ovalis type of ASD may extend to the orifice of the inferior vena cava (IVC) (**Figure 25.1e**). A posterior ASD is located posteriorly and inferiorly; the right pulmonary veins are close to the rim of the defect (**Figure 25.1f**). A sinus venosus defect (superior vena cava (SVC) defect) is located superiorly, under the orifice of the superior vena cava (**Figure 25.1g**). It is almost always associated with the anomalous drainage of one or two right superior pulmonary veins to the SVC. A coronary sinus ASD (**Figure 25.1h**) is part of an unroofed coronary sinus syndrome. This is described in Chapter 23.

TYPES OF PARTIAL ANOMALOUS PULMONARY VENOUS DRAINAGE

It is important to assess the drainage of the pulmonary veins and their relationship to the ASD. The most common type of partial anomalous drainage is associated with a sinus venous defect (**Figure 25.2a**). The right upper and sometimes the middle pulmonary vein may be connected to the SVC at the junction of the SVC and the right atrial junction (**Figures 25.2a,b**). Sometimes the pulmonary veins may connect higher up to the lateral aspect of the SVC. In rare instances, a right superior pulmonary vein may be connected to the SVC in the absence of a sinus venous defect.

All pulmonary veins from the right lung may drain to the right atrium (**Figure 25.2c**); the orifices of individual veins may be close together or separated. Alternatively, the superior pulmonary vein may drain to the SVC and the others to the right atrium. All right pulmonary veins may also drain to the IVC (scimitar syndrome); this is a rare anomaly (Kiely *et al*., 1967). The anomalous pulmonary venous trunk is usually anterior to the right hilum and enters the IVC just above or below the diaphragm (**Figure 25.2d**). The orifice of the pulmonary veins is usually very close to the orifices of the hepatic veins. Scimitar syndrome is often associated with right lung hypoplasia and dextroposition of the heart. The blood supply to the part of the right lung may be via aortopulmonary collateral vessels originating from the descending aorta. On occasion, true sequestration may be associated with the scimitar syndrome (Alivizatos *et al*., 1985).

Anomalous drainage from the left lung may be either to the innominate vein or via an anomalous vertical vein (**Figures 25.2e,f**) or to the coronary sinus.

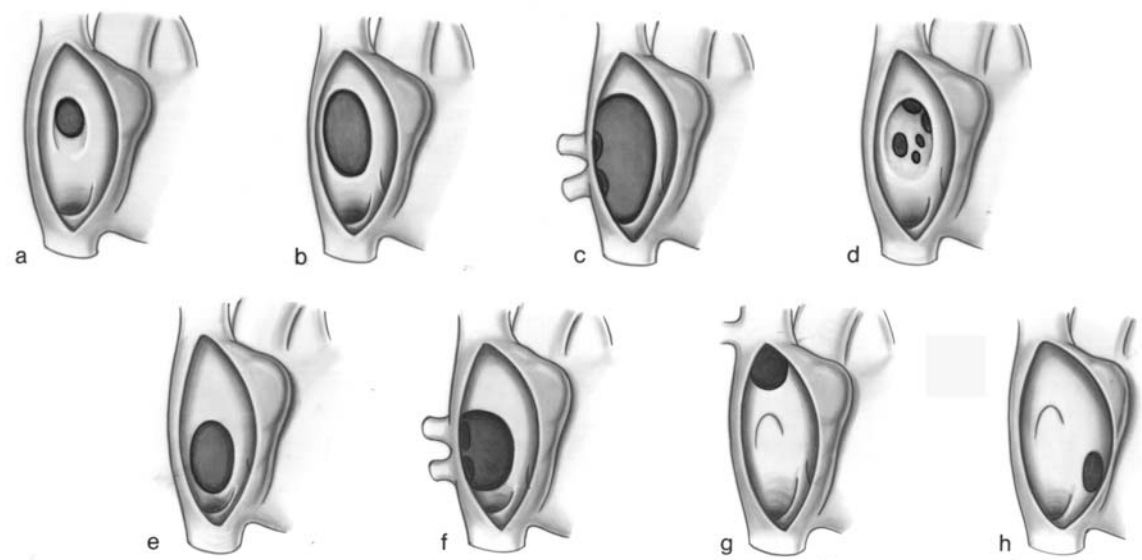

Figure 25.1

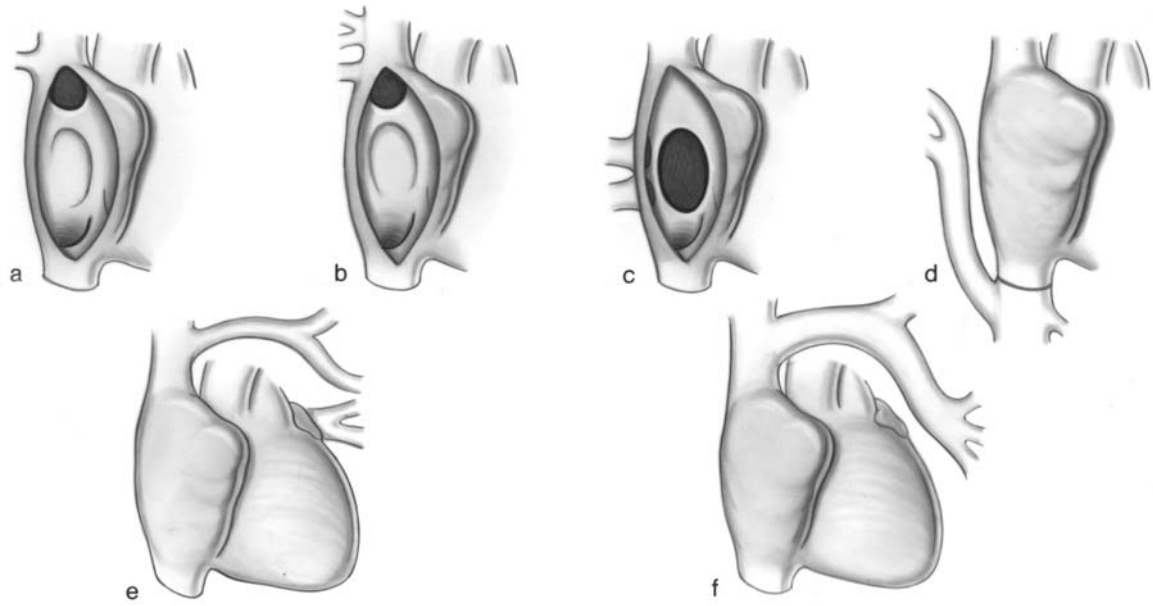

Figure 25.2

SURGICAL TECHNIQUE

The operation is performed on cardiopulmonary bypass; the closed techniques are not used any more. The usual approach is through a limited midline sternotomy. Bilateral submammary incision followed by a midline sternotomy (Brutel de la Riviere *et al*., 1981) or a right anterior thoracotomy through the fourth intercostal space (Rosengart and Stark, 1993) can be used for cosmetic reasons. If the right thoracotomy approach is used, it is important to rule out other lesions, such as pulmonary stenosis or persistent ductus arteriosus, which would be difficult to approach from the right side.

If a midline sternotomy is used, purse-string sutures are placed on the ascending aorta, directly on the SVC, and on the IVC. Heparin is given, the aorta is cannulated and right-angled cannulae are used for the SVC and IVC.

For a right thoracotomy, the patient is positioned on the operating table with the right side elevated by 45°. The incision is short: anteriorly it stops below the right breast about 3 cm from the lateral edge of the sternum, and laterally it extends to the edge of the latissimus dorsi muscle. The fourth intercostal space is opened. The right thymic lobe is resected. The pericardium is opened about 2 cm in front of the right phrenic nerve from the IVC towards the ascending aorta. Traction stitches are placed on both sides of the pericardium. The two most cephalad stitches are tied to the periosteum of the rib; this brings the ascending aorta forward and facilitates the cannulation (**Figure 25.3**). If the aorta is too deep and not easily accessible, the iliac artery can be cannulated instead. In 60 consecutive cases of ASD closure through the right thoracotomy, the authors were always able to cannulate the ascending aorta.

When all cannulae are placed, perfusion is started with a perfusate temperature of 34°C, the aorta is cross-clamped and cardioplegic solution is infused into the ascending aorta. For an uncomplicated ASD, mild hypothermia with electrical fibrillation can be used. Great care must be taken not to place a sump sucker through the ASD to entrap air in the left side of the heart.

The right atrium is opened with an oblique incision (**inset, Figure 25.3**). The atrial anatomy is carefully inspected. If there is any doubt about the competence of the tricuspid or the mitral valves, they should be tested. The small defect can often be closed by direct suture, but some very large defects and the defects in certain locations (large posterior defect, sinus venosus defect, and some defects associated with partial anomalous venous drainage) may necessitate placement of a patch.

Direct Closure

Small and medium-sized defects are closed by direct suture (**Figure 25.4**). The author and associates use 5-0 Prolene continuous mattress sutures in one layer, followed by an over-and-over stitch in the second layer.

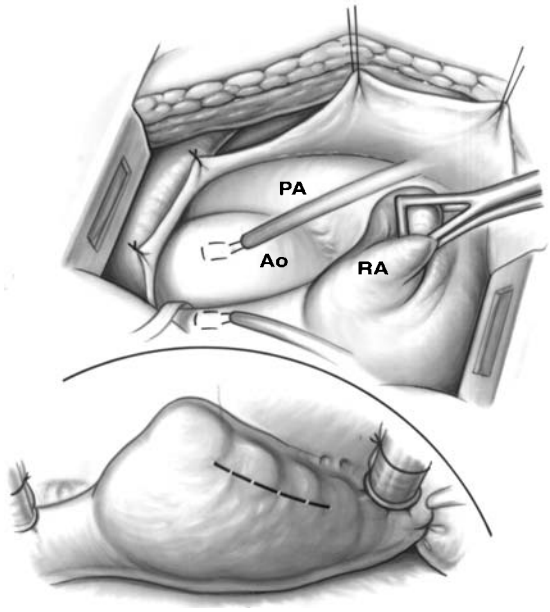

Figure 25.3

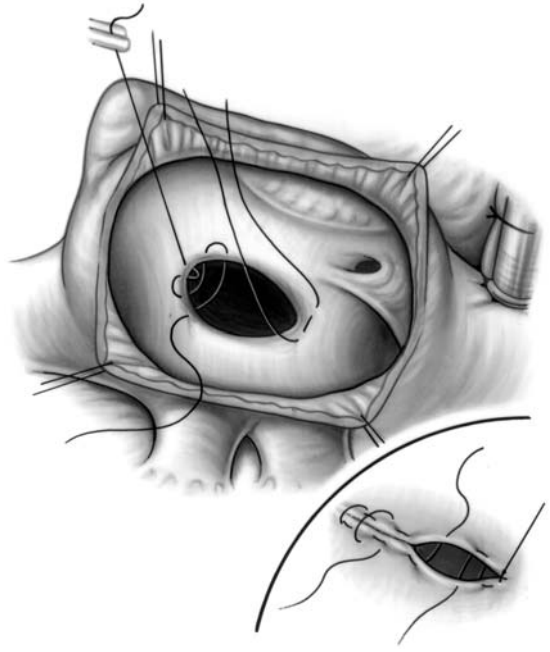

Figure 25.4

It is important not to close the defect under tension, because postoperative arrhythmias or residual defect may result.

Patch Closure

Of the various patch materials available, the authors prefer the patient's own pericardium or Gore-Tex. Certain technical points are worth mentioning. In the upper pole of the defect, deep stitches should be avoided. The aorta lies just underneath the posterior right atrial wall and can be damaged. In low IVC-type ASD, care must be taken to visualize the lower pole of the defect. Failure to visualize this corner, or placing the stitches close to the IVC cannula and not tying them securely, may result either in a residual shunt or, more significantly, in partial or total drainage of IVC into the left atrium (Bedford *et al*., 1957).

The suture line is started with an open technique in the lower corner of the ASD, the orifice of the IVC and the Eustachian valve being carefully visualized. When the patching is almost completed, the aortic clamp is removed, the anaesthetist inflates the lungs, and any residual air is expelled from the left atrium (**inset, Figure 25.5**). The last stitch on the patch is then tied. Because of the policy not to aspirate blood below the rim of the ASD, air should not enter the left side. Additional measures for evacuating the air include placement of an aortic needle vent at the cardioplegic needle site, inversion of the left atrial appendage, and aspiration of the right upper pulmonary veins and the right ventricle. We currently create a right pleuro-pericardial window to avoid late pericardial effusion (incidence 16%) or

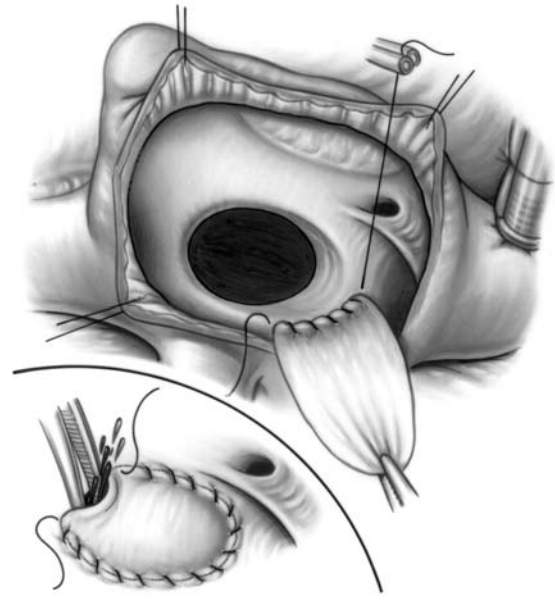

Figure 25.5

tamponade (incidence 1.5%), which can be fatal (Yip *et al*., 1997).

TECHNIQUES FOR SPECIAL CONDITIONS

Sinus Venosus Defect

Anomalous connexion of the pulmonary veins to the SVC is assessed from outside the heart. The SVC cannula is placed well above the anomalous vein. Mild systemic hypothermia at 32°C is routinely used, the aorta is cross-clamped, and cold blood cardioplegia is infused into the root of the aorta. Placement of an atrial incision depends on the position of the pulmonary veins. If the superior pulmonary vein drains to the SVC–atrial junction or just above it, a standard oblique atriotomy can be used (**Figure 25.6a**). If the veins drain high into the SVC, a posterior incision (**Figure 25.6b**) can be used; it runs on the lateral wall of the SVC and stays behind the sinus node to open the right atrium posteriorly. Care must be taken not to injure the tail of the sinus node.

When the pulmonary veins drain to the junction of the SVC and the right atrium, the pericardial patch is stitched with an open technique to the upper rim of the vein (**Figure 25.7**) with a 5-0 or 6-0 polypropylene suture. The patch is then lowered into position and sutured around the ASD, diverging from the orifice of the pulmonary vein toward the edge of the ASD. If the pulmonary veins drain high onto the SVC (**Figure 25.8a**), the posterior incision offers excellent exposure (**Figure 25.6b**). If the ASD is small, it is enlarged inferiorly. Care must be taken not to cut superiorly, as this would open the atrium to

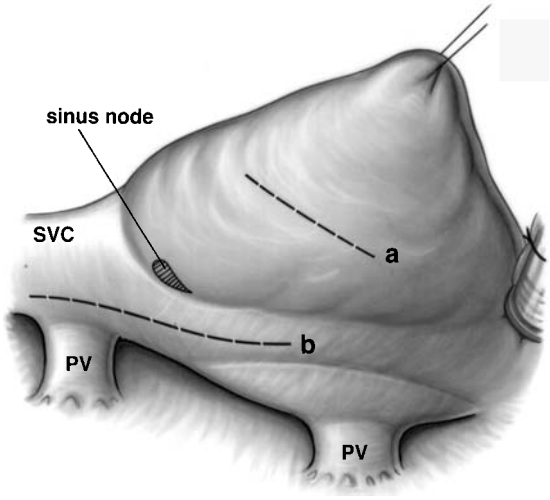

Figure 25.6

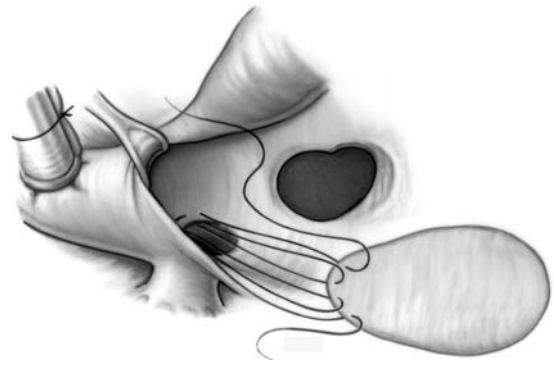

Figure 25.7

the pericardial cavity. Blood from the pulmonary veins is redirected with a pericardial patch (**Figure 25.8b**). We usually close the posterior right atrial incision with another pericardial patch (**Figure 25.8c**) to minimize the risk of SVC narrowing/obstruction.

Another approach to the anomalous right pulmonary vein drainage into the high SVC is the Warden procedure (Warden *et al*., 1984) (**Figure 25.9**). The junction of the SVC/innominate vein is cannulated, and the posteriorly sited azygous vein is located. A standard right atriotomy is used. The SVC is divided upstream to the anomalous pulmonary vein. Preferably, closure of the cardiac end of the divided SVC is undertaken with an autologous pericardial patch without compromising the pulmonary venous drainage. From within the right atrium, the SVC orifice is connected with the atrial communication using an autologous pericardial baffle, sutured by continuous 5-0 or 6-0 Prolene. A purse-string effect must be avoided. The cephalic end of the SVC is then anastomosed to the amputated right atrial appendage with fine 6-0 Prolene suture.

Repair of Coronary Sinus Atrial Septal Defect

This rare defect is repaired by the excision of the septum between the enlarged coronary sinus ostium and the ASD, and by placing an autologous pericardial patch over this area. The details of the technique are described in Chapter 24. Because of the proximity of the atrioventricular node, fine superficially placed 6-0 Prolene sutures are placed within the orifice of the coronary sinus. The coronary sinus blood will drain into the left atrium.

Repair of Partial Anomalous Pulmonary Venous Return

Anomalous drainage associated with a sinus venosus defect was described earlier.

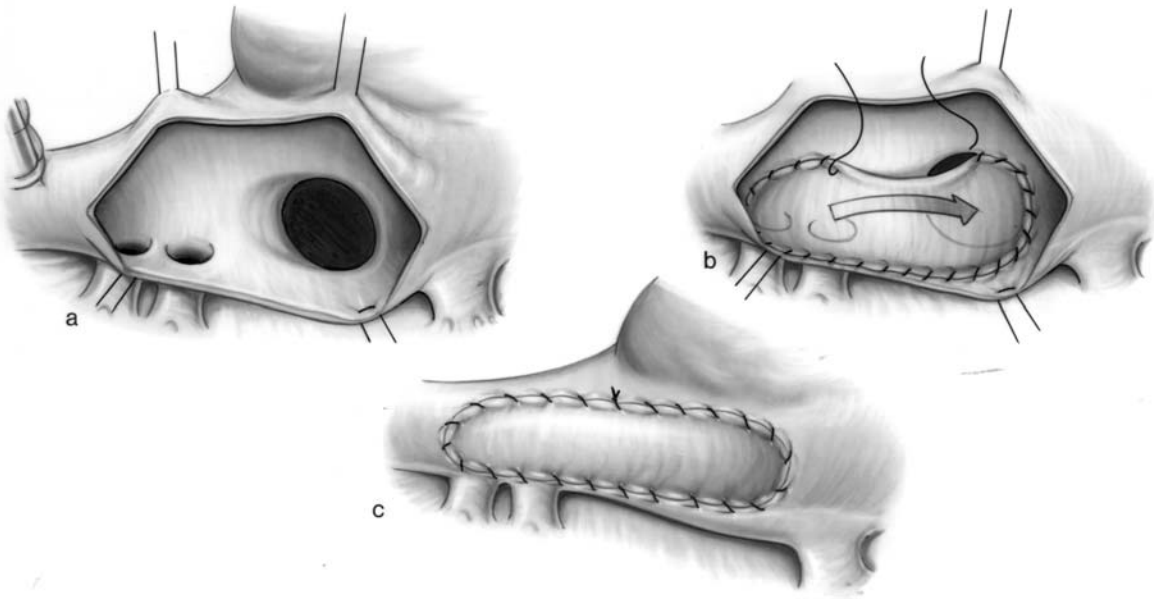

Figure 25.8

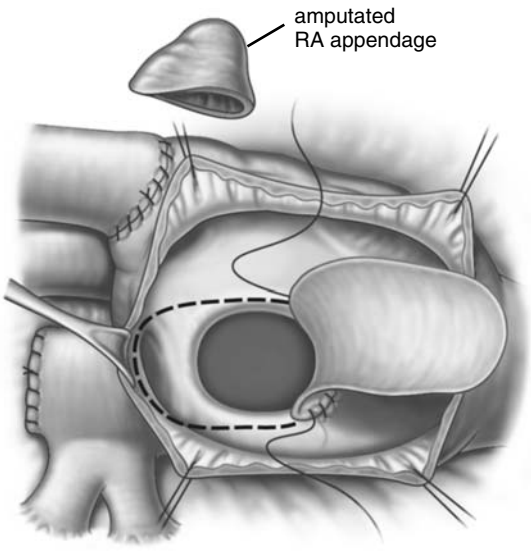

Figure 25.9

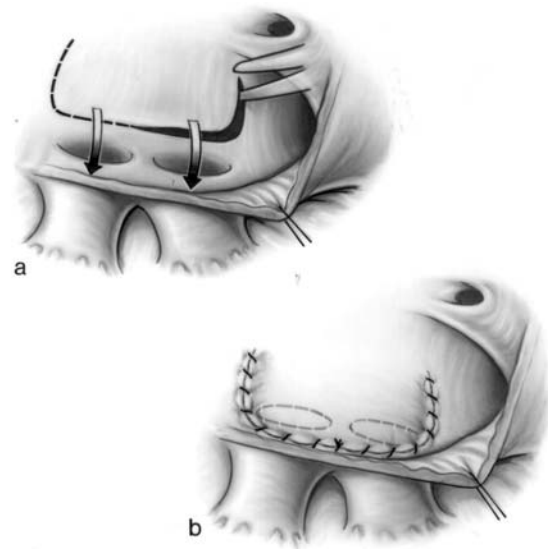

Figure 25.10

Repair of Drainage from the Right Pulmonary Veins to the Right Atrium

The right atrium is opened with the usual oblique incision. If a posterior defect is present, it is simply closed with a pericardial patch. Laterally, the suture runs above the orifices of the pulmonary veins to divert the pulmonary venous blood through the ASD into the left atrium. If the atrial septum is intact, it can be opened with a longitudinal incision posteriorly with two side cuts (**Figure 25.10a**); this creates a flap, which can be sutured to the lateral atrial wall above the orifice of the pulmonary veins (**Figure 25.10b**). Alternatively, a large defect in the fossa ovalis with posterior extension may be created. The pulmonary veins are then redirected to the

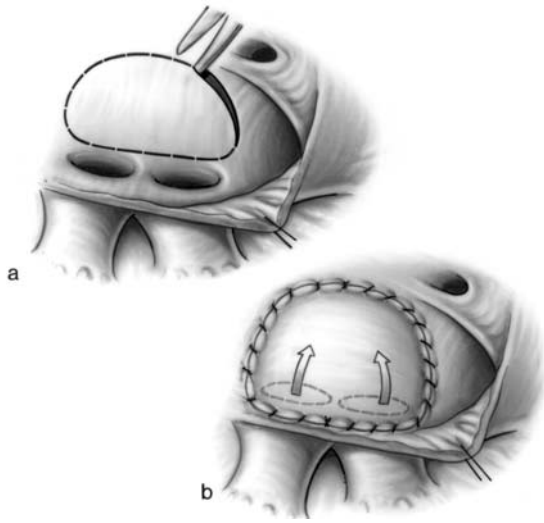

Figure 25.11

left atrium, through the ASD, using a pericardial patch (**Figures 25.11a,b**).

Repair of Drainage from the Right Pulmonary Veins to the Inferior Vena Cava (Scimitar Syndrome)

Presentation is variable according to age at which the diagnosis is made. Infants presenting in heart failure usually have multiple anomalies. Usually, all pulmonary veins form a common curved channel (scimitar), which drains to the IVC either above or below the diaphragm (**Figure 25.2d**). Scimitar syndrome is often associated with right lung hypoplasia and dextroposition of the heart. The blood supply to the part of the right lung may be via aortopulmonary collateral vessels originating from the descending aorta. Association with lung sequestration has been reported (Alizivatos *et al*., 1985). Significant collaterals, if present at cardiac catheterization, should be coil-embolized. An ASD or a patent foramen ovale may or may not be present.

Two techniques can be used: tunnelling from the orifice of the right pulmonary veins in the IVC through an enlarged ASD to the left atrium with a baffle, or disconnecting the anomalous venous channel and anastomosing it directly to the left atrium (Murphy *et al*., 1971; Pelleier and Spray, 2001).

Tunnelling

Tunnelling is preferred because of the tendency of venous anastomoses to constrict with growth. The patient is placed on cardiopulmonary bypass, and the SVC and IVC are cannulated (**Figure 25.12**). The latter can be spared by using femoral cannulation. The authors prefer to undertake the surgery, which can be difficult, using deep hypothermic circulatory arrest. The aorta is cross-clamped and blood cardioplegic solution is infused into the root of the aorta. While the patient is being further cooled (to 18°C), the ASD is inspected and enlarged or, if absent, it is created (**Figure 25.13**). This may not be straightforward because of dextroposition of the smallish LA.

Inferior enlargement of the ASD is performed, and care must be taken not to cut through the posterior wall of the atrium and not to cause damage to the area anterior to the coronary sinus. Native pericardium or Gore-Tex baffle trimmed from a tube graft is prepared. When the

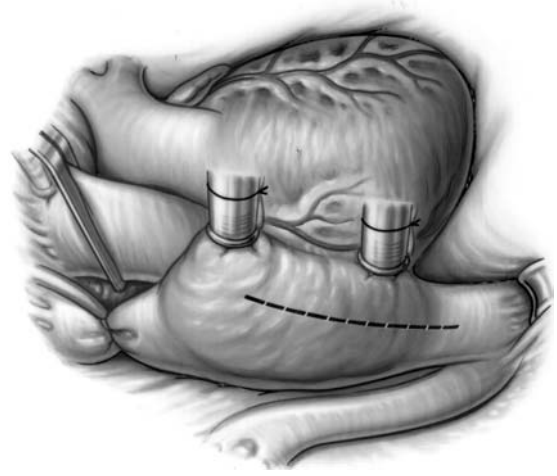

Figure 25.12

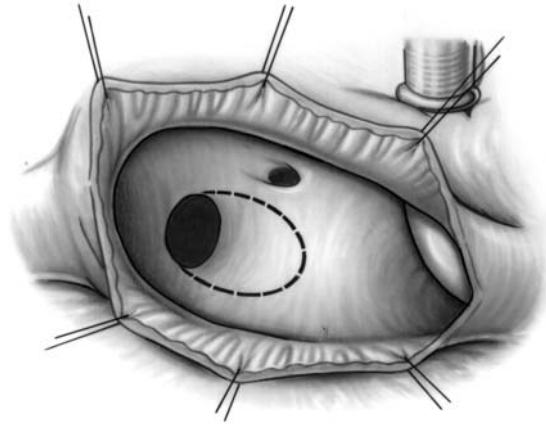

Figure 25.13

nasopharyngeal temperature reaches 18°C, perfusion is temporarily stopped, blood is drained into the oxygenator and the IVC cannula, if present, is removed. The atrial incision is then extended into the IVC, and the openings of the anomalous pulmonary vein (PV) and the hepatic veins (HV) are identified under direct vision (**inset, Figure 25.14**). If the orifice of the pulmonary vein cannot be easily identified because its entrance to the IVC is deep below the diaphragm, the diaphragm can be divided. Another strategy which the authors have used to correctly identify the opening of the anomalous pulmonary vein is the percutaneous placement of a soft wire by the interventional cardiologist immediately before surgery.

The channel is then constructed by suturing the patch around the orifice of the pulmonary veins, using an open technique (**Figure 25.14**). The patch is sutured toward the ASD; an adequate channel must be created for pulmonary venous drainage. A purse-string effect should be avoided. When the suture line reaches the level of the coronary sinus, the IVC cannula is replaced (**Figure 25.15a**), and cardiopulmonary bypass is restarted. Rewarming of the patient may be started. The patch is then completed around the ASD, the aortic clamp is removed, and usual de-airing is accomplished. If the incision had to be extended deep into the IVC it may be advantageous to repair the IVC incision with a small pericardial patch (**Figures 25.15b,c**).

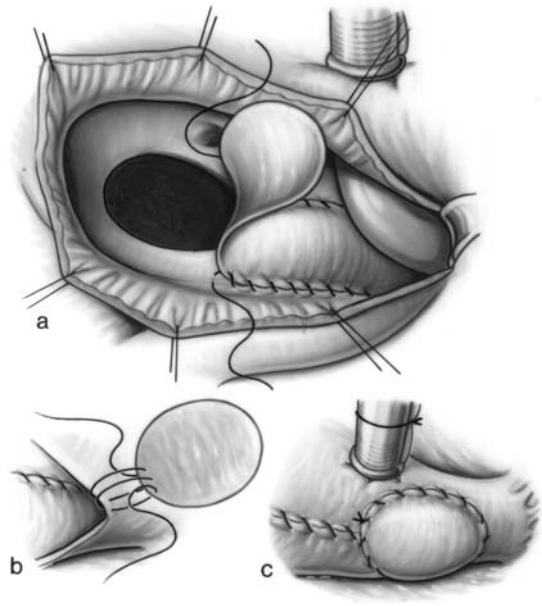

Figure 25.15

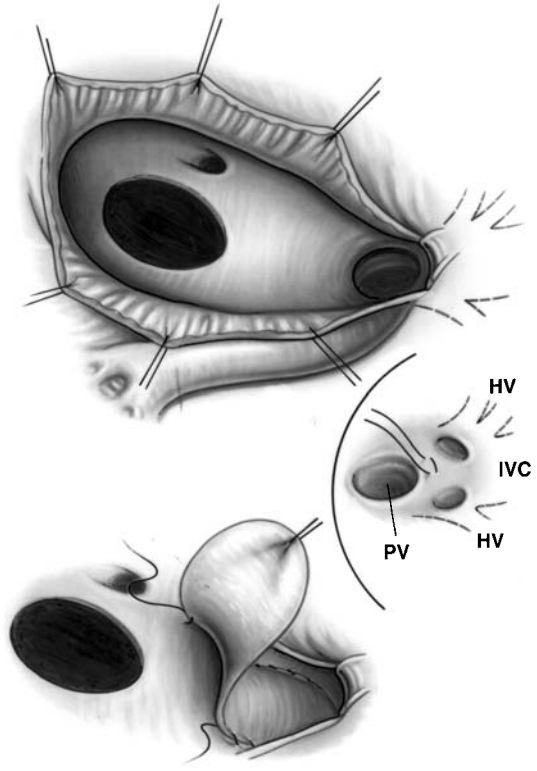

Figure 25.14

Anastomosis of the Anomalous Pulmonary Vein to the Left Atrium

If the anomalous vein joins the IVC too deep below the diaphragm, it can be ligated, divided and anastomosed to the left atrium. Operation is again performed on cardiopulmonary bypass because clamping of the vein during the anastomosis may cause severe congestion of the right lung, with damage to the pulmonary capillaries. Both venae cavae are cannulated separately and the anomalous pulmonary vein is also dissected. The temperature of the perfusate is lowered, the aorta is cross-clamped, and cardioplegic solution is infused. The right atrium is then opened and the atrial anatomy visualized. The right pulmonary vein may be anastomosed side-to-side to the left atrium to achieve a long anastomosis, and then the vein is ligated distally (**Figure 25.16**). Alternatively, the vein may be divided and anastomosed end-to-side onto the left atrium. A wide, cobra-like anastomosis must be constructed because the venous anastomosis has a tendency to shrink (**Figure 25.17**). Kinking of the vein must be avoided.

An alternative approach through the right thoracotomy without the use of cardiopulmonary bypass was recently reported by Brown *et al*. (2003). Anastomosis between

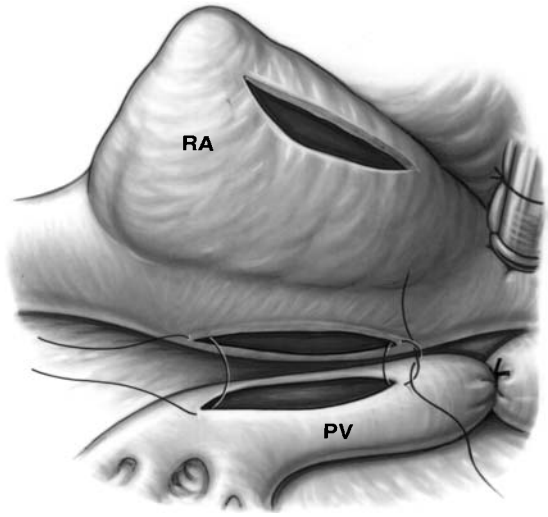

Figure 25.16

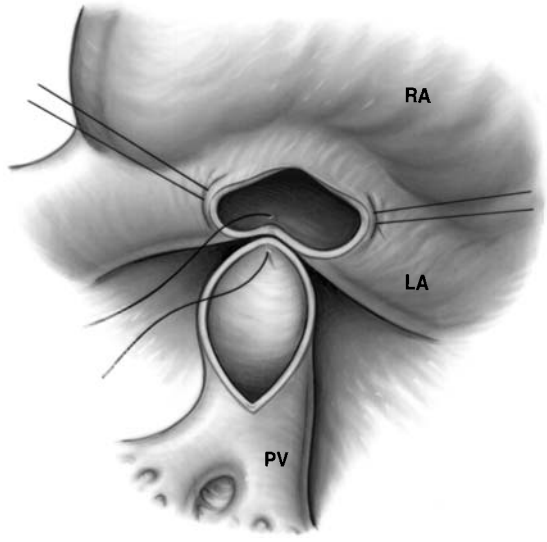

Figure 25.17

the pulmonary veins and the right atrium, if technically easier, can be accomplished. The ASD is enlarged inferiorly. An obliquely cut end of the pulmonary vein is then anastomosed to the lateral aspect of the right atrium. Redirection of blood flow from the right pulmonary veins into the left atrium is then achieved by suturing a wide patch around the ASD and the orifice of the right pulmonary veins (**Figure 25.18a**). The completed operation is shown in **Figure 25.18b**.

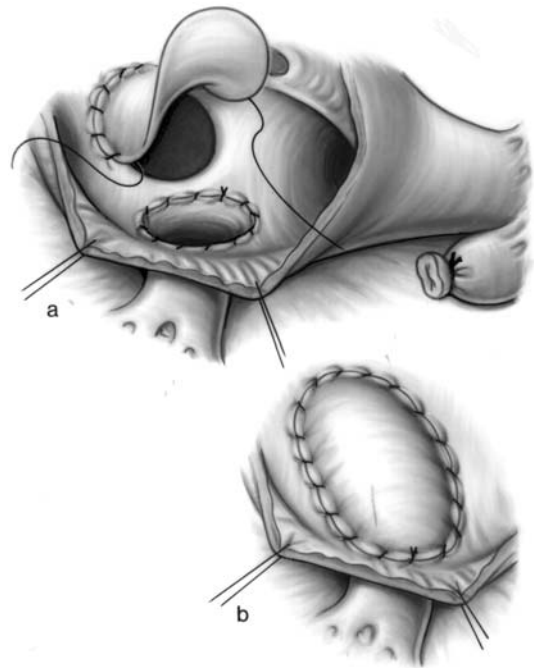

Figure 25.18

Anomalous Drainage from the Left Lung into the Innominate Vein

This is a rare isolated anomaly (Ports *et al*., 1979). It is most often a part of mixed-type total anomalous pulmonary venous drainage: the right pulmonary veins drain into the right atrium or coronary sinus, and the left pulmonary veins drain via the left vertical vein into the innominate vein.

Isolated drainage of the left upper-lobe vein to the innominate vein may be left uncorrected. If it is part of a complex defect and we consider this lesion physiologically important, we would correct it. If, however, the whole left lung drains anomalously, the physiological consequences are similar to those encountered in large ASD, and correction is indicated. Although the operation can be performed through the left side of the chest as a closed procedure, we prefer repair from the midline sternotomy on cardiopulmonary bypass. The pulmonary veins and the vertical vein are carefully dissected. Hypothermia and aortic cross-clamping are used. The vertical vein is ligated close to the junction with the innominate vein. It is important not to construct the anastomosis with the left atrial appendage. The appendage is therefore amputated, and the incision is extended into to the left atrium. The vertical vein is spatulated and approximated to the opening in the left atrium (**Figure 25.19**). Anastomosis with 7-0 or 6-0

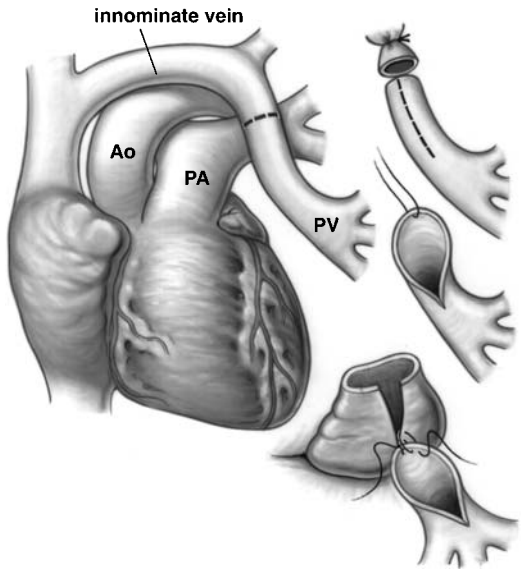

Figure 25.19

polypropylene sutures is performed; care is taken to avoid any purse-string effect. This is best achieved by tying sutures at both ends and running the continuous suture in between.

''RESIDUAL'' ASD

A small residual defect may result from faulty surgical technique. It is more likely to occur in older patients with congestive heart failure. Reoperation for a residual or recurrent ASD is required only rarely. In the current era, device closure during cardiac catheterization would deal with the problem, if indicated.

In patients with elevated right ventricular end diastolic pressure, such as those who have undergone repair of tetralogy of Fallot or pulmonary atresia, a small ''residual'' ASD or patent foramen ovale may optimize the cardiac output at the expense of a right to left shunt. The shunt usually diminishes after a few days when the right ventricular compliance improves. Some surgeons recommend leaving the foramen ovale open in patients in whom pulmonary hypertensive crisis (e.g. truncus arteriosus) may develop. The patent foramen ovale provides a useful valved mechanism during a crisis; in the long term, it does not cause problems.

COMPLICATIONS

Late Pericardial Effusion and Tamponade

This occurs commonly after repair on cardiopulmonary bypass. The incidence has been quoted as 16% and 1.5%, respectively (Yip *et al*., 1997). Creation of the right pleuro-pericardial window should prevent this complication. Routine postoperative echocardiogram should be performed prior to discharge. If effusion is detected, its progression or regression should be carefully monitored.

Arrhythmias

Patients with ASD are at risk of developing arrhythmias both before and after the repair, particularly if repair is undertaken later in life (Konstantides *et al*., 1995). In the postoperative patient, atypical flutter or incisional re-entry tachycardia (Kalman *et al*., 1996) may occur around the right atriotomy scar or the ASD patch suture line. Atrial fibrillation may become the dominant rhythm during the long-term follow-up. The management of arrhythmias is discussed in Chapter 10.

RESULTS

ASD Closure

Even in the early years of cardiac surgery, good results of surgical closure of ASD were reported. Rahimtoola *et al*. (1968) reported 696 patients operated on between 1954 and 1965, of whom 22 (3.2%) died; Hallman and Cooley (1975) 10 deaths (1.4%) among 707 patients. In a combined series from Birmingham, Alabama, USA, and Green Lane Hospital, Auckland, New Zealand, Kirklin and Barratt-Boyes (1986) reported five deaths (1%) among 518 patients.

The authors closed ASDs in 150 children between 1965 and 1969; three (2%) died, and two of those three were infants. In the most recent series (2000–2004), 79 patients underwent isolated ASD repair. 15 were infants, two of those had concomitant tracheal repair. There was no hospital mortality.

Survival after repair of ASD was influenced by early age (Bull *et al*., 1981) and increased pulmonary arterial resistance. In the series of Steele *et al*. (1987), patients with pulmonary arterial resistance of 15 U/m^2 did very poorly, whereas those with resistance under 10 U/m^2 did reasonably well. However, resistance increased to these levels is very rare. Follow-up was not long enough; the long-term prognosis in this group of patients remains uncertain. Currently, in our practice, patients with borderline pulmonary vascular resistance, but with evidence of L–R shunt, are considered to have their defect closed with a fenestrated patch.

Closure via Right Thoracotomy

During 1985–1992, one author (J.S.) closed ASDs through right thoracotomy in 72 children aged 1.0–14.3

years (mean 5.7 years). The average hospital stay was 5 days. There was no early or late mortality. Däbritz *et al*. (1999) reported 87 female patients, age range 3–56 years, operated on for all types of ASD via limited right anterolateral thoracotomy during 1982–1993. There were no intraoperative complications. However, areas of anaesthesia and scar pain were quite frequent (16.5%) in their experience.

Device Closure of ASD

Berger *et al*. (1999) reported the results and complications of surgical vs. trans-catheter occlusion of ASD. Complete closure rates and complications were identical. Hospital stay was shorter in the device group.

Repair of Sinus Venosus Defect

Closure of sinus venosus ASD with partial anomalous pulmonary venous return does not carry an increased risk. Trusler *et al*. (1980) operated on 52 patients between 1962 and 1975, and there were no early or late deaths. One patient had a significant SVC–right atrial gradient (6 mmHg); four had small L–R shunts. Russell *et al*. (2002) reported similar results in 44 patients; 16% of patients were not in sinus rhythm at follow-up to 15 years. In our results (2000–2004), 14 patients aged 2 months–9 years underwent repair of sinus venosus defect, using either a Warden procedure or pulmonary vein to left atrium baffle without hospital mortality or any early complication. However, long-term follow up will be necessary to assess possible SA node dysfunction SVC PV obstruction.

Repair of Scimitar Syndrome

Najm *et al*. (1996) reported the results of repair of this anomaly in 17 patients. Six were infants, 11 were older children. There were no deaths; however, eight patients (47%) developed pulmonary venous stenosis. All six infants developed stenosis. Two required reoperation. Brown *et al*. (2003) used the right thoracotomy approach for the repair in nine patients, mean age 11.5 ± 7.6 years. There were no early or late deaths or reoperations. Echocardiography demonstrated patent anastomosis in all patients. Occasionally lung resection may be necessary.

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26

Ventricular Septal Defects

C. van Doorn and M. R. de Leval

Found either as an isolated lesion or in combination with other cardiac anomalies, ventricular septal defects (VSDs) are the most common congenital intracardiac defects of clinical importance. First described as an entity by Roger in 1879, the first successful closure of a VSD under direct vision, using cross-circulation, was reported by Lillehei *et al*. in 1955. Staged repair with pulmonary artery banding was recommended because of the high risk of open heart surgery in infancy. Today, pulmonary artery banding still has a role in the management of VSD in the setting of associated complex cardiac lesions. Device closure of VSD has been recently added to our armamentarium.

DIAGNOSIS

The pathophysiology of isolated VSDs is determined by the size of the defect and the pulmonary vascular resistance. The majority of candidates for surgical treatment can be divided into two groups. The first group includes patients, usually infants, in congestive heart failure, experiencing failure to thrive, and who frequently have a history of repeated chest infections. They have large defects with low pulmonary vascular resistance and a large left-to-right (L–R) shunt.

The second group is made up of older children, who are less symptomatic and who may still have a large defect and a large L–R shunt and/or an increased pulmonary vascular pressure and resistance. On physical examination, a systolic thrill over the left precordium is usually present. A pan-systolic murmur, maximal in the fourth left intercostal space, is characteristic. The second sound is normally split unless there is a pulmonary vascular obstructive disease, when it becomes single. When pulmonary vascular resistance is not seriously elevated, there is a prominent third sound and a diastolic filling rumble is heard at the apex, indicating a large flow across the mitral valve.

Chest X-ray shows cardiac enlargement and increased pulmonary vascular markings. The electrocardiogram reveals overloading of both ventricles. The site and position of the defects is assessed by two-dimensional (2D) echocardiogram (Bierman *et al*., 1980), which also should provide detailed morphological information on the remainder of the heart, including the aortic arch and ductus arteriosus. Echo Doppler interrogation is useful to estimate the right ventricular pressure and the magnitude of the L–R shunt.

Colour Doppler echocardiography is particularly useful to visualize multiple VSDs. In infants with a large L–R shunt and, on combined 2D echocardiography and Doppler flow evaluation, with evidence of an isolated ventricular septal defect, surgical repair can safely proceed on the basis of echo only (Carotti *et al*., 1997).

In borderline cases, cardiac catheterization with assessment of pulmonary vascular resistance remains necessary to obtain more precise information on the magnitude of the L–R shunt, to measure the pulmonary arterial pressure and calculate the pulmonary vascular resistance. Left ventricular-angled angiocardiograms, showing in profile the interventricular septum (Soto *et al*., 1978), have proved to be useful in defining the precise anatomy of the defects. Detailed cardiac morphology, often with three-dimensional (3D) reconstruction, can be increasingly demonstrated using MRI and CT, especially in the setting of complex VSDs. In addition, pulmonary vascular resistance can be quantified by combining phase contrast magnetic resonance flow with invasive pressure monitoring (Muthurangu *et al*. 2004).

INDICATIONS

Large VSD

Early surgical treatment is indicated for infants with large VSDs who remain in intractable congestive heart failure despite medical treatment. Primary closure of large isolated VSDs, rather than banding of the pulmonary artery, has long been accepted as the treatment of choice. We continue to band infants with multiple VSDs of the

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Swiss-cheese variety, some VSDs and coarctation of the aorta, and VSDs with straddling of one atrio-ventricular valve. In addition, infants with very low body weight and those who are not fit for cardiopulmonary bypass because of an intercurrent illness are considered for pulmonary artery banding.

For those VSDs that are planned for possible future interventional closure, we have recently introduced a dilatable pulmonary artery band. A 3–4 mm wide Gore-Tex strip is placed around the main pulmonary artery and tightened using two surgical clips. Debanding can be achieved in the catheter laboratory by balloon dilatation of the band. In some instances VSD may be closed (by device or surgery).

Severe pulmonary hypertension leads to pulmonary vascular disease, which tends to worsen with age and is eventually fatal. Pulmonary vascular remodelling towards normal has been demonstrated at a young age and the most crucial factor in determining late outcome is the age at which repair is carried out. Young infants with echo demonstration of high pulmonary artery pressure should be operated on within 3 months of age. Early repair would provide the best chance of surgical cure with a normal pulmonary vascular resistance 5 years later (Blackstone *et al*., 1976).

Moderately Large VSD

The risk of VSD closure is low and the development of pulmonary vascular disease is unpredictable. Therefore, patients who for some reason still show a large L–R shunt ($> 2{:}1$) by the age of 2 years are operated early. In older patients with large VSDs, pulmonary hypertension and increased pulmonary vascular resistance, the probability of surgical cure is uncertain (duShane and Kirklin, 1973). In practice, we recommend closure of VSDs if the pulmonary:systemic blood flow ratio is 1:1.5 or greater. Patients with pulmonary arteriolar resistance of 10 units/m^2 or more are considered to be inoperable; those with the resistance of 6–8 U/m^2 have to be evaluated very carefully. Improved understanding of the genetics and cell biology of pulmonary hypertension, and recent advances in the treatment of primary pulmonary hypertension in adults, indicate that it could be possible to arrest and even perhaps reverse the disease process. The potential is likely to be greater in those children in whom the vasculature is still remodelling. Although the field is moving rapidly, the indications for treatment and therapeutic modalities remain to be established in the young (Howarth, 2002).

Small to Moderate VSDs

Small and moderately large VSDs with normal pulmonary vascular resistance have a natural tendency to become smaller and may eventually close. Surgery is not normally indicated for persistant small defects. Their closure remains controversial, especially for grown-up patients. It is nevertheless recommended that even small VSDs be closed after one episode of infective endocarditis if the defect remains open once the infection has been cured.

Doubly Committed Subarterial VSDs

Doubly committed subarterial VSDs must be discussed separately because of the particular aspects of their clinical presentation and surgical treatment. They occur within the outlet septum that is shared by both the right and left ventricle. The upper border of the defect is formed by the pulmonary and aortic valve cusps, which are frequently at the same level—rather than the usual cephalad offset of the pulmonary valve—and are only separated by a thin rim of fibrous tissue. Large defects tend to produce severe congestive heart failure in infancy. They have no tendency to close, and are prone to induce early pulmonary vascular changes. These defects should be closed primarily early in infancy. Small and moderately large doubly-committed subarterial VSDs may cause aortic valve prolapse and regurgitation (Tatsuno *et al*., 1973; Van Praagh and McNamara, 1968). This is the result of a lack of support of the aortic valve in the absence of infundibular septum and the pressure drop from the Venturi effect beneath the aortic valve. Only VSDs of moderate or small size are likely to produce the rapid flow velocities needed to pull the right coronary cusp into the VSD, which will eventually cause aortic valve prolapse and regurgitation. These defects should be closed as soon as aortic valve regurgitation is detected or even earlier in the presence of aortic valve prolapse (de Leval *et al*., 1988).

SURGICAL TECHNIQUES

Operations for VSDs are performed through a midline sternotomy incision, using conventional hypothermic cardiopulmonary bypass with cold blood cardioplegia. Bicaval cannulation with direct cannulation of the superior vena cava facilitates the exposure of the defects through the right atrium. Rarely, in very small infants, we use the technique of deep hypothermia and total circulatory arrest.

The procedure aims at obtaining a secure and complete closure of the defect without damaging any important structure and, in particular, the conduction tissue. The rules that should enable the surgeon to predict the course of the conduction system with a fair degree of accuracy are described in Chapter 8. They are based on differentiating perimembranous defects from other

defects. In perimembranous defects, the penetrating and branching bundles are closely related to the posterior and inferior margin of the defect. Placement of the sutures on the right ventricular septal side and away from the postero-inferior margin of the defect is always safe.

Numerous surgeons, however, have now closed large numbers of perimembranous VSDs by placing superficial sutures on the free margin of the defect without damaging the conduction system. This technique appears to be safe, in particular for the closure of perimembranous outlet VSDs in which a prominent posterior extension of the trabecula septomarginalis extends to the area of the membranous septum covering the branching bundle, which becomes more left-sided. In these circumstances, it is therefore safe to place the sutures superficially on the right side of the crest of the defect, even in its postero-inferior angle.

This is the anatomic variant most commonly seen in the perimembranous outflow VSD of tetralogy of Fallot, in which the major part of the main conduction axis remains left-sided and remote from the crest of the defect. This technique, however, is not recommended for perimembranous inlet and trabecular defects in which the conduction axis may run superficially in close relationship to the postero-inferior angle of the defect. Although the branching bundle is usually left-sided, it sometimes runs along the crest of the antero-inferior aspect of a perimembranous defect near the site of origin of the right bundle branch. It is therefore important to stay to the right of the crest of the septum, not only along its posterior or inferior margin but up to and beyond the antero-inferior angle.

Muscular VSDs are characterized by the presence of a bar of muscle separating the defect from the area containing the membranous septum. The conduction tissue is therefore remote from the margin of the defect, onto which it is safe to place the sutures to anchor the patch. Sometimes, however, the bar of muscle separating a trabecular or outlet muscular defect and the membranous septum is so narrow that the penetrating and the branching bundles are very near the crest of the defect. In these cases, it is safer to consider the defect as a perimembranous defect, remembering that it is always safe to place the sutures through the base of the septal leaflet of the tricuspid valve. Similarly, an inlet muscular defect that extends towards the area of the membranous septum may come very close to the conduction axis, which in this particular situation will be to the surgeon's left-hand side when seen through the right atrium (Kurosawa and Becker, 1987).

Occasionally, VSDs are surrounded by fibrous tissue resulting from endocardial fibrosis with or without tricuspid valve leaflet tissue. In such instances, the fibrous tissue can be used to anchor the patch (Kurosawa and Becker, 1987). Care must be taken, however, not to leave a residual defect between the fibrous ridges.

Muscular defects can occur anywhere in the ventricular septum, but are most common in the middle portion of the trabecular septum and may be overlaid by the septal band (trabecula septomarginalis). Thus, even when single on the left ventricular side, some defects have at least two openings on the RV side.

Depending on their location in the interventricular septum and the presence or absence of associated cardiac anomalies, VSDs are closed through the right atrium, the right ventricle, one of the great arteries or rarely through the left ventricle. Techniques of closure described here apply only to patients with atrio-ventricular concordance.

Closure through the Right Atrium

The right atrial approach is chosen to close most isolated VSDs of the perimembranous or inlet muscular types. Trabecular muscular defects and infundibular muscular defects are often, but not always, accessible through the right atrium.

An oblique incision well anterior to the sulcus terminalis parallel to the atrio-ventricular groove is made from the appendage inferiorly toward the inferior vena cava. Two stay sutures are placed on the posterior margin of the incision, taking care to avoid the sinus node that lies subepicardially in the sulcus terminalis at the junction of the superior vena cava with the right atrium. The tricuspid valve ring is exposed by everting the anterior margin of the atriotomy. This can be achieved by placing two stay sutures on the inner wall of the right atrium, close to the tricuspid valve ring, 1–2 cm from the cut edge or by using a small retractor (**Figure 26.1**).

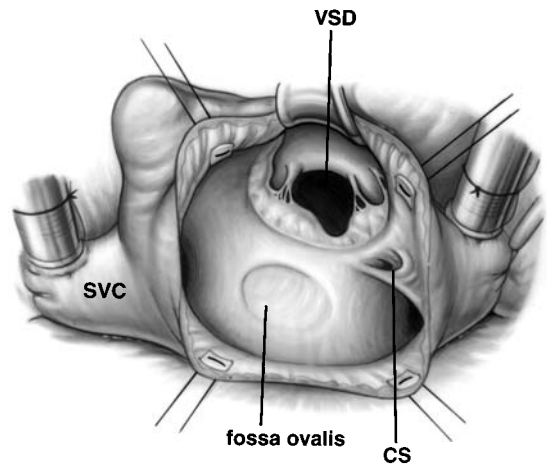

Figure 26.1

The ventricular septum is then explored through the tricuspid valve, so that the defects can be seen and the anatomy assessed. If the central fibrous body (area of continuity of the membranous septum and the annulus of the tricuspid, mitral, and aortic valves) forms part of the rim of the defect, one is dealing with a perimembranous defect, which is therefore closely related to the atrio-ventricular conduction tissue. In practice, the anteroseptal commissure of the tricuspid valve, which is supported by the medial papillary muscle, is immediately to the left side of the conduction tissue. Sometimes, a cleft in the septal leaflet points directly toward the central fibrous body. From the atrio-ventricular node (apex of the triangle of Koch), the penetrating bundle passes through the central fibrous body and then continues into the branching bundle on the inlet side (right side) of the defect.

Perimembranous Trabecular Ventricular Septal Defects

From the area of the central fibrous body, the long axis of these defects is orientated toward the cardiac apex. Clockwise around the defect from the central body, the boundaries are the central fibrous body and the septal leaflet directly adjacent to it, the infundibular septum, the trabecular septum and the inlet septum. As illustrated in **Figure 26.2**, the antero-septal commissure of the tricuspid valve attached to the medial papillary muscle is more often above and to the left side of the defect. In some cases it is attached toward the apex, and the chordae lying across the defect may therefore compromise its exposure and render the closure more difficult. A cleft (arrow) in the septal leaflet of the tricuspid valve (**Figure 26.2**) frequently extends towards the central fibrous body, indicating the position of the penetrating bundle.

Beneath the caudad aspect (right side) of the septal leaflet, there may be a remnant of the interventricular membranous septum buttressing the defect beneath the tricuspid valve that may contain the penetrating bundle. Once it has penetrated the fibrous body, the bundle tends to veer toward the left ventricular side of the septum and bifurcate immediately, so that the crest of the inlet and trabecular septa are not occupied by the branching bundle (**Figure 26.3**). This, however, is not consistent, and in other patients the branching bundle is more closely related to the rim of the defect.

Once the anatomical diagnosis of perimembranous trabecular VSD has been made, two fine traction sutures may be placed on the septal leaflet and held in mosquito clamps to facilitate exposure of that part of the defect hidden beneath the valve tissue (**Figure 26.3**). Small defects partially covered by tricuspid valve may be repaired by direct sutures. In most cases, however, a patch of xenograft pericardium, Gore-Tex (0.4 mm in infants; 0.6 mm in older children) or Dacron velour is inserted with interrupted or continuous sutures.

Patch Closure with Interrupted Sutures

For this technique, pledget-supported interrupted mattress sutures are first placed around all margins of the defect and then passed through an appropriately tailored

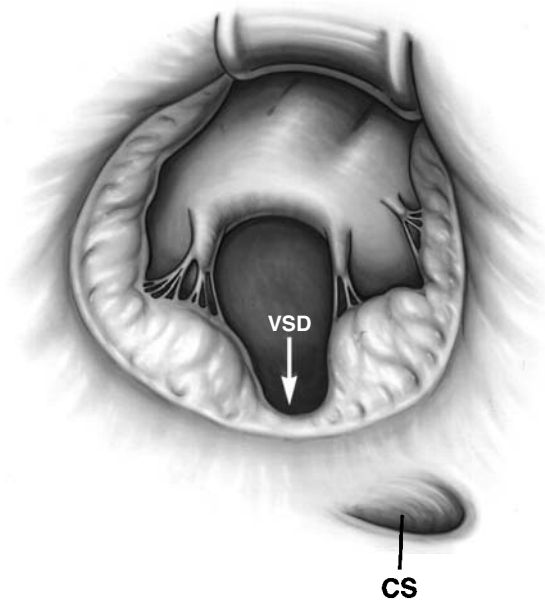

Figure 26.2

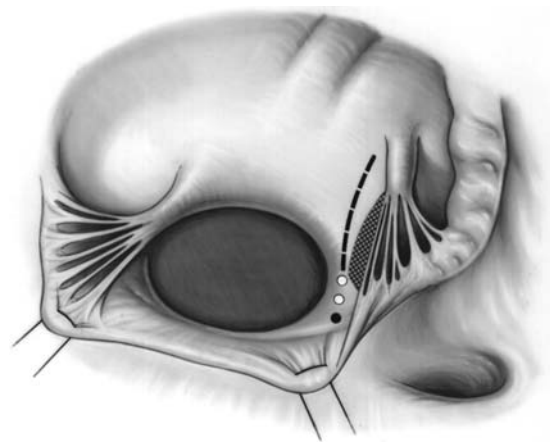

Figure 26.3

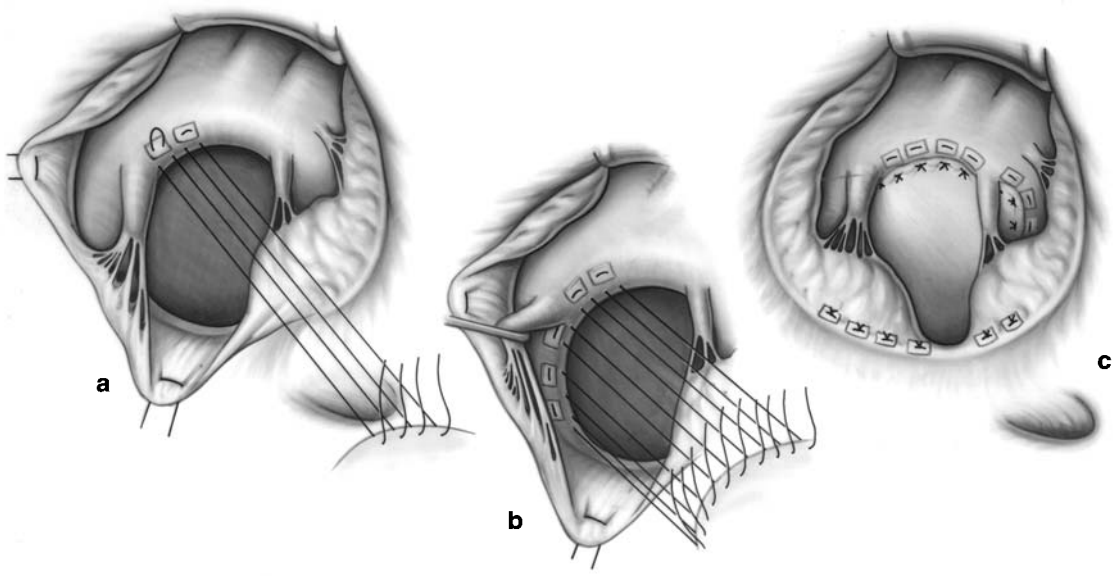

Figure 26.4

patch, which is lowered down and tied in. In small infants, 6-0 double-ended synthetic sutures are used, mounted on small semi-circle needles and buttressed with small Teflon or pericardial pledgets. In older children we use 5-0 sutures (Doty and McGoon, 1983). The first stitch is deeply placed in that part of the margin of the defect most distant from the surgeon, on the trabecula septomarginalis (**Figure 26.4a**).

The sutures are placed on a rubber-shod clamp (not illustrated on **Figure 26.4a,b**). Traction on that first stitch and on the successive sutures exposes the cephalad margin of the defect (left side), through which the sutures can be safely placed, until the area of the central fibrous body is reached (**Figure 26.4b**). The next two or three stitches are placed through the septal leaflet of the tricuspid valve (**Figure 26.4c**) about 1–2 mm from the annulus. If a defect is present in the septal leaflet, care must be taken to avoid the bottom of the cleft, which may reach back to the penetrating bundle. It is best to place the stitches on each side of the cleft on the leaflet tissue. If a remnant of the membranous septum is buttressed beneath the septal leaflet, it may also contain the penetrating bundle and the sutures must therefore be placed through the valve itself.

Next, sutures are placed on the inlet septum and back to the trabecula septomarginalis on the caudad (right side) margin of the defect. These sutures do not pass through the full thickness of the septum but grasp only its right surface; they should be placed 3–5 mm away from the edge of the defect to avoid the branching bundle in case it does not bifurcate immediately after penetrating the central fibrous body. The margins of the defect have now been completely covered with sutures and the prosthetic patch is trimmed to an appropriate shape and size. The latter must be greater than the actual defect, as the suture line in the posterior–inferior angle is away from the margin of the defect. The interrupted sutures are passed through the patch, which is lowered into position, and are then tied (**Figure 26.4c**). If the defect is crossed by chordae that cannot be easily retracted, the patch is lowered after the cephalad stitches have been passed through it so as to position it beneath the chordae before attaching the other stitches.

Patch Closure with Continuous Sutures

The patch is tailored to the appropriate shape and size. Posteriorly or inferiorly, the patch should be wider than the defect, so as to cover the triangular area containing the conduction tissue (**Figure 26.5**). The patch is initially held away from the heart on the drapes covering the right sternal edge. In small infants, we usually use a suture of 6-0 Surgilene on a 10 mm semicircular, rounded-bodied needle. The suturing starts at the 12 o'clock position, taking a deep bite in the infundibular septum. Continuous suturing is then carried out superiorly. Traction on the first stitch and on the successive sutures exposes the cephalad part of the defect. The patch is then pulled down into position as the surgeon works towards the tricuspid valve annulus. When this point has been reached, the suture is passed through the base of the septal leaflet of

the tricuspid valve and then through the right side of the edge of the pericardium. Two or three mattress stitches are placed back and forth through the pericardial edge, the tricuspid leaflet, and the patch. The suture is held with a rubber-shod clamp (**Figure 26.6a**).

Working now with the second arm of the suture and towards himself/herself, the surgeon takes a few deep bites in the trabecula septomarginalis. When the medial papillary muscle or the antero-inferior angle of the defect is reached (see point A, **Figure 26.5**), the stitches are placed superficially along line AB 4–6 mm away from the inferior margin of the defect, to avoid the branching bundle in case it does not bifurcate immediately. When the tricuspid valve is reached (point B), the stitch penetrates the base of the septal leaflet and is then passed into the pericardial edge at a distance from the superior arm of the suture equivalent to the remaining length of tricuspid valve tissue, over which the patch will be anchored. The needle penetrates the septal leaflets about 2 mm away from the annulus. When the superior limb of the suture is reached, the two arms are held on a rubber-shod clamp, and a strip of pericardium into which the sutures have been placed is trimmed. The sutures are then pulled taut, and the pericardial strip is snugged down along the base of the attachment of the tricuspid leaflet, which is sandwiched securely between the pericardium and the patch (**Figure 26.6b,c**).

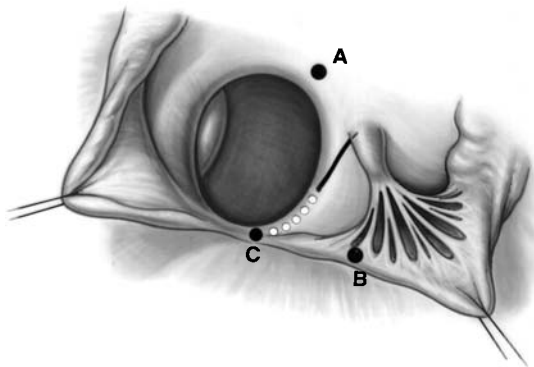

Figure 26.5

In the presence of numerous chordae tendineae obscuring the VSD (**Figure 26.7**), the closure can be facilitated by detaching the septal leaflet, so as to have direct access to the margin of the VSD beneath the chordae (**Figure 26.8a**). The incision is made parallel to, and 2–3 mm from, the annulus, taking great care not to disconnect the leaflets one from the other. The detached leaflets are reflected into the right atrium, and the VSD is closed (**Figure 26.8b**); the tricuspid valve is then repaired with a continuous suture of fine material (**Figure 26.8c**). We do not recommend this technique in small infants in whom valve tissue is thin and fragile.

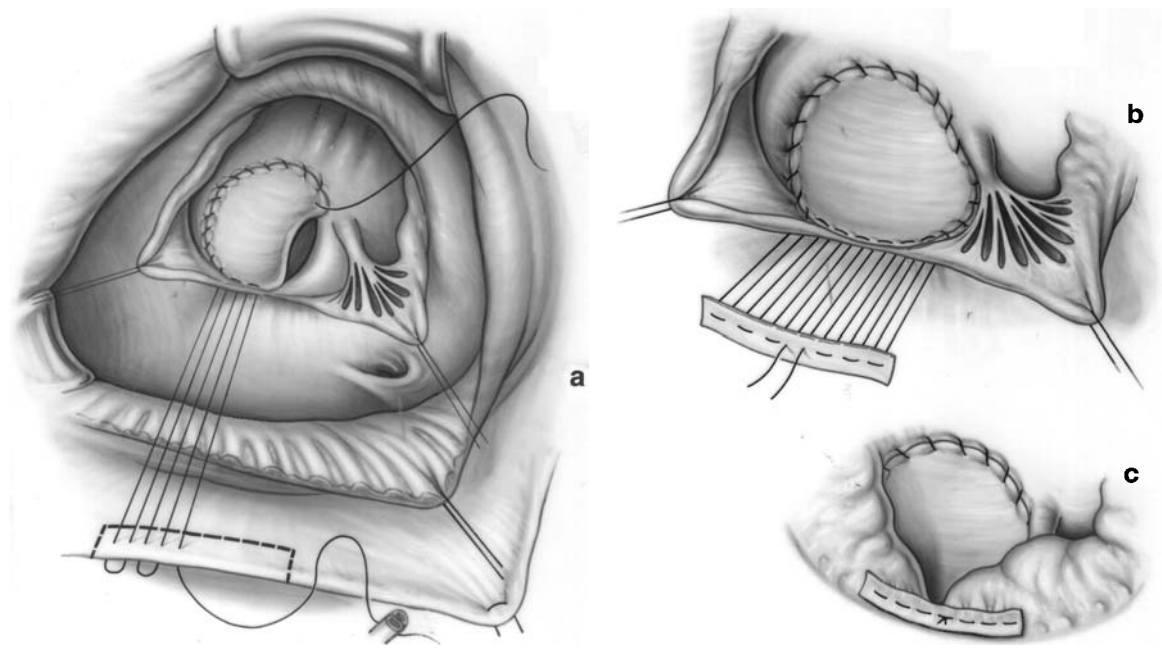

Figure 26.6

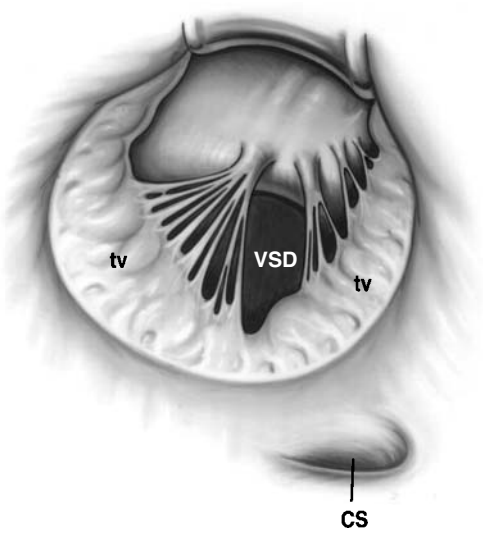

Figure 26.7

We have found it useful in bigger children on some occasions.

So-called Gerbode's defects constitute a haemodynamic entity with a left-ventricle-to-right-atrial shunt. They are considered from an anatomical point of view as a variety of perimembranous trabecular defects. This is illustrated in **Figure 26.9a**, which shows an intact atrio-ventricular component of the membranous septum and the annulus of the tricuspid valve on the atrial side of the defect. If the defect were truly in the atrio-ventricular component, the tricuspid valve annulus would be on the ventricular aspect of the defect, as depicted in **Figure 26.9b**. In closing these defects, the proximity of the conduction system has to be kept in mind. The often redundant tricuspid valve tissue attached on the crest of the ventricular septum can be used to place the sutures. This must be distinguished from the remnant of the interventricular membranous septum that may harbour the non-branching bundle.

Perimembranous Inlet Defects

Instead of extending towards the apex, these defects extend into the inlet septum. In addition to the central fibrous body, the contiguous septal leaflet of the tricuspid valve and the anterior leaflet of the mitral valve also form the fibrous rim of the defect. The medial papillary muscle is attached above the defect. Chordae supporting the septal leaflet and attached to small papillary muscles arising from the posterior division of the trabecula septomarginalis may cross the defect. As in complete atrio-ventricular defect, the apex of the triangle of Koch, and therefore the atrio-ventricular node, is deviated toward the coronary sinus (**Figure 26.10**). The penetrating and branching bundles are situated along the inlet margin (to the surgeon's right). Exposure of the VSD is also facilitated by placing two traction sutures of fine material on the septal leaflet of the tricuspid valve.

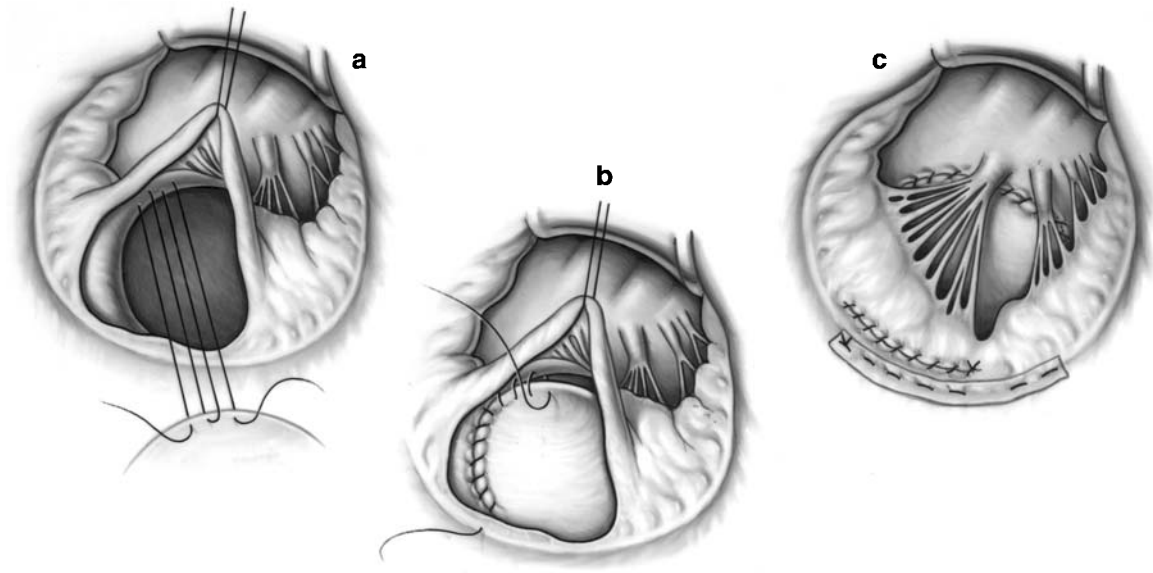

Figure 26.8

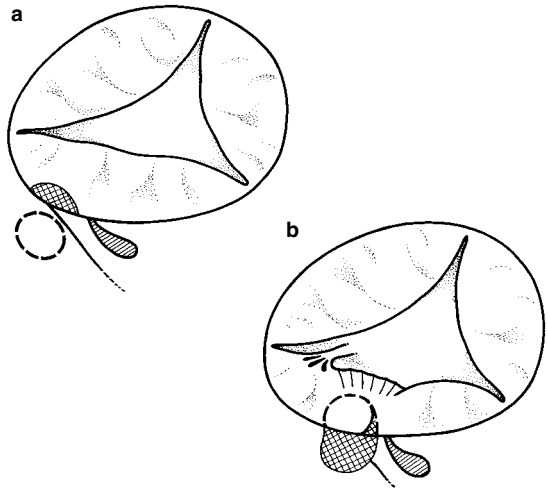

Figure 26.9

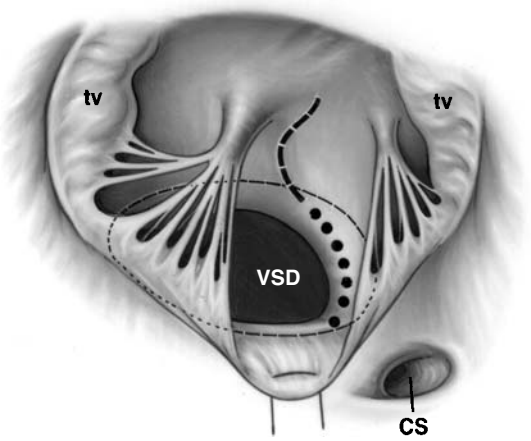

Figure 26.10

Continuous or interrupted sutures can be used to attach the prosthetic patch. Stitches are deeply placed through the cephalad margin of the defect from the area immediately above the medial papillary muscle to the central fibrous body. On the caudad margin, sutures are placed away from the rim to avoid the penetrating bundle and superficially to avoid the left bundle. Several stitches are also placed through the septal leaflet above the annulus of the tricuspid valve.

Perimembranous Infundibular Defects

The deficiency of the muscular septum is in the area of the infundibular septum, which is less well developed

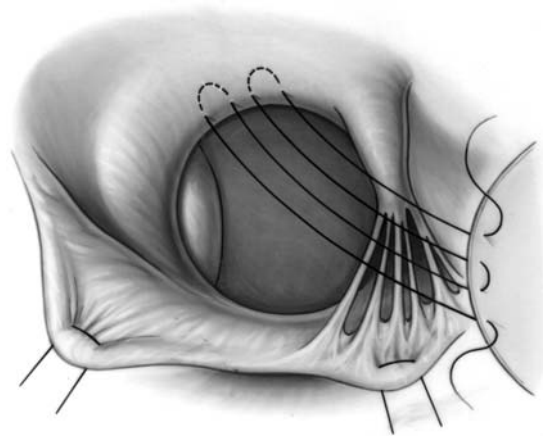

Figure 26.11

and is usually deviated to the right, so that the aortic valve frequently overrides the right ventricular cavity. This is the characteristic septal defect of tetralogy of Fallot, closure of which will also be described through a right ventriculotomy. Viewed from the right atrium, these defects are bordered by the central fibrous body, the adjacent portion of the anterior and septal leaflets of the tricuspid valve, the ventriculo-infundibular fold, the annulus of the overriding aortic valve, and the infundibular septum merging with the trabecula septomarginalis. The medial papillary muscle is below the defect (**Figure 26.11**). The exposure is facilitated by placing traction sutures on the anterior and septal leaflets of the tricuspid valve.

Attachment of the patch is similar to that described above. In some patients, the ventriculo-infundibular fold is poorly developed and there is an area of continuity between the aortic and the tricuspid valve that may be difficult to expose from a right atriotomy approach. In these cases, we have occasionally found it useful to close the defect through an incision in the anterior and septal leaflet of the tricuspid valve (**Figure 26.12**). Excessive traction of the tricuspid valve for exposure of high infundibular VSD may predispose to the development of junctional ectopic tachycardia (JET).

Trabecular Muscular Septal Defects

Some of these defects can be closed through the right atrium, when positioned to either side of the body of the trabecula septomarginalis. The main problem in the repair of these VSDs is the difficulty identifying the true margins among the trabeculations of the right ventricular surface of the septum. Often, trabeculae must be divided to expose the margins.

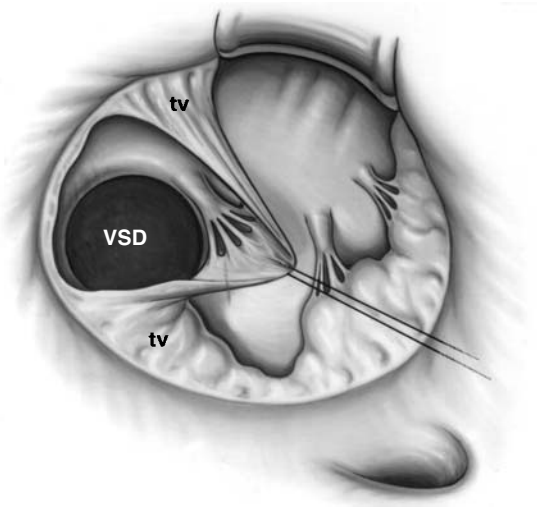

Figure 26.12

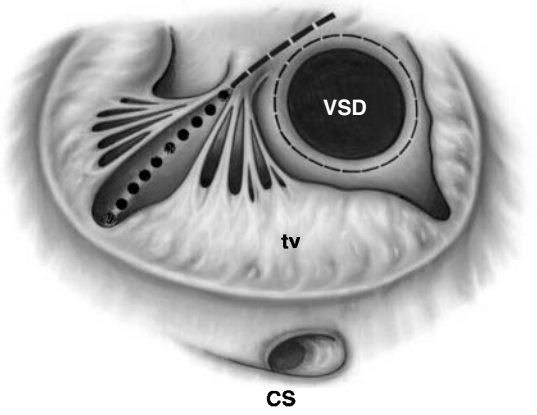

Figure 26.13

Large defects are closed with a patch using interrupted or continuous sutures (**Figure 26.13**). These VSDs have no direct relationship to the penetrating or branching segments of the conduction tissue but may be related to terminal bundle branches. The stitches are placed along the margin of the defect on the right side of the septum. Small defects are closed with interrupted buttressed sutures.

Multiple muscular VSDs can be closed individually, but a single patch to cover multiple defects has also been described (Seddio *et al*., 1999). The patch is sewn in with continuous 4-0 or 5-0 Prolene with a larger than usual needle to bulk up the thick trabecular muscle. Additional interrupted pledget-supported sutures are used to reinforce the suture line as necessary.

Identification and closure of multiple VSDs can be challenging. A novel approach was recently described by Brizard *et al*. (2004) using epicardial echocardiography on the beating heart to locate the defects which were then transfixed with a guide wire through the right ventricular free wall. On cardiopulmonary bypass and with a cardiopleged heart the VSDs were subsequently approached through the right atrium and closed using an oversized custom-made double patch that sandwiched the ventricular septum.

Inlet Muscular Ventricular Septal Defects

Compared with the perimembranous defects, these VSDs have a rim of muscular tissue, and the atrio-ventricular conduction axis penetrates into the ventricles on the outlet aspect of the defect (to the left hand of the surgeon). In this area, the stitches are placed superficially on the right ventricular side of the rim of the defect.

Infundibular Muscular Defects

When bounded by entirely muscular rims, infundibular defects are easily exposed through a right ventriculotomy incision but can sometimes be closed through the right atrium.

Association of Perimembranous and Inlet Muscular Defects

In this association, the penetrating and branching bundles are in their usual position in relation to the perimembranous VSD. The bridge of muscular septum that separates the two defects and contains the conduction tissue can be so narrow that there would be a serious risk of heart block should the defects be closed with two separate patches. A large patch covering the two defects and inserted along the line indicated in **Figure 26.14** constitutes the safest option for those defects (Bharati *et al*., 1983).

Malalignment Defects

These defects are characterized by a deviation of the outlet septum in relation to its adjoining inlet and trabecular parts. An anterior deviation is commonly seen in tetralogy of Fallot and contributes to the right ventricular outflow tract obstruction (**Figure 26.15a**), whereas a posterior deviation is often associated with aortic arch malformation and may result in subaortic stenosis (**Figure 26.15b**). A resection of the septal and parietal extensions of the infundibular septum is part

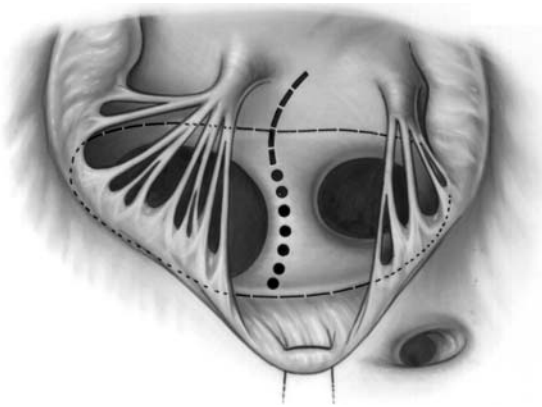

Figure 26.14

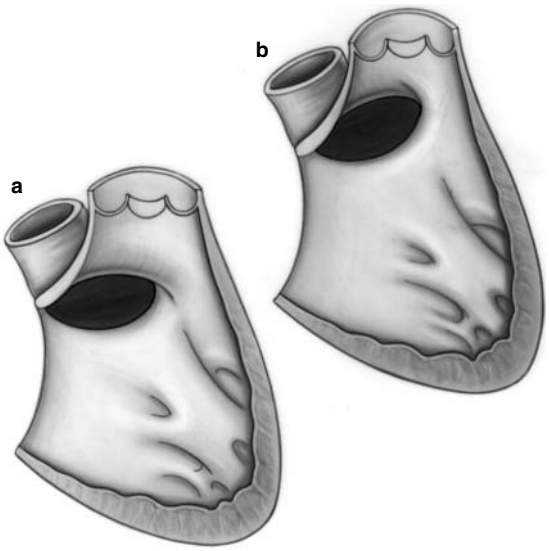

Figure 26.15

of the repair of tetralogy of Fallot, as indicated in Chapter 29. This can be performed through the right atrium and/or right ventricle.

In cases with significant posterior deviation of the outlet septum, resection of the infundibular septum along the superior margin of the VSD, up to the level of the aortic annulus, through the right atrium, has been suggested (Bove *et al*., 1993). The resulting enlarged VSD is then closed with a patch that widens the subaortic area.

Closure through the Right Ventricle

The right ventricular approach can be used for the perimembranous infundibular, infundibular and some trabecular defects.

The right ventricle is entered through a transverse incision when there is no need for patch enlargement of the RV outflow tract and when the repair of associated anomalies does not include insertion of an external cardiac conduit. In those cases, a vertical incision is preferred. Two traction stitches, taking the full thickness of the right ventricular wall, are placed on the infundibulum in an area free of major coronary arteries, remaining at least 5–10 mm away from the left anterior descending coronary artery. The incision is then made between these stitches. Two small cat's paw retractors are placed on each side of the ventriculotomy, and an eyelid retractor is placed in the apical part of it. Traction sutures placed on the ventriculotomy and tied on the skin edge may also be used for the exposure. The position and size of the VSD is then assessed. For perimembranous and low infundibular defects, exposure of the lower margin is greatly facilitated by a small retractor placed into the defect and pulled on the infundibular septum and its septal extension.

Perimembranous Defects

The most common type of perimembranous VSD closed through the right ventricle is the perimembranous infundibular defect seen in tetralogy of Fallot. **Figure 26.16** is a conceptual visualization of such a VSD. The central fibrous body, the anterior and septal leaflets of the tricuspid valvé and the aortic valve annulus

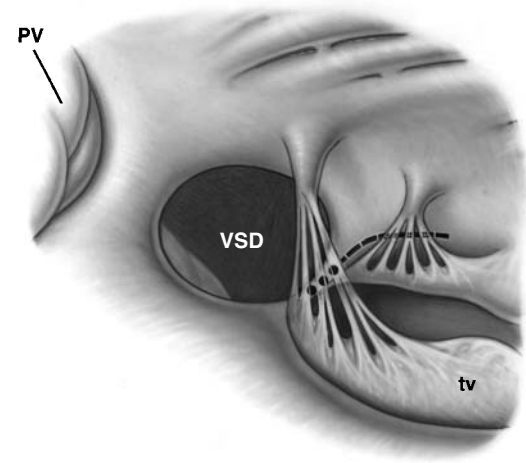

Figure 26.16

can all be part of the fibrous component of the margin of the defect. The penetrating bundle perforates the central fibrous body and comes close to the defect, particularly in hearts with aortic–tricuspid fibrous continuity. Then the branching bundle runs, in the majority of cases, below the inferior rim of the defect on the left septal side. In other hearts, however, the branching is more closely related to the margin of the VSD.

A retractor is placed at the bottom of the ventriculotomy with its tip through the tricuspid valve, retracting the posterior leaflet caudally. Two traction stitches of fine suture are placed on the anterior and septal leaflets of the tricuspid valve. A patch of Dacron velour, Gore-Tex, or glutaraldehyde-preserved xenograft pericardium is then appropriately tailored and inserted, using interrupted or continuous sutures.

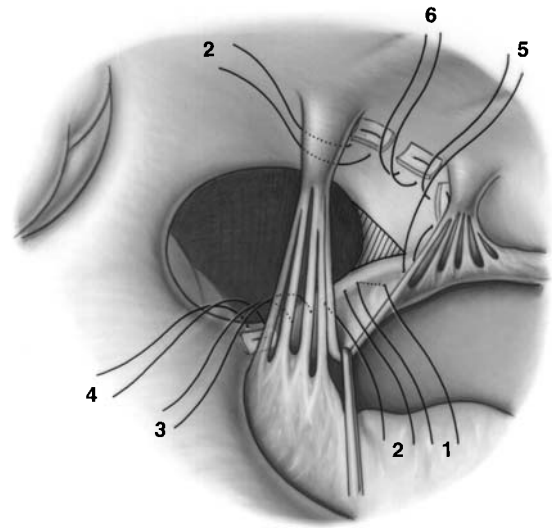

Figure 26.17

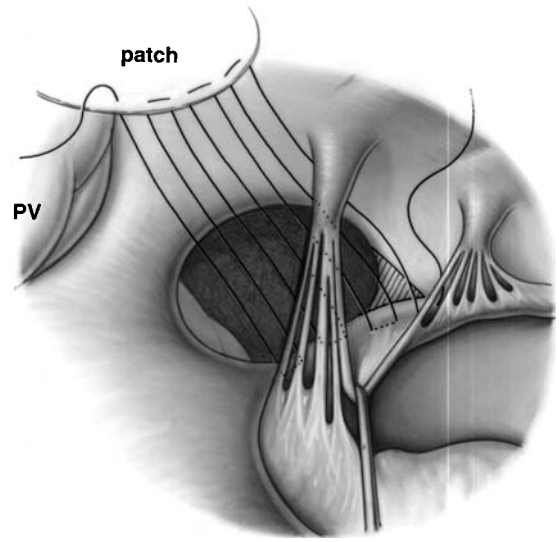

Figure 26.18

Patch Closure with Interrupted Sutures

Similar to the approach via the right atrium, pledget-supported interrupted mattress sutures can be used to repair the defect. All the sutures are placed around the defect before they are passed through an appropriately tailored patch. The sutures are placed superficially and away from the margin of the defect, postero-inferiorly from the junction between the septal leaflet and the ventricular septum and the medial papillary muscle. A transitional stitch is sometimes used to bridge the area between the septum and the septal leaflet. One arm of the transitional suture is placed superficially on the ventricular septum, away from its margin, and then through the tricuspid leaflet, entering the right atrium and then back into the ventricle. The second arm of the suture is passed through the ventricular septum (**Figure 26.17**). An alternative technique, using a combination of mattress and simple interrupted sutures, was described by McGoon (1968).

Patch Closure with Continuous Sutures

Exposure of the defect is identical to that described above. The patch is clipped on the left upper corner of the sternal incision. The first three or four mattress stitches of a double-armed half-circle needle, using monofilament polypropylene suture, are placed on the patch and along the margin of the defect. We routinely start at the junction of the posterior limb of the trabecula septomarginalis and the base of the septal leaflet of the tricuspid valve. The suture line is then carried along the base of the tricuspid valve beyond the central fibrous body, so that the stitching in the area of the conducting tissue is completed before lowering the patch into position (**Figure 26.18**). The suture line has to be carried beneath the chordae that are frequently attached to the tricuspid leaflet. The remainder of the closure is then carried out, using a mattress or running over-and-over suture. The closure may be reinforced with a few interrupted buttressed sutures in the areas where the muscle is particularly friable.

Combination of Interrupted and Continuous Sutures

We often use three or four interrupted mattress sutures, reinforced with Teflon pledgets posteriorly, to anchor the patch on the tricuspid valve securely and in the area

near the conduction system. A continuous suture is then used for the rest of the circumference of the defect.

Trabecular Muscular Defects

When located in the anterior part of the trabecular ventricular septum, VSDs can be repaired by an external buttress technique (Breckenridge *et al*., 1972). The position of the defect or defects is assessed through the right atrium. A vertical right ventriculotomy incision is then made opposite to the VSDs, at least 5 mm away from the left anterior descending coronary artery. Two strips of Teflon or Dacron felt are then tightly approximated with several mattress sutures passed from below the defects through the anterior wall of the right ventricle and tied outside the heart (**Figure 26.19**).

Infundibular Muscular Defects

When bounded by entirely muscular rims (**Figure 26.20**), these VSDs are closed with a patch sewn on the free margin of the defect, which is separated inferiorly and posteriorly from the conduction tissue by the fusion of the posterior limb of the trabecula septomarginalis with the ventriculo-infundibular fold. This type of VSD is encountered in approximately 15–20% of cases of tetralogy of Fallot.

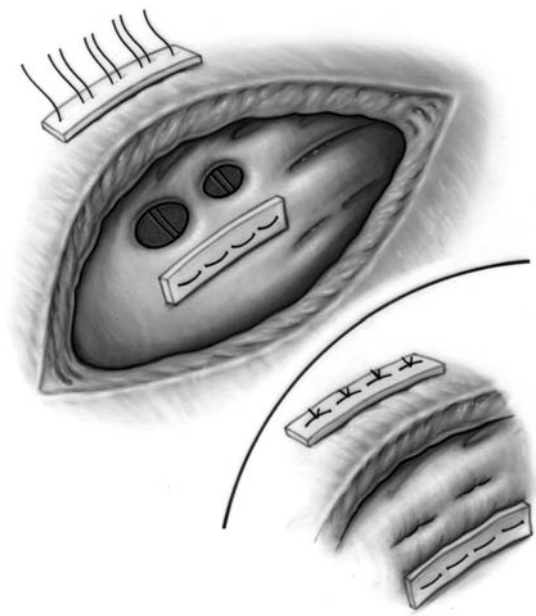

Figure 26.19

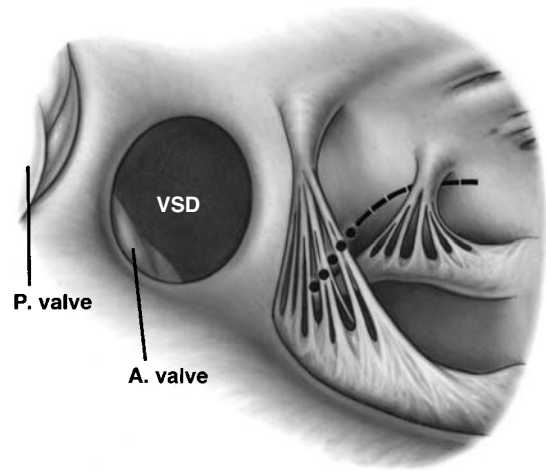

Figure 26.20

Closure through a Great Artery

Doubly-committed Subarterial Ventricular Septal Defects

The approach to the VSD depends on its exact location and the presence of other anomalies. Isolated defects are closed through a great artery. In the small child or in the infant, we prefer the trans-pulmonary approach. Doubly-committed subarterial VSDs have been closed through the aorta in older children with aortic valve incompetence, particularly if concomitant aortic valve repair or replacement is required. In the presence of subvalvar pulmonary stenosis, an additional right atriotomy may be required for infundibular muscle resection or the entire operation can be performed through a right ventriculotomy.

The operation is performed on hypothermic cardiopulmonary bypass. The left ventricle is vented via the right superior pulmonary vein. The cardioplegic solution is infused into the aortic root. If the aortic incompetence is more than moderate and the cardioplegic solution returns through the left heart vent, an oblique aortotomy incision is made towards the non-coronary sinus and the coronary artery ostia are directly perfused with cardioplegic solution.

The main challenge in repairing this type of VSD is the accurate attachment of the patch between the arterial valves. The main pulmonary artery is opened transversely above the commissures. A small eyelid retractor is placed through the pulmonary valve so as to expose the VSD. A large doubly-committed subarterial defect is illustrated in **Figure 26.21**. Occasionally, there is a fibrous ridge separating the arterial valves. Most commonly, however, there is direct continuity

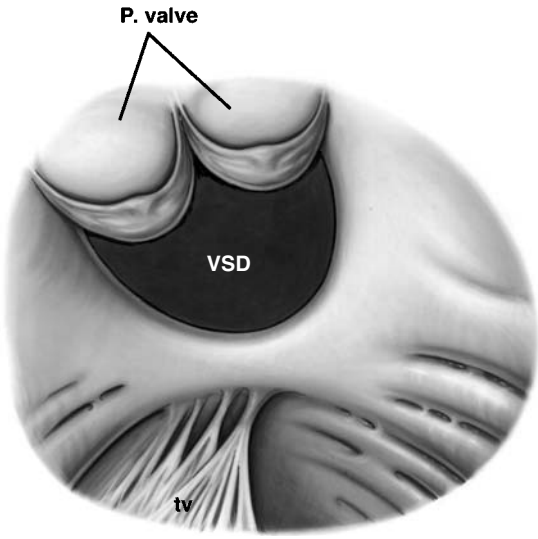

Figure 26.21

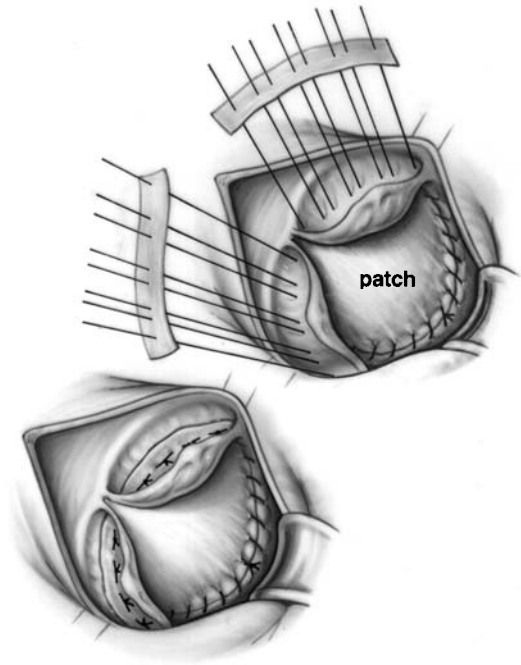

Figure 26.22

between the two arterial valves, and the patch must be anchored superiorly to the base of the posterior cusps of the pulmonary valve. This can be done with interrupted mattress sutures reinforced with small Teflon or autologous pericardial pledgets carefully placed from within the pulmonary sinuses. The rest of the defect can be closed with either interrupted or continuous sutures (Kawashima *et al*., 1977). Inferiorly, medially and laterally, the patch is anchored to the free muscular margin of the defect with a running over-and-over stitch. Two strips of autologous pericardium are used to reinforce the closure of the defect along the base of the posterior leaflets of the pulmonary valve, through which sutures are sometimes placed (**Figure 26.22**). We have used a Dacron or a Gore-Tex patch to close these defects to support the prolapsing aortic valve.

In the presence of aortic incompetence, the aortic valve may also require repair. Over the last decades a number of surgical techniques have been described. Trans-oesophageal echo, after induction of anaesthesia, is helpful in identifying the anatomy of the VSD, the mechanism of aortic incompetence and degree of regurgitation, and to check the repair after completion. Following exposure of the valve during the operation, an evaluation of the degree of incompetence and repair of the valve is facilitated by temporarily suturing together the midpoints (nodules of Arantius) of the three aortic cusps with a fine suture (Frater, 1967). The prolapsed leaflet, usually the right, is then inspected, and the length of the free margin at each commissure is measured. The

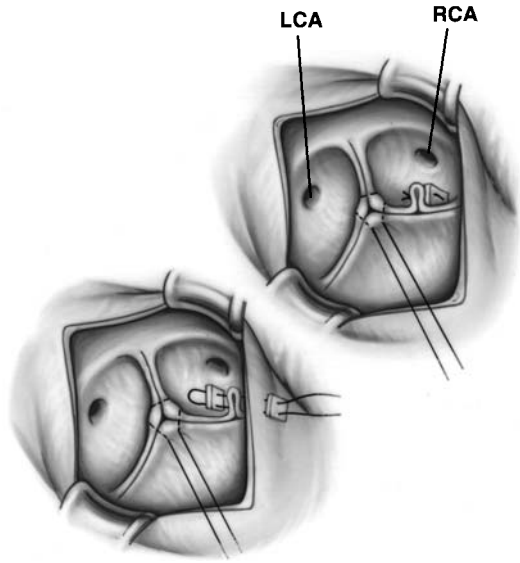

Figure 26.23

elongated commissures are then plicated either along the free margin of the leaflet (Spencer *et al*., 1973) or on the aortic wall (Trusler *et al*., 1973) (**Figure 26.23**). In

the latter technique excess leaflet tissue, folded at the commissure, is secured to the aortic wall with two or three mattress sutures. These sutures are reinforced with pericardial or Teflon pledgets, one within the aortic sinus and one outside the aortic wall. Trusler *et al*. (1973) also suggested that the commissure should be reinforced with a tiny patch held with a mattress suture to secure the new commissure to the aortic wall.

Carpentier (1983) described patch closure of the VSD and repair of the aortic valve by triangular resection of the redundant segment of the prolapsing leaflet **Figure 26.24**. In the presence of annular dilatation, he also advocated circular annuloplasty of the left ventriculo-arterial junction as well as thinning of thickened leaflets to relieve restricted leaflet motion. Yacoub and colleagues (1997) proposed a technique that aims to correct the basic morphologic abnormality of this lesion, which results from a ''discontinuity'' of the aortic media from the aortic annulus and ventricular septum in the region of the right coronary sinus, rather than a true defect in the ventricular septum. This leads to progressive dilatation of the unsupported sinus and downward and outward displacement of the aortic valve. Repair is achieved by elevating the crest of the ventricular septum to the aortic media and plicating the unsupported sinus of Valsalva. Working through the aortic valve, the extent of the dilatation of the right coronary sinus and the thin area of the sinus resulting from the discontinuity between the aortic valve annulus and the media of the aorta are accurately defined. A series of interrupted vertical 4-0 Ethibond (Ethicon Inc., Somerville, NJ) mattress sutures supported by small pericardial pledgets are then inserted slightly on the right side of the crest of the septum (to avoid the conduction tissue that runs on the left ventricular side 4–5 mm below the edge of the crest) (**Figure 26.25a**). The sutures are then passed through the annulus of the aortic valve, and are used to plicate the thin portion of the sinus of Valsalva until the normal aortic tissue supported by the media is reached. Tying the sutures results in closing the VSD, elevating the right coronary annulus and cusp, and reducing the size of the right coronary sinus and right ventricular outflow tract. (**Figure 26.25b**).

If the aortic valve cannot be repaired, the leaflets are excised and the valve replaced with a prosthetic valve. An alternative would be to use a free aortic homograft or an aortic root replacement in smaller children. If the aortic valve has to be replaced, the VSD is always closed first because it may be necessary to place sutures through the upper margin of the VSD patch to help anchor the valve prosthesis in the area between the right and non-coronary cusps.

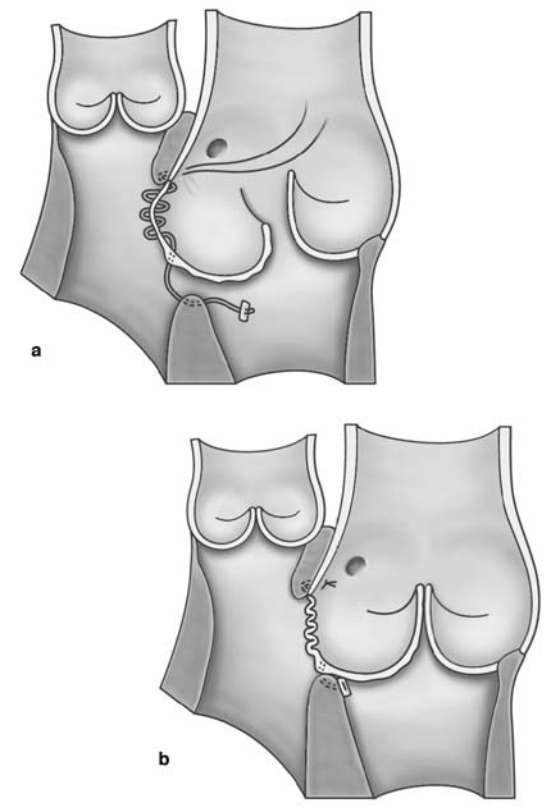

Figure 26.25

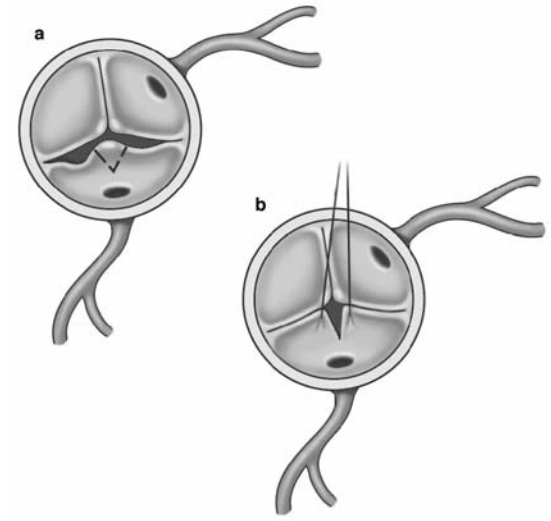

Figure 26.24

Closure through the Apex

Exposure and closure of muscular defects in the lower part of the trabecular septum may be easier when approached from the left ventricle. It is also easier to identify a single left ventricular opening as opposed to multiple openings into the right ventricle. However, the debate continues with regard to the potential negative sequels. Observations of late myocardial dysfunction, formation of apical aneurysms and arrhythmogenesis have been reported (Kitagawa *et al*. 1998).

Apical VSDs are often multiple and can be associated with higher defects that might need to be closed through the right atrium and/or through the right ventricle. The defects are first inspected through the tricuspid valve to confirm their position. A good relaxation of the heart, and in some cases the division of the trabecula septomarginalis, allows access to the lower part of the ventricular septum from a right atrial approach (Serraf *et al*., 1992). Apical right ventriculotomy has also been reported for closure of these defects, using a sandwich technique (Tsang *et al*. 2002).

For a left apical approach, a short fish-mouth incision is made at the apex away from and parallel to the anterior descending coronary artery. (**Figure 26.26**) Care must be taken to avoid damaging the anterior papillary muscle of the mitral valve. The margins of the defect are easily defined on the smooth left side of the ventricular septum. The defect is closed with a patch. If multiple defects are near to each other, a large patch covering all the defects can be inserted with a continuous suture, the higher left ventricular pressure keeping the patch against the septum (Singh *et al*., 1977). The ventriculotomy is closed with a double layer of continuous monofilament suture. Residual VSDs and left ventricular dysfunction are not uncommon following closure of lower VSDs through the left ventricle.

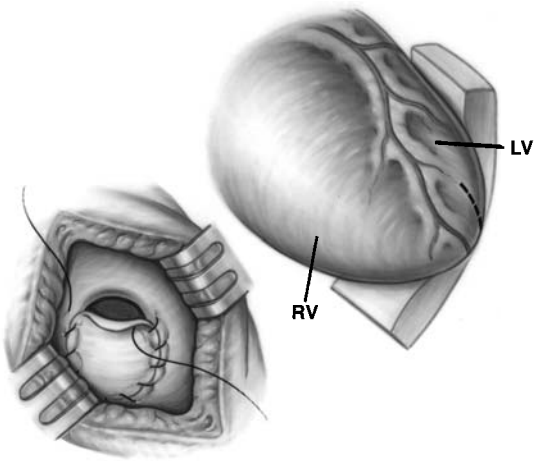

Figure 26.26

Ventricular Septal Defect and Aortic Arch Anomalies

Neonates and infants with severe coarctation of the aorta and large VSDs used to be subjected to a staged repair, consisting of a repair of the aortic arch with banding of the pulmonary artery and a subsequent debanding and closure of the VSD. Currently, many centres proceed with a one-stage repair, using techniques very similar to those described to repair interrupted aortic arches (see Chapter 21). The ascending aorta and its branches are fully mobilized. The innominate, left carotid and left subclavian arteries are encircled with tourniquet clamps. The descending thoracic aorta is dissected, starting along the right side of the horizontal aorta and going down by sharp and blunt dissection into the descending thoracic aorta. The ductus arteriosus is dissected and closed with a heavy silk suture. A single cannula is introduced into the ascending aorta, unless there is a near-atresia of the aortic arch, in which case a second cannula is introduced into the descending thoracic aorta, through the main pulmonary artery and the patent ductus arteriosus.

In the neonate, a single venous cannula is placed in the right atrium and the operation is performed under profound hypothermia and total circulatory arrest. The dissection of the ascending aorta is completed while the patient is being cooled. The tourniquets on the head vessels are snugged down and the circulation is arrested. The arterial cannula may or may not be removed, depending on the extension of the aortic arch hypoplasia. The ductus arteriosus is divided, and the ductal tissue is completely removed from the aorta. The descending thoracic aorta is brought superiorly. The inferior aspect of the horizontal aorta is widely incised if hypoplastic. An incision is made on the lateral aspect of the descending aorta so as to obtain an extensive aortoplasty. The suture line is done with 6-0 or 7-0 polypropylene. More recently, with increasing experience with the Norwood operation, techniques for aortic arch patch augmentation have been applied (see Chapter 20, Coarctation of the Aorta). The VSD is then closed through the right atrium or through the right ventricle, depending on its position. It is not uncommon for these VSDs to present with a posterior displacement of the infundibular septum, in which case it may be necessary to resect part of the infundibular septum to prevent subaortic obstruction.

If one is concerned about the length of the total circulatory arrest time, the operation can be performed with bicaval cannulation, so as to reinstitute cardiopulmonary bypass after the arch reconstruction, or alternatively be performed with selective cerebral perfusion throughout the arch reconstruction period, thus totally avoiding circulatory arrest.

Trans-catheter Device Closure of Ventricular Septal Defects

In selected cases, device closure of VSDs has been successfully performed. This has been particularly useful in apical muscular defects and residual VSDs (Lock *et al*., 1988). Potential for development of heart block in a perimembraneous VSD device closure remains a concern. The largest reported experience to date is a multicentre study of 170 patients, 168 of whom underwent successful device implantation for either congenital or postoperative residual VSDs (Knauth *et al*., 2004). Multiple devices were placed in 40% of patients. Device implantation resulted in a significant decrease in L–R shunting and improvement in VSD size/severity, and device position proved stable. There were 332 adverse events; 39 were related to the device, 261 to catheterization and all but five events occurred in the periprocedural period; 14 patients in this series died; 18 had the devices explanted. With increasing experience and improved equipment, the incidence of adverse events is expected to come down in the future. Devices have also been placed intraoperatively (Murzi *et al*., 1997). The technique continues to develop and may become the approach in the future (Bridges *et al*., 1991). Details are described in Chapter 7.

RESULTS

Residual VSD

Residual interventricular shunting may be related to undiagnosed defects, incomplete closure, or reopening of a defect. Intraoperative echocardiogram (trans-oesophageal or epicardial) is now performed routinely in many centres to check on the accuracy of the repair. If a residual VSD is found and thought to be haemodynamically important, a decision has to be made whether to go back on cardiopulmonary bypass to close the defect, band the pulmonary artery or proceed to early trans-catheter closure.

Conduction Disturbances

Conduction disturbances are frequent after repair of VSDs. Right bundle branch block is commonly seen following closure of a VSD, not only through the right ventricle but also through the right atrium. A small proportion of patients develop the electrocardiographic pattern of right bundle branch block and left anterior hemi-block (Rein *et al*., 1977). The incidence of surgically induced permanent heart block in isolated VSD is now approaching zero in most centres. In our retrospective study (1976–2001), permanent heart block was observed in seven (0.7%) of 996 patients operated for isolated VSD (Anderson *et al*., 2006). Despite such a low incidence of heart block, all patients have atrial and ventricular pacemaker wires inserted at the end of the procedure. If the sinus rhythm does not return within 2 weeks, we would currently insert a permanent pacing system.

Hospital Mortality

The hospital mortality rate for repair of isolated VSDs in infancy currently approaches zero in most specialized centres. Incremental risk factors for hospital death are multiple VSDs and major associated cardiac anomalies. In a large series published by Serraf *et al*. (1992), there was a hospital mortality rate of 7.7%. Half of the patients who did not survive had a Swiss-cheese type of defect. The actuarial survival rate at 7 years was 89.6%. Our current results for isolated VSD (1997–2001) show a mortality rate of 0.7% (2/263 patients). Recent results from the Central Cardiac Audit Database, a prospectively collected and validated dataset, showed an overall 30 day mortality of 1.4% (99% confidence limits, 0.6–3.5%) for the 698 patients that underwent VSD closure in the UK in the year April 2000–March 2001 (Gibbs *et al*., 2004). Young age (neonate) did not appear to be a risk factor; however, the number of operated neonates was very small.

LONG-TERM RESULTS

Subsequent to successful correction, the majority of patients live virtually normal lives and have normal exercise capacity (Meijboom *et al*., 1994). A study of 176 patients operated for isolated VSD in 1968–1980 (Roos-Hesselink *et al*., 2004) showed an incidence of reoperations of 6% and development of sick sinus syndrome in 4%. Aortic insufficiency was observed in 16% of operated patients.

A significant cause of morbidity and mortality is pulmonary vascular disease. Patients with high pulmonary vascular resistance operated on beyond infancy have a poor long-term prognosis because of progression of their pulmonary vascular disease, and it is not currently known whether new medical pharmacological treatments for pulmonary hypertension will change these results.

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27

Atrio-ventricular Septal Defects

A. D. Pacifico

Atrio-ventricular septal defects include a spectrum of cardiac malformations characterized by varying degrees of incomplete development of the inferior portion of the atrial septum, the inflow portion of the ventricular septum and the atrio-ventricular (A-V) valves. This group of defects can be subdivided into partial, intermediate and complete forms. In the *partial form* of the A-V septal defect, there is usually an ostium primum type of atrial septal defect occupying the portion of the atrial septum just superior to separately formed tricuspid and mitral valves. The defect may be of varying size but typically is large and haemodynamically unrestrictive. The mitral and tricuspid valves are displaced into the ventricles and are attached to the crest of the ventricular septum. The mitral valve in this defect was originally considered to be a bileaflet structure, similar to the normal mitral valve but with a cleft in the midportion of its anterior leaflet. Later, this mitral valve was described as a trileaflet structure (Carpentier, 1977), whose anatomy and mode of function are different from those of the normal mitral valve. In keeping with the nomenclature for leaflets in the entire spectrum of A-V septal defects, the three mitral leaflets are designated left lateral, left superior and left inferior. There are anterolateral, posterolateral and septal commissures; the septal commissure corresponds to the so-called ''cleft''. The lateral leaflet occupies only 25% of the annular circumference, and thus it is distinctly different from the corresponding posterior leaflet of the normal mitral valve, which occupies 65%. Similarly, the left superior and left inferior leaflets together occupy 75% of the annular circumference, and thus they are distinctly different from the anterior leaflet of the normal mitral valve, which occupies 35%. The anatomy of the septal commissure varies considerably; when very well developed, the commissure may almost completely separate the left superior and inferior leaflets. In this instance, the leaflet edges may be thickened and curled, which can lead to incompetence of the mitral valve at this site. For repair of the incompetent mitral valve in the partial AV-septal defect, however, it is important to consider all of its elements and not assume that simple suture closure of the septal commissure will result in a competent valve. The tricuspid valve also has three leaflets, which are designated right superior, right lateral and right inferior. Usually, no ventricular septal defect is present, although tiny defects in the attachment of the mitral valve to the crest of the ventricular septum may result in small interventricular communications. In some patients, a coexistent ventricular septal defect is found. In the partial form, the major element that differentiates it from the other forms of A-V septal defects is the presence of two separate A-V valves.

In the *complete form* of the A-V septal defect, the key element is the presence of a common (single) A-V valve that is common to the right and left A-V chambers. The central aspect of this orifice is usually contiguous, with an ostium primum type of atrial septal defect above and a defect in the ventricular septum below. Looking through the right atrium at the A-V valve, a surgeon can always identify a completely bare area of the crest of the ventricular septum. The common A-V valve has six leaflets: left superior and inferior, left and right lateral, and right superior and inferior. Neither the left superior and inferior leaflets nor the right superior and inferior leaflets are attached to each other. The left superior leaflet may cross the crest of the ventricular septum and lie partially on the right ventricular side. Rastelli *et al*. (1966) classified complete A-V septal defects into types A, B and C, which serve only to highlight the variable degree of such bridging by the left superior leaflet in a continuous spectrum of leaflet and chordal anatomy.

In type A, the left superior leaflet is entirely over the left ventricle and, together with the right superior leaflet, is attached by chordae tendineae to the crest of the ventricular septum.

In type C, there is marked rightward displacement of the left superior leaflet, which floats freely over the ventricular septal crest and is not attached to it by chordae tendineae. The size and prominence of the right superior leaflet is inversely related to the

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degree of bridging of the left superior leaflet. The anatomy of the left inferior leaflet often does not mirror the anatomy of the left superior leaflet, and the left inferior leaflet is usually variably attached to the crest of the ventricular septum directly or by chordae tendineae. The left and right superior and inferior leaflets usually have appropriate chordal connections laterally to papillary muscles within the left and right ventricles. An accessory orifice, whose own chordae are attached to its own papillary muscle, may exist at the left superolateral or inferolateral commissure and tends to reduce the size of the main orifice of the A-V valve. The size of the interventricular communication may vary considerably. In some patients it is only a small deficiency between the septal attachments of the left superior and inferior leaflets, but more commonly it extends completely beneath the left superior leaflet and sometimes completely beneath the left inferior leaflet as well. The position of the common A-V valve in relation to the plane of the ventricular septum is also variable. In the right dominant form of the complete A-V septal defect, the right ventricle is large, the left ventricle is smaller than normal and the position of the common A-V valve in relation to the septum is right-ward. In the left dominant form, the left ventricle is larger than normal, the right ventricle is smaller than normal, and the position of the valve is left-ward. These forms are less common than the balanced type, in which there is bilateral enlargement of each ventricle and a centrally positioned common A-V valve.

The *intermediate form* of the common A-V septal defect lies between the partial and complete forms. These hearts have an ostium primum type of atrial septal defect, with a fibrous connection between the left superior and inferior leaflets over the ventricular septum. This fibrous tissue connection somewhat obliterates the bare area of ventricular septum, which is always found in the complete form but is not sufficient for the mitral and tricuspid portions of the valve to be considered separately formed entities. The intermediate form is sometimes amenable to the surgical repair used for the partial A-V septal defect, but more often must be converted at operation to the complete form and repaired as such. A-V septal defects may coexist with major associated cardiac anomalies, including: tetralogy of Fallot; double-outlet right ventricle; transposition of the great arteries; total anomalous pulmonary venous connection; and valvar, subvalvar and supravalvar aortic or pulmonary stenosis. It may also be found in hearts with A-V discordance. Atrial isomerism (situs ambiguus) is often accompanied by complete A-V septal defects, which may be associated with partial and total anomalous pulmonary and systemic venous connection. Bilateral superior venae cavae are often found in patients with complete A-V septal defects, with or without common atrium. When the atrial septum is developed, the left superior vena cava usually drains to the coronary sinus, but sometimes drains to the upper left corner of the left atrium (unroofed coronary sinus). When a common atrium is present, the left-sided superior vena cava usually drains to the left upper corner of the atrium, the coronary sinus is unroofed, and there may be interruption of the inferior vena cava, with hemi-azygos continuation to the left superior vena cava and separate entrance of hepatic veins into the atrium.

DIAGNOSIS

Clinical manifestations of A-V septal defects depend on the magnitude and direction of the shunt. This in turn is determined by the size of the interatrial or interventricular defect or both, the magnitude of A-V valve incompetence, and the relative preload and afterload of each ventricle. Patients with small or moderately increased pulmonary:systemic blood flow ratios (QP:QS < 3) are often asymptomatic, whereas those with large left-to-right shunts (QP:QS > 3) may exhibit fatigue, dyspnea or congestive heart failure. Those with elevated pulmonary vascular resistance may have no symptoms, but when elevation is severe, cyanosis may develop as a result of right-to-left shunting. Clinical manifestations, physical findings, conventional radiographic features and haemodynamic data are similar to those of patients with isolated atrial or ventricular septal defects. The electrocardiogram may be helpful in the diagnosis of these malformations because 90% contain a counter-clockwise frontal plane loop, with the initial vector directed inferiorly and to the right. Definitive diagnosis is made by real-time echocardiography and by colour Doppler echocardiography. Cardiac catheterization and angiocardiography is required in special situations only: the main reason is assessment of severely elevated pulmonary vascular resistance. The anteroposterior view of the left ventriculogram shows the typical ''gooseneck'' deformity, which results from a narrow and elongated outflow tract. More precise information can be obtained from axial cineangiographic projections (Bargeron *et al*., 1977; Elliot *et al*., 1977; Soto *et al*., 1981) and from oblique projections (Brandt *et al*., 1972; Macartney *et al*., 1979).

PARTIAL FORM

Indications

A few patients with the partial A-V septal defect have severe mitral valve incompetence and usually exhibit significant symptoms of congestive heart failure or growth failure during the first year of life. A small percentage exhibit severe symptoms in infancy and may

require urgent operation. Pulmonary vascular disease is uncommon in this malformation. The majority of patients with mild to moderate mitral valve incompetence have more severe symptoms at an earlier age than do patients with secundum atrial septal defects. Congestive heart failure occurs in about 20% but may not be present until the fourth decade of life. Precise natural history data obtained prospectively are not currently available, but for the majority of patients, the natural history is probably similar to that of patients with ostium secundum defects. Operation is advised electively for most patients during the first year of life. Only a small number of patients at this age have severe mitral incompetence and usually this can be improved by repair.

Surgical Technique

A median sternotomy incision is made, preserving the left side of the pericardium, which can be left in place for later excision as a patch to be used for closure of the ostium primum defect. Alternatively, it can be exised at this time, cleansed and placed on a trimmed towel soaked in saline. It remains adherent to the towel, which improves its handling quality, and later, after both are simultaneously trimmed to the final shape, suturing is begun by peeling the working edge of pericardium away from the towel.

The heart is externally inspected in order to assess chamber size, anomalies of systemic or pulmonary venous connection and patency of the ductus arteriosus. Cardiopulmonary bypass is established with ascending aortic perfusion, and direct caval cannulation is accomplished with the use of thin-walled, angled metal cannulae (Medtronic Inc., Minneapolis, MN). The details of perfusion have been described previously (Pacifico, 1989). When a left-sided superior vena cava is present, it may be directly cannulated (after partial bypass is established), or its drainage into the coronary sinus can be collected with a sump sucker. Perfusate temperature is adjusted to 34°C. Cold cardioplegic solution is injected into the ascending aorta after cross-clamping. Carbon dioxide field flooding is employed throughout.

The right atrium is opened longitudinally from the midportion of the right atrial appendage towards the inferior vena cava cannula (**Figure 27.1**). Four stay sutures are placed to secure the edges of the atriotomy to the subcutaneous tissue or pericardial edges. A sump tip sucker (Medtronic Inc.) is placed through the foramen ovale, atrial septal defect or, if this defect is not present, through a stab wound deliberately made in this location. The anatomy is best inspected when the aorta is cross-clamped and residual blood is aspirated from the cardiac chambers.

Cold saline is injected into the left ventricle, causing the mitral valve to float to its closed position, so that

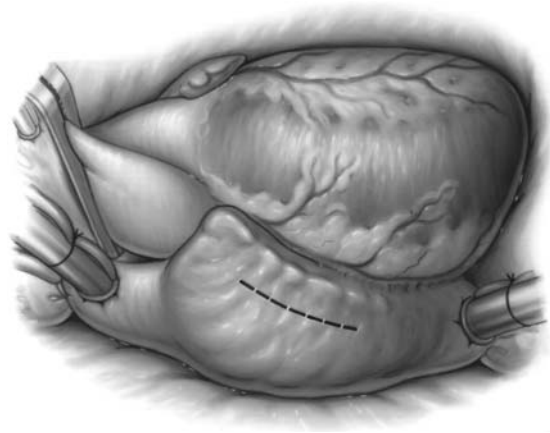


Figure 27.1

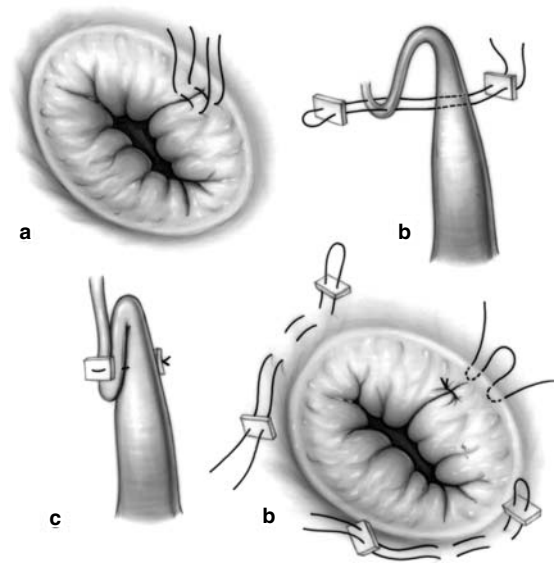


Figure 27.2

valve architecture, the extent of the septal commissure and the location and presence of incompetence and jet lesions can be assessed. The septal commissure is closed with interrupted 5-0 or 6-0 Prolene sutures. The initial stitch is first placed to approximate the left superior and inferior leaflets nearest their septal attachment. This procedure serves as a commissuroplasty, bringing the leaflets closer together, and additional interrupted sutures are placed to approximate the touching edges of the leaflets (**Figure 27.2a**). Saline is reinjected into the ventricle to verify valve competency. Care is exercised

to avoid excessive closure of the commissure, which can further distort the leaflets and possibly create a stenotic orifice. The orifice is sized with Hegar dilators in relation to the mean normal value **(Table 27.1).** An orifice of one standard deviation smaller than the mean normal value is acceptable.

In addition to an incompetent septal commissure, the pathological features of the mitral valve in this malformation may include incompetence of the superolateral or inferolateral commissures. There may be elongation of the chordae tendineae with leaflet prolapse, or annular dilatation with resultant central incompetence. Saline is again injected into the left ventricle in order to reveal these additional but less frequently found abnormalities. Should prolapse of the leaflet because of elongated chordae be present and be responsible for persistent incompetence, shortening of involved chordae is accomplished by the technique shown in **Figure 27.2b,c**. Central incompetence from annular dilatation is repaired by a local annuloplasty at the superolateral and inferolateral commissures, as depicted in **Figure 27.2d**. Incompetence localized to either the superolateral or inferolateral commissure is repaired by a single commissuroplasty. Final valve architecture and competency are reviewed after saline is injected into the left ventricle.

The patch of pericardium is trimmed to size, oriented with its smooth surface leftward, and stitched to close the interatrial defect with continuous double-armed 4-0 (5-0 in infants) Prolene sutures. The suture line is on the base of the left superior and inferior leaflets; it begins at the inferior aspect of the left superior leaflet adjacent to the septal commissure and continues anteriorly to the junction of the atrial septum with the A-V valve annulus. A rubber-shod clamp is placed on this suture, and the other end used to attach the patch to the base of the left inferior leaflet (**Figure 27.3**). The conduction tissue lies in the atrial wall just above the crux cordis. It is in the area between the coronary sinus and the junction of the left and right inferior leaflets; it extends on the crest of the septum and divides into bundle branches before reaching the septal commissure (see Chapter 8). Near the posterior junction of the left inferior leaflet and A-V valve annulus, the suture line continues anteriorly on the base of the right inferior leaflet for about 1 cm and then anteriorly around the coronary sinus, leaving it and the conduction tissue left-ward.

Closure is completed by suturing the patch to the remaining free edge of the ostium primum type defect (**Figure 27.4**). When the coronary sinus receives a left-sided superior vena cava, it must remain on the right side of the patch. The path of the suture line is thus altered to include the left-ward margin of the coronary sinus. The

Table 27.1 Mean normal diameter of atrio-ventricular valves.

BSA (m)	Mitral* (mm)	Tricuspid* (mm)
0.25	11.2	13.4
0.3	12.6	14.9
0.35	13.6	16.2
0.4	14.4	17.3
0.45	15.2	18.2
0.5	15.8	19.2
0.6	16.9	20.7
0.7	17.9	21.9
0.8	18.8	23.0
0.9	19.7	24.0
1.0	20.2	24.9
1.2	21.4	26.2
1.4	22.3	27.7
1.6	23.1	28.9
1.8	23.8	29.1
2.0	24.2	20.0

BSA, body surface area.

* The approximate standard deviations are 0.3 m = 1.9 mm and 0.3 m = 1.6 mm for the mitral valve and 1.0 m = 1.7 mm and 1.0 m = 1.5 mm for the tricuspid valve.

Modified from Rowlatt UF, Rimoldi HJA, Lev M. The quantitative anatomy of the normal child's heart. *Pediatr Clin North Am* 1963; **10**: 499.

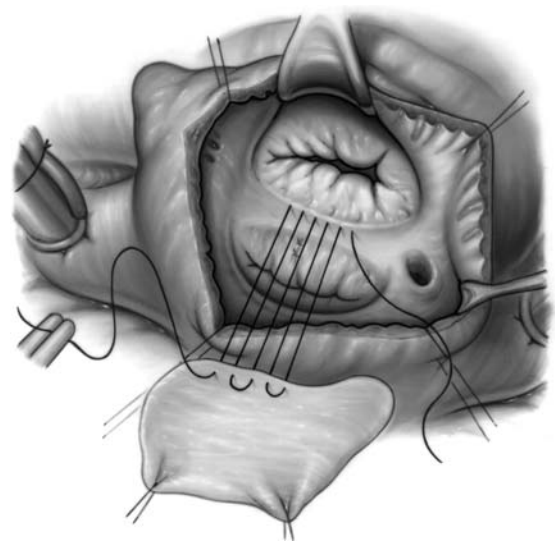


Figure 27.3

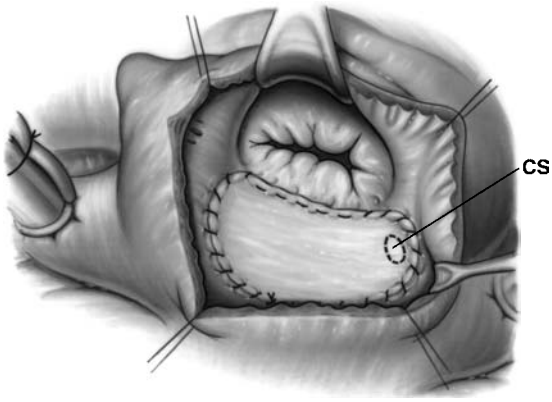


Figure 27.4

foramen ovale, if patent or incised for venting, is closed directly with a separate Prolene suture; air is evacuated from the left atrium by inflating the lungs before completion of closure. Rewarming is commenced, the aortic cross-clamp is released while air is aspirated from the aorta, the right atriotomy is closed, and the usual debubbling technique is employed, with or without intraoperative trans-oesophageal guidance. Bypass is discontinued when nasopharyngeal temperature reaches 37°C.

Results

The combined surgical experiences reported by Rastelli *et al*. (1965), McMullan *et al*. (1973), Levy *et al*. (1974), and Losay *et al*. (1978) yielded information on 424 patients undergoing repair of the partial atrio-ventricular septal defect. Hospital mortality rates in these series for elective operation were less than 6%, and low cardiac output was the most common cause of death. The data suggest that patients requiring operation in infancy and those with enlarged hearts had a higher operative risk. Among patients with severe preoperative mitral valve incompetence, the hospital mortality rate was higher and mitral valve replacement was more frequently required.

Studer *et al*. (1982) analysed the University of Alabama at Birmingham experience during 1967–1982, with 310 consecutive patients undergoing repair of A-V septal defects; 154 patients did not have an interventricular communication, and the hospital mortality rates were 3.6% among the 140 patients without major associated anomalies and 14.3% among the 14 patients with them. Significant incremental risk factors for hospital death identified by multivariate analysis were earlier date of operation, increasing severity of preoperative A-V valve incompetence, increasing level of preoperative disability, and the presence of an accessory valve orifice. Younger age at operation was an incremental risk during the earlier years of this experience, but this effect had been neutralized by 1977.

King *et al*. (1986) studied the Mayo Clinic experience during January 1962–January 1984; 199 patients, age range 5 months–71 years (mean, 11.2 years), underwent correction of partial atrio-ventricular canal defects. The overall 30 day mortality rate was 5.5%, and significant determinants for these deaths were congestive heart failure, cyanosis, failure to thrive, age of less than 4 years and moderate to severe mitral valve incompetence ($p < 0.01$). El-Najdawi *et al*. (2000) reviewed the Mayo Clinic experience with 334 patients undergoing repair of partial A-V septal defects prior to 1995 and principally focused on the long-term results. The 30 day survival rate was 98%.

Complications and Late Results

Surgical heart block complicates repair of partial atrio-ventricular defect in fewer than 1% of patients. Losay and associates (1978) reported atrial dysrhythmias occurring in 18 of 83 patients (22%) in the early postoperative period and in 13 of 75 patients (17%) in whom late postoperative follow-up information was available. McMullan *et al*. (1973) showed an actuarial late survival rate over a 20 year period of 94%, the majority of patients being asymptomatic. Secondary mitral valve surgery was advised late in the postoperative period for about 5% of patients.

In the Mayo Clinic experience reported by El-Najdawi *et al*. (2000), actuarial late survival rates for 334 patients operated prior to 1995 were 93% at 10 years, 87% at 20 years and 76% at 40 years. These were significantly different from 97%, 94% and 92% at similar intervals for an age- and sex-matched control population ($p < 0.001$).

Postoperative left A-V valve regurgitation was documented in 230 patients (69%). It was severe in 19 (8%), moderately severe in 2 (1%) and moderate in 21(9%). Reoperation was performed in 38 patients (11%) and the estimated incidence was 3% at 1 year, 6% at 5 years, 8% at 10 years, 12% at 20 years, 17% at 30 years and 22% at 40 years. The commonest reason for reoperation was left A-V valve regurgitation, subaortic stenosis and left A-V valve stenosis. Postoperative supraventricular arrythmias were more common in older patients.

COMPLETE FORM

Indications

Tandon *et al*. (1974) reviewed an autopsy series of 91 patients with the complete A-V septal defect and found

that 65% had died within the first year of life. Although information obtained from autopsy data may not precisely parallel prospectively obtained data, it is believed that if the latter were available, it would probably be similar to Tandon *et al*.'s age-related death rates. Newfeld *et al*. (1977) studied the incidence and time course of pulmonary vascular disease in 67 children with the complete form of A-V septal defect. They found that advanced pulmonary vascular disease began to develop during the first year of life and was commonly found after 2 years of age. The rate of progression of pulmonary vascular disease was no different from that found in a group of 40 children with isolated large ventricular septal defects, similarly studied. Newfeld *et al*. (1977) concluded that surgery must be performed during the first year of life in order to alter the natural progression of pulmonary vascular disease. Blackstone *et al*. (1976) reached a similar conclusion for patients with isolated large ventricular septal defects. Berger *et al*. (1979) presented data to define survival and probability of cure with and without operation for complete A-V septal defects. These natural history data were derived from published autopsy studies and analysed by the parametric survival distribution method, confirming that 65% of untreated patients die by the age of 12 months, 85% by 24 months and 96% by 5 years. They reviewed the rates of early and late survival after repair of complete A-V septal defects at the University of Alabama at Birmingham (to be described) and assumed that the probability of having no major pulmonary hypertension 5 or more years after repair, as a function of age at operation, was similar in this group to that in patients with isolated ventricular septal defect. These data were combined and used to relate parametrically the probability of surgical ''cure'' (alive 5 years after operation and with a mean pulmonary artery pressure of 25 mmHg) with age at operation; the findings were that maximal surgical cure is achieved when operation is performed in patients aged 3–31 months (peak at 14 months) of age. Analysis by Studer *et al*. (1982) of the University of Alabama at Birmingham experience, involving 156 patients with A-V septal defects and interventricular communication, revealed neutralization of the effect of young age on hospital mortality since 1977. The author therefore currently advises elective corrective surgery for all patients during the first year of life, preferably at age between 3–6 months. Severe elevation of pulmonary arteriolar resistance (above 10 U/m^2) contraindicates operation. When pulmonary arteriolar resistance is elevated (6–8 U/m′) in patients under age 3–6 months, prompt operation is advised. Trisomy 21 is present in about 25% of patients with complete A-V septal defects, and management of these patients is identical to that of others with A-V septal defects.

Surgical Technique

Surgical repair in the author's unit consists basically of closure of the interventricular communication with a Dacron patch, attachment of the common A-V valve to the crest of this patch, and closure of the atrial septal defect with a patch of pericardium. The basic architecture of the common A-V valve should not be altered, and use of the two-patch technique facilitates achieving this goal. Prosthetic valve replacement can be avoided in the overwhelming majority of patients and is not discussed because it is rarely necessary.

The location of the specialized conduction tissue in this form is similar to that in the partial form and is shown in **Figure 27.5**. The A-V node lies in the atrial wall just above the crux cordis. The penetrating bundle then passes through the tricuspid valve annulus (TVA) and divides into the left and right bundle branches (LBB and RBB).

It is essential for the operating surgeon to have a sound understanding of the variations in pathological anatomy found in this group of malformations, particularly in its complete form. A number of excellent descriptions are available (Wakai and Edwards, 1956; Lev, 1958; Rastelli *et al*., 1966; Tenckhoff and Stamm, 1973; Ugarte *et al*., 1976; Piccoli *et al*., 1979a, 1979b; Bharati *et al*., 1980).

Cardiopulmonary bypass and support techniques are similar to those employed for repair of the partial form. Cold cardioplegic myocardial protection is always employed, not only for its protective advantages but also because operating conditions are optimal in the soft, arrested heart. This aids in accurate suture placement

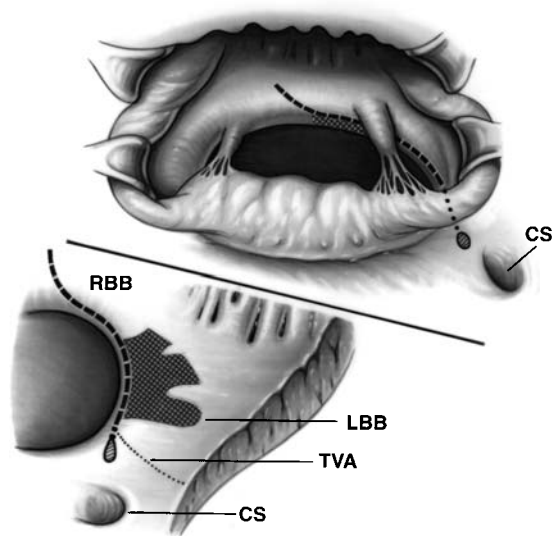


Figure 27.5

and reduced cardiac trauma. The right atrium is opened as shown in **Figure 27.1**. Cold saline is injected into the ventricles to approximate the A-V valve in its closed position. The anatomy of the valve leaflets, the presence of accessory orifices, the sizes and locations of the interatrial and interventricular defects, the location of the coronary sinus, and the possible presence of pulmonary or systemic venous anomalies are specifically discerned. The position of the common A-V valve in relation to the plane of the ventricular septum is determined, in order to ensure that repair will leave adequate orifices to each ventricle. When this adequacy is in doubt, it is helpful to compare the diameters of these orifices (measured with Hegar dilators) with the normal mitral and tricuspid dimensions, described by Rowlatt *et al*. (1963) and shown in **Table 27.1**. The morphological features of the left superior leaflet are studied to determine the position of the leaflet in relation to the crest of the ventricular septum, the origin and insertion of its chordal connections, the amount of its tissue and the size of the underlying interventricular communication. Some surgeons may find it helpful to classify the A-V valve according to Rastelli types A, B or C, but the author and associates believe that it is better to consider the anatomy of the A-V valve on a continuous pathological spectrum.

Cold saline is injected into the ventricles, and the valve architecture is studied in the closed position. The surgeon must identify the precise site of coaptation of the left superior and inferior leaflets above the crest of the septum. A 6-0 Prolene suture is placed through each leaflet at this coaptation site to mark the position of separation of the common A-V valve into mitral and tricuspid components. This site later becomes the apex of the septal commissure of the newly created mitral valve (**Figure 27.6**). The suture is left untied and is secured by a rubbershod clamp. When there is no rightward bridging of the left superior leaflet (Rastelli type A defect), this suture is usually, but not always, placed at the medial corner of this leaflet. When the left superior leaflet bridges right-ward (Rastelli type C defect), this suture is usually placed at a site directly above the crest of the ventricular septum. Hegar dilators are then placed through the valve on the left and right side of the suture to measure the size of the mitral and tricuspid portions, for comparison with the normal valve of similar-sized patients **(Table 27.1)**. If either the mitral or tricuspid portion is smaller than normal, this coapting suture is moved to a more appropriate site. An orifice one standard deviation smaller than the mean normal mitral valve is acceptable. Mild traction is placed on this suture, and saline is again reinjected in order to ascertain that the natural leaflet coaptation has not been disturbed and that valve incompetency has not been created or worsened. This important initial step sets the stage for a proper repair.

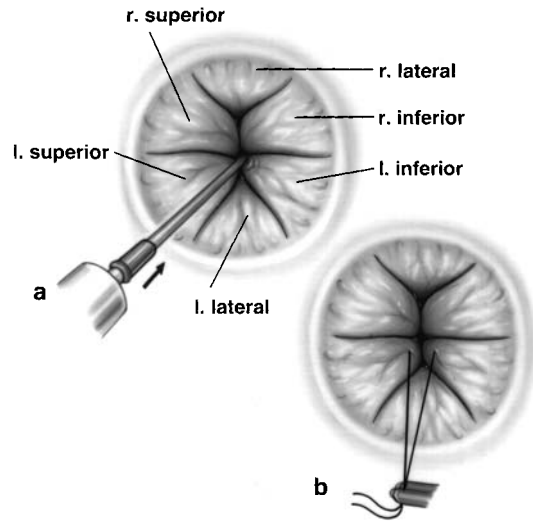


Figure 27.6

A separate patch of knitted polyester fibre (Dacron) is used to close the defect in the ventricular septum below the common A-V valve, and a second patch of pericardium is used to close the atrial component above. The previously placed coapting suture marks the place of attachment of the leaflets to the Dacron ventricular patch below and the pericardial atrial patch above. Stay sutures (6-0 Prolene) are placed on the right superior and inferior leaflets to permit gentle traction, in order to expose the defect in the ventricular septum. When there is marked rightward displacement of the left superior leaflet, one suture is placed on its rightward edge instead of on the right superior leaflet. When the ventricular septal deficiency extends beneath the left inferior leaflet, the leaflet is not incised and usually no chordae are divided. Sometimes, however, troublesome chordae attach the leaflet to the right side of the ventricular septal crest, and they are divided to permit complete exposure of the right side of the crest of the septum. This is sometimes necessary for improving exposure and aiding the surgeon in avoiding injury to the conduction tissue. When the left inferior leaflet is attached to the crest of the underlying septum by a membrane or by many closely placed chordae that permit only tiny communications, the chordae are left undisturbed because they will remain to the left of the Dacron ventricular patch. Similarly, when chordae are present attaching either the left or right superior leaflet to the right side of the crest of the septum or attaching the left superior leaflet to an anomalously placed right ventricular papillary muscle (Rastelli type B defect) and interfere with placement of the ventricular patch, they are divided (Pacifico and Kirklin, 1973).

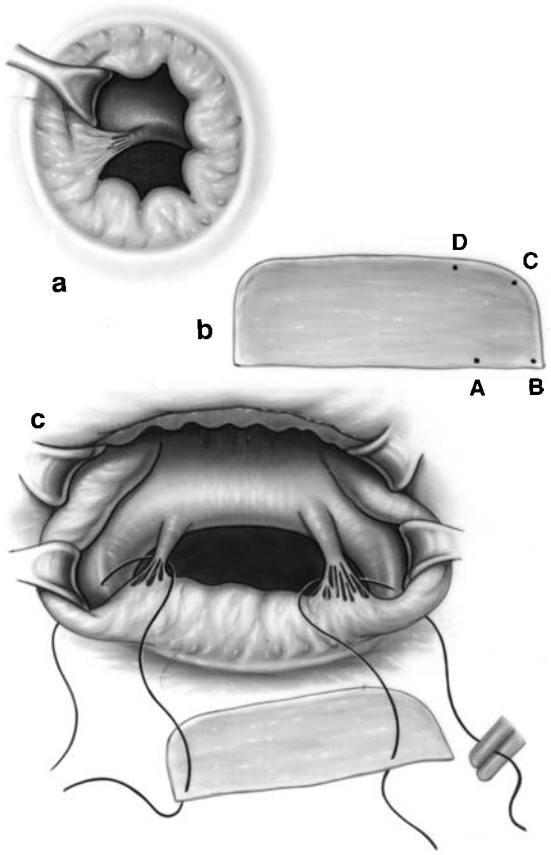


Figure 27.7

The next step in the procedure is to close the defect in the ventricular septum with the Dacron patch (**Figure 27.7**). Initially, the distance between the A-V valve annulus over the septum, superiorly and inferiorly, is measured with forceps. This measurement is used to mark the width of the Dacron patch. To judge the height of the patch, cold saline is injected into the ventricles, causing the leaflets to float to their closed position, and a second measurement is made. This is marked on the Dacron patch. The shape of the defect in the ventricular septum is studied and outlined on the Dacron patch, using a sterile pen.

The defect in the septum may extend more anteriorly than the position of the annulus, as is the rule when tetralogy of Fallot is associated. The measurements just described are very important and useful. If the patch is too narrow, the superior and inferior aspects of the A-V valve annulus will later be brought closer together. If there is significant central incompetence of the A-V valve, this closeness would be useful (and sometimes is purposely done), because it represents a type of annuloplasty and is desirable. Otherwise, the new circumference of the mitral and tricuspid valves is narrowed inappropriately, and leaflet prolapse later results. When the size and shape of the desired Dacron patch have been obtained, the patch is further extended 1 cm on its posterior aspect (**Figure 27.7**). This allows additional width to the patch, which can later be folded at this point (A–D) in a 90° angle to allow the extended segment of the patch to be attached to the base of the right inferior leaflet, or the bridging left inferior leaflet, so as to completely avoid suturing in the area of the conduction tissue. Double-armed 4-0 Prolene sutures are passed through the Dacron patch at the appropriate sites and then through the respective points of the A-V valve annulus, superiorly and inferiorly (**Figure 27.7**). The Dacron patch is continuously stitched with double-armed 4-0 Prolene sutures, beginning at the posterior annulus (**Figure 27.8a**). The patch is initially held away from the heart, and the suture that had been placed through it posteriorly is used to attach it to the base of the leaflet overlying the right side of the posterior septal crest (point A–B in **Figure 27.8a**). This is similar to the technique used in repair of perimembranous ventricular septal defects and of tetralogy of Fallot. The objective of this manoeuvre is to bring the suture line completely away from the area of the conduction tissue. When the corner of the patch is reached (point B), it is lowered

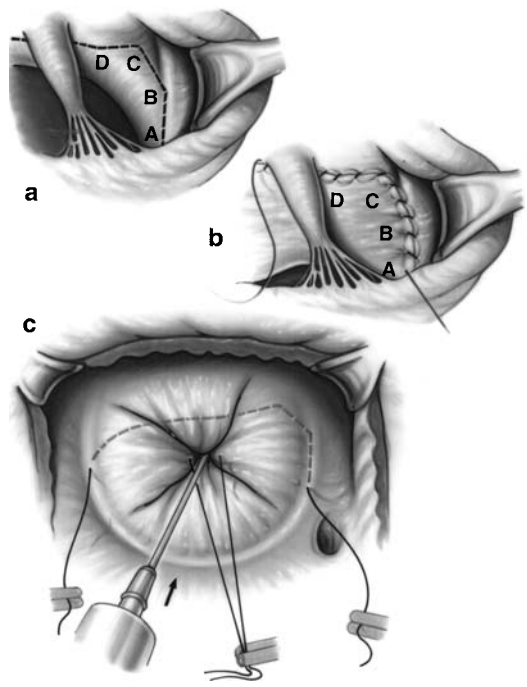


Figure 27.8

into place. Placement of points A, B, C and D of the patch on the heart are illustrated in **Figure 27.8a**; **Figure 27.8b** shows this part of the patch completed. Suturing then continues anteriorly, keeping initially on the right ventricular wall and then moving onto the right side of the ventricular septum until the superior aspect of the annulus is reached. At that point, the stitching is brought through the annulus into the atrium, tied to the previously placed and tied double-armed suture, and secured with a rubber-shod clamp.

The height of the patch is studied, appropriately contoured and trimmed if necessary (**Figure 27.8c**). If this is properly done, the patch should not interfere in any way with the contour of the closed A-V valve. It must not be too short, because excessive displacement of the valve into the ventricle can result in subaortic obstruction. The next step in the procedure is to attach the valve leaflets to the crest of the Dacron patch. This is accomplished with the use of interrupted 6-0 Prolene sutures placed at intervals of about 5 mm (**Figure 27.9**). The initially placed coaptation suture is tied, and usually one or more additional sutures are placed in the septal commissure to ensure appropriate leaflet coaptation and commissural stability. The entire group of sutures is left long and secured by a rubber-shod clamp.

The pericardial patch is next fashioned by measuring the distances necessary for it to close the atrial septal defect and also leave the coronary sinus to the left (**Figure 27.10a**). Suturing of this patch into place begins superiorly at the annulus and continues as a simple over-and-over stitch that incorporates the A-V valve leaflet (L), the underlying crest of the Dacron ventricular patch (D) and the pericardial patch (P) (**Figure 27.10b**). Placement of traction on the previously placed interrupted sutures in sequence aids inclusion of the Dacron patch into this suture line. When the inferior aspect of the A-V valve annulus is reached, the suture line continues anteriorly, incorporating the base of the right-sided leaflet and its underlying Dacron patch, before continuing around and rightward of the coronary sinus and around the edges of the atrial septal defect. Before completion of the attachment (**Figure 27.10c**), the interrupted sutures are cut and saline is injected into the mitral valve in order to assess its degree of competency. If additional valvoplasty is indicated, it is performed at that time, as shown on **Figure 27.2**. When there is a left-sided superior vena cava draining to the coronary sinus, the suture line for the pericardial patch must not leave the coronary sinus left-ward because this would result in postrepair cyanosis. In this circumstance, the suture line for the pericardial patch inferior to the septal commissure of the left A-V valve is placed on the base of the left inferior leaflet, as shown in **Figure 27.11**. The suture line then continues on the free edge of the atrial septum and leaves the coronary sinus right-ward. This technique is essential when there is a left-sided superior vena cava draining to the coronary sinus, but it is also the preferred technique of some surgeons.

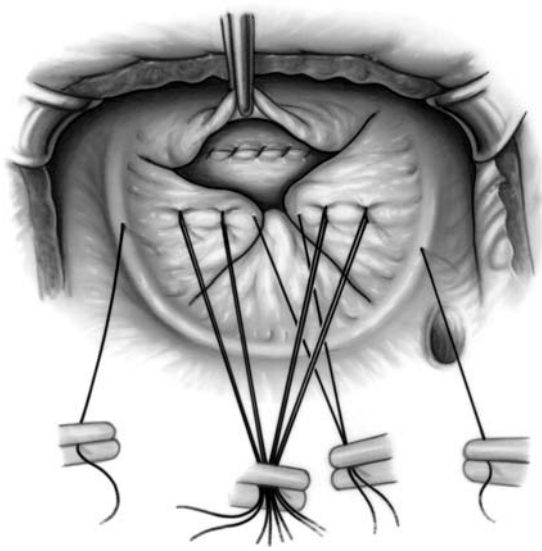


Figure 27.9

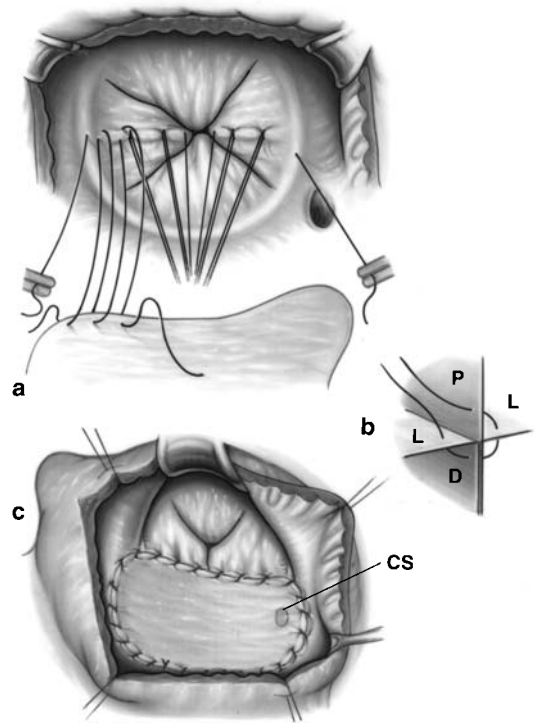


Figure 27.10

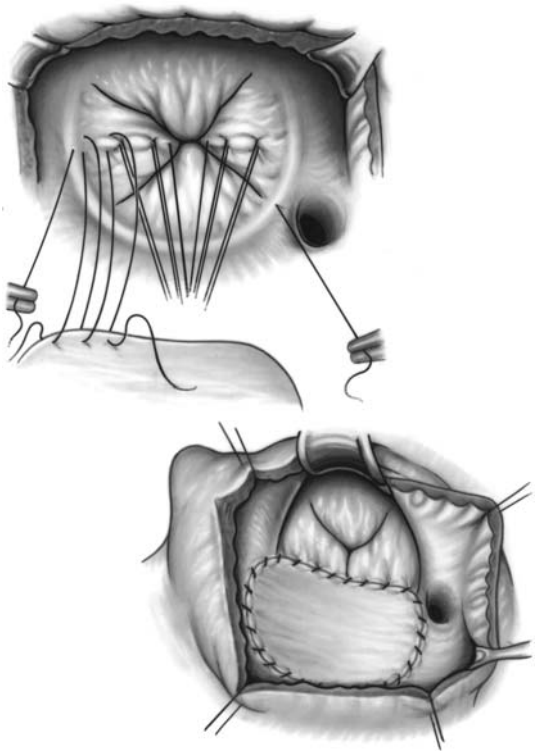


Figure 27.11

The type of patch material, and whether a single patch or separate patches are used to close the defects in the atrial and ventricular septa, remain controversial. The author and associates prefer separate patches, because this allows the surgeon more elasticity in order to avoid any significant alteration of the A-V valve and also to completely avoid suturing near the conduction tissue. A pericardial patch is preferred for closure of the atrial defect to avoid the small risk of severe postoperative haemolysis, which can result when a jet of residual mitral incompetence strikes the patch. The technique that the author and associates used previously is described only briefly. It is no longer used because it is believed to alter the basic architecture of the A-V valve more than does the two-patch technique, and it may be associated with an increased risk of A-V valve leaflet dehiscence. However, as the one-patch technique is occasionally useful, it will be described. Testing of the valve and placement of the initial suture is similar to the technique described earlier. The left superior leaflet (anterior bridging leaflet), if markedly bridging right-ward, or the commissure between the left and right superior leaflets may be incised superiorly to the valve annulus, as shown in **Figure 27.12**. If there is a large interventricular

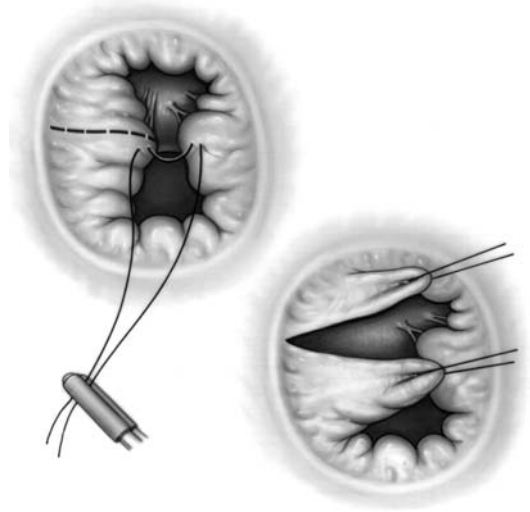
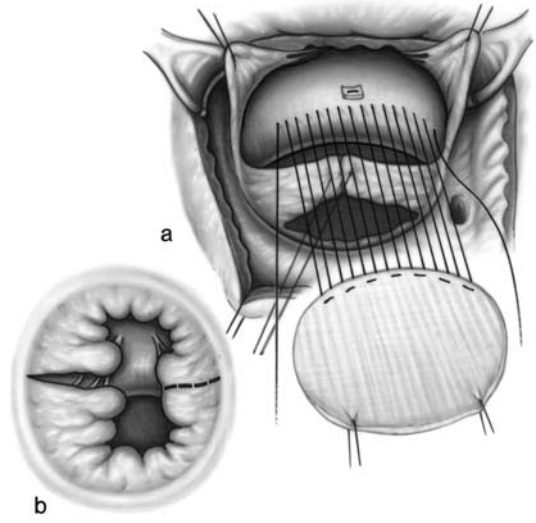


Figure 27.12

communication posteriorly, the inferior leaflets may also be similarly incised (**Figure 27.13b**). A pericardial or Dacron patch is then constructed and attached to the right side of the ventricular septum with either continuous (**Figure 27.13a**) or single sutures (**Figure 27.14**). The coronary sinus is left to drain to the left atrium (**inset, Figure 27.14**). The patch is then directed right-ward, and the initially placed coapting suture is tied to approximate the superior and inferior components of the newly created mitral valve. The base of this leaflet, and usually the right inferior leaflet, are attached at the appropriate level to the patch with interrupted horizontal 5-0 Prolene mattress



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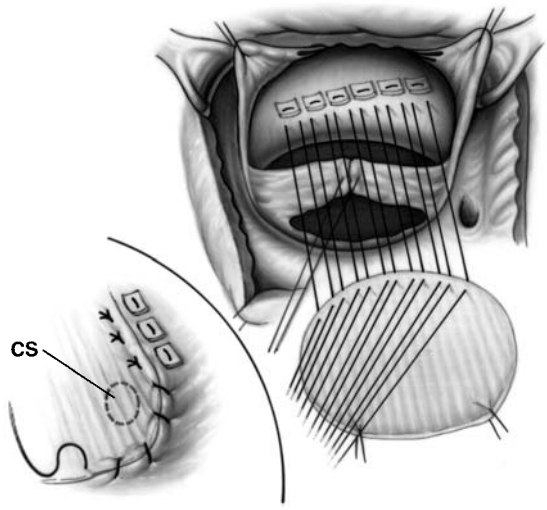

Figure 27.14

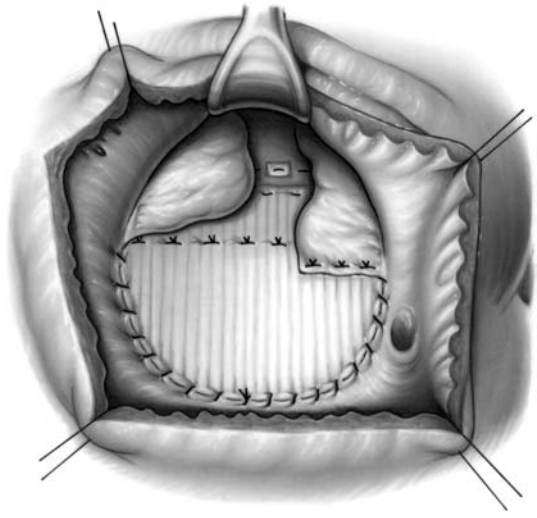

Figure 27.16

sutures (**Figures 27.15 and 27.16**). The precise level of attachment is usually judged to be on the same plane as the rest of the A-V valve annulus. Inappropriate inferior attachment may potentially result in subaortic obstruction. When the commissure between the left and right superior leaflets is completely developed (Rastelli type A defect), attachment of its tricuspid portion to the patch often seems inappropriate and may result in tricuspid valve stenosis (**Figure 27.16**). Wilcox *et al*. (1997) described a technique for repair of complete A-V septal defect which consists of suturing the left superior and left inferior leaflets directly to the crest of the septum, to obliterate the interventricular communication without using a patch. The ostium primum defect is then closed with a pericardial patch. These authors restricted this technique to patients having a shallow interventricular communication, but Nicholson *et al*. (1999) expanded its use to all types of interventricular communications without creating subaortic stenosis or A-V valve dysfunction. The author has used this technique and found it useful in selected patients.

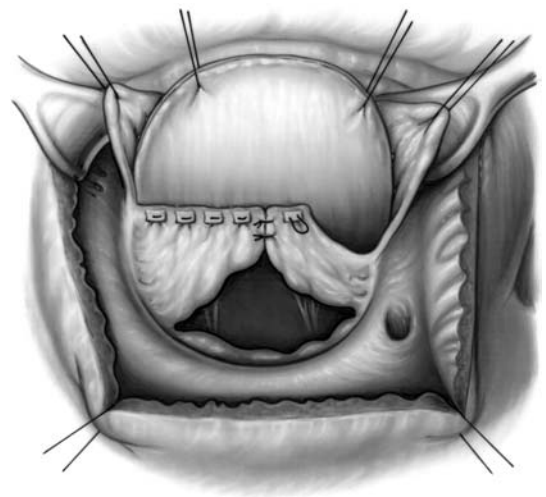

Figure 27.15

When an intermediate form of the A-V septal defect is present, the repair may be identical to that described for the partial form. However, if a significant interventricular communication is present, the fibrous bridge between the anterior and posterior leaflets is divided, which creates the complete form of the A-V septal defect and the defect is repaired as such. A-V septal defects are uncommonly accompanied by an intact atrial septum. Such hearts have all of the typical features of a complete A-V septal defect with a common A-V valve orifice, but the septum primum is intact, with the A-V valve leaflets either attached to the underside of the atrial septum or abutting it during ventricular systole. In either case, significant interatrial shunting is prevented. This anatomy may be confusing to the surgeon viewing it through a right atrial incision, and is clarified by opening the atrial septum to inspect, test and repair the left-ward components of the A-V valve. The right-ward portions of the superior and inferior bridging leaflets are retracted with stay sutures, and the interventricular communication is closed with a Dacron patch. The bridging leaflets are then attached to the crest of the Dacron patch, but because the plane of the ventricular septum is usually to the right of the atrial septum, it is also necessary to obliterate the communication from the

left ventricle to the right atrium by closure of the touching edges of the superior and inferior bridging leaflets, between their attachment to the crest of the Dacron patch and their attachment to the atrial septal edge (Stark *et al*., 1988). The presence of an anomalous left-sided superior vena cava draining to the upper left corner of the left atrium is managed as described by Sand *et al*. (1986). Repair of tetralogy of Fallot or double-outlet right ventricle associated with complete A-V septal defects is repaired as described by Pacifico *et al*. (1988).

Results

Progressive improvement in the early results after repair of complete A-V septal defects began in the 1970s and has continued into the modern era. Studer *et al*. (1982) analysed the 310 patients undergoing repair of all forms of A-V septal defects at the University of Alabama at Birmingham during 1967–1982. Over the years of this experience, hospital mortality rates progressively improved and the incremental risk associated with young age disappeared after 1976. The severity of preoperative A-V valve incompetence and New York Heart Association functional class affected hospital mortality rates. In 1984–1985 at the University of Alabama at Birmingham, there was 1 hospital death (2%) among 41 patients who underwent repair of isolated complete A-V septal defects (Kirklin *et al*., 1986). The absence of an incremental risk in association with young age in this later experience was again demonstrated, and lends confidence to the advisability of early surgical repair within the first 6 months of life. Severe ventricular dominance (hypoplasia of the right or left ventricle), increasing severity of A-V valve incompetence, increased functional disability and associated major cardiac anomalies probably still remain risk factors. Results in the last category, however, have significantly improved according to Pacifico *et al*. (1988), who reported 1 hospital death (3%) among 29 consecutive patients with complete A-V septal defects associated with tetralogy of Fallot, double-outlet right ventricle or additional anomalies associated with atrial isomerism. Improved hospital survival rates after repair of the isolated form as well as of forms associated with major cardiac anomalies also were reported by other groups (Binet *et al*., 1980; Bove *et al*., 1984; Castaneda *et al*., 1985; He and Mee, 1986; Urban, 1990; Backer *et al*., 1995; Bando *et al*., 1995; Reddy *et al*., 1998; Crawford and Stroud, 2001; Al-Hay *et al*., 2003).

Complications and Late Results

Complete heart block occurred in 1 of 96 patients (1%; 70% confidence limits, 0.1–3.5%) in the experience at the University of Alabama at Birmingham (Studer *et al*., 1982). Current methods of repair are designed to routinely avoid the area of the atrio-ventricular node and His bundle and should further reduce this occurrence. Kirklin *et al*. (1986) used equations to predict the 10 year survival rates according to the date of operation (**Figure 27.17**). The predicted 10 year survival rates were 99% for patients operated upon in 1985 with partial A-V septal defects and 95% for those with complete A-V septal defects.

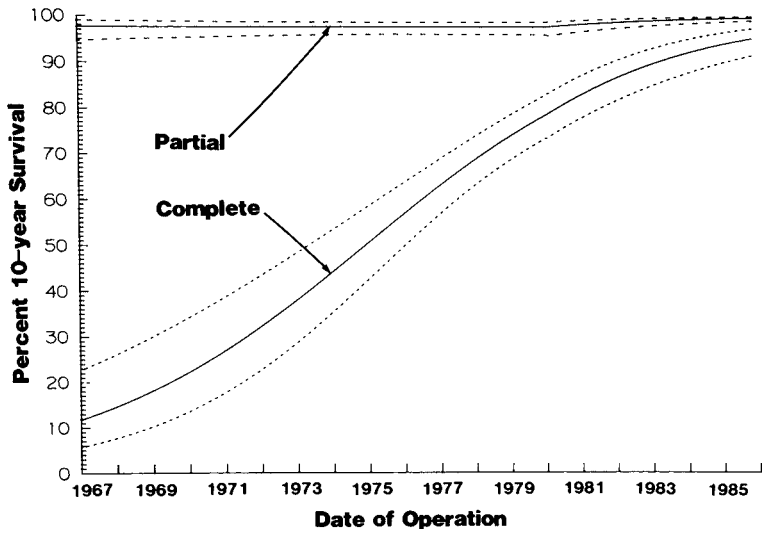

Figure 27.17

The actuarial late survival rate at 60 months among 23 patients operated upon at the University of Alabama at Birmingham before October 1976 was projected to be 91%, and each patient was asymptomatic at the time of follow-up (Berger *et al*., 1978, 1979). Two late deaths occurred in this group: one from recurrent pneumonia and sepsis 2.1 months postoperatively, and the second after a respiratory infection 2.9 months after operation. The latter patient had a pulmonary arteriolar resistance of 16 U/m^2 preoperatively and persistent congestive heart failure postoperatively. Although the magnitude of pulmonary vascular disease in that patient would ordinarily contraindicate repair (see the section on indications), the operating surgeon believed that the magnitude of symptoms, the poor prognosis without surgery and the young age of the patient warranted an attempt at repair.

In the experience at the Mayo Clinic with 25 hospital survivors, all but two were asymptomatic late postoperatively, as reported by McMullan *et al*. (1972). Late reoperations for residual or recurrent mitral valve incompetence were not required.

There were 19 deaths after hospital discharge among 258 traced patients surviving initial repair for the entire spectrum of A-V septal defects (Studer *et al*., 1982). The postoperative actuarial survival rate at 12.5 years was 84%. Of the 19 late deaths, 10 occurred in patients known to have left A-V valve repair failure. A Cox multivariate analysis identified severe preoperative left A-V valve incompetence, poor preoperative status, accessory A-V valve orifice and trisomy 21 as independent incremental risk factors for premature late death.

In the current era, there is support for advising complete repair at a very early age (2–3 months). Reddy *et al*. (1998), with a median follow-up of 24 months, do not believe this will adversely affect late A-V valve function. This is supported by the 12 year experience reported by Al-Hay *et al*. (2003), who found that the need for late A-V valve reoperation was less among patients operated before 3 months of age than in older subjects.

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28

Right Ventricular Outflow Tract Obstruction with Intact Ventricular Septum

C. van Doorn and M. R. de Leval

Pulmonary stenosis and pulmonary atresia with intact ventricular septum are classically described as two separate entities. However, there is a consensus that in the neonatal period, critical pulmonary stenosis and pulmonary atresia often have a similar clinical presentation, and hearts with pinhole pulmonary stenosis may indeed exhibit the anatomical features of right ventricular (RV) hypertrophy and cavity hypoplasia commonly seen in pulmonary atresia with intact ventricular septum. Right ventricular outflow tract obstruction (RVOTO) without RV hypertrophy is also included.

ANATOMICAL SUBSTRATE

Pulmonary Valve

In pulmonary atresia, the pulmonary valve is replaced by an imperforate, fibrous membrane. In some cases, prominent commissural ridges converge and meet at the centre of the imperforate valve. This is the form of imperforate valve observed in patients with severe infundibular stenosis or atresia. The other type of imperforate pulmonary valve is characterized by commissural ridges present only at the periphery of the valve, the centre being a smooth fibrous membrane, having the potential to bulge into the pulmonary trunk. This type of valve is usually associated with a lesser degree of right ventricular hypoplasia and with an open infundibulum (Zuberbuhler and Anderson, 1979). In severe pulmonary stenosis, there is commissural fusion at the periphery of the valve. Centrally, there is a smooth fibrous dome with an orifice that is usually central but sometimes eccentric. In less severe forms, two, three or sometimes four leaflets are relatively well formed, with only partial commissural fusion. The valve is usually thickened and may have a myxomatous appearance, particularly in neonates and small infants. Pulmonary stenosis due to mucoid thickening of the valve leaflets without commissural fusion is known as pulmonary valvular dysplasia. The valve ''ring'' may or may not be hypoplastic. Hypoplastic ring is often seen in the Noonan syndrome (Noonan and Ehmke, 1963).

Pulmonary Arteries

A good-sized main pulmonary artery with confluent branches is the usual situation in pulmonary atresia with intact ventricular septum. Beyond infancy, poststenotic dilatation of the main and proximal left pulmonary arteries is a common finding in patients with pulmonary stenosis. Stenoses of pulmonary arterial branches can occur in isolation or in association with other cardiac malformations. These can be central or peripheral, discrete or tubular, single or multiple. Multiple peripheral pulmonary stenoses are often a part of the rubella syndrome.

Right Ventricle and Tricuspid Valve

In pulmonary atresia with intact ventricular septum, right ventricular cavity size is variable with different degrees of myocardial hypertrophy and tricuspid valve anomalies. Most patients have both right ventricular hypoplasia and hypertrophy, both of which may be severe. In these hearts, the size of the tricuspid valve annulus has been found to relate to the degree of cavity hypoplasia. In addition, the tricuspid valve leaflets are often thickened and chordae are abnormal in number and attachments. At the other end of the spectrum are those hearts with greatly enlarged right ventricular cavities and a regurgitant dysplastic tricuspid valve, with or without displacement (Ebstein's anomaly). In this rare anomaly

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the right ventricular wall is usually thinned and, on occasion, may be devoid of myocardium (Uhl's anomaly and pulmonary atresia).

In valvar pulmonary stenosis, the obstruction produces concentric hypertrophy of the right ventricle, which may result in secondary infundibular stenosis. There is usually mild to moderate reduction in right ventricular cavity size, which is partly related to the concentric ventricular hypertrophy. Severe right ventricular hypoplasia or enlargement is rare. Isolated subvalvar pulmonary stenosis produced by a large anomalous muscle bands account for 10–20% of patients with pulmonary stenosis and intact ventricular septum (Kouchoukos *et al*., 2003).

Right Atrium

An atrial communication is present in all cases of pulmonary atresia with intact ventricular septum and in most neonates with critical pulmonary stenosis. The communication is restrictive in about 5–10% of neonates with pulmonary atresia with intact ventricular septum (Freedom *et al*., 1983).

Coronary Arteries

A substantial number of patients with pulmonary atresia with intact ventricular septum and diminutive and hypertensive right ventricle have persistence of right ventricular myocardial sinusoidal coronary artery connections (Freedom *et al*., 1983; Gittenberger-de Groot *et al*., 1988). The fistulous communications can connect the hypertensive right ventricle to one or both coronary arteries. Connection to the left anterior descending coronary artery is the commonest. Sometimes the proximal connection between the coronary artery and the aorta is absent, and perfusion of the involved coronary circulation is retrograde from the right ventricle and is dependent on right ventricular hypertension (right ventricular-dependent coronary circulation). Intimal fibromuscular hyperplasia often occurs in the coronary arteries linked with the sinusoids and may be responsible for myocardial ischaemia and infarction, involving either the right or the left ventricle or both.

PRINCIPLES OF MANAGEMENT

In recent years, a more aggressive approach towards early relief of right ventricular outflow obstruction was adopted. The aim is to provide pulmonary forward flow and stimulate right ventricular growth and enable the right ventricle to serve the pulmonary circulation whenever possible. In addition, there is good evidence that left ventricular function can be compromised by the presence of a hypertrophied, hypertensive right ventricle (Freedom *et al*., 1983; Elzinga *et al*., 1974). We feel that right ventricular decompression should be considered, even if the right ventricle is too small to be incorporated later into a biventricular circulation.

However, the right ventricle should not be decompressed in the presence of a right ventricular-dependent coronary circulation. Under these circumstances, the high-pressure right ventricle perfuses portions of the right and left ventricular myocardium with desaturated blood in systole. During diastole, the lower right ventricular diastolic pressure may allow a steal of blood from the coronary arteries into the right ventricle, to the detriment of the coronary circulation. Furthermore, the coronary circulation may be right ventricular-dependent in the case of a proximal discontinuity between one or both coronary arteries and the aorta and/or in the case of peripheral stenoses caused by intimal hyperplasia. Decompression of such ventricles may result in myocardial ischaemia and/or infarction in the distribution of the involved coronary arteries.

The decision between univentricular and biventricular repair depends on the morphological characteristics of the right ventricle, in particular the size of the right ventricular cavity and the presence or absence of coronary artery fistulae. The adequacy of the hypoplastic right ventricle for biventricular repair is difficult to assess and different methods have been used.

In patients with pulmonary atresia and intact ventricular septum, we have found it useful to classify the hypoplastic, hypertrophied right ventricles into three categories on the basis of the tripartite concept of the right ventricle (Goor and Lillehei, 1975; Bull *et al*., 1982): the hearts with all three components present; those with the absence of the trabecular portion; and those with an inlet zone only. In these hearts, the size of the tricuspid valve annulus was found to relate to the degree of cavity obliteration. Our studies (de Leval *et al*., 1985) have suggested that a right ventricle whose tricuspid valve diameter is within 99% (three standard deviations) of mean normal is capable of providing adequate pulmonary blood flow and of being incorporated into a biventricular circulation. Our initial measurements of the tricuspid valve diameter were made on angiocardiograms. Currently they are made on two-dimensional (2D) echocardiograms. **Table 28.1** gives the mean normal values for a given body weight and their lower 99% confidence limits. The mean normals were derived from the studies of Rowlatt and adjusted by Bull *et al*. (1982).

Hanley *et al*. (1993) suggested using the z value (standard deviation units) of the tricuspid valve diameter,

Table 28.1 Measurements of valve diameter used in decision making.

Weight (kg)	Angiographically estimated diameter of tricuspid valve (mm)	
	Normal mean*	Lower 99% CL
2	13	5
3	16	8
4	19	11
5	20	12
6	22	14
7	23	15
8	25	17
9	26	18
10	27	19
12	28	20
14	29	21
16	30	22
18	31	23
20	32	24
25	34	26
30	36	28

CL, confidence limit.
* Derived from autopsy data of Rowlatt *et al*. (1963). Adjusted for angiographic assessment according to Bull *et al*. (1982).

calculated as follows:

$$z \text{ value} = \frac{\text{measured diameter} - \text{mean normal diameter}}{\text{standard deviation of mean normal diameter}}$$

The normal diameter is the diameter in a normal individual of the same body surface area. The data for the mean normal diameters and their standard deviations also come from the anatomical data of Rowlatt. A nomogram for use in deriving the z value of the tricuspid valve diameter from the echocardiographically determined diameter and body surface area published by Hanley *et al*. (1993) is reproduced in **Figure 28.1**.

Patients with a three-portion right ventricle, whose tricuspid valve diameter is within three standard deviations of mean normal or whose z value of the tricuspid valve diameter is -1.5 or more, will have valvotomy either by interventional cardiological procedure or by surgery. The majority of these patients will not require a systemic-to-pulmonary artery shunt. If there is infundibular stenosis in addition to the valvar obstruction, the patient should receive a trans-annular patch, placed using cardiopulmonary bypass. The patent foramen ovale is usually left open. By and large, in patients with a patent infundibulum whose tricuspid valve diameter is below the 99%

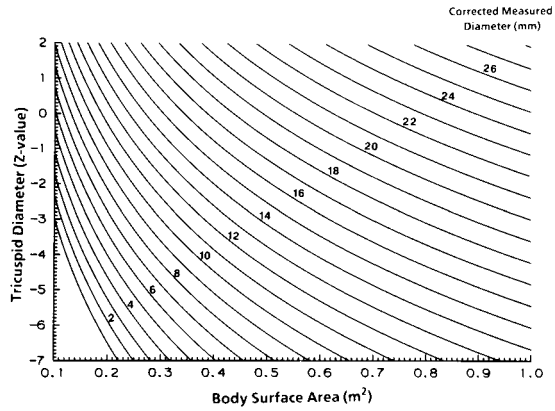

Figure 28.1

confidence limit of mean normal, there is infundibular stenosis in addition to the valvar atresia. These patients are best treated with a trans-annular patch without closure of the patent foramen ovale and with a concomitant systemic-to-pulmonary-artery shunt. In Hanley's protocol, this applies to patients in whom the z value of the tricuspid valve diameter is between -1.5 and -4. In patients without a patent infundibulum or those with a tricuspid diameter that has a z value of -4 or less, and those with significant anomalous coronary connections, a systemic-to-pulmonary artery shunt only is performed. A balloon atrial septostomy should be carried out in these patients if the atrial septal defect is restrictive.

In terms of timing of intervention, patients with right ventricular outflow tract obstruction and intact ventricular septum fall into one of the three categories:

- The symptomatic neonate, who by and large will receive a palliative treatment.
- The child who survived palliation in infancy.
- The asymptomatic child with pulmonary stenosis.

Symptomatic Neonate with Critical Pulmonary Stenosis or Pulmonary Atresia

Cyanosis is usually present in such infants on the first day of life and rapidly becomes more severe as the ductus arteriosus closes. The suspected diagnosis of critical pulmonary stenosis or pulmonary atresia with intact ventricular septum is easily confirmed non-invasively by 2D echocardiography. In some cases, the diagnosis may already have been made prenatally (Daubeney *et al*., 1998). Echocardiography must provide the following information: size and type of right ventricular cavity; level of atresia (valvar or infundibular and valvar); size and competence of the tricuspid valve; size and

confluence of the pulmonary arteries; size of the interatrial communication; size of the patent ductus arteriosus; and left ventricular size and function. Cardiac catheterization and angiography are indicated when right ventricular sinusoidal coronary artery connections are suspected. A right ventricular angiogram will disclose connections between the right ventricular myocardial sinusoids and the coronary arteries. If there is retrograde opacification of the aortic root, there is certainly proximal continuity between the aorta and the coronary arteries. When dense opacifications of the coronary circulation from the right ventricle do not lead to visualization of the aortic root, it is necessary to exclude proximal discontinuity between the aorta and the coronary arteries (Freedom *et al*., 1983). A left ventriculography or aortogram is then necessary to study the origin of the coronary arteries and to identify stenoses of the coronary branches linked to the sinusoids.

The primary goal of treatment is to maintain an adequate pulmonary blood flow. This ensures survival beyond the neonatal period. In addition, treatment should aim to enable the right ventricle to serve the pulmonary circulation whenever possible, to enlarge a restrictive atrial septal defect, to decompress a hypertensive right ventricle, even if not large enough to serve the pulmonary circulation, and to deal with coronary anomalies if present.

Our current management protocol for neonates with pulmonary atresia and intact ventricular septum is as follows. Prostaglandin E is started, to maintain patency of the ductus arteriosus, as soon as the diagnosis is suspected. For patients with the smallest right ventricles, whose cavity is made of an inlet portion only, a right-modified Blalock–Taussig shunt is our procedure of choice. In patients with a patent infundibulum, relief of right ventricular outflow obstruction can be accomplished using a variety of procedures. In many centres, including our own, percutaneous pulmonary balloon valvuloplasty has replaced open valvotomy for most patients. Using mechanical, laser or radiofrequency energy, the valve is perforated and then dilated with a balloon. Pulmonary regurgitation introduced by the balloon dilatation is generally mild, is usually well tolerated and does not appear to be more severe than that introduced by surgical valvotomy. The prostaglandin infusion is slowly reduced after the procedure and discontinued over the following few days. Surgical intervention may occasionally still be necessary for severely dysplastic valves that cannot be opened by balloon valvuloplasty. Surgical resection of the valve cusps with/without trans-annular patching may then be required. Under these circumstances the procedure is usually performed via median sternotomy on cardiopulmonary bypass.

Because postoperative right ventricular performance is not easy to predict, it may be difficult to know whether the patent ductus arteriosus and the atrial septal defect should be left patent or closed at the time of the operation. An outflow tract patch inevitably creates pulmonary regurgitation that interferes with right ventricular diastolic function. It may increase the right-to-left shunt at atrial level, if the atrial septal defect is left open or causes congestive right ventricular failure, if it is closed. Occasionally, if cardiopulmonary bypass is contraindicated, closed surgical pulmonary valvotomy can be performed. The necessity and the timing of a systemic-to-PA shunt remains controversial in patients presenting with mild to moderate RV hypoplasia and a patent infundibulum (Alwi *et al*., 2005). A smaller tricuspid valve size was the only predictive factor for the need of systemic-to-PA shunt after RV decompression.

Management of patients with right ventricular-to-coronary artery connection remains difficult. Discrete fistulae can sometimes be ligated. Right ventricular thrombo-exclusion was advocated (Williams *et al*., 1991) in patients with intramural connections, to prevent a steal phenomenon into the right ventricle. Patients with right ventricular-dependent coronary circulation might be inoperable and considered for heart transplantation.

Definitive Repair of Patients with Pulmonary Atresia with Intact Ventricular Septum

The main strategies for definitive surgery are:

- Biventricular repair in patients with two adequate ventricles.
- Single ventricle repair for very small right ventricle and/or right ventricle-dependent coronary circulation.
- One-and-a-half ventricle for varying degrees of RV hypoplasia.
- Transplantation in very high-risk patients for the above strategies.

Our data (de Leval *et al*., 1985) have suggested that a three-portion right ventricle with a tricuspid valve diameter above the lower 99% confidence limit can safely be used to support the pulmonary circulation. This would also apply to patients whose z value of the tricuspid valve diameter is -1.5 or more. Patients who fulfil these criteria, either since birth or as a result of a satisfactory decompression, will undergo radical repair, leading to a biventricular circulation. The operation consists of relieving any residual right ventricular outflow tract obstruction and closing the atrial septal defect and the extracardiac shunt, if present. Some or all of these procedures can be sometimes carried out by interventional

cardiac catheterization. Recruitment of the right ventricular cavity by excision of hypertrophic muscle of both the trabecular and infundibular portions may render some ventricles suitable for biventricular repair (Pawade *et al*., 1993). An outflow tract patch enlargement or homograft valve placement may be necessary in some patients. If tricuspid valve regurgitation is present, repair of the tricuspid valve is undertaken.

Patients whose decompressed right ventricle has not grown sufficiently to provide a right ventricular-dependent pulmonary circulation can benefit from a variety of procedures. If it is believed that the right ventricular cavity is large enough to deal with two-thirds of the systemic venous return, the superior vena cava can be connected to the right pulmonary artery (classical Glenn procedure), and the inferior vena caval blood can be diverted to the left pulmonary artery via the right ventricle, following complete relief of any residual right ventricular outflow tract obstruction. The atrial septal defect should ideally be closed to separate completely the systemic and the pulmonary circulations (**Figure 28.2a**). Because of the known complications, we no longer perform classical Glenn.

An alternative approach for the creation of a one-and-a-half ventricle circulation, suggested by Billingsley *et al*. (1989), consists of performing a bidirectional cavopulmonary anastomosis, inserting a homograft in the right ventricular outflow tract, and closing the atrial septal defect (**Figure 28.2b**). The ASD can also be left partially opened, with a small defect created centrally in the patch so that at a later date closure with a device would be possible. Alternatively, an adjustable atrial septal defect can be used (Laks *et al*. 1991) (**Figure 28.3**). The theoretical concern about one-and-a-half ventricle repair is that in ventricular systole the peak pulmonary arterial pressure can be higher than the superior vena caval pressure, thus producing systemic venous hypertension with retrograde flow in the superior vena cava. Incorporating too small a right ventricle in the circulation can become more of a hindrance than a benefit, and a Fontan type of procedure might be preferable.

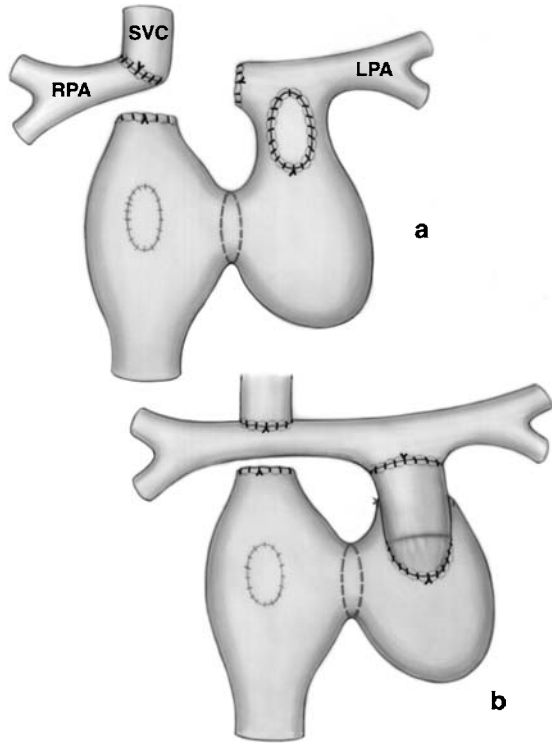

Figure 28.2

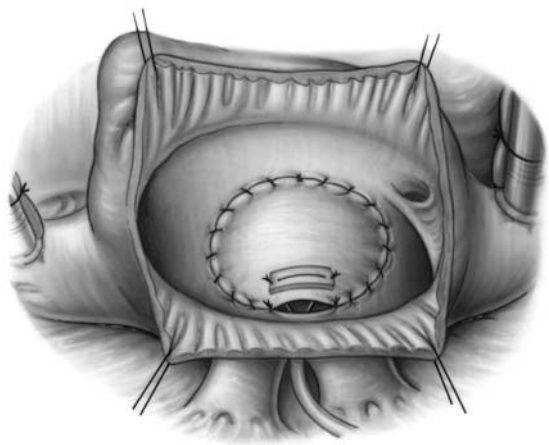

Figure 28.3

Repair of Right Ventricular Outflow Tract Obstructions in Children

There is no unanimity concerning the level of right ventricular systolic pressure at which relief of right ventricular outflow obstruction is indicated. In general, pulmonary stenosis is well tolerated and the risk of sudden death is much lower than with obstruction to the left ventricular outflow. A cooperative study (Nugent *et al*., 1977) recommended surgery in children with gradients greater than 50 mmHg. The advent of trans-catheter balloon valvuloplasty (Kan *et al*., 1982) has reduced considerably the number of patients who require surgical treatment for isolated pulmonary valve stenosis. For balloon valvuloplasty, the combination of right ventricular hypertrophy and a peak velocity of 4 m/s would encourage most cardiologists in the UK to intervene (Gibbs, 2000).

OPERATIVE TECHNIQUES

Pulmonary Valvotomy/Valvectomy under Inflow Occlusion

This procedure is usually carried out at normothermia through a midline sternotomy. Details of surgical technique are illustrated in **Figure 28.4**. An incision is made, and pericardial stay sutures are inserted as usual on the edges of the pericardium. Tapes are placed around the superior and inferior venae cavae. A suture tied around the tip of the on the left atrial appendage is used to keep it away from the anterior aspect of the pulmonary artery. The aorta and the pulmonary arteries are not dissected apart. A side-biting clamp is then placed on the main pulmonary artery, onto which a longitudinal incision of about 1 cm in length is now made. Two stay sutures of 6-0 polypropylene are then placed on the edges of this arteriotomy incision.

It is important to emphasize the details of preparations made for inflow occlusion. The patient is hyperventilated with 100% oxygen; sodium bicarbonate is given to counteract the metabolic acidosis that may develop during the arrest. The IVC and SVC tapes are snugged. When the systemic arterial pressure falls to 30 or 40 mmHg, a large side-biting clamp is introduced into the transverse sinus, so as to occlude the ascending aorta and the bifurcation of the pulmonary artery in such a way that the flow from the ductus arteriosus to the pulmonary arteries is interrupted. If the ductal flow is not interrupted during the period of inflow occlusion, blood will continue to flow from the aorta into the pulmonary arteries, from there into the left atrium, then into the right atrium through the patent foramen ovale, and then into the right ventricle and the pulmonary artery, thus allowing blood to pour through the pulmonary artery and making the valvotomy difficult.

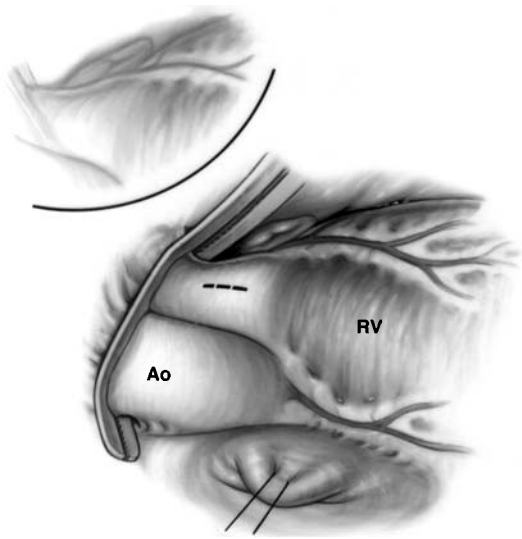

Figure 28.4

The valve is opened as widely as possible. If the valve is very dysplastic and remains obstructive in spite of the valvotomy, the cusps are excised. The caval tapes are then released. The lateral stay sutures are held so as to place the side-biting clamp on the main pulmonary artery. The side-biting clamp placed into the transverse sinus is released, and the pulmonary arteriotomy incision is closed with a running suture of 6-0 polypropylene. If pulmonary blood flow is still felt not to be satisfactory after the valvotomy, a modified Blalock–Taussig shunt can be added during the same procedure.

Relief of Right Ventricular Outflow Tract Obstruction on Cardiopulmonary Bypass

A midline sternotomy incision is made. A patch of pericardium is harvested if a patch enlargement is expected to be required. Bovine pericardium or a Gore-Tex patch can also be used. The pulmonary trunk and the aorta are separated from each other by sharp dissection, so that the aorta may be cross-clamped without impinging on the pulmonary artery. The aorta is cross-clamped because there is an ASD. The usual techniques for cardiopulmonary bypass are used. Hypothermic cardiopulmonary bypass is initiated; the degree of hypothermia depends on the expected length of the procedure. If a patent duct is present, it is closed. The aorta is cross-clamped and the cardioplegic solution is infused into the aorta. The left side of the heart is vented either across the atrial septum or by placing a sump sucker into the opened pulmonary artery.

Pulmonary Valvotomy/Valvectomy

A vertical incision is made in the proximal main pulmonary artery. The incision is carried down into the anterior sinus of Valsalva. The exposure is facilitated by the placement of a small nerve root retractor in that sinus. The anatomy of the valve is inspected. If the valve is tricuspid, the fused commissures are incised sharply with a knife. If the valve is bicuspid, it is often necessary to detach the cusps from their lateral attachment to the arterial wall to obtain a satisfactory relief of the obstruction. The detachment of the cusps from the arterial wall is done both in bicuspid and tricuspid settings (**Figure 28.5**).

Some degree of incompetence will inevitably result from this manoeuvre. If the valve cusps are bulky and obstructive, valvectomy may be necessary to relieve the obstruction (**Figure 28.6**). The pulmonary arteriotomy

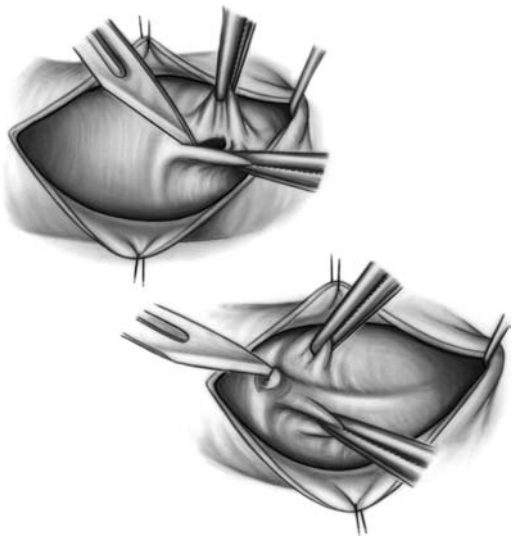

Figure 28.5

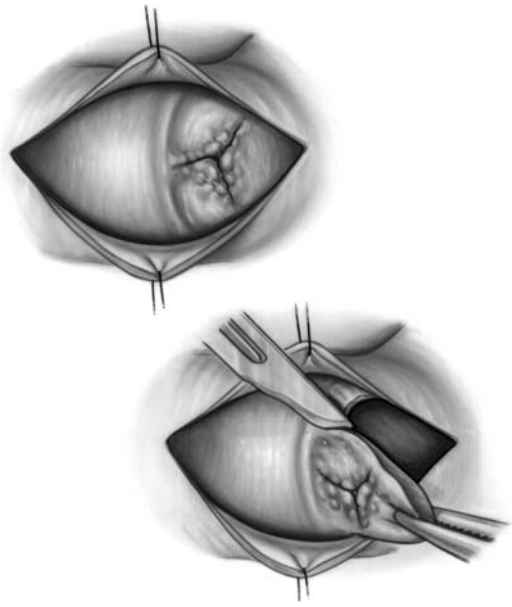

Figure 28.6

incision is usually closed directly with a continuous suture of fine polypropylene. If there is concern about the size of the pulmonary artery, a patch of pericardium is used. If the atrial septal defect is to be closed, the caval snares are tightened and the right atrium opened. The left heart is allowed to fill with blood and the interatrial communication closed, either directly or with a small patch. The right atrium is closed. The aorta is unclamped and the heart is de-aired.

Secondary infundibular hypertrophy is a common cause of residual gradient between the right ventricle and the pulmonary artery. It is sometimes difficult to decide whether an infundibular resection with, or without, patch enlargement is necessary. In the setting of biventricular repair a small dose of propranolol (0.01–0.02 mg/kg) can be given intravenously when the RV/LV pressure ratio is greater than 0.75, or when the RV/PA gradient is greater than 50 mmHg. This manoeuvre can identify patients in whom the residual gradient is reversible and those who need further surgical relief of the obstruction (Wensley *et al*., 1987). Intraoperative echocardiography may also be useful.

Infundibular Resection

A trans-atrial–trans-pulmonary approach can be used initially, and only if access is inadequate is the right ventricle opened. The right ventricle is opened through a transverse incision, which is closed directly, or alternatively through a vertical incision, which is usually closed with a small patch. A vertical ventriculotomy allows for extension across the pulmonary valve annulus, if trans-annular patching is required. Infundibular obstruction is most commonly due to hypertrophy of the parietal and septal extensions of the infundibular septum. The hypertrophied muscular bands are widely excised. The resection usually starts on the septal limb, taking care to remain fairly superficial to avoid perforation of the septum or injury to the septal branches of the left anterior descending coronary artery. Passing a clamp behind each hypertrophied mass before transection shows that the ventricular septum is neither thinned nor perforated (**Figure 28.7**).

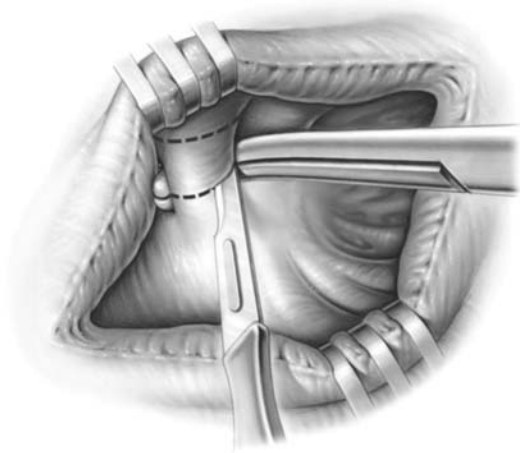

Figure 28.7

The base of the medial papillary muscle of the tricuspid valve may attach to one of the hypertrophied septal bands, and care must be taken not to transect it. The parietal wall of the right ventricle is mobilized in a similar fashion, extending the resection as far caudally as the papillary muscle of the tricuspid valve. The infundibular septum itself may be trimmed where it bulges into the floor of the outflow tract. When the infundibular resection of the right ventricle is carried out from the right atrium, the parietal extension of the infundibular septum is situated superiorly and can be approached inferiorly with an angled clamp, so as to transect it from the infundibular septum and the free wall of the right.

Relief of Mid-right Ventricular Obstruction (Double-chambered Right Ventricle)

This intraventricular obstruction is situated between the inlet and the outlet portions of the right ventricle. It is usually, but not always, associated with a small or moderately large ventricular septal defect. The repair is done on cardiopulmonary bypass with bicaval cannulation. The examination of the heart may reveal the presence of a thin-walled distal chamber.

There is often a branch for the right coronary artery that courses over the underlying obstruction. The obstruction can often be relieved via a right atrial approach. The ventricular cavity is explored via the tricuspid valve, entering the proximal chamber. The muscular obstruction in the roof of the chamber often leaves a small circular orifice, covered with fibrous tissue. This looks very similar to a small ventricular septal defect. However, a ventricular septal defect tends to be situated more leftward and inferior and may bound onto the tricuspid valve. Gentle probing of the opening will reveal connection with the distal chamber. Resection, initially starting anterior and to the left of the opening, will start to open up the connection with the distal chamber, taking care not to divide the papillary muscles of the tricuspid valve. Alternatively, the resection of the obstruction can be done through a transverse right ventriculotomy, immediately above the site of the obstruction, and of the coronary branch (if present). The appearance of the muscular obstruction in the bottom of the infundibular chamber is illustrated in **Figure 28.8**, often very similar to a small ventricular septal defect. The tricuspid valve, which is situated upstream to the obstruction, cannot be visualized, and this indicates that the orifice is not a ventricular septal defect. An incision is first made into the obstructing muscle anteriorly and to the left. This allows one to see the proximal chamber of the right ventricle and the tricuspid valve apparatus. The resection is then completed, taking care to preserve the papillary muscles of the tricuspid valve.

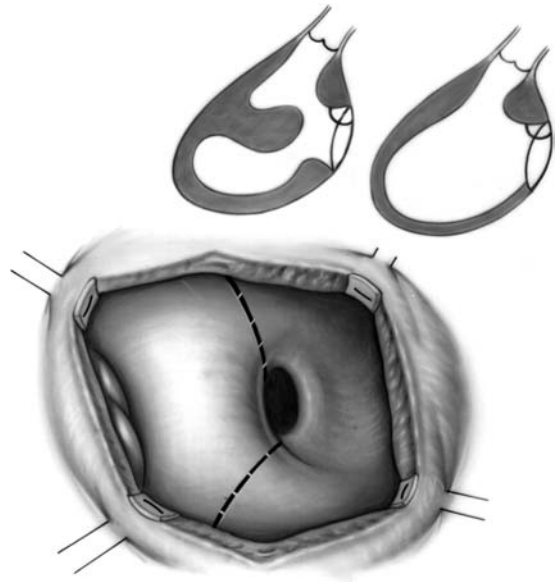

Figure 28.8

If present, the ventricular septal defect is then closed. The right ventriculotomy incision is closed directly.

Trans-annular Patch Enlargement of Right Ventricular Outflow Tract

When the right ventricular outflow tract obstruction is due to a diffuse hypoplasia, involving the infundibular chamber and the pulmonary valve ring, a trans-annular patch may be required. The annulus is sized with Hegar dilators. If the diameter is more than two standard deviations below the mean normal value established by Rowlatt *et al*. (1963), as shown in **Table 28.1**, a trans-annular patch is inserted. The patch may be fashioned from pericardium or from prosthetic material (Dacron or Gore-Tex). It should be oval; diamond-shaped patches increase the risk of stenosis at the end of the patch (**Figure 28.9**). The original incision in the pulmonary artery is extended inferiorly across the pulmonary valve ring. The pulmonary valve is at least partially excised anteriorly. The posterior cusps might be left *in situ* if they are not obstructive. The patch is positioned, starting the suturing with a mattress suture at the most cephalad end of the incision (6-0 polypropylene sutures for neonates and small infants; 5-0 in older children). The suturing then continues with a running over-and-over suture, placing the first two or three bites along each side before pulling the pericardial patch into position as the suture is tightened. Suturing is continued on each side of the pulmonary artery and then on the right ventriculotomy

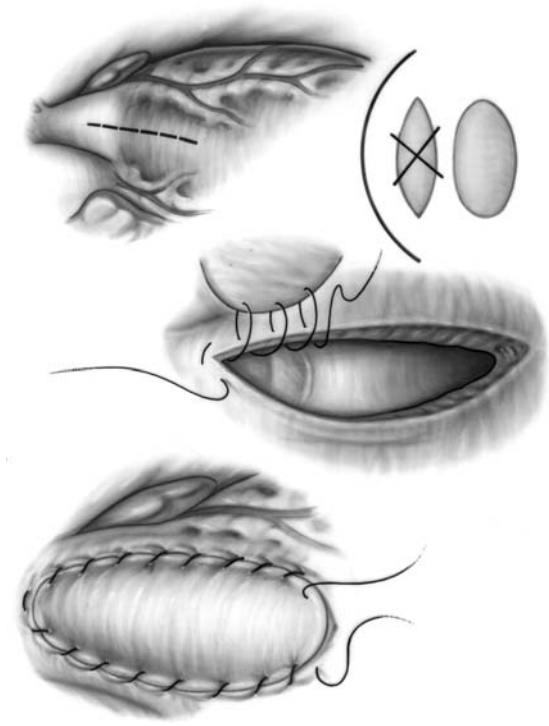

Figure 28.9

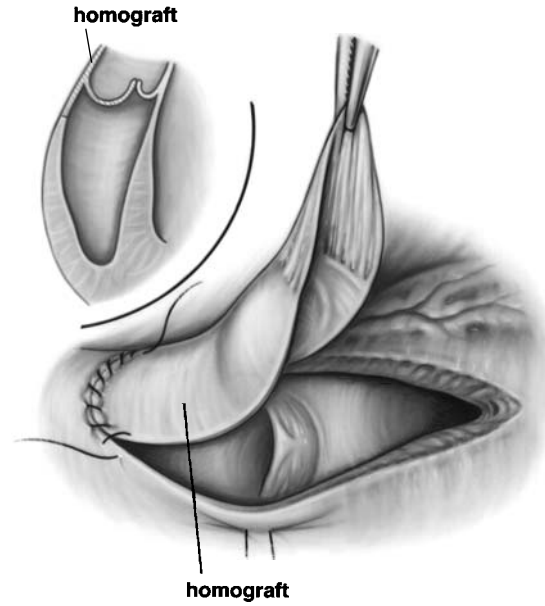

Figure 28.10

incision, taking deep bites into the muscle. This part of the operation can be performed on the beating heart while rewarming the patient, if air has been completely evacuated from the left side of the heart and if there is no residual communication between the two atria. Before completing suturing in the patch, the right ventricular outflow tract is reinspected to ascertain that the size is still adequate after the heart muscle has regained tone. Monocusp patches fashioned from homografts or pericardium have been used; recently Gore-Tex has been recommended (Turrentine *et al*. 2002) to minimize postoperative pulmonary regurgitation. The insertion of a homograft unicusp patch is illustrated in **Figure 28.10**. In placing the patch, it is important to position the valve at the same level as the posterior leaflet of the patient's own valve cusp for optimal effect. Usefulness of these valved patches remains controversial.

If it were considered that a competent pulmonary valve was required, we would use an aortic or a pulmonary homograft. In the presence of pulmonary hypertension, a porcine valve Dacron conduit may be preferable. Similarly, poor right ventricular function, tricuspid valve incompetence and small right ventricular cavity are other indications for pulmonary valve insertion at the time of right ventricular outflow tract reconstruction. This may consist of an orthotopic pulmonary valve replacement or the interposition of a homograft between the right ventricle and the pulmonary artery.

Pulmonary Valve Replacement Using Homograft Interposition

The pulmonary artery is dissected from the ascending aorta. If the main pulmonary artery and pulmonary annulus are very hypoplastic, further dissection is necessary: The pulmonary artery branches are fully mobilized down to the hilum on both sides and the ligamentum arteriosum is doubly ligated and divided. A full mobilization of the pulmonary arteries will permit the use of a short homograft without the need for prosthetic material extension. The pulmonary artery is transected upstream to the bifurcation. The opening might need to be extended into the proximal right and/or left pulmonary arteries to match the diameter of the homograft. The homograft is tailored to the appropriate length (allowing for the heart regaining its tone after the release of the aortic clamp) and the distal anastomosis is made with a running 5-0 or 6-0 polypropylene suture. The proximal anastomosis is then carried out, starting the suturing posteriorly on the superior margin of the right ventriculotomy incision (in case of pulmonary atresia) or on the posterior aspect of the pulmonary valve ring. This is usually done with a 5-0 or 4-0 polypropylene suture. The suture is continued from the midline along both sides around half of

the circumference of the conduit. If the conduit is an aortic homograft, the anterior leaflet of the mitral valve may sometimes be used as an onlay patch to complete the insertion of the conduit to the right ventricle. On the other hand, the mitral valve leaflet may be resected, as this would give the aortic homograft a more natural curve. Whenever possible, we try to avoid extensions of these homograft conduits with autologous pericardium because of aneurysmal dilatation of the patch over time.

When the hypoplasia of the pulmonary artery extends to its bifurcation and the origin of both the right and the left pulmonary arteries, a long pulmonary artery homograft taken with its bifurcation can be anastomosed to the transected right and left pulmonary arteries and to the right ventriculotomy. A patch of pericardium (patient's own or bovine) or prosthetic material is cut to a semilunar shape and is sutured between the homograft and the margins of the right ventriculotomy (**Figure 28.11**). The exposure is sometimes facilitated by a transection of the ascending aorta. In our experience this is rarely necessary after ample mobilization of the aorta and the pulmonary arteries.

Repair of Supravalvar Pulmonary Stenosis

Stenosis of the origin of the left pulmonary artery is dealt with by extending the incision from the main pulmonary artery well beyond the obstruction. Stenosis of the origin of the right pulmonary artery is more difficult to repair. The incision must extend from the main pulmonary artery, well across the stenosis (**Figure 28.12**). Adequate mobilization of the ascending aorta usually permits a good exposure of the origin of the right pulmonary artery. Stenosis of the pulmonary artery bifurcation is usually dealt with by extending the incision from the main pulmonary artery, well into both the left and the right pulmonary arteries. It is often difficult to tailor a rather complex T-shaped patch to the appropriate size; it may be easier to use two patches. A rectangular patch is utilized to widen the right and left pulmonary arteries, and a second patch to widen the main pulmonary artery, as shown in **Figure 28.13**. Cooperation with an interventional cardiologist is essential; he/she may be able to relieve some of these obstructions by balloon dilatation and/or stenting, if indicated.

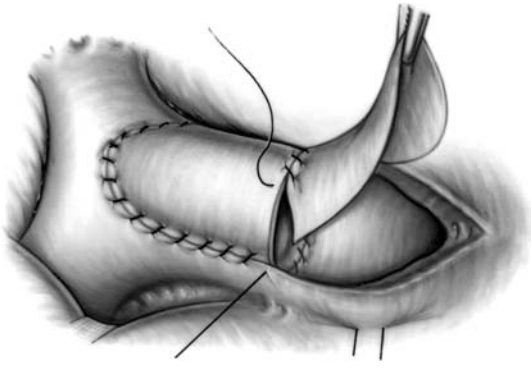

Figure 28.11

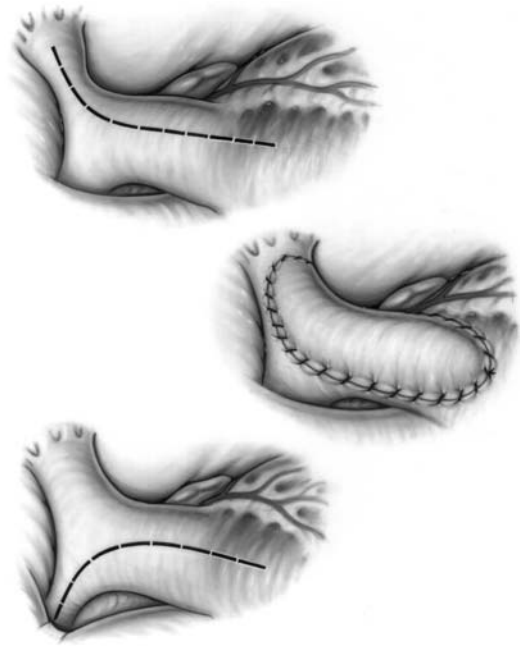

Figure 28.12

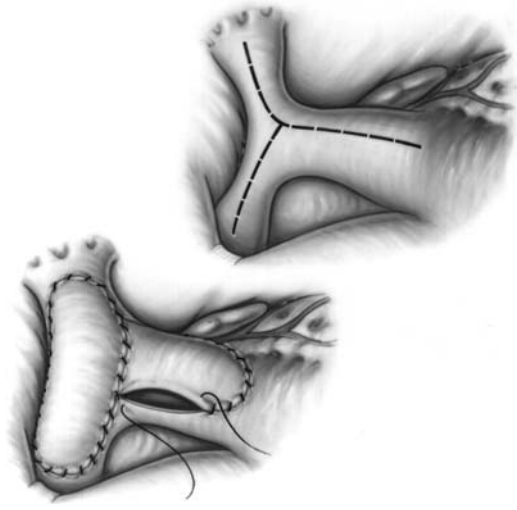

Figure 28.13

RESULTS

RVOTO with Normal Sized Right Ventricle

In the absence of right ventricular hypoplasia, the mortality rate for repair of pulmonary valve stenosis and intact ventricular septum is very low and approaches zero for patients with normal right ventricular function. The postoperative mortality and morbidity rates are higher for patients in chronic right heart failure. The 20–30 year postoperative follow-up is excellent for patients operated on before the age of 21 years. In older patients, late survival and functional status were poorer than in the control populations (Kopecky *et al*., 1988).

Pulmonary Atresia with Intact Ventricular Septum

Neonatal Palliation

Because of the different patients groups and various surgical methods to open the RV outflow tract comparisons of results is very difficult. Introduction of trans-catheter relief of the obstruction improved the outcomes. A recent study by Alwi *et al*. (2000) of 33 children showed that in selected patients with patent infundibulum, percutaneous radiofrequency perforation of the atretic pulmonary valve associated with balloon dilatation was safer than closed surgical valvotomy and BT shunt (total mortality, 15.7% vs. 42.8%). Similar results were presented by others (Ovaert *et al*., 1998). The authors reported a 17% mortality using percutaneous perforation in a more heterogenous groups. In the past, surgical closed valvotomy carried 33% mortality (Bull *et al*., 1994).

Agnoletti *et al*. (2003) reported the result of the strategy currently employed in Paris. In anatomically favourable cases (patent infundibulum and non RV-dependent coronary circulation), catheter decompression of the RV was first attempted (39 patients, two deaths, four failures leading to urgent surgery with no death). At a median follow-up of 15 days, 21 patients required further surgery (12 BT shunts, eight BT shunts plus RVOT patch, one RVOT patch) with four deaths (19%). Two more patients had a RVOT patch enlargement after the neonatal period. Repeated interventional catheterization was needed in 59% of the patients requiring surgery. The authors demonstrated the importance of further interventions, whether surgical or cardiological, required during the interim period.

RVOT Patch

Results of RVOT patch enlargement (Jahangiri *et al*., 1999) in patients without RV-dependent coronary circulation were excellent (no mortality in 25 patients). The authors emphasized the importance of patient selection for this procedure, which was based on coronary artery anatomy. Mainwaring *et al*. (1993) reported no operative mortality for RVOT enlargement in 14 patients with a well-developed RV without RV-dependent coronary circulation.

Definitive Repair

The initial morphology should direct the management from the beginning toward a biventricular repair or toward a Fontan circulation to avoid attrition in the interim interval. The overall survival has improved over recent years. Earlier reports from our own institution (de Leval *et al*., 1982; Bull *et al*., 1994) showed a 5 year survival of less than 50% with 25% of children determined to be suitable for definitive repair.

The latest multicentre study from the CHSS (Ashburn *et al*., 2004), prospectively enrolling 408 neonates from 33 institutions, reported a 5 year survival of 60% (79% for the later years) with 52% of children reaching a definitive repair. This study could demonstrate the prevalence of six end-points 15 years after entry in the study: for two-ventricle repair, 33%; Fontan circulation, 20%; 1.5-ventricle repair, 5%; heart transplant, 2%; death before reaching definitive repair: 38%; alive without definitive repair, 2%. Overall survival was 77% at 1 month, 70% at 6 months, 60% at 5 years and 58% at 15 years.

Among the risk factors for reaching definitive stages were RV size, tricuspid valve size and competence, birth weight, date of admission, role of institution, prior atrial balloon septostomy. This study estimated that with current management, definitive repair could be achieved in 85% of neonates within 3–5 years after admission. On the basis of factors predicting repair pathways, they expect that 50% of neonates will be suitable for a two-ventricle repair and 35% will be more appropriately managed on a Fontan pathway. Rychik *et al*. (1998) demonstrated that, using three different surgical strategies (two-ventricle repair, Fontan palliation or heart transplant) depending on morphology, they could achieve the same survival in the three patients groups (overall actuarial survivals at 1, 5 and 8 years were 82%, 76% and 76%, respectively).

The role of a one-and-a-half ventricle repair is more controversial. The advantage of providing a pulsatile pulmonary flow could be counterbalanced by a competition flow with the SVC flow through the bidirectional cavopulmonary connection. The behaviour of this type of palliation during exercise is unknown (Numata *et al*., 2003; Stellin *et al*., 2002).

Acknowledgements

Figure 28.1, the nomogram for determining the z value of the tricuspid valve from the measured diameter on the echocardiogram, is reprinted from Hanley FL,

Sade RM, Blackstone EH *et al*. Outcomes in neonatal pulmonary atresia with intact ventricular septum. *J Thorac Cardiovasc Surg* 1993; **105**: 406–423.

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29

Tetralogy of Fallot

E. L. Bove and J. C. Hirsch

The first complete description of tetralogy of Fallot is credited to the French physician Etienne Fallot, who published his findings in 1888 (Fallot, 1888). It was not until 1945, however, that the first surgical treatment for tetralogy of Fallot was performed by Blalock at Johns Hopkins University (Blalock and Taussig, 1945). A number of innovative systemic-to-pulmonary shunt procedures were soon developed, followed by the first successful intracardiac repair using human cross-circulation. The first successful repair using a pump oxygenator was performed by Kirklin at the Mayo Clinic 1 year later. Numerous contributions have been made in the management of this defect since these initial pioneering efforts, including one-stage vs. two-stage repair, trans-annular patching, conduit repairs and trans-atrial repair, to name a few.

The classic components of the ''tetrad'' that comprise this defect are a ventricular septal defect (VSD), right ventricular outflow tract obstruction, aortic override and right ventricular hypertrophy (**Figure 29.1**). All of these individual components result from one basic morphological abnormality: anterior and left-ward displacement of the infundibular septum (Van Praagh *et al*., 1970). The VSD in tetralogy of Fallot is a large, non-restrictive defect that results from the malalignment of the left-ward or septal extent of the infundibular septum with the septal band (trabecula septomarginalis). Because the left-ward extent of the infundibular septum is displaced anterior to the anterior limb of the septal band, rather than between its anterior and posterior limbs, a large malalignment VSD results (Suzuki *et al*., 1990). The superior border of the VSD is the anteriorly rotated right-ward (parietal) extent of the infundibular septum (IS = interventricular septum, VIF = ventriculo-infundibular fold). Posteriorly, it is bounded by the anteroseptal leaflet of the tricuspid valve. The inferior border is the posterior limb of the septal band, and the anterior border is the anterior limb of the septal band. The bundle of His penetrates at the posteroinferior edge of the defect (Dickinson *et al*., 1982; Kurosawa *et al*., 1988). Although the VSD is generally subaortic in position, it may extend to the subpulmonary region when the infundibular septum is absent or deficient (Anderson, 1981; Vargas *et al*., 1986) (**Figure 29.2**). Additional VSDs may exist in approximately 5% of patients and generally occur in the muscular septum.

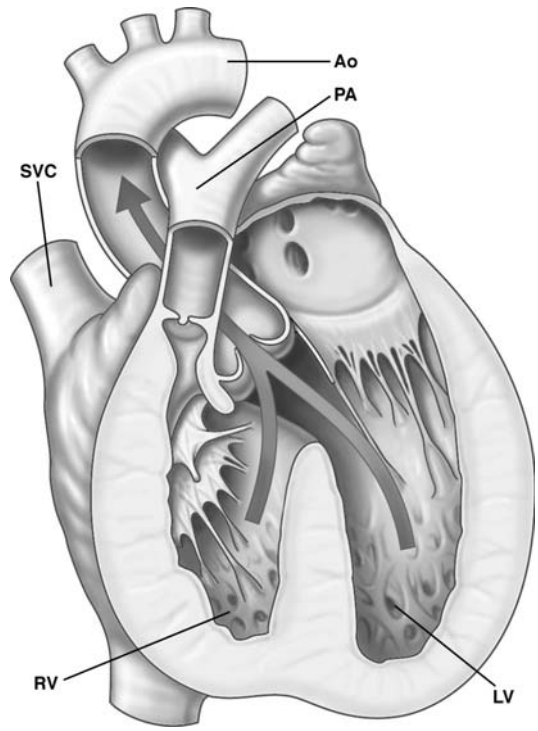

Figure 29.1

The anterior and left-ward displacement of the infundibular septum also results in right ventricular outflow tract obstruction from hypoplasia of the right ventricular infundibulum. Prominent muscle bands extend from the septal extent of the infundibular

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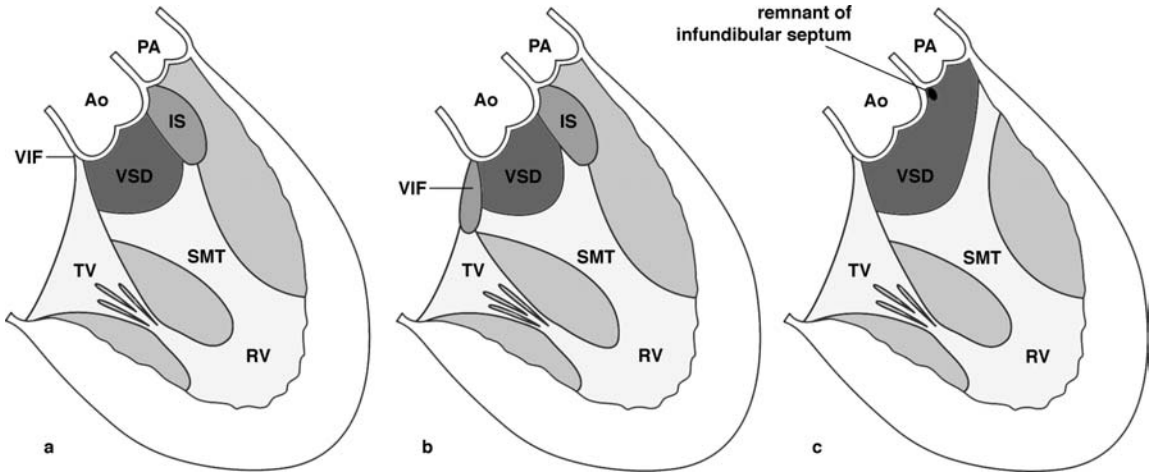

Figure 29.2

septum to the right ventricular free wall and contribute to the obstruction. A localized narrowing, or os infundibuli, frequently occurs at the inferior border of the infundibular septum. The pulmonary valve is nearly always involved in the obstruction. The leaflets are usually thickened and tethered to the pulmonary artery wall. The pulmonary valve is bicuspid in 58% of patients, but it is the narrowest part of the outflow tract in a small minority of patients (Altrichter *et al*., 1989; Shimazaki *et al*., 1992). The ''annulus'' of the pulmonary valve, although not a true fibrous structure, is nearly always smaller than normal. Important degrees of obstruction may also occur at the level of the right and left branch pulmonary arteries. Uncommonly, the left pulmonary artery may take origin from the ductus arteriosus and its intrapericardial portion may be completely absent. More commonly, localized narrowing of the origin of the right or left pulmonary arteries will be present and may be difficult to fully appreciate without special preoperative angiographic views. In extreme cases of anterior displacement of the infundibular septum, complete atresia of the distal right ventricular infundibulum and main pulmonary artery trunk may result.

Pulmonary atresia is present in approximately 7% of patients with tetralogy of Fallot (Chiariello *et al*., 1975). Multiple aortopulmonary collateral arteries (MAPCAs) are usually found in those patients without an associated patent ductus arteriosus and provide a variable degree of the pulmonary blood flow. Unlike patients without pulmonary atresia, who possess centrally located areas of discrete pulmonary arterial stenoses, those with pulmonary atresia and MAPCAs are more likely to have peripheral pulmonary stenoses (for details, see Chapter 30).

Approximately 5% of patients with tetralogy of Fallot will have complete absence of the pulmonary valve leaflets (Miller *et al*., 1962). Right ventricular outflow obstruction occurs at the level of the hypoplastic annulus and severe pulmonary regurgitation is generally present. Aneurysmal dilatation of the main pulmonary artery trunk and the right and left branch pulmonary arteries occurs and may result in compression of the distal tracheobronchial tree (Bove *et al*., 1972). This anomaly is discussed in detail in Chapter 31.

The origin of the left anterior descending coronary artery from the right coronary artery occurs in approximately 3–5% of patients with tetralogy of Fallot (Humes *et al*., 1987). The left anterior descending coronary artery will cross the right ventricular outflow tract a short distance below the pulmonary valve annulus to reach the anterior interventricular septum and is susceptible to injury from an incision in this area. Rarely, a single right coronary artery gives rise to the left main artery which then crosses the right ventricular outflow tract.

Major associated cardiac defects are relatively uncommon in tetralogy of Fallot. The most frequently associated lesions are atrial septal defect, patent ductus arteriosus, complete atrio-ventricular septal defect and multiple ventricular septal defects. Other less common defects include persistent left superior vena cava, anomalous origin of the left anterior descending coronary artery and aberrant origin of the right or left pulmonary artery.

DIAGNOSIS

The initial presentation of the patient with tetralogy of Fallot is dependent on the degree of right ventricular

outflow tract obstruction. Most commonly, cyanosis is mild at birth and gradually progresses with age as the obstruction increases due to increasing hypertrophy of the right ventricular infundibulum. Cyanosis tends to become significant within the first 6–12 months of life in these patients. In such situations, the obstruction is entirely or predominantly at the infundibular level. In addition, these patients may develop characteristic cyanotic ''spells'', which are periods of profound systemic hypoxaemia. These spells are characterized by a marked decrease in pulmonary blood flow and an increase in the right-to-left shunt across the VSD, directing desaturated blood into the aorta.

A smaller percentage of patients will present with significant cyanosis at or shortly after birth. In this group the outflow tract obstruction is nearly always due to a hypoplastic pulmonary valve annulus, with or without severe right ventricular infundibular obstruction or hypoplasia. Cyanosis is constant in these patients, due to the fixed nature of the obstruction to pulmonary blood flow. Patients with atresia of the pulmonary valve and main pulmonary trunk will be dependent on a patent ductus arteriosus or systemic aortopulmonary collateral arteries for pulmonary blood flow. In the latter situation, the collateral arteries may be such that pulmonary overcirculation exists and congestive heart failure is present (Ramsay *et al*., 1985; Shimazaki *et al*., 1988).

Cyanosis is the main physical finding. The characteristic systolic murmur results from the right ventricular outflow tract obstruction and is usually moderate in intensity. Typically the murmur disappears in the presence of a ''spell''. Continuous murmurs, best heard over the back and bilateral lung fields, will be heard in patients with a large amount of systemic aortopulmonary collateral artery flow.

The characteristic electrocardiographic finding is that of right ventricular hypertrophy from pressure overload of the right ventricle. Right axis deviation will also be found. Left ventricular hypertrophy may be seen in those patients with increased pulmonary blood flow from large shunts or collaterals.

On chest radiography, the heart size is generally normal and the pulmonary artery segment may be small. The aortic arch is right-sided in approximately 25% of patients. The characteristic ''boot-shaped'' heart results from elevation of the cardiac apex from the hypertrophied right ventricle and a concave upper left heart border caused by a narrow main pulmonary artery.

The diagnosis is usually easily established by echocardiography (McConnell, 1990). The typical malalignment VSD with aortic override and right ventricular outflow tract obstruction is well visualized. Often the location of the left anterior descending coronary artery can be visualized as well. The peripheral pulmonary artery anatomy is poorly seen, however, and delineation of pulmonary artery anomalies constitutes one of the main indications for cardiac catheterization. The presence and precise anatomy of any significant aortopulmonary collateral arteries are determined by aortography. Branch right and/or left pulmonary artery stenoses, as well as non-confluent pulmonary arteries, must be sought by special cranial tilt views. Crossing the right ventricular outflow tract with the catheter may precipitate a spell and is generally not advised. In patients with long-standing cyanosis and those with pulmonary overcirculation, measurement of the pulmonary artery pressure and pulmonary vascular resistance may also be required to exclude fixed pulmonary hypertension, which might preclude repair.

INDICATIONS FOR OPERATION

Most patients with tetralogy of Fallot have satisfactory systemic arterial oxygen saturation at birth and require no treatment. Progression of hypoxaemia will ultimately occur, and when the oxygen saturation falls below 75–80% operative intervention should be performed. Hypoxaemic spells may occur from transient reductions in pulmonary blood flow, due to a sudden increase in right ventricular outflow tract obstruction and a decrease in systemic vascular resistance. The occurrence of hypoxaemic spells is also generally considered an indication for operation, although in some cases medical management with propranolol may be used to delay operation. In those patients where specific indications have not yet developed, elective complete repair is recommended in most institutions by 1 year of age and, in our own institution, preferably by 3–6 months of age (Castaneda *et al*., 1990) (**Table 29.1**). Single-stage complete repair is being done in many centres, regardless of age (Di Donato *et al*, 1991; Gustafson *et al*., 1988) but an initial systemic-to-pulmonary artery shunt procedure is preferred by others (Karl *et al*., 1992), particularly when symptoms occur within the first 6 months of life. The presence of pulmonary atresia, branch pulmonary artery hypoplasia, anomalous origin of the anterior descending coronary artery from the right coronary artery, or severe associated non-cardiac

Table 29.1 Benefits of early complete repair.

Promote normal growth and development of organs
Eliminate hypoxaemia
Less need for extensive right ventricular muscle excision
Better late left ventricular function
Decreased incidence of late dysrhythmias

anomalies are other generally accepted indications for a shunt rather than primary repair in the infant at several institutions. Many other institutions, however, would still prefer primary repair, even when repair must be accomplished with the insertion of a conduit between the right ventricle and the pulmonary arteries. The need for a trans-annular patch because of significant hypoplasia of the pulmonary valve annulus was formerly considered a contraindication to complete repair in the infant, but this risk has now been neutralized (Kirklin *et al*., 1992; Hennein *et al*., 1995).

When an initial systemic-to-pulmonary shunt procedure is chosen as part of a staged repair, the classic or modified form of the Blalock–Taussig procedure is most commonly selected (Ullom *et al*., 1987). The classic Blalock–Taussig procedure is performed on the side opposite the aortic arch (ipsilateral to the innominate artery) in order to allow the most favourable angle for the subclavian artery to reach the pulmonary artery without kinking. Although this anastomosis can be satisfactorily constructed in patients of any age or size (Guyton *et al*., 1983) many surgeons currently prefer the modified shunt in very small neonates because of the small size of the subclavian artery (de Leval *et al*., 1981). In the modified procedure, an interposition polytetrafluoroethylene conduit is placed between the subclavian and pulmonary arteries (Di Benedetto *et al*., 1981). A 4 mm graft is generally preferred in neonates because early complete repair may be performed and larger shunts may result in congestive heart failure. The results achieved by this procedure have been excellent, with an extremely low shunt failure rate and an acceptable duration of palliation (Bove *et al*., 1987; McKay *et al*., 1980).

In the presence of severe hypoplasia of the pulmonary arteries, palliation with a right ventricular outflow tract patch or conduit, leaving the VSD open, may be used (Okita *et al*., 1990). This promotes symmetrical pulmonary artery blood flow and reduces the likelihood of branch pulmonary artery distortion from a shunt procedure. However, in most patients with tetralogy of Fallot and pulmonary stenosis, the hypoplasia of the pulmonary arteries is due to the lack of pulmonary artery flow itself and responds promptly to procedures that increase this flow. Therefore, leaving the VSD open may result in severe congestive heart failure and pulmonary oedema from a sudden increase in pulmonary blood flow. Branch left pulmonary artery stenosis is well known to develop in some patients after repair and should be looked for during a routine follow-up. This is more likely to occur when repair is performed in the neonate, as the presence of a patent ductus arteriosus can mask the origin of the left pulmonary artery, which may become stenotic after the ductal tissue involutes.

Palliation of the small or unstable neonate with catheter-based approaches has been employed at some centres. Balloon dilatation of the pulmonary valve and stenting of ductus arteriosus can be utilized for stabilization prior to definitive operative intervention or in place of a systemic-to-pulmonary artery shunt (Gibbs *et al*., 1992). This approach, however, has not been widely accepted or utilized at this time.

SURGICAL TECHNIQUE

Anaesthesia

Monitoring lines include a femoral or radial arterial catheter and a central venous catheter (femoral or SVC). Additional peripheral intravenous lines and bladder catheter are placed. Nasal, cutaneous, and rectal temperature probes are utilized. Standard cardiac anaesthesia with high-dose narcotics and paralysis is employed. Care is taken to avoid significant changes in systemic and pulmonary vascular resistance that may precipitate a ''spell'' or increase the right-to-left (R–L) shunt.

Should important hypoxaemia occur, α-agonist drugs are recommended to increase systemic vascular resistance and minimize the R–L shunt. A transoesophageal echocardiogram probe is placed after anaesthesia is induced. Additional intracardiac lines are placed at the termination of the procedure. Details of anaesthesia are discussed in Chapter 12.

Surgical Technique

The techniques of creating systemic-to-pulmonary shunts are discussed in Chapter 16. For repair, a midline sternotomy incision is performed and the heart is exposed. The precise distribution of the coronary artery branches is confirmed and preparation is made for cardiopulmonary bypass. Little manipulation of the heart is done in order to avoid precipitating severe hypoxaemia from a ''spell''. Existing systemic-to-pulmonary artery shunts are exposed for subsequent interruption, although they may be dissected after cardiopulmonary bypass is established if the exposure is particularly difficult. Heparin is administered (300 U/kg). The aorta is cannulated just proximal to the level of the innominate artery and each vena cava is directly cannulated with small right-angled metal-tipped cannulae (DLP, Grand Rapids, MI). Alternatively, a single cannula may be placed in the right atrium if the repair is to be performed using deep hypothermia and circulatory arrest. The latter approach may be utilized in patients $< 2.0–2.5$ kg in weight. In this situation, the VSD is closed under a brief period of circulatory arrest and the outflow tract repair is performed on cardiopulmonary bypass. When repair

is done using continuous cardiopulmonary bypass, the systemic temperature is lowered to 25–28°C and the left ventricle is vented through the right superior pulmonary vein. All shunts are ligated and/or divided, the main pulmonary trunk and bifurcation (if branch stenoses are present) are mobilized, and the ductus arteriosus is ligated. After the aortic cross-clamp is applied, the cardioplegic solution (30 ml/kg) is administered. At the University of Michigan, we utilize a dilute blood solution with added potassium to a concentration of 25 meq/l. Repeat doses (15 ml/kg) are administered at 20–30 minute intervals. The ASD and VSD are closed and the RVOTO is alleviated. The heart is de-aired via the left ventricular vent and the aortic air needle. The cross-clamp is removed and the trans-annular patch or RV–PA conduit, as necessary, are completed with the heart beating. A patent foramen ovale may be left open in neonates, particularly when post-repair haemodynamic residua remain, including pulmonary insufficiency and elevated right ventricular pressure. Because the neonate tolerates volume overload poorly, due to a non-compliant right ventricle and elevated pulmonary vascular resistance, oxygen delivery can be maintained by allowing a limited R–L atrial shunt at the expense of mild to moderate cyanosis. This is not necessary in older patients.

In order to achieve a satisfactory repair, the surgeon must be completely familiar with all details of the anatomy, including the size and distribution of the branch pulmonary arteries, the nature and size of the pulmonary valve annulus (junction between the right ventricle and main pulmonary trunk), extent of the right ventricular outflow tract obstruction, coronary artery distribution, anatomy of the VSD, and the presence of any associated defects. After aortic cross-clamping, a right atriotomy is made to assess the anatomy (**Figure 29.3**). Stay stitches are placed on the anterior and septal leaflet of the tricuspid valve. Gentle traction on these stitches will facilitate the exposure. The dotted line indicates the position of the VSD (TV, tricuspid valve; SVC, superior vena cava; CS, coronary sinus). If an atrial septal defect or patent foramen ovale is present, it is closed at this time. The anatomy of the VSD and right ventricular outflow tract obstruction is viewed through the tricuspid valve (**Figure 29.4**). A retractor placed anteriorly through the tricuspid valve and the stay stitches in the septal and anterior leaflet aid in the exposure of the distal outflow tract. When the repair is accomplished entirely through the right atrial approach, as is preferred in our institution, the outflow tract obstruction is approached first. A traction suture placed in the anterior and septal leaflets of the tricuspid valve aids exposure of the distal infundibulum. The position of the anterior margin of the VSD and the aortic valve leaflets are noted, and

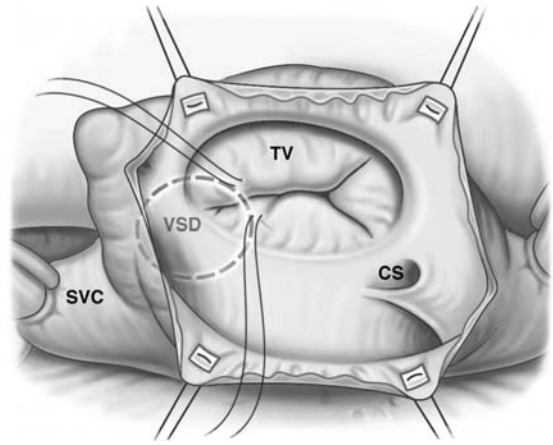

Figure 29.3

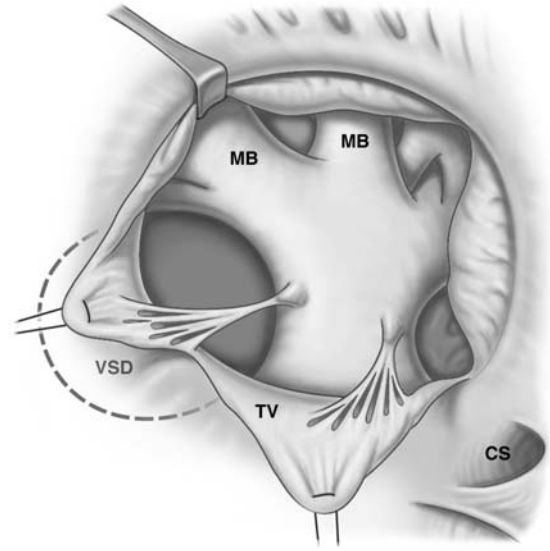

Figure 29.4

the parietal extent of the anterosuperiorly malpositioned infundibular septum is visualized (Pacifico *et al*., 1987; Binet, 1988). Invaginating the right ventricular free wall with a finger placed from outside the heart facilitates this exposure. Muscle trabeculations along the anterior limb of the septal band (trabecula septomarginalis) are divided on the right-angled dissector down to the level of the moderator band if necessary (**Figure 29.5**). When repair is performed in infancy, excision of the parietal extent of the infundibular septum (trabecula parietomarginalis) is rarely necessary and simple division of the obstructing muscle bundles is all that is required (**Figure 29.6**). Note

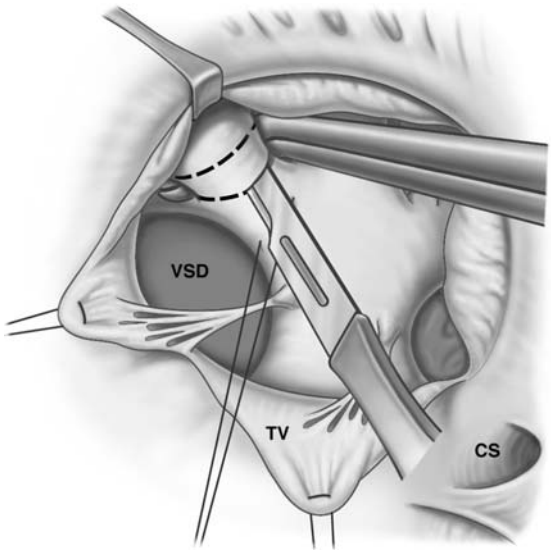

Figure 29.5

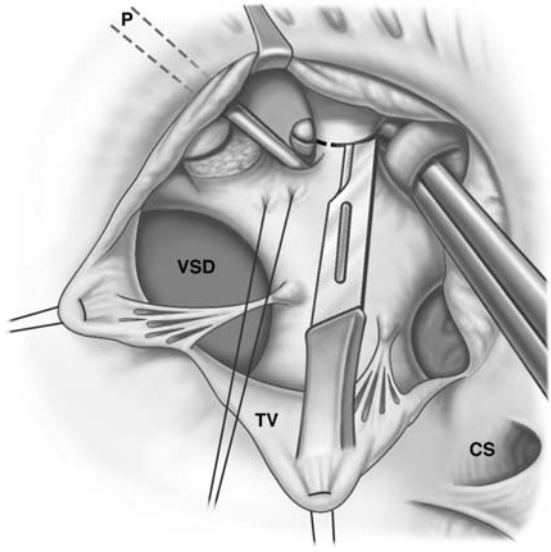

Figure 29.6

the probe passed (P) from the pulmonary artery through the valve to the ventricle. A pulmonary valvotomy can now be performed through the right atrial approach. If exposure is not adequate, a vertical incision is made in the main pulmonary artery, through which a pulmonary valvotomy may be performed (Pacifico *et al*., 1990; Coles *et al*., 1988). Valve leaflets may be mobilized and fused commissures divided all the way to the pulmonary artery wall. At this time, an assessment of the diameter of the pulmonary valve annulus is made by inserting calibrated dilators across the right ventricular outflow tract into the pulmonary artery trunk. The decision to place a trans-annular patch is made if the estimated post-repair RV/LV pressure is predicted to exceed 0.7 (Naito *et al*., 1980). In this situation the main pulmonary artery incision is extended onto the right ventricular outflow tract across the pulmonary valve annulus. It can be kept quite short, extending only a few millimetres proximal to the annulus, as the infundibular obstruction has been adequately relieved trans-atrially. Whenever possible, this incision is placed directly through the anterior commissure of the valve to allow the pulmonary valve leaflets to remain functional and decrease the amount of pulmonary regurgitation. In some centres, the use of a monocusp outflow tract patch is preferred to prevent, or at least minimize, pulmonary regurgitation. Monocusp pulmonary valves, however, will only provide competence for a relatively short period of time, but may be particularly useful with associated pulmonary artery hypoplasia, distal pulmonary obstruction or decreased ventricular function. Care needs to be taken when placing a right ventriculotomy that an aberrant origin of the LAD from the RCA is not injured. It is often possible to limit the incision in such a way that it remains superior to an anomalous LAD when a trans-atrial repair is done (hockey stick incision). A monocusp valve constructed of Gore-Tex, pericardium or homograft tissue may be placed in the outflow tract if pulmonary insufficiency is to be avoided. However, these valves provide only short-term competence and require a longer incision in the right ventricular infundibulum. In certain conditions, including hypoplastic branch pulmonary arteries, distal stenoses and poor ventricular function, the early pulmonary valve competence may confer an important haemodynamic advantage and improve the postoperative course.

Closure of the VSD is accomplished from the trans-atrial approach, regardless of whether or not a trans-annular patch is needed, as that allows the ventricular extent of the incision to be minimized to the length necessary only for relief of obstruction, not for VSD exposure (Kavey *et al*., 1987). Visualization of the VSD is generally adequate through the tricuspid valve and is even easier after dividing obstructing muscle bundles. A patch of polytetrafluoroethylene (Gore-Tex) is cut to the appropriate size and sutured to the right side of the septum, utilizing a continuous suture technique (**Figure 29.7**). Suturing is commenced at the angle between the anterior and posterior limbs of the septal band (trabecula septomarginalis), directly opposite the perimembranous rim, and begun superiorly over the infundibular septum and aortic valve. The sutures are

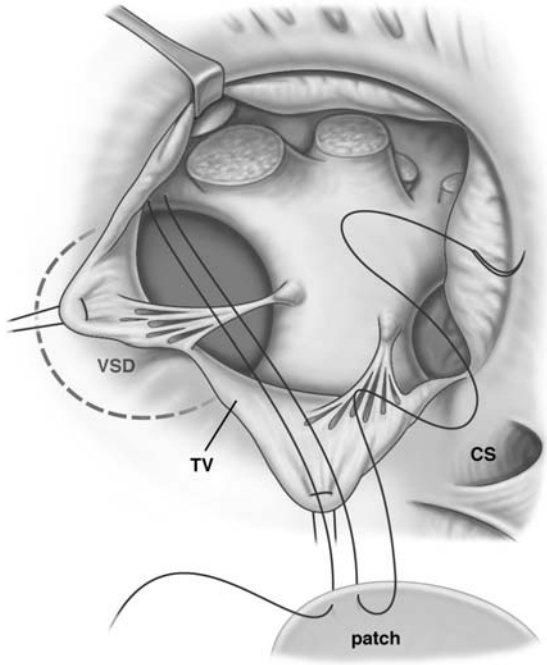

Figure 29.7

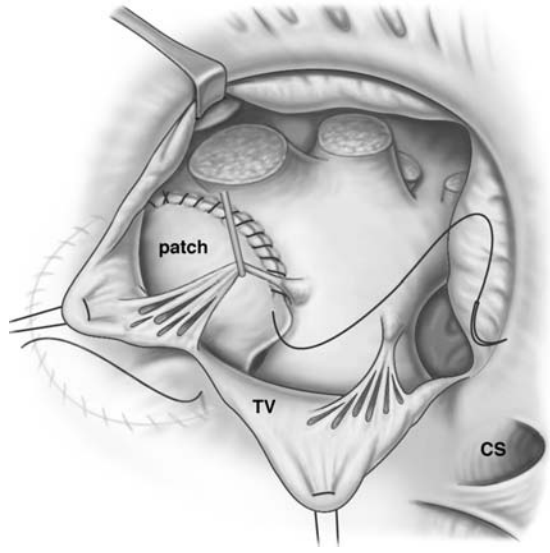

Figure 29.8

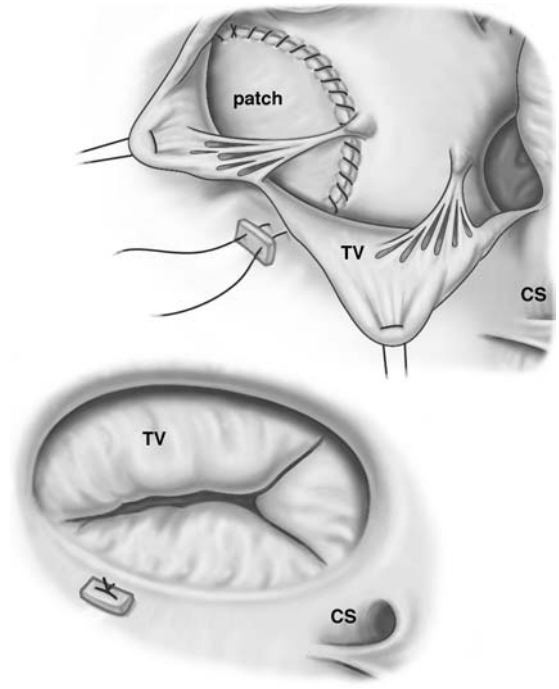

Figure 29.9

kept close to the aortic valve annulus itself in order to avoid residual defects in muscle trabeculations. This initial arm of the suture is brought into the right atrium by passing the needle through the anterior leaflet of the tricuspid valve at its junction with the ventriculo-infundibular fold (**Figure 29.8**). The other needle is then brought inferiorly, past the medial papillary muscle and under any chordae tendinae from the septal leaflet of the tricuspid valve, until the posteroinferior rim of the defect is reached. At this point, suturing must be done approximately 5 mm away from the crest of the VSD itself and only on the right ventricular side. This is done in order not to injure the bundle of His, which penetrates the floor of the atrial septum in the apex of the triangle of Koch and runs adjacent to this margin of the VSD (see Chapter 8). Attaching the patch to the septal leaflet of the tricuspid valve away from the penetrating bundle completes suturing. The suture is tied over a pledglet on the right atrial side (**Figure 29.9**).

Although the trans-atrial approach can be accomplished in the majority of patients undergoing repair of tetralogy of Fallot, even in infants, it is not suitable when the right ventricular outflow tract is diffusely hypoplastic (McGrath *et al*., 1988). In these situations, repair is best done with an outflow tract patch to enlarge the diameter of the infundibulum (**Figure 29.10**). Severe hypoplasia of the pulmonary valve annulus and main pulmonary trunk are common in this situation, and the outflow patch is then carried across the valve onto the pulmonary artery. It may be necessary to extend the patch, or tailor a separate patch, onto the left pulmonary artery in order

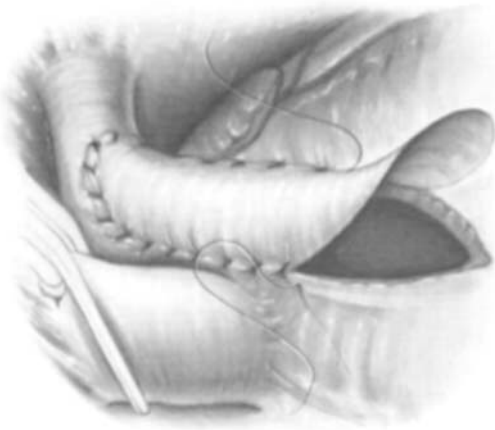

Figure 29.10

to ensure that the obstruction is sufficiently relieved, because the main pulmonary trunk and origin of the left (or right) pulmonary artery branch are frequently hypoplastic. When an outflow patch is required, either for relief of severe infundibular and annular obstruction or for isolated enlargement of the pulmonary valve annulus alone (when the repair has been accomplished trans-atrially), it should be kept to the shortest length possible on the right ventricular aspect in order to avoid damaging right ventricular function (Kavey *et al*., 1987; Miura *et al*., 1992).

Upon completion of systemic re-warming, cardiopulmonary bypass is discontinued in the usual fashion and the haemodynamics are assessed for important residual lesions. The peak RV/LV pressure ratio is measured to ensure that significant residual outflow tract obstruction does not exist. If the post-repair RV/LV pressure is in excess of 0.7 and a trans-annular patch has not been placed, bypass is resumed and a patch is inserted across the pulmonary valve annulus. If a trans-annular patch has been placed, other causes of persistent elevation of right ventricular pressure must be considered, including branch pulmonary artery stenoses, hypoplastic peripheral pulmonary arteries, residual VSD or residual infundibular obstruction. In the absence of these conditions, it may be assumed that the right ventricular hypertension will be well tolerated and will, in fact, improve over the next 24–48 hours (Goor *et al*., 1981). Often this elevation in right ventricular pressure results from dynamic right ventricular outflow tract obstruction, particularly when an outflow patch is avoided, as in the case of a trans-atrial repair. Administration of an ultra-short acting β-blocking agent, such as Esmolol, can help to differentiate dynamic vs. fixed residual right ventricular outflow tract obstruction intraoperatively (Wensley *et al*., 1987).

SPECIAL CIRCUMSTANCES

Pulmonary Artery Abnormalities

Stenosis of the origin of the left and/or right pulmonary arteries is frequently encountered in patients with tetralogy of Fallot. Left pulmonary artery stenosis is best augmented with placement of a separate patch. Simple extension of the right ventricular outflow patch onto the left pulmonary artery can cause flow disturbances, as well as distortion of the left pulmonary artery take-off, due to the acute posterior course which the vessel follows (Hennein *et al*., 1995). Stenosis at the origin of the right pulmonary artery is more difficult to repair, because of the right angle that this vessel takes from the main pulmonary artery and the more difficult exposure resulting from the overlying ascending aorta. In this situation, a separate patch may be necessary to enlarge the proximal right pulmonary artery. Alternatively, resection of the stenotic area, if it is relatively localized, with end-to-end anastomosis using absorbable suture, may also give good results. Bifurcation stenoses involving both pulmonary artery origins may be repaired with a resection and end-to-end anastomoses of both pulmonary arteries. Alternatively, each branch of a bifurcated pulmonary artery allograft may be anastomosed to the distal pulmonary arteries beyond their stenoses. Less commonly, one of the branch pulmonary arteries may have an anomalous systemic arterial origin. Usually it is the left pulmonary artery that arises from a normally positioned ductus arteriosus and proceeds directly to the hilum of the lung without entering the pericardium (Shanley *et al*., 1993). Exposure of the left pulmonary artery is best accomplished by first dissecting under the arch of the aorta to isolate the ductus. The pulmonary artery can then be followed toward the hilum to gain sufficient length for primary anastomosis to the side of the main pulmonary artery trunk. When the right pulmonary artery is not in continuity with the left, it is likely to originate from a right-sided ductus off the innominate artery. Rarely, it may take origin from the descending aorta. When the branch pulmonary arteries are non-confluent, or confluent but with a significant stenosis centrally (usually at the insertion of the ductus itself), palliation with a systemic-to-pulmonary artery shunt is a poor choice of treatment. In these cases the shunt flow will be predominantly to one lung, while the other receives little or no flow. Reconstruction of the central pulmonary artery bifurcation using one or more of the techniques outlined above is preferred in order to provide symmetric pulmonary blood flow. This is best

combined with complete intracardiac repair, although placing a shunt to the reconstructed pulmonary arteries may also be done (Shanley *et al*., 1993).

Pulmonary Atresia with Multiple Aortopulmonary Collateral Arteries (MAPCAs)

This challenging group of patients is covered in more detail in Chapter 30. In brief, the preferred initial approach at the University of Michigan includes placement of a right ventricle-to-pulmonary artery conduit or outflow patch by 3–6 months of age to improve pulmonary blood flow and to stimulate growth of the central pulmonary arteries. Unifocalization is delayed unless MAPCAs are clearly demonstrated to provide the sole blood supply to major areas of the lung or are severely stenotic and at risk of thrombosis. Initial RV–PA conduit placement will allow better evaluation of dual blood supply from MAPCAs at subsequent catheterization and will minimize or eliminate the need for unifocalization in many patients.

Tetralogy of Fallot with Complete Atrio-ventricular Septal Defect

Complete atrio-ventricular septal defect (CAVSD) occurs in approximately 2% of patients with tetralogy of Fallot and is more common in patients with Down's syndrome. The anatomy is that of the typical CAVSD, although the left anterior bridging leaflet is always undivided and unattached to the crest of the ventricular septum (Rastelli type ''C''). There is anterosuperior displacement of the infundibular septum, such that the VSD has a large outlet component in addition to the inlet portion associated with the CAVSD. The aortic valve overrides the outlet component of the VSD. The right ventricular outflow tract obstruction is the same as that for isolated tetralogy of Fallot. The repair is complicated by the increased difficulty of placing an intraventricular patch without causing left ventricular outflow tract obstruction or atrio-ventricular valve regurgitation. Marked overriding of the aortic valve makes this more difficult, as exposure is not easily obtained via the atrial approach. The need for a valve in the pulmonary position when a trans-annular patch is required has also been debated. It appears to be unnecessary unless significant pulmonary artery branch hypoplasia is present. This combination of defects is also discussed in Chapter 27.

INTENSIVE CARE UNIT MANAGEMENT

The postoperative care of the patient with tetralogy of Fallot can be challenging. A right atrial pressure line is routinely placed for monitoring of volume status and as a guide to right ventricular function. Occasionally, particularly when left ventricular function is compromised, the addition of a left atrial monitoring line is also useful. Significant volume resuscitation is often necessary to provide adequate filling of the hypertrophied and non-compliant right ventricle. Inotropes are instituted to maintain adequate cardiac output and perfusion pressure once filling pressures are optimized. A patent foramen ovale, if present, is generally left open in the neonate to allow for R–L shunting, to maintain cardiac output at the expense of mild systemic arterial desaturation in the early postoperative period. Atrial and ventricular pacing wires are placed for the diagnosis and management of postoperative arrhythmias. Junctional ectopic tachycardia (JET) is a relatively common postoperative arrhythmia and is treated with surface cooling, overdrive pacing and intravenous amiodarone administration.

In addition to arrhythmias, important postoperative complications include tamponade, residual outflow tract obstruction, residual VSD, tricuspid insufficiency, and phrenic nerve palsy. Even small residual VSDs are poorly tolerated by a left ventricle not accustomed to volume overload, and tamponade may cause haemodynamic compromise early in the presence of a dilated right ventricle, as seen with pulmonary insufficiency. Tricuspid insufficiency may occur secondary to the sutures used for VSD patch placement and is poorly tolerated when pulmonary insufficiency and elevated right ventricular pressure coexist.

RESULTS

The early (hospital) mortality after repair of tetralogy of Fallot is currently 1–5% in most reported series (Groh *et al*., 1991; Castaneda, 1990; Gustafson *et al*., 1988; Karl *et al*., 1992). A review of 399 patients undergoing complete repair for tetralogy of Fallot (excluding repairs performed concomitant with or subsequent to unifocalization of MAPCAs) at the University of Michigan between January 1993 and November 2004 is depicted in **Table 29.2**. The overall early hospital mortality rate for complete repair of tetralogy of Fallot was 3% (95% confidence interval 1.2–4.6%). Repair within the first year of life did not influence the early outcome, as had been reported in a number of earlier series. The neutralization of this risk factor is largely due to improved intraoperative techniques (particularly the avoidance of excessive right ventricular outflow tract muscle excision), cardiopulmonary bypass management and refinements in postoperative care.

Among 814 patients undergoing complete repair at the University of Alabama at Birmingham, survivorship at 1 month and at 1, 5 and 20 years was 93%, 92%, 92% and 87%, respectively (Kirklin *et al*., 1989). Survival

Table 29.2 Hospital survival after complete repair for tetralogy of Fallot, University of Michigan,* January 1993–November 2004 ($n = 399$).

	Total (n)	Alive n (%)	95% CI (%)
TOF (All)			
< 1 month	61	60 (98)	91.2–100
> 1 month < 1 year	266	260 (98)	95.2–99.2
> 1 year	72	69 (96)	88.3–99.1
Total	399	389 (97)	95.4–98.8
TOF with PS			
< 1 month	37	36 (97)	85.8–99.9
> 1 month < 1 year	236	231 (98)	95.1–99.3
> 1 year	55	52 (96)	84.9–98.9
Total	328	319 (97)	94.9–98.7
TOF with PA			
< 1 month	24	24 (100)	N/A
> 1 month < 1 year	15	14 (93)	68.1–99.8
> 1 year	9	9 (100)	N/A
Total	48	47 (98)	88.9–100
TOF with CAVSD			
< 1 month	0	0	N/A
> 1 month < 1 year	15	15 (100)	N/A
> 1 year	8	8 (100)	N/A
Total	23	23 (100)	N/A

*Excluding repairs performed concomitant with or subsequent to unifocalization procedures for MAPCAs.
95% CI, 95% confidence interval; TOF, tetralogy of Fallot; PS, pulmonary stenosis; PA, pulmonary atresia; CAVSD, complete atrio-ventricular septal defect.

was slightly less than that for an age-, race- and gender-matched control population. In their analysis, the risk factors for late death were older age at repair, high peak right/left ventricular pressure immediately after repair (> 0.85) and the presence of a Potts shunt. The use of a trans-annular patch was not found to be a risk factor for premature late death.

LATE COMPLICATIONS

The most common indications for reoperation are the result of long-term complications of the right ventricular outflow tract, such as severe pulmonary regurgitation, residual outflow tract obstruction or conduit failure (Oechslin *et al*., 1999). Less commonly, reoperation is necessary for a residual VSD. Even relatively small residual defects are poorly tolerated after tetralogy repair, and reoperation is recommended when the pulmonary:systemic flow ratio exceeds 1.5. The most common location for a residual VSD is at the posteroinferior margin of the patch, presumably because suturing in this area is done superficially to avoid heart block. Additional reasons for reoperation include pulmonary valve stenosis, left pulmonary artery stenosis and aortic valve insufficiency. The exact amount of residual outflow tract obstruction required for reoperation is controversial, but when right ventricular pressure exceeds 60 mmHg, relief of residual obstruction is generally indicated.

An earlier review from the University of Michigan of tetralogy of Fallot patients undergoing complete repair in the first month of life demonstrated a 1 month, 1 year and 5 year freedom from reoperation rates of 100%, 93% and 63% (Hennein *et al*., 1995). The relatively high reoperation rate in this age group, however, was influenced by the complex anatomy associated with symptomatic neonates with tetralogy of Fallot, including a high incidence of non-confluent pulmonary arteries, branch pulmonary artery stenoses and conduit insertion. An intraoperative pressure ratio between the right and left ventricles, at the time of initial repair, of 0.75 or greater is an independent risk factor for reoperation. The precise surgical intervention depends on the nature and location of the obstruction, but a careful search for branch pulmonary stenosis should be done. When the obstruction is located at the annular level, a trans-annular patch should be inserted. Overall survival and functional status following reoperation is very good, with a 10 year actuarial survival of 92%, with 93% of these patients in a New York Heart Association classification of I or II (Oechslin *et al*., 1999). When significant residual pulmonary artery branch stenoses are diagnosed late after repair, treatment by balloon dilatation and/or stent insertion can be very effective.

Although the effect of pulmonary insufficiency on early and late mortality has been neutralized in recent series, long-standing pulmonary insufficiency appears to have certain deleterious effects on ventricular function and exercise capacity. This is particularly true when additional residual lesions are present, such as a VSD or peripheral pulmonary stenosis (Finck *et al*., 1988). In a series of 20 patients with isolated pulmonary insufficiency studied an average of 9 years after repair, we noted significantly lower right and left ventricular ejection fractions when compared with a group of patients with competent pulmonary valves (Bove *et al*., 1983). This was found despite the fact that all the patients analysed in this study were asymptomatic and had excellent haemodynamic repairs. Thus, it is likely that some patients with pulmonary insufficiency will develop symptoms many years after repair, even in the absence of other significant residual defects (Shimazaki *et al*., 1984). When symptoms develop, the insertion of a pulmonary valve has been shown by a number of groups

to improve functional status and ventricular function (Chandar *et al*., 1990; Ilbawi *et al*., 1987; Bove *et al*., 1985). Following reoperation, a statistically significant drop in RV:LV pressure ratio has been demonstrated, along with a reduction by at least one NYHA classification grade. The indications for pulmonary valve replacement in the absence of symptoms of exercise intolerance or congestive heart failure are not well defined, but this procedure should be considered in the presence of poor ventricular function, tricuspid insufficiency and progressive right ventricular dilatation. Early reoperation for asymptomatic right ventricular dysfunction improves the chance for full recovery of ventricular function, as well as decreasing the prevalence of ventricular arrhythmias (Ilbawi *et al*., 1987). A review of patients undergoing elective pulmonary valve replacement has demonstrated that the operative risk was low (1.1%). In addition, functional status was NYHA I for 90% of patients following repair, with a 10 year survival of 95% (Yemets *et al*., 1997).

Interest has again developed in the use of monocusp valves in the right ventricular outflow tract. The objective is to minimize pulmonary regurgitation and thereby reduce the long-term effects on the right ventricle. Short term data has failed to consistently demonstrate improved postoperative recovery in patients with monocusps vs. those with standard trans-annular patches, particularly when the trans-annular patch is kept short and right ventricular muscle excision is avoided (Bigras *et al*., 1996). Additionally, longer-term follow-up has shown a failure of monocusps to maintain satisfactory function after a few months.

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30

Pulmonary Atresia and Ventricular Septal Defect

W. J. Brawn and M. R. de Leval

In pulmonary atresia there is absence of functional continuity between the right ventricle and the pulmonary arteries. The blood supply to the lungs most commonly comes from the ductus arteriosus and in a small number of cases from the aorta as systemic collaterals or major aortopulmonary collateral arteries (MAPCAs). The origin and development of the pulmonary artery tree and the MAPCAs has caused much debate.

Boyden (1970), showed that in early foetal development, the vascular plexus forming in the lung buds is connected to segmental arteries from the dorsal aorta. At the same time the lung is also perfused by the sixth aortic arch (the central pulmonary arteries). When, for whatever reason, normal development is arrested, these segmental arteries from the aorta may persist as MAPCAs (Macartney *et al*., 1973). However, more recent studies in the normal human embryonic and foetal lungs have shown that the peripheral pulmonary arteries are fed from the heart at 38 days of gestation and that there was no evidence of connections to the developing lung buds from the dorsal aorta (Hall *et al*., 2000); thus the embryonic origin of the MAPCAs is to be regarded as speculative until the normal development of the pulmonary vascular tree is more clearly understood (Rossi *et al*., 2002).

The intracardiac anatomy of patients with pulmonary atresia and VSD is similar to that of tetralogy of Fallot. Rarely patients with pulmonary atresia, VSD and MAPCAs can have congenitally corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum and univentricular hearts.

There is also a strong association between chromosome 22q11 microdeletion and pulmonary atresia with VSD and MAPCAS, occurring in at least 40% of patients (Hofbeck *et al*., 1998; Chessa *et al*., 1998).

GENERAL CONSIDERATIONS

The size and distribution of the intrapericardial and intrapulmonary arteries together with their blood supply from a ductus arteriosus or MAPCAs, determine the surgical management options for each patient. In addition their clinical condition, age, weight and previous surgical history has to be taken into account. The patients can generally be divided into three main groups. The first and by far the commonest group are patients with normal-sized intra pericardial confluent pulmonary arteries, normally distributing to both lung fields. The second, quite rare, group are those patients with moderate hypoplasia of intrapericardial pulmonary arteries but with normal distribution to both lung fields. In these first two groups the pulmonary vessels are supplied by a ductus arteriosus. The third and the smallest group, about 25% of patients with pulmonary atresia and VSD (Puga *et al*., 1989), usually have diminutive or absent intrapericardial pulmonary arteries with marked arborization abnormalities in the lungs fields. It is this group of patients with pulmonary atresia and ventricular septal defect that is dependent on MAPCA blood supply to the lungs, which remains a challenge (**Figure 30.1**).

MORPHOLOGY OF THE INTRA PERICARDIAL AND INTRA-PULMONARY ARTERIES

The variations in the pulmonary artery morphology seen in pulmonary atresia are diagrammatically illustrated in **Figure 30.1**, which shows that the intra pericardial pulmonary arteries are of normal size, supplied by a ductus arteriosus and connected by the main pulmonary artery to the right ventricular infundibulum. The pulmonary valve is atretic. **Figure 30.1a** the separation of the main pulmonary artery to right

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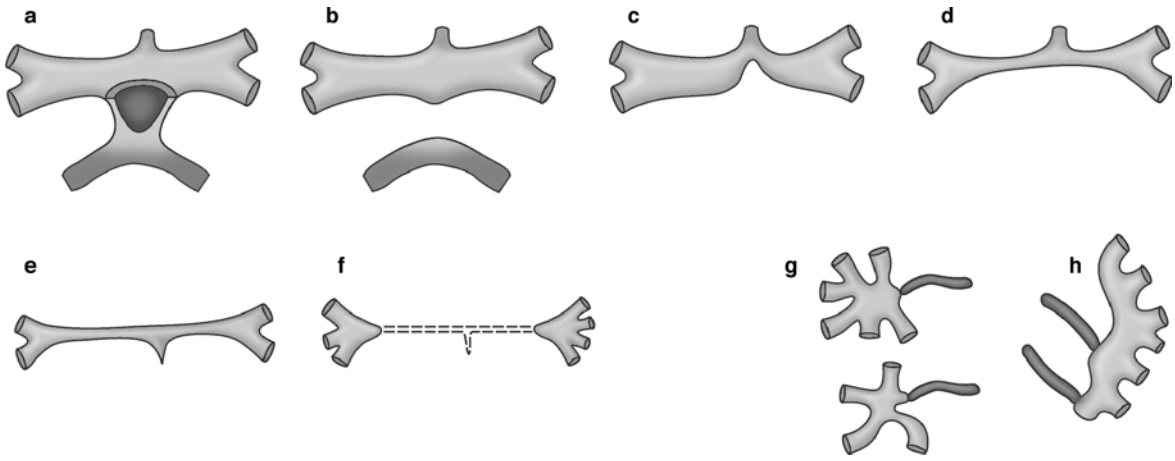


Figure 30.1

ventricular infundibulum is illustrated in **Figure 30.1b**. In **Figure 30.1c** the intrapericardial pulmonary arteries are supplied by a ductus arteriosus. There is absence of the main pulmonary artery and the right ventricular infundibulum. The ductus arteriosus supplies confluent intrapericardial pulmonary arteries; but there is a stenosis of the pulmonary arteries at the point of the entry of the ductus. This anatomy may be associated with or without the presence of a main pulmonary artery remnant attached to the infundibulum. **Figures 30.1d and 30.1e** show generalized hypoplasia of the intrapericardial pulmonary arteries with the absence of a main pulmonary artery. **Figure 30.1f** shows just a fibrous cord connecting both the left and right pulmonary arteries at the hilar region. These fibrous cords may pull in the hilar pulmonary arteries towards the midline. In **Figures 30.1g and 30.1h** there is absence of the intrapericardial pulmonary arteries with a MAPCA supply to non confluent intrapulmonary arteries as shown in **Figure 30.1g** and confluent intrapulmonary arteries as shown in **Figure 30.1h**.

The main pulmonary artery may be patent down to an atretic pulmonary valve, obstructed by right ventricular muscle, entirely separated from the right ventricle or absent. The entrance point of the ductus arteriosus into the pulmonary arteries may be associated with a stenosis of the pulmonary arteries, particularly the left one. Marked diminution in the size of the intrapericardial pulmonary arteries or replacement by fibrous cords or their total absence is usually associated with blood supply to the lungs by MAPCAs. The intrapulmonary arteries may themselves be confluent across both upper and lower lobes or discontinuous within the lung. This can have important implications for surgical outcome.

The majority of patients with pulmonary atresia and VSD will be seen as young infants or even neonates with ductus arteriosus-dependent pulmonary circulation and good-sized pulmonary arteries.

The minority will have MAPCAs supply to the lungs with arborization abnormalities of the pulmonary arteries. It is this small number of patients with MAPCAs supply to the lungs which require major investigations and provide the great difficulties in decision making. Patients with ductus-dependant pulmonary circulation are well saturated and clinically stable, while those with MAPCAs supply to the lungs show a great variability in their clinical status. They may be stable, with oxygen saturations of 75–80%, very cyanosed or in congestive cardiac failure with marked pulmonary overcirculation.

INVESTIGATIONS

Detailed investigation of patients with pulmonary atresia and VSD is usually limited to those in whom the intrapericardial pulmonary arteries cannot be clearly seen to be of normal size and are ductus-dependent. In patients with good-sized confluent intrapericardial pulmonary arteries and ductus supply, surgery can be planned without the need for invasive investigations. If there is any doubt about the size of the pulmonary arteries and the distribution and arterial supply to the pulmonary arteries, then a cardiac catheterization is indicated. Likewise, angiography will be necessary following creation of systemic-to-pulmonary artery shunts, to demonstrate that there is no iatrogenic damage requiring repair to the pulmonary arteries. Thus, in the majority of cases with pulmonary atresia and VSD, diagnosis can be confirmed and a management plan decided upon echocardiographic evidence alone.

Where there is doubt about the anatomy of the pulmonary arteries or absence of a ductus arteriosus or presence of MAPCAs supply to the lung, an early cardiac catheterization within the first few weeks of life is advised in order to delineate the exact anatomy. The age at investigation is determined by the clinical condition of the patient. When cyanosis or marked heart failure is present, then invasive investigation is indicated in the first few days of life. In those patients who are stable without heart failure or cyanosis, usually with saturations of about 80%, invasive investigation can be delayed for a few weeks.

During invasive investigation, contrast is injected into the aortic arch as well as into the descending thoracic aorta and directly into the MAPCAs in order to fully delineate the vessels. Retrograde pulmonary venous wedge injections can be useful to delineate a native true pulmonary artery system and show intrapericardial pulmonary arteries. The small intrapericardial pulmonary arteries may be demonstrated as a characteristic seagull pattern ''flapping up and down'', moving with the heart. It is also possible to see the washout of dye from the pulmonary arteries, suggesting collateral flow from different vessels. It is often necessary to repeat the cardiac catheterization to assess the anatomy accurately and with certainty.

Whilst cardiac catheterization is the mainstay of investigating patients with MAPCAs, magnetic resonance imaging and computed axial tomography are becoming more widely used. These investigations are usually used to supplement cardiac catheterization data and can have the advantage of showing the course of the MAPCAs as they pass around such structures as the trachea and the oesophagus (Geva *et al*., 2002).

PULMONARY ATRESIA AND VSD, CONFLUENT NORMAL-SIZED OR SLIGHTLY HYPOPLASTIC CENTRAL PULMONARY ARTERIES

Patients with good-sized confluent intrapericardial pulmonary arteries are suitable for repair. They usually present as neonates with a ductus arteriosus supplying the pulmonary arteries. Patency of the PDA is maintained with prostaglandin. These patients usually have saturations around 80% and are all suitable for repair with closure of the VSD. When investigation shows hypoplasia of the intrapericardial pulmonary arteries, various methods have been described to assess pulmonary artery sizes. This enables a post RV:LV pressure ratio to be estimated, which has been historically the strongest factor related to survival after repair of pulmonary atresia with VSD (Kirklin *et al*., 1988). One of the methods most commonly used is the McGoon ratio (the sum of the diameters of the proximal extrapericardial right and left pulmonary arteries are divided by the diameter of the descending thoracic aorta at the diaphragm; McGoon *et al*., 1975). The pulmonary arteries are considered to be adequate for repair when the ratio is more than 2. When the ratio is 1 the predicted pressure ratio between the right and left ventricle is about 0.7. For a McGoon ratio of about 0.8, the post-repair pressure ratio is slightly higher than 0.8 (Kirklin *et al*., 1986).

Another method for prediction of post-repair RV:LV pressure ratios is the Nakata index [the sum of the right and left pulmonary artery cross-sectional areas is divided by the body surface area (normal value $330 \pm 30\,mm^2/m^2$; Nakata *et al*., 1984)]. The size of the vessels is unlikely to be a problem in this group of patients. It is possible to repair intrapericardial pulmonary arteries way out beyond the upper lobe branches into both left and right lungs.

Decision making largely revolves around whether to repair these patients primarily with a valved conduit from the right ventricle to the pulmonary artery or to stage the repair where a systemic-to-pulmonary artery shunt is placed to delay the repair. Excellent results have been reported with primary repair (Pigula *et al*., 1999). Most centres in the UK prefer a systemic-to-pulmonary artery shunt in a neonate to provide pulmonary flow and defer the closure of the VSD and placement of valved conduit until the patient is older.

Surgical Techniques

Systemic-to-Pulmonary Shunts

A classic or modified Blalock–Taussig shunt is usually placed on the right side, through either a right thoracotomy or a midline sternotomy. Following the shunting procedure, cardiac catheterization is performed prior to repair to exclude any distortion of the pulmonary arteries which might require repair.

In addition, even if the pulmonary arteries are of adequate size and have good peripheral distribution, there can be problems with stenosis at the site of ductus insertion into the pulmonary artery. The stenosis at this site can be difficult to repair. We patch this stenosis with native pericardium or with a patch tailored from pulmonary homograft. The stenosis may be an indication for complete repair with forward flow through the valved conduit from the right ventricle. Recurrent stenosis at this site remains a problem.

Repair

Surgery involves cardiopulmonary bypass and cooling to 25°C nasopharyngeal temperature. Shunt is occluded and PDA ligated. The aorta is cross-clamped and blood

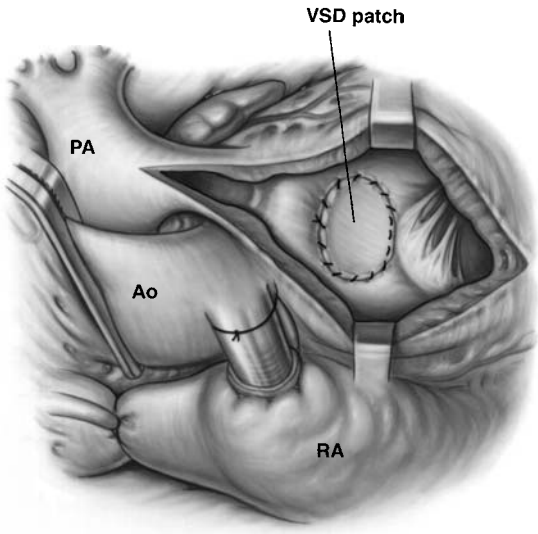


Figure 30.2

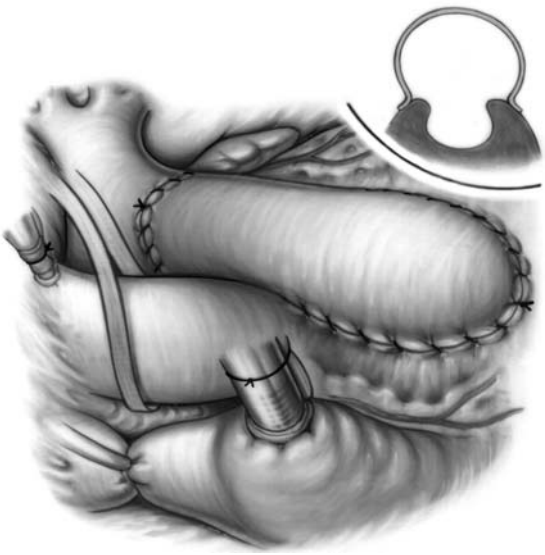


Figure 30.3

or crystalloid cardioplegia is given. The authors use topical cooling of the heart with iced slush. VSD is closed with a double velour Dacron patch inserted with interrupted reinforced mattress sutures, either through the right atrium or through the right ventriculotomy. A right atrial approach avoids traction on the ventriculotomy and reduces the size of the ventriculotomy. Aortic or pulmonary homografts are usually available for the older patient. In the neonate the availability of homografts is a limiting factor when considering primary repair. In addition, small valved conduits in neonates have not lasted well and required early replacement. For this reason the authors prefer a staged procedure for this group of patients.

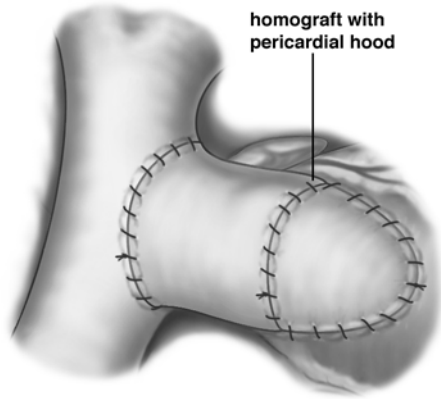


Figure 30.4

In patients with localized atresia of the pulmonary valve, the right ventricle can be opened across the atretic area and patched either with Gore-Tex or bovine pericardium, a monocusp from a homograft valve or bridged with a valved conduit. **Figure 30.2** shows the incision in the right ventricle and the VSD closed with a patch. Completed outflow tract patch is illustrated in **Figure 30.3**. When the gap is longer, a bridging valved conduit is placed. **Figure 30.4** shows a pulmonary homograft conduit extended with a hood, constructed from pericardium or a homograft. We do not recommend the use of the anterior mitral leaflet of the aortic homograft to complete the repair of the RVOTO.

PULMONARY ATRESIA WITH MARKEDLY HYPOPLASTIC CENTRAL PULMONARY ARTERIES WITHOUT MAPCAs

Marked hypoplasia of the intrapericardial pulmonary arteries and good distribution to both lung fields is unusual. It is not possible to close the VSD without first enlarging, or restoring, continuity of the central intrapericardial pulmonary arteries. It is wise to avoid peripherally placed shunts in the pulmonary arteries, as these can cause stenosis of the distal vessels. **Figure 30.5** shows a technique for constructing a central shunt between the pulmonary artery and the ascending aorta, using interposition Gore-Tex tube. **Figure 30.6** demonstrates direct implantation of the pulmonary artery onto the ascending aorta (Watterson *et al*., 1991).

Alternatively, patch enlargement of the right ventricular outflow tract can be performed (**Figure 30.7**)

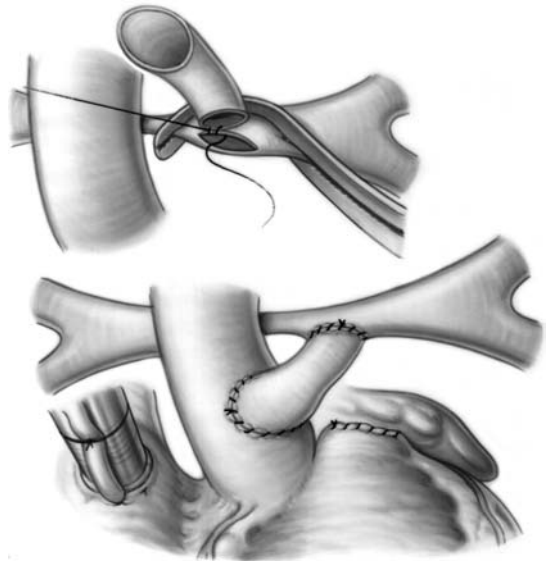


Figure 30.5

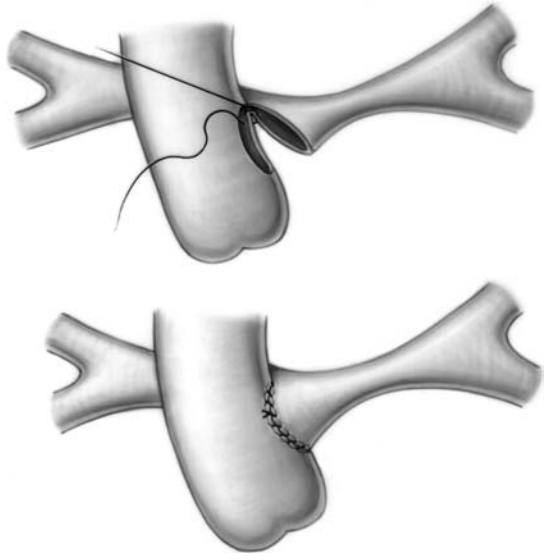


Figure 30.6

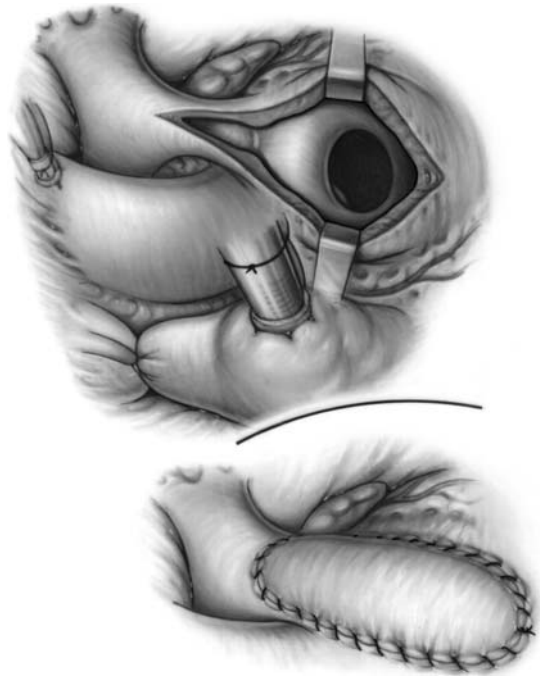


Figure 30.7

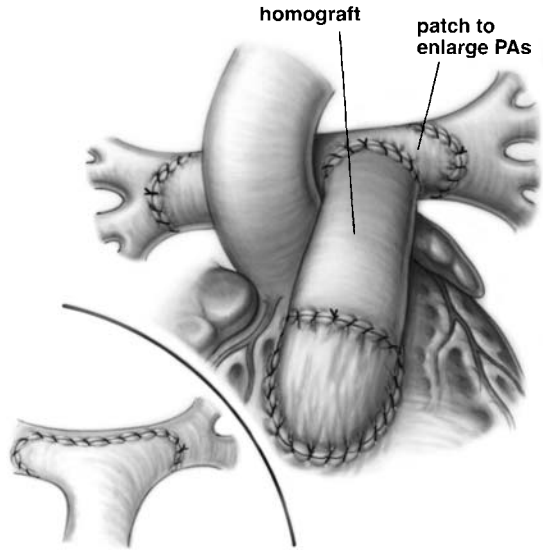


Figure 30.8

or the right ventricle is connected to the pulmonary artery with a valved conduit (**Figure 30.8**). In patients with discontinuity of the intrapericardial pulmonary arteries it may be necessary to patch the vessels extensively or place an interposition tube between the hilar vessels (**Figure 30.9**). When the pulmonary artery reconstruction is considered adequate the VSD can be closed. If the pulmonary artery reconstruction distally is regarded as inadequate then the VSD can be left open. A restrictive right ventricle to pulmonary artery conduit prevents lung overflow.

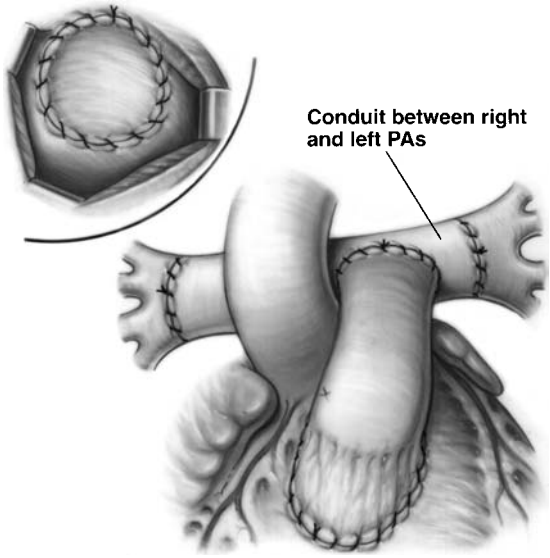


Figure 30.9

PULMONARY ATRESIA WITH VSD AND MAPCAs

Management has evolved from shunting for cyanosis, to occlusion of MAPCAs in the presence of marked heart failure, to non-intervention when patients are in a stable condition. Now all patients with pulmonary atresia, VSD and MAPCAS undergo extensive investigation, to delineate the underlying pulmonary artery tree and MAPCAs supply, before deciding whether or not it is possible or necessary to embark on a surgical programme to repair the vessels so that the pulmonary arteries can be restored to the size to accommodate repair with VSD closure. This decision making can be very difficult, as 30% of the patients when first seen as infants are often stable, with saturations in the 80% range and without evidence of marked effort intolerance or heart failure. However, the knowledge that in the future they may deteriorate, become cyanosed and run the risks of stroke, pulmonary haemorrhage and heart failure make it very important to analyse carefully the surgical possibilities at an early age. One must also be aware that any intervention may jeopardize otherwise good pulmonary arteries and make further surgery more difficult. Considerable experience with this group of patients, and particularly experience in small vessel surgery, is a prerequisite for successful treatment.

An additional problem is that the patients' native tissues available for reconstruction may be limited and the materials used for valved conduits, patch or tubed repair of vessels also have limitations. The valved conduits may need replacing, the patch material may develop stenosis at anastomotic sites and may develop an intimal peel, which can narrow anastamoses and jeopardize outcome. One must also be aware that embarking on a surgical programme may require multiple operations and multiple catheter interventions over the years without necessarily assuring satisfactory outcome. One must be aware that with closure of the VSD in patients with poor arborization pattern of the pulmonary arteries, the high RV pressures will cause RV failure, which in itself is probably more damaging and clinically difficult to deal with than ongoing cyanosis. It can also shorten the life expectancy.

Morphology of the Pulmonary Arteries and MAPCAs

Pulmonary atresia with VSD and MAPCAs shows the greatest variation in size and distribution of the native pulmonary arteries (**Figure 30.1**). MAPCAs generally originate from the descending thoracic aorta at the level of the carina and course in a variable way through the posterior mediastinum to the lungs (**Figures 30.10 and 30.11**). They are often intimately related to the tracheobronchial tree and the oesophagus and may require dissection from the airways or separation from the outer layers of the oesophageal musculature. Less commonly the MAPCAs originate from the aortic arch or coronary arteries.

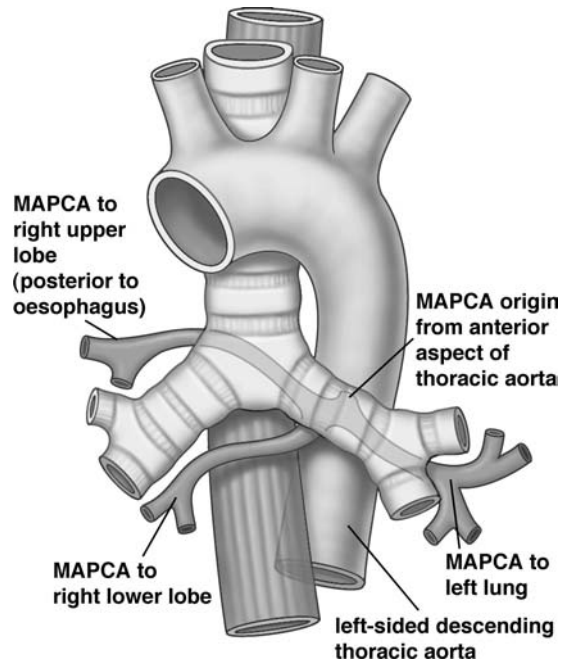


Figure 30.10

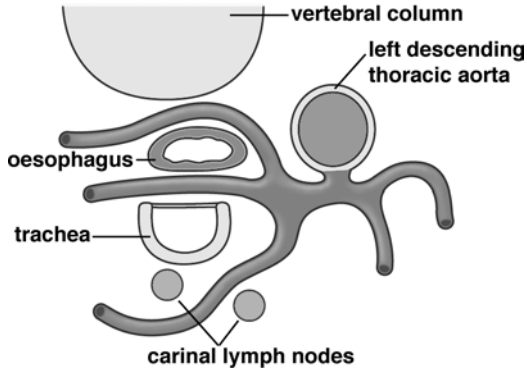


Figure 30.11

Classification of pulmonary atresia with VSD has been difficult. Tchervenkov *et al*. (2000) described three main groups:

- *Group A* has central intrapericardial pulmonary arteries, which are ductus-dependent without MAPCAs.
- *Group B* has both central pulmonary arteries and MAPCAs.
- *Group C* has no central intrapericardial pulmonary arteries, with MAPCAs providing the only supply to the lungs.

In our operative series we identified three important groups:

- *Group I*. Native intrapericardial pulmonary arteries as well as MAPCAs were present.
- *Group II*. Intrapericardial pulmonary arteries were absent but there were confluent intrapulmonary arteries in one or both lungs.
- *Group III*. Both intrapericardial and confluent intrapulmonary arteries were absent.

Surgical outcomes were significantly better in groups I and II than in group III. Thus, the presence of confluent intrapulmonary pulmonary arteries seems to be an important positive predictor of outcome (Griselli *et al*., 2004).

GENERAL CONSIDERATIONS

The age of the patient determines the type of reconstruction undertaken. In infancy tissue-to-tissue anastamoses without interposition tubes are possible because of the elastic vessels and small anatomy. However, only small tubes and valved conduits can be placed, which will need changing with the growth of the patient.

The young adult may have had multiple surgical procedures with marked secondary collateral development. This can make surgical dissection difficult and hazardous. As mobilization of vessels is not as easy as in infants, interposition tubes are necessary to unifocalize the pulmonary circulation. However, the tubes and conduits can be adult-sized.

In all cases it is wise to plan for reoperation by reconstituting the pericardial envelope, either directly or with a Gore-Tex membrane. If surgery should be staged, the method of staging depends on institutional preference. For example, Duncan *et al*. (2003) strongly advocated a staged approach, whereas Reddy *et al*. (2000) and Lofland (2000) favoured primary repair with VSD closure.

Our own preference (Griselli *et al*., 2004) is for focalization of the pulmonary arteries to the right ventricle via a valved conduit with VSD closure if possible. If the VSD is not closed, a small valved conduit may be placed to restrict lung flow or, in a small infant, clipped or banded to restrict pulmonary overcirculation. This artificial pulmonary stenosis restricts lung flow and prevents flooding of the lungs or development of pulmonary hypertension when distal stenoses are opened by interventional catheterization. Bilateral focalization of PAs to the lung hila with systemic shunts is unusual in our practice.

Surgery in Infants and Young Children

Surgery in infancy (median age 10 months) has been our commonest form of surgical repair in the last 10 years (Griselli *et al*., 2004) and has gone through an evolutionary process. Currently we aim to centralize the vessels to the right ventricle, using a valved conduit. This is achieved by mobilization of the collaterals and exploration of the pulmonary arteries within the lung and outside the lung, both through a thoracotomy based on the side of the descending thoracic aorta and via a midline sternotomy. The initial exploration through a thoracotomy is rapid, allows control of collaterals, with development of interlobar fissures, which may be advantageous in terms of reconstruction and allows less chance of damage to phrenic or vagus nerves. A midline sternotomy is then performed and the heart exposed. Through the posterior mediastinum the previously dissected collaterals are located. The placement of ligatures around the collaterals allows ready control of these vessels so as not to flood the lungs or distend the heart when cardiopulmonary bypass is initiated. (**Figure 30.12**). Extensive mobilization of the collaterals may be necessary, with occasional removal of lymph nodes from the subcarinal position. Collaterals can be moved anterior to the bronchi or left in a posterior position if they are large and mobilization is difficult. In the majority of cases, where there are three or four collaterals, these can be joined together to create a posterior wall which can be enlarged with a anteriorly placed patch, usually tailored from pulmonary homograft (**Figure 30.13**).

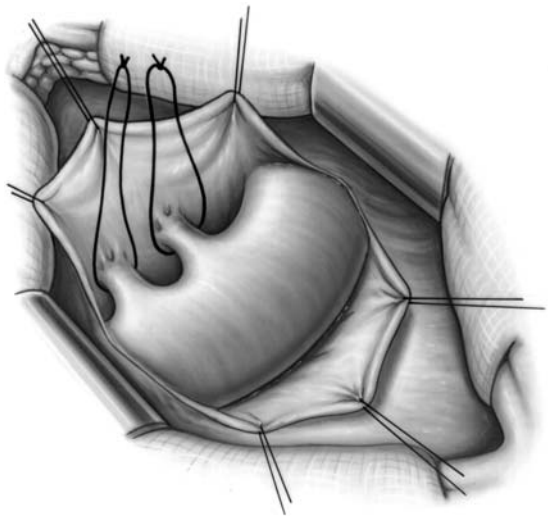


Figure 30.12

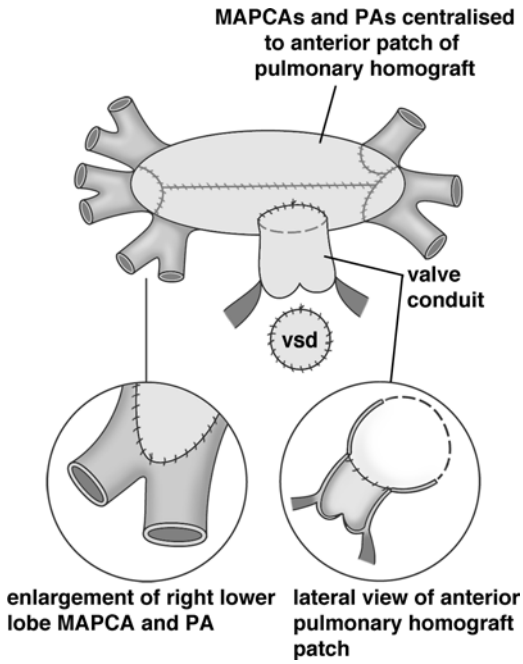


Figure 30.13

When mobilizing the vessels, care is taken not to stretch them unduly, twist or turn them, or create torsion which could cause obstruction. In addition, where the back wall of the collateral may be anastomosed to the major reconstructed central sump, a pulmonary homograft patch enlargement of the distal vessel may be advantageous, particularly in the lower vessels on the left and right side (**inset, Figure 30.13**). When dissecting the vessels, care must be taken not to damage the oesophagus. It is not unusual for the collaterals to pass through the outer layers of the oesophageal musculature, either posteriorly or anteriorly. It is often necessary to dissect the vessels carefully from the trachea and bronchi, as they can be intimately adherent to these structures. Some of the smaller vessels are often very thin-walled and it may not be possible to connect them. In addition there may be adventitial tissue which may have to be resected, so that when the vessels are joined kinking or narrowing does not occur. This is particularly so in patients who have very small vessels reconstructed with a large anterior flap of pulmonary homograft; when the blood flows under high pressure into these vessels, they can twist and create peripheral stenoses.

When collaterals are mobilized from the back of the hilum over the main bronchus, particularly the right main bronchus, care has to be taken to maintain the orientation of the vessels so that they are not twisted or kinked as they enter into the upper or lower lobe. This is particularly the case where there may be just two collaterals, one supplying the left lung and one supplying the right lung entering the posterior hilum, and these vessels are used as conduits into the lungs. In mobilizing over the upper border of the bronchus, one has to be careful not to put too much tension on these vessels, and it may well be worthwhile supplementing the connection across the midline with a tube or patches to take tension off these vessels.

The reconstruction is performed on cardiopulmonary bypass, usually cooling the patient to 28°C nasopharyngeal temperature. It is usually possible to control the vessels distally, either with vascular clamps or with small Yasargil aneurysm clips, so that the reconstruction and patching of the central pulmonary arteries can be performed whilst on cardiopulmonary bypass, without the need for low flow or circulatory arrest. The aim in small children is to reconstruct with as much native tissue as possible, to allow for potential growth of the vessels. A valved conduit is attached to the newly created pulmonary artery confluence on the left side of the aorta and then the aorta is cross-clamped and cardioplegic solution is administered. The right ventriculotomy is then performed. VSD is closed via the right atrium or via the right ventriculotomy and then the conduit is attached to the right ventricle. When high RV pressure is anticipated (when the VSD was left-opened) the RV anastamosis is carefully reinforced with mattress sutures with Teflon pledgets.

In the majority of patients the reconstructed intrapericardial pulmonary arteries are behind the ascending aorta and above the bronchi in the normal hilar position. Sometimes the pulmonary arteries cannot be reconstructed to the lung hila, as they are too posterior

and the MAPCAs enter the lung posteriorly and cannot be mobilized to an anterior position. If there are confluent intrapulmonary arteries (Griselli *et al*., 2004), it is possible to connect a tube from the pulmonary arteries in the oblique fissure of each lung across the midline and then via a valved conduit to the heart. A subhilar reconstruction of intrapericardial pulmonary arteries in the oblique fissure using a non-valved conduit from hilum to hilum is illustrated in **Figure 30.14**).

The Young Adult with Pulmonary Atresia, VSD and MAPCAs

This is a very difficult group of patients. Because they have survived, there is a possibility that they have some reasonably good MAPCAs and pulmonary vessels. They may also have developed pulmonary vascular disease through unprotected blood flow over the years. Careful evaluation is necessary with measurement of distal PA pressures. Often these patients have been stable over their lifetime and they are reluctant to come forward for surgery, despite increasing cyanosis and polycythaemia. The possibilities are of total correction with VSD closure or palliation with a shunt or RV–PA conduit to low pressure vessels to improve oxygenation.

Surgical reconstruction of the pulmonary arteries can be very difficult and, unlike in infants, the collaterals and pulmonary arteries are merely dissected and not mobilized. The vessels are friable and there is secondary collateral development. It is better to place tubes between vessels and connect them across the midline. Because of the size of the patient, this can be achieved with adult-sized grafts. Again, some ingenuity in surgical procedures may be necessary, with use of circulatory arrest to gain access to pulmonary arteries which may be of reasonable size but on the posterior aspect of the lung, and in this situation hypothermia with circulatory arrest may allow access to these vessels with collapse of the lung far more readily than continuing on bypass or dissecting through a thoracotomy. However, we would still advocate mobilization of MAPCAs through a thoracotomy in the first instance.

Some of the techniques more applicable to the older patient are described here. Sawatari *et al*. (1989) advocated the use of intrapulmonary bridges made of xenograft pericardium for peripheral unifocalization (**Figure 30.15**). Permut and Laks (1994) created tubes from heterologous pericardium to create artificial pulmonary arteries to which the MAPCAs can be anastomosed (**Figure 30.16**) A separate intrapericardial conduit is then connected from hilum to hilum, then to the heart with a valved conduit (**Figure 30.17**).

If it is not possible to close the VSD, palliative procedures are performed to increase lung flow and reduce cyanosis. These include a modified Blalock–Taussig shunt, central shunts (**Figure 30.5**) or a restrictive valved conduit from the RV to a hypoplastic intrapericardial pulmonary artery.

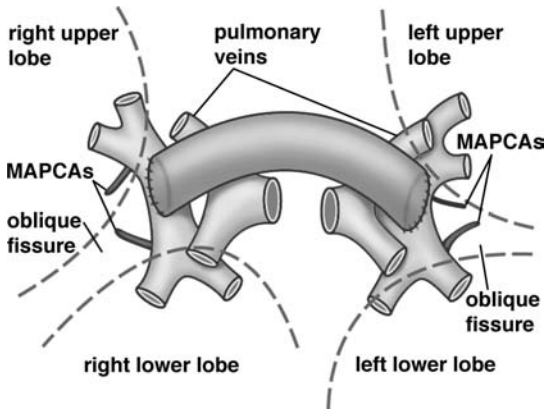
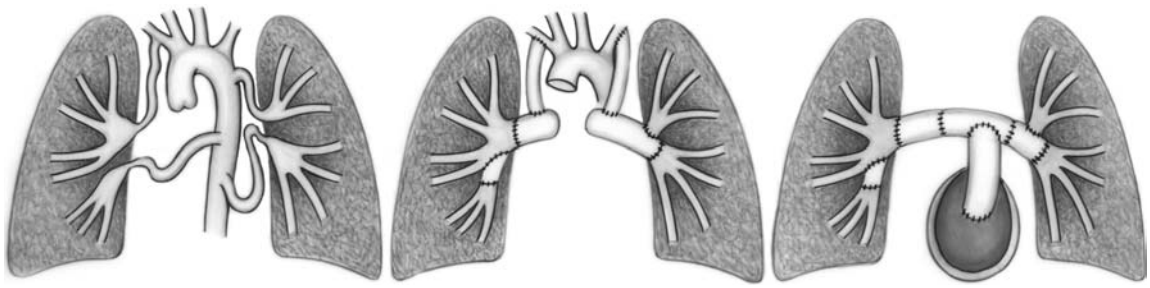


Figure 30.14

SURGICAL COMMENTS

The conduct of cardiopulmonary bypass is as usual; however, our own practice (W.B.) is not to use modified



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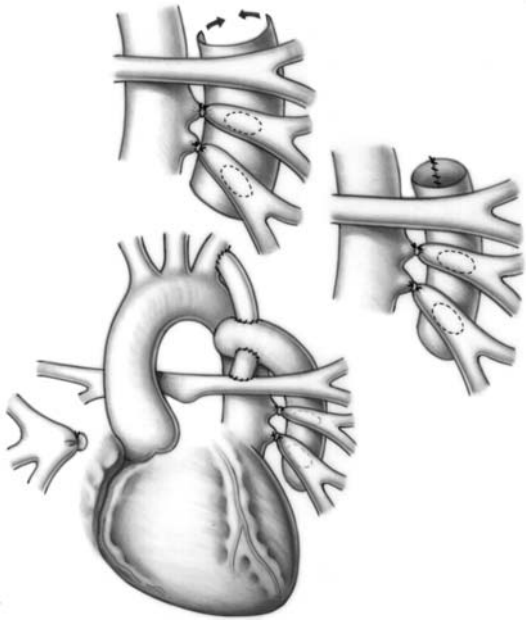

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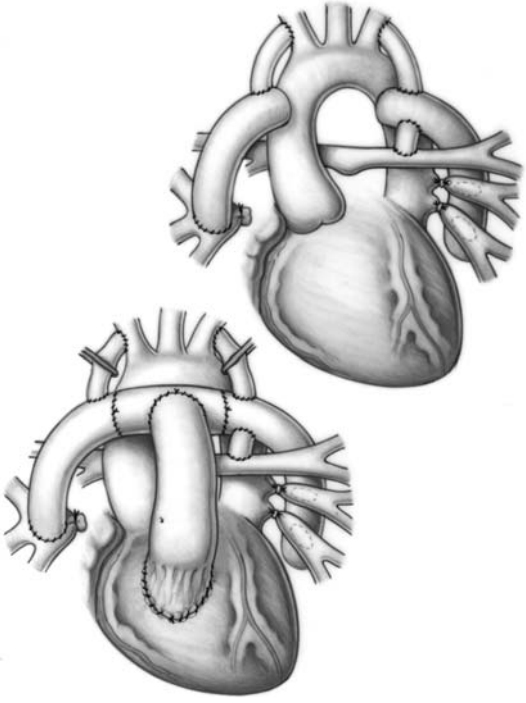

Figure 30.17

ultrafiltration, although other institutions consider this may be helpful. We do not cannulate the superior vena cava directly but through the right atrial appendage. This allows manipulation of the superior vena cava for work deep in the posterior mediastinum and behind the upper border of the right upper lobe pulmonary veins. Short periods of circulatory arrest and/or low flow may be necessary to visualize vessels clearly and place very accurate sutures into quite often flimsy vessels, deep in the lung. Aprotinin is infused during the operation and tissue glue is used around the suturelines, since at the end of the procedure many sutures lines are distally placed and are not accessible. Previous surgery can make dissection very difficult and one has to weigh up very carefully the pros and cons of reoperating on the thorax where previous thoracotomies have been performed. Staging the procedure has sometimes proved advantageous. We dissect the vessels, achieve haemostasis and then, rather than heparinizing, we delay for a few days before reoperating through the midline sternotomy in order to gain the advantage of the previously dissected field but not to have bleeding problems with heparinization on cardiopulmonary bypass. We have no experience of the clam shell incision, although others have found that it gives good exposure (Luciani *et al*., 1995; Moritz *et al*., 1996).

Can the VSD be Closed?

The ideal aim is to close the VSD, with resultant low RV pressures well maintained over the long term. However, it can be difficult to predict the postoperative RV pressure. The McGoon and Nakata calculations are not of value beyond the lung hila; however, the predictive pulmonary artery sizes in the Kirklin and Barratt-Boyes textbook (1993) are helpful in assessing the required sizes of the reconstructed intrapericardial pulmonary arteries. We depend on the preoperative evaluation of the vessels, together with the assessment of success of the operative reconstruction of the pulmonary arteries, to determine whether the VSD can be closed. Since Fallot's tetralogy can be repaired to one lung with resultant low RV pressure, we consider perfusion of 15 lung segments, i.e. approximately one lung, as safe for VSD closure. If we are unsure, we do not close the VSD (Griselli *et al*., 2004). Reddy *et al*. (1997) have devised an intraoperative pulmonary blood flow study to assess the possibility for VSD closure. This has been very successful. Usefulness of this technique has also been demonstrated by Carroti *et al*. (2003).

POSTOPERATIVE MANAGEMENT AND PROBLEMS

The patients are monitored with a left atrial line together with the central venous and arterial lines. Sternal closure

may be delayed in small children, so as not to compress the heart or the conduit. Usually dobutamine and adrenaline and more recently milrinone have been used for inotropic support. Pulmonary problems are common, with oedema of part or whole lungs because of increased flow after the repair. Occasionally frank haemorrhage occurs, requiring positive-pressure ventilation for longer periods.

Postoperative high central venous pressures may indicate RV hypertension or pulmonary hypertension. High LA pressures, especially during weaning from ventilation, suggest residual left-to-right shunting, particularly a significant residual MAPCA. When there is concern about haemodynamic status, then cardiac catheterization is performed to define the reconstruction. Any residual mechanical problems can be resolved by reoperating on the reconstructed pulmonary arteries or on the residual VSD. Occasionally it may be necessary to reopen the VSD. Postoperative cardiac catheterization to plan future interventions and further surgery if needed are performed in the first 3 months after surgery. At this time, balloon dilatation of stenotic vessels can be commenced.

COMBINED MANAGEMENT WITH SURGERY AND INTERVENTIONAL CARDIOLOGY

It is important to consider a combined approach to these patients, so that surgery can give access for the cardiologist with interventional balloons, stents and cutting balloons to improve the surgical repair. This is particularly important for the stenoses of intrapulmonary arteries. Whilst stenting may be of limited value, it can be useful in the older patients to open out vessels that are not accessible surgically. Direct connection of the pulmonary arteries to the right ventricle also allows the interventional cardiologist ready access to the peripheral vessels. The shunt may give difficult, limited or no access at all for the interventionist. A proactive approach is taken to intervene before stenoses become severe. Usually the patient is catheterized before discharge after reconstructive surgery, then further catheters are planned, depending on the anatomy seen at the first study.

VALVED CONDUITS AND PATCHES

Valved conduits are unsatisfactory. Available ones are the homografts, the Hancock valved conduits and Contegra. If low RV pressures are anticipated, we favour use of the pulmonary over the aortic homograft. If high RV pressures are anticipated then an aortic homograft will tolerate higher pressures better. The Contegra is occasionally utilized, but we have been concerned about fibrous in-growth into parts of the anastomoses. The Hancock valved conduit is reliable for repair. When the rigid ring is removed, this allows for manipulation of the conduit. Fibrous in-growth or a peel formation on the inner surface of the conduit is a problem. However, it is satisfactory for clipping down and for providing a small limiting valved conduit under high pressures; it will not distend. We have had three instances of false aneurysms developing at the site of insertion of the RV conduit into the right ventricle, despite reinforcing the suture line. One of these patients died at the time of reoperation. We have used native pericardium in the past, together with homograft material, aortic tubes and bovine pericardium for reconstruction. We have found the pulmonary homograft patch, in light of our experience with the Norwood procedure, to be a satisfactory substitute to withstand pressures in small infants well and not to develop peel on the inside, which might encroach onto the distal pulmonary artery reconstruction.

RESULTS

Pulmonary Atresia and VSD

Early and long-term results after repair of tetralogy of Fallot and the pulmonary arteries are less satisfactory than after repair of tetralogy of Fallot with pulmonary stenosis. In a series of 139 patients who underwent repair of tetralogy of Fallot and pulmonary arteries in 1967–1986 (Kirklin *et al*., 1988), the operative mortality rate was 15% and the survival rates at 1.5, 5, 10 and 20 years were, respectively, 82%, 76%, 69% and 58%. The probability of death was inversely related to the size of the pulmonary arteries and directly related to the postrepair peak right-to-left ventricular pressure ratio and to the number of MAPCAs. At Great Ormond Street Hospital (2000–2004) we repaired isolated PA + VSD (tetralogy of Fallot type) in 29 children. One patient with major postoperative lung parenchymal haemorhage died (3.4%).

Pulmonary Atresia, VSD and MAPCAs

In the group of 164 patients managed at Birmingham, UK, during 1989–2002, we found that pulmonary artery confluence both in the pericardium and within one or both lungs gave a lower risk of death than the group of patients with absent intrapericardial pulmonary arteries and non-confluent intrapulmonary arteries. Actuarial survival at 10 years was $80 \pm 4\%$ for all patients and $86 \pm 4\%$ after VSD closure. There was no statistical difference in risk between the primary repair with VSD closure and the groups undergoing staged repair with VSD closure later (Griselli *et al*. 2004).

Reddy *et al*. (2000), who popularized the primary unifocalization and VSD closure technique, reported 85 patients during 1992–2000, with an actuarial survival

of 80% at 3 years. 66% of patients were suitable for primary repair with VSD closure, with a survival rate of 88% at 2 years. Carotti *et al*. (2003) reported 37 patients. The strategy was based on the total neopulmonary arterial index. 25 underwent primary one-stage repair. Survival at 7 years was 81%. The authors found intraoperative flow studies helpful to determine the feasibility of the VSD closure. Duncan *et al*. (2003) are strong advocates of a staged approach, with excellent results: 46 consecutive patients with one late death.

All groups have noted the association of 22q11.2 deletion with pulmonary atresia and VSD. Mahle *et al*. (2003), in a series of 67 patients with pulmonary atresia and VSD ± MAPCAs, noted an incidence of 34% of patients with 22q11.2 deletion. 65% of patients with MAPCAs (20 patients) had this anomaly.

All reports have noted the high incidence of reoperations and catheter reinterventions in these complex patients. In our own series, freedom from intervention after VSD closure was only 46 ± 8% at 10 years. There is little long-term information on the RV:LV pressure ratios. Griselli *et al*. (2004) reported a median RV:LV pressure ratio of 0.6 (0.3–0.9) during the perioperative period and Duncan *et al*. (2003) reported an RV:LV pressure ratio of 0.5 ± 0.22 in the early follow-up period. Comparison of these results with the natural history of the condition is difficult (Bull *et al*., 1995). It seems that surgery is at least improving the early outlook for these patients. However, medium- to long-term quality of life after surgery has to be assessed.

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31

Absent Pulmonary Valve Syndrome

M. J. Elliott

Absent pulmonary valve syndrome (APVS) is a relatively rare condition, representing a subset of around 3% of cases of tetralogy of Fallot in both autopsy and clinical reviews (Lev and Eckner, 1964; Stafford *et al*., 1973). The syndrome is characterized by a ring-like and usually stenotic malformation, rather than absence, of the pulmonary valve, with failure of development of the valve cusps. The central pulmonary arteries are usually hugely dilated or aneurysmal and 93% of the patients (Calder *et al*., 1980) have the intracardiac characteristics of tetralogy of Fallot (**Figure 31.1**). The pulmonary arterial abnormalities are often not limited to the central vessels. Postmortem angiography has demonstrated abnormal ''tufted'' segmental and subsegmental pulmonary arterial branching patterns (Karl *et al*., 1986; Milanesi *et al*., 1984; Rabinovitch *et al*., 1982). These abnormally branching vessels spread peripherally with little change in their size, compressing their associate bronchi. **Figure 31.2a** shows normal branching; **Figure 31.2b** illustrates the abnormalities often associated with this syndrome. The proximal pulmonary arteries also show reduced lamina formation. The bronchial compression and subsequent airway problems are exaggerated by deficient or defective cartilage formation in the bronchi (Karl *et al*., 1986), and broncho-alveolar multiplication may be abnormal (Rabinovitch *et al*., 1982).

Since the syndrome was first described by Cheevers in 1847, many other associated cardiac defects have been described; intact ventricular septum (Horigome *et al*., 1997), ventricular and atrial septal defects (Sethia *et al*., 1986), absent ductus arteriosus (Emmanoulides *et al*., 1976; Milanesi *et al*., 1984), transposition of the great vessels (Rabinovitch *et al*., 1982), tricuspid atresia and intact ventricular septum (Freedom *et al*., 1979), single atrio-ventricular valve (Opie *et al*., 1983) and interrupted aortic arch (Mignosa *et al*., 1998).

There has been considerable debate about the aetiology of this condition. For some time, the apparent agenesis of the ductus artreriosus in many cases was thought to contribute to the pulmonary artery dilatation (Emmanoulides *et al*., 1976) as a result of increased pulmonary artery pressure and flow in the absence of diastolic run-off through a ductus in foetal life. A more complicated explanation of the prenatal role of the ductus has been provided by Yeager *et al*. (2002). However, the existence of several cases in which the ductus was seen both to be present and widely patent casts doubt on this theory, despite recent revival of interest in the complex role of the ductus in aetiology (Podzimkova *et al*., 1997). More favoured is the argument that the tightly stenotic pulmonary annulus, together with free pulmonary regurgitation, acts against the high pulmonary vascular resistance *in utero* (Alpert and Moore, 1985; Kron *et al*., 1988; Lakier *et al*., 1974; Thanopoulos *et al*., 1986). Associated deletion of chromosome 22 has also been reported (Johnson *et al*., 1995), but the importance of this observation is not yet clear. Extrathoracic associations, such as has been described (Beck *et al*., 2002) with Crouzon syndrome, should be considered.

DIAGNOSIS

The diagnosis of absent pulmonary valve syndrome can be made prenatally using foetal echocardiography (Sameshima *et al*., 1993; Callan and Kan, 1991; Momma *et al*., 1990). However, the outcome of prenatally diagnosed absent pulmonary valve syndrome is very poor (Razavi *et al*., 2003). Postnatally, it is self-evident from the variation in morphology described above that the clinical presentation may also vary. Patients with absent pulmonary valve syndrome are probably best categorized into two groups, based on the age at presentation (Snir *et al*., 1991):

1. Infants (often neonates) with severe respiratory symptoms of tachypnoea, air-trapping, cyanosis and wheezing. Sometimes there is actual respiratory failure. In these patients the bronchial compression

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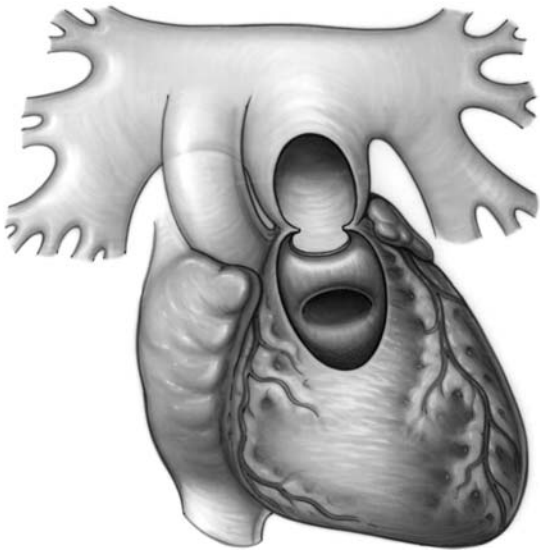

Figure 31.1

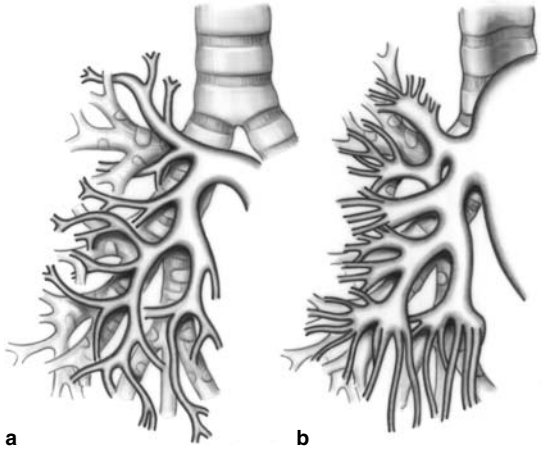

Figure 31.2

is the dominant pathology, and may precipitate attempts at life-saving surgery.

2. Older children with less severe symptoms, often those of tetralogy of Fallot, in whom surgery can often be deferred.

The physical signs usually include a pan-systolic murmur at the left sternal border, together with a variable length diastolic rumble of pulmonary regurgitation. The pulmonary second sound is absent. In infants particularly, wheezing sounds are common, and areas of air-trapping or consolidation may be evident. The chest may be hyperexpanded and the use of accessory muscles of respiration obvious.

The diagnosis can readily be defined by echocardiography and high-resolution computed tomography (CT), particularly with three-dimensional (3D) reconstruction, is an excellent method (Kim *et al*., 2002; Sagy *et al*., 1996) of demonstrating the complex relationships between the dilated central pulmonary arteries and the major airways. These images can be used to plan surgery. Unfortunately, there seems (Moon-Grady *et al*., 2002) to be little correlation between measurements of the pulmonary arteries and outcome, implying that the sum of the whole ''complex'' is more important than its individual parts. CT has largely superseded cardiac catheterization in diagnosis, but in some patients the very distal pulmonary arteries may require morphological demonstration by angiography. Aortopulmonary collaterals have also been described (Knauth *et al*., 2004) in absent pulmonary valve syndrome, and may play a particular role in delaying recovery from surgery. They should be sought on preoperative investigation and repeat cardiac catheterization or MRI may be required in the postoperative period if recovery is slow.

INDICATIONS FOR SURGERY

The indications for surgery vary with age, the severity and cause of airway obstruction and the nature of the intracardiac problem. Medical management of these children has no value other than as supportive therapy. However, excellent intensive care is vital to minimize the pulmonary problems, particularly in the neonate. The complex intrapulmonary balance between areas of high pulmonary blood flow and others with air trapping can be difficult to manage (Stayer *et al*., 2002), and skilled manipulation of ventilator strategies may be required.

The goals of surgery are to correct any intracardiac anomaly, prevent right-sided heart failure and to alleviate or prevent bronchial compression and peripheral lung damage. There is also no role for palliative operations which, historically, have not been successful. Surgery is best carried out as a one-stage procedure, including correction of the intracardiac anomalies and plication of the pulmonary arteries, sometimes with suspension or repositioning to relieve as much airway obstruction as possible. Al-Halees *et al*. (1997) have argued in favour of a two-stage approach with preliminary pulmonary artery banding, but this is not widely supported. The most common intracardiac morphology is that of tetralogy of Fallot. In these cases it is necessary to close the ventricular septal defect and relieve any right ventricular or pulmonary valve obstruction. In infants, it may also be

necessary to insert an RV–PA valve conduit to protect the RV from the damaging effects of pulmonary regurgitation from the high resistances associated with the complex vascular and bronchial anomalies. The timing of the surgery relates to the severity of symptoms, with neonates and infants often requiring relatively urgent surgery, as the airway problems predominate. Neonatal presentation can cause particular difficulty in decision making. Older children can wait a while, and we prefer to treat them exactly as other tetralogy patients, with elective reparative surgery at around 6–12 months of age.

Because of the incidence of arborization anomalies in the pulmonary circuit, and the presence of bronchial compression (especially in the symptomatic neonate or small infant), conventional treatment of the Fallot morphology (small pulmonary annulus and right ventricular outflow tract obstruction) using a small trans-annular patch may be doomed to failure because of severe pulmonary regurgitation, due to the relatively high peripheral pulmonary resistances. In such patients, a valved conduit should be inserted between the right ventricle and pulmonary arteries. Those patients with absent pulmonary valve syndrome presenting later, without airway symptoms, can usually be managed as any other case of tetralogy of Fallot.

SURGICAL TECHNIQUE

Choosing the type of procedure to perform for absent pulmonary syndrome can be challenging, and will be influenced by personal experience and confidence. The surgical technique must be dependent on the underlying morphology. For the purposes of this review, I will consider the most common morphology of tetralogy of Fallot (TOF). The underlying principles of repair are those of TOF, which are described in Chapter 29. The specific features of APVS are the dilated pulmonary arteries, airway compression and abnormal pulmonary valve. Repair of these features usually involves reduction or modification of the shape of the proximal branch PAs, with or without the insertion of a valved conduit from RV to PA. A variety of reconstructive strategies have been described, and will be reviewed later in this section. Recently, Hraska *et al*. (2002) have proposed a variant of the Lecompte manoeuvre, usually performed in the arterial switch operation, for relief of bronchial compression. This operation is particularly suitable for neonates, may produce excellent relief of airway obstruction in well-selected patients and may avoid the need for an RV–PA conduit in such a small child.

Preparation for Repair

Common to all single-stage repairs of APVS, the approach to the heart is via a median sternotomy. It is usually necessary to remove the thymus to maximize the space available to decompress the central airways. Particular care should be taken to avoid the phrenic nerves, as the additional burden of diaphragmatic paralysis would be very badly tolerated in this group of patients. Thus, blunt dissection of the lateral margins of the thymus with avoidance of diathermy is sensible. The pericardium is opened longitudinally, slightly to the right, to facilitate the taking of a pericardial patch later in the operation if required. Stay sutures are applied to the pericardial margins and attached to the skin edges, burying the knots so that they do not catch on the sutures later. The external anatomy of the heart and pulmonary arteries is carefully defined and decisions as to the best approach confirmed.

The aorta is then separated form the pulmonary artery. I prefer to use diathermy. A nylon tape is passed around the aorta to facilitate traction and aortic cross-clamping in the presence of the large, dilated pulmonary arteries. In absent pulmonary valve syndrome, the lymphatic vessels between the aorta and pulmonary artery are sometimes particularly prominent and it is advisable to ligate them to prevent later chylothorax. The pulmonary arteries should then be dissected out to the hilum on each side, once again taking particular care to avoid the phrenic nerves. If a patent ductus arteriosus is present, it should be ligated. If the Lecompte manoeuvre is to be performed, as in the Hraska *et al*. (2002) approach, then the mobilization must be complete and, if present, the ductus divided between ligatures, and the aorta too must be mobilized to its first branches.

Once the pulmonary arteries have been mobilized, cannulation sutures should be inserted. The aortic cannulation site should be positioned as distally as possible and preferably in anterior midline of the aorta, to facilitate access to both sides of the vessel to permit pulmonary artery reconstruction. We prefer to use bicaval cannulation, using Medtronic Pacifico wire-reinforced venous cannulae. The purse-strings are placed as described in Chapter 11. Cardiopulmonary bypass is established, patent ductus ligated, and the patient is cooled on full flow to (usually) 28°C. This temperature is chosen so that it does not commit the child to deep hypothermia, but facilitates rapid cooling if it proves necessary to reduce flow in order to have a clear operating field. Circulatory arrest is rarely required to deal with complex pulmonary artery reconstruction, if there is significant collateral flow. The aorta is cross-clamped and cold blood cardioplegia is instilled into the aortic root. During the infusion of cardioplegia, a vent is inserted into the left atrium at the junction of the right superior pulmonary vein. This will greatly improve visibility during the operation. A purse-string suture placed around the insertion site facilitates later

placement of a left atrial pressure line if required. We use a disposable DLP 10 Fr gauge vent which has a malleable wire core to permit easy and stable positioning.

Repair of Intracardiac Defects

The right atrium is opened parallel to the atrio-ventricular groove and stay sutures are applied to its margins to improve exposure. The intracardiac anatomy is inspected, noting particularly the position and margins of the VSD and the presence or absence of obstructing right ventricular outflow tract muscle bundles. I prefer to close the VSD through the right atrium in almost all cases. To do so, I first place a Teflon-buttressed horizontal mattress suture at the 12 o'clock position of the VSD, and use that as a retractor to pull the margin of the VSD towards me. This has the double effect of protecting the aortic valve and exposing the septal and parietal extensions of the trabecula septomarginalis, which usually make the obstructing component of the tetralogy of Fallot. Whilst retracting on the Teflon buttresses traction suture, any obstructing muscle bundles can be divided, following which the VSD can be closed using the traction suture as the start of a continuous suture line. It is easier to complete the closure of the upper part of the VSD first, using external pressure with the left index finger to expose the upper margin of the VSD if access is difficult. Further details of the repair can be found in Chapter 29 and will not be repeated here.

Approaches to Repair the Pulmonary Arteries

The approaches to the pulmonary arterial component of the syndrome can be divided into three main groups:

1. Plication or resection of pulmonary arterial wall.
2. Resection of central pulmonary arteries.
3. Repositioning of pulmonary arteries anterior to the aorta.

Each of these approaches can be combined with the insertion of some sort of valve between the RV and the reconstructed pulmonary artery.

Plication/Resection of Pulmonary Arterial Wall

This technique was originally described by Dunnigan *et al*. (1981), and has been since adapted by many surgeons. The technique has been well described by Mee's group (Stellin *et al*., 1983; Watterson *et al*., 1992) and more recently by Planche's group (Conte *et al*., 1997). The amount of PA tissue resected depends on the degree of dilatation, and must be individualized. However, the techniques are best described by imaging severe dilatation of the pulmonary arteries. Good exposure of the pulmonary arteries is mandatory and, following aortic cross-clamping, aortic transection may be necessary to achieve this (Conte *et al*., 1997). Stay sutures applied to the transected aortic margins can be used to retract the aorta to improve access. Once the PAs are exposed, the lines of resection should be determined, and I have found it useful to mark out these lines directly onto the PA using a sterile surgical pen. Once marked (or imagined), a vertical incision is made in the anterior wall of the MPA from the infundibulum of the RV out to the bifurcation of the main pulmonary artery, and the resection is continued as marked in **Figure 31.3a**.

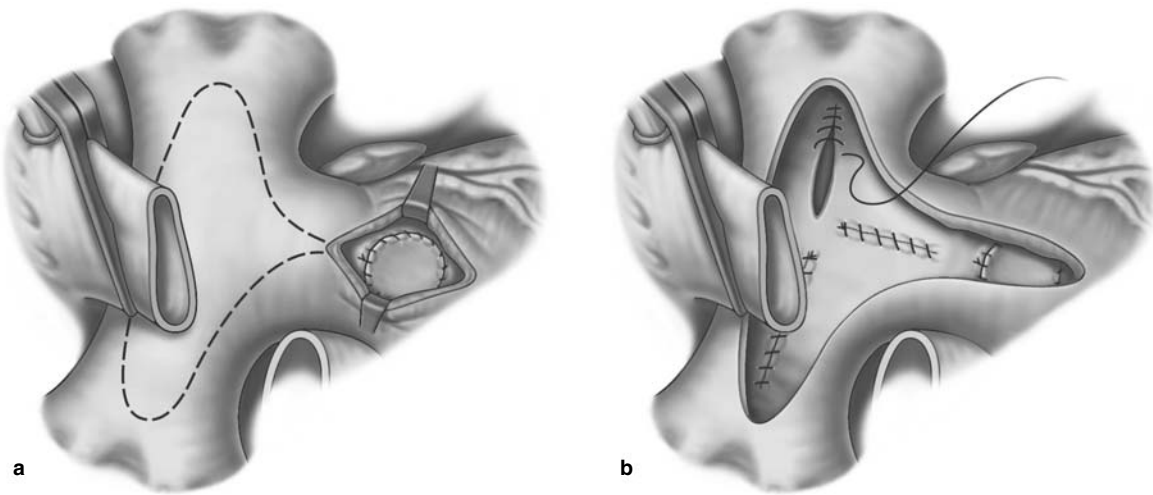

Figure 31.3

If the dilatation is severe, plication or resection of the posterior walls of the pulmonary arteries can be carried out via the wide open pulmonary arteries, as indicated in **Figure 31.3b**. Plication can be achieved by pulling up a ridge of the posterior wall and oversewing it with two layers of 5-0 or 6-0 Prolene, with the first layer as a horizontal mattress technique to establish the depth of plication and the second layer an over-and-over stitch to limit the vertical size of the plicated ridge. I prefer to resect tissue from the posterior wall, using a similar technique to the anterior wall. Once the resection is complete and the posterior plication performed, the margins of the anterior wall are reconstituted using Prolene sutures. The repair of the outflow tract will depend on the decision as to whether or not a valve is necessary in the circuit, or whether a trans-annular patch can be used, as shown in **Figure 31.4**.

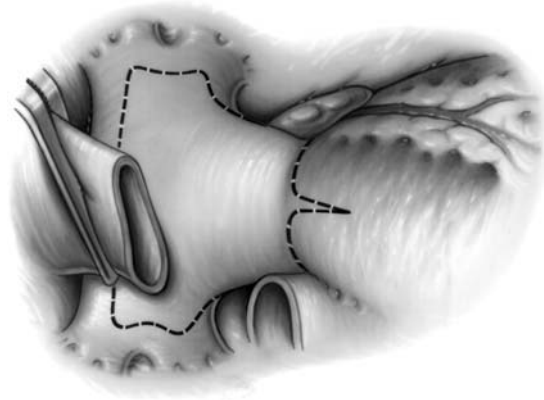

Figure 31.5

Resection of Central Pulmonary Arteries

If this technique is chosen, then the central pulmonary arteries are resected and plicated if necessary. The main pulmonary artery is excised, with transection of its proximal end just above the pulmonary annulus and of its distal end at the level of the bifurcation. **Figure 31.5** shows the aorta transected, but this should only be done if exposure by mobilization and retraction is inadequate to permit repair of the pulmonary arteries. These distal lines of excision can be extended laterally along the anterior wall of the pulmonary arteries as far as the first bifurcation on each side, and can be plicated as shown in **Figure 31.6a**. This approach also allows plication

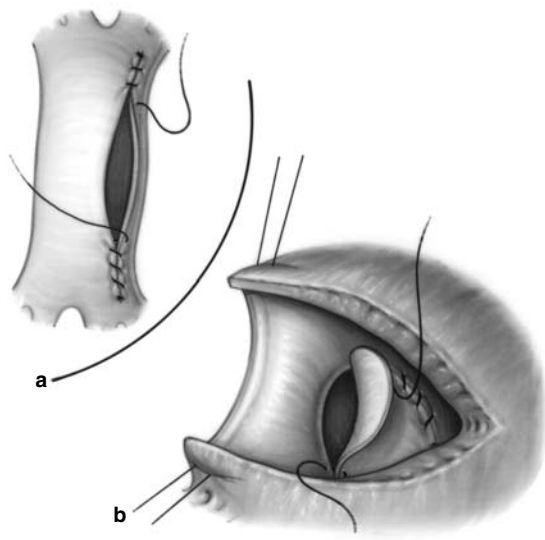

Figure 31.6

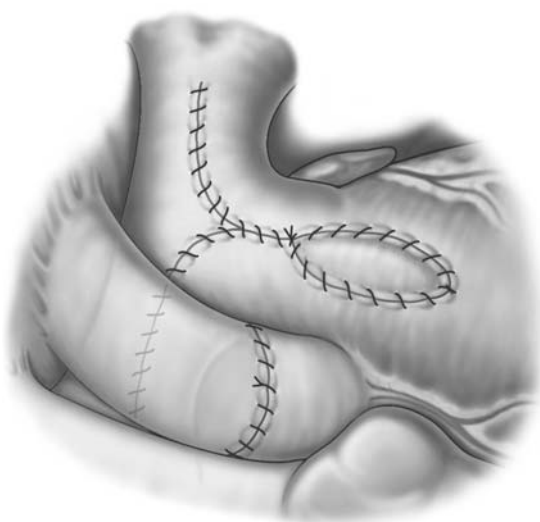

Figure 31.4

of the posterior pulmonary arteries, as described above. **Figure 31.6b** also shows that the VSD can be approached via the infundibulotomy if preferred, or if the access via the right atrium is too difficult or traumatic.

The simplest method of replacing the lost tissue from the pulmonary arteries is to insert a valved homograft. Once the intracardiac part of the repair is complete and, if previously divided, the aorta is reconstituted, the homograft implantation can often be undertaken with the aortic cross-clamp removed. A suction catheter inserted via the right atrial appendage can be used to aspirate coronary sinus blood. **Figure 31.7** shows the method of insertion of an aortic homograft, which is prepared as

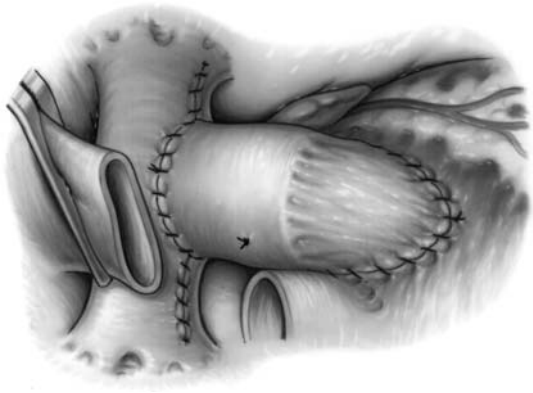

Figure 31.7

described in Chapter 35. It does not really matter whether the suturing starts distally or proximally, as long as the length of the homograft is judged well. In the context of absent pulmonary valve syndrome, it is probably better to err slightly on the too-short side than the too-long, since there would be a risk of kinking of the vessel if it were too long, and also the risk of failure adequately to relieve the airway obstruction. It is useful to use a large (25 mm) needle 4-0 Prolene suture for the proximal end of the homograft, as it greatly speeds up the anastomosis. If the anterior mitral leaflet is present on the aortic homograft, then it may be used to patch the anterior wall of the ventriculotomy incision. However, currently we prefer a patch of bovine pericardium or Gore-Tex to complete the anterior wall. The patch should be cut carefully, with the shape shown in **Figure 31.8** being ideal.

If a valved conduit is required, I prefer to use a pulmonary homograft with its bifurcation intact if one

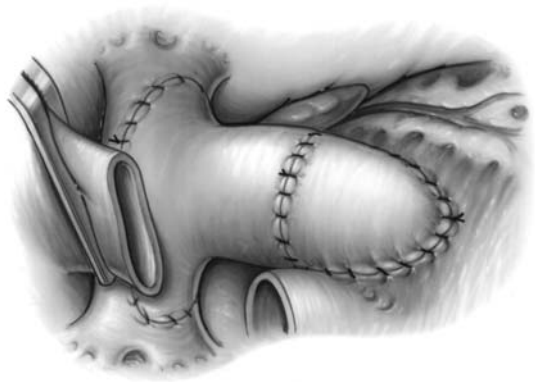

Figure 31.8

is available. This allows the distal bifurcation to be used as an on-lay flap, if necessary optimizing the flow dynamics of the RV outflow. Whilst this technique is usually applied when there is pulmonary artery stenosis, the manipulation of the PAs in absent pulmonary valve syndrome repairs can result in minor distortions of the PA, which can be corrected by such a conduit. The bifurcated pulmonary artery can also be used as true branch pulmonary arteries, as described by the Boston group (Hew *et al*., 2002), anastomosing the distal branches of the bifurcated homograft to transected pulmonary arteries of the patient close to the bifurcation on each side. One can also resect the central branch pulmonary arteries, reanastomose their posterior walls centrally and attach the distal end of a homograft to this area, as suggested by Ungerleider's group (Kirshbom *et al*., 1999). Homografts are not the only valved conduits that have been used, and many surgeons now like to use the Contegra bovine jugular vein graft (Boudjemline *et al*., 2003). However, there are inadequate long-term data to justify recommending these conduits, and reports of distal stenosis of these grafts are worrying.

It is not always necessary to insert a full valved conduit, and several workers have reported the use of monocusp valves in this setting. Notably, Kreutzer's group in Buenos Aires has described (Kreutzer *et al*., 1999) an elegant combined pulmonary arterioplasty and monocusp insertion. Too few of these have been reported to know whether it is a superior technique to that described above.

Repositioning of the Central Pulmonary Arteries Anterior to the Aorta

This innovative and effective technique was first described by Hraska (2000), specifically to address the problems of airway compression in small infants. In his technique, the mediastinal vessels including the ascending aorta, aortic arch and its branches, superior vena cava and the pulmonary arteries must be freely and extensively mobilized. The azygos vein should be divided to further mobilize the superior vena cava. Following repair of the intracardiac lesions, the aorta is transected above the sinotubular junction, and a further transection is made a centimetre or so distally, thus resecting a tubular or triangular segment of the ascending aorta (**Figure 31.9**).

The pulmonary artery is also transected, just above the pulmonary valve. The aorta is then reconstituted using continuous Prolene behind the pulmonary arteries (as in the Lecompte manoeuvre), drawing the aorta downwards and to the left. The pulmonary artery may

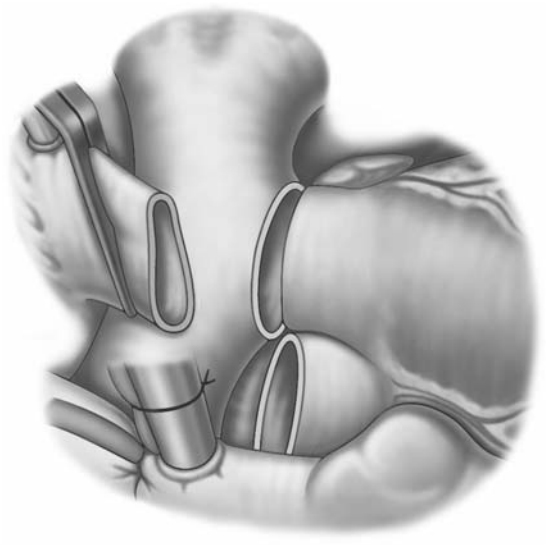

Figure 31.9

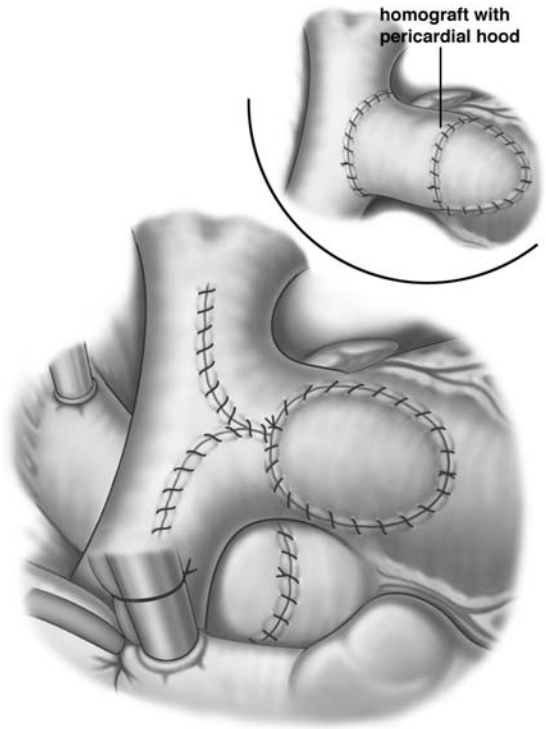

Figure 31.10

then be plicated, resected or attached to a homograft as required to complete the repair (**Figure 31.10**). This technique is especially useful in small infants with major airway problems.

COMPLICATIONS

Complications include the ones that can occur after any repair of tetralogy of Fallot. However, it is the dilatation of the pulmonary arteries, airway compression and presence of any associated bronchomalacia that are responsible for most of the residual complications. The details of how to deal with the airway complications were described in Chapter 22. The altered vascular–bronchial relationships can result in significant distal air trapping and resultant true lobar or segmental emphysema. This condition is particularly common in neonates and small infants, and needs aggressive management. Obviously, repair of the absent pulmonary valve syndrome is the treatment of first choice, with relief of airway obstruction being a primary goal. However, air-trapping may persist and lobectomy has proved necessary (Snir *et al*., 1991) in some patients to allow the remaining lung to function well. It has been suggested that bronchial stenting might have a role, but this technique performs poorly in the presence of external vascular compression (McLaren *et al*., 2005) and should be reserved purely for palliative therapy. In the postoperative period, very careful management of ventilation is required and a clear understanding of the complexities of the pathophysiology is vital to a good outcome.

RESULTS

The results of surgical treatment of absent pulmonary valve syndrome from selected series from major centres are shown in **Table 31.1**. Hew *et al*. (2002) demonstrate clearly in this large series that the risk of death was estimated to be 12 times greater for a neonate and for infants weighing less than 3 kg. Surgery in the current era has reduced the risk, and homograft insertion in this group was protective.

In older children, one might get away with a more conventional repair, without the primary use of a valved conduit, reserving this for proven pulmonary regurgitation in the future. The data do not exist to be certain which is the better approach in this older age group.

The neonate with absent pulmonary valve syndrome remains a formidable challenge for both the surgeon and the intensivist. There are no long-term quality-of-life studies available to inform our practice or advise families as to what the future holds.

Table 31.1 Absent pulmonary valve syndrome: selected reports.

Study	Era	No. of patients			Deaths in hospital			Late deaths (*n*)
		< 1 year (*n*)	> 1 year (*n*)	***Total***	< 1 year *n* (%)	> 1 year	***Total (%)***	
(Fischer *et al*., 1984)	1958–1981	10	5	15	6 (60)	0	40	0
(Ilbawi *et al*., 1986)	1962–1978 1982–1985	5	10	15	1 (20)	0	7	3
(Snir *et al*., 1991)	1982–1988	8	14	22	2 (25)	0	9	0
(Godart *et al*., 1996)	1977–1995	10	27	37	2 (20)	1	8	1
(Hew *et al*., 2002)	1960–1998	39	15	54	8 (21)	0	15	5
(Hraska *et al*., 2002)	1998–2002	5	1	6	0	0	0	0

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32

Double-outlet Right Ventricle

V. T. Tsang and J. Stark

The concept of conotruncal malseptation to explain double-outlet right ventricle (DORV) was proposed by Lev *et al*. in 1972. According to their concept, DORV encompasses a spectrum of morphological disorders from ventricular septal defect (VSD) with overriding aorta, with or without pulmonary stenosis (true DORV) to the Taussig–Bing heart and complete transposition of the great arteries (TGA) with VSD.

We use the term ''double-outlet right ventricle'' for hearts in which more than 50% of each great artery arises from the morphologic right ventricle. It is such a heterogeneous group that some have questioned the usefulness of the term. DORV may be present in the context of abnormalities of atrial situs or atrio-ventricular connection. For example, DORV with an anterior aorta and pulmonary stenosis is the typical ventriculo-arterial connection of hearts with right isomerism and common atrioventricular orifice. This chapter deals with DORV associated with normal atrial situs, concordant atrio-ventricular connection and balanced right and left ventricles.

The main determinant of the haemodynamic situation in DORV, and hence the surgical strategy, is the position of the VSD. This may be subaortic (a), subpulmonary (b), doubly-committed (c) or non-committed (d) (remote from both aorta and pulmonary artery) (**Figure 32.1**). A subaortic VSD is present in tetralogy of Fallot with DORV, whereas a subpulmonary VSD is present in the form of DORV sometimes referred to as a Taussig–Bing heart, in which the physiology is similar to TGA with VSD. It is important to realize that in DORV, the outlet septum is a right ventricular structure, separating the aortic and pulmonary outflow tracts, rather than separating right ventricle from left ventricle, and perhaps might be better termed the ''right ventricular outlet septum'' in this situation.

The terminology used to describe VSD morphology can be confusing and is not always used consistently. It may be helpful to reserve the terms ''subaortic'' and ''subpulmonary'' VSD for hearts in which the relevant artery overrides the trabecular septum. In hearts where

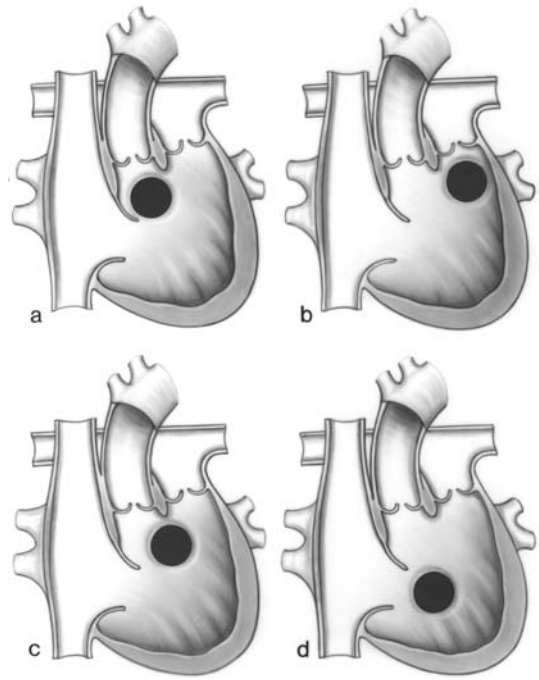

Figure 32.1

both aorta and pulmonary artery arise exclusively from the right ventricle, both great arteries will be surrounded by a ring of muscle (''double conus'' or ''bilateral infundibulum''); in this situation the VSD is usually best described as non-committed, even if it is closer to one great artery than the other. It is necessary to connect each ventricle to a great artery by constructing the surgical baffle within the right ventricle.

Apart from the position and size of the VSD, other determinants of surgical strategy include the relation of the great arteries, obstruction to either aortic or pulmonary outflow tract, and chordal connections of the tricuspid valve. For example, tricuspid valve chordae

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attached to the outlet septum may contraindicate an intraventricular repair which would be feasible. DORV is, therefore, a term which provides a useful and convenient nomenclature only if the details of the highly variable morphology in an individual are also specified.

Many hearts with DORV have other malformations, such as absence of one atrio-ventricular (AV) valve, absence or hypoplasia of one ventricle, complete AV septal defect, total anomalous pulmonary venous connection, and straddling of the mitral or tricuspid valve. In an anatomical study (Wilcox *et al*., 1981), 36% of 63 hearts studied would not have been operable, one-third would have been correctable by means of intracardiac repair and one-third would have required an extracardiac valved conduit.

DIAGNOSIS

Understanding the anatomy of patients with DORV is very important, both for indications for operation and for surgical treatment. If it is accepted that DORV is a spectrum between VSD and complete TGA, it is clear that borderline cases may be difficult to diagnose with certainty. The important features to be assessed are the size of the VSD and its relationship to the origin of the great arteries and tricuspid valve, the spatial relationship of the two great arteries, the presence or absence of pulmonary valvar or subvalvar stenosis or both, and straddling of the AV valve. In the very young, echocardiography should be sufficient to provide the necessary information for surgery, including the coronary and the aortic arch anatomy. In the presence of TGA physiology, echocardiographic guided balloon atrial septostomy is routinely performed at our unit to decompress the left atrium at the time of diagnosis. If PVR is being questioned in the setting of pulmonary overcirculation beyond infancy, cardiac catheterization would be indicated.

INDICATIONS

Indications for operation are similar to those for defects that lie on each side of the DORV spectrum (VSD, tetralogy of Fallot and TGA). The presence or absence of associated cardiac defects may modify the approach. The position of the VSD and the presence and severity of pulmonary stenosis are probably the most important considerations.

Kirklin *et al*. (1986) analysed the data of 127 patients who underwent repair of DORV between 1967 and 1984. On the basis of their detailed analysis, they suggested treatment protocols for each subset of patients with DORV. Some modifications have been made to reflect the current practice of early repair. Children with DORV, a large left-to-right shunt, pulmonary hypertension and heart failure should be treated in the same way as patients with VSD and should undergo early operation.

Taussig–Bing hearts should be corrected using an arterial switch operation and closure of subpulmonary VSD during the first months of life, regardless of the relationship of the great arteries. Any other associated lesions, such as arch hypoplasia or coarctation, can be repaired concurrently.

Patients with DORV and pulmonary stenosis are protected from the development of pulmonary vascular obstructive disease. If the infant becomes severely cyanotic in the first 3 months of life, a systemic-to-pulmonary shunt (usually a modified Blalock–Taussig shunt) is constructed. Intracardiac repair is undertaken within 6–9 months of age. Early primary repair, especially in young infants, may require a more frequent use of trans-annular patch or the use of external valved conduit.

When the VSD is uncommitted, the optimal management must be considered for each individual case. The aim is to create an unobstructed pathway connecting the VSD to the distant aorta without compromising the inflow and the outflow of the right ventricle. Repair in infancy may be feasible, with arterial switch and baffle repair of the VSD. In some cases the VSD may need enlargement. When using an intraventricular tunnel approach, the patch is often bulky and consequently an outflow patch or an external valved conduit may have to be used. For this reason, a two-staged approach may be preferable—a shunt or pulmonary artery banding followed by intraventricular repair in a bigger child.

SURGICAL TECHNIQUE

Double-outlet Right Ventricle with Subaortic Ventricular Septal Defect without Pulmonary Stenosis

It is usually possible to perform an intraventricular repair. The patch does not close the VSD; rather, it forms an intraventricular tunnel to divert blood from the left ventricle, via the VSD, into the aorta. The first repair was performed in 1957 (McGoon, 1961; Kirklin *et al*., 1964).

The operation is performed on cardiopulmonary bypass with hypothermia and cardioplegic arrest. In small infants, deep hypothermia with circulatory arrest may be preferred. The anatomy is first inspected through the tricuspid valve. It is important to find the exact position of the VSD and assess its size and suitability for connecting the left ventricle, through the VSD, to the aorta. Attachment of the tricuspid valve chordae to the infundibular septum can make the repair difficult, occasionally impossible. The chordae can be detached, and then reattached adjacent to subvalvar tissue to preserve the tricuspid valve function.

Repair can be performed through the tricuspid valve. If there is any difficulty exposing the anterior aorta,

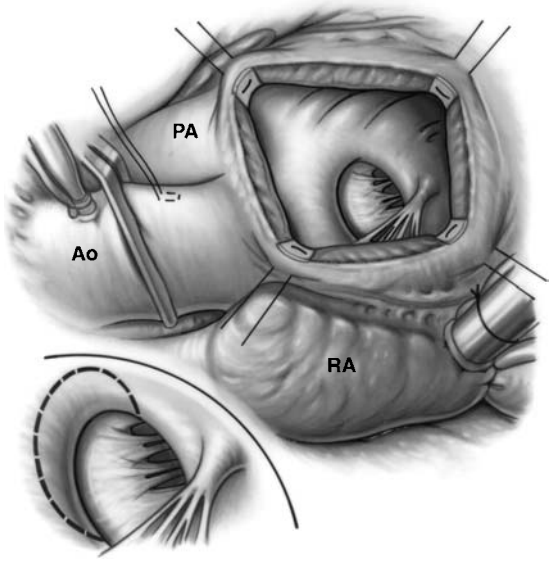

Figure 32.2

the authors use the trans-ventricular approach. If the diameter of the VSD is smaller than that of the aortic valve ring, the VSD is enlarged by excision of the septum in its superior and lateral margin. This area is free from conduction tissue and, if the resection is not too deep, damage to the conduction tissue and the mitral valve is easily avoided (**inset, Figure 32.2**). It is important to excise the septum rather than the anterior ventricular wall because of the proximity of the left coronary artery. The patch can be tailored from a Gore-Tex or Dacron tube (**Figure 32.3**). We select the tube size that corresponds approximately to the size of the ascending aorta. The advantage of this material is its tendency to keep the required curve in the patch. A patch cut from a piece of Gore-Tex or Dacron can kink unless an exact size and shape are used. The patch is inserted with interrupted 5-0 Surgilene (10 mm semi-circular needle) horizontal mattress stitches buttressed with Dacron pledgets. Stitches are placed all around the VSD and around the aortic valve ring to avoid muscular trabeculations. The stitches are then passed through the patch; the authors prefer to start at the midpoint, between the VSD and the aorta (**Figure 32.3a**). The optimal curve of the patch is assessed, and both the inferior and superior portions are trimmed as necessary. Anteriorly, it may be advantageous to bring the stitches through the anterior wall of the right ventricle above the aortic valve (**Figure 32.3b**). In this part, the patch may be secured with a running stitch of 5-0 Surgilene. It is buttressed on the outside of the ventricle with a strip of Teflon felt. The completed intraventricular tunnel is shown in **Figure 32.4**.

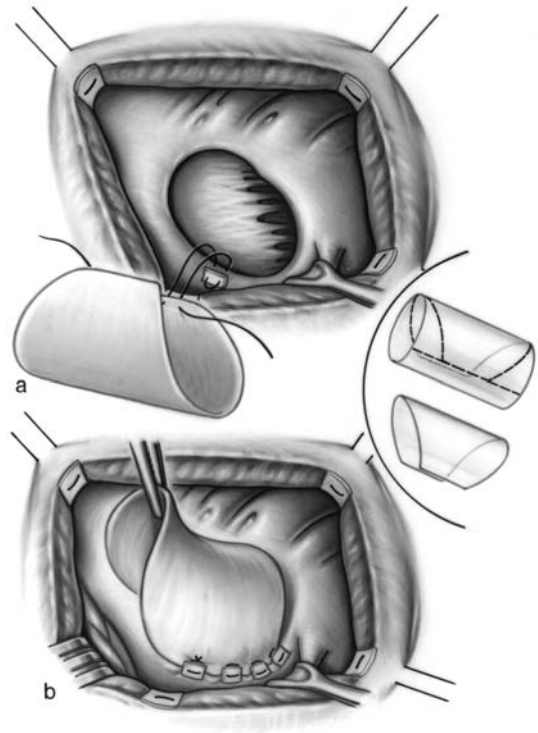

Figure 32.3

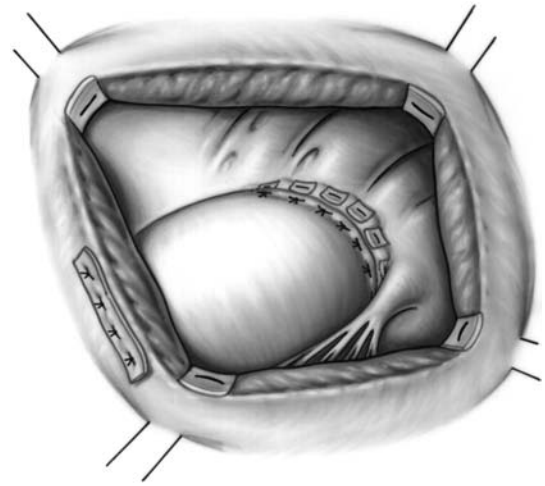

Figure 32.4

Double-outlet Right Ventricle, Ventricular Septal Defect and Pulmonary Valvar or Subvalvar Stenosis

If the pulmonary valve ring is adequate (Rowlatt *et al*., 1963) and stenosis is at the valve level, pulmonary valvotomy through the pulmonary artery is performed.

Intracardiac repair does not differ from that described for DORV without pulmonary stenosis. If, however, subvalvar obstruction is present, a different technique may have to be used. In some patients, infundibular resection and RVOT patching combined with intraventricular repair may be possible. In others, the combination of subpulmonary obstruction and a generous intraventricular patch leaves a residual obstruction from the right ventricle to the pulmonary artery. In these circumstances, either a trans-annular RVOT patch or an extracardiac valved conduit may have to be used. The decision to use one or the other of these methods depends on the position and distribution of the left coronary artery (usually behind the pulmonary artery) and the age of the patient, as discussed earlier.

If a major coronary artery branch runs across the outflow tract of the right ventricle under the pulmonary valve ring, the use of an outflow tract patch is precluded and a conduit must be used. When pulmonary arteriolar resistance is high, which may be the case in patients with long-standing large aortopulmonary shunts or only mild pulmonary stenosis, a valved conduit is preferred.

Double-outlet Right Ventricle with Doubly-committed Ventricular Septal Defect

Most infants with this type of VSD can be treated in the same way as that described for DORV with subaortic VSD. The defect is usually large, and connecting the left ventricle to the aorta does not present special problems.

Double-outlet Right Ventricle with Subpulmonary Ventricular Septal Defect (Taussig–Bing Type)

This lesion was first described by Taussig and Bing in 1949. The aorta is usually situated anteriorly, but the great arteries may lie side-by-side. The VSD is in the subpulmonary position; the pulmonary artery may override it. Pulmonary valve stenosis is rare. Subaortic stenosis, coarctation/arch hypoplasia, and multiple VSDs are not uncommon. These lesions are dealt with at the time of arterial switch. The management of multiple VSDs in the setting of an arterial switch is described in Chapter 36.

Arterial Switch and VSD Closure

Since the advance of arterial switch operation, it is possible to streamline the intracardiac repair in some complex DORV. In the case of Taussig–Bing hearts, arterial switch operation and closure of the VSD are currently considered the operations of choice (Brawn and Mee, 1988; Serraf *et al*., 1991). The authors prefer to use bicaval cannulation to repair the VSD via the right atrium or one of the great arteries without circulatory arrest. The transfer of coronary arteries may be more demanding, as there are often unusual coronary artery patterns. The importance of an adequate infundibular resection to relieve the subaortic stenosis has been emphasized by several authors (Serraf *et al*.,1991; Smolinsky *et al*., 1988). Arch repair for hypoplasia/interruption, if required, is performed using a pulmonary homograft patch with a brief period of circulatory arrest. This would even out the disparity between the great arteries at the time of aortic reconnection (**Figure 32.5**). The transfer of the coronary arteries, which are partially inverted, is illustrated in **Figure 32.6**. We routinely use the Lecompte manoeuvre as long as the neo-aortic root is posterior to the neopulmonary root. Extensive mobilization of the branch pulmonary arteries allows a neopulmonary artery anastomosis without undue pressure on the reimplanted coronary arteries and tension on the pulmonary arteries.

Intraventricular Tunnel

If the great vessels are side-by-side, the left ventricle can be occasionally redirected into the aorta via the VSD in a way similar to that described for subaortic VSD (Kawashima *et al*., 1971). The VSD is enlarged, the subpulmonary part of the infundibular septum between the two great arteries is resected, and a generous baffle is sewn in (**Figure 32.7**). There should be sufficient distance between the tricuspid valve and the pulmonary

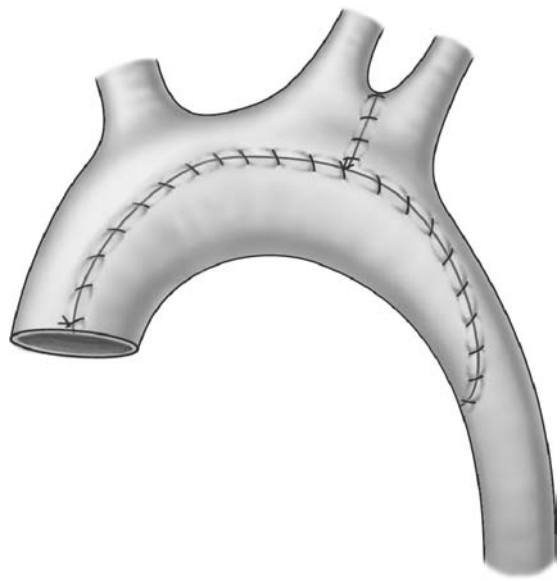

Figure 32.5

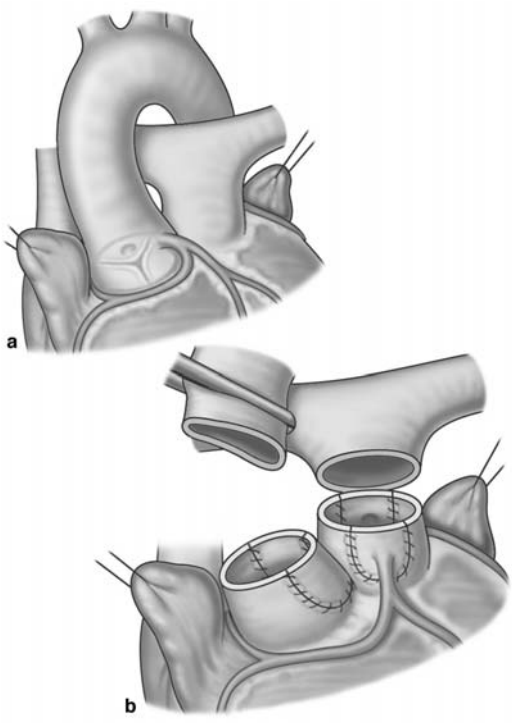

Figure 32.6

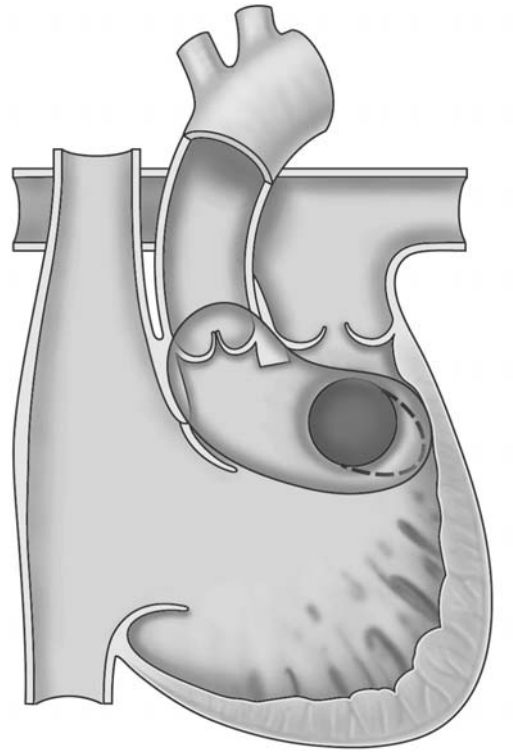

Figure 32.7

root to accommodate the baffle; this is illustrated in **Figure 32.8a** (adequate distance) and **Figure 32.8b** (inadequate distance). An intraventricular patch may be rather long and bulky and may obstruct the outlet from the right ventricle into the pulmonary artery. For this reason, an external valved conduit may be required. The risk of residual subaortic stenosis and the use of a conduit make the tunnel repair rather unattractive.

Double-outlet Right Ventricle with Uncommitted Ventricular Septal Defect

Repair of this variant may be very difficult. The use of a large and long intraventricular baffle almost certainly dictates the use of a RVOT patch or an external valved conduit. The patch may also bulge into the tricuspid valve orifice, which may become obstructed. If the

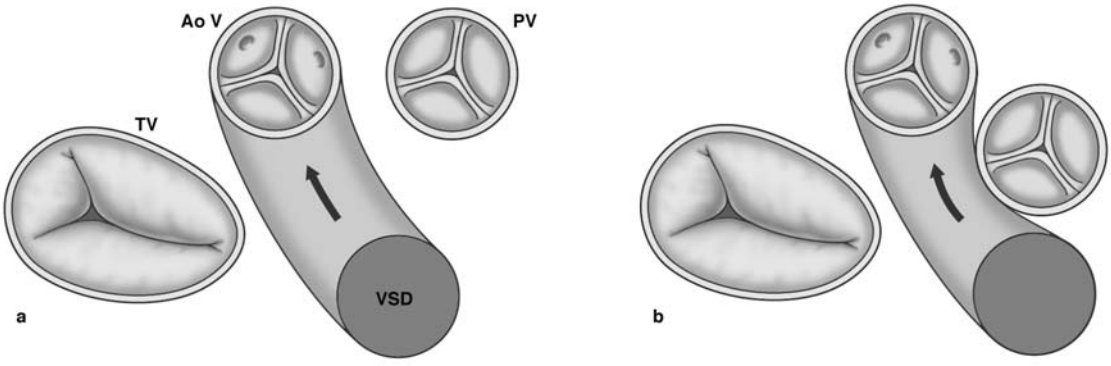

Figure 32.8

uncommitted VSD is small, it must be enlarged. It is important to recognize which type of VSD is present. As described in Chapter 8, the conduction system is located at the medial and inferior margins of the perimembranous and trabecular defects. In muscular inlet defects, the conduction mechanism is located superiorly and laterally. The perimembranous inflow or trabecular defect is enlarged in the superior lateral margins. From the surgeon's view, it is between the 10 and 1 o'clock positions (**inset, Figure 32.9**). The intraventricular patch is usually tailored from a Gore-Tex tube. Occasionally the complete tube may be employed; one end is sutured around the VSD and the other end around the aortic valve ring. An external valved conduit is often necessary in this situation. Defects in the upper part of the muscular ventricular septum are usually far removed from the aortic or pulmonary valve. If small, they have to be enlarged. Enlargement should be done inferiorly (**Figure 32.10**) because the conduction mechanism is located superiorly to the defect (**Figure 32.11**). Use of this long tunnel technique is limited by the presence of tricuspid chordae and by the distance between the tricuspid valve and the pulmonary valve (**Figure 32.8**). It is associated with an important risk of subaortic obstruction. Rerouting into the pulmonary artery followed by the arterial switch seems a more satisfactory solution (Lacour-Gayet *et al*., 2002).

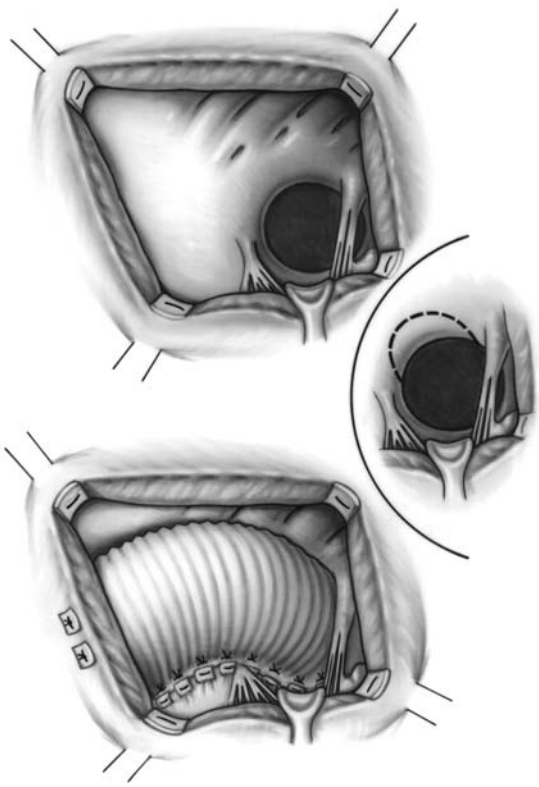

Figure 32.9

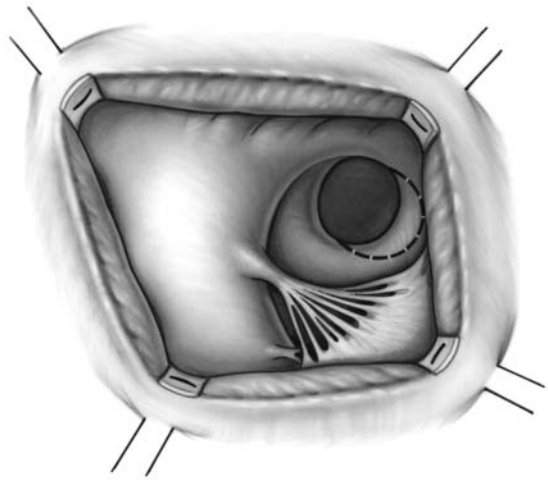

Figure 32.10

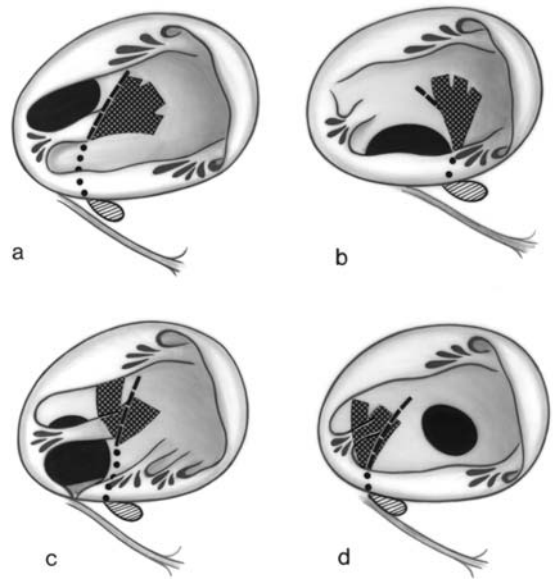

Figure 32.11

When the VSD is distant from the aorta, it is almost always quite close to the pulmonary artery. Rerouting to the pulmonary artery creates a shorter channel, and its use is not limited by the presence of tricuspid chordae or the tricuspid to pulmonary valve distance. However, the

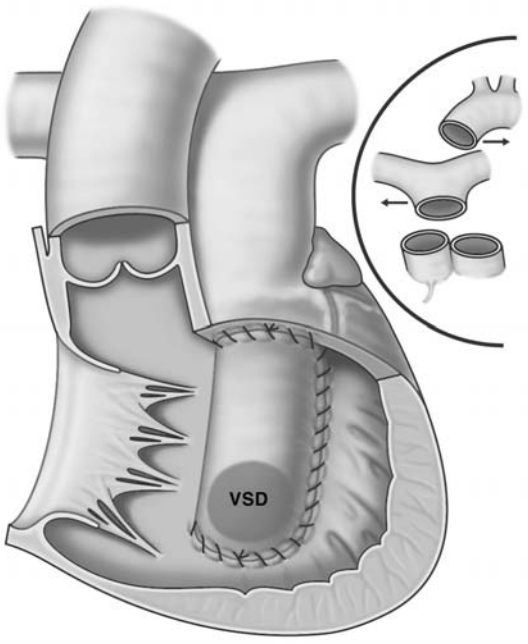

Figure 32.12

arterial switch (**inset Figure 32.12**) frequently involves relocating complex coronaries arteries. Alternatively, some hearts with complex DORV associated with pulmonary stenosis or hearts with straddling of the mitral or tricuspid valve can be treated with total cavopulmonary connection or other variations of the Fontan operation (Delius *et al*., 1996).

COMPLICATIONS

Severe haemolysis may occur after intraventricular repair of DORV with a patch. Residual VSDs, early and late complete heart block, late development of left ventricular outflow tract obstruction, false left ventricular aneurysm and damage of the left coronary artery after enlargement of the restrictive VSD in DORV have been reported.

RESULTS

In the early years the mortality rate for intraventricular repair of DORV was high. It was 50% for DORV associated with subpulmonary defects and 16% for subaortic defects (Harvey *et al*., 1977). Non-committed VSD also carried an increased mortality (Kirklin *et al*., 1986). The year of operation and the position of the VSD were significant factors influencing hospital survival. It was illustrated by the results reported from the Brompton Hospital, London (Musumeci *et al*., 1988). Among 93 patients who underwent reparative procedure between 1973 and 1986, the surgical mortality rate was 27%.

Between 1981 and 1990 we have treated 98 patients with DORV. Of 35 patients who underwent palliative procedures, two (6%) died; of 63 who underwent complete repair, 17 (27%) died. Survival rates were better among 55 patients with restricted pulmonary blood flow (90%) than among 43 patients with high pulmonary blood flow (75%). DORV associated with complete atrioventricular septal defect was successfully corrected by Pacifico *et al*. (1988); nine of 10 patients (90%) survived the operation.

Late results from the Mayo Clinic were evaluated by Shen *et al*. (1990). Among 118 patients undergoing intraventricular repair for DORV between 1965 and 1985, there were 23 hospital deaths (19%); six patients were lost to follow-up observation. Data from 89 late survivors were reviewed: there were 22 late deaths (25%), of which 16 (73%) were sudden. The risk factors for late death included older age at the time of operation, peri- or postoperative ventricular tachyarrhythmias, and complete atrio-ventricular block.

In the current era, the results of repair of DORV with subpulmonary VSD (Taussig–Bing complex) have improved since the introduction of the arterial switch operation for the treatment of this condition. Serraf *et al*. (1991) reported a series of 27 patients undergoing operation between 1978 and 1990. Intraventricular repair was achieved in seven patients; one (14%) died. Twenty patients were treated with the arterial switch operation and VSD closure; one (5%) died. Of the seven patients who underwent reoperation, none died. There were two late deaths in their series. Results from Melbourne on 28 consecutive infants were reported by Comas *et al*. (1996); 11 had associated aortic arch obstruction; staged approach was used in four. The reoperation rate was 73% at 6 years. In the group without arch obstruction, there were two early deaths after one-stage repair. No reoperation was reported up to 6 years. Griselli *et al*. from Birmingham, UK (2005), presented results for Taussig–Bing hearts between 1988 and 2003. Thirty-three patients underwent arterial switch operation and VSD closure. 25% of patients (with arch obstruction) underwent staged repair. Overall, there were three early deaths and one late death. There were two early and four late reoperations.

Lacour-Gayet *et al*. (2002) reported results in patients with DORV and uncommitted VSD using arterial switch approach in 10 patients. Median age at surgery was 16 months. VSD was rerouted into the pulmonary infundibulum. There was one non-cardiac death. At the mean follow-up of 20 months all nine survivors were

in sinus rhythm and had no subaortic gradient more than 15 mmHg.

DOUBLE-OUTLET LEFT VENTRICLE

DOLV is extremely rare (Wilkinson, 2002). Both great arteries originate from the left ventricle, and the 50% rule also applies here. The commonly associated cardiac anomalies include VSD (usually in the subaortic position), pulmonary stenosis, Ebstein's malformation of the tricuspid valve, and hypoplastic right ventricle (Brandt *et al*., 1976). It has been observed with both situs solitus and situs inversus (Urban *et al*., 1977) and with the AV discordance. Indications for operation are similar to those in DORV: early repair or pulmonary artery banding in infants with DOLV and high pulmonary blood flow and systemic-to-pulmonary shunt and subsequent repair, with external valued conduit in patients with associated pulmonary or subpulmonary stenosis.

Because the aorta arises completely from the left ventricle, there is usually no need for a long, tortuous intraventricular baffle. The VSD is simply closed with a patch, the pulmonary artery is transected and oversewn. Continuity between the right ventricle and the pulmonary artery is established with a valued conduit. On occasion, an intraventricular conduit may be placed for directing the right ventricular blood through the VSD into the pulmonary artery (Sakakibara *et al*., 1967). Kirklin and Barratt-Boyes (1986) reported a combined series from their two institutes: of 22 operated patients, five (23%) died.

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33

Mustard Operation

J. Stark

Surgical treatment of patients with transposition of the great arteries (TGA) has evolved rapidly since the last edition of this book. Physiological correction with the Senning (1959) and Mustard (1964) techniques was to a great extent replaced by anatomical repair (Jatene *et al*., 1975). There continue, however, to be a few situations in which either the arterial switch operation (ASO) is contraindicated (pulmonary valve stenosis, non-resectable left ventricular outflow tract obstruction) or the patient presents at an age that is considered too old for ASO and the institution does not perform two-stage ASOs. However, the main indication for physiological repair today is the so-called double-switch operation for congenitally corrected transposition, which is described in Chapter 39. Mustard operation has been largely superseded by Senning. It was generally felt that Senning operation is more reproducible; however, in certain conditions, such as juxtaposed atrial appendages, where the volume of the right atrium is small, Mustard operation is still preferred. In dextrocardia, surgical access is difficult and both procedures would be rather demanding. For these reasons, the techniques of the Mustard and the Senning operation are described in this book.

DIAGNOSIS

The diagnosis of TGA is established by echocardiography. A severe degree of pulmonary vascular obstructive disease may be present in patients with TGA as early as the first year of life, even in the absence of a ventricular septal defect (VSD) or a persistent ductus arteriosus (PDA). Patients with increased pulmonary arteriolar resistance must be evaluated very carefully to assess their operability.

INDICATIONS

Current indications are limited to patients with simple TGA who present after the optimal age for arterial switch operation, to those with pulmonary vascular obstructive disease (as an alternative to palliative arterial switch) and as a part of double-switch operation (see Chapter 39).

SURGICAL TECHNIQUE

The heart is approached through a midline sternotomy. The pericardium is dissected from the pleura and the thymus by both sharp and blunt dissection. The thymus is partially resected. The pericardium is then resected to obtain a large patch (**Figure 33.1**). A small incision is first made near the diaphragm and is extended to the left. A stay stitch is placed and the incision is continued upwards. The incision must be kept away from the phrenic nerves, and care should be taken with the diathermy to avoid phrenic nerve injury. On the left side, a small pocket of pericardium is left for the apex of the heart, as shown in **Figure 33.1**. The pericardial patch is carefully cleaned and placed in heparinized saline. After

Figure 33.1

Surgery for Congenital Heart Defects, Third Edition. Edited by J. F. Stark, M. R. de Leval and V. T. Tsang
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previous intrapericardial operations (atrial septectomy, pulmonary artery banding), there may be some difficulty in obtaining the patch, and sometimes resection of the patch must be completed on cardiopulmonary bypass. The author prefers pericardium to other materials for the patch. If the pericardium is scarred or if holes were made in it during previous operations, synthetic material (Gore-Tex) is used. The patch is cut into the shape of trousers, and its size is estimated according to the criteria suggested by Brom (1975), on the basis of the diameters of the superior vena cava (SVC) and inferior vena cava (IVC) (**Figure 33.2a, b**). The rationale for these measurements is to create a pathway in which at least 50% of the circumference is formed by the atrium and 50% or less is formed by the patch (**Figure 33.2c**). A sufficient amount of the heart's own tissue is left for subsequent growth, and some deposition of tissue on the patch or contraction of the patch should not impair the adequacy of the newly created channel.

The measurements and the way in which the patch is pre-cut are illustrated in **Figure 33.2d**. The distance from points A to B is the distance between the left upper and lower pulmonary veins, which is not measured but only estimated. In a neonate it is about 1 cm; in a 1 year-old, about 1.5 cm; and in an older child, about 2 cm. The distances from points E to D and from points D to F are twice the flat diameters of the SVC and IVC, respectively. The angle α is about 30° and the angle ***R*** is 90°. The distance from points C to D equals:

$$\frac{(\text{E to D}) + (\text{D to F})}{2}$$

It may be necessary to adjust the size of the patch during insertion, because the shape of each atrium differs. The same applies to the length of each of the two legs of the ''trousers''; these are trimmed during insertion, if necessary.

If a PDA is present or suspected, the PDA or ligamentum arteriosum is dissected before the institution of cardiopulmonary bypass and before heparin administration. If access to the pulmonary artery is difficult or if the heart does not tolerate retraction, completion of the dissection and PDA ligation can be accomplished on cardiopulmonary bypass (Chapter 19.)

The aorta is cannulated; a purse-string suture is placed directly on the SVC. It starts about 1 cm above the junction of the SVC and the right atrium to avoid any damage to the sinus node or the sinus node artery, and it should be more oblong than circular, so as to avoid SVC constriction when it is tied after removal of the cannula. Two fine artery forceps are placed inside the suture, the vein is opened with a pointed knife, and the opening is enlarged with another artery forceps. The SVC is then cannulated with a metal tipped right-angled cannula (**Figure 33.3**).

Bypass is started, the IVC purse-string suture is placed as low as possible, and the IVC is cannulated with a metal tipped right-angled cannula. If the heart is irritable, one cannula can be placed into the right atrial appendage and bypass started before caval cannulation. In neonates/small infants, this operation can be performed using DHCA. Only one venous cannula is placed in the right atrial appendage. It is removed during DHCA and replaced after completion of the procedure as a pulmonary venous atrial vent. The rest of the procedure

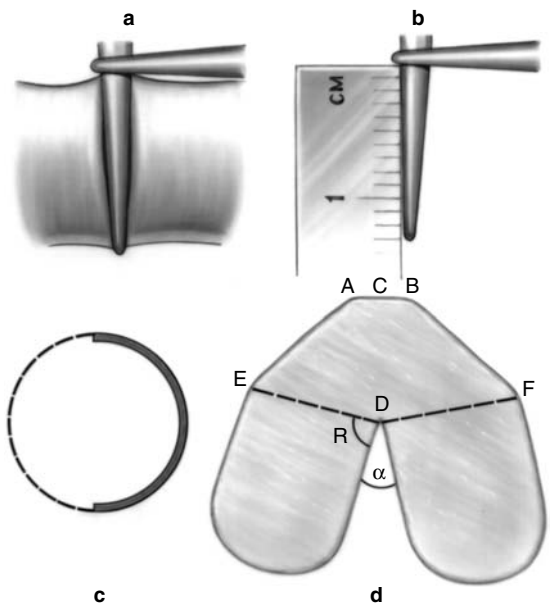


Figure 33.2

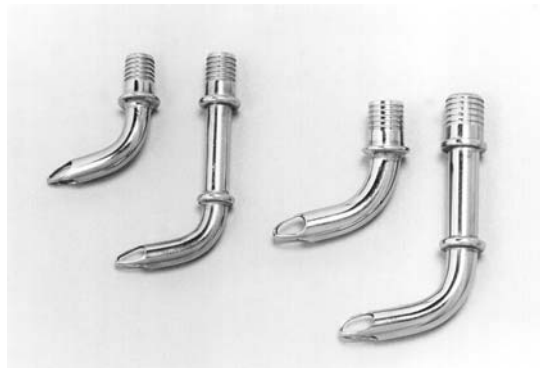


Figure 33.3

is performed on cardiopulmonary bypass using bicaval venous return. At the end of bypass, air is evacuated via the right atrial appendage (physiological left atrium) and through a large-bore aortic needle vent.

Management of perfusion follows the principles outlined in Chapter 11. Surgery is done using moderate hypothermia and cold blood cardioplegia. PFO or ASD is often present, and this should allow the drainage of excessive pulmonary venous return. Oblique right atriotomy (**line b in Figure 33.4**.) is performed. The atrial anatomy is assessed and the atriotomy extended to provided adequate access for insertion of the patch. An alternative is an atriotomy from the base of the right atrial appendage toward the junction of the right upper and lower pulmonary veins (**line a in Figure 33.4**). This incision can be extended across the crista terminalis toward the left atrium if enlargement of the pulmonary venous atrium is desired. The author has used this incision routinely in infants under the age of 6 months and for all older patients in whom the cavity of the right atrium seems to be inadequate (e.g. as in juxtaposition of the atrial appendages) or if the capacity of the pulmonary venous atrium is in doubt. The Y–V incision (**inset, Figure 33.4**) was recommended by Kirklin and Barrat Boyes (1986). After completion of the procedure, the tip of the Y is sutured deep between the right upper and lower pulmonary vein, which changes it into a V shape. Atrial tissue provides for adequate enlargement of the atria. Because multiple incisions in the right atrium could cause arrhythmias, the author does not use them.

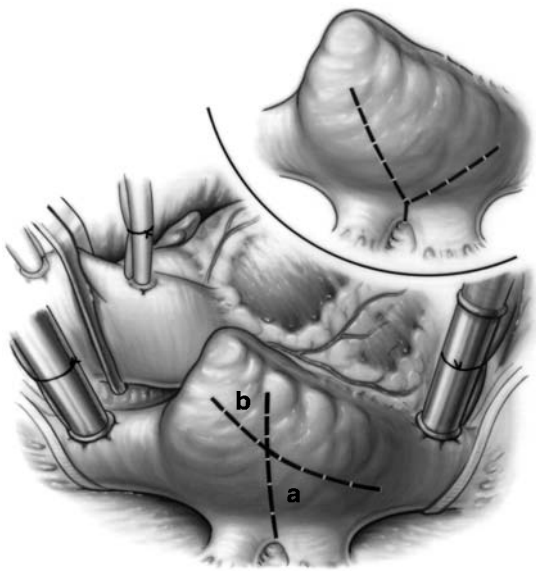


Figure 33.4

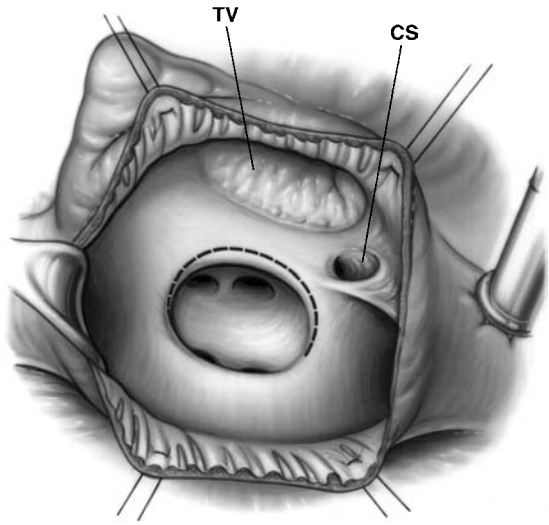


Figure 33.5

Two atrial stay sutures are placed near the crista terminalis on the right side, and two more are placed close to the tricuspid valve ring on the left (**Figure 33.5**). Well-placed stay sutures render the use of intracardiac retractors unnecessary. The atrial septal remnant is excised, as illustrated by the dotted line in **Figure 33.5**. It is important to recognize any perforation of the atrial wall by an overextensive resection. If such a perforation is repaired before discontinuation of cardiopulmonary bypass, it is of no consequence.

Controversy exists about coronary sinus cut-back. We cut the coronary sinus posteriorly and deep into the left atrium in patients in whom the distance between the right lower pulmonary vein and coronary sinus seems short, and also in patients in whom a left SVC drains into the coronary sinus. This cutting back provides a wider IVC pathway. The cutting back of the coronary sinus is usually not necessary when the pulmonary venous atrium is enlarged with a patch. **Figure 33.6** illustrates the excision of the atrial septum close to the coronary sinus. Care is taken not to excise the septum anterior to the coronary sinus, so as to avoid injury to the atrioventricular node. For coronary sinus cutback, scissors are placed inside the sinus and posteriorly toward the left atrium (**Figure 33.6**).

A double-armed 5-0 or 6-0 Prolene suture is used to insert the patch. The first stitch is placed between the left pulmonary veins and the orifice of the left atrial appendage (**Figure 33.7**). On the patch, the suture starts as shown in **Figure 33.7**. The upper suture line then goes around the left upper pulmonary vein and across to the upper edge of the orifice of the right upper pulmonary

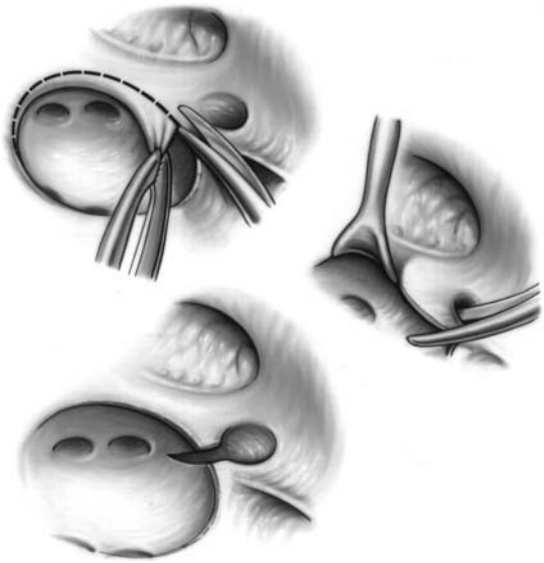


Figure 33.6

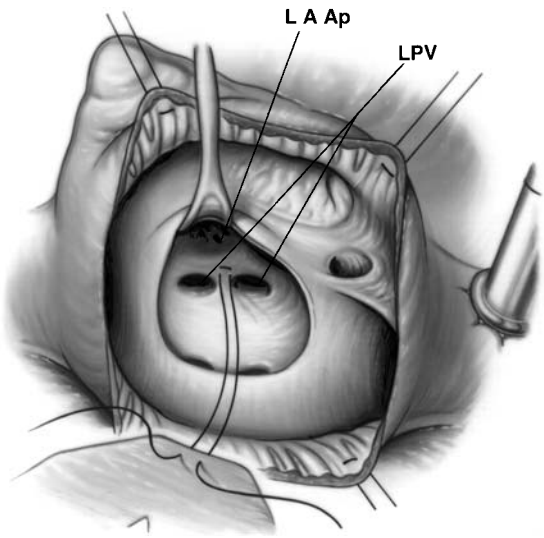


Figure 33.7

vein (**Figure 33.8**). Care is taken not to run this suture line too high on the left atrial wall and to stay away from the cut edge of the atrial septum. If the suture line is placed too close to the cut edge of the septum, an inadequate SVC pathway may result. However, if the suture line stays too deep in the floor of the left atrium, an hourglass configuration may be produced, which could lead to subsequent obstruction of the left pulmonary

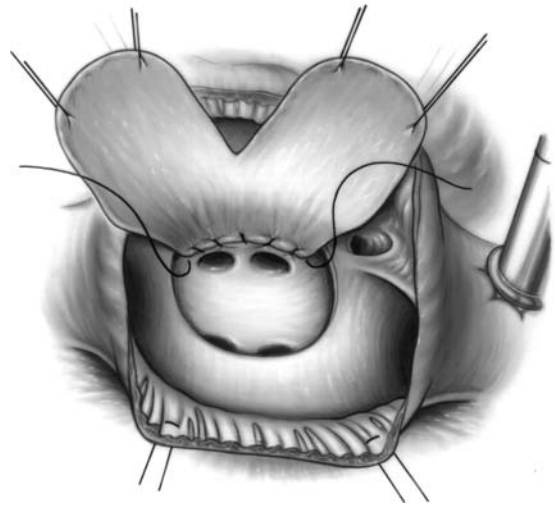


Figure 33.8

veins. The lower suture line is placed around the left lower pulmonary vein and then across toward the lower edge of the right lower pulmonary vein (**Figure 33.8**). It is helpful to place an additional stay suture above and below the right pulmonary veins. Traction on these sutures produces folds inside the atrium, which help the surgeon in following the correct direction when inserting the patch. The two suture lines should diverge toward the SVC and IVC (Stark *et al*., 1974a). If the suture lines come too close together, a constricting ring may be formed, and pulmonary venous obstruction may develop (**Figure 33.9**).

Point D on the patch (**Figure 33.2d**) is then approximated to the edge of the atrial septum. A right-angled dissector pushes the patch from behind toward the lateral atrial wall (**Figure 33.10**). The patch should not touch the lateral atrial wall; if it does, it will balloon excessively and increase the risk of pulmonary venous obstruction. The anterior suture line is then commenced with a second double-armed suture (**Figure 33.11**). Care is taken not to leave communications behind the atrial trabeculations; these may cause atrial shunt later. This problem can be avoided by placing a continuous mattress suture through the wall of the atrium. The lower part of the patch is sutured first to the cut edge of the septum and then through the floor of an opened coronary sinus (**Figure 33.12**). If the coronary sinus has not been cut open, the suture line stays behind it (**inset, Figure 33.12**). The anterior rim of the coronary sinus should be avoided because a suture in this area may damage the tail of the atrio-ventricular node.

If enlargement of the pulmonary venous atrium is planned, an atriotomy from the base of the right atrial

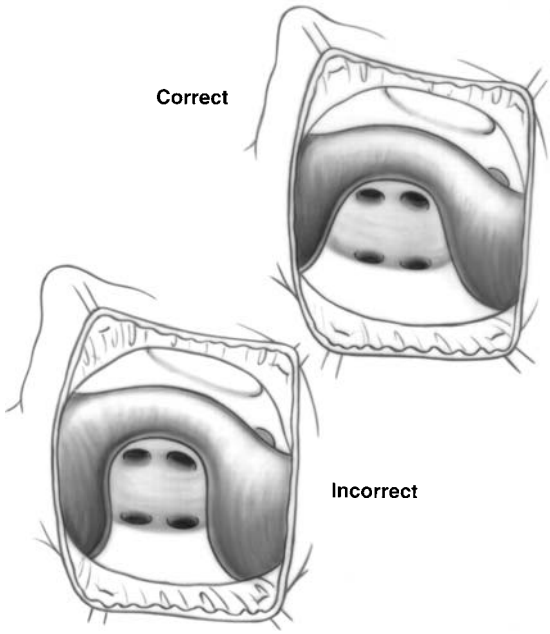
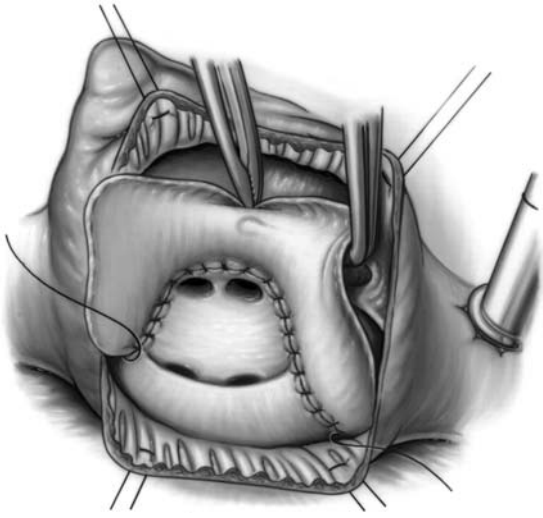


Figure 33.9



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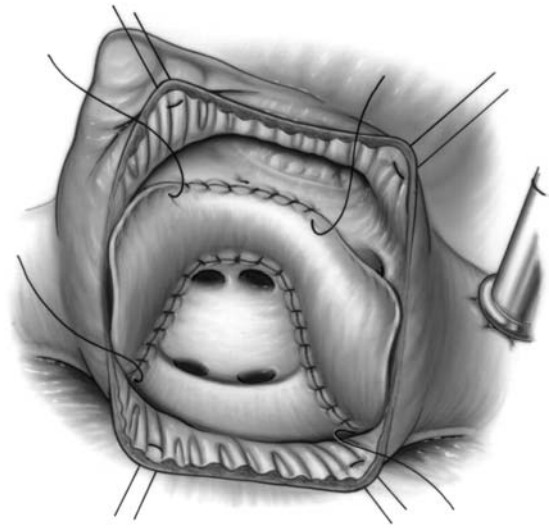


Figure 33.11

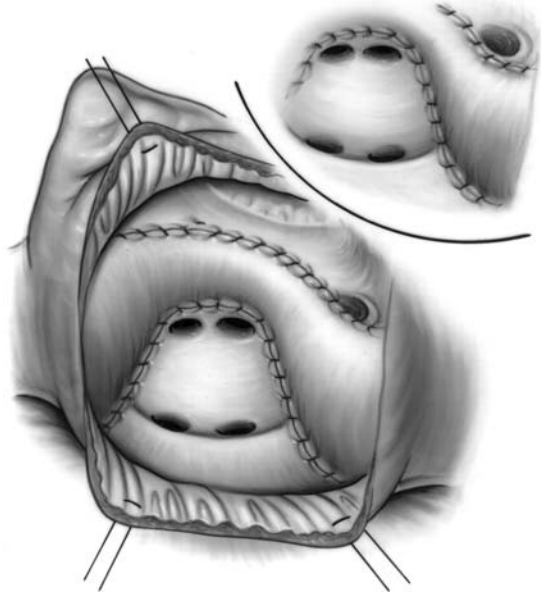


Figure 33.12

appendage to between the right upper and right lower pulmonary veins may be used (line a in **Figure 33.4**). The incision is extended across the crista terminalis, between the veins, to the posterior wall of the left atrium. The atrium is enlarged with a pericardial or Gore-Tex patch. The purpose of the patch is to separate the right upper and lower pulmonary veins; therefore, a wide rather than pointed patch is used (**Figure 33.16b**).

Before the atrial patch or direct closure of the atrium is completed, the caval snares are released, the right atrium (pulmonary venous atrium) is allowed to fill with blood prior to tying down the suture. Air is carefully evacuated, and the aortic clamp is removed. An aortic

needle vent is connected to suction and the patient is completely rewarmed. It is important to keep a slightly positive pressure in the pulmonary venous atrium at this stage. If the pressure becomes negative, air can be sucked into the atrium through the needle holes and embolize into the aorta. Ventilation is restarted and the de-airing procedure is repeated. The aortic vent is then clamped and bypass gradually discontinued. Pressure in the pulmonary venous atrium is kept at 8–10 mmHg by transfusion. Protamine is given, and atrial and ventricular pacemaker wires are placed. The chest is closed in the usual manner with drains.

RESULTS

The Mustard and Senning operations have been largely replaced by the arterial switch operation. Mortality after the Mustard operation reported at the end of the ''Mustard era'' has been 5% or less (Trusler *et al*., 1980; Kirklin and Barrat-Boyes, 1986). The reported long-term results are satisfying. Helbing *et al*. (1994) reported the results after 16 years. Significant independent risk factors for late death were complex transposition and active arrhythmias. Mustard procedure carries a high risk of late sinus node dysfunction (Deanfield *et al*., 1988).

COMPLICATIONS

Both surgical and non-surgical complications of Mustard operations have been described. Surgical complications include constriction or obstruction of the systemic venous pathway (Mazzei and Mulder 1971, Stark *et al*., 1974b), or the pulmonary venous pathway (Stark *et al*., 1972, Clarkson *et al*., 1976). Pathway obstruction used to be related mainly to the surgical technique, with the refinements of the surgical technique, it became rare. Residual or recurrent VSD, and residual or recurrent left ventricular outflow extraction were also described.

Some of these complications may not manifest themselves clinically and may be discovered only on a routine restudy. *Tricuspid valve incompetence* may be related to the VSD closure, may be caused by the malformation of the valve itself or may be secondary to the right ventricular dysfunction. In a series of 563 Mustard operations, tricuspid valve replacement was required in only six patients (Stark and Pacifico, 1989). *Leakages in the baffle* are not common; important leakages necessitating reoperations are very rare. Other surgical complications are discussed in detail elsewhere (Stark and Pacifico, 1989).

Of the non-surgical complications, *dysrhythmias* and right ventricular dysfunction are perhaps the most important. With the refinement of surgical techniques, the incidence of arrhythmias has been greatly reduced. With careful follow-up and with 24 hour Holter monitoring over a long period of time, a gradual decrease in the number of patients remaining in sinus rhythm has been observed after both Mustard and Senning operations (Deanfield *et al*., 1988).

Right ventricular dysfunction (Graham *et al*., 1975) may be the result of inadequate right ventricular protection during the operation or may be the consequence of the right ventricle's functioning as a systemic ventricle. It may lead to the incompetence of the tricuspid valve. Severe right ventricle dysfunction may be treated by heart transplantation or by changing the atrial repair into an arterial switch operation (Mee, 1986).

RE-OPERATIONS AFTER MUSTARD OPERATION

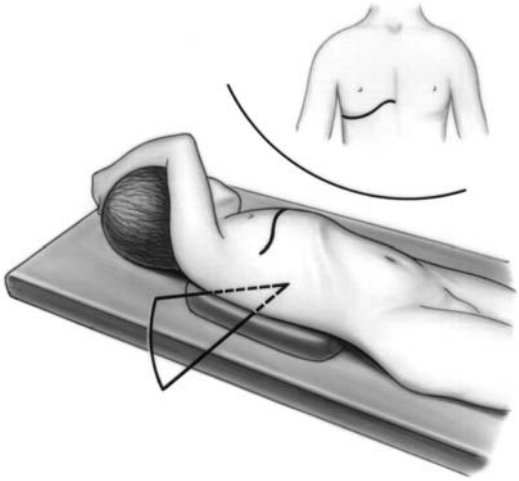
Two approaches can be used. Both sternotomy and right thoracotomy provide good access for operations involving the tricuspid valve or VSD and for systemic and pulmonary venous obstructions (Szarnicki *et al*., 1978). However, the disadvantages of resternotomy are well known but surmountable. One should be aware of the close proximity of the anterior right ventricle to the sternum, dense adhesions in the area of SVC and IVC, and the risk of damaging the right phrenic nerve.

Dissection from a right thoracotomy is not difficult. It is easy to separate the lung from the right atrium, to identify both the remnant of the pericardium and the phrenic nerve, and to dissect the SVC and IVC. It is not necessary to dissect the right ventricle; this avoids the possibility of injury to the coronary arteries. The aorta is usually situated to the right of the pulmonary artery; therefore, dissection and cannulation of the aorta does not present problems either. The right groin may be prepared for all reoperations. The vessels are dissected only if a problem is encountered.

Right Thoracotomy Approach

The patient is positioned with the right side of the chest elevated at about a 45° angle and the right arm is held above the head (**Figure 33.13**). A right-sided anterolateral thoracotomy through the fifth intercostal space is performed. The lung is freed from the heart and the aorta, SVC, right atrium and IVC are dissected.

Purse-string sutures are placed and the vessels are cannulated. Another purse-string suture is placed on the right atrium (pulmonary venous atrium) for placement of a vent (**Figure 33.14**). Cardiopulmonary bypass is started, ventilation stopped, and the patient's temperature is lowered to 32°C. The aorta is cross-clamped, and cold blood cardioplegic solution is infused. An atrial incision is made from the site of the vent insertion down to between the right upper and right lower pulmonary veins (**inset, Figure 33.15**). The vent is advanced through the



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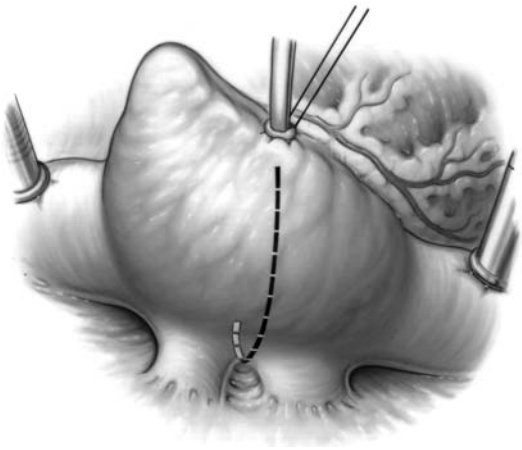

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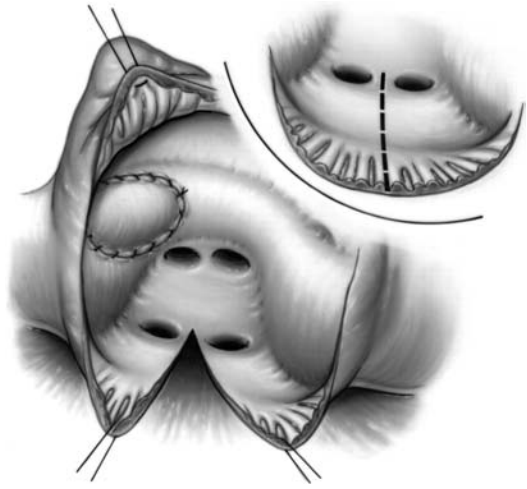

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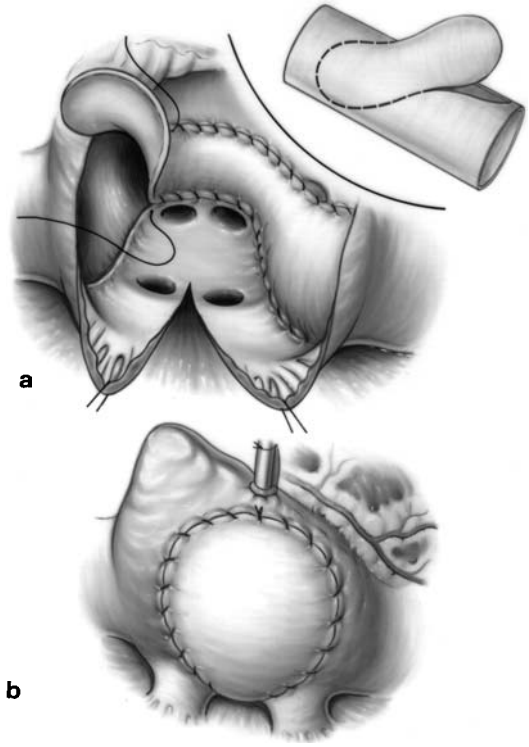

Figure 33.16

tricuspid valve to the right ventricle. Stay sutures are placed on the edges of the atriotomy. In the presence of localized obstruction to the SVC pathway, the narrow part can be incised and enlarged with a small patch (**Figure 33.15**). If both venous channels are compromised, the whole patch is excised, leaving a narrow rim of the ''old'' patch to facilitate secure insertion of a ''new'' patch (**Figure 33.16a**). The patch is tailored obliquely from a Gore-Tex tube (**inset, Figure 33.16**). Because the crimps of the Dacron patch are orientated obliquely, the patch does not kink.

The pulmonary venous atrium is enlarged with a large autologous pericardial patch or Gore-Tex patch (**Figure 33.16b**). When the intracardiac repair is complete, air is carefully aspirated from the ascending aorta, and the aortic clamp is released. If spontaneous cardiac rhythm does not occur, the heart is defibrillated. Further details of re-operations for TGA are discussed elsewhere (Stark and Pacifico, 1989).

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34

Senning Operation

A. D. Pacifico

The arterial switch operation (Jatene *et al*., 1975) has become the procedure of choice for patients with transposition of the great arteries (TGA) in most medical centres (Castaneda *et al*., 1988). Although venous switching may occasionally be employed in some centres for this entity, its principal application is in the double-switch operation for patients with corrected transposition and some other unusual entities (Ilbawi *et al*., 1990; Karl *et al*., 1997; Hibino *et al*., 2001; Langley *et al*., 2003; see also Chapter 39). Knowledge of the venous switch operation described by Senning (1959) and Mustard (1964) should be maintained by surgeons treating patients with congenital cardiac defects. In the Senning operation, viable autologous atrial tissue is used to accomplish intra-atrial transposition of venous return. The development of balloon atrial septostomy by Rashkind and Miller (1966) and the demonstration by Stark *et al*. (1974), that the Mustard technique could be performed during the first year of life without increasing the mortality rate, led to early elective Mustard operations. Brom (Quaegebeur *et al*., 1977) pioneered the revival of the original Senning operation, with some technical modifications, in a series of 20 patients, and this restored interest in this type of venous switching.

INDICATIONS

Indications for operation are identical to those for the Mustard technique of interatrial transposition of venous return (see Chapter 33). The presence of a large atrial septal defect (ASD), previous atrial septectomy or a left-sided superior vena cava do not contraindicate the procedure. The author and associates recognize left-sided juxtaposition of the atrial appendages and dextroversion as contraindications to the standard Senning technique.

SURGICAL TECHNIQUE

The heart is exposed through a median sternotomy incision, and purse-string sutures are placed in the ascending aorta for arterial cannulation and directly upon each vena cava for venous cannulation. When a VSD or some other associated malformation coexists, it is usually repaired initially. The vena caval purse-string sutures are oval in order to minimize any effect toward narrowing the vena cava when they are later tied. If the superior vena cava (SVC) is particularly small, the suture is used only to secure the cannula and later, after removing the cannula, the caval opening is stitched directly with 7-0 Prolene sutures. On occasion, patients less than 3 months old are operated upon through the use of limited cardiopulmonary bypass with a single venous cannula and total circulatory arrest techniques.

Before cardiopulmonary bypass is established, the circumferences of the SVC and the inferior vena cava (IVC) are measured and recorded. Marking sutures are placed on the interatrial groove to define the cephalic (point C in **Figure 34.1**) and caudal (point D) extent of the left atriotomy. These sutures lie at the lateral margin of each vena cava and not posterior to them. A marking 6-0 Prolene suture (point B) is placed a few millimetres cephalad from the junction of the IVC and the right atrium, at a measured distance from the caudal marking suture (point D) on the interatrial groove that is equal to two-thirds of the circumference of the IVC, or at a minimal distance of 15 mm (**Figure 34.1**). This defines the inferior extent of the right atrial incision.

Cardiopulmonary bypass is established with the IVC cannula in place and the SVC cannula (Medtronic Inc., Minneapolis, MN) filled and ready for immediate insertion. The details of the perfusion techniques have been described by Pacifico (1989). For bypass strategy in double-switch operation, see Chapter 39.

While the patient's temperature is lowered to 24°C, ligatures are placed around each vena cava for later occlusion around each cannula, and the midportion of the interatrial groove is dissected about 1 cm left-ward (**Figure 34.2**). This dissection should be confined by the previously placed marking sutures and must not be extended beneath either vena cava; otherwise, it

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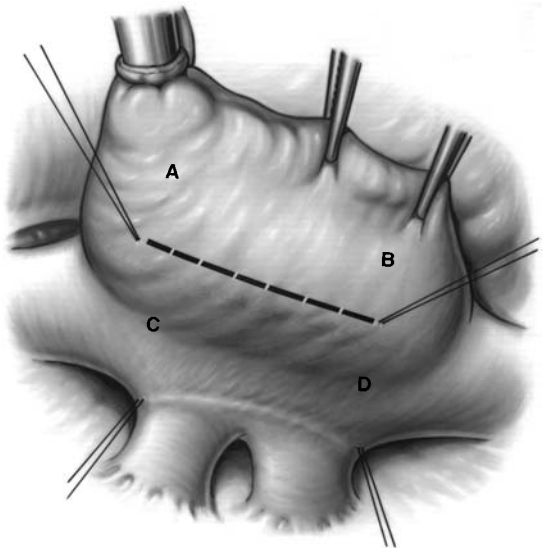

Figure 34.1

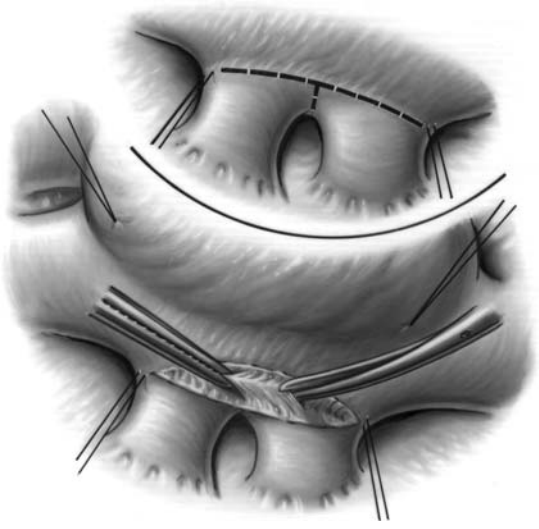

Figure 34.2

may later contribute to caval obstruction and distortion by the suture line used to construct the pulmonary venous pathway. If two venous cannulae are used, a small incision is made into the left atrium, and a disposable sump tip sucker (Medtronic Inc.) is placed into it. The aorta is cross-clamped, and cold cardioplegic solution is infused. If a single venous cannula was used, it is removed from the right atrial appendage after circulatory arrest is induced and partial exsanguination is accomplished.

Systemic Venous Pathway

The right atrium is opened superiorly 1 cm anterior to the crista terminalis (point A in **Figure 34.1**), and the incision is extended anteriorly to the previously placed marking suture (point B) near the junction of the IVC and the right atrium. The left-ward margin of the atriotomy is sutured to the subcutaneous tissue for traction, and the right-ward margin is retracted by two sutures held in a curved clamp. The atrial septum is inspected, and a flap is created from the limbic tissue anteriorly toward the superior and inferior aspects of each respective right pulmonary vein. The flap remains attached at the interatrial groove (**Figure 34.3**). This flap has a trapezoid shape with a defect created by the existing interatrial communication. In rare instances (to be described), a small patch of pericardium or polyester (Dacron) is sutured to the septal flap to make up for this deficiency, leaving a trapezoid configuration (**inset, Figure 34.3**). When the ASD is small and centrally positioned, it is closed primarily before the septal flap is created, which completely avoids the need for foreign material. If a previous surgical atrial septectomy has been performed, a larger patch is sometimes required in order to reconstruct the septum.

The use of non-viable material is most commonly avoided by the creation of a flap of coronary sinus

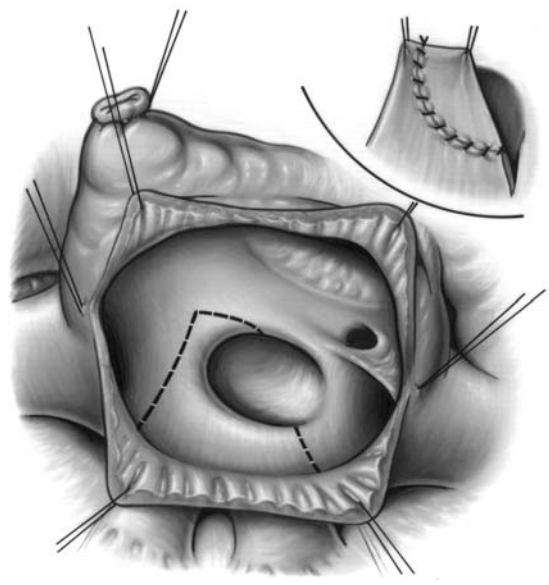

Figure 34.3

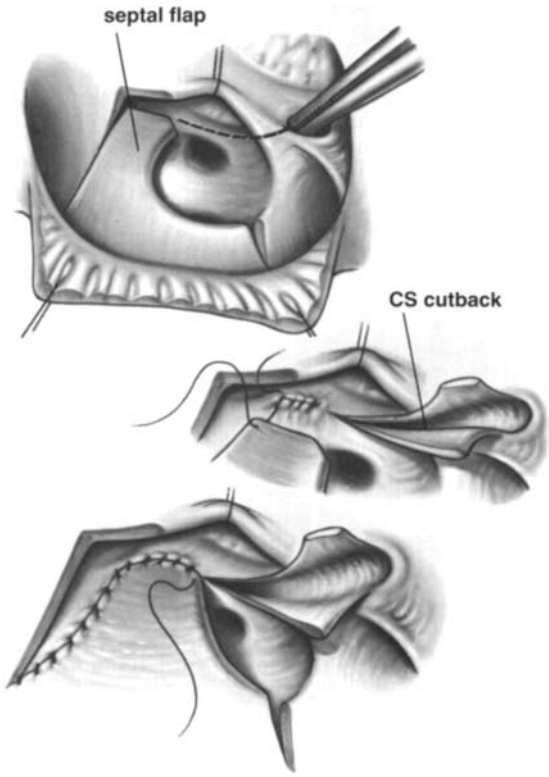

Figure 34.4

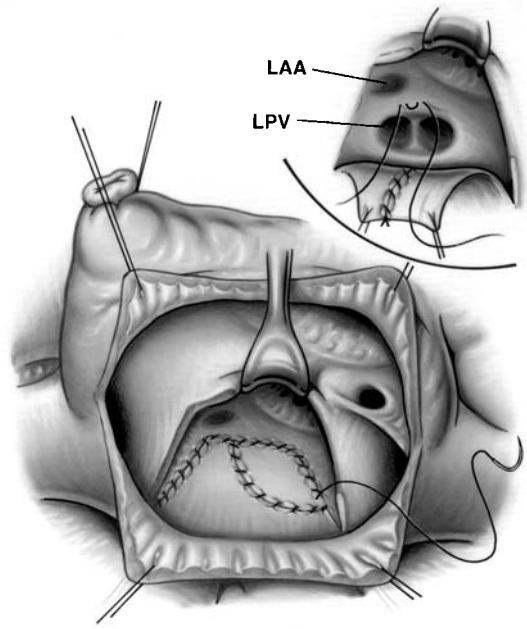

Figure 34.5

tissue by anteriorly incising the common wall between it and the left atrium (**Figure 34.4**) (Pacifico, 1990). The tissue at the left-ward apex of the coronary sinus flap and the left-ward wall of the left atrium, anterior to the left pulmonary veins and posterior to the orifice of the left atrial appendage, is imbricated with a mattress suture to create a small roof above the left pulmonary veins. The coronary sinus flap is then combined with the atrial septal flap to form the roof of the left pulmonary septal flap venous pathway (**Figure 34.4**). This technique has proved useful even after a previous surgical atrial septectomy. The left atrial incision is extended between the marking sutures (points C and D in **Figure 34.1**) on the interatrial groove. If two venous cannulae are used, all of the manoeuvres just described are accomplished with the patient on cardiopulmonary bypass and during core cooling.

In small hearts, it is sometimes convenient at this point to establish a brief period of total circulatory arrest in order to facilitate suturing of the septal flap. A double-armed 4-0 Prolene suture is placed above the left pulmonary veins and brought through the apex of the septal flap (**inset, Figure 34.5**). The inferior border of this flap is sutured within the left atrium to its origin at the IVC junction, and the superior portion is similarly sutured to its origin at the SVC. These suture lines should diverge, so as not to constrict the posterior pathway for left pulmonary venous return (**See Figure 34.5**). Early in the author's experience, he would open the occlusive ligature about the IVC, remove the cannula temporarily, and insert a stent constructed from an appropriate-sized segment of an Argyle chest tube (16-F for infants less than 6 months old, 20-F for infants 6–12 months old, and 24-F for children more than 12 months old) into the IVC. This procedure avoids potential distortion from the IVC tape and cannula during construction of the anterior portion of this venous pathway. Cardiopulmonary bypass is re-established with the patient's temperature at 24°C, after suturing of the septal flap.

The caval pathway is completed anteriorly by stitching (with continuous 4-0 Prolene sutures) the caudal extent of the right side of the free right atrial wall to the atrial tissue about the IVC orifice and continuing to the coronary sinus (**Figure 34.6**). The coronary sinus is left to drain with pulmonary venous blood or, if it has been incised, within the systemic venous pathway. In the latter case, the suture line is placed posterior to the remaining rim of the coronary sinus, so as to avoid the area of the conduction tissue. The stent, if used, is removed from the IVC and passed into the SVC. A second suture is used to complete the superior attachment around the SVC and

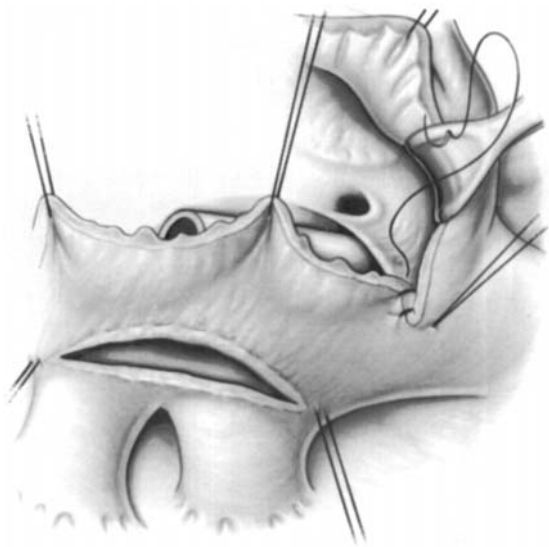

Figure 34.6

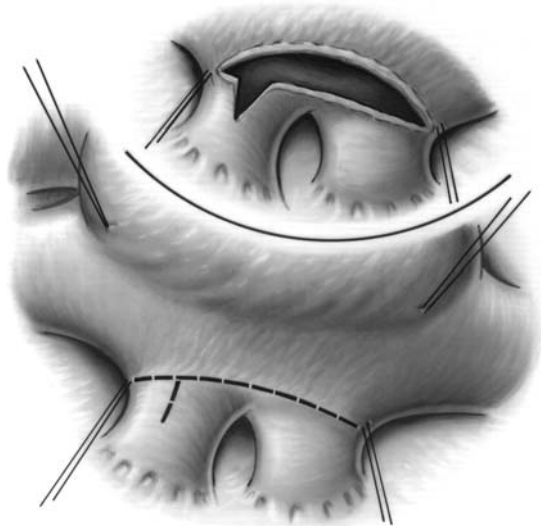

Figure 34.8

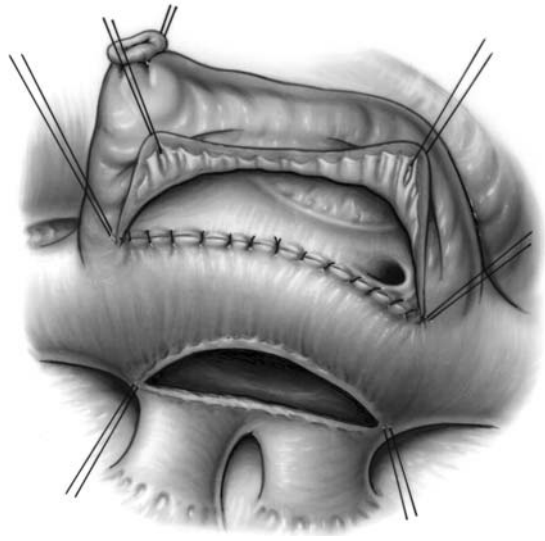

Figure 34.7

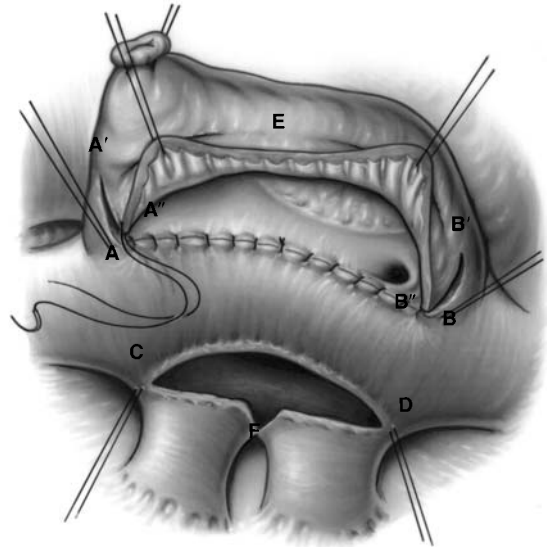

Figure 34.9

along the limbic tissue; the stent is removed before the second suture joins the previously placed inferior suture (**Figure 34.7**).

Pulmonary Venous Pathway

The perimeter of the left atriotomy is extended by incising onto the right superior pulmonary vein for a distance of about 1 cm (**Figure 34.8**). When the right superior and inferior pulmonary veins are more horizontally orientated, the incision is made between them (**Figure 34.9**). The original right atrial incision is extended anteriorly, from points A to A′ and from points B to B′ (**Figure 34.9**). The length of each incision is about one-quarter the circumferences of the SVC and IVC, respectively (or more if required), which increases the perimeter of this flap by half the

circumference of each cava and leaves additional length for attachment above the right pulmonary veins. The pulmonary venous pathway is constructed by initially placing an interrupted 6-0 Prolene suture from the appropriate point E (**Figure 34.9**) on the free right atrial wall (which may not be its midpoint) to the edge of the incision (point F) between the right pulmonary veins. A suture is placed at point A and retracted left-ward while the appropriate site of the cephalad edge of the free right atrial wall (approximately at point A″) is sutured to point C (**Figure 34.10**). Traction in opposite directions tenses the tissue from points A to C, and this segment of the flap is superficially sutured directly on the SVC to avoid injury to the sinus node, which lies in the subepicardial layer. Similarly, sutures at points B and D are placed on opposite traction, and 6-0 Prolene sutures are placed over the IVC pathway, closing this segment of the pulmonary venous pathway (**Figure 34.11**). Unequal tension on the right atrial flap and the caval pathway, or the purse-string effect of a continuously placed suture, must be avoided in order to prevent subsequent narrowing of the caval pathway by these suture lines. In small hearts, interrupted 6-0 Prolene sutures are preferred. Interrupted sutures attach the remaining segment of this flap to the rightward edge of the left atriotomy in order to complete the pulmonary venous pathway (**Figures 34.12** and **34.13**). Interrupted sutures theoretically should not restrict future growth of this area.

When a single venous cannula is employed, it is re-inserted into the morphological right atrial appendage (new left atrium), cardiopulmonary bypass is re-established, and rewarming is commenced. When two venous cannulae are employed, rewarming begins while the roof of the pulmonary venous pathway is

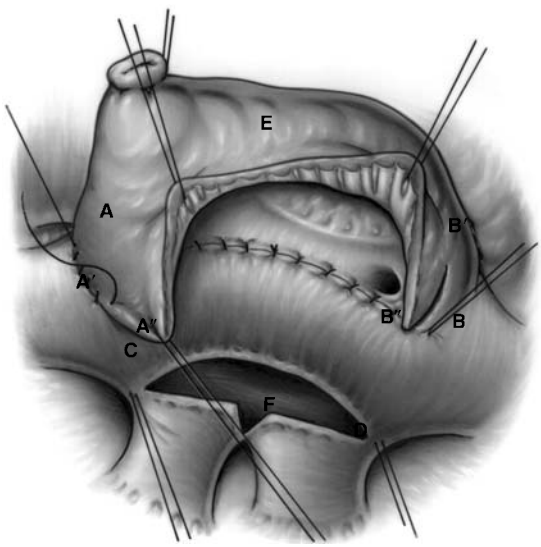

Figure 34.10

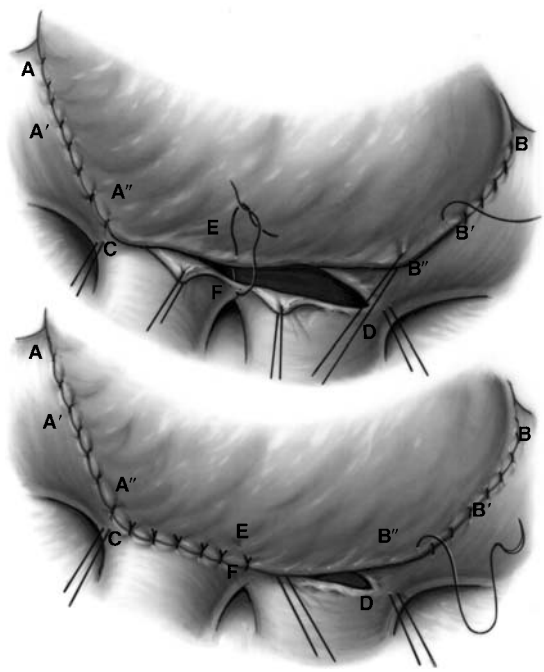

Figure 34.11

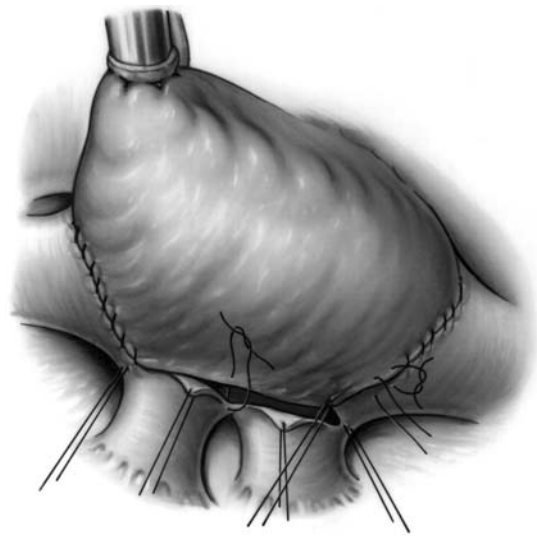

Figure 34.12

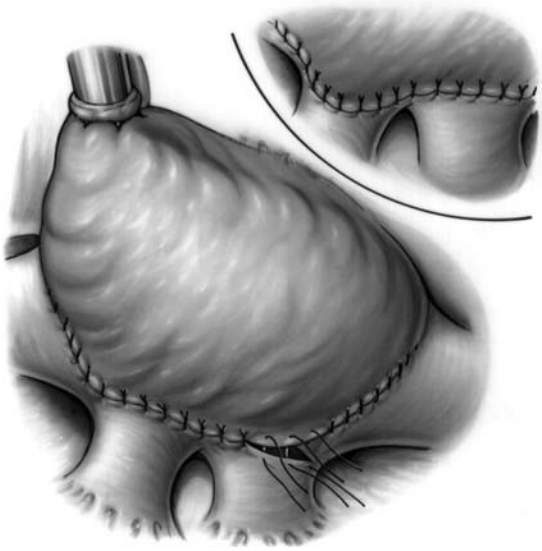

Figure 34.13

being constructed. The usual debubbling techniques are employed as the aorta is declamped. The surgical technique just described differs from that reported by Quaegebeur *et al*. (1977) primarily in that the Eustachian valve is not used in constructing the IVC pathway. The Eustachian valve is well developed in some patients and essentially non-existent in others, and therefore the author and associates prefer not to rely on it. In addition, it is sometimes injured by direct cannulation of the IVC. Avoiding the use of the Eustachian valve requires directing the caudal aspect of the right atriotomy more anteriorly (i.e. farther left-ward from the interatrial groove) in order to leave sufficient right atrial tissue for construction of this segment of the caval pathway. The incision is similar to that originally used by Senning (1959), but a longer perimeter of the remaining free right atrial wall is required for proper construction of the pulmonary venous pathway. This is easily achieved by using an advancement flap technique after making anterior incisions at each end of the original right atriotomy (**Figure 34.9**) or enlarged with an autologous pericardial patch.

RESULTS

Among 146 patients with various forms of transposition of the great arteries from three institutions, the hospital mortality rate after the Senning procedure was 2.7% (Pacifico, 1983). This population consisted of 58 patients operated upon in Leiden, The Netherlands (Brom, 1984); 53 patients operated upon in London (Stark, 1984); and 35 patients operated upon in Bergamo, Italy (Locatelli *et al*., 1979). This rate reflected the early experience with the Senning operation after its revival in 1977 by Quaegebeur *et al*. in Leiden. In general, the early and late results of the Senning operation combined with closure of a large VSD in infants with TGA were not as good as those for infants with TGA and intact ventricular septum.

LATE RESULTS AND COMPLICATIONS

Complications that can occur after the Senning operation include systemic or pulmonary venous obstruction or both, baffle suture line leakage, tricuspid valve incompetence, residual or recurrent VSD or left ventricular outflow tract obstruction or both, arrhythmias, and reduction of systemic (right) ventricular function.

The incidence of ***systemic venous obstruction*** (SVC) after the Senning operation is 0–13% (George *et al*., 1987; Marx *et al*., 1983). This complication can result from technical error during initial construction of the systemic venous pathways and also can be related to narrowing of the perimeter of the SVC by the suture line for the roof of the pulmonary venous pathway. Reoperation to repair systemic venous obstruction is uncommonly required, and in some patients, the obstruction may be managed effectively by percutaneous catheter balloon dilatation.

Pulmonary venous obstruction after the Senning operation has been rare in most series. It developed in six (11%) of 57 patients who underwent the Senning operation at a mean age of 6.6 months (Marx *et al*., 1983). It has not occurred in most series reported in the literature.

Baffle leakage can occur after the Senning operation but is uncommon. It was present in one (2%) of 54 survivors of the Senning operation in one series (Marx *et al*., 1983).

Tricuspid valve incompetence and systemic ventricular dysfunction can also occur after atrial switch operations. In the Boston experience, mild and severe tricuspid valve incompetence each was reported in three patients among 39 survivors of the Senning operation and VSD closure (Penkoske *et al*., 1983). Graham *et al*. (1985) studied right ventricular function after interatrial repair of TGA. The postoperative right ventricular ejection fraction was subnormal (< 0.49) in 16 of 32 patients who underwent the Senning procedure. The mean postoperative right ventricular ejection fraction was lower for patients operated upon during 1971–1974 (0.39 ± 0.11) than for either of the patients who underwent the Mustard operation during 1975–1978 (0.47 ± 0.13) or those who underwent the Senning operation (0.48 ± 0.09). Postoperative right ventricular performance was improved in patients who more recently underwent intra-atrial

repair, which leads to the speculation that younger age at surgery, better preoperative function and improved methods of myocardial protection may be responsible for improved postoperative ventricular performance. Initial pulmonary artery banding and subsequent dismantling of the venous switch procedure coupled with arterial switching has been successfully employed in severely symptomatic patients with severe right ventricular dysfunction (Mee, 1990).

Helbing *et al*. (1994) reviewed the long-term results of 122 patients who received either a Mustard (60 patients) or Senning (62 patients) operation. Median duration at follow-up was 16 years (maximum 25 years) for the Mustard operation, and 11 years (maximum 20 years) for the Senning procedure. There were no reoperations except for pacemaker implantation, and no differences were found between the two groups concerning baffle-associated problems, right ventricular failure, sudden death (6% in both groups) or functional status. However, survival in the absence of rhythm disturbance at 16 year follow-up was 18% for the Mustard group and 53% for the Senning group ($p < 0.001$).

The Senning operation is an excellent technique for venous switching and is of significant historical interest. Its application in the modern era is limited and principally warranted in double-switch procedures for some patients with corrected transposition and other selected unusual malformations.

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35

Surgery for Complex Transposition

V. T. Tsang and J. Stark

The majority of patients with simple transposition of the great arteries (TGA) and atrial septal defect (ASD) with or without persistent ductus arteriosus (PDA) are treated during the neonatal period with an arterial switch procedure (Chapter 36). We use primary arterial switch up to 2 months of age, with mechanical assist as a standby to support the LV if required (Kang *et al*., 2004). Older infants may be best treated by a Senning operation, although a rapid two-stage arterial switch to prepare the low pressure, thin-walled LV has been advocated by the Boston group (Jonas *et al*., 1989). For patients presenting late with simple TGA and pulmonary hypertension, palliative atrial or arterial switch can be employed.

TGA associated with other cardiac anomalies presents different challenges. In some patients, additional malformations must be corrected at the time of arterial repair. In other patients, correction by these techniques may not be feasible, and an alternative strategy may have to be employed. In this chapter, the discussion focuses on TGA associated with the following malformations:

1. Ventricular septal defect (VSD).
2. Pulmonary vascular obstructive disease (PVOD).
3. VSD and left ventricular outflow tract obstruction (LVOTO).
4. LVOTO and intact ventricular septum.

TGA AND VSD

About 30% of all patients with TGA have associated VSD (Fyler, 1980). About one-third of the VSDs are perimembranous, and another one-third are outlet muscular VSDs with malalignment. The remaining VSDs comprise other muscular VSDs, and the rare subarterial types. The aorta tends to be smaller than the pulmonary artery in the presence of VSD, and the discrepancy between the great vessels is greater with anterior malalignment VSD. In contrast, in the presence of posterior deviation of the outlet septum, LVOTO is common.

Infants with TGA and VSD are usually mildly cyanotic but with signs of heart failure due to the transposition physiology, the lowering of pulmonary vascular resistance soon after birth and pulmonary recirculation. Pulmonary plethora and cardiomegaly are evident on chest X-ray.

The diagnosis is made by echocardiography. The coronary anatomy, the size and the relationship of the great vessels are delineated. The VSD, the LV outflow tract, the aortic arch and the ventricular cavity size are evaluated. These investigations are considered sufficient for indicating surgery during the first 3 months. BAS may not be necessary in the presence of VSD, but the advantages of improved mixing at atrial level and decompression of left atrium should be considered. As pulmonary vascular disease develops early in infants with TGA and VSD (Clarkson *et al*., 1976), operation during the first 2 months of life is advisable.

When coarctation is present, primary or staged repair may be performed. For staged repair, coarctation can be repaired from a left thoracotomy, and the pulmonary artery banded in the presence of a significant VSD. An arterial switch operation and PA reconstruction can be performed within a few weeks. For primary repair, the patient is placed on cardiopulmonary bypass through a midline sternotomy. The aortic arch is dissected during the cooling period and then repaired from the front. The arterial switch operation and closure of the VSD completes the procedure (Sano and Mee, 1990; Hazekamp *et al*., 1991). Primary repair is a more attractive option when there is an important aortic arch hypoplasia.

The arterial switch procedure is described in Chapter 36. Using bicaval cannulation, most VSDs can be closed through the tricuspid valve by using techniques described in Chapter 26. The approaches to the different types of VSD in TGA/DORV are separately described in Chapter 32. In small neonates, single right atrial venous cannulation and a short period of deep hypothermic circulatory arrest can be used for the VSD closure. The intracardiac repair is usually performed before the coronary reimplantation. A synthetic patch may prove to be somewhat stiff in neonates and young infants; for

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this reason, we prefer to use the patient's own untreated pericardium (in small neonates), bovine pericardium, or a thin (0.4 mm thickness) Gore-Tex patch. Continuous suture with 6-0 or 7-0 Prolene is employed. Care must be exercised not to distort or damage the tricuspid valve or its chordae.

Multiple VSDs are usually small and do not require closure. If haemodynamically significant (rarely), they should be closed. Some muscular VSDs located near the apex may not be accessible through the tricuspid valve. In such cases a small apical right ventriculotomy can be used to close a large defect. Occasionally, banding of the neopulmonary artery may be required. However, the discrepancy between the great arteries in TGA/VSD with a relatively smallish right ventricular outflow tract following arterial switch may provide some impedence to pulmonary overcirculation without banding of the neopulmonary artery. Spontaneous reduction in size and closure of those muscular VSDs may follow. If the VSDs remain significant at follow-up, closure may be indicated. Some muscular VSD can be closed by interventional procedure.

Results

A mortality rate of less than 10% was reported in earlier years (Bove *et al*., 1988; Brawn and Mee, 1988; Planche *et al*., 1988). Duncan *et al*. (2004) reported recent experience during 1993–2001 on 35 infants with TGA and VSD. There were no hospital deaths in their series. We operated on 26 infants during 2000–2004 with one death. This infant had left and right intramural coronary arteries.

TGA AND PULMONARY VASCULAR OBSTRUCTIVE DISEASE

TGA, VSD and Pulmonary Vascular Disease

The development of pulmonary vascular obstructive disease (PVOD) is faster and the incidence more common among children with TGA and associated VSD with or without PDA. Most factors presumed to contribute to the development of PVOD are present in those patients: high pulmonary blood flow, high pulmonary artery pressure, hypoxaemia and polycythaemia.

This should not delay an early arterial switch procedure. In patients who are diagnosed late, severe PVOD may be present. Cardiac catheterization will establish the degree of increased pulmonary arteriolar resistance. As a guide, patients with a pulmonary arteriolar resistance of more than 8 units/m^2 are considered inoperable. Those with a resistance of 6–8 units/m^2 are evaluated individually. Some patients with gross elevation of pulmonary vasculer resistance who are subjected to surgery may survive; however, pulmonary vascular disease usually progresses, and late morbidity and mortality rates are high (Nakajima *et al*., 1996).

In patients who are diagnosed late, a severe degree of pulmonary vascular obstructive disease may have developed. Such patients can be treated with a palliative repair (Mustard, Senning, or arterial switch) without closure of the VSD. The concept of the palliative Mustard operation (Lindesmith *et al*., 1972). consisted of atrial redirection of venous return but leaving the VSD open. It was presumed that the VSD could decompress the left ventricle if the pulmonary arteriolar resistance suddenly increased, for example during intercurrent lung infection. The child could become temporarily more cyanotic but less likely to die suddenly in heart failure. The same principle may be applied to other operations, and a palliative Senning or arterial switch operation may be performed.

The operative technique used in the palliative Senning, Mustard or arterial switch operation does not differ from that described in Chapters 33, 34 and 36.

Results

The hospital mortality rates after palliative atrial repair have been low. Dhasmana *et al*. (1985) evaluated a group of 41 patients who had undergone the palliative Mustard operation between 1973 and 1980. The VSD was not closed in 34 patients, and VSDs were created in seven others. There were three in-hospital and two late deaths. Survivors were observed for 3–10 years. Clinical improvement was striking in most patients. Arterial oxygen saturation increased and the haemoglobin decreased. Eighteen patients were in New York Heart Association functional class I, 17 were in class II, and only one was in class III. No change in pulmonary artery pressure and resistance was noted in 21 recatheterized patients. Burkhart *et al*. (2004) reported similar results in 28 patients with palliative atrial switch between 1965 and 2000. The improvement in arterial oxygen saturation and quality of life in selected patients with TGA, VSD and severe PVOD was significant.

TGA, Intact Ventricular Septum and Pulmonary Vascular Disease

Severe PVOD can be present during the first year of life even in patients with TGA and intact ventricular septum. Our group extended the concept of the palliative Mustard operation to children with TGA and intact ventricular septum (Stark *et al*., 1976). The Mustard, Senning or arterial switch operation was performed, and a VSD was created in the apical portion of the interventricular septum.

The heart is elevated, and a small ventriculotomy is created at the apex of the left ventricle, between the branches of the coronary arteries (**Figure 35.1**). The anatomy of the right ventricle is assessed through the tricuspid valve, and a suitable site for creation of a VSD is selected in the lower part of the septum. It is important to avoid damage to the papillary muscles and chordae of the tricuspid valve. A needle is therefore passed through the septum from the left side. The heart is elevated again, and the septum is excised around the needle (**Figure 35.2**). The left ventricle is closed with 4-0 Prolene sutures in two layers, and the atrial or arterial switch is performed. The aortic clamp is removed, the heart is de-aired, the patient is completely rewarmed, and perfusion is terminated. We believe that in children with TGA, PDA and PVOD, the PDA does not provide an adequate vent, and therefore in such patients a VSD is also created. At the time of discharge from the hospital, arterial oxygen saturation is usually 85–90%.

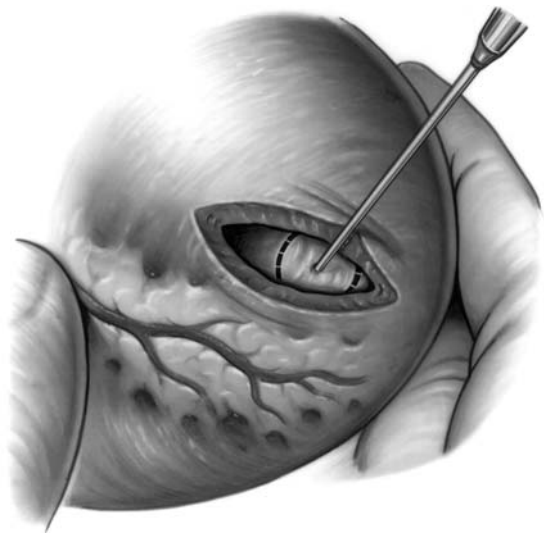


Figure 35.2

TGA, VSD AND LVOTO

Obstruction of flow from the left ventricle to the pulmonary artery may occur at various levels and may be caused by several anatomical structures. Fibromuscular tunnel, fibrous diaphragm, abnormal insertion or abnormal movement or both of the mitral valve, aneurysm of the membranous interventricular septum or tricuspid valve tissue and pulmonary valve stenosis have all been described as causes of LVOTO. The development of an anatomical correction using an RV-to-PA conduit (Rastelli, 1969; Rastelli *et al*., 1969) improved the survival rate of these patients. However, the rate of required reoperations/reintervention is disappointing, even in the medium term (Lee *et al*., 2004).

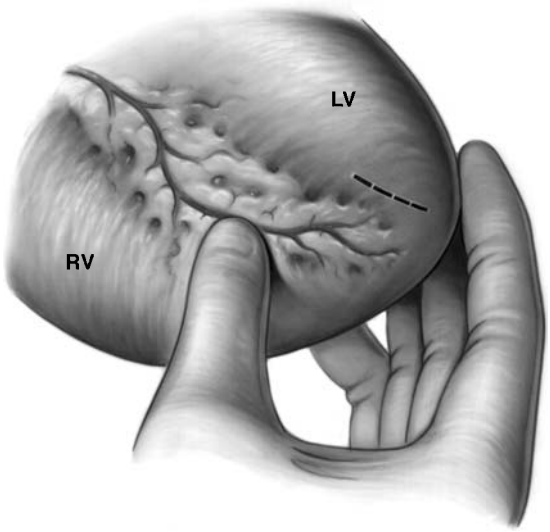


Figure 35.1

Recent data (Wernovsky *et al*., 1990; Sohn *et al*., 1998) and our own experience has demonstrated that pulmonary valve and LVOT abnormalities are not contraindications to arterial switch operation and concurrent direct relief of LVOTO in selected patients. The anatomical criteria of LVOTO are determined. The left ventricular to pulmonary artery gradient alone should not be the basis to judge the severity of the obstruction. The presence of right to left flow across the VSD infers that the obstruction may not be severe and might be amenable to resection or valvotomy. In the presence of severe, fixed LVOTO, it may still be necessary to consider the Rastelli procedure. We try to avoid assigning patients with TGA and LVOTO to treatment strategies less satisfactory than the arterial switch procedure. However, late outcome data in terms of the neo-aortic valve regurgitation remains a concern in these patients (Sharma *et al*., 2002).

The Lecompte procedure (Lecompte *et al*., 1982), an alternative to the Rastelli operation, involves extensive resection of infundibular septum between the aortic orifice and VSD. Pulmonary artery is then reimplanted directly on the RV without an extracardiac conduit. Utilization of the Lecompte manoeuvre depends on the position of the great arteries (Vouhe *et al*., 1992).

In patients in whom a Rastelli operation is considered the best option, a systemic-to-pulmonary artery shunt may be required in the presence of severe cyanosis

in infancy. It is important to ensure that interatrial communication is adequate when a shunt operation is performed. For this reason, a right-sided shunt is preferred, so that atrial septectomy, if required, can be performed at the same time. The shunt operation carries a low risk and enables the surgeon to delay the Rastelli operation until the child is bigger and a larger conduit can be used. Weight above 10 kg is desirable.

Children with a VSD in the trabecular muscular septum in whom the distance between the VSD and the root of the aorta is long may not be suitable for the Rastelli operation. Short chordae crossing the VSD, straddling of the tricuspid or mitral valve, abnormal attachment of the tricuspid valve chordae to the outlet septum, or a combination of these conditions may also preclude creation of an adequate intraventricular tunnel. Under such circumstances, the patient can be treated with total cavopulmonary anastomoses (de Leval *et al*., 1988). Another alternative treatment consists of closure of the VSD, atrial redirection of flow (Mustard or Senning operation), and left-ventricle-to-pulmonary-artery conduit.

Rastelli Operation

The principle of the Rastelli operation is illustrated in **Figure 35.3**. **Figure 35.3a** illustrates the anatomy of TGA, VSD and LVOTO. The VSD is not closed but is used to connect the left ventricle to the aorta with a large prosthetic patch (**Figure 35.3b**). The pulmonary valve is oversewn, the pulmonary artery is ligated or transected, and continuity between the right ventricle and pulmonary artery is re-established with an external valve-bearing conduit (**Figure 35.3c**). This conduit can be an aortic or a pulmonary homograft with a Gore-Tex graft extension. Heterografts, conduits containing prosthetic valves or valveless conduits are other, although less optimal alternatives.

The operation is performed on hypothermic (28°C) bypass. The pericardium is opened on the right side to create a flap which can be used later for covering the RV–PA conduit. The aorta and pulmonary artery are dissected and completely separated. Purse-string sutures are placed for cannulation of the aorta, the superior vena cava and the inferior vena cava, as shown in **Figure 35.4**. At this stage, a shunt, if previously constructed, is identified and prepared for occlusion by ligation or by a Ligaclip. It is preferable to start dissecting the shunt when all purse-string sutures are placed. Should any bleeding occur during the dissection of the often dense adhesions, heparin may be given and the heart cannulated. Dissection may be then completed while the patient is cooled on bypass. The aorta is cross-clamped.

Cold blood cardioplegic solution is administered to the root of the aorta. Caval snares are tightened, the right atrium is opened. The left side of the heart is decompressed with a sump placed through the patent foramen ovale (PFO)/ASD into the left atrium. The intracardiac anatomy is then assessed through the right atrium and tricuspid valve. The position and the size of the VSD are noted, and any chordal

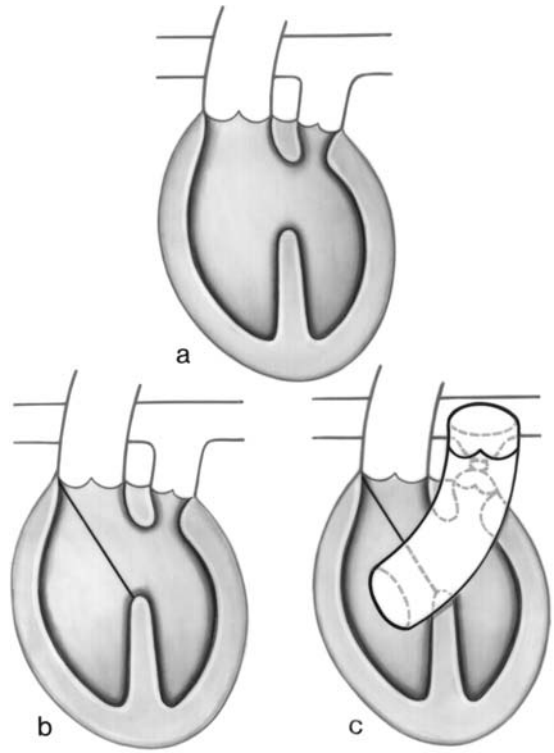


Figure 35.3

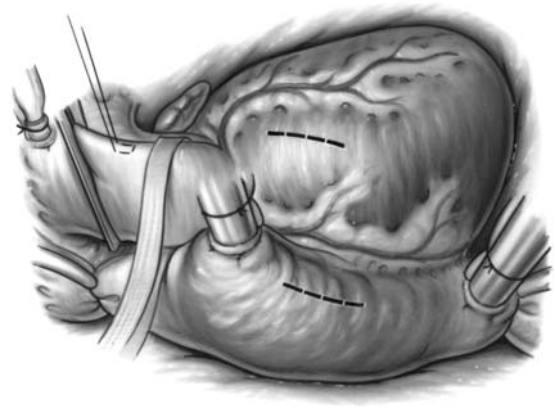


Figure 35.4

attachments of the tricuspid valve into the infundibular septum are looked for. These attachments may preclude creation of the interventricular tunnel. If the surgeon is sure that the procedure is possible, the optimal place for the right ventriculotomy is selected from the right atrial approach. Ideally, the incision should be oblique, high up on the ventricle. It should be pointing to the left so that once the conduit is inserted, it will not cross the midline (**Figure 35.4**).

In most patients, the pulmonary artery is behind the aorta and slightly to the left; placement of the conduit on the left side is therefore straightforward. When the pulmonary artery is to the right of the aorta, a conduit to the main pulmonary artery anastomosis is performed on the right side. Then the conduit is brought under the aorta to the left side. Anastomosis of the conduit to the ventricle is then on the left side to avoid conduit compression by the sternum. Under such circumstances, it is important to ensure that the left pulmonary artery does not kink.

The VSD is inspected, and if it is smaller than the aortic root, it is enlarged to provide a wide outflow from the left ventricle. Either a wedge resection (**Figure 35.5**) or a simple incision in the laterosuperior margin is made (**inset, Figure 35.5**). It is possible to enlarge even a small, restrictive VSD to a suitable diameter if the VSD is located in the upper interventricular septum. We have enlarged the VSD in 97 (89%) of 109 patients undergoing Rastelli operation between 1971 and 1990, without adverse effects.

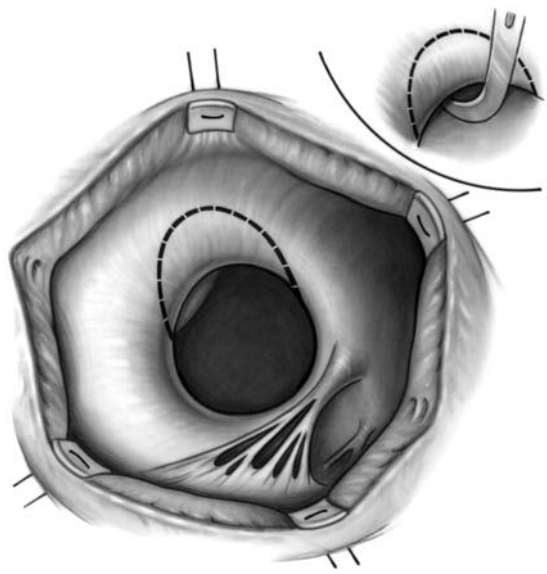


Figure 35.5

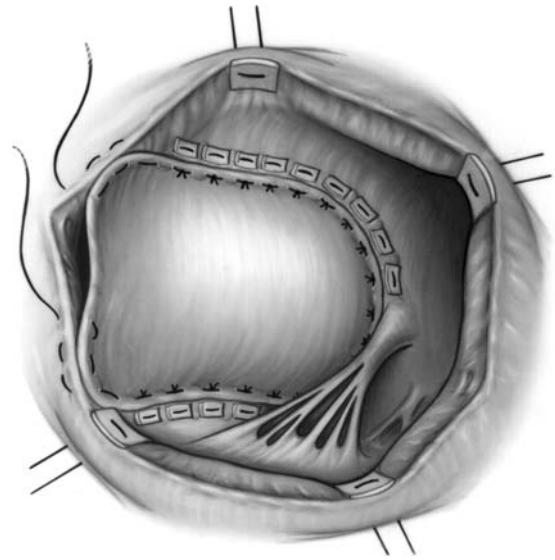


Figure 35.6

The length and the width of the channel from the left ventricle to the aorta are measured, and a generous Gore-Tex or Dacron patch is prepared. It may be tailored from a tube. Because of the possibility of a residual or recurrent VSD, we do not use a running Prolene suture; rather, we prefer single mattress stitches buttressed with Teflon pledgets (**Figure 35.6**). The sutures start at the lower corner of the VSD near the tricuspid valve and run around the VSD to the anterior right ventricular wall in order to provide a wide left ventricular outflow. The last two stitches are left untied so as to provide access to the aortic valve for de-airing later.

The pulmonary artery is then transected, the pulmonary valve is closed with a running 4-0 Prolene suture, and then the pulmonary artery is closed formally in two layers with continuous mattress and over-and-over stitches (**Figure 35.7**). The ASD, if present, is closed, or a PFO stitch is placed. De-airing is performed, and the aorta is unclamped. The stitch on the PFO/ASD is then tied. The aortic needle vent is connected to suction, and rewarming is started.

The conduit is then measured and trimmed. In general, there is a tendency to leave the conduit too long; when it is filled with blood, it may kink and become partially obstructed. A sump is placed through the conduit to the pulmonary artery so as to facilitate the distal anastomosis (**Figure 35.8**). The size of the homograft can be estimated according to the size and weight of the child. In early years we tried to fit in the largest possible homograft. We now realize that that was possibly a mistake: too large a homograft may become compressed, and the

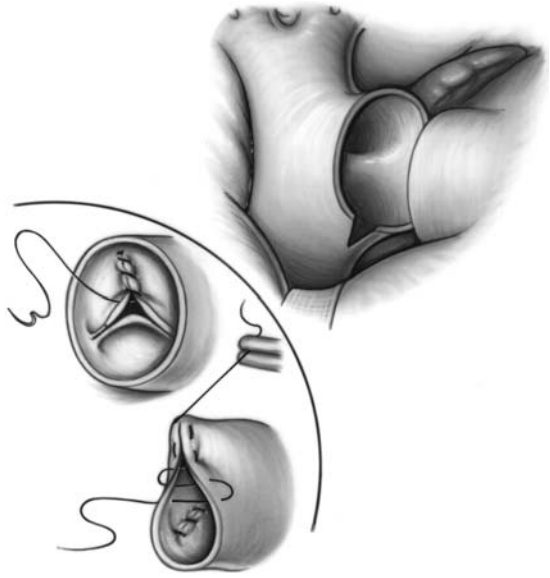


Figure 35.7

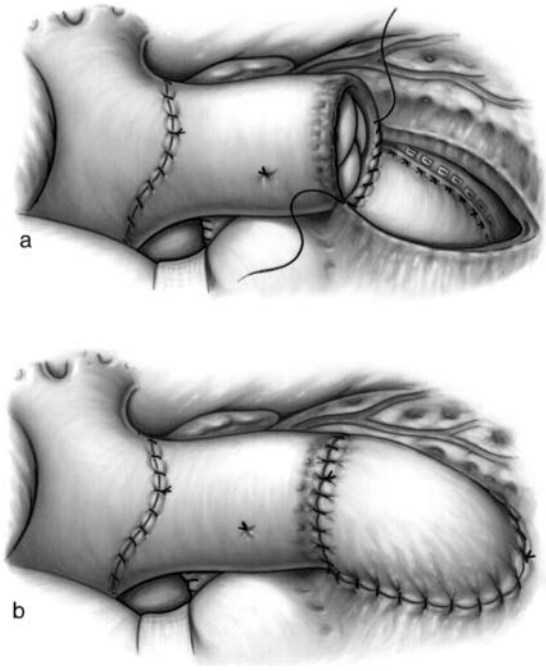


Figure 35.9

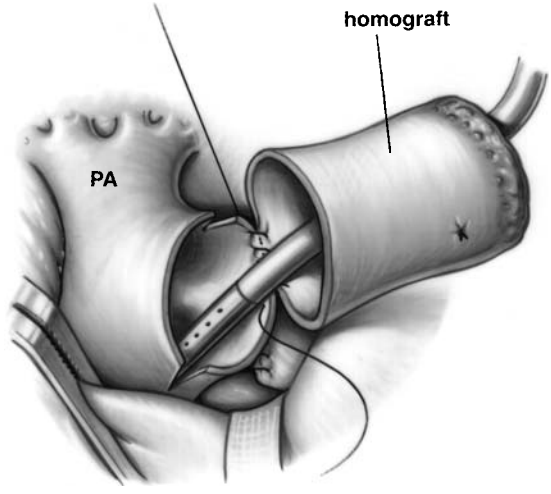


Figure 35.8

resulting turbulence may contribute to early dysfunction. In general, 18 or 20 mm homografts are usually placed in 4 year-old patients.

Conduit orientation is important. If an aortic homograft is used, the anterior cusp of the mitral valve is cut away, leaving a generous rim on the homograft. This part is sutured to the superior aspect of the ventriculotomy (**Figure 35.9a**). It is important to check that the edges of the ventriculotomy are not too thick and that there are no large muscle bundles within the ventricle. The muscle bundles may contribute to the development of obstruction at this proximal anastomosis; they may have to be resected or at least transected, and the edges of the ventriculotomy may have to be thinned. The right ventricular outflow tract is then reconstructed with a hood of Gore-Tex or bovine pericardium (**Figure 35.9b**).

When the conduit attachment to the right ventricle is completed, the right atriotomy is closed, and the caval snares are released and the heart is de-aired. Bypass is discontinued in the usual manner. The usual atrial and ventricular pacemaker wires and drains are placed.

If the conduit does not cross the midline, compression is usually avoided. If the conduit lies too close to the midline, it is possible to open the left pleura and the posterolateral pericardium widely; this enables the heart to rotate to the left, moving the conduit to the left as well. Because the incision must be made posteriorly, it is better to make it before discontinuation of cardiopulmonary bypass. To facilitate reoperations, especially conduit replacement, closure of the pericardium with a Gore-Tex membrane is recommended.

Complications

The numerous suture lines and the long conduit increase the risk of postoperative bleeding and tamponade. Residual or recurrent VSD, development of true or

false aneurysms of the right ventricular outflow tract obstruction, left-ventricle-to-aorta gradients, and late infections have also been described.

REV Procedure ("Reparation á l'Etage Ventriculaire")

The principle of this operation, which was proposed by Lecompte *et al*. (1981) is illustrated in **Figure 35.10**. On hypothermic bypass with cold blood cardioplegia, the aorta is cross-clamped and the right ventricle opened. As much as possible of the infundibular septum is resected (**Figure 35.11**). The resection is helped by placement of a Hegar dilator through the transected pulmonary artery and pulmonary valve into the left ventricle. In patients with inlet VSD, the septum anterior to the VSD is resected (**Figure 35.12**). Because of the extensive resection of the septum, the patch connecting the left ventricle to the aorta can be shorter than the patch in the Rastelli procedure (**Figure 35.13**). The Lecompte manoeuvre, as described in Chapter 36, is performed, the aorta reanastomosed and the pulmonary artery is sutured directly onto the right ventricle, possibly with a unicusp pericardial patch (**Figure 35.14**).

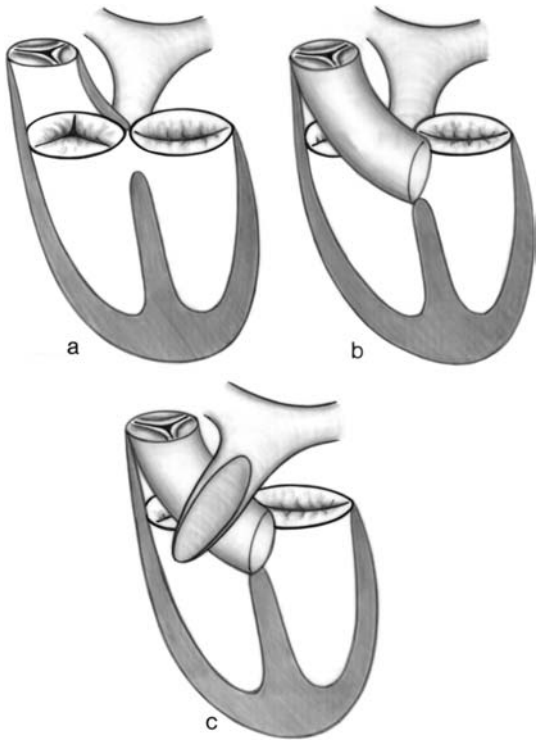
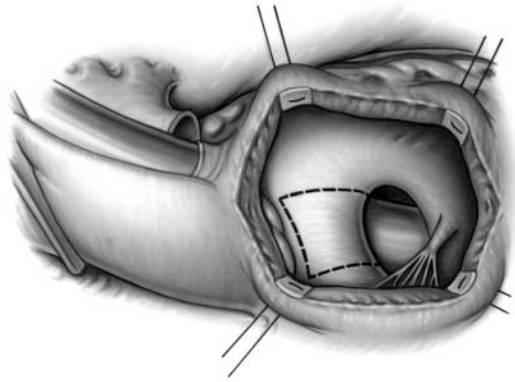


Figure 35.10



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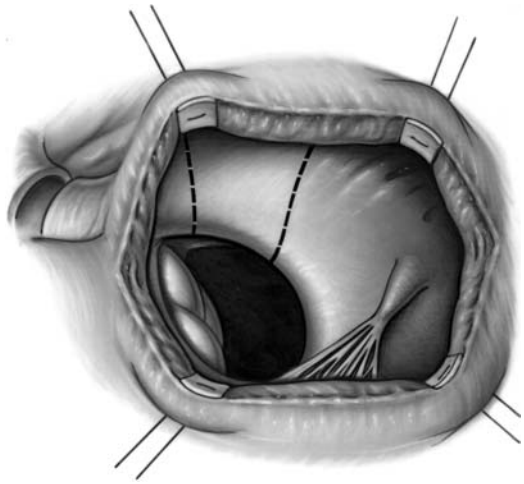

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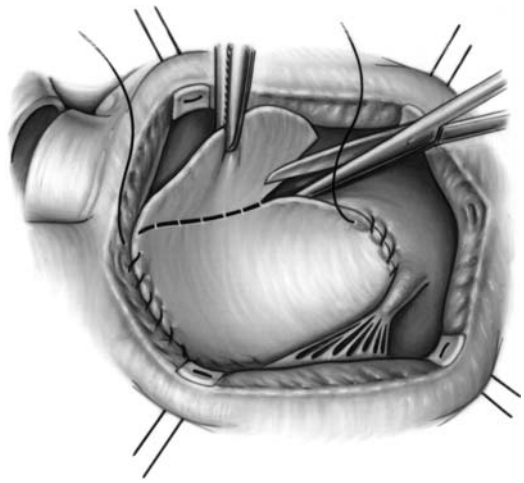

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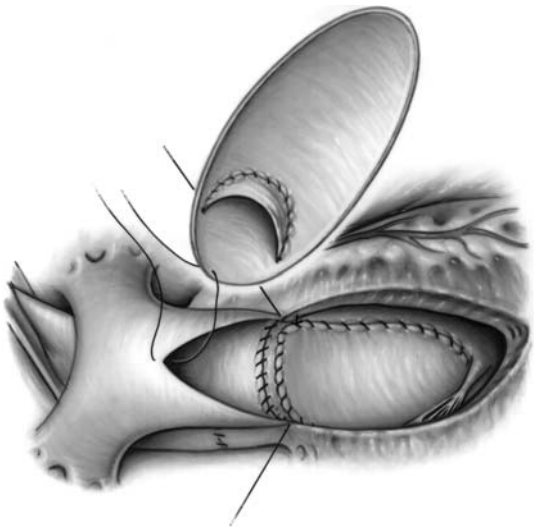

Figure 35.14

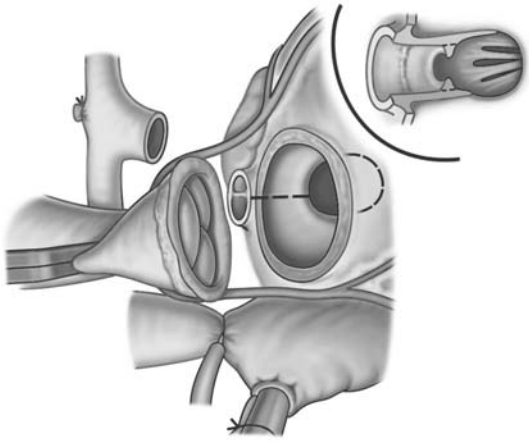

Figure 35.15

Aortic Translocation

Another alternative was described by Nikaidoh (1984). It consists of posterior translocation of the aortic root with biventricular ouflow tract reconstruction. Division of the outlet septum towards the VSD is illustrated in **Figure 35.15**. The narrowed LVOT is augmented with a prosthetic patch. The Lecompte procedure can also be added. This procedure is probably suitable for some patients who may have suboptimal anatomy for Rastelli operation.

RESULTS

The hospital mortality rate of the ***Rastelli operation*** in the earlier years was approximately 14% (Marcelletti *et al*., 1976; Moulton *et al*., 1981). The underdevelopment of the pulmonary arteries, the position of the VSD and its relationship to the tricuspid valve mechanism constitute other important risk factors. If it is not easy to construct a pathway from the left ventricle to the aorta, it may be better to abandon biventricular repair and to perform a total cavopulmonary connection.

The in-hospital mortality rate has been reported to be lower in more recent years. The author and associates have operated on 109 patients between 1971 and 1990; 14 patients died (12.8%). During 1986–1990, only two (6%) of 33 operated patients died. Even in the current era the late morbidity and mortality related mainly to obstruction of the conduit and late LVOTO (Lee *et al*., 2004).

In patients in whom relief of LVOTO seems feasible, we would prefer to achieve a less than perfect relief of the LVOT, rather than place a conduit for the Rastelli procedure. In such patients arterial switch operation, closure of VSD and relief of LVOTO is our operation of choice.

Lecompte performed 94 ***REV procedures*** between 1985 and 1992. Fourteen patients died (14%). Vouhe *et al*. (1992) compared the results of REV and Rastelli operations in their institution. Twenty-two patients underwent Rastelli procedure, 40 patients underwent the Lecompte operation (REV). Two patients died after Rastelli (9%) and five after REV (12.5%). The difference was not statistically significant. There were eight late reoperations (five in the Rastelli group, three in REV). The authors concluded that both procedures provide satisfactory results. However, the Lecompte operation is feasible in infancy and it may reduce the need for reoperation for the outflow tract obstruction.

Morell *et al*. (2005) reported 12 patients who underwent ***aortic translocation*** between 1996 and 2005. One patient died after the operation (8.3%). Three patients have undergone four reoperations.

TGA, LVOTO AND INTACT VENTRICULAR SEPTUM

If the obstruction is dynamic rather than anatomical, the arterial switch operation can be safely performed. When the obstruction is anatomical, the approach depends on the underlying cause. If complete or nearly complete relief is expected (as with fibrous shelf, aneurysm of the membranous interventricular septum, or bicuspid pulmonary valve), the arterial switch operation is appropriate. If surgical relief is likely to be difficult

(as with septal attachment of the mitral valve and fibromuscular tunnel), arterial switch may not be feasible and different treatment strategies are required. If there is severe cyanosis in infancy, a systemic-to-pulmonary shunt can be created, but it is important in such patients that inter-atrial communication is adequate. If the efficiency of balloon atrial septostomy is in doubt, atrial septectomy may be required at the time of the shunt operation. Definitive surgery may then consist of a left-ventricle-to-pulmonary-artery conduit with concurrent Senning or Mustard operation (Singh *et al*., 1976; Dasmahapatra *et al*., 1989). We do not consider this to be the preferred strategy. Careful assessment of each individual patient is mandatory; it may be preferable for a patient to undergo direct relief of LVOTO and a concomitant arterial switch operation, rather than an inflow types of repair as well as a conduit insertion. Residual pressure gradients in the LVOT have been shown to be well tolerated clinically up to 10 years after the arterial switch approach (Wernovsky *et al*., 1990; Sohn *et al*., 1998).

The left ventricular outflow tract can be approached from above through the pulmonary artery and pulmonary valve, from the left atrium through the mitral valve, or from an apical left ventriculotomy. The approach through the pulmonary artery is most commonly used (**Figure 35.16**). The pulmonary artery is opened longitudinally, and a modified nasal speculum is inserted through the valve (**Figure 35.17**). Alternatively, two small nerve hook retractors may be used. A fibrous shelf is resected in a manner similar to that described for patients with subaortic membrane in ventriculo-arterial concordance. Excision of the muscle in patients with fibromuscular obstruction is limited to the area indicated in **Figure 35.17**. This area is between the 11 o'clock and 2 o'clock positions from the surgeon's viewpoint.

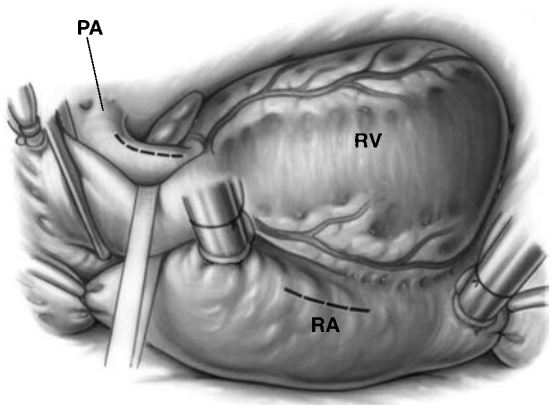

Figure 35.16

Figure 35.17

LV–PA Conduit

In patients in whom the relief of obstruction is inadequate or not attempted, the obstruction can be bypassed with a valved conduit inserted between the apex of the left ventricle and the pulmonary artery (Singh *et al*., 1976). Although rarely performed, the procedure is described for completeness.

First, a Mustard or Senning operation is performed. The pericardium and the left pleura are opened widely posterior and parallel to the left phrenic nerve. As the conduit lies behind the heart in the left pleural space, compression of the conduit is avoided. A left ventriculotomy is made from the apex upward, parallel to the anterior descending coronary artery. Care is taken to interrupt as few branches of the coronary arteries as possible (**Figure 35.18**). At first, a small opening is made and then enlarged under direct vision to avoid any damage to the papillary muscles. The obstruction may be visualized from below and, on occasion, relieved from this approach. However, it is important to remember that on the relaxed heart it is difficult to judge the degree of obstruction. The surgeon may be able to pass a large Hegar dilator through the outflow tract into the pulmonary artery and, when the heart regains its tone, a severe residual gradient can be measured. The ventricular end of the conduit is trimmed obliquely, and the anastomosis between the ventricle and the conduit is created with 5-0 Prolene sutures buttressed with two strips of Teflon felt (**Figure 35.19**). The distal end of the conduit is attached to the side of the main pulmonary

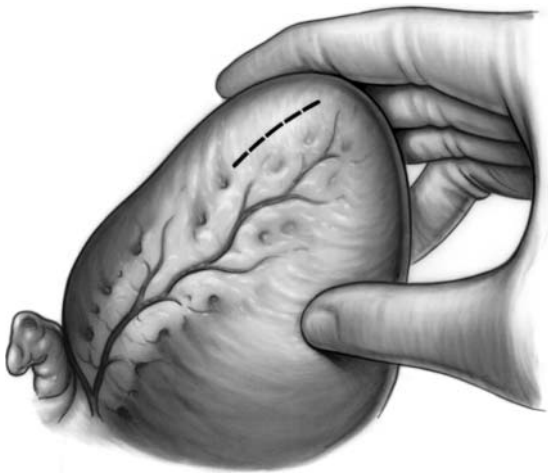

Figure 35.18

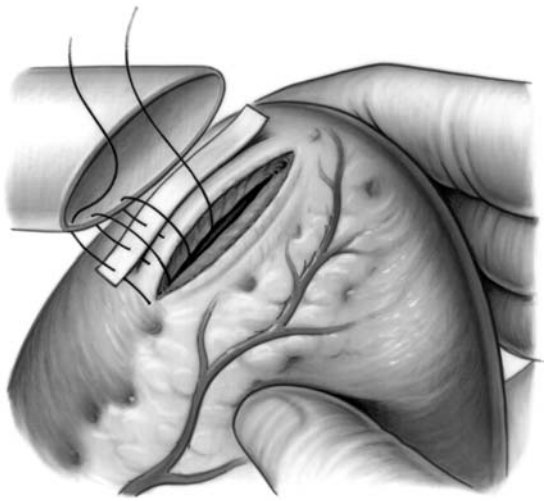

Figure 35.19

artery (**Figure 35.20**). This conduit may be smaller than the conduit used in the Rastelli operation. Even in severe obstruction, there is some way through the left ventricular outflow tract and pulmonary valve, so that the conduit functions as an additional outlet.

RESULTS

In the atrial switch era, Idriss *et al*. (1977) reported only two deaths in their series of 12 operated patients with TGA, LVOTO and intact ventricular septum; all 10 patients in Oelert *et al*.'s series survived (1977). Both

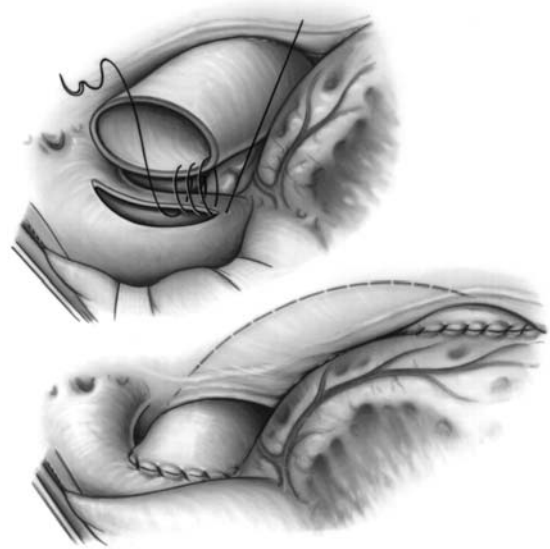

Figure 35.20

groups of authors mentioned, however, that residual gradients were present, and in patients with an abnormal mitral valve, the relief of obstruction was not attempted. The Toronto group reported similar experience in 46 infants and children with direct relief of LVOTO and concurrent Mustard operation (Dasmahapatra *et al*., 1989), but the results of LV-to-PA conduit were discouraging. Long-term results of surviving patients presumably depend on the fate of the conduit, which may be more favourable than those of valve conduits used in other conditions, as the conduit is far away from the sternum. It also provides only alternative pathway from the left ventricle to the pulmonary artery.

In the arterial switch era, Sohn *et al*. (1998) from Melbourne reported encouraging results with direct relief of LVOTO and concurrent arterial switch in 26 TGA patients (median age 69 days) with organic LVOT (four bicuspid valves, five dysplastic valves, 17 LVOT anomalies including accessory atrio-ventricular valve tissue, fibromuscular ring, anomalous muscle bands, and septal malalignment). There were two perioperative deaths. The actuarial freedom from reoperation for the neo-aortic valve or residual LVOT was 87% at 130 months, representing two reoperations at the time of reporting.

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36

The Arterial Switch Operation

R. B. B. Mee

Direct repair of ventriculo-arterial discordance (arterial switch) in transposition of the great arteries (TGA), as opposed to inflow repair (Mustard and Senning operation), has remained intellectually appealing throughout the history of surgery for TGA. Direct repair was first attempted by Mustard in 1954 in seven patients, in whom only the left coronary artery was transposed attached to the ascending aorta; none survived. Senning (1959) had capitalized on Albert's (1954) proposal of physiological repair by inflow baffling and achieved the first success by using an ingenious rearrangement of the atrial walls. This procedure was superseded by the simpler Mustard operation in 1963 (Mustard, 1964).

The arterial switch procedure continued to be appealing because of persisting shortcomings of the atrial inflow repairs, viz. atrial arrhythmias, right ventricular dysfunction, tricuspid valve incompetence, morbidity and mortality among patients awaiting atrial repair, persisting dynamic left ventricular outflow tract obstruction (LVOTO) and baffle obstructions. The last problem has largely been obviated by the reintroduction of the Senning operation by Quaegebeur *et al*. (1977).

The first successful arterial switch for TGA and ventricular septal defect (VSD) was reported by Jatene *et al*. (1975), who performed an operation similar in design to that of Baffes (1961). This was followed quickly by reports of further successes, also for TGA and VSD (Yacoub and Radley-Smith, 1976; Jatene *et al*., 1976). In 1981, Lecompte *et al*. described a manoeuvre whereby the pulmonary artery bifurcation was brought in front of the neo-ascending aorta, obviating the need for interposition conduit. This was an important milestone in the evolution of the arterial switch operation, greatly expanding its appeal to centres in which its progress was critically monitored and in which surgeons persisted in using the well-tried atrial repairs involving low early mortality rates.

There remained the hurdle of introducing the arterial switch for simple TGA. After his failures in 1972, Yacoub introduced a two-stage repair, comprising initial pulmonary artery banding to retrain the left ventricle, supplemented by a systemic-to-pulmonary-artery shunt if banding resulted in excessive desaturation, followed months later by arterial switch (Yacoub *et al*., 1977). Yacoub and Radley-Smith (1976) described five basic coronary artery configurations and methods for their transfer (**Figure 36.1**). In simple TGA, if the patient was young enough, arterial switch was possible with a single-stage approach (Mauck *et al*., 1977; Abe *et al*., 1978). By 1984, a small series of primary arterial switch repairs for simple TGA with reasonable mortality rate was reported by Castaneda *et al*. (1984).

Controversy revolves around indications and contraindications for primary arterial switch for simple TGA, in terms of age of the patient, left ventricular (LV) pressure, LV dimensions, shape, wall thickness, and estimated LV mass. A most important contribution was made by the Boston group (Jonas *et al*., 1989), who quantified the very rapid increase in LV mass in neonates and older infants after pulmonary artery banding; the increase was such that adequate retraining to

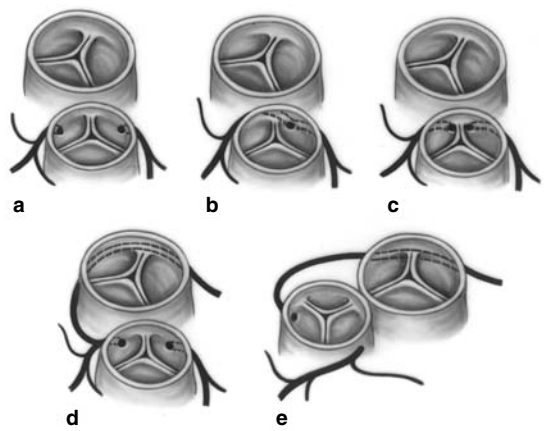

Figure 36.1

Surgery for Congenital Heart Defects, Third Edition. Edited by J. F. Stark, M. R. de Leval and V. T. Tsang
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systemic workload could be effected in as little as 6–7 days. This finding supports the possibility of performing primary arterial switch for simple TGA in older neonates or young infants, with the use of non-banding pharmacological and intensive care unit support methods for retraining the left ventricle. This possibility is also supported by the author's own experience, in which primary arterial switch for simple TGA has been performed on all patients who have presented under the age of 2 months at the author's institutions (Royal Children's Hospital, Melbourne, and Cleveland Clinic Foundation, Cleveland, OH), regardless of the LV considerations just described, since January 1985 (Davis *et al*., 1993; Duncan *et al*., 2004).

INDICATIONS FOR THE ARTERIAL SWITCH OPERATION

TGA and VSD or Double-outlet Right Ventricle with Subpulmonary VSD

In this group of patients, late results of inflow atrial repair have been disappointing, particularly from the point of view of subsequent right ventricular (RV) dysfunction and tricuspid valve incompetence. In most centres, the arterial switch experience began with such patients, and most surgeons would currently agree that arterial switching represents the procedure of choice. Coarctation of the aorta or arch interruption with the frequently associated right ventricular outflow tract obstruction (RVOTO) may all be dealt with simultaneously, through the midline incision.

Simple TGA

Only a few centres worldwide have achieved an operative mortality rate for the arterial switch operation similar to that of the inflow repair established by top centres over the last decade. However, elective inflow repair following neonatal palliation (balloon atrial septostomy and/or Blalock–Hanlon septectomy) was associated with a significant interim mortality—based on intention to treat—providing ''room'' for the higher mortality for early arterial switch operation. The late results of inflow repair are better for simple transposition than for complex TGA or ventricle (DORV) where a VSD has been patched. In those centres with very low mortality for arterial switch, the early risk is now equal to or better than the best earlier reported for inflow repair, and the late results of arterial switch are clearly superior to those of inflow repair for either simple or complex TGA.

Ideally, arterial switching for simple transposition is performed within the first 2 weeks of life, before involution of the left ventricle begins (**Table 36.1**). (Norwood *et al*., 1988). Note that **Table 36.1** actually suggests an increased risk over the age of 1 week and

Table 36.1 Hospital mortality related to age at operation: 20-institution study.

Age at arterial switch repair (Days)	Simple TGA				TGA with VSD				
		Deaths				Deaths			
≤<	*n*	No.	(%)	CL (%)	*n*	No.	%	CL (%)	P(χ^2)
4	38	1	3	0.3–9	3	0	0	0–47	
4–8	52	3	6	3–11	9	2	22	8–45	
8–15	32	6	19	11–29	3	0	0	0–47	
15–31	3	1	33	4–76	3	1	33	4–76	
31–40	0	—	—	—	1	0	0	0–85	
40–50	0	—	—	—	1	0	0	0–85	
50–60	0	—	—	—	2	1	50	7–93	
60	0	—	—	—	8	1	12	2–36	
Total	125	11	9	6–12	30	5	17	9–27	0.20
p (logistic)			0.001				0.96		

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This depiction includes all patients, including the 12 (one death) in whom the morphology (simple TGA vs. TGA with VSD) was not known when all the other depictions and the multivariate analyses were performed, but was known when this depiction was assembled.

TGA, transposition of the great arteries; VSD, ventricular septal defect; CL, confidence limits (70%).

the data include only three patients, aged 2–4 weeks, with one death. This was not the experience of the author over a 20-year experience and most centres performing arterial switches for simple TGA would not recognize that the risk in the second week exceeds that in the first week.

LV involution may be deferred by at least moderate patency of the ductus arteriosus or by dynamic LVOTO, which extends the safe primary arterial switching period. Dynamic LVOTO is distinguished from organic LVOTO by ultrasound interrogation. There is septal bulge to the left without evidence of endocardial thickening, ridges or nodules.

Patency of the ductus arteriosus may be maintained by prostaglandin E1 (PGE_1) infusion if deferment of arterial switch is desired. If PGE1 infusion is administered on presentation, arterial oxygen saturations improve, and if the institution's policy is to perform the arterial switch within a few days, balloon atrial septostomy (BAS) is considered unnecessary. With other management regimens, BAS is performed routinely. The author's preference is routine BAS, which constitutes adequate palliation in about 85% of patients. In the remaining patients, in whom there is failure of mixing, PGE_1 infusion is recommenced, the patient is stabilized and early arterial switching is arranged.

Certain criteria have been recommended for distinguishing between patients who can safely undergo primary arterial switch and those who need a two-stage approach. These criteria remain challengeable and relative. However, a patient with simple TGA who presents at a ''switching centre'' at the age of 4 months, with normal pulmonary artery pressure and no LVOTO, would uncontroversially be a candidate for the two-stage switch approach. However, for a patient of this sort of age with simple TGA having good RV function and minimal tricuspid valve incompetence, the wisdom of a rapid two-stage switch with its risk of late LV dysfunction vs. an inflow repair remains highly controversial on a worldwide basis. The results of more gentle banding and delayed arterial switch (allowing the patient to ''grow'' into the band) will probably prove to be less deleterious to the LV but may risk more damage to what is to become the neo-aortic valve.

Double-inlet Univentricular Heart of LV Type, TGA with Severe Subaortic Stenosis, Coarctation, Severe Arch Hypoplasia, or Interruption with Duct-dependent Descending Aortic Flow in Neonates

These patients may be palliated in various ways with a view to subsequent Fontan type repair. The arterial switch as an initial palliation has the advantage over a Norwood-type procedure, by placing the pulmonary artery bifurcation in front of the neo-aorta, obviating the compression, particularly of the left pulmonary artery observed when it remains behind the large posteriorly placed proximal MPA. In addition, an initial systemic shunt physiology is avoided, with the restrictive bulboventricular foramen limiting pulmonary blood flow in systole and avoiding the systemic diastolic run-off. Technically, the arterial switch is more difficult because of the huge discrepancy in sizes of the proximal PA and distal ascending aorta, which is usually absolutely hypoplastic. This problem can be overcome by excising all of the distal ascending aorta and anastomosing the proximal MPA to a combination of the distal ascending aortic stump, the proximal aortic arch and the descending aorta—identical to the technique developed by the author in the modified Norwood procedure, performed without non-vital tissue for arch reconstruction for hypoplastic left heart syndrome. This same technique can be applied for biventricular arterial switches in the presence of coarctation or interruption of the aortic arch associated with RV outflow tract obstruction, small aortic valve, ascending aortic and transverse arch hypoplasia, a not uncommon finding in DORV subpulmonary VSD, and arch obstruction (Mee, 1994).

RV Dysfunction and Tricuspid Incompetence after Mustard or Senning Operation

Two-stage conversion to an arterial switch has been performed in only a relatively few patients worldwide. The first stage (main pulmonary artery banding) should be timed to allow an adequate interval for LV retraining, which may well be slower and less predictable in older patients than in rapidly growing neonates or infants. Banding of large pulmonary arteries with much infolding of the wall results in a later loss of effective banding as these folds smooth out within the band, and early band tightening is necessary. The presence of concomitant LV dysfunction and mitral valve incompetence is a relative contraindication to two-stage conversion to an arterial switch.

Results are variable, particularly in patients older than their early teens. During banding there should be no signs of LV dysfunction, early or late, as this appears to jeopardize the result of the eventual switch conversion. In teenagers and young adults it is unusual to be able to raise the peak LV pressure by more than 20–25 mmHg without seeing the first signs of LV dysfunction (a rise in central venous pressure and/or echo evidence of reduced LV free wall contractility), either on the operating table or within the first 1–2 days postoperatively. Such an observed change requires immediate loosening of the PAB until LV function returns to normal, in order to avoid damage

and subsequent fibrosis. We have observed myocardial oedema by magnetic resonance imaging (MRI) T2-weighted scanning, early post-banding, proportional to the degree of observed LV dysfunction. A programme of LV retraining with a view to arterial switch conversion is complemented by a policy of parallel heart transplantation if LV retraining fails in the presence of progressive RV failure and TR; or after conversion to an arterial switch if progressive LV failure becomes apparent. An update on the consecutive experience of two institutions is available from Poirier *et al*. (2004).

Progressive Dynamic LVOTO after Inflow Atrial Repair

This problem is well handled, in theory, by one-stage conversion to an arterial switch. However, the proximal main pulmonary artery is thin-walled and it may be difficult to achieve haemostasis.

Patients with Congenitally Corrected TGA and VSD, Unrestricted Pulmonary Blood Flow with or without Ebstein's Anomaly of the Morphological RV and Tricuspid Valve

This small group of patients may be best served by combined VSD closure, atrial switch and arterial switch (see Chapter 39).

MANAGEMENT METHODOLOGY FOR ARTERIAL SWITCH

The methods used in managing patients who are to undergo an arterial switch, as applied to neonates, infants and young children (for the first three indications), are well standardized. The method of conversion of failed Mustard or Senning operations to arterial switch is handled in detail elsewhere (Mee, 1986, 1989; Cochran *et al*., 1993; Poirier *et al*., 2000, 2004), but the results are briefly updated here.

Diagnosis

Reliance is placed on ultrasonography for full segmental assessment, atrial situs, atrio-ventricular connection characteristics, ventricular size, shape, wall thickness and outlets, ASD size, VSD size and position (single or multiple), great vessel position and anatomy, ductal size, aortic arch and coronary artery anatomy. BAS, if required for inadequate mixing, is usually performed at the bedside (ICU) under ultrasound control. Angiography is rarely required in the neonatal setting, but may be indicated in older infants suspected of increased pulmonary vascular resistance. BAS does assist with operative technique for neonates with simple TGA, when a single venous cannula is used providing good venting of both sides of the heart. BAS is used in complex TGA, DORV presenting after duct closure with inadequate saturations, and poor mixing.

Preoperative Preparation

If BAS has resulted in good palliation, the patient is sent to the ward, oral feeding is established and, if simple TGA is present, an arterial switch operation is performed at a convenient time within 2 weeks of birth. If palliation by BAS is poor, PGE_1 infusion is recommenced; if the ductus arteriosus opens, the arterial switch operation is performed within the next day or two. If the duct has not opened and oxygen saturations do not improve, an emergency arterial switch operation is performed. Patients from other centres with simple TGA may be referred late, but if they are under 2 months of age, a primary arterial switch operation is performed. After BAS for TGA and VSD or DORV, the patient may be stabilized with decongestive therapy, be sent home, and undergo elective repair by arterial switch at 6–8 weeks of age. If congestive heart failure can be controlled only by management in the intensive care unit, repair is performed as a matter of urgency (within a day or two of the patient's being declared intensive care unit-dependent). If a patient with TGA and VSD or DORV is not intensive care unit-dependent but hospital ward-dependent, the repair is scheduled earlier than for patients undergoing elective repair.

Patients with TGA and VSD or DORV with coarctation but well-formed arch and no RVOTO undergo coarctation repair through the left chest, without main pulmonary artery banding, and are then managed in the same way as patients with TGA and VSD or DORV without coarctation. Banding of the main pulmonary artery (MPA) is performed only when multiple VSDs are present, and the surgeon believes that ventricular septation cannot be reliably achieved at that time. Neonates with severe arch hypoplasia and coarctation, or interruption, usually also have subaortic stenosis and only infrequently can achieve independence from the intensive care unit. These patients are stabilized with paralysis, intermittent positive-pressure ventilation, PGE_1 infusion, dopamine infusion (5–10 μg/kg/minute) manipulation of systemic vascular resistance with vasodilators (nitroprusside followed by phenoxybenzamine), if appropriate, and undergo correction of acid–base and electrolyte imbalances. While improvement is occurring, repair is scheduled within 2–4 days. Failure to improve or produce urine leads to emergency repair. At operation, the aortic arch is repaired, combined with atrial and ventricular septation, arterial switch and routine RVOT resection.

Arch repair is performed under circulatory arrest at 18°C nasopharyngeal temperature, and all the rest of the repair under CPB and full flow. The author's policy for midline repair of coarctation with hypoplastic aortic arch is to select neonatal patients with arch diameter (in millimeters) less than the patient's weight (in kilograms) plus 1 (i.e. an arch diameter of less than 4 mm in a 3 kg infant or less than 3.5 mm in a 2.5 kg infant).

Anaesthesia

Types of anaesthesia and routes of administration vary little. Morphine or fentanyl induction with non-depolarizing muscle relaxation and nasotracheal intubation are followed by insertion of catheters into the right radial artery and right internal jugular vein. Nasopharyngeal, oesophageal, and toe temperature probes, along with surface electrocardiographic electrodes, are established with the patient lying on a water heating-cooling blanket and the neck extended with the head turned to the left. The patient is draped with the use of a head frame, which provides an instrument table for the surgeon and the first assistant. Phenoxybenzamine (1–2 mg/kg) is infused slowly while the chest is opened. A fall in blood pressure is countered by infusion of 5% albumin in normal saline. Cephazolin (25 mg/kg) is given intravenously at induction, and a urinary catheter is inserted.

Surgical Exposure

A midline sternotomy is performed, followed by amputation of the thymic lobes at the level of the innominate vein. The anterior pericardium is harvested in two rectangles, stayed at each corner with a silk suture and stored in a pack soaked in cold Hartman's Solution. The aorta is routinely mobilized beyond the origin of the innominate artery and separated from the main pulmonary artery to the level of the coronary ostia. The right and left pulmonary arteries and duct are partially dissected out. If arch reconstruction is required, all arch vessels are dissected and the upper descending aorta is partially mobilized.

Cannulation

Heparin (2 mg/kg body weight) is given and the aorta cannulated with a Bardic cannula (10 Fr. for neonates) with a rubber ring placed 2–3 mm from the end, via a stab incision within a longitudinally placed purse-string U-suture centred on the origin of the innominate artery. If the arch is atretic or severely hypoplastic, a second arterial cannula of the same size is placed, using the same technique, in the proximal duct and joined by a Y-connector to the aortic cannula and the arterial pump line. For simple TGA cases, a single right-angled venous cannula is placed through the right atrial appendage (**Figure 36.2**). For complex cases the cavae are cannulated separately.

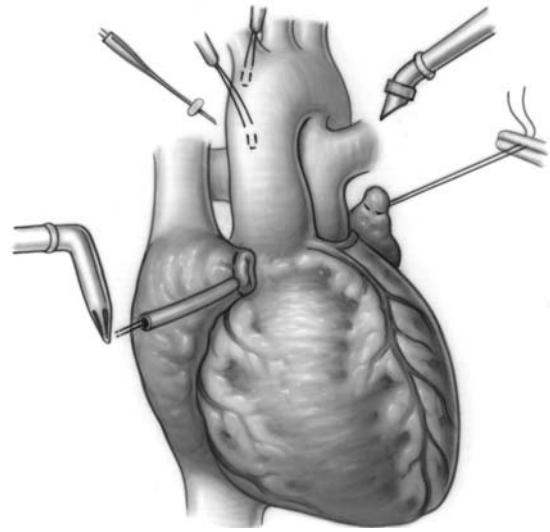

Figure 36.2

Perfusion

The pump circuit is primed with fresh (less than 24 hours old) heparinized blood, adjusted with clear volume expanders to achieve a mixture of the patient's and prime haemoglobin of 9 g/dl. The patient undergoes core cooling to 22°C nasopharyngeal temperature and is rewarmed with a blood–nasopharyngeal temperature gradient not exceeding 7°C. In simple TGA the ASD is closed during a brief period of circulatory arrest (less than 10 minutes) at 22°C. If the aortic arch is being reconstructed, the VSD is closed and the coronaries translocated during cooling to 18°C, and then under conditions of circulatory arrest the arch is repaired, cardiopulmonary bypass re-established at full flow, the ASD closed before the heart starts beating, the RA closed and caval snares released and the neopulmonary artery reconstructed with the heart beating. This will include trans-aortic valve RVOT resection. Neo-ascending aortic continuity is achieved after the Lecompte manoeuvre just prior to circulatory arrest if the distal ascending aorta is of good size, and during circulatory arrest if the very small size of the distal ascending aorta requires excision up to the innominate artery, in which case the proximal MPA bearing the translocated coronaries is anastomosed to a combination of the distal ascending aortic stump and medial aspect of the descending aorta, which was

brought up to repair the aortic arch. Bypass flows are maintained at 150–220 ml/kg/minute or more to attain a perfusion pressure of 35 mmHg in neonates, throughout the operation, except for periods of circulatory arrest.

On the basis of an estimated blood volume of 100 ml/kg in infants less than 6 months old, the pump is primed with fresh heparinized blood; 10 mmol/l sodium bicarbonate, 30 mg/l heparin, 20 mg/kg methylprednisolone, and Plasma-Lyte 148 injection in water (Travenol Labs, Deerfield, IL) are added to create a combined prime and patient's blood haemoglobin of 9 g/dl. The basic perfusion flow rate is 150 ml/kg/minute.

Ideal perfusion pressures depend on the patient's weight:

- 25–35 mmHg for infants weighing 2–3 kg.
- 35–45 mmHg for infants weighing 3–5 kg.
- 45–55 mmHg for infants weighing 5–10 kg.

Most neonates and infants are treated with 1–2 mg/kg phenoxybenzamine intravenously before or just as cardiopulmonary bypass is begun, to maintain the appropriate pressure range. Under these conditions, flow rates not uncommonly exceed 150 ml/kg/minute (150–250 ml/kg/minute). In general, the author uses full flow, more than full flow, or circulatory arrest. Low flow is not used except momentarily.

Cardioplegia

The cardioplegic solution used by the author has remained unchanged for 25 years and is a crystalloid solution containing 6 g% of albumin for patients weighing less than 10 kg; it is composed of the following substances:

1. One bag of base solution, 385 ml:

Na*	23 mmol/l
K^+	15 mmol/l
Ca^{++}	0.35 mmol/l
Cl^-	39 mmol/l
Glucose	2.52 g
Mannitol	2.48 g
Total	106 mmol/l

2. 100 ml 25% human albumin
3. 26 ml (1 ampoule) modified cardioplegic buffer solution

Na_2CO_3	0.28 g
$NaHCO_3$	0.81 g

The cardioplegic solution is administered as initial dose of 110 ml/min/m^2 of body surface area for 2–4 minutes, depending on cardiopulmonary bypass perfusate temperature at the time (4 min if perfusate temperature is higher than 22°C). Subsequent doses of 110 ml/min/m^2 of body surface for 2 minutes are given at approximately 20-min intervals. Delivery temperature at aortic root is 5–6°C. Cardioplegic solution is infused by using an occlusive roller pump with in-catheter pressure monitoring. An infusion pressure of 30 mmHg is not exceeded for neonates. In neonates and small infants, additional cardioplegia is not given during coronary transfer and neo-aortic reconstruction to avoid coronary trauma from a hand-held cardioplegia cannula.

SURGICAL TECHNIQUE

When cardiopulmonary bypass is established, the ductus arteriosus is fully dissected out and divided between 2-0 silk ligatures, securing the aortic end with an additional 7-0 Prolene purse-string suture (**Figure 36.3**). Right and left pulmonary arteries are each surrounded with a Silastic vessel loop and dissected out until first-order branches are clearly seen. The approximate prospective sites of implantation of the coronary arteries into the proximal MPA are determined and marked with a 6-0 Prolene suture (**Figure 36.4**). If the circumflex artery arises from the right-facing sinus (the usual origin of the right coronary artery), the marking suture is placed 2–3 mm higher on the right side than it is in the presence of normal (Yacoub type A) coronary anatomy. This prevents kinking of the circumflex artery after translocation (**Figure 36.5a**). Patients with Yacoub type A coronary anatomy but who have

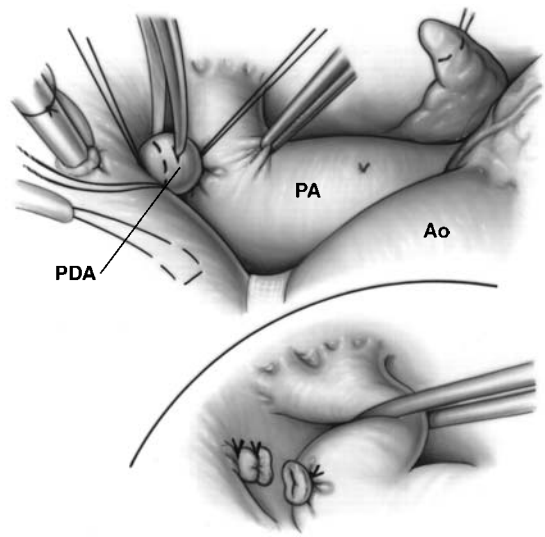

Figure 36.3

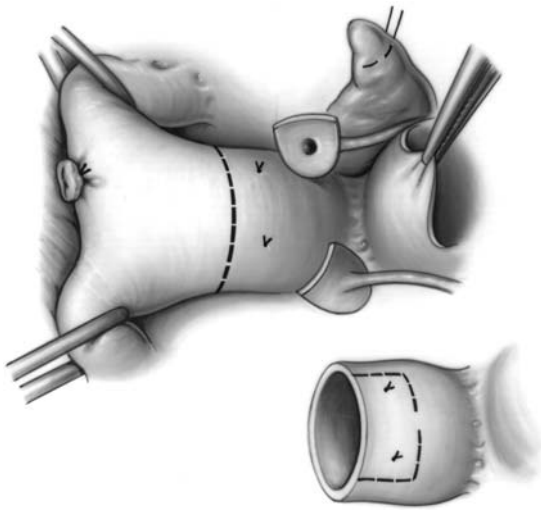

Figure 36.4

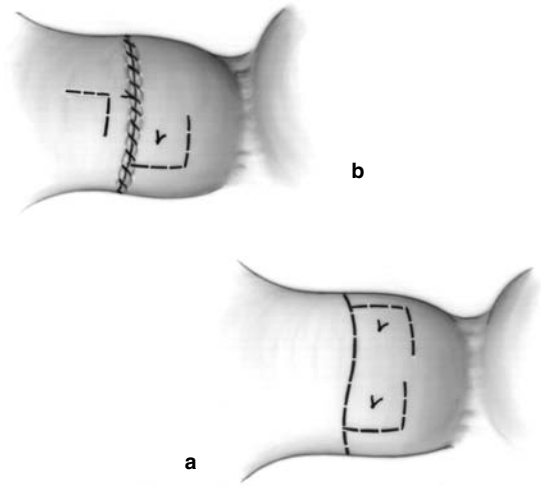

Figure 36.5

a long tortuous left main coronary artery—usually with minimal surrounding fat and suspended on a ''mesentery''—require a high, more right-ward site of reimplantation above the neo-aortic anastomotic suture line (**Figure 36.5b**).

If arch reconstruction is required, the ductus arteriosus is ligated proximal to the ductal cannula soon after cardiopulmonary bypass is established, and the duct, the aortic arch, and the upper descending aorta are fully mobilized. At a nasopharyngeal temperature of 18°C or less, bypass flows are reduced to 10%, arch vessel snares are tightened, the ductal cannula is clamped and removed, and a cross-clamp is placed obliquely just distal to the aortic cannula. Pump flows are adjusted to achieve an arterial bypass line pressure of 30 mmHg. With only the heart perfused and beating (isolated coronary perfusion technique; Sano and Mee, 1990), all ductal tissue is excised, and the descending aorta is anastomosed to the underside of the proximal arch with running 7-0 Prolene sutures. The proximal end of this anastomosis should lie opposite the innominate artery origin. When aortic reconstruction is complete, the clamp is shifted proximal to the aortic cannula, cardioplegic solution is administered and full bypass is reinstated. If the distal ascending aorta is very small and needs excision, then this technique of isolated myocardial perfusion is not used.

Coronary Artery Translocation

The cardioplegic needle is removed, and the ascending aorta is opened anteriorly 3 mm superior to the upper edge of the coronary ostial bulge and inspected internally for coronary ostia before the transection is complete (**Figure 36.6**). Absence of the typical coronary ostial bulge externally hints at an intramural course of the coronary artery.

Excision of the Coronary Ostia

Coronary ostia are excised with a D-shaped cuff; in neonates, all but a 0.5–1.0 mm rim of aortic sinus wall along the line of leaflet attachment is removed.

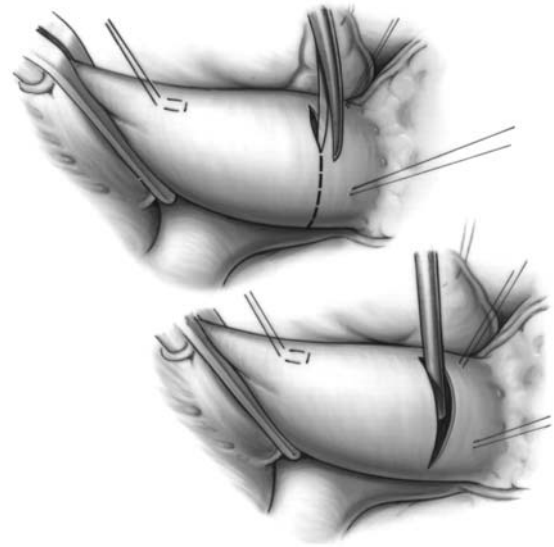

Figure 36.6

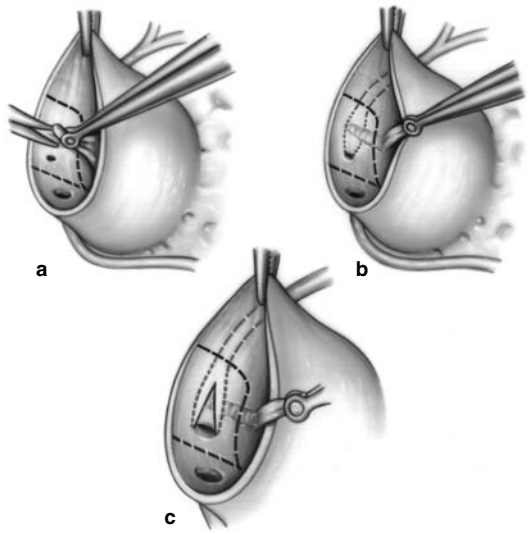

Figure 36.7

Eccentrically placed ostia are treated on their merits. If a 1.0–1.5 mm margin of aortic sinus wall can be included in the coronary cuff by excising up the leaflet attachment, this is satisfactory. Otherwise, the adjacent commissure is sharply dissected away from the aortic wall in order to allow this minimal cuff to be obtained (the commissure is later resuspended to the aortic sinus pericardial patch). When an intramural coronary artery runs in the aortic wall behind a commissure, the same technique is used (**Figure 36.7a**). Usually the intramural coronary artery has a smaller ostium than normal, or the ostium is frankly stenotic. A triangular portion of internal aortic wall is excised until full coronary diameter is reached without extending this excision beyond the point of entry of the coronary into its intramural course (**Figure 36.7b, c**).

Yacoub type B coronary anatomy is difficult to manage. If the two ostia are 2 mm or more apart, two separate coronary cuffs can be created, usually with takedown of the commissure between the facing sinuses. Both coronaries can then be handled in the usual way. If the ostia are inseparable and there is no intramural course, they are left together and handled in a different way (to be described).

Reimplantation of the Coronary Arteries

The MPA is transected 2 mm above the marker sutures in cases in which the coronary arteries are to be reimplanted proximal to the neo-aortic suture line. At this time, operable LVOTO (e.g. accessory endocardial cushion tissue) is resected. Medially hinged trap-door flaps are created in the proximal MPA in the appropriate sinuses (**insert, Figure 36.4**, for Yacoub type A coronary anatomy; **Figure 36.5a** for Yacoub type D coronary anatomy; and **Figure 36.5b** for Yacoub type A coronary anatomy with long, tortuous left main coronary artery).

The coronary artery cuffs are then sewn with running 7-0 Prolene sutures into the defects created by the trapdoor flaps, without axial rotation, starting at the edge away from the hinged flap (**Figure 36.8a,b**). The suture line finishes with the flap sewn back one-third of the distance along the top of the D-shaped cuff (**insert, Figure 36.8**). This creates a cone-like extension of the proximal neo-aortic wall from which the coronary artery arises, and it reduces the angle through which the transposed coronary artery rotates in the plane at right angles to the axis of the neo-aorta. At the same time, the diameter of proximal neo-aorta does not become excessive. If the flap appears too long, 1 mm is trimmed off before the suture line is completed. The ''dog ears'' project at right angles to the reconstituted neo-aortic wall (**Figure 36.8c**).

The two ways of handling Yacoub type B coronary anatomy are described, as follows. Both coronary ostia are excised together, a generous cuff of aortic wall having mobilized the commissure between facing sinuses if necessary. The flat (superior) edge of the D-shaped cuff is sewn directly to the adjacent edge of the proximal MPA. A pericardial patch is then sewn to the inferior curved part of the cuff to create a pocket. The coronary artery-bearing cuff is therefore not rotated in any direction. The ascending aorta is sewn to the anterior wall of this pocket during neo-aortic anastomosis (Yacoub and Radley-Smith, 1978).

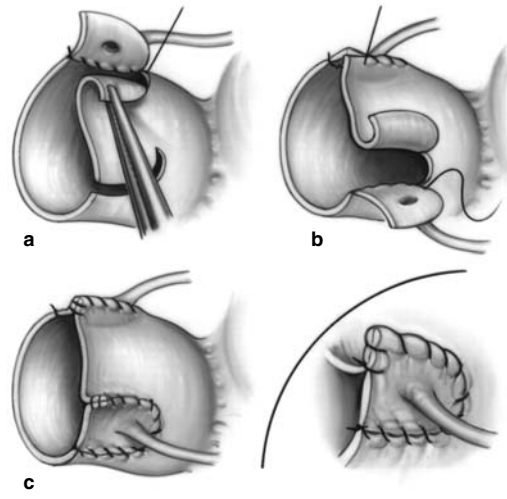

Figure 36.8

An alternative technique has been described by Imai of Tokyo (personal communication, 1990). The coronary arteries are left *in situ*. The aortic wall above the ostia is excised to within 1 mm of the superior edge of the ostia. A slightly deeper excision is made in the adjacent proximal MPA wall, and the adjacent top edges of the proximal aorta and MPA are sewn together for the full width of the sinus (**Figure 36.9a**). Next, a D-shaped cuff from the non-facing aortic sinus is excised but left hinged just above the commissure between the coronary artery-bearing sinus and the non-facing sinus. The curved part of this D-shaped hinged patch is then sewn into the depth of the coronary artery-bearing sinus, again creating a pocket with a superior broad opening (**insert, Figure 36.9** and **Figure 36.9b**). The ascending aorta is then anastomosed to the front edge of this pocket, so that the pocket bearing the coronary arteries opens into the neo-aorta. With neo-pulmonary artery construction, the anterior wall of the pocket becomes the posterior wall of the neo-pulmonary artery (**Figure 36.10**).

The Neo-aorta

In all cases, the Lecompte manoeuvre (Lecompte *et al*., 1981) is performed in such a way as to bring the ascending aorta behind the bifurcation of the pulmonary artery.

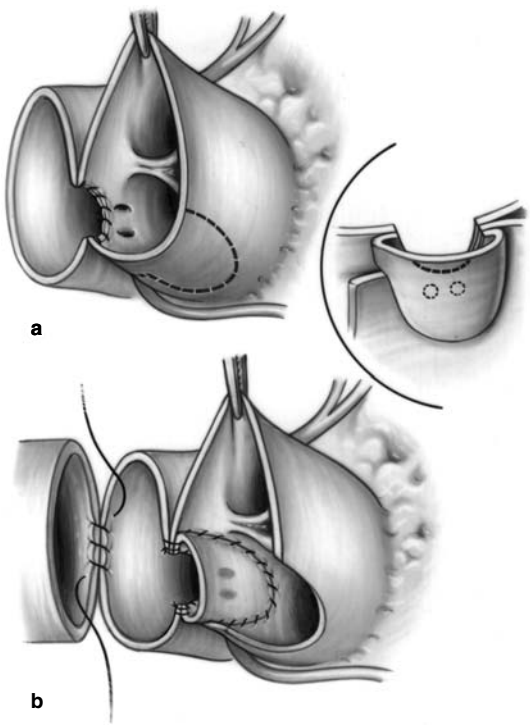

Figure 36.9

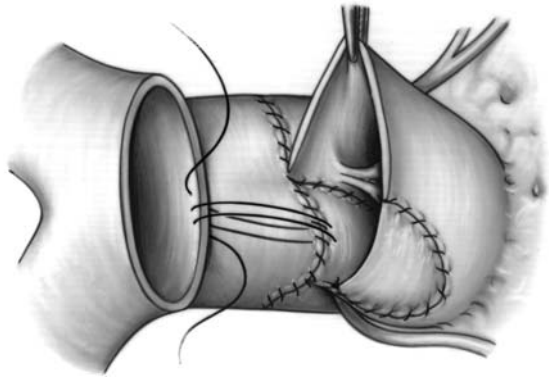

Figure 36.10

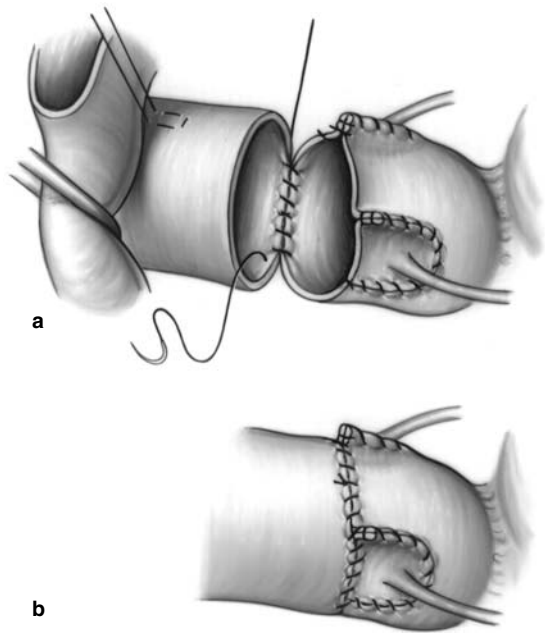

Figure 36.11

The cardioplegic purse-string suture is also passed behind the bifurcation of the MPA and then pulled superiorly to hold the bifurcation up against the aortic cross-clamp, which is not repositioned. The proximal MPA, which bears the translocated coronary arteries, is then anastomosed to the ascending aorta with running 7-0 Prolene sutures in neonates and small infants (**Figure 36.11a,b**). The cardioplegic purse-string suture is passed back up under the MPA bifurcation, which is pulled down to allow de-airing of the aortic root through the cardioplegic needle hole, the ASD closed by direct suture, the LV de-aired, and the aortic cross-clamp is removed.

The heart is observed to turn pink and almost invariably starts beating slowly at the low temperature. All coronary artery branches are observed to confirm that they are filling. Failure of an area of myocardium supplied by a major coronary artery to become pink must be assumed to be due to kinking during translocation; the problem is identified and is then rectified by reimplantation or ostial adjustment with horizontal plicating mattress sutures. Small distal areas of unperfused myocardium may be attributed to intracoronary air, and the offending coronary branches are gently massaged distally until the blotches disappear.

In simple TGA, the proximal MPA is usually a little larger than the distal ascending aorta. The back wall of the MPA is gathered into the posterior wall of the aorta, leaving a good match between vessel ends anteriorly and hence avoiding the need for gathering on the proximal MPA anteriorly, which causes the translocated coronary arteries to bunch up. In complex TGA or DORV, particularly when there is associated coarctation of the aorta, the discrepancies are major; nevertheless, the posterior gathering technique can be used and the placement of an anterior triangular patch in the ascending aorta avoided.

In extreme cases of discrepancy, as in complex TGA, DORV with aortic arch interruption or double-inlet ventricle in neonates, the author has brought the descending aortic anastomosis lower down into the ascending aorta, trimmed off some of the ascending aorta, and anastomosed the proximal MPA to the conjoined ascending and descending aorta. In this situation, the MPA has been transected higher than usual, and triangular, medially based flaps for coronary artery translocation have been created proximal to the neo-aortic suture line. The theoretical benefit of this modification is avoidance of excessive tapering of the proximal MPA at commissure level and avoidance of neo-aortic valve incompetence (a confirmed cause of early death in one of our neonates with double-inlet ventricle).

Repair of Aortic Sinus Defect

With the cross-clamp off, the heart beating and a coronary sinus sucker placed through the aortic valve, the D-shaped aortic sinus defects are repaired with generous untreated autologous pericardial patches sewn in with running 7-0 Prolene sutures in neonates and small infants (**Figure 36.12a,b,c**). The patches are sewn together posteriorly for about 5–7 mm above the posterior commissure to lengthen the back wall of the neo-MPA and allow the MPA bifurcation to sit high above the translocated coronary arteries (**Figure 36.12d**). A detached commissure is resuspended at this time (**Figure 36.13a,b**). Detail of completed resuspension is shown in the inset of **Figure 36.13**.

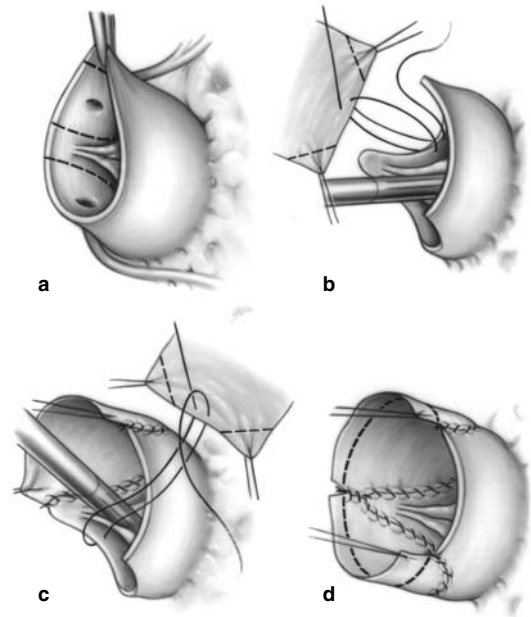

Figure 36.12

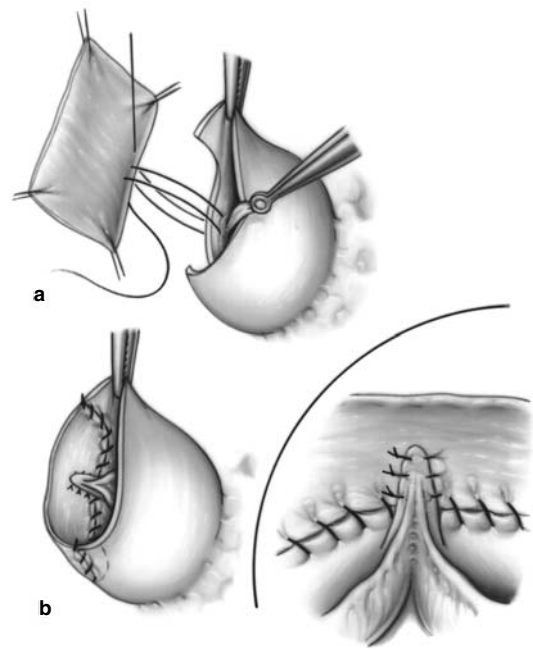

Figure 36.13

Intracardiac Repair

For simple TGA, the aortic cross-clamp is reapplied when the nasopharyngeal temperature is 22°C; cardioplegic solution is run into the neo-aortic root for 2 min at a standard flow rate; cardiopulmonary bypass is discontinued; blood is drained from the patient; the single venous cannula is removed; a coronary sinus sucker is placed through the right atrial appendage; and the right atrium is opened low down with a short longitudinal incision. The ASD is closed with running 6-0 Prolene sutures from below up. In neonatal patients with double-inlet ventricle, the septum is excised. Cardiopulmonary bypass is then restarted, along with rewarming. The heart is de-aired and the aortic cross-clamp removed. Silastic vessel loops on the right and left pulmonary arteries are relaxed to allow open venting of bronchial collateral flow, and the heart is observed for adequate coronary filling and perfusion.

With the heart beating and blood return aspirated by the coronary sinus sucker, the right atrium is closed with running 7-0 or 6-0 Prolene sutures, and the venous cannula is inserted. For complex TGA or DORV, two venous cannulae are used. RVOTO resection is performed from below through the tricuspid valve and from above through the aortic valve after coronary excisions. The VSD or VSDs are closed through the tricuspid valve with the use of a knitted Dacron patch secured with pledgetted horizontal Prolene mattress sutures (6-0 in neonates and 5-0 in older infants). The ASD is closed by direct suture just before cross-clamp removal. The trans-tricuspid valve work can be done at the beginning of the cross-clamp period or after coronary transfer and completion of the neo-aortic part of the switch operation. The author's preference is at the beginning.

With rewarming proceeding on full cardiopulmonary bypass and with the heart beating, the pericardial patches are sewn into the proximal aorta, joined together posteriorly, trimmed (**Figures 36.12** and **36.13**) and an end-to-end anastomosis is created between the proximal aorta and distal MPA with running Prolene sutures (7-0 in neonates and small infants) (**Figures 36.14** and **36.15**). In the side-by-side relationship of the great vessels, with a right-sided aorta, the left half or two-thirds of the distal MPA is closed and an incision is made into the inferior aspect of the RPA, the right pericardial patch is trimmed longer and the proximal reconstructed aorta is sewn into a combination of the RPA and the rightward portion of the distal MPA. The importance of generous pericardial patches is stressed. Early attempts to reconstruct the neo-MPA without patches or with small patches were responsible for almost all of our reoperations for neo-MPA stenosis; neo-MPA stenosis is now rare.

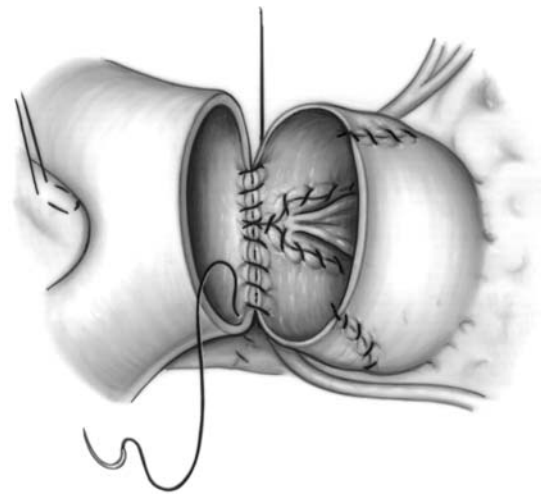

Figure 36.14

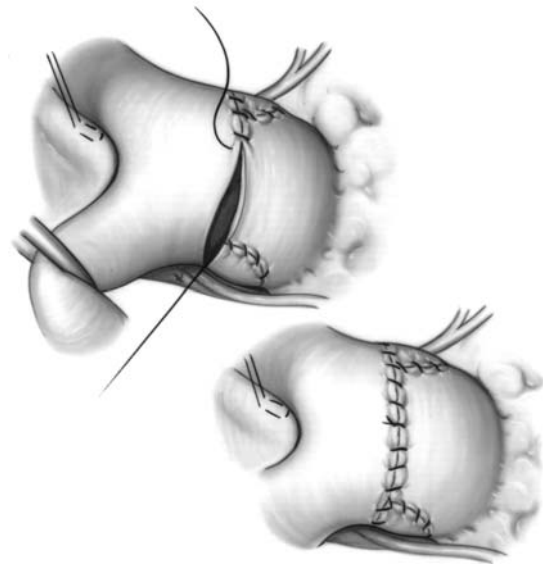

Figure 36.15

Completing the Operation

With the heart repaired, pressure-monitoring catheters are placed in the left atrial appendage and the neo-MPA via the suture line anteriorly. Both catheters are tied by a 6-0 Prolene suture to the heart with the use of a very fine bite of epicardium. RV and right atrial temporary pacing wires are sewn into place. When rewarming is complete and dopamine is infused at 5–10 mg/kg/min, cardiopulmonary bypass is quite rapidly discontinued. The heart is carefully observed for perfusion, and the

left atrial pressure is monitored closely. Ischaemia of the myocardium is reflected by poor colour or a rise in left atrial pressure before electrocardiographic changes are noted. Any rise in left atrial pressure above 10 mmHg necessitates careful scrutiny of the myocardium and surface coronary vessels. A progressive rise in left atrial pressure should be interpreted as the result of mechanical coronary kinking, rather than of inadequate left ventricle in simple TGA, in order to identify and immediately correct a coronary problem at an early stage.

Bleeding

Because of the many external and composite suture lines needed, bleeding has been a widely recognized problem. It becomes less of a problem with experience but is best dealt with definitively and aggressively. Major leakages are first identified after neo-aortic connection and the first aortic declamping and should be dealt with then, preferably with horizontal adventitial 7-0 Prolene mattress sutures. Major bleeding after complete repair may necessitate the reinstitution of cardiopulmonary bypass and even takedown of the neo-MPA anastomosis in order to obtain access to the site of bleeding. Multiple small bleeding sites are managed by application of gelatin foam soaked in topical thrombin. These foam pieces are removed before sternal closure.

Chest Closure

Two soft rubber chest drains are inserted: a short one on the left, trimmed off at the level of the diaphragm, and a longer one on the right. The anterior slip of diaphragm that is attached to the back of the right xiphisternum is detached, so the longer drain can lie more right-ward and not in front of the heart. A Silastic peritoneal dialysis catheter is placed through a stab incision above and to the left of the umbilicus and drawn into the abdominal cavity by a right-angle forceps passed down through a hole made in the peritoneum in front of the liver. After the catheter is positioned, the peritoneal hole is closed. **Figure 36.16** shows the layout before routine sternotomy closure.

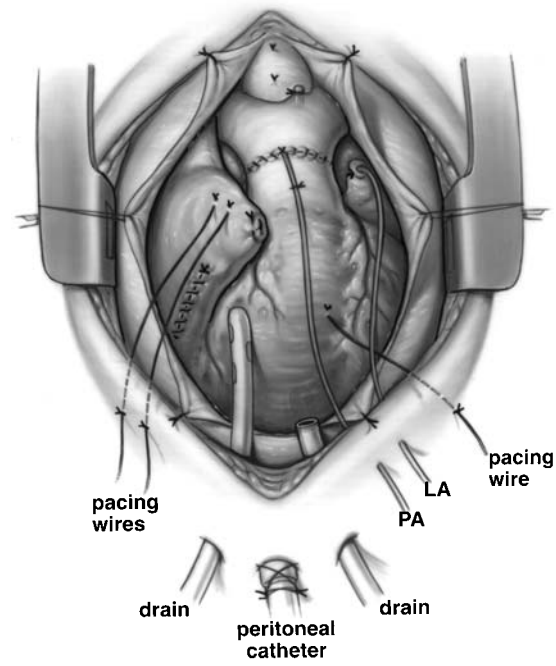

Figure 36.16

POSTOPERATIVE MANAGEMENT

General

The patient is returned to the intensive care unit still paralysed and ventilated. Arterial PO_2 readings commonly are low initially but improve within 12–36 hours. In general, there has been little benefit in terms of blood pressure from volume loading beyond a mean left atrial pressure of 10 mmHg, and so this is carefully avoided. Intravenous phenoxybenzamine is continued every 8 hours at doses of 0.3–0.5 mg/kg. The dopamine infusion is only rarely reduced to less than 5 μg/kg/minute before the patient is weaned from the respirator, and it is routinely continued at diminishing doses for 1–3 days after non-assisted spontaneous respiration is established.

In patients with simple TGA who are referred after 2–3 weeks of age and do not have significant patent ductus arteriosus or an element of dynamic LVOTO, it is common for mean arterial pressure to hover at 40–45 mmHg for 2–3 days, in the presence of peripheral warmth. In such patients, particular care is taken to avoid volume overloading and, if possible, the mean left atrial pressure is maintained at 6–8 mmHg while the development of LV hypertrophy is awaited. The author has observed a gradual rise in mean arterial pressure, which allowed stepwise reduction in ventilatory support and enabled extubation by 3–6 days after operation. Any significant rise in left atrial pressure (more than 10 mmHg) during weaning is handled by reparalysis and a wait of 24 hours before the process is repeated. Phenoxybenzamine is generally discontinued about 3–4 days after the patient leaves the intensive care room.

The Peritoneal Dialysis Catheter

Initially, this catheter is used simply as a continuous drain. If urine output falls below 0.5 ml/kg/hour and is

unresponsive to one or two intravenous 1 mg/kg doses of frusemide, or if the serum potassium level rises about 5.0 mmol/litre, peritoneal minidialysis is commenced. This consists of a regime of 30 minute cycles involving 10 ml/kg alternating isotonic (1.5%) and hypertonic (4.25%) standard dialysis solutions without added antibiotics or heparin. The ''in and dwell'' cycle is 20 minutes long, and the ''drain'' cycle is 10 minutes long. A negative dialysis balance is initially replaced 100% with fresh-frozen plasma, 5% albumin in normal saline and one-third the volume of concentrated (25%) albumin, alternated hourly to create a 3-hour cycle.

If 100% replacement subsequently produces a rise in the left atrial pressure, the replacement is reduced first to 80% and then to 50% of losses. The potassium added to the dialysate fluid is adjusted to 0–3.5 mmol/litre. Dialysis is continued for periods of several hours to several days, as deemed necessary. The temperature of the dialysis fluid is controlled carefully in order to add or remove heat from the patient, as indicated by core (rectal) temperature monitoring.

Left atrial and pulmonary arterial catheters are tied off when no longer required for monitoring and are removed before the chest drains under conditions of early extubation, but if chest drains have already been removed the lines are capped and removed on the fifth postoperative day, followed by a precordial echo to exclude early bleeding. Blood glucose levels are carefully monitored (hourly) for the first 24 hours in neonates, and levels are maintained above 7 mmol/litre for the first few days with intravenous infusion of 50% glucose (1–2 ml/hour). The serum ionized calcium level is similarly monitored and maintained at 1.18–1.25 mmol/litre by molar calcium gluconate infusion (0.5–1.0 ml/hour). Patients over the age of 1 month with simple TGA are given 50% of the normal maintenance dose of thyroxin orally, which commences early postoperatively and continues for 2–3 days after the patient leaves the intensive care unit.

RESULTS

The arterial switch operation is technically more challenging than inflow atrial repairs, and so the learning curve has been longer and more hazardous. Risk is related to experience (**Table 36.2**) (Norwood *et al*., 1988); the crucial part of the operation remains coronary artery translocation, some coronary configurations being more difficult to treat than others. The author maintains that all coronary configurations can be handled with reasonable safety.

Indication 1: Complex TGA and DORV

In May 1983, the author decided to offer arterial switching to patients with complex TGA in whom it was

Table 36.2 Hospital mortality related to number of operations performed.

No. of arterial switch repairs per institution		Total patients			
			Deaths		
≤<	No. of institutions	(*n*)	No.	(%)	CL (%)
10	9*	20	11	55	41–68
10–20	4†	59	21	36	29–93
20–30	1‡	22	2	9	3–20
30–40	0	—	—	—	—
40–50	—	—	—	—	—
50	2	111	10	9	6–13
Total	16	212	44	21	18–24
p (logistic)				<0.0001	

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CL, confidence limit.

* One of the nine institutions performed five operations, with one death (20%; CL = 7–93%); these were considered low-risk institutions.

† One of the four institutions performed 15 operations, with two deaths (13%; CL = 4–29%) and was considered a low-risk institution.

‡ This institution was considered a low-risk institution.

known that the results of atrial repair would be poor, viz. those with a small right ventricle. Until December 1991, the total experience had comprised 98 patients weighing 1.6–15 kg (median, 3.8 kg). Ages at operation ranged from 2 days to 7 years (median, 2.8 months). Fourteen patients had undergone pulmonary artery banding, 10 coarctation repair, one creation of classical Blalock–Taussig shunt and one a Blalock–Hanlon septectomy.

Additional procedures performed at time of arterial switch and VSD closure were aortic arch repair in 13 patients, resection of obstructive right ventricular outflow tract muscle in six, LVOTO resection in four and repair of total anomalous pulmonary venous drainage to the coronary sinus in one. Of 98 patients, 17 had DORV; 12 had DORV with subpulmonary VSD, two with doubly-committed VSD, three with uncommitted VSD, and three with multiple VSDs. One patient also had hypoplastic left ventricle. The other 81 patients had complex TGA, 20 with uncomplicated perimembranous VSD and 14 with perimembranous VSD associated with some other complexity. Twenty-one patients had malalignment VSD, eight with inlet VSD, 18 with muscular VSD, and nine with multiple VSDs. Additional associated problems comprised ''criss-cross'' atrio-ventricular connection to two, hypoplastic right ventricle in seven, posterior TGA in one, situs inversus in one, and total anomalous pulmonary venous drainage to the coronary sinus in one; one 4-month-old who was a Jehovah's Witness underwent repair without blood products.

Yacoub type A coronary anatomy was present in 54 patients (three intramural, including one with ostial stenosis), type D in 24 (one intramural), type B in seven (one intramural with ostial stenosis), and type E in four; the anatomy in 11 other patients did not fit into Yacoub's classification. Great vessel relationship was largely anterioposterior in 69 and largely side-by-side in 29. There were six early deaths (6%).

The follow-up period was 2 weeks–103 months; there was one late death in a patient who died overseas from recurrent sternal infection. Eight patients have required reoperation, and all are currently well. Four of these patients required MPA reconstruction; two of these four also required RVOTO resection, and one in addition required repair of supravalvar neo-aortic stenosis. One patient required closure of residual VSDs as well as RVOTO resection. One patient required LVOTO resection. One, who had undergone LVOTO resection of accessory endocardial cushion tissue, also required early repair of a perforated mitral valve. The eighth patient required repair of proximal aortic arch stenosis.

The author's additional experience from July 1993 to June 2004 comprises 44 additional complex transpositions, anatomically repaired under the age of 3 months; 35 had TGA + VSD and nine had DORV. Arch reconstruction was required in three with TGA + VSD and in six with DORV. There was one death at 6 weeks postoperatively with renal shut-down preceding repair and continuous postoperatively. This patient had DORV, arch obstruction, smallish LV and multiple VSDs. Sepsis was the eventual cause of death. There were no deaths early or late in complex TGA repaired after 3 months of age, many of whom had been debanded.

The principal risk factors appear to be the following:

1. Inexperience of the surgical staff (Norwood *et al*., 1988).
2. Miscalculating ventricular sizes.
3. Multiple VSDs not adequately dealt with.
4. Attempted resection of organic (fibrous) LVOTO.
5. Inadequate management of aortic arch hypoplasia.
6. Poor siting of translocated coronary arteries.
7. Pulmonary vascular obstructive disease in patients who present late and have not undergone pulmonary artery banding.

Organic LVOTO is a relative contraindication. Ease of resection should be taken into account. A bicuspid pulmonary valve creates unease but is not necessarily a contraindication. The author and associates advocate aggressive management of RVOTO at the subvalvar level and enlargement of a poorly committed or restrictive VSD in treating DORV. The aim is to preserve the function of the neo-pulmonary valve (by resuspending commissures instead of cutting across them during explantation of eccentrically placed or intramural coronary arteries). They believe that intramural coronaries can be handled satisfactorily (Asou *et al*., 1994).

Indication 2: Simple TGA, Including Some with Small or Tiny VSDs that Were Not Closed

At the author's institutions, the first arterial switch operation for simple TGA was performed in December 1983. Two patients who presented did not undergo an operation; both arrived at the hospital in moribund condition, and both died within 2 hours of arrival during attempts at BAS under cardiac massage. In all, by December 1991, 134 patients with simple TGA had undergone an arterial switch operation. Weights at operation were 1.8–9.5 kg (median, 3.2 kg); ages were 2 days–10 months (median, 11 days). Thirty-nine patients were in the first week of life, 68 in the second week, nine in the third week, eight in the fourth week, five in the sixth week, three in the eighth week, one in the fourth month (dynamic LVOTO), and one in the tenth month (pulmonary artery banding). Yacoub type A coronary anatomy was present in 81 (with four intramural), type D in 36, type B in

eight (with three intramural), and type E in one; other configurations not identifiable by Yacoub classification were present in eight. The great vessel relationship was anterioposterior in 133 and side-by-side in one.

There was one early death (day 1) in a 1.8 kg premature girl who was 22 days old. The death was sudden, after adequate progress with excellent haemodynamics, and was clearly of an ischaemic nature. The coronary anatomy was Yacoub type A with a long, tortuous left main coronary artery.

The follow-up period ranged from two weeks to 96 months (mean, 38 months). There have been no late deaths. Twenty-seven patients underwent primary arterial switch when they were over the age of 2 weeks. The single early death for simple TGA was in this subgroup, but the mode of death was not LV inadequacy. This experience is at odds with Norwood *et al.*'s (1988) 20-institute study (see **Table 36.1**), in which only three of 125 infants who underwent switch operations in low-risk institutions were over the age of 15 days, and one of the three died.

Reoperations have been necessary in seven of these patients. All survived and were well at the time of review in December 1991. Three patients required repair for coarctation, which was diagnosed weeks to months after the arterial switch. Two patients required MPA reconstruction, and one of these patients also required repair of supravalvar neo-aortic valve stenosis (Yacoub type B coronary anatomy). One patient underwent repair of proximal left pulmonary artery stenosis, and one patient underwent repair of a cleft mitral valve.

Of 133 survivors, 55 were catheterized 10–47 months (mean, 22 months) after the operation. The results are shown in **Table 36.3**.

In our series of simple TGAs, the only identifiable risk factor was Yacoub type A coronary anatomy with long, tortuous left main coronary artery; there were three such patients. In one, the left coronary artery had to be relocated three times, and the patient survived and did well after 6 hours 8 minutes on cardiopulmonary bypass; in the one patient, the left coronary artery was kinked but was adjusted by the application of a horizontal plicating mattress suture to the edge of the coronary cuff; and the third patient, a 1.8 kg premature girl, did well initially but died suddenly of myocardial ischaemia toward the end of the first day. In this form of coronary anatomy, the bifurcation into left anterior descending and circumflex arteries is fixed and partly buried in the ventricular muscle. The tortuous left main coronary artery is almost devoid of surrounding fat and is suspended on a ''mesentery''. It appears that this artery requires translocation high (above the neo-aortic anastomotic suture line) and more right-wardly.

Table 36.3 Postoperative catheter studies for simple TGA (55 patients).

Gradient (mmHg)	Patients (*n*)
Left ventricle to aorta	
0–10	11
20	1*
50	1†‡
Right ventricle to pulmonary artery	
0–9	30
10–19	19
20–30	4 (1†)
50	1†
75	1†
Coarctation	3†

Postangiocardiographic left ventricular end-diastolic pressure recorded in 31 patients.
In no case has an obstructed coronary artery been identified.
* With bicuspid valve.
† Reoperations.
‡ With Yacoub type B coronary anatomy.

Additional experience gained by the author from July 1993 to June 2004 comprised 105 patients undergoing primary switch up to the age of 2 months. Four required coarctation repair, either at initial surgery or within 2 months of the arterial switch. There have been no early or late deaths in this 11-year series.

Indication 3: Double-inlet Ventricle or LV Type with TGA and Severe Subaortic Stenosis, Severe Arch Hypoplasia and Coarctation of the Aorta, or Interrupted Aortic Arch

After various attempts at palliation, including creating aortopulmonary windows with distal MPA banding and various staged arch reconstructions, we combined arch reconstruction, arterial switch and atrial septectomy as a preliminary palliation, to be followed eventually by a Fontan-type operation. Eight neonates in whom the descending aortic supply was dependent on the ductus arteriosus were maintained by PGE_1 infusion, were resuscitated, and then underwent this form of palliation early. The subaortic obstruction became subpulmonary obstruction after the arterial switch and adequately limited pulmonary blood flow. This obstruction progressed and subsequently systemic-to-pulmonary shunting or bidirectional cavopulmonary shunting was usually necessary.

There were two early deaths, one caused by neo-aortic valve incompetence. This was believed to be a result

of matching the very large proximal MPA to the small distal ascending aorta by gathering and thus bringing the pulmonary valve commissures inwards, which caused prolapse of the cups. The author thinks that this can be avoided by dividing the MPA high and translocating the coronary arteries below the neo-aortic suture line. The other early death was caused by bleeding. There was one late death from known residual aortic arch stenosis, which was neglected. Four patients underwent creation of Gore-Tex shunts from the right subclavian artery to the right pulmonary artery. Five underwent creation of bidirectional cavopulmonary shunts, and two of these patients underwent completion of a total cavopulmonary anastomosis.

Creating neo-aortic valve incompetence from commissural gathering can be avoided by high transection of the MPA. If a short segment of the small distal ascending aorta is left intact proximal to the arch reconstruction, it is likely to become a site of obstruction. In the last two patents of this series, the highly transected proximal MPA was anastomosed to the combined distal ascending aorta, proximal aortic arch and proximal descending aorta.

Indications 4 and 5: Conversion of Previous Mustard and Senning Operations to Arterial Switch

A two-stage approach is used for patients with RV dysfunction, tricuspid valve incompetence, and no significant dynamic LVOTO. The first stage involves MPA banding via a midline sternotomy and is followed after a variable interval by removal of the band, reversal of the Senning or Mustard procedure, and arterial switching. Operative details are described elsewhere (Mee 1986, 1993; Cochrane *et al*., 1993; Poirier *et al*., 2000, 2004). Retraining of an involuted LV has proved easier and more reliable in patients presenting for PA banding before their mid-teens, as opposed to older age. Associated cardiopulmonary abnormalities appear to play an important role in switch conversion failures. A review of the author's personal experience from 1981–1993 at the Royal Children's Hospital (RCH) Melbourne and from 1993–2003 at the Children's Hospital at the Cleveland Clinic Foundation (CCF) includes 39 patients (19 at RCH and 20 at CCF) presenting for conversion to an arterial switch at a median age of 10.8 years (13 months–24 years) and a median of 10.3 years (0.5–24 years) after a Mustard or Senning operation. Four patients had LVOT obstruction and proceeded to arterial switch conversion without pulmonary artery banding, with two early deaths from uncontrolled bleeding from the thin-walled proximal MPA. Thirty-five patients underwent PA banding with one early death and four late deaths before LV training was completed. Four patients had progressive RV dysfunction before LV retraining was adequate and underwent heart transplantation. Twenty-five patients underwent conversion to arterial switch, a median of 13 months (0.5–5.4 years) after banding, with four early deaths, three late deaths and two patients progressing to heart transplantation because of postconversion progressive LV dysfunction. One patient has had a PAB for 5.4 years, has RV function restored to almost normal with regression of TR to minimal. There appears to be no clear advantage to converting this patient until such time that RV dysfunction returns. It should be noted that 11 of the 19 RCH patients preceded the development of the National Pediatric Transplant Center at RCH in 1988 and four of the 20 CCF patients were clinically not heart transplant candidates. We have learned that LV retraining and conversion to arterial switch is more successful in patients younger than 12 years ($p = 0.02$). The older age group is clearly more difficult to retrain and convert, and currently the majority of patients now being put forward for this protocol are over 20 years old. Pre-existing LV dysfunction or MR, significant arrhythmias requiring drug control and pacing, late stage RV dilatation, dysfunction and TR, complex coronary anatomy, and a history of large VSD closure or left ventriculotomy are relative contraindications to attempted LV retraining and conversion to arterial switch in the older age group. A parallel heart transplantation programme ensures customized selection of patients and the ability to move from the switch conversion protocol to transplantation if warranted. Finally, older patients may sometimes benefit from moderate pulmonary artery banding in terms of RV dysfunction and TR regression, acting as a bridge to subsequent transplantation or a decision to proceed along the conversion protocol.

Indication 6: Congenitally Corrected TGA with Morphological RV Dysfunction and Tricuspid Valve Incompetence

Results for this group of patients are described in Chapter 39.

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37

Surgery of the Left Ventricular Outflow Tract

V. T. Tsang and M. R. de Leval

ANATOMICAL SUBSTRATE

Normal Left Ventricular Outflow Tract

The left ventricular outflow tract is a complex structure that includes the aortic valve, the sinuses of Valsalva and the subvalvar fibromuscular outflow from the left ventricle. The aortic root is a dynamic unit in which all components contribute to the opening and closure of the aortic valve and are continuously changing during a cardiac cycle in response to the aortic pressure, the ventricular pressure and the ventricular geometry (Thubrikar *et al*., 1981). Congenital or acquired anomalies of any component of the aortic root can eventually lead to aortic valve dysfunction.

The normal aortic valve is made of three cusps that have a crescentic shape, from which the name ''semilunar valve'' originates. There is a small fibrous thickening in the middle of the free edge of each cusp (nodule of Arantius). The cusps overlap in their closed position. The closure line produced by the apposition of the adjacent cusps is visible on their ventricular surface, 2–3 mm from the free edge (Angelini *et al*., 1989). The three cusps are attached to the aortic wall in a crescentic manner without a circular structure that could be assimilated to a ring or an annulus. The cusps are separated by the commissures. Above the valve, the aortic wall is expanded to form the sinuses of Valsalva. The dilatation of the sinuses ends at the level of the tip of the commissures. At that level, there is a discrete ridge marking the sinotubular junction of the aorta. On its ventricular aspect, the insertion of the valve delineates three triangular areas between each cusp where the aortic wall is thinner. These triangles separate the left ventricular outflow tract from the pericardial cavity. The sinuses and the cusps are named according to the origin of the coronary arteries, namely the left, right and non-coronary sinuses and cusps. In a normal three-leaflet aortic valve, the length of the free edge of each cusp equals the diameter of the aorta. The combined length of the free edges of the three cusps equals the circumference of the aorta at the sinotubular junction. In a bicuspid valve the combined length of the free edge is at best twice the diameter of the aorta. The orifice is therefore significantly smaller than the circumference of the aorta (Doty, 1987).

A precise knowledge of the anatomy of the aortic root and its relationship with the surrounding cardiac structures is of great importance to the surgeon (**Figures 37.1** and **37.2**). This is particularly true for the repair of congenital malformations necessitating the enlargement of the left ventricular outflow tract (Sud *et al*., 1984). The understanding of the hinge mechanism of the left and right fibrous trigones in the subaortic region would also improve the surgery for left ventricular outflow tract obstruction (Yacoub *et al*., 1999).

The entire right coronary sinus lies adjacent to the right ventricular outflow tract. Inferiorly, it relates to the interventricular septum. The non-coronary sinus lies adjacent to the right and the left atrial chambers and the interatrial septum. Inferiorly, the commissure between the right and the non-coronary cusps is related to the membranous septum and to the penetrating atrio-ventricular bundle. The left part of the non-coronary sinus inserts into the anterior mitral leaflet. The commissure between the non-coronary and the left coronary leaflets is positioned above the area of aortic mitral valve continuity. The left coronary sinus lies adjacent to the left atrium on the right and to the lateral aspect of the base of the left ventricle to the left. Inferiorly, it relates to the anterior leaflet of the mitral valve. To the right, this is the only part of the aortic root that does not relate to a cardiac chamber but to the free wall of the left ventricle. The commissure between the left and the right coronary cusps lies adjacent to the corresponding commissure of the pulmonary valve, and there is an area of dense fibrous tissue between these commissures.

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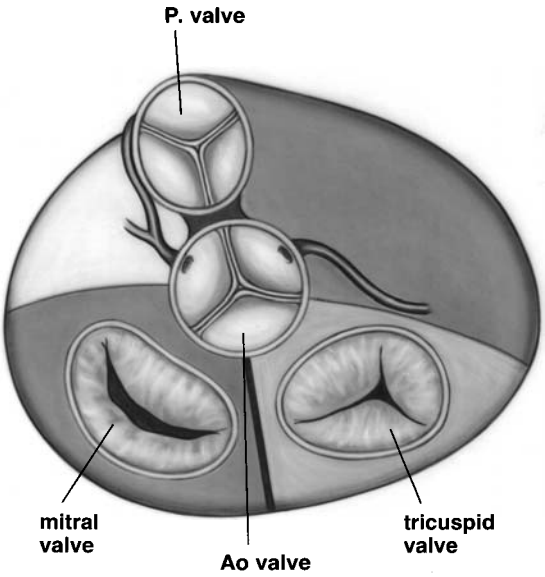

Figure 37.1

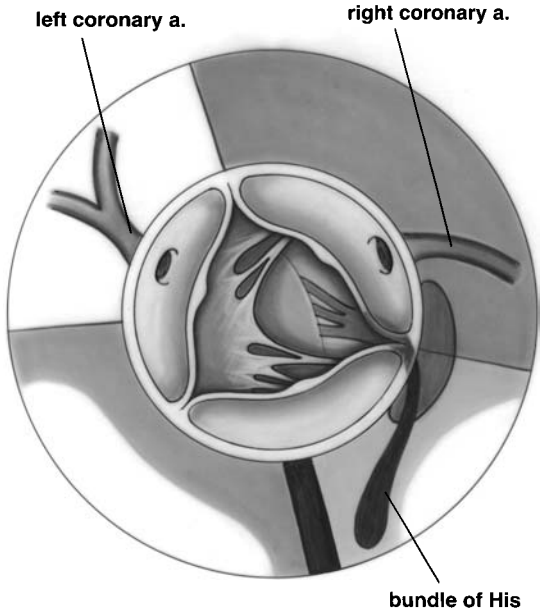

Figure 37.2

Abnormal Left Ventricular Outflow Tract

Obstruction to the left ventricular outflow tract may be found at valvar, subvalvar, supravalvar or combined levels. The left ventricular outflow tract obstruction may be isolated or associated with other defects, such as coarctation of the aorta, interruption of the aortic arch, ventricular septal defect, pulmonary stenosis, patent ductus arteriosus, mitral valve abnormalities or, less commonly, atrio-ventricular septal defects, atrial septal defects and anomalies of the pulmonary venous return. The association of aortic stenosis with parachute mitral valve and coarctation of the aorta is known as the Shone syndrome (Shone *et al*., 1963).

Aortic valve regurgitation is rarely an isolated congenital lesion. More often, the aortic regurgitation develops as a complication of valvar or subvalvar aortic stenosis. It can also be associated with other congenital cardiac anomalies, such as subarterial ventricular septal defects. Aortic valve regurgitation is more frequently an acquired disease due to an inflammatory or degenerative process. It may also be related to a dilated aortic root in connective tissue disorders. Nowadays, balloon valvuloplasty and surgical valvotomy is the main cause of aortic regurgitation.

Congenital Valvar Aortic Stenosis

The various anatomical forms of aortic valve stenosis have been well described (Edwards, 1961). The valve is bicuspid in about 70% of cases. The leaflets, which are usually thickened, are orientated either laterally, with the commissures positioned anteriorly and posteriorly, or they are arranged anteroposteriorly, with the commissures to the right and to the left. A fibrous ridge or raphe, which varies in length and thickness, is often present on one of the two cusps. There are usually variable degrees of peripheral fusion of one or both commissures. Stenosis can occur without commissural fusion simply as a result of thickened leaflets and a bicuspid configuration. If the free edges of both thickened bicuspid leaflets are taut and equal in length to the diameter of the aortic root, they cannot open and they produce a significant gradient. Stenosis may result from the unequal size of the cusps, which is frequently seen in bicuspid aortic valves. When the commissures are situated in an anteroposterior plane, the coronary arteries usually arise from each sinus of Valsalva. When the commissures are situated in a frontal plane, the coronary arteries more frequently arise from the posterior sinus.

In about 30% of cases, the stenotic valve is tricuspid, with thickened leaflets that may or may not be of even size and variable degrees of peripheral commissural fusion. This type of valve produces a dome with a central stenotic orifice, and it constitutes the most favourable configuration for valvotomy. In rare instances, the valve may be unicuspid, with a single commissure that may or may not present with peripheral fusion. A unicuspid valve is usually severely stenotic, even in the absence of commissural fusion.

In aortic valve stenosis, there is always a secondary concentric left ventricular hypertrophy, which sometimes results in virtual obliteration of the left ventricular cavity. Endocardial fibroelastosis as a result of chronic ischaemia from high LV wall stress is a common feature of severe critical aortic stenosis of the neonate.

Supravalvar Aortic Stenosis

Supravalvar aortic stenosis can be localized or diffuse (Rastelli *et al*., 1956). In the localized type, the aortic narrowing is made by an annular ridge protruding into the lumen of the aorta immediately above the commissural attachment of the aortic cusps. The outer diameter of the aorta may be normal or reduced, giving an hourglass appearance to the ascending aorta. In the diffuse type, the narrowing of the lumen and the abnormal thickness of the wall usually involves the ascending aorta and extends to the origin of the head vessels. The stenotic region has a thickened intima and hypertrophy of the media with an increase in fibrous and elastic tissues.

Abnormalities of the aortic cusps are present in at least one-third of cases, the most common being underdevelopment of the left coronary cusp. Sometimes, the supravalvar ring may almost cover the already diminutive entry into the sinus of Valsalva. At times, the free edge of the cusp is adherent in places to the ring. This may impair the filling of the left coronary artery. The coronary arteries are often dilated and tortuous. Peripheral stenoses of the pulmonary arteries are the most common associated anomalies. Supravalvar aortic stenosis has also been described in association with unusual ''elfin'' facies, mental retardation and infantile hypercalcaemia. This syndrome is often known as the Williams syndrome (Williams *et al*., 1961).

Subvalvar Aortic Stenosis

Fixed subaortic stenosis can be discrete or diffuse. The commonest type of discrete stenosis is made of a crescent or, less commonly, a complete ring of fibrous tissue that lies 5–10 mm beneath the aortic valve. The orifice is usually eccentric. The fibrous shelf is attached to the anterior cusp of the mitral valve and the thickened endocardium of the ventricular septum. Sometimes, the shelf may also be attached to the aortic cusps. The left ventricular myocardium shows concentric hypertrophy when the lesion is severe and long-standing. There is often excessive septal bulging. There is some evidence to suggest that this type of discrete subvalvar aortic stenosis is not congenital but an acquired obstruction secondary to flow disturbances in the left ventricular outflow tract (Gewillig *et al*., 1992).

Diffuse subvalvar aortic stenosis is caused by a circumferential zone of fibromuscular hypertrophy below the aortic valve and extending downward into the left ventricular cavity. This type of obstruction is to be differentiated from hypertrophic obstructive cardiomyopathy (HOCM), a genetic disease involving the myosins and troponin. The clinical spectrum (Wigle *et al*., 1995) is characterized by basal septal ventricular hypertrophy and a dynamic obstruction associated with abnormal systolic anterior motion (SAM) of the anterior leaflet of the mitral valve into the LVOT.

CLINICAL PRESENTATION AND DIAGNOSIS

Neonates and infants with critical *aortic valve stenosis* present in severe congestive heart failure with reduced-volume peripheral pulses and a systolic ejection murmur in the aortic area that may sometimes be unimpressive because of low cardiac output. The chest X-ray shows cardiomegaly, and the electrocardiogram reveals left ventricular or biventricular hypertrophy. T-wave changes in the left chest leads are present on the electrocardiogram of most patients. The diagnosis is confirmed by cross-sectional echocardiography and Doppler study. A doming dysplastic aortic valve and a hypertrophied and often dilated left ventricle are the typical echocardiographic features of critical aortic valve stenosis. The aortic valve annulus is frequently measured at less than 6 mm in diameter. There is often some degree of endocardial fibrosis, hypoplasia of the left ventricle and/or aortic annulus.

Care must be taken to rule out a coarctation of the aorta with associated bicuspid aortic valve because, in the presence of low cardiac output, the clinical and echocardiographic features may be misleading. Patients with congestive cardiomyopathy resulting from primary myocardial disease or anomalous origin of the left coronary artery from the pulmonary artery have a thin-walled dilated left ventricle and a normal aortic valve. The differentiation between critical aortic stenosis and left ventricular hypoplasia may be difficult (see later).

The Doppler gradient across the aortic valve can be misleading in a situation of low cardiac output. A patent ductus arteriosus is not uncommon in neonatal aortic stenosis. A ductal-dependent systemic circulation with right-to-left shunt at ductal level can be seen in the most severe cases. Mitral valve anomalies are common in this group of patients with multilevel left heart lesions. In the majority of cases, the diagnosis can be made without cardiac catheterization.

Children and young adults with aortic stenosis may remain asymptomatic, even with important gradients. However, angina and/or breathlessness, and/or syncope

during exercise usually indicate significant obstruction. Congestive heart failure occurs exceptionally in the absence of previous warning symptoms. On the chest X-ray, the ascending aorta is often prominent, indicating a poststenotic dilatation. Serial Doppler echocardiographic techniques are currently sufficiently reliable to estimate aortic valvar gradients and LV dimensions, thus avoiding the need for cardiac catheterization.

The symptoms of congenital *subvalvar aortic stenosis* are similar to those of the valvar variety. The echocardiogram is usually diagnostic in demonstrating a discrete localized or diffuse stenosis and can also easily differentiate the discrete obstruction from the characteristic features of obstructive hypertrophic cardiomyopathy. Trivial or mild aortic valve incompetence is present in about two-thirds of cases with subaortic stenosis.

The clinical features and physical signs of *supravalvar aortic stenosis* are similar to those in other types of congenital aortic stenosis. It is necessary to outline the type and extension of the obstruction and to assess the pulmonary arteries, in view of the frequent association of supravalvar aortic stenosis with peripheral pulmonary artery stenoses. However, cardiac catheterization and angiocardiography may interfere with the coronary flow in the setting of compromised coronary ostia. Magnetic resonance imaging (MRI) is currently our preferred choice of safe imaging to obtain the necessary information in this condition.

Patients with **aortic incompetence** frequently present without symptoms because of its long asymptomatic phase unless the lesion is serious or develops acutely. The wide pulse pressure, the early diastolic murmur that radiates toward the apex often accompanied by an ejection systolic murmur, and the mid-diastolic murmur at the apex caused by fluttering of the anterior mitral leaflet are the classical physical signs of aortic valve regurgitation. The chest X-ray shows left ventricular enlargement with or without enlargement of the ascending aorta. The electrocardiographic changes depend on the severity and duration of aortic regurgitation. Serial two-dimensional and colour Doppler echocardiography are often sufficient to define the degree of regurgitation and the LV cavity dimensions. However, the optimal timing for surgery is not clearly defined (Cheung *et al*., 2003).

INDICATIONS FOR OPERATION AND INTERVENTIONS

Symptomatic neonates and infants with *critical aortic stenosis* need urgent treatment. They often need resuscitation and intensive care management, including mechanical ventilation, correction of the metabolic acidosis and inotropic support. An infusion of prostaglandin to maintain the patency of the ductus arteriosus often improves the circulation. Although critical aortic stenosis in the neonate is still often managed surgically in some centres (Gaynor *et al*., 1995), balloon dilatation of the aortic valve in the catheterization laboratory is currently the standard method of treatment (Lababidi *et al*., 1984). However, one should be aware of the occasional risk of damage to the aortic and mitral valves with the interventional approach, which may lead to emergency surgery.

More babies are now born with a borderline LV. It is often difficult to determine whether *the small or borderline LV* is adequate to sustain the systemic circulation without a patent ductus arteriosus. Even more difficult is to determine whether biventricular repair is feasible. Echocardiographic indices have been proposed for surgical decision, such as aortic and mitral valve annular dimensions, indexed LV outflow and inflow dimension, the ratio between the LV:RV apex-to-base dimensions, LV long axis: heart total long axis (AV valve annulus: apex) ratio, and LVEDV (Kirklin and Barratt-Boyes, 1993). Rhodes *et al*. (1991) developed a multivariate equation using echo measurements to produce a score that allows prediction of mortality after biventricular repair for critical aortic stenosis. However the score should be used with caution and other anatomical variables, such as the LV mass, presence of endocardial fibroelastosis and non-apex forming LV, should also be considered. Kovalchin *et al*. (1998) found that retrograde flow through the transverse arch was associated with higher risk after a biventricular repair. CHSS has come up with the idea of a ''calculator'', based on a multi-institutional study of management, outcomes and risk factors of a large group of neonates with critical aortic stenosis, to stratify these complex patients to single ventricle vs. two ventricle repair (Lofland *et al*., 2001). We have recently recognized the persistence of pulmonary hypertension due to the restriction of LV filling, despite an apparently ''successful'' biventricular repair (Burch *et al*., 2004). Nevertheless, the surgical options (Norwood, neonatal Ross $\pm$ LV endocardial fibroelastosis resection, or cardiac transplantation) are difficult in this group of patients. Longer-term follow-up data are essential to decide whether it is preferential to have a good single ventricle repair or a biventricular repair with possible subsequent reoperations. Conversely, it may be a disadvantage to have a borderline hypertensive LV contributing to an inefficient function of the neo-systemic right ventricle (Cohen and Rychik, 1999).

In childhood, the indications for aortic valvotomy are: (a) electrocardiographic changes at rest or after moderate exercise, even in the asymptomatic patient; (b) a resting peak systolic gradient greater than 60 mmHg; and (c) symptoms related to valve stenosis (Kirklin

and Barratt-Boyes, 1986). The increasing experience with aortic balloon angioplasty is bound to lower the threshold for the indications for treatment of aortic valve stenosis. Ideally it should be dealt with before the left ventricle develops irreversible myocardial changes. However, it is also important to keep in mind that premature valvuloplasty or valvotomy on the mildly to moderately stenotic aortic valve may result in an important regurgitation.

The indications for surgical relief of discrete *subvalvar stenosis* are reasonably well defined. Because of the progressive nature of this malformation, it has been suggested that surgery is indicated even with gradients as small as 20 mmHg (Somerville, 1987). Serial echocardiographic data with increasing gradient, and the detection of aortic regurgitation, would be our indications for surgery.

Supravalvar aortic stenosis is usually a progressive lesion, with an increase in left ventricular outflow tract pressure gradient related to poor growth of the ascending aorta. Hence, early surgical intervention is indicated (Wren *et al*., 1990).

Asymptomatic patients with *aortic valve incompetence* present a difficult problem. When left ventricular enlargement reaches a certain point, the secondary cardiomyopathy may not be completely reversible. It has been suggested that surgery is indicated even in asymptomatic patients when the cardiothoracic ratio exceeds 55% (Kirklin and Barratt-Boyes, 1986). Our data would also suggest that recovery of LV function is less likely when the *z* score of the left ventricular end diastolic dimensions has reached the value of 4. Therefore, early intervention is recommended (Cheung *et al*., 2003).

SURGICAL TECHNIQUES

Valvotomy in Neonates and Infants

Valvotomy in neonates and small infants, performed as a closed trans-ventricular aortic procedure (Trinkle *et al*., 1978) or as an open procedure under inflow occlusion (Sink *et al*., 1984), is outdated because of the risk of refractory ventricular fibrillation or inadequate ventricular performance at the completion of the valvotomy.

Open Valvotomy with Cardiopulmonary Bypass

The use of cardiopulmonary bypass to perform an aortic valvotomy in neonates and young infants provides more time to examine and repair the valve. The surgical technique is essentially similar to that described for older patients. The ductus arteriosus is always ligated. In the neonate, the aortic valve is sometimes made of immature tissue with no commissural structures, in which case the valve orifice is simply enlarged.

Valvotomy in Childhood

The heart is exposed through a midline incision. A purse-string suture is placed at the base of the atrial appendage for the venous cannulation. This suture can be used as a traction to facilitate the dissection and the exposure of the aortic root. The patient is heparinized, the aortic cannula is introduced near the innominate artery, and a single venous cannula is placed in the right atrium. The operation is performed under mild hypothermia.

The aorta is cross-clamped, and the cold cardioplegic solution is infused. An oblique incision is made from the anterior aspect of the aorta downward and to the right to enter the non-coronary sinus of Valsalva posterior to the commissure between the right and the non-coronary cusps. Stay sutures are applied to the edges of the aortotomy for exposure.

The aortic valve is carefully examined. Most commonly the valve is bicuspid, with an eccentric orifice situated between the right and the non-coronary cusps. In about half of cases, a raphe of varying height is present. In occasional patients, it is high enough to simulate a true secondary fused commissure. These raphes do not extend to the free edge of the valve and do not provide lateral support for it. These rudimentary ''third commissures'' should not be incised because this would produce incompetence. The fused commissures are carefully divided with a knife to within 1 or 2 mm of the aortic wall. Cusp mobility and the effective orifice area can be further improved by thinning out areas of leaflet thickening or myxomatous nodules.

In unicuspid valvar stenosis, only one commissure is attached to the aortic wall. The commissure may be any of the three possible normal commissures; in addition, one or two raphes representing abortive commissures may be present. A unicuspid aortic valve is intrinsically stenotic. If the commissure is partly fused, the valve orifice can be enlarged by a commissurotomy (**Figure 37.3**). If not, a short incision at each raphe may be necessary to enlarge the aortic valve orifice. A short incision may result in an appreciable opening of the outflow orifice, because flow is a function of the fourth power of the radius. This is not dissimilar to what balloon valvuloplasty can often achieve with a good opening of a stenotic valve and a small degree of regurgitation, thus providing the platform for further growth. Stenosis of a tricuspid aortic valve is a rare entity in infancy and childhood. The valve is often deformed, and commissures are fused to varying extents. Incisions are made along the fused commissures. If there is doubt as to whether a stenotic valve is tricuspid or bicuspid, it should always be treated as a bicuspid valve.

An anatomical and echocardiographic analysis of critically stenotic aortic valves in infancy (Leung *et al*., 1991) led to the development of techniques of extended valvuloplasty (Ilbawi *et al*., 1991). The incision of the

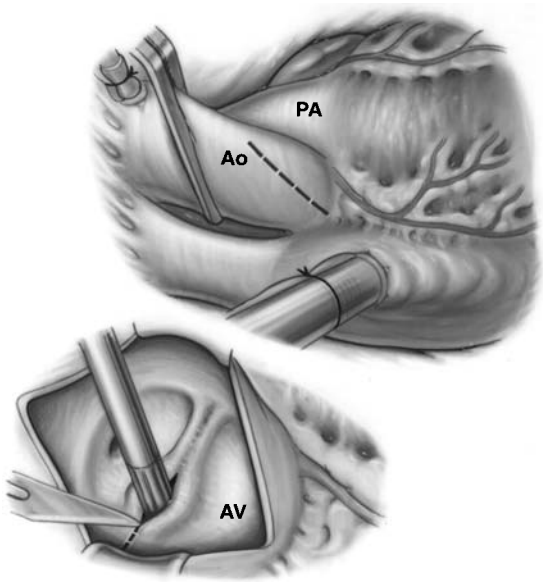

Figure 37.3

fused commissure is extended in a circumferential fashion beyond the commissure into the aortic wall around the insertion of the aortic leaflet for a distance of 1–3 mm on both sides, splitting the aortic wall in two layers and thus giving more mobility to the valve.

Aortic Valve Repair in Children

The advantages of aortic valve repair include growth potential, avoidance of anticoagulation and delay of at times inevitable aortic valve replacement. No single technique can reliably achieve suitable results in all patients (Tweddell *et al*., 2005). Available techniques for aortic valve repair require are commissural resuspension, partial commissural closure, triangular resection of redundant leaflet tissue, annuloplasty, resection of raphe, and autologous or bovine pericardial augmentation of aortic valve cusp. Repair techniques also include procedure on the sinus of Valsalva, sinotubular junction and supra-aortic region in patients with dilated aortic root. Intraoperative trans-oesophageal echocardiography is the mainstay of assessment in operating room. This may make aortic valve repair an exercise of trial and error. The most important issue is to achieve effective delay of the inevitable valve replacement without undue compromise of myocardial function.

Aortic Valve/Root Replacement in the Young

Replacement of the aortic valve in the young age group is one of the most challenging problems for the cardiac surgeon. In infancy and childhood, as there is no other suitable alternative in terms of valve growth, the Ross operation is the procedure of choice at our unit, with excellent functional results in the majority. We have also found the pulmonary autograft procedure very versatile in the setting of concomitant complex left ventricular outflow tract obstruction. However, some current work provide evidence that autograft root dilatation is not an uncommon phenomenon late after the Ross operation. It may be associated with autograft valve dysfunction, leading to regurgitation that increases with time (Luciani *et al*., 2003). Possible explanations include technical error, lack of support of the annulus and sinotubular junction, and valve mismatch with dilated native aortic root.

Another important issue is aortic valve substitution in the adolescents and young adults with adequate aortic annular dimension. The options are prosthetic aortic valve replacement, insertion of an aortic valve homograft, homograft aortic root replacement, and pulmonary autograft aortic root replacement. The disadvantages of aortic homograft are early calcification, no growth, and difficult reoperation with coronary reimplantation. Regardless of the type of mechanical valve implantation, the constant hazard is the risk of haemorrhage and thromboembolism. Bioprosthetic valve is not used at our unit because of the known risk of early calcification in children and adolescents. Nevertheless, pulmonary autograft in this age group is not without concern, because patients with bicuspid aortic valve often have a dilated aortic root (David *et al*., 2000). This may be a contributory factor for late autograft valve dysfunction. Annular hypoplasia, discrete or diffuse left ventricular outflow tract obstruction, and previous prosthetic valve or homograft root surgery would also warrant concomitant enlargement of the aortic root and outflow tract surgery regardless of the types of valve substitution.

Aortic Valve Replacement

Aortic valve replacement operations are performed on hypothermic cardiopulmonary bypass with cold blood cardioplegic arrest. The aorta is cannulated high at the level of the innominate artery. A single arterial venous cannula or two caval cannulae are used for the venous return. The cardioplegic solution is first infused into the root of the aorta, if there is no aortic valve regurgitation, and later directly into the coronary arteries when the aorta is opened. If there is aortic valve regurgitation, the first dose of cardioplegia is also infused directly into the coronary arteries. The left heart is vented via a left atrial catheter. For a prosthetic valve replacement, the aortotomy incision starts as a vertical incision below the aortic clamp and curves down obliquely into the centre of

the non-coronary sinus. Two traction sutures are placed on the lower margin of the aortotomy incision and tied to either side of the pericardial edge, and one suture is placed on the superior aspect of the aortotomy incision. If a homograft valve is to be used, the aortotomy incision stops 10 mm above the top of the commissures, to allow room to sew on the pillars of the homograft above each commissure.

Although the free-hand subcoronary aortic homograft valve insertion is currently seldom performed, its surgical principle, upon which many aortic procedures have developed, is described for completeness.

Aortic Homograft Valve Insertion

The technique of insertion of homograft valves was initially described by Duran and Gunning in 1962 and by Ross, who reported his initial clinical experience in the same year. The aortic valve is excised. The cusps are trimmed completely flat except for the region of the membranous septum, where a small margin of cusp tissue is preserved. The diameter of the aortic valve ring is then measured with an obturator. It is important to choose a sizer that will rest just within the annulus rather than falling into the left ventricle. The selected homograft should have an internal diameter about 2–3 mm less than the diameter of the aortic annulus.

The cryopreserved homograft is then taken through the standard thawing process. When it is completely thawed it is trimmed of excessive tissue. The lower margin consists of the anterior leaflet of the mitral valve and the muscular septum. The mitral valve is first cut straight 2 mm below the nadir of the aortic cusps (**Figure 37.4a**). The cardiac muscle is trimmed to leave a sewing edge 2–3 mm thick, which is strong enough to hold the sutures. The valve is held on the index finger. The myocardial tissue is thinned out. The depth of the thinning is gauged with the thumb and index finger. Superiorly, the aortic homograft is transected 5 mm above the tip of the commissures (**Figure 37.4b**) and then the aortic sinuses are removed, making U-shaped incisions and leaving a rim of 3 or 4 mm of aortic wall for the attachment of the homograft to the aorta (**Figure 37.4c,d**).

For the orientation of the homograft, it is easy to remember that the right cusp is sitting entirely on muscle. Conventionally, the homograft is rotated 120° counter-clockwise so as to bring the homograft's right coronary sinus against the recipient's left coronary sinus. The advantage of the rotation is that the muscular rim of the homograft posteriorly overlies the fibrous tissue of the anterior leaflet of the mitral valve, while the fibrous portion of the homograft lies anteriorly and to the right across the ventricular septum of the recipient.

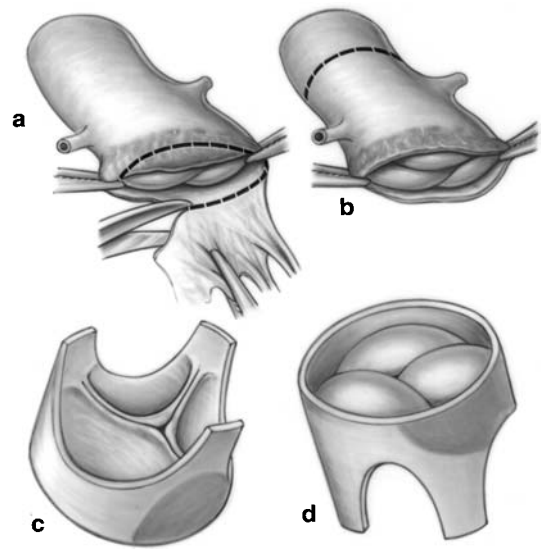

Figure 37.4

The original right cusp becomes the new left cusp. This avoids sewing muscle to muscle anteriorly and results in a more secure suture line that does not bulk posteriorly.

The first of the three guarding stay sutures is now placed immediately beneath the nadir of the remnant of the left cusp and beneath the right cusp of the homograft. The other two sutures are similarly placed beneath the nadir of the other two cusps (**Figure 37.5**). The homograft is then lowered into the left ventricular outflow tract and the valve is turned inside out and inverted, so that the lower edge of the homograft is exposed. The three stay sutures are tied down, and the proximal suture line is begun in a continuous over-and-over technique. This suture line does not follow the scallops of the recipient annulus but proceeds in a horizontal line beneath the commissures. The only exception to this is in the region of the membranous septum, where the suture line should be through or just below the annulus to avoid injury to the conduction system (**Figure 37.6**).

The homograft valve is then pulled back into the aorta, throwing the pillars up into the aortic root. A mattress suture of 4–0 Prolene is placed through the top of each pillar and through the recipient aortic wall about 5 mm above the normal location of the commissures. The ends of each commissural mattress suture are tagged and not tied, to facilitate the manipulation of the graft away from the recipient aortic wall when placing the upper suture line (**Figure 37.7**). The position of the pillars must be judged carefully in both the vertical and the horizontal planes, particularly the two pillars on each side of the

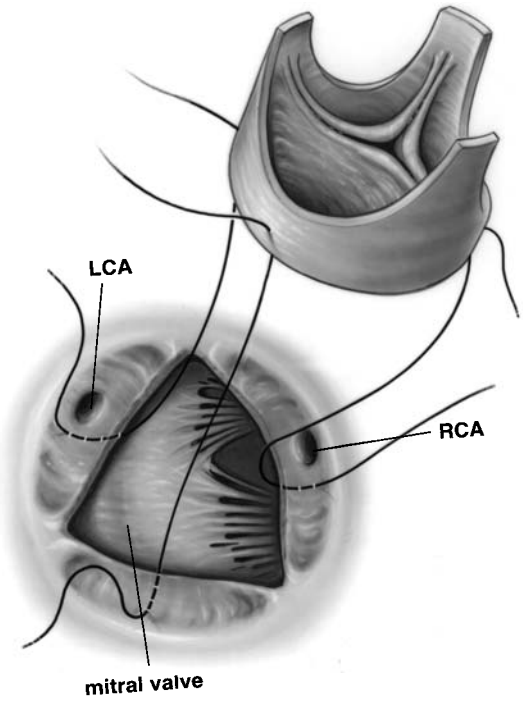

Figure 37.5

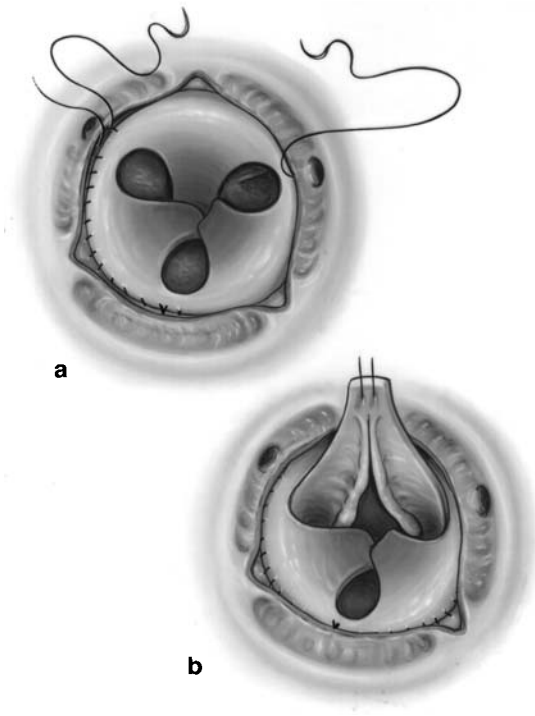

Figure 37.7

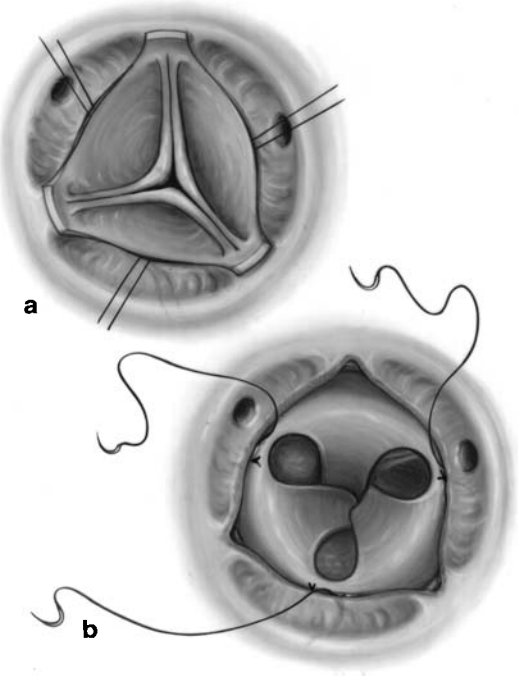

Figure 37.6

aortotomy incision. The distance between these pillars should be a little greater than the distance between the other pillars, to allow for the aortotomy closure. The distal suture line is then carried out, starting at the bottom of the left sinus. The suture line is in a subcoronary position and takes the host's aortic wall and the upper margin of the homograft. The running over-and-over sutures proceed along each half of the left sinus. Each of the sutures is tied to the commissural mattress stitch. The suturing along the right and non-coronary sinus proceeds in a similar fashion (**Figures 37.8** and **37.9**).

The aortotomy incision is closed with a simple over-and-over stitch of 4–0 polypropylene suture, placing a pledgeted mattress suture at each extremity of the incision. In his classic description of the freehand subcoronary homograft insertion, Ross (1962) retained the aortic wall of the non-coronary sinus and did not rotate the homograft into the left ventricular outflow tract of the recipient (**Figure 37.10**). By retaining the non-coronary sinus, one reduces the risk of malalignment of the three pillars and maintains the cylindrical configuration which is so important to avoid valvar regurgitation. Retaining the aortic wall of the sinus, which will occupy the host's non-coronary sinus, does not preclude inversion of the homograft for the proximal

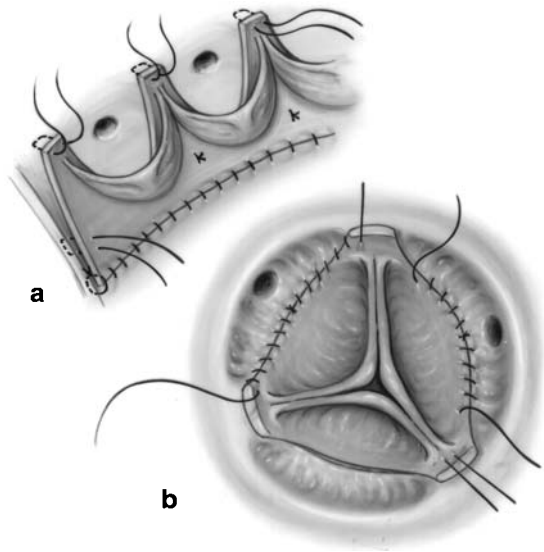

Figure 37.8

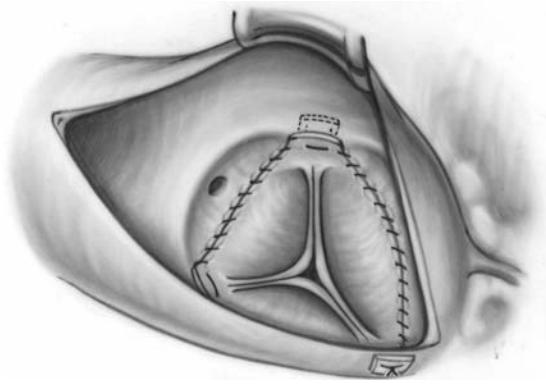

Figure 37.9

suture line. The closure of the aortotomy incision incorporates the wall of the homograft to obliterate the space behind the non-coronary sinus.

In children with small aortic roots, the authors prefer not to invert the homograft while performing the proximal suture line and to use simple interrupted sutures (4–0 Tycron), holding the homograft out of the aortic root to facilitate the exposure. This is indeed the technique that was recommended by Ross even for large aortic roots.

A homograft with an internal diameter of 16 mm can be inserted into a 19 mm root without a haemodynamically significant gradient. However, in paediatric practice, the aortic annulus is often smaller and/or the disease is not

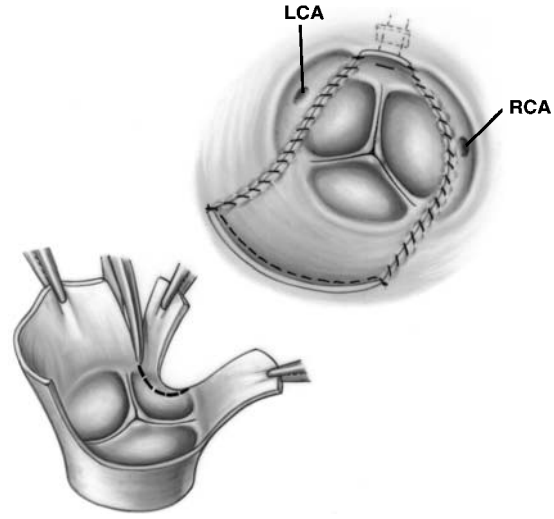

Figure 37.10

limited to the aortic valve itself but to the entirety of the left ventricular outflow tract. In these patients, the techniques of enlargement of the aortic root have to be considered.

Prosthetic Aortic Valve Replacement

Following excision of the native valve, the annulus is sized. The sizer should sit *in* the annulus, not *on* the annulus. Trying to force in a valve that is too large can cause disaster, such as coronary ostial impingement or tear of the annulus. We have now completely abandoned bioprostheses in the young age group because of their early degeneration. We have used a number of low-profile mechanical prostheses. The most commonly used is the St. Jude valve and, more recently, the Carbomedics valve for the small annulus. If too small, the aortic root may have to be enlarged (see aortic root enlargement section).

The prosthesis is inserted with either simple interrupted or mattress sutures (Tycron). They are first placed in the area of the left coronary sinus, starting at the commissure between the right and the left sinus, then in the right coronary sinus and finally in the non-coronary sinus. The sutures are placed through the remnant of the annulus. Care must be taken to avoid deep suturing in the area of the membranous septum beneath the junction between the non-coronary and right coronary cusps, to prevent injury to the conduction system. The sutures are placed through both annulus and prosthesis as the replacement proceeds (**Figures 37.11** and **37.12**). When all the sutures are placed, the valve is lowered into

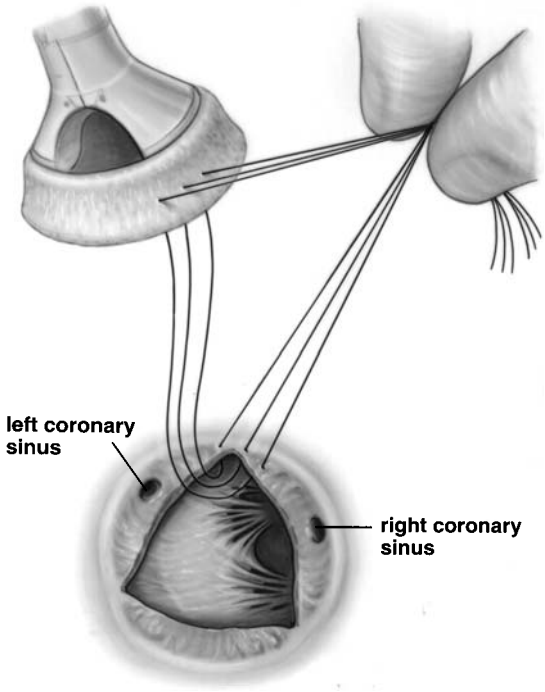

Figure 37.11

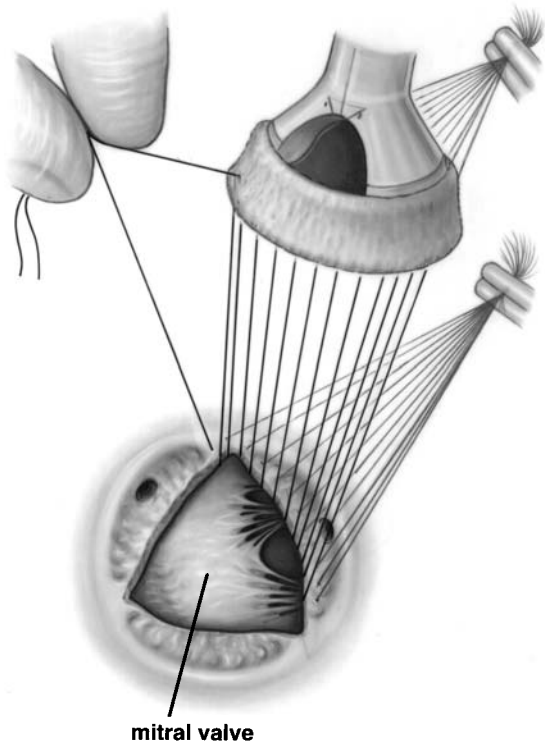

Figure 37.12

position and the sutures are tied. If mattress sutures are used, these may or may not be reinforced with Teflon pledgets. In children, because of the frequent small size of the annulus, the mattress sutures are placed from below the annulus and up through the prosthesis to place the annulus between the sutures and the prosthetic device using the maximum diameter of the aortic root.

Aortic Root Replacement

Homograft Aortic Root Replacement

Homograft aortic root replacement introduced by Ross (Somerville and Ross, 1982) has been performed to treat lesions of the ascending aorta, such as aneurysms and dissections, to treat complicated bacterial endocarditis with aortic root abscesses, and in the management of complex left ventricular outflow tract obstructions. In the latter group, the removal of the native stenotic and fibrotic aortic root opens up the left ventricular outflow tract and can often relieve subaortic gradients, with or without an additional septal myotomy or myectomy. Aortic root replacement has two main advantages over the free-hand homograft replacement. First, the valve is more likely to remain competent, as it is inserted as a functional unit. Second, a valve of greater diameter than the native valve can be inserted if implanted as a root (Stelzer *et al*., 1989).

The operation is performed using hypothermic cardiopulmonary bypass with a single atrial cannula for venous drainage and a high aortic cannulation. The left heart is vented. The pulmonary artery is separated from the aorta. The cold blood cardioplegic solution is infused every 20–30 min. The first infusion is into the aortic root if the aortic valve is competent.

A proximal transverse aortotomy is first carried out to examine the aortic valve, the left ventricular outflow tract and the coronary anatomy. If the decision is made to proceed with the aortic root replacement, the proximal transverse aortotomy is completed. At this stage, protection of the coronary arteries is of utmost importance. First, the back of the aorta is cut away from the left coronary orifice, leaving a disc of at least 3 or 4 mm of aortic wall as a cuff. The right coronary artery is also removed with a shield of aortic wall, and the rest of the aorta is now cut away freely, leaving some remnant of fibrous tissue anteriorly on the septum for later attachment of sutures. Any subaortic obstruction is now fully accessible and should be dealt with by enucleation or excision of fibrous tissue or muscle. Root enlargement

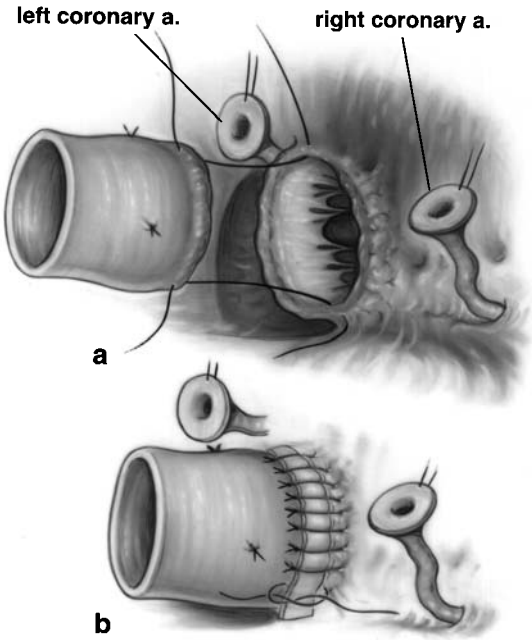

Figure 37.13

is performed, if necessary. The proximal coronary arteries are then fully mobilized, using sharp dissection. Care must be taken not to perforate the main pulmonary artery during this dissection. This is easily done during a reoperation. A full mobilization of the coronary arteries is important, as they have to be reimplanted slightly higher than their original position in the native root. The authors tend to leave the superior edge of the coronary cuffs straight (like a shield) for orientation at the time of reimplantation. Stay sutures are then placed on the aortic cuff surrounding the coronary arteries, so as to retract them outside the operating field (**Figure 37.13a**).

An aortic homograft of appropriate size is chosen. In children, it is always possible to use a larger homograft than the native aortic root. The larger size of the new aorta emphasizes the importance of fully mobilizing the coronary arteries, to avoid subsequent tension when they are reattached to the new aortic root. The lower margin of the homograft is sewn into the left ventricular outflow tract. We prefer, as Ross suggested, to have the valve orientated into the anatomical position so that the left coronary sinus of the homograft corresponds with the native left coronary artery. The circumference of the outflow tract is first divided into three by a set of commissural marking sutures, and subannular interrupted sutures are placed in each sinus in a sequential manner. With all the sutures in place, a strip of autologous pericardium can be positioned around the aortic root, and interrupted sutures are snugged down onto it (**Figure 37.13b**). The

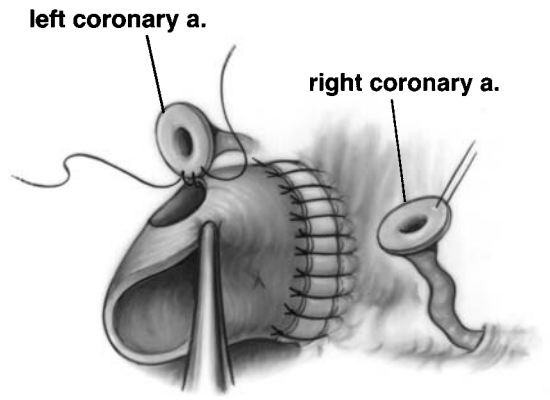

Figure 37.14

left coronary stump on the homograft is widely excised, and the button of the native aortic wall surrounding the left coronary artery is anastomosed to the homograft with a running 6–0 Prolene suture (**Figure 37.14**).

An alternative technique consists of rotating the homograft 120°, so that the right coronary artery ostium faces the recipient's left coronary artery (McGriffin and O'Brien, 1989). This has the advantage of avoiding sewing muscle to muscle and probably results in a more secure suture line, which can be done with interrupted sutures or with a running suture of 4–0 Prolene. The left coronary artery is then sewn into the enlarged ostium of the homograft's right coronary artery. The left coronary artery of the homograft is closed with a 5–0 Prolene suture. As a result of the rotation of the homograft, the aorta has a tendency to curve to the right. This can be rectified by appropriate bevelling of the graft.

Ross suggested reimplanting the right coronary artery after completion of the upper anastomoses of the aorta, which is then placed under tension by temporary release of the aortic clamp to facilitate the siting of the right coronary artery, the origin of which is highly variable (**Figure 37.15**). Care must be exercised not to damage the aortic cusp during this manoeuvre. A marking suture, placed before the aorta is closed, may be useful.

Pulmonary Autograft Root Replacement

The idea of transferring the freehand pulmonary valve to the aortic position, combined with right ventricular outflow tract reconstruction, was introduced by Ross in 1967. The pulmonary autograft root has one major advantage, as the neo-aortic valve is more likely to remain competent as it is inserted as a functional unit. However, the prerequisite for the success of this operation requires the utmost attention to the construction of the many suture lines. Contraindications for the Ross procedure

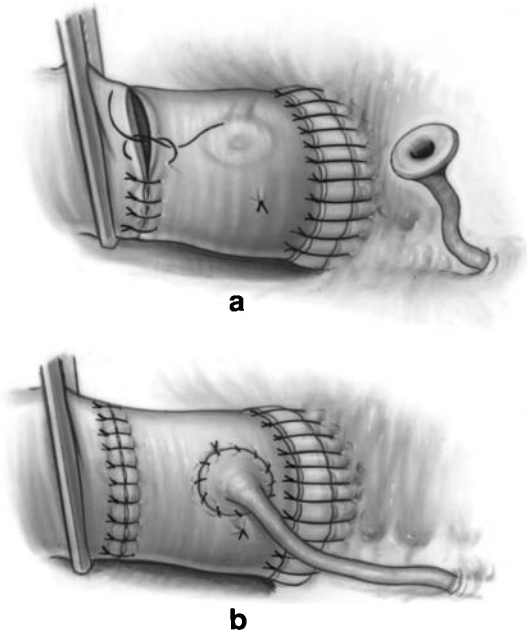

Figure 37.15

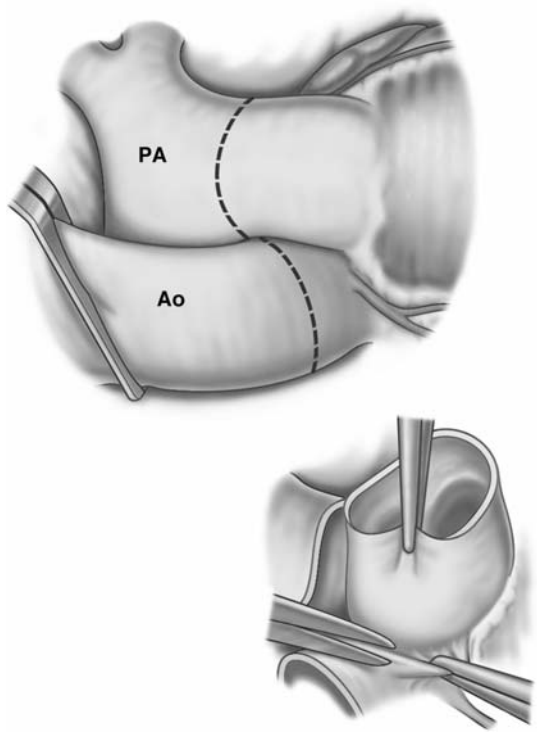

Figure 37.16

considered in our institution, are doubly-committed subarterial VSD, dysplastic pulmonary valve and significant coronary artery crossing the proximal RVOT. Any major discrepancy between the sizes of the two outlet valves is also a relative contraindication.

The operation is performed using hypothermic cardiopulmonary bypass with bicaval cannulation. Before the cannulation, the ascending aorta and the pulmonary artery and its branches are extensively mobilized. The left heart is vented via the right pulmonary vein and, later on, through the transected pulmonary artery. If the decision to replace the aortic valve with a pulmonary autograft has been made, and if the aortic valve is competent, the pulmonary valve can be harvested before the aorta is clamped. Otherwise, the aortic clamp is applied and a transverse aortotomy is made. The cold blood cardioplegic solution is administered every 20–30 minutes. The first infusion can be made directly in the aortic root if the aortic valve is competent. The preparation and dissection of the left ventricular outflow tract and the dissection of the coronary arteries are the same as for the homograft root replacement.

Harvesting of the Autograft

The main pulmonary artery is transected just proximal to the bifurcation. The pulmonary valve is inspected to make sure that it is normal. The base of the pulmonary valve is then separated from the root of the aorta by sharp dissection (**Figure 37.16**). A curved clamp is then passed retrogradely through the pulmonary valve, as a guide to locate the incision on the anterior aspect of the right ventricular outflow tract 3 or 4 mm below the lowest point of leaflet attachment (**Figure 37.17**). The incision is carried out medially, taking the full thickness of the anterior wall of the right ventricle. One should try to avoid the conus branch of the right coronary artery as the incision is carried toward the aorta. Posteriorly and to the left lie the left main coronary artery, the left anterior descending coronary artery and the first septal branch, which are all at risk during this dissection (**inset, Figure 37.17**). The septal branch is avoided by making an oblique incision deeper on the endocardial than the epicardial side. The endocardial incision is first made with fine scissors from within the right ventricular outflow tract. The enucleation of the pulmonary valve is completed with scissors angled to leave the epicardium intact (**Figure 37.18**). Any bleeding points in this dissected RVOT area, shown up at the time of cardioplegia delivery into the left coronary artery, should be diathermized or carefully oversewn with fine Prolene sutures.

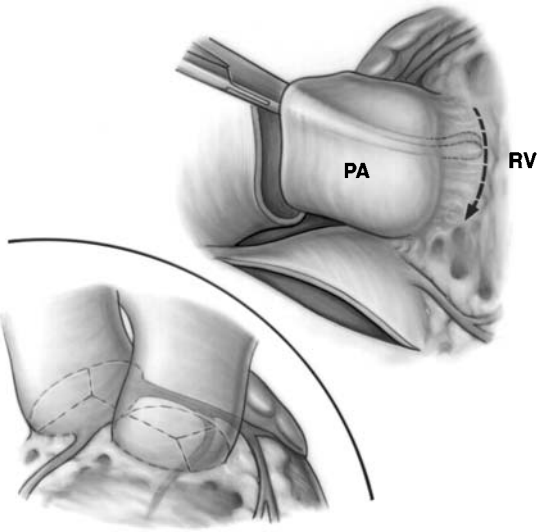

Figure 37.17

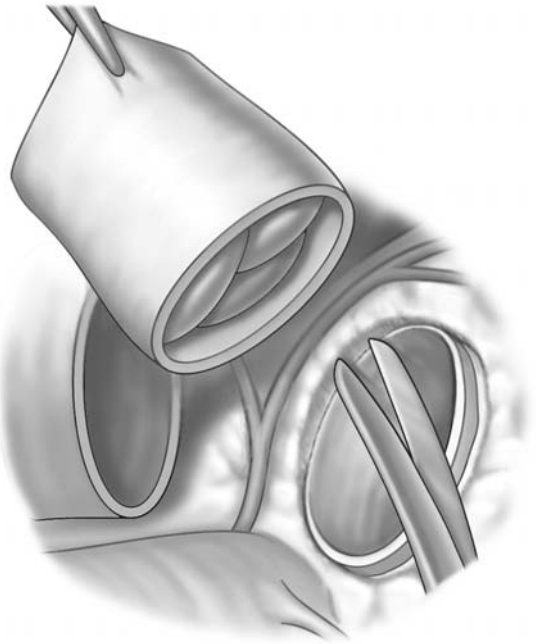

Figure 37.18

The aortic valve is excised, and the pulmonary autograft root is inserted into the subannular aortic position, using the technique described for the insertion of an aortic root homograft. The autograft is trimmed

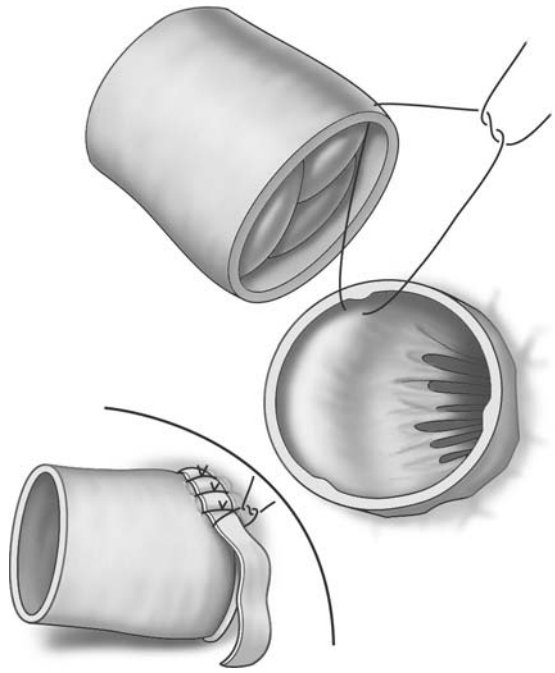

Figure 37.19

of all excessive muscle up to within 2 mm of the cusp insertions, and any loose tissue is removed from the pulmonary arterial wall. The proximal suture line can be constructed in a variety of ways, including continuous sutures, single interrupted sutures or mattress sutures, with or without reinforcement. We prefer the use of interrupted 4-0 Tycron sutures reinforced with a strip of autologous pericardium for the proximal suture line of the autograft into the left ventricular outflow tract. The suture line is subannular (**Figure 37.19**), so that the autograft root is well supported by the fibromuscular LVOT. The orientation of the autograft follows the slight curve, and the left and right coronary arteries face a sinus (**Figure 37.20**). The coronary arteries are implanted into the autograft, avoiding rotation, tension and kinking (**Figure 37.21**).

The right ventricular outflow tract is reconstructed with a pulmonary or an aortic homograft of appropriate size. The distal suture is carried out first, placing the sinuses of the homograft into their appropriate positions (two sinuses facing the aorta) (**Figure 37.22**). The distal aortic suture line is completed without tension. The authors would also reduce the dimension of the dilated distal ascending aorta, if present, to protect the newly created sinotubular junction. This can be easily achieved by either a triangular anterior aortic wall resection

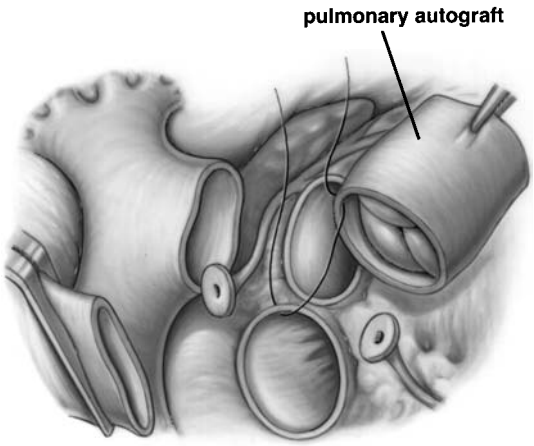

Figure 37.20

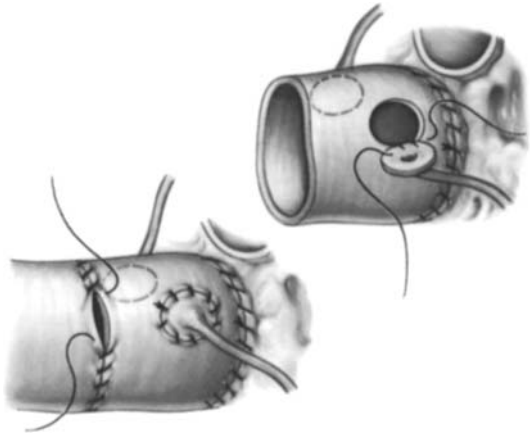

Figure 37.21

(see **inset, Figure 37.22**.) or the placement of several plicating horizontal prolene sutures.

The proximal RVOT homograft suture line is carried out before the aorta is unclamped (**Figure 37.23**). Haemostasis and avoidance of damaging the left coronary artery system are particularly crucial during this anastomosis, which has to be performed on a relaxed heart and in a bloodless field. Anteriorly, the suture line takes the full thickness of the right ventricular outflow tract. The completed operation is shown in **Figure 37.24**.

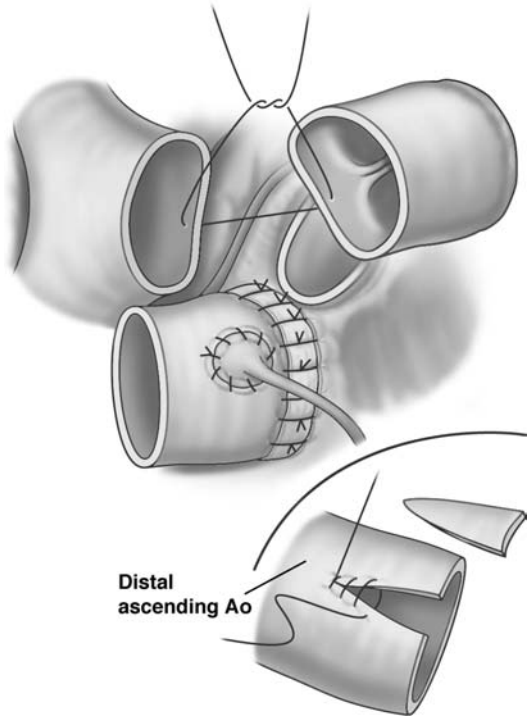

Figure 37.22

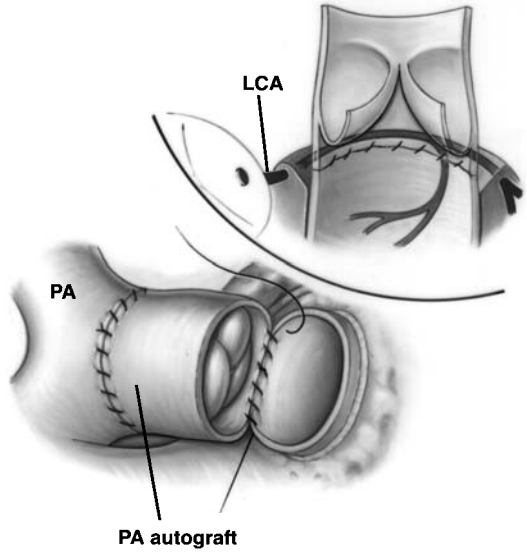

Figure 37.23

Relief of Subvalvar Aortic Stenosis

Resection of Subaortic Shelf

This operation is performed on hypothermic cardiopulmonary bypass with bicaval cannulation or a single atrial cannula for the venous return. The cold blood cardioplegic solution is infused into the aortic root if the aortic valve is competent, or directly into the coronary arteries if there is more than mild aortic valve

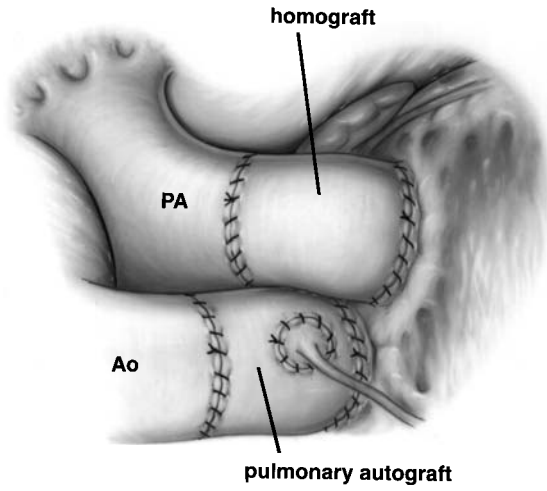

Figure 37.24

regurgitation. An oblique aortotomy incision extending into the non-coronary sinus is performed. The aortic valve is inspected. The right coronary cusp is retracted anteriorly, exposing the subvalvar obstruction. A modified nasal speculum (Alfieri and Subramanian, 1975) is sometimes useful to retract the aortic cusps and expose the subvalvar area in small children. The fibrous shelf is grasped with forceps and a vertical incision is made into the shelf to the left of the right coronary cusp. A combination of sharp and blunt dissection is used to enucleate the circumferential shelf off the ventricular septum and the anterior leaflet of the mitral valve (**Figure 37.25**). McKay and Ross (1982) described a technique of blunt dissection, using a Watson–Cheyne dissector to peel off the fibrous tissue from the subaortic area. Limiting the dissection to the fibrous tissue in the area of the membranous septum is particularly important, to avoid the conduction system. When the fibrous shelf is attached to the undersurface of one or more of the aortic cusps, it is carefully shaved away from the cusp tissue with a knife. Any fibrous tethering of the left and right fibrous trigones in the subaortic region, including the anterior mitral leaflet, should be carefully released to widen the left ventricular outflow tract (Yacoub *et al*., 1999). Once the fibrous component of the obstruction has been enucleated, we routinely perform septal myotomy/myectomy because the ventricular septum is nearly always significantly hypertrophied. The technique is similar to that utilized for diffuse fibromuscular stenosis or obstructive hypertrophic cardiomyopathy (**Figure 37.26**), although the resection may be less extensive. In a narrow aortic root, myectomy is sometimes not feasible; myotomy with a scimitar blade may be sufficient.

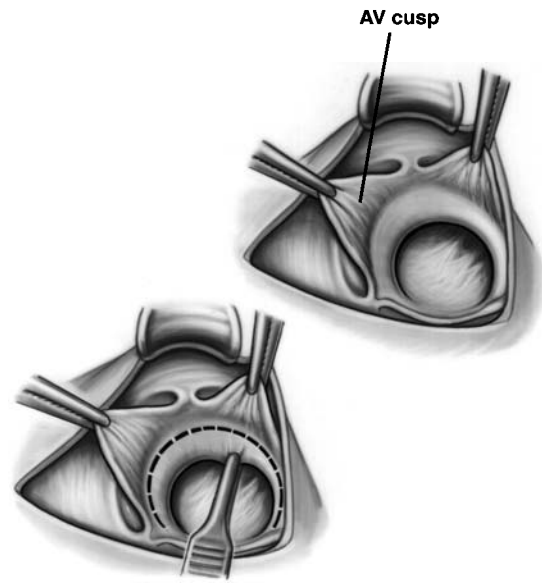

Figure 37.25

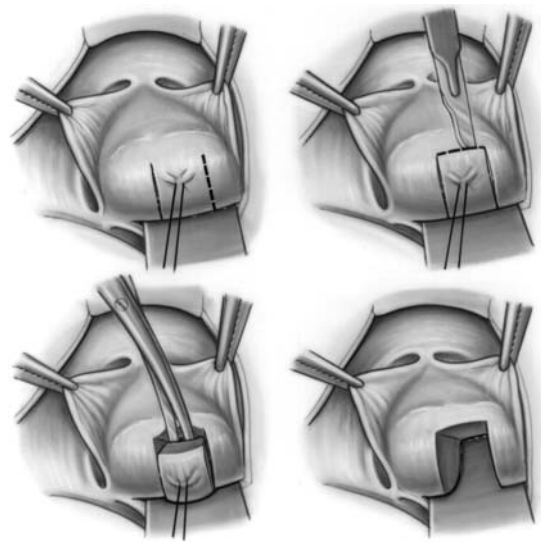

Figure 37.26

Fibromuscular Stenosis

Good myocardial protection is paramount in the setting of a very hypertrophied heart. The aortic valve is gently retracted, and the area of the membraneous septum must be avoided. Venting of the left ventricle provides

good visual approach for extended septal myectomy, as the risk of creating an iatrogenic VSD is not insignificant.

For a limited wedge excision of the fibromuscular ring, two parallel incisions are made into the ventricular septum. The first incision is made beneath the commissure between the left and the right coronary cusps, immediately to the right of the mitral apparatus, and the second incision is made beneath the belly of the right coronary cusp. Both incisions are deepened and carried inferiorly as far as needed. The two incisions are then connected by a transverse incision beneath the right coronary cusp, and a chunk of ventricular septum is excised with scissors (**Figure 37.26**).

For HOCM, the extent of the LV septal myectomy would be guided by the trans-oesophageal findings. The aim is to abolish the dynamic obstruction due to systolic anterior motion of the anterior mitral leaflet, and to create a wide open LVOT. The left and right fibrous trigones should be unhinged as described above, and any fibromuscular accessory mitral attachments to the septum should be divided (Dearani and Danielson, 2005). Any echocardiographic evidence of residual narrow LVOT or systolic anterior motion of the anterior mitral leaflet would warrant further septal resection. Discontinuation of cardiopulmonary bypass requires optimal preload, atrio-ventricular synchrony; inotropes are not indicated.

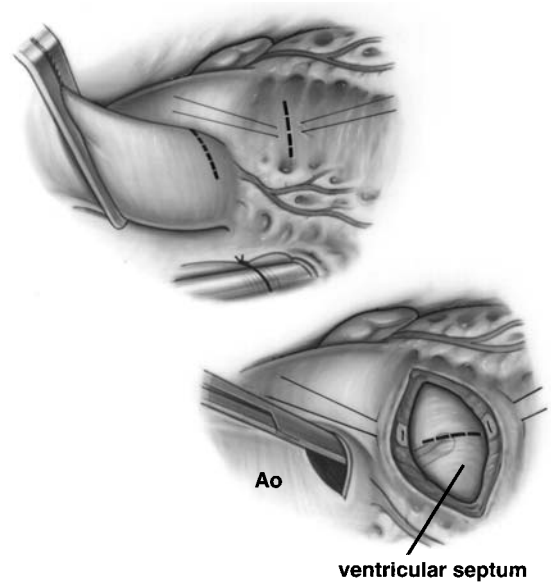

Figure 37.27

''Tunnel-like'' Stenosis with Normal Aortic Valve

If the severity of the subvalvar obstruction is such that the previously described technique is not sufficient to relieve it, a modification of the Konno operation, leaving the aortic valve annulus intact (partial Konno) may be indicated. Aortotomy incision, pointing toward the commissure between the right and the left aortic cusps, is carried out. The aortic valve and the subvalvar area are inspected. If the stenosis cannot be resected through the aortic valve, a short transverse incision is made in the right ventricular outflow tract below the pulmonary valve. A right-angled clamp is passed through the aortic valve into the left ventricular outflow tract pointing toward the interventricular septum, in which an incision is made through the right ventriculotomy incision. The incision should remain above the muscle of Lancisi in order to avoid the conduction system. Superiorly, the incision is carried to within a few mm of the aortic valve annulus (**Figure 37.27**). The fibromuscular components of the subvalvar stenosis are excised. A patch of Dacron or Gore-Tex is then inserted to widen the left ventricular outflow tract (**Figure 37.28**). The right ventriculotomy can be closed by direct suture.

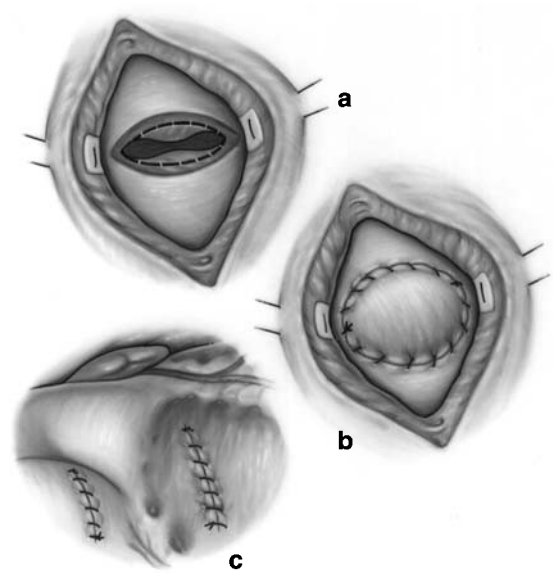

Figure 37.28

Small Aortic Roots and Diffuse Left Ventricular Outflow Tract Obstructions

Hypoplasia of the aortic valve annulus, diffuse obstruction of the left ventricular outflow tract, and the lack of potential for growth after prosthetic valve insertion have

led to the design of a number of procedures aiming to enlarge the aortic root.

Annular Enlargement towards the Mitral Valve

Two posterior techniques have been described. The first consists of extending the aortotomy incision through the midpoint of the non-coronary sinus across the annulus, into the anterior leaflet of the mitral valve (Nicks *et al*., 1970). The original description involved an incision made in the roof of the left atrium opposite to the aortotomy incision. A composite teardrop-shaped patch of pericardium internally lining a knitted Dacron patch was sutured onto the mitral valve and then to the aortotomy incision. We prefer to maintain the integrity of the mitral valve leaflet. One should also keep in mind the possibility of an abnormal circumflex coronary artery arising from the right coronary artery and coursing posteriorly, or a sinus node artery arising from the circumflex coronary artery. Therefore the incision into the aortomitral curtain is stopped well short of the mitral fibrous annulus (**Figure 37.29**) and a patch of bovine pericardium is used. The enlarged aortic annulus is calibrated, and a valve substitute is selected. Pledgeted horizontal mattress sutures are first passed through the patch, and finally through the sewing ring of the valve. The rest of the prosthetic valve is then sewn in, in the usual fashion, to the aortic ring of the left and right coronary sinuses (**Figure 37.30**). The proximal aorta is concomitantly enlarged using the teardrop-shaped patch.

Another technique of posterior annular enlargement was described by Manouguian and Seybold-Epting (1979). The aortotomy incision is directed toward the tip of the commissure between the left and the non-coronary sinuses (**Figure 37.31**). The left atrium is dissected away, and the incision is carried through the area of fibrous continuity between the aortic and the mitral valves. It is often possible to stop the incision immediately above the hinge of the anterior leaflet of the mitral valve without opening the left atrium. A teardrop-shaped bovine pericardial patch is then inserted with a continuous 4-0 Prolene suture. Once the enlarged aortic root is prepared, the insertion of an aortic valve substitute should follow the technique described earlier. Some surgeons prefer to use the mitral leaflet of the aortic homograft into the patient's anterior leaflet of the mitral valve.

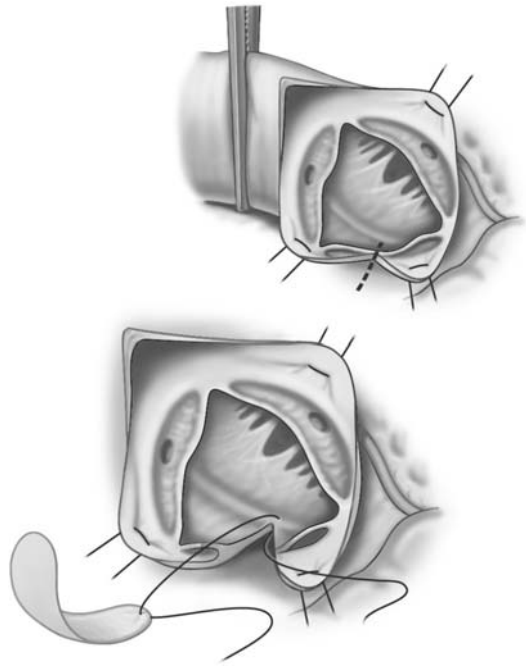

Figure 37.29

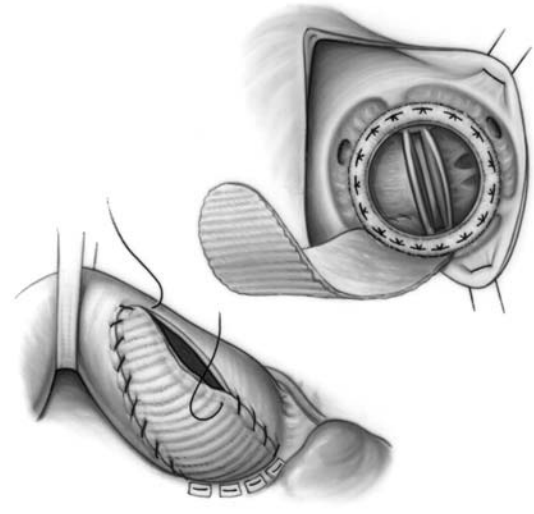

Figure 37.30

Aortoventriculoplasty (Konno Operation)

This approach was developed simultaneously in the late 1970s by Konno and Rastan to treat tunnel-type subaortic stenoses and hypoplasia of the aortic valve annulus (Rastan and Koncz, 1976; Konno *et al*., 1975).

The position of the right coronary artery is accurately noted, and a longitudinal aortotomy incision is made down to the aortic annulus well to the left of the right coronary artery toward the commissure between the right and the left coronary arteries. A transverse incision is

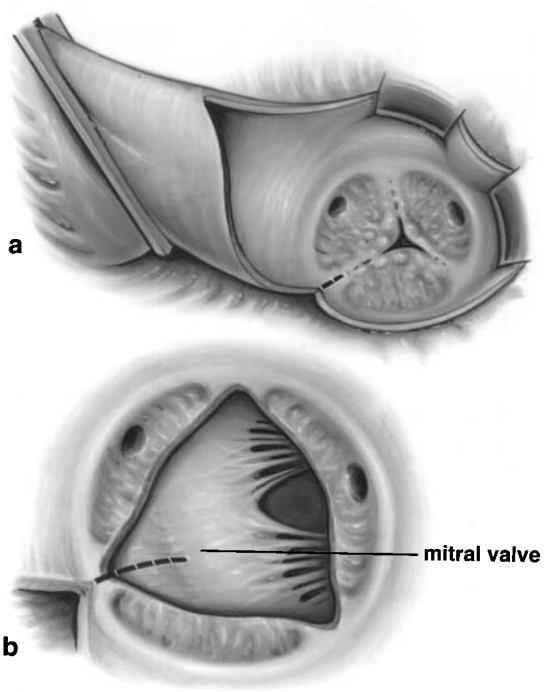

Figure 37.31

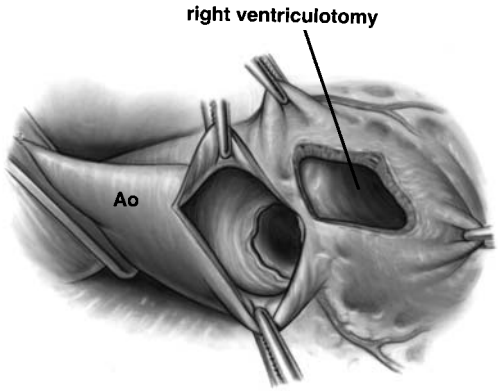

Figure 37.32

made in the outflow tract of the right ventricle, just proximal to the pulmonary annulus (**Figure 37.32**). Care must be taken to avoid damaging the pulmonary valve cusps, which lie very near the point of entry of the incision into the right ventricle. Scissors are then positioned, with one blade into the left ventricle through the aortotomy, and one in the right ventricle through the ventriculotomy, so as to join the incision between the right ventricle and the aorta, opening the interventricular septum. This opening must be extended far enough below the tunnel stenosis, staying above the muscle of Lancisi to avoid the conduction system.

The aortic valve is completely removed, and a valve sizer is then laid in the area of the divided annulus to estimate the amount of prosthetic patch required to enlarge the aortic orifice and to accommodate a predetermined valve size. An oblong-shaped Dacron or Gore-Tex patch is then trimmed and sutured to the ventricular septum by horizontal mattress sutures buttressed with a strip of Teflon felt on the right ventricular side of the septum (**Figure 37.33**). The suturing of the patch is interrupted just beyond the aortic annulus, and the appropriately sized aortic valve prosthesis is then sutured in place in the standard manner, with simple interrupted sutures or mattress sutures through the aortic annulus and the valve ring (**Figure 37.34**). The part of the valve covered by Dacron is attached with horizontal mattress sutures in the valve ring, which are then brought through the Dacron and tied on the right ventricular side of the patch. At this time, each of the two previously placed sutures at the aortic annulus are tied and then run onto the aorta so as to enlarge and close the aortotomy incision (**Figure 37.35**).

After completion of the suture lines, routine de-airing is done and the aortic clamp released. Any leakages in the patch can be repaired. The defect in the right ventricular outflow tract is then closed with a large patch of autologous or bovine pericardium (**Figure 37.36**), which is brought over the entire Dacron patch so that

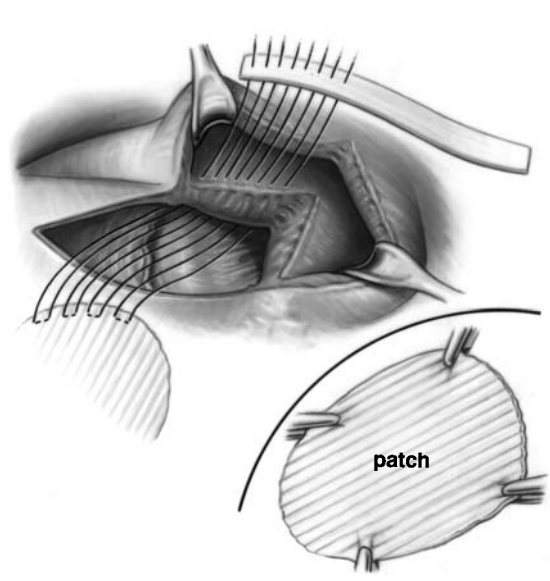

Figure 37.33

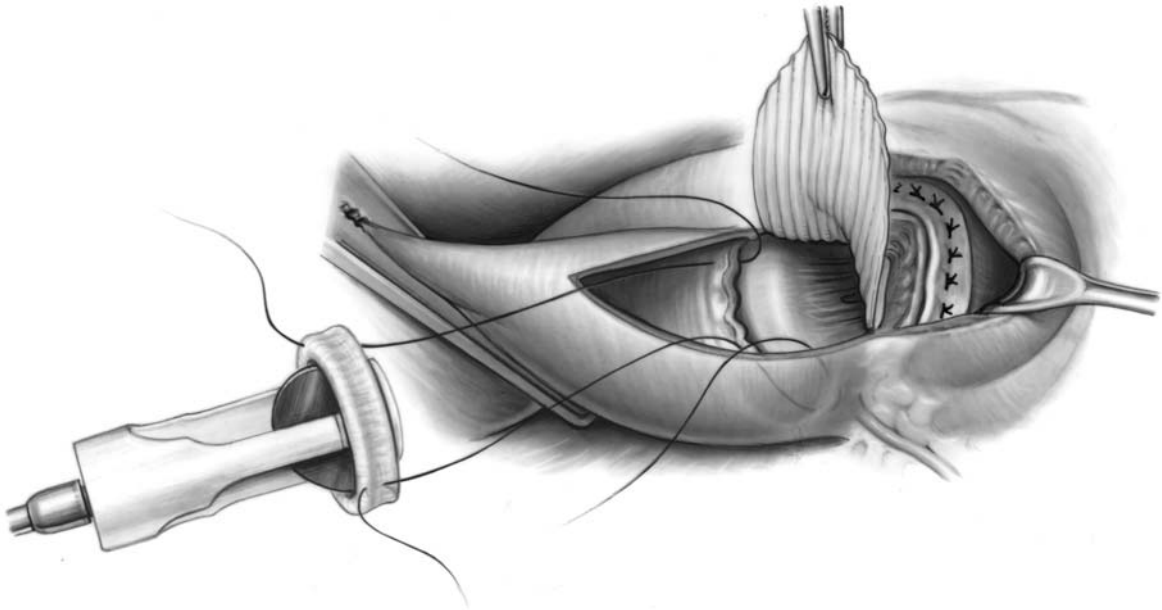

Figure 37.34

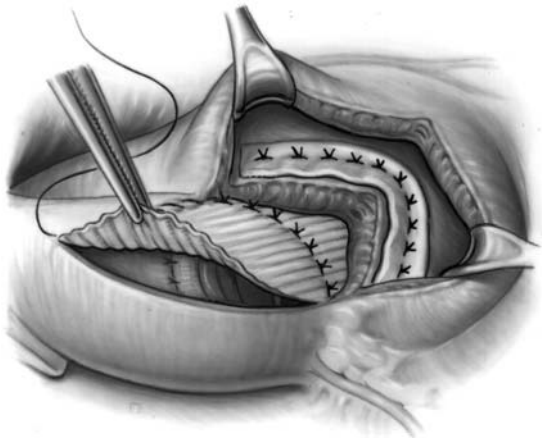

Figure 37.35

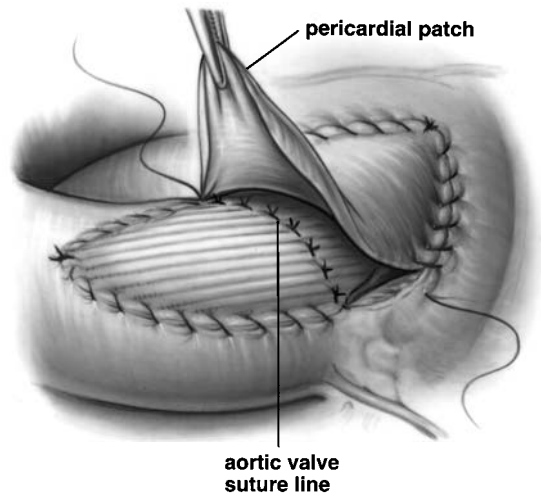

Figure 37.36

the prosthetic patch is totally within the right ventricular cavity. Any leakage though the Dacron patch would thus be into the right ventricle. This greatly reduces postoperative bleeding (**Figure 37.37**).

Extended Aortic Root Replacement with Aortic Homograft

As an alternative to the Konno operation, Clarke (1987) applied the concept of aortic root replacement combined with the concept of aortoventriculoplasty to describe the extended aortic root replacement, which has the advantage of not using a prosthetic valve. The aortic and the cardiac incisions are similar to those used in aortoventriculoplasty: a vertical aortotomy and an oblique incision across the right ventricular outflow tract. These two incisions come together at the aortic annulus between the left and the right

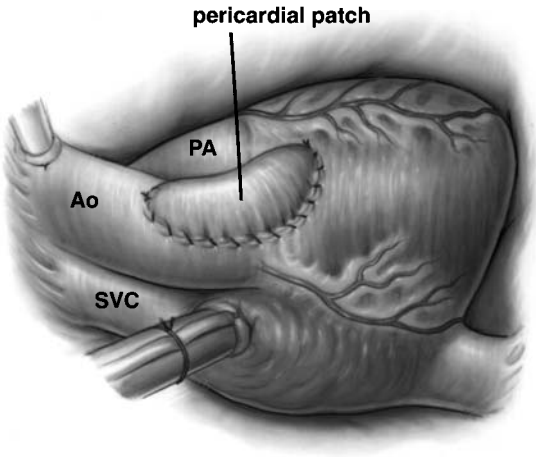

Figure 37.37

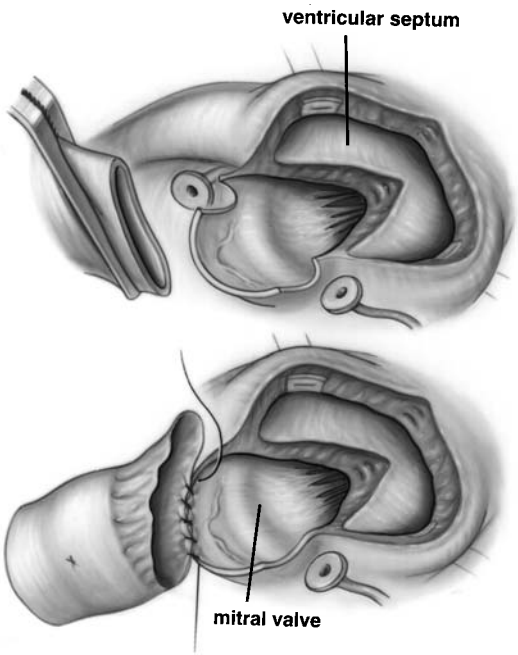

Figure 37.38

coronary ostia. In this area, the incision is carried across the aortic annulus and down into the infundibular septum. The ascending aorta is transected, and the coronary arteries are excised with a cuff of aortic wall and mobilized as for an aortic root replacement (**Figure 37.38**).

Aortic homografts of at least 18 mm internal diameter can be used even in early childhood. Excess muscle is trimmed from the proximal portion of the homograft. The anterior leaflet of the mitral valve is left attached. The homograft is then orientated so as to have the mitral valve covering the incision in the infundibular septum. The proximal anastomosis of the homograft to the posterior aspect of the outflow tract is then carried out with a running suture of 4-0 Prolene (**Figure 37.38**). This is interrupted and secured on either side of the aortotomy incision. The anterior leaflet of the mitral valve is then trimmed so as to fill exactly the septal incision, onto which the leaflet is sewn with several interrupted horizontal mattress sutures, reinforced with Teflon pledgets.

Next, the coronary arteries are anastomosed into the two defects created in the homograft (**Figure 37.39**). The left coronary artery may be just opposite to the origin of the right coronary artery of the homograft, in which case this ostium is enlarged. A hole has to be made in the anterior aspect of the homograft for the right coronary artery. Care must be taken not to damage the aortic valve leaflet during that manoeuvre.

Next, the distal anastomosis of the homograft to the host's ascending aorta is carried out with a running suture of 4-0 Prolene (**Figure 37.39**). At this point, the aortic clamp is released. The left side of the heart is de-aired, and the patient is rewarmed while the incision in the right ventricular outflow tract is closed. This is done with a

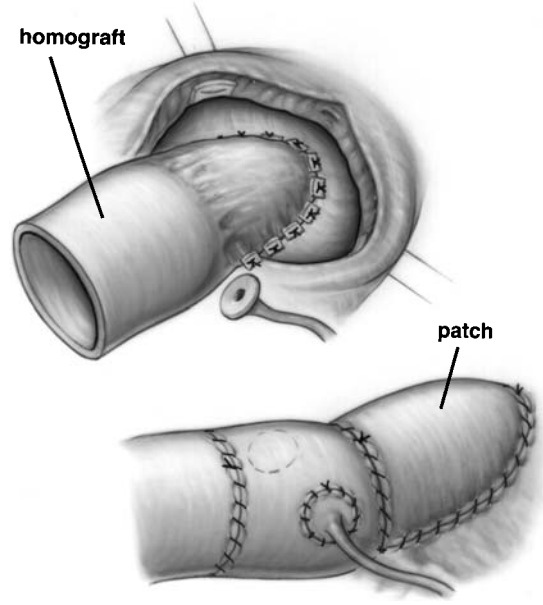

Figure 37.39

triangular patch of pericardium or with a piece of aortic homograft. The patch is sutured to the margin of the right ventriculotomy incision and along the annulus of the aortic valve of the homograft. Particular care is taken to anchor the patch securely at the junction of the right ventricle and the homograft.

Combined Ross/Modified Konno Procedure

This is our preferred option for infants/children with aortic valve disease complicated by complex LVOTO. The operative procedure is similar to what has been described under the extended aortic root replacement with an aortic root homograft. However, the removal of the native fibrotic aortic valve and stenotic aortic root often opens up the LVOT and can effectively relieve subaortic obstruction without an additional annular enlargement procedure. In some cases, an incision into the aortic annulus along the commissure between the right and left coronary cusps (similar to the direction of the classical Konno septal incision) may be needed. This is followed by a LV myotomy incision, to release the restrictiveness of the bulky muscular septum without opening up the interventricular septum. Our personal experience with this approach, to create a widely opened LVOT without a patch to the interventricular septum, is good. This technique should cause less distortion to the integrity of the right ventricle, taking into consideration that the pulmonary valve would be harvested as an autograft. The latter can be prepared for a standard Ross procedure without an excessive length of anterior infundibular septum. The autograft is inserted with an interrupted 4-0 Tycron proximal suture line in the subannular position, reinforced with a strip of autologous pericardium. Coronary transfer has already been described. The distal autograft is reconnected with the distal ascending aorta, with or without a reduction aortoplasty. For the ease of completing the surgery, a slightly larger sized pulmonary homograft can be used to re-establish RV-to-PA continuity.

Apical–Aortic Bypass

The use of an extracardiac valved conduit between the left ventricular apex and the aorta was reported (McGoon, 1976; Cooley *et al*., 1976) for diffuse left ventricular outflow tract obstructions. The conduit graft can be implanted from the apex of the left ventricle to the ascending, descending thoracic or abdominal aorta. These operations are now very rarely performed, as preference is given to the more logical approach of directly relieving the obstruction of the native outflow tract. On the other hand, the surgical approach described below is similar to what has been used for the apical placement of LVAD, which some readers may find useful.

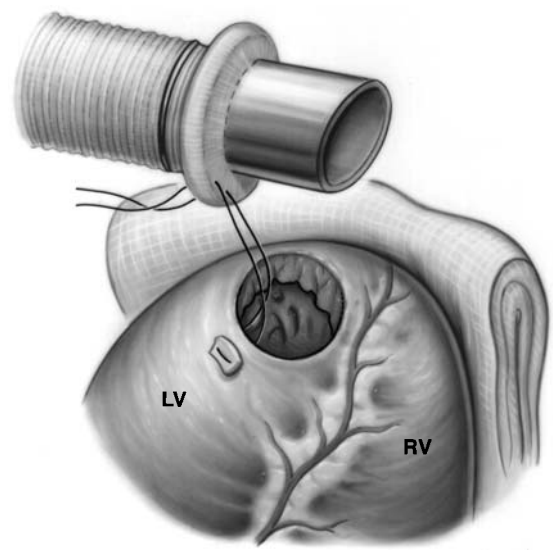

Figure 37.40

The operation is performed on cardiopulmonary bypass. An incision is made at the apex of the left ventricle, parallel and to the left of the left anterior descending coronary artery. Care must be taken not to damage the papillary muscle of the mitral valve. To overcome constriction or even obliteration of the graft during peak systole, the use of a rigid inlet tube has been recommended (**Figure 37.40**). The Dacron conduit containing a heterograft is then appropriately trimmed. If it is anastomosed to the ascending aorta, the left lateral portion of the pericardial sac is incised longitudinally to allow the conduit to bulge into the left pleural space. The superior anastomosis is then made on the left lateral aspect of the ascending aorta (**Figure 37.41**).

The median sternotomy incision can also be extended into the linea alba to a point just above the umbilicus. The liver is mobilized toward the right by dividing the triangular ligament, and the abdominal aorta proximal to the coeliac axis is exposed. The aorta is partially occluded with a side-biting clamp. A Dacron graft containing a valve is sutured to the aorta with a continuous suture. The ventricle with its graft attached is replaced into the pericardium. A small aperture is made in the left hemidiaphragm at a point closest to the left ventricular apex, the graft is drawn into the abdomen, and an end-to-end anastomosis is constructed between the two conduits. Air is evacuated from the composite conduit proximal and distal to the valve, from the left ventricular cavity,

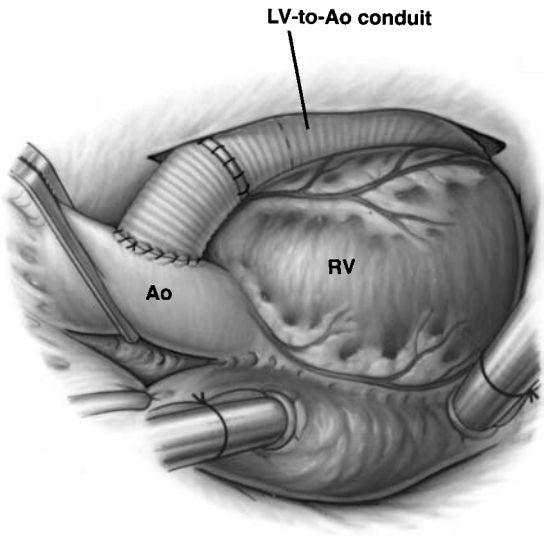

Figure 37.41

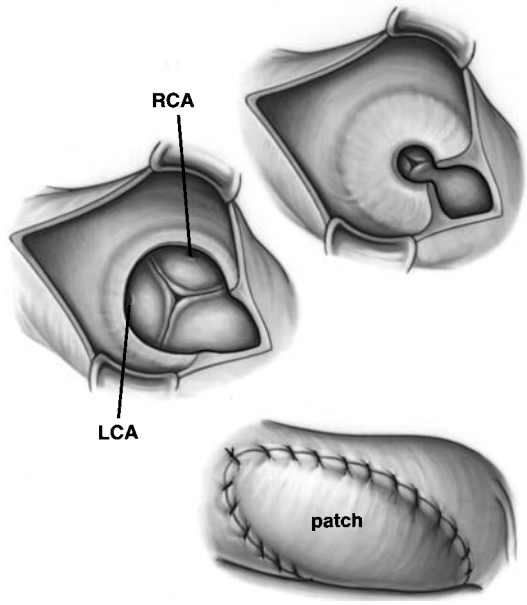

Figure 37.42

and from the ascending aorta before release of the aortic cross-clamp and restoration of cardiac activity.

Relief of Supravalvar Aortic Stenosis

The surgical relief of *localized* stenosis is classically done by partial resection of the stricture. The incision is in the aortic root across the stenotic region and down into the non-coronary sinus of Valsalva. The narrowing is enlarged by a patch. An oblong-shaped patch of pericardium is incorporated into the incision with a running suture of polypropylene (**Figure 37.42**). A more extensive aortoplasty has been recommended by Doty *et al*. (1977). An incision is made in the ascending aorta across the fibrosing ring into the non-coronary sinus. This incision is then extended as an inverted Y through the ring into the right coronary sinus, between the origin of the right coronary artery and the commissure between the right and the left coronary cusps. Half of a tubular Dacron prosthesis, about the same diameter as the ascending aorta, is trimmed so as to take a wedge from its lower end and to accommodate the area of the aorta containing the right coronary artery. This graft is then used to widen the ascending aorta. The tips of the graft are placed into the apex of each incision, near the aortic annulus (**Figure 37.43**). Currently, we prefer to use bovine pericardium for this procedure.

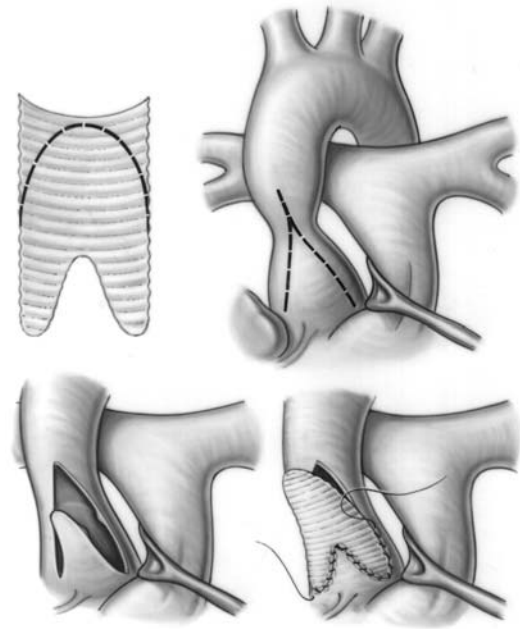

Figure 37.43

Brom (1988) has advocated a more extensive repair, consisting of a transection of the ascending aorta with complete excision of the fibrous ridge and a patch enlargement of all the three sinuses with three separate pericardial patches (**Figure 37.44**). This approach aims to restore the whole aortic root and deals with the often crowded left coronary sinus related to the fibrotic shelf. This in itself can interfere with the orifice of the left

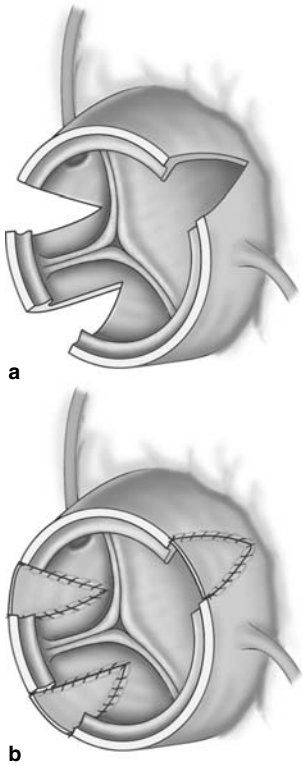

Figure 37.44

coronary artery. Depending on the extent of coronary artery involvement, additional procedures, such as ostial plasty, freeing the aortic valve cusp from the shelf or possibly coronary bypass grafting, may be required.

In a *diffuse* supravalvar aortic stenosis extending up to the head vessels, extensive reconstruction of the aorta is necessary. The operation is performed under deep hypothermia and circulatory arrest. The head vessels are dissected and snared or clamped during the period of deep hypothermia and total circulatory arrest. A long incision is made in the ascending aorta, well down into the non-coronary sinus of Valsalva and well up into the aortic arch, the transverse portion and the upper descending thoracic aorta if necessary. A patch of homograft, bovine pericardium, or Gore-Tex is tailored to appropriate size and shape and sewn with a running suture of polypropylene.

RESULTS

Aortic Valvar Stenosis in Neonates

Since its first use in 1986, primary balloon aortic valvotomy for neonatal aortic stenosis has replaced surgical valvotomy (open or closed) at many centres. McElhinney *et al*. (2005), in a series of 113 neonatal balloon aortic valvotomies, reported 14% early mortality and 15% incidence of significant aortic regurgitation. They identified young age, pre- and post-balloon aortic valvotomy gradient and large balloon: annulus ratio as significant factors predicting future reintervention.

Neonatal surgical aortic valvotomy has been reported to have mortality in the range 10–40% (Vida *et al*., 2005; Brown *et al*., 2003). Bhabra and colleagues (2003) have suggested that surgical aortic valvotomy has better results when trileaflet anatomy can be achieved at valvotomy. Reintervention is common following either procedure (25–40% at 5 years; McCrindle *et al*., 2001; Brown *et al*., 2003).

Aortic Valve Repair

Some published reports include aortic valvotomy and commissurotomy as aortic valve repair procedures. This at times complicates the evaluation of results of aortic valve repair in general.

Hasaniya *et al*. (2004) reported 17.6% and 35.2% incidence of mild and moderate aortic regurgitation, respectively, with 47.1% incidence of reoperation on aortic valve during follow-up of 5.3 ± 2.4 years. Tweddell and colleagues (2005) reported comparable results of complex aortic valve repair to aortic valve replacement, with freedom from reintervention at 1, 5 and 10 years of $94 \pm 3\%$, $85 \pm 6\%$ and $44 \pm 5\%$ compared to $96 \pm 3\%$, $77 \pm 9\%$ and $77 \pm 9\%$, respectively, for aortic valve replacement. Aortic valve repair has been carried out for aortic regurgitation associated with bicuspid aortic valve. Fraser and Mee reported freedom from aortic valve reoperation of 94% and 89% at 12 and 24 months, respectively (Fraser *et al*., 1994).

Aortic Valve Stenosis in Children

Heterograft and Homograft Aortic Valve Replacement

Use of heterograft/xenograft for aortic valve replacement in children has been discarded following reports of early degeneration, calcification and structural failure. Homograft replacement of the aortic valve can be achieved with low morbidity and mortality. However, it suffers from the same problems as heterograft/xenograft, but over a longer period of time (Turrentine *et al*., 2001). O'Brien *et al*. (2001) reported 47% freedom from reoperation at 10 years in young patients receiving homograft for aortic valve replacement.

Mechanical Aortic Valve Replacement

In children this carries low morbidity and mortality with reasonable long-term outcome (Turrentine *et al*.,

2001). Alexiou *et al*. (2000) reported 5.3% mortality with survival at 10 and 20 years of 91% and 84.9%, respectively. Freedom from thromboembolism was 93%, freedom from valve-related events 86.6% and freedom from reoperation at 20 years 86.4%.

Pulmonary Autograft

Pulmonary autograft has been performed with low morbidity and low mortality by many centres. Hraska and colleagues (2004) reported mortality approaching zero, freedom from reoperation on autograft of 93% at 5 years and freedom from homograft replacement of the right ventricular outflow tract of 90% at 10 years.

Unfortunately, autograft root dilatation is common late after implantation. This may affect nearly one-third of patients receiving pulmonary autograft. Freedom from reoperation on autograft of 92–95% at 5 years and 88–91% at 7 years has been reported in children (David *et al*., 2000; Luciani *et al*., 2003). Younger age at operation, bicuspid aortic valve, geometric mismatch between aortic and pulmonary roots and the technique of autograft insertion have been suggested as important factors. Homograft dysfunction due to calcification and degeneration is reported in 6–20% of patients at 10 years. Factors leading to homograft dysfunction are the use of aortic homograft, use of a small-sized graft, age less than 10 years, homograft storage times and possible immune reaction to the homograft.

During 1994–2005, 126 patients underwent Ross or Ross/Konno procedure at Great Ormond Street. Median age was 14 years (5 days to 43 years). 80% of patients had undergone previous interventions, such as balloon aortic valvotomy, surgical valvotomy, aortic valve repair, aortic valve replacement, resection of subaortic stenosis, Konno procedure and repair for other congenital heart defects; 27 (21.4%) patients had undergone previously two or more procedures on left ventricular outflow tract; nine (7.14%) patients had undergone previous aortic valve replacement; nine (7.14%) patients required Konno or modified Konno procedure with pulmonary autograft for complex left ventricular outflow tract obstruction. There were four (3%) early deaths and no late deaths. Three of four early deaths were in our early experience. At median follow-up of 28 months, there were 11 (8.7%) reinterventions; seven (5.5%) were autograft-related—aortic root dilatation in one patient, infective endocarditis in five patients and cusp perforation in one patient. There were two homograft-related reinterventions (2%).

Subaortic Stenosis

Early results of relief of discrete subvalvar aortic stenosis are generally satisfactory, with a low operative mortality. However, there is significant late morbidity related to residual/recurrent left ventricular outflow tract obstruction. Recurrence rates of 7–27% and reoperation rates of 4–35% have been reported (Rohlicek *et al*., 1999; Serraf *et al*., 1999).

Diffuse subaortic stenosis or tunnel-like subaortic obstruction is uncommon. Jahangiri and colleagues (2000) have suggested that the majority are secondary to other congenital heart defects. A recurrence rate of 14% at 5 years has been reported.

Supravalvar Aortic Stenosis

Early mortality of 1–10% has been reported (Stamm *et al*., 1999; Hazekamp *et al*., 1999). Long-term outcome depends on the status of aortic valve, subaortic region and involvement of coronary arteries. Recurrence rates of up to 25% have been reported, with freedom from re-operation of 86% at 10 years and 66% at 20 years. Hazekamp *et al*. have reported no difference with type of surgical technique but showed that three-sinus reconstruction achieves better symmetry and restores normal aortic root anatomy.

Rheumatic Aortic Valve Disease

Aortic valve repair has been attempted for rheumatic aortic regurgitation with reasonable success. In patients undergoing aortic valve repair for rheumatic aortic regurgitation, Talwar *et al*. (2005) reported survival of $95.2 \pm 2.8\%$ and freedom from reoperation of $85.4 \pm 6.7\%$ at follow-up of 160 months. However, freedom from significant aortic stenosis or aortic regurgitation was $52.4 \pm 16.9\%$.

Pulmonary autograft has also been used in the treatment of rheumatic aortic valve disease (Pieters *et al*., 1996). However, the results of pulmonary autograft in rheumatic aortic valve disease are poor compared to its use with other causes of aortic valve disease at any age. Kumar *et al*. (2005) have reported freedom from significant aortic stenosis or regurgitation of $91.5 \pm 2.8\%$ in non-rheumatics and $80.6 \pm 4.8\%$ in rheumatics. At median follow-up of 90 months, event-free survival was $86.2 \pm 4.9\%$ for non-rheumatics and $68.9 \pm 5.3\%$ for rheumatics. They have also shown the rheumatic effect in explanted autografts. They do not recommend pulmonary autograft for younger patients (< 30 years) with rheumatic heart disease, opting for aortic valve repair as far as possible in such circumstances.

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38

Persistent Truncus Arteriosus

M. R. de Leval

Persistent truncus arteriosus accounts for fewer than 3% of all congenital heart defects. It is characterized by the presence of a single arterial trunk that arises from the base of both ventricles by way of a semi-lunar valve, a high ventricular septal defect (VSD) subjacent to the valve, and by the origination of the pulmonary arteries from the truncus (**Figure 38.1**). In type I truncus arteriosus, a single main pulmonary trunk connects the main left and right pulmonary arteries to the truncus; in type II, the pulmonary arteries have a common origin from the truncus; and in type III, the right and left pulmonary arteries originate separately from the lateral aspect of the truncus. Patients with type IV truncus arteriosus (described by Collett and Edwards, 1949), in whom the lungs are supplied by way of systemic arteries, should be grouped with patients with pulmonary atresia and VSD. In rare instances, one of the pulmonary arteries arises from a ductus arteriosus. A vascular sling made by the left pulmonary artery, which arises from the right pulmonary artery and compresses the tracheobronchial tree, has also been reported.

The aortic arch is left-sided in about 60%, right-sided in about 30% and interrupted in about 10% of cases of persistent truncus arteriosus. The interruption is usually of type B (interruption distal to the origin of the left carotid artery).

The truncal valve is tricuspid in approximately 50%, bicuspid in about 30% and quadricuspid in about 15–20% of cases of persistent truncus arteriosus. In exceptional cases, the valve is unicuspid. Various degrees of truncal valve dysplasia are often present, resulting in truncal valve regurgitation, truncal valve stenosis or both. The coronary arteries exhibit various patterns of origin. There are usually two coronary arteries, but a single coronary artery is not uncommon. Sometimes the left coronary artery originates high from the posterior wall of the truncus.

The VSD is classically a subarterial defect, the roof of which is made by the truncal valve. Two types of arrangement can be seen posteriorly (Crupi *et al*., 1977).

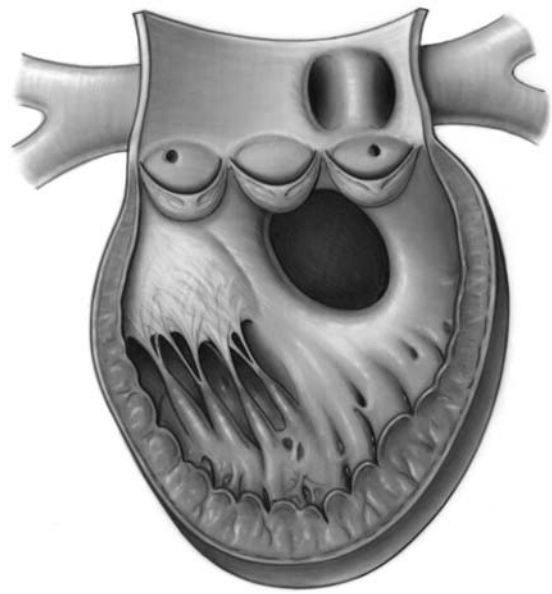

Figure 38.1

In approximately 80% of patients with persistent truncus arteriosus, the posterior margin of the defect is muscular, completely separated from the anterior leaflet of the tricuspid valve, and made by the posterior limb of the trabecula septomarginalis fused with the ventriculo-infundibular fold (**Figure 38.2a**) (infundibular subarterial defect). The other 20% of patients have a perimembranous infundibular defect, the posterior margin of which extends back to the anterior leaflet of the tricuspid valve (**Figure 38.2b**). In some cases of persistent truncus arteriosus, there is an effacement of the ventriculo-infundibular fold that accounts for a wide area of continuity between the truncal valve and the tricuspid valve. There is always continuity between the truncal valve and the mitral valve. The

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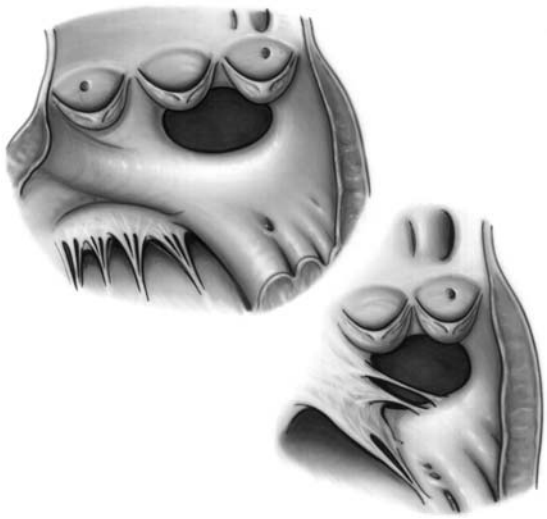


Figure 38.2

truncal valve overrides the VSD and relates equally to the two ventricles in about 50% of cases. In the other cases, the truncal valve originates predominantly from the right ventricle but in some instances from the left ventricle.

DiGeorge's Syndrome

A significant proportion of patients with persistent truncus arteriosus have DiGeorge's syndrome, which consists of aplasia (complete form) or hypoplasia (partial form) of the thymus and parathyroid glands, which results from malformation of the third and fourth pharyngeal pouches. T cell immune deficiency, hypocalcaemia and predilection to infection are the commonest features of DiGeorge's syndrome. Facial dysmorphism is also often associated with the syndrome. A reduction in the total number of T cells is the result of a reduction in the T helper cell subset (Radford *et al*., 1988). Because fatal graft-vs.-host reactions have been observed with blood transfusions in patients with DiGeorge's syndrome, it is recommended that irradiated blood be used in affected children (Parshuram *et al*., 2002). Calcium supplementation and particular attention to the prevention of infections are also part of the management of patients with DiGeorge's syndrome.

DIAGNOSIS AND INDICATIONS FOR SURGERY

Congestive heart failure secondary to excessive pulmonary blood flow, with or without truncal valve regurgitation, usually develops in early infancy. Lowness of the diastolic pressure, caused by the run-off into the low-resistance pulmonary arteriolar resistance system, may result in an impairment of the coronary perfusion with decreased ventricular performance. Truncal valve regurgitation further increases the haemodynamic burden on the ventricles. The median age of death reported in autopsy series of untreated patients has varied from a few weeks to 6 months (Marcelletti *et al*., 1976). In those who survive beyond the age of 1 year, progressive pulmonary arteriolar resistance obstructive disease is likely to develop.

Accurate preoperative haemodynamic and anatomical evaluation is mandatory. From an anatomical standpoint, non-invasive methods of investigation, such as echocardiography and Doppler interrogation, have rendered angiography superfluous in the majority of the cases. The investigators must be aware, however, of the possibility of an interrupted aortic arch, a ductal origin of one pulmonary artery or the absence of one pulmonary artery, and must be aware of associated defects such as multiple VSDs. Haemodynamic investigations are usually not required in young infants with congestive heart failure and large pulmonary blood flow. Patients with persistent truncus arteriosus should undergo operation early, generally before the age of 2 or 3 months, even if they are asymptomatic. It is perhaps useful to wait for the pulmonary arteriolar resistance to fall and not to operate during the first 2 or 3 weeks of life unless a patient is in intractable congestive heart failure, in which case surgery should take place regardless of the patient's age. There is a general consensus that banding of the pulmonary artery as treatment for persistent truncus arteriosus is obsolete.

In older patients, the preoperative haemodynamic evaluation is primarily concerned with the status of the pulmonary arterial bed. Although the left and right ventricular blood is ejected into the truncal root, which gives rise to the pulmonary arteries, the pressure and oxygen content of blood in the pulmonary arteries are not necessarily the same as those in the truncus. Considerable streaming of blood may occur. It is therefore mandatory that both pulmonary arteries be catheterized to obtain the pressure and oxygen content within them in order to assess pulmonary arteriolar resistance. In children, the criteria for operability are closely related to the pulmonary arteriolar resistance. Patients in whom the pulmonary arteriolar resistance is higher than 10 units/m^2 have a better prognosis if they do not undergo operation (Mair *et al*., 1974).

In exceptional patients with banded pulmonary arteries, the possible inequalities of pressure and flow between the two pulmonary arteries often make it difficult to precisely calculate pulmonary resistance. In practice, if one pulmonary artery has a mean pressure of less than half the systemic pressure, the lung supplied by that artery

has been protected and does not have severe arterial obstructive changes (McFaul *et al*., 1976).

For patients with persistent truncus arteriosus and a single pulmonary artery, Mair *et al*. (1977) suggested that, at a given degree of obstructive pulmonary vascular disease confirmed by histological studies, the calculated resistance was twice that of patients with two pulmonary arteries. The criteria for operability could therefore be based on the calculated resistance divided by 2. The long-term results of repairs on a single pulmonary artery, however, have been significantly worse than those of repairs on two pulmonary arteries (Fyfe *et al*., 1985).

SURGICAL TECHNIQUES

Repair of Truncus Arteriosus

After a midline sternotomy incision, a patch of pericardium is harvested to be used if a proximal extension of the homograft conduit onto the right ventricle is necessary. A purse-string suture is placed on the right atrial appendage as a retraction to expose the truncus and is used later for atrial cannulation. The anatomy of the truncus is assessed. Both pulmonary arteries are fully dissected down to their branching in the hilus of both lungs (**Figure 38.3**). The aorta is also fully mobilized, particularly if it is intended to transect the truncus in order to detach the pulmonary arteries (for repair of types II and III). The head vessels are dissected and encircled with snares for cases of interrupted aortic arch. Dissection of the aorta is carried down onto the descending thoracic aorta from the right lateral aspect of the aorta; this dissection is often completed during cardiopulmonary bypass.

The main pulmonary arterial trunk (type I) or the proximal pulmonary arteries (types II and III) are also encircled with snares, to be occluded as soon as cardiopulmonary bypass is initiated, in order to prevent run-off and flooding in the pulmonary circulation.

The aortic cannula is placed as distally as possible. Bicaval cannulation is used more and more frequently in neonatal cardiac surgery and the operation is performed on high-flow cardiopulmonary bypass, with various degrees of hypothermia. For very small infants we continue to use a single cannula in the right atrium and perform the procedure under deep hypothermia, with a period of total circulatory arrest to close the ventricular septal defect. If there is significant truncal valve regurgitation the aorta must be cross-clamped as soon as the bypass is initiated. The cold blood cardioplegia is infused directly into the truncal root if the truncal valve is competent, directly into the coronary ostia if the truncal valve is incompetent.

Detaching the Pulmonary Arteries

The very important first step of the repair consists of detaching the pulmonary arteries from the truncus. In

Figure 38.3

type I, a limited incision is made on the anterior or superior aspect of the main pulmonary trunk (**Figure 38.4**). The pulmonary arterial orifices are visualized, as are the truncal valve and the ostium of the left coronary artery, which on occasion arises near the pulmonary trunk. The incision is then carried inferiorly and posteriorly so as to detach the pulmonary arteries completely (**Figure 38.5**). The defect in the aorta is usually closed transversely by direct suture. Sometimes it is better to use a small prosthetic patch of polyester (Dacron) or polytetrafluoroethylene (Gore-Tex) to repair the defect, in order to prevent a distortion of the truncal valve (**inset, Figure 38.6**).

In types II and III truncus arteriosus, I now prefer to transect completely the aorta distal to the truncus. First, the distal aorta is opened transversely on its anterior surface. Then the incision is continued posteriorly above the orifices of the pulmonary arteries, which are detached as an elliptical button containing both orifices. The ascending aorta, which had been fully mobilized, is reconstructed by an end-to-end anastomosis, wider bites being taken on the proximal segment to make up for the discrepancy in the size of the two ends of this aorta (**Figure 38.7**).

Closure of the Ventricular Septal Defect

An oblique right ventriculotomy incision is then made, from about the midpoint of the anterior right ventricular wall to the base near the left aspect of the annulus of

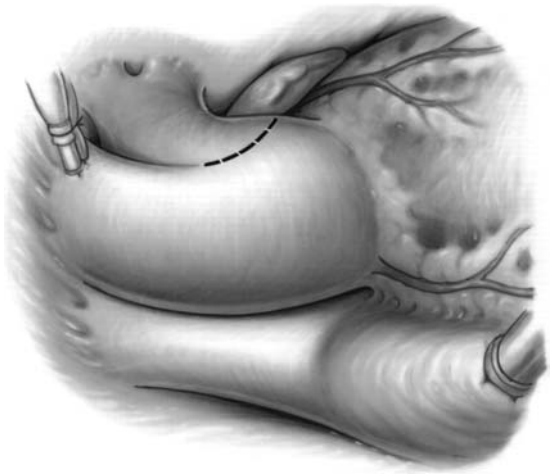


Figure 38.4

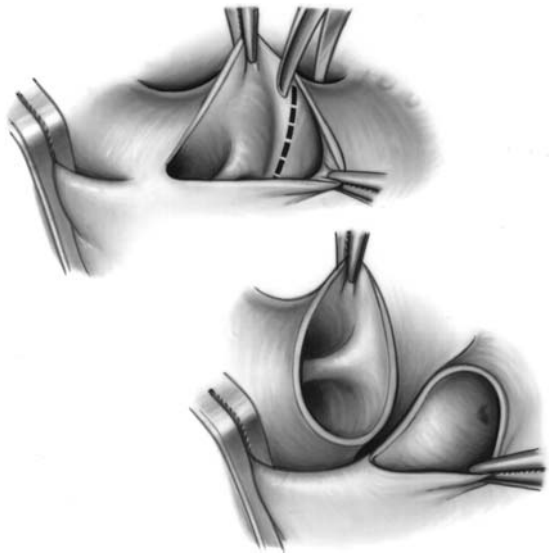


Figure 38.5

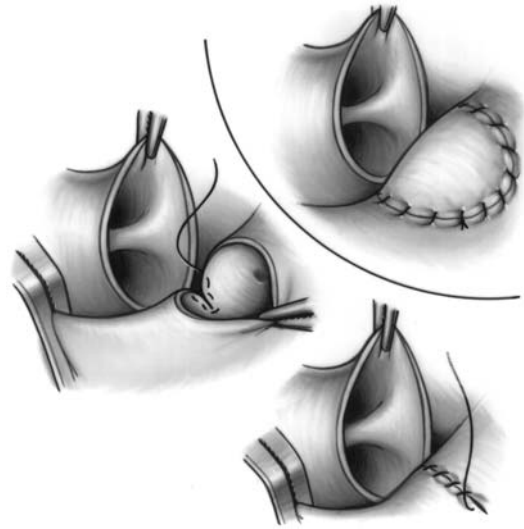


Figure 38.6

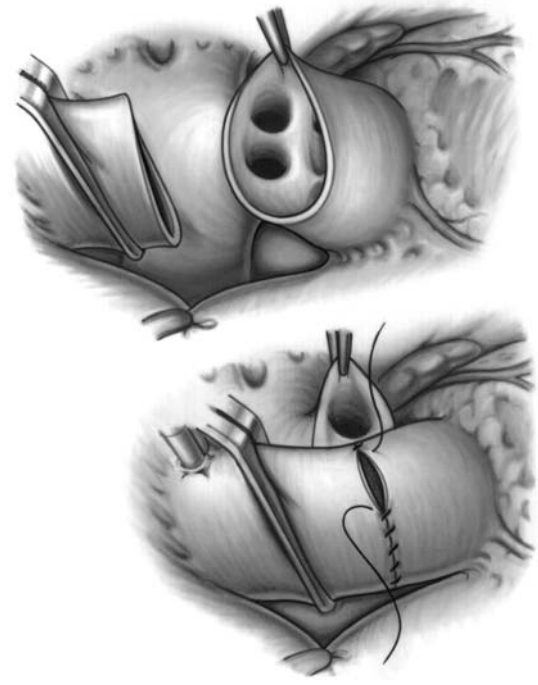


Figure 38.7

the truncal valve. The type of ventricular septal defect (infundibular, muscular or perimembranous) is identified, as well as its relationship with the truncal valve. Heavy trabeculations of the parietal and septal extensions of the infundibular septum are divided to facilitate the exposure of the ventricular septal defect and prevent late occurrence of subvalvar pulmonary stenosis. The ventricular septal defect is then closed with a patch of xenograft pericardium, Gore-Tex or Dacron, with a running suture of 6-0 or 7-0 Prolene. The sutures are placed on the free margin of the defect if it is a muscular defect, away from the posterior margin if it is a perimembranous defect. Anteriorly and superiorly, the sutures are placed through the anterior wall of the right ventricle as horizontal mattress sutures, which can be reinforced

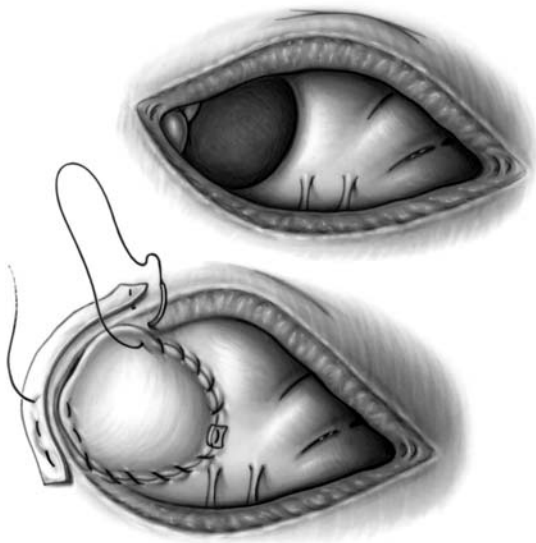


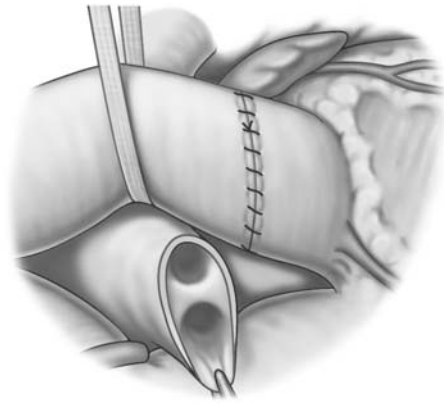
Figure 38.8

with a strip of pericardium (**Figure 38.8**). In trimming the patch, the surgeon must account for the degree of overriding of the truncal valve to prevent a narrowing of the pathway from the left ventricle to the aorta.

Opinions vary with regard to the closure of the patent foramen ovale. Some surgeons believe that it may constitute an escape route for the blood to reach the left ventricle in case of postoperative pulmonary hypertensive crises. They routinely leave the PFO opened.

Establishment of Continuity between the Right Ventricle and the Pulmonary Arteries

Most surgeons use an interposition extracardiac valved conduit to establish continuity between the right ventricle and the pulmonary arteries. Some have recommended a direct connection using autologous tissues. Various types of extracardiac valved conduits are available: aortic or pulmonary cryopreserved homografts, porcine valved Dacron conduits, bovine jugular vein grafts, pericardial composite valve conduits. The aorta is unclamped after the closure of the right atrium and the patient is rewarmed while the conduit is being placed. The pulmonary anastomosis is done first. After full mobilization of the pulmonary arteries, it is easy to bring them to the right side of the aorta (**Figure 38.9**) and take them in a side-biting clamp to perform the anastomosis. The valve is placed as distally as possible to prevent sternal compression later. The anastomosis is performed with a continuous suture of 6-0 or 7-0 Prolene. The conduit is then brought behind the aorta and to the left. If a



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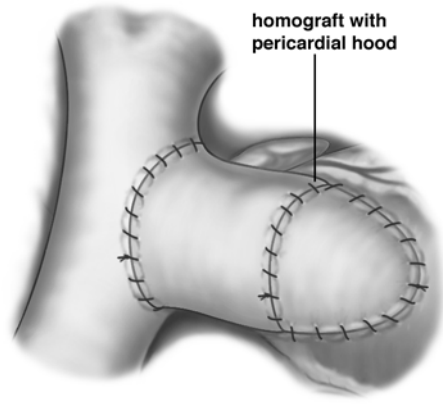


Figure 38.10

homograft is used, there is a need to place a hood or a gusset for the ventricular anastomosis. This can be made of autologous or heterologous pericardium or Gore-Tex if a pulmonary homograft is used. If an aortic homograft has been used, which is currently our conduit of choice, a portion of the ascending aorta is used for the proximal hood (**Figure 38.10**). If a Dacron or heterologous conduit is used, it is appropriately trimmed to be anastomosed to the right ventriculotomy incision. The suture line on the right ventricle can be reinforced with a strip of autologous pericardium or a Teflon strip. 5-0 or 6-0 Prolene is usually used for this anastomosis.

A reconstruction of the pulmonary outflow tract without prosthetic conduit was described by Lecompte in 1982 for various conditions, including persistent truncus arteriosus. The technique consists of bringing the bifurcation of the pulmonary artery in front of the reconstructed aorta and to anastomose the pulmonary

arteries directly to the right ventriculotomy incision. In 1986, Reid *et al*. proposed a technique of direct connection, which was later popularized by Barbero-Marcial *et al*. (1990). The technique consists of making a vertical pulmonary arteriotomy incision through which the communication with the truncus is closed with a patch. The back wall of the ventriculo-arterial connection is made either of a direct anastomosis between the pulmonary artery and the right ventriculotomy incision, if the arteriotomy is immediately adjacent to the upper portion of the right ventriculotomy; otherwise, a trapdoor-type incision is made in the pulmonary artery and turned down towards the ventricle to bridge the gap between the pulmonary artery and the right ventricle. Several types of materials can be used to construct an anterior hood between the right ventricle and the pulmonary arteries.

SPECIAL CONSIDERATIONS

Repair of Truncus Arteriosus with Truncal Valve Regurgitation

Truncal valve regurgitation remains a challenging problem. Mild to moderate truncal valve regurgitation should be dealt with conservatively. Once severely regurgitant, the truncal valve must be repaired or, rarely, replaced. Regurgitation often results from a prolapsing vestigial leaflet in a quadricuspid valve. The prolapsing leaflet can be sutured to one or both adjacent leaflets (**Figure 38.11**), thereby creating a functional tricuspid or bicuspid valve (Mavroudis and Backer, 2001).

An annular valvuloplasty consisting of excision of the small prolapsing leaflet with annular remodelling has also been described (Imamura *et al*., 1999; Mavroudis and Backer, 2001).

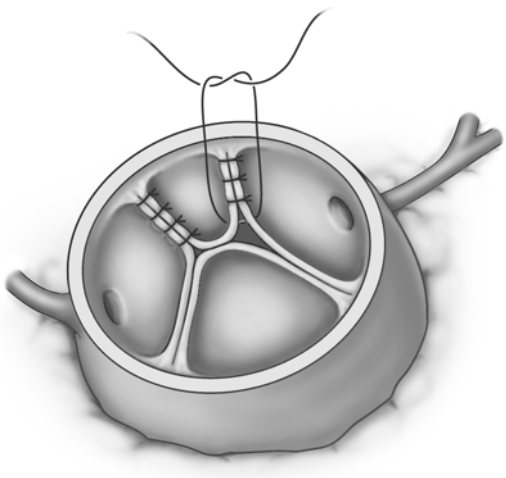


Figure 38.11

Severe truncal valve regurgitation resulting from gross dysplasia of the truncal valve must be treated by valve replacement: either a prosthetic valve or an aortic homograft. Even with a large truncal valve, it may be difficult to accommodate a prosthetic valve in small infants. It is sometimes necessary to make a vertical incision in the truncus across the annulus into the ventricular septal defect to perform a repair similar to the aortoventriculoplasty described by Kono (Bove *et al*., 1989). The results of homograft root replacement have been disappointing (McElhinney *et al*., 1998).

If the truncal valve is regurgitant, it is important to avoid ventricular distension when the myocardial contractility weakens with cooling. In these circumstances, the left heart should be vented and the aorta must be readily cross-clamped. Truncal valve regurgitation can also impair the delivery of cardioplegia. If this is the case, it should be infused directly into the coronary ostia. If there is residual truncal valve regurgitation at the end of the procedure, there is again a risk of left ventricular distension when the aorta is unclamped. In these circumstances it is useful to start rewarming earlier, so that the myocardium can regain its contractility sooner. If distension occurs, it should be dealt with by massage of the left ventricle and placement of a vent through the extracardiac conduit before completing the ventricular anastomosis.

Repair of Truncus Arteriosus with Interrupted Aortic Arch

Persistent truncus arteriosus can be associated with interruption of the aortic arch of all three types, but most commonly of type B (interruption distal to the origin of the left carotid artery). The first successful repair of this condition was reported by Gomes and McGoon in 1971. They described a technique whereby the patent ductus arteriosus is left in place for the aortic arch reconstruction. This technique is not applicable in young infants, in whom the ductus arteriosus is likely to close and thus produce aortic stenosis or atresia. The conventional approach has been to excise the ductal tissue as completely as possible and bring the descending thoracic aorta to the ascending aorta.

The ascending and descending thoracic aorta, the pulmonary arteries, the ductus arteriosus, and the head vessels are fully mobilized The arterial cannula is introduced into the distal ascending aorta and another one through the PDA into the distal descending aorta. A single cannula is introduced into the right atrium. The pulmonary arteries are snared as soon as the bypass has begun. The patient's temperature is lowered to 18°C, the circulation is arrested, and the head vessels are snared. Alternatively, the technique of continuous cerebral perfusion described in Chapter 11 can be utilized.

The truncus is transected, the pulmonary arteries are excised from the truncus, the ductus arteriosus is transected at its pulmonary end and followed down to the descending thoracic aorta, from which it is also excised. The mobilization of the descending thoracic aorta is completed, and the vessel is brought superiorly to be anastomosed to a longitudinal incision in the posterolateral ascending aorta (**Figure 38.12**). This incision can be extended superiorly to the origin of the left carotid artery. It is sometimes useful to divide the left subclavian artery to facilitate the mobilization of the descending aorta.

Our current approach uses either a complete or partial transection of the truncus (**Figure 38.13**) and augmentation of the aortic arch and the small ascending aorta with a patch of pulmonary homograft. This will allow for a better size match with the proximal aorta (**Figure 38.14**), which would allow tension-free anastomosis. An anterior translocation of the pulmonary bifurcation (Lecompte manoeuvre; Lecompte *et al*., 1982) has been recommended (see also Chapter 36, Arterial Switch Operation) to prevent a compression of the right pulmonary artery and facilitate the placement of the right ventricle-to-pulmonary artery conduit (Imamura *et al*., 1999).

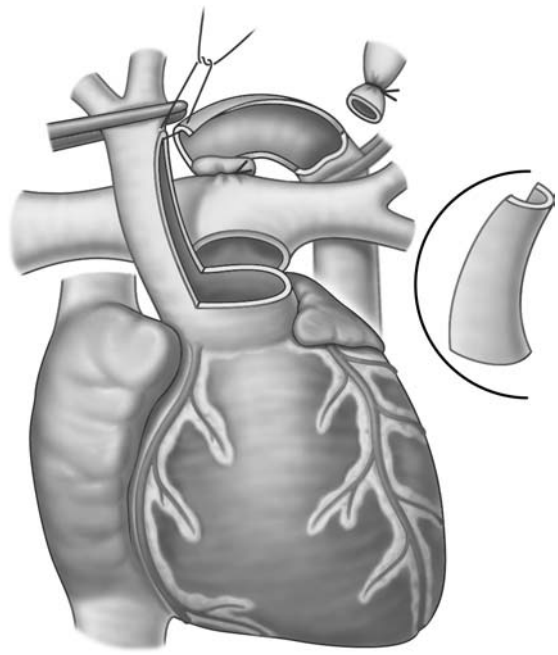
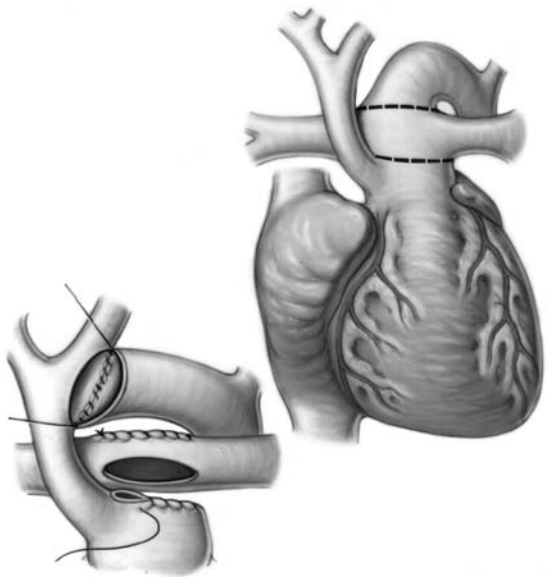


Figure 38.13

Repair of Truncus Arteriosus with Discontinuous Pulmonary Arteries

Most commonly, the left pulmonary artery arises from the ductus arteriosus and the right pulmonary artery arises



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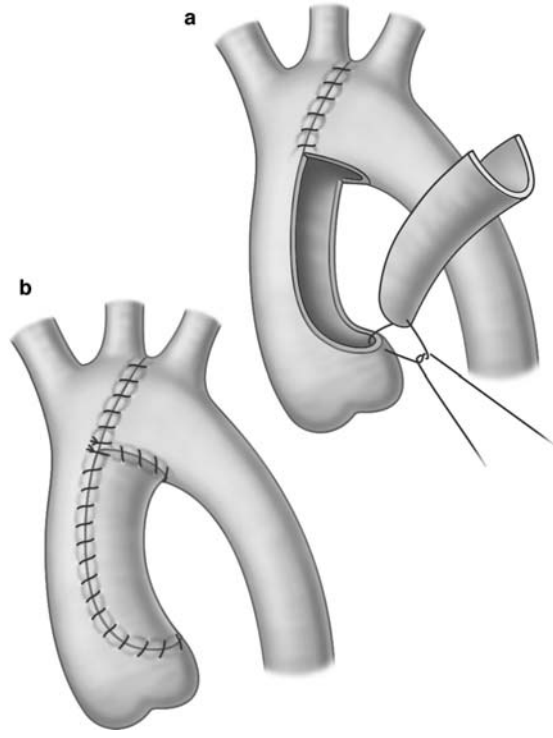

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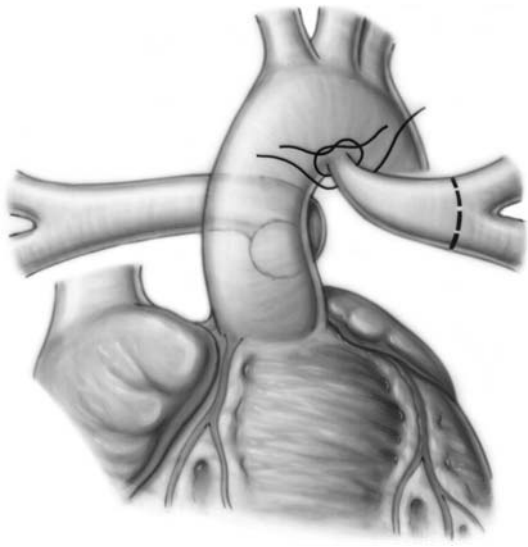

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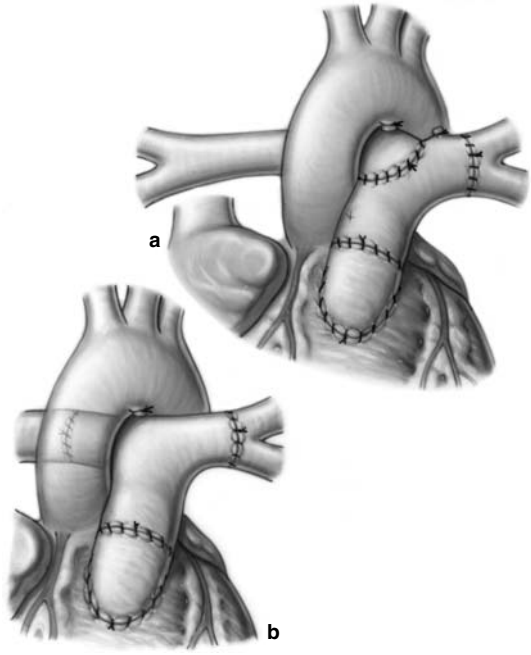


Figure 38.16

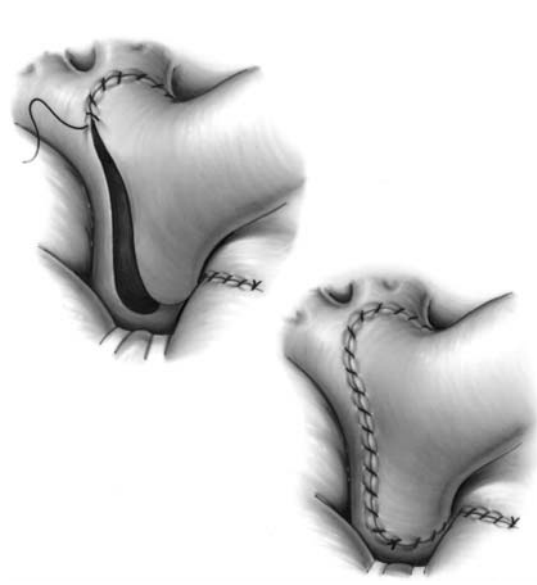
from the truncus arteriosus (**Figure 38.15**). Both arteries are detached from their aortic connections. Ideally, the two pulmonary arteries should be anastomosed together directly. However, if this is not feasible, the larger artery is sewn end-to-end to the homograft, and the other one is sewn end-to-side (**Figure 38.16a**). Alternatively, the pulmonary homograft with its bifurcation can be used with an end-to-end anastomosis for both pulmonary arteries (**Figure 38.16b**).

Repair of Truncus Arteriosus with Prior Banding of the Pulmonary Arteries

The pulmonary arteries are detached from the truncus and the resulting aortic defect is closed, as previously described. The pulmonary arterial orifice is then enlarged by incising laterally on the right and left pulmonary arteries through and well beyond the fibrotic banded zone. If the pulmonary arterial wall is of good quality, the homograft conduit can be trimmed in a fishmouth manner, so that the resulting flap-like extension can be sutured as a gusset-type patch to enlarge the proximal banded and fibrotic pulmonary arteries (Parker *et al*., 1975) (**Figure 38.17**). The pulmonary arteries beyond the band are sometimes so thin and fragile that a direct anastomosis would tear the vessels. In such cases, it is useful to sew a patch of pericardium or pulmonary homograft from hilum to hilum, across the banded area of both pulmonary arteries, as a large patch onto which the homograft is later anchored (**Figure 38.18**).

POSTOPERATIVE MANAGEMENT

Pulmonary hypertensive crises used to be the main causes of postoperative morbidity and mortality. Their incidence and severity has significantly decreased, since most of these patients are now treated electively during the neonatal period, or at least under the age of 3 months.



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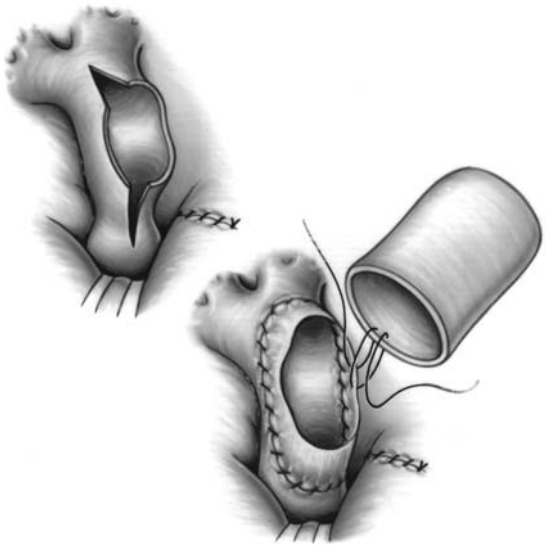

Figure 38.18

Hyperventilation with a high fraction of inspired oxygen and heavy sedation is often sufficient to overcome these crises. Should they be more severe, the use of pulmonary vasodilators, such as nitric oxide, is indicated.

RESULTS

The operative mortality following repair of persistent truncus arteriosus has decreased significantly during the past decade. Although multifactorial, this improvement is certainly due in part to the introduction of elective repair in the neonatal period or early infancy. A review of the recent literature (Miyamoto *et al*., 2005) shows that the published operative mortality varies (3–17%) for individual institutions. Multi-institutional reports published in 1998 by the European Congenital Heart Surgeons Society and by the Consortium USA reported mortalities of 27% and 44%, respectively. Truncal valve regurgitation and interruption of the aortic arch remain significant risk factors.

Between 2000 and 2004 we have operated on 18 infants with truncus arteriosus; two had associated interrupted arch, one died. This patient had a repair of associated interrupted arch and long segment congenital tracheal stenosis.

As far as the long-term results are concerned, all these patients require further surgery on the right ventricular outflow tract. The durability of a variety of conduits, including homografts inserted in the neonatal period or in early infancy, have proved disappointing (Wells *et al*., 2002; Perron *et al*., 1999; Meyns *et al*., 2004). A recent report (Chen *et al*., 2005) compared the results of homograft repair and direct connection and showed that the valved homograft recipients were more likely to require reoperation than patients receiving direct connection.

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39

Atrio-ventricular Discordance and Congenitally Corrected Transposition

R. B. B. Mee and J. Stark

Atrio-ventricular (AV) discordant connection is an anomaly in which the morphological right atrium (mRA) connects to the morphological left ventricle (mLV) and the morphological left atrium (mLA) connects to the morphological right ventricle (mRV). AV discordance may be associated with either situs solitus or situs inversus. The ventriculo-arterial connection in patients with AV discordance also varies. It may be discordant, as in congenitally corrected transposition of the great arteries (ccTGA), or it may be double-outlet right or left ventricle (DORV or DOLV). Isolated AV discordance (without TGA, DORV, DOLV) is very rare and when seen is usually associated with heterotaxy-type hearts with venous and other atrial connection anomalies.

Associated defects, such as atrial or ventricular septal defect (VSD) and pulmonary or subpulmonary stenosis or atresia, are usually present. These defects determine the clinical picture and are the subject of the surgical treatment. The AV node, the bundle of His and the peripheral branching of coronary arteries are also abnormal in patients with AV discordance. In ccTGA with atrial situs solitus there is an anterior AV node in the atrial septum at the point where the anterior limbus of the atrial septum joins the AV valve ring. The penetrating bundle then passes over the pulmonary annulus and descends onto the anterior part of the infundibular septum (see **Figures 8.29** and **8.30**) (Monckeberg, 1913). In situs inversus, the penetrating bundle arises from the regular posterior node, and its course is similar to that in AV concordance (Wilkinson *et al*., 1978). For more details about the conduction system in AV discordance, see Chapter 8.

AV discordance with ventriculo-arterial discordance was probably first described by Rokitansky in 1875. The first clinical descriptions were published by Anderson *et al*. (1957) and Schiebler *et al*. (1961). Positional anomalies of the heart may complicate the surgical procedure (Kinsley *et al*., 1974). If the heart is rotated to the right, cannulation for cardiopulmonary bypass and approach to the VSD may be difficult. If the heart is rotated around a horizontal axis, one ventricle may lie below the other one (overunder ventricles).

A discordant relationship between the apex of the ventricular mass and the atrial situs (situs solitus with dextrocardia or situs inversus with laevocardia) appears to be more common when there is severe subpulmonary stenosis, pulmonary valve stenosis or pulmonary valve atresia. When the ventricular mass lies in front of the morphological right atrium, the free wall of the right atrium is narrow and the right atrial appendage is usually small.

DIAGNOSIS

Diagnosis can be made accurately by two-dimensional (2D) echocardiography and Doppler echocardiography (Hagler *et al*., 1981; Sutherland *et al*., 1983). Cardiac catheterization provides important information about physiology. Pressures and flows are measured to assess the degree of pulmonary stenosis and shunts and to calculate pulmonary arteriolar resistance. Angiography is particularly important for assessment of peripheral pulmonary arteries. Straddling of AV valves is often associated with AV discordance. Because the severe form of straddling may alter the surgical plan, it is important to diagnose it preoperatively. 2D echocardiography is the most reliable technique for assessing straddling.

In the initial investigation, the AV and ventriculo-arterial (VA) connection must be established, the positional anomalies of the heart defined, and associated intra- and extracardiac malformations confirmed or ruled out. The investigation must provide information about the sizes of ventricles, the size and position of the VSD or VSDs, the degree of pulmonary stenosis, the magnitude of intracardiac shunts, and the pulmonary arteriolar

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resistance. It is also important to evaluate the origins of the coronary arteries and initial branching pattern if possible, the venoatrial connections, the absolute sizes and morphology of the semi-lunar valves, the presence or absence of coarctation of the aorta and arch obstruction, and the morphology and function of the AV valves, particularly the left-sided tricuspid valve, which is often abnormal (Allwork *et al*., 1976). Some patients show features of Ebstein's anomaly, with downward displacement of the origin of the posterior and septal leaflets (Becu *et al*., 1955).

INDICATIONS

The natural history of the disease is not favourable. About 10% of infants with AV discordance are born with complete heart block (Friedberg and Nadas, 1970). Another 40–50% are born with first- or second-degree heart block (Bonfils-Roberts *et al*., 1974).

The closure of the VSD is more difficult in small hearts than in large hearts, and an extracardiac valved conduit is required for treating patients with significant unresectable subpulmonary valve stenosis or stenosis and hypoplasia of the pulmonary valve. It may be advisable to perform palliative procedures (systemic-to-pulmonary shunts) in such patients in infancy and defer definitive treatment until the organs are larger. When pulmonary blood flow is unrestricted, biventricular repair may be indicated within the first 3 months of life, or a pulmonary artery band (PAB) should be applied to protect the pulmonary circuit when the possibility of biventricular repair is in doubt, or if the patient presents after 2 months of age with a restrictive VSD, and low mLV pressures in order to retrain the mLV and to preserve the options for biventricular repair (either VSD closure alone or combined with a double-switch operation—Senning or Mustard plus arterial switch operation). The PAB should be positioned carefully to preserve the pulmonary valve and sinotubular junction. Patients tolerate moderate to severe tricuspid valve incompetence poorly and, when present, this should accelerate surgical management decision making and implementation (Lundstrom *et al*., 1990).

SURGERY FOR CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Surgical Technique

Surgery for ccTGA can be approached in two ways. The so-called classical or physiological approach is to focus on individual associated anomalies, such as the VSD, ASD and mLV outflow tract obstruction, tricuspid valve regurgitation, PDA, coarctation, arrhythmias, etc, leaving the mRV and TV in the systemic circuit. Early mortality for this approach has been low but late problems with RV and TV have been significant and have increased with time (Sano *et al*., 1995). This is similar to what has been observed after atrial repair of simple TGA and more commonly with complex TGA or DORV with subpulmonary VSD. One could theoretically surmise that mRV and TV failure are even more likely in ccTGA when approached physiologically, because of the higher incidence of VSD (70%), the markedly higher incidence of organic TV disease and the higher propensity for arrhythmias (accessory conduction pathways associated with Ebstein's anomaly) and the inherent trend to spontaneous complete heart block (30%).

The second approach is to restore the mLV to the systemic circuit (double switch) whenever possible. This is the author's preference, and much of this chapter will be devoted to the anatomical correction approach. For atrial repair of atrio-ventricular discordance, the author's preference is a Senning-type procedure, with or without a modification of the Schumacker's modification (Schumacker, 1961) according to the position of the ventricular mass and the size of the free wall of the mRA.

Pulmonary Artery Banding

This is most accurately performed through a midline sternotomy, using the Trussler formula to calculate the PAB circumference for patients with a non-restrictive VSD, and a larger PAB carefully adjusted to avoid immediate or early mLV dysfunction when the mLV is at low pressure and needs retraining. In the latter case, one aims for the highest mLV pressure without trans-oesophageal echo signs of impaired contractility of the mLV free wall, but probably, more importantly, *no* rise in central venous pressure (we consider this the most reliable sign of impending mLV strain). If this occurs, the PAB should be loosened immediately, or if central venous pressure rises and decreased mLV free wall contractility is confirmed by echo after leaving the operating room, loosening of the PAB should be considered forthwith. We have identified an increase in myocardial oedema by T2-weighted magnetic resonance imaging (MRI) 1 week post-PAB, proportional to the degree of post-PAB mLV dysfunction. When the correct size of the PAB has been ascertained in the operating room, a dopamine infusion at 2.5–3.0 μg/kg/min is commenced and run empirically for 5 days. The PAB is carefully sutured to the MPA wall and the PA bifurcation inspected for possible compromise. Overall, it is probably preferable to err towards some compromise of the bifurcation than of the sinotubular junction. An

evolved empirical routine has been to keep the patient asleep and ventilated overnight. If echocardiography confirms unimpaired mLV function the next morning, the patient is weaned from the ventilator. If there is some doubt about mLV function on echocardiography and from central venous pressure measurement, a decision is made the morning after surgery to either loosen the PAB or keep the patient asleep and ventilated for another 24 h and then repeat the assessment. Neglected early mLV dysfunction and its association with myocardial oedema probably indicates myocardial damage and therefore a compromise of subsequent mLV retraining and function. Late mLV dysfunction has been observed after rapid two-stage switch for simple TGA, where in the early experience it was common to observe severe mLV dysfunction for a few days after the first stage (tight PAB and shunt).

Systemic-to-Pulmonary Artery Shunting

There are many approaches to shunting. The author's preference for a neonate or young infant is to place a 5 mm PTFE shunt from the subclavian artery to the ipsilateral branch pulmonary artery on the side of the morphological right lung, irrespective of the side of the aortic arch. A small shunt from a large innominate artery will not last as long as a larger shunt from a smaller but growing subclavian artery. There is some theoretical value, if the patient remains stable, in being able to defer definitive repair when a conduit is necessary, and particularly so if a Senning/Rastelli is being contemplated, to minimize growth discrepancy of the subaortic tunnel. Furthermore, in those patients with a discordant relationship between the atrial situs and the position of the apex of the heart, with a narrow mRA free wall the modified Senning procedure becomes less critical in a bigger patient. If the branch pulmonary artery on the side of the morphological left lung is hypoplastic at the time of presentation, then that is the side that should be shunted. It is the author's opinion that definitive biventricular repair for ccTGA with PVS or pulmonary atresia should be performed while the patient is still somewhat volume-loaded. A prolonged state of significant cyanosis with Qp:Qs < 1 in a growing child may well mean the ventricles are less than normal size. In this case, the result of biventricular repair is immediate volume loading for a heart that has experienced a significant period of aortic cross-clamp time. A pre-existing bidirectional cavopulmonary shunt will accentuate the phenomenon of chronic volume underloading in a growing patient. If the proposed ventricle for pulmonary blood flow is small, then a bidirectional cavopulmonary shunt at the time of septation becomes more appropriate.

Tricuspid Valve Replacement and Repair

A variable incidence of Ebstenoid-type abnormality of the tricuspid valve is reported and is not uncommon in ccTGA, but not all significantly incompetent tricuspid valves have this anomaly. Moderate or severe TR is seen more commonly when pulmonary blood flow is unrestricted or when mRV dysfunction has developed. Repair of a systemic tricuspid valve is difficult and the results unreliable. Replacement can be considered, and if so, a mechanical prosthesis is the device of choice in children. Alternatively, the TR can be considered as a primary indication for the double-switch operation. The mRV and TV are returned to the pulmonary circuit (double switch), under which circumstances the state of the TV can usually be ignored with a high probability of immediate decrease in TR, mRV volume and mRV dysfunction, provided pulmonary resistance is low or can be counted on to fall quickly after the double-switch procedure.

Left Ventricular Outflow Tract Obstruction (mLVOTO)

A stenotic bicuspid pulmonary valve of adequate size may still be suitable for an arterial switch after valvotomy and measurement. Subpulmonary obstruction takes several forms. Anterior muscular obstruction cannot be removed without creating complete heart block in ccTGA in situs solitus (SLL), but if heart block is already present and the pulmonary valve ring is of adequate size for an arterial switch operation, then this muscle can be resected to relieve mLVOTO. Quite commonly, important mLVOTO is on the basis of accessory valve tissue, and if at operation the accessory tissue is not deemed integral to the function of the mitral valve (MV), it can be removed. Occasionally, the mLVOTO is due to aneurysm of the membranous component of the interventricular septum, which always seems larger in ccTGA. This aneurysm can also be resected.

VSD Closure

The majority of VSDs are perimembranous, but are often reported as inlet-type from echocardiography evaluation. Inlet extension of a perimembranous VSD is certainly seen particularly in the presence of an overriding or straddling MV. The size of the VSD is very important when considering the Rastelli component of a double-switch operation. It is difficult to enlarge an undersized perimembranous VSD superiorly in ccTGA situs solitus (SLL) without creating CHB. It can be inspected and measured first via the initial mRA incision and the MV. If the Rastelli operation is feasible, actual closure is approached via the ventriculotomy in the

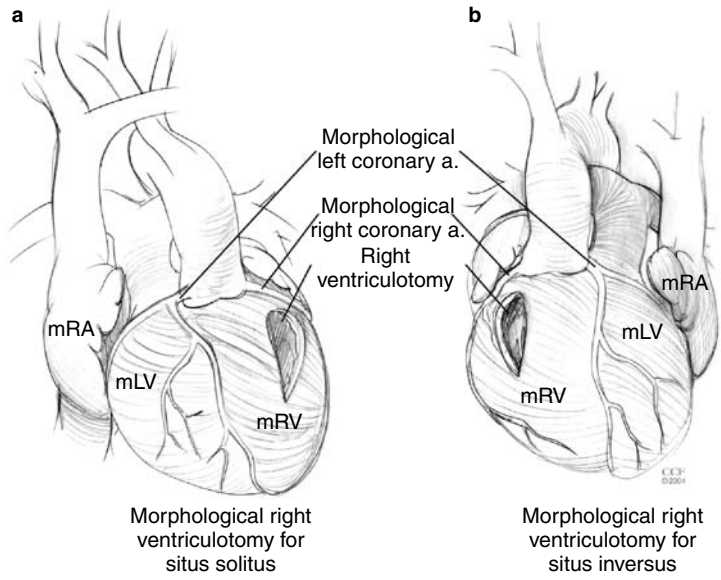

Figure 39.1

mRV (**Figure 39.1a,b**), under which circumstances, the conducting bundle is remote from the proposed suture line and relatively safe in ccTGA with atrial situs solitus (SLL) configuration, but in the usual inferior position, and vulnerable in ccTGA with atrial situs inversus (IDD) configuration. Sutures (interrupted or running) are placed through the attachment of the tricuspid valve, then around the VSD (**Figure 39.2a**) and around the aortic valve. Completed VSD patch is shown on **Figure 39.2b**. This way, the mLV will be connected to the aorta and the RV will be connected to the PA using a valved conduit. When both great arteries arise from the mRV (DORV), the aortic valve is further from the VSD, the subaortic tunnel is longer after Rastelli-type operation, the proposed suture line more difficult to map, and the risk of creating subaortic stenosis is higher.

In hearts with mildly obstructed or unobstructed pulmonary blood flow, or with resectable mLVOTO and an adequate sized PV, in which a Senning plus arterial switch operation is proposed, the VSD is closed by direct patching of the defect. There are several techniques for patching a VSD (interrupted pledgeted or continuous sutures, or a combination), but in ccTGA, VSD closure is associated with a higher risk of CHB, either from suture damage to the conducting bundle or from retraction on the anteriorly placed AV node and bundle. When access to the VSD is via the MV the risk of CHB is greatly reduced by gentle anterior retraction, but more importantly by using the technique described by de Leval *et al*. (1979), placing sutures superiorly and left-wardly on the morphological right ventricular side of the septum (**Figures 39.3a,b,c** and **39.4a,b**). For this technique it is probably easier and more reliable to use interrupted sutures. Care should be taken in planning that part of the suture line below the PV, keeping the suture line as inferior as possible and avoiding any shortening of this perimeter of the VSD, in order to reduce the tendency to create distal LVOTO. In the SLL configuration associated with dextrocardia, access through the MV is more difficult and requires more anterior traction. The VSD is, therefore, more easily approached from the left side through the mLA and tricuspid valve. An additional benefit is that the VSD patch is on the morphological right side of the interventricular septum and the conducting bundle is remote from the proposed suture line. Conversely, in the IDD configuration with trans-mitral valve VSD closure, the left bundle branch is inferior, where sutures should be placed in fibrous tissue if possible. Trans-mLA and TV VSD closure (IDD with laevocardia) requires the usual precautions of VSD closure with an inferior conducting bundle.

Persistent Morphological Left Superior Vena Cava (mLSVC)

During the Senning procedure, the systemic venous baffle suture line passes in front of the coronary sinus without danger to the conducting system in SLL configuration (superior AV node and bundle) and the

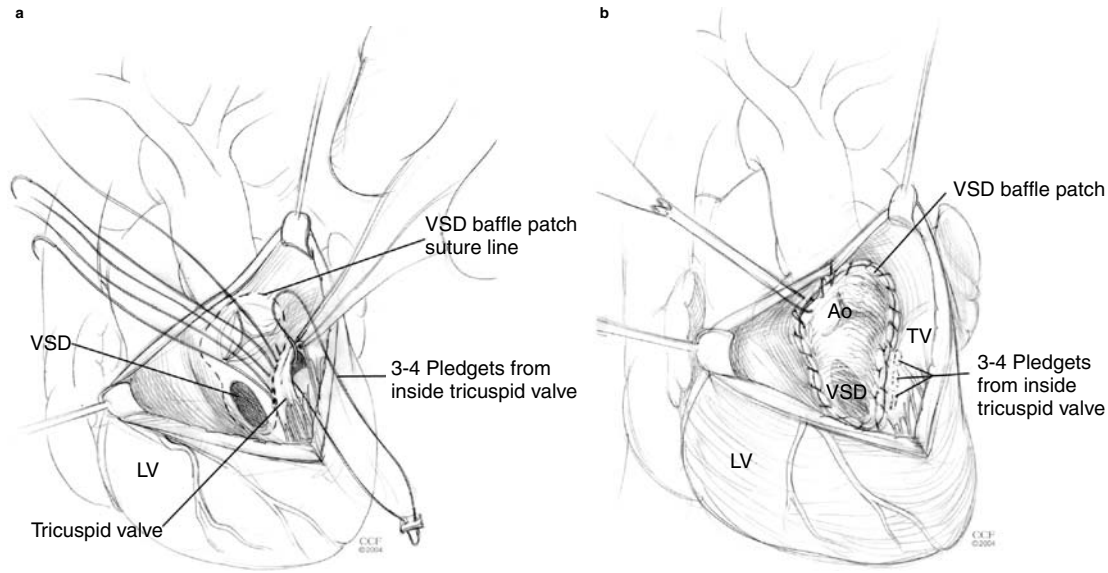

Figure 39.2

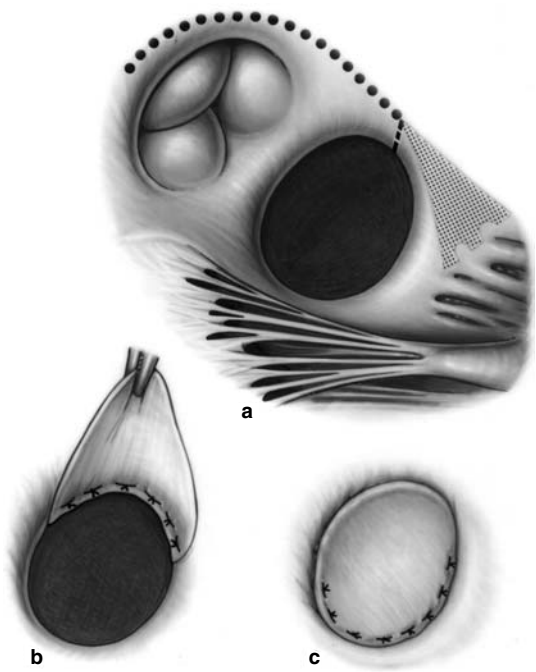

Figure 39.3

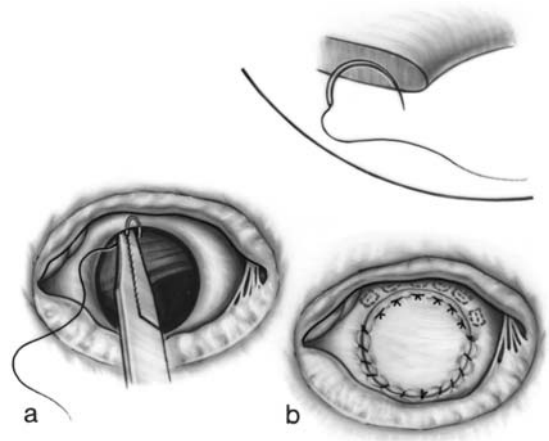

Figure 39.4

coronary sinus drains into the new systemic venous atrium. In IDD configuration with the AV node and bundle lying inferiorly, and in the absence of complete heart block, another strategy is suggested. If the mLSVC is small, or if on clamping the proximal mLSVC pressure remains below 15 mmHg, it can be ligated. Otherwise, the coronary sinus is widely unroofed into the mLA, the coronary sinus ostium sutured closed from within and the systemic venous baffle suture line placed posterior to the closed coronary sinus ostium and the ligament of Todaro. A third approach is to divide the mLSVC and perform a bidirectional cavopulmonary shunt, but this will be directly opposite the distal anastomosis of the

mRV to PA conduit when a Senning/Rastelli is required. The author does not favour this third option.

If the mLSVC opens directly into the mLA, then the posterior suture line of the systemic venous baffle runs in front of the morphological left pulmonary veins (as usual) and curves posterior to the internal orifice of the mLSVC to include the mLSVC in the systemic venous atrium.

Separate Drainage of the Hepatic Veins

A hepatic vein draining separately into the mLA may significantly complicate the posterior suture line of the systemic venous baffle, depending on its spatial relationship to the tricuspid valve annulus and the ipsilateral pulmonary veins. Almost always this would only be seen in a heterotaxy-type heart. The anomalously draining vein can sometimes be excised and transplanted closer to the IVC, or it can be banded to reduce right-to-left shunting and allow time for intrahepatic veno-venous connections to develop. Simple primary ligation is not recommended.

Reconstruction of Distorted Pulmonary Arteries

Previous shunting, PA banding and aberrant ductal tissue may result in important PA bifurcation or branch PA distortions and require accurate reconstruction before completion of the mRV to PA connection. The author's preference is to use fresh autologous pericardium for PA reconstruction, using continuous fine (7-0–5-0) Prolene with small suture bites.

The RV–PA Conduit

The anterior pericardium is opened to the right of the midline in SLL and to the left of midline in IDD to provide a pericardial flap to cover the conduit (**Figure 39.5a,b**). The morphological right ventriculotomy should be placed as far to the left as possible in ccTGA (SLL) and as far to the right as possible in ccTGA (IDD) (**Figure 39.1a,b**). This may make the placement of the VSD baffle a little more difficult, but maximizes the opportunity of keeping the conduit well away from the midline, increasing the safety of the inevitable subsequent conduit changes. The ventriculotomy is made by excising a button of free wall and excising underlying obstructive trabeculae. Occasionally, an obstructing TV papillary muscle needs division for VSD access and then reattachment. The ventriculotomy is sized with graded dilators to easily accommodate the oblique proximal conduit anastomosis, having selected the conduit size from the PA nomogram chart. In the author's opinion, the conduit should always lie to the left of the ascending aorta in ccTGA (SLL) and to the right of the ascending aorta in ccTGA (IDD), irrespective of the position of the ventricular apex. The author's preference is to select a woven Dacron conduit containing a preserved porcine pulmonary valve, 3–4 mm diameter larger than the normal MPA size

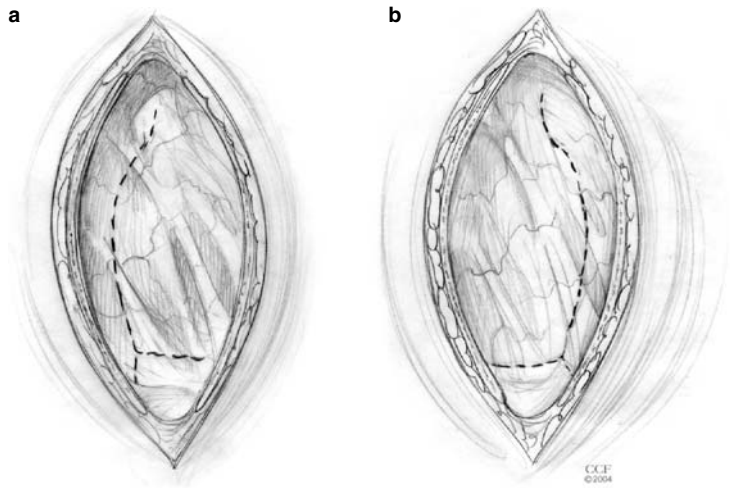

Figure 39.5

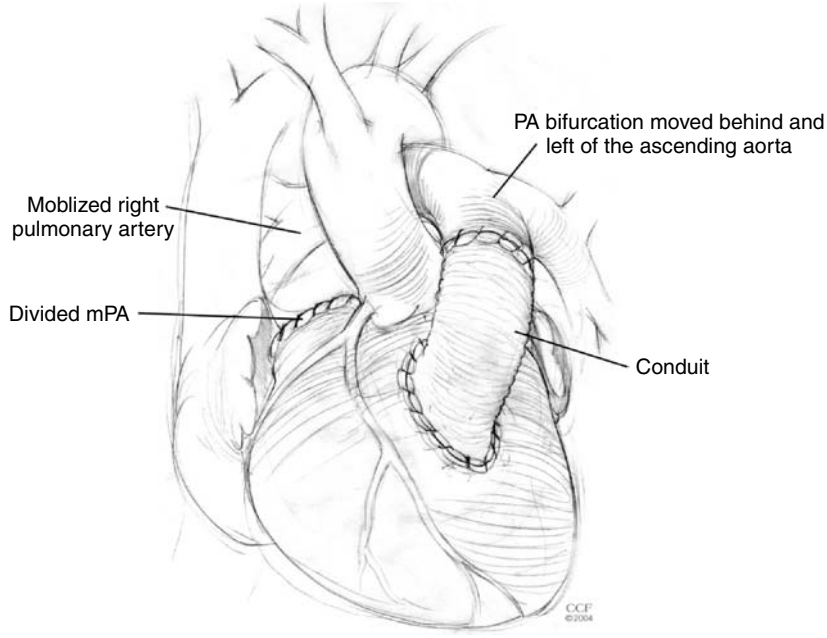

Figure 39.6

for that patient. To create more room for the conduit, the pericardium is split anterioposteriorly down to the mLPA before doing so. This allows easier extension of the PA incision into the mLPA if necessary. Under most circumstances the divided distal MPA has been brought behind the ascending aorta to the side of the mLPA, after mobilization of both branch PAs and MPA bifurcation. The distal conduit anastomosis is achieved with running 5-0 Prolene. The length of the conduit is then carefully judged so as not to lie tense across the morphological right coronary artery (on the left side in ccTGA (SLL), bevelled appropriately and sewn to the right ventriculotomy with running 5-0 or 4-0 Prolene (**Figure 39.6**).

Subsequent conduit changes are easily performed on CPB with the heart beating, shelling out the Dacron conduit, removing all of the distal conduit and leaving a 2 mm Dacron rim superiorly, right-wardly and left-wardly at the proximal anastomosis, to provide a safe sewing ring for the new conduit, remote from nearby and no longer identifiable coronary arteries.

Arterial Switch Procedure

The methodology is very similar to that described in Chapter 36. The coronary arteries are usually ''mirror image'' to those of TGA without AV discordance. In ccTGA (SLL) the mRCA passes to the left and in ccTGA (IDD) the mRCA passes to the right. On the whole, in ccTGA, the aorta and MPA are a little less anterioposterior than in TGA, unless the heart is rotated to the right in ccTGA (SLL) or to the left in ccTGA (IDD). Thus, after the Lecompte manoeuvre the neo-MPA anastomosis is more likely to extend into the mLPA, with partial closure of the contralateral side of the distal MPA. For Lecompte manoeuvre, see **Figure 44.11**.

The Senning Operation with Modifications

There are multiple technical variations used by different surgeons when performing the Mustard or Senning operation. Younger surgeons may have observed few or none of these operations. The author prefers the Senning procedure over the Mustard procedure, because in the former the widest part of the systemic venous baffle is at its midpoint, the proportions are set by anatomical landmarks, with baffle size being dependent on these landmarks and because the right heart border is effectively shifted to the right, broadening the distance between right heart border and the left-ward remnant of the interatrial septum. It is this distance that has to accommodate both pulmonary and systemic venous channels at their midpoint (Jonas *et al*., 1980). The author will focus on some of the technical aspects of the Senning operation and modifications.

The Systemic Venous Baffle

The right atrial incision, when there is concordance between atrial situs and apex of the heart (e.g. SLL with laevocardia), is commenced just in front of the crista terminalis, through a characteristically very thin part of the right atrial wall about one-third of the way from the superior to inferior cavoatrial junctions. The incision is extended superiorly parallel and several millimetres in front of the crista, and then posteriorly to within a few millimetres of the anterior aspect of the superior cavoatrial junction. Looking inside the mRA at the inferior cavoatrial junction, if there is a large, well formed anterior ''valve'', the incision is continued inferiorly parallel to the crista and then swung posteriorly to meet the right-ward attachment of the IVC ''valve'' (Eustachian valve) (**Figure 39.7**). If the ''valve'' is absent, the mRA incision is swung forward as one proceeds inferiorly and then posteriorly at the level of the inferior cavoatrial junction to the right-ward aspect of this junction, thus providing a longer free edge to compensate for the absence of the IVC valve. When there is discordance between atrial situs and ventricular apex (e.g. SLL dextrocardia) and the mRA free wall is narrow, this initial mRA incision is similar in its superior one-third, but runs within 3–4 mm of the anterior atrio-ventricular junction in order to harvest as much mRA free wall as is available to dedicate to the systemic venous baffle (**Figure 39.8**).

A counter-incision is made in front of the right pulmonary veins after developing the right interatrial

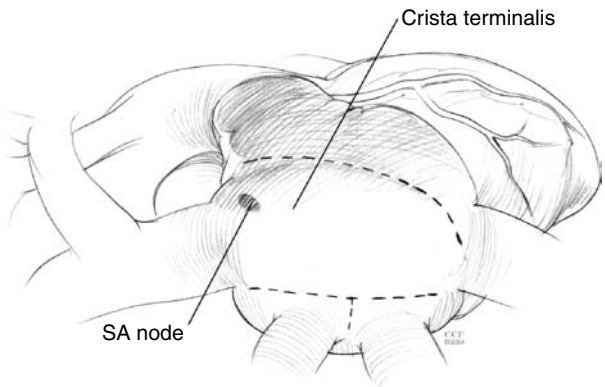

Figure 39.7

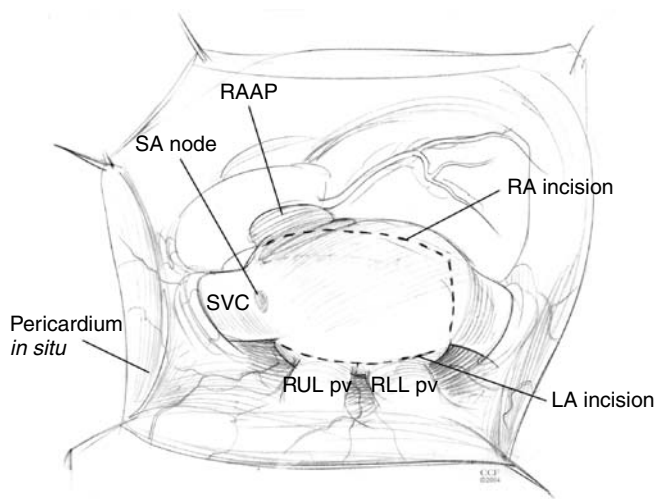

Figure 39.8

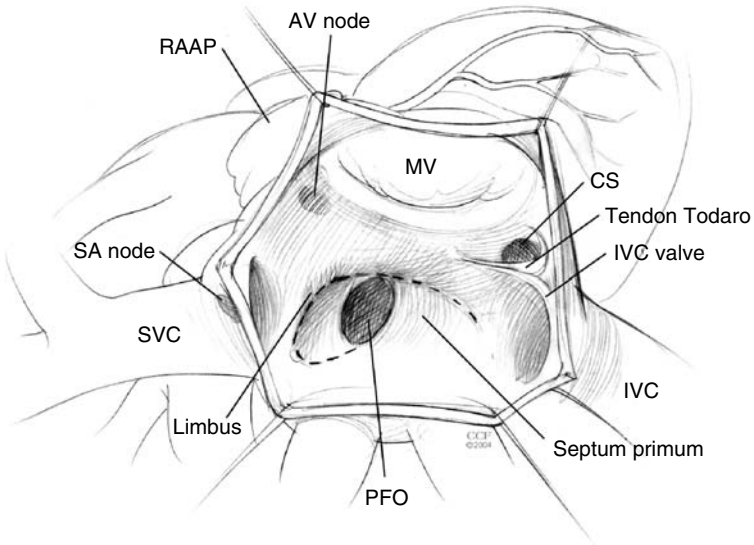

Figure 39.9

groove to the upper border of the superior pulmonary vein and the inferior border of the inferior pulmonary vein, and a short cut back made in the posterior edge of this incision to a point just in front of the two pulmonary veins, effectively lengthening this edge and enlarging the available aperture for the waist of the pulmonary venous atrium (**Figure 39.7**).

The remnant of septum primum is then mobilized left-wardly and inferiorly and superiorly (in the absence of a PFO), remaining hinged right-wardly. A portion of the limbus is then excised (**Figure 39.9**) to enhance SVC drainage towards the mTV. The hinged interatrial septum is then sewn along into place **(dotted line in Figure 39.10**) using two-thirds of its free edge superior to point A (**Figure 39.10**). The length of left atrial wall inferior to point A will be significantly longer than the inferior one-third of the atrial septal flap and must be gathered to the atrial flap, using longitudinal bites in the mLA wall and horizontal bites in the atrial septal flap. This is particularly true in the presence of TR and mLA enlargement.

The completion of the posterior suture line of the systemic venous baffle is shown in **Figure 39.11**. The anterior suture line of the systemic venous baffle is shown by the dotted line (**Figure 39.11**; note points B, B′ and C, C′). It is completed (**Figure 39.12**) running anterior to the coronary sinus in SLL but not in IDD, where the suture line runs posterior to the coronary sinus to avoid atrio-ventricular nodal damage. The anterior suture line is commenced inferiorly. Superiorly, if sewing into the crista terminalis, include no more than the inner one-third of this muscle bundle in the suture line to preserve the superficially placed SA node. Preferably, suturing into the crista is avoided altogether.

The Pulmonary Venous Atrium

In concordant atrial situs and ventricular apex position (SLL laevocardia, IDD dextrocardia) there is enough right atrial free wall to complete the pulmonary venous atrium. In **Figure 39.12**, note points D, D′ and E, E′ on the prospective suture line. The distance from D′ to the apex of the suture line inferiorly is shorter than that of D to the apex, to preserve as much length as possible of the mRA free edge for the upper part of the pulmonary venous suture line. The distance from E′ to the superior apex of the suture line is longer than the distance of point E from the superior apex, to avoid bowstringing across and constriction of the lateral wall of the SVC. **Figure 39.13** shows the completion of the pulmonary venous atrial suture line.

In discordant atrial situs and ventricular apex configuration (SLL dextrocardia, or IDD laevocardia), when the mRA free wall is narrow (**Figure 39.14**) the pericardium *in situ* is used to complete the pulmonary venous atrium (Schumacker, 1961). Note points F, F′, G, G′ and H, H′ in **Figure 39.14** on the dotted prospective suture line. Prior to commencing the pulmonary venous atrial suture line, some of the excess tissue anterior to the right pulmonary veins is excised. **Figure 39.15**. shows the pericardium *in situ*, turned forward to illustrate the

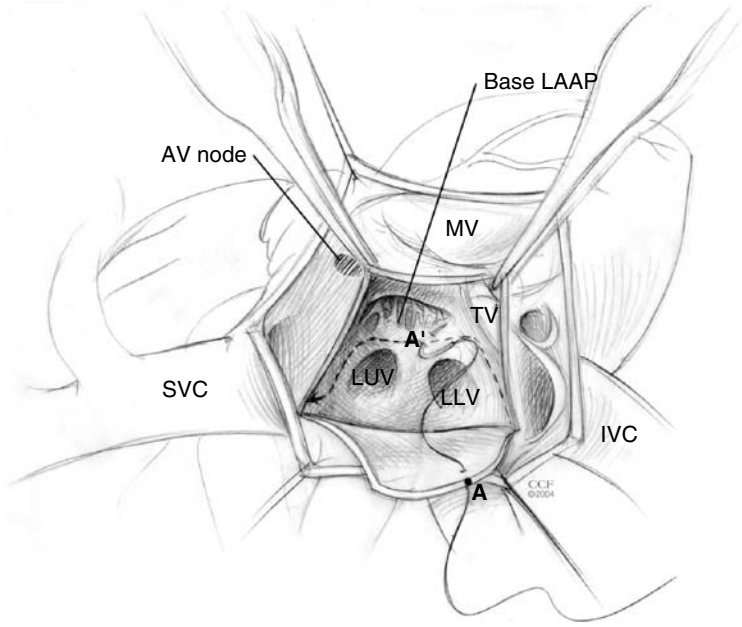

Figure 39.10

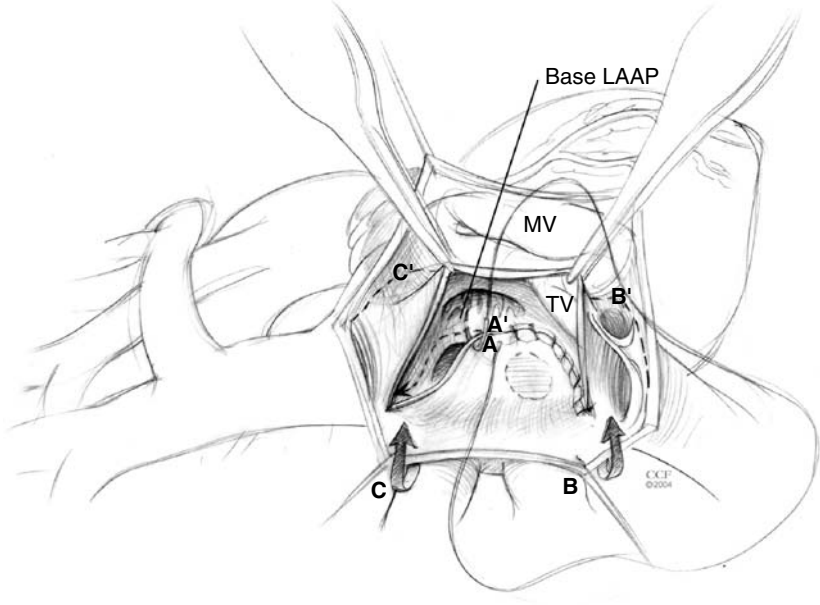

Figure 39.11

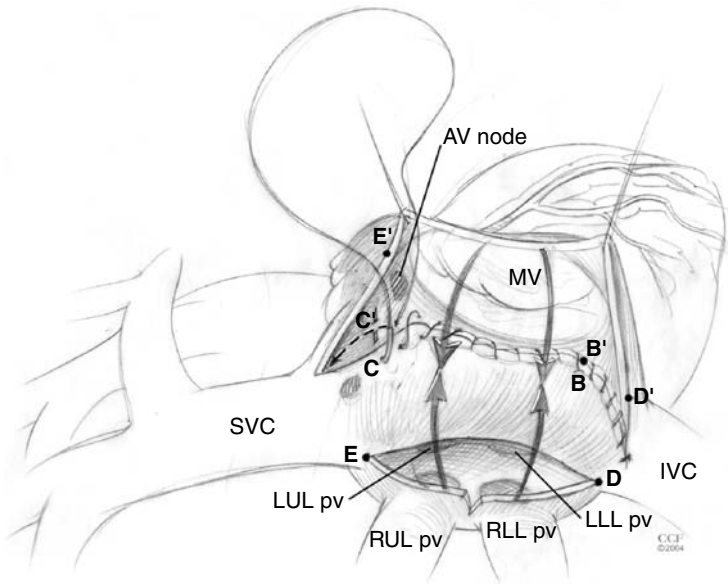

Figure 39.12

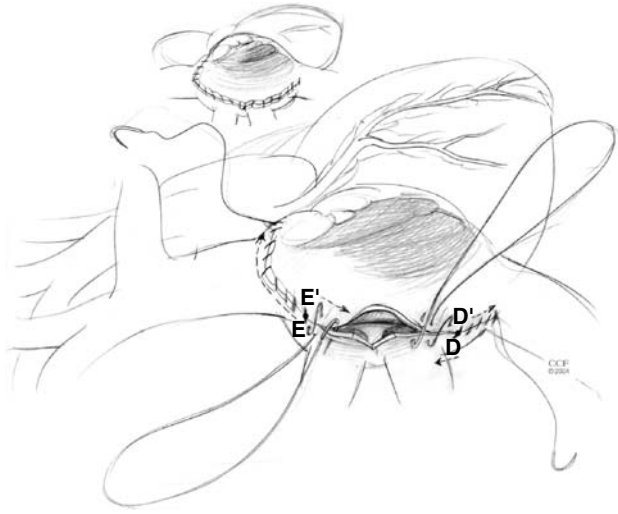

Figure 39.13

partial thickness pericardial sutures sparing the phrenic nerve on the pleural side.

SURGICAL PREPARATION AND METHOD

The patient is anaesthetized, prepped and draped, with lines inserted in the usual fashion. Midline sternotomy is performed, the pericardium is opened to one side if a conduit is to be inserted, leaving an anterior flap to cover the conduit and separate it from the sternum at the time of closure (**Figure 39.5a,b**).

If an arterial switch is to be performed, the required pericardial patches are harvested with stay stitches in

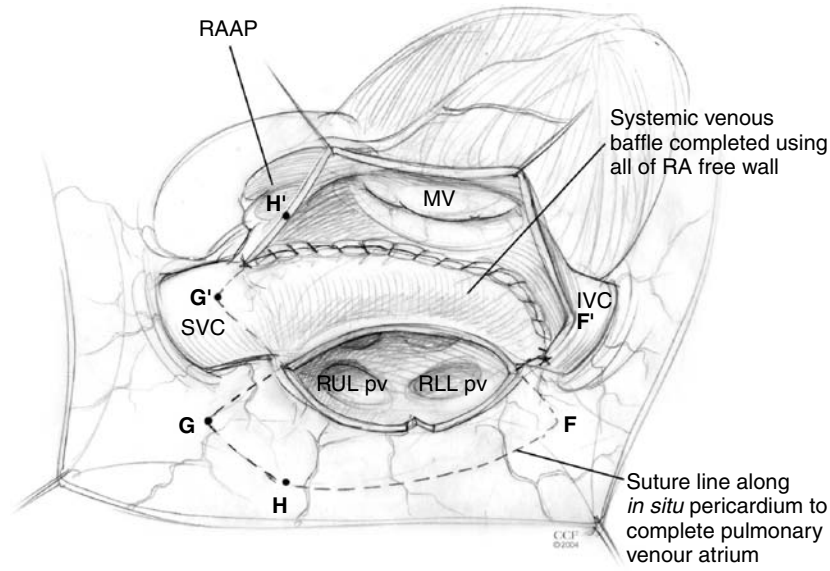

Figure 39.14

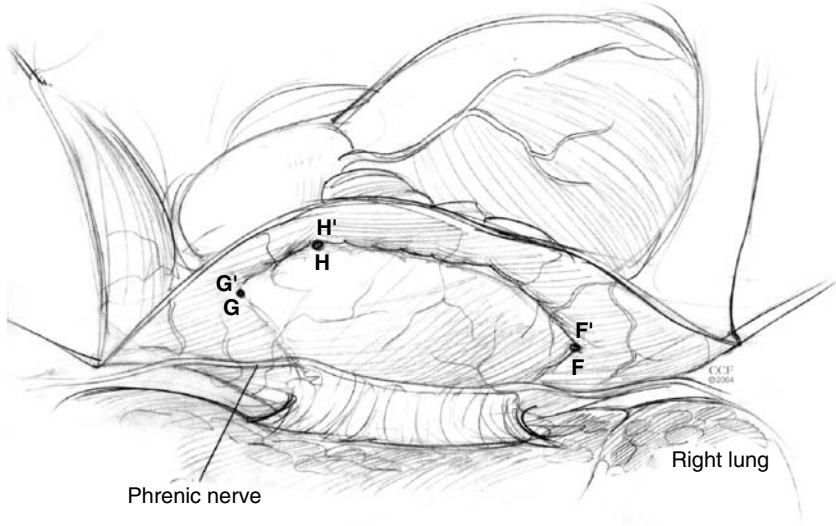

Figure 39.15

each corner and stored in cold lactated Ringer's solution. Pericardial adhesions are taken down under tension with low-voltage coagulation diathermy, and using the same technique the ascending aorta is completely separated from the MPA. The duct/ligament is divided and MPA bifurcation and branch PAs mobilized for both kinds of double switch. Shunts or PAB are dissected out, heparin given and bypass cannulae placed high in

the ascending aorta, high in the SVC and low in the IVC. CPB is commenced, cooling the patient to 22°C nasopharyngeal temperature. During cooling the PAB is removed, shunts divided between silk ligatures and dissection of adhesions and PA branches completed. The morphological right interatrial groove is developed in preparation for the Senning. The aortic cross-clamp is applied and cardioplegia solution infused into the aortic root (see Chapter 36 for details of the arterial switch operation), during which the initial Senning incision is made in the mRA, and the interatrial septum is mobilized medially, superiorly and inferiorly. The VSD is patched via the mRA and MV or via the mLA and TV (Senning and ASO) or via the morphological right ventriculotomy (Senning/Rastelli) using a stretch-knitted Dacron patch. Cardioplegia solution is administered at 20 min intervals. The Senning operation is performed with or without modifications. The tip of the mRA appendage is amputated and a purse-string with snugger applied. If we are performing a Senning/Rastelli operation, rewarming is commenced, the MPA divided and the proximal end oversewn, the heart de-aired and the aortic cross-clamp removed. Pulmonary artery reconstruction and conduit insertion is achieved with the heart empty and beating. For a Senning/arterial switch operation, cardioplegia solution is infused just before transecting the aorta, the open-heart sucker remains in the mRA appendage and cardioplegia is repeated using hand-held cannulae after 20 min. With completion of the neo-aortic suture line, the heart is de-aired and the cross-clamp removed. Reconstruction of the neo-pulmonary artery is performed with the heart beating—either spontaneously or by pacing with temporary pacing wires. In all cases a pulmonary venous atrial pressure line is placed through the mRA appendage, and if indicated a fine PA pressure line is inserted through the neo-MPA suture line. Two atrial and two mRV temporary pacing wires are applied. The patient is weaned from CPB. A high LA pressure is assumed to be due to coronary kinking or compression until proved otherwise. Haemostasis usually takes some time, and intravenous clotting factors other than fresh frozen plasma are not used unless absolutely necessary. The chest is closed in a routine fashion over two mediastinal drains and usually a Tenckhoff peritoneal dialysis catheter is placed in the abdominal cavity. Trans-oesophageal echocardiography is performed to assess contractility of the ventricles, atrio-ventricular valve function, semi-lunar valve function, patency of ventricular outflow tracts, the Senning pathways and any residual interventricular or interatrial defects. There is a low threshold for repeating echocardiography postoperatively for any haemodynamic concerns. Postoperative weaning from the ventilator is not attempted during the ensuing night and is subsequently considered after establishing good haemodynamic and pulmonary stability. Low-dose heparin (6–10 units/kg/h) infusion is commenced postoperatively as soon as the chest drain effluent becomes clearly serosanguinous. Low-dose aspirin (3–5 mg/kg/day) is commenced on the first postoperative day, and for an arterial switch a nitroglycerine patch (18 h on, 6 h off) is commenced the day after surgery. The LA pressure line is maintained patent with low-dose heparin in saline infusion, which is discontinued on day 5 postoperatively and the line pulled a few hours later.

ATRIO-VENTRICULAR DISCORDANCE WITH DOUBLE-OUTLET RIGHT OR LEFT VENTRICLE

Double-outlet right ventricle may coexist with atrio-ventricular (AV) discordance. Most such hearts have associated VSD and pulmonary or subpulmonary stenosis. The first description of this combination of lesions was published by Monckeberg (1913); the first surgical repair was reported by Kiser *et al*. in 1968.

The position of the conduction system may differ from that in ccTGA. Both posterior and anterior AV nodes may be present. They may be connected by a sling of conduction tissue around the VSD (Losekoot *et al*., 1983). In some patients, only the anterior or posterior AV node continues as a penetrating bundle. For these reasons, it may be safer to place all stitches on the morphological right side of the septum, as occurs when the combined Senning/Rastelli operation is performed. The intraventricular baffling of the VSD to the aorta is also performed through the ventriculotomy in the mRV, but is more difficult than in ccTGA. A longer tunnel is required, with the aorta displaced further from the VSD. The back wall of the tunnel may require partial resection to avoid its protrusion into the tunnel pathway. A conduit is then placed between the mRV and divided distal MPA.

Double-outlet left ventricle may also coexist with AV discordance (Brandt *et al*., 1976; Subirana *et al*., 1984). It may occur with situs solitus or situs inversus, but this combination of lesions is rather rare. In repair, the VSD is closed from the mRV side, the MPA divided, the proximal end oversewn and a conduit placed from mRV to distal MPA. In both DORV and DOLV with AV discordance, the anatomical correction is completed by performing a Senning or modified Senning procedure.

ISOLATED VENTRICULAR INVERSION

AV discordance with ventriculo-arterial concordance (isolated AV discordance, or isolated ventricular inversion) is a very rare congenital heart anomaly (Quero-Jimenez and Raposo-Sonnenfeld, 1975; Arciprete *et al*., 1985). It is usually associated with VSD (Van Praagh and Van Praagh, 1966). The haemodynamics are similar to those of TGA (AV discordance with ventriculo-arterial discordance). Repair can be achieved by atrial repair (Mustard or Senning operation), which is appealing because the morphological left ventricle remains a systemic ventricle and the morphological right ventricle remains a subpulmonary ventricle.

The VSD is closed through the mRA, but if this is difficult because of a discordant relationship between the atrial situs and side of the ventricular apex, the VSD is approached through the mLA and TV prior to performing the Senning operation. The position of the conducting system is uncertain in this rare anomaly, which is most commonly seen in heterotaxy with additional anomalies of systemic and pulmonary venous connection. It is wise to assume that the AV node and penetrating conducting bundle could be superior or inferior and plan sutures to avoid both these sites during VSD closure and in performing the Senning operation.

RESULTS

Physiological Repair

For physiological repair of AV discordance and VA discordance or malposition, the results had already improved since the early 1980s (McGrath *et al*. 1985). Ninety patients were operated on at Great Ormond Street Hospital for Sick Children with AV and VA discordance during 1975–1990. There were 13 in-hospital deaths (14%) and four late deaths (4.5%). The early and late mortality rates were higher in the earlier years (1975–1984) (22% and 6%, respectively). During 1985–1990, 39 patients underwent operations; two died early (5.1%) and there was one late death (2.6%).

At the University of Alabama at Birmingham (Kirklin and Barratt-Boyes, 1986), 75 patients underwent repair during 1967–1983; there were seven early deaths (9%) and 10 late deaths (15%). Williams *et al*. (1981) and Westerman *et al*. (1982) reported in-hospital mortality rates of 9% in their respective series. The risk of operation seems to be higher in patients with AV discordance and double-outlet right ventricle. Kirklin and Barratt-Boyes (1986) reported 16 patients, of whom six (38%) died. Tabry *et al*. (1978) reported 20 patients, or whom three (15%) died. Great Ormond Street Hospital has operated on 17 such patients; four (24%) have died.

The risk of complete heart block remains higher in AV discordance than in other congenital heart defects. Incidences between 17% and 46% have been reported (Kirklin and Barratt-Boyes, 1986; Hwang *et al*., 1982; Westerman *et al*., 1982). In the Great Ormond Street Hospital series, five patients had complete heart block before surgery. Of 85 patients at risk, 17 (20%) developed complete heart block after the operation. The date of operation was an important risk factor. Of 44 patients at risk during 1975–1983, 11 (25%) developed complete heart block, whereas of 41 at risk during 1984–1990, six (15%) developed complete heart block. McGrath *et al*. (1985) reported a preoperative incidence of complete heart block of 11%. Twenty-three patients (26%) developed complete heart block during surgery.

Late results were analysed in detail by the Birmingham, Alabama, group (McGrath *et al*., 1985). One year and 10 year actuarial rates of survival among their 99 patients were 75% and 68%, respectively. The factors for premature death were double-outlet right ventricle, preoperative or perioperative complete heart block, young or old age at operation, high haematocrit levels, and early date of operation; 57 (91%) of 70 surviving patients were in New York Heart Association class I, and 12 (17%) were in class II. Williams *et al*. (1981) reported a late mortality rate of 19% with an average follow-up period of 4.4 years. Reoperation was required in nine of 32 survivors, and the rate of mortality from this procedure was 33%. The most common reason for reoperation was tricuspid valve incompetence (seven of nine reoperations).

From The Royal Children's Hospital, Melbourne (Sano *et al*., 1995), 27 patients with ccTGA had undergone VSD closure at age 1–10 years, with two early deaths (7%). At mean follow-up of 72 months (6–84 months) there were three late deaths (11%). Four of 19 patients with serial echocardiogram had moderate or severe mRV dysfunction, and seven of these 19 patients had moderate or severe tricuspid valve incompetence. This analysis reinforced our programme of pursuing the double-switch approach, which had already been applied to patients presenting with a large VSD associated with pre-existing TR and RV dysfunction. A study documenting progressive TR in ccTGA was published by Prieto *et al*. (1998).

Anatomical Repair (Double Switch)

A total personal series of 73 double switches were performed from July 1989 to July 1993 at The Royal

Children's Hospital, Melbourne, and from July 1993 to September 2004 at The Cleveland Clinic Foundation. The age range was 1 week–16.3 years. There were 47 Senning/arterial switch and 26 Senning/Rastelli operations. The segmental configuration was SLL in 67 and IDD in six. Twenty-seven of 67 patients with ccTGA (SLL) had dextrocardia and six with mesocardia. Of the six patients with ccTGA (IDD) four had laevocardia and one had mesocardia. Sixty-one patients (84%) had a VSD. Of the 36 patients with mLVOTO (PVS in 19, pulmonary atresia in 17) 10 patients had adequate-sized pulmonary valves and correctable mLVOTO, permitting a Senning/arterial switch operation. Six of 73 patients had coarctation of the aorta or interrupted aortic arch. Two patients had small pulmonary arteries and MAPCAs requiring staged unifocalization prior to the Senning/Rastelli operation. Four patients already had pacemakers for complete heart block, two of which resulted from previous VSD closure. Three patents had pacemakers placed following the double-switch operation, one of whom had unpaced complete heart block preoperatively. One of two patients with double-switch-induced CHB returned to sinus rhythm 1 month after pacemaker insertion.

Twenty-four patients had prior PAB to control CHF in 13 and to retrain to MLV in 11. Previous shunting was performed in 25 patients, 22 with systemic-to-pulmonary shunts and three with cavopulmonary shunts. One of the PAB patients had repair of total anomalous pulmonary venous return at the same time. Two patients required unifocalization of MAPCAs before double-switch.

The double-switch is a long operation. For the Senning/ASO, the mean CPB time was 200 min and mean aortic cross-clamp time 104 min. A mean of 192 min CPB time and 101 min cross-clamp time was required for the Senning/Rastelli. Median ICU stay was 3.5 days (2–60 days) and median hospital stay 8 days (5–60 days).

There was one 30 day/hospital death at 60 days, an infant who had previously undergone a TV repair and loose PAB and was transferred after 6 weeks on a high-pressure ventilator with ongoing severe TR, recurrent pulmonary haemorrhage, pneumothoraces, and an estimated LA pressure from PFO Doppler velocities of 30–55 mmHg. Post-double switch, very poor lung function eventually led to 14 days of ECMO and then death 60 days after operation. Two patients were placed on LVAD—one electively to retrain a deconditioned mLV and one for small mLV and MV, eventually leading to conversion of the Senning/Rastelli to a Fontan operation. Both LVAD patients are long-term survivors. Early complications include one reoperation for PA bifurcation stenosis, two for mediastinal bleeding, and two temporary tracheostomies. Other complications include sepsis in three, pleural effusion requiring drainage in 10, upper gastrointestinal bleeding in one, one with hemidiaphragm paralysis and one renal failure. Late reinterventions include MV cleft closure in one, SVC dilatation in two, conduit replacement in two, subneo-aortic resection in two and late pacemaker insertion in two.

LATE RESULTS

There have been three late deaths (two from the Senning/ASO group). Two were sudden and perhaps due to coronary events or arrhythmias, and one patient with retrained mLV represented with severe biventricular failure and died after heart transplantation at another centre. Therefore, of the 11 patients undergoing Senning/ASO after LV retraining, one died later after heart transplant, two have mild and one has moderate mLV dysfunction. On the other hand, at the last follow-up one patient has mild and two have moderate mRV dysfunction. Similarly, 14 have trivial, four have mild, three have moderate and two have severe TR.

During the Cleveland Clinic Foundation experience of double-switching, a further 31 patients with ccTGA have been assessed. Eleven have been banded, with two late deaths and nine awaiting full LV retraining. Five patients with end-stage disease have undergone heart transplantation, four patients underwent a Fontan operation for non-septatable heart and 10 adult patients have had either TV replacement or partial mLVOTO resection as a palliation.

Other early series of double-switch operations were reported from Birmingham, UK, by Stumper *et al*. (1995), and from the Tokyo Woman's Hospital by Yamagishi *et al*. (1993) and Imai (1997).

In summary, our double-switch programme has resulted in satisfactory intermediate-term survival, has resulted in improvement in mRV dysfunction and TR in most patients, preserved mLV and MV function (except in three with late mLV retraining patients). With time we anticipate an increasing incidence of atrial arrhythmias, additional cases of late spontaneous CHB, additional cases of late subaortic stenosis in Senning/Rastelli patients, and late neo-aortic valve dysfunction. Conduit changes will be necessary in all the Senning/Rastelli patients eventually, but these same patients would require MLV-to-PA conduit changes if they had undergone physiological repair. An unanswered question is whether or not arrhythmias will be better tolerated with the MLV rather than the MRV in the systemic circuit. The following references refer to the topic of ccTGA and

mLV retraining: Mee and Harada (1991), Ranjit *et al*. (1991), Karl *et al*. (1997), Helvind *et al*. (1998), Imamura *et al*. (2000), Poirier *et al*. (2000), Ando *et al*. (2003) and Duncan *et al*. (2003).

Acknowledgements

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40

Single Ventricle

C. van Doorn and M. R. de Leval

HISTORICAL NOTE

Ever since the masterful description of the circulation by William Harvey in *De Motu Cordis*, the importance of the right ventricular function has always been debated. From the early 1940s, numerous experiments were performed to determine whether the pumping action of the right ventricle is necessary to maintain a normal circulation. Starr *et al*. (1943) noted no significant rise in the venous pressure following the destruction of a large portion of the right ventricle by cautery. In addition, Bakos (1950) demonstrated that there was no fall in pulmonary arterial pressure. Right ventricular exclusion was successfully achieved by Rodbard and Wagner in 1949, who ligated the main pulmonary artery and anastomosed the right atrial appendage to the pulmonary artery. The right ventricle, however, was still thought to contribute to the pulmonary circulation because of functional tricuspid regurgitation. Several other experimental studies confirmed the feasibility of right ventricular exclusion when the right atrium had been prepared to sustain an additional workload. Haller *et al*. (1966) performed, as a first stage, a side-to-side anastomosis of the superior vena cava to the right pulmonary artery and, as a second stage, closed the tricuspid valve so as to achieve a complete bypass of the right ventricle. In these experiments, the assumption was made that the right atrium was used as the pumping chamber for the pulmonary circulation. It was on the same principle of the pumping function of the right atrium that Fontan performed his first atrial pulmonary connection in a case of tricuspid atresia in 1968 (Fontan and Baudet, 1971).

Partial bypass of both the right atrium and the right ventricle was achieved by Carlon *et al*. in 1950, when they described the anastomosis of the superior vena cava to the right pulmonary artery. Cavopulmonary anastomoses were then performed and studied experimentally and clinically by independent groups in the Soviet Union (Galankin and Darbinian, 1956) and the USA (Glenn and Patino, 1954). Haller *et al*. (1966) also studied experimentally the feasibility of anastomosing the superior vena cava end-to-side to the pulmonary artery, thus shunting the superior vena caval blood into both lungs instead of to the right lung only. This bidirectional cavopulmonary anastomosis was then applied clinically by Azzolina *et al*. (1972) as a definitive palliation or in preparation for a Fontan type of operation.

The Fontan concept has been applied to increasingly complex single ventricle circulations (Puga *et al*., 1987) and has become the definitive surgical palliation for single ventricle circulations. The Fontan operation itself has also continued to evolve.

The study of flow characteristics through the reconstructed circulation has led to a complete or near-complete exclusion of the right side of the heart, to achieve the more energy efficient total cavopulmonary connection (de Leval *et al*., 1988). This initially involved the creation of an intra-atrial baffle between the inferior vena cava and pulmononary artery. The lateral tunnel technique remains the procedure of choice in some centres, in particular as the final part of the Norwood strategy. More recently, the use of an extracardiac interposition graft between the transected inferior vena cava and the pulmonary artery (Marcelletti *et al*., 1990) has gained popularity. This technique was originally designed to avoid pulmonary and systemic venous obstruction in patients with a small atrium, but has the additional advantage that it avoids extensive atrial suture lines that are potentially arrhythmogenic. In addition, the procedure can be performed without cross-clamping the aorta and in selected patients also without the use of cardiopulmonary bypass (Uemura *et al*., 1998). Concern about the lack of growth potential of the prosthetic extracardiac conduit has led to further modifications, using either an extracardiac tube of autologous pediculated pericardium (Hvass *et al*., 1992) or direct anastomosis between the inferior vena cava and pulmonary arteries (Yamagishi *et al*., 1997). However, these techniques are currently not in widespread use.

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INDICATIONS FOR THE FONTAN OPERATION

The Fontan procedure, initially described for the treatment of absent right atrio-ventricular connection, has been extended to many complex cardiac malformations with only one well developed ventricle. These include left atrio-ventricular valve atresia, double-inlet left or right ventricle, pulmonary atresia with intact ventricular septum and hypoplastic right ventricle, biventricular hearts with hypoplastic right or left ventricle with ventricular septal defects, with or without straddling atrio-ventricular valve, and as part of the strategy for the treatment of hypoplastic left heart syndrome (hypoplastic left heart syndrome is described in detail in Chapter 41).

SELECTION CRITERIA FOR FONTAN OPERATION

In their initial description of the procedure for patients with tricuspid atresia, Fontan and colleagues have established criteria for selection of patients, known as the ''ten commandments'' (**Table 40.1**). Time, growing experience, and extension of the Fontan procedure to numerous other cardiac malformations have established that these criteria are not as rigid as first estimated.

The age for the Fontan operation has progressively come down, but the minimum and optimal age for the procedure have not yet been established. The operation should perhaps not be performed before a child is walking, or at least crawling, to benefit from the role of the muscular contraction on the venous return. Preoperative sinus rhythm does not appear to be indispensable for a successful outcome. Facilities for sequential pacing in the immediate postoperative period are, however, important. Abnormal systemic venous drainage is compatible with a Fontan type of operation. Pulmonary arteriolar resistance is probably the most important preoperative criterion and should be less than 4 units/m^2. Similarly, significant impairment of the systolic and/or diastolic function of the main ventricular chamber is a significant risk factor for failure of the Fontan circulation. The importance of preservation of ventricular function has now been recognized, and severe or long-standing volume overloading prior to the Fontan circulation should be avoided. The same applies for increased systemic ventricular afterload, such as in the presence of subaortic stenosis or coarctation of the aorta. More than mild atrio-ventricular valve regurgitation is also detrimental for ventricular function. Assessment of the pulmonary vascular bed can be difficult, especially in the presence of accessory sources of pulmonary blood flow, such as aortopulmonary collaterals and systemic-to-pulmonary artery shunts. A mean pulmonary arterial pressure not greater than 15 mmHg has been suggested by Fontan and colleagues; however, this number should not be taken in isolation without knowing the pulmonary blood flow. We believe that patients with a very low pulmonary blood flow and a mean pulmonary arterial pressure of less than 15 mmHg might indeed have too high a pulmonary arteriolar resistance. On the other hand, patients with very large pulmonary blood flow and a mean pulmonary arterial pressure greater than 15 mmHg may be compatible with a successful Fontan circulation. The importance of the size of the pulmonary arteries has been diversely appreciated. In a multivariate analysis of patients who had undergone a Fontan operation at the University of Bordeaux, France, and the University of Birmingham, Alabama (Fontan *et al*., 1989), the dimensions of the right and left pulmonary arteries, expressed as a McGoon ratio, were the most powerful risk factors for death or take-down of the Fontan operation. The McGoon ratio is obtained by summing the diameter of' the immediately prebranching portion of the right pulmonary artery and that of the left, and dividing this sum by the diameter of the descending aorta just above the diaphragm, the measurements being taken during systole. The risks of failure of the Fontan circulation were higher for a McGoon ratio less than 1.8 (Fontan *et al*., 1989). Another expression for the size of the pulmonary vascular bed is the Nakata index, which is derived from the sum of the diameter of the prebranching left and right pulmonary arteries, divided by body surface area. A lower Nakata index has been identified by some (Knott-Craig *et al*., 1993) as a risk factor for Fontan outcome, but not by others (Reddy *et al*., 1996). The usefulness of these indices has been questioned, because they do not take into account the compliance of the pulmonary vascular bed and any distortion of the pulmonary arteries related to previous shunts.

Table 40.1 The 'Ten Commandments' for Selection of Patients for the Fontan Procedure.

1.	Minimum age 4 years
2.	Sinus rhythm
3.	Normal caval drainage
4.	Right atrium of normal volume
5.	Mean pulmonary artery pressure $\leq$ 15 mmHg
6.	Pulmonary arteriolar resistance $<$ 4 units/m^2
7.	Pulmonary artery to aorta diameter ration $\geq$ 0.75
8.	Normal ventricular functions (ejection fraction $\geq$ 0.6)
9.	Competent left atrio-ventricular valve
10.	No impairing effects of previous shunt

SURGICAL PREPARATION FOR THE FONTAN CIRCULATION

In patients born with a single ventricle, the systemic and pulmonary circulations are in parallel with mixing of the circulations within the heart. In the majority of patients, blood flow is preferentially to the systemic circulation, but there may be pulmonary overflow and, rarely are the systemic and pulmonary circulations balanced. In the Fontan circulation, the systemic and pulmonary circulations are separated and placed in series, with the single ventricle connected to the systemic circulation. Therefore, to reach the Fontan state, many patients will need preliminary operations to balance the pulmonary and systemic circulations to improve the natural history of the disease, and to provide the anatomical set-up for the Fontan circuit. Furthermore, other significant haemodynamic abnormalities (such as pulmonary venous obstruction or systemic ventricular outflow obstruction) may have to be addressed. The choice of procedure is guided by the underlying anatomy and pulmonary resistance, both of which are subject to changes over time. Often a series of procedures may be necessary. It is important to remember that inappropriate or poorly performed preliminary procedures can result in loss of Fontan suitability.

Preparation for the Fontan Circulation in the Neonatal and Early Infancy

Systemic to Pulmonary Artery Shunt

If augmentation of pulmonary blood flow is required early in life, when the pulmonary vascular resistance is still high, a systemic-to-pulmonary-artery shunt is performed. In the past, direct connections between the aorta and pulmonary artery were used, but these shunts were abandoned because of unpredictability of shunt flow and a high incidence of pulmonary artery distortion. These risks are less with the currently used prosthetic interposition shunt, which is usually placed between the subclavian or innominate artery and the pulmonary artery (modified Blalock–Taussig shunt). Systemic-to-pulmonary artery shunts are described in more detail in Chapter 16.

Pulmonary Artery Banding

In patients with high pulmonary blood flow, the main pulmonary artery is banded to restrict pulmonary blood flow and to protect the patient from developing pulmonary vascular disease and ventricular dysfunction. Adequate tightness of the band may be difficult to achieve, and a band that is loose will fail to protect against the development of pulmonary vascular disease. Other complications associated with pulmonary artery banding are band migration, pulmonary artery distortion and erosion of the pulmonary artery by the band. Pulmonary artery banding may also promote or exacerbate the development of subaortic stenosis (Franklin *et al*., 1990), thought to be related to myocardial hypertrophy as a result of the combined effect of an increased afterload and a reduction in volume load of the systemic ventricle. This is a major adverse factor for the Fontan circulation (Kirklin *et al*., 1986). Patients at risk are those in whom the aorta arises above a small outlet chamber, such as tricuspid atresia or double-inlet left ventricle and discordant ventriculo-arterial connection, particularly if the ventricular septal defect is small or if there is associated aortic arch obstruction. In these patients, pulmonary artery banding is contraindicated and instead the Lamberti modification of the Damus–Kay–Stansel operation should be performed to establish an unobstructed arterial outflow (Waldman *et al*., 1988). Depending on the pulmonary arteriolar resistance, blood flow to the pulmonary arteries is re-established via a systemic or venous shunt. Alternatively, a Norwood type strategy can be used (see Chapter 41). The technical details of pulmonary artery banding are described in Chapter 17.

Other surgical interventions may also be required, either to provide the anatomical set-up for the Fontan operation or to correct other significant haemodynamic abnormalities, such as obstructed pulmonary venous drainage or coarctation of the aorta.

Over time, pulmonary blood flow will become inadequate because of the fixed diameter of the prosthetic shunt or pulmonary artery band, and further augmentation of pulmonary blood flow may be needed. Many centres will now not wait until the patient becomes symptomatic, but perform early evaluation for suitability for a Fontan circulation once the pulmonary vasculature has matured. In patients with good Fontan characteristics, a Fontan operation may then be done in one stage. In those with less favourable characteristics, and also as a preferential strategy for good Fontan candidates in some units, the Fontan circulation is achieved via a staged procedure. This initially involves a cavopulmonary anastomosis, followed at a later stage by completion of the total cavopulmonary connection.

PHYSIOLOGY OF RIGHT HEART BYPASS OPERATIONS

Cavopulmonary Anastomosis

The fundamental differences and advantages of a systemic venous-to-pulmonary artery shunt over a systemic artery to pulmonary artery shunt are:

1. More desaturated blood (systemic venous rather than arterial) is shunted to the lungs, with therefore

a much greater efficacy on increments of arterial oxygen saturation.
2. Diversion of approximately one-third of the systemic venous return to the lungs reduces the volume load on the heart, whereas an arterial shunt constitutes an additional ventricular volume and workload.

The classical cavopulmonary anastomosis, diverting the superior vena caval blood to the right lung, is performed rarely because it results in right-to-left pulmonary artery discontinuity. If a Fontan type of procedure is to be carried out later, major reconstructive surgery is required to establish continuity between the two pulmonary arteries, or the inferior vena caval blood (~two-thirds of the systemic venous return) has to be diverted to the smaller left lung. We prefer the use of the bidirectional cavopulmonary anastomosis, in which the superior vena cava is connected end-to-side to the (undivided) right pulmonary artery, thus supplying blood to both the right and the left lung. An alternative to the bidirectional cavopulmonary anastomosis is the hemi-Fontan modification, which involves a connection between the right atrial–superior vena caval junction and the pulmonary arteries and patch augmentation of the central pulmonary arteries. This operation has particularly gained popularity in patients with hypoplastic left heart syndrome, where, after the first stage Norwood operation, pulmonary artery hypoplasia and distortion are common. Whether any additional sources of pulmonary blood flow (such as forward flow through a patent right ventricular outflow tract, or flow from natural or surgically created systemic-to-pulmonary artery shunts) should be taken down at the time of the cavopulmonary shunt remains open to discussion. Retrospective studies have shown that the presence of additional blood flow offers no difference in long-term survival and suitability for completion of the Fontan circulation (van de Wal *et al*., 1999; Berdat *et al*., 2005). If preoperative investigation revealed any other adverse anatomical features for the Fontan circulation, such as subaortic obstruction, this should be addressed at the time of the cavopulmonary anastomosis, or earlier if necessary.

The criteria for successful bidirectional cavopulmonary anastomosis are not uniformly agreed upon, but it is generally accepted that they are less rigid than for a Fontan type of procedure. The bidirectional cavopulmonary shunt, often combined with additional reconstructive procedures, has provided excellent palliation in high-risk Fontan candidates and has also been useful in reducing risk factors for subsequent Fontan completion (Bridges *et al*., 1990a). However, the routine staging of good risk candidates for the Fontan procedure remains open to discussion.

We currently perform a bidirectional cavopulmonary anastomosis as either a temporary or a definitive palliation in cases that are less than optimal for a Fontan operation. This includes:

1. Young patients ($< 1–2$ years).
2. Patients requiring extensive pulmonary arterial reconstruction.
3. Patients with subaortic obstruction with severe systemic ventricular hypertrophy.
4. Patients with borderline pulmonary vascular resistance or systemic ventricular function.

Patients with interrupted inferior vena cava and azygos continuation (usually left isomerism) are particularly good candidates for a bidirectional Glenn procedure, as the superior vena cava carries all the systemic venous blood except the hepatic and coronary venous return (Kawashima *et al*., 1984). It is not uncommon for these patients to have two superior vena cavae, in which case a bilateral bidirectional cavopulmonary anastomosis has to be constructed.

As might be expected, the late problems after the cavopulmonary anastomosis are similar to those described previously after classical cavopulmonary anastomosis: (a) preferential perfusion of the lower part of the lungs; (b) development of venous collaterals between the superior and the inferior vena caval territories; (c) development of pulmonary arteriovenous fistulae; and (d) progressive increase in pulmonary vascular resistance due to rising haematocrit. In addition, it appears that the efficiency of a bidirectional cavopulmonary anastomosis decreases with age. This probably relates to the fact that the cerebral blood flow is proportionally greater in the younger child.

Profound cyanosis early after the Kawashima procedure is also a well-recognized complication and may, at least in part, be related to abnormal systemic venous connections in isomeric hearts. In one reported case it was found to relate to abnormal intrahepatic systemic-to-hepatic venous communications, and saturations increased significantly after trans-catheter occlusion of the abnormal connections (Slavik *et al*., 1995). In another instance there was early development of diffuse pulmonary arteriovenous fistulae (Knight and Mee, 1995). Redirection of hepatic venous blood to the confluent pulmonary arteries led to regression of the fistulae.

Atrio-ventricular Connection

Atrio-ventricular connection used to be performed on hearts with a subpulmonary ventricular chamber, assuming that it might contribute positively to the circulation. This was the case for diminutive right ventricle in absent right atrio-ventricular connection (tricuspid atresia) or

small right ventricle in pulmonary atresia with intact ventricular septum. By and large, the atrio-ventricular connection has now been abandoned because of poor late haemodynamic performance.

Atriopulmonary Connection

Fontan and Baudet (1971) described with great precision the mechanical requirements for a contractile chamber to act as a pump, viz. a power source generating sufficient stroke work, an inlet valve, and an outlet valve. It soon became evident that inlet and outlet valves were dispensable, and that the non-hypertrophied right atrium could also be incorporated successfully into the Fontan circulation.

However, the efficiency of the atrium as a pump in the Fontan circulation has also been questioned. There is no evidence of step-up in pressure energy between the atrium and the pulmonary artery. Furthermore, our studies in hydrodynamic models have revealed that the atriopulmonary connection performs poorly in terms of flow energetics, because of turbulence that is further exacerbated by pulsation (de Leval *et al*., 1988). In addition, long-term atrial distension contributes to atrial arrhythmias, thrombo-embolic complications and further deterioration of the haemodynamics that can lead to failure of the Fontan circulation.

Total Cavopulmonary Connection (TCPC)

Total cavopulmonary connection consists of driving the superior vena caval blood directly into the pulmonary arteries (bidirectional cavopulmonary anastomosis or hemi-Fontan) and channelling the inferior vena caval blood to the pulmonary artery through a lateral tunnel, an intra-atrial tunnel, or an extracardiac conduit. The advantages of the total cavopulmonary connection are better flow dynamics, and reduction of the risk of thrombosis and arrhythmias due to less atrial distension.

SURGICAL TECHNIQUES

Superior Cavopulmonary Anastomosis

Bidirectional Cavopulmonary Anastomosis

We prefer to do the bidirectional Glenn procedure on cardiopulmonary bypass from a midline sternotomy. Most of the right lobe of the thymus gland is removed to expose the innominate vein. The superior vena cava is fully mobilized. Attention must be paid to the right phrenic nerve during dissection. The azygos vein is ligated or divided between ligatures to prevent a run-off from the higher pressure SVC to the lower-pressure IVC. If there is interruption of the IVC with azygos continuation, the azygos vein must be kept patent. The main and right pulmonary arteries are then dissected free

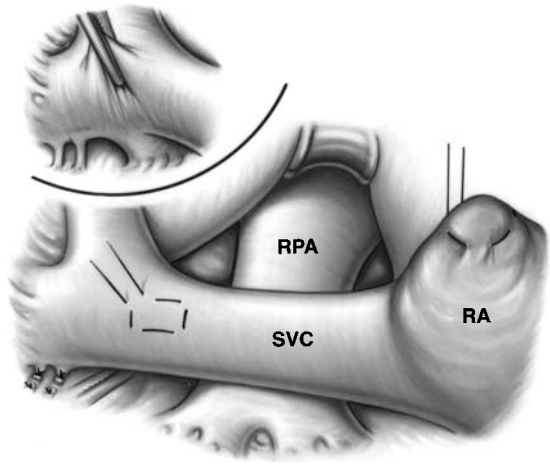

Figure 40.1

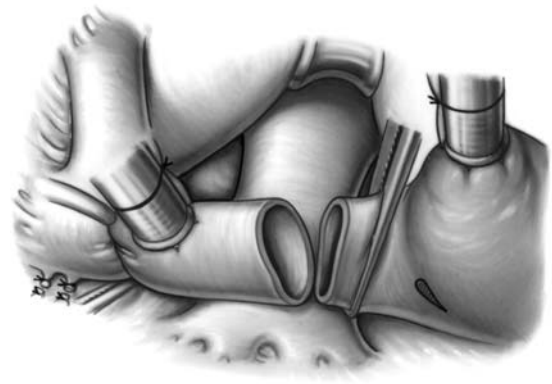

Figure 40.2

from the surrounding tissues (**Figure 40.1**). If present, the shunt is also dissected and closed through the posterior pericardium after cardiopulmonary bypass has been established. It is useful to place marking sutures on the SVC and right pulmonary artery before bypass, because after the vessels have been divided it may be difficult to re-establish their normal alignment, and twisting or kinking of the anastomosis may occur. The aortic cannulation purse-string suture is placed as usual. One purse-string suture is placed on the right atrial appendage, and a second on the SVC near the innominate vein. The aorta is cannulated after full heparinization. A cannula is introduced into the right atrium through the right atrial appendage. Cardiopulmonary bypass is initiated, and the SVC is cannulated with a right-angled venous cannula (**Figure 40.2**). The operation is performed on the beating heart with mild hypothermia.

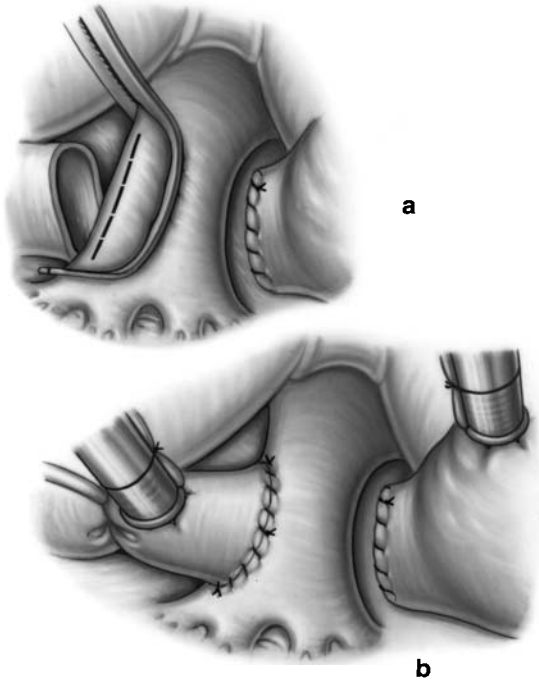

Figure 40.3

If present, a shunt is now occluded. A snare is passed around the SVC and snugged down onto the venous cannula. Alternatively, the SVC is occluded with a vascular clamp just proximal to the cannula. If there is azygos continuation of the IVC, the site of occlusion of the SVC must be proximal to the azygos connection. This may significantly reduce the length of the vein available for the anastomosis, and there may be a risk of obstructing the azygos return during the operation. A vascular clamp is then applied immediately above the cavoatrial junction; care must be taken not to damage the sinus node. The SVC is divided immediately above the clamp, and the cardiac end of the SVC is oversewn with a continuous suture of 6-0 Prolene. The clamp is released. The mobilization of the SVC and the right pulmonary artery and its branches is then completed. In order to perform the anastomosis in a bloodless field, a large side-biting clamp is applied over the superior aspect of the right pulmonary artery, and a long incision is made on the right pulmonary artery from its origin to its branching (**Figure 40.3**) If a right shunt is present, it is disconnected from the pulmonary artery and its proximal end is oversewn with a fine Prolene suture. The opening in the right pulmonary artery is then extended centrally. If the shunt has been placed near the right upper lobe artery, it is sometimes necessary to perform a pericardial patch angioplasty of the right pulmonary artery before the cavopulmonary anastomosis. The cavopulmonary anastomosis is then carried out with a running suture of 6-0 Prolene, which is interrupted in two areas to avoid a purse-string effect of the suture line and to maintain a wide anastomosis (**Figure 40.3**). The patient is rewarmed, and the bypass is discontinued. Additional sources of blood supply, such as the forward flow across a stenosed pulmonary outflow tract or a left systemic-to-pulmonary artery shunt, are usually left patent to maintain higher systemic oxygen saturations. In the case of bilateral superior vena cavae, it is necessary to do a bilateral bidirectional cavopulmonary anastomosis, unless there is a large bridging vein and one of the two cavae is small. Both cavae must be cannulated separately. The hemi-azygos vein is ligated unless there is hemi-azygos continuation of the IVC, and the left cavopulmonary anastomosis is constructed as described above for the right side.

Some surgeons prefer to avoid direct cannulation of the SVC, particularly if the vessel is small, and to perform the cavopulmonary anastomosis during deep hypothermic circulatory arrest. Another technique for the construction of a superior cavopulmonary anastomosis is the use of a temporary shunt between the superior vena cava and the right atrium, thus avoiding the use of cardiopulmonary bypass and its deleterious effects.

Hemi-Fontan Operation

The description and illustrations of this procedure are included in Chapter 41 (Hypoplastic Left Heart Syndrome).

Total Cavopulmonary Connection

Intracardiac Connection

A bidirectional Glenn shunt may already be in place, otherwise the beginning of this operation is similar to that described for bidirectional Glenn procedure using cardiopulmonary bypass. A piece of pericardium is harvested in case of its need as an onlay patch on the pulmonary artery. The SVC is fully mobilized, taking care not to damage the right phrenic nerve. The azygos vein is also dissected and may be later temporarily occluded with a bulldog clamp during the anastomosis of the SVC to the pulmonary artery. The main pulmonary artery and its bifurcation, the right and left branches, are circumferentially dissected and mobilized beyond the lateral pericardium and their branching. The ligamentum arteriosum is doubly ligated and divided. If present, the systemic-to-pulmonary artery shunts are controlled in the usual manner. A right shunt will be disconnected from the pulmonary artery, as described above. The ascending aorta is cannulated.

The cavae are cannulated with right-angled cannulae superiorly near the innominate vein and inferiorly at the cavoatrial junction. Cardiopulmonary bypass is initiated, and the patient is cooled to 28–30°C. The dissection of the pulmonary arteries and the shunts is completed if necessary. A snare is snugged around the SVC, or alternatively the vein is controlled with a vascular clamp placed just proximal to the cannula. Another vascular clamp is applied just above the SVC/atrial junction, and the SVC is transected. The bidirectional cavopulmonary anastomosis is then constructed on the beating heart, as described earlier. When the anastomosis is nearly completed, the patient is cooled to 28°C. The aorta is cross-clamped, and cold cardioplegic solution is infused. A needle vent is placed in the ascending aorta. The main pulmonary artery is transected. To prevent bleeding and aneurysm formation, the proximal stump is closed with a double row of continuous 5-0 polypropylene suture, reinforced with two small Teflon felt strips, and the pulmonary valve leaflets incorporated in the suture line. If the distal main pulmonary artery is not used for the cavopulmonary connection, it is closed with a patch, so as not to narrow or distort the branch pulmonary arteries. The right atrium is then opened. The atrial septal defect is left open or is sometimes enlarged when the left atrio-ventricular valve is atretic or stenotic, to allow the pulmonary venous return to reach the right atrio-ventricular valve in an unobstructed fashion. The intra-atrial cavo-caval pathway may consist of the interposition of a conduit of polytetrafluoroethylene (Gore-Tex) between the caval orifices, or the creation of a composite conduit made of the lateral atrial wall and a prosthetic patch, or the construction of an atrial baffle, establishing the continuity between the IVC and SVC.

1. ***Conduit insertion***. The right atrium is opened along the crest of the appendage. The bypass flow is reduced to quarter flow. The IVC cannula is clamped and removed and the IVC snare released. A sump sucker is placed into the IVC through the purse-string suture of the cannulation site (**Figure 40.4**). This allows good visualization of the IVC orifice from within the right atrium. The length of the atrial conduit is measured between the Eustachian valve and the crista terminalis. The conduit should be as short as possible. A Gore-Tex prosthesis of at least 18 mm is then sewn in the IVC at its junction with the right atrium, with a running suture of 4-0 or 5-0 polypropylene. The conduit is then trimmed obliquely and shortened, to be sewn on the prominent ridge made by the crista terminalis in front of the SVC (**Figure 40.5**). The right atriotomy is then closed with a running 5-0 Prolene suture.

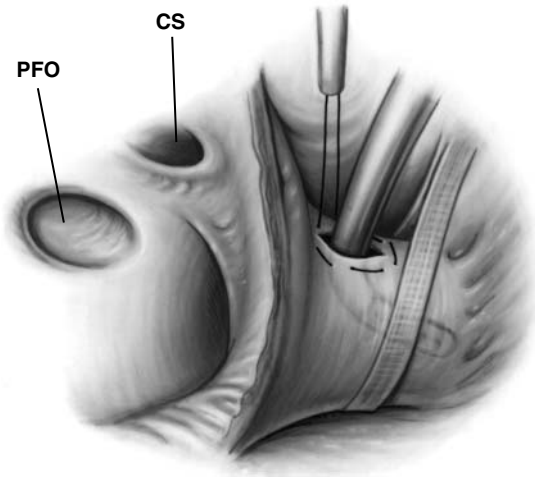

Figure 40.4

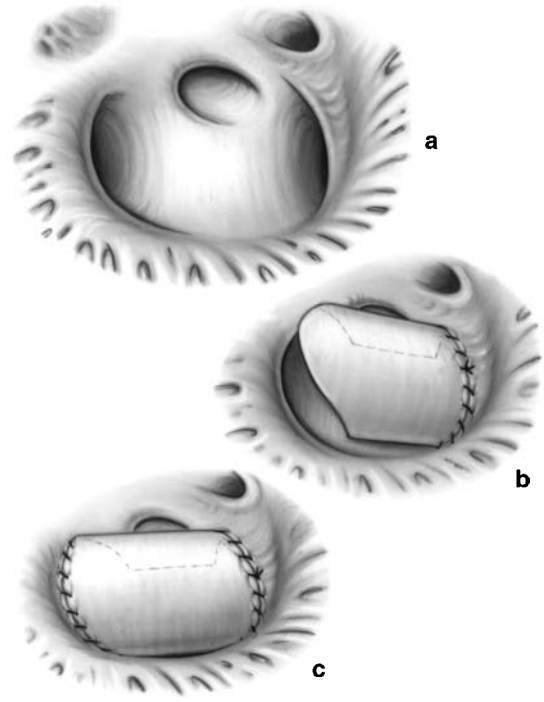

Figure 40.5

2. ***Lateral tunnel***. When a lateral tunnel is created, a Gore-Tex conduit of at least 16 mm in diameter is sized as before and split longitudinally. The baffle is sewn halfway around the junction of the IVC with the right atrium, along the posterior atrial wall,

crista terminalis and half-way around the junction with the SVC. Care is taken to avoid the sinus node. The right atriotomy incision is then closed with a running suture of 5-0 polypropylene.

The rest of the operation is identical in all cases. The IVC cannula is replaced, and the perfusion flow is increased. The distal opening of the main pulmonary artery is enlarged, making an incision toward the right pulmonary artery. Alternatively, the anastomosis can be made on the undersurface of the right pulmonary artery only. At the beginning of our experience, we used to make an incision on the medial aspect of the SVC to enlarge it to the diameter of the IVC. This does not deal with the restriction produced by the crista terminalis within the right atrium. If this incision was carried out into the right atrial chamber, there would be a risk of damaging the sinus node artery in a significant proportion of patients. We do not enlarge the cardiac end of the SVC.

The anastomosis between the cardiac end of the SVC and the pulmonary artery is done with a running suture of 5-0 or 6-0 polypropylene. The suturing starts with the right pulmonary artery, taking wider bites on the pulmonary artery than on the SVC, so as to make up for the discrepancy in the diameter of the two structures (**Figure 40.6a**). Usually, this anastomosis can be made without a patch. However, it is sometimes necessary to use a small pericardial patch medially (**Figure 40.6b**). It is a mistake to simply anastomose the transected main pulmonary artery and the transected SVC, as this distorts the pulmonary artery, which has to be brought right-ward and anteriorly (**inset, Figure 40.6**). The patient is fully rewarmed, and air is evacuated from the heart. It is particularly important to aspirate the air from that portion of the right atrium that is now part of the systemic circulation. A left atrial line is inserted into the right upper pulmonary vein or directly into the left atrium before cardiopulmonary bypass is discontinued.

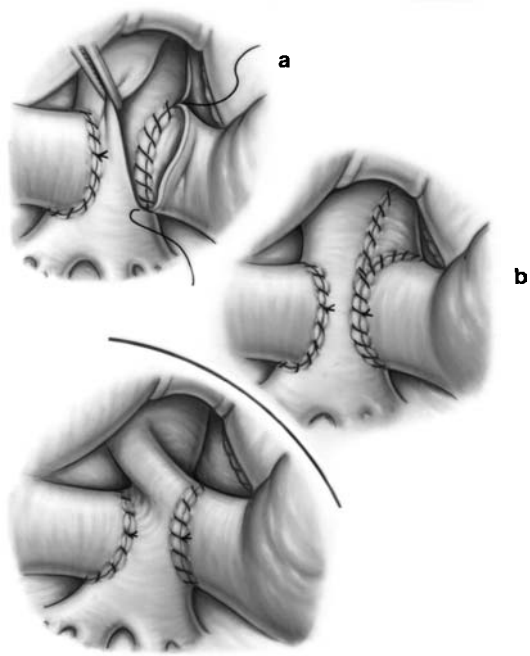

Figure 40.6

In patients who have had a prior hemi-Fontan operation, the beginning of the operation proceeds in the same way. Following opening of the right atrium the previously patched SVC orifice is opened. Placement of the intra-atrial tunnel to complete the total cavopulmonary connection then proceeds as described above.

Extracardiac Connection on Cardiopulmonary Bypass

Cannulation for cardiopulmonary bypass and the first part of the operation are the same as described for the intracardiac connection. It is particularly important that the IVC is cannulated low down. Alternatively, in larger patients, decompression of the IVC territory can be achieved by cannulation of a femoral vein. The pulmonary arteries are dissected, any systemic-to-pulmonary artery shunts taken down, and a bidirectional cavopulmonary shunt, if not already present, is constructed on the beating heart with mild hypothermia. A needle vent is placed in the ascending aorta. The main pulmonary artery is transected above a vascular clamp. However, if control of the proximal pulmonary artery is difficult and the risk of introducing air into the single ventricle is judged to be high, the procedure should be carried out during a short period of cardioplegic arrest. The space between the right lower pulmonary vein and the IVC is dissected and, in case of a re-do operation, adhesions between the right side of the heart and pericardium are taken down to create space for the conduit. Care is taken to avoid injury to the phrenic nerve. The conduit is brought in a gentle curve to the undersurface of the right pulmonary artery, extending onto the main or left pulmonary artery as necessary. The conduit is trimmed obliquely, so that it lies flush with the pulmonary arteries. The superior caval shunt is temporarely occluded and the inferior surface of the pulmonary artery incised. The top end of the Gore-Tex conduit is anastomosed end-to-side to the undersurface of the pulmonary artery, with continuous 5-0 polypropylene. The cannula in the IVC is snared. A vascular clamp is placed across the cavoatrial junction, taking care to avoid obstruction of coronary sinus flow to the atrium. The cavoatrial junction is transected on the clamp and the cardiac end oversawn with continuous 4-0 polypropylene in two layers. A Gore-Tex conduit of at

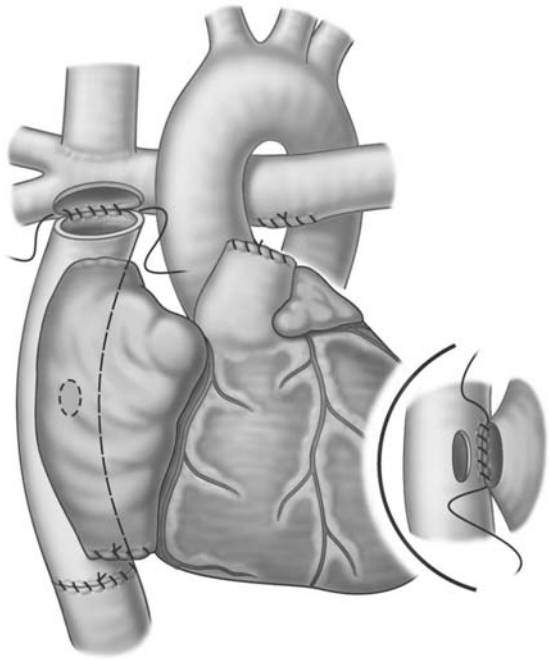

Figure 40.7

least 16 mm diameter (22 mm in adults) is anastomosed end-to-end to the transected IVC (**Figure 40.7**), using continuous 4-0 or 5-0 polypropylene.

Extracardiac Connection without Cardiopulmonary Bypass

In selected patients the operation can be performed without exposing the patient to the damaging effects of cardiopulmonary bypass (Uemura *et al*., 1998). We have no personal experience with this technique. In patients without a prior SVC-to-pulmonary artery connection, this is performed using a temporary shunt from the SVC to the right atrium. For completion of the extracardiac total cavopulmonary connection (TCPC), the anastomosis between the conduit and undersurface of the pulmonary artery is performed next. A segment of the central pulmonary arteries is isolated by placing two clamps. One is from the medial aspect of the cavopulmonary shunt obliquely across the right pulmonary artery, thus diverting blood flow from the shunt to the right lung only; care has to be taken not to obstruct the shunt flow, which is necessary to provide systemic oxygenation. The other clamp is placed across the left pulmonary artery. Following completion of the anastomosis, the clamps are replaced with a single clamp half-way on the conduit and bidirectional cavopulmonary shunt is re-opened. Decompression of the IVC for the distal anastomosis is achieved by constructing a temporary right atrial-to-IVC shunt, by connecting two venous cannulae together.

ADDITIONAL PROCEDURES

Fenestration of the Intra-atrial Baffle

Early mortality after the Fontan procedure can be related to a transient haemodynamic disturbance, such as a postoperative increase in the pulmonary vascular resistance or systemic ventricular dysfunction. Whatever the cause of the failure, systemic venous hypertension is a common denominator that in itself may be responsible for a vicious circle. It causes capillary leakage in the interstitial compartment and in the peritoneal, the pleural, and sometimes the pericardial cavities. Systemic venous hypertension is also responsible for hypertension in the lymphatic system, including the lungs, which may become congested, with further increase in the pulmonary vascular resistance. Immediately after a Fontan procedure, there is usually a reduction in the end-diastolic volume, with significant increase in ventricular wall thickness (Gewillig *et al*., 1990) and stiffness. This is in the presence of inflow restriction to the left ventricle resulting from the increased pulmonary vascular resistance, another step in the vicious circle.

An ingenious and elegant method to prevent this vicious circle was simultaneously developed by the Boston group (Bridges *et al*., 1990b) and the Los Angeles group (Laks *et al*., 1991) for patients at increased risk. The technique consists of fenestrating the atrial baffle or conduit or making an adjustable atrial septal defect. Right-to-left shunting through this defect allows the cardiac output to be maintained at the expense of some arterial desaturation and prevents undue elevation of the systemic venous pressure.

The size of the interatrial communication can be calculated from the Gorlin formula. If one-third of the systemic venous return is diverted to the left atrium, the arterial oxygen saturation will be greater than 85%, assuming a mixed venous oxygen saturation of 60%. For a right atrial pressure of 15 mmHg and a left atrial pressure of 10 mmHg, and if one-third of the venous return traverses the atrial communication, a defect of 6 mm/m^2 is required. In practice, a defect of 4–6 mm in diameter unloads the systemic venous circulation while the systemic arterial saturation remains around 85%.

In patients with an intra-atrial baffle, a fenestration is made in the Gore-Tex conduit with a 4–6 mm punch. The adjustable atrial septal defect is rarely used any more. If an extracardiac conduit has been used, a 4–6 mm hole is cut in the Gore-Tex tube and a slightly larger hole in the opposite free atrial wall. To avoid obstruction of the fenestration by atrial tissue, the atrial wall is anastomosed

to the Gore-Tex tube a few millimetres away from the margin of hole in the prosthesis (**inset, Figure 40.7**).

In a high percentage of cases, the fenestration can be subsequently closed using a trans-catheter technique. In their early experience, Bridges *et al*. (1990b) were able to close the fenestration in 11 of 19 patients within 20 days of surgery. A test occlusion is first carried out. A rapid fall in the right atrial saturation with test occlusion is an indication to leave the fenestration open. In some of their cases, failure to tolerate test occlusion was attributed to residual distal pulmonary stenoses, which were relieved by trans-catheter dilatation; to aortopulmonary collaterals, which were embolized; or to systemic ventricular dysfunction, which improved with time and medical management. We have also adopted this technique of baffle fenestration for high-risk patients. This includes patients who had undergone banding of the pulmonary artery and patients with borderline pulmonary vascular resistance and/or systemic ventricular dysfunction.

Relief of Subaortic Stenosis

Subaortic stenosis often complicates the clinical course of patients with double-inlet left ventricle or tricuspid atresia and discordant ventriculoarterial connection. An increasingly restrictive muscular ventricular septal defect is the most common site of obstruction. Sometimes, there is a subaortic muscular obstruction within the rudimentary ventricle. Pulmonary artery banding is known to exacerbate or promote development of subaortic stenosis, and the risk of this occurring is even greater in the presence of associated aortic arch obstruction (Franklin *et al*., 1990). Ventricular hypertrophy and poor ventricular compliance resulting from subaortic obstruction are major risk factors of the Fontan operation (Kirklin *et al*., 1986). With increasing awareness of these risk factors, the focus has now changed from treatment of subaortic obstruction to the prevention of this complication. Also, growing confidence with the first stage of the Norwood operation has resulted in the more aggressive surgical treatment of subaortic obstruction early in life.

Subaortic obstruction is considered to be significant when the size of the ventricular septal defect is less than half the diameter of the aortic valve in end systole on angiography or two-dimensional echocardiography, as well as when a gradient greater than 10 mmHg at rest is measured at cardiac catheterization or Doppler interrogation. Isoprenaline challenge during the cardiac catheter study has also been suggested to detect significant obstructions (Freedom *et al*., 1986). The subaortic gradient also increases when there is less volume loading of the systemic ventricle following TCPC. Various techniques for the relief of subaortic obstruction have been suggested.

1) Enlargement of Restrictive Ventricular Septal Defect

This can be carried out through the right atrium, the aortic root, or the diminutive right ventricle. The main risk of the enlargement of the ventricular septal defect is the creation of surgical heart block. In double-inlet left ventricle or tricuspid atresia, the atrio-ventricular conduction axis is situated on the left ventricular aspect of the apical and right-sided border of the defect. The safe area for surgical enlargement, therefore, is either the roof of the defect (the outlet septum) or the segment of the septum closest to the obtuse margin of the heart (left border of the defect) (**Figure 40.8**) (Cheung *et al*., 1990).

If the ventricular septal defect enlargement is done through the right ventricle, a vertical incision is made well below the aortic root, to avoid damaging the aortic cusps. Any infundibular obstruction is resected, and the myotomy or myectomy is carried out. The ventriculotomy incision is then closed with a prosthetic patch, to relieve any potential infundibular obstruction. We have now abandoned this technique because of high incidence of ventricular aneurysms. The transaortic approach is prefered.

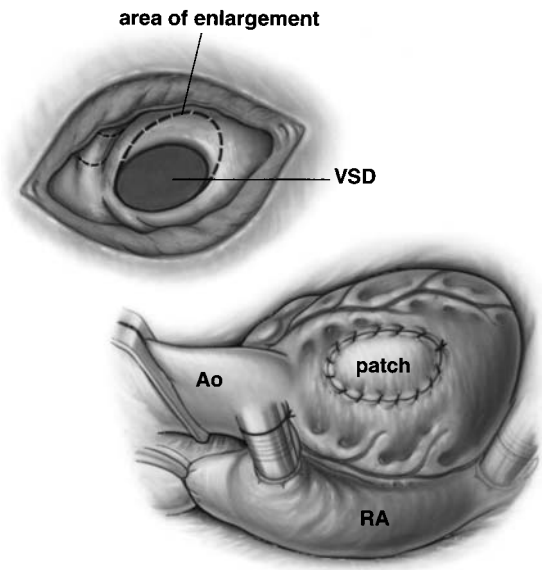

Figure 40.8

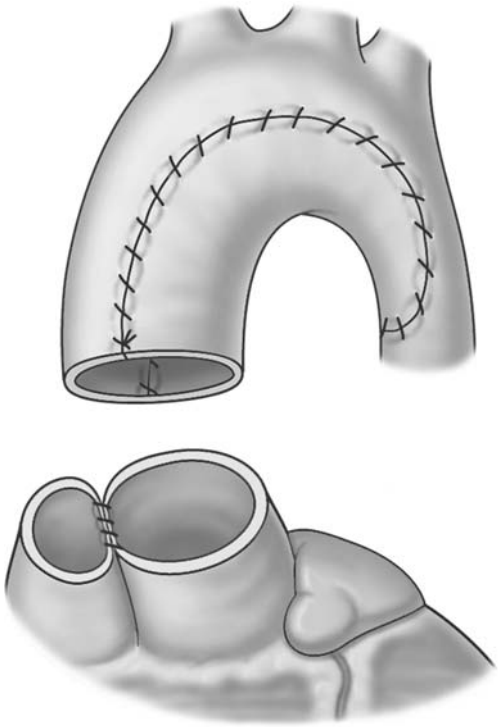

Figure 40.9

2) Use of the Subpulmonary Outflow Tract for Relief of Systemic Outflow Obstruction

The Lamberti modification of the Damus–Kaye–Stansel procedure is currently the most commonly employed solution for relief of subaortic obstruction without pulmonary stenosis (Waldman *et al*., 1988). The pulmonary artery is transected at the bifurcation and anastomosed to a circular aortotomy incision (**Figure 40.9**). It can be combined with a Fontan procedure or one of its modifications, a systemic-to-pulmonary artery shunt, or a bidirectional Glenn anastomosis.

TCPC IN ISOMERIC HEARTS

Abnormalities of the systemic venous drainage are often encountered in isomeric hearts. In the case of azygos continuation, often seen in left isomerism, most of the lower body venous drainage will enter the pulmonary arteries via the SVC anastomosis. However, the hepatic venous return enters the atrial mass from below. In right atrial isomerism, the hepatic veins may enter the atrial mass separately from the IVC. In these cases the hepatic venous return often has to be directed via a separate channel to the pulmonary arteries. Depending on local anatomy and personal preference, this may involve the use of an intra- and a combined extracardiac conduit.

RESULTS

Death before or Unsuitability for the Fontan Procedure

In a cohort of patients presenting with tricuspid atresia between 1971 and 1999, Sittiwankul *et al*. (2004) found that 30% were either ineligible or died before the Fontan procedure. Risk factors included earlier birth date, lower birth weight, aortic arch anomaly, severe right ventricular hypoplasia, and no palliative procedure. Attrition in other forms of univentricular heart is considerably worse and in particular those with heterotaxy syndrome do badly, with overall survival estimates of 49% at 1 year (Hashmi *et al*., 1998). Even fewer patients in the subset with obstructed pulmonary venous connections and severe obstruction of the pulmonary or systemic outflow tracts reach the Fontan operation, especially if surgical intervention is required in the neonatal period, which carries around 75% mortality (Hashmi *et al*., 1998).

Outcome after the Fontan Operation

Early Mortality and Morbidity

Over the three decades since the introduction of the Fontan operation, the operative mortality has decreased from around 20% to around 5%. These improved results apply both to the intracardiac tunnel techniques (Stamm *et al*., 2001; Azakie *et al*., 2001) and extracardiac conduit Fontans (Azakie *et al*., 2001; Alexi-Meskishvili *et al*., 2004) and have occurred in spite of liberating the original Fontan selection criteria and extending the procedure to many forms of complex single ventricle. Factors thought to have contributed to the improved outcome are the more energy-efficient circuit design with the use of a lateral tunnel (de Leval *et al*., 1988) or extracardiac conduit (Marcelletti *et al*., 1990), and the reduction of cross-clamp time and bypass time, both of which can sometimes be avoided in the extracardiac Fontan (Yetman *et al*., 2000). The beneficial effects of the routine use of an atrial fenestration, staging the Fontan procedure with a bidirectional cavopulmonary shunt, and preoperative occlusion of collateral vessels remain open to discussion. Patients with single ventricle and heterotaxy syndrome remain a high risk population for the Fontan circulation because of their multiple associated cardiovascular abnormalities, which include variable anatomy of the sinus node and conducting system, variable systemic venous drainage, potential for pulmonary venous obstruction, atrio-ventricular valve regurgitation, and recurrent or persistent cyanosis in

the presence of arteriovenous shunting. A recent paper from Toronto reported that surgical mortality had been reduced to 13% in this group of patients, and the authors indicated that further improvements could be achieved by early detection and repair of obstructed pulmonary venous return and correction of atrio-ventricular valve regurgitation (Azakie *et al*., 2001).

The importance of unobstructed surgical pathways for the adequate functioning of the single ventricle circulation cannot be overemphasized. A gradient of 1 or 2 mmHg over a cavopulmonary anastomosis may indicate a significant stenosis with energy loss, and this must be revised. Caval and left atrial pressures are monitored postoperatively. Often, there is a modest and temporary increase in the trans-pulmonary gradient during the first 24 h. When this becomes important, it is usually a sign of failure of the circulation. In the event of a low output state in the early postoperative period, mechanical assist devices are occasionally helpful in the recovery if the underlying problem is temporary myocardial dysfunction. If it is not possible to break the haemodynamic vicious circle of the failed Fontan circulation, then the only chance for survival is early take-down of the Fontan circuit.

The Fontan patient is particularly sensitive to a loss of sinus rhythm and also has impaired ability to increase the heart rate. Atrial and ventricular temporary pacing wires are routinely inserted at the end of the Fontan operation to allow sequential pacing. In our experience, arrhythmias, such as His bundle tachycardia, have a very poor prognosis in the Fontan patient.

Atrial thrombosis has been described in several series of Fontan operations. We now recommend anticoagulating these patients routinely, initially with heparin and later coumarin, for a period of 6 weeks, particularly if prosthetic material was used in the right atrium, such as a Gore-Tex conduit for the TCPC.

Pleural and pericardial effusions and ascites are relatively common. The right pleural space is routinely drained for at least 2 or 3 days after the Fontan operation. Pleural effusions are usually serous but occasionally chylous. The chyle probably escapes from the lymphatic system at multiple points, rather than through an area of trauma of the thoracic duct (Kirklin and Barratt-Boyes, 1986), as a result of systemic venous hypertension. On occasion, chylothorax may develop without an exceedingly high venous pressure. If there are significant drain losses, half the volume is replaced with albumin solutions. In the case of chylous drainage, a diet containing medium-chain triglycerides is started. Severe malnutrition may result from persistent chylous leak, and total parenteral nutrition should be considered before deterioration of the patient's general condition. There has been anecdotal evidence that the use of intravenous octreotide (1 to 5 μg/kg/h), a synthetic somatostatin analogue, is beneficial in abolishing the postoperative chylothorax in cardiac patients (Pratap *et al*., 2001). If there are ongoing drain losses, the Fontan haemodynamics should be carefully studied and any pathway obstructions corrected. Surgical control of a chylothorax is rarely possible either by oversewing the area of the leakage or by identifying and ligating the thoracic duct. Scarification of the pleura, injection of sclerosing agents such as tetracycline, or use of fibrin glue have all been tried to enhance adherences between the visceral and the parietal pleura.

Late Outcome after the Fontan Circulation

Twenty years after the introduction of the procedure, Fontan and his colleagues (1990) reported that the ''Fontan state'' was associated with a premature decline in functional status and survival. Even when the operation had been performed under perfect conditions, the predicted survival was only 86%, 81% and 73% at 5, 10 and 15 years, respectively. The reasons for the ongoing late attrition are not known, but they are likely to be complex and multifactorial. A significant proportion of the late complications may be related to the surgical pathways used in the earlier Fontan operations. For the more recently introduced lateral tunnel modification of the Fontan operation, survival has been reported as 93% and 91% at 5 and 10 years, respectively (Stamm *et al*., 2001). Chronic elevation of the systemic venous pressures is likely to play an important role in the late morbidity and is reflected in some of the late complications, such as atrial arrhythmias, hepatic dysfunction and elevated splanchnic venous pressures.

Atrial arrhythmias are a frequent problem at long-term follow-up, probably related to the distended atrium and the presence of extensive suture lines. The incidence has been reduced with the introduction of the lateral tunnel and extracardiac Fontan modifications. Freedom from supraventricular arrthythmias in a recent series of lateral tunnel Fontan operations was 86% at 5 years and 91% at 10 years (Stamm *et al*., 2001, 2002). Heterotaxy syndrome and atrio-ventricular valve abnormalities were identified as risk factors. Atrial fibrillation and supraventricular re-entry tachycardia may be difficult to control medically and lead to ventricular failure. Fontan conversion to a total cavopulmonary connection with extracardiac conduit and concomitant cryoablation of the atrium may be successful in controlling the arrhythmias (Sheikh *et al*., 2004).

Various types of collateral vessels may develop in the single-ventricle circulation. Systemic venous collateral communications develop in up to one-third of patients following a cavopulmonary shunt.

Pulmonary atriovenous fistulae have been reported following cavopulmonary anastomosis and the Fontan operation. Their aetiology remains unclear, but lack of a hepatic factor in the pulmonary circuit may be important (Srivastava *et al*., 1995) and resolution of the malformations has been observed after including hepatic venous return in the Fontan circulation (Shah *et al*., 1997). Peculiar venous communications in many different organ systems have been reported in patients with atrial isomerism. All of the above collateral channels can result in significant right-to-left shunting, with resultant hypoxaemia and cyanosis. Systemic-to-pulmonary artery collaterals are also common and can create a significant left-to-right shunt, imposing a volume load on the single ventricle. In patients with progressive cyanosis or heart failure, an aggressive search for collateral vessels should be made and, if possible, followed by occlusion of the channels.

Thromboembolic complications occur both early and late after the Fontan operation, and involve pathway thrombosis, pulmonary artery thrombosis and cerebral emboli or stroke. The reported clinical incidence varies widely, but has been reported up to 16% for venous thrombosis and 19% for stroke. Many events probably go unrecognized. The effect of prophylactic anticoagulation on the incidence of thrombolism has not been assessed in a systematic way, and permanent anticoagulation after the Fontan operation remains a contentious issue. At our institution we currently use coumadin for the first 6 weeks and then convert to aspirin unless there are clinical reasons to modify this regimen.

Protein-losing enteropathy is a debilitating complication of unknown aetiology that involves loss of protein from the gastrointestinal tract. The reported incidence is up to 15%, with 50% mortality at 5 years following diagnosis (Mertens *et al*., 1998). Non-selective protein loss may result in peripheral oedema, ascites and effusions, immunodeficiency and coagulopathy. An elevated α 1-antitrypsin level in the stools is diagnostic (Hill *et al*. 1981). Medical treatment is with diuretics and protein supplements. High-dose corticosteroids (1–2 mg/kg/day for a minimum of 2–3 weeks) and high molecular weight heparin (5000 U/m^2/day for several weeks) have been successful in reducing protein loss in some patients. Fontan haemodynamics should be optimized if possible, and this may involve revision of Fontan pathways or pacing. None of the above treatments or even cardiac transplantation (Mertens *et al*., 1998) has been universally successful in treating the problem.

Problems related to the surgical pathways are common and increase with time. Baffle leaks and localized stenoses can increasingly be managed by catheter intervention, thus avoiding reoperation in these complex patients. The recent introduction of the extracardiac conduit is potentially a problem for small children, in whom the prosthesis may become too small with somatic growth, with resultant systemic venous pathway obstruction. This has so far not been reported in the literature; however, oversizing of the conduit has been complicated by poor haemodynamics and conduit thrombosis (Alexi-Meskishvili *et al*., 2000).

Ventricular failure is also a frequent problem. There may be many reasons why the performance of the single ventricle may be impaired in the Fontan circulation. The myocardium may be intrinsically abnormal or may be impaired because of the adverse working conditions prior to the Fontan circulation, such as chronic cyanosis, volume overload or subaortic stenosis. Also, the working conditions for the myocardium in the single ventricle circulation are less favourable than in a biventricular circulation: the acute preload reduction at the time of the Fontan circulation leads to an increase in mass:volume ratio and impaired diastolic performance. At the same time there is an increase in afterload because the systemic and pulmonary circulations are now in series. Thus, single ventricles need to spend more energy to generate a unit of cardiac output, but also have less flow reserve because of limited preload.

MANAGEMENT OF THE FAILING FONTAN

The failing Fontan circulation is an increasing problem. In patients with a failing atriopulmonary connection and arrhythmias, marked improvement has been reported following conversion to a total cavopulmonary connection with concomitant arrhythmia surgery, and was associated with a low operative risk (Mavroudis *et al*., 1998). The selection criteria for this operation, however, have not yet been defined in detail.

If ventricular dysfunction is the main reason for Fontan failure, the patient should be assessed for cardiac transplantation. Assessment of the pulmonary vascular bed is difficult, because of non-pulsatile pulmonary flow, low cardiac output and possible aortopulmonary collaterals. Post-transplant right heart failure due to elevated pulmonary vascular resistance is a well-recognized problem and is associated with high mortality. The transplantation can be challenging because of multiple previous interventions, the need for extensive vascular reconstructions and the risk of bleeding. In a recent review of patients who underwent orthotopic cardiac transplantation for failing single ventricle physiology, the 30 day mortality was 68%, but there were no further deaths up to 14 years follow-up (Michielon *et al*., 2003). Because of the long waiting time for transplantation, it is not uncommon for these patients to die on the transplantation waiting list.

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41

Hypoplastic Left Heart Syndrome

C. Pizarro and W. I. Norwood

Hypoplastic left heart syndrome (HLHS) is the fourth most common congenital cardiac anomaly presenting in the first year of life. It has been reported to occur in at least 0.016–0.036% of live births, and thus it is the most common defect in which there is only one ventricle (Fyler, 1980). In autopsy series, it represents 1.4–3.8% of cases of congenital heart disease (Edwards, 1953) and has been reported to cause 23% of the deaths from congenital heart disease in the neonatal period (Watson and Rowe, 1962). Lev (1952) first described the pathological features of hypoplasia of the aortic tract complexes, and Noonan and Nadas (1958) referred to these lesions as hypoplastic left heart syndrome.

Although the anatomical features in this categorization are somewhat varied, the physiological similarities within this anatomical spectrum have resulted in acceptance of this term. With the advent of surgical management for patients with HLHS, the anatomy has been more precisely defined. The central anatomical features are aortic valve atresia or hypoplasia with associated marked hypoplasia or absence of the left ventricle. The aortic arch and ascending aorta are functionally a branch of the ductus arteriosus–thoracic aorta continuum, with retrograde flow through the aortic arch into a diminutive ascending aorta, which is usually 1.5–3 mm in diameter. A less common variation includes malalignment to the right of the common atrio-ventricular canal with regard to the muscular ventricular septum. Although 10% of patients with HLHS have double-outlet right ventricle rather than the usual normal relation of the great arteries, cases involving transposition of the great arteries with hypoplasia of the left ventricle and pulmonary artery or hypoplasia of the right ventricle and aorta are not considered to be hypoplastic left heart syndrome.

The anatomy of patients with aortic stenosis or atresia has been thoroughly described (Bharati *et al*., 1984; Noonan and Nadas, 1958; Weinberg *et al*., 1986). In 2.5–4.3% of patients, a persistent left-sided superior vena cava is found (Bharati *et al*., 1984; Roberts *et al*., 1976). The right atrium is dilated, and the tricuspid valve orifice is enlarged. The right ventricle is also somewhat enlarged, as are the pulmonary orifice and main pulmonary artery. The pulmonary venous connection is usually normal, but occasionally patients have an anomalous or accessory pulmonary venous connection, usually through a persistent left vertical vein to the left innominate vein (Watson and Rowe, 1962). It is important to distinguish total anomalous pulmonary venous connection, when present, from normal connection to a diminutive left atrium but anomalous drainage (Seliem *et al*., 1992). The left atrium is small and frequently hypertrophied. An interatrial communication is common, although a congenitally small foramen ovale may be present (Bharati *et al*., 1984). Mitral stenosis or hypoplasia is present in approximately 60% of patients, and the remaining 40% have mitral atresia. Many patients with mitral hypoplasia or stenosis (patent left ventricular inflow) have prominent endocardial fibroelastosis (Bharati *et al*., 1984; Roberts *et al*., 1976) with myofibril disarray (Bulkley *et al*., 1983). Short- or long-term outcome has not proved to be different between patients with a patent left ventricular inflow compared to those with an atretic one. In a study by Bharati *et al*. (1984), 87% of the patients had aortic atresia and 13% had aortic stenosis. The left ventricle generally does not form the apex of the heart. This is a key anatomical feature that can aid in distinguishing between HLHS and critical aortic stenosis with a small left ventricle. The ascending and transverse portions of the aorta are hypoplastic. When aortic atresia is present, the diameter of the ascending aorta is usually 1–3 mm. In cases of aortic annular hypoplasia without atresia, the mean diameter of the ascending aorta has been reported to be as large as 5–6 mm. True coarctation of the aorta is not a common finding; there is, however, a posterolateral intimal ridge at the junction of the aortic isthmus, ductus arteriosus and thoracic aorta in most patients.

The coronary arteries originate normally from the aortic root and usually have a normal distribution. In

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the unusual case of a coronary anomaly, left ventricle to coronary artery fistula has the highest incidence (Baffa *et al*., 1992).

The anatomical features of atrio-ventricular septal defect (AVSD; malaligned atrio-ventricular canal) are similar to those of any AVSD. Atrial communication is via an ostium primum atrial septal defect. An inlet ventricular septal defect may or may not be present. The development of the left ventricle depends on the degree of malalignment of the common atrio-ventricular valve. This ventricle, however, still does not form the apex of the heart. The diameter of the ascending and transverse aorta may be larger than in patients with aortic atresia but smaller than in patients with isolated aortic stenosis.

In the rare case of left ventricular outflow obstruction with an unrestrictive ventricular septal defect, left ventricular development may be normal; thus, a primary myocardial abnormality is unlikely to be the cause of HLHS. The elevation in left ventricular pressure that results from unrestricted left ventricular inflow and obstructed left ventricular outflow in the absence of a ventricular septal defect probably causes the endocardial fibroelastosis encountered in some patients. Although congenital smallness or absence of the foramen ovale has been suggested as a possible cause of HLHS (Lehman, 1927; Lev *et al*., 1963), this occasional finding is more likely to be the result of an increase in left atrial pressure secondary to the left ventricular outflow obstruction.

DIAGNOSIS

The advent of foetal echocardiography has allowed the diagnosis of HLHS with accuracy, as early as 18–20 weeks of gestation. This development has influenced the management of these patients in several areas:

1. It has resulted in a smaller number of cases at risk for clinical compromise secondary to delayed diagnosis.
2. It has simplified the transport of the patient and mother before birth to a neonatal cardiac centre.
3. It has provided additional time for the parents to understand and become familiar with the management options.
4. It has provided a diagnostic and monitoring tool to explore the feasibility and usefulness of foetal intervention.

Physical examination of the newborn with HLHS typically reveals mild cyanosis with tachypnoea and tachycardia. Depending on the degree of ductal patency at the time of evaluation, peripheral pulses may be normal, diminished or absent. Rales may be heard, although the lung fields generally are clear. Cardiac examination reveals a dominant right ventricular impulse on palpation with a decreased left ventricular (apical) impulse. S1 is normal, and S2 is usually single and increased in intensity. One-third of patients may have a gallop rhythm at the apex. Many patients have a non-specific soft grade 1/6–3/6 systolic murmur at the left sternal border. An apical mid-diastolic flow rumble is heard in 20% of patients. The liver is commonly enlarged.

The electrocardiogram often reflects the underlying pathology. Right atrial enlargement is common, and right ventricular hypertrophy is usually present. More than half of patients have a QR pattern in lead V1. In patients with malaligned common atrio-ventricular canal defect, the QRS axis is to the left and superior. The findings on the chest film are non-specific. Cardiomegaly is common, and pulmonary vascular markings may be prominent. In the occasional patient with a severely restrictive atrial septal defect, a reticular pattern may resemble that seen in total anomalous pulmonary venous connection with pulmonary venous obstruction. *Two-dimensional* (2D) *echocardiography* is a diagnostic study of HLHS. The intracardiac anatomy should be examined with subcostal frontal, sagittal, left oblique and right oblique views (Chin *et al*., 1985; Isaaz *et al*., 1985; Marino *et al*., 1985). In accordance with some obstruction at the interatrial level and an obligatory left-to-right atrial shunt, the atrial septum is usually bowed from left to right. When the atrial septum is bowed from right to left, either severe tricuspid regurgitation or anomalous pulmonary venous connexion should be suspected. When the atrial septum is thickened and severely bowed from left to right, congenital smallness or absence of the foramen ovale should be suspected. The septum primum commonly shows evidence of posterior and left-sided deviation of its superior attachment (Helton *et al*., 1986), although this feature does not influence surgery or its outcome. In suprasternal imaging, the anatomy of the ascending aorta, aortic arch, and upper descending aorta should be assessed with frontal, sagittal and left oblique views. A diminutive ascending aorta is characteristic of HLHS. After the anatomical details are determined, colour-flow imaging, pulsed Doppler imaging and continuous-wave Doppler imaging are used to evaluate physiological features. Of primary concern is the assessment of the tricuspid or common atrio-ventricular valve for regurgitation (Barber *et al*., 1988). Fifty-six percent of patients with hypoplastic left heart syndrome have tricuspid regurgitation preoperatively (mild in 35%, moderate in 16%, and severe in 5%). Because accurate diagnosis is possible by 2D and Doppler echocardiography, cardiac catheterization is not routinely performed except in the occasional

patient with evidence of a significantly restrictive atrial septal communication and anatomy suitable for balloon septotomy.

PREOPERATIVE CONSIDERATIONS

The most obvious physiological consequence of HLHS is that systemic blood flow is provided virtually in its entirety from the right ventricle via a patent ductus arteriosus. The foetus with HLHS is generally healthy and unaffected by the anatomical abnormality, but two of the natural changes from foetal to neonatal physiology are life-threatening. As the ductus arteriosus closes, systemic perfusion is impaired. In addition, the natural decrease in pulmonary vascular resistance shortly after birth results in a volume shift from the systemic to the pulmonary circulation, also impairing systemic perfusion. Satisfactory systemic perfusion and oxygenation can be maintained in the newborn by ensuring patency of the ductus arteriosus, maintaining an adequate intravascular volume and avoiding large differences between systemic and pulmonary vascular resistances. The last two principles are critically important in the management after palliative surgery for HLHS and before surgery as well. In all neonates, a continuous infusion of prostaglandin E1 at a dose of 0.01–0.05 μg/kg/min maintains patency of the ductus arteriosus. Only rarely, when there is virtual absence of an interatrial communication, does metabolically important hypoxaemia occur, as a consequence of a low pulmonary-to-systemic flow ratio. Most patients are often threatened not by metabolically important hypoxaemia but by decreased systemic perfusion, as a consequence of a low pulmonary vascular resistance and accompanying increased systemic vascular resistance. In this circumstance, addition of 1–4% carbon dioxide to the inspired gas mixture (Gullquist *et al*., 1992) or nitrogen (Day *et al*., 1998) results in an increase in the pulmonary vascular resistance, favourably influencing the ratio of pulmonary to systemic flow and promoting adequate systemic perfusion. In some instances a continuous infusion of sodium nitroprusside has been used to decrease an abnormally elevated systemic vascular resistance, and the perioperative use of the systemic vasodilator phenoxybenzamine has gained popularity in some centres (Tweddell *et al*., 1999).

The preoperative use of catecholamines is almost never necessary or appropriate, with the exception of the occasional patient who presents in extremis with a high pulmonary:systemic flow ratio. We have rescued a few neonates who could not be resuscitated with the usual medical and pharmacological measures by rapid placement of pulmonary artery bands on the individual pulmonary arteries over a right-angled dissector (equivalent to a 7-Fr. dilator) via median sternotomy (**Figure 41.1**) followed by Stage I Norwood procedure several days after banding, once the patient's condition stabilized and end-organ function has improved (Pizarro *et al*., 2003b). In some cases when the patient condition warranted avoidance of exposure to cardiopulmonary bypass, these patients underwent placement of a stent in the ductus arteriosus and had further surgery delayed for 4–6 months, at which time aortic arch reconstruction was combined with superior cavopulmonary connection (hemi-Fontan), with excellent outcomes. Additionally, some institutions are exploring the relative merits of a systematic approach, placing flow-limiting occluders in the branch pulmonary arteries combined with the elective deployment of a stent in the ductus arteriosus in the catheterization laboratory, either as a bridge to transplantation or to delay surgery for several months (Mitchell *et al*., 2003).

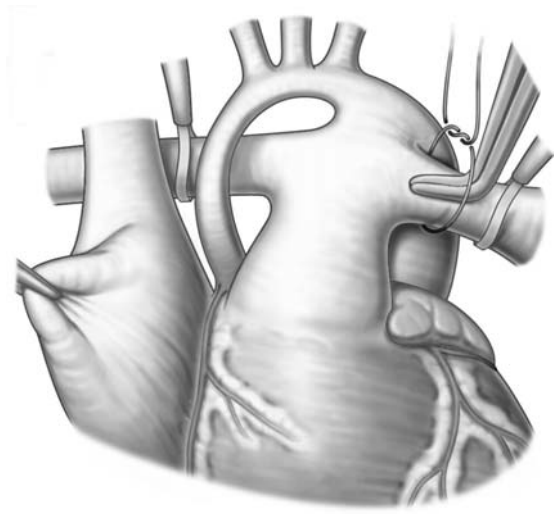

Figure 41.1

Pulmonary venous return through a restrictive atrial septal communication could complicate the management of patients with HLHS. Some degree of obstruction to pulmonary venous return limits pulmonary overcirculation, therefore a balloon atrial septotomy performed during cardiac catheterization may result in haemodynamic deterioration and should be avoided. On the other hand, extreme hypoxaemia secondary to a highly restrictive or intact atrial septum is an indication for urgent intervention. In this scenario and according to the anatomic substrate (Rychik *et al*., 1999), an atrial septal communication needs to be created or enlarged, either in the cardiac catheterization laboratory or during surgery, to allow the patient's stabilization prior to reconstructive surgery. Recent experience demonstrates that, despite

prenatal diagnosis, advances in cardiac catheterization techniques and a policy of immediate intervention after birth, neonatal mortality in the subgroup of patients with intact or very restrictive atrial septum remains quite high (Vlahos *et al*., 2004). With the idea that early intervention may change the flow-related development of the cardiac structures, the feasibility of foetal intervention is being explored in these patients.

Like other single-ventricle complexes, HLHS is ultimately treatable by some modification of the Fontan procedure. However, at birth the lungs are immature and the vascular resistance is naturally high, precluding a Fontan procedure in the neonatal period. Yet affected infants are dependent on patency of the ductus arteriosus. Although the ductus arteriosus can be opened and maintained with continuous infusion of prostaglandin E1, prolonged natural association of the pulmonary circulation with the systemic circulation results in either intractable congestive heart failure or, ultimately, development of pulmonary vascular obstructive disease. Therefore, it has been clear even from the beginning of our surgical development that staged management is necessary. Three basic principles underlie the initial palliation:

1. The aorta must be associated directly with the right ventricle in a manner guaranteeing unobstructed flow from the right ventricle to the systemic circulation, with growth potential obviating further aortic surgery.
2. Pulmonary blood flow must be regulated for proper growth, development and maturation of the pulmonary vasculature, in order to avoid the development of pulmonary vascular obstructive disease and to minimize the excessive volume load on the right ventricle.
3. A large interatrial communication is necessary in order to avoid pulmonary venous hypertension and its consequences.

The surgical details of the initial palliation evolved during the 1980s. Although the initial surgical effort to regulate pulmonary blood flow was accomplished through the interposition of a graft between the right ventricular free wall and the central pulmonary arteries, this was soon replaced with a modified right Blalock–Taussig shunt, largely because of technical issues related to haemostasis, control during the second procedure and because such a shunt could be placed during the period of rewarming, thus minimizing circulatory arrest time (Norwood *et al*., 1981). In recent years, and following preliminary reports of remarkable circulatory stability, there has been a resurgence of interest in the use of a right ventricle-to-pulmonary artery shunt (Kishimoto *et al*., 1999; Imoto *et al*., 1999; Sano *et al*., 2003; Pizarro *et al*., 2003a). This renewed interest also stems from the theoretical advantage of elimination of diastolic run-off from the systemic (coronary) arterial system to the lower-resistance pulmonary arterial bed, thereby providing for a more normal systemic flow pattern. This has been documented when a 5 mm right ventricle–pulmonary artery conduit was compared to the use of a 3.5 or 4.0 mm modified BT shunt as the source of pulmonary blood flow (Maher *et al*., 2003; Mair *et al*., 2003).

Additionally, it has been suggested that the use of a right ventricle-to-pulmonary artery conduit may have the potential to increase the circulatory stability, not only improving the immediate postoperative period, but also reducing the risk of interim mortality between the first and second operations, possibly due to a more favourable physiology by virtue of eliminating the connection of the systemic and the pulmonary circulation at the arterial level, allowing a reduction of the volume load on the single ventricle, a higher diastolic pressure, and the potential for a favourable impact on coronary perfusion (Pizarro *et al*., 2004).

NORWOOD PROCEDURE (STAGE ONE)

Anaesthetic Considerations

As suggested earlier, the preoperative management of patients with HLHS depends mainly on the ratio between pulmonary and systemic blood flow exhibited by the individual patient. Fortunately, the vast majority of patients present with a relatively balanced Qp:Qs, evidenced by adequate peripheral perfusion and the absence of metabolic acidaemia. Alternatively, patients can present with excessive pulmonary blood flow associated with systemic hypoperfusion and metabolic acidaemia, or with severely decreased pulmonary blood flow, usually as a consequence of a very restrictive atrial septal communication or an intact atrial septum. In this latter situation, the clinical condition is dominated by severe hypoxaemia and requires immediate intervention. In any of these scenarios, the balance between the pulmonary and systemic circulations should be maintained during transport and initiation of anaesthesia.

The preoperative management is the same for patients undergoing *Stage I Norwood* or *heart transplantation*, and includes an infusion of PGE1, appropriate intravascular volume expansion, and balancing the pulmonary and systemic blood flows. There is no set guideline regarding the timing of reconstructive surgery, which in our unit is performed in the first days of life; meanwhile, most patients are allowed to breathe spontaneously and receive enteral feedings. The close proximity of the operating room and the effective

half-life of PGE1 allow the patient's transport after discontinuation of the infusion in the ICU. If the patient is already intubated, ventilation through a Mapleson system is used to deliver the same mixture of gases used in the intensive care unit preoperatively. Hyperventilation would favour an increase in Qp:Qs, reflected by an increase in the PaO_2 and a decrease in the arterial blood pressure, and should be avoided.

The anaesthetic technique is aimed at preservation of the balance between systemic and pulmonary resistances and to avoid myocardial depression. General anaesthesia is induced with sevofluorane as the inhalational agent, followed by 0.2 mg/kg pancuronium (Pavulon) intravenously, which is used for its vagolytic properties. Nasotracheal intubation is used and 10 μg/kg fentanyl are administered, followed by another 30 μg/kg after the initiation of cardiopulmonary bypass. More recently, remifentanyl, a short-acting opioid, has been utilized as a continuous infusion, at a dose of 0.3 μg/kg/min, for increased control over the level of anaesthesia intra- and postoperatively. If an endotracheal tube is already in place, this is changed to eliminate the potential for airway obstruction due to inspissated secretions. In order to preserve vascular access and minimize the potential for vascular thrombosis, placement of femoral arterial and central venous lines are avoided, and access is usually obtained by placement of a peripheral venous line and a right radial artery line percutaneously, or by cut-down in case an umbilical arterial line is not available. A urinary bladder drainage tube, a nasogastric tube, and nasopharyngeal, oesophageal and rectal temperature probes are placed. During this preparatory phase, surface cooling is in progress, as the OR is maintained at 20°C and the rectal temperature usually drifts down to 32–34°C. Again, care must be taken to avoid significant changes in Qp:Qs secondary to the effects of general anaesthesia and neuromuscular blockade on minute ventilation.

Once the patient is ready for surgery, the cooling blanket set at 15°C, the temperature in the operating room is maintained at 20°C and ice bags are applied to the head. During this phase, a decrease in the CO_2 production should be noted; however, its potential impact on Qp:Qs are minimized by the expeditious control of pulmonary blood flow after institution of cardiopulmonary bypass.

Surgical Technique

A conventional midline sternotomy incision is made, and the thymus is removed to facilitate exposure of the diminutive aortic arch and its branch vessels. Arterial cannulation is most conveniently achieved in the proximal main pulmonary artery, just above the sinuses of Valsalva, by placing a single diamond-shaped purse-string suture through a small rubber tourniquet. A 10-French aortic cannula is fitted with a rubber bumper approximately 3–4 mm from the tip. After heparinization, while the first assistant holds and stretches the arterial wall in its long axis proximal and distal to the purse string, the pulmonary artery is incised, and the cannula is introduced. Threading of the cannula through the ductus arteriosus followed by ligation around the arterial cannula could be performed; however, it is unnecessary and requires excessive manipulation of the cardiovascular structures. Distortion and trauma to the delicate neonatal cardiovascular structures must be minimized in order to preserve anatomy and function. A 16-French venous cannula is placed through the right atrial appendage, and cardiopulmonary bypass is instituted. The right and left pulmonary artery branches are rapidly exposed and occluded with 2–0 Tevdek tourniquets to ensure systemic perfusion through the ductus arteriosus. The infant is cooled to 18°C while oesophageal, nasopharyngeal and rectal temperatures are monitored. During this time, the branch vessels of the aortic arch are exposed and looped with 2–0 Tevdek tourniquets in preparation for circulatory arrest (**Figure 41.2**). Dissection is extended around the aortic arch onto the thoracic aorta in the posterior mediastinum. At this point, the branch vessels of the aortic arch are occluded, the circulation is discontinued, and the blood is drained into the venous reservoir. The arterial and venous cannulae are removed, and cold crystalloid cardioplegic solution is infused directly in the ascending aorta if the size permits (aortic stenosis/hypoplasia), in the innominate artery proximal to the

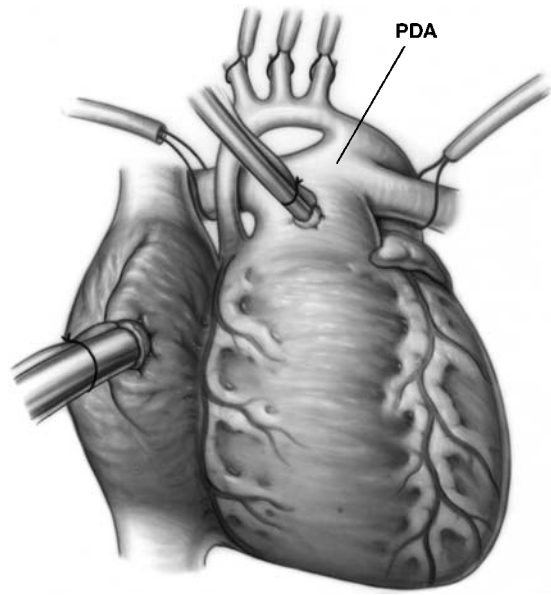

Figure 41.2

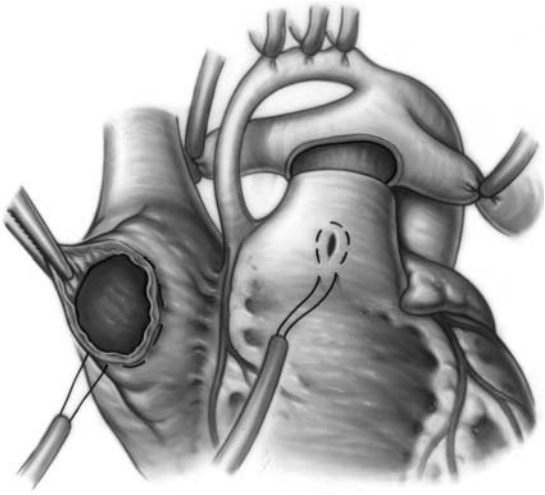

Figure 41.3

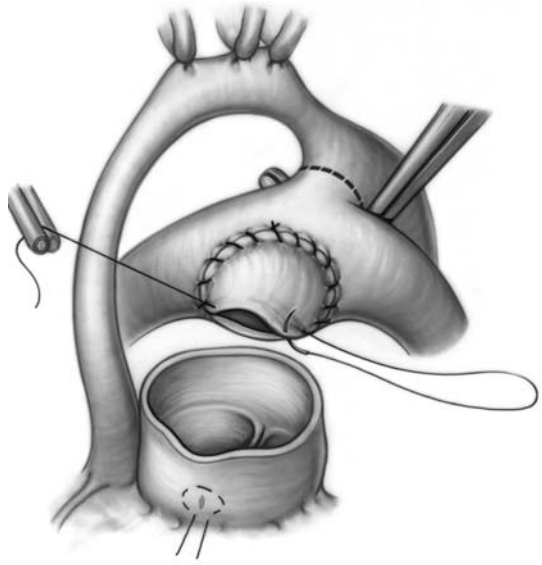

Figure 41.4

tourniquet, or through the pulmonary arterial cannulation site while the descending thoracic aorta is temporarily occluded with forceps. Through the right atrial cannulation site, the septum primum is identified and excised. Occasionally, particularly when the interatrial communication is very small or virtually absent, the extra time to access the atrial septum through a lateral right atriotomy is advantageous. Attention is then turned to the main pulmonary artery, which has been separated from the diminutive ascending aorta during the cooling phase of cardiopulmonary bypass. Immediately proximal to the take-off of the right pulmonary artery, the main pulmonary artery is transected (**Figure 41.3**), and the distal main pulmonary artery is closed with a patch of pulmonary artery homograft by a running monofilament suture. Patch closure is recommended in order to maintain better continuity between the right and left pulmonary artery branches (**Figure 41.4**). The ductus arteriosus is then exposed, ligated and transected at its entrance into the thoracic aorta (**Figure 41.5**). An incision in the aorta is extended distally 1–2 cm into the thoracic aorta, and also proximally into the aortic arch and ascending aorta, to the level of the rim of the transected proximal main pulmonary artery. Because the isthmus of the aorta and the aortic arch actually function as a branch of the continuum of the main pulmonary artery, ductus arteriosus and thoracic aorta, the junction of the isthmus and the thoracic aorta should be gusseted with a patch which extends towards the ascending aorta (**Figure 41.6**). Cryopreserved pulmonary artery homograft is used for this patch, due to its advantages of being thin, pliable, easy to handle and haemostatic. The pulmonary artery homograft patch is then extended to

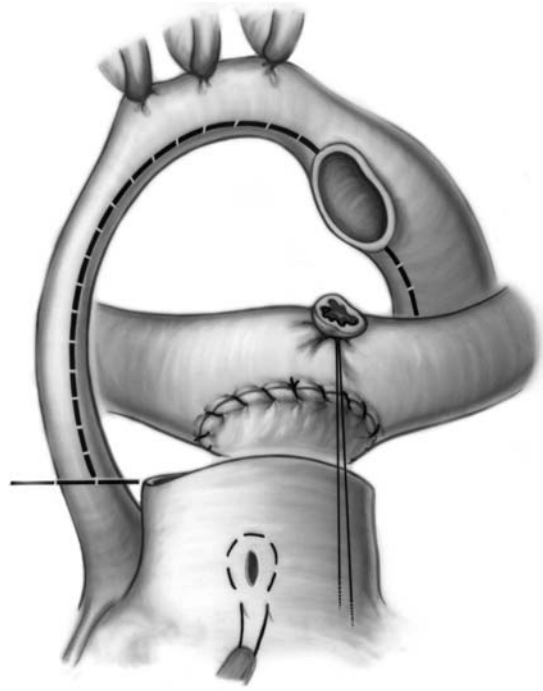

Figure 41.5

approximately 5 mm above the end of the most proximal incision in the ascending aorta (**Figure 41.7**). When the ascending aorta is very small, the proximal anastomosis of the main pulmonary artery to the ascending aorta can be begun, with multiple interrupted fine monofilament

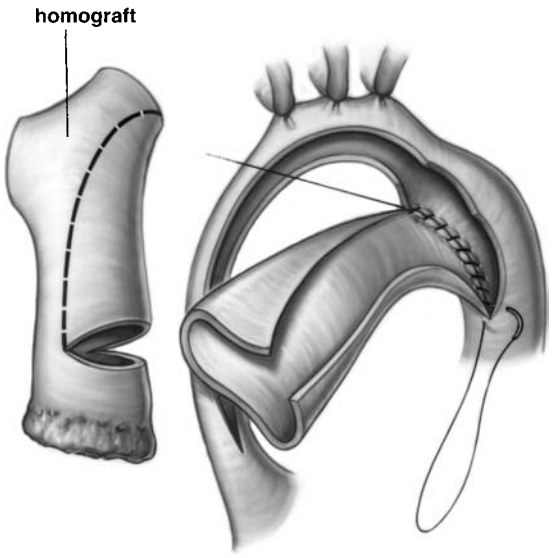

Figure 41.6

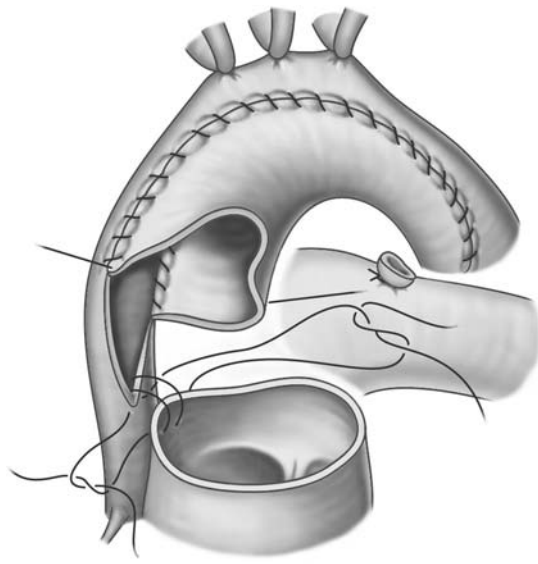

Figure 41.8

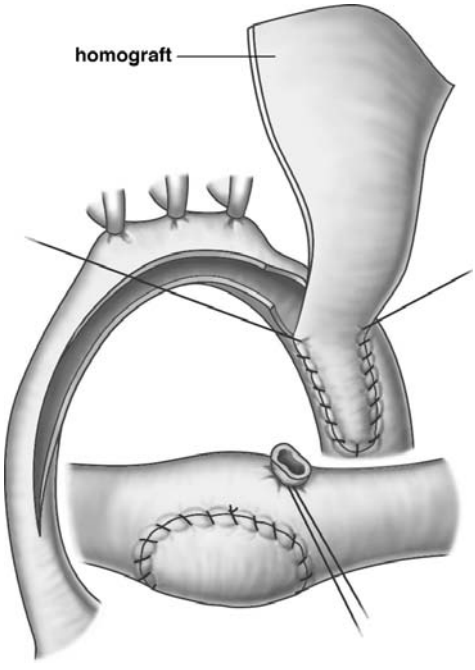

Figure 41.7

sutures to avoid ''purse-stringing'' of the inlet into the diminutive aortic root (**Figure 41.8**). Generally, however, this connection is accomplished quite satisfactorily with a running suture technique. The reconstruction is completed by anastomosis of the ascending aorta, main pulmonary artery and pulmonary homograft, thus creating outflow from the right ventricle to the augmented aorta through the pulmonary valve (**Figure 41.9**).

The construction of a short shunt of 4 mm Gore-Tex tube between the innominate artery and a confluence of the pulmonary arteries was favoured, thinking that it may provide a more even distribution of flow and thus balanced growth of the right and left pulmonary arteries.

More recently, we have used a 5 mm PTFE tube graft interposed between the right ventricle and central pulmonary arteries as the source of pulmonary blood flow, for the reasons previously outlined. In this case, the PTFE conduit is sewn to an oval patch of pulmonary homograft with a central opening and used to close the distal end of the transected main pulmonary artery (**Figure 41.10**). Following completion of the aortic reconstruction, the proximal anastomosis of the PTFE conduit is made to an incision in the free wall of the RV infundibulum, carefully avoiding the coronary arteries and ventricular septum when present (**Figure 41.11**). Passing a right-angled clamp through the pulmonary valve prior to completion of the neo-aortic reconstruction could assist in finding the appropriate site for the infundibulotomy, particularly in the case when the left ventricle is only moderately hypoplastic. A 2–4 mm ventriculotomy is made and enlarged to the appropriate size by separating the muscle fibres with a small mosquito

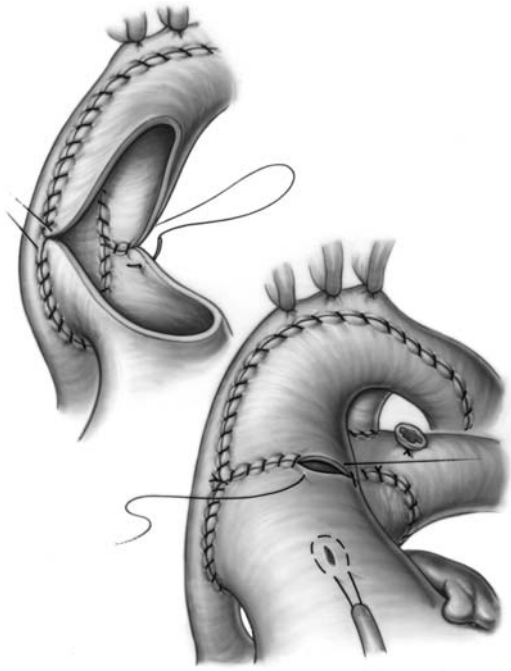

Figure 41.9

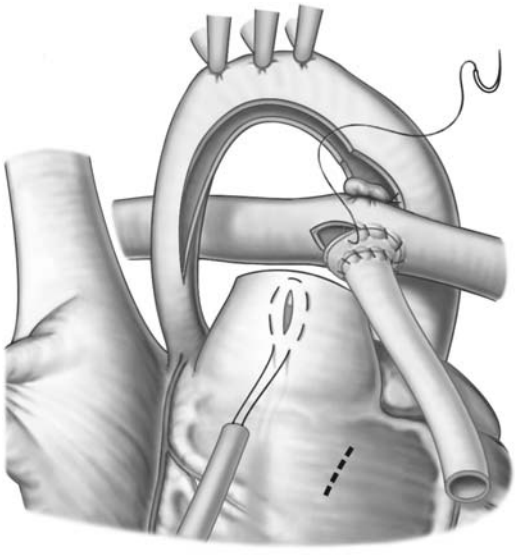

Figure 41.10

clamp. Resection and thinning of the myocardium is seldom necessary and can lead to the development of aneurysmal complications. Cardiopulmonary bypass is reinstituted, the tourniquets on the brachiocephalic vessels of the aortic arch are removed, and the patient is warmed to 37°C. However, in order to minimize the period of circulatory arrest, the proximal anastomosis between the conduit and the infundibulum can be performed while on cardiopulmonary bypass with the heart beating. Good visualization to allow inclusion of epicardium and endocardium during the construction of the proximal anastomosis is quite important, in order to avoid obstruction of the conduit inflow by muscle growth.

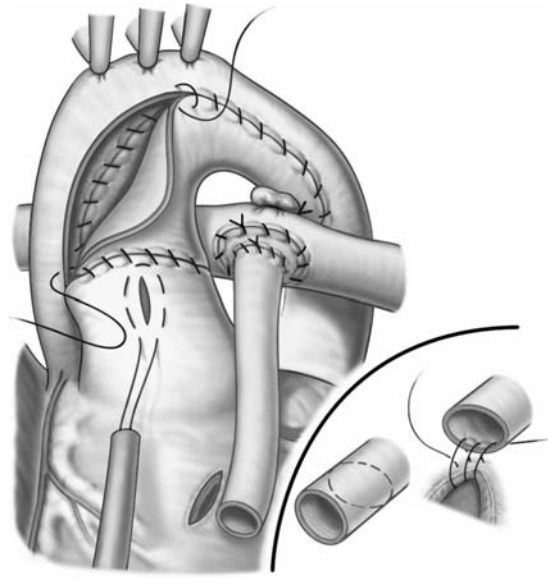

Figure 41.11

After the patient is weaned from cardiopulmonary bypass, the cannulae are removed, a pressure-monitoring catheter is placed through the right atrial appendage cannulation site and the chest is usually closed. If for any reason the surgery has resulted in excessive swelling, or if use of extracorporeal support is anticipated, the sternal closure is delayed. Upon transfer to the intensive care unit, the patient is placed on volume-cycled ventilation with a total volume of 20 ml/kg/minute, a ventilatory rate of approximately 20 breaths/minute and a fraction of inspired oxygen between 0.21 and 0.3. The use of a right ventricle-to-pulmonary artery conduit has been associated with very stable haemodynamics in the postoperative period. Clinical evidence of excessive pulmonary blood flow is rare, therefore the addition of carbon dioxide or nitrogen to the inspired gas mixture, commonly used among patients who received a modified Blalock–Taussig shunt, is unnecessary.

After the Norwood procedure the right ventricle is still subjected to an excess volume load. With a goal of long-term preservation of ventricular function, prior to the late 1980s, suitability for a modified Fontan procedure was assessed when the patient was 12–18 months old. Poor outcome from this approach was attributable in some patients to rapid changes in ventricular geometry, which resulted in decreased ventricular compliance after such a volume unloading procedure. These observations led to dividing the process of connecting the venae cavae to the pulmonary arteries into two separate operations, a strategy which resulted in a significant decrease in morbidity and mortality. The authors' current practice is to evaluate the patient's anatomy and physiology at 5–6 months of age, prior to associating the superior vena cava to the central pulmonary arteries. At this age, the pulmonary vascular resistance is likely to be low (< 2.5 Wood's units), ventricular end-diastolic pressure normal (< 7–8 mmHg) and development and maturation of the pulmonary vasculature complete. At the time of hemi-Fontan, the shunt is closed, the central pulmonary arteries are augmented with a patch of pulmonary artery homograft, and the superior vena cava is connected to the branch pulmonary arteries as described. The superior vena cava is not separated anatomically from the right atrium; rather, the junction of the superior vena cava with the atrium is closed with a portion of the homograft patch that is used to augment the pulmonary arteries. Although many practitioners use some form of bidirectional Glenn procedure at this juncture, the authors favour the hemi-Fontan procedure because it facilitates the association of the inferior vena cava to the pulmonary arteries and at the same time allows enlargement of the central pulmonary arteries (Norwood and Jacobs, 1993).

HEMI-FONTAN (STAGE TWO)

The heart is exposed through a midline sternotomy, and cannulation for cardiopulmonary bypass is achieved by placing an arterial cannula in the neo-ascending aorta and a single venous cannula through the right atrial appendage. The systemic-to-pulmonary artery shunt or right ventricle-to-pulmonary artery conduit is exposed and occluded after cardiopulmonary bypass is instituted, and the patient's core temperature is reduced to 18°C. During this cooling phase, the right and left pulmonary artery branches are exposed in preparation for generous patch augmentation, so as to avoid proximal pulmonary arterial obstruction from unrecognized irregularities in size. The aorta is then cross-clamped, and cardioplegic solution is infused in the aortic root after the circulation is arrested and blood is drained into the venous reservoir. The pulmonary arteries are opened by a single incision extending from behind the right-sided superior vena cava, and just proximal to the right upper lobe branch, across the midline to the left lower lobe branch. An incision is then made in the dome of the right atrium and extended across the right atrium superior vena cava junction, towards the medial aspect of the right-sided superior vena cava immediately adjacent to the incision in the right pulmonary artery. The interatrial communication is inspected and enlarged if necessary. A side-to-side anastomosis between the superior vena cava and the pulmonary arteries is begun between the apex of the incised right pulmonary artery and the apex of the incision on the medial side of the right-sided superior vena cava. This is performed with a running monofilament suture (**Figure 41.12**). The construction of the superior vena cava/pulmonary artery confluence is completed by patch augmentation of the pulmonary arterial incision with an elongated patch of pulmonary homograft material, creating a roof over the anastomosis between the SVC and the pulmonary arteries. This patch begins on the left pulmonary artery and extends under the neo-aorta until it reaches the anastomosis between the right pulmonary artery and right-sided superior vena cava (**Figure 41.13**). The sutures are tied to the suture line of the SVC–RPA anastomosis. The suture on the lower lip of the pulmonary arteriotomy is used to anchor the base of the triangular pulmonary homograft patch to the floor of the superior vena cava–right atrial junction. A separate suture line is carried between the superior lip of the incision in the right atrium and the homograft patch, closing the junction of the superior vena cava

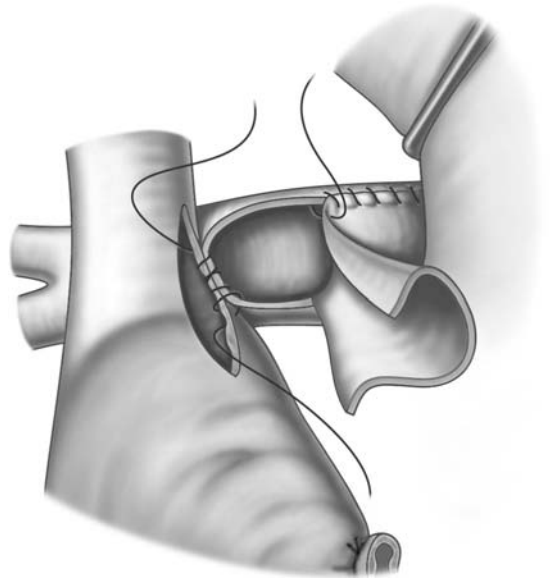

Figure 41.12

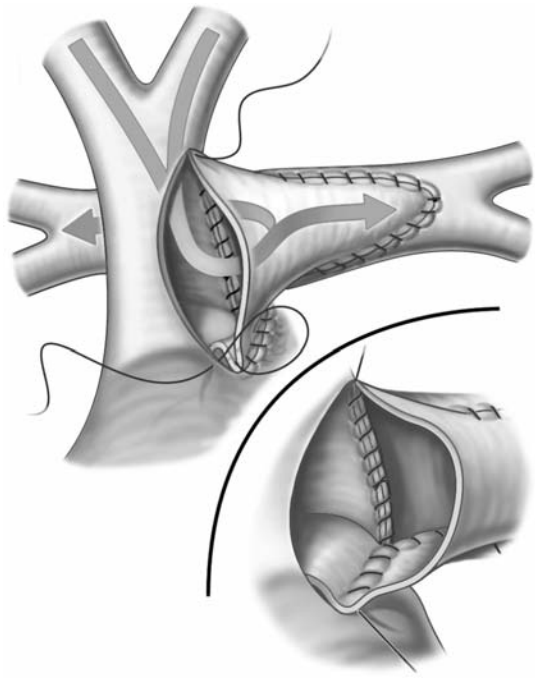

Figure 41.13

with the right atrium, thereby creating a dam which occludes the inflow of the SVC into the right atrium and directs the flow from the superior vena cava into the augmented pulmonary arteries (**inset, Figure 41.13**). If a left SVC is present, it is connected to the left pulmonary artery in the same fashion. When bilateral superior vena cavae are present, the left SVC is transected, the cardiac end closed and the distal end anastomosed end-to-side to the left pulmonary artery. The central pulmonary artery augmentation starts on the right side of this anastomosis and is carried towards the right SVC; the remaining reconstruction is as described. Cardiopulmonary bypass is re-established after the suture line closing the right atriotomy is completed, and the patient is warmed to 37°C. Placement of a pressure-monitoring catheter through the right atrial appendage enables postoperative assessment of the end-diastolic pressure of the single ventricle.

Haemodynamic issues related to aortic arch obstruction, atrio-ventricular valve insuffiency, anomalous pulmonary venous drainage, and additional sources of pulmonary blood flow resulting in additional volume or pressure load of the single ventricle should be recognized and promptly addressed, in order to achieve optimal outcome at the time of Fontan completion.

The interval between stages must permit restoration of the optimal ventricular compliance at its new end-diastolic dimension. After remodelling of the right ventricle for a period of 6–12 months following volume-unloading surgery (hemi-Fontan), patients undergo evaluation in consideration for Fontan completion. The data obtained is interpreted in light of the changes anticipated as a result of the Fontan completion.

MODIFIED FONTAN PROCEDURE

The goals of the Fontan procedure are to restore near-normal oxygen saturation, eliminate the risk of paradoxical embolization, and prevent the development of pulmonary arteriovenous malformations. Following the hemi-Fontan procedure, Fontan completion is a rather simple technical exercise, requiring little dissection, which minimizes the risk of bleeding and injury to important structures. We usually accomplish this by channelling the inferior vena cava to the pulmonary arteries with an intra-atrial PTFE baffle (lateral tunnel). Using cardiopulmonary bypass and a brief period of circulatory arrest, a right atriotomy is made parallel and anterior to the sulcus terminalis. To connect the inferior vena cava to the pulmonary arteries, a piece of PTFE tubing 7–10 mm in diameter is cut in half lengthwise and used as a baffle for channelling inferior vena cava flow along the lateral aspect of the right atrial wall to the dam created to close the superior vena cava entrance into the right atrium. The baffle is sewn with a running 5–0 monofilament suture around the orifice of the inferior vena cava, along the floor of the right atrium, immediately anterior to the edge of the atrial septal defect, and around the patulous orifice in the superior vena cava entrance, created by merely excising the homograft dam placed 6 months earlier (**Figures 41.14** and **41.15**). The superior aspect of this lateral tunnel is then completed by incorporating the upper edge of the PTFE baffle in the suture line closing the atriotomy. The lateral tunnel technique was introduced to minimize complications associated with tricuspid prolapse or regurgitation or with obstruction of pulmonary venous return to the right ventricle, which was experienced early in our series with an alternative technique. In order to lower the systemic venous pressure, a deliberate right-to-left shunt could be created by placing a fenestration in the mid-portion of the PTFE baffle. Our preference is to routinely place a 4 mm fenestration in all patients, which, if it remains patent and depending on the haemodynamic characteristics (arterial oxygen saturation, haematocrit level, exercise capacity) could be closed in the cardiac catheterization laboratory.

The rationale for this staged approach is that in some patients, diastolic volume diminishes rapidly after removal of the excess volume load in association

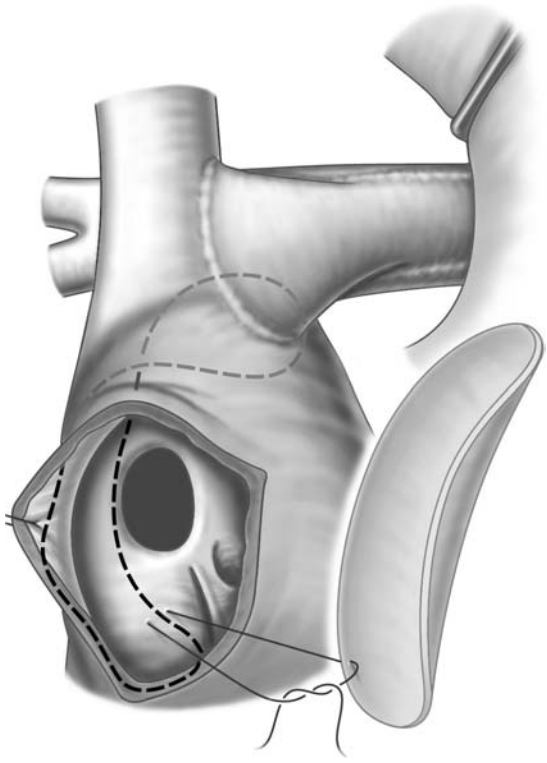

Figure 41.14

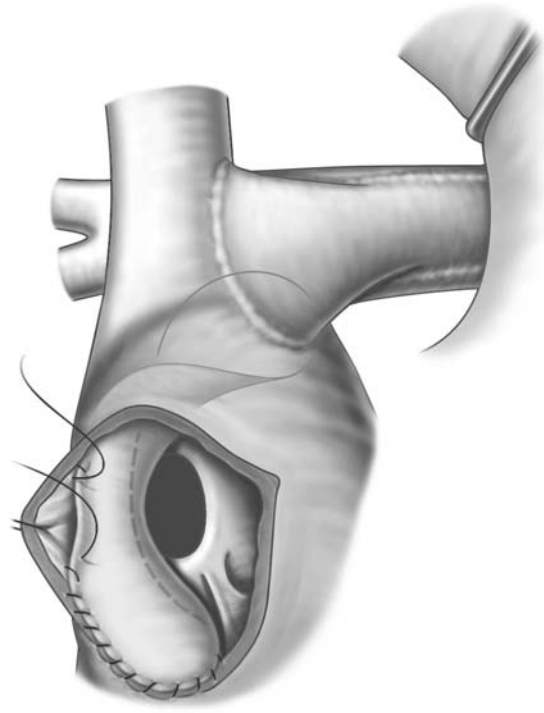

Figure 41.15

with a systemic-to-pulmonary artery shunt. Moreover, preoperative physiological information does not enable the surgeon to predict which patients will develop rapid contraction of end-diastolic volume. The anatomical and physiological consequences of this process are an apparent increase in wall thickness, with impairment of the diastolic function of the ventricle, which results in an increase in end-diastolic pressure, an increase in central venous pressure and a decrease in cardiac output. To minimize the physiological significance of such a rapid geometrical change, dividing the Fontan procedure into two parts is recommended in order to allow a decrease in end-diastolic volume and a commensurate decrease in muscle mass, while only half of the systemic venous return is obliged to flow passively through the pulmonary vascular bed. When the ratio between the end-diastolic volume and wall thickness has normalized, the Fontan procedure may be completed more safely.

RESULTS

The knowledge and experience gained in the management of patients with hypoplastic left heart syndrome have led to a significant improvement in the outcome of reconstructive surgery. The most recent report from the Congenital Heart Surgeon Society (Ashburn *et al*., 2003), which includes 710 neonates with critical aortic stenosis or aortic atresia undergoing Stage I Norwood between 1994 and 2000, revealed a 72%, 60% and 54% survival at 1 month, 1 year and 5 years, respectively, which constitutes an overall improvement compared to the 64%, 50% and $< 47\%$ survival for reconstructive surgery in the study undertaken between 1994 and 1997. Incremental risk factors for death after Stage I Norwood included lower birth weight, older age at operation, associated anomalies, smaller size of the ascending aorta and longer circulatory arrest time. A number of strategies introduced to assist in the perioperative management of these patients have resulted in improved survival (Gullquist *et al*., 1992; Tweddell *et al*., 1999); however, none of them have modified the challenging physiology present in these patients until the time of a second-stage procedure. The use of a right ventricle-to-pulmonary artery conduit as part of the Norwood reconstruction has been revisited, with hospital survival rates of 80–90% in several centres worldwide (Pizarro *et al*., 2003; Sano *et al*., 2003; Mair *et al*., 2003). However, due to the non-randomized and retrospective nature of these single-centre reports, the

true impact of this surgical modification could only be elucidated with longer follow-up and a multicentre study, such as the forthcoming NIH sponsored single ventricle repair trial.

At present, our institutional management scheme is palliation of the newborn, a hemi-Fontan procedure at the age of 5–6 months, followed by completion of the Fontan at the age of 12–18 months. The majority of patients we see today have the diagnosis made by foetal echocardiogram and the incidence of moribund presentation has decreased to the betterment of overall outcome. Notwithstanding, even the sickest patients can often be rescued by urgent pulmonary artery banding, followed by Stage I Norwood or deferral of reconstructive surgery for several months after stenting of the ductus arteriosus, which has become a common approach for patients with important comorbidities or with very low weight. Our recent experience between May 2001 and June 2004 with 85 patients who underwent Stage I Norwood, using a conduit between the right ventricle and pulmonary arteries, resulted in 8 early deaths. These deaths were related to technical issues in two cases at the beginning of the series and the remaining cases involved patients with a variety of high-risk conditions, including low birth weight, prematurity, associated genetic conditions, etc. Moreover, interim mortality between the Norwood procedure and hemi-Fontan is very rare, accounting for only one death. Among 77 survivors, 50 had the hemi-Fontan operation at a median age of 5.5 months, with two deaths. A few patients (6/50) underwent the hemi-Fontan procedure at a median age of 4.3 ± 0.8 months, due to progressive hypoxaemia, commonly related to distal conduit stenosis, without mortality. Fifteen patients had a completion Fontan without mortality.

The approach outlined in this chapter for reconstructive surgery of hypoplastic left heart syndrome will most certainly continue to evolve. The ever-increasing power of catheter-driven therapy in the catheterization laboratory will be increasingly brought to bear on this complex group of patients, not eliminating surgery but rather enhancing it. Based on recent follow-up studies, the challenge for the future is to develop management strategies which could not only improve survival rates, but result in a good functional outcomes for this challenging group of patients.

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42

Congenital Malformations of the Mitral Valve

A. Carpentier and C. Brizard

Congenital malformations of the mitral valve are rare, complex and frequently associated with other congenital malformations of the heart or the aorta that may hide or be hidden by the valve malformation. Malformations of the mitral valve in atrio-ventricular septal defect, atrio-ventricular discordance and straddling mitral valve are excluded from this chapter. Echocardiography allows a precise diagnosis of the type of malformation and its functional consequences. The surgical management has been transformed by modern reconstructive techniques introduced by the senior author in the past 30 years.

CLINICAL PRESENTATION

The malformations may present as predominant valve stenosis or predominant valve regurgitation.

Mitral Valve Stenosis

Congenital mitral valve stenosis belongs to the category of anomalies that causes pulmonary venous hypertension. A mitral diastolic murmur with right ventricular and left atrium hypertrophy seen on electrocardiograms suggests a mitral valve stenosis. Establishing the diagnosis may be difficult in the presence of a systolic murmur, which may or may not be related to associated lesions. Atrial fibrillation is extremely rare in children, even in the advanced forms. Echocardiography study is essential, not only for the initial diagnosis of mitral valve stenosis but also in the recognition of the anatomy. The optimal age for operation depends on several factors, such as physical disability, growth retardation and the presence of pulmonary hypertension, but mostly on the expected difficulty of surgery and on the annular diameter. Operation is often considered necessary within the first few years of life. The average age in our initial clinical series was 5.1 years (Carpentier *et al*., 1976). Whenever possible, it is preferable not to operate before the age of 6 months because in infancy, particularly under 3 months of age, the collagen has not matured adequately and the valve tissues are particularly fragile and difficult to manipulate. In children with severe pulmonary hypertension, the operation should not be delayed beyond the age of 18 months.

Mitral Valve Regurgitation

Congenital mitral valve regurgitation may lead to repeated pulmonary infections, cardiac failure and eventually to failure to thrive. The disease is, however, tolerated slightly better than is congenital mitral valve stenosis, and whenever affected, children usually undergo operation at a later age (average 6.1 years in the author's early clinical series).

Whenever possible, the operation should be delayed until ideally as late as 6 or 8 years, an age at which a relatively large annuloplasty ring can be used. In practice, repeated episodes of cardiac failure and progressive enlargement of the systolic diameter (evaluated with the Z value) indicates a deterioration of the systolic function. This must be recognized and should indicate an operation, whatever the age.

PREOPERATIVE VALVE ANALYSIS. THE FUNCTIONAL CLASSIFICATION

Trans-thoracic and trans-oesophageal echocardiography allows classification of the malformations according to the motion of the leaflets and the morphology of the papillary muscles, in one of the three following types (Carpentier *et al*., 1976) (**Figure 42.1**):

- Type I: Normal leaflet motion. The regurgitation results from a lack of coaptation between the leaflets.
- Type II: Leaflet prolapse. The free edge of one or the two leaflets overrides the plane of the orifice during systole.

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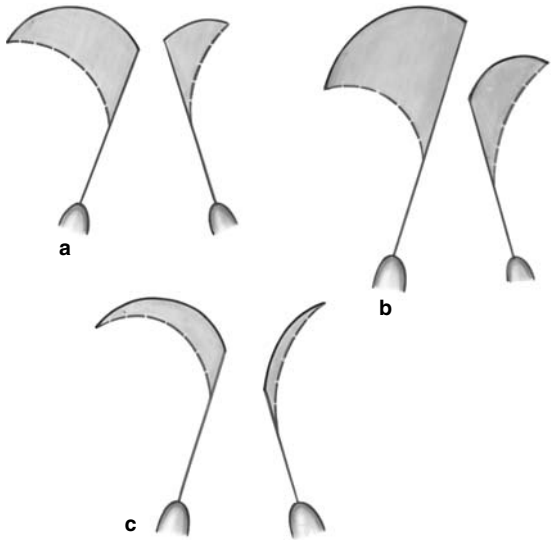

Figure 42.1

- Type III: Restricted leaflet motion. The motion of one or the two leaflets is limited. Two subgroups are described, depending upon the morphology of the papillary muscles.
- Type III_1: Restricted leaflet motion, with two clearly visible papillary muscles.
- Type III_2: Restricted leaflet motion, with abnormal papillary muscles.

In addition to the ''functional analysis'', ''segmental valve analysis'' by trans-oesophageal echocardiography allows precise localization of the leaflet dysfunction (Carpentier *et al*., 1995). The mitral valve is arbitrarily segmented in eight areas (**Figure 42.2**), clearly identifiable by echocardiography using a transgastric view.

In addition to valve analysis, echocardiography allows assessment of the volumes and contractility of the two ventricles and recognition of associated lesions, which were present in 60% of the cases in our series.

It is rarely possible by echocardiography to make a precise diagnosis of all the lesions responsible for the valvar dysfunction. The precise diagnosis is made intraopoeratively.

SURGICAL TECHNIQUE

Approach

The approach is through a midline sternotomy. Associated coarctation can be treated during the same operation, either through the midline sternotomy incision, using cardiopulmonary bypass, or through a separate left thoracotomy as a first-stage operation. Minimally invasive surgery, although becoming more popular in adult patients, is not recommended in children.

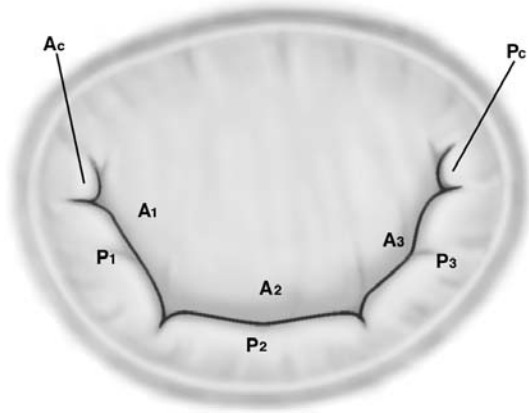

Figure 42.2

Cardiopulmonary Bypass and Myocardial Protection

Using conventional sternotomy approach, an aortic cannula is inserted into the ascending aorta just below the innominate artery. A needle for removing air is also inserted into the ascending aorta, 1 cm below the aortic cannula. This needle is connected to both a suction line and a cardioplegia line through an Y connector. The venae cavae are cannulated separately and surrounded by tapes. The child's temperature is lowered to 32°C in simple repair and to 28°C in complex repair. Myocardial protection is achieved by blood cardioplegia. The myocardial temperature is monitored. The amount of cardioplegic solution given is the minimum required for reaching a myocardial temperature of 10°C. Topical cooling with iced water is used to obviate repeated doses of cardioplegic solution. Only whenever the myocardial temperature reaches 15°C is another volume of cardioplegia given.

Intraoperative Valve Analysis and Repair

After dissection of the interatrial groove to approach the mitral valve, the left atrium is opened. The atrial cavity is inspected to recognize jet lesions or thrombus formation. The components of the mitral valve leaflets, chordae and papillary muscles are then examined. The aim of this analysis is to confirm the echocardiography findings, to recognize the type of malformation and to make final precise diagnosis.

MITRAL VALVE STENOSIS

Congenital Mitral Valve Stenosis with Normal Papillary Muscles

In this group of mitral stenoses, four lesions can be encountered: commissure fusion, excess valvar tissue, supravalvar ring and annular hypoplasia (**Table 42.1**).

Commissure Fusion/Short Chordae

In this common condition, one or two papillary muscles are directly implanted on the commissure without any intermediate chordae tendineae (**Figure 42.3a**). The commissures are fused and thickened. The motion of the leaflets is restricted. The central orifice of the mitral valve between the two papillary muscles is severely narrowed, and the valve may have a funnel-shaped appearance. A *forme fruste* may exist, with short chordae instead of papillary commissure fusion. Another form associates one papillary commissure fusion and short chordae from the other papillary muscles. This condition is treated by commissurotomy, papillary muscle fenestration and resection of secondary chordae, so as to restore adequate leaflet motion (**Figure 42.3b**). The repair is not difficult and good results can be expected. Valve replacement is necessary only in exceptional cases.

Table 42.1 Valve dysfunction and anomalies in a series of 50 children with congenital mitral valve stenosis (Carpentier, 1994).

Valve stenosis with normal papillary muscle	
Commissure fusion	17
Excess valvar tissue	2
Supra-valvar ring	9
Annular hypoplasia	2
Valve stenosis with abnormal papillary muscle	
Parachute valve	9
Hammock valve	11

Excess Valvar Tissue and Double Mitral Orifice

In this condition, the interchordal spaces are obstructed by abnormal valvar tissue (**Figure 42.4a**). The mitral valve apparatus is otherwise normal. Angiocardiography of this lesion may show a funnel-shaped orifice. Echocardiography may be normal, or abnormal echoes underneath the leaflets may be present. In some cases, a large bridge of valvar tissue joins the anterior and posterior leaflets, thus delineating an accessory orifice of the mitral valve. Interchordal space obliteration must be corrected by chordal fenestration, i.e. removal of the excess tissue which obliterates the interchordal spaces (**Figure 42.4b**). Whenever a double orifice is present, the cause of the stenosis must be searched for at the subvalvar level (imperforated interchordal spaces), since the double orifice itself is usually not stenotic and must be left intact in order not to create a regurgitation.

Supravalvar Ring

This circumferential ridge of fibrous tissue is truly supravalvar at the level of the mural leaflet (2–3 mm above the annulus) but intravalvar at the level of the

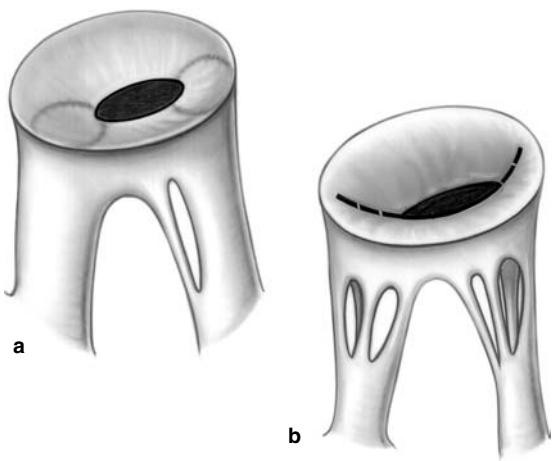

Figure 42.3

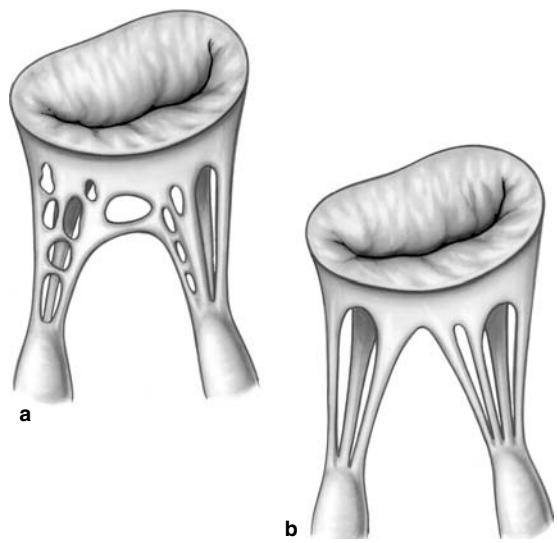

Figure 42.4

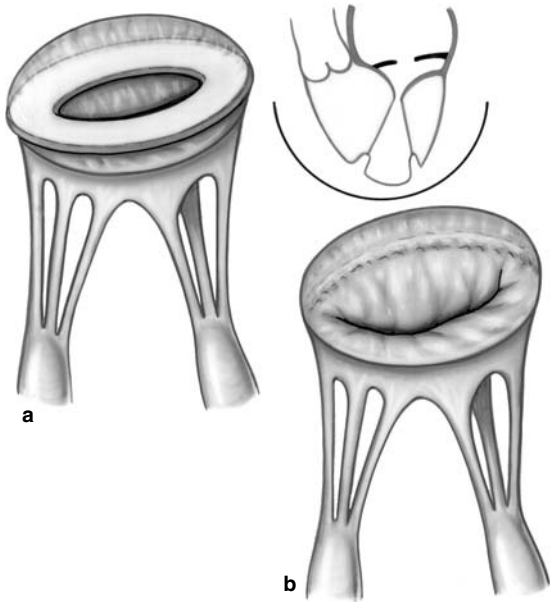

Figure 42.5

anterior leaflet to which it is attached, 3–5 mm below the annulus (**Figure 42.5**). This malformation can be recognized by angiocardiography, echocardiography or both. However, it may be discovered only at operation. Depending on the diameter of the ring, various degrees of obstruction may exist. The supravalvar ring may be the only malformation, the mitral valve underneath being otherwise normal, although slightly hypoplastic. More often, the supravalvar ring is associated with a parachute valve, a hammock valve, or any other mitral valve malformation. In the latter conditions, the ring may play a part in the functional obstruction. Consequently, when a supravalvar ring is found at operation, the entire valve apparatus should be examined in order to recognize associated lesions. Surgical correction consists of resection of the fibrous tissue, taking care not to damage the anterior leaflet (**Figure 42.5a,b**).

Hypoplasia of the Annulus

Underdevelopment of the mitral valve annulus can be associated with each of the malformations just described. It should be clearly distinguished from mitral valve hypoplasia and mitral valve atresia, which constitute part of the hypoplastic left heart syndrome. The size of the mitral valve is compared with ''normal'' references for age and weight (Rowlatt *et al*., 1963) (**Table 42.2**).

Table 42.2 Mean normal valve diameters.

Body surface area	Valve diameter (mm)			
(m^2)	Mitral	Tricuspid	Aortic	Pulmonary
0.25	11.2	13.4	7.4	8.4
0.30	12.6	14.9	8.1	9.3
0.35	13.6	16.2	8.9	10.1
0.40	14.4	17.3	9.5	10.7
0.45	15.2	18.2	10.1	11.3
0.50	15.8	19.2	10.7	11.9
0.60	16.9	20.7	11.5	12.8
0.70	17.9	21.9	12.3	13.5
0.80	18.8	23.0	13.0	14.2
0.90	19.7	24.0	13.4	14.8
1.00	20.2	24.9	14.0	15.3
1.20	21.4	26.2	14.8	16.2
1.40	22.3	27.7	15.5	17.0
1.60	23.1	28.9	16.1	17.6
1.80	23.8	29.1	16.5	18.2
2.00	24.2	30.0	17.2	18.0

Modified from Rowlatt UF, Rimoldi HJA and Lev M: The quantitative anatomy of the normal child. *Pediatr. Clin North Am 10:499,1963*. The approximate standard deviations are 1.9 mm (body surface area $< 0.3\,m^2$) and 1.6 mm (body surface area $> 0.3\,m^2$) for mitral valve and 1.7 mm (body surface area $< 1.0\,m^2$) and 1.5 mm (body surface area $> 1.0\,m^2$) for tricuspid valve.

Mitral Valve Stenosis with Abnormal Papillary Muscles

This group consists of three malformations:

- Parachute mitral valve (single papillary muscle).
- Hammock valve (multiple papillary muscles).
- Absence of one or both papillary muscles.

Parachute Mitral Valve

The parachute mitral valve is one of the most common malformations causing mitral valve stenosis. In this condition, all chordae are attached to a single papillary muscle (**Figure 42.6a**). The single papillary muscle is formed either from the fusion of the two papillary muscles or from the development of one of the papillary muscles, the second being either hypoplastic or adherent to the muscular ventricular wall without any chordal connection. The stenosis results from obliteration of the interchordal spaces by excess valvar tissue. In some cases, a commissural cleft, usually anterior, corresponding to the absent papillary muscle, is the

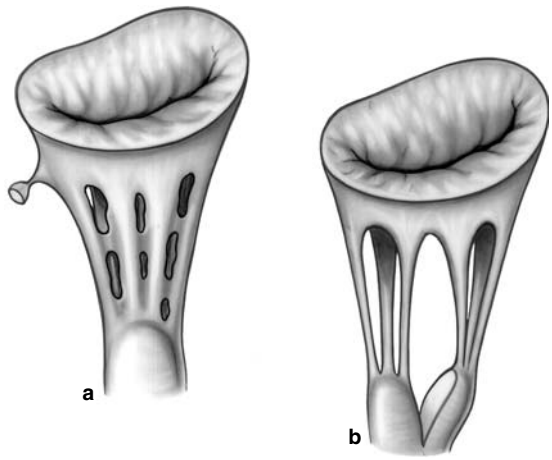

Figure 42.6

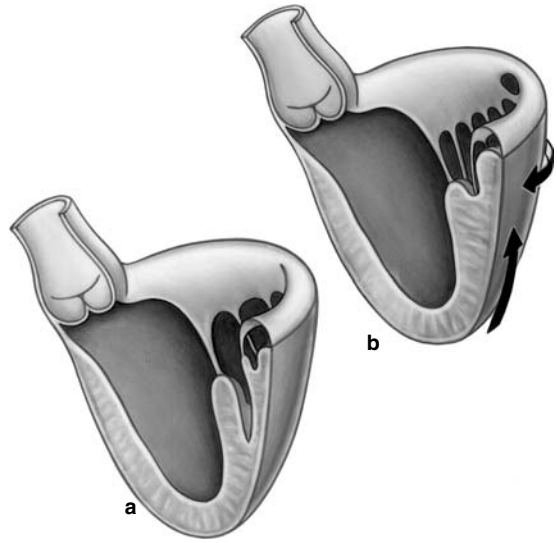

Figure 42.7

only orifice that allows blood to flow from the left atrium to the left ventricle. The presence of a parachute mitral valve anatomy does not automatically warrant a surgical indication. Some parachute mitral valves are functionally adequate. The gradient through a restrictive paprachute valve should remain stable over the follow-up. If the gradient increases, one should suspect a supravalvar mitral ring (Schaverien *et al*., 2004).

The parachute mitral valve can be corrected by splitting the papillary muscle and fenestrating the interchordal spaces (**Figure 42.6b**). A parachute valve is often associated with left ventricular outflow tract obstruction or an aortic coarctation, and more rarely these two malformations and a supra-annular ring (Shone *et al*., 1963).

Hammock Valve

The term ''hammock valve'' refers to the atrial aspect of various subvalvar anomalies (Carpentier *et al*., 1976). The valvar orifice is partially obstructed by intermixed chordae and abnormal papillary muscles, characteristically implanted underneath the mural leaflet (**Figure 42.7a**). This malformation includes various lesions that have been described in the literature as mitral arcade, obstructive papillary muscles, hypertrophied papillary muscles, and ''typical'' congenital stenosis (Carney *et al*., 1962; Castaneda *et al*., 1969; Davachi *et al*., 1971). When looking carefully at the description given by these authors, these malformations actually follow a similar pattern: the two normal papillary muscles are absent, in their place are numerous papillary muscles and fibrous bands implanted high on the posterior wall of the ventricle underneath the mural leaflet. **Figure 42.7a** shows such a valve; the arrows indicate the lateral and upward displacement of the papillary muscles with, as a result, obstruction of the mitral valve orifice. The chordae tendineae of the anterior leaflet cross the orifice toward the posteriorly implanted papillary muscles, producing the hammock appearance. The leaflet tissue is often normal and pliable. There is not one central orifice but several orifices, in contrast to other types of mitral stenosis. In extreme cases, the hammock valve contains a fibrous diaphragm with scattered holes that allow the blood to flow from the left atrium to the left ventricle. Echocardiography usually reveals either restricted or normal motion of the anterior leaflet, and multiple echoes underneath a restricted posterior leaflet. The angiogram may show a characteristic filling defect on the posterior wall of the ventricle, just beneath the mural leaflet (Carpentier *et al*., 1976). This malformation is the most difficult to correct by reconstructive techniques (**Figure 42.7b**). The surgeon should try to separate the anterior leaflet from the posterior leaflet by splitting the muscular mass transversely. Then all of the chordae and papillary muscle that are not attached to the free edges of the leaflets must be removed in order to free the inflow chamber. A temporary incision in the mural leaflet, followed by patch enlargement, may provide better access to the subvalvar apparatus. Valve replacement is often necessary. The hammock valve is frequently associated with aortic stenosis, coarctation of the aorta, persistent ductus arteriosus or ventricular septal defect.

Absent Papillary Muscles

Absence of the papillary muscles rarely gives rise to mitral valve stenosis. In this entity, numerous intermixed

chordae are attached to the ventricular wall with imperforate interchordal spaces. Although this condition can be treated by interchordal space fenestration in selected cases, valve replacement is often necessary.

MITRAL VALVE REGURGITATION

The lesions are classified into three groups according to the leaflet motion: normal, prolapsed or restricted (Carpentier *et al*., 1976; Chauvaud *et al*., 1998) (**Table 42.3**).

Normal Leaflet Motion

In this group, the motion of the leaflets is normal. The regurgitation is due to a lack of apposition of the opposing leaflets or to a cleft or to a leaflet defect.

Annular Dilatation and Deformation

The existence of congenital primary dilatation of the mitral annulus has been questioned by various authors. One of us, however, has encountered this malformation in four cases of mitral valve regurgitation diagnosed within the first few days of life. There was no associated coarctation or subvalvar aortic stenosis. The dilatation mainly affected the mural leaflet. The leaflet tissue was slightly thickened as a result of secondary lesions. In two cases, minor abnormalities of the mitral valve tissue, such as excess tissue or imperforated interchordal spaces, were present and further supported the diagnosis of congenital origin. An ostium secundum atrial septal defect was associated with this annular anomaly in two cases.

Table 42.3 Functional anomalies and lesions in a series of 105 congenital mitral valve regurgitation (Carpentier *et al*., 1976).

Type I: Normal leaflet motion	
Annulus dilatation	11
Cleft leaflet	16
Leaflet defect	3
Type II: Prolapsed leaflet	
Elongated chordae	32
Elongated papillary muscle	18
Absent chordae	3
Type III: Restricted leaflet motion	
Normal papillary muscles	
Papillary muscle commissural fusion	5
Short chordae	5
Abnormal papillary muscles	
Parachute	4
Hammock	8

Children suffering from this malformation usually do not require surgery before the age of 5 years. The heart is enlarged, as well as the mitral valve tissue, so that a prosthetic ring annuloplasty is possible in most cases. Paediatric prosthetic rings (Heart Institute, Ho Chi Minh City, Vietnam) 22–30 mm in diameter are used, with a corresponding orifice area of 2.5–4 cm. The technique of insertion of the prosthetic ring in children does not differ from that already described for adults. The selection of the ring is based on the measurement of the base of the aortic leaflet. One stay suture is placed at each commissure, and the distance between these two points is measured with obturators. A prosthetic ring of appropriate size is then selected. Fifteen sutures are placed through the annulus. These sutures are then passed through the sewing ring of the prosthesis, care being taken to keep the same spacing for the sutures of the anterior leaflet while spacing is reduced for the sutures of the posterior leaflet. This measured annuloplasty corrects not only the dilatation but also the deformation of the annulus without impairing leaflet motion or reducing the orifice area. Long-term experience has shown that prosthetic rings sizes 22–26 mm are well tolerated and only slightly stenotic up to age 20–25, at which age they can be replaced by larger rings. Leaflet extension using glutaraldehyde-treated autologous pericardium helps when using larger ring sizes. The buffered glutaraldehyde solution can be prepared by the pharmacy of the hospital according to the formula described previously (Carpentier *et al*., 1969).

Before the age of 6, it is preferable to avoid the use of a prosthetic rings. In this situation, mitral valve incompetence may be corrected by one or two triangular resections at the base of the mural leaflet, followed by plication of the annulus and suture of the leaflet edges (**Figure 42.8a**). The resection should be performed electively in the areas of indentations of the mural leaflet. Whenever the dilatation is too important to be treated by this technique, leaflet expansion using glutaraldehyde-treated autologous pericardium is performed (**Figure 42.8a**). A prosthetic ring with leaflet expansion using glutaraldehyde-treated autologous pericardium is shown in **Figure 42.8b**.

Cleft Leaflet

The condition called cleft leaflet usually affects the anterior leaflet. It may be of two types: cleft anterior leaflet or the atrio-ventricular canal mitral valve. The latter could be more appropriately called ''three-leaflet mitral valve'', because it may exist in the absence of atrio-ventricular septal defect (Carpentier, 1978).

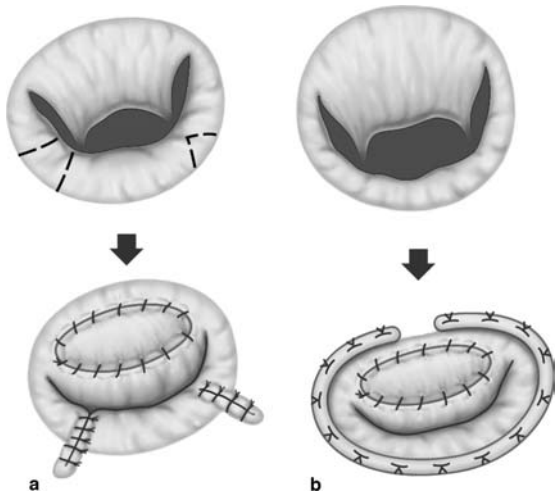

Figure 42.8

Cleft Anterior Leaflet

In this condition, contrary to the mitral valve in atrioventricular septal defect, the mitral valve apparatus is normal, with the exception of a cleft that vertically separates the anterior leaflet into two hemi-leaflets (**Figure 42.9**). The cleft is directed towards the right and non-coronary interleaflet triangle. The two papillary muscles are normal and normally located, with the two strut chordae connected to the respective aortic fibrous trigone. The commissures are correctly positioned, and the mural leaflet attachment represents two-thirds of the circumference of the mitral orifice, as in normal hearts. Abnormal chordae tendineae are often attached to the free edges of the cleft.

In most cases, the cleft can be closed after resection of the abnormal chordae. A prosthetic ring may also be necessary (**Figure 42.9**). In some instances, it is necessary to use a glutaraldehyde-treated pericardial patch to fill the gap between the two edges of the leaflets, particularly if the annulus is dilated (**Figure 42.10**). Commissuroplasties at the anterolateral and posterolateral commissures may be added. Accurate analysis of the valve after injection of saline into the left ventricle is crucial for demonstrating the location of the leakages.

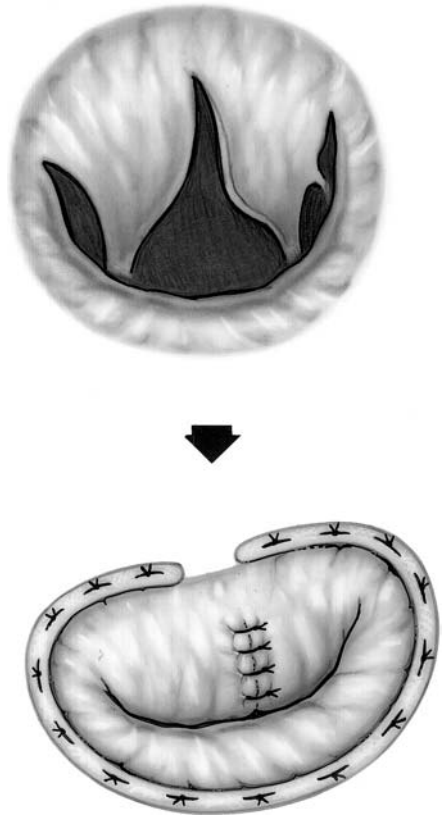

Figure 42.9

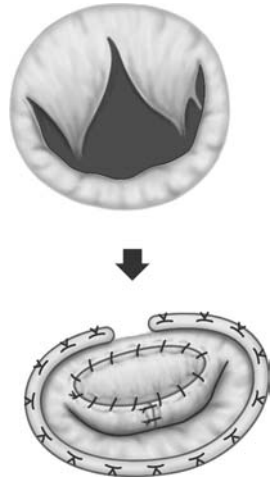

Figure 42.10

Three-leaflet Mitral Valve

A three-leaflet mitral valve may be encountered in the absence of septal defects. The regurgitation results from poor coaptation of the edges of the valvular components and from enlargement of the annulus. The closure of the cleft is mandatory, either by interrupted sutures or by patching with glutardehyde-treated autologous pericardium. Commissuroplasties at the anterolateral and posterolateral comissures are often indicated.

Cleft Posterior Leaflet

This condition is rare and is often confused with enlargement of the normal indentations of the mural leaflet, secondary to annular dilatation. These indentations, well described by Ranganathan *et al*. (1970), are normal features of the mural leaflet. Only clefts that affect the leaflet tissue between these indentations may be regarded as true clefts. Correction is performed by suturing the free edges after plication of the annulus.

Leaflet Defect

Various defects, such as holes or localized agenesis of the leaflet tissue, may be encountered, particularly in the mural leaflet. The free edge of the defect may be free of chordae or may attach thin chordae tendineae. The combination of young age of the child and absence of any history of bacterial endocarditis is evidence of the congenital nature of these lesions. A defect in the mural leaflet is treated by rectangular resection of the corresponding part of the leaflet and suturing of the free edge of the leaflet remnants after a sliding plasty of these remnants has been carried out (Carpentier, 1988) (**Figure 42.11**). A defect in the anterior leaflet is treated by direct suturing, or pericardial patching whenever the size of the defect does not make possible a direct suture without tension.

Leaflet Prolapse

The term ''leaflet prolapse'' should be restricted to the condition in which the free edge of one or two leaflets override the plane of the orifice during systole (Carpentier *et al*., 1976). This functional definition excludes the billowing mitral valve (Barlow's syndrome), in which excess valvar tissue balloons within the atrium during systole, while the free edge of the leaflets remain in apposition underneath the plane of the orifice. The lesions responsible for leaflet prolapse are of three types:

Figure 42.11

Figure 42.12

absence of chordae, elongation of chordae, and elongation of papillary muscle.

Absence of Chordae

Only the margin or a whole leaflet segment may be free of chordae. The first condition can be treated by transposition of secondary or basal chordae to the free edge. The second condition is treated by leaflet quadrangular resection after annulus plication (**Figure 42.12**). Gore-Tex chordae may be useful, with the limitation that they do not grow with age. However, some adaptation mechanism may take place, as recently reported (Matsumoto *et al*., 1999; Privitera *et al*., 2005).

Elongated Chordae Tendineae

Chordae elongation usually involves all of the chordae tendineae arising from one papillary muscle. The corresponding portion of the leaflet tissue is prolapsed within the atrium during systole. The cause of the chordal elongation is unknown in most instances, except whenever excess tissue, billowing leaflets and myxoid degeneration suggest Barlow's disease. Such a condition has been encountered by one of us in an 8 year-old child in whom a mitral valve regurgitation had been diagnosed at the age of 3 months. This case supports the hypothesis that Barlow's syndrome may be a genetic disorder (Disse *et al*., 1999). Elongation of chordae has also been described in association with Marfan's syndrome.

Elongation of chordae can be treated by the following technique. A groove is made within the anterior aspect of the papillary muscle (dotted line, **Figure 42.13**); a 5-0 suture is then passed through each side of the groove and around the elongated chordae (**Figure 42.13b**). Tying of the suture buries the excessive length of the chordae in the groove, which is subsequently closed (**Figure 42.13c**). Alternatively, papillary muscle sliding plasty, in which the papillary muscle supporting the elongated chordae is sutured to an adjacent shorter papillary muscle, can be performed. When using any of these sliding papillary muscle techniques, one should make sure not to have

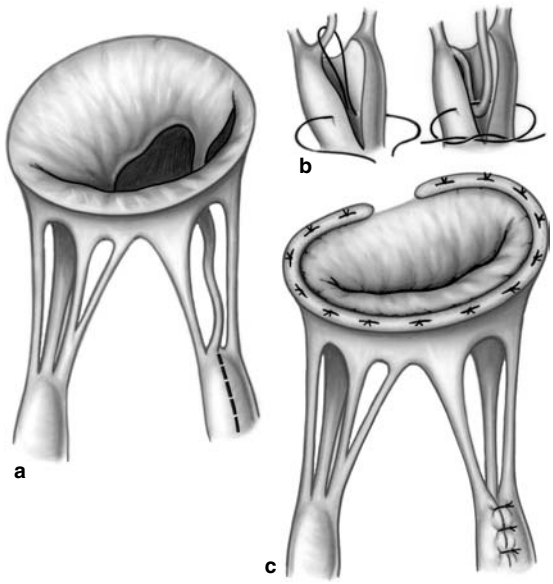

Figure 42.13

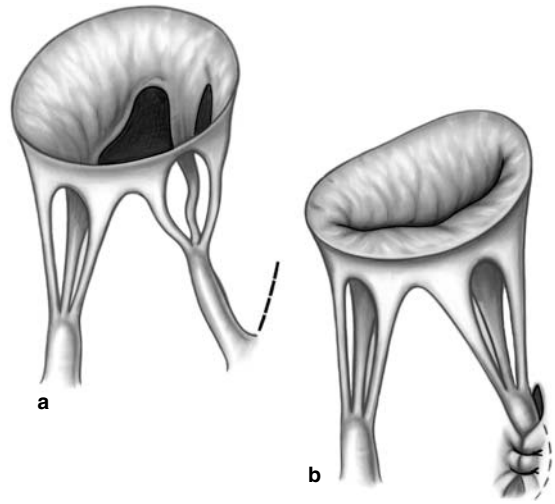

Figure 42.14

any suture in contact with chordae, to avoid chordae abrasion. Associated dilatation of the annulus, if present, is corrected by annulus remodelling.

Elongation of Papillary Muscle

Elongation of one papillary muscle that leads to leaflet prolapse is usually associated with other cardiac anomalies, such as anomalous origin of the left coronary artery from the pulmonary trunk or aortic stenosis. Papillary muscle elongation may exist in the absence of other intracardiac or coronary artery abnormalities, which justifies the categorization of this lesion as a congenital malformation. Yet there are good reasons to think that this lesion results from a papillary muscle ischaemia. The papillary muscle is thin, flattened and elongated to such an extent that its tip can prolapse through the mitral orifice. The papillary muscle either has a white fibrotic appearance or is yellowish with a necrotic appearance. Histological specimens usually show scar tissue similar to that seen in a healed infarction. Infarcted areas may also be seen in the adjacent subendocardial layer of the left ventricle, demonstrating abnormal vascularization. Papillary muscle elongation can be corrected by plication of the papillary muscle whenever it is fibrotic, or by papillary muscle repositioning whenever it is necrotic. In the latter technique, a trench is created within the muscular wall of the left ventricle, just above the implantation of the papillary muscle (**Figure 42.14a,b**). The body of the papillary muscle, with the exception of its extremity, is buried within the trench, restoring appropriate tension on the chordae. The trench is subsequently closed (Carpentier *et al*., 1976).

Restricted Leaflet Motion

Restricted Leaflet Motion with Two Well-identified Papillary Muscles

Two lesions belong to this subgroup: commissural fusion and short chordae.

Commissural Fusion

At first glance, the lesions seem similar to those described for rheumatic valve disease. Detailed analysis, however, shows that the tissue obliterating the commissural area is covered by an abnormal endothelial tissue, sometimes multiperforated, without the characteristic fibrosis and calcifications seen in rheumatic valvular disease. Underneath the commissural area, chordae and hypertrophied papillary muscles may adhere to the commissure. Mitral valve regurgitation results from either hypoplasia or a defect of the anterior leaflet or annular dilatation, or both. This condition is treated by commissurotomy and leaflet extension (**Figure 42.15**). Incision in the abnormal commissural tissue may create commissural incompetence, which can be treated by secondary chordae transposition to the free edges of the commissure. Valve extension is preferred to annulus remodelling but a prosthetic ring may still be necessary in case of severe annulus dilatation (**Figure 42.15b**).

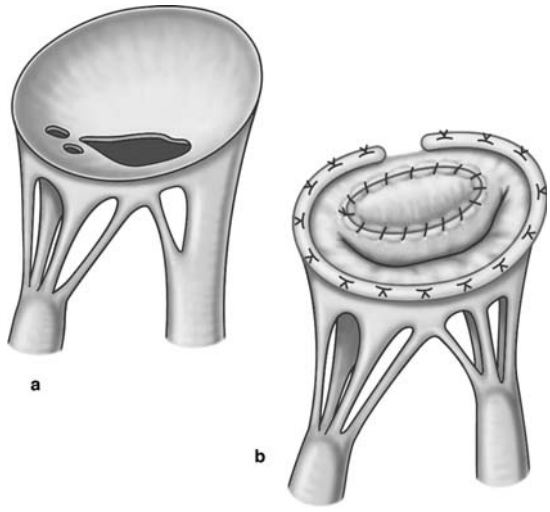

Figure 42.15

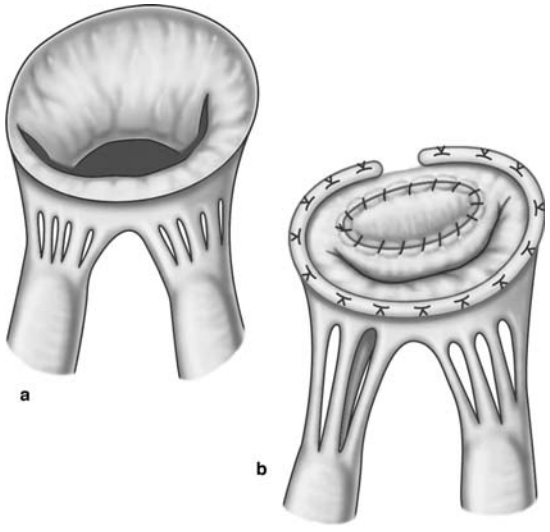

Figure 42.16

Short Chordae

Short chordae tendineae may limit the mobility of the leaflet, thus creating mitral valve regurgitation (**Figure 42.16**). In contrast to rheumatic valve disease, these chordae are well delineated, only slightly thickened and not fused. The interchordal spaces are reduced, which, in conjunction with papillary muscle hypertrophy, may create an associated subvalvar stenosis. The annulus is often enlarged secondary to the mitral valve regurgitation. Surgical correction of these malformations consists of fenestration of the papillary muscle, to enlarge the interchordal spaces, restore the mobility of the leaflets and relieve the subvalvar stenosis. Leaflet extension and/or annular remodelling is then performed to restore the apposition of the mitral valve leaflets (**Figure 42.16b**).

Restricted Leaflet Motion with Abnormal Papillary Muscles

Echocardiography and, whenever possible, MRI make it possible to analyse both the location and the morphology of the papillary muscles and thus enable recognition of the three malformations: parachute valve, hammock valve and papillary muscle agenesis.

Parachute Mitral Valve

Although usually purely stenotic, a parachute mitral valve can be associated with a regurgitation. The regurgitation is usually due to the hypoplasia of the anterior leaflet near one commissure, usually the anterior one, a malformation also called ''cleft commissure'' (Carpentier *et al*., 1976). In addition to the release of the valve stenosis as described above, the repair of the regurgitation is achieved by anterior leaflet extension, using glutaraldehyde-treated autologous pericardium. Prosthetic ring annuloplasty may be added.

Hammock Valve

As in the parachute mitral valve, the hammock valve is usually stenotic. Whenever a regurgitation is associated, it results from leaflet hypoplasia or restricted leaflet motion. Posterior leaflet extension and prosthetic ring annuloplasty may be used to correct the regurgitation in addition to the techniques described to release the stenosis. Temporary detachment of the posterior leaflet greatly facilitates the release of subvalvar stenosis.

Papillary Muscle Hypoplasia or Agenesis

Poor development or absence of one or both papillary muscles may lead to mitral valve incompetence. Chordae are attached to tiny papillary muscles or directly to various points of the ventricular wall. Valve regurgitation is caused by either abnormal tension on these chordae or underdevelopment of the leaflet tissue. Valve repair may necessitate leaflet extension, Gore-Tex chordae, and annular remodelling. Surgical correction may be performed with the use of a rigid ''classic'' prosthetic ring, the curvature of which may be modified so as to compensate the lack of tissue. Valve replacement, however, is often necessary.

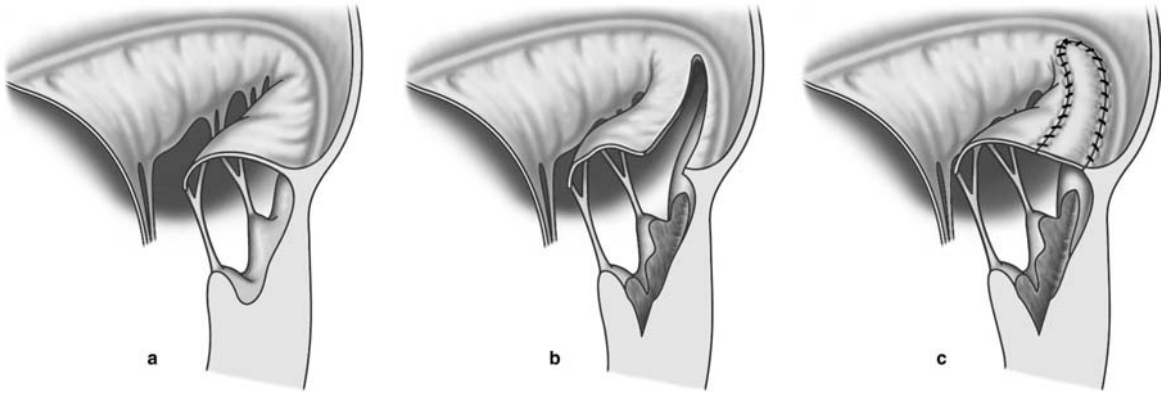

Figure 42.17

ASSESSMENT OF VALVE REPAIR

The repair is tested by injecting saline through the valve into the ventricular cavity, using a 250 ml bulb syringe after suction is established through the aortic needle. The result is judged to be satisfactory when the following conditions are achieved:

1. The closure line of the two leaflets lies parallel to the posterior portion of annulus.
2. The surface of coaptation between the two leaflets is greater than 3 mm.
3. Leaflet mobility is satisfactory.

An asymmetrical line of closure indicates residual prolapse or residual restricted motion of one leaflet. Small leakages have little importance as long as the three previously exposed criteria are present: they usually disappear or remain trivial once the valve is working under normal systolic pressure.

VALVE REPLACEMENT

Valve replacement is necessary in 10–15% of cases of congenital mitral valve malformation. The choice of an artificial valve in children used to present a dilemma. In fact, mechanical valves are the only option, as mitral valve homografts have not produced satisfactory results in children (Chauvaud *et al*., 2003) and have to be considered as a palliation to allow for annular growth. The early models of bioprosthesis have led to disappointing results in children, with an incidence of prosthesis failure as high as 50% at 4 years (Antunes and Santos, 1984). For this reason, valvular bioprostheses remain contraindicated in children. It remains to be seen whether the most recent methods of calcium mitigation will make it possible to change this policy (Carpentier *et al*., 1998).

CONGENITAL MITRAL VALVE ANOMALIES IN NEWBORNS AND INFANTS

The main technical difficulties in surgical treatment of the mitral valve in this age group are the size of the annulus and the size of the valvar orifice to gain access to the subvalvar apparatus. On the other hand, the tissues are usually free of secondary lesions, i.e. thickening or retraction, as would be seen in older patients. Surgical constraints due to the size of the annulus lead to the more liberal use of detaching the posterior leaflet or the anterior leaflet to gain access to the subvalvar apparatus (**Figure 42.17**).

The annuloplasty technique is also adapted to the size of the annulus. All surgical strategies in some way are designed to gain time before a definitive procedure can be performed. Congenital mitral valves which require surgery in infancy have been associated in the past with poor results. This has been improved with the evolution of this surgery in the last decade. On the one hand, valve repairs have significantly improved; on the other hand, recent publications have shown that mitral valve replacements with mechanical prostheses have improved as a result of better management of anticoagulation and a better knowledge of the consequences of using small-sized prostheses. Anticoagulation therapy with warfarin, when adequately monitored, is safe in children beyond 6 years of age but ''younger children have a significantly more complicated course'' (Streif *et al*., 1999).

Mechanical valve replacement has a good prognosis, provided that the prosthesis is implanted in anatomical position and that the concept of valve re-replacement is accepted. The short- and long-term survival is related to the ratio of prosthetic valve size to patient native mitral annulus (Caldarone *et al*., 2001). The need for prosthesis re-replacement is dictated by the size of the

first prosthesis implanted and the age at first implantation (Raghuveer *et al*., 2003).

Accordingly, for each congenital mitral valve patient, a strategy must be devised in which the replacement of the mitral valve using supra-annular implantation or an oversized prosthesis is avoided. Ideally, mitral valve replacement should be planned, after one or two palliative mitral valve repairs in order to allow the annulus to grow. Then one should try to place as large a prosthesis as possible in the anatomical position. In the interim, an imperfect result should be accepted. Residual regurgitation is better tolerated than stenosis and maximizes the growth of the annulus. In experienced hands, the valve in congenital mitral regurgitation, presenting beyond infancy, can almost always be successfully repaired.

Anatomy

The most frequent group of congenital mitral valve problems in this age group is the one with abnormal anatomy. The lesions are dominated by a lack of valvar tissue and the same anatomy can be found in predominant stenosis or predominant regurgitation. The three main anatomical anomalies in this age group are: papillary muscle to commissure fusion, hammock valve and parachute mitral valve.

The second most frequent group are valves with normal anatomy and associated congenital cardiac anomalies, generating large shunts. Functionally, there is severe regurgitation, with moderate or severe type II prolapse of the anterior leaflet. The lesion is an elongated chordae/papillary muscle. Although an acquired lesion, the supravalvar ring can be encountered very early in life. It has to be suspected every time the transvalvular gradient increases during follow-up, or when the gradient is greater than what the echocardiographic Doppler investigation has suggested.

The Shone syndrome deserves special attention in this age group. Patients symptomatic with the association are symptomatic from birth, with duct-dependent circulation. The mitral valve anatomy is most often a parachute mitral valve, but functionally it can be predominantly regurgitant. It is often difficult to link the severity of the clinical condition to one of the two systemic valves involved. In our experience in Melbourne, we accept that at some stage both valves will have to be treated.

Clinical Presentation and Investigations

In this age group, for both stenosis and regurgitation, the clinical presentation is of cardiac failure, with dyspnoea on exertion (feeding) and tachypnoea. A severe failure to thrive is usually present. Patients who present at 2–4 months of age with normal mitral anatomy, severe valve regurgitation and depressed systolic function suggest, and should be suspected to have, an anomalous coronary artery arising from the pulmonary artery (ALCAPA), which has to be ruled out before considering a functional aetiology. Clinical examination shows a large liver and cold extremities. Auscultation is difficult. A strong apical systolic murmur should indicate a mitral regurgitation, whereas a diastolic murmur in the case of mitral stenosis can be difficult to pick up or absent in the context of low cardiac output or associated left-to-right shunting at the atrial level. ECG demonstrates left atrial enlargement and left ventricular hypertrophy in the presence of mitral regurgitation. Right ventricular hypertrophy is constant. The chest X-ray shows pulmonary congestion and a cardiac enlargement. The echocardiographic study is essential in establishing the diagnosis, the functional consequences and, to a lesser degree, the anatomy. Because of the size of the mitral valve, the differential diagnosis between the three main groups of anomalous anatomy can be very difficult and should be accepted with caution. The functional consequences of the anomaly (trans-mitral gradient and degree of pulmonary hypertension) are important data to collect for evaluation of the severity of the anomaly and for follow-up but, *at this age*, do not represent an indication for surgical intervention.

Medical Treatment

Medical treatment is given to delay surgery. Diuretics are given in the context of mitral stenosis and vasodilators and transfusion in the presence of mitral regurgitation. Elective surgery at this age is indicated for severe failure to thrive despite maximum therapy. Alternatively, inotropic support and mechanical ventilation are indicated for acute severe cardiac failure episodes. Surgery is indicated if the baby fails to wean from ventilation support.

Surgical Treatment

Surgical techniques used for the correction of mitral valve regurgitation or stenosis do not differ from those described earlier in this chapter; they are adapted for size. Emphasis has to be placed on adequate surgical exposure. We use a self-retaining retractor adapted to the size of the patient in all cases. Venous drainage is achieved through bicaval cannulation. The bypass temperature is routinely 32°C. Myocardial protection is achieved with repeated cold/tepid blood cardioplegia. Topical cooling is facilitated by division of left ventricular adhesions when necessary. The valve is approached through the interatrial groove. The approach through the interatrial septum is avoided, as it does not allow for adequate room for the retractor hooks and exposes the conduction

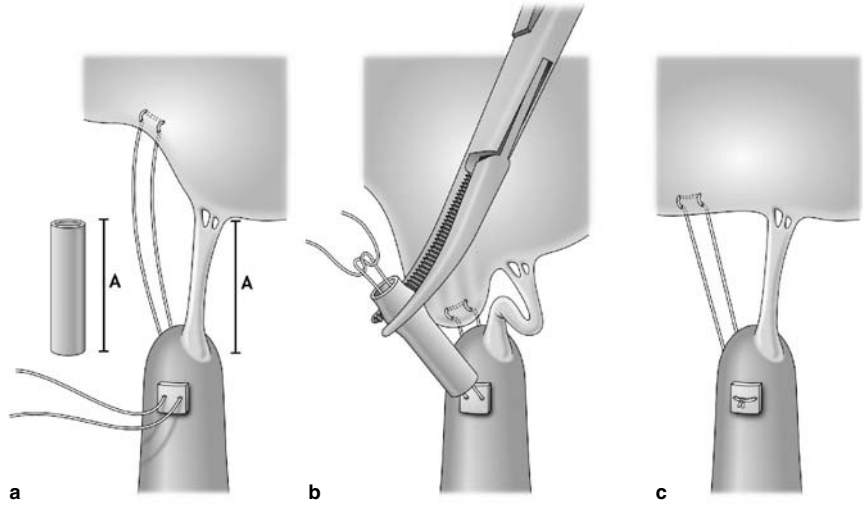

Figure 42.18

tissue to greater stretch and pressure. The exposure is completed by the placement of mattress sutures into the posterior annulus to pull the valve anterior and to the right. The snugger around the inferior vena cava has to be pulled anterior and to the left. Autologous pericardium is obtained every time a leaflet extension is deemed necessary. The pericardium is trimmed of fat and treated with 0.625% glutaraldehyde for 8 minutes.

In patients with abnormal mitral valve anatomy, correction of moderately enhanced leaflet motion is rarely necessary. In patients with normal anatomy and prolapsed anterior leaflet (type II) associated with a large shunt (VSD or PDA), it is corrected usually by wedge resection or, more rarely, by chordal shortening when there are chordae available. When none of these techniques is appropriate, we use Gore-Tex chordae. In this age group special attention should be being taken to avoid overcorrection, which is the usual pitfall of this technique. **Figure 42.18a–c** shows our technique for Gore-Tex chordae implantation.

Annuloplasty is mandatory for all cases of mitral valve regurgitation. In this age group, with mitral valve annuli less than 24 mm, no remodelling annuloplasty rings are available commercially. We use a posterior annuloplasty with a Gore-Tex conduit of 3, 3.5 or 4 mm (**Figure 42.19**). The continuity of the annuloplasty can be interrupted once or twice to allow for subsequent growth. If only a less than perfect result can be achieved, a complete posterior annuloplasty will jeopardize the annular growth and subsequent valve replacement. In this situation, we compromise with an annuloplasty limited to the area of greatest restriction of the

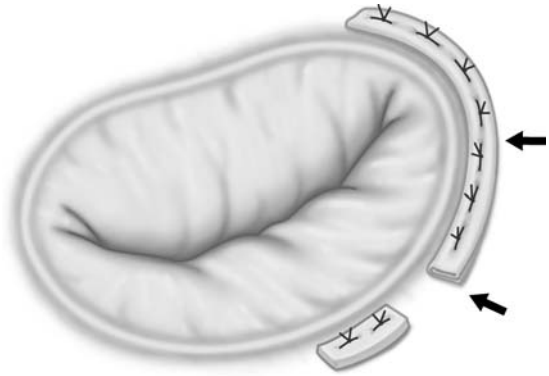

Figure 42.19

posterior leaflet and augmentation of the latter. The correction of a restricted leaflet motion (type III) is usually done by patch augmentation of the posterior leaflet. This technique allows for the preservation of a larger mitral annulus and is used in combination with the detachment of the posterior leaflet for exposure purposes (**Figure 42.17a–c**). Lack of anterior leaflet tissue is compensated by anterior leaflet extension, usually without annuloplasty.

RESULTS

Many publications reporting valve repair or replacement in the paediatric age group include left atrio-ventricular valves or even rheumatic valves. This may convey an incorrect notion of favourable prognosis or unnecessary

need for valve replacement. The authors' experiences encompass three series of children with true congenital malformations of the mitral valve who benefited from a systematic attempt to repair rather than to replace. These series are only summarized here, the details being found in the referenced articles (Carpentier *et al*., 1976; Chauvaud *et al*., 1998).

In a series of 155 children below the age of 12 operated on for congenital mitral valve regurgitation ($n = 105$) or congenital mitral valve stenosis ($n = 50$) between March 1970 and August 1991, valve repair was possible in 98 children (93%) in the regurgitation group, and 47 (94%) in the stenotic group, with a hospital mortality of 5% and 26%, respectively. The main cause of death was low cardiac output in the regurgitation group and pulmonary hypertension in the stenotic group. In the follow-up of this series (maximum 21 years, mean 7 years), reoperation was necessary in 6% of the children with valve regurgitation and 11.7% of the children with valve stenosis. The main cause of reoperation was residual or recurrent valve regurgitation in those patients having not had a ring annuloplasty, i.e. 100% of valve stenosis and 50% of valve regurgitation. It is interesting to underline that in three patients a re-do valve repair could be performed thanks to a generous use of leaflet extension and the systematic use of ring annuloplasty. The actuarial rate of freedom from all events including death and reoperation was 47% at 10 years for mitral valve stenosis and 62% at 15 years for valve regurgitation (**Figure 42.20**).

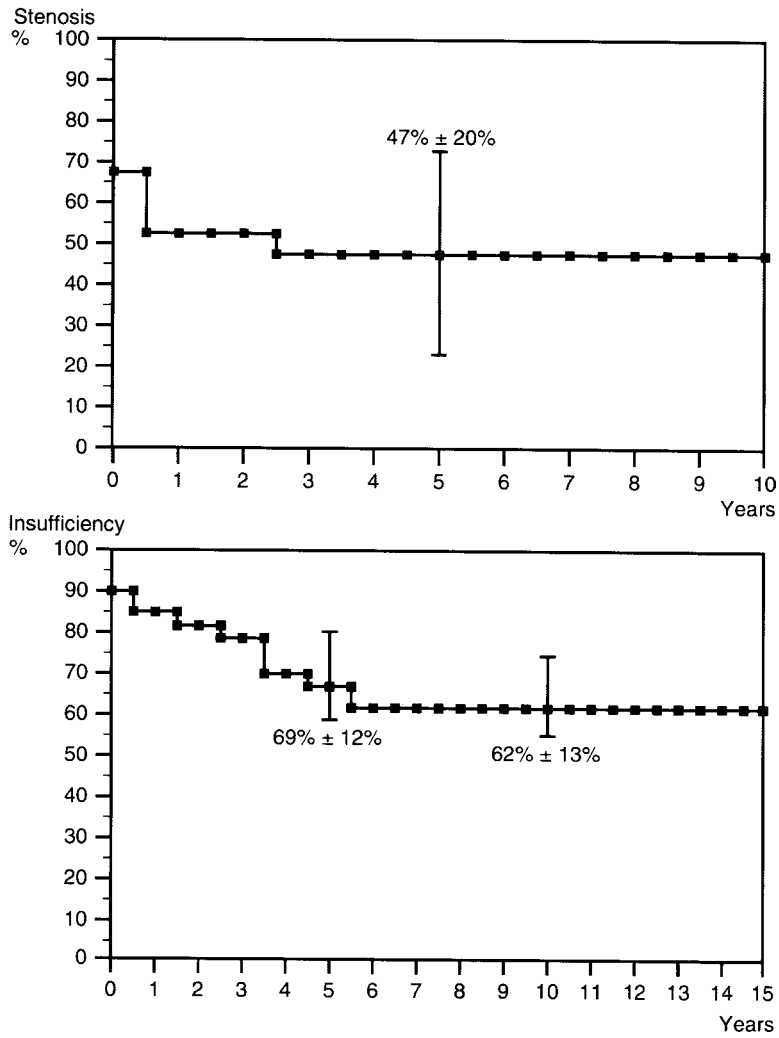

Figure 42.20

In order to compare valve repair vs. replacement and to analyse in detail the causes of reoperations, the valve regurgitation group of 105 children has been reassessed with a longer follow-up and completed by an additional number of 40 children, so that the new study comprises 145 children operated upon for congenital mitral valve regurgitation between 1970 and 1995 (Chauvaud *et al*., 1998). The age range was 0.17–12 years (mean 5.7 ± 3.1). Valve dysfunction and pathology did not differ significantly from the first series (**Table 42.4**).

Valve repair was possible in 138 patients (95%). Among these, 70 patients required a prosthetic ring annuloplasty and 21 patients a valve extension with a pericardial patch (**Table 42.5**). Valve replacement was necessary in only seven patients (5%). The in-hospital mortality was 5% (95% CL, 2.5–9.9%) (seven patients). Mean follow-up was 9.3 ± 6.9 years (1–26 years), and cumulative follow-up was 1142 patient-years. No early death occurred in the group of patients who underwent valvular replacement. Actuarial survival at 24 years was 88% in patients who underwent valve repair and 51% in patients who underwent valve replacement (**Figure 42.21**).

Late operation was necessary in two patients who had a valve replacement with a bioprosthesis (40%) and in 21 patients (15%) who had undergone a valve repair. Actuarial freedom from reoperation was 68% (95% CL, 80.5–51.5%) at 15 years, and the linearized

Table 42.4 Mitral valve dysfunction and lesions in a series of 145 children classified according to the Carpentier functional approach. Reprinted from Journal of Thoracic Cardiovascular Surgery, volume 115, Chavaud, S. Reconstructive surgery in congential mitral valve insufficiency (Carpentier's techniques): Long-term results. p 84–92 ♥ Copyright (1988), with permission from American Association for Thoracic Surgery.

Type	(*n*)	(%)
Type I: Normal leaflet motion	31	21.4
Annular dilatation	7	4.8
Cleft leaflet	18	12.4
Leaflet defect	6	4.1
Type II: Leaflet prolapse	79	54.5
Chordal elongation	46	31.7
Papillary muscle elongation	24	16.5
Absence of chordae	9	6.2
Type III: Restricted leaflet motion	35	24.1
Normal papillary muscles	15	
Commissure fusion	8	5.5
Short chordae	7	4.8
Abnormal papillary muscles	20	
Parachute mitral valve	5	3.4
Hammock mitral valve	12	8.2
Papillary muscle hypoplasia	3	2

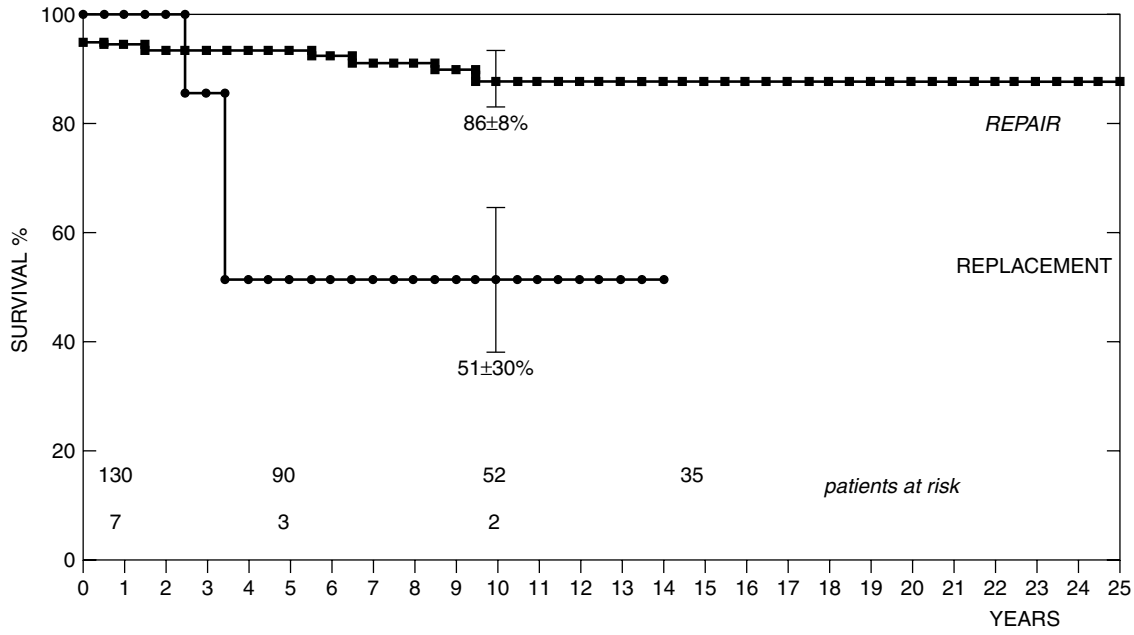

Figure 42.21

Table 42.5 List of the different Carpentier's repair techniques used in 138 patients who underwent valve repair for mitral valve regurgitation (Reprinted from Journal of Thoracic Cardiovascular Surgery, volume 115, Chavaud, S. Reconstructive surgery in congential mitral valve insufficiency (Carpentier's techniques): Long-term results. p 84–92 ♥ Copyright (1988), with permission from American Association for Thoracic Surgery).

Procedures	(*n*)	(%)
Annuloplasty		
Carpentier prosthetic ring	70	48
Plication of the commissures	6	4
Plication of the annulus	5	3
Semicircular suture	5	3
Cleft suture without patch	14	10
Pericardial patch enlargement		
Anterior leaflet	14	10
Posterior leaflet	8	5
Chordal shortening	46	33
Papillary muscle shortening	24	17
Chordal transposition	12	8
Commissurotomy	15	10
Splitting of papillary muscle	13	9
Fenestration of papillary muscle	9	6

rate of exposure to reoperation was 1.9%/patient-year (**Figure 42.22**).

The causes of reoperations included a left ventricular failure developed 18 years after the initial repair in one patient. In this patient the mitral valve was functioning well and a cardiomyoplasty was performed, with a good result. Residual or recurrent regurgitation was present in 17 patients and mitral valve stenosis in three but only four patients required an early reoperation (1–10 days) after the initial repair. Among these, one patient died after the reoperation and is included in the hospital mortality. Five patients in whom a prosthetic ring was not implanted during the initial operation underwent an implantation of a prosthetic ring during the reoperation, with a good result. Amongst those having had a ring implanted, only three patients (4.2%) required a reoperation for mitral valve stenosis, 14.2, 6.5 and 4 years after the initial operation. The size of the ring was 26 mm in two patients. In these patients an adult-sized prosthetic ring could be implanted after leaflet enlargement, with an excellent result. None of the children who underwent a leaflet enlargement by autologous pericardium needed a reoperation for calcification or retraction. Amongst the 20 patients reoperated on for mitral valve dysfunction, 11 could benefit from a second repair and nine required a valve replacement. A mechanical valve was used in seven and a biological prosthesis in two. The latter two had yet another (a third) operation for degeneration of the bioprosthesis at 3.5 and 5 years.

At the Royal Children's Hospital in Melbourne, nine neonates or infants less than 12 months old had 12 mitral valve procedures for mitral valve regurgitation between 1996 and December 2004. All underwent valve repair. Two reoperations involved valve replacement. One patient received a 21 mm bileaflet prosthesis 7 months after the initial repair. The moderate residual regurgitation with limited posterior annuloplasty became severe but allowed the annulus to grow from 13 mm to 21 mm at the time of reoperation. The second patient with Shone syndrome had the mitral valve replaced after initial mitral valve repair (papillary muscle-to-commissure fusion) and Ross procedure in the neonatal period. The initial surgery with limited posterior annuloplasty generated moderate residual regurgitation. A 12 mm homograft mitral valve was implanted following early failure of the initial repair. The patient died from the complications of severe bilateral bronchomalacia; there was moderate residual MR. The third reoperation involved a repeated repair of a flail anterior leaflet after papillary muscle splitting. Artificial chordae were used with a good stable result. There was one early death in an extreme neonatal Marfan patient. The severe mitral regurgitation had been successfully corrected.

During the same time span (1996–2004), five patients less than 10 months old were operated on for congenital mitral stenosis. The median age was 5 months (range 1 week–10 months). All had severe failure to thrive and severe pulmonary hypertension. The mean preoperative gradient was 13 ± 2.3 mmHg. The malformations included papillary muscle-to-commissure fusion ($n = 2$), parachute mitral valve ($n = 2$) excess tissue ($n = 1$) and supravalvular ring ($n = 1$). One patient had Shone's syndrome. Two patients underwent reoperations. The first patient had three reoperations, leading ultimately to valve replacement with a mechanical valve (diameter 23 mm). The second patient (neonatal Shone syndrome valve and interrupted aortic arch) had a reoperation at 2 years for recurrent obstruction. He had a supravalvular membrane resected. So far, all patients have a satisfactory result.

CONCLUSION

Congenital mitral valve regurgitation can be repaired in infancy with a low mortality. Valve repair is achievable in the majority of cases for both congenital mitral valve insufficiency and stenosis. These techniques offer stable long-term results with a low rate of reoperation. A larger use of leaflet extension since 1980 has allowed us to

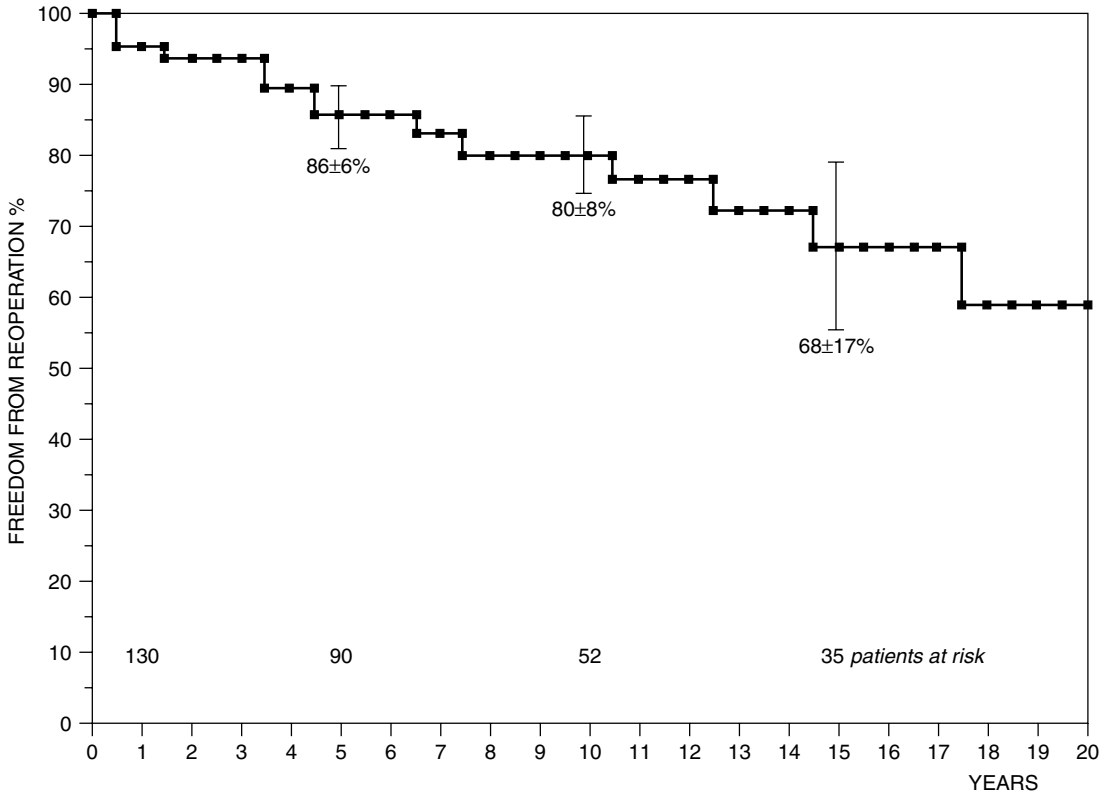

Figure 42.22

implant larger prosthetic rings, a policy which has been associated with a reduced risk of reoperations.

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43

Malformations of the Tricuspid Valve and Ebstein's Anomaly

A. Carpentier and C. Brizard

Tricuspid valve stenosis, tricuspid valve regurgitation and Ebstein's anomaly are described in this chapter, which does not cover the tricuspid valve when severe stenosis or atresia of the right ventricular outflow tract is present. It also excludes right atrio-ventricular valve in complete or partial atrio-ventricular septal defect.

Ebstein's anomaly is the first described and most common malformation of the tricuspid valve. It is characterized by the downward displacement of the septal and the posterior leaflets into the right ventricle at the junction of the inlet and trabecular portions (Ebstein, 1866). The incidence is low amongst patients with congenital heart disease, but earlier diagnosis has been seen since the advent of two-dimensional (2D) echocardiography. The aetiology of other congenital tricuspid regurgitation conditions is more rare. There are no displacements of the valve insertions but malformations or anomalies of some of the valve components (leaflets, chordae or papillary muscles). Isolated congenital tricuspid valve stenosis is very rare.

CONGENITAL TRICUSPID VALVE STENOSIS

Isolated congenital tricuspid valve stenosis with normal annulus is exceedingly rare. More often the valve, however well formed, is severely hypoplastic and the clinical manifestations are those of tricuspid atresia and should be treated as such.

Minor tricuspid valve hypoplasia with a small annulus has also been described in association with other defects, mainly ventricular septal defect. The gradient across the valve evolves as the child grows. Surgery is rarely indicated.

Generally, tricuspid valve stenosis is associated with stenosis or atresia of the right ventricular outflow tract, which is treated in the relevant chapter of this book.

CONGENITAL TRICUSPID VALVE REGURGITATION

Isolated congenital tricuspid valve incompetence is also very uncommon. Transient tricuspid regurgitation in a neonate without right ventricular outflow tract obstruction might reflect a form of ventricular or papillary muscle dysfunction, or may be a result of the late development of the septal leaflet. This rare entity should be treated conservatively. Severe tricuspid insufficiency seen from birth, related to papillary muscle ischaemic elongation or rupture, can also be seen and is amenable to simple and reliable repair. When associated with right ventricular hypoplasia and outflow tract obstruction, tricuspid valve incompetence can be functional or organic. In patients with organic lesions, there is great variation in the severity of the dysplasia, from slight nodular thickening of the leaflets to focal agenesis, thickening of the chordae, and direct attachment of the leaflets is a rare entity that should be treated conservatively. There are three main anomalies: (a) agenesis of one or two leaflets (**Figure 43.1a**); (b) annular distension (**Figure 43.1b**); and (c) absence of papillary muscles (**Figure 43.1c**). Surgical treatment is rarely indicated in neonates and infants; in older patients with symptomatic isolated tricuspid valve incompetence, tricuspid valve reconstruction is almost always possible. A patch of glutaraldehyde-treated autologous pericardium may be required in order to enlarge a hypoplastic leaflet, and artificial chordae are necessary to replace absent chordae or papillary muscle.

EBSTEIN'S ANOMALY

Ebstein's anomaly of the tricuspid valve is a rare congenital malformation that accounts for less than 1% of all congenital heart defects. The essence of the malformation is the downward displacement of the

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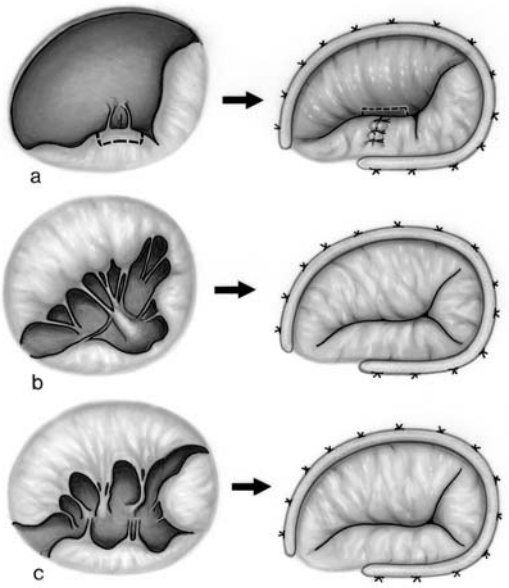

Figure 43.1

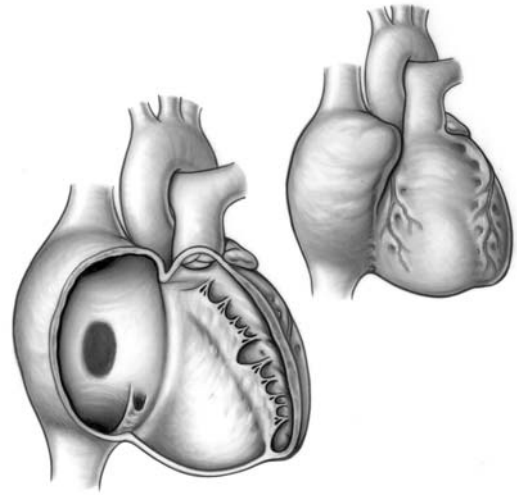

Figure 43.2

septal and part or whole of the posterior leaflets into the inlet portion. The apposition of the tricuspid leaflets is displaced towards the infundibulum and the apical trabecular portion of the right ventricle, creating the distal orifice of the tricuspid funnel. The part of the right ventricle beyond is the functional right ventricle; the part upstream is the atrialized inlet portion of the right ventricle (**Figure 43.2**). The tricuspid valve is often incompetent and, at the level of the distal orifice, sometimes stenotic. The anterosuperior leaflet seems to be in continuity with the posterior leaflet, without a clear commissure or attachment to the tricuspid annulus. Often it is large and thin. Close to the anteroseptal commissure, the leaflet is usually freely mobile but as the observer scans towards the posterior leaflet, the ventricular part of the valve tends to be tethered and then fused to the right ventricular free wall, while the hinge point itself becomes progressively displaced towards the apex of the right ventricle and away from the tricuspid annulus. There is marked obliteration of the interchordal space, almost constituting a veil. Rarely, the normal anterior and medial papillary muscles can be identified; more often, the anterior leaflet is attached directly to the right ventricle cavity (Carpentier *et al*., 1988).

SURGICAL ANATOMY

Ebstein's anomaly has three characteristic features: (a) a downward displacement of the septal and posterior leaflets into the right ventricle; (b) an atrialized portion of the right ventricle between the tricuspid annulus and the attachment of the posterior and septal leaflets (inlet portion); and (c) a malformation of the right ventricle, where the functional right ventricle is small and thin-walled and often has an abnormal systolic function. The trabecular portion is reduced; the infundibular portion may be partially obstructed by excess valvar tissue attachments of the anterior leaflet to the infundibulum. This is best appreciated with magnetic resonance imaging (MRI).

Depending on the severity of these malformations, Ebstein's anomaly can be categorized into four types (**Figure 43.3**):

- *Type a*. The anomaly is limited to the displacement of the septal leaflet with a small atrialized chamber. The anterior leaflet is large, moves freely and has a free leading edge between the anteroseptal and anterolateral commissures. (This type is not illustrated on **Figure 43.3**).
- *Type b*. The anterior leaflet is restricted, with some interchordal space obliteration and attachment of the ventricular surface to the ventricle by fibrous bands. The posterolateral and anteroseptal commissures are clearly delineated, the latter serving as a communication between the atrialized chamber and the functional right ventricle. The atrialized chamber is large **Figure 43.3b**.
- *Type c*. The anterior leaflet is partially fused to the infundibulum and the trabecular portion of the right ventricle. Interchordal spaces are obliterated.

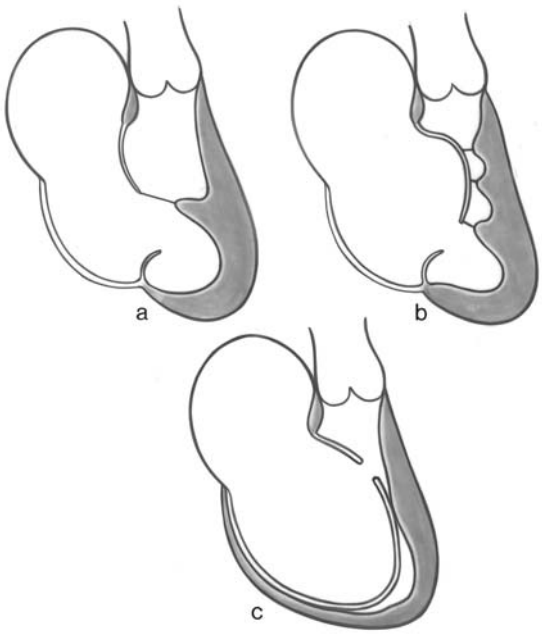

Figure 43.3

The ventricular edge of the valve is adherent to the ventricle, in continuity with the adherent septal and posterior leaflets. There is a marked displacement of the posterior and septal leaflets, which may be severely hypoplastic. The atrialized chamber is large and has a thin, non-contractile wall. The functional right ventricle is small and its contractility may be diminished **Figure 43.3c**.

- *Type d*. No anterior or posterior leaflets can be recognized, as they are totally adherent to the ventricular wall. The tricuspid orifice is deeply displaced towards the infundibulum and is often stenotic. The right ventricle is entirely made of the atrialized chamber **Figure 43.3d**.

DIAGNOSIS

The diagnosis of tricuspid disease is based mainly on echocardiographic findings. As for the mitral valve, the tricuspid valve can be analysed by means of the functional classification (see Chapter 42), which also serves as a guideline in valve reconstruction.

In addition to leaflet motion, valve analysis must define the level of attachment of the leaflets, the number and position of the papillary muscles and the geometry and function of the right ventricle. Associated anomalies, such as septal defects and pulmonary valve stenosis, are common and must also be recognized.

PRESENTATION

Cardiomegaly, at the expense of the right atrium and atrialized chamber, is the most typical radiological feature. It is almost constant and can be extreme. Cardiography will demonstrate right atrial hypertrophy, while AV conduction delay and intraventricular delay are common. A Wolff–Parkinson–White pre-excitation pattern is present in up to 25% of patients. Atrial fibrillation or atrial flutter, acute or recurrent, are often found in symptomatic patients. Atrio-ventricular re-entry tachycardia or pre-excitation require an electrophysiological study before surgery is performed. Often, ablation can be achieved successfully. Cardiac catheterization is no longer indicated. If performed, it would demonstrate relatively low right atrial and ventricular pressure and usually a right-to-left shunt at the atrial level.

The planning of Ebstein's anomaly repair requires morphological and functional information on the valve. These are best obtained with echocardiography.

Not only the functional result of a successful valve repair, but also the difficulty of the postoperative course or indications for an associated bidirectional cavopulmonary connection will depend on the size and the function of the right ventricle after the repair. This can best be predicted using MRI or Computed Tomography (CT). The surgical indication should not be dictated by symptoms or presence of arrhythmias but should be refined with exercise testing, associated if possible with calculation of maximal oxygen consumption.

MEDICAL MANAGEMENT

With the advent of 2D echocardiography and foetal echocardiography, the scope of presentation for Ebstein's anomaly has greatly evolved. Patients are more likely to present during foetal life or as neonates and infants than during childhood or as adults. The prognosis as well as the therapeutic options may vary greatly according to the age at presentation.

Various degrees of contribution of tricuspid valve incompetence, obstruction between the two parts of the right ventricle (tricuspid stenosis), small functional right ventricle, right-to-left shunt at the atrial level or arrhythmias to the clinical presentation, depending on the age, pulmonary resistance and anatomy, explain the great diversity of the latter. Symptomatic patients present with limitations of exercise ability, cyanosis, right heart failure or arrhythmias, or a combination of these.

Neonates and Infants

Neonates may have had antenatal diagnosis and are cyanotic at birth. High pulmonary resistance, with a combination of low systolic function of the functional right

ventricle and tricuspid regurgitation, generates a functional pulmonary atresia. Medical management relies on prostaglandin infusion and ventilation when necessary until the natural decline in the pulmonary resistances allows for anterograde pulmonary blood flow. Infants usually present with some degree of right heart failure and this can be controlled with modest antifailure therapy. This failure decreases with time. Indications for surgery at this age are rare unless associated lesions are present.

Management at the Royal Children's Hospital in Melbourne has remained conservative. We have not embarked on a programme of exclusion of the right ventricle and shunting (Starnes *et al*., 1991), neither have we indicated tricuspid valve repair at this age (Knott–Craig *et al*., 2002). Indications for these neonatal and infant repairs are often dictated by the grade of severity derived from the 2D echocardiography examination in four-chamber view (Celermajer *et al*., 1994). At this age there is a high correlation between survival and ratio of the combined right atrial and atrialized right ventricular area to the area of the functional right ventricle and left heart. An echocardiographic ratio > 1.0 was 100% predictive of mortality in a series of neonates (Yetman *et al*., 1998).

Children and Teenagers

This group often has no symptoms and should be medically followed-up. Subtle decline of exercise capacity occurs with time. Exercise testing should be used as a follow-up tool to document this. Earlier and timely surgical indications should emerge from these.

Young Adults

Often become symptomatic when recurrent paroxysmal or chronic arrhythmias develop. Surgery is indicated but should be performed earlier to prevent these events.

GENERAL OPERATIVE CONSIDERATIONS

A midline sternotomy incision is the approach of choice.

CARDIOPULMONARY BYPASS AND MYOCARDIAL PROTECTION

Cardiopulmonary bypass is conducted between the ascending aorta and the two vena cavae, cannulated directly and away from the body of the right atrium. Clear prime is used above 9 or 10 kg, depending on the haematocrit of the patient. The bypass is conducted at 32°C at 200–250 ml/kg below 1 m^2 body surface area, or 2.5 l/m^2. The repair is conducted with cardioplegic arrest, using tepid blood cardioplegia injected every 20 minutes. The K^+ concentration varies (18–8 mmol/l). After the cross-clamp is applied, cardioplegia is delivered. The right atriotomy is performed parallel to the atrioventricular groove from the right atrial appendage to the space between the cannulation site for the IVC and the right coronary artery. The left-sided cavities are vented through the foramen ovale or the ASD. Stay stiches are applied on the endocardium of the superior edge of the right atriotomy for exposure.

Inspection of the heart during cardioplegia delivery and after its completion will allow appreciation of the size and extension of the atrialized portion of the right ventricle, the relationship with the right coronary artery and the posterior descending artery. Analysis of the valve itself aims at evaluating the area of available valvar tissue after mobilization and establishing the ratio of that tissue to the valvar orifice. This will determine the need and the amplitude of the annular reduction or the need for leaflet extension. As for a mitral valve analysis, the inspection finishes with the suspension apparatus: size, position and number of the papillary muscles, distribution and quality of the chordae. Prolapses and restriction of the free edge of the leaflets are best analysed on the preoperative transthoracic echocardiographic or trans-oesophageal studies. Together with the echocardiographic study, the analysis allows classification of the valve into one of the three functional groups and recognition of the anomalies.

Assessment of Repair

Whilst the heart is arrested, the repair is tested by injecting saline into the ventricular cavity through the valve at low pressure. The repair is judged satisfactory whenever both apposition and mobility of the leaflets are adequate. There should be a high zone of apposition between the free edge of the anterior leaflet and the septum (or the septal leaflet if there is one). This zone of apposition should deploy spontaneously or with minimal pressure from the testing syringe. Most often, only a large monocusp of anterior leaflet can be reconstructed. The quality of the repair is confirmed with trans-oesophageal echocardiography. A residual regurgitation greater than mild should mandate a redo of the repair, provided that the mechanism of the residual regurgitation is understood.

Indications

It is necessary to dissociate the feasibility of the tricuspid valve repair from the surgical risk and functional result. With good understanding of the surgical anatomy and when sufficient valvar tissue is available, the tricuspid valve repair can now almost always be achieved. The

surgical risk and functional result depend on the size and function of the functional right ventricle after the repair and its interaction with the left ventricle. For example, a very large functional right ventricle with poor systolic function associated with a competent tricuspid valve repair will impinge on the diastolic function of the left ventricle and generate high end-diastolic pressures. In return, this will generate higher pulmonary artery pressure. A negative vicious circle is therefore initiated.

Classical surgical treatment is indicated in the presence of severe cyanosis, congestive heart failure or severe arrhythmia. We recommend an earlier indication for surgery when severe tricuspid regurgitation is associated with restriction of exercise ability, even limited, depending on the feasibility of the repair. Re-entry supraventricular tachycardia caused by an accessory pathway can be successfully treated by electrophysiological mapping and ablation before surgery. Alternatively, they can be ablated surgically. Paroxysmic flutter or atrial fibrillation is treated with the right atrial Maze procedure (**Figure 43.4**) and resection of the right atrial wall.

Patients with a large functional right ventricle with good systolic function are the best candidates for repair. Patients with a small functional right ventricle or a functional right ventricle with poor systolic function may need to have an associated bidirectional cavopulmonary connection. When some degree of obstruction is present, it is imperative to relieve this. In patients with severe hypoplasia of the functional right ventricle or Carpentier type D a total cavopulmonary connection may be indicated.

SURGICAL TECHNIQUE

The atrial septal defect is directly closed. Resection of the redundant atrial wall is performed when the atrium is grossly dilated, more so when anti-arrhythmic surgery is associated.

Lillehei–Hardy–Hunter Operation

This operation, modified by Danielson, used to include a transverse plication of the atrialized portion of the right ventricle, parallel to the anterior tricuspid annulus, and a posterior tricuspid annuloplasty (Hunter and Lillehei,1958; Danielson, 1996) (**Figure 43.5**). The proportion of patients requiring tricuspid valve replacement with a bioprosthesis is 35%. The procedure, as described by Danielson, has itself been modified by several authors who do not include a plication (Hetzer *et al*., 1998).

Carpentier Technique (Carpentier et al. 1988)

The procedure consists of the following steps:

1. Temporary detachment of the anterior leaflet and the adjacent part of the posterior leaflet to achieve extensive mobilization of the leaflet tissue (**Figure 43.6a,b**).
2. Longitudinal plication of the atrialized chamber and the adjacent right atrium, to reconstruct the right

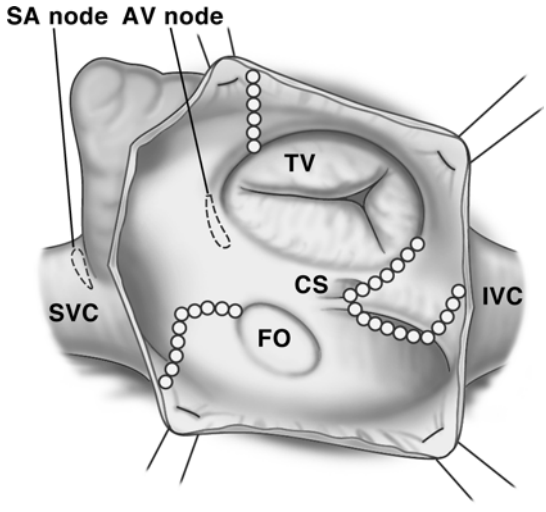

Figure 43.4

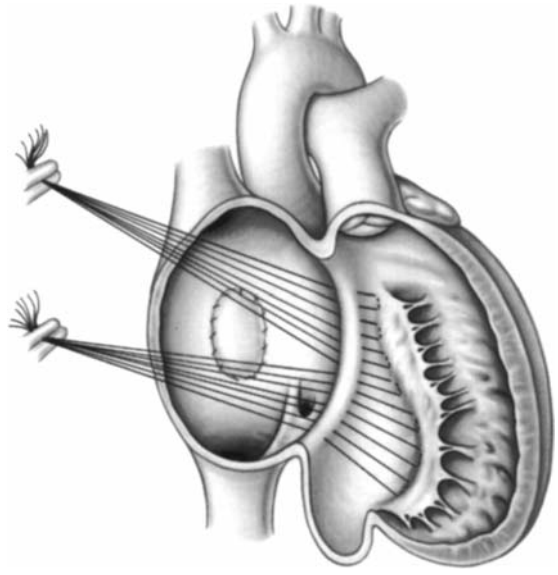

Figure 43.5

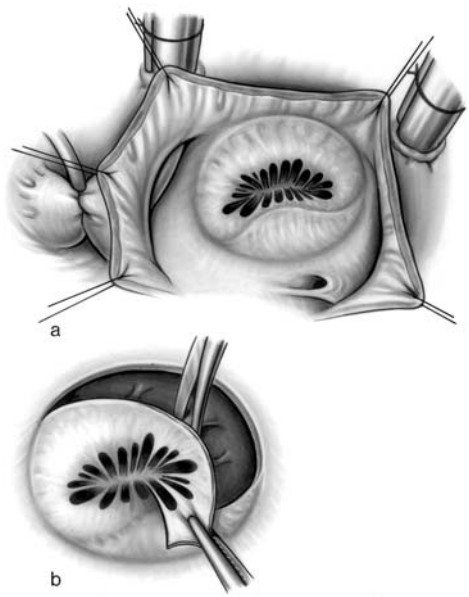

Figure 43.6

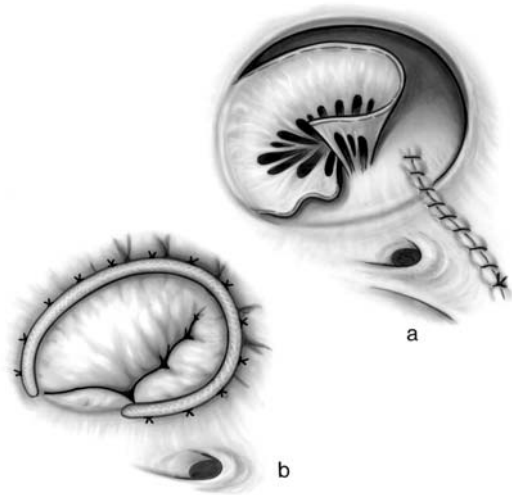

Figure 43.7

ventricle and reduce the size of the dilated right atrium (**Figure 43.7a**).

3. Repositioning of the anterior and posterior leaflets at the tricuspid annulus to cover the entire orifice area with leaflet tissue at a normal level.
4. Reinforcement of the tricuspid annulus with a prosthetic ring when a large one can be inserted (i.e. in children above age 15) (**Figure 43.7b**). Otherwise, reinforcement of the annular reconstruction is done with Gore-Tex strips or not at all (Quaegebeur *et al*., 1991).

The anterior leaflet and the adjacent portion of the posterior leaflet are detached from the annulus fibrosus and the ventricle. However, about one-third of the anterior leaflet attachment proximal to the anterior commissure is left attached to the annulus fibrosus (see **Figure 43.7a**), as it has normal mobility and attachments. Leaflet mobilization of the anterior leaflet and of the adjacent part of the posterior leaflet is achieved, first, by resection of the fibrous bands connecting the ventricular surface of these leaflets to the muscular wall of the right ventricle and, second, by fenestration of the interchordal spaces. Longitudinal plication of the atrialized chamber is achieved with interrupted 3-0 sutures passed at the edges of the atrialized portion to approximate them. A plication suture is also passed through the annulus to reduce its size. Size 4-0 sutures are then used to plicate the right atrium behind the coronary sinus to avoid the conduction system (see **Figure 43.7a**). At the Royal Children's Hospital in Melbourne, we use transfixing stiches mattressed over Teflon felt to perform this plication, carefully avoiding the right coronary artery branches. This allows prevention of secondary rupture of the plication.

Ventricular plication is essential to reduce the volume of the right ventricle and minimize the paradoxical motion of that area after it has been replaced on the ventricular side of the tricuspid valve. The right atrium is reduced by radical resection of redundant tissue.

The anterior leaflet and adjacent portion of the posterior leaflet are then reattached to the annulus with a gathering motion to cover the distance to the posteroseptal commissure. If tension on the anterior leaflet is excessive, then either the leaflet tissue has to be enlarged, using patch of autologous pericardium (**Figure 43.8**), or the suspension apparatus of the anterior leaflet has to be repositioned, using one of multiple techniques available. We have used repositioning of the papillary muscle higher into the septum, closer to the septal leaflet, with transection and reimplantation or simple fixation with a mattress suture. Artificial chordae are a simple and safe alternative (see Chapter 42 for technique).

Leaflet coaptation is tested by injecting saline under pressure into the ventricular cavity using a bulb syringe. If leaflet coaptation is adequate, a prosthetic ring is selected whose size should correspond to the size of the already plicated tricuspid annulus and to the area of the

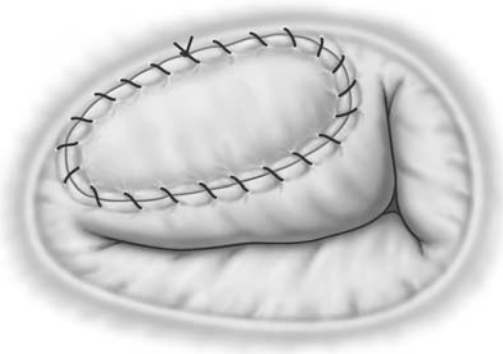

Figure 43.8

anterior leaflet. If coaptation is poor, a prosthetic ring one size smaller is inserted. In patients over 15 years of age, 36 mm and, more rarely, 34 mm rings are generally used. In younger patients with a smaller annulus, the ring should be avoided.

Association of a Cavopulmonary Connection (Chauvaud et al., 2003)

The rationale behind the association of a cavopulmonary connection to an Ebstein's valve repair is to unload the right ventricle. Patients who benefit most from this adjunction are those with a large functional right ventricle of poor systolic function. A cavopulmonary connection allows a direct preloading of the left ventricle and avoids the negative effect of a distended right ventricle on the compliance and diastolic function of the left ventricle.

On the other hand, a small functional right ventricle with preserved systolic function would not benefit from association with a bidirectional cavopulmonary shunt (BCPS) and should be managed with temporary atrial pacing.

The need for a BCPS should be anticipated with the morphological evaluation of the functional right ventricle preoperatively and the assessment of its systolic function.

Eventually, the decision is made after weaning from the cardiopulmonary bypass. High right atrial pressure, low mixed-venous saturations and high left atrial pressure are good indicators for a BCPS. Chauvaud *et al*. (2003) have associated a BCPS in 34% of their patients.

Valve Replacement

Rarely a valve replacement may be necessary. The use of a bioprosthesis (**Figure 43.9**) is generally recommended. A valve replacement is indicated when valve repair is

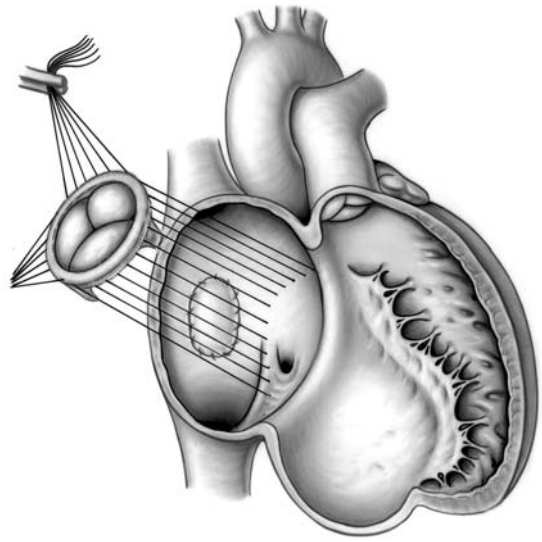

Figure 43.9

impossible or has failed, but only when the systolic function and the size of the functional right ventricle are adequate. Otherwise, a partial or a total cavopulmonary connection is a more sound indication.

To prevent damage to the penetrating bundle and a iatrogenic atrio-ventricular block, the valve is sutured to the right of the coronary sinus orifice, which is left in a subvalvar position (Barnard and Schrire, 1963) (see **Figure 43.9**).

RESULTS

Two series are reported here. The first is the series from Chauvaud and Carpentier in Paris (Chauvaud *et al*., 2003). This series illustrates the original technique of longitudinal plication and repositioning of the anterior leaflet to the native tricuspid annulus. The technique has been significantly modified by the adjunction of the bidirectional cavopulmonary connection in a large proportion of patients. The second is our series at the Royal Children's Hospital in Melbourne.

From 1980 to July 2002, 191 patients (mean age 24.4 ± 15 years, range 1–65 years) were operated on. Anterior leaflet function was assessed on preoperative echocardiography and surgical examination. Conservative surgery was possible in 187 patients and included mobilization of the anterior leaflet, longitudinal plication of the right ventricle and prosthetic annuloplasty in adults. Bidirectional cavopulmonary shunt was associated in 60 patients. Four patients had their valve replaced. There were 18 hospital deaths—this relates to

187 of patients who underwent conservative surgery :9% (95%CL: 6–15%). Mean follow-up was 6.4 years (0–22 years) with an actuarial survival rate of 86 ± 2.5% at 10 years (38 patients at risk). Tricuspid valve insufficiency was trivial or mild in 80% of the cases. Freedom from reoperation in hospital survivors was 89 ± 3.5% at 10 years (36 patients at risk). A successful second repair was obtained in 10 patients. Electron beam CT (20 patients) demonstrated improvement of the left ventricle ejection fraction from 56% to 66% ($p < 0.05\%$). Supraventricular tachycardia and pre-excitation syndromes were reduced from 23% to 5%. The long-term results suggest sustained improvement of the functional class and the exercise tolerance.

This series demonstrates that the adjunction of cavopulmonary connection improves the preload on the left ventricle in patients who would otherwise have a difficult postoperative course. This is shown by the increased ejection fraction of the left ventricle. No mapping and manoeuvres are done to localize or treat the re-entry tachycardia when present, but the surgery itself reduces the rate from 25% to 5%. As most re-entry pathways are anterior to the sinus node along the posterior annulus of the tricuspid valve, the mobilization and reattachment of the anterior leaflet along the tricuspid annulus interrupts them. A mapping prior to surgery should diagnose the posterior pathway and allow successful surgical ablation (Khositseth *et al*., 2004).

At the Royal Children's Hospital, Melbourne, the approach to Ebstein's anomaly repair has been guided by the technique demonstrated by Carpentier and Chauvaud. We have younger patients who belong to two main age groups: infants and toddlers, very symptomatic with cardiac failure; and teenagers with limited exercise tolerance but who claim to be asymptomatic. We operate on patients with few or no symptoms when tricuspid regurgitation is severe and exercise tolerance is measurably diminished. We expect to improve exercise ability and protect the functional right ventricle from further degradation of systolic function.

During 1996–2005 we have operated on 15 patients, of whom four were infants or toddlers. All four were severely symptomatic and have been repaired without BCPS. One patient with complex double-outlet right ventricle required secondary BCPS for degradation of the haemodynamics, and died despite her reoperation. The 10 teenagers have had good functional results and are in functional class 1. Their exercise ability has improved, even for the three patients with moderate residual tricuspid valve regurgitation. One patient required reoperating for endocarditis.

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44

Aortopulmonary Window

J. Stark and V. T. Tsang

Aortopulmonary (AP) window is a rare malformation with serious haemodynamic consequences. It is caused by incomplete formation of the septum in the developing truncus arteriosus. This results in an abnormal communication between the aorta and the pulmonary artery distal to their semilunar valves. Elliotson described (1830) a patient with AP window. The first successful operative correction was achieved in 1948 by Gross (1952). AP window was classified by Mori *et al*. (1978) into three types: proximal, distal and total defects. Alternative classification was proposed by Richardson *et al*. (1979): type I is proximal defect, type II is more distal defect close to the right pulmonary artery, and type III is actually anomalous origin of either pulmonary artery from the ascending aorta.

DIAGNOSIS

Symptoms and signs of congestive heart failure are present. Pulses are bounding, and the pulse pressure is wide. Because of pulmonary hypertension, the murmur is rarely continuous. The pulmonary second sound is usually split, and the pulmonary component is loud. The heart is enlarged clinically and on chest X-ray and the lung fields are plethoric. Pulmonary oedema may be present and may be the cause of cyanosis. The electrocardiogram shows biventricular hypertrophy and right axis deviation in the majority of patients. The differential diagnosis of AP window is persistent ductus arteriosus (PDA), persistent truncus arteriosus and ventricular septal defect (VSD), with or without aortic incompetence. The diagnosis can be established by two-dimensional echocardiography, demonstrating echo-free communication with the adjacent ascending aorta. Confirmation of two separate arterial valves is important; it distinguishes this condition from truncus arteriosus. Colour-flow mapping may confirm flow from the aorta to the pulmonary trunk via the window. Magnetic resonance imaging (MRI) is also useful, but cardiac catheterization may be required for assessing pulmonary arteriolar resistance in older patients; occasionally, it may be helpful for identifying other intracardiac anomalies.

Associated cardiac anomalies are common. Anomalous origin of the right coronary artery was described by Burroughs *et al*. (1962); anomalous origin of the left coronary artery was described by Doty *et al*. (1981). AP window can be associated with PDA (Deverall *et al*., 1969) and should be confirmed or ruled out by preoperative investigation. Association with ventricular septal defect (Faulkner *et al*., 1974), aortic atresia (Rosenquist *et al*., 1974) and tetralogy of Fallot (Castaneda and Kirklin, 1977) has also been reported. The association of interrupted aortic arch with AP window is not an infrequent finding (Braunlin *et al*., 1982).

INDICATIONS

Indications for surgery follow principles similar to those outlined for patients with VSD (Chapter 26). Infants in severe heart failure are operated upon as soon as the diagnosis is established. Those in whom heart failure is controlled medically should be operated upon early, because the progression of pulmonary vascular disease is difficult to monitor.

SURGICAL TECHNIQUE

AP window has been treated by ligation, by clamping and division, and by division and suture during inflow occlusion or cardiopulmonary bypass. There are certain hazards in using these techniques. Dissection of the high-pressure, sometimes thin-walled communication may be difficult. The lower margin of the defect is often close to the origin of the left coronary artery, or the coronary artery may arise from the AP window. For these reasons, we prefer and recommend the trans-aortic approach with the use of cardiopulmonary bypass (Wright *et al*., 1968). In a neonate or a sick infant, deep hypothermia with circulatory arrest could be considered.

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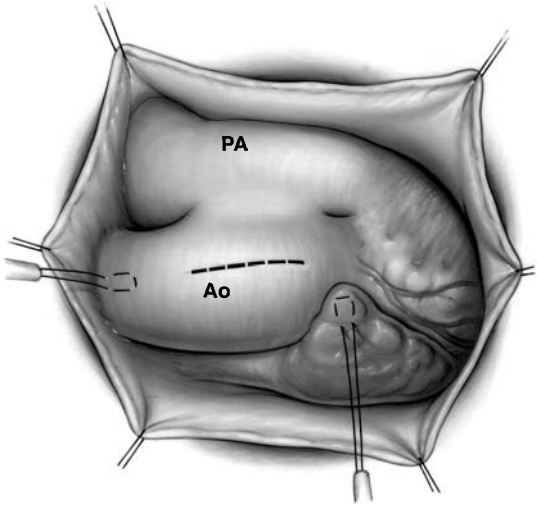

Figure 44.1

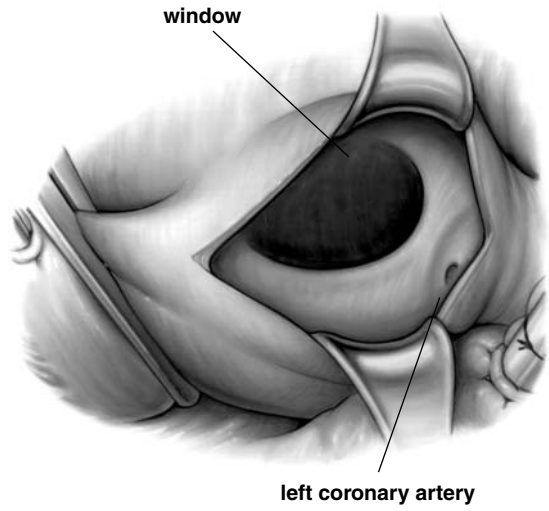

Figure 44.2

The heart is approached through a midline sternotomy. Findings are usually typical. The pulmonary artery is large, and there is a large communication between the aorta and the pulmonary artery just above the valves (**Figure 44.1**). The aorta should be cannulated well above the window, so that a clamp can be applied without difficulty and without impairing visualization of the defect. In infants, a single venous cannula may be used. Venting of the left heart is desirable.

The left and right pulmonary arteries are dissected and encircled with a Silastic sling. In infants, the superior and inferior venae cavae are cannulated separately and the left heart is vented, either through the right superior pulmonary vein or through the patent foramen ovale, after opening the right atrium. Bypass is started and both pulmonary arteries are snared. Ventilation is stopped, and the aorta is cross-clamped well above the window. Cold blood cardioplegic solution is infused to the root of the aorta. It is repeated at 20-minute intervals if required.

A vertical incision is made on the aortic side of the window (**Figure 44.1**) and the defect is visualized (**Figure 44.2**). It is important to identify the origins of the coronary and pulmonary arteries. The defect is closed with a prosthetic patch (Gore-Tex) and 5-0 or 6-0 continuous polypropylene suture (**Figure 44.3**). This approach provides good exposure to ensure that the patch does not interfere with the aortic valve, the coronary artery ostia and the origin of the right pulmonary artery. The patient is then rewarmed and the aortotomy closed (**Figure 44.4**).

Alternatively, a sandwich technique (Johansson *et al*., 1978) can be used. An incision is made through the

Figure 44.3

anterior part of the window (**Figure 44.5**). Before the incision is extended toward the proximal end of the window, the orifices of the coronary arteries are identified. A Gore-Tex patch is tailored and sutured to the posterior rim of the defect with a running polypropylene stitch (**Figure 44.6a**). When the suture reaches the edges of the incision, a continuous mattress suture incorporates the wall of the pulmonary artery, the patch and the aortic wall (**Figure 44.6b**). For better haemostasis, an over-and-over stitch may be added (**Figure 44.6c**).

While the patch is being sutured in place, rewarming is started. The atrial septal defect/patent foramen ovale is then closed through a small right atriotomy. When

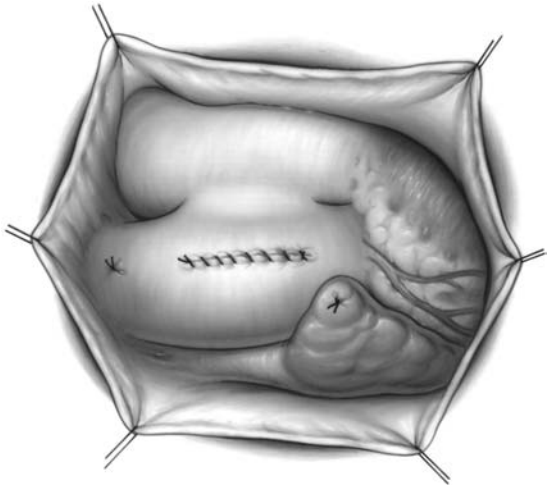

Figure 44.4

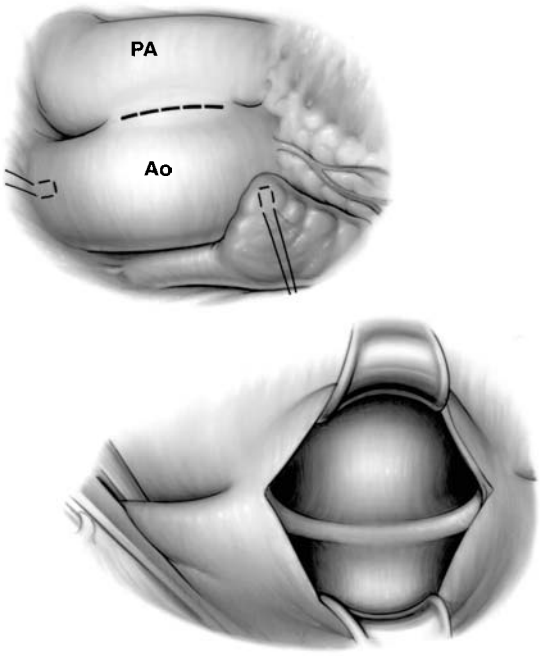

Figure 44.5

the aorta is closed, an aortic needle vent is attached to suction. The usual de-airing procedures are repeated and the aortic clamp is removed. Ventilation is restarted. When the patient is completely rewarmed, perfusion is terminated in the usual way. Postoperative care follows the principles outlined for other patients with large

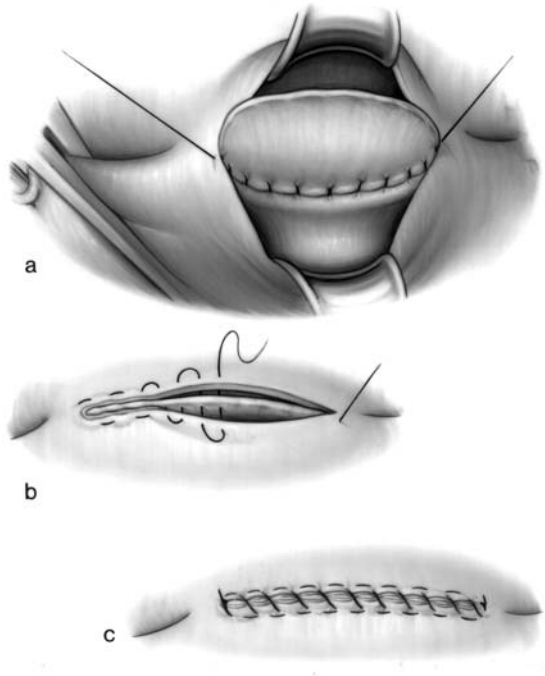

Figure 44.6

left-to-right shunts. Appropriate measures to prevent pulmonary hypertensive crises in early postoperative period, including sedation, paralysis, hyperventilation and attention to acid–base balance, are used.

AP Window Associated with Interrupted Aortic Arch

AP window associated with an interrupted aortic arch (IAA) would need a special approach. The repair of IAA would require DHCA or regional low-flow cerebral perfusion, according to the surgeon's preference. The AP window needs to be patched. Extensive mobilization of the distal pulmonary arteries should be undertaken. Patching of the aortotomy would minimize the risk of supravalvar aortic stenosis in this setting.

RESULTS

Since the first report from our hospital (Deverall *et al*., 1969), the authors and associates operated on 24 children with AP window during 1970–1990. Four had associated PDA; in three, the PDA was ligated in a separate operation. Two patients had tetralogy of Fallot, one had discrete subaortic stenosis, one had a large VSD and one had interrupted aortic arch. Of the six patients who died, two had tetralogy of Fallot, one had a large VSD and one had interrupted aortic arch; one 2 kg premature infant

suffered fibrillation before cardiopulmonary bypass was instituted and died on the operating table. The sixth death occurred early in our series in a patient who had a diagnosis of PDA; an AP window was found (the correct initial diagnosis was missed) and an attempt was made to divide it through the left thoracotomy with a partial occlusion. The aortic clamp slipped, and bleeding could not be controlled. At follow-up in 1993, all survivors were well.

We reviewed our current data for 2000–2004. Repair was undertaken in six patients (one isolated AP window and five complex: two with VSD, two with tetralogy of Fallot, and one with TGA). There was one hospital death, of the patient with TGA.

There were no deaths among 12 patients with simple AP window in a series of McElhinney and colleagues during 1961–2001. Of 12 patients with complex associated anomalies, four died early and there were four additional late deaths (McElhinney *et al*., 1998). Bagtharia *et al*. (2004) reported their results for 1969–1999. There was no deaths in the group of 14 isolated AP window, but three deaths among 17 patients with repair of other defects. Kaplan–Meier estimates of survival were 81% up to 11.5 years, and 69% up to 21 years. The outcome for an isolated lesion is excellent. However, mortality still occurs with complex associated lesions, particularly interrupted aortic arch. Successful trans-catheter umbrella closure of a small AP window was reported (Tulloh and Rigby, 1997). Although the technique is feasible in infancy, its application would probably be suitable only in a few cases.

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45

Congenital Anomalies of Sinuses of Valsalva and Aorticoventricular Tunnel

V. T. Tsang and J. Stark

ANEURYSM AND FISTULA OF SINUS OF VALSALVA

Abnormalities of the sinus of Valsalva may be congenital or acquired. Congenital aneurysms of the sinus of Valsalva are confined to one aortic sinus, usually the right, and consist of the localized diverticular outpouching of the aortic sinus. The basic lesion is thinning of the wall of the aortic sinus just above the annulus. This may be the result of deficiency of elastic and muscular tissue in the aortic root (Edwards and Burchell, 1957). The aneurysm develops in this weakened area; eventually it may rupture into one of the lowered-pressure cardiac chambers, usually the right ventricle, less commonly the right atrium, pulmonary artery, and very rarely the left atrium and ventricle (Chu *et al*., 1990).

The aneurysms may be associated with other congenital anomalies, the most common being a ventricular septal defect (VSD). Congenital aneurysms may be asymptomatic, or cause symptoms by rupture and fistula formation, by distortion of the aortic valve that may become incompetent, or by compression that may cause the right ventricular outflow tract obstruction. Very rarely it may be intrapericardial rupture.

A fistula may also result from rupture of an acquired aneurysm of the aortic sinus, from penetrating trauma, or from congenital tracts in otherwise normal sinuses. Rupture of a congenital aneurysm nearly always produces a fistula between the aorta and one cardiac chamber, whereas rupture of an acquired aneurysm is usually extracardiac, into the pericardial, mediastinal or pleural space.

DIAGNOSIS

Aneurysms are diagnosed early in life during investigation of other congenital cardiac defects, such as VSD, aortic regurgitation, or if bacterial endocarditis develops, or if symptoms are produced by compression. Most commonly, aneurysms enlarge until they rupture into a cardiac chamber during the fourth decade of life. Intracardiac rupture of the aneurysm usually causes abrupt onset of congestive heart failure, with a continuous murmur best heard at auscultation over the sternum or the left sternal border. The diagnosis is established by two-dimensional (2D) echocardiography with colour-Doppler (Terdjman *et al*., 1984). MRI or multislice CT with contrast angiography demonstrate the close proximity of the proximal right coronary artery. Before rupture, the aneurysm is visualized as a diverticulum of the sinus. After rupture, aortography shows a jet of contrast material from the aneurysm entering one of the cardiac chambers. The majority of the fistulae arise from the right aortic sinus; fistulae arising from the non-coronary and left coronary sinuses are less common. Communication between the aortic root and the right ventricle is most common in patients with right aortic sinus aneurysm. Aneurysms arising from the non-coronary sinus more often rupture into the right atrium. Once the diagnosis has been established, early surgical intervention is indicated for prevention of cardiac complications, such as aortic regurgitation, that may ensue, and the treatment of associated cardiac lesions (Qiang *et al*., 1994).

SURGICAL TREATMENT

Operations are performed on cardiopulmonary bypass and mild hypothermia. Cold blood cardioplegic solution is infused into the ascending aorta or, for cases of fistula, directly into the coronary arteries. The aorta is opened with an oblique incision and the aortic valve closely inspected. If a VSD is also present, it may be examined from this approach.

The principle of surgery is simple. The defect in the aortic wall is closed, the aneurysm is resected,

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and the fistula tract is obliterated. All this is usually accomplished by a combined approach of opening the chamber into which the aneurysm protruded or ruptured (Spencer *et al*., 1960). The windsock-like deformity produced by the aneurysm, fistula or both is resected and obliterated via the cardiac chamber involved, and the defect in the sinus is closed through the aorta. This approach enables accurate placement of fine interrupted sutures, which the authors prefer, without injury to a coronary artery or distorting an aortic valve cusp. Reconstruction or replacement of the aortic valve is sometimes necessary because of valvar or annular deformity (Azakie *et al*., 2000).

Figure 45.1 depicts an aneurysm of the right coronary sinus with a fistula in the right ventricle and the sites of the aortic, pulmonary artery and right ventricular incisions. High aortic and bicaval cannulation is recommended. An oblique aortotomy incision toward the non-coronary cusp is made, so as to expose the origin of the aneurysm or fistula. The fistula can be closed by direct suture, which may be buttressed between pledgets (**Figure 45.2a,b**), or is preferably patched (**Figure 45.2c**). The technique depends on the size of the orifice, the firmness of the edges and the degree of distortion of the aortic valve. The pericardial or Gore-Tex patch is inserted with fine interrupted or continuous sutures. When the fistula terminates in the right ventricular infundibulum, the aneurysm or fistula may be resected through a transverse pulmonary arteriotomy (**Figure 45.3a**). The anterior pulmonary cusp is retracted, the aneurysmal sac is excised (**Figure 45.3b**) and the fistula is closed by continuous suture (**Figure 45.3c**). Sometimes a transverse right ventriculotomy incision provides better exposure of the fistula, as shown in (**Figure 45.4**).

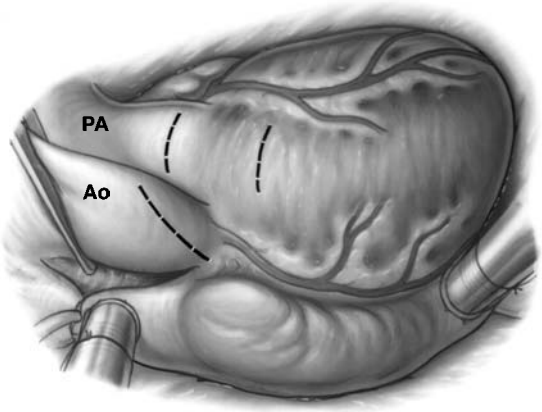

Figure 45.1

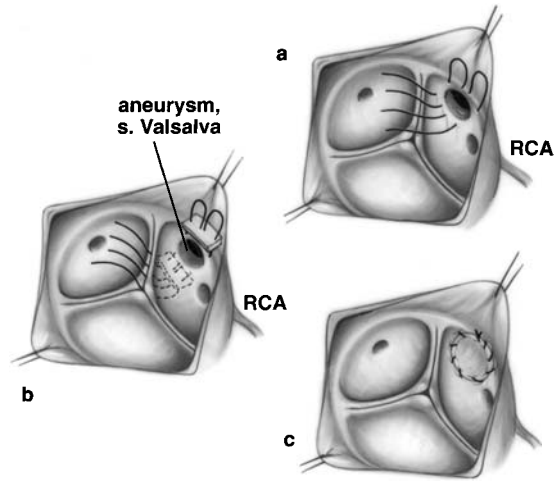

Figure 45.2

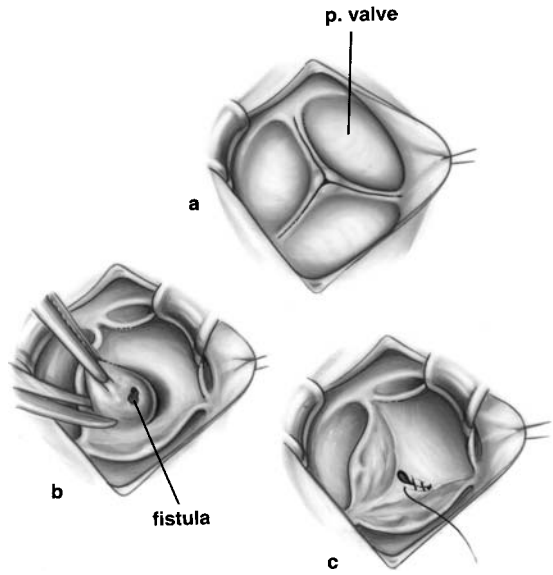

Figure 45.3

Figure 45.5 depicts repair of an aneurysm with protrusion and formation of a fistula into the right atrium. The aneurysm or fistula is exposed through the right atriotomy (**Figure 45.5a**). The aneurysmal sac is opened (**Figure 45.5b**) and the fistula is closed primarily (**Figure 45.5c**) and this closure is further reinforced by approximating the remnants of the aneurysmal sac (**Figure 45.5d**).

Associated anomalies, such as VSD, are also repaired. When present, a VSD is nearly always associated with a

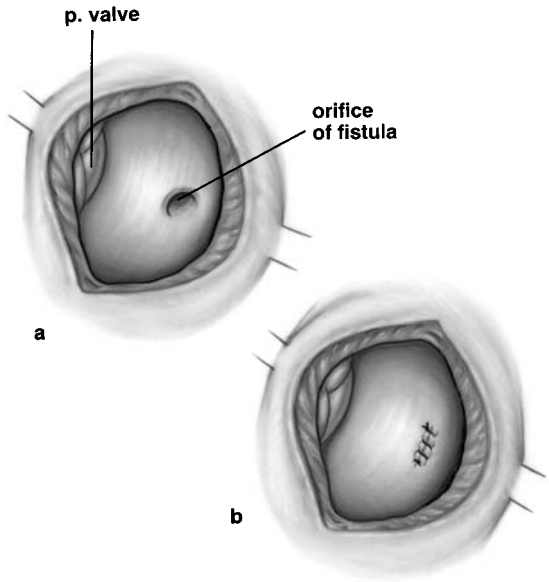

Figure 45.4

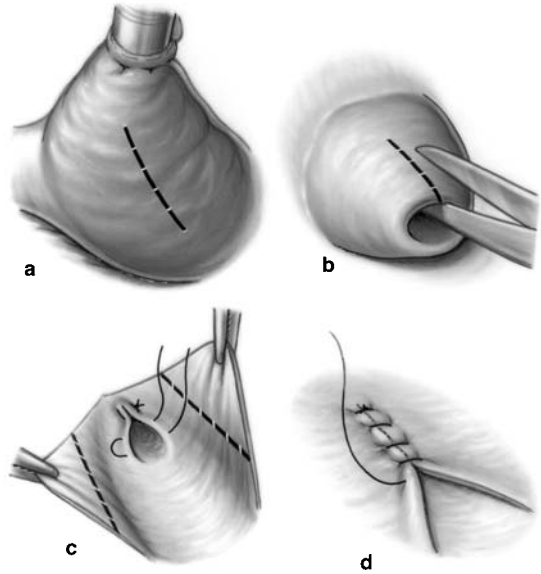

Figure 45.5

right sinus fistula. These defects are usually infundibular and subarterial (Sakakibara and Konno, 1968). They can be closed through either the pulmonary artery or the right ventricle. The long-term outlook of the repair depends on the presence or absence of aortic valve prolapse (Azakie *et al*., 2000). The natural history of aortic valve regurgitation in this setting is progressive. Early surgery is therefore indicated to minimize the risk of valve replacement. The techniques used for the aortic valve repair include commissural resuspension, partial commissural plication, triangular resection of redundant leaflet tissue or a combination of these techniques.

Results

Okada *et al*. (1977) reported eight deaths (4.1%) in a series of 195 patients. There was no mortality among 29 patients reported by Kirklin and Barratt-Boyes (1986). Azaki *et al*. (2000) employed two techniques for the repair; simple closure in 10 and patch closure in 24. Concomitant aortic valve surgery was needed in 11 of 24 patients with aortic valve regurgitation. Five early fistula recurrences occurred with primary closure rather than with a patch repair. Late aortic valve surgery was performed in six patients, with $83 \pm 9\%$ freedom from reoperation at 10 years. Au *et al*. (1998) reported long-term survival after repair in 53 patients, with no early deaths. Actuarial survival was 84% at 15 years. Reoperation, aneurysm draining into the LV, aortic prosthesis dehiscence and bacterial endocarditis were important complications influencing late survival.

AORTICO–LEFT VENTRICULAR TUNNEL

Aortico–left ventricular tunnel is a very rare congenital anomaly. It was described by Levy *et al*. in 1963. The tunnel is a congenital communication between the ascending aorta and the left ventricle; rarely it is a communication between the aorta and the right ventricle (Rosengart *et al*., 1993; Westaby and Archer, 1992). The tunnel originates in the aorta, just above the sinus of Valsalva, and passes down towards the left or right ventricle. It can be seen externally as a bulge (**Figure 45.6**). Tuna and Edwards (1988) described deficient support of the aortic root in relation to the right aortic sinus in this condition, which may result in aortic regurgitation. Severe stenosis of the aortic valve, necessitating aortic root replacement, was also described in a neonatal aorta-to-left ventricle tunnel (Weldner *et al*., 1996).

Diagnosis

Symptoms depend on the size of the communication. Severe congestive heart failure may be present in neonates. In our series of 13 patients, five presented as neonates; two presented in the first day of life (Horvath *et al*., 1991). Findings are very similar to those seen in aortic incompetence: systolic and diastolic murmurs, wide pulse pressure and bounding pulses. The X-ray

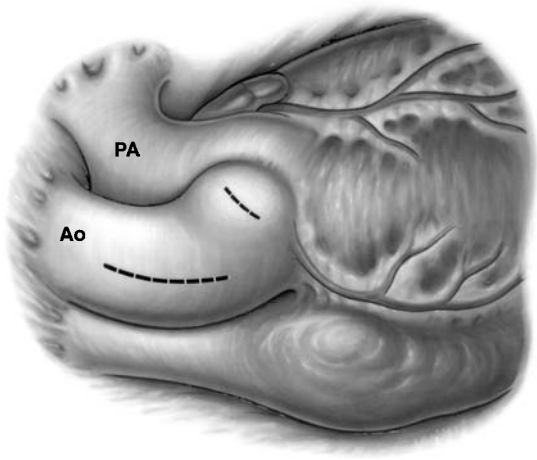

Figure 45.6

shows cardiomegaly with a broad upper mediastinal shadow. Marked left ventricular hypertrophy is visible on the electrocardiogram. In 12 of our 13 patients, an ischaemic pattern over the left ventricle was present. Diagnosis is confirmed by 2D echocardiography (Humes *et al*., 1986), and colour-flow mapping can demonstrate the massive regurgitation, bypassing the aortic valve. Angiocardiography (Horvath *et al*., 1991) is now rarely indicated unless the non-invasive findings are inconclusive or the lesion seems to be suitable for trans-catheter closure. Associated lesions include persistent ductus arteriosus, atrial septal defect, and VSD. Aortic valve anomalies are quite common.

INDICATIONS

Urgent or even emergency operation is indicated in sick neonates. Older children can be operated electively once the diagnosis is made. We do not recommend delaying surgery because the turbulence caused by the tunnel may lead to aortic valve incompetence.

Surgical Technique

Several surgical techniques have been reported in the literature (Hovaguimian *et al*., 1988). Our technique of choice is the closure of both the aortic and the ventricular openings using two separate patches. The operation is performed on cardiopulmonary bypass with hypothermia and cardioplegia. To minimize LV distension on bypass, early aortic cross-clamping and venting of the LV via the right pulmonary vein is required. We prefer the direct delivery of blood cardioplegia into the coronary arteries, using a DLP coronary cannula. In neonates, deep hypothermia with circulatory arrest is an alternative. The tunnel can be seen as a bulge between the aorta and the right ventricular outflow tract as soon as the pericardium is opened (**Figure 45.6**). The aorta is opened with an oblique incision, and the aortic valve and the aortic opening of the tunnel are inspected. The opening of the aortic end of the tunnel is then closed with a prosthetic patch, attached with fine monofilament suture (**Figure 45.7a**). The bulging tunnel outside the heart is then opened, and the ventricular opening of the tunnel is identified. Another patch is used for the closure of the ventricular opening of the tunnel to provide ''support'' for the neo-aortic root (**Figure 45.7b**). The surgeon should be aware of very close proximity of both patches to the aortic valve and particularly to the right coronary cusp. The principle of the operation is schematically illustrated in **Figure 45.8**. The suture is attached to a bar of arterial tissue, not to the cusp of the aortic valve. The advantage of this two-patch technique is a complete closure of the tunnel and identification of the coronary arteries, so that injury to the coronary arteries is avoided.

Results

A mortality rate of 100% was reported in medically-treated patients, whereas the mortality rate among surgically-treated patients was 16% (Hovaguimian *et al*., 1988). In a review of the literature (Duveau *et al*., 1989),

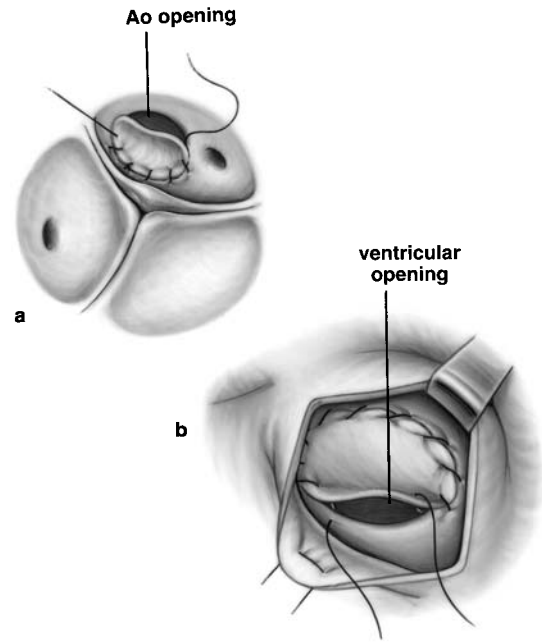

Figure 45.7

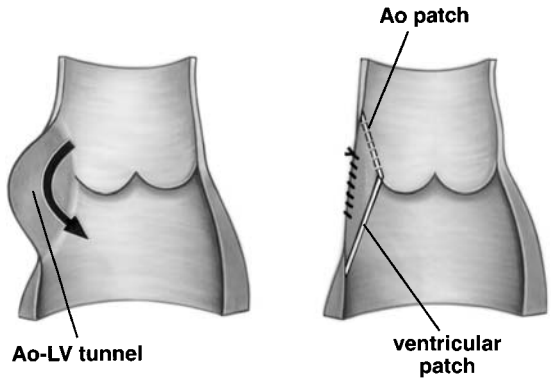

Figure 45.8

57 patients were reported; 51 underwent operation, and the mortality rate was 21.5%. A high rate of late aortic incompetence was reported after direct closure (Serino *et al*., 1986; Duveau *et al*., 1989).

There was one operative death in our series of 13 patients (Horvath *et al*., 1991). One late death was from non-cardiac causes (neuroblastoma). Aortic regurgitation developed in one patient after direct closure of only the aortic opening. The remaining patients were operated using the two-patch technique. Martins *et al*. (2004) reviewed their 35-year experience with nine patients; seven with direct suture and two with patch closures. At follow-up (median 5 years), all were asymptomatic, three with residual tunnel and mild aortic regurgitation. For this reason, we recommend the patch closure, rather than direct closure.

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46

Congenital Coronary Artery Fistula and Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

V. T. Tsang and J. Stark

CONGENITAL CORONARY ARTERY FISTULA

A congenital coronary artery fistula is a communication between the coronary artery and one of the four cardiac chambers, the coronary sinus, superior vena cava, pulmonary artery or pulmonary vein close to the heart. It was first reported by Krause in 1865, and the first successful surgical correction was performed by Biorck and Crafoord in 1947. Isolated fistulae originating from the right coronary artery are more common than those originating from the left coronary artery. A fistula may occur either at the side of the main vessel or at the termination of the vessel. In 90% of cases, the fistula opens into the right side of the heart, most commonly into the right ventricle.

Complex coronary artery fistulae, rather than an isolated lesions, can compound the management of some major cardiac lesions. Fistulae arising from hypertensive right ventricle in pulmonary atresia with intact ventricular septum, and from hypertensive left ventricle with mitral stenosis and aortic atresia in hypoplastic left heart syndrome, should be referred to the respective chapters.

The individual anatomy of these fistulae varies considerably. Often the fistula is a single vessel, frequently tortuous, with a single origin and a single termination (terminal fistula). **Figure 46.1a** depicts a terminal fistula from the right coronary artery into the right ventricle. A right coronary artery fistula may enter the posterior aspect of the right ventricle, as illustrated in **Figure 46.1b**. A fistula originating from the left coronary artery and opening into the right ventricle outflow tract is depicted in **Figure 46.1c**. In some patients, the fistula forms a plexus of tortuous vessels that may terminate in the cardiac chamber at multiple sites (**Figure 46.2**). Occasionally, the fistula may dilate due to high flow, or part of it may even become aneurysmal (**Figure 46.3**). In rare instances, a giant aneurysm involves the whole artery.

Diagnosis

The majority of patients are asymptomatic, have relatively small shunts, and do not require intervention (Liberthson *et al*., 1979; Sherwood *et al*., 1999). The physiological effects of a coronary artery fistula depend on the size of the shunt and whether it empties into the right or left side of the heart. The most common symptoms are dyspnoea and fatigue, caused by a left-to-right shunt. Though rare, congestive heart failure has been reported in infancy (Cooley and Ellis, 1962) and then after the age of 40 years due to left ventricular volume overload. A coronary steal producing myocardial ischaemia is uncommon. Subacute bacterial endocarditis occurs in 5–10% of patients with coronary artery fistula (Kirklin and Barratt-Boyes, 1986).

The diagnosis is suggested by the presence of precordial continuous murmur. In comparison with the murmur of persistent ductus arteriosus, a coronary artery fistula murmur is heard lower than in the second intercostal space. The fistula can be seen on two-dimensional echocardiography and colour-flow Doppler. Cardiac catheterization, aortogram and, in particular, selective coronary artery/fistula cineangiography provide the detailed information needed for preoperative assessment. Most of these shunts are only of moderate degree; therefore, congestive heart failure does not develop until the fourth or fifth decade of life. Among 173 reported cases, death was attributable to fistulae in 6% (Liberthson *et al*., 1979). The mortality rates were 1% in patients younger than 20 years and 14% in those presenting over the age of 20 years. In view of the low

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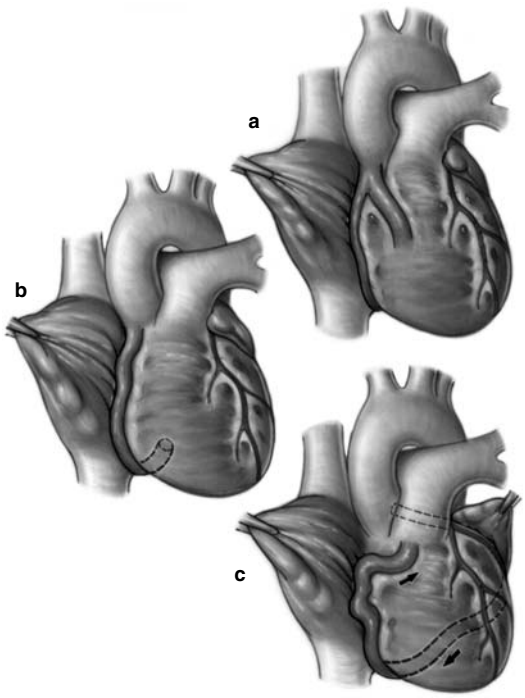

Figure 46.1

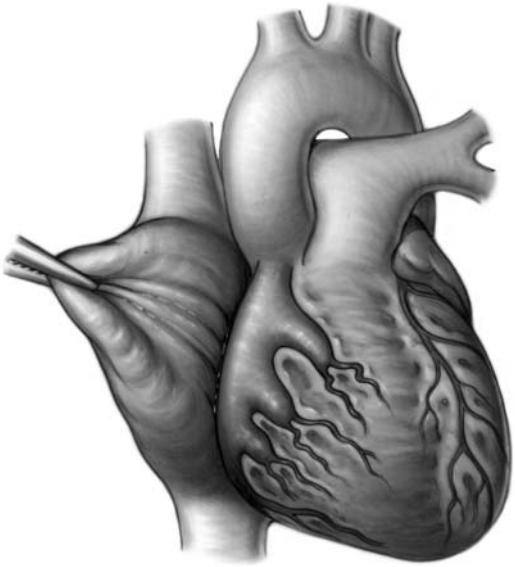

Figure 46.2

surgical risk and increasing incidence of symptoms in older patients, surgical treatment for large fistulas in childhood should be considered. Another option is interventional catheterization (Qureshi *et al*., 1996), but the complexity of the fistula and the origins of the coronary arteries should be part of the consideration.

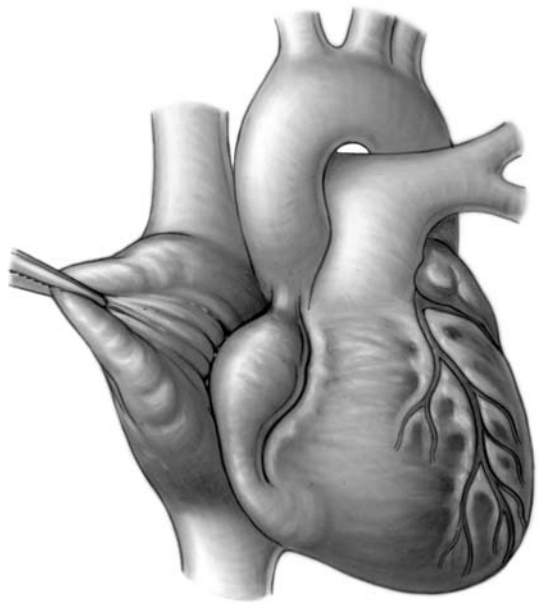

Figure 46.3

Surgical Technique

Angiographic delineation of the entry and exit points of the fistula, and its relationship with the distal coronary arteries, must be obtained preoperatively. Some fistulae may be closed during the diagnostic cardiac catheterization by placing coils. If the placement of the coil is considered difficult or if it may occlude an important coronary artery branch, operation is indicated. Some fistulae can be closed without cardiopulmonary bypass. However, cardiopulmonary bypass is required for patients with fistulae opening into the posterior aspect of the heart, such as the right ventricular inflow tract or the coronary sinus; for patients in whom the coronary artery or the fistula has to be opened; and for patients in whom fistulae are closed from within the heart chambers.

The heart is approached through a midline sternotomy. The arteries involved in forming the fistula are usually recognized as dilated tortuous vessels. The procedures used for closure of the coronary artery fistula are ligation or division, trans-arterial closure, tangential arteriography and intracardiac repair.

Ligation or Division

This is the approach of choice for terminal fistulae when a large coronary artery communicates with a cardiac chamber or the pulmonary artery. The site of communication

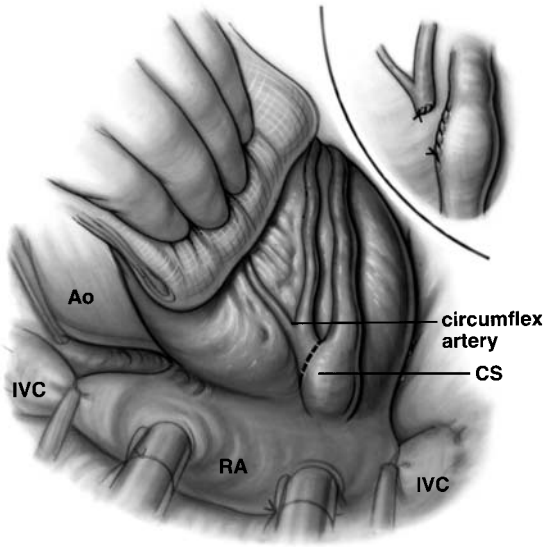

Figure 46.4

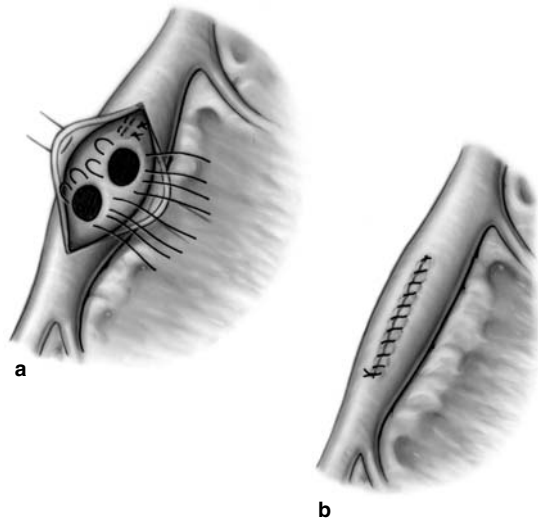

Figure 46.5

must be adequately exposed, and the fistula is temporarily occluded. Any thrill should be eliminated, and no adverse changes should be observed on the electrocardiogram. The fistula can then be ligated or divided, as illustrated for the posterior fistula on **Figure 46.4**. Because of the posterior location, the operation for the fistula depicted was performed with the patient on cardiopulmonary bypass.

Trans-arterial Closure

This approach is used for largely dilated coronary arteries and for aneurysmal dilatations of the coronary arteries and fistulae. The operation is performed on cardiopulmonary bypass. The site of fistula is marked before cardiopulmonary bypass is instituted. The aorta is cross-clamped, the fistula is digitally occluded, and cardioplegic solution is infused. The coronary artery or the aneurysm is then incised longitudinally, and the fistulae are closed by direct sewing with a fine polypropylene suture from within the artery (**Figure 46.5**). The ''aneurymal'' coronary artery is then tailored and repaired with a 7-0 continuous monofilament suture.

Tangential Arterioplasty

This technique was first described by Cooley and Ellis in 1962 for lateral fistulae and is illustrated in **Figure 46.6**. It can be used without cardiopulmonary bypass. Several horizontal mattress sutures are passed beneath the coronary artery to close the fistula, while the patency of the coronary artery is maintained.

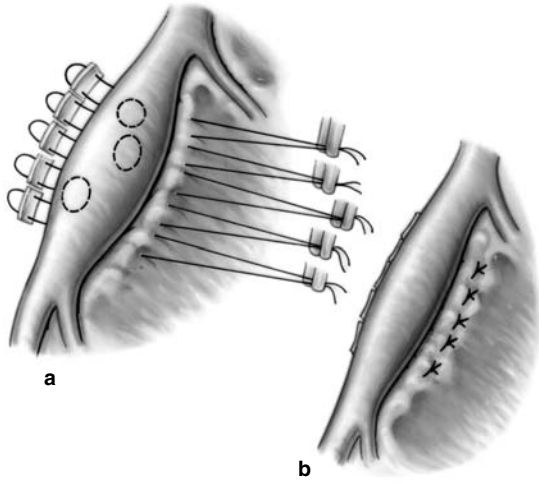

Figure 46.6

Intracardiac Repair

If a fistula cannot be safely exposed on the surface of the heart, the chamber into which it drains is opened on cardiopulmonary bypass. The fistula is closed with simple sutures, which may be reinforced with pledgeted mattress sutures (**Figure 46.7**). The adequacy of closure can be tested by infusion of cardioplegic solution into the root of the aorta. If the closure of the terminal fistula may compromise the flow of an important distal coronary artery, such as the posterior descending branch, coronary

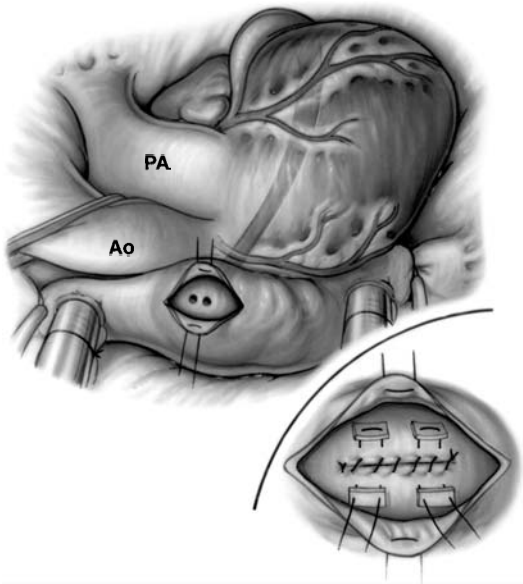

Figure 46.7

artery bypass grafting would be indicated to prevent myocardial ischaemia.

Results

Even in the early years, the rate of hospital mortality from repair of coronary artery fistulae was low. In 1975, Rittenhouse *et al*. reported an early mortality rate of 2% in a collective series of 170 cases reported in the English literature. There were no deaths among Kirklin and Barratt-Boyes's (1986) 25 operated patients. However, in the presence of giant aneurysms, the risk of death from operation and subsequent complications was higher (Kirklin and Barratt-Boyes, 1986).

Kamiya *et al*. (2002) reviewed 25 patients (2–69 years old) operated between 1973–2000. There was no mortality or perioperative complication. Postoperative angiogram showed a residual coronary artery fistular flow in one patient. The average follow-up was 10 years, and all patients were asymptomatic.

ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA)

Anomalous left coronary artery may arise from the proximal main pulmonary artery or, in rare instances, from the right pulmonary artery. The vessel involved may be the entire left main coronary or only the left anterior descending or circumflex artery. Origin of the right coronary artery or both coronary arteries from the main pulmonary artery is very rare (Goldblatt *et al*., 1984; Bregman *et al*., 1976). The anomalous left coronary artery usually arises from the left pulmonary sinus of Valsalva; it then assumes the course and distribution of a normal left coronary artery.

Physiology and Diagnosis

The pathophysiology of this condition depends on the pressure difference between the systemic and pulmonary circulations and on the presence or absence of the collateral vessels between the left and right coronary artery systems. In foetal life and in a newborn, when the pulmonary vascular resistance is elevated, the left coronary artery pressure is adequate for perfusion of the myocardium. Although the left ventricular myocardium is perfused with desaturated blood, adequate oxygenation may be provided. When the pulmonary vascular resistance falls, the oxygenation of the left ventricular myocardium depends on the adequacy of the collateral circulation. If the collateral circulation is inadequate, myocardial infarction is inevitable.

Symptoms may appear soon after birth but become more severe when pulmonary vascular resistance decreases, at approximately 2–3 months of age. Heart failure with breathlessness, poor feeding, sweating and tachycardia are typical features. Severe cardiomegaly is evident on chest X-ray, and signs of mitral valve regurgitation caused by papillary muscle ischaemia/infarction may be present. This situation is known as Bland–White–Garland syndrome (Bland *et al*., 1953).

Electrocardiograms usually show anterolateral infarction with Q-wave and ST elevation in lateral chest leads. Echocardiography shows a dilated and poorly contracting left ventricle, often with ischaemic mitral regurgitation. This must be clearly distinguished from dilated cardiomyopathy. Colour-flow Doppler echocardiography should be able to demonstrate the anomalous coronary origin and the retrograde flow from the anomalous left coronary artery into the pulmonary artery. Aortography, rarely necessary in infancy, shows only the right coronary artery arising from the aorta. With the development of adequate collateral circulation, contrast material can be followed through the right coronary artery, through the collateral vessels and into the pulmonary artery. Selective angiography provides the exact information about the origin of the artery and the extent of the collateral circulation. With large collateral vessels, a step-up in oxygen saturation may be detected in the pulmonary artery.

When severe symptoms do not occur in infancy, manifestation of the defect may be delayed to the second or third decade of life. Some adult patients may remain asymptomatic or complain only of fatigue and dyspnoea.

Indications for Operation

Infants with anomalous origin of the coronary artery are at risk of sudden death. Development of acute myocardial infarction or diffuse myocardial fibrosis can lead to heart failure. Infarction of the papillary muscle produces mitral regurgitation. For these reasons, operation is indicated as soon as the diagnosis is made. Unfortunately, a number of patients are still diagnosed too late, and they present with major myocardial injury and often severe mitral regurgitation. Heart transplantation may be the only viable option. Elective operation should be performed in older asymptomatic children and adults to establish a dual coronary arterial system.

Surgical Technique

Several surgical techniques have been described and used: ligation of the anomalous coronary artery in patients with adequate collateral circulation and aorto-coronary connection with the use of a subclavian artery, a graft or a tunnel. However, direct reimplantation of the coronary artery into the aorta has become the operation of choice, as a result of experience with the transfer of the coronary arteries in the arterial switch operation (Chapter 36). This technique has gained in popularity and the results have improved markedly. The improvement in results is also partly related to the availability of extra-corporeal circulatory support of a very compromised left ventricle post repair (Chapter 14). For completeness, some of the early palliative and bypass procedures are included here as part of the surgical considerations.

Ligation of the Left Coronary Artery

Ligation of the anomalous left coronary artery can be performed through a left thoracotomy without cardiopulmonary bypass. The mortality rate used to be high when this technique was used in patients without adequate intercoronary circulation. Ligation is indicated only in patients with a significant left-to-right shunt, in order to eliminate coronary steal and thus improve myocardial bloodflow. Kirklin and Barratt-Boyes (1986) recommended this procedure for very sick infants. However, today restoration of two coronary artery systems is preferred (Bunton *et al*., 1987).

The pericardium is opened in front of the left phrenic nerve, and the left coronary artery is identified and dissected near the origin from the pulmonary artery (**Figure 46.8**). During trial occlusion, heart action and the electrocardiogram are carefully observed; the coronary artery is then doubly ligated, the pericardium is loosely closed, and the chest is closed with a drain.

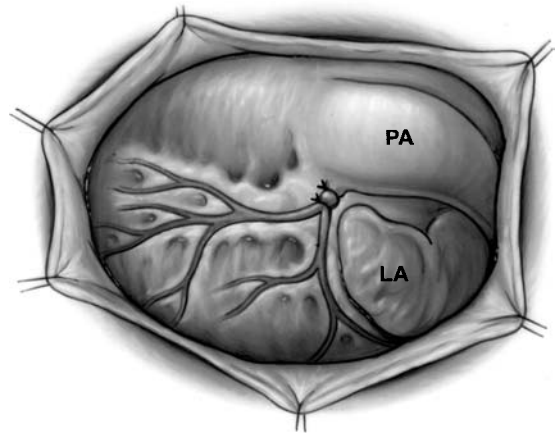

Figure 46.8

Subclavian–Coronary Artery Anastomosis

This procedure was described by Apley *et al*. (1957) and successfully performed by Meyer *et al*. (1968). It can be performed with or without cardiopulmonary bypass. The subclavian artery is mobilized and divided at the primary branches. The anastomosis can be performed end-to-side after ligation of the coronary artery at its origin (Monro *et al*., 1978) (**Figure 46.9a, b**); alternatively, the left coronary artery is excised from the pulmonary artery with a cuff of pulmonary arterial wall. This cuff is then anastomosed to the subclavian artery; the defect in the pulmonary artery is either patched or closed directly (**Figure 46.9c**). The operation, with the patient on bypass, can be performed through a midline sternotomy (Suzuki *et al*., 1978) or through a left-sided anterolateral thoracotomy (Richardson and Doty, 1979).

Undue tension and kinking at the origin of the left subclavian artery are considered the major disadvantages of this technique, when compared with the use of internal mammary artery. Although long-term patency of the subclavian to coronary artery anastomoses has been documented angiographically in some patients, this technique has been replaced by direct reimplantation (Neches *et al*., 1974; Turley *et al*., 1995) or tunnel construction, described by Takeuchi *et al*. (1979).

Aortocoronary Connections

Direct Reimplantation

The operation is performed on cardiopulmonary bypass with moderate hypothermia, aortic cross-clamping, venting of the dilated left ventricle and cold blood cardioplegia. Prior to institution of bypass, the main

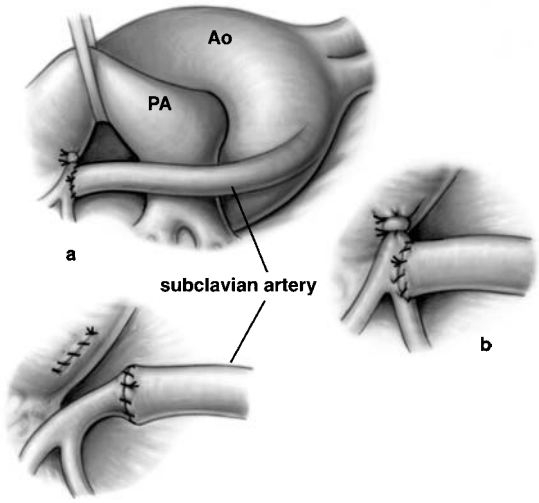

Figure 46.9

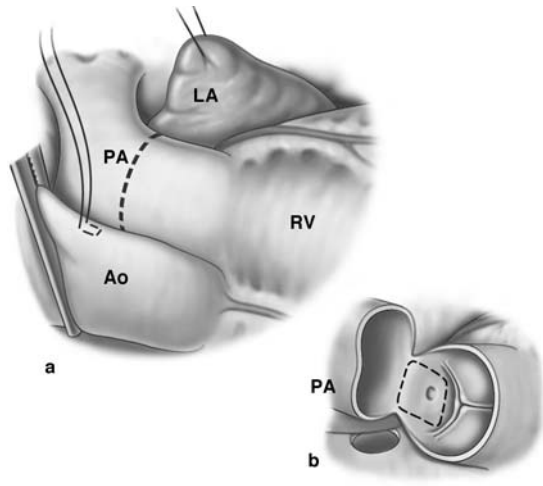

Figure 46.10

pulmonary artery surrounded by many vascular collaterals is carefully dissected out. Once on bypass, the ''coronary steal'' from the decompressed right heart into the pulmonary artery must be minimized. The branch pulmonary arteries are snared to prevent damaging run-off. The aorta is cross-clamped swiftly and concomitant cardioplegic solution can be administered into the root of the aorta and the main pulmonary artery. The authors also use the additional direct delivery of cardioplegia into the origin of the left coronary artery, immediately after the pulmonary artery is opened. The pulmonary artery is opened anteriorly, and the origin of the left coronary artery is identified; it is usually located in the left sinus of Valsalva.

If the anomalous left coronary orifice is adjacent to the aorta, a large button of pulmonary arterial wall around the coronary artery is excised, and the pulmonary artery is completely transected 3–4 mm above the commissures (**Figure 46.10**). The optimal place for reimplantation of the coronary artery to the left lateral aspect of the ascending aorta is selected and marked with a fine suture. A button of aortic wall is excised which may suffice for a tension-free direct anastomosis (**Figure 46.11**). Otherwise, a trapdoor opening is created to accommodate the slight rotation (Ando *et al*., 2002). An anastomosis between the coronary artery and the aorta is then constructed with 7–0 monofilament suture material. Earlier patching of the posterior pulmonary root is not shown in the figure for the purposes of clarity.

In cases where the left coronary orifice is too far from the aorta, the authors would harvest the posterior pulmonary arterial wall extensively to recreate

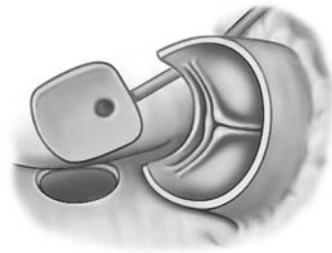

Figure 46.11

a ''tubular'' proximal left anterior descending artery (**Figure 46.12a–c**)

The pulmonary artery is divided at the level of bifurcation. The posterior commissure may have to be temporarily detached in order to provide a good length of posterior pulmonary artery flap. The superior and inferior edges are sutured together with 7-0 Prolene to form a proximal tubular extension of the left coronary artery. This should provide a tension-free transfer onto the left lateral aspect of the aorta. Barth *et al*. (2003) described a technique of isolating the anterior and posterior transverse segment of pulmonary artery in continuity with the origin of the anomalous coronary artery. The segments are folded and the edges are sutured together to form an extension tube.

Direct approximation of the posterior wall of the transected pulmonary artery is not recommended, as this may

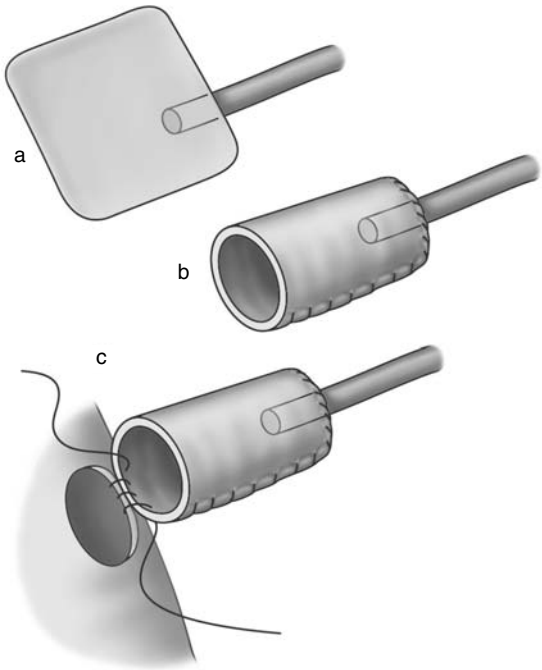

Figure 46.12

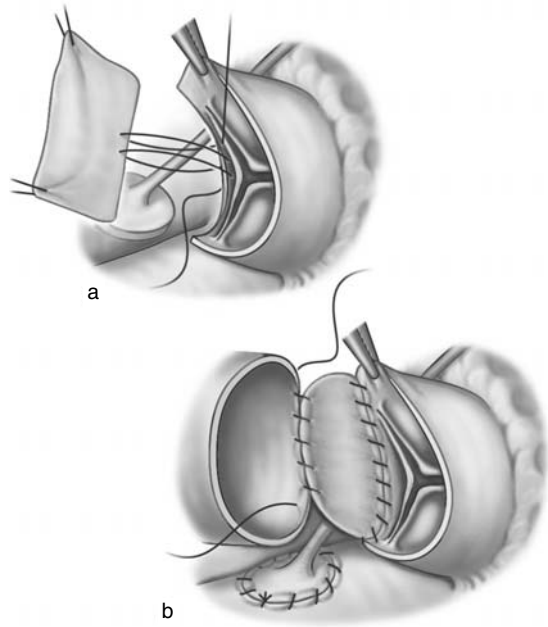

Figure 46.13

put pressure on the relocated coronary artery. We therefore prefer to interpose a patch of autologous pericardium (**Figure 46.13**). The patching to the posterior pulmonary root and, if required, the resuspension of the detached commissure should be undertaken prior to the coronary-to-aorta anastomosis. Reconstruction of the pulmonary artery can be performed easily if the bifurcation and the branched pulmonary arteries are fully mobilized and the ligamentum arteriosum divided.

Use of mechanical circulatory assistance (see Chapter 14) is briefly mentioned here as part of the surgical strategy in the successful management of the often very poor left ventricular function after repair. The technique used depends on the expertise of individual institutions. Del Nido *et al*. (1999) uses LVAD; in our unit, ECMO is employed (Chaturvedi *et al*., 2004). The important principle is to use the mechanical assistance early. Increasing inotropic support, progressive elevation of the left atrial pressure and metabolic acidosis with a rising serum lactate level would be strong indications for early mechanical assistance. The left ventricle should be sufficiently recovered to cope with the afterload within 48 hours. The mechanical support is discontinued gradually, with the heart function supported on a combination of low-dose adrenaline and milrinone.

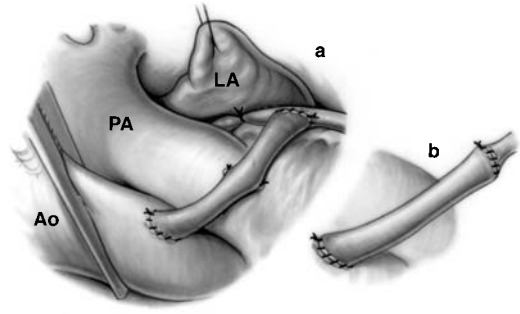

Figure 46.14

Coronary Artery Bypass Grafting

Autologous reverse saphenous vein graft, or a free segment of a subclavian artery, or a free radial artery graft have been used in preference to Dacron grafts since the original reports by Cooley *et al*. (1966), Neches *et al*. (1974) and Arciniegas *et al*. (1980). The graft can be implanted end-to-side after ligation of the origin of the coronary artery (**Figure 46.14a**) or end-to-end after division of the artery (**Figure 46.14.b**). However, coronary artery bypass vein grafting in the young should be avoided, because all vein grafts ultimately do become occluded. Internal mammary artery grafting, however, is

a good alternative because of better patency, at least in the medium term, in the young. Long-term patency has yet to be determined (Mavroudis *et al*., 1996).

Intrapulmonary Tunnel

Creation of an aortopulmonary window and intrapulmonary tunnel was suggested by Takeuchi *et al*. (1979) and by Hamilton *et al*. (1979). The operation suggested by Takeuchi *et al*. consists of the creation of an aortopulmonary window. The flap is cut from the anterior wall of the pulmonary artery adjacent to the aortopulmonary window (**Figure 46.15**). This flap is then sutured to the posterior wall of the pulmonary artery and around the ostium of the left coronary artery (**Figure 46.16a**). A direct tunnel from the aortopulmonary window to the left coronary artery is thus created. The defect in the pulmonary artery is then closed with a pericardial patch (**Figure 46.16b**).

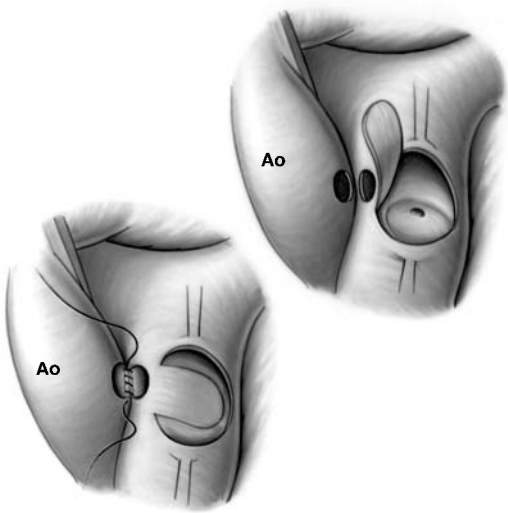

Figure 46.15

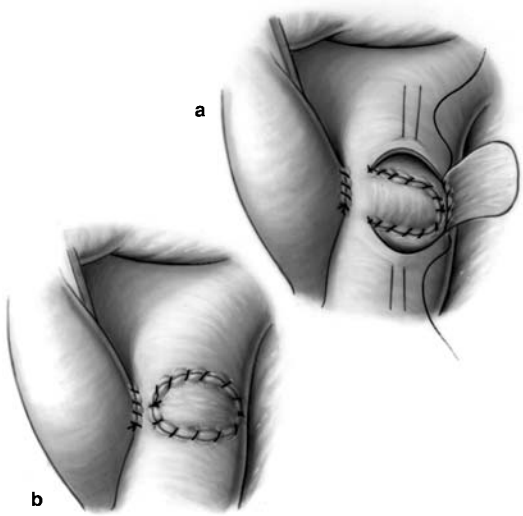

Figure 46.16

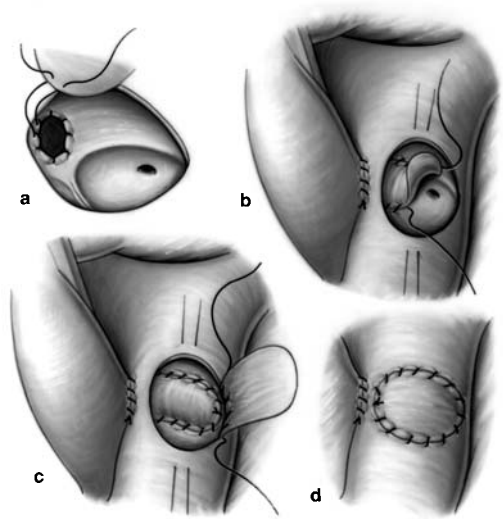

Figure 46.17

To avoid obstruction of the right ventricular outflow as a result of the intrapulmonary tunnel, Hamilton *et al*. (1979) used a pericardial baffle to connect the aortopulmonary window to the coronary artery (**Figure 46.17**), whereas Arciniegas *et al*. (1980) used a free segment of the subclavian artery. Alternatively a Gore-Tex tube may be used (**Figure 46.18**). However, the significant late supravalvar pulmonary stenosis deters the widespread use of this technique.

Management of Mitral Regurgitation

Although the establishment of a two-coronary artery system is the procedure of choice, it remains controversial as to how the regurgitant mitral valve should be handled. Based on his study of 31 children with ALCAPA, Vouhe *et al*. (1992) observed some important improvements in mitral valve function in most patients after successful coronary reimplantation. The mitral valve should not be interfered with at the initial procedure unless absolutely necessary. However, persistent mitral regurgitation may necessitate later operation. Huddleston *et al*. (2001) suggested that in the presence of late mitral regurgitation

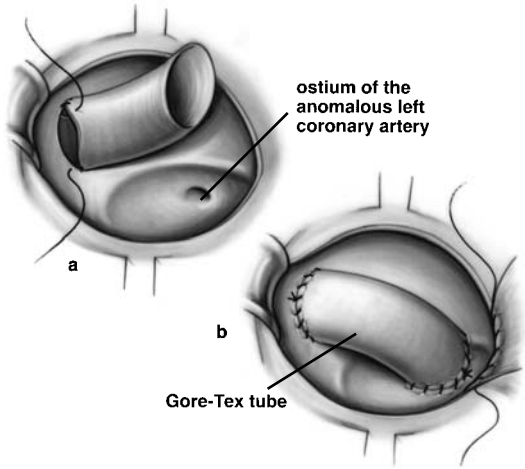

Figure 46.18

the coronary artery should be reassessed for stenosis. Concomitant revascularization and mitral valve repair may be necessary. On the other hand, Isomatsu *et al*. (2001) reported their experience of simultaneous mitral annuloplasty at the anterolateral commissure at the time of ALCAPA repair in all 24 patients with incompetence. Preoperative mitral regurgitation decreased in all but one of the operative survivors at follow-up. Late normalization of the left ventricular function after two-coronary repair was also noted.

Results

Results reported in earlier years were rather poor (Driscoll *et al*., 1981; Laborde *et al*., 1981) Arciniegas *et al*. reported two deaths among 12 operated patients, but six sick infants, who did not undergo an operation, all died.

Results with coronary artery reimplantation are more encouraging. Vouhe *et al*. (1992) operated on 31 consecutive patients between 1983 and 1991. There were three hospital deaths (10%); two patients (6%) died after discharge from the hospital, one from severe pulmonary infection and the other suddenly, presumably from arrhythmia, 2 months after discharge. The severity of the preoperative left ventricular dysfunction was the only risk factor for operative deaths. Of the patients with a preoperative shortening fraction of less than 20%, 31% died. None of those with a shortening fraction equal to or greater than 20% died ($p = 0.03$). There were no late deaths up to 6 years. Twenty-three patients had follow-up (mean, 4.9 years $\pm$ 2.2 years): 22 were free of symptoms and were taking no medication; one patient was in NYHA class II because of mitral regurgitation; 100% late patency of the reimplanted coronary artery was demonstrated on angiocardiography and Doppler echocardiography.

We performed 22 coronary reimplantations with three deaths (14%) during 1990–2005, The median age at repair was 6 months (range 57 days–5 years). There have been no hospital deaths during the last 10 years. Postoperative mechanical assistance was employed in five patients; they all survived. This has coincided with improved results of neonatal and infant cardiac surgery, especially with the introduction of postoperative mechanical assistance to support recoverable myocardium.

Few studies have reported the mid-term results in a reasonably sized patient population. Lambert *et al*. (1996) reviewed the outcomes of 39 consecutive patients with ALCAPA operated on during 1980–1995. The hospital mortality was 13% (five patients). There were no late deaths, but one patient required mitral valve repair 18 months after surgery. LV shortening fraction was normal in 84%. In some patients LV dilation and ischaemic segments of the LV persisted at long-term follow-up.

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47

Surgery for Arrhythmias

Y. Ishii, J. L. Cox and R. J. Damiano

Arrhythmia surgery began in the late 1960s with the successful division of a right free-wall accessory pathway for Wolff–Parkinson–White (WPW) syndrome (Cobb *et al*., 1968). Since that time, surgery has played an important role in the treatment of arrhythmias. The development of anatomically precise surgical techniques based on mapping data allowed for surgical correction of many medically refractory arrhythmias. Indications for arrhythmia surgery have been extended to include supraventricular and ventricular tachyarrhythmias. Knowledge and experience from surgical therapy for arrhythmias has fostered the development of catheter ablation techniques. Since the initial clinical application of catheter ablation in the early 1980s, the remarkable evolution of this therapy has changed the indications for surgery. The success of catheter ablation for WPW syndrome has made surgery unnecessary in patients with normally structured hearts. In spite of this development, surgery still plays a role in the treatment of some arrhythmias and demonstrates a high restoration rate to sinus rhythm. Surgical therapy for atrial fibrillation provides higher cure rates than catheter ablation. Patients with arrhythmias and congenital heart defects also benefit from arrhythmia surgery performed in conjunction with surgical repair of congenital heart defects, since correction of defect alone often will not reliably decrease the probability of an arrhythmia (Gatzoulis *et al*., 1999).

The principle strategy of arrhythmia surgery is to block conduction. The cut-and-sew technique provides the most reliable conduction block. Cryoablation has also been used historically to ablate tissue to create conduction block. Recently, to reduce surgical invasiveness, several new ablation technologies have been developed, such as unipolar and bipolar radiofrequency energy, microwave, laser and ultrasound. However, the reliability and safety of these alternative energy sources still need to be assessed. This chapter will describe the present state-of-the-art arrhythmia surgery.

SURGICAL ANATOMY

The anatomical considerations for surgical treatment of arrhythmias in children are the same as in adults and are independent of surgical technique.

The sinus node is located within the sulcus terminalis, between the superior vena cava and the right atrial appendage (James, 1965). Specialized cells within the sinus node generate calcium-dependent action potentials. While activation propagates through the whole atrium from the sinus node to the atrio-ventricular (AV) node, there are three preferential conduction pathways, which are composed of ordinary atrial fibres; the superior, middle and inferior pathways (James, 1963; Waldo *et al*., 1973). Atrial conduction propagates faster through these pathways because the muscle bundles are thicker and the long axes of the cells are parallel to each other (Spach *et al*., 1980). The superior interatrial pathway proceeds from the sinus node through the anterior portion of the atrial septum to the AV node. The middle pathway (Wenckebach's bundle) travels around the anterior limbus to the AV node from the sinus node. The inferior pathway runs along the sulcus terminalis from the sinus node to the AV node. Bachmann's bundle also provides a preferential connection from the sinus node to the left atrium in sinus rhythm.

The triangle of Koch is an important consideration in arrhythmia surgery, particularly in surgery for AV nodal re-entry (**Figure 47.1**). The anatomy of the triangle of Koch is described precisely in Chapter 2. The AV node lies between the upper part of the coronary sinus and septal leaflet of the tricuspid valve in the triangle of Koch, which is described in Chapter 8.

Patients with accessory AV connections have congenital abnormal muscle connections between the atrium and ventricle. Anatomically, the heart can be sectioned at the level of the AV groove and divided into four discrete areas in this horizontal plane. These four discrete areas are: the *left free-wall space*, which encompasses the majority of the tissue adjacent to the

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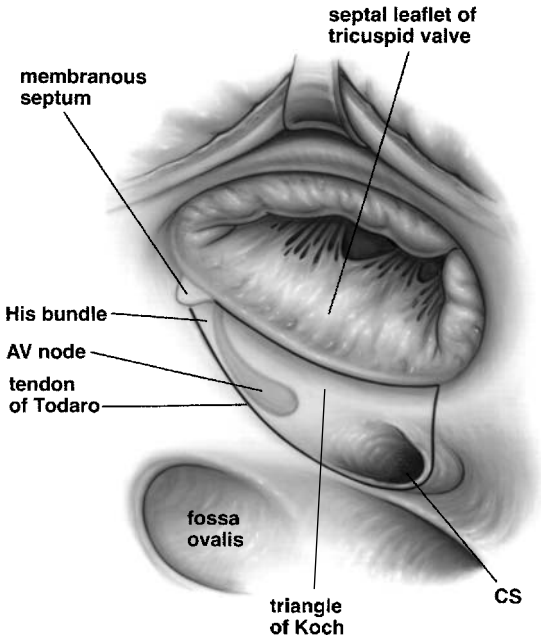

Figure 47.1

posterior leaflet of the mitral valve; the *right free-wall space*, which encompasses the anterior and posterior leaflets of the tricuspid valve; the *posterior septal space*, which is between the tricuspid and mitral annuli posterior to the central fibrous body of the cardiac skeleton; and the *anterior septal space*, which is between the aorta and anterior tricuspid annulus behind the pulmonary outflow tract (**Figure 47.2**). The two fixed boundaries defining these spaces are the left and right fibrous trigones of the skeletal structure of the heart. The other boundaries are defined by adjacent anatomical landmarks, which include the following:

1. The junction of the left free wall and the left posterior interventricular septum, separating the left free wall and posterior septal spaces and defined endocardially 1 cm posterior to the medial commissure of the mitral valve.
2. The junction of the right free wall and right posterior interventricular septum, separating the right free wall and posterior septal spaces and defined endocardially 1 cm anterior (onto the free wall) to the ostium of the coronary sinus. The junction of the right free wall and the pulmonary infundibulum, separating the right free wall and anterior septal spaces, is arbitrarily defined. It has been learned that in order to ablate a pathway surgically in one of these regions with a success rate approaching 100%, the entire anatomical space in which the pathway resides must be dissected in every patient.

The second anatomical fact about the AV groove that must be appreciated is that the fibrous skeleton of the heart is not completely horizontal. The cardiac skeleton is strongest at the central fibrous body, where the annuli of the mitral, tricuspid and aortic valves meet (**Figure 47.3**). The tricuspid annulus is more apical in position than the mitral annulus and is not in the same horizontal plane. Because of this, the anterior part of the central fibrous body extends into the ventricles beneath the attachment

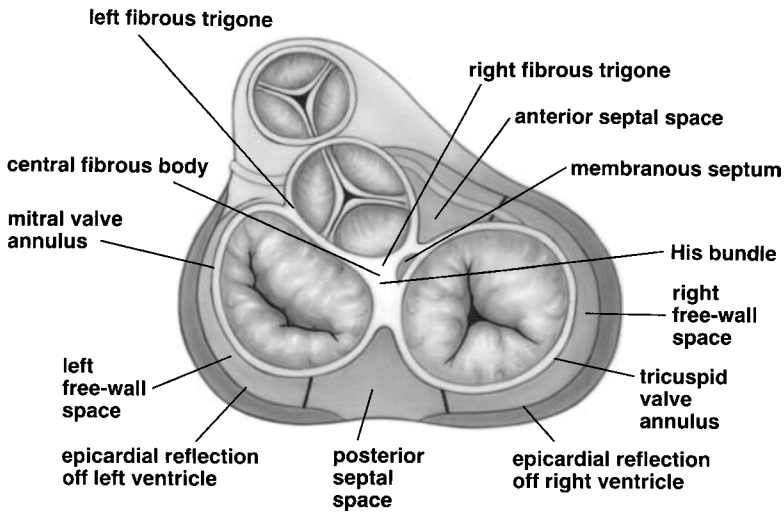

Figure 47.2

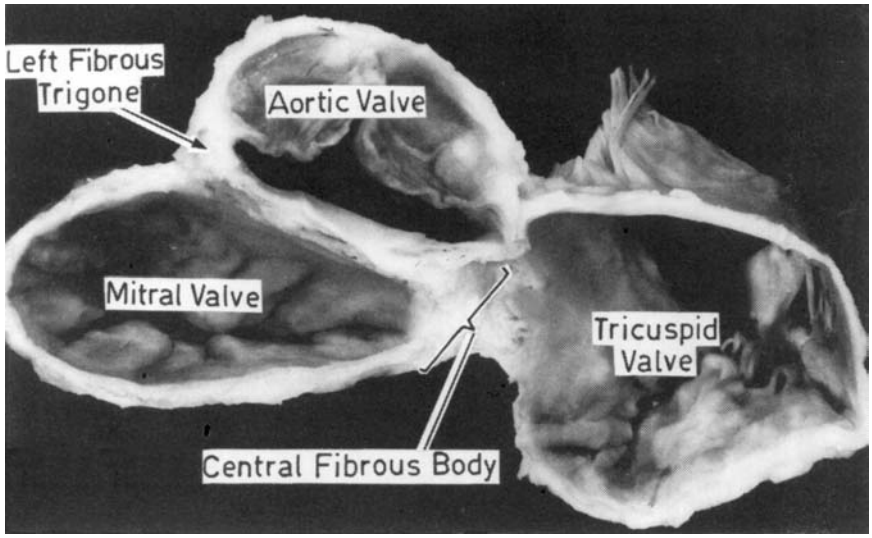

Figure 47.3

of the tricuspid valve, and the interventricular component of the membranous septum, between the aortic outflow tract and the right atrium, lies cephalad to the tricuspid annulus. This anatomical landmark is important, because the His bundle penetrates the central fibrous body just posterior to the membranous septum.

The mitral and aortic valve annuli contribute significantly to the structural integrity of the fibrous skeleton and are further strengthened at their junction to form the left fibrous trigone. The AV groove between the left fibrous trigone and the right fibrous trigone (the anterior portion of the central fibrous body) represents the site of continuity between the anterior leaflet of the mitral valve and the aortic valve annulus. This is the only area in the AV groove in which atrial muscle is not in juxtaposition to ventricular muscle and, for this reason, accessory AV pathways are not found between the left and right fibrous trigones. This part of the fibrous skeleton does lie in the horizontal plane of the AV groove.

Conceptualization of the anatomical relationships of these pathways in the vertical plane is more difficult. Because of the anatomical limitations imposed by the valve annulus on the inside of the AV groove, and by the epicardium on the outside of the AV groove in the vertical plane, all accessory pathways must connect atrium to ventricle somewhere between these two boundaries (**Figure 47.4**). No accessory pathway in a human being has ever been shown to pass through the tissue of the valve annulus. Therefore, in the vertical plane, accessory pathways are confined to locations:

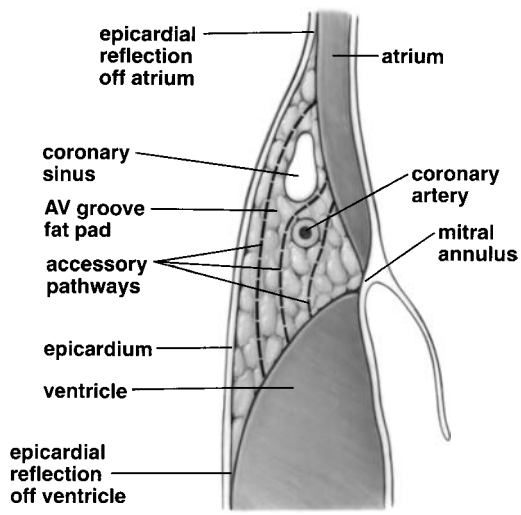

Figure 47.4

1. Adjacent to or near the valve annulus.
2. Within the fat pad of the AV groove.
3. Just beneath the epicardium overlying the AV groove.

Anatomically, ectopic focus or automatic atrial tachycardias can originate from foci occurring anywhere within the left or right atrial tissue or atrial septum. In addition, there may be multiple foci. Numerous studies have shown tachycardia originating from the right (70%)

and left (30%) atria (Mehta *et al*., 1988; Walsh, 1994). The ectopic foci cluster in a few specific areas of the right and left atrial appendages, and near or within the pulmonary venous ostia (Salerno *et al*., 2004). These findings suggest that the tachycardia involves subtle electrical changes in trabeculated atria or connections between the primitive atria and systemic veins.

In infants and children, ventricular tachycardia that is amenable to surgery is unusual and is most often associated with a cardiomyopathy, prior surgery for congenital heart defects, and cardiac tumours. There are anatomical considerations that relate to each of these entities.

Arrhythmogenic right ventricular dysplasia (ARVD) is a congenital myopathy remarkable pathologically for transmural infiltration of adipose tissue (Fontaine *et al*., 1999). As illustrated in **Figure 47.5**, this infiltration results in thinning (shown by the dotted line) or aneurysmal bulging, or both, of three areas of the right ventricle: the infundibulum, the apex and the posterior basilar region. Ventriculography demonstrates diffuse dilatation of the right ventricle with a significant reduction in contractility. Ventricular bulges or frank aneurysms are often seen in the right ventricle. ARVD is rarely treated surgically and drug-refractory patients are usually treated with implantable defibrillators.

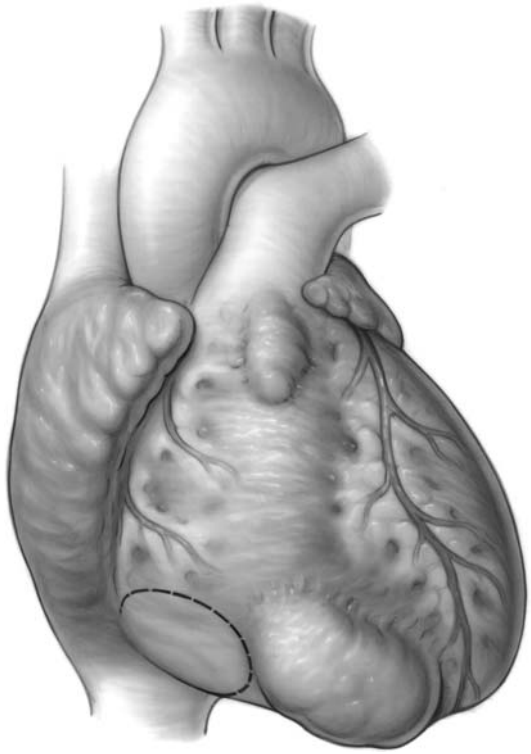

Figure 47.5

Cardiac tumours can be localized to the left or right ventricle, or septum in association with isolated arrhythmogenic tissue, or they can diffusely involve cardiac muscle and conduction tissue. Localized tumours may be epicardial, intramyocardial or on the endocardial surface of the heart.

In patients with congenital heart defects, especially tetralogy of Fallot, ventricular arrhythmias may develop years after the corrective procedure. The occurrence of these arrhythmias appears to be related in part to the haemodynamic result achieved with the operative procedure. Some of these arrhythmias have been shown to originate from the site of the right ventriculotomy (Gatzoulis *et al*., 2000).

ELECTROPHYSIOLOGICAL EVALUATION

Preoperative Mapping

A preoperative catheter electrophysiological study is often indicated. The difference between arrhythmia surgery and other cardiac surgery lies in the inability to visually confirm the pathology. Therefore, an electrophysiological study is important in understanding the mechanism of the arrhythmia. The goal of this study is to define the anatomical substrate responsible for the arrhythmia and to locate it as precisely as possible.

Re-entrant pathways, focal activation or accessory connections may be found by electrophysiological study in patients with supraventricular arrhythmias. While the evaluation of paediatric patients with supraventricular tachyarrhythmias is identical to that of adults, the incidence of these arrhythmias among children is different. Ectopic focus atrial or junctional tachycardia is uncommon in adults, but constitutes approximately 10% of supraventricular tachycardia in children (Ko *et al*., 1992). Localization of the ectopic focus at the time of the preoperative study is particularly important if surgical ablation is contemplated, because these tachycardias are frequently suppressed by general anaesthesia, which makes intraoperative localization difficult.

Tachycardias resulting from the presence of accessory pathways are the most common supraventricular arrhythmias (approximately 70%) in children (Ko *et al*., 1992). An additional 15% of supraventricular tachycardias in children are the permanent form of junctional reciprocating tachycardia (PJRT), which is uncommon in adults. This re-entrant mechanism involves an accessory connection just posterior to the AV node–His bundle tissue as the retrograde limb of the circuit. Finally, although AV nodal re-entry tachycardia does occur in children, it is more common in adults.

In patients with ventricular tachycardia, electrophysiological study is most often performed to establish the diagnosis of a wide QRS tachycardia and to localize

the earliest site of origin of the ventricular tachycardia before surgical ablation. Ventricular tachycardia in children less than 4 years old is usually caused by a definable lesion, most often tumours, which are very rare (Purkinje cell tumour, histocytic cardiomyopathy or myocardial hamartoma). Other causative tumours include rhabdomyoma, fibroma and myoma. Electrophysiological study in patients with ventricular ectopy after tetralogy of Fallot repair is important to affect a cure of the tachycardia. Patients with medically refractory ventricular ectopy or inducible ventricular tachycardia should be considered for surgical therapy or catheter ablation. Excision of the ventriculotomy scar has been curative in a number of patients (Harken *et al*., 1980).

Development of an electroanatomic mapping system (CARTO) has allowed collection of more detailed activation mapping data than the traditional electrophysiological study (Ben-Haim *et al*., 1996). The information obtained from the preoperative electrophysiological study helps to determine the indications for arrhythmia surgery and the type of surgical procedure that should be used.

Intraoperative Mapping

The goals of intraoperative mapping are to confirm the findings of the preoperative electrophysiological study and to rule out the presence of additional pathology. The complexity of the system used for intraoperative mapping varies from institution to institution. Meticulous intraoperative mapping performed with a hand-held mobile mapping electrode can be satisfactory in many circumstances. A number of limitations exist with this technique, however. More sophisticated computerized mapping systems have been used since the mid-1980s in both adults and children. The advantages of these systems include successful data acquisition and localization by mapping only a single beat of tachycardia, and the ability to define complex activation patterns such as atrial fibrillation. At our institution, we have used both single point and multipoint mapping systems.

SURGICAL TECHNIQUES

Supraventricular Tachyarrhythmias Atrial Fibrillation

Atrial fibrillation is the most common supraventricular tachycardia in the world, occurring in over 2 million people in the USA. The lifetime risk for development of atrial fibrillation is 1 in 4 for men and women 40 years of age and older (Lloyd-Jones *et al*., 2004). There is a significant incidence in adult congenital heart patients. For instance, atrial fibrillation has been shown to occur in 9–19% of patients with atrial septal defects (Gatzoulis *et al*., 1999).

This arrhythmia is responsible for substantial morbidity and mortality. Symptoms can include palpitations, dizziness, fatigue, shortness of breath and impaired exercise tolerance. Thromboembolism is frequently caused by atrial fibrillation. The rate of ischaemic stroke in patients with atrial fibrillation is six times that of the normal population (Wolf *et al*., 1991). Atrial transport function is also an important determinant of haemodynamic function in patients with ventricular diastolic dysfunction. Absence of the atrial contribution during atrial fibrillation impairs ventricular diastolic filling and has been shown to decrease cardiac output and stroke volume.

Drug therapy for atrial fibrillation has significant limitations. Medical management includes control of ventricular rate and pharmacological restoration of sinus rhythm. The Atrial Fibrillation Follow-up Investigation of Rhythm Management (AFFIRM) and Rate Control vs. Electrical Cardioversion for Persistent Atrial Fibrillation (RACE) studies evaluated strategies of rate control and rhythm control in atrial fibrillation (Corley *et al*., 2004; Hagens *et al*., 2004). These studies showed that rate control was as effective as rhythm control in terms of event-free survival. However, patients in both therapy groups were documented to need lifetime anticoagulation therapy. The Stroke Prevention in Atrial Fibrillation study actually showed that antiarrhythmic drugs were associated with increased mortality, and there was a trend toward this in AFFIRM (Corley *et al*., 2004). The limitations of medical therapy, and the potential side-effects of lifetime drug usage for children, make surgical therapy an attractive option for treatment of atrial fibrillation in this patient population.

INDICATIONS

In patients with congenital heart defects and atrial fibrillation, it is advisable to perform surgical therapy for atrial fibrillation concurrently with repair of the structural heart defects. Postoperative restoration of sinus rhythm provides more effective atrial transport function and allows patients to discontinue anticoagulation therapy. The strongest predictors of surgical success include shorter durations of preoperative atrial fibrillation, a cardiothoracic ratio $< 70\%$ and left atrial dimension < 80 mm (Kosakai *et al*., 1994).

Catheter ablation for atrial fibrillation has been used extensively throughout the world. Although the cure rates of atrial fibrillation by catheter ablation are improving, there is still a significant recurrence rate (15–20%) of atrial fibrillation (Stabile *et al*., 2003). Surgical therapy for atrial fibrillation should be considered for failed catheter ablation or in instances in which patients are referred for concomitant cardiac surgery.

The indications for surgical therapy for atrial fibrillation at our institution include those patients who have documented atrial fibrillation of over 3 months duration, with one or more of the following indications:

1. Failed antiarrhythmic drug therapy, electrical cardioversion or catheter ablation.
2. Severe symptoms and/or the development of a tachycardia-induced cardiomyopathy.
3. Development of a contraindication to anticoagulation.
4. Cerebral vascular accident in spite of adequate anticoagulation.
5. Patients with permanent atrial fibrillation undergoing concomitant congenital heart surgery.

The Cox–Maze procedure is the gold standard of surgical therapy for atrial fibrillation (Cox *et al*., 1991). Its most recent modification has been termed the Cox–Maze III procedure. The atrial incisions of the Cox–Maze III procedure are designed to block potential macrore-entrant pathways and propagation of microre-entrant wavelets (**Figure 47.6**). Moreover, focal activation from the pulmonary veins is blocked by the isolation of the left atrial posterior wall and all of the pulmonary veins (Cox *et al*., 1991). Studies have shown that the Cox–Maze III procedure restores normal sinus rhythm in over 90% of patients at late follow-up (Prasad *et al*., 2003). Recent modifications of the Cox–Maze procedure, using new ablation technologies, have been shown to be as effective and much simpler to perform. At our institution, we have developed a new modification (the Cox–Maze IV) of this procedure (Damiano *et al*., 2004). This new procedure has cut the cross-clamp time in half and is widely applicable to even the most complex patients.

In patients with right atrial volume and pressure overload, such as in Ebstein's anomaly, atrial fibrillation can arise from the right atrium. In these cases, surgery on the right atrium alone converts atrial fibrillation to sinus rhythm. The Mayo Clinic has reported a 100% cure of atrial fibrillation in patients ($n = 18$) with Ebstein's anomaly or tricuspid valve atresia by the right-sided Maze procedure (Theodoro *et al*., 1998). However, the cure rate of atrial fibrillation by using the right-sided Maze procedure for all forms of congenital heart disease is only 49% (Kosakai, 2000). In patients with left or biatrial pathology, the complete Cox–Maze III procedure lesion is recommended.

SURGICAL TECHNIQUE OF THE COX–MAZE III PROCEDURE

After median strenotomy and heparinization, the patient is placed on cardiopulmonary bypass with bicaval cannulation. A lateral incision of the right atrial (RA) free wall, parallel to the right AV groove, is placed from the RA appendage toward the IVC. The RA appendage is usually spared. A posterior longitudinal right atriotomy is then placed from the orifice of the SVC to a point near the pericardial reflection onto the IVC. The lower portion of the posterior longitudinal incision is closed immediately to prevent inadvertent tearing and extension into the IVC.

The first of two incisions in the RA free wall toward the tricuspid valve annulus is then performed. A T-shaped incision is made from the posterior longitudinal atriotomy, beginning 1–2 cm above the IVC cannula across the lower portion of the RA free wall, transversely to the right AV groove (**Figure 47.7**). The remainder of the incision, extending to the tricuspid valve annulus, is made endocardially with a scalpel, and any remaining muscles are divided with a nerve hook. Care should

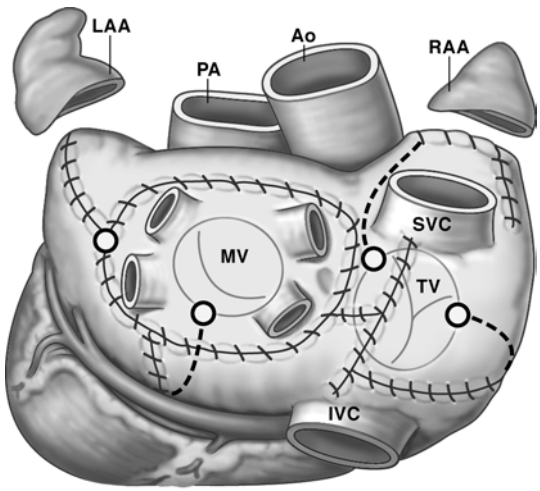

Figure 47.6

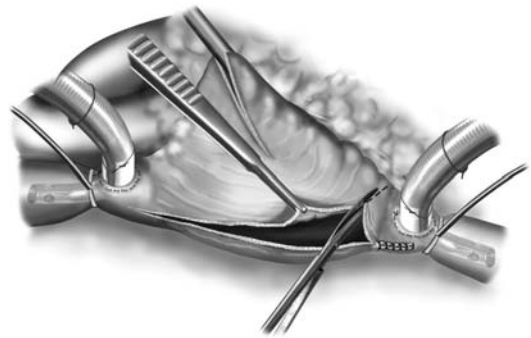

Figure 47.7

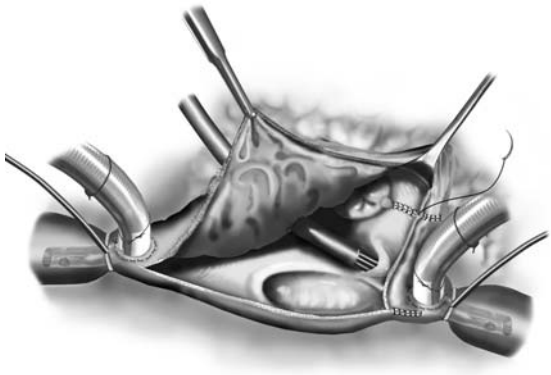

Figure 47.8

be taken to avoid injury to the right coronary artery. A cryolesion is placed at the end of the incision at the tricuspid valve annulus for 2 minutes at −60°C (**Figure 47.8**).

An anterior RA incision is then placed from the base of the RA appendage toward the AV groove. This incision is extended to the anteromedial tricuspid valve annulus by dividing the remaining atrial myocardial fibres with a scalpel from an endocardial approach. After a cryolesion is placed on the tricuspid annulus, the entire incision is closed from the annulus to the RA appendage (**Figure 47.9**). The RA incisions can be performed on the beating heart if patients do not have any interatrial or interventricular communications.

After cross-clamping of the aorta and cardioplegic arrest, a standard left atriotomy is carried out in the interatrial groove. The atrial septum is divided from 2–3 cm below the orifice of the SVC across the anterior limbus of the fossa ovalis (**Figure 47.10**).

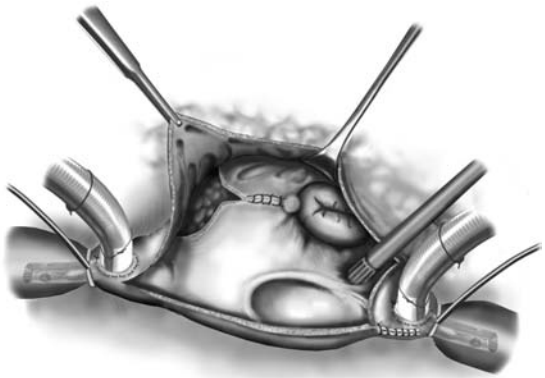

Figure 47.9

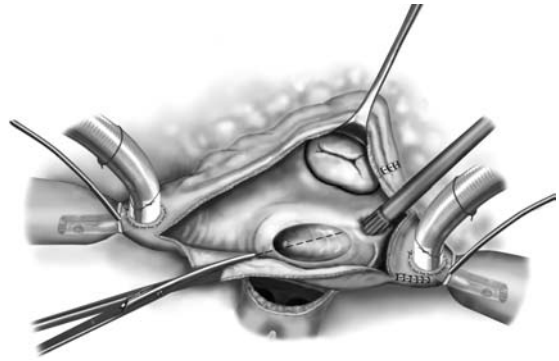

Figure 47.10

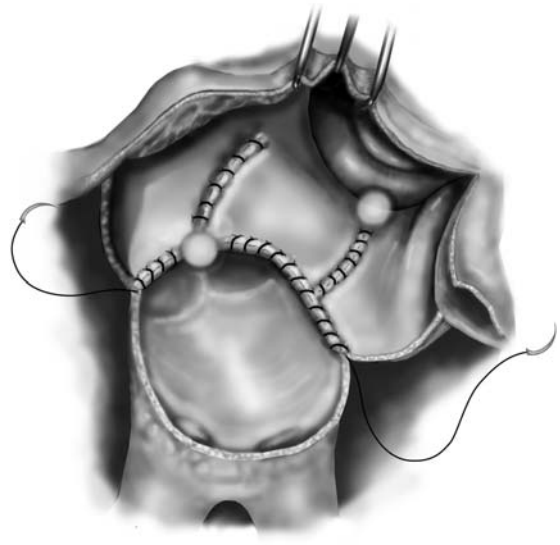

Figure 47.11

The atrial septum is retracted, and the superior and inferior ends of a standard left atriotomy are extended around the LA posterior wall, encircling the pulmonary veins. Before the two ends of the left atriotomy are joined below the LA appendage, the inverted LA appendage is excised. A 1.5 cm cryoprobe (3 minutes at −60°C) is applied between the two ends of the left atriotomy and the LA appendage incision. The LA appendage incision and several centimetres of the left atriotomy encircling the pulmonary veins are then closed (**Figure 47.11**).

After the inferior left atriotomy is closed to a point midway between the right and left inferior pulmonary veins, a perpendicular incision is extended to the mitral valve annulus. All visible left atrial myocardial fibres are

divided with a scalpel or nerve hook. Care must be taken not to injure the coronary sinus or circumflex coronary artery. Trans-mural cyoablations (3 minutes at -60°C) are placed on the mitral annulus and coronary sinus, because the mitral annulus is thicker than the tricuspid annulus. Incomplete ablation allows postoperative conduction and results in recurrence of atrial flutter around the mitral valve. If retrograde cardioplegia is used, the cannula should be removed prior to cryoablation. If mitral valve repair or replacement is required, it can be performed at this time. The left atriotomy and septal incision are then closed. The remaining incisions of the RA are then closed to complete the procedure (**Figure 47.12**).

Recently, investigators have used various different ablation technologies, including microwave, radiofrequency energy, cryoablation, laser and ultrasound (Gillinov *et al*., 2002; Williams, 2002). All of these energy sources have been used successfully in the operating room. The goal of these new devices is to create conduction block and replace the surgical incisions. Depending on their particular application, it is important for surgeons to ensure that they are capable of producing conduction block, either from the endocardial surface when used in arrested heart cases, or epicardially on a beating heart. The only reliable way to create conduction block in myocardial tissue is a trans-mural lesion. It is important for surgeons to understand the effect of these energy sources on surrounding vital structures, and to develop knowledge of the dose–response curves, in order to avoid collateral injury.

In our centre, bipolar radiofrequency energy has been favoured over other sources, because it can create rapid trans-mural lesions in a reliable and consistent manner (Prasad *et al*., 2002). These devices use online measurement of conductance as a surrogate for trans-murality. Experimental and clinical evidence has validated this approach (Prasad *et al*., 2002; Mokadam *et al*., 2004). In our series, the use of this ablation device has cut our cross-clamp time in half for a lone Maze procedure, and by over 30 minutes with concomitant procedures. More importantly, the procedure has been greatly simplified. These new ablation devices may have significant application in the paediatric population and enable surgeons to perform atrial fibrillation surgery more easily and quickly.

Atrial Septal Defect and Atrio-ventricular Septal Defect with Atrial Fibrillation

Surgical treatment for atrial fibrillation carries a risk of interatrial conduction block in patients with more complex atrial anomalies, resulting in permanent pacemaker implantation. It is important to give due consideration to the atrial conduction from the sinus node to the AV node prior to surgery, in order to prevent conduction block. In patients with an *atrial septal defect*, none of the conduction pathways are interrupted by the defect. If they undergo the Maze procedure for atrial fibrillation concomitant with repair of the atrial septal defect, the posterior longitudinal, septal and lower right atrial incisions block atrial conduction pathways through the anterior free wall and posterior septum, as in patients with no atrial septal defect (**Figure 47.13**). Atrial conduction can still propagate from the sinus node to the AV node through the anterosuperior septum without any interatrial conduction block after surgery. However, in patients with *atrio-ventricular septal defect*, atrial conduction through the anterosuperior septum is already interrupted by the defect (Waldo *et al*., 1973). Atrial conduction propagates through the anterior free wall and posterior septum from the sinus node to the AV node. The complete incisions of the Maze procedure in patients with atrio-ventricular septal defect may risk dividing atrial conduction pathways through the anterior free wall and posterior septum (**Figure 47.14**), resulting in isola-

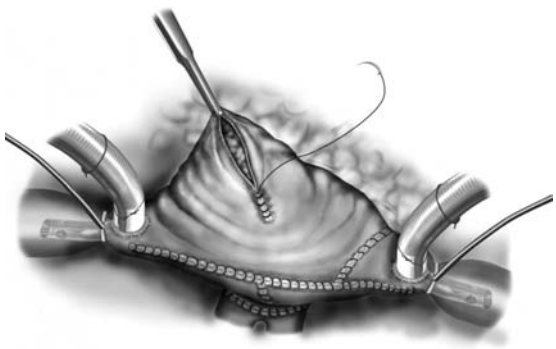

Figure 47.12

Figure 47.13

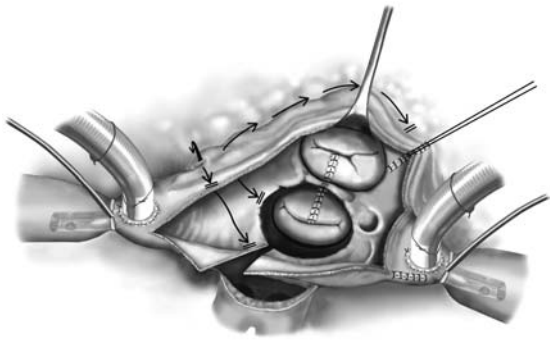

Figure 47.14

tion of the AV node. Therefore, patients have interatrial conduction block and require pacemaker implantation. Omitting the posterior septal incision preserves atrial conduction through the posterior septum and effectively prevents interatrial conduction block (Nitta *et al*., 2002).

Management of Postoperative Medications

Surgeons should be aware of several unique aspects of the postoperative management of patients following surgery for atrial fibrillation. These patients are prone to fluid retention, which may occur in up to one-third of patients. Atrial natriuretic peptide secretion has been reported to be severely reduced by some investigators (Yoshihara *et al*., 1998). Moreover, ADH and aldosterone have been shown to be significantly elevated following these procedures (Ad *et al*., 2002). The problem is managed effectively with diuretics and spironolactone (50–75 mg/day) for 2–4 weeks.

These patients have a very high incidence of early postoperative atrial tachyarrhythmias (Ishii *et al*., 2004). These arrhythmias include atrial fibrillation, atrial flutter and atrial tachycardia. Their peak incidence occurs on postoperative day 8, and 87% of patients who have these arrhythmias will experience them in the first 2 weeks. The incidence is higher in patients undergoing a concomitant Maze procedure vs. a lone procedure (54% vs. 39%). In a follow-up study from our series, the late recurrence of atrial fibrillation was not affected by whether or not the patients had early postoperative atrial arrhythmias. These arrhythmias are treated with antiarrhythmic drugs, most often amiodarone. These are continued for the first 2 months, and then discontinued if the patient is in normal sinus rhythm. Elective cardioversion is performed, if necessary, immediately prior to discharge or at the 1-month follow-up visit. Because of the high incidence of atrial tachyarrhythmias in these patients, the patients are placed on coumadin for 2–3 months.

Accessory Atrio-ventricular Connections in Infants and Children

Indications

The success rate for the surgical division of accessory AV connections has been high (Cox *et al*., 1985). In general, the surgical techniques which have been developed for the adult population have worked well in the paediatric age group. Catheter radiofrequency ablation also has had a high success rate for treatment of accessory AV connections (Jackman *et al*., 1991) and currently is the first choice of therapy. Surgical therapy is performed in patients who have failed catheter ablation and in patients who are having concomitant surgery for structural heart defects.

Electrophysiological Evaluation

Preoperative electrophysiological evaluation provides the surgeon with important information about the location of accessory pathway(s). This may be particularly important in the up to 9% of patients with multiple pathways (Weng *et al*., 2003). Intraoperative mapping, when available, should be performed epicardially and/or endocardially to confirm the findings of the preoperative evaluation. The atrial activation sequence can be determined by ventricular pacing and induced tachycardia.

Surgical Technique

The purpose of surgery for accessory atrio-ventricular connections is to block the accessory connections that are responsible for re-entrant tachycardia, by ablating them with either surgical incisions or cryoablation. Endocardial (**Figure 47.15**) and epicardial surgical approaches have been developed. Although the endocardial approach is performed with cardiopulmonary bypass, the epicardial approach can often be performed on the beating heart.

Left Free Wall Accessory Connections—Endocardial Approach

Accessory connections on the left free wall are approached through a standard left atriotomy. A supra-annular incision is placed 2 mm above the posterior mitral valve annulus (**Figure 47.16**). The AV groove fat pad is dissected from the top of the left ventricle through the entire length of the supra-annular incision (**Figure 47.17**). Both ends of the supra-annular incision should be carried to the mitral annulus to block juxta-annular connections. The supra-annular incision is closed with a continuous suture.

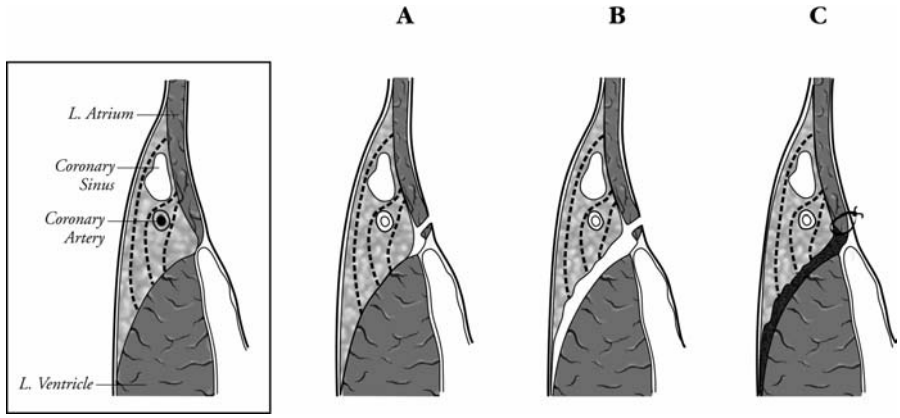

Figure 47.15

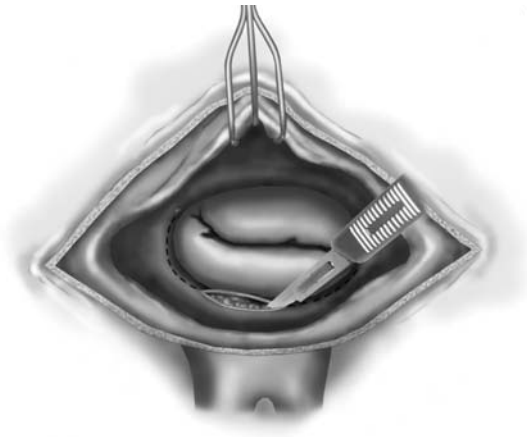

Figure 47.16

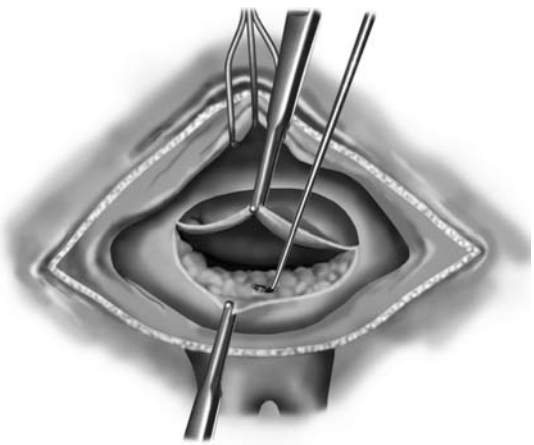

Figure 47.17

Left Free Wall Accessory Connections—Epicardial Approach

The AV groove is exposed by using right-ward cardiac retraction. An apical suction device can aid in this exposure. The epicardial reflection of the atrium is incised, and a plane of dissection is established between the AV groove fat pad and the atrial wall. The dissection plane is extended to the level of the posterior mitral valve annulus and slightly onto the posterior left ventricle. Care should be taken to avoid injury of the coronary artery and coronary sinus. Cryoablation may be necessary to complete the dissection. If a patient develops unstable haemodynamics, cardiopulmonary bypass should be initiated.

Posterior Septal Accessory Connections

Accessory connections in the posterior septal space can be divided during normothermic cardiopulmonary bypass. A supra-annular incision is placed 2 mm above the posterior medial tricuspid valve annulus (**Figure 47.18a**). This supra-annular incision is extended in a counter-clockwise direction onto the posterior right atrial free wall. A plane of dissection is established between the fat pad of the AV groove and the top of the posterior ventricular septum (**Figure 47.18b**). The fat pad is dissected off the posterior aspect of the central fibrous body (**Figure 47.18c**). The plane of dissection between the fat pad and the top of the posterior ventricular septum is developed by following the mitral annulus over to the posterosuperior process of the left ventricle and by following the epicardial reflection from the posterior right ventricle across the posterior crux onto the posterior left ventricle (**Figure 47.19**).

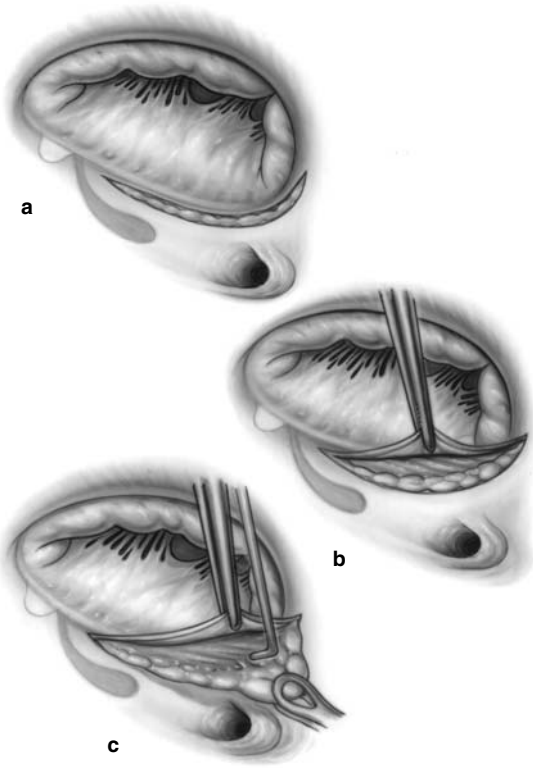

Figure 47.18

Right Free Wall Accessory Connections—Endocardial Approach

The endocardial approach for right free wall accessory connections should be performed in patients in conjunction with repair of other congenital defects. A supra-annular incision is placed 2 mm above the tricuspid valve annulus, around the entire right atrial free wall. A plane of dissection is established between the AV groove fat pad and the top of the right ventricle throughout the length of the supra-annular incision.

Right Free Wall Accessory Connections—Epicardial Approach

In most patients with right free wall accessory connections, it is possible to perform an epicardial dissection without cardiopulmonary bypass. The epicardial reflection of the atrium is incised, and a plane of dissection is established between the AV groove fat pad and the atrial wall. The dissection plane is extended to the level of the tricuspid valve annulus and carried slightly onto the right ventricle.

Anterior Septal Accessory Connections—Endocardial Approach

Accessory pathways in this location are frequently adjacent to the right fibrous trigone and are typically situated just anterior to the recorded His deflection. Endocardial

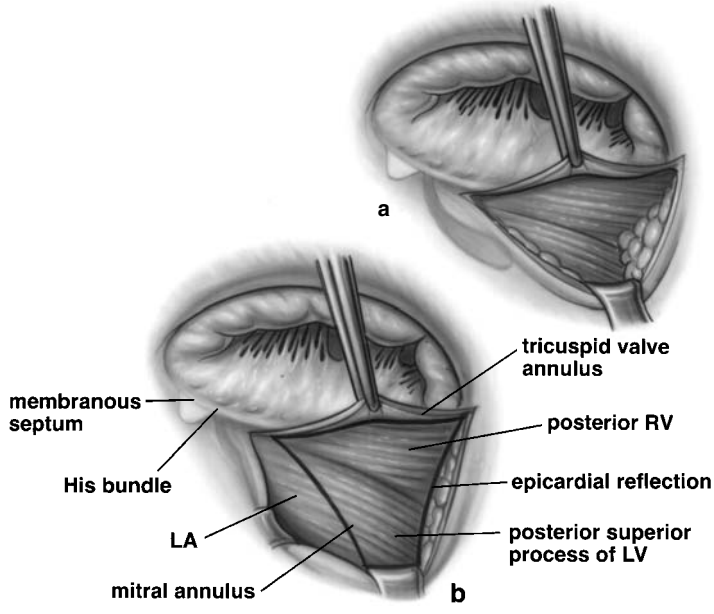

Figure 47.19

mapping around the annulus of the tricuspid valve is particularly helpful in defining the precise location of the accessory pathway in relation to the His bundle.

A supra-annular incision is placed anterior to the His bundle, 2 mm above the annulus, and extended in a clockwise direction onto the free wall of the right atrium. The extensive fat pad occupying this anatomical space is dissected to the epicardial reflection of the right ventricle. The medial boundary of the dissection is the aortic wall beneath the right coronary artery orifice, and this portion of the aorta is quite thin. The fat pad contains the proximal right coronary artery before it inserts into the AV groove, and retraction must be performed carefully. The dissection is carried anteriorly to the epicardial ventricular refraction. The initial portion of this dissection is usually performed while the heart is in sinus rhythm, in order to monitor for changes in AV conduction. The dissection can be completed with the heart arrested, if necessary. Cryosurgery can be used to isolate the atrial rim of tissue in this dissection, and the supra-annular incision is closed in the usual fashion.

Another approach using cryoablation has been described to treat anterior and posterior septal pathways in infants and children (Lee *et al*., 1989). In this technique, the exposure is similar to that described above. Careful epicardial and endocardial intraoperative mapping is performed. The anterior or posterior septal region is cryoablated at the site of earliest activation. Repeated overlapping cryoablation is extended progressively toward the AV node. Ablation of the accessory pathway is completed when the delta wave disappears from the electrocardiogram. When PR interval prolongs significantly, or when transient complete heart block occurs, cryoablation should be terminated.

Concealed Accessory Pathways and Permanent Form of Junctional Reciprocating Tachycardia (PJRT)

Concealed accessory pathways conduct only in a retrograde direction. The ventricle is activated through the normal AV node–His bundle complex, and thus the electrocardiogram is normal during sinus rhythm. The reciprocating tachycardia that results is termed ''paroxysmal supraventricular tachycardia''. The intraoperative approach to these pathways involves only retrograde atrial epicardial mapping during ventricular pacing and reciprocating tachycardia, and the surgical techniques are identical to those described earlier.

As mentioned earlier, PJRT is a relatively common arrhythmia in children. Because the concealed accessory connection lies in the posterior septal space just posterior to the His bundle, patients with PJRT are managed in the same way as other patients with posterior septal pathways. In most cases, it is possible to separate the pathway from the normal conduction tissue and not to have to resort to ablation of the His bundle for control of the arrhythmia.

Ectopic Focus (Automatic) Atrial Tachycardias

Surgical intervention in a patient with ectopic focus atrial tachycardia is warranted in cases of failure of medical and catheter-based therapy. Preoperative localization of the origin of the tachycardia is critical, since intraoperative induction of the tachycardia may be difficult.

If the tachycardia focus can be localized, a variety of techniques can be used for surgical treatment. Cryoablation of the ectopic focus, with or without cardiopulmonary bypass, is effective (Bredikis *et al*., 1991). However, it should be realized that epicardial cryoablation may not produce a transmural lesion in the normothermic beating heart (Doll *et al*., 2003). A combination of cryoablation and resection might be effective in some cases (Graffigna *et al*., 1992). For multiple ectopic focal tachycardias, more extensive techniques could be required, such as right or left atrial isolation or pulmonary vein isolation (Bredikis *et al*., 1991; Doll *et al*., 2003).

Incisional Atrial Re-entrant Tachycardia After Surgery for Congenital Heart Disease

Incisional atrial re-entrant tachycardia (IART) is a life-threatening complication that occurs after repair of congenital heart defects (Girod *et al*., 1987). This tachycardia is often medically refractory (Balaji *et al*., 1994). The incidence of IART has been found to be 20–38% in patients followed late after the Fontan procedure (Girod *et al*., 1987; Porter and Garson, 1993). In patients who have undergone atrial septal defect repair, it has been shown that 4–18% of patients experience this tachycardia during a 10- to 32-year follow-up period (Murphy *et al*., 1990). These patients can suffer from palpitations, fatigue, dyspnoea, syncope and/or haemodynamic collapse during tachycardia. The re-entrant tachycardia results from slow conduction activating through an isthmus between two anatomical barriers (Ishii *et al*., 2003). The right atrial scars or atrial septal baffle after the Fontan procedure can also be a substrate responsible for re-entrant tachycardia (Gandhi *et al*., 1996). Catheter ablation therapy has successfully treated this tachycardia. However, 33–53% of patients treated by catheter ablation suffer recurrence (Kalman *et al*., 1996). Therefore, surgery to prevent incisional atrial re-entrant tachycardia is an important treatment option.

It has been demonstrated that the mechanism of IART is associated with slow conduction through a critical narrow isthmus (5–15 mm) (Ishii *et al*., 2003). Electroanatomical mapping after surgical repair of congenital

heart defects has shown that re-entrant activation traversed through the narrow isthmus between surgical scars in patients with this arrhythmia (Nakagawa *et al*., 2001). IART can be prevented during surgery simply by leaving a sufficiently large isthmus. In a small infant, however, it may be impossible to maintain an isthmus of more than 15 mm between the atriotomy and the cannulation site on the right atrium. One option is to use a bicaval cannulation technique, inserting cannulas directly into the superior and inferior vena cavae to avoid creating multiple incisions on the right atrium. If this can not be done, the tissue between the incisions should be ablated so that no isthmus remains between them.

After a Fontan procedure, the right atrial scar or atrial septal baffle can cause IART (Gandhi *et al*., 1996). The incidence of IART after the Fontan procedure is higher than that after simple atriotomy, such as that used in surgery for atrial septal defect or tetralogy of Fallot (Balaji *et al*., 1994; Porter and Garson, 1993). Factors that lead to the development of IART after the Fontan procedure include not only the atrial scars due to the multiple atriotomies, but also the elevated atrial wall stress or atrial hypertrophy resulting from the atrial pressure and volume overload (Ghai *et al*., 2001). Non-trans-mural suture lines, as in those placed during the lateral tunnel procedure, also cause slow conduction and re-entrant circuits (Gandhi *et al*., 1996). After the Fontan procedure, the right atrium has several isthmuses lying between incisional scars. An extracardiac conduit Fontan procedure can reduce the number of incisional scars on the right atrium, and this modification of the procedure may lower the postoperative incidence of IART (Haas *et al*., 2000). For patients with postoperative refractory atrial arrhythmias, total cavopulmonary artery conversion and concomitant arrhythmia surgery (isthmus cryoablation, right-sided Maze procedure or the Cox–Maze III procedure) is also excellent therapy (Mavroudis *et al*., 2002). This procedure has a low mortality and the reported arrhythmia recurrence rate was 12%.

VENTRICULAR TACHYARRHYTHMIAS AFTER REPAIR OF CONGENITAL HEART DEFECTS

Ventricular tachyarrhythmias that occur after repair of tetralogy of Fallot are a life-threatening complication (Gatzoulis *et al*., 2000). Ventricular tachycardia is one of the main causes of late morbidity and sudden death in these patients. The incidence of sudden death for patients after repair of tetralogy of Fallot is 2–6% at 20–30 years of follow-up (Gatzoulis *et al*., 2000; Murphy *et al*., 1993). This arrhythmia is associated with elevated right ventricular end-diastolic volumes, pulmonary regurgitation, or a residual shunt from a ventricular septal defect patch after surgery (Gatzoulis *et al*., 2000; Marie *et al*., 1992). Right ventricular myocardial stretch due to an enlarged right ventricle has been associated with inhomogeneous ventricular activation (Hansen *et al*., 1990). Ventricular scar tissue from the right ventriculotomy also plays an important role. The incisional scar creates a critical isthmus with slow ventricular conduction that facilitates ventricular tachycardia (Dietl *et al*., 1994).

Preservation of pulmonary valve function during repair of tetralogy of Fallot may reduce the incidence of ventricular tachycardia. Patients who receive a trans-annular patch for the reconstruction of the right ventricular outflow tract, which results in pulmonary regurgitation, have been found to have a significantly higher late incidence of ventricular tachycardia after surgical correction than patients who did not receive a trans-annular patch (Gatzoulis *et al*., 2000). If patients have pulmonary insufficiency after surgery, early reoperation for asymptomatic right ventricular dysfunction improves ventricular function and decreases the incidence of ventricular tachycardia (Ilbawi *et al*., 1987).

If ventricular tachycardia occurs in patients after repair of tetralogy of Fallot, intraoperative endocardial mapping can aid in determining the activation pathway (Therrien *et al*., 2001). Ideally, surgical correction of structural abnormalities should be performed, accompanied by intraoperative map-guided surgical ablation (Schalij *et al*., 1998).

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48

Paediatric Heart and Heart–Lung Transplantation

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HEART TRANSPLANTATION

Paediatric heart transplantation was first attempted in 1967 in a 16-day-old child with tricuspid atresia (Kantrowitz *et al*., 1968). The following year, a 2-month-old child with atrio-ventricular septal defect also received a heart transplant (Cooley *et al*., 1969). However, the level of interest in this field was low throughout the 1970s, due to poor outcome in children as well as in adults, essentially from the lack of adequate immunosuppression. This disinterest is evident in the registry of the International Society of Heart and Lung Transplantation (ISHLT), which reveals that, before the mid-1980s, very few children received transplants each year, and the majority of these children were adolescents (Boucek *et al*., 2004). Activity started to increase after various factors in the 1970s and 1980s and led to a marked improvement in survival. These included improvements in patient and donor selection, myocardial preservation, surgical technique, and the development of surveillance programmes for the detection of rejection by endomyocardial biopsy. The most important factor, however, was the introduction of cyclosporine, and hence less reliance on corticosteroids for immunosuppression.

The total number of transplants has steadily increased from 1982, reaching a plateau in 1990. Since then, 300–400 cases have been performed worldwide every year. Another noticeable change was an increase in the proportion of younger recipients. Since 1990, about one-third of paediatric heart transplants every year have been performed in patients younger than 1 year, one-third in recipients aged 1–10 years, and one-third in those aged 11–17 years. The number of centres has also increased, and there are now over 70 centres that perform paediatric heart transplantation. However, it may now be beneficial to restrict this specialized transplant activity to fewer centres with greater and more concentrated expertise.

INDICATIONS

Cardiac transplantation is offered to patients in isolated end-stage irreversible heart failure from whatever cause, having exhausted all other medical and surgical options. The primary diagnosis in the majority of children undergoing heart transplantation (**Table 48.1**) is either congenital heart disease or cardiomyopathy, with congenital heart disease being the more common diagnosis in the younger age group and cardiomyopathy in older children (Boucek *et al*., 2004). Other conditions, such as cardiac tumours and retransplantation, account for a very small percentage of transplant activities.

The majority of cardiomyopathies in children requiring transplantation are dilated cardiomyopathies. These are mainly idiopathic, although sometimes familial. Ischaemic cardiomyopathy is very rare in children. Transplantation is rarely required for hypertrophic cardiomyopathy in children. Restrictive cardiomyopathy is problematic, as the high pulmonary vascular resistance causes early right ventricular failure following transplantation. We reported recently a series of paediatric transplants for restrictive cardiomyopathy and although mortality was low, there was a high early morbidity (Fenton *et al*., 2006).

The majority of congenital cardiac defects are not indications for transplantation, as they can be successfully corrected or palliated. However, some children may develop heart failure at a later stage and become transplant candidates. Examples of referrals from adult congenital practice include ventricular dysfunction in transposition of the great vessel following atrial repair, and ventricular dysfunction following repair of tetralogy of Fallot. In children, the failing Fontan procedure now represents a growing need for heart transplantation (Mitchell *et al*., 2004). Although the Fontan procedure achieves good results in staged palliation of most complex congenital heart defects, it is associated with a

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Table 48.1 The indications for heart transplantation in different age groups. The prevalence of each indication is described as a percentage of the total number of heart transplants in that age group. Data obtained from the seventh official paediatric report of the International Society for Heart and Lung Transplantation (Boucek *et al*., 2004).

Indications	Age < 1 year (%)	Age 1–10 years (%)	Age 11–17 years (%)
Congenital	66	37	28
Cardiomyopathy	30	52	62
Other	3	4	4
Retransplant	1	7	6

steady decline in survival, even in the best candidates, in whom 5-year and 15-year survivals are only 86% and 74%, respectively (Fontan *et al*., 1990). Although recent modifications of the Fontan procedure have improved its long-term outcome, they have not eliminated its relentless attrition rate (Gentles *et al*., 1997; de Leval, 1998; Freedom *et al*., 2000). Furthermore, interstage attrition is also expected, with some patients requiring transplantation having already undergone a superior cavopulmonary anastomosis but before the Fontan circuit is completed as the final stage procedure (Michielon *et al*., 2003; Jayakumar *et al*., 2004).

Patients with failing Fontan circulation represent a very high risk for transplantation, for several reasons. First, the assessment of pulmonary vascular resistance in a Fontan circulation is inaccurate (see below). Consequently, an element of doubt will always exist as to whether the right ventricle of the donor heart will function well or not following transplantation. Second, an immunologically suitable organ may not be found because of the presence of raised recipient lymphocytotoxic IgG class antibodies, which have arisen in response to previous blood transfusions or possibly the previous use of a homograft (Carey *et al*., 1998). Third, many patients with a failing Fontan circulation suffer from protein-losing enteropathy (Michielon *et al*., 2003; Mitchell *et al*., 2004). Fourth, they also suffer from hepatic dysfunction, resulting in coagulopathy. Fifth, there will be technical difficulties in these patients who have had at least three previous operations, with altered pulmonary artery anatomy. It is, therefore, not surprising that the ISHLT data show that congenital heart disease is the number one independent risk factor for mortality within 1 year for paediatric heart transplantation. The Fontan patient represents one of the highest risk groups, with most reports quoting an operative mortality of around 30%, although the Denver group have reported good results (Mitchell *et al*., 2004).

Despite the difficulties of transplantation in patients with a failing Fontan circulation, a time comes when it is the only option left for these patients (Gamba *et al*., 2004). However, the results of transplantation are better if offered to those who have had a superior cavopulmonary anastomosis but before the completion of the Fontan circulation (Michielon *et al*., 2003; Jayakumar *et al*., 2004). Transplantation should therefore be considered as an alternative to Fontan completion in high-risk candidates (severe atrio-ventricular valvar incompetence and ventricular dysfunction).

CONTRAINDICATIONS

Absolute contraindications are multiorgan failure, active infection, malignancy, and severely elevated pulmonary vascular resistance. Controversy exists regarding transplantation in children with prior malignancy because of the risk of recurrent disease or of new malignancies, and also determining the length of time that the child should be free of disease prior to transplantation. All of these would be a function of the type and stage and response to treatment of each malignancy. A recent review suggested reducing the time of remission before transplant could be considered from 5 years (Ward *et al*., 2004).

Various chromosomal anomalies that may prevent functional life would also be considered relative (but not absolute) contraindications to transplantation. Similarly, lack of family support and various psychosocial problems would be also significant factors, since life after transplantation requires compliance with treatment. Nonadherence to medication is a leading cause of morbidity in children and adolescents who have had a transplant (Shemesh *et al*., 2004). ISHLT survival figures show a much worse survival for teenagers, and this is likely to be related to compliance. We have regular psychological input with the teenagers and have started a summer school by the psychology team. This aims to improve confidence and independence and reinforce the need for good compliance.

The presence of HIV infection is considered a relative contraindication to transplantation by most centres. We have transplanted hepatitis C-positive children, as with both hepatitis B and C the natural history of these conditions in the paediatric population is not clear (Webber, 2003).

PREOPERATIVE EVALUATION

Preoperative evaluation consists of the confirmation of the indications and ruling out of the contraindications for transplantation, starting the matching process once the patient has been accepted as a candidate, and the

education of the patient and the family of the patient regarding life after transplantation.

Cardiac catheterization is performed to measure the trans-pulmonary gradient as well as the pulmonary vascular resistance. In general, a trans-pulmonary gradient of less than 15 and an indexed pulmonary vascular resistance of no more than 6 Wood's units are required for heart transplantation. If the pulmonary vascular resistance is elevated, its reversibility would also be assessed (Webber 2003; Gajarski *et al*., 1994). Pulmonary vascular resistance greater than 12 Wood's units would be a contraindication to heart transplantation, but such a patient may be suitable for heart–lung transplantation. In borderline cases, the patient may be kept in hospital and treated with an intravenous inotrope, such as dobutamine, and a pulmonary vasodilator, such as prostacycline, for 6 weeks before recatheterization.

Patients with a failing Fontan circulation are particularly difficult to evaluate. The calculation of the pulmonary vascular resistance and trans-pulmonary gradients is unreliable in this group. The main reason for this is that the pulmonary flow is low, due to the loss of hydrodynamic energy. This is made worse by the presence of microvascular sludging and thromboemboli, as well as the presence of arteriovenous malformations (Kawashima *et al*., 1994).

The patient considered suitable for transplantation is then screened for the presence of active infectious diseases. Immunological evaluation for donor–recipient match includes blood group typing and determining the presence and percentage of lymphocytotoxic reactive antibodies. The presence of reactive antibodies greater than 10% would ideally necessitate a lymphocytotoxic cross-match with a potential donor. This is particularly important in those who have had previous blood transfusions. Raised levels of lymphocytotoxic IgG class antibodies may lead to hyperacute rejection of the graft (Carey *et al*., 1998). In infants younger than 3 months, reactive antibodies are probably maternal and their significance is not clear. Due to the limited donor pool in the UK, we do not carry out a lymphocytotoxic cross-match prospectively. Instead, we cross-match after transplantation with donor lymph nodes. Despite this approach, we have not seen acute graft failure from pre-existing antibodies and have successfully transplanted patients with multiple previous transfusions, including ECMO-assisted children.

INFANT TRANSPLANTATION AND ABO MISMATCH

Blood group compatibility between donor and recipient has always been considered essential because of the risk of hyperacute rejection from antibodies to the blood group antigens of the donor. These pre-existing serum anti-A and anti-B antibodies are referred to as natural isohaemagglutinins. They would bind to the respective blood group antigens on the vascular endothelial cells of the donor organ and result in the activation of the complement cascade, leading to rapid thrombosis.

A clinical survey by Cooper (1990) identified eight cases of heart transplantation between ABO blood group-incompatible recipients and donors worldwide. Five of these suffered hyperacute rejection, of which two died and three were retransplanted. Three patients showed no hyperacute rejection and were alive 1 year later.

The problem of hyperacute rejection does not appear to apply to infants who have not yet produced sufficient levels of isohaemagglutinins. In fact, titres are usually low until the age of 12–14 months (Fong *et al*., 1974). This, together with the knowledge that the complement system is not fully developed in infants, and the problem of shortage of donor organs, encouraged the Toronto group to perform ABO mismatch transplantation in 10 infants (West *et al*., 2001). All 10 were very ill. Serum titres of isohaemagglutinins were measured before transplantation. The titres were also measured intraoperatively after performing plasma exchange on bypass, as well as after the transplantation. The bypass circuit was primed with group AB blood, unless the recipient was group O, in which case the prime could also be done with the blood of the donor's group. Thus, no antibody to the donor's or the recipient's blood group was introduced. Similarly, all plasma products used were of the AB group, or the donor's blood group if the recipient was group O. The results have been encouraging. There were no cases of hyperacute rejection. There were two deaths unrelated to rejection. There was no rapid production of isohaemagglutinins after transplantation. Two cases developed relatively high titres but with no evidence of rejection. Two infants aged 14 months had relatively high titres before transplantation but both did well with intraoperative plasma exchange. In addition, this strategy reduced their waiting list mortality from 58% to 7%. We ourselves have adopted the ABO mismatch transplantation for young children. All of our seven recipients are alive at follow-up. The average age of our patients is in excess of 1 year and the oldest was 2.5 years. This reflects the shortage of suitable donors for small children in the UK. Unlike the American centres, we do not offer ECMO bridging to transplant for children under 10 kg because of the shortage of donors. The highest titre of isohaemagglutinins we have accepted for transplantation is 1 in 16.

PRETRANSPLANTATION MANAGEMENT

Pretransplant care in paediatrics is usually shared with referring cardiologists if the child is well enough to be

an outpatient. At the transplant centre, a heart failure clinic with specialist nursing and cardiologist support is valuable for this vulnerable group of children. Medical therapy can be optimized, psychological support offered to the family and nutritional advice given. If necessary, biventricular pacing can be offered, although our experience is that this offers little to children who have deteriorated enough to be considered for transplantation.

In children awaiting transplantation, live vaccines, particularly BCG, are best avoided because of the potential for complications with immunosuppresssion. Some centres avoid all vaccines when a child is on the active list, for fear of stimulation of the immune system and possible early graft rejection. After transplantation all live vaccines are contraindicated, although it is likely that some attenuated live vaccines, such as MMR (measles, mumps and rubella) will be tolerated, as it has been given safely to paediatric AIDS patients in the USA.

SURGICAL TECHNIQUES

There are two standard operations for performing heart transplantation—orthotopic, in which the recipient heart is excised and replaced by the donor heart in the anatomical position, and heterotopic (the so-called ''piggyback transplant''), in which the donor heart is placed in the right side of the chest, alongside the recipient organ, and anastomosed so as to allow blood flow through either or both hearts (Cooper and Novitzky, 1991). Currently, the heterotopic technique is very rarely used.

Donor Considerations

After the diagnosis of brain death has been established in a potential donor, there are three main issues to consider before accepting organ donation. The first refers to the previous general health of the donor, the second to his/her cardiac function, and the third to whether the donor matches any of the recipients awaiting transplantation.

Relevant issues in the donor's previous health refer to anything that could jeopardize the recipient's health, especially illnesses that could possibly be transmitted, such as malignancy and infection. Examples include hepatitis B and C, HIV and cytomegalovirus (CMV) infections.

The function of the donor heart can be compromised due to the physiological changes during and after brainstem death (Smith, 2004). The haemodynamic data is obtained (where possible) through pulmonary artery catheterization and echocardiography. The ideal donor heart would produce excellent data (good cardiac output and blood pressure, with low filling pressures) in the absence of inotropic support. However, such ''perfect'' donor hearts are scarce. Consequently, the reluctance to accept less-than-perfect hearts would only be met with an increased mortality on the waiting list. Therefore, attempts have been made to improve the function of ''marginal'' hearts, by reversing the deleterious physiological effects of brainstem death, so that they can be accepted for transplantation (Smith, 2004). Cardiovascular support may include the use of a short-acting β-blocker, such as esmolol, to counteract the initial short-lived sympathetic overactivity. Adequate filling and an infusion of vasopressin may be needed in order to maintain blood pressure.

Respiratory support of the donor aims to maximize oxygen delivery to the transplantable organs. At the same time, barotrauma to the lungs, due to high tidal volumes and positive end-expiratory pressure (PEEP), are to be avoided, especially if the lungs are also being considered for transplantation.

Hormonal support may be deemed beneficial in the presence of otherwise unexplained cardiac dysfunction. When indicated, it consists of a bolus of methylprednisolone (15 mg/kg), triiodothyronine (4 g bolus followed by an infusion at a rate of 3 g/hour), vasopressin (1 U bolus followed by an infusion of 0.5–4 U/hour), and an infusion of insulin.

Although one would ideally aim for a close size–weight match, this would prohibitively restrict the supply of donor organs. In an effort to find suitable donors for more infants, a donor–recipient mismatch of up to 3:1 has generally been accepted, with good results (Fullerton *et al*., 1992).

Donor Heart Excision

A median sternotomy is performed, and the pericardium is opened longitudinally. The heart is inspected for external signs of major damage caused by trauma or external cardiac massage. The superior vena cava (SVC) and IVC are mobilized. The donor is fully heparinized (3 mg/kg). The SVC is doubly ligated and divided between the two ligatures. The IVC is clamped at the diaphragm, completing inflow occlusion. The ascending aorta is then cross-clamped at the level of the innominate artery and 10 ml/kg cold (4°C) cardioplegic solution (e.g. St Thomas' solution) is infused into the root of the aorta. Topical cooling is also used.

Division of the four pulmonary veins is completed. The IVC is divided immediately proximal to the clamp, decompressing the right side of the heart, and the inferior pulmonary vein is divided to decompress the left side. The aorta is divided high, and the pulmonary artery is divided across the major right and left branches. The apex of the heart is then lifted, and mediastinal tissue posterior to the atrium and major vessels is divided, allowing the heart to be removed from the pericardial cavity. It is then kept bathed in cold saline and transported to the receiving hospital.

Preparation of the Donor Heart

The preparation can be for either a right atrial connection or bicaval anastomosis. For the right atrial connection, the right atrial cavity is opened posterolaterally at the IVC orifice, continuing the incision into the base of the right atrial appendage. The tissue between the orifices of the four pulmonary veins on the posterior aspect of the left atrium is excised, leaving one large opening (**Figure 48.1**). The aorta and the pulmonary arteries should not be trimmed to ideal lengths until the atrial anastomoses have been completed during insertion. Simple defects, such as patent foramen ovale, should be repaired prior to implantation.

If the bicaval anastomosis technique is envisaged, the right atrial incision is omitted but the left atrial preparation is the same as that of the standard connection.

Recipient Operation

A median sternotomy is performed. After heparinization, cardiopulmonary bypass is initiated by venous drainage via cannulae in the SVC and IVC, and arterial return via an ascending aorta cannula at the level of the innominate artery. Body cooling to 28°C helps to prevent early rewarming of the donor heart. However, heart transplantation for certain complex congenital heart diseases may require periods of circulatory arrest in order to establish good exposure (such as aortic arch reconstruction), in which case further cooling is required.

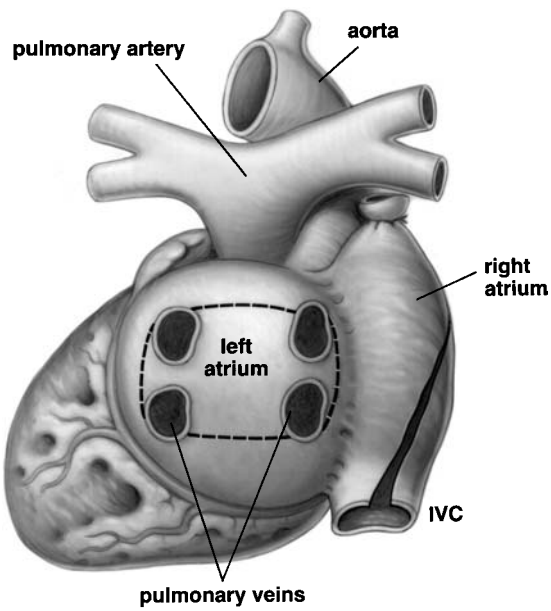

Figure 48.1

The aorta is cross-clamped. The heart is excised by dividing the right and left atrial walls close to the atrio-ventricular groove and the atrial septum, leaving a cuff of atrial wall to allow suture of the donor heart (**Figure 48.2**). Both atrial appendages should be excised to prevent subsequent thrombus formation (**Figure 48.3**). For the bicaval anastomosis technique, the SVC and IVC are transected with a rim of atrial tissue on each. The rest of the right atrium is excised. The aorta and the main pulmonary artery are divided as close to their respective valves as possible. Subsequently, these vessels may be trimmed before being sutured to their counterparts of the donor heart.

The donor heart is placed over the left side of the divided sternum, parallel to the remnant of the excised

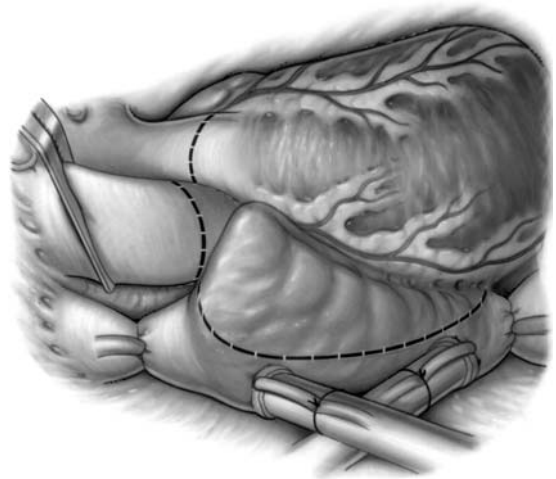

Figure 48.2

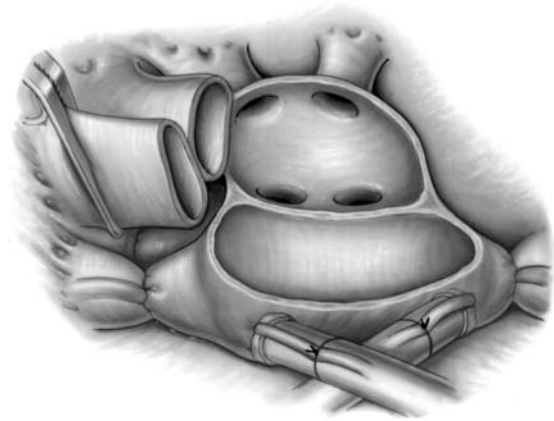

Figure 48.3

recipient heart. The donor heart is rotated to the left, so that its posterior surface faces anteromedially, such that the free walls of both recipient and donor left atria lie adjacent to each other. They are sutured to each other, using continuous 4/0 or 5/0 polypropylene, starting at the base of the left atrial appendage of the donor heart and at a point close to the caudal end of the recipient left superior pulmonary vein (**Figure 48.4**). At a convenient stage, the donor heart is drawn down into the pericardium and the suture tightened. The suturing is continued around the superior and inferior walls of the left atrium onto the atrial septum.

The right atria are anastomosed, starting at a convenient point in the posterior lip of the incision in the recipient right atrium. The anastomosis is continued first inferiorly because of the small cuff of recipient right atrium that remains in the region of the IVC (**Figure 48.5**). The superior part of the anastomosis is then completed.

For the bicaval anastomosis, each vena cava is anastomosed individually. This technique requires meticulous attention to avoid purse-stringing or kinking of the SVC. Because of this risk, right atrial anastomosis may be the preferred method of connection in small infants.

Aortic and pulmonary anastomoses are done in standard fashion. The left heart is meticulously de-aired and the cross-clamp is removed. Rewarming is started at this stage (**Figure 48.6**). The caval snares are released and the right heart is also de-aired.

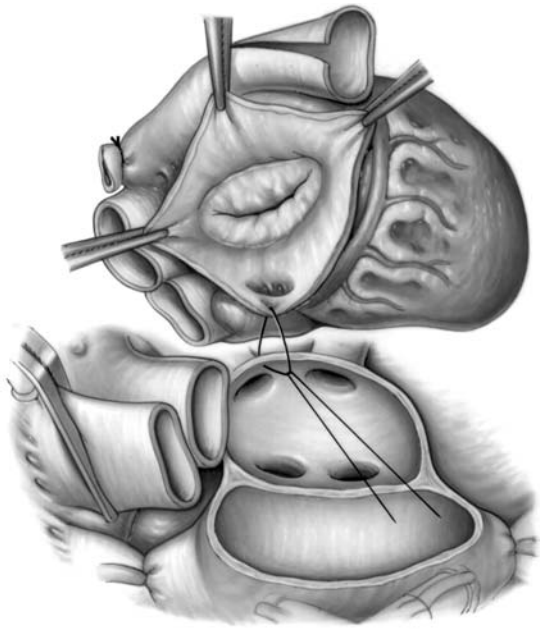

Figure 48.4

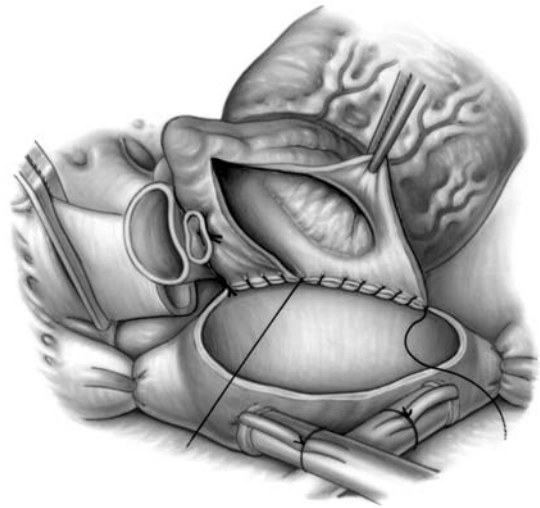

Figure 48.5

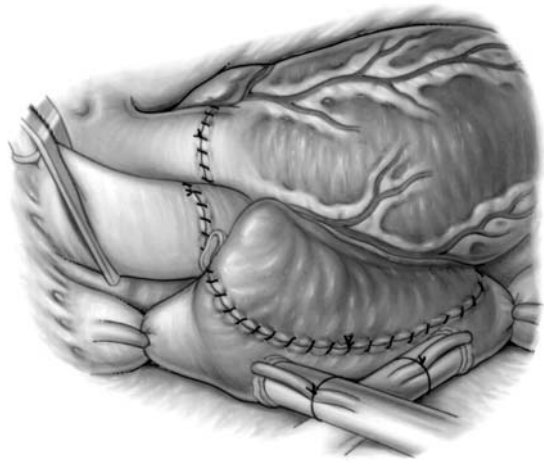

Figure 48.6

Monitoring lines are placed in the left atrium and the pulmonary artery. Right atrial and right ventricular pacing wires are inserted. When the myocardial function appears satisfactory, the patient can be weaned off the cardiopulmonary bypass. Haemostasis can be difficult. The cell-saver and aprotinin are routinely used.

TECHNICAL ASPECTS IN COMPLEX CONGENITAL HEART DEFECTS

Technical adjustments may be needed to deal with unusual cardiac morphology and the anatomical

distortions imposed by previous surgery (Allard *et al*., 1991; Chartrand, 1991; Menkis *et al*., 1991; Michler and Rose, 1991). For the assessment of the required technical adjustments, we recommend a sequential approach in the following order: pulmonary venous return; systemic venous return; atrial morphology; ascending aorta and aortic arch; main pulmonary artery and its two main branches; and spatial relationship between the aorta and the pulmonary artery.

Pulmonary Venous Return

Total anomalous pulmonary venous drainage is likely to have been corrected previously. In this case, the anastomosis can be taken down, and a new anastomosis constructed to the back of the donor left atrium. For partial anomalous pulmonary venous drainage, no special measure is necessary, other than making sure the atrial anastomosis incorporates the opening of the baffle into the left atrium. In the absence of previous corrective surgery, such a baffle can be constructed at the time of transplantation, using the atrial tissue of the donor heart.

Systemic Venous Return

The presence of a left-sided SVC is not usually a problem if there is a bridging (innominate) vein connecting it to the right-sided SVC. The left-sided SVC can then be ligated and divided below the bridging vein. The presence of a left-sided SVC with no bridging vein would require the harvesting of an extra length of donor SVC as well as the innominate vein. The proximal end of the left SVC of the recipient can be anastomosed to the innominate vein of the donor, while its distal end (connected to the left atrium or coronary sinus) can be ligated. This may not be suitable in the rare instances of interruption of the IVC with hemi-azygos continuation connecting to the left SVC. In this situation, the drainage of the left SVC into the right atrium should be preserved. If the left SVC drains into the coronary sinus and there is no bridging vein, the coronary sinus must be preserved and incorporated into the new (transplanted) right atrial chamber. If the coronary sinus is unroofed, it must be repaired with a patch. If the left SVC opens independently into the left atrium, a baffle can be constructed, connecting it to the right atrium via the atrial septum (**Figure 48.7**).

A short SVC due to previous cavopulmonary anastomosis/Fontan operation is managed by harvesting an extra length of SVC with the donor heart. The donor and recipient SVCs are then anastomosed to each other, and the recipient IVC is connected to the IVC of the donor right atrium.

Atrial Morphology

Situs solitus presents no difficulty. Situs inversus is more challenging (Doty *et al*., 1990; Vouhé *et al*., 1993). The

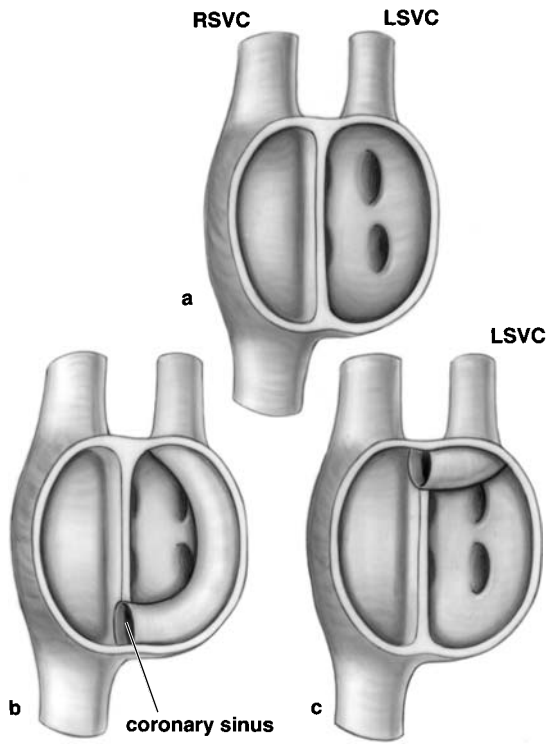

Figure 48.7

atria should be repositioned to allow standard transplantation with a situs solitus donor heart. Two techniques may be used: modified Senning or a true atrial transposition manoeuvre.

In the presence of situs ambiguous, the patient usually has a single atrium that receives all the systemic and pulmonary veins. This can be managed by using a prosthetic patch to partition the atrium into systemic and pulmonary atria (**Figure 48.8**). Standard orthotopic transplantation is then carried out.

Ascending Aorta and Aortic Arch

In hypoplastic left heart, the recipient's arch reconstruction can be done using one of the two techniques: the hemi-arch repair, or complete replacement of the arch. In the hemi-arch repair, a long spatulated anastomosis is performed, utilizing the donor aortic arch and descending aorta as an onlay patch to create continuity with the recipient aorta, as well as to repair any coarctation (**Figure 48.9**).

Complete arch replacement involves an anastomosis between the descending aorta of the donor and the recipient below the level of the ductus arteriosus. The head

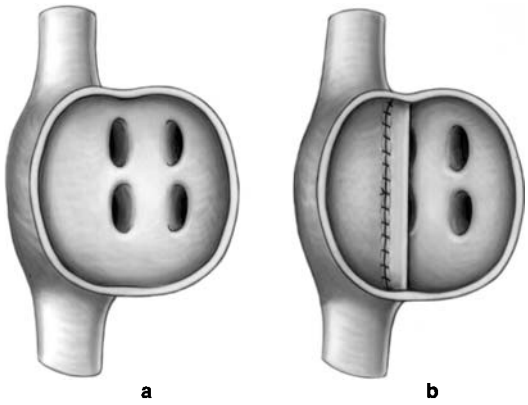

Figure 48.8

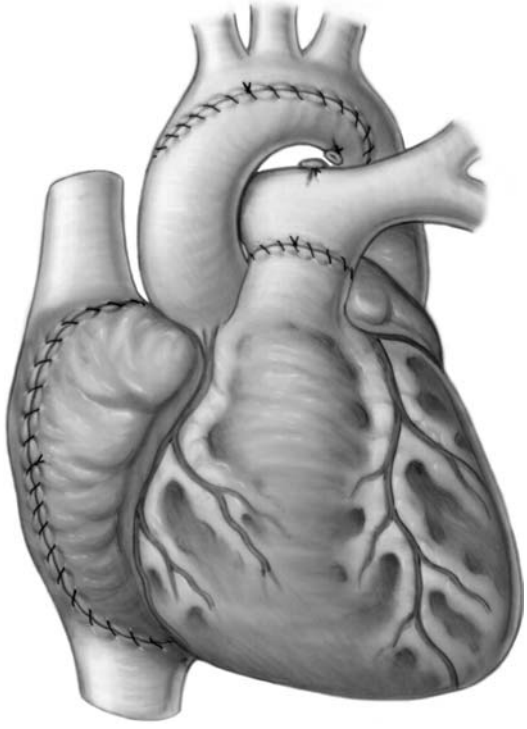

Figure 48.9

and neck vessels of the recipient are reimplanted onto the new arch as a Carrel patch. Complete arch replacement should be reserved for complex cases, not amenable to a hemi-arch repair.

Main Pulmonary Artery and Its Two Main Branches

The donor pulmonary artery can be harvested well beyond the bifurcation. This would allow direct anastomoses to the recipient's pulmonary arteries in cases where the main pulmonary artery is distorted by previous surgery.

Spatial Relationship between the Aorta and the Pulmonary Artery

Transposition of the great arteries may have been corrected previously, and the correction may involve the Lecompte manoeuvre. Two technical adjustments may be needed. First, if the recipient had previously undergone surgery that included a Lecompte manoeuvre, it would be useful to reverse this after the excision of the recipient heart. Second, it may be necessary to shift the main pulmonary artery sideways.

POSTOPERATIVE MANAGEMENT

Haemorrhage can be a major problem if the transplant involves complex reconstruction. Many of these patients may have coagulopathy secondary to prolonged bypass. The appropriate use of blood products, aprotinin, and possibly recombinant factor VIIa, may be helpful.

Haemodynamic stability is usually the norm; nevertheless, inotropic (adrenaline) and chronotropic (isoprenaline) support is usually required. A phosphodiesterase inhibitor (milrinone) is often used. Optimal heart rate can also be managed using external pacing. Pulmonary hypertension should be managed with nitric oxide. Occasionally, pulmonary hypertension may be sufficiently severe to lead to right ventricular dysfunction. In such circumstances, mechanical assistance of the right ventricle may be the only option.

Immunosuppression

The maintenance immunosuppressive regimes used in children traditionally include a calcineurin inhibitor, an antiproliferative agent and corticosteroids (triple therapy). The calcineurin inhibitors available are cyclosporine and tacrolimus. Cyclosporine is the most commonly used immunosuppressant in adult practice, but does have significant side-effects, such as hypertension, hirsutism, gingival hyperplasia and abnormal facial bone growth (Boucek and Boucek, 2002). These problems are difficult for children, and can cause noncompliance. These disfiguring problems can be avoided by the use of tacrolimus, which otherwise has a similar side-effect profile. In adult liver and kidney transplant patients, tacrolimus appears to be a significantly better immunosuppressant. As yet, this is unproven in heart transplantation, as the trials have had smaller numbers, but there is a trend towards significance. The ISHLT data shows that approximately 55% of

patients receive cyclosporine and 45% tacrolimus at 5-year follow-up. Patients treated with tacrolimus may be more rapidly weaned from corticosteroids (Armitage *et al*., 1993). However, tacrolimus may be associated with insulin-dependent diabetes (Wagner *et al*., 1997) and potentiation of lymphoproliferative disease (Younes *et al*., 2000).

The antiproliferative agents commonly used are azathioprine and mycophenolate mofetil. The latter shows a survival advantage (Eisen *et al*., 2005). Other antiproliferative agents have also been introduced, such as sirolimus (rapamycin). This may help to withdraw or reduce the dose of calcineurin inhibitors (Sindhi *et al*., 2001).

There is reticence about the long-term use of corticosteroids in children, because of their side-effects, especially the problems of growth retardation and inhibition of development. We use steroids in most cases, but aim to stop them prior to the biopsy at 6 months, unless there has been rejection of grade III/IV. Even when children have had rejection, we try to discontinue steroids after 1 year. Some children need steroids to be added when calcineurin inhibitors are stopped because of late renal dysfunction. The ISHLT data shows that approximately 60% paediatric patients are on prednisolone 1 year after transplantation. This is reduced to 40% at 5-year follow-up.

Antibody induction therapy remains controversial. It involves the use of monoclonal or polyclonal antithymocyte globulin preparations, the true value of which remains to be determined by randomized trials. However, the ISHLT data shows that the use of induction therapy is gradually increasing. Our recent approach has been to give basiliximab—a mouse monoclonal antibody that has been ''humanized'' by changing the variable region, so that it is not highly immunogenic. We have found this to be a safe therapy. It has been particularly useful in very sick children with compromised renal function, where calcineurin inhibitors can be delayed (Ford *et al*., 2005).

Surveillance and Treatment of Acute Rejection

It has been reported that rejection is seen in two-thirds of children in the first year following transplantation (Webber, 2003) but after 1 year acute rejection is rarer, with 82% being free of further episodes at 2 years and 73% at 3 years. However, with newer drug therapies the incidence of acute rejection is much lower. In our experience, symptomatic acute rejection is uncommon in the first year after transplantation. The strongest risk factor for rejection appears to be older age at transplantation (Pahl *et al*., 2001). Genetic risk factors have also been investigated, with particular emphasis on genetic polymorphism for cytokines and other genes involved in the immune response (Awad *et al*., 2001). Genetic polymorphism is also thought to exist with respect to response to immunosuppressive drugs (Zheng *et al*., 2002).

Rejection is diagnosed by histological examination of endomyocardial biopsies, although echocardiography as a diagnostic tool is evolving. The limiting factor in the use of echocardiography is the fact that it relies on the detection of ventricular dysfunction, which is a late event in the rejection process and is load-dependent. Histology, on the other hand, relies on the extent of myocardial lymphocyte infiltration and the presence of myocyte injury.

Rejection surveillance remains controversial (Boucek and Boucek, 2002). It is not clear whether biopsies should be scheduled routinely or dictated by symptoms (Mills *et al*., 1997). Our policy is to perform three biopsies in the first 6 months post-transplantation, with the initial biopsy being prior to discharge. We only perform further biopsies if there has been rejection above grade I or there have been clinical concerns. No routine biopsies are performed at annual review. Clinical assessment and echocardiography remain important tools as some children with symptomatic rejection have a normal biopsy (Pahl *et al*., 2001), but this has not been our experience.

The treatment of acute rejection would be 10–15 mg/kg/day methylprednisolone for 3 days. We would increase (or restart) oral steroids and change to myocophenolate if on azothiaprine. Resistant cases are very rare with tacrolimus/mycophenolate/steroids, but if there is haemodynamic compromise or poor ventricular function at any time, the early use of antithymoctye/lymphocyte globulin should be considered. In biopsy-negative rejection, plasmapheresis could be considered, as it reduces the high circulating levels of cytokines and immune complexes (Pahl *et al*., 2001).

Cardiac Allograft Vasculopathy

Cardiac allograft vasculopathy is the leading cause of death in the late survivors of heart transplantation. Unlike atherosclerosis, it consists of diffuse concentric myointimal proliferation. Generally, it does not cause angina, as the transplanted hearts are denervated, but it can occur in some patients. Instead, they may present with heart failure or even sudden death. It is common in the adult population, where it affects 30–50% of recipients at 5 years. In the paediatric population, 10–20% appear to be affected 5 years following transplantation. Risk factors are older recipient age, donor age above 30 years, and greater number of acute rejections in the first year (Webber, 2003). Bypass grafting or angioplasty is usually ineffective in view of the diffuse nature of the disease,

but some centres are encouraged by the use of drug-eluting stents. The best option for significant disease is probably retransplantation, which is limited by the availability of donor organs. Drug treatments are offering some hope of reducing the incidence of this problem. Both statins and mycophenolate have been encouraging in this respect (Eisen *et al*., 2005; Wenke *et al*., 2003). Sirolimus and everolimus have been particularly useful in both reducing the incidence and improving the outcome for those with established disease.

Infection

This risk is highest during the first 6 months after transplantation, when patients are most intensely immunosuppressed and steroid use is still common. Infections most commonly seen are wound infections, pneumonia, urinary tract infections and generalized sepsis. Causative organisms include *Staphylococcus aureus*, *Escherichia coli* and the *Pseudomonas* species. Later occurring serious infections are usually opportunistic infections such as Herpes zoster, *Pneumocystis carini* infection, and aspergillosis.

Viral infections, particularly Cytomegalovirus (CMV) pneumonia, may be associated with subsequent graft vascular disease. CMV-negative recipients who receive CMV-positive donor hearts may benefit from prophylactic treatment with gancyclovir. We undertake regular screening using quantitative polymerase chain reaction (PCR) for CMV in all transplants and will treat rising levels. CMV infections are treated with gancyclovir. These infections, although they may cause considerable morbidity, are now less often fatal.

Lymphoproliferative Disease and Tumours

Another major side-effect of immunosuppression is malignant disease, which, in children, is post-transplant lymphoproliferative disease (PTLD). This is caused by a clonal expansion of B cells infected with Epstein–Barr virus. The incidence of PTLD in the first year after transplantation is significantly higher compared with other solid organ recipients, and is higher in children than in adults (Smets *et al*., 2002). The incidence among paediatric heart transplant recipients in North America was found to be 4.7% (Webber, 2003).

Our recent approach has been to repeatedly quantify EBV load in peripheral blood by real-time PCR. Initial treatment of PTLD is to reduce immunosuppression. Clearly, the major side-effect of this treatment is graft rejection. Anti B cell (CD20) monoclonal antibody treatment (ritoximab) may be given. However, more complex chemotherapy may be required, in which case the mortality is high.

Other malignancies are uncommon in paediatric heart transplant recipients but are well recognized in adults. With increased survival, skin malignancies are likely to be a problem, as in renal transplant recipients.

OUTCOME

Survival

The ISHLT data shows an overall survival of 50% at 11–12 years. The conditional half-life of paediatric heart transplantation is 17.5 years for those aged 1–10 years, and 13.5 years for those aged 11–17 years. However, there is significant improvement by era, and the data from 1998 onwards appear to be very promising. Current survival by diagnosis and era in our experience is shown in **Figures 48.10** and **48.11**, respectively. The improved early survival is likely to be related to better surgical technique, particularly with the congenital heart disease recipients, improved paediatric intensive care and better organ preservation. The late survival is more influenced by improved immunosuppression. The causes of death include graft vascular disease, acute rejection, malignancy, infection, graft failure, technical reasons, renal failure, multi-organ failure and cerebrovascular complications.

Morbidity

The ISHLT registry shows that the functional status of the children who have undergone transplantation is good, with over 90% having no activity limitation at 5-years follow-up. In the first year following transplantation, 50% of recipients have not been hospitalized. From the second to the fifth year, over 70% have had no hospitalization.

Hypertension is less frequently seen in the absence of corticosteroids but other drugs, such as cyclosporine, contribute to it. Renal dysfunction is caused by cyclosporine and tacrolimus, although very few have had a renal transplant. Hyperlipidaemia and diabetes would require monitoring. The incidence of cardiac allograft vasculopathy amongst survivors is 2.5% at 1 year and 11.4% at 5 years, although it is significantly less common in the younger age group. This complication dramatically affects survival. Malignancy affects 1.9% of survivors at 1 year and 4.6% at 5 years. The majority of these are lymphomas. At our unit, we have observed an overall incidence of PTLD of 2%. Somatic growth can be affected by immunosuppressive drugs, particularly corticosteroids but possibly also calcineurin inhibitors (cyclosporine and tacrolimus) (Boucek and Boucek, 2002). Consequently, every effort is made to manage these patients without maintenance by corticosteroids.

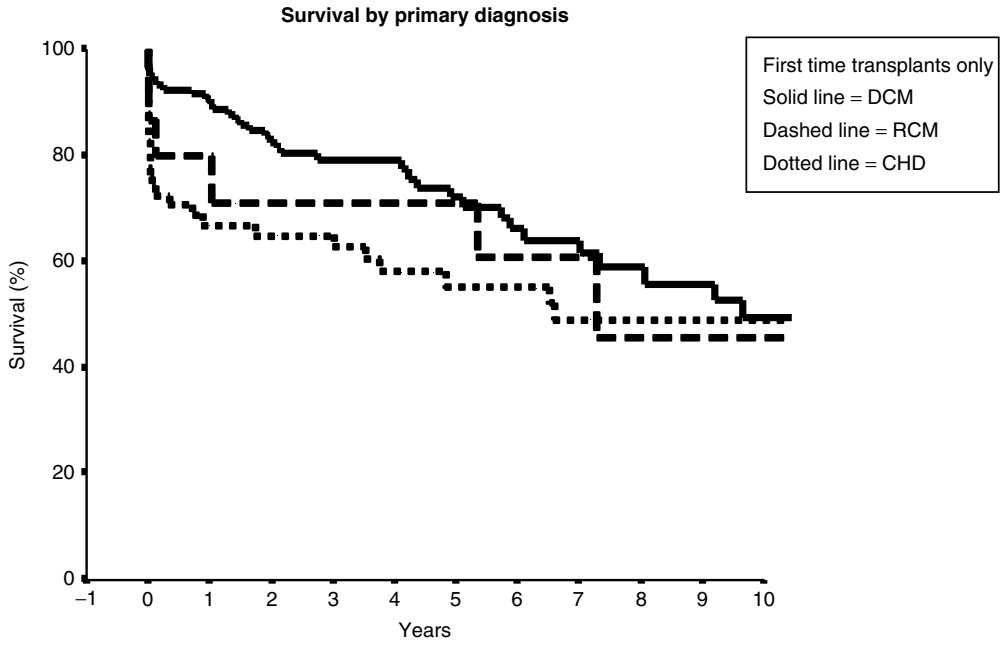

Figure 48.10

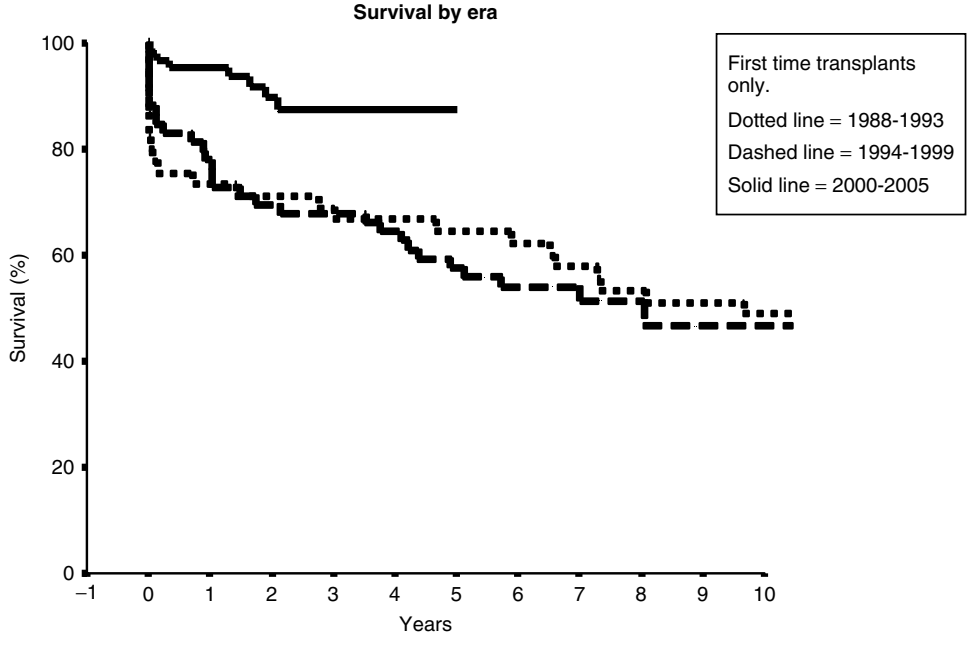

Figure 48.11

HEART–LUNG TRANSPLANTATION

Heart–lung transplantation is a recognized treatment for primary end-stage lung disease, and end-stage heart failure complicated with secondary pulmonary vascular disease.

Primary end-stage lung disease includes conditions such as cystic fibrosis and primary pulmonary hypertension. Although these patients do not suffer from heart disease and only need lung transplantation, it was felt, in the 1980s, that the outcome would be superior if heart–lung transplantation was performed. This was due to the fear of anastomotic complications at the site of bronchial anastomoses. Refinements in surgical technique have gradually shifted away from heart–lung transplantation in favour of double lung transplantation over the past 10 years. If heart–lung transplantation is performed, the explanted heart of the recipient can be offered to another patient awaiting cardiac transplantation (the domino procedure).

End-stage heart failure with the additional complication of pulmonary vascular disease precludes heart transplantation. For these patients, heart–lung transplantation is the only option. End-stage heart failure of any cause can result in pulmonary vascular disease, since the pulmonary vasculature is known to constrict in response to an elevated left atrial pressure. This includes all forms of cardiomyopathy (dilated, hypertrophic, restrictive, arrhythmogenic and other specific ones) as well as end-stage congenital heart disease. When congenital heart disease is the primary diagnosis, it is typically a defect with left-to-right shunt. The shunt diminishes as pulmonary hypertension develops. With time, the pulmonary hypertension increases in severity and becomes fixed, with reversal of the shunt and cyanosis (Eisenmenger syndrome).

The ISHLT data shows that paediatric heart–lung transplantation is performed in 9 centres worldwide. The largest age group is 11–17 years. The primary diagnosis is congenital heart disease, with Eisenmenger syndrome in 35% of cases, cystic fibrosis in 25%, primary pulmonary hypertension in 20% and retransplantation in 13%. Donor shortages of suitable heart–lung blocks have severely curtailed the number of transplants offered.

SELECTION OF PATIENTS

Three factors are considered: life expectancy of 2 years or less; poor quality of life; and no contraindications to transplantation.

Life expectancy is not easy to assess. A number of studies on cystic fibrosis have identified young age, female sex, low FEV-1 (30% or lower), low arterial saturation during the 12 minute walk test, high resting heart rate (age-adjusted), low plasma albumin concentration and low haemoglobin as the predictors of poor survival (Aurora, 2004). The prediction of life expectancy is even more difficult for patients with congenital heart disease and Eisenmenger syndrome. Therefore, the deteriorating quality of life and the need for additional hospital admissions and medication are the key indicators for determining acceptance to the waiting list, rather than predictions of survival.

Assessment of quality of life in the paediatric population is also difficult and inexact. Assessments are made according to the ability to partake in daily activities, exercise tolerance, hospital admissions and the requirement for oxygen therapy.

The contraindications to heart–lung transplantation are similar to those of heart. Patients with cystic fibrosis, however, merit further consideration, as they are usually colonized with pathogens. Most of these pathogens do not contraindicate transplantation, although patients with aspergillosis or multiresistant organisms, e.g. *Burkholderia cepacia* (LiPuma, 2001), represent a high-risk group and should probably not be transplanted.

DONORS

The quality of donor organs is directly linked to the subsequent success of any transplant operation. Probably fewer than 20% of donors who are suitable for cardiac transplantation are also suitable for heart–lung transplantation, because the lungs deteriorate rapidly in the brain-dead ventilated donor. The donor criteria for heart and lung donation include ABO compatibility, normal gas exchange, good pulmonary compliance, clear chest X-ray and no infection. A short ventilation period and appropriate size match of the donor and the recipient are also important. CMV compatibility is important to avoid primary CMV pneumonitis, which can cause morbidity and mortality.

Good donor care can be time-consuming and difficult. Bronchoscopy for bronchial toilet is required, and the correct ventilation to avoid atelectasis is important. Fluid control and physiological support of the circulation may prevent pulmonary oedema. Hormonal imbalances associated with brain death can be treated by the administration of tri-iodothyronine and insulin. These measures have been shown to be beneficial in stabilizing the brain-dead donor.

Donor Procurement

Removal of heart and lung for transplantation is performed through a median sternotomy. Both pleurae and the pericardium are widely opened. The heart and both lungs can then be inspected. The donor is heparinized. Cannulae are placed in the ascending aorta

and in the main pulmonary artery, in preparation for the administration of cardioplegia and pneumoplegia. Prostacyclin, a potent vasodilator, is flushed through the lungs via the pulmonary artery cannula. This is immediately followed by myocardial preservation, as described earlier in the chapter, except that the left heart is decompressed via the left atrial appendage rather than venting through a pulmonary vein. The lungs, flushed with cold (4°C) pneumoplegic solution, are vented through the left atrial appendage. Centres differ in their choice of pneumoplegic solutions, which can be extracellular (Ringer's solution) or intracellular (e.g. Euro-Collins solution). We use an extracellular solution, which, for an adult-sized donor, consists of 700 ml Ringer's solution, 100 ml 20% human albumin solution, 200 ml 20% mannitol, 10 000 U heparin and 400 ml donor's blood anticoagulated with citrate phosphate dextrose. In smaller donors, these volumes are proportionately reduced.

The heart–lung block is excised in its entirety, with the lungs partially inflated and the trachea divided high. Total ischaemic time should be as short as possible, generally less than 4 hours. Good early function is the norm for good-quality organs carefully dissected and properly protected during the ischaemic period.

RECIPIENT OPERATION

Combined heart and lung transplantation may be performed through either a median sternotomy or a bilateral throacosternotomy (clamshell incision). The latter provides excellent exposure and is valuable when adhesions may be difficult to mobilize, such as in patients who have had previous thoracotomies. The exposure offered by the clamshell approach is also useful in the presence of large bronchopulmonary and aortopulmonary vascular collaterals, which will need to be dealt with as early as possible in the procedure and certainly before the heart is excised on bypass, as the whole blood volume can be lost through these vessels after the heart is excised. These possible technical difficulties highlight the vital importance of good timing and coordination between the recipient and the donor teams, in order to avoid an unnecessarily prolonged ischaemic time. As much dissection as possible should take place before heparinization, provided that manipulation of the organs does not cause ventilatory or haemodynamic instability.

Explantation

Bypass is usually established with routine aortic and bicaval cannulation. If the heart is being excised for subsequent transplantation (the domino procedure), or there are other anatomical considerations, venous cannulation can be modified. For example, innominate vein and low IVC cannulation may be useful, and separate left SVC cannulation may be necessary. Standard bypass is used for heart–lung transplantation.

The heart is excised first (**Figure 48.12**); the left lung is mobilized by division of the pulmonary ligament and dissection of the hila around the pulmonary artery and the pulmonary veins. A small cuff of pulmonary artery is left at the site of the duct to protect the recurrent laryngeal nerve (**Figure 48.13**), and the phrenic nerve is protected

Figure 48.12

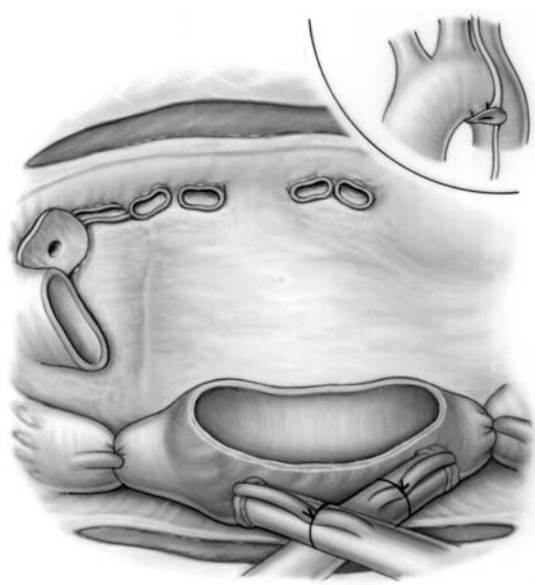

Figure 48.13

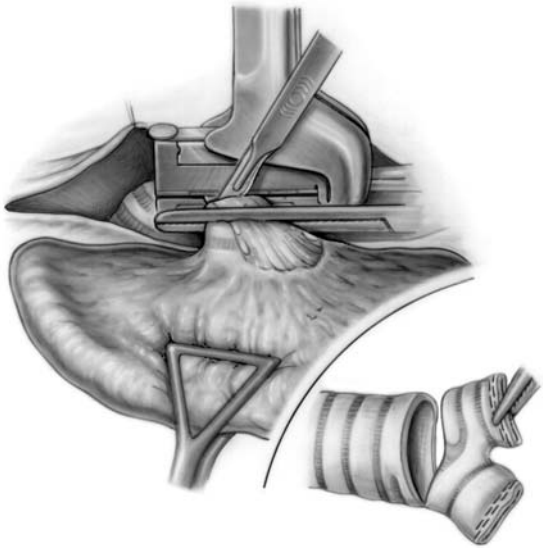

Figure 48.14

on a wide pericardial pedicle. The vagus nerve, which comes high up into the hilum, is dissected backwards into the mediastinum before the left lung is removed by clamping the bronchus and dividing it (**Figure 48.14**).

The right lung is similarly mobilized. The right phrenic nerve runs directly on the SVC and pulmonary veins and is protected by careful dissection of the hilum. The vagus nerve is swept back into the mediastinal tissue. After the right bronchus has been clamped and the lung removed, mediastinal dissection can be completed. It is worth taking some time at this stage to identify and clip or cauterize all posterior mediastinal bleeding vessels. The bifurcation of the trachea is mobilized and the vagus nerve is again swept back into the mediastinum. The recipient trachea is divided just above the carina, and at this stage the thoracic cavity can be washed with a weak antiseptic solution. Often, the removal of the recipient's diseased organs is the most difficult and time-consuming part of the heart–lung transplant procedure.

Implantation

The donor organs are placed into the chest of the recipient (**Figure 48.15**). The donor trachea is cut one ring above the bifurcation, and the tracheal anastomosis (**Figure 48.16**) is performed using continuous absorbable PDS or, in large children and adults, continuous polypropylene sutures. The donor trachea is usually invaginated into the recipient and, when the tracheal anastomosis has been completed, the pretracheal fasciae from the donor and recipient are apposed.

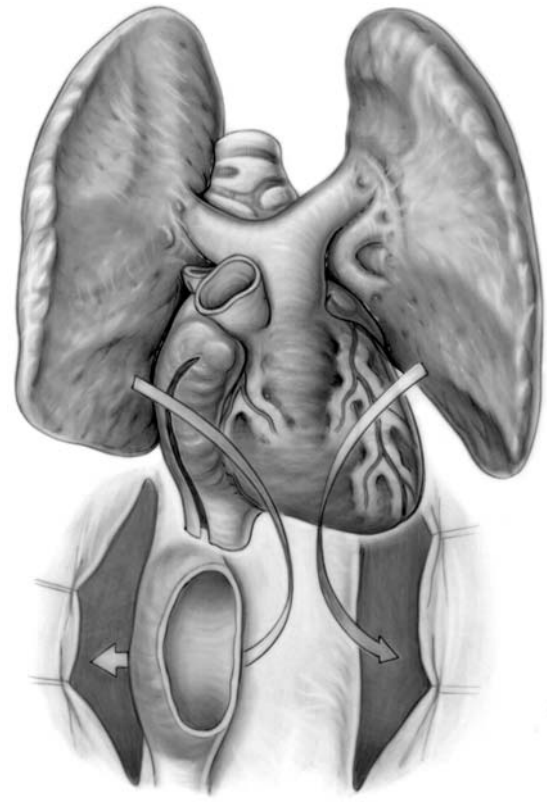

Figure 48.15

For heart–lung transplantation, even in complex congenital heart disease, only aortic and systemic venous connections are left *in situ*. This leaves enough room for an adequate conduit to be made for connecting the donor heart to the recipient venous system, regardless of the pattern of systemic atrial connections. This is usually right atrium-to-right atrium (**Figure 48.17**), but can be separate caval connections. Even a large left SVC can be swung under the aorta and connected to the SVC of the donor organs.

After all the anastomoses have been completed, the organs are reperfused and de-aired and vent sites closed. Usually, spontaneous sinus rhythm occurs and bypass is discontinued without difficulty.

Two mediastinal and two pleural chest drains are inserted, the latter reaching to the apex and lying posterolaterally within the chest. Management of these drains in conjunction with appropriate ventilation of the patient is important. If the size of the donor lungs matches the recipient's lung size, no special problems are encountered and usually the lungs can be ventilated with 5 cmH_2O positive end-expiratory pressure.

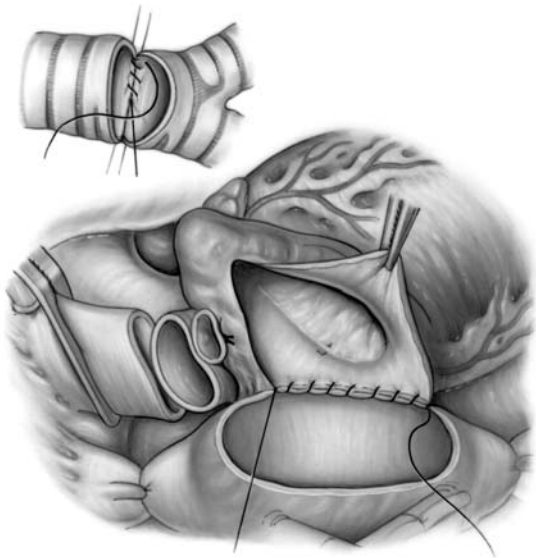

Figure 48.16

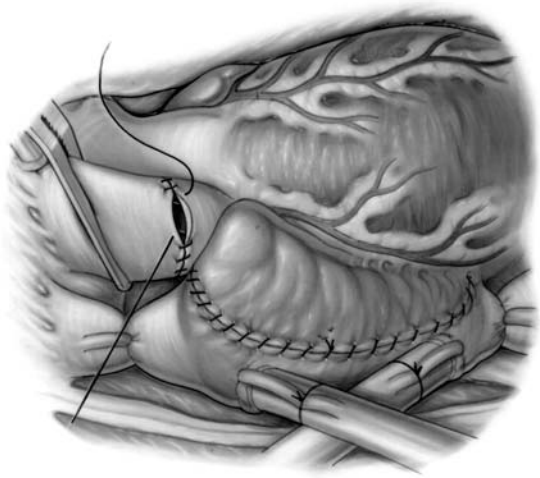

Figure 48.17

If the lungs are too large, some atelectasis is expected after extubation. When the lungs are too small for the chest cavity, all will be well until the chest is closed and suction is applied to the drains. Then the lungs will be at full lung capacity and the intrathoracic pressure and ventilatory pressure may rise precipitously and can cause tamponade, with a dramatic and potentially fatal drop in cardiac output. If this is not recognized as a problem of lung size, inappropriate resuscitation with high-dose inotropic drugs and increased ventilation may be instituted. The problem is easily resolved by stopping the suction applied to the drains and ventilating the patient, using low tidal volume with a rapid rate. After extubation, the problem resolves spontaneously as a result of the different dynamics of self-ventilation. Pneumothorax and air leaks may persist for some days and the drains should be left *in situ* until the lungs are fully expanded and all air leakage has stopped.

Administration of aprotinin (Trasylol) has reduced postoperative bleeding in heart–lung transplantation. This is particularly useful in patients with pleural and mediastinal adhesions. The use of recombinant factor VIIa is another potentially powerful tool in the management of refractory bleeding (Hedner and Erhardtsen, 2002). However, its efficacy and safety in cardiac surgical patients are not yet proven, and its dosage and indications for use remain to be defined.

POSTOPERATIVE MANAGEMENT

The majority of patients are extubated within 24 hours following heart–lung transplantation and a regime similar to that for heart transplantation is followed, including the immunosuppression regimen. We use intravenous tacrolimus for lung recipients, rather than oral (as in the hearts), until enteral feeds are established. Also, higher levels of tacrolimus are used at follow-up. We also use more CMV and *Aspergillus* prophylaxis in the lung recipients. In addition, most of our lung and heart–lung recipients will have a fundoplication in the first year following transplantation because of our concern about chronic silent aspiration. The pattern of other post-transplant complications is very similar to that of heart transplantation, including infection, rejection, tumour occurrence, effects on growth and other side-effects of immunosuppression. However, there are a few additional issues that must be mentioned.

Diagnosis of Acute Rejection

Rejection of the lungs in heart–lung transplantation occurs most frequently in the first 3 months following transplantation. Although patients can be treated on a clinical and radiological suspicion of rejection, in the absence of any infection, trans-bronchial biopsy confirms the diagnosis. Lung function tests, particularly the forced expiratory volume in 1 second (FEV-1), are non-specific for rejection, but a fall in FEV-1 in an otherwise well patient indicates that further investigations should be performed. Trans-bronchial biopsy is easily done in adolescents and adults through a flexible bronchoscope, but in children, it may necessitate a general anaesthetic and rigid bronchoscopy with subsequent passage of a flexible bronchoscope, to obtain adequate biopsy

specimens. The trans-bronchial biopsy is useful not only in confirming the rejection but also in isolating opportunistic pathogens and assessing the presence of infection and rejection, which may occur at the same time. We treat rejection episodes with pulsed intravenous steroids, and the dose of tacrolimus is increased. Usually, azothiaprine is changed to mycophenolate mofetil. Severe rejection is treated with antithymocyte globulin.

Infection

Most considerations of infection with regard to heart transplantation remain pertinent for the heart–lung transplant patient. While sputum culture and viral serological study results are useful guides to pulmonary infection, these can be misleading in the immunosuppressed host. Broncho-alveolar lavage and trans-bronchial biopsy allows culture of both lavage fluid and lung tissues. Cystic fibrosis patients who have received a heart–lung transplant may have upper airways that have been chronically infected with pathogens, particularly *Pseudomonas*. Antibiotic therapy is guided by the culture and sensitivity findings of the specimens obtained at surgery from the recipient and the donor. Antibiotics are continued for 7–10 days. All patients who are seropositive for CMV or Herpes simplex virus are given prophylactically acyclovir daily. CMV infections are treated with intravenous gancyclovir. Patients receiving steroids are given co-trimoxazole for *P. carini* pneumonia prophylaxis.

OUTCOME

The ISHLT data show an overall survival of 50% at 3 years (Trulock *et al*., 2004). The conditional survival, however, is 50% at 6–8 years. Patients with Eisenmenger syndrome or cystic fibrosis do not seem to differ from the main statistical group. The leading cause of death varies depending on the time period since transplantation. In the first month after transplantation, acute graft failure and infection are the leading causes of death. From 30 days up to 1 year, it is infection and beyond 1 year, bronchiolitis obliterans becomes the leading cause of death, although infection and graft failure also contribute significantly to the morbidity and mortality.

Chronic Rejection

Bronchiolitis obliterans is the manifestation of chronic rejection. It is the most important long-term determinant of the outcome of transplantation. The disease is characterized by progressive loss of small airways and diminishing lung function. It is clinically manifested by wheezing and increasing shortness of breath on exercise. Formal testing shows a decline in respiratory function, especially the FEV-1. The diagnosis is confirmed by histological findings of lung biopsy. There is no effective cure, although the avoidance of acute rejection and the prevention of CMV infections would be helpful, since these are recognized risk factors. Retransplantation is also a possibility, but donor organs are scarce, and the results of retransplantation are not as good as those of the initial organ transplantation.

THE FUTURE

Many obstacles to transplantation have been overcome in the last two decades. However, organ availability remains a major limitation. The shortage of donor organs will be best tackled by the education of the public and by improvements in the management of potential donors and organ preservation techniques. This shortage may also be improved, to some extent, by the more widespread practice of ABO mismatched transplantation. This would benefit infants, but the upper age limit for ABO mismatched transplantation has not yet been determined. Xenotransplantation has not materialized. Replacing organs by artificial means is also far from reality, and the use of mechanical support as a bridge to transplantation in children is limited.

Increasing understanding of the immune system and pharmacological development will lead to better management of immunosuppression. It should also improve the management of rejection, especially chronic rejection. This increasing knowledge will hopefully one day make the induction of tolerance a reality.

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49

Paediatric Lung Transplantation

J. J. Nigro, R. D. Bart and V. A. Starnes

Lung transplantation has evolved to become an effective and reliable treatment for (non-malignant) end-stage lung disease in children (Starnes *et al*., 1996, 1997, 1999, 2004; Huddleston *et al*., 1999, 2002; Sweet, 2003; Boucek *et al*., 2004). Paediatric lung transplant information is compiled by the International Society of Heart Lung Transplantation (ISHLT), which collects data from voluntarily reporting centres. The following information and figures are based on data made available by ISHLT (Boucek *et al*., 2004). Paediatric lung transplantation remains rare (**Figure 49.1**) and is performed in a limited number of highly specialized centres (**Figure 49.2**) (Starnes *et al*., 1996, 1997, 1999, 2004; Huddleston *et al*., 1999, 2002; Sweet, 2003; Boucek *et al*., 2004). Most transplants occur in the adolescent age group (**Figure 49.3**) and are due to cystic fibrosis or pulmonary hypertension. A small number of infants and pre-teen children undergo lung transplantation and these patients are likely to have pulmonary failure due to congenital heart disease (**Figure 49.4**). Paediatric lung transplantation is a very effective palliation for end-stage lung disease and provides the vast majority of living patients with an excellent functional classification (**Figure 49.5**). Survival is related to pretransplant diagnosis and overall is 70–80% at 1 year and 50% at 5 years (**Figure 49.6**) (Starnes *et al*., 1996, 1997, 1999, 2004; Huddleston *et al*., 1999, 2002; Sweet, 2003; Boucek *et al*., 2004). Bronchiolitis obliterans ultimately affects the majority of survivors, limits graft function and remains a significant impediment to long-term survival (**Figure 49.7**).

INDICATIONS

The primary indication for paediatric lung transplantation is the presence of progressive pulmonary insufficiency, likely resulting in death shortly after the expected waiting time for donor lung availability (approximately 2 years). Most paediatric lung transplantations occur in the adolescent age group, and are performed for end-stage lung disease due to cystic fibrosis, pulmonary hypertension or congenital heart disease (Boucek *et al*., 2004). Although the traditional criterion for referral of a cystic fibrosis patient is a forced expiratory volume in 1 second (FEV_1) of less than 30% of the predicted value, the Cystic Fibrosis Consensus Conference established more detailed guidelines for referral to lung transplantation (**Table 49.1**) (Yankaskas and Mallory, 1998; Liou *et al*., 1999, 2002a, 2002b; Yankaskas *et al*., 2004). A statistical model has recently been developed, which includes age, gender, predicted ideal weight for age, pancreatic sufficiency, presence of diabetes, FEV_1, *Staphylococcus aureus* colonization status, *Burkholderia cepacia* status and the number of acute (respiratory) exacerbations within 1 calendar year, to provide a more accurate way to select (cystic fibrosis) patients for lung transplantation (Liou *et al*., 1999, 2002a, 2002b).

Indications (for lung transplantation) for pulmonary hypertension include: progressive exercise intolerance,

Table 49.1 Criteria/guidelines for patients with cystic fibrosis who are likely to benefit with lung transplantation (Yankaskas and Mallory, 1998; Liou *et al*., 1999, 2002a, 2002b; Yankaskas *et al*., 2004).

1. Progressive pulmonary function impairment manifested by $FEV_1 < 30\%$ predicted, severe hypoxaemia (< 7.3 kPa or 55 mmHg), and hypercapnia (> 6.7 kPa or 50 mmHg)
2. Increasing functional impairment, evidenced by increasing frequency and duration of inpatient treatment for pulmonary decompensations
3. Significant haemoptysis or other life-threatening pulmonary complication
4. Progressive bacterial resistance to antibiotic therapy

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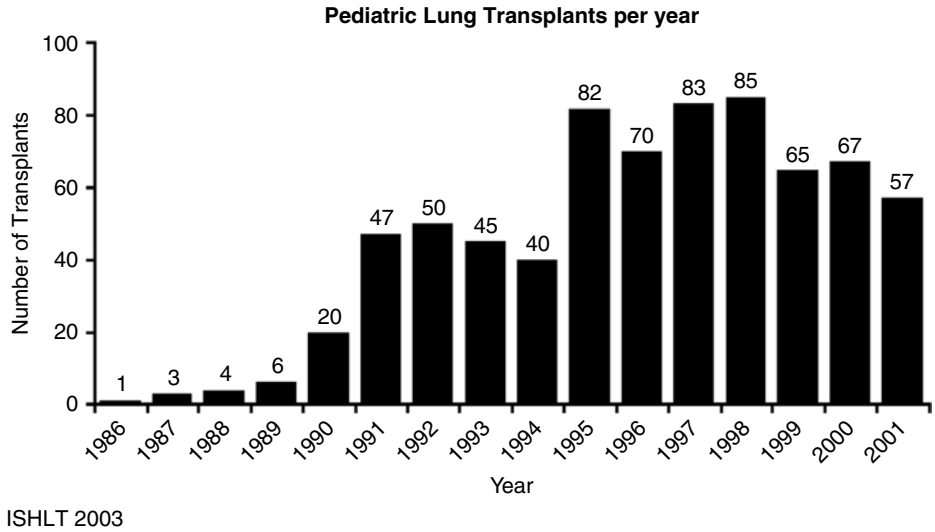

Figure 49.1

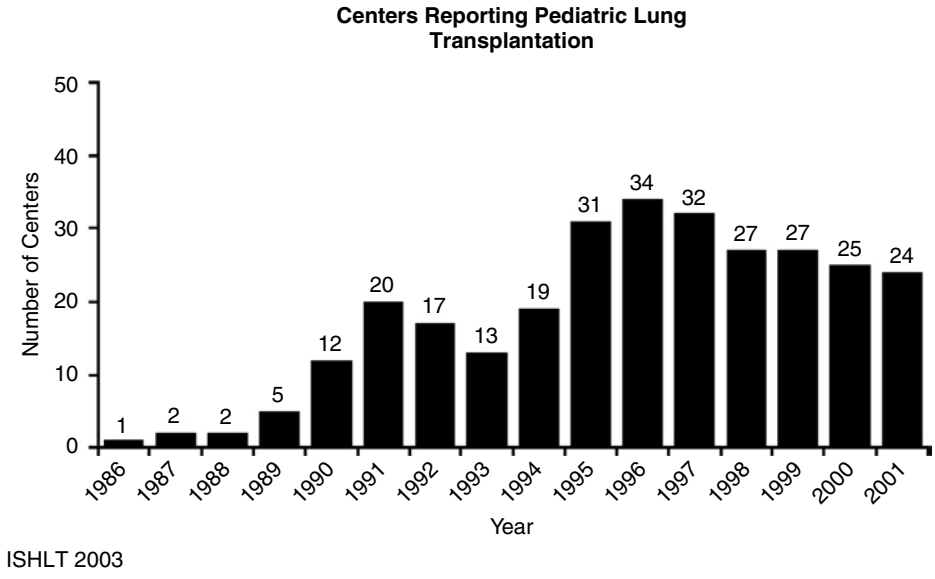

Figure 49.2

syncope, haemoptysis, angina pectoris or significant right ventricular failure (Spray, 1998). Haemodynamics generally associated with these symptoms are a right atrial pressure of greater than 8 mmHg with a decreased cardiac index and a pulmonary vascular resistance of greater than 20 Woods units/m^2 (Spray, 1998). A product of mean right atrial pressure and pulmonary vascular resistance index greater than 360 correlates with limited survival and suggests the utility of lung transplantation (Spray, 1998). Pulmonary vasodilators, including prostacyclin, sildenafil and bosantin, can be used to stabilize patients while awaiting donor organs.

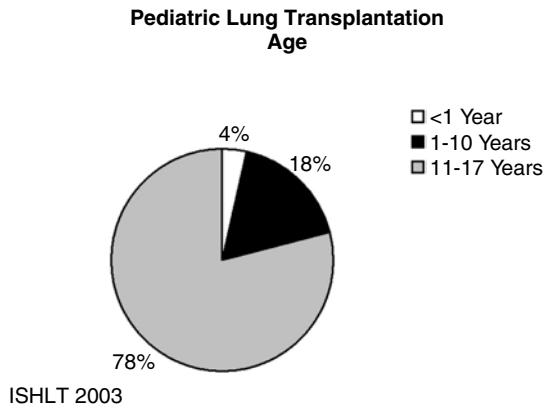

Figure 49.3

Indications for lung transplantation in patients with end-stage lung disease due to congenital heart disease and Eisenmenger's syndrome include the onset of severe polycythaemia, with haemoptysis, right ventricular failure and progressive exercise intolerance. Patients with other rare aetiologies of end-stage lung failure, including pulmonary fibrosis, pulmonary veno-occlusive disease, alveolar capillary dysplasia, surfactant protein B deficiency, pulmonary arteriovenous malformations and idiopathic pulmonary haemosiderosis, are evaluated on an individual basis and referred for transplantation when the expected survival without transplantation is 2 years or there is a significant impact on quality of life.

CONTRAINDICATIONS

The presence of severe irreversible injury to any other organ system or of a non-curable or active malignancy within the last 2 years are contraindications to lung transplantation (Starnes *et al*., 1992, 1994, 1996, 1997, 2004; Spray, 1994; Spray *et al*., 1994; Bridges *et al*., 1995; Koutlas *et al*., 1997; Woo *et al*., 1998; Mendeloff *et al*., 1998, 2002; Huddleston *et al*., 1999, 2002; Sweet, 2003). Infections including HIV, tuberculosis, viral hepatitis (B or C) and colonization with completely resistant bacteria, such as multiresistant *Pseudomonas*, are also contraindications. Mechanical factors, including the presence of a shifted mediastinum due to prior pneumonectomy, severe chest wall deformity (severe kyphoscoliosis) or the presence of extensive chest wall-to-lung collaterals complicate lung transplantation and are relative contraindications. Other factors which complicate lung transplantation, requiring intervention prior to transplantation and which may be contraindications, include severe malnutrition, high-dose steroid dependence, uncontrolled collagen vascular disease, severe musculoskeletal disease, prolonged mechanical ventilation, fungal (*Aspergillus*) and atypical mycobacterial colonization. A psychosocial evaluation prior to transplantation ensures the greatest likelihood of optimal utilization of scarce resources. Presence of a major psychoaffective disorder, poor compliance, inability to follow a complex medical regime, and limited access to follow-up care (financial or geographic) are factors that require pretransplant interventions and may become relative contraindications (Durst *et al*., 2001).

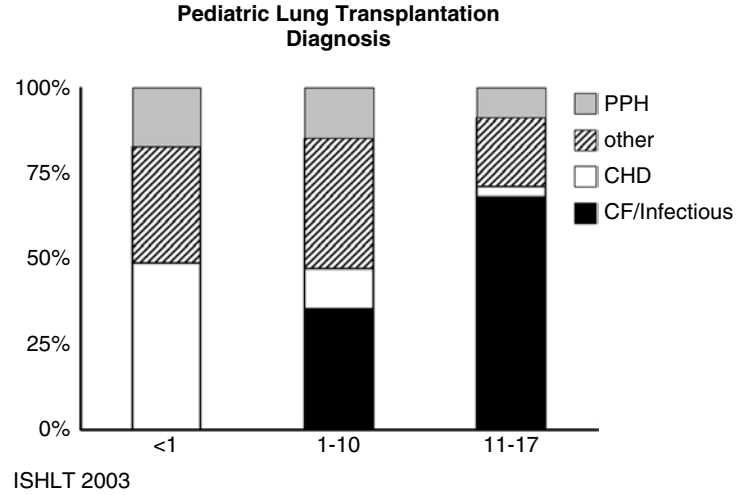

Figure 49.4

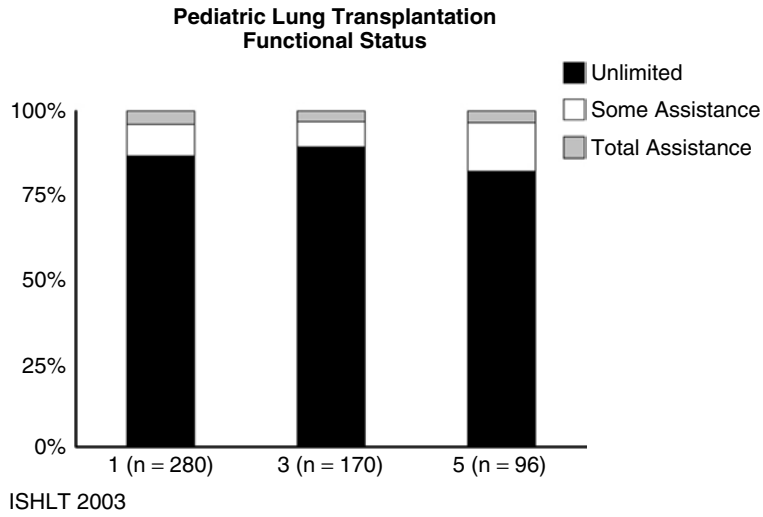

Figure 49.5

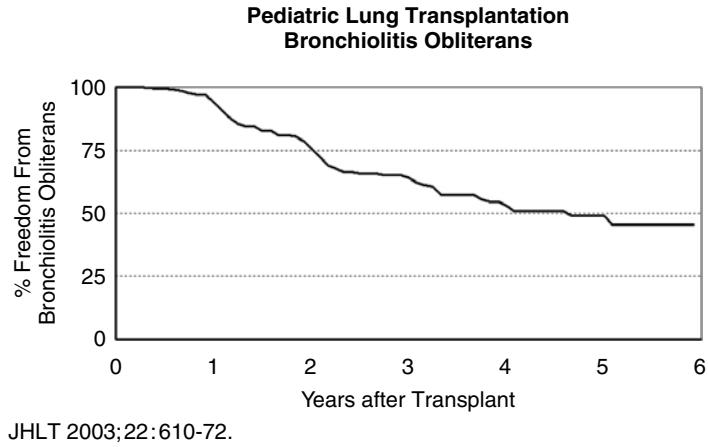

Figure 49.6

Mechanical pleurodesis and prior thoracic operations are no longer strict contraindications to lung transplantation.

PSYCHOSOCIAL EVALUATION

Medically appropriate candidates and their families undergo a formal psychosocial assessment to ensure that scarce donor organs are optimally utilized to obtain successful outcomes. This comprehensive evaluation includes an assessment of social and financial factors that are predictive of poor outcomes (**Table 49.2**) (Durst *et al*., 2001). Interventions are rendered to correct identified problems, and patients who are refractory to these interventions will not benefit from lung transplantation.

TYPES OF LUNG TRANSPLANTS

Heart–Lung Transplant

In this procedure the combined heart and lung are harvested from the donor and transplanted into the recipient *en bloc*. This is accomplished by removal of the diseased heart and lungs with careful preservation of the phrenic nerves. The donor organs are implanted with a single tracheal anastomosis and caval and great vessel anastomoses. Prior to successful (isolated) lung

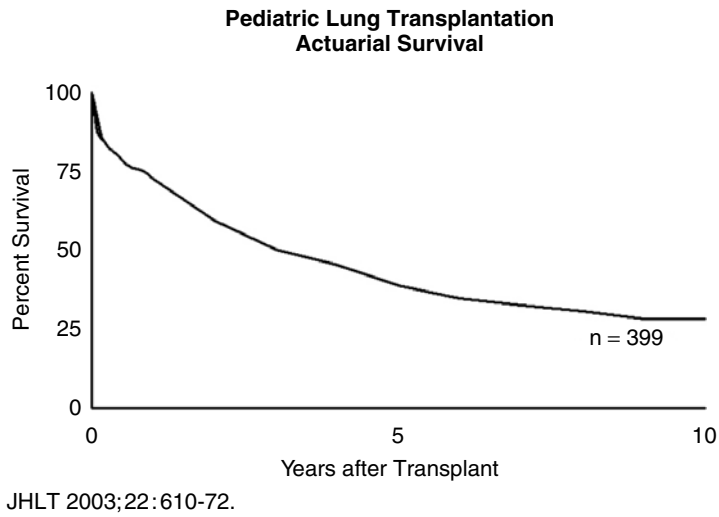

Figure 49.7

Table 49.2 Psychosocial considerations for successful lung transplantation. Reprinted with permission from Durst, C.L., Horn, M.V., MacLaughlin, E.F., *et al*., 'Psychosocial responses of adolescent cystic fibrosis patients to lung transportation.' Pediatric Transplantation. Blackwell Publishing.

Demonstrated non-compliance
Psychiatric disturbance
Lack of commitment to the transplant process
Lack of financial resources
Dysfunctional relationships
Poor or dysfunctional support systems
Inability to follow instructions or to understand transplantation
Poor problem-coping skills
Poor home environment
Inability to communicate or work with the transplant team

transplantation, heart–lung block transplantation was the only way to treat end-stage pulmonary disease. With the limited number of donor organs and the success of isolated lung transplants, this procedure (heart–lung transplant) is reserved for patients with combined unrepairable end-stage heart and lung disease. Lung transplantation is the focus of this chapter; a more detailed description of heart–lung transplantation is given in Chapter 48.

Bilateral Lung Transplant

Bilateral sequential lung transplantation is the most common type of lung transplant performed for children. Both lungs must be replaced in the presence of suppurative lung disease, such as cystic fibrosis, to protect the implanted graft from infection by the contralateral infected native lung (Spray *et al*., 1992, 1994; Spray, 1994; Yankaskas and Mallory, 1998; Mendeloff *et al*., 1998, 2002; Huddleson *et al*., 2002; Sweet, 2003; Starnes *et al*., 2004). For pulmonary hypertension, maximal relief of pulmonary artery pressures with better long-term pulmonary function and outcomes are obtained if both lungs are transplanted.

Single Lung Transplant

Single lung transplantation is rarely utilized for children and is considered in the unusual patient with a significant contraindication to entering one of the pleural spaces (Spray, 1998). Bilateral lung transplantation offers the advantages of complete removal of suppurative lung disease, symmetrical chest growth and improved long-term outcomes for pulmonary hypertension relative to single lung transplants (Spray, 1998).

Living Lobar Lung Transplant

The scarcity of donor (cadaveric) lungs and the precarious state of patients with end-stage lung disease provided the impetus to develop living related lobar lung transplantation. In this elective procedure, a right lower lobe is harvested from one living donor and a left lower

lobe is harvested from a second; the two lobes (grafts) are subsequently implanted in the recipient as right and left (respectively) lung transplants. This procedure effectively expands the donor pool, but is available at a limited number of transplant centres. Although this procedure provides some predictability for transplant candidates, it is not a procedure that should be applied as an emergency or last-minute referral. This procedure requires careful identification of donors and puts two additional patients (donors) at potential risk. The results at our centre have been excellent, with a lower incidence of bronchiolitis obliterans relative to matched cadaveric transplants (Starnes *et al*., 1996, 1999, 2004; Woo *et al*., 2001).

DONOR SELECTION

Quality donor lungs are quite scarce compared to other organs because of aspiration, pulmonary contusion and pulmonary oedema. General guidelines for the selection of donor lungs include an arterial oxygen partial pressure (PaO_2) greater than 100 mmHg on 40% inspired oxygen with a positive end-expiratory pressure of 5 cmH_2O, a clear chest roentgenogram, absence of lung injury due to aspiration or trauma, absence of a history of significant primary pulmonary disease or malignancy, and ABO compatibility (Wagner and Reichenspurner, 2002). HIV infection and other intractable communicable diseases are contraindications to organ donation. Size matching of donor and recipient is critical for successful paediatric lung transplantation. For heart–lung blocks, sizing is based on donor and recipient weights, which should be within 20–30% of each other (Spray, 1998). Sizing for isolated cadaveric lung transplants correlates best with height and age, and patients are listed for donors who are 4 inches above or below the height of the recipient (Spray, 1998). Sizing for adolescents is generally less sensitive and acceptable donors are usually less than 35 years old (Spray, 1998). Elevated panel reactive antibodies should be treated in the donor and a prospective cross-match is indicated for donor matching.

Donor selection for living lobar lung transplantation is more complex, because it involves the selection of two donors who have adequate lung function and are the appropriate size to tolerate donation of a quality lobe. Donors have traditionally been family members; others are considered but they should have a long-standing emotional attachment to the recipient. Guidelines for the selection of living lobar donors are presented in **Table 49.3** (Woo *et al*., 1998).

Optimal size matching between donor and recipient is difficult, but it is currently accomplished based on estimated lung volumes, determined by computed tomography (CT), spirometry and quantitative ventilation/perfusion scans. Once two donors are identified, right and left lobar donors are designated; the larger donor is assigned as the right lung donor. If donors are of equal size, the one with the most complete fissure on the left is designated to donate that side.

Table 49.3 Guidelines for the selection of living lobar lung donors.

No coercion (emotional or financial)
ABO compatibility
Height at least 4 inches taller than recipient
Age 18–55 years
No smoking history
No significant past medical history
No recent viral illnesses (no active EBV, hepatitis, HIV)
Normal electrocardiogram and echocardiogram
Normal chest radiograph
$PaO_2 > 80$ mmHg on room air
FEV_1 and FVC > 85% predicted
Normal high-resolution chest CT
Quantitative ventilation/perfusion scan evidence to tolerate donation of a quality lobe

ORGAN PROCUREMENT

Once appropriate organs are identified, donor procurement is initiated. Most centres send a procurement team to complete a final donor evaluation and to ensure that organs are retrieved in a reproducible and predictable state. Final evaluation includes a review of chest radiographs, ventilator settings, arterial blood gasses, ABO compatibility, and the chart for appropriate brain-death notations and factors potentially affecting lung quality. Donor harvest usually involves multiple organ procurement teams, and a coordinated effort is crucial to ensure optimal harvest of utilizable organs. The donor is positioned in the operating room for median sternotomy. Once the sternum has been opened, both pleural spaces are carefully opened and the lungs are both visually inspected and carefully palpated. Large contusions, malignant lesions, unexplained masses or large infarcts are contraindications to harvest. The transplant (implant) team is notified once a final decision is rendered regarding the donor lungs, and donor cross-clamp time is estimated. For acceptable lung donors, the pericardium is opened and the heart and great vessels are exposed. Details of the procurement operation depend on whether the heart will be harvested; we will focus on

the preservation and removal of the lungs for isolated lung transplantation.

An excellent flush with pulmonoplegic solution is critical to the harvest. This ice-cold solution must be thoroughly distributed throughout the lung fields to provide even cooling and as complete removal of donor blood from the pulmonary vascular tree as possible. This needs to be accomplished expeditiously without hydrostatic trauma to the lungs. The currently preferred preservative solution is low-potassium dextran solution (LPD, Perfadex Vitrolife Gothenburg, Sweden) (Struber *et al*., 2000; Fischer *et al*., 2001; Kelly *et al*., 2003), which is broadly categorized as an extracellular solution. The solution is usually buffered by the retrieval team with 1 meq sodium bicarbonate (or THAM)/litre LPD (Perfadex), and does not require the use of prostaglandin. The solution is infused at 4°C (Mora and Patterson, 2002) and for a total of 60 ml/kg. Critical aspects of the harvest include adequate venting of the left atrium by resection of the tip of the left atrial appendage, and venting the inferior vena caval blood below the diaphragm (if possible) by the abdominal organ procurement team, to limit contact between the relatively warm venous blood and the lungs. Prior to aortic cross-clamp, the donor is fully heparinized. Superior vena cava inflow to the heart is occluded, the inferior vena cava is opened on its anterior aspect, the left atrial appendage resected and the ascending aorta is cross-clamped. Once ejection of blood from the right heart has stopped, pulmonoplegia solution is infused through the main pulmonary artery under physiological pressures and vented through the left atrial appendage. A complete dose of pulmonoplegia (60 ml/kg) requires about 10 minutes to infuse. During this time the lungs are ventilated with 100% oxygen, ensuring optimal distribution of the solution through the pulmonary tree, and cold solution or ice is placed into the pleural spaces to provide some topical cooling and to insulate against warming. Once the infusion is complete and the heart has been removed, a retrograde flush with 10–15 ml/kg pulmonoplegia is infused via the pulmonary veins (the total dose is divided evenly between the veins). Recent reports suggest that this retrograde flush improves organ preservation (Wittwer *et al*., 2004) by flushing out blood clots or other matter that has embolized to the pulmonary arteries and reaches vascular beds not well perfused via the antegrade route. Clear fluid return from the pulmonary artery suggests a complete lung flush. Once the lungs have been completely flushed, they are removed by incising the bilateral pleura, starting at the diaphragm and proceeding in a cephalad direction to the apex of the chest. The aortic arch and azygous vein are transected during this mobilization and the entire lung block is carefully dissected off the oesophagus and aorta. Paratracheal tissues are removed and the trachea mobilized. The endotracheal tube is then pulled back to the superior part of the trachea and the lungs are carefully insufflated by the anaesthesiologist to eliminate atelectasis; inspiratory pressures are monitored to limit parenchymal lung injury. Once the lungs are well inflated, the trachea is staple transected well above the carina and the lungs are removed to a back table (**Figure 49.8**). Care must be taken to ensure that there are not significant areas of atelectasis but also that the lungs are not over distended under high pressures; they should be soft and moderately inflated to avoid lung injury. The lungs are placed in a bag with 1–2 litres cold Perfadex solution; this bag is then placed in another bag with cold saline and transported to the recipient operation on ice. Donor lungs can safely sustain an ischaemic time of up to 8 hours in this state (Mora and Patterson, 2002; Conte and Reitz, 2002; Kelly *et al*., 2003).

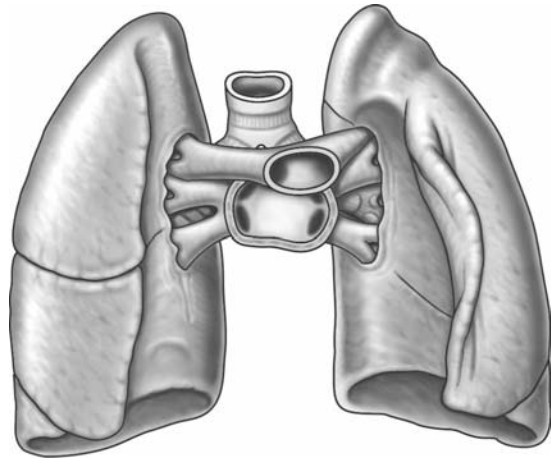

Figure 49.8

GRAFT PREPARATION

Donor lungs are brought into the recipient operating room and are maintained under sterile and cold conditions. Each of the individual donor lungs is subsequently prepared for implantation (**Figure 49.9**). This preparation includes tailoring of the pulmonary venous cuff to form an optimal anastomosis with the recipient. The graft pulmonary artery is isolated from the other hilar structures and its length is tailored to ensure that the anastomosis does not impinge on the

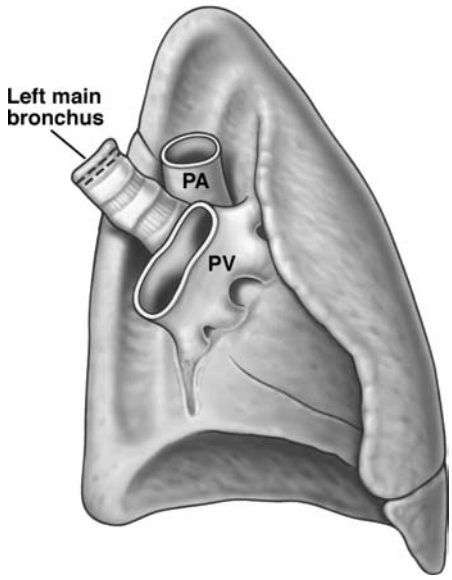

Figure 49.9

lobar arterial branches and is not too long and therefore result in arterial kinking. The donor airway is transected about two rings from the first airway arborization for a direct end-to-end anastomosis with the recipient airway.

LIVING LOBAR LUNG TRANSPLANTATION

The concept of living lobar lung transplantation is to provide the recipient with the physiological equivalent of at least one lung and to expose the living donors to minimal risk. The donor left and right lower lobes are implanted on their respective sides in the recipient. Vascular and bronchial lobar anatomy has consistently allowed for a safe donor procedure and for adequate recipient pulmonary function.

Technique

The donors and recipients are all placed in separate operating rooms for simultaneous surgery. Epidural catheters are placed in the donors for postoperative pain management. After induction of general anaesthetic, donors undergo fibre-optic bronchoscopy to exclude obvious infection and inflammation and to evaluate airway anatomy. A double-lumen endotracheal tube is subsequently placed and the donor is prepared for thoracotomy by positioning in the lateral decubitus position. Prostaglandin E_1 infusion is initiated to provide pulmonary vasodilatation and the dose is adjusted to maintain adequate donor systemic blood pressure. The goal of the donor operation is to harvest a graft with adequate cuffs for transplantation and to provide minimal damage to the remaining portions of the donor's lung.

Donor Right Lobectomy

A right posterior lateral thoracotomy is performed and the chest is entered through the sixth intercostal space, with the lung deflated by the anaesthesia team. The lung is inspected and mobilized by incising the inferior pulmonary ligament and a careful dissection of the pleura overlying the hilar structures. The fissure is then dissected to expose the arterial supply to the lower and middle lobes. Adequate distance between the superior segmental artery and the artery to the right middle lobe for placement of a vascular clamp between them is ideal (**Figure 49.10a**). However, lobar arterial anatomy is variable and occasionally there are two middle lobe arteries, with one arising distal to the superior segmental artery. In this situation, the distal middle lobe pulmonary artery will need to be sacrificed. Next the inferior pulmonary vein is mobilized into the pericardium, which will ultimately allow placement of a vascular clamp onto the donor atrium and provide for an adequate venous cuff for the graft (**Figure 49.10b**).

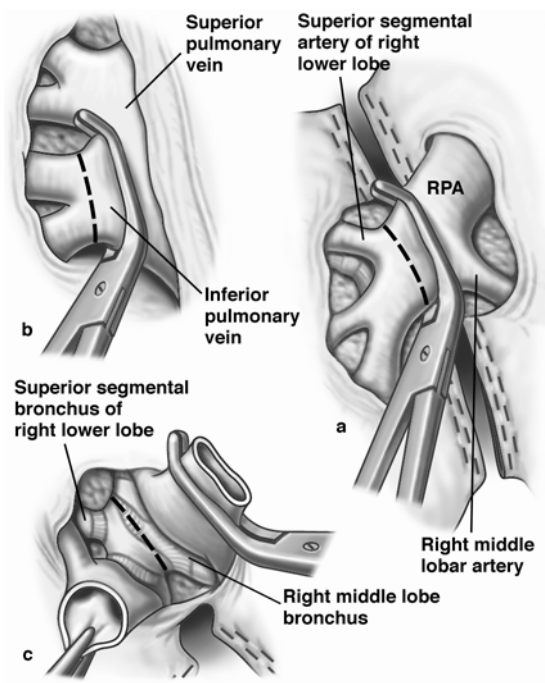

Figure 49.10

Once the vasculature has been prepared, the fissure is completed (if necessary) with a vascular stapler. The lung is subsequently ventilated and inflated for 5–10 min and 10 000 units heparin and 500 mg methylprednisolone are administered intravenously. After adequate circulation, the lung is deflated and the pulmonary artery is clamped, followed by clamping of pulmonary vein. The artery is transected to allow for an adequate (donor) graft cuff and to allow for repair without compromise to the remaining branches. The inferior pulmonary vein is next transected, allowing for an adequate cuff. The airway is then exposed and transected in a oblique manner, starting from above the bronchus to the superior segment of the lower lobe down to a point below the take-off of the right middle lobe (**Figure 49.10c**). The entire lobar harvest is performed expeditiously to limit warm ischaemic time and, once complete, the donor graft is wrapped in a cold moist sponge and taken to a separate sterile table for preservation. The donor pulmonary artery and vein are repaired with a running double-layer polypropylene suture. The airway is closed with interrupted polypropylene sutures, avoiding compromise of the bronchus intermedius or of the airway to the middle lobe. Two pleural drainage tubes are placed and the chest closed.

Donor Left Lower Lobectomy

The donor is positioned in the left lateral thoracotomy position, the chest entered through the sixth intercostal space and the left lung is mobilized. The fissure is dissected and the pulmonary arterial anatomy is evaluated. The relationship between the artery to the lingula and the superior segmental artery to the lower lobe is critical (**Figure 49.11a**). Ideally, the pulmonary artery can be transected to provide an adequate cuff and not compromise either the superior segmental artery or the lingular artery. The lingular artery can be sacrificed if it is small and if there is no impairment of absorption atelectasis when it is occluded. It can be reimplanted if necessary. The lower lobe pulmonary vein is mobilized into the pericardium and the fissure is completed with staplers. The lung is then reinflated and ventilated for 5–10 minutes and heparin and steroid are administered, as described for the right lobar donor. The vessels are subsequently clamped with vascular clamps and transected, as described for the right donor lobe. The airway is exposed and transected in an oblique manner, so that the superior segmental bronchus to the donor lobe and the left upper lobe bronchus are not compromised (**Figure 49.11b**). The donor lobe is removed to a back table for preservation and the pulmonary vessels and bronchus are repaired as previously described.

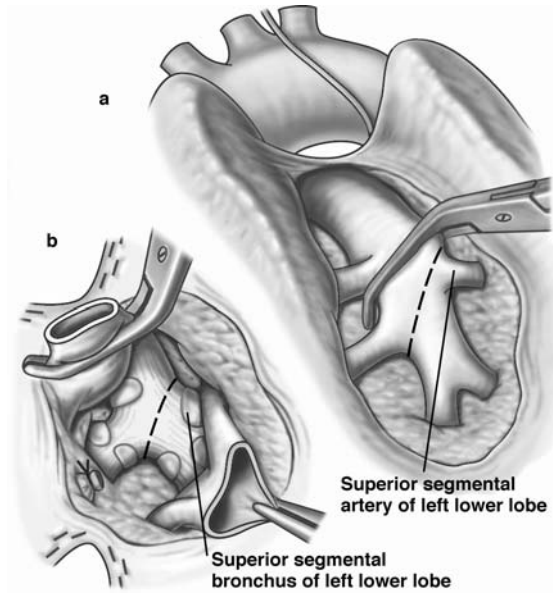

Figure 49.11

Lobar Allograft Preservation

Immediately after harvest, the donor lobe is taken to a back table and immersed in cold crystalloid solution. The airway is carefully intubated with an appropriately sized endotracheal tube and carefully ventilated with a hand-bag and room air. An inline manometer is used and the lung is inflated to 20–25 cm H_2O to avoid barotrauma. During this time 1 litre of pulmonoplegia (LPD) is administered antegrade through the pulmonary artery and another 0.5–1.0 litre of pulmonoplegia is administered retrograde through the pulmonary vein until the effluent is clear and the graft is blanched white. When flushing is complete, the lobe is inflated to 75% maximal inflation, the tube is removed and the airway is occluded with a non-crushing vascular clamp. The lobe is then stored in cold solutions, as described for the cadaveric lungs, and transported to the recipient operating room.

RECIPIENT OPERATION

The vast majority of paediatric lung transplants are bilateral and we will focus on the implantation technique for bilateral sequential lung transplantation; single lung transplant techniques are described elsewhere (Conte and Reitz, 2002).

Anaesthesia and Positioning

The patient is positioned supine, anaesthesia induced, and a double lumen endotracheal tube utilized to provide

selective lung ventilation when possible. Arterial and venous monitoring lines are placed and a urinary catheter positioned in the bladder. The arms are cushioned and supported above the patient's face, using a crossing bar so that there is unobstructed access to the bilateral chest. Alternatively, the arms can remain at the patient's side.

Cardiopulmonary Bypass

Bilateral sequential lung transplantation without cardiopulmonary bypass is possible and has strong proponents (Adatia *et al*., 1994; Date *et al*., 1996; Waters *et al*., 1996). However, it requires the ability to tolerate single lung ventilation and the increased right ventricular load present when one pulmonary artery is clamped. The aetiology and nature of pulmonary failure in children dictates that most paediatric lung transplants require utilization of cardiopulmonary bypass.

Operative Technique

A bilateral subcostal incision is made and the pleural spaces are entered through the fourth intercostal space. The internal mammary arteries are ligated bilaterally, the sternum is transected through the intercostal space in a horizontal manner and the entire anterior mediastinum and bilateral pleural spaces are exposed (**Figure 49.12**). Adhesions are taken down in both pleural spaces and the pericardium is entered vertically. Proponents of off-bypass lung transplantation will initiate pulmonary resection based on split-lung ventilation perfusion scan results. The inferior pulmonary ligament is mobilized and the hilar pleura are incised exposing the hilar structures. The patient's ability to tolerate single lung ventilation is assessed, which may require transient occlusion of one of the main pulmonary arteries. Most children will require cardiopulmonary bypass, but as much mobilization as possible is completed prior to the initiation of bypass. For bypass, the patient is heparinized with 300 u heparin/kg, with a goal of achieving an activated clotting time of 400; the ascending aorta is cannulated and a single two-stage venous cannula is placed in the right atrium. Normothermic bypass is initiated. The right lung is then removed by staple transection of the pulmonary veins, pulmonary artery and the mainstem bronchus at the hilum. The pulmonary artery is subsequently circumferentially mobilized into the pericardium and under the superior vena cava. The pulmonary veins are mobilized by circumferentially incising the surrounding pericardium, taking care not to damage the phrenic nerve. Sondegaard's groove is developed to provide a large pulmonary venous cuff. The airway is carefully mobilized, enough to completely visualize the anatomy, but also to conserve its vasculature; the posterior aspect rarely requires mobilization and the adjacent pleura acts to reinforce the airway anastomosis. The contralateral lung is usually removed, the existing endotracheal tube removed, the airway lavaged and a new endotracheal tube is placed to reduce the potential for contamination in suppurative lung disease. The right mainstem bronchus is opened adjacent to the previously placed staple line, and is prepared by trimming to two rings distal to the carina. During this time, exposure is facilitated by careful placement of an Alice clamp on the mainstem (bronchial stump) cartilaginous rings anteriorly. The graft is brought up into the field, orientated and prepared for implantation as previously described. It is very important to identify the proper orientation for each cuff pedicle, to avoid kinking or twisting of the anastamoses. The graft is placed into the chest and insulated by a cold laparotomy sponge. The airway anastomosis is completed first. A 4-0 polypropylene suture (5-0 is utilized in children less than 20 kg) is run across the membranous portion of the airway, employing large evenly spaced bites so that there are roughly five to eight bites in this area (**Figure 49.13**). The suture is continued in a simple running manner across the cartilaginous portions, with bites encompassing at least one ring on each side. After completion of the anastomosis, areas of concern are reinforced with simple interrupted sutures. Next, a side-biting clamp is placed on the left atrium and the pulmonary veins are incised to create a donor cuff. The pulmonary venous anastomosis is completed, utilizing a running polypropylene suture (**Figure 49.14**), and is left slightly patent for subsequent flushing. A vascular clamp

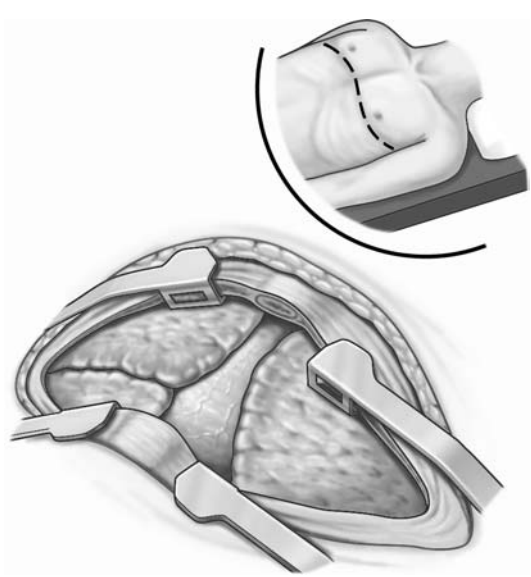

Figure 49.12

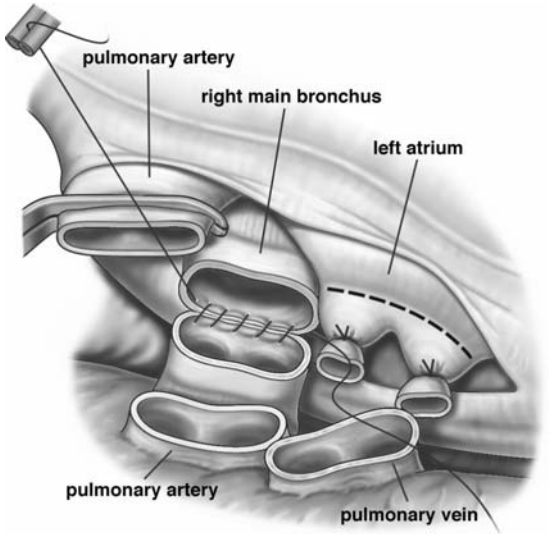

Figure 49.13

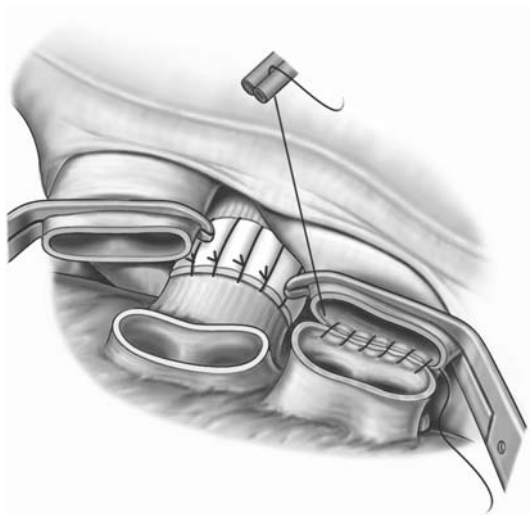

Figure 49.14

is placed on the proximal branch pulmonary artery, a cuff is created by resecting the previously placed staple line, and an end-to-end anastomosis is completed with a running polypropylene suture (**Figure 49.15**). Care must be taken to ensure that this anastomosis is not twisted or kinked due to redundancy; this can be avoided by careful orientation of the donor and recipient arteries, utilizing stay sutures, and by properly preparing the length of both arteries. The pulmonary artery clamp is carefully opened and the graft flushed

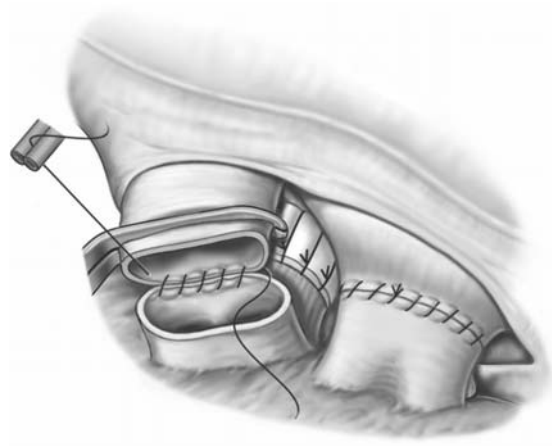

Figure 49.15

through the (patent) venous anastomosis until the effluent is blood. The pulmonary artery is reclamped, the venous anastomosis is completed by tying the previously placed suture, and the pulmonary venous clamp is removed. Steroids (10 mg/kg solumedrol) are administered and the pulmonary arterial clamp is carefully removed to provide a controlled reperfusion of the graft. A pulmonary artery vent can be placed through the main pulmonary artery if flow appears to be extravagant. The contralateral lung is implanted in an analogous manner. Ventilation is initiated, utilizing 30–40% inspired oxygen, and atelectasis is carefully cleared from the lungs, avoiding excessive airway pressures and barotrauma. Weaning from cardiopulmonary bypass slowly but progressively increases the pulmonary blood flow. High oxygen concentrations are avoided, due to concern about (oxygen) free radical-induced reperfusion injury, and a positive end expiratory pressure of 6–8 cmH_2O aids in obtaining oxygen saturations above 92%. Once the patient is stable, the bypass cannulae are removed and heparin is reversed with protamine. Two pleural drainage tubes are placed on each side and the chest is closed once bleeding has been controlled. The sternal edges are approximated, using a steel wire ''figure-of-eight'', and the ribs are apposed with multiple interrupted absorbable sutures. Intercostal blocks or the use of local anaesthetic infusion pumps can help to alleviate postoperative pain. The wound is subsequently closed with absorbable sutures and dressed. Trans-oesophageal echocardiography is performed to assess pulmonary vein patency. Bronchoscopy is also performed to evaluate the bronchial anastomoses and to clear any blood or secretions. The patient is subsequently transported to the intensive care unit.

Living Related Lobar Transplantation

This operation requires the utilization of cardiopulmonary bypass to ensure that the grafts are reperfused in a controlled manner. The implantation operation is as described previously, with adjustments made for the size of donor pedicles. A slow and controlled wean from CPB is critical to avoid reperfusion injury and subsequent damage to the lobar grafts, which now handle an entire cardiac output (Starnes *et al*., 1994, 1996, 1997, 2004; Haddy *et al*., 2002; Haddy and Starnes, 2003). Nitric oxide is liberally employed to help control pulmonary arterial pressures and the patient is maintained mildy hypovolaemic to avoid overflow to the grafts.

POSTOPERATIVE MANAGEMENT

Reperfusion injury and cardiopulmonary bypass contribute to capillary leak and increased permeability in the transplanted lungs. Pulmonary lymphatic disruption makes these lungs susceptible to pulmonary oedema; consequently, fluid administration is limited and mild hypovolaemia is accepted immediately after surgery. Adequate perfusion is important but volume boluses are discouraged and systemic blood pressure is supported with vasoactive drugs, such as dopamine or epinephrine. These agents are also utilized to support the right heart in patients with reduced function due to pulmonary hypertension. Mild diuretic therapy is initiated within 24 hours after the transplantation, to ensure that fluid does not accumulate in the grafts (lungs).

Ventilator settings are chosen to minimize atelectasis and barotrauma; the patient is usually ventilated with 7–10 ml/kg and a positive end expiratory pressure of 5–8 cmH_2O. Ventilator rate is adjusted based on carbon dioxide levels from the arterial blood gas. The fraction of inspired oxygen concentration (FiO_2) is adjusted to the lowest level, which will provide for saturations of 92% and above. The FiO_2 is minimized to limit free radical-induced lung injury and to limit atelectasis. Early graft dysfunction traditionally occurs in approximately 10% of lung transplants. It results in increased capillary permeability, alveolar oedema, poor gas exchange, impaired compliance and increased pulmonary vascular resistance. The resulting hypoxaemia is initially treated with increases in FiO_2, positive end expiratory pressure, and patient sedation. Despite these interventions, the process can progress and early intervention with nitric oxide or extracorporeal oxygenation may be necessary. Nitric oxide is routinely employed at our centre after lung transplantation, and has been shown to improve oxygenation and reduce pulmonary artery pressures in patients after lung transplantation (Adatia *et al*., 1994).

Nitric oxide is weaned off and patients are extubated once there is no evidence of significant bleeding, they are alert, breathing spontaneously at an acceptable rate, have adequate pain control, and have sufficient oxygenation and ventilation at a physiological positive end expiratory pressure. Pain control is critical at this point and can optimally be controlled with an epidural catheter. Aggressive respiratory therapy, including chest physiotherapy, coughing, deep breathing and incentive spirometry, is employed. Intermittent positive-pressure breathing (IPPB) and other methods to encourage recruitment of alveoli are occasionally required. Chest tubes are progressively removed once drainage is less than 100–200 ml/day (approximately less than 2 ml/kg/day) and there are no air leaks.

Management for the patient after living related lobar lung transplantation differs from cadaveric lung transplant in several ways. Because the entire cardiac output flows through only two pulmonary lobes, post-transplant efforts are made to minimize the risk of reperfusion injury and pulmonary oedema by maintaining a moderate state of hypovolaemia. Meticulous fluid management is crucial. Nitroglycerin infusion and inhaled nitric oxide are both employed routinely prior to separation from bypass and for about 48 h postoperatively. Positive end expiratory pressure is used aggressively at levels of 5–10 cmH_2O for 48 hours to optimally recruit alveoli and decrease atelectasis. Because of size and shape discrepancy between the donor lobe and the recipient thorax, abrupt application of suction to the chest tubes can result in graft distortion and kinking of the pulmonary artery or veins. The tubes are placed to low suction (10 cmH_2O) sequentially for 1 hour intervals in a rotational fashion for the first 24 hours and are then placed on continuous suction with progressive increase to 20 cmH_2O over the next 48 hours. These tubes remain for 2–3 weeks after transplantation because of residual pleural spaces related to graft shape.

IMMUNOSUPPRESSION

During preparation for the operating room immunosuppression is initiated with oral tacrolimus and mycophenolate mofetil (MMF). The combination of tacrolimus, MMF and prednisone has been shown to provide for optimal long-term results (Keenan *et al*., 1995; Mulligan and Wood, 2003; Taylor, 2003) and our postoperative immunosuppression is based on these three agents (**Table 49.4**).

Some centres employ induction therapy utilizing polyclonal rabbit antithymocyte globulin in an attempt to minimize incidence of acute rejection and the subsequent development of bronchiolitis obliterans (Moon *et al*., 2002). This is believed to increase the risk

Table 49.4 Immunosuppression protocol.

Preoperative	
Tacrolimus	0.075–0.1 mg/kg/dose, enteral, 1–4 hours prior to operation
MMF	15 mg/kg, enteral, 1–4 hours prior to operation
Interoperative	
Methylprednisolone	10 mg/kg intravenous
Postoperative	
Tacrolimus	0.075 mg/kg, enteral, every 12 hours, levels checked daily and doses adjusted accordingly
MMF	10–15 mg/kg/dose, enteral, every 12 hours
Methylprednisolone	5 mg/kg, intravenous, every 8 hours for 3 doses, then 0.5 mg/kg, intravenous, every 12 hours and convert to prednisone once tolerating oral medications

MMF, myophendate moferil.

Table 49.5 Complications related to immunosuppressants (Starnes *et al*., 1996, 1997, 1999, 2004; Huddleston *et al*., 1999, 2002; Dobrolet *et al*., 2001; Fiser *et al*., 2001; Sweet, 2003; Boucek *et al*., 2004).

Complication	Approximate prevalence (%)	Responsible agent
Infection (opportunistic, bacterial)	40	All immunosuppressives
Hypertension	61	Tacrolimus, cyclosporin, steroids
Renal insufficiency	34	Tacrolimus, cyclosporin
Hyperlipidaemia	15–60	Cyclosporin, steroids
Neurological (seizures, tremor, headache, leukoencephalopathy)	20–30	Tacrolimus, cyclosporin
Skeletal (growth retardation, osteoporosis, avascular necrosis)	25–75	Steroids
Haematological (leukopenia, anaemia)	50	Azathioprin, gancyclovir
Diabetes insipidus	20–30	Tacrolimus, cyclosporin, steroids
Malignancy (post-transplant lymphoprolipherative disorder)	5–10	Tacrolimus, cyclosporin, steroids

of infection and possibly increase the risk of subsequent development of post-transplant lymphoproliferative disorder (PTLD). Our centre has avoided the use of antithymocyte antibody, and triple-drug therapy (tacrolimus, MMF, steroids) is initiated perioperatively. Methylprednisolone is administered at a dose of 5 mg/kg intravenously every 8 hours for the first 24 hours and is subsequently weaned to oral prednisone. MMF and tacrolimus are dosed to obtain therapeutic serum levels and are adjusted based on measured levels, renal function, fluid status, and drug interaction. These medications have significant side effects (**Table 49.5**) and doses are weaned as possible to maintain adequate immunosuppression.

COMPLICATIONS

Acute Rejection

Acute rejection is most likely to occur within the first few weeks to months after transplantation (Sweet, 2003; Boucek *et al*., 2004). Signs of rejection are subtle and include fever, leukocytosis, infiltrates, effusion, dyspnoea and deteriorating function on spirometry. Trans-bronchial biopsy can confirm the diagnosis of rejection and bolus steroid therapy is subsequently initiated. Some advocate routine surveillance biopsy and bronchioalveolar lavage to evaluate for rejection (Girgis *et al*., 1995). The frequency of surveillance is determined by the proximity to transplantation, with decreasing

frequency 1 year post-transplant (Girgis *et al*., 1995). Our centre employs a selective approach to biopsy, based on clinical evidence of rejection and on decreases in pulmonary function by spirometry. A decrease in FEV_1 of 15–20% or greater, or a decrease of forced expiratory flow of 25–75 seconds (FEF_{25-75}) of 25% or greater from previous baseline (or predicted values for those who are growing) is an indication for bronchoscopy and biopsy.

Infection

Infection remains a significant cause of post-transplantation morbidity and mortality and the recipient is at particularly high risk immediately after transplantation (Sweet, 2003). Pulmonary infections have immediate life-threatening consequences in the immunosuppressed patient but they also predispose to bronchiolitis obliterans and affect long-term pulmonary function (Sweet, 2003). Bacterial and opportunistic infections are common after lung transplantation. Prophylactic antibiotics are initiated prior to transplant surgery, based on organisms harboured by the recipient, and are ultimately adjusted based on the results of cultures obtained from the donor. Recipients with cystic fibrosis often harbour multiple organisms, including multiresistant *Pseudomonas*, requiring the use of synergistic antimicrobials. Fungal infection after transplantation can be devastating and requires aggressive therapy with amphotericin and other antifungal agents. The most common viral infection after transplantation is cytomegalovirus (CMV), which can cause a localized pneumonitis, affect the gastrointestinal tract or cause a systemic viremia. Most CMV-related infections occur within the first 6–12 months after transplantation (Moon *et al*., 2002). Diagnosis is based on seroconversion to anti-CMV immunoglobulin M (IgM-positive), and increase in CMV immunoglobulin G titres by four-fold, positive viral urine cultures, buffy coat, bronchopulmonary lavage, or CMV inclusion bodies in tissue (lung) biopsy (Moon *et al*., 2002). Recipients who are CMV-negative but receive lungs from a CMV-infected donor are at highest risk and 90% will develop infection (Moon *et al*., 2002). Prophylactic therapy varies among centres but generally all patients who receive grafts from a CMV-positive donor are treated proactively with gancyclovir (Moon *et al*., 2002; Sweet, 2003). Patients also receive sulfamethoxazole–trimethoprim as prophylactic therapy from *Pneumocystis carinii* pneumonitis.

OUTCOMES

Survival and functional classification at follow-up have been addressed in the introduction to this chapter, and they indicate that lung transplantation is an effective palliation for end-stage lung disease. The primary aetiology of death (after lung transplantation) is related to time after transplantation and is presented in **Table 49.6**, which represents data reported to the International Society of Heart and Lung Transplantation (Cooper *et al*., 1993; Boucek *et al*., 2004).

Table 49.6 Cause of death for early and late mortalities after lung transplantation (data from ISHLT).

30 days or less post-transplantation	5 years or greater post-transplantation
Graft failure (50%)	Infection (31%)
Infection (25%)	Graft failure (31%)
Technical (15%)	Bronchiolitis obliterans (25%)

Bronchiolitis obliterans probably represents the most significant factor limiting long-term survival and has been reported to be responsible for 40% of mortalities at long-term follow-up after lung transplantation. This process, which may represent chronic rejection, is a histological diagnosis based on lymphocytic infiltration of the small airways, subendothelial fibrosis, fibromyxoid deposits and fibrous obliteration of the bronchioles. Since this is a focal process and potentially difficult to biopsy, criteria which clinically describe bronchiolitis obliterans have been defined and are referred to as the *bronchiolitis obliterans syndrome*. This syndrome represents the otherwise unexplained development of obstructive changes in pulmonary function (Estenne *et al*., 2002). A decline in FEV_1 of 20% or greater from previous baseline defines the presence of bronchiolitis obliterans syndrome, and a staging system based on the amount of decline (in FEV_1) exists. Decreases in the FEF_{25-75} have recently been incorporated to provide more sensitivity and specificity in the diagnosis and staging of bronchiolitis obliterans syndrome (Estenne *et al*., 2002). Due to lung (airway) growth in paediatric patients, spirometry results are evaluated relative to percentage predicted values, instead of absolute values. Newer techniques to facilitate measurement of these parameters in infants are being developed.

Factors associated with the risk of developing bronchiolitis obliterans include: frequency or severity of rejection episodes, graft ischaemic time, immune mechanisms related to mismatch or the presence of antibodies, infection with CMV (Starnes *et al*., 1999, 2004; Woo *et al*., 2001; Bowdish *et al*., 2004) and chronic aspiration (Starnes *et al*., 1999, 2004; Woo *et al*., 2001; Bowdish *et al*., 2004). Reduced incidence of bronchiolitis obliterans has been observed after

living lobar lung transplantation for children (relative to cadaveric), and this is believed to be due to reduced graft ischaemic times and higher quality donor grafts (Woo *et al*., 1998; Starnes *et al*., 1999, 2004; Woo *et al*., 2001; Bowdish *et al*., 2004). There exists no consistent and reliable treatment for bronchiolitis obliterans and when it leads to end-stage lung function, retransplantation may be the only therapy.

Living lobar lung transplantation has proved to be safe and has provided good outcomes (Woo *et al*., 1998, 2001; Starnes *et al*., 1999, 2004; Bowdish *et al*., 2004). This procedure has effectively enlarged our donor pool and the number of recipients who are successfully transplanted (Starnes *et al*., 1994, 1996, 1997, 1999, 2004; Bowdish *et al*., 2004); however, it does place donors at risk for potential complications. After approximately 250 donor operations at our centre there is a 20% donor complication rate (for living lobar lung donation) and there have been no donor deaths (Bowdish *et al*., 2004; Starnes *et al*., 2004). Our experience has indicated that living lobar lung transplantation provides excellent results for properly selected children who require lung transplantation.

SUMMARY

Paediatric lung transplantation remains an effective (palliative) treatment for end-stage lung disease and is available at a limited number of centres. The evolution of living lobar lung transplantation has effectively increased the number of children who can benefit from this therapy. Bronchiolitis obliterans continues to be a significant barrier to long-term survival after lung transplantation. Future advances in immunosuppression, surgical techniques and the basic understanding of bronchiolitis obliterans are expected to provide significant improvements for paediatric lung transplantation.

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50

Miscellaneous: Straddling Atrio-ventricular Valves, Pericardium, Tumours, Diverticula and Ectopia Cordis

J. Stark and V. T. Tsang

STRADDLING ATRIO-VENTRICULAR VALVES

Straddling atrio-ventricular (AV) valves are rare but significant malformations across a wide spectrum of congenital heart disease. This anomaly was first described by Lambert in 1952, but only since the 1970s has it been diagnosed more often, especially through the use of cross-sectional echocardiography (Liberthson *et al*., 1971; Tandon *et al*., 1974). Milo *et al*. (1979) extensively studied 57 hearts with straddling AV valves. They described several subgroups and commented on the disposition of the conduction mechanism in the presence of straddling valves. In principle, they distinguish four major groups: straddling of mitral or tricuspid valves in either AV concordance or AV discordance. The position of the AV conduction system in hearts with straddling valves depends on whether the septum extends to the crux of the heart (see Chapter 8). The advent of three-dimensional echocardiography has provided a technique that could offer significant additional information in the assessment of the straddling AV valves and the atrio-ventricular junction before the operation (Vogel *et al*., 2000).

An AV valve straddles if its tension apparatus originates from both sides of the ventricular septum (**Figure 50.1**). In straddling mitral valve, some of the chordae of the mitral valve cross the ventricular septal defect (VSD) and attach to the endocardial surface of the morphologically right ventricle. In straddling tricuspid valves, some of the chordae attach through the VSD on the endocardial surface of the morphologically left ventricle.

Tabry *et al*. (1979) distinguished three types of straddling. In type A, the chordae attach into the opposite ventricle but close to the defect (**Figure 50.1a**). In type B, the attachment is farther away from the defect but still on the septum of the other ventricle (**Figure 50.1b**). In type C, the chordae and its papillary muscle are attached to the free wall of the opposite ventricle (**Figure 50.1c**).

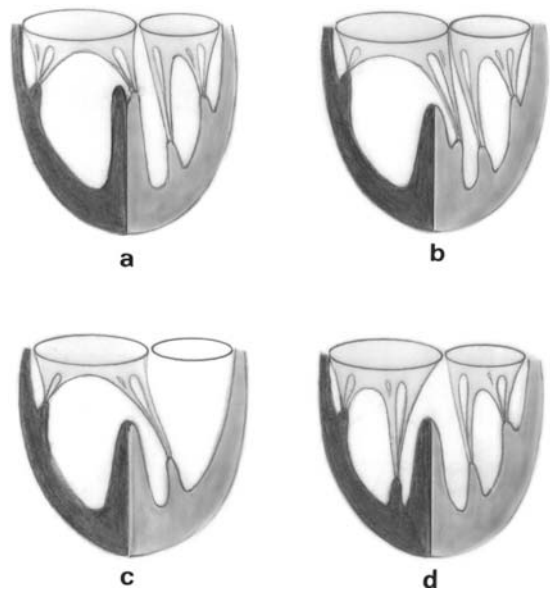

Figure 50.1

Straddling must be distinguished from overriding. If the valve orifice is committed to both ventricles but the papillary muscles and chordae of that valve are attached into the one ventricle, overriding, but not straddling, is present (**Figure 50.1d**). In other words, straddling is related to the tension apparatus, whereas overriding is related to the annulus. Some valves both straddle

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and override, and others straddle without overriding or override without straddling (Milo *et al*., 1979). In some hearts, the degree of overriding of the straddling valve may be such that the AV connection should be classified as double-inlet ventricle, rather than AV concordance or discordance.

Surgical Implications

Hearts with straddling AV valve present a major operative challenge. Straddling adds to the difficulty of the repair, which in itself is often already complex (Danielson *et al*., 1979). Tabry *et al*. (1979) reported a series of 19 patients who underwent operation in the presence of straddling AV valves. Of the 14 hearts with two ventricles, two had type A, two had type B, and 10 had type C straddling. Ten patients underwent a corrective procedure, and nine received palliative treatment.

If opting for repair, the issue is whether to use a Fontan type of surgery or to perform complex biventricular repair. The sizes and the morphology of the ventricles, septal malaligment and valvar morphology will influence decision-making. Fontan operation has been advocated on the assumption that it carries a lower operative risk then the biventricular repair. Delius *et al*. (1996) demonstrated that sometimes the early risk of the complex biventricular repair might outweigh the long-term disadvantages of a single-ventricle approach.

Tabry *et al*. (1979) used four basic types of corrective operation:

1. ***The Rastelli operation*** for patients with transposition of the great arteries. The intraventricular patch was placed either to the left of the straddling tricuspid valve or to the right of the straddling mitral valve. The former is not so simple, even with the straddling cords near the septal crest. With the patch placed on the left side of the septum, the conduction tissue would be at risk.
2. ***Closure of the VSD*** was used when straddling was minimal; the patch was again deviated so as to accommodate the straddling valve.
3. ***Replacement of the straddling valve*** and either closure of the defect or septation of the univentricular heart.
4. ***A modified Fontan operation***.

Patients with VSD or with double-outlet right ventricle and VSD and straddling tricuspid valve may be treated by closure of VSD, but a slot is made in a patch for the chordae tendineae arising from the left ventricular side of the septum. We would also consider excluding the straddling cord with its involved leaflet onto the ''wrong'' side of the VSD patch, without compromising the tricuspid valve function. This approach is further supported by the results reported by Serraf *et al*. (1996). Kirklin and Barratt-Boyes (1993) suggested the detachment of the papillary muscle from the septum on the inappropriate side of the ventricle. The septal defect is then closed, and the papillary muscle is reattached to the patch. Necrosis of the reattached papillary muscle is a real danger of this approach (Reddy *et al*., 1997).

With the straddling chordae attached to the outlet septum, Serraf *et al*. (1996) suggested the creation of myocardial flaps, so that an unobstructed pathway is left for tunnel construction. When the chordae arise further down on the ''wrong'' side of the septum or from the parietal wall, biventricular repair involving replacement of the straddling valve would carry a high risk. We currently consider biventricular repair unsuitable for complex lesions with restricted pulmonary blood flow and major straddling (Delius *et al*., 1996). Total cavopulmonary connection can be performed in such cases with low risk if the haemodynamic criteria for this operation are met (see also Chapter 39).

Results

Only limited information about results is available. In the Tabry *et al*. (1979) series, there were three early deaths and one late death among 10 patients who underwent corrective operations. The highest mortality rate was observed among patients who underwent replacement of the straddling valve, closure of the VSD and placement of an extracardiac valued conduit. All three patients treated in this way died. In addition, seven patients in the series (five who underwent corrective operation and two who received palliative treatment) required implantation of a pacemaker for complete heart block. There were no early deaths among the nine patients who received palliative treatment. Pacifico *et al*. (1979) operated on six patients in whom the cardiac malformations included straddling tricuspid or mitral valves. There was one early death and one late death. Total cavopulmonary connection performed in patients with a straddling valve or one hypoplastic ventricle or both carries a mortality rate of about 10%.

More recently, Serraf *et al*. (1996) operated on 34 patients with double-outlet right ventricle or transposition of the great arteries and straddling AV valves, between 1984 and 1995. All but three patients had ventricles of an adequate size. Thirty patients underwent biventricular repair, four had a Fontan-type repair. Four died early and there was one late death. All occurred in the group with biventricular repair. In the authors' experience, curtain-like tricuspid valve chordae were the only contraindication to biventricular repair.

We have reported (Delius *et al*., 1996) 11 patients with complex diagnoses and AV valve straddling.

All patients in this subgroup of patients underwent Fontan-type operations. There was one early death. This raises an important question: is a high-risk biventricular repair always preferable to a single-ventricle repair? At present the decision depends on individual surgeon's and institution's preference, but there are no hard data supporting one or the other approach.

PERICARDIUM

Congenital Defects

Congenital defects of the pericardium are rare. Both total absence of the pericardium and smaller localized defects have been reported (Fosburg *et al*., 1968). Most patients are asymptomatic; however, pericardial defects may cause chronic chest pain, atypical angina, arrhythmias and vague, non-specific symptoms. Herniation and sudden death may occur, especially with left-sided deficiencies of the pericardium. For these reasons, the diagnosed presence of a pericardial defect is an indication for operation. Preoperative diagnosis may be made through chest X-ray, echocardiography and CT/MRI to demonstrate herniation. During repair, care must be taken to avoid the phrenic nerve, which may be in the vicinity of the anterior border of the defect. The defect may be closed either directly or with a synthetic material such as Gore-Tex.

Cysts

Pericardial cysts are rare; they are most often asymptomatic (Shidler and Holman, 1952) and are usually discovered on a routine chest X-ray; however, they may cause cardiac compression, chronic inflammatory symptoms or cardiac chamber erosion. The differential diagnosis consists of lung tumours, thymoma and other mediastinal lesions. Diagnosis is aided by echocardiography and CT/MRI. However, exploration may be required in order to establish a definitive diagnosis (Basso *et al*., 1997). Trans-oesophageal echocardiography may be useful preoperatively to plan the surgical approach. Cyst excision is usually accomplished without cardiopulmonary bypass, but bypass and even cardioplegic arrest may be required if there is preoperative evidence of the involvement of cardiac chambers (Chopra *et al*., 1991).

Tumours

Tumours of the pericardium are extremely rare. Benign tumours (lipomas, fibrous polyps, haemangiomas), primary mesotheliomas, sarcomas and teratomas have been reported (Ebert, 1976; Basso *et al*., 1997).

Tamponade

Tamponade most commonly occurs after cardiac surgery, but it also may occur after penetrating injuries to the heart or with acute effusion into the pericardial cavity. Cardiac function is compromised mainly in diastole by increased pressure from the pericardial fluid. In acute tamponade, especially in infants, a small amount of blood or fluid may cause severe symptoms.

Increased venous pressure, tachycardia, decreased cardiac output, and signs of shock may develop rapidly. High venous pressure in the presence of other signs of shock is very suggestive of tamponade. Often oliguria or even anuria is one of the first signs. Cardiac tamponade is a clinical and not an echocardiological diagnosis. The latter findings, which would support tamponade, are diastolic collapse of the right atrial, right ventricular and left atrial walls (Armstrong *et al*., 1982).

Tamponade after surgery should be treated by immediate reopening of the sternotomy wound, evacuation of blood clots and control of the bleeding. In a critical situation, opening the lower end of the sternotomy wound to evacuate part of the haematoma and then transferring the child to the operating room for formal revision and resuture of the sternum may be life-saving.

Treatment of cardiac tamponade from acute effusion must be prompt. Pericardial aspiration may be lifesaving. The standard approach is to aspirate from the subxiphoid position of the patient (**Figure 50.2**). The aspirating

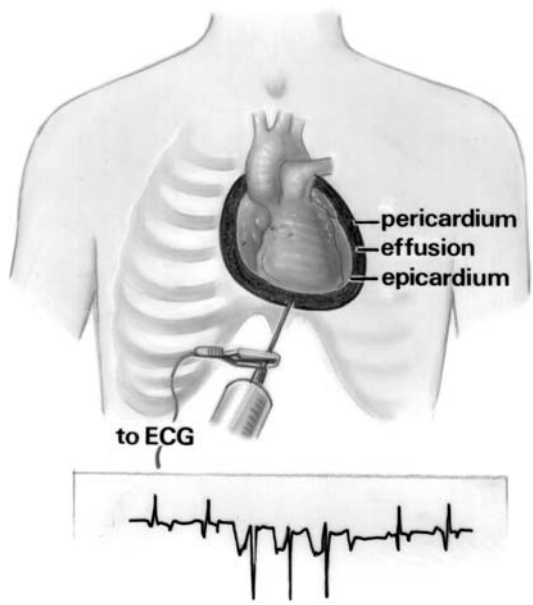

Figure 50.2

needle can be attached to the electrocardiographic (ECG) lead, so that the contact of the needle with the heart (usually the left ventricle) can be detected immediately. However, pericardial aspiration is currently best performed under two-dimensional (2D) echocardiography control. This makes the procedure very safe. Even small amounts of fluid evacuated by aspiration would decrease cardiac filling pressure markedly. If a large amount of fluid is present or if the effusion is recurrent, a drain may be inserted by means of the Seldinger technique.

Chylopericardium may cause either acute or chronic tamponade. It was observed in patients with increased superior vena caval (SVC) pressure, such as in SVC thrombosis or in SVC obstruction after the Mustard operation. Chylous or lymphatic effusion can also complicate the postoperative course of patients after the Fontan operation or after total cavopulmonary connection. Increased SVC pressure or transection of the lymphatic channels in the mediastinum are two possible causative mechanisms.

Aspiration of the pericardial fluid and institution of a medium chain triglyceride diet is our treatment of choice. If a large quantity of drainage persists, chest tube insertion may be required and total parenteral nutrition may be instituted; a chest drain may have to stay in place for several weeks in rare cases. The use of somatostatin (Pratap *et al*., 2001) has been reported. In persistent cases, a pericardial window is created to avoid the danger of acute tamponade when the fluid reaccumulates. If the chylopericardium or chylothorax is caused by SVC occlusion or constriction, treatment should concentrate on re-establishing free SVC drainage. If surgical cause (after operations for PDA, vascular ring, coarctation of aorta, etc.) is suspected and the accumulation of the chylus does not stop after one or two aspirations, surgical revision and ligation of lymphatic channels is indicated.

Pericarditis

Aetiology

Pericarditis may occur as a primary disease or as a secondary manifestation of a systemic disease. Primary pericarditis may have many different aetiological agents. Viruses are probably the most common cause; viral pericarditis is often accompanied by myocarditis. Other causes include bacteria (pyogenic or tuberculous pericarditis). Pyogenic pericarditis in infants is a rare disease but can be fatal (Gersony and MacCracken, 1967). The primary source may pass unnoticed. The organisms commonly involved are *Haemophilus influenzae, Staphylococcus, Pneumococcus* and *Streptococcus* spp. Secondary pericarditis can occur with scleroderma, rheumatic fever, lupus erythematosus, sarcoidosis, autoimmune disease and irradiation. Cholesterol pericarditis has been observed in patients with hypothyroidism.

Diagnosis and Treatment

Early treatment is important because constrictive pericarditis may develop in many of the conditions just listed. First, pericardial fluid is aspirated so as to establish the diagnosis. Antibiotics are used according to the sensitivity, for a period of 4–6 weeks. In chronic effusion or in the presence of pus, a drain is inserted. Adequate drainage is extremely important. As mentioned, pericardiotomy should be considered if adequate drainage is not achieved after insertion of a drain.

In tuberculous pericarditis, early treatment with antituberculous drugs is recommended. Wood (1956) emphasized that constrictive pericarditis developed in most patients in whom treatment was delayed for more than 4 months. Even in the current era, up to 80% of patients with TB pericarditis develop constrictive pericarditis despite anti-TB treatment (Yang *et al*., 2005). Constrictive pericarditis develops after other chronic inflammatory processes; however, tuberculosis is the most common aetiological factor. A fibrous, thickened pericardium constricts the heart; this impairs the diastolic filling of the ventricles. Further constriction limits the systolic ejection by both ventricles.

Physical signs of constriction vary with the severity of the lesion. High venous pressure, decreased cardiac output and low blood pressure are commonly observed. Liver enlargement and ascites are often present. The peripheral pulse may be paradoxical. Chest X-rays show the heart to be normal in size or slightly enlarged. Calcium deposits may be seen, especially in the lateral projection. The ECG study shows low voltage; T-waves may be flat or inverted. The diagnosis is best confirmed by echocardiography. Cardiac catheterization, if indicated, shows a rapid rise in diastolic pressure in the right ventricle, with a plateau effect and a small A-wave. Right atrial pressure is markedly elevated.

Pericardiectomy

Treatment of constrictive pericarditis is surgical. In patients with proven diagnosis of tuberculosis, early resection of the pericardium should be considered. Operation is easier at the effusion stage and should prevent the development of severe constriction. Pericardiectomy is performed after adequate preparation of the patient. If a tuberculous origin of pericarditis is proved, an adequate antituberculosis regimen should be established before surgery. Different surgical approaches (bilateral thoracotomy, midline sternotomy, and left

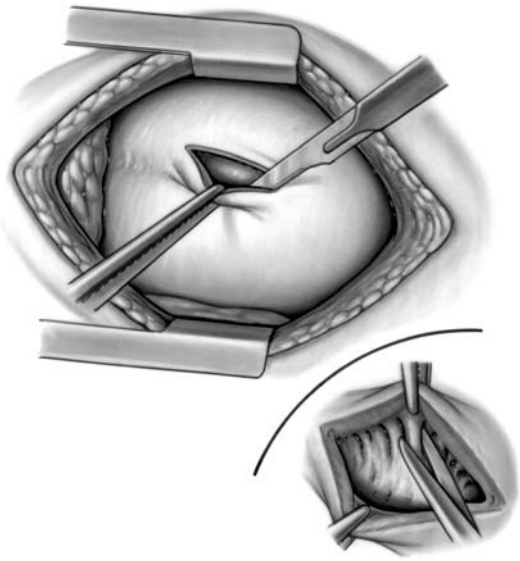

Figure 50.3

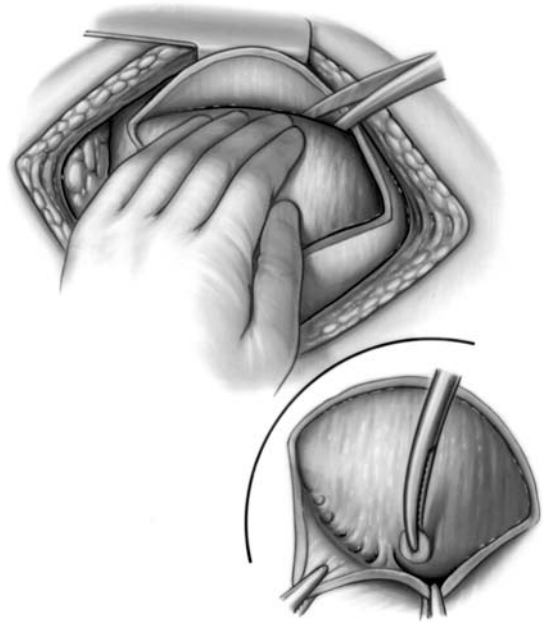

Figure 50.4

thoracotomy) have been suggested. An inadequate approach does not enable complete removal of the diseased pericardium and also may pose difficulties should complications (usually bleeding) occur during surgery. For these reasons, a midline sternotomy is preferred. The thymus is dissected from the pericardium, and the pleura is pushed laterally. The pericardium is then incised longitudinally with a scalpel (**Figure 50.3**).

Careful dissection with a sharp and blunt technique is used to identify the epicardium. Once the correct plane of dissection is found, the dissection becomes easier. The pericardium is first freed over the left ventricle and the left atrium. If the right ventricle is mobilized first, filling of the heart improves, but the left ventricle could possibly not cope. Dissection in the correct plane is relatively bloodless; excessive blood loss often indicates that the dissection is too deep into the myocardium. Extreme care must be exercised to avoid injury to the coronary vessels. Bleeding from the heart is controlled by very fine stitches or by compression with warm saline. Control of such bleeding has been facilitated by the use of human fibrin seal (Haverich and Borst, 1981). The left ventricle should be freed at least partially over its posterior aspect. Gentle retraction of the heart to the right (**Figure 50.4**) facilitates the exposure. Care must be taken to identify and mobilize the phrenic nerve in order to avoid damage to it.

The right ventricle and the right atrium are often thin-walled, and so pericardial resection over these chambers must be undertaken with extreme caution (**Figure 50.5**). When the right ventricle and the right atrium are freed,

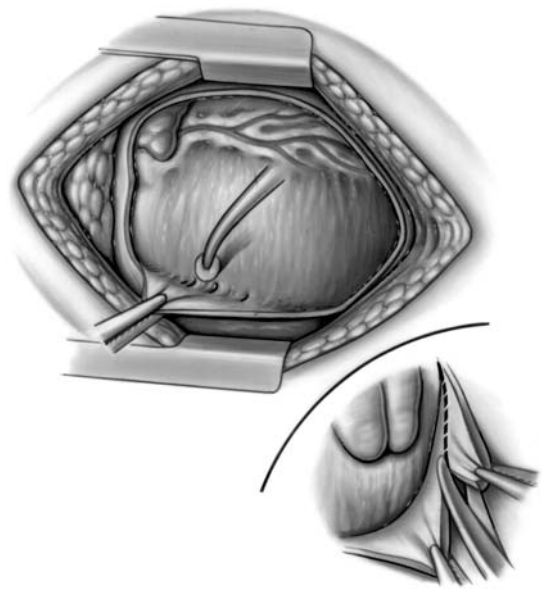

Figure 50.5

the pericardium is resected from the aorta, pulmonary artery and both SVC and inferior vena cava. When the resection is completed, both pleural spaces and the mediastinum are drained, and the sternotomy is closed in the routine manner.

The results are satisfactory in patients in whom resection is adequate (Simcha and Taylor, 1971; McCaughlin *et al*., 1985). Yang *et al*. (2005) reported favourable outcomes following a combination of antituberculosis chemotherapy and timely pericardiectomy. Even today it is important for the clinicians to maintain a high degree of suspicion of TB pericarditis in patients with pericardial effusion.

TUMOURS

Tumours of the heart are uncommon in children (Basso *et al*., 1997). The majority of such tumours are histologically benign: rhabdomyomas, lipomas, fibromas, teratomas and myxomas. Spontaneous regression is possible in some, but they can produce life-threatening complications. Malignant tumours (Engle and Glenn, 1955) and secondary tumours have also been reported (Edwards, 1972). Early echocardiography, including foetal echocardiography, has resulted in better understanding of the history, the improvement of management and the improvement of overall outcome (Hwa *et al*., 1994). Magnetic resonance imaging (MRI) has the additional advantage of providing a wider view of the extracardiac field, as well as the cardiac tumour characteristics (Kiaffas *et al*., 2002).

Rhabdomyomas are the most common cardiac tumours in childhood. They are usually associated with tuberous sclerosis. Because they are often multiple, evaluation by 2D echocardiography of all four chambers is essential. Severe and even fatal arrhythmias have been caused by cardiac catheterization in patients with rhabdomyomas (Corno *et al*., 1984).

Clinically benign tumours present either with symptoms of left or right ventricular outflow tract obstruction or with complete heart block or arrhythmias. Surgery may be considered for obstructing intracavity rhabdomyomas. Complete removal is not always possible. Intramural ventricular rhabdomyomas have limited potential for growth and may regress with time; therefore, management should be conservative. If severe arrhythmias are noted, myocardial mapping can be performed. Depending on the location of the tumour, they may be amenable to surgery (Goldman *et al*., 1985).

Fibromas are the second most common cardiac tumours in childhood. They originate either from the intraventricular septum or from the wall of the left or right ventricle. They are solitary and intramural, and they entrap and displace cardiac segments. The clinical presentation includes congestive heart failure, arrhythmias, and murmurs of significant aortic or mitral valve dysfunction. Sudden death from conduction system involvement has been reported (Folger, 1992). 2D echocardiography is important for establishing the diagnosis. Trans-oesophageal echocardiogram and MRI can provide important additional information. Cardiac catheterization is rarely indicated nowadays. Surgery may be indicated for treating arrhythmias and for preventing sudden death. Because these tumours do not destroy but do displace myocardium, the remaining muscle is usually sufficient to afford adequate cardiac performance. Long-term survival is possible with complete excision. Asymptomatic survivors 10 years after incomplete excision have also been noted. A 10 month-old patient with endocardial fibroma operated upon by Waterston in 1967 (Lincoln *et al*., 1968) remained asymptomatic 21 years after the operation. Unresectable left ventricular fibroma has been successfully treated with cardiac transplantation (Jones *et al*., 1986).

Atrial myxomas are rare in children. They usually rise from the fossa ovalis and project into the left atrium. Patients with atrial myxomas may present with symptoms of atrio-ventricular (AV) valve obstruction, tumour embolization or systemic illness.

The diagnosis is sometimes based on an incidental finding of a mass on plain chest X-ray or histological studies of the thrombus removed after thrombectomy in tumours that embolize. Echocardiography establishes the accurate site and size of the tumour (Miralles *et al*., 1991). Trans-oesophageal echocardiogram is an obvious adjunct to assess the septal anatomy and the AV valves. MRI can provide characteristics of the tumour. It is recommended that the operation be performed as soon as the diagnosis of atrial myxoma has been made.

The concept of familial occurrence of myxoma and its association with the NAME syndrome (naevi, atrial myxoma, myxoid neurofibromata and ephilides) has been espoused. Patients with the syndrome are younger than those with non-familial myxomas; these myxomas are found in atypical or multicentric locations, and without large enough resection of the intra-atrial septum, the incidence of recurrence is increased (Miralles *et al*., 1991).

SURGICAL TECHNIQUE

Operations are performed on cardiopulmonary bypass, hypothermia and cardioplegia. The approach is through a midline sternotomy. Care must be taken to remove the tumour without damaging the adjacent important structures; this is often difficult and sometimes impossible.

Minimal manipulation of the heart is recommended. The aorta, SVC and inferior vena cava are cannulated; the patient is cooled; the aorta is cross-clamped; and cardioplegic solution is infused before the tumour is

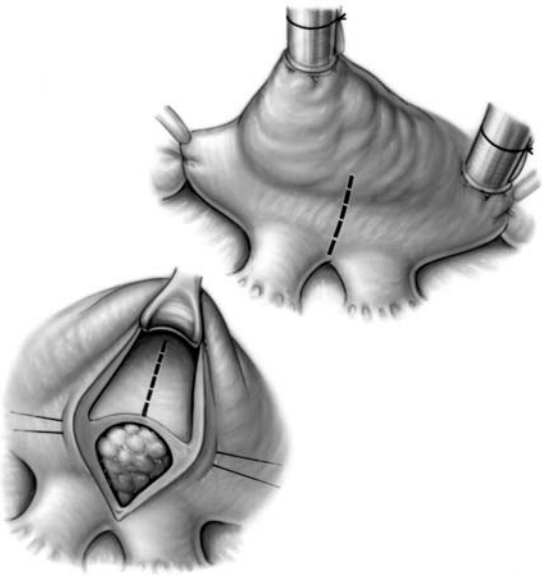

Figure 50.6

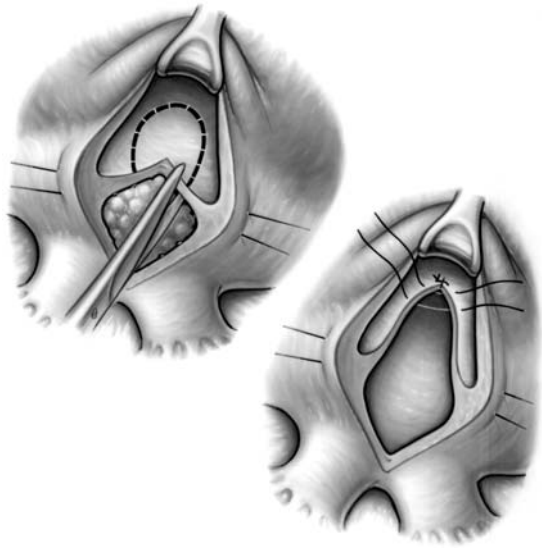

Figure 50.7

approached. These actions should avoid embolization of the tumour.

Bi-atrial incision gives an excellent exposure. It starts at the base of the right atrial appendage and extends across the crista terminalis and interatrial groove and into the left atrium. It can also be extended to between the right upper and lower pulmonary veins (**Figure 50.6**). Stay stitches are placed, and the tumour is excised with the part of the atrial septum to which it is attached (**Figure 50.7**). The atrial septum is then sutured directly, or a small patch is inserted. The atriotomy is closed in the usual manner.

An alternative approach is from the left atrium, behind the interatrial groove. Using the standard incision behind the interatrial groove opens the left atrium, and the tumour is visualized (**Figure 50.8**). The tumour is removed after excision of the part of the interatrial septum to which it is attached (**Figure 50.9**). The defect in the septum is sutured directly or with a small patch, and the left atriotomy is closed in a routine manner (**Figure 50.10**).

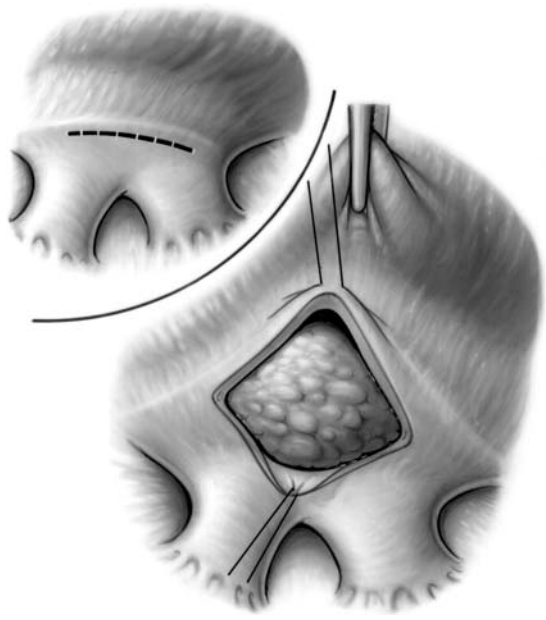

Figure 50.8

DIVERTICULA

Congenital diverticula of the heart are rare. They may involve any of the four cardiac chambers (Copeland *et al*., 1975; Szarnicki *et al*., 1981). Symptoms and signs depend on the size and the location of the diverticula. Chest X-ray may reveal abnormal cardiac shadow. Echocardiography should be diagnostic, if necessary, further supported by CT.

Atrial Diverticula

Atrial diverticula may cause arrhythmias and may be a source of embolization. Atrial thrombi are often present, and cerebral embolization constitutes a major risk. Behrendt (1979) mentioned that of 12 reported

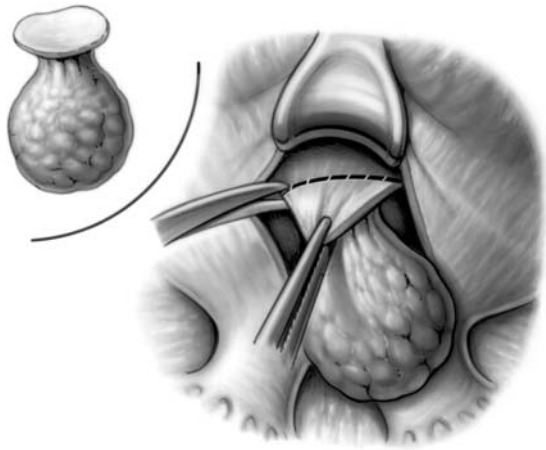

Figure 50.9

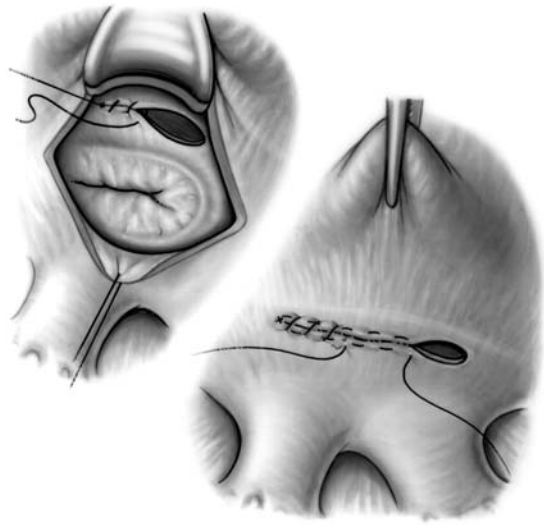

Figure 50.10

patients with the left atrial diverticula, six suffered cerebral embolus. Because of the risk of thrombosis and embolization, operation is recommended. The aorta should be cross-clamped before the diverticulum is manipulated, so as to avoid dislocation of a possible thrombus.

Ventricular Diverticula

A ventricular diverticulum should communicate with the ventricle through a narrow neck, unlike an aneurysm, which has a wide base with the ventricle. The narrow muscular diverticula are frequently associated with midline defects in the chest or abdominal walls or both (Cantrell *et al*., 1958). Associated intracardiac anomalies are common in affected patients (Murphy *et al*., 1968).

Operative treatment of ventricular diverticula is recommended because of the possibility of rupture, thrombosis or impairment of ventricular function with mitral regurgitation. The diverticulum is resected and the opening is closed with a patch on cardiopulmonary bypass.

ECTOPIA CORDIS

In true ectopia cordis, the heart is completely outside the chest (**Figure 50.11**), and the pericardium is missing. In most patients, the ectopic heart has other, usually complex, congenital malformations. Probably for this reason, the operative treatment was almost uniformly

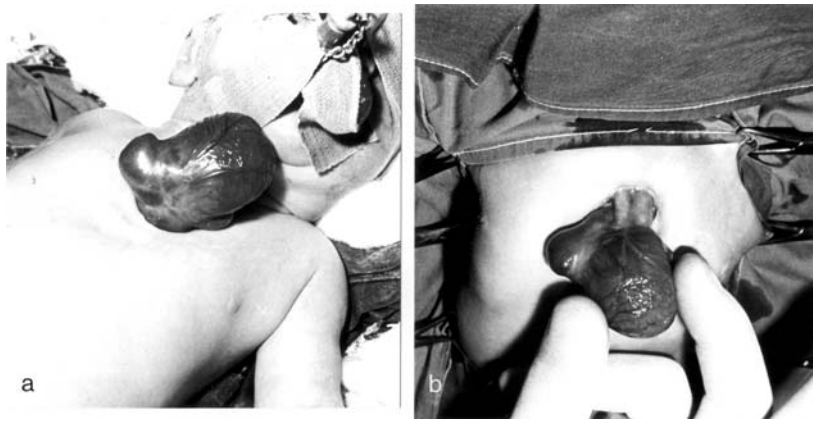

Figure 50.11

unsuccessful. Ravitch (1979) mentioned only one survivor from among 18 published cases. A successful two-stage repair involving the use of a plastic chest shield at the initial operation was reported by Tachibana *et al*. in 1991. However, the presence of significant extracardiac defects would strongly influence the outcome of these infants, despite the modest success with the staged approach (Hornberger *et al*., 1996).

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51

Data Collection and Assessment of Performance in Congenital Heart Surgery

S. Gallivan, J. Stark, N. Kang and V. T. Tsang

INTRODUCTION

In 1908, the English surgeon Ernest Hey Groves published an article in the *British Medical Journal*, entitled ''A plea for a uniform registration of operation results'' (Hey Groves, 1908). In it, he remarked:

> To keep a tally of operations and their outcome, to have these results available for reflection, to be shared with colleagues, and for inspection by others, should be a simple matter and entirely appropriate.

Despite enormous progress in the development of information technology, electronic databases and sophisticated statistical software, there are still numerous challenges faced in the seemingly simple task of such data collection.

It is increasingly being accepted that the routine monitoring of outcomes has an important role to play in the provision of safe and effective health care services, whether in congenital heart surgery (CHD) or in any other speciality. A variety of different systems have been developed and implemented, with different degrees of success. However, as far as we are aware, there has been no systematic evaluation or comparison of the value of systems in current use (Stark and Stark, 2003).

In this chapter we shall summarize the main components of systems for monitoring outcomes, what these systems are trying to achieve, and what effects, both positive and negative, they have had.

Outcomes monitoring can be performed at different levels, ranging from an individual surgeon to international institutions. A number of different systems have been developed and, despite their differences, the majority share a number of common features. In particular, they break down the monitoring process into two main components: ***data collection and storage*** and the subsequent ***assessment of the data***, usually using some form of statistical analysis. Both of these processes involve many complex issues, particularly in relation to CHS, and form the main themes of this chapter.

It may seem a relatively easy matter to develop standardized data collection procedures and analyses and then to compare outcomes between different surgeons or institutions, thus allowing those with good or bad performance to be identified. However, there are many pitfalls in this seemingly simple process. While the authors certainly subscribe to the view that more data should be collected and more learned about factors that affect outcomes, this enthusiasm needs to be tempered by an appreciation of the inherent difficulties in this process. This chapter attempts to discuss both sides of the matter.

ISSUES ASSOCIATED WITH DATA COLLECTION

Golden Rules of Data Collection Studies

Data collection is a complex and time-consuming process and should not be embarked upon lightly. Golden rules, before ever embarking on a data collection exercise, are:

- Make sure that resources are available to ensure that accurate data collection and compilation can and will be achieved.
- Be sure that one knows precisely what the data are going to be used for.
- Make detailed plans, in advance, concerning what analysis will be carried out, in order to address the questions that the data are supposed to shed light on.

Sadly, all too often these rules are broken. Often, ambitious large-scale data collection exercises are initiated that flounder once initial enthusiasm has waned,

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since data collection takes too long and suffers due to competing commitments. Data collection studies that run their course may then fall foul of the sad fact that, just because there are a lot of data, does not mean that there is any useful information. Even if there is useful information present, if one waits until the end of data collection to decide what analysis to do, one may then discover that there are far too few data to infer anything with a useful degree of certainty. Equally, one might find that one has collected far too much and thus wasted a considerable amount of time and energy.

Specific Purposes for Collecting Data Relating to CHS

Paediatric cardiac surgeons may want to use a database for a number of reasons (Stark and Stark, 2000):

1. To monitor results of their operations.
2. To give the parents of their patients current information about the risk of the proposed operations.
3. To evaluate long-term results.
4. To initiate research projects.
5. To participate in the preparation of guidelines and standards and to facilitate quality assurance procedures.
6. To provide a basis for management decisions regarding the allocation of resources.

A database is a structured collection of information. It need not necessarily be implemented on a computer system, but doing so offers many advantages. In particular, a well designed computer database can greatly simplify and speed up data entry and data retrieval, can offer a wide variety of sorting and searching operations, can facilitate a range of methods for viewing and presenting the data, can allow better error checking and can permit non-local access.

ISSUES ASSOCIATED WITH THE SCALE OF DATA COLLECTION RELATED TO CHS

Williams *et al*. (2002) described several types of databases with different magnitudes of collected data:

1. Some of the data for all of the patients—**registry database**
2. All of the data for a defined subset of patients—**academic database**
3. Some of the data for some of the patients—
4. All of the data for all of the patients—**integrated database**

Clearly, the larger the scale of data collection, the more resources are needed. It is common to underestimate the time and effort required to set up and maintain databases, thus it is essential to determine an appropriate scale of data collection dependent on one's data needs and available resources. While the notion of the integrated database might seem the most appealing from Williams's classification, it would be by far the most expensive option and one should consider the issue of what analyses would ever be done that require all of the data for all of the patients. One should avoid collecting data for data collection's sake. It is very easy to compile a large list of data items. The skill lies in devising a short list of data items that suffice for the purpose one has.

In setting up a CHS database, one has to decide what data to collect (Willliams *et al*., 2002). It is important that this choice is based on the uses to which the database will be put, and it is essential that these are specified before data collection begins. The larger the dataset, the more resources are required for data collection, the higher the probability of errors in the database and the lower the likelihood that it will be used conscientiously. If clinicians are going to take central responsibility for data collection, we would recommend that at most 20–30 data items per case are collected, indeed it may well be appropriate to gather fewer, depending on the purpose of the data collection. Although clearly this does not encompass all the data that would typically be available concerning a patient, such a simple dataset is more likely to be understood and accepted by busy clinicians, and hence more likely to be used correctly. It still requires a dedicated part-time data manager (preferably a clinician) and appropriate training for all clinical staff. Finite resources thus currently preclude more comprehensive datasets in almost all institutions.

NOMENCLATURE AND MAPPING

A major difficulty in comparing outcomes between different institutions is the lack of a single accepted nomenclature for diagnoses and procedures. Paediatric cardiac surgery deals with a relatively large number of heart defects and their variations, which require a large number of different operations. In 2000, several groups of congenital heart surgeons from USA and Europe agreed on a nomenclature which was subsequently adopted by both the Society of Thoracic Surgeons (STS) and the European Association for Cardiothoracic Surgery (EACTS). (Mavroudis and Jacobs, 2000). The Association of European Paediatric Cardiologists (AEPC), adopted a nomenclature which was different from the surgical STS/EACTS nomenclature (Franklin *et al*., 1999). Efforts are currently being put into mapping between these different nomenclatures (Jacobs, 2002).

Unfortunately, mapping is not an ideal solution. Whilst it may be feasible in principle to move from a large detailed nomenclature to a more condensed one, the reverse is impossible. Inevitably there are more ''names'' in the more detailed nomenclature, so whatever mapping is considered, some of these ''names'' will be unassigned yet correspond to diagnoses or procedures that might occur in practice. Even trying to map between two different nomenclatures of comparable size can give rise to ambiguities. One should stress that, in general, such mapping is undesirable, since there will almost certainly be no way of verifying the resulting data.

ERRORS AND DATABASE VALIDATION

No matter how well designed the database and how well trained the users, it has to be accepted that most databases contain errors. Errors can arise in a variety of ways. The simplest are pure typographical mistakes, such as mis-spelling a complicated name in a foreign language, or mis-keying a date of birth. Even such innocuous mistakes can have serious repercussions. If a patient has several operations and his/her name is mis-spelled only for some of them, then he/she may appear as two different patients in the database, whilst an incorrectly entered date of birth can turn a neonate into a teenager. A second class of error can occur as a result of misunderstanding. For example, a patient with an atrial septal defect and also jaundice may have the latter miscoded by an inexperienced coding clerk as the reason for surgery. The third class of errors arises from honest mistakes that are sometimes inevitable when recording or entering complex information: the age entered into the weight field, a belated blood test result entered into the data for the wrong ''John Smith'', etc.

A very common form of error concerns the way in which missing data are treated and interpreted. A blank in a field indicating whether a patient was an emergency admission might mean that was not recorded; equally it might mean ''no'' if another field confirms that the patient was an elective case.

The final category arises from structural flaws in a database. A particularly serious example in the context of CHS databases occurs with patients who die following a number of procedures during one admission. Such a death is often not due to the last procedure carried out, and hence should not be ascribed solely to that operation; on the other hand, recording the death under all the operations in that admission can easily result in a single patient giving rise to several deaths in any statistical summary.

Rate of errors increases alarmingly with the number of items collected. This is illustrated in **Figure 51.1** reproduced with kind permission from Stark and Stark (2000). Supposing that after validation the per item error rate is 0.1%, then if 25 items are recorded per operation, on average 2.5% of the operations in the database will contain one or more faulty items of data. If, however, 300 items are recorded, then the proportion rises to 26%. If the per item error rate is 1% (which is still not overly pessimistic), then the figure rises to 95%, so data for virtually no operations would be entered completely correctly.

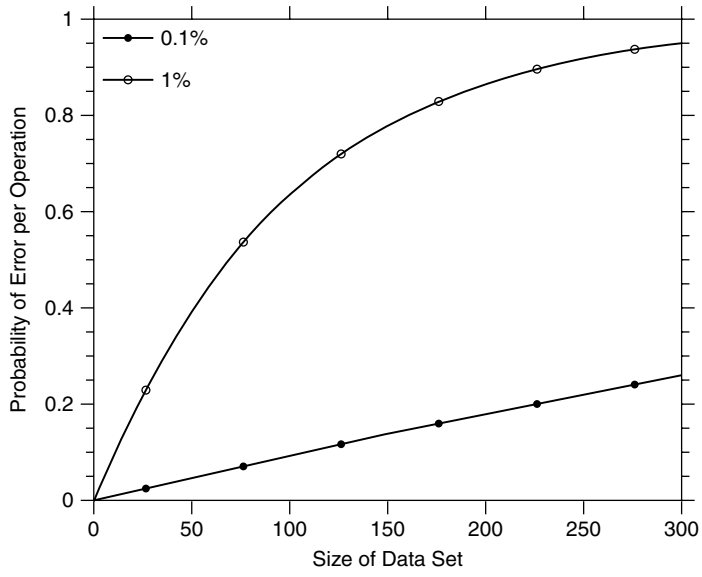

Figure 51.1

Most commercial databases include facilities to assist in reducing errors by, for example, checking that age data lie in an appropriate range, issuing a warning at the time of data entry if this is not so. Extensive use should be made of such facilities.

Database Validation

Once the data are entered, it is essential that a validation exercise is carried out to try and eliminate as many errors as possible. A well designed database can help in this task, for instance by allowing searches for empty fields, for duplicated data records, for particular combinations of operations, etc. Another mechanism for uncovering errors is simply frequent use of the database. This can reveal inconsistent or aberrant data, which can then be checked and corrected.

Another somewhat resource-intensive data validation method is to arrange for data (or a random subset of data) to be entered twice, by two separate data entry clerks. It is then possible to compare the two databases and determine discrepancies.

Many such methods of data validation and data cleaning are common, yet at present there are no formal recommendations concerning such data validation for clinical systems.

At some stage it has to be decided that the data are of sufficient quality to be used for subsequent analysis, yet often this is done in the knowledge that residual errors almost certainly persist; thus, such analysis requires something of an act of faith that such errors are unlikely to have a major effect on the analysis.

There is currently very little information on what error rates to expect in clinical databases and on the effect of different error rates on subsequent conclusions. Although some work has been done in this area (Gallivan, 2005), further research in this area is clearly needed.

EXISTING SYSTEMS

Data have been collected in a number of departmental databases for years. Perhaps the most efficient was the one used at the Hospital for Sick Children in Toronto (Williams *et al*., 2002).Valuable information about collecting data in a multi-institutional database was published by Moller (1998). The data are validated locally and the results are available within 1–2 years. This provides an excellent means for examining trends in mortality rates, but is not suitable for assessment of surgeons' performance. Since 2000, many European countries have submitted data to the database of the European Association for Cardiothoracic Surgery (EACTS). This database uses EACTS/STS nomenclature, but at present data are not validated. In the USA, the Society of Thoracic Surgeons collects data from many departments of congenital heart surgery, using the same nomenclature. Unfortunately, several European countries, including the UK, use different nomenclature (Franklin *et al*., 1999), thus making current comparison of results impossible. In the UK, all departments operating on congenital heart defects submit data to Central Cardiac Audit Database (CCAD) (Gibbs *et al*., 2004). Although the data are validated by visiting teams, the method of validation was not formally tested.

RISK STRATIFICATION

The Rationale for Risk Stratification

In cardiac surgery, the risk of perioperative death is high in peoples' minds, not least the parents of a child referred for complex surgery, who have a right to be given a fair appraisal of the risks involved. Risk is also a major issue for the surgeon to consider and should be an integral part of the decision processes involved in case planning. Risk also plays a major role in issues associated with a surgeon's own self-interest. Given that caseloads vary between surgeons, with the more experienced surgeons typically taking on the more complex cases, there is a possibility that audit results may unfairly penalize a surgeon whose mortality rates are relatively high, simply due to the fact that his caseload has been skewed towards riskier cases. In view of this, when assessing overall outcome, it is important to take case mix into account.

Unfortunately, assessing the risks associated with an individual operation is by no means straightforward. Different procedures with different degrees of complexity have different risks, and risk is also affected by factors related to the patient. Surgical experience tells us that a younger, low-weight baby requiring a repair of the truncus arteriosus would be far more at risk than an older child of normal weight requiring the repair of a VSD.

While surgical experience and subjective opinion give some insight into the factors that affect risk, there has been growing interest in devising quantitative methods for assessing risk. It is appealing to believe that a quantitative and objective method for assessing risks should be preferable to subjective opinion alone, indeed some may take this as self-evident. In view of this, there has been growing interest in the development of risk-scoring systems. However, as with all views that are put forward as self-evident, it is wise to question this and to recognize that there can be deficiencies in risk-scoring systems and that they need to be used with due care.

For adult cardiac surgery, scoring systems have been developed that take account of age, sex of the patient, presence of associated defects and co-morbidities. Given the uses to which such risk scores are put (informing patients and carers, case planning, within audit), it is

sensible that risk scoring should only be based on factors that are known preoperatively. Thus, although the occurrence of a postoperative wound infection may well add substantially to risk, it is not something that would be known in advance. Risk information conveyed to a patient or a carer should be based on what is known, not on some uncertain future event. Also, the occurrence of infections may possibly be a marker of poor surgery, so audit should not be based on a risk model that includes this as a factor, since this would tend to obscure potentially poor outcomes.

Much work concerned with risk scoring in adult cardiac surgery has been done (e.g. Hannan *et al*., 1990; Higgins *et al*., 1992; Hammermeister *et al*., 1994; Edwards *et al*., 1997). The most commonly used risk-scoring methods are the Parsonet score (Parsonet *et al*., 1989) and the euroSCORE (Nashef *et al*., 1999), and these methods are used extensively.

Risk stratification for congenital heart defects has proved more difficult. There are several reasons for these difficulties and congenital heart defects are relatively rare compared with acquired heart disease; many diagnostic groups are treated by a great variety of operations. Many patients undergo several procedures during one operation or during multiple admissions and some patients may require operations for other congenital, non-cardiac defects. Even for the more common congenital abnormalities, relatively small numbers of cases may be available for analysis. For rarer lesions the numbers of cases may be extremely small. Unfortunately, statistical inference tends to be very imprecise when sample sizes are small. In view of this, analysis of so many subgroups, many with relatively few cases, cannot be expected to be all that informative. Furthermore, age and weight variation of several orders of magnitude compound the difficulty. Prematurity, chromosomal and syndromal anomalies add to the plethora of variables that one would expect to be included as risk factors, based on surgical experience.

In such circumstances it is unwise to place too much faith in the ability of even the most sophisticated statistical analysis to discern all the factors that have a major effect on risk. If statistical analysis fails to identify weight as a risk factor, this may merely reflect the sparseness of the available data.

One should be wary about putting undue faith in impressively low p values. Surgical experience would indicate that any operation on a grossly underweight 6 month-old would be far riskier that the same operation on a 6 month-old of normal weight. It would be folly to ignore this during the surgical planning process, purely on the grounds that analysis failed to find a statistically significant association. Statistical analyses are designed to do the best that can be done with the interpretation of the available data, but ''best'' does not necessarily mean adequate. An acid test far more important than any of the p values that such an analysis may produce is to consider the face validity of the risk stratification suggested. It is useful to recall a quotation from the eminent statistician George Box: ''All models are wrong. Some are useful''.

CATEGORIZING CONGENITAL HEART OPERATIONS ACCORDING TO THEIR COMPLEXITY

Jenkins *et al*. (1994) suggested grouping operations according to subjective opinion of their intrinsic complexity. Later a modification of this grouping was devised (Gallivan *et al*., 2001) and analytical methods suggested to assess surgeons' case mix. All operations for congenital heart defects were placed in six ''complexity'' categories, based on a subjective perception of complexity of the operations rather than the mortality data. However, examining the average mortality rates in the six groups using data from one department, mortality increased progressively from group one to group six. This makes it plausible but does not prove that risk is related to the complexity category.

An attempt to provide a more inclusive system was made by Jenkins *et al*. in 2002. They developed a system of six categories of operations, based on a consensus of surgeons/cardiologists, which was based on their previous classification (Jenkins *et al*., 1994). They estimated mortality rates in each category from the Paediatric Cardiac Care Consortium (PCCC) database (4602 patients) and 4493 patients from three state hospital discharge databases. They found that 98.5% of patients from PCCC could be assigned to one of the six categories; this was possible only for 89.2% of the patients in hospital discharge datasets. This inability to assess all the patients presents a clear disadvantage of the system.

In 2004, Lacour-Gayet *et al*. published an alternative system, known as the ''Aristotle score'', to reflect complexity determined as a composite of mortality, morbidity and technical difficulty of a given procedure. A group of 50 surgeons rated 145 procedures based on these three elements, and a ''complexity score'' of 3–14.5 was arrived at by consensus. This was termed the ''basic'' score. A further 248 variables (procedure-dependent and procedure-independent) were also rated to add further points to the original score, giving a so-called ''comprehensive'' score (range 3–25). The scoring system is purely subjective in nature and was not tested during its development to see whether it did indeed predict any (or all) of the three end-points. Also, although numbers may be used as an intermediate step in deriving the final ''score'', the process is essentially a rule-based algorithm for ranking cases in order of complexity.

Pragmatically, it is eminently sensible to group operations in terms of expert subjective opinions concerning their intrinsic complexities. However, the resulting classification needs to be interpreted with caution, particularly from a statistical viewpoint. Unfortunately, confusion can easily arise, since numbers are used to label complexity categories rather than, say, letters of the alphabet. Such labels merely express an ordering rather than being in any sense quantitative. In statistical terms, complexity categories give what is called ordinal data. Unlike tangible physical measurements, such as a weight in grams, one should not use ordinal data as though they obey the laws of arithmetic. Put two 2 g weights onto a balance pan and it will balance with a 4 g weight. However, an operation in risk category 4 does not carry twice the risk as an operation in category 2. Equally, one would not expect the same mortality from a group of 200 operations equally split between categories 1 and 5 as for 200 cases from category 3. Taking averages of ordinal data or carrying out other arithmetical operations is mathematically nonsensical. Unfortunately there are occasions when this is done.

Another caveat is that, even though efforts have been made to group operations into sensible categories, mortality rates almost certainly vary between individual operations within each group. It is also entirely possible that an operation in one complexity category might have a lower mortality than an operation in a less complex category. In view of these caveats, great care is required in the interpretation and analysis of such complexity data.

Attempts to Stratify Risks

Gallivan *et al*. (2001) used a basic risk stratification, categorizing patients according to their age band (neonate, 0–28 days; infant, 29 days–1 year; child, > 1 year) and whether the operation was open or closed. The mortality risk for each of the six categories was then calculated from a study of results in congenital cardiac surgery from five UK departments over a 2 year period (Stark *et al*., 2001).

Given the relatively small sample of data available, no attempt was made to model risks for individual operations or operations with similar complexity. In view of the simple nature of the risk stratification, the method was termed ''*partial risk scoring*'' and great caution was urged in interpreting the resulting risk estimates.

Later, Jenkins *et al*. (2002) published the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. This uses six risk categories agreed upon by consensus panel of 11 surgeons and cardiologists, who subjectively rated the risk of in-hospital mortality for an initial 207 surgical procedures. The method was tested on two different datasets, amounting to approximately 9000 patients and judged to be effective. Taking other factors into account, such as age group, prematurity and presence of a major non-cardiac structural anomaly improved the discrimination of the model.

The RACHS-1 method has been studied by other groups. Both Boethig *et al*. (2004) and Kang *et al*. (2004) found the RACHS-1 risk categories to be significantly associated with postoperative mortality.

The predictive ability of the other risk stratification method, the Aristotle score, has also been studied. Al-Radi *et al*. (2005) examined results from approximately 13 000 cases that took place over 22 years and found that, although the basic Aristotle score was associated with mortality, its forecasting ability was inferior the RACHS-1 method. There were similar findings in a study by Kang *et al*. (2005a).

ASSESSMENT OF OUTCOMES

Used sensibly and sensitively, risk forecasts such as those discussed above can assist in informing parents and may also have a role in the planning and decision processes that precede surgery. In these contexts the absolute precision of a risk forecast is not a crucial issue and, in any case, information conveyed by the risk forecast should be tempered by surgical experience and judgement.

The third main use of risk forecasting is within the audit process, and the question arises how best this can be done and how much faith to place in current risk-forecasting methods in relation to audit. This starts to get into areas that are still subject to on-going research and a detailed picture has yet to emerge. Certainly there are analytical methods that can use risk forecasts to assist the audit process and these have some advantages. What is less clear is whether there are potential deficiencies of such methods and, as yet, there is insufficient evidence to judge this. Given the nature of existing risk models, which are still relatively simplistic, the advice to be given is that such methods may assist audit but that their results should be used with caution.

Graphical Methods for Assessing Outcome

A commonly used method of displaying performance is with league tables. These rankings provide an outcome measure, such as early postoperative mortality.

However, league tables have their drawbacks. Tables of numbers are not easy to interpret and it can be easy to misjudge a surgeon as performing poorly just because his mortality rate falls into the higher tail of the league. It is an indisputable mathematical truth that with any such list of numbers, there will be one number that is the biggest. This does not of itself mean that this is unacceptably high. Poloniecki (1998) discusses such misinterpretation

in his article, ''Half of all doctors are below average'', although it should be noted in passing that, strictly speaking, this statement isn't true. The proportion of doctors with less than the average number of legs is not a half but close to zero.

An alternative way of assessing outcomes is to use a graphical method to compare outcomes against a defined standard, so that any divergence in observed outcomes from the expected benchmark can be readily visualized. Such graphical methods were first introduced to surgeons by de Leval *et al*. in 1994, preceding the availability of risk stratification methods.

Since then, various analytical methods have been developed that can assist the assessment of risk-stratified outcome data (Lovegrove *et al*., 1997; Gallivan *et al*., 2001).

One such method, originally developed in the context of adult cardiac surgery, is a monitoring method, ''Variable Life Adjusted Displays'', known by the acronym VLAD. This is based on consideration of a series of operations carried out within a period being audited. A risk model is used to estimate the cumulative number of deaths ''expected'' during the series. In golfing parlance, if the underlying risk model is unbiased, then this represents ''par for the course''. The VLAD method involves tracking how a surgeon's actual cumulative mortality compares with this expected tally. This is illustrated in **Figure 51.2**, in which VLAD charts for several surgeons are displayed. The series above the horizontal axis indicate outcomes that are generally better that might be expected, according to the underlying risk model. The series that shows a sharp downward trend indicates outcomes that are worse than might be expected. Sadly, in this case the surgeon concerned was found to be suffering from a brain tumour.

The merits of VLAD lie not so much in its mathematical sophistication, which is slight, but in its simplicity, which enables rather complex case-mix adjustment analysis to be displayed in a graphical format that is readily comprehensible.

For those who are yet to be convinced by the accuracy of risk models for congenital cardiac surgery, it is not possible to use VLAD charts as they stand. However, it is possible to use the VLAD method as a basis for examining a surgeon's outcomes retrospectively over, say, a 1- or 2-year period, in order to identify periods of divergent performance that might be investigated further using other methods. To do this, a different form of VLAD chart has been developed, known as a standardized VLAD plot (Gallivan *et al*., 2001). Using the partial risk stratification based on age-band and whether operations are open or closed, simple risk adjustment can be carried out to derive an estimate for the ''expected cumulative deaths'' in the form required for a usual VLAD analysis. The only difference between this and the usual VLAD chart is that all preoperative risks are scaled by the same factor, to ensure that the resulting VLAD chart ends precisely on the horizontal axis. Similar analysis can be carried out using the RACHS-1 risk-scoring method (Kang *et al*., 2005b).

The reason for adopting this scaling is that in using this new version of VLAD, one accepts that the underlying risk stratification is only partial and is very unlikely to present an accurate picture. Scaling avoids the erroneous interpretation of such charts as an indication in absolute terms of whether a surgeon's overall outcomes accord with known standards. Use of partial risk stratification

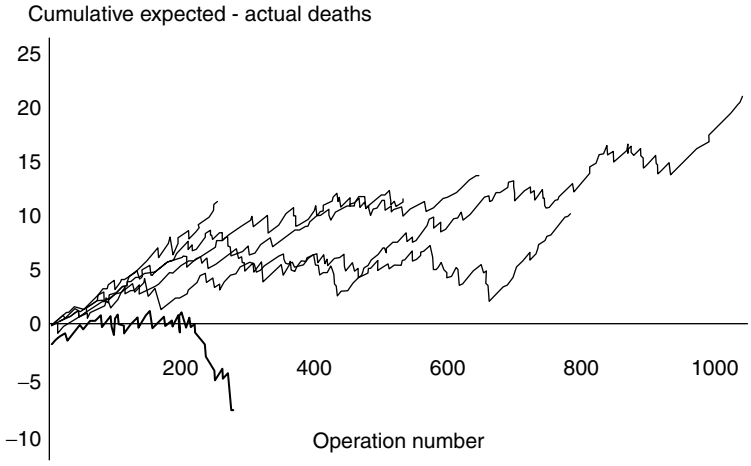

Figure 51.2

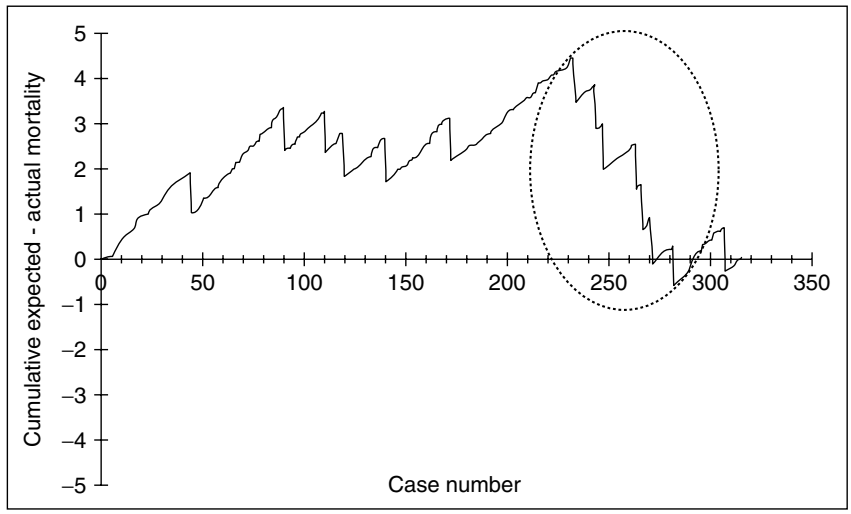

Figure 51.3

information helps to identify runs of apparently poor outcome, as shown in **Figure 51.3**. This shows a standardized VLAD curve for a series of cases of a congenital cardiac surgeon. The analysis is based on partial risk stratification of cases, according to age of the child and whether or not the operation was conducted on cardiopulmonary bypass. The chart is adjusted to the surgeon's overall mortality to ensure that the final point of the chart lies on the horizontal axis. This is certainly not statistical proof that this actually was a run of poor outcome, but it identifies cases that might benefit from further investigation using alternative audit methods.

The chart shown in **Figure 51.3** highlights a period during which there appears to be an anomalous series of outcomes. To investigate this further, a natural question to ask is whether the case load of the surgeon was particularly complex during that period. Another graphical method has been suggested to assist with investigating this. The complexity profile chart (Gallivan *et al*., 2001) is a simple chart, based on considering the number of operations carried out in each of six categories of increasing complexity. A cumulative percentage is plotted for each complexity level, showing the percentage of cases at that level or lower. All such profile charts rise to the 100% level; the faster the rise, the less complex the case load. Two such profile charts are shown in **Figure 51.4**, based on cases for the surgeon whose outcomes data were shown in **Figure 51.3**. This shows that the operations during the period of divergent outcomes were in fact less complex than during the remainder of the audit period.

LIMITATIONS OF RISK MODELLING

It is very important to recognize the inherent limitations of any risk model when interpreting outcomes. First, and perhaps most importantly, the model is only as good as the data upon which it is based. Data quality is critical to constructing a reliable risk model. All databases contain errors and omissions which reduce the ability of the model to accurately predict outcome. For example, Gibbs *et al*. (2004) reported that 22% of deaths following paediatric cardiac surgery in the UK were under-reported by participating institutions between April 2000 and March 2001. Where possible, several different sources should be used to verify, cross-validate and ensure completeness of the data.

A more subtle problem with retrospective data is ''selection bias''. Because patients have been selected for operation based on clinical decision-making and judgement as to the suitability for surgery made at the time, the data collected from this retrospective cohort is affected by bias. Put simply, the surgeon's decision whether or not to operate on a particular patient at a particular time was not a randomized event, but rather was influenced by a host of carefully considered deliberations. This has important implications for the conclusions to be drawn from the risk model. For example, identifying young age as a risk factor for closure of ventricular septal defect (VSD) does not imply that VSD closure should be postponed for as long as possible. It simply indicates that patients with large VSDs and severe heart failure required surgery earlier and were therefore sicker and less likely to do well than patients with smaller VSDs, who could be observed

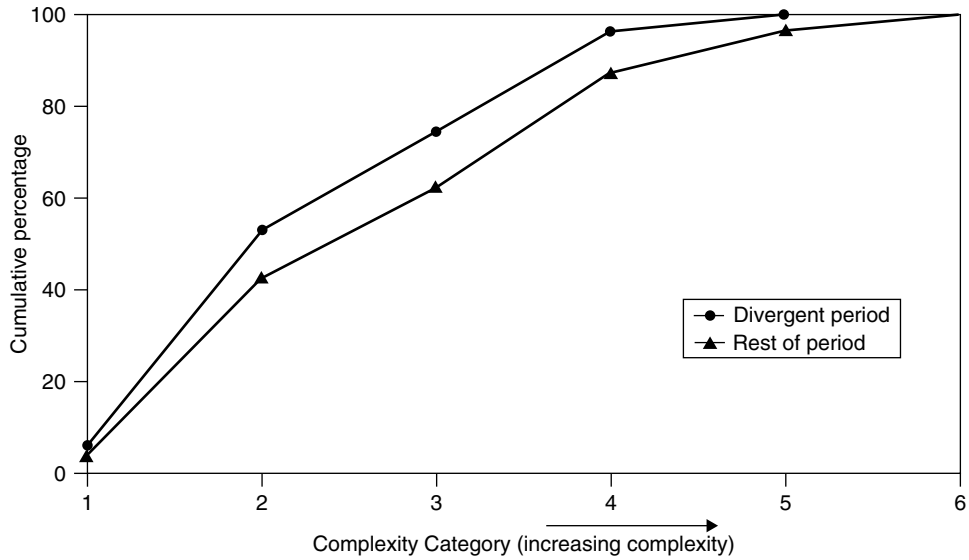

Figure 51.4

for longer, treated medically initially and operated more electively at an older age.

It is also important to realize that risk modelling itself is not an exact science. Different statisticians may produce slightly different models using the same data. Ultimately, all models fall short of perfect accuracy in predicting outcome; even with the best, around 15–30% of outcomes may not be accounted for by the risk model. This suggests that there are other factors, besides those identified in the model, that play a role in determining some outcomes.

A final note of caution concerns the view that assessing outcomes is synonymous with assessing performance. This can be a very misleading assumption, particularly when it is taken further, ascribing all the blame for poor outcome to the surgeon. There are many things that can contribute to perioperative death that are beyond the control of the surgeon or the surgical team: a case arising in emergency circumstances, death due to co-morbidity, errors in the intensive care process. Risk stratification and outcomes assessment are relatively imprecise methods and results should certainly be treated with caution.

Acknowledgements

Figure 51.1 reproduced with kind permission from Stark, J. F., 'Databases for Congenital Heart Defects: Beginners Guide.' Annual in Cardiac Surgery. Edited by W. G. Williams. W. B. Saunders, Philadelphia. Copyright 2000.

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Index

Abdominal pain, 293
Abnormally open structures, closure of, 109–114
Absent pulmonary valve syndrome (APVS), 425
  categorization of, 425–426
  characterization of, 425
  complications, 431
  diagnosis of, 425–426
  lymphatic vessels, ligation, 427
  neonate with, 431
  plication/resection of pulmonary arterial wall, 428–429
  repair, preparation for, 427–428
  results, 431
  specific features of, 427
  surgery, indications for, 426–427
  surgical technique, 427–431
Accessory connections
  anterior septal, endocardial approach, 629–630
  left free wall
    endocardial approach, 627
    epicardial approach, 628
  posterior septal, 628
  right free wall
    endocardial approach, 629
    epicardial approach, 629
Accessory pathways, 621, 622, 630
ACE inhibitors. *See* Angiotensin-converting enzyme (ACE) inhibitors
Acidaemia, 97
  metabolic, 562
ACT. *See* Activated clotting time (ACT)
Activated clotting time (ACT), 174, 211, 234
  kaolin, 174
Acute endocarditis, 280
Acute rejection, 665–666
  symptomatic, 643
Acute renal failure (ARF), 221
  children with, 221
  hyperkalemia, 221
  management of, 221
    dopamine, 221
    frusemide, 221
    mannitol, 221
  occurrence of, 221
  renal replacement therapies, 221
Acute respiratory distress syndrome (ARDS), 215
Acute vessel rupture, 108
Acyanotic heart defects, 74–77
Adenosine, intravenous, 213
Adrenaline, 194, 421
  higher-dose, 205
Adrenergic agonists, 205
AEPC. *See* Association of European Paediatric Cardiologists (AEPC)
Aerobic gram-negative bacteria, 222
AFFIRM. *See* Atrial Fibrillation Follow-up Investigation of Rhythm Management (AFFIRM)
Airways
  assessment of, 313
  compression, 313
  with pulmonary artery sling, management of, 313–316
Albumin, 483
ALCAPA. *See* Anomalous origin of left coronary artery from pulmonary artery (ALCAPA)
Alfentanil, 217
Alpha Stat/pH Stat Mode, 179
α-adrenergic agonist, 191
α-2 adrenergic agonists, 217
α-adrenergic blocker, 208
α-adrenergic receptor, 208
α-agonist drugs, 402
Altered perfusion, 169
Alveolar ventilation, 93, 187
Ametop, 191
Amiodarone, 149, 213
  intravenous, 213
Amniocentesis, 63
Amphotericin, 666
Amplatzer duct occluder, 110
Amplatzer muscular VSD occluder (AMVSDO), 112
Amplatzer septal occluder (ASO), 110
Anaerobic threshold, 98

Surgery for Congenital Heart Defects, Third Edition. Edited by J. F. Stark, M. R. de Leval and V. T. Tsang
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Anaesthesia, 187–201, 402, 475
basic principles of, 187
Cardiac defects, 188–189
complex shunt, 188
dependent shunt, 188
with high pulmonary blood flow, 188
with low pulmonary blood flow, 189
obligatory shunt, 188
outflow tract obstruction, left ventricular, 189
cardiovascular stability, provides, 200
drugs, short-acting, 197
with high-dose narcotics, 402
induction of, 188, 192–193
inhalation , 189
intravenous, 189
maintenance of, 194
spinal, 199
systemic vascular resistance, 191
Anaesthetic, 94
Anaesthetic agents, 198
insoluble, 189
Anaesthetic considerations, 562–563
Anaesthetic drugs, 217
Anaesthetic implications, of cardiac pathophysiology, 190
Anaesthetist, 187, 188
Analgesia, 187, 197, 216–217
and sedation, balance of, 217
Analgesic drugs, 217
Anastomosis, 301, 316, 395
end-to-end, 287–289
end-to-side, 312
neopulmonary artery, 438
tissue-to-tissue, 417
Anastomotic obstruction, 304
Anastomotic stenosis, 301
Anastomotic stricture, 336
Aneurysm, 296, 603
intracardiac rupture of, 603
Aneurysmal, 138
Aneurysmal dilatation, 396, 400
Aneurysmal sac, 604
Angiocardiogram, 96, 388
Angiocardiography, 95, 96, 275, 327, 339, 374, 390, 575, 617
Angiographic delineation, 610
Angiography, 60, 91, 95–96, 116, 390, 412, 525
Angioplasty, 104–106, 268
Angiotensin-converting enzyme (ACE) inhibitors, 208
Annular dilatation, 578
Annular hypoplasia, 494
Annular valvuloplasty, 520
Annuloplasty, 376, 585
type of, 380
Annulus
of atrio-ventricular valve, 20
hypoplasia of, 576
Annulus plication, 580
Anomalous drainage, 347
from left lung into innominate vein, 351–352
Anomalous innominate artery, 307, 311
Anomalous left carotid artery, 307
Anomalous left coronary artery, 612
Anomalous origin of left coronary artery from pulmonary artery (ALCAPA), 612–617
diagnosis of, 612
aortocoronary connections, 613–616
indications for operation, 613
physiology of, 612
results of, 617
surgical techniques for, 613–617
Anomalous pulmonary vein, anastomosis to left atrium, 350–351
Anomalous systemic venous return
to both atria, 325
exceptional, 320
to pulmonary venous atrium, 322–325
anatomy, 322–323
surgical technique, 323–325
surgical repair of, results of, 326
to systemic venous atrium, 320–322
anatomy, 320
surgical situations, 321–322
surgical technique, 320–322
Ansa subclavia, 13
Anterior leaflet, chordae tendineae of, 577
Anterior muscular obstruction, 527
Anti-CMV immunoglobulin M (IgM-positive), 666
Antiarrhythmic drugs, patients dependent preoperatively on, 213
Antibiotic prophylaxis, 107, 192
for cardiac surgery, 192
Antibiotic therapy, 222, 650
Antibiotic treatment, 222
Antibiotic-resistant organisms, 222
Antibody induction therapy, 643
Anticoagulants, 219
Anticoagulation, 106
Anticoagulation therapy, 583
Antiproliferative agents, 643
Antithymocyte antibody, use of, 665
Antithymoctye/lymphocyte globulin, 643
Aorta
anterior translocation of, 466
branches of, 30
coronary sinuses of, 30
foetal, imaging of, 64
bypass cannulae in, 234
cannulation of, 444, 462
central pulmonary arteries anterior to, repositioning of, 430–431
cervical, 309, 311
coarctation of, 69, 76–77, 104–105, 261, 285–296
diagnosis of, 285–286
early complications of, 293–294
late complications of, 294–295
long-term follow-up of, 296
medical management of, 285–286
neonates, 199–200
older children, 200, 286
results of, 295–296
stent implant for, 108–109

surgical techniques in, 286–293
symptomatic infants, 285–286
velocity-encoded phase-contrast MR, 77
congenital narrowing of, 285
echocardiographic examination of, 69
mirror-image branching of, 308, 309
mobilization of, 301
and pulmonary artery
spatial relationship between, 642
sinotubular junction of, 489
suprasternal imaging, 560
Aortic annulus, 495
Aortic arch, 401, 459, 515, 659
anomalies of, fetal, imaging of, 65
hypoplasia of, 290
interruption of, 299–304
anatomy of, 299
diagnosis and medical management of, 299–300
midline approach, 300–302
reoperation, 304
repair of
results, 303–304
surgical technique, 300–303
type A, 301, 302
type B, 301, 302
repair of
through the left chest, 302–303
Aortic atresia, 67, 559, 560
arterial connection in, 8
Aortic cannula, 175, 196, 243, 493, 517, 574
Aortic clamp, 200
Aortic homograft, 396, 430, 464, 508
disadvantages of, 494
valve, insertion of, 495–497
Aortic hypoplasia, 197
Aortic incompetence, 367, 492
bicuspid valve assessment of, 77
Aortic isthmus, 108
tubular hypoplasia of, 287
Aortic needle vent, 447
Aortic orifice, 506
Aortic outflow tract, 121
Aortic pathology, 85
Aortic regurgitation, 493
Aortic root abscess, 498
Aortic root, opacification of, 390
Aortic root replacement, 498–502
homograft, 498
Aortic sinus defect, repair of, 480
Aortic stenosis
isolated, 560
neonatal, 491, 511
subvalvar, 491
supravalvar, 491
Aortic translocation, 466
Aortic valvar orifice, 134
Aortic valve, 299, 489, 493, 501, 506, 604
atresia, 559
annulus, hypoplasia of, 504
balloon dilatation of, 492
cusps, 356
dysfunction of, 489
homograft replacement of, 511
incompetence, 493, 606
leaflets of, 25–27
orifice, 493
overriding of, 407
prolapse of, 127
regurgitation, 490, 494
repair of, 511–512
in children, 494
replacement of, 494–495
heterograft/homograft, 511
mechanical, 511
prosthetic, 497–498
ring, 437
root replacement, 494–498
stenosis, 103–104, 491
in children, 511–512
congenital
balloon dilatation valvuloplasty for, 103
unicuspid, 493
Aortic valvotomy, 492, 493, 511
Aortic valvuloplasty, 103
Aortic–tricuspid continuity, 131
Aortico–left ventricular tunnel, 605–607
congenital anomalies of, 603–607
diagnosis of, 605–606
indications for, 606
results of, 606–607
surgical technique for, 606
Aortogram, 390
Aortography, 103, 401
Aortopulmonary (AP) window, 599–602
diagnosis of, 599
indications for, 599
with interrupted aortic arch, 601
results of, 601–602
surgical technique for, 599–601
Aortopulmonary blood flow, temporary balloon occlusion of 95
Aortopulmonary collaterals, 426
large, cyanotic patients with, 180
Aortotomy, 276
proximal transverse, 498
Aortotomy incision, 496, 506
Aortoventriculoplasty, 505–507
Apex
closure through, 369
Apical aneurysms, 369
Apical–aortic bypass, 509–510
Apical trabecular septum
defects in, 128
Aprotinin, 170, 194, 197, 211, 234, 420, 640, 649
high-dose, 211
use of, 174
APVS. *See* Absent pulmonary valve syndrome (APVS)
Arch anomalies, diagnosis of, 69
Arch hypoplasia, 69, 436
Arch replacement, complete, 641

Archiving echocardiography records, 40–41
digital, 41
advantages of, 41
ARDS. *See* Acute respiratory distress syndrome (ARDS)
Argyle chest tube, 453
Aristotle score, 684, 685
Arrhythmias, 97, 211, 352
ablation strategies of, 164–165
atrial, 148, 161, 162
catheter ablation of, 161–165
advanced mapping techniques of, 164
case selection and preparation of, 163
conditions of, 161–163
conventional contact mapping, 163–164
defects of, 163
electrophysiological study of, 163–164
surgical anatomy of, 163
Fontan, 165
management, 212
postoperative, 407
provocation, 98
supraventricular, 161, 622
surgery for, 619–631
electrophysiological evaluation, 622–623
indications for, 619
principle strategy of, 619
surgical anatomy, 619–622
surgical techniques, 623–631
triangle of Koch, 619
Arrhythmogenesis, 161, 369
Arrhythmogenic right ventricular dysplasia (ARVD), 622
Arterial cannula, 369
Arterial cannulation, 174–176
Arterial catheter, 286
Arterial connection, 9
modes of, 9
types of, 9
Arterial oxygen saturation, 460, 461
Arterial switch, 473
diagnosis of, 474
management methodology for, 474–476
perfusion, 475–476
postoperative management of, 482–483
preoperative preparation of, 474–475
results of, 483–486
indication 1, 483–484
indication 2, 484–485
indication 3, 485–486
indication 4 and 5, 486
indication 6, 486
surgical
exposure of, 475
technique for, 476–482
and ventricular septal defect (VSD) closure, 438
Arterial switch operation (ASO), 81, 441, 443, 459, 472–486, 528
complication of, 80
indications for, 472–474
Arterial switch procedure, 531
Arterial valve, anatomy of abnormalities 20
Arteriography, 95
Arterioscelorotic lesions, dilatation of, 101
Artificial chordae, 596
ARVD. *See* Arrhythmogenic right ventricular dysplasia (ARVD)
ASD. *See* Atrial septal defect (ASD)
ASO. *See* Arterial switch operation (ASO)
Asplenia, 4
polysplenia syndrome, 327
syndrome, 319
Association of European Paediatric Cardiologists (AEPC), 681
Atelectasis, 214, 216, 310, 659, 663
Atherosclerotic aorta, 171
Atria, 48
Atrial appendage, 319
left, anatomy of, 4
right atrium and, junction between, 15–19
right, anatomy of, 4
Atrial cannula, 330
Atrial conduction, 619, 626
Atrial diverticula, 675
Atrial dysrhythmias, 377
Atrial fibrillation (AF), 161, 165, 593, 623–627
catheter ablation for, 623
drug therapy for, 623
indications for, 623–624
postoperative medications, management of, 627
stroke prevention in, 623
surgical therapy for
Cox–Maze procedure, 624
indications for, 624
Atrial Fibrillation Follow-up Investigation of Rhythm Management (AFFIRM), 623
Atrial flutter, 161, 213
Atrial isomerism, 319–320, 374
left, 319
right, 319
Atrial lead insertion, 151
Atrial morphology, 641
Atrial myocardial fibres, 625
Atrial myocardium, right, 18
Atrial myxoma, 674
Atrial pacing, 214
Atrial patch, 447
Atrial sensing, bipolar leads, 148
Atrial septal baffle, 630
Atrial septal defect (ASD), 74–75, 110–112, 164, 322, 324, 381, 391, 451, 626
closure of, 378
device closure of, 353
direct closure, 345
evaluation of, 75
management of, 75
patch closure, 346–347
quantification of left-to-right (L–R) shunts, 75
residual, 352
results of, 352
secundum, 343–353
surgical techniques, 345–352

trans-catheter techniques, 75
types of, 344
Atrial septal defect (ASD) anatomy, 75
Atrial septectomy, 268, 271–274, 462, 467
indications for, 271
results of, 274
surgical techniques in, 271–273
Atrial septum, 48, 106, 322, 330, 374, 452
bowed, 560
Atrial situs, 139, 525
abnormalities of, 435
assessment of, 5
morphology and, 4–5
discordant, 533
and venous connections, 41–44
and ventricular apex, discordance between, 532
Atrial stay sutures, 445
Atrial switch, 321
Atrial systole, 213
Atrial tachycardias, 161, 621
ectopic focus, 630
Atrial thrombi, 675
Atrial transport function, 623
Atriopulmonary anastomosis, 164
Atriopulmonary connection, 321
Atrioseptostomy, 106
Atriotomy, 161, 273, 446
Atrio-ventricular block, 214
Atrio-ventricular bundle, anatomic course of, 29
Atrio-ventricular canal defect, 126
Atrio-ventricular concordance, 357
Atrio-ventricular conduction, 134, 212
loss of, 214
Atrio-ventricular connection, 44–45
absent, 8
modes of, 8
types of, 8
accessory
in infants and children, 627–630
electrophysiological evaluation, 627
indications for, 627
surgical technique, 627–630
biventricular, 44
concordant, 44
discordant, 44
univentricular, 44
Atrio-ventricular discordance
atrial repair of, 526
conduction system, position of, 537
and congenitally corrected transposition, 525–540
associated defects, 525
diagnosis of, 525–526
indications of, 526
with double-outlet left ventricle (DOLV), 537
with double-outlet right ventricle (DORV), 537
double-switch approach, 538
heart block, risk of, 538
physiological repair of, 538
results, 538–540
situs solitus/situs inversus ,associated with, 525
with ventriculo-arterial discordance, 525, 538
Atrio-ventricular groove, 357, 620, 621
heart, sectioning of, 619
Atrio-ventricular junction, 28
Atrio-ventricular nodal re-entry tachycardia (AVNRT), 161
Atrio-ventricular node 17, 18, 121
artery, 31
dysfunction, 214
Atrio-ventricular re-entry tachycardia (AVRT), 161, 213
Atrio-ventricular septal defect (AVSD), 66, 321, 373–385, 560, 579, 626
cardiac anomalies, associated with, 374
complete form, 373, 374, 377–385
classification of, 373
complications, 384–385
indications of, 377–378
results of, 384
superior vena cava, bilateral, 374
surgical technique, 378–384
type A, 373
type C, 373
diagnosis of, 374
intermediate form, 374, 383
late survival, 378
ostium primum type (partial AVSD), 374
partial form, 373–377
complications of, 377
indications of, 374–375
results of, 377
surgical technique, 375–377
subdivisions, 373–374
with atrial fibrillation (AF) 626–627
Atrio-ventricular septum, 48
in normal hearts, muscular, 17, 23–24, 27
Atrio-ventricular synchrony, 145, 148, 213
Atrio-ventricular valve, 35, 50
abnormalities of
anatomy of, 20
straddling, 5–6
overriding vs., 6
common, 373
leaflets, 373
position of, 374, 379
Rastelli types, 379
regurgitation, 377
regurgitation, left, 377
straddling of, 139–141, 525
Atrio-ventricular valve orifice, 385
Atrio-ventricular valve rings, 35
Atrium(a), anomalous venous return to
left, anatomy of, 19
components of, 19
surgical access to, 19
morphology and, 4–5
right, anatomy of, 15–19
atrial appendage and, junction between, 15–19
components of, 15
floor of, 15, 17
Autograft, harvesting of, 500
Autograft root replacement, pulmonary, 499–500

AVRT. *See* Atrio-ventricular re-entry tachycardia (AVRT)
Azathioprine, 643, 650
Azygos vein, 319, 430, 659
  orifice of, 327
Azygos/hemi-azygos continuation, 319, 320, 321, 325

Balloon angioplasty, 106, 108, 109, 304
  use of, 286
Balloon atrial septostomy (BAS), 268, 271, 389, 436, 451, 467, 473, 561
Balloon catheter, 103
Balloon dilatation, 101, 316, 390, 397, 402, 456
Balloon valvuloplasty, 490
  trans-catheter, 391
Barium oesophagogram, 53
Barlow's syndrome, 580
Barotrauma, 638, 661
BAS. *See* Balloon atrial septostomy (BAS)
BCPS. *See* Bidirectional cavopulmonary shunt (BCPS)
Benign tumour, 671, 674
β-adrenergic agonists, 198
β-adrenergic blocking agent, 189
β-adrenergic drugs, 208
β-blockade, 209
  intravenous, 213
β-blocker, 638
β-blocking agent, 406
Benzodiazepine, 217
  weaning of, 217
Bi-atrial incision, 330, 675
Bicarbonate, 171
Bicaval anastomosis, 639, 640
Bicaval cannulation, 176–178, 356, 427, 438, 459, 500
  direct, 177
  indirect, 177
Bicaval venous return, 445
Bicuspid aortic valve, 285
Bicuspid valve, 392, 489
Bidirectional cavopulmonary shunt (BCPS), 597
Bifurcated homograft, 430
Bifurcation, 429
Bilateral infundibulum, 134, 435
Bilateral submammary incision, 345
Bilateral thoracotomy, 246
  incision, 239
Biocompatible tubing, development of, 236
Biological emboli, 171
Biomedicus cannulae, 239
Bioprosthesis, 583
Bioprosthetic valve, 494
Biopsy-negative rejection, 643
Bipolar leads, 148
Bipolar pacing, 151
Bipolar system, 149
Biventricular circulation, 390
Biventricular hypertrophy, 599
Biventricular pacing (Cardiac resynchronization therapy), 154–156
Biventricular repair, 397
Blalock–Hanlon septectomy, 271, 274, 472, 484
Blalock–Taussig shunt, 58, 95, 114, 183, 194, 199, 200, 235, 243, 251, 390, 392, 484
  classic, 251–253, 256, 402, 413
  closure of functioning, 253–254
  modified
    via lateral thoracotomy, 256–257
    via median sternotomy, 257–258
  obstruction of, 82
  right-sided, 253
Bleeding, 241, 243, 282, 293, 482
  and blood transfusion, 211
  management of, 234
  to reduce, 197
Block conduction, 619
  cut-and-sew technique, 619
Blockers, 209
Blood cardioplegia, 195
Blood drains, 172
Blood gas
  management of, 179–180
  measurements of, 93–94
Blood loss, 97
Blood oxygen, saturation measurements, 93
Blood pumps, 173
  centrifugal, 173
Blunt dissection, 153
Bolus steroid therapy, 665
Bovine jugular venous valve, 115
Bovine pericardium, 392, 414, 430, 460
Brachiocephalic artery, 7
Brachiocephalic vein, 13
Bradyarrhythmias
  and pacing, postoperative, 214
  perioperative, 162
Bradycardia, 148, 194, 200
  and pacing
    postoperative, 214
Branch pulmonary artery stenosis, 78, 82
Bronchi, bilateral short eparterial, 319
Bronchial arrangement, 319
Bronchial compression, 427
Bronchial rings, 315
Bronchial situs, 5
Bronchial stenting, 431
Bronchioalveolar lavage, 665
Bronchiolitis obliterans, 650, 653, 664, 666, 667
  developing, risk of, 666
Bronchiolitis obliterans syndrome, 666
Bronchopulmonary lavage, 666
Bronchoscopy, 313, 316, 646, 663
  fibre-optic, 313, 316
  virtual, 313
Bronchus, 15
Brugada syndrome, 98, 159
Bundle of His, 399, 525, 621
Bupivacaine, 199
Burkholderia cepacia, 646
Bypass, 301, 304, 377, 444, 647
  grafting, 643

Calcineurin inhibitors, 642–644
Calcium, 220
Caloric requirements
  critically ill children, 218
  modifications to, 218
Calories, recommended daily intake (RDI), 218
Cannula, 402
  near-occlusive, 194
Cannulation, 174–182, 233–234, 242–243, 475
  arterial, 174–176
  bicaval, 176–178
  direct, 175, 182, 320
  indirect, 182
  other techniques, 182
  retrograde, 176
  venting, 178–179
Cannulation sutures, 427
Capillary leak, 180, 211
  magnitude of, 220
Capillary leak syndrome, 211, 220
Cardiac abnormalities
  diagnosing, echocardiographic method, 66
  prenatal series, chromosomal defects, associated with, 69
Cardiac allograft vasculopathy, 643–644
  incidence of, 644
Cardiac anatomy, 85–86
Cardiac arrest
  congenital cardiac disease 210
Cardiac catheterization, 60, 73, 91, 96, 97, 254, 275, 286, 294, 327, 339, 343, 349, 355, 374, 390, 413, 460, 525, 609, 672
  with projectional angiography, 84
Cardiac compression, rate, 210
Cardiac ejection, 175
Cardiac failure, 584
Cardiac function
  impaired, 204
  ventilatory manipulation of, 216
Cardiac index, 205
  arterial switch operations (ASO), 205
  fall in, 205
Cardiac inflow stasis, 167
Cardiac malformations, 66–69
Cardiac mechanical support, aims of, 229
Cardiac MR, 75–77, 80, 83
Cardiac output, 37
Cardiac position, nomenclature and, 9
Cardiac rhythm, 449
Cardiac segments, connections between, 8
  identification of, 4–7
Cardiac skeleton, 29
  atrio-ventricular part of, 29
  central fibrous body and, 29
  fibrous trigone and, 29
Cardiac surgery, 168
  preoperative diagnosis, 203
  risk stratification, 683
Cardiac tamponade, 58, 204, 210, 671
  treatment of, 671
Cardiac teratomas, 69
Cardiac transplantation, 322
Cardiac tumours, 69, 622
Cardiology, interventional, 421
Cardiomegaly, 275, 286, 459, 560, 593
Cardiomyopathy, 155, 231, 233, 236, 635
  congestive, 491
  dilated, 612, 635
  restrictive, 635
Cardiomyoplasty, 588
Cardioplegia, 476, 494, 520, 606
  cardioplegic solution, 321, 375, 462
  blood, 195
  infusion of, 427
Cardiopulmonary bypass (CPB), 167, 194–196, 198, 232, 239, 243, 268, 271, 276, 328, 330, 349, 351, 402, 406, 445, 476, 481, 509, 574, 599, 610, 613
  avoiding of, 116
  bleeding, 196
  cannulation for, 320–321
  conduct of, 174
  conventional hypothermia, 356
  extracorporeal circuit, 171–174
  hypothermic, 366, 392, 494, 498
  importance of pulsatility during, 169
  initiation of, 169, 170
  low-flow, 180–181
  and myocardial protection, 594–595
    assessment of repair, 594
    indications for, 594–595
  in neonates, 167
  with normothermia, 253
  pathophysiology of, 167–171
  pump flow rates during, 169
  special circumstances of, 182–183
  surgical aspects of, 174
Cardiopulmonary exercise testing
  in children and adolescents, 98–99
    limitations of, 98
    potential impact of, 98
Cardiopulmonary resuscitation, 210–211
CardioSEAL, 109
Cardiotomy suction, 170
Cardiovascular MDCT, 87
  indications for, 84–85
  non-cardiac gated, 87
Cardiovascular MR imaging, 87
Cardiovascular system, monitoring, 203–204
Carotid artery, 301
Carpentier technique, 595–597
Castaneda clamp, 289, 314
  side-biting, 314
Catecholamine
  higher dose, 205
  preoperative use of, 561
  receptors, 187, 192
Catheter
  displacement, 97
  heparin-bonded, 204
  placement, 94–95
  pressure-monitoring, 566
  radiofrequency ablation, 627

Catheter ablation
complications of, 165
therapy, 630
Causative organisms, 644
Caval cannula, 196
Caval cannulation, 375
Caval pathway, 453
Caval snares, 464
Cavopulmonary anastomoses, 259, 262, 321–323, 462
bidirectional, 391, 546
bilateral, bidirectional, 321
Cavopulmonary connection
association of, 597
bidirectional, 397
total, 325
Cavopulmonary shunt, bidirectional, 527
CBF. *See* Cerebral blood flow (CBF)
CCAD. *See* Central Cardiac Audit Database (CCAD)
ccTGA. *See* Congenitally corrected transposition of the great arteries (ccTGA)
Cell-saver, 640
Central Cardiac Audit Database (CCAD), 683
Central fibrous body, 15
cardiac skeleton and, 29
Central pulmonary arteries
resection of, 429–430
Central venous catheter, 402
Central venous pressure, end-diastolic pressure 569
Centrifugal blood pumps, 173
Cephalic vein, 152
Cephalosporin, 222
Cerebral autoregulation, mechanism of, 169
Cerebral blood flow (CBF), 168
maintenance of, 223
Cerebral embolism, 323
Cerebral metabolism,
cervical aorta, 309, 311
at low temperatures 195
Chest closure, 482
Chest X-ray, 355
Cholesterol pericarditis, 672
Chordae tendineae, 20, 373, 379, 670
absence of, 580
basal, 20
cleft, 20
elongation of, 376, 580–581
in imperforate valve recognition, 8
shortening of, 376
Chordal fenestration, 575
Chorion villus sampling, 63
Chromosomal anomalies, 70
Chronic hypoxaemia, 189
Chronic pulmonary regurgitation, 161
Chronic rejection, 666
Chyle, 219
chylopericardium, 672
chylothorax, 219, 282, 294, 427, 672
initial management of, 219
chylous effusion
diagnosis of, 219
pleural, 219
chylous leak, 294
Circular annuloplasty, 368
Circulatory arrest, 176, 180, 300, 320, 328, 427, 475, 566
in deep hypothermia, 271
hypothermic, 171, 180–181, 331
optimal temperature for, 180
Circulatory failure
due to myocarditis/cardiomyopathy, 232–233
postcardiotomy, 232
Circumflex artery, 31
Citrate phosphatedextrose (CPD), 169
Cleft, 373
Cleft leaflet, 578–579
anterior, 579
posterior, 580
Closed cardiac operations, 199
Closed system, 173
CMV. *See* Cytomegalovirus (CMV)
Coagulation screen, 211
Coagulopathy, 168, 201, 642
Coarctation, 436, 459
aortic, cardiac MR evaluation of, 76
children undergoing, 204
isolated, 285–286
and ventricular septal defect (VSD), 286
of aorta, 69, 76–77, 104–105, 261, 285–296
diagnosis of, 285–286
early complications of, 293–294
late complications of, 294–295
long-term follow-up of, 296
medical management of, 285–286
neonates, 199–200
older children, 200, 286
results of, 295–296
stent implant for, 108–109
surgical techniques in, 286–293
symptomatic infants, 285–286
velocity-encoded phase-contrast MR, 77
Cold cardioplegic myocardial protection, 378
Collaterals
anastomoses, 418
mobilization of, 417, 418
Colour Doppler echocardiography, 355
Colour-flow Doppler echocardiography, 612
Colour-flow Doppler interrogation, 35
Combined anomalies of pulmonary and systemic venous return, 325–326
Commissural fusion, 387, 581, 575
Commissurotomy, 493, 511
Common atrium, 374
Complete arch replacement, 642
Complex transposition, surgery for, 459–468
Weaning from ventilator,
complex, 215
routine, 215
Computed tomography (CT)
congenital heart defects, 84–86
high-resolution of, 313

Concealed accessory pathways, 630
Concomitant revascularization, 617
Concordant ventriculo-arterial connections, 139
Conduction block, 626
Conduction pathways, 619
Conduction system
  atrio-ventricular, bundle branches and, 29–30
    interatrial, 19
  disposition of
    basic rule, 121–122
Conduction tissue, 134, 141, 376, 378
Conduit, 464, 467, 530
  insertion, 537
  intrapericardial, 419
  obstruction, 79
  orientation, 464
  pulmonary homograft, 414
  stenosis, 81
  ventricular end of, 467
Congenital aneurysm, rupture of, 603
Congenital aortic stenosis, balloon valvuloplasty of, 104
Congenital cardiac anomaly, 338
Congenital coronary artery fistula, 609–612
  diagnosis of, 609–610
  ligation/division, 610
  results of, 612
  surgical techniques for, 610–612
Congenital diverticula, 675
Congenital heart defects, 38
  complex, technical aspects in, 640–642
  computed tomography (CT), 84–86
  echocardiography, 73
  gradient-echo imaging, 74
    three-dimensional, 74
    two-dimensional, 74
  interventional catheterization in, 101–117
    indications of, 101
  magnetic resonance imaging (MRI), 73–84
    velocity-encoded Phase contrast MR, 74
  spin-echo imaging, 73–74
Congenital heart disease, 70, 93, 98, 146, 152, 156, 161, 635
  cyanotic, 251
Congenital heart lesion, 94, 95
Congenital heart surgery
  assessment of performance in, 679–688
  data collection in, 679–688
    golden rules of, 679–680
    issues associated with, 679–683
  outcomes
    assessment of, 685–687
    graphical methods for assessment, 686–687
  risk modelling
    limitations of, 687–688
  risk stratification, 683–685
    attempts for, 685
    rationale for, 683–684
Congenital mitral valve anomalies
  in newborns and infants, 583–585
    anatomy of, 584
    clinical presentation of, 584
    investigations of, 584
    medical treatment for, 584
    special considerations of, 583–584
    surgical treatment for, 584–585
Congenital mitral valve stenosis, with normal papillary muscles, 575
Congenital stenosis, of pulmonary veins, 336
Congenital valvular diseases, valvuloplasty for, 101–104
Congenitally corrected transposition, 136
Congenitally corrected transposition of the great arteries (ccTGA), 81–82, 525
  with situs inversus (IDD) configuration, 528, 530, 531
  with situs solitus (SLL) configuration, 528, 530, 531
  surgery for, 526–537
Congestive heart failure, 275, 285, 327, 375, 491, 516, 609
  intractable, 355
Conotruncal malseptation, 435
Constrictive pericarditis, 672
  treatment of, 672
Contegra bovine jugular vein graft, 430
  Contegra, 421
Continuous positive airway pressure (CPAP), 215
Continuous sutures, combined with interrupted sutures, 365
Contralateral lung, 663
Conventional ultrafiltration (CUF), 183
Cor triatriatum, 338–340
  diagnosis of, 338–339
  indications for, 339
  results of, 339
  surgical techniques, 339
Core cooling, 208
Coronary anatomy, 459
Coronary artery 53–54
  anatomy of, 30–32
    anterior descending, 31
    blood supply of, 32
    branches of, 31–32
    dominant, 31
    epicardial, 32
    main stem of, 32
    major branches of, 32
    orifices of, 31
    origin of, 7
    right, branches of, 31
  bypass grafting, 615
  fistula, 388
    congenital, 609–612
  fistula murmur, 609
  left anterior, descending , 400
  origination, 559
  reimplantation of, 478–479, 494, 613
  relocating, 441
  translocation, 477–479, 483
  Yacoub type B anatomy, 478
Coronary artery stenosis, 80
Coronary circulation, 388
  anatomy of, 30–32
Coronary MR angiography, 81

Coronary ostia, 477, 520
excision of, 477–478
Coronary sinus, 16, 19, 155, 319, 376, 446, 453, 529
atrial septal defect, 324
cannulation of, 320
cutting back of, 445
flap, 452
orifice, 321, 597
atresia, 320
ostium, 165
unroofed, 322
Coronary sinus atrial septal defect, repair of, 347
Coronary vein(s), 32
Corticosteroids, 642
long-term use of, 643
use of, 170
Cox–Maze III procedure, 624
atrial incisions of, 624
surgical technique of, 624–626
Cox–Maze procedure, modifications of, 624
CPAP. *See* Continuous positive airway pressure (CPAP)
CPD. *See* Citrate phosphatedextrose (CPD)
Crista supraventricularis, 6, 21–24, 122
Crista terminalis, 16, 165, 319, 445, 532
Critical aortic stenosis, symptomatic neonates and infants with, 492
Critical pulmonary stenosis/pulmonary atresia, diagnosis of, 389
Crouzon syndrome, 425
Crux cordis, 376, 378
Cryoablation, 619, 630
Cryolesion, 625
Cryopreserved homograft, 495
Cryoprobe, 625
Cryosurgery, 630
Crystalloid cardioplegia, 181
Crystalloid solutions, 211
CUF. *See* Conventional ultrafiltration (CUF)
Cusp
detachment of, 392
mobility, 493
Cyanosis, 168, 189, 310, 322, 374, 381, 389, 401, 461, 527, 560
long-standing, patients with, 401
Cyanotic ''spells'', 401
Cyanotic heart defects, 77–83
Cyclosporine, 642, 644
Cystic fibrosis, 646, 650, 657, 666
patients with, 646
Cystic hygroma, association of, 69
Cytomegalovirus (CMV), 666
pneumonia, 644

Dacron conduit, 395
woven, 530
Dacron grafts, 615
Dacron patch, 292, 378, 380–384, 404, 505
stretch-knitted, 537
ventricular, 379
Dacron pledgets, 437
Dacron velour, patch of, 365
Damus–Kaye–Stansel (DKS) procedure, 263
Data collection
errors in, 682
existing systems, 683
nomenclature and mapping, 681–682
related to CHD
issues associated with, 680–681
purposes for, 680
Database validation, 682–683
De-airing, 463
Debanding, 268, 356
Decannulation, 226
Deep hypothermia, 436
Deep hypothermic circulatory arrest (DHCA), 180, 328
Defibrillators, implantable, 158–159
Deformation, 578
q11 deletion syndrome,D
220, 299
Deltopectoral groove, 152
Denis Browne dissector, 240, 242, 277
Dexamethasone, 201
Dextran, 171
Dextrocardia, 41, 443, 539
Dextrose, 196
DHCA. *See* Deep hypothermic circulatory arrest (DHCA)
Diabetes mellitus, maternal, as risk factor for CHD, 63
Diaphragmatic paralysis, 427
Diastole, 178
Diastolic pulmonary regurgitation, 35
Diathermy, 223, 226, 241, 252, 281, 427, 443
Diffuse left ventricular outflow tract, obstruction, 504–510
DiGeorge's syndrome, 299, 516
Digital subtraction angiography (DSA), 95
Dilatation, 68
Dilated cardiomyopathy, with epicardial pacing, 155
Direct cannulation, 175, 182
Discordant atrio-ventricular connections, 136, 140
Dissection, 223
Distal aorta, to the undersurface of aortic arch
anastomosis of, 291
extended resection of, 291
Distal conduit, anastomosis, 531
Diuretics, 197, 209
Diverticula, 675–677
atrial, 675
congenital, 675
ventricular, 676
DLP 10 Fr gauge vent, 428
DLP arterial cannula, 175
Dobutamine, 421
DOLV. *See* Double-outlet left ventricle (DOLV)
Domperidone, 219
Donor,
harvest, 658
left lower lobectomy, 661
lobe, 661
operation, goal of, 660
organs, 656

right lobectomy, 660–661
recipient match, immunological evaluation for, 637
Dopamine, 196, 221, 526, 664
low-dose, 205
Dopamine infusion, 263, 482
Doppler,
colour-flow mapping, 310
echocardiography, 74, 83, 294, 374, 617
benefits of, 37
flow echocardiography, 33–34
imaging, 37, 156
tissue, 35
interrogation, colour-flow, 35
shift, 34
signals, 33, 35
system
continuous-wave, 34
pulsed, 34
Doppler waveforms, 34
Double aortic arch, 307, 310–311, 313
Double mitral orifice, 575
Double-inlet univentricular heart, 473
Double-inlet ventricle, 262–263, 267–268
Double-outlet left ventricle (DOLV), 442
Double-outlet right ventricle (DORV), 82, 134, 435–442
with anterior aorta, 435
atrio-ventricular septal defect, associated with, 441
complications, 441
diagnosis of, 435–436
Fallot's-type, 82
great arteries, origin of, 67
haemodynamic situation in, 435
indications of, 436
interventricular communication in, 133–136
malformations, 435
and pulmonary stenosis, patients with, 436
results, 441–442
surgical technique, 436–441
trans-ventricular approach, 436
with ventricular septal defect (VSD) 439–441
doubly-committed, 438
with pulmonary stenosis, 437–438
without pulmonary stenosis, 436–437
subpulmonary, 438–439
uncommitted, 441
Double switch, 538–539
Doubly-committed juxta-arterial defects, 127, 132
Down's syndrome, 407
Drug-eluting stents, 644
DSA. *See* Digital subtraction angiography (DSA)
Dual-chamber pacing, 151
Ductus arteriosus, 52, 204, 285, 287, 288, 301, 307, 308, 311, 390, 401, 412, 425, 427, 474, 562
bilateral, 307
left-sided, 308
ligation, 276
occluder coils, 109
patency, 198
thoracic aorta continuum, 559
Dye dilution, 94
Dynamic infundibular stenosis, 102
Dyspnoea, 584, 609
Dysrhythmia, 187, 196, 448

EACTS. *See* European Association for Cardiothoracic Surgery (EACTS)
Ebstein's anomaly, 68, 526, 591–594, 624
categorization of, 592–593
characteristic features of, 592
in children and teenagers, 594
general operative considerations in, 594
malformations of, 591–598
in neonates and infants, 593–594
presentation and medical management of, 593–594
surgical anatomy of, 592–593
in young adults, 594
Ebstenoid-type abnormality, 527
ECG-gated MDCT, 86
Echo Doppler interrogation, 355
Echocardiography, 33–60, 73, 116, 199, 300, 313, 389, 575
archiving, 40–41
atrial situs, and venous connections, 41–44
atrio-ventricular connection, 44–45
basic principles of, 33–37
conventional, 38
Doppler, 275
flow, 33–34
echocardiogram, 204
epicardial, 54
foetal, 63–70, 425
heart malformations, 70
implications of, 69–70
normal foetal cardiac anatomy in, 64–66
organization of, 63–64
intraoperative, 54–57
M-mode, 33
oesophageal, 75
postoperative assessment of, 57–60
practical considerations of, 37–41
quantitative measurement of, 35–37
sequential segmental anatomy, 41–54
serial, 50
single-line, 33
three-dimensional, 35
trans-oesophageal, 38–40, 54, 77, 537, 663
trans-thoracic, 77, 80
transducer, 33
two-dimensional (2D), 33, 275, 310
use of, limiting factor , 643
ventriculo-arterial connection, 45–48
ECMO. *See* Extracorporeal membrane oxygenation (ECMO)
Ectopia cordis, 676
Ectopic foci cluster, 622
Ectopic focus, localization of, 622
Eisenmenger syndrome, 343, 646, 650, 655
Elective generator replacement, 158
Elective surgery, 584
Electroanatomic mapping system, 623
Electrocardiogram, 214, 374, 560, 612

Electrocardiography, 401
Electrode dislodgement, 156
Electrolytes, 220
  calcium, 220
  magnesium, 220
  potassium, 220
Electronic inversion, 38
ELSO. *See* Extracorporeal Life Support Organization (ELSO)
Emboli, 171
  biological, 171
  gaseous, 171
  intravascular, 171
  non-biological, 171
Emphysema, 310
End-stage heart failure, 646
End-stage lung disease, 667
Endocardial fibroelastosis, 491, 492, 560
Endocardial implantation, 148–153
Endocardial lead extraction, 157
Endocardial mapping, 630
Endocarditis, subacute bacterial, 276
Endoscopic approach, 247
Endotracheal tube
  tip of, 214
  uncuffed, 214
Endovascular stents, 106–109
  general considerations of, 106–107
Enteral feeding, 218
Epicardial cryoablation, 630
Epicardial implantation, 148, 153–154
  in children, indications of, 149
Epicardial leads, 157
Epicardial pacing wires, 212
Epicardium, anatomy of, 11
Epidural analgesia, 194
Epidural catheters, 660
Epinephrine, 664
Epstein–Barr virus, 644
Erosion, 157
Erythromycin, 219
Esmolol, 213, 406, 638
Ethibond synthetic sutures, 243
Etomidate, 193
European Association for Cardiothoracic Surgery (EACTS), 681, 683
Euroscore, 683
Eustachian valve, 17, 177, 456, 532
Everolimus, 644
Excessive bleeding, 234
Exomphalos, as risk factor for CHD, 63
Extended aortic root replacement, with aortic homograft, 507–509
Extracardiac anomalies, 70
  foetal, 63
Extracardiac valved conduit, 59, 509, 519, 526
Extracorporeal cardiopulmonary resuscitation (ECPR), 233
Extracorporeal centrifugal VAD, vs. extracorporeal membrane oxygenation (ECMO), 230
Extracorporeal circuit, 171–174
  prime, 171–172
Extracorporeal circulation, 169
Extracorporeal Life Support Organization (ELSO), 229
Extracorporeal membrane oxygenation (ECMO), 173, 229, 230
  determining adequacy of, 234–235
  management in patients with systemic–pulmonary shunt, 235
  veno-arterial, 230
  veno-venous, 230
  weaning from, 235
Extubation, 197

Facial dysmorphism, 516
Fallot's tetralogy, 67, 208
  prenatal life, 67
Fast-tracking, 197
Fatigue, 609
Femoral arterial sheath, 103
Femoral cannulation, 349
Femoral pulses, 299
Femoro-femoral bypass, 197
Fentanyl, 193, 194, 198, 217
  High doses of, 191
FFP. *See* Fresh frozen plasma (FFP)
Fibre-optic bronchoscopy, 660
Fibrinolysis, 104, 196
Fibroma, 674
Fibromuscular,
  hyperplasia, 388
  hypertrophy, 491
  obstruction, 467
  stenosis, 503–504
Fibrous tissue, 49
  circumferential ridge of, 575
Fick principle, 94, 205
Finochietto retractors, 246
Fistula, 603
Flecainide, 213
Flow metabolism coupling, 168
FlowSeal, 196
Fluid management, postoperative, 219–220
Fluid requirements, 220
Fluid therapy, 211
  volume replacement, 211
Foetal,
  arrhythmia
    bradycardia, 63
    as risk factor for CHD, 63
  cardiac anomaly, 63
  channels, 197
  echocardiogram, 570
  echocardiography, 425
  hydrops, 69
    nonimmune, as risk factor for CHD, 63
Foetal karyotype, 66
Foley catheter, balloon-tipped, 182
Fontan,
  circulation, 208, 216
  completion, 101, 568

operation, 162–163, 321–322, 441
procedure, 83
extracardiac conduit, 631
modified, 567–569
Fontan-type procedure, 195
Foramen ovale, 377, 389
flap, 64
Fossa ovalis, 16, 178, 272, 332
inferior margin of, 17
Fresh frozen plasma (FFP), 196, 201
Frusemide, 209, 221, 483
continuous infusions of, 209
Functional single ventricle, 162–163
Fungal septicaemia, 218

Gadolinium-enhanced MR angiography (Gd-MRA), 74, 76
drawback, 74
pulmonary venous abnormalities, detecting, 75
Gancyclovir, 644, 666
Gaseous emboli and gaseous microemboli, 171, 173
Gastric promotilant drug, 219
Gastroduodenal paralysis, 218
Gastrointestinal bleeding, management of, 219
Gastrointestinal system, 217–219
Gd-MRA. *See* Gadolinium-enhanced MR angiography (Gd-MRA)
Gelofusin, 171, 211
Generator
dual-chamber, 148
placement, 154
sub-xiphoid approach, 154
suprarenal approach, 154
pocket, 153
single-chamber, 148
Generic pacemaker code
NASPE/BPEG, 147
Genesis stents, 106
Gianturco, 109
Glenn shunt, 82, 198
bidirectional, 85, 117, 263, 267
Classical Glenn procedure, 391
visualization of, 83
Glomerular filtration, 168
Glyceryl trinitrate (GTN), 195, 208
Gore-Tex, 316
chordae, 580, 585
patch, 292, 392, 395, 404, 449, 460
shunt, 200
closure of, 259
sleeve, 157
tube, 440
Gradient-echo imaging, 74
three-dimensional, 74
two-dimensional, 74
Graft
dysfunction, 664
ischaemic time, 667
preparation, 659–660
Granulation tissue, risk of, 316
Great arteries, 51–53
anatomic relationships of, morphology of, 7
closure through, 366–368
double-outlet right ventricle, 67
transposition of, 67, 162, 165
Great vessels, anatomic relationships of, 12
Gross malalignment, 132
Grown-up congenital heart disease (GUCH), 73, 84
GTN. *See* Glyceryl trinitrate (GTN)
Guanylate cyclase, 197
GUCH. *See* Grown-up congenital heart disease (GUCH)

H_2 receptor antagonist, 219
Haematocrit, 189, 211
Haemodialysis, 221
Haemodilution, 169–171
rationale for, 169
Haemodynamic measurement, 94–95
Haemodynamic stability, 642
Haemofiltration (HF), 203, 221
Haemoglobinuria, 197
Haemolysis, 179, 441
Haemorrhage, 219, 642
Haemostasis, 243, 474, 502
Haemothorax, 116
Hammock valve, 577, 582
Hand ventilation, 210
Hand-held mobile mapping electrode, 623
Hanley's protocol, 389
Heart
acute margin of, 11
anatomic relationships of, 9, 13–15
connection, abnormalities of, 66–69
crux of, 5
with discordant atrio-ventricular connections, features of, 137
with doubly-committed defects, 134
fetal, chamber identification in, four-chamber view of, 64
fibrous skeleton of, 620
foetal, chamber identification in, 64
normal anatomy of, 64–66
heterotaxy-type, 530
lymphatic network of, 32
normal morphogenesis of, 4
with pinhole pulmonary stenosis, 387
sequential localization of, 4
shape of, 11
single outlet, 9
straddling atrio-ventricular valves 139–141
left, 140–141
right, 141
with straddling tricuspid valves, 141
thoracic location of, 11
univentricular, 198
Heart transplantation, 231, 236, 562, 635–644
ABO Mismatch, 637
acute rejection
surveillance and treatment of, 643
treatment of, 643

Heart transplantation, (*continued*)
for congenital heart diseases, complex, 639
contradictions, 636
donor considerations, 638
donor heart, 639
excision, 638–640
ideal, 638
preparation of, 639
heart block, 114, 363, 370, 377, 408, 538
late survival, 644
indications for, 635–636
infant transplantation, 637
infection, 644
morbidity, 644
postoperative management, 642–644
preoperative evaluation, 636–637
pretransplantation, management, 637–638
recipient operation, 639–640
surgical techniques, 638–640
survival, 644
early, 644
late, 644
Heart valves, trans-catheter replacement of, 101
Heart–lung transplantation, 646–650
acute rejection
diagnosis of, 649–650
infection, 650
chronic rejection, 650
donor procurement, 646–647
donors, 646–647
heart–lung block, 647
transplantation, 657
implantation, 648–649
outcome, 650
patients, selection of, 646
postoperative management, 649–650
recipient operation, 647–649
Heat exchanger, 173–174
Hegar dilators, 376, 379, 394, 465, 467
Hemi-arch repair, 641
Hemi-azygos continuation, 641
Hemi-Fontan (Stage Two), 567–568
Heparin, 170, 194, 201, 257, 279, 328, 402, 462, 475, 536, 663
dosage, 174
low-dose, 537
neutralization, 174
and protamine pharmacokinetics, variability in, 168
Heparin-bonded shunts, 200
Hepatic veins, 320
drain, 321
drainage of, 530
intracardiac opening of, 322
Herpes simplex virus, 650
Heterografts, 462
Heterotaxy, 319, 337
HF. *See* Haemofiltration (HF)
HFOV. *See* High-frequency oscillatory ventilation (HFOV)
High pacing threshold, 156
High-frequency oscillatory ventilation (HFOV), 215
High-frequency shifts, 34
His bundle tachycardia, 213
HIV infection, presence of, 636
HLH. *See* Hypoplastic left heart (HLH)
HLHS. *See* Hypoplastic left heart syndrome (HLHS)
Hockey stick incision, 404
HOCM. *See* Hypertrophic obstructive cardiomyopathy (HOCM)
Hollow fibre oxygenator, 173
Homograft
conduit, 197
dysfunction, 512
implantation, 429
insertion, 431
in neonate, 414
size of, 463
unicusp patch, insertion of, 395
Homograft valve, 495
Horner's syndrome, 252
Hourglass configuration, 446
Human albumin, 171
Hyaline membrane disease, 199, 338
Hypercalcaemia, infantile, 491
Hypercapnia, 211
Hypercyanotic attack, 189
Hyperglycaemia, 171
Hyperkalemia, 221
potassium administration, 221
Hyperlactaemia, 205
Hypertension, 296, 644
paradoxical/rebound, 293
persistent postoperative, 296
systemic, management of, 208
Hypertrophic cardiomyopathy, high-risk, 159
Hypertrophic obstructive cardiomyopathy (HOCM), 491
Hypertrophied septal bands, 394
Hypertrophy, 387
Hyperventilation, 523
Hypocalcaemia, 220
ionized, 208
Hypocapnia, 211
Hypokalaemia, 220
Hypomagnesaemia, 220
Hypoparathyroidism, 220
Hypoplasia, 68, 309
arch repair for, 438
Hypoplastic left heart (HLH), 263
Hypoplastic left heart syndrome (HLHS), 82, 559–570
diagnosis of, 560–561
foetus with, 561
initial palliation, 562
palliative surgery, management after, 561
preoperative considerations, 561–562
results of, 569–570
Hypoplastic ring, 387
Hypotension, 188, 191
systemic, 189, 208
Hypothermia, 97, 168–169, 171, 174, 195, 301, 303, 351, 606
degree of, 392
intraoperative, 168
neuroprotective effects of, 168

postischaemic, 169
Hypothermic bypass, 465
neonatal deep, 171
Hypothermic circulatory arrest, 171, 180–181, 331
circulatory arrest, 176, 180, 300, 320, 328, 427, 475, 566
Hypovolaemia, 193, 664
Hypoxaemia, 191, 664
arterial, 189
progression of, 401
Hypoxaemic spells, 401
occurrence of, 401
Hypoxic–ischaemic encephalopathy, 233

IABP. *See* Intra-aortic balloon pump (IABP)
IART. *See* Incisional atrial re-entrant tachycardia (IART)
Iatrogenic atrio-ventricular block, 597
Iatrogenic pulmonary artery, 259
Ibuprofen, 275
Idiopathic pulmonary haemosiderosis, 655
Immunosuppression, 642–643
drugs, 198, 643, 644
side-effect of, 644
Implantable defibrillators, 158–159
indications for primary prevention of, 159
technique for, 159
Incision, muscle-sparing, 246
Incisional atrial re-entrant tachycardia (IART)
after Fontan procedure, 631
mechanism of, 630
prevention, 631
after surgery for congenital heart disease, 630–631
Indirect cannulation, 182
Indomethacin, 275
Induction agent, 198
Induction therapy, 664
Infant, normothermia, 188
Infection, 157
prevention of, 222
Inferior pulmonary vein, 661
Inferior vena cava (IVC), 41, 92, 93, 164, 174, 239, 319, 327
anatomic relationship to heart, 12
left superior, ligament of, 13
superior, anatomic relationship to heart of, 13
interrupted, 320
Inflammatory response, 170
awareness of, 170
Inflow occlusion, 599
Infundibular hypertrophy, secondary, 393
Infundibular muscular defects, 363, 366
Infundibular obstruction, 393
Infundibular resection, 393–394
Infundibular septum, 6, 7, 299, 362, 363, 394, 399, 508
displacement of, 399
Infundibular stenosis, 77, 387
secondary, 388
subpulmonary, 130
Infundibulotomy, 429
Infundibulum, pulmonary, 22–24
subpulmonary, 27
Inhaled nitric oxide (iNO), 210
Inlet muscular defects, association with perimembranous muscular defects, 363
Inlet septum, 6
Innominate artery, 41, 52, 311, 314, 493, 494
anatomy of, 14
Innominate vein, 41, 327
anatomic course of, 14
iNO. *See* Inhaled nitric oxide (iNO)
Ino-dilator milrinone, 195
Inotropes, 407
Inotropic drugs, 205–208
first-line, 205
Inotropic therapy, 187
Insulin infusions, 220
Intact ventricular septum, right ventricular outflow tract obstruction, 387–397
Intensive care management,
post-operative patient, 203–226
paediatric cardiac, 203
management, 407
Interatrial communication, 467, 564
Interatrial groove, 330, 453, 574
Interatrial septum, 5, 533
Interatrial shunting, 383
Interclavicular ligament, 240
Intercostal blocks, 663
Intercostal vein, superior, 256
Intermittent positive-pressure breathing (IPPB), 664
International Society of Heart and Lung Transplantation (ISHLT), 635, 653
Interruption,
of aortic arch, 299–304
anatomy of, 299
diagnosis and medical management of, 299–300
midline approach, 300–302
reoperation, 304
repair of
results, 303–304
surgical technique, 300–303
type A, 301, 302
type B, 301, 302
Interventional angioplasty, 294
Interventional cardiology, 101, 106, 281, 294
hybrid approaches in, 116–117
Interventional catheterization
abnormally or normally closed structures, opening of, 101–109
indications of, 101
Interventricular communication, 378, 383
closure of, 378
double-outlet ventricle, 133–136
size of, 374
Interventricular septum, 504, 506
anatomy of, 29–30
membranous component of, 27
subaortic outflow tract of, 27
Intra pericardial and intra-pulmonary arteries, morphology of, 411–412

Intra-aortic balloon pump (IABP), 231
  aim of, 231
Intra-atrial baffle, 324, 325
  in infants, 324
Intra-atrial prosthetic tube, 324
Intra-atrial redirection, 323–324
Intra-atrial re-entry tachycardia (IART), 161
Intracardiac anatomy, 428
Intracardiac defects
  and coarctation of aorta, simultaneous repair of, 292–293
  repair of, 428
Intracardiac suction, 179
Intraoperative cryoablation, 165
Intraoperative echocardiography, 54–57, 393
Intraoperative mapping, 623
Intraoperative valve, analysis and repair of, 574
Intrapericardial pulmonary arteries
  discontinuity of, 415
  hypoplasia of, 411–413
  subhilar reconstruction, 419
Intrapulmonary tunnel, 615–616
Intravascular emboli, 171
Intraventricular conduit, 442
Intraventricular patch, 439, 440
Invasive catheterization, 75
Invasive investigation, 91–98
  general conduct of, 91
  risks of, 97
Investigation, post-operative, 96–97
IPPB. *See* Intermittent positive-pressure breathing (IPPB)
Ischaemia
  coronary, 208
  myocardial, 189, 208
Ischaemic cardiomyopathy, 635
Ischaemic heart disease, 98
Ischaemic mitral regurgitation, 612
Ischaemic stroke, rate of, 623
ISHLT. *See* International Society of Heart and Lung Transplantation (ISHLT)
Isoflurane, 192
Isohaemagglutinins, 637
  serum titres of, 637
Isomerism, 4–5
  indication of, 5
  right, 4–5
Isoprenaline, 196, 199, 205
Isthmoplasty
  direct, 292
  indirect, 292
IVC. *See* Inferior vena cava (IVC)

JET. *See* Junctional ectopic tachycardia (JET)
Jugular vein, 193
  internal, 149
  internal/external, 152
Junctional ectopic tachycardia (JET), 149, 213, 362, 407
  managing, 213
  mechanism, 213
  ‘‘warm up’’, 213
Junctional tachycardia, 622
Juxta-arterial doubly-committed defect, 139
Juxtaductal coarctation, 291
Juxtaposed atrial appendages, 443

KAVD. *See* kinetic-assisted venous drainage (KAVD)
Kawasaki disease, 98
Ketamine, 191, 192, 198
Kinetic-assisted venous drainage (KAVD), 173
Kommerell’s diverticulum, 311

Labetalol infusion, 200
Lactate, 205
Laevocardia, 539
Laparotomy, 219
Laryngeal nerve, 314
  recurrent, 276
Laryngospasm, 189
Late pericardial effusion, 352
Lateral tunnel, 568
Lateralization, 4
Lateralized situs, indication of, 4
LCO. *See* Low cardiac output (LCO)
LCOS. *See* Low cardiac output syndrome (LCOS)
Lead fracture, 156
Lead implantation, optimal guideline measurements at, 151
Leaflet coaptation, 596
Leaflet defect, 580
Leaflet prolapse, 580–581
Lecompte manoeuvre, 57, 80, 427, 438, 461, 465, 475, 479, 531, 642
  utilization of, 461
Left aortic arch, with right-sided upper descending aorta, 311
Left atrial and central venous pressures, monitoring, 198
Left atrial appendage thrombus, assessment of, 38
Left atriotomy, 454, 625
Left atrium
  drainage of inferior vena cava/hepatic veins, 325
  enlargement of, 335
Left coronary artery, ligation of, 613
Left coronary cusp, 491
Left inferior leaflet, anatomy of, 374
Left pulmonary artery, implantation of, 314
Left thoracotomy, 239, 263–265
Left upper-lobe vein, isolated drainage of, 351
Left ventricular apex, direct venting of, 179
Left ventricular lead malposition, 157
Left ventricular outflow obstruction, with ventricular septal defect (VSD), 560
Left ventricular outflow tract, 467
  abnormal, 490–491
  anatomical structure of, 489–491
  clinical presentation of, 491–492
  diagnosis of, 491–492
  indications for operation, 492–493
  interventions of, 492–493
  normal, 489
  obstruction, 299, 302, 304, 490
  results of, 511–512

surgery of, 489–512
Left ventricular outflow tract obstruction (LVOTO), 81, 466, 471
anatomical, 466
late survival, 303 , 385
progressive dynamic, after inflow atrial repair, 474
relief of, 466
residual pressure gradients in, 467
Left ventriculography, 390
Left ventriculotomy, 467
Levosimendan, 208
Ligaclip, 462
Ligaclip occlusion, in premature infants, 280
Ligamentum arteriosum, 263, 265, 288, 310, 312, 395, 444
dividing of, 311
Lillehei–Hardy–Hunter operation, 595
Linea alba, 240
Lingular artery, 661
Lithium iodide cell, development of, 145
Liver, immaturity of, 168
Living lobar lung transplantation, 660–661, 667
evolution of, 667
related lobar transplantation, 664
technique, 660–661
Lobar allograft preservation, 661
Lobar anatomy, vascular and bronchials, 660
Lobar arterial anatomy, 660
Lobar emphysema, 431
Lobar harvest, 661
Lobectomy
donor left lower, 661
donor right, 660–661
Local anaesthesia cream, 191
Long QT syndrome, 98, 159
Long-segment congenital tracheal stenosis (LSCTS), 313
Low cardiac output (LCO), 204–210
arteriovenous extraction, 205
clinical manifestations of, 205
metabolic indicators, 205
non-pharmacological management of, 208–209
core cooling, 208
ventilatory strategies, 208–209
pharmacological management of, 205–208
inotropic drugs, 205–208
Systemic vasodilators, 208
Low cardiac output syndrome (LCOS), 204, 208
occurrence of, 204
LSCTS. *See* Long-segment congenital tracheal stenosis (LSCTS)
Lung donors, acceptable, 658
Lung transplant
bilateral, 657
early and late mortalities, cause of death, 666
heart, 656–657
living lobar, 657–658
donor selection for, 658
single, 657
types of, 656–658
Lung transplantation
off-bypass, proponents of, 662
psychosocial considerations for, 657
Lungs
blood supply to, 411
ventilation of, 211
LV–PA conduit, 467–468
LVOTO. *See* Left ventricular outflow tract obstruction (LVOTO)
Lymphocytes, 219
Lymphocytotoxic IgG class antibodies, 636, 637
Lymphomas, 644
Lymphoproliferative disease, 644

MAC. *See* Minimum alveolar concentration (MAC)
Magnesium, 220
Magnetic resonance imaging (MRI), 77, 91, 254, 474, 492, 599
cardiac, 73
congenital heart defects, 73–84
dynamic imaging, 73
static imaging, 73
velocity-encoded Phase contrast MR, 75
vs Multidetector computed tomography (MDCT), 86–87
Main pulmonary artery (MPA), banding of, 474
Major aortopulmonary collateral arteries (MAPCAs), 411, 416, 421
investigations, 413
patient with, 412
Malacia, 316
Malalignment defects, 363–364
Mannitol, 171, 221
MAPCAs. *See* Multiple aortopulmonary collateral arteries (MAPCAs)
Mapleson system, 563
Marfan syndrome, 35, 580
Marking sutures, 451
Mayo syndrome, 139
McGoon ratio, 413, 420
MDCT. *See* Multidetector computed tomography (MDCT)
Mechanical assistance device, choice of, 229–231
Medial muscle, contraction of, 275
Median sternotomy, 265, 328, 390, 509, 561
Mediastinal pleura, 252, 286
Membranous septum, 137, 357, 495
atrio-ventricular component, defects in, 124
interventricular, 123
component, 138
remnant of, 123–125
Mesocardia, 539
Methicillin-resistant staphylococcus aureus (MRSA), 222
Methoxamine, 195
Methylprednisolone, 665
Micro-air embolism, 173
Microporous membrane oxygenators, 167
Microre-entrant wavelets, 624
Mid-right ventricular obstruction, relief of, 394
Midazolam, 191
Midline sternotomy, 239–244, 271, 292, 333, 345, 351, 417, 443, 475, 535, 574, 600, 610
cannulation, 242–243

Midline sternotomy, (*continued*)
closure from, 279–280
Midline sternotomy incision, 356, 517, 594
Mild valve stenosis, 64
Milrinone, 196, 208, 421
Minimum alveolar concentration (MAC), 192
Mitral and tricuspid valves, contiguous leaflets of, 126
Mitral,
annulus, 578
atresia, 45
with atrial septum, 323
diastolic murmur, 573
inflow velocity, Doppler assessments of, 155
leaflet, anterior, 489
regurgitation
assessment of, 38
management of, 616–617
stenosis, 559, 584
valve, 65, 575, 616
anatomy of, 27
leaflets of, 25
surgical access to, 24
annular enlargement, 505
anterior leaflet of, 361, 495
bileaflet structure, 373
congenital malformations of, 573–588
clinical presentation of, 573
surgical techniques for, 574
dysfunction, 587
incompetence, 578
leaflets, 574
pathological features of, 376
regurgitation, 573, 578–582
congenital, 573, 586
normal leaflet motion, 578
repair of, 373
restricted leaflet motion, 581–582
with abnormal papillary muscles, 582
with papillary muscles, 581–582
results of, 585–588
stenosis, 573, 575–578
with abnormal papillary muscles, 576–578
congenital, 573
straddling, 669
surgery, secondary, 377
three-leaflet, 579–580
trileaflet structure, 373
Mixed Venous oxygen saturation, 205
mLA. *See* morphological left atrium (mLA)
mLV. *See* morphological left ventricle (mLV)
MMF. *See* Mycophenolate mofetil (MMF)
Modified ultrafiltration (MUF), 183
Monocusp patches, 395
Monocusp pulmonary valves, 404
Morphine, 187, 189, 197
intravenous, 217
pharmacokinetics of, 217
morphological left atrium (mLA), 525
morphological left superior vena cava (mLSVC)
persistent, 528–530
morphological left ventricle (mLV), 525, 539
morphological left ventricle (mLV) dysfunction
early, 527
late, 527
morphological right atrium (mRA), 525
incision, 532
morphological right ventricle (mRV), 525
morphological right ventricle (mRV) dysfunction, 527, 539
morphologically left ventricular outflow tract obstruction (mLVOTO), 527
MR imaging. *See* Magnetic resonance imaging (MRI)
MR signal
echo, 73
gradient echo, 73
producing echo, 73
spin echo, 73
mRA. *See* morphological right atrium (mRA)
MRI. *See* Magnetic resonance imaging (MRI)
MRSA. *See* Methicillin-resistant staphylococcus aureus (MRSA)
MUF. *See* Modified ultrafiltration (MUF)
Mullins sheath, 106
Multidetector computed tomography (MDCT), 84
acquisition, triggering of, 84
disadvantages of, 87
scanning, 87
techniques, 84
Multiple aortopulmonary collateral arteries (MAPCAs), 400, 407
Multitrack catheter, 102–104
Mural leaflet, 578
defect in, 580
Murmur, 401
crescendo–decrescendo, 275
Muscle of Lancisi, 21
Muscle twitching, 157
Muscle-sparing incision, 246
Muscular defects, 135
Muscular inlet defects, 440
Muscular obstruction, appearance of, 394
Muscular septal hypertrophy, 261
Muscular septum, 495
deficiency of, 362
ventricular, 141
Muscular ventricular septal defects, 112–113
Muscular ventricular septum, 121
Mustard operation, 443–449, 451, 460, 643, 644
complications of, 448
re-operations after, 448–449
results, 448
Mustard/Senning repair, 321
Mycophenolate mofetil (MMF), 664
tacrolimus, combination of, 664
Myocardial dysfunction, 369
left ventricular, 232
postcardiotomy, 232
Myocardial fibrosis, diffuse, 613
Myocardial flaps, 670
Myocardial infarction, acute, 613
Myocardial ischaemia, 181, 189, 388, 609

Myocardial ischaemia–reperfusion, 232
Myocardial oedema, 474, 526
Myocardial oxygenation, 189
Myocardial preservation, 167, 181–182
Myocardial protection, 574, 584
Myocarditis, 231, 236
Myocardium, 178, 181, 189, 200
  infant, 187
  ischaemia of, 482

Nakata index, 413, 420
NAME syndrome (naevi, atrial myxoma, myxoid neurofibromata and ephilides), 674
Nasal Continuous positive airway pressure (CPAP), 215
Nasal speculum, 467
Nasogastric tube, 199
Nasopharynx, 174
Nasotracheal intubation, 563
Necrotizing enterocolitis (NEC), 218–219
  in congenital heart disease, 219
  occurrence, 219
Negative-pressure ventilation, clinical use of, 209
Neo-aorta, 479–480, 499
  arch obstruction, 82
  regurgitation, 461
  suture line, 537
Neo-pulmonary artery,
  banding of, 460
  reconstruction of, 537
Neointimal proliferation, 107
Neonatal myocardium, 168
Neonatal palliation, 397
Neonates
  with isolated coarctation, 53
  with pulmonary atresia, management protocol, 390
Nerve hook retractors, 467
Neuroblastoma, 607
Neurological dysfunction, 222–226
Neurological injury, 222
Newborns, with critical pulmonary stenosis or atresia, 102
Nitroglycerin infusion, 664
Nitroglycerin patch, 537
NMDA receptor antagonist, 193
Nodule of Arantius, 489
Nomenclature, 3–9
  connections vs. relations in, 3–4
  function of, 3
Nomogram, 389
Non-biological emboli, 171
Non-blood fluids, 211
Non-functioning wire, 157
Non-invasive CPAP, 215
Non-invasive positive pressure support, 215
Noonan syndrome, 387
Noradrenaline, 189, 191, 205
Normal cardiac anatomy, 70
Normal leaflet motion, 578
Normothermia, 167, 188, 392
Normothermic bypass, 662
Norwood operation, 197–198
Norwood procedure, 82, 562–567
  surgical technique, 563
Nutrition, 217–218

Oblique right atriotomy, 445
Oblique sinus, 12
Obstructive hypertrophic cardiomyopathy, 492
Obtuse margin, of heart, 11
Oesophageal echocardiography, 75
Oesophagogram, barium, 310
Oesophagus
  anatomic relationship to heart of, 15
  vascular compression of, 53
Oliguria, 221
  children with, 221
One-and-a-half ventricle repair, 397
Open valvotomy, with cardiopulmonary bypass (CPB), 493
Opioid, 192, 199
  analgesic, 192
  intravenous, use of, 216
  sedative effects of, 217
Organ dysfunction, postoperative, 169
Orthotopic transplantation, 641
Ostium primum, 373
Ostium primum atrial septal defect, 383
Ostium primum patch, 321
Ostium secundum, 322
Ostium secundum atrial septal defect, 75
Outcomes monitoring, 679
Outflow tract, repair of, 429
Oxygen consumption, 205
Oxygen therapy, 214
Oxygenators, 173
  hollow fibre, 173
  silastic, 173

PAB. *See* Pulmonary artery banding (PAB),
Pacemaker, 408
  and defibrillators, use of, in children 145–159
  follow-up, 158
  implantation techniques for, 149–156
    permanent systems, 149–156
    temporary systems, 149
  problems of, 156–158
    elective generator replacement, 158
    electrode dislodgement, 156
    endocardial lead extraction, 157
    erosion, 157
    high pacing threshold, 156
    infection, 157
    lead fracture, 156
    left ventricular lead malposition, 157
    muscle twitching, 157
    non-functioning wire, 157
  terminology of, 146
Pacemaker implantation, 214
  choice of system to be implanted, 148

Pacemaker implantation, (*continued*)
in children, indications for, 146–148
Pacemaker wires, 448, 464
Pacing
dual-chamber, 151
post-shock ventricular, 159
Pacing system analyser (PSA), 151
Paediatric Cardiac Care Consortium (PCCC), 684
Paediatric cardiac mechanical support, 229–236
complications of, 236
duration for, 235–236
future developments of, 236
indications for, 232–233
management of patients, 235
mechanical assistance device, choice of, 229–231
outcomes of, 236
patient selection criteria, 233
practical aspects of, 233–236
timing for, 232–233
univentricular, vs. biventricular anatomy, 232
Paediatric lung transplantation, 653–667
acute rejection, 665–666
contradictions, 655–656
donor selection, 658
graft preparation, 659–660
immunosuppression, 664–665
indications for, 653–655
infection, 666
fungal, 666
viral, 666
operative technique, 662–663
organ procurement, 658–659
outcomes of, 666–667
postoperative management, 664
nitric oxide, 664
pain control, 664
psychosocial evaluation, 656
recipient operation, 661–664
anaesthesia, 661
cardiopulmonary bypass, 662
Palmaz, 316
Pancuronium, 193, 198
Papillary muscle, 374, 580
absence of, 577
elongation of, 581
medial, 22, 126
Papillary muscle hypoplasia/agenesis, 582
PAPVR. *See* Partial anomalous pulmonary venous return (PAPVR)
Paracetamol, 217
Parachute mitral valve, 576–577, 582
Paracorporeal device, cannulation for, 234
Paradoxical/rebound hypertension, 293
Paraplegia, 287, 293
Parenteral fluids, 219
Parenteral nutrition (PN), 218
central venous access for, 218
critically ill children, 218
Parietal band, 6
Parietal pleura, 281
Paroxysmal supraventricular tachycardia, 630
Paroxysmic flutter, 595
Parsonet score, 683
Partial anomalous pulmonary venous drainage
diagnosis of, 343
indications for, 343
surgical techniques, 345–352
types of, 344
Partial anomalous pulmonary venous return (PAPVR), 327, 343–353
complications of, 352
repair of, 347
tunnelling, 349–350
Partial risk scoring, 685
Patch graft aortoplasty, 292
Patent ductus arteriosus (PDA), 109–110, 178, 199, 263, 369, 492
complications of surgery, 282
diagnosis, 275
indications for operation, 275
methods for closure, 276
closure through midline sternotomy, 279
closure from inside pulmonary artery, 280
closure in the presence of endocarditis, 280
division, 279
ligaclip occlusion in premature infants, 280
ligation, 276
patch closure, 280
transcatheter closure of PDA, 281
results, 178
Patent foramen ovale (PFO), 462
PCCC. *See* Paediatric Cardiac Care Consortium (PCCC)
PD. *See* Peritoneal dialysis (PD)
PDA. *See* Persistent ductus arteriosus and patent ductus arteriosus
PDA closure with a patch, 280
with continuous sutures, 359–361, 365
with interrupted sutures, 358–359, 365
Pectoralis muscle, 157, 246
PEEP. *See* Positive end-expiratory pressure (PEEP)
Percutaneous cannulation, site for, 204
Perfusion, 475–476
techniques, 167–184
Pericardial cavity, 54, 329
anatomy of, 11–13
Pericardial cysts, 671
Pericardial effusion, 58, 210, 235
Pericardial flap, 530
Pericardial patch, 316, 381–384, 443, 449, 535
Pericardial sac, 333
Pericardial sutures, 535
Pericardiectomy, 672–674
Pericarditis, 672–674
aetiology of, 672
cholesterol, 672
constrictive, 672
diagnosis and treatment of, 672–674
primary, 672
pyogenic, 672
secondary, 672

tuberculous, 672
viral, 672
Pericardium, 242, 314, 444, 671–674
anatomy of, 11
congenital defects of, 671
patch of, 376, 392, 393, 396
tamponade, 671–672
tumors of, 671
Perimembranous defects, 123, 134, 135, 139, 364–366
Perimembranous infundibular defects, 362
Perimembranous inlet defects, 361–362
Perimembranous trabecular ventricular septal defects, 358–361
Perioperative renal dysfunction, 220–221
Peripheral pulmonary stenoses, stent implant for, 107–108
Peripheral stenoses, 388
Peritoneal dialysis (PD), 220–221, 293
Peritoneal dialysis (PD) catheter, 482–483
Permanent form of junctional reciprocating tachycardia (PJRT), 622, 630
Permanent pacing, in children, recommendations for, 147
Permissive ventilatory strategies, 215
Persistent ductus arteriosus (PDA), 189, 275–282, 289, 443, 444, 459, 599
complications in, 282
diagnosis of, 275
indications for, 275–276
ligation, 444
methods for closure of, 276–282
division, 279
ligation, 276–278
medical, 276
surgical techniques of, 276
murmur of, 609
perioperative mortality for, 282
recanalization of, 282
residual patency of, 282
results of, 282
transcatheter closure of, 281
Persistent systemic hypertension, 294
Persistent truncus arteriosus, 515–523
diagnosis for surgery, 516–517
indications for surgery, 516–517
postoperative management of, 522
results of, 523
surgical techniques for, 517–522
PFO. *See* Patent foramen ovale (PFO)
Phenothiazine-related drugs, 217
Phenoxybenzamine, 208, 328, 475, 476
intravenous, 482
Phenylephrine, 191
Phosphodiesterase inhibitors, 197, 208, 642
Phosphodiesterase ino-dilator, 198
Phrenic nerve, 673
anatomic course of, 13–15
left, 613
Phrenic nerve damage, 336
PHT. *See* Pulmonary hypertension (PHT)
Piggyback transplant, 638
Pigtail catheter, 103
Plasma lactate
high, 205
normal, 205
PlasmaLyte 148, 171
Plasmapheresis, 643
Platelet count, 211
Pleural cavities, anatomic relationship to heart, 13–15
Pleural fluid, aspiration of, 672
PN. *See* Parenteral nutrition (PN)
Pneumocystis carinii, 666
Pneumothorax, 282, 649
Polycythaemia, 189
in children, 191
Polydioxanone, 301
ribbon, absorbable, 267
Polymerase chain reaction (PCR), 644
Polymeric silicone (Silastic) membrane, 244, 266
Polypropylene sutures, 661
Polysplenia syndrome, 4, 319
Polytetrafluoroethylene (PTFE) conduit, 565
Portex, 193
Positive end-expiratory pressure (PEEP), 191, 199, 638
Post-arrest myocardium, 211
Post-bypass, 196–197
ultrafiltration, 196
Post-transplant efforts, 664
Post-transplant lymphoproliferative disease (PTLD), 644, 665
initial treatment of, 644
Postcardiotomy bleeding, management of, 234
Postcardiotomy circulatory failure, 232
Posterior pleura, 276
Posterolateral thoracotomy, 244–246
Posteroseptal commissure, 596
Postoperative
cardiac catheterization, 421
central venous pressures, 421
management, 420–421
Postoperative analgesia, 197
Postoperative infections
causes of, 222
Postoperative investigation, 96–97
Postoperative patient, intensive care management of, 203–226
Postoperative surveillance, 316
Postresuscitation stabilization, 211
Posttracheostomy care, 225–226
Potassium, 220
Potts shunt, 251, 254–255, 408
Precordial continuous murmur, 609
Pregnancy
and increased risk for CHD, 63–64
termination of, 70
Prenatal diagnosis of CHD, haemodynamic stability, 70
Preoperative, 191–192
anaesthetist's visit, 191
starvation protocol, 191
Preoperative mapping, 622–623
Preoperative valve analysis, functional classification of, 573–574

Prepectoral fascia, 153
Pressure, measurements of, 93
  direct pulmonary artery, 195
Pressure-flow, 313
Pressure-monitoring catheter, 481, 568
Pressure–volume loops, 313
Primary end-stage lung disease, 646
Primary pericarditis, 672
Profound arterial hypoxaemia, 97
Prolene, 176, 289, 376, 379, 380, 445, 480, 481, 495, 530
Prolonged intensive antifailure therapy, 188
Prophylactic antibiotics, 666
Prophylactic therapy, 666
Propofol, 193, 194, 217
Propranolol, 189, 209, 393, 401
Prostacyclin, 647
Prostaglandin, 285, 300, 304
  infusion of, 198, 390, 492
  synthesis
    inhibitors, 276
Prostaglandin E, 188, 390
Prostaglandin E1, 562
  infusion, 660
Protamine, 174, 211, 448
  infusion of, 196
PSA. *See* Pacing system analyser (PSA)
Pseudomonas, 650
PTFE baffle, intra-atrial, 568
Pulmonary arterial branches, stenoses of, 387
Pulmonary arterial wall, plication/resection of, 428–429
Pulmonary artery, 69, 102, 252, 256, 272, 280, 614, 642
  abnormalities, 406–407, 425
  in adults, surgical reconstruction of, 419
  allograft, 406
  anastomosis of, origin of, 14
  anatomy, 85
  bifurcation, 255, 471, 479
  detaching, 517
  distal middle lobe, 660
  distortion, 402
    reconstruction of, 530
  exposure of, 428
  focalization of, 417
    bilateral, 417
  graft, 659
  homograft, cryopreserved, 564
  hypoplasia of, 396, 402
  hypoplastic branch, 404
  intrapericardial, 412, 414
  ipsilateral branch, 527
  left
    exposure of, 406
    origin of, 400
  lost tissue, replacing of, 429
  and major aortopulmonary collateral arteries (MAPCAs)
    general considerations, 417
    morphology of, 416–417
  mobilization of, 395
  opacification of, 102
  peripheral, 411
  pressure, 204, 210
  reconstruction, 418–419, 537
    after banding, 268
  repair, approaches to, 428–431
  rerouting to, 440
  stenosis, 105–106, 406, 412, 408, 430
    branch left, 402
    left, 406
Pulmonary arteriolar resistance, 343, 460, 525
Pulmonary artery banding (PAB), 200, 261–269, 356, 526–527
  absorbable, 267
  adjustable, 267
  anterior extrapleural approach, 265
  approaches in, 263, 267
  classical indications for, 261
  complications in, 268
  current indications for, 261–263
  degree of, 265–268
  dilatable, 267
  material used for, 267
  operative techniques, 263–265
  results of, 268–269
  in very small/young infants, 268
  special considerations in, 267–268
  techniques in, 263–265
Pulmonary artery sling, 309, 311–312
  diagnosis of, 310
  repair of, 314
Pulmonary artery trans-catheter insertion, 114–116
Pulmonary artery tree, 416
  origin and development of, 411
Pulmonary atresia, 48, 54, 66, 78–79, 102, 139, 387, 400
  classification of, 417
  hypoplastic pulmonary arteries, without major aortopulmonary collateral arteries (MAPCAs), 414–415
  with intact ventricular septum, 79, 397
    anatomical substrate, 387–388
    coronary arteries, 388
    patients with, repair of, 390–391
    pulmonary arteries, 387
    right atrium, 388
    and tricuspid valve, 387–388
  with Multiple aortopulmonary collaterals (MAPCAs), 407
  patients with, intracardiac anatomy of, 411
  presence of, 401
  and ventricular septal defect (VSD), 79, 411–422
    investigations, 412–413
    pulmonary arteries, 413–414
    repair, 413–414
    surgical technique, 413
  with VSD and MAPCAs , 416–419
    in adults, 419
    results of, 421–422
Pulmonary autograft, 512
Pulmonary balloon valvuloplasty, 390
Pulmonary blood flow, 189, 201, 526
  low, 189
Pulmonary circulation, ductus-dependant, 412

Pulmonary failure, nature of, 662
Pulmonary haemorrhage, 416, 539
Pulmonary homograft, 395, 413, 421, 430, 522
Pulmonary hypertension (PHT), 168, 197, 209–210, 267, 278, 330, 356, 395, 492, 599, 642, 646
  management of, 209–210
  refractory, 230
Pulmonary hypertensive crises, 335–336
Pulmonary incompetence, 77
Pulmonary infections, 666
Pulmonary infundibulum, 22–24
Pulmonary insufficiency, 409
  effect of, 408
Pulmonary ligament, inferior, 662
Pulmonary lymphatic disruption, 664
Pulmonary oedema, 97, 215, 285, 335, 339, 599, 664
  evidence of, 275
Pulmonary plethora, 275, 459
Pulmonary regurgitation, 77, 80, 390, 400, 404, 409, 431, 631
Pulmonary stenosis, 102, 139, 200, 387, 391, 417
  subvalvar, isolated, 388
  supravalvar, repair of, 396–397
  valvar, 388
Pulmonary to systemic flow, ratio, 561
Pulmonary valve 500
  absent, leaflets of, 26
  atresia, 525
  function, 631
  hypoplastic, 401
  imperforate, 387
  leaflet, 400
  monocusp, 404
  relief, 102
  replacement of, 395, 409
    homograft interposition, 395–396
  ring, 437, 438, 527
  stenosis, 68, 461, 525
  stenotic bicuspid, 527
  trans-catheter implantation of, 116
Pulmonary valve annulus, 400, 403
    diameter of, 404
    hypoplasia of, 402
Pulmonary valvotomy, 404, 437
Pulmonary valvotomy/valvectomy, 392–393
  under inflow occlusion, 392
Pulmonary valvular dysplasia, 387
Pulmonary vascular constriction, 335
Pulmonary vascular obstructive disease (PVOD), 191, 251, 370, 375, 459, 443, 460
  irreversible, 188
  magnitude of, 385
Pulmonary vascular remodelling, 356
Pulmonary vascular resistance, 95, 187, 189–191, 355, 567, 637
  assessment of, 636
  calculation of, 637
  effect of, 192
  elevated, 374
  fall in, 197
  Fick principle, 94
  lowering of, 459
  measurements of, 94
  rise in, 197
Pulmonary vascular response, testing of, 94
Pulmonary vasculature, 328, 646
  reactivity of, 168
Pulmonary vasodilator, 210, 335, 523, 654
Pulmonary vein
  confluence of, 330
  congenital stenosis of, 336
  lower lobe, 661
  stenosis, congenital, 336
Pulmonary venous anatomy, 85
Pulmonary venous atrial pressure line, 537
Pulmonary venous atrial suture line, 533
Pulmonary venous atrial vent, 444
Pulmonary venous atrium, 449, 533–535
  enlargement of, 446
  pressure in, 448
Pulmonary venous connection, 559
Pulmonary venous cuff, tailoring of, 659
Pulmonary venous drainage, 209, 327, 641
  patterns of, 43
Pulmonary venous hypertension, 338, 573
Pulmonary venous obstruction (PVO), 336, 338
  repair of, 336–337
  risk of, 446
Pulmonary venous pathway, 454–456
  construction, 455
Pulmonary venous return, 332, 561, 641
  partial anomalous, 343–353
Pulmonary venous stenosis, 336
Pulmonary venous suture line, 533
Pulmonary venous wedge injections, 413
Pulmonoplegia solution, infusion of, 659
Pulse oximetry, 200
Pulse oxymeter, 265
Pulsus paradoxus, 210
Pump prime, 171
Purse-string suture, 176, 265, 266, 427, 444, 448, 462, 493
  for cannulation, 242
  vena caval, 451
PVOD. *See* Pulmonary vascular obstructive disease (PVOD)
Pyogenic pericarditis, 672

RACE. *See* Rate Control vs. Electrical Cardioversion for Persistent Atrial Fibrillation (RACE)
Radiation dosage, 97
Radiofrequency energy (RF)
  bipolar, 626
  pulse, 73
Radiofrequency perforation, 397
Rashkind's balloon septostomy, 189
Rastelli operation, 462–465, 527
  complications, 464
  results, 466
Rastelli procedure, 82, 461
Rate Control vs. Electrical Cardioversion for Persistent Atrial Fibrillation (RACE), 623

Re-coarctation, 294
balloon dilatation of, 294
incidence of, 294
repair of, 294–295
Re-do surgery, 197
Reattached papillary muscle, necrosis of, 670
Rectus muscle, 154
Reflex apnoea, 310
Refractory ventricular ectopy, 623
Remifentanyl, 194, 217, 563
Renal dysfunction, 644
perioperative, 220–221
Renal failure, 293
Renal perfusion, 168
Renal replacement therapy (RRT), 221
in neonates and infants, choice of, 221
Renal system, and fluid therapy, 219–221
Renal vascular resistance, 168
Reparation á l'Etage Ventriculaire (REV) Procedure, 465–466
Residual atrial septal defect, 352
Residual interventricular shunting, 370
Respiratory support, 188
Respiratory system, 214–217
Respiratory therapy, 664
Resternotomy, 176
disadvantages of, 448
Resuscitation drugs, 210
Retinopathy, development of, 199
Retro-oesophageal aorta, 311
Retrograde cannulation, 176
RF pulse. *See* Radiofrequency energy (RF) pulse
Rhabdomyoma, 674
Rheumatic aortic valve disease, 512
Rheumatic valve disease, 581
Right atrium, 452
VSD closure through, 176, 357–364, 384, 481
Right pulmonary artery stenosis, 396
Right pulmonary veins stenosis 336
Right thoracotomy, 239, 448–449
anterolateral, 448
dissection from, 448
results of, 352
Right ventricle
for biventricular repair, 388
catheter decompression, 397
closure through, 364–366
morphological characteristics of, 388
and pulmonary arteries, establishment of continuity between, 519–520
Right ventricle-to-pulmonary artery conduit 108, 407, 415, 521, 562
use of, 562, 566
stenosis, stent implant for, 108
Right ventricle-to-pulmonary artery shunt, 562
Right ventricular
hypertension, 406
outlet septum, 435
thrombo-exclusion, 390
Right ventricular apex (RVA), 163
Right ventricular apical pacing, 151
Right ventricular decompression, 388
Right ventricular dysfunction, 77, 208, 409, 448, 631, 642
Right ventricular hypoplasia, 66, 387, 388, 591
Right ventricular outflow tract, 464
trans-annular patch enlargement of, 394–395
Right ventricular outflow tract obstruction (RVOTO), 77, 387, 400, 401, 407, 472
on cardiopulmonary bypass (CPB), relief of, 392–396
in children, repair of, 391
and intact ventricular septum, 389
with intact ventricular septum, 387–397
with normal sized right ventricle, 397
operative techniques, 392–397
patch, 397
principles of management, 388–389
results, 397
Right ventriculotomy, 394, 395, 418, 463, 530
incision, 394, 395
Risk Adjustment in Congenital Heart Surgery (RACHS-1) method, 685
Risk forecasting, 685
Ritoximab, 644
Roller pump, 173
RRT. *See* Renal replacement therapy (RRT)
Rubella syndrome, 387
Right ventricular (RV) hypoplasia, 79
Right ventricular (RV) restriction, adverse effects of, 216
Right ventricular outflow tract (RVOT)
anatomy, delineation of, 78
patch, 165, 391, 394–395, 439
transannular, 438
RVOTO. *See* Right ventricular outflow tract obstruction (RVOTO)
Rygg cannula, right-angled, 176
RV–PA conduit, 530–531

Saline, 375, 376
SAM. *See* Systolic anterior motion (SAM)
Sano modification, 198
Saphenous vein graft, autologous reverse, 615
Sarcoplasmic reticulum, 187
Scar-related flutter, 161
Scimitar syndrome, 344, 349, 353
Secondary pericarditis, 672
Secundum atrial septal defect, 64, 324, 343–353
complications of, 352
indications for surgery, 343
Sedation, 216–217
Sedative drugs, 217
Segmental emphysema, 431
Seldinger technique, modification of, 149
Selective pulmonary angiography, 254
Senning operation, 443, 451–457, 459, 643, 644
complications of, 456
indications for, 451
modified, 531–535
procedure, 528
preference of, 531

results of, 456
surgical preparation, 535–537
surgical technique, 451–456
Senning/arterial switch operation (ASO), 537, 539
Senning/Rastelli operation, 537
Sepsis, 221–222
Septal band, 6
Septal bulging, 491
Septal commissure, 373, 375
anatomy of, 373
Septal flap, suturing of, 453
Septoparietal trabeculations, 130
Septum primum, 560
remnant of, mobilization, 533
Septum secundum, 18
Sequential segmental anatomy, 41–54
Serine protease inhibitors, 170
Serum potassium, 195
Sevoflurane, 192, 198, 563
Shone's syndrome, 285, 490, 584
Shunts
control of, 183
systemic-to-pulmonary, 251–259
Side-biting clamp, 392
Signal, spectral display of, 34
Silastic oxygenator, 173
Silastic peritoneal dialysis, 482
Sildenafil, 335
oral, 210
Sinus bradycardia, 214
Sinus node, 18
artery, origin and course of, 31
dysfunction of, 148, 214
vascular supply to, 18
Sinus of Valsalva, 368, 489, 490, 614
aneurysm and fistula of, 603–605
congenital anomalies of, 603–607
diagnosis of, 603
results of surgery, 605
surgical treatment of, 603–605
Sinus rhythm, 149, 648
postoperative restoration of, 623
Sinus septum, 19
Sinus venarum, 15
Sinus venosus defect, 75, 347, 353
Sirolimus, 643, 644
SIRS. *See* Systemic inflammatory response syndrome (SIRS)
Situs ambiguus, 4, 319, 323
Situs inversus (IDD) configuration, 4, 442, 525, 528
Situs solitus (SLL) configuration, 4, 442, 528, 641
dextrocardia, associated with, 528
Skin incision, 240
Society of Thoracic Surgeons (STS), 681
Sondegaard's groove, 662
Spatial encoding, 73
Spin-echo imaging, 73–74
spin-echo black blood,
imaging, 82
sequence 73, 79
benefits, 74
Spin-echo sequences, 81
Spinal analgesia, 194
Spinal cord injury, 223
Spleen, 319
isomerism and, 4
Split-lung ventilation, 662
SSFP. *See* Steady-state free precession (SSFP)
Staphylococcus aureus, 222
Staphylococcus epidermidis, 193, 222
Steady-state free precession (SSFP), 74
2D balanced, 79
multislice short-axis balanced , 78
techniques, 3D balanced, 75
Stent implant
for coarctation of aorta, 108–109
for peripheral pulmonary stenoses, 107–108
for right ventricle-to-pulmonary artery conduit stenosis, 108
to relieve obstructions to other vessels, 109
technique for, 106
Steri-Drape, 239
Sternal retractor, 242
Sternotomy, 96, 153, 448, 311, 312, 402
closure of, 243–244
median, 375
midline, 417
rapid transfusion, 194
Steroid, 643, 644, 663
Strabecula septomarginalis, 127
Straddling valve, 669
atrio-ventricular valves, 669–671
Rastelli operation, 670
replacement of, 670
results of, 670–671
surgical implications for, 669–670
types of, 669
Stress ulceration, 219
Stridor, 201, 215, 310
biphasic, 312
STS. *See* Society of Thoracic Surgeons (STS)
Subacute bacterial endocarditis, 609
Subaortic defect, morphology of, 133
Subaortic obstruction, 381, 383
Subaortic shelf, resection of, 502–503
Subaortic stenosis, 50, 303, 377, 438, 439, 492, 512
Subaortic tunnel, 527
Subcapsular haemorrhage, 153
Subclavian artery, 256, 288, 613
left, 285, 302
right, aberrant, 307, 311
trans-pleural dissection of, 254
Subclavian–coronary artery anastomosis, 613
Subclavian flap
reversed, 290–291
technique, 289–290
Subclavian loop, 13
Subclavian steal syndrome, 311
Subclavian vein, 149, 153, 157
Subglottic stenosis, 214
Subpulmonary obstruction, 527

Subpulmonary outflow tract, 133
Subpulmonary stenosis, 525
Subvalvar aortic stenosis, 491
  congenital, symptoms of, 492
  diffuse, 491
  relief of, 502–504
Subvalvar fibromuscular outflow, 489
Subvalvar obstruction, 438
Subvalvar stenosis, 493, 504, 582
  pulmonary 518
Subxiphoid, 153
Sulcus terminalis, 15, 619
Sump tip sucker, 375
Superior interatrial pathway, 619
Superior segmental artery, 661
Superior vena cava (SVC), 41, 172, 239, 252, 272, 327
  Bilateral, 319
  bridging vein, 641
  coronary sinus, drains into, 641
  left, 182
    re-implantation of, 323
  left-sided, 320, 321
    ligation of, 323
  right-sided, 320
    draining, 322
Superolateral/inferolateral commissure, 376
Supra-annular incision, 627
Supracristal defect, 127
Suprasternal ligament, 240
Supravalvar aortic stenosis, 491, 493, 512
  clinical features and physical signs of, 492
  diffuse, 511
  relief of, 510–511
Supravalvar mitral ring, 575–576
Supraventricular arrhythmias, 161
  postoperative, 377
Surface cooling, 300
Surgery, in infants and young children, 417–419
Surgical approaches, 239–247
Surgical pen, 428
Surgical septectomy, 271
Surgilene, 437
Swiss-cheese septum, 128
Symptomatic neonate
  with critical pulmonary stenosis/pulmonary atresia, 389–390
  with tetralogy of Fallot (ToF), 408
Systemic and pulmonary venous connections, anomalies of, 325
Systemic and pulmonary venous return, combined anomalous, 325–326
Systemic antibiotics, 157
Systemic blood flow ratio (Qp:Qs), 74
Systemic collaterals, 411
Systemic hypoxaemia, 401
Systemic inflammatory response syndrome (SIRS), 170
Systemic re-warming, 406
Systemic vasodilatation, 188
Systemic vasodilators, 208
Systemic veins, cannulation, 321
Systemic venous atrium, 321
Systemic venous baffle, 530, 532–533
  suture line of, 533
Systemic venous drainage, abnormalities of, 322
Systemic venous hypertension, 391
Systemic venous obstruction (SVC), 456
  repair, reoperation, 456
Systemic venous pathway, 452–454
  constriction/obstruction of, 448
Systemic venous return, 641
  anomalies of, 319–326
    classification, 320
  roofing procedure, 323
    risk of, 324
Systemic-to-pulmonary artery shunt, 194, 200–201, 251–259, 267, 389, 413, 461, 467, 527, 567
  disadvantages of, 251
  indications for, 251
  prosthetic, 255–259
  results of, 259
  surgical techniques of, 251–259
Systolic anterior motion (SAM), 491

Tachyarrhythmias, 213
  early postoperative, 212–213
  pacing of, 212
  supraventricular, 623–631
Tachycardia, 203, 622, 623
  atrial ectopic, 212–213
  automatic, 212
  catacholamine-sensitive, 159
  junctional, 196
  junctional ectopic, 204
  supraventricular, 622
Tachypnoea, 425, 584
Tacrolimus, 649
  Oral, 664
  use of, 642
Tamponade, 671–672
Tangential arterioplasty, 611
TAPVD. *See* Totally anomalous pulmonary venous drainage (TAPVD)
Taussig–Bing,
  anomaly, 82
  heart, 133, 435, 436, 438
  malformation, 140
TCPC. *See* Total cavopulmonary connection (TCPC),
Teflon cannula, 193
Teflon graft, 300
Teflon pledgets, 365, 418, 463, 498, 508
Temporary pacemaker wires, 196
Temporary pacing, 212
Tenckhoff peritoneal dialysis catheter, 537
Tendon of Todaro, 17
Tension pneumothorax, 210
Teratogens, exposure to, 63
Tetralogy of Fallot (ToF), 41, 50, 53, 77–78, 93, 156, 161–162, 164, 191, 380, 399–409, 601, 622
  cardiac defects, associated with, 400

with Complete atrio-ventricular septal defect (CAVSD), 407
diagnosis of, 400–401
late complications, 408–409
magnetic resonance imaging, role of, 79
morphology of, 427
nomenclature considerations in, 7
operation
indications for, 401–402
repair of, 384
results of, 407–408
surgical technique, 402–406
velocity-encoded phase-contrast MR, 79
TGA. *See* Transposition of the great arteries (TGA).
Thebesian valve, 17
Thermodilution, 94
Thiazide diuretics, 209
Thiopentone, 193
Thoracic duct, 294
Thoracic epidural infusions, 200
Thoracotomy, 153, 155, 199, 200, 246, 417
advantage of, 153
anterolateral, 276
bilateral, 246
left, 239, 256, 276
posterolateral, 244–246, 276
retractors, 287
right, 239
Throacosternotomy, bilateral, 647
Thrombocytopenia, 196
Thromboelastography, 196, 211
Thromboembolism, 623
Thrombosis, 407
Thymic hypoplasia, result of, 220
Thymus, 443
anatomic location of, 13
Tidal volume, 187
Tisseal fibrin seal, 316
Tissue
Doppler imaging, 35
hypoxaemia, 205
metabolism, shifts, 205
oedema, 169
tamponade, 210
ToF. *See* Tetralogy of Fallot (ToF)
Topical agents, 196
Topical cooling, 195
Torsades des pointes, 213
Total anomalous pulmonary venous connection, 559
Total anomalous pulmonary venous return (TAPVR), 327–338
anatomical types of, 327
cardiac, 327, 331–332
diagnosis of, 327
early complications in, 335–336
indications for, 327–328
infracardiac, 327, 332–334
late complications in, 336
mixed, 327, 334–335
palliation for, 328
results of, 337–338
after primary repair, 337–338
after reoperations for pulmonary venous obstruction (PVO), 338
supracardiac, 327, 329–331
surgical techniques for, 328–335
Total cavopulmonary connection (TCPC), 82, 175, 198, 262, 321, 441
Total neopulmonary arterial index, 422
Trabecula septomarginalis, 7, 21–24, 125, 127, 131, 134, 357, 359, 362, 366, 369, 399, 428
divisions, 403
Trabecular muscular septal defects, 362–363, 366, 440
Trabecular septum, 49, 369
anatomy of, 6
displacement of, 6
recognition of, 8
Trachea
mobilization of, 315
retraction, 201
vascular compression of, 53
Tracheal
compression, 310
homograft, 316
obstruction, degree of, 201
repair, 314–316
rings, 223, 309, 313
stenosis, 201, 313
children with, 313
suction, 201
Tracheo-oesophageal compression, 311
Tracheo-oesophageal fistula, 63
Tracheobronchial stenosis, 307
Tracheobronchomalacia, 216, 313
management of, 316–317
Tracheomalacia, 201
Tracheoplasty, 315
Tracheostomy, 223, 316
surgical techniques, 223–225
tube, 224, 226
insertion of, 225
Trans-annular patch, 389, 394, 402, 406, 409, 427, 436
Trans-arterial closure, of fistula, 611
Trans-atrial approach, to tetralogy, 404, 405
Trans-bronchial biopsy, 649, 650, 665
Trans-catheter pulmonary valve insertion, 114–116
Trans-catheter techniques, 75
Trans-catheter valve
implantation, percutaneous, 115
insertion, presentation of, 117
Trans-cutaneous intracardiac lines
removal, 204
Trans-hepatic venous access, 153
Trans-mural cryoablations, 626
Trans-mural lesions, 626
Trans-oesophageal echocardiography, 38–40, 77, 112, 402
parasagittal imaging, 38
Trans-pulmonary pressure, 94
Trans-thoracic echocardiography (TTE), 75, 77, 80
Transducer, 38

Transplantation, 198–199
paediatric, recipients, 198
progressive cardiomyopathy, patients with, 198
Transposition of the great arteries (TGA), 80–82, 189, 262, 271, 302, 443, 451, 459
arterial repair (switch), 80, 163, 189, 262, 471–487
atrial repair of, 526
cardiac anomalies, associated with, 459
diagnosis of, 443
indications of, 443
intact ventricular septum and pulmonary vascular disease, 460–461
left ventricular outflow tract obstruction (LVOTO) and intact ventricular septum, 466
results, 468
malformations, associated with, 459
neonates with, 460
and pulmonary vascular obstructive disease (PVOD), 460–461
simple, 472–473
with subpulmonary stenosis, 80, 81
and ventricular septal defect (VSD), 459–460
and left ventricular outflow tract obstruction (LVOTO), 461–466
results, 460
Transverse sinus, 13
Transverse submammary incision, 240
Triangle of Koch, 17, 121
apex of, 125
Tricuspid annulus, 596, 620
Tricuspid aortic valve
stenosis of, 493
Tricuspid atresia, 45, 66, 261, 267–268
atrio-ventricular connection in, 7–8
without pulmonary stenosis, 262–263
Tricuspid disease
diagnosis of, 593
results of, 597–598
Tricuspid incompetence, after Mustard/Senning operation, 473–474
Tricuspid valve, 16, 44, 48, 50, 150, 157, 365, 369, 392, 394
annulus, 164, 378, 624
antero-superior leaflet of, 21
chordae, 435, 436
cleft in septal leaflet, 123
contiguous septal leaflet of, 361
diameter, 389
measurements of, 388
dysplasia, 68
dysplastic, 387
Ebstein's anomaly of, 51, 161, 163
hypoplasia, 591
incompetence, 448, 593
isolated congenital, 591
leaflets of, 23, 373, 387
displacement of, 68
malformations of, 591–598
mechanism, 466
overhang of, 128
regurgitation, 82, 133, 391, 560
congenital, 591
replacement of
double-switch procedure, 527
and repair, 527
right atrium approach to, 21
septal leaflet of, 16–19, 131
stenosis
congenital, 591
isolated congenital, 591
straddling, 669
supero-inferior commissure of, morphology of, 25–27
valvotomy, 101
Triple-drug therapy, 665
Trisomy 21, 378, 385
Truncus arteriosus, 67, 301–303
arterial connection in, 9
with discontinuous pulmonary arteries, repair of, 521–522
with interrupted aortic arch, repair of, 520–521
with prior banding of pulmonary arteries, repair of, 522
repair of, 517
with truncal valve regurgitation, repair of, 520
truncal valve, regurgitation, 520, 523
Trusler method, 300
Trussler formula, 526
TTE. *See* Trans-thoracic echocardiography (TTE)
Tuberculosis, 672
Tuberculous pericarditis, 672
Tumours, 674
Tunnel-like stenosis, with normal aortic valve, 504
Tunnelling, for Scimitar syndrome 349–350
Turner's syndrome, 69
TVA. *See* Tricuspid valve annulus (TVA)
Tympanic membrane, 174

UF. *See* Ultrafiltration (UF)
Ulnar artery, 204
Ultrafiltration (UF), 183–184, 223
prebypass, 173
Ultrasonography, 92
Ultrasound waves, 33
burst of, 34
Umbilical arterial catheter, 204
Unicusp pericardial patch, for right ventricular outflow tract 465
Unipolar twitching, 154
Univentricular and biventricular repair, decision between, 388
Univentricular heart, 271
Unroofed coronary Sinus
with left-sided superior vena cava, 322
without left-sided superior vena cava, 323, 324

Vacuum-assisted venous drainage (VAVD), 173
Vagus nerve, 286, 648
anatomic course of, 13
Valvar aortic stenosis, 104
congenital, 490–491
unicuspid, 493
Valvar orifice, 577
Valvar pulmonic stenosis, balloon dilatation of, 101

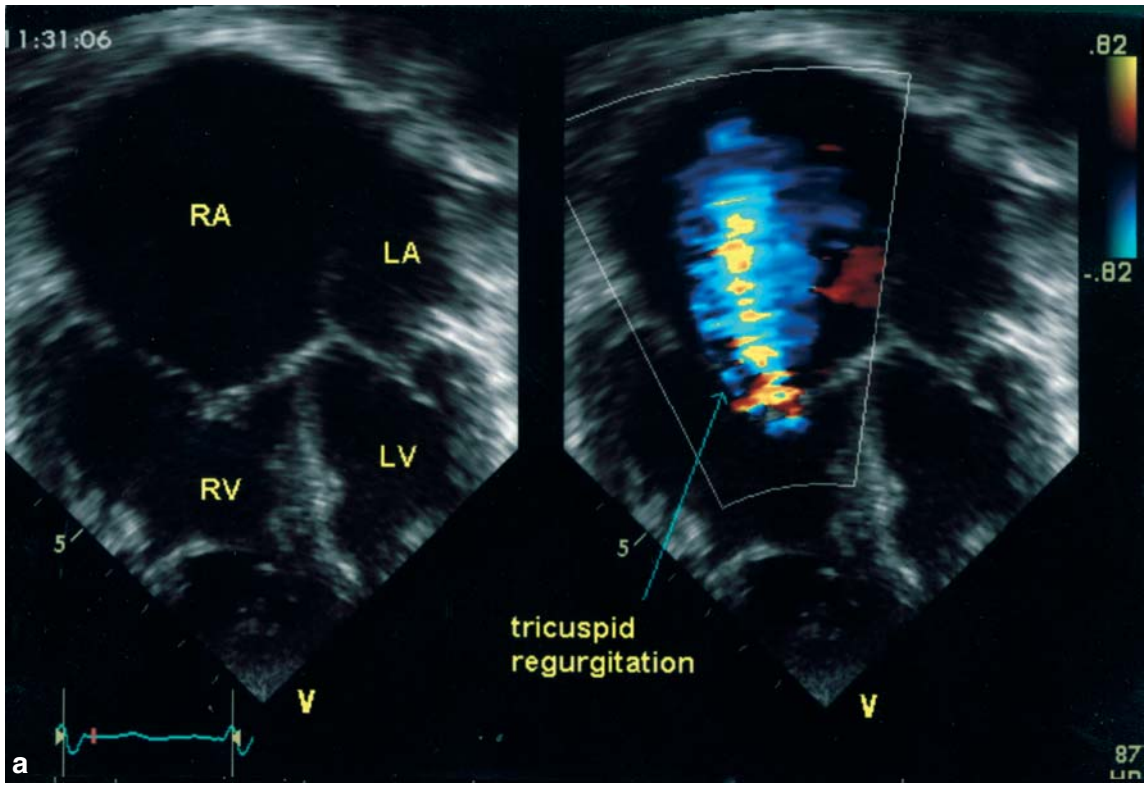
Valve dysfunction, atrio-ventricular, 112
Valve leaflets
anatomy of, 379, 383
bridging, degree of, 374
Valve orifice, 669
Valve repair, assessment of, 583
Valve replacement, congenital mitral valve malformation, 583
Valve stenosis, 68
pulmonary, 68
Valve-bearing conduit, 462
Valve. See also(individual types
imperforate, recognition of), 8
Valved conduits, 416, 418, 421, 427, 431, 438, 468
external, 439
Hancock, 421
Valved homograft, 429
Valvoplasty, mitral valve, 381
Valvotomy, tricuspid valve, 389, 461
in childhood, 493–494
in neonates and infants, 493
Valvular regurgitation, quantification of, 74
Valvuloplasty, pulmonary artery stenosis, 105
Variable life adjusted displays (VLAD), 686
Vascular access, 91–93, 204
Vascular access lines, 193–194
Vascular anatomy, 85
Vascular anomaly, 307, 310
Vascular clamps, 277, 289
Vascular compression
left-sided aortic arch, associated with, 307, 311
right-sided aortic arch, associated with, 308–309, 311
Vascular narrowing (pulmonary veins, baffles, surgical pathway), 106
Vascular obstructions, 106
Vascular rings, 201, 307–317
diagnosis of, 309–310
surgical anatomy, 307–309
surgical technique, 310–312
tracheal stenosis and malacia, associated with, 312–316
Vascular stapler, 661
Vasoactive drugs, 664
Vasoconstriction
peripheral, 195
pulmonary, 196
Vasoconstrictors, 195
Vasodilatation
refractory, 205
Vasodilator, 195, 208, 584
pulmonary, 214
use of, 235
Vasodilator therapy, 208
Vasopressin, 205, 638
VATS. *See* Video-assisted thoracoscopic surgery (VATS)
VAVD. *See* Vacuum-assisted venous drainage (VAVD)
Vecuronium, 193
Velocity-encoded phase-contrast MR, 77
Veno-arterial ECMO, in children, 233
Venodilator, 208
Venogram, 150
Venous access, 152
trans-hepatic, 153
Venous anastomosis, 350, 663
Venous cannula, 178
central, 204
insertion-related risks, 204
small lumen, 173
Venous cannulation, 176–178
single, 176
Venous drainage, 172–173
Venous reservoirs, 172–173
Venous sinus, 15
Venous thrombosis, 151
Venous/cardiotomy reservoir, 172
Ventilation, 91
parallel circulations, 216
positive pressure, 215, 216
postoperative, 214–215
Ventilator
intensive care, 215
management of, 182
settings, 664
strategies, 208–209
Venting, 178–179, 243
Ventricle, 48–50
anatomy of, 15–27
components of, 5, 23
definition of, 5–7
features of, 20, 24
inlet and outlet tract of, 25–27
inlet portion of, 29
left, anatomy of, 20
morphology and, 5–7
muscle bundles within, 6–7
outflow tract of, 25–27
recognition of, 7–8
right, anatomy of, 15–19
surgical trauma vulnerability of, 26
trabecular zone of, 23, 25
Ventricular
arrhythmias, 162, 409, 622
assistance devices (VAD), 229, 230–231
centrifugal, 231
paracorporeal, 231
univentricular, 231
bulges, 622
cavity, 583
diverticula, 676
dominance, 384
dysfunction, 635
epicardium, 145
inversion, isolated, 538
outflow tract obstruction, 399
pacing, 148, 150–151
plication, 596
septal crest, 379
Ventricular septal defect (VSD), 37, 64, 68, 75–76, 189, 261–262, 285, 299, 355–370, 394, 515, 599, 603, 669, 688
anatomy of, 121–142
and aortic arch anomalies, 369

Ventricular septal defect (VSD), (*continued*)
with aortic pulmonary fibrous continuity, 127–128
apical, 369
muscular, 49
closing of, 301, 414, 420
closure of, 404, 517–519, 526–528, 670
through morphological left atrium (mLA), 538
risk associated with, 528
coexistent, 373
conduction disturbances, 370
diagnosis of, 355
with discordant atrio-ventricular connections, 136–139
doubly committed subarterial, 49, 356, 366–368
enlargement of, 136
with fibrous continuity, 123–127
in hearts, 131–133
hospital mortality, 370
indications for, 355–356
inlet muscular, 363
inlet-type, 527
isolated
anatomy of, 122–130
pathophysiology of, 355
large, 355–356
membranous defects, 123
moderately large, 356
morphology of, 136
terminology, 435
multiple, 261, 460
muscular, 363
management of, 76
muscular, 49, 112–113, 357
muscular defects, 128–140
opening to the apical trabecular component, 128–129
opening to the inlet of the right ventricle, 128
opening to the right ventricle, 129
non-committed, 441
patching of, 304
perimembranous, 49, 113–114, 357
perimembranous defects
opening centrally, 123–125
opening to the inlet of the right ventricle, 125–126
opening to the outlet of the right ventricle, 126–127
and pulmonary vascular disease, 460
results, 460
quantification of left-to-right (L–R) shunts, 75
recurrent, 463
residual, 370
results of, 370
long-term, 370
small to moderate, 356
subaortic, 435
subpulmonary, 435
surgical techniques in, 356–370
in tetralogy of Fallot (ToF), 399
in trabecular muscular septum
children with, 462
trans-catheter device closure of, 370
Ventricular septum, 44, 48–50
components of, 5
closing the defect, 379
Ventricular systole, 103
Ventricular systolic pressure, 37
Ventricular tachyarrhythmias, 441
after repair of congenital heart defects, 631
Ventricular tachycardia (VT), 114, 162, 165, 213, 622
in children, 623
polymorphic, 213
Ventriculo-arterial connection, 45–48
Ventriculo-arterial discordance, 50, 325
direct repair of, 471
Ventriculo-arterial junctions, anatomic and haemodynamic, 20
Ventriculo-infundibular fold, 6, 127, 362
Ventriculography, 622
Ventriculotomy, 138, 364, 369, 394, 414, 461, 464, 506
vertical, 393
Vertical vein, 329
Video-assisted thoracoscopic surgery (VATS), 281
Viral pericarditis, 672
VLAD. *See* Variable life adjusted displays (VLAD)
Volume resuscitation, 407
VSD. *See* Ventricular septal defects (VSD)
VT. *See* Ventricular tachycardia (VT)

Warden procedure, 347
Waterston shunt, 254–255
anastomosis, 251
closure of, 254–255
Weaning and extubation, 215–216
Wenckebach's bundle, 619
Williams syndrome, 189, 491
Wire implantation, 148–149
Wire position, practical difficulties with, 151–152
Wolff–Parkinson–White (WPW) syndrome, 619

X-ray angiography, 74, 76
X-ray fluoroscopy, 73
Xenograft patch, 255
Xenograft pericardium, 419, 518
Xenotransplantation, 650

Yasargil aneurysm clips, 418


1:31:06
.82
RA
LA
LV
RV
5
5
tricuspid
regurgitation
V
V
87
a

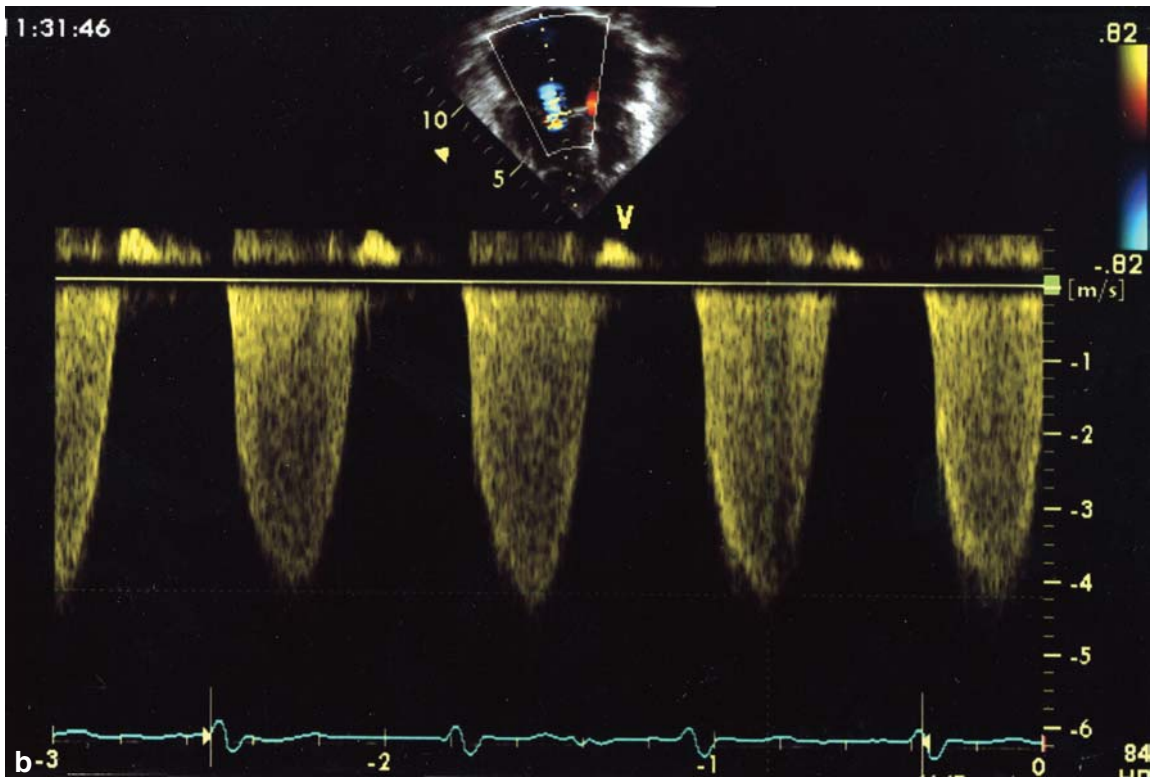

1:31:46
.82
10
5
V
-.82
[m/s]
-1
-2
-3
-4
-5
-6
b
-3
-2
-1
0
84

Plate 1

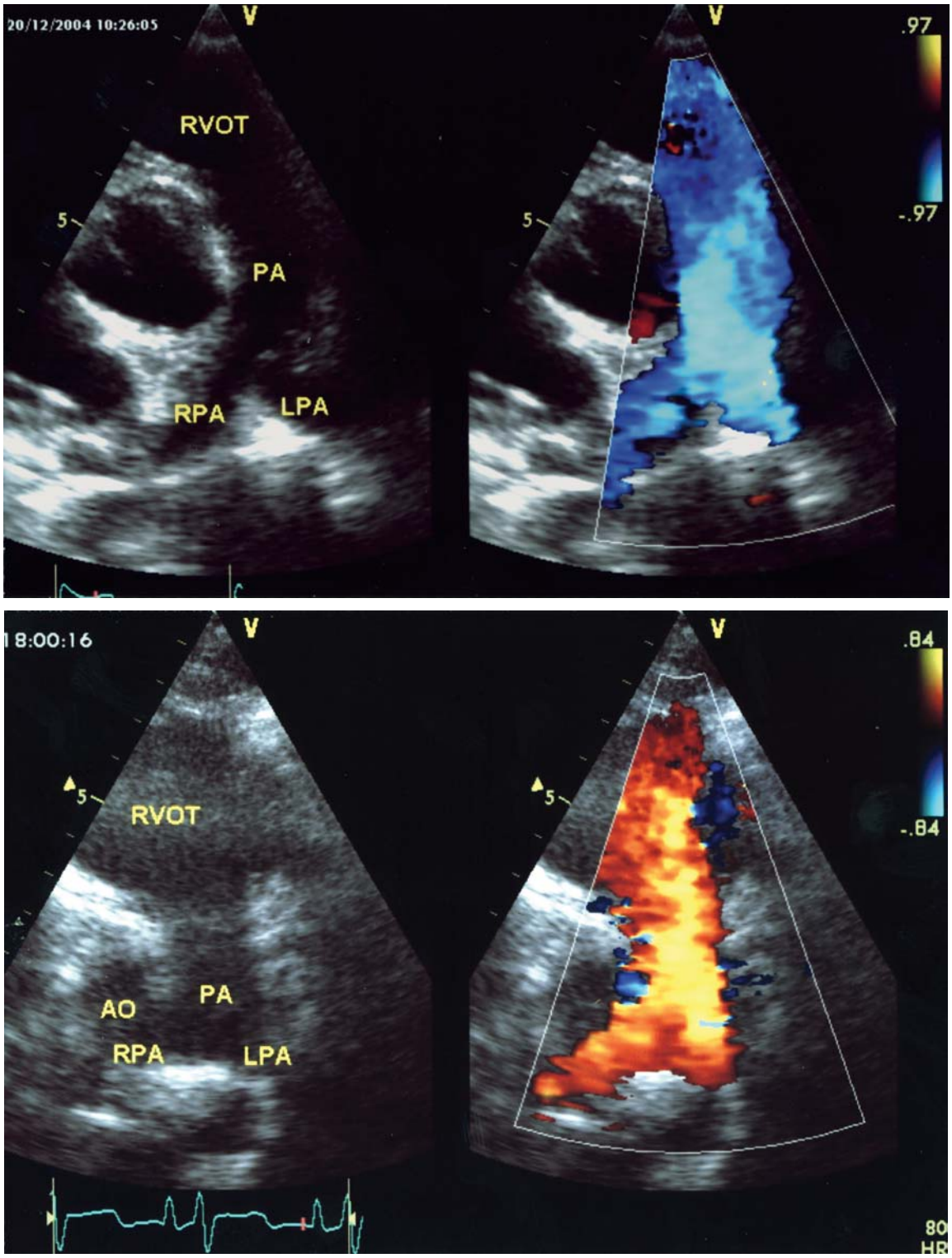
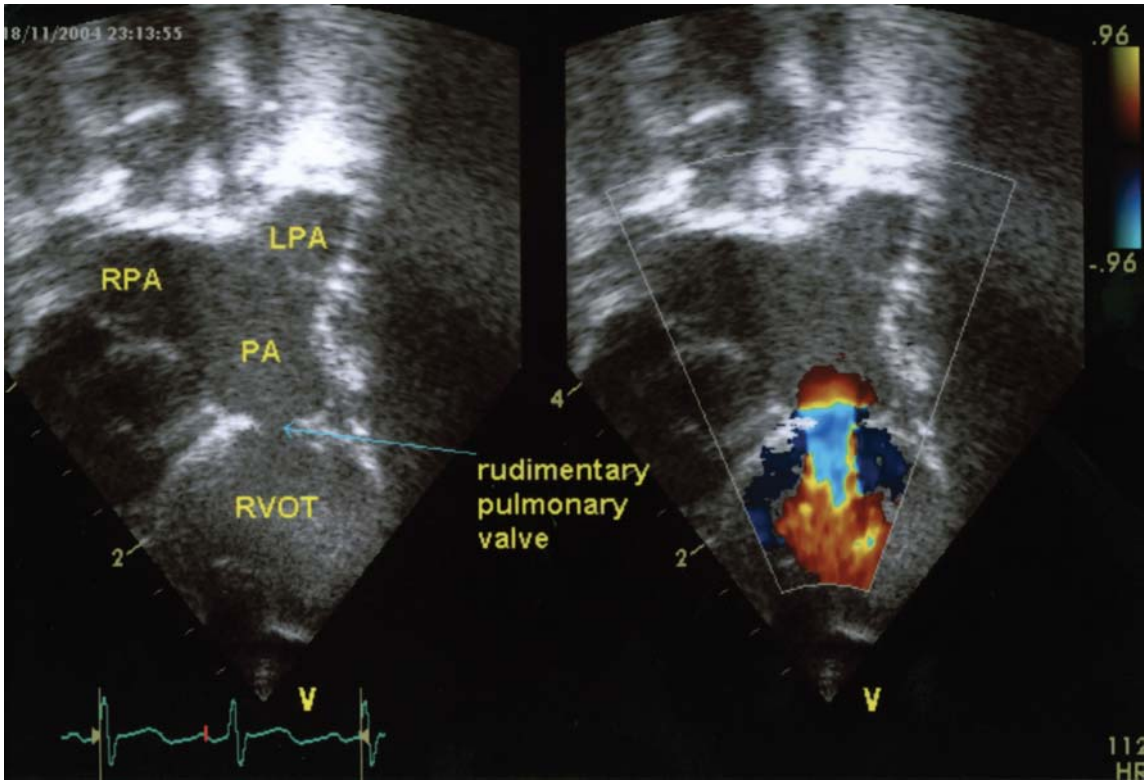

20/12/2004 10:26:05
RVOT
PA
RPA
LPA
.97
-.97
18:00:16
RVOT
AO
PA
RPA
LPA
.84
-.84

Plate 2


18/11/2004 23:13:55
.96
-.96
LPA
RPA
PA
RVOT
rudimentary
pulmonary
valve
4
2
V
V

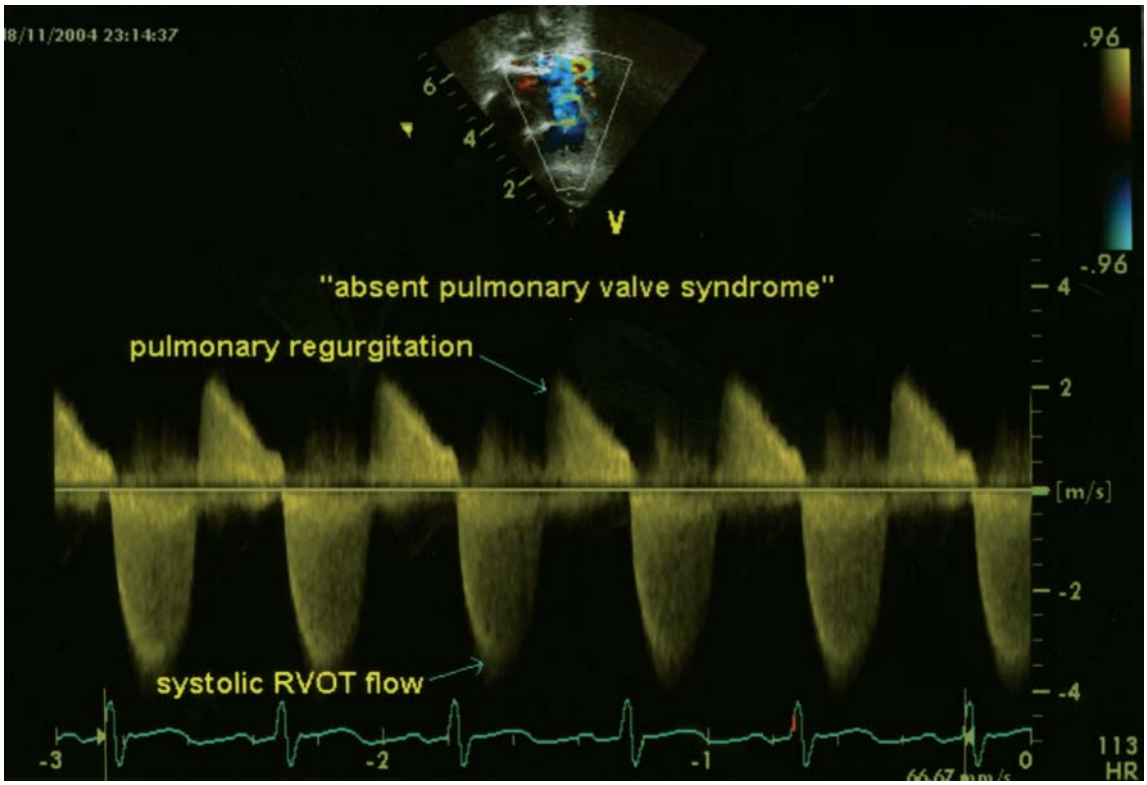

8/11/2004 23:14:37
.96
-.96
6
4
2
V
"absent pulmonary valve syndrome"
pulmonary regurgitation
4
2
[m/s]
-2
-4
systolic RVOT flow
-3
-2
-1
0
113
HR

Plate 3

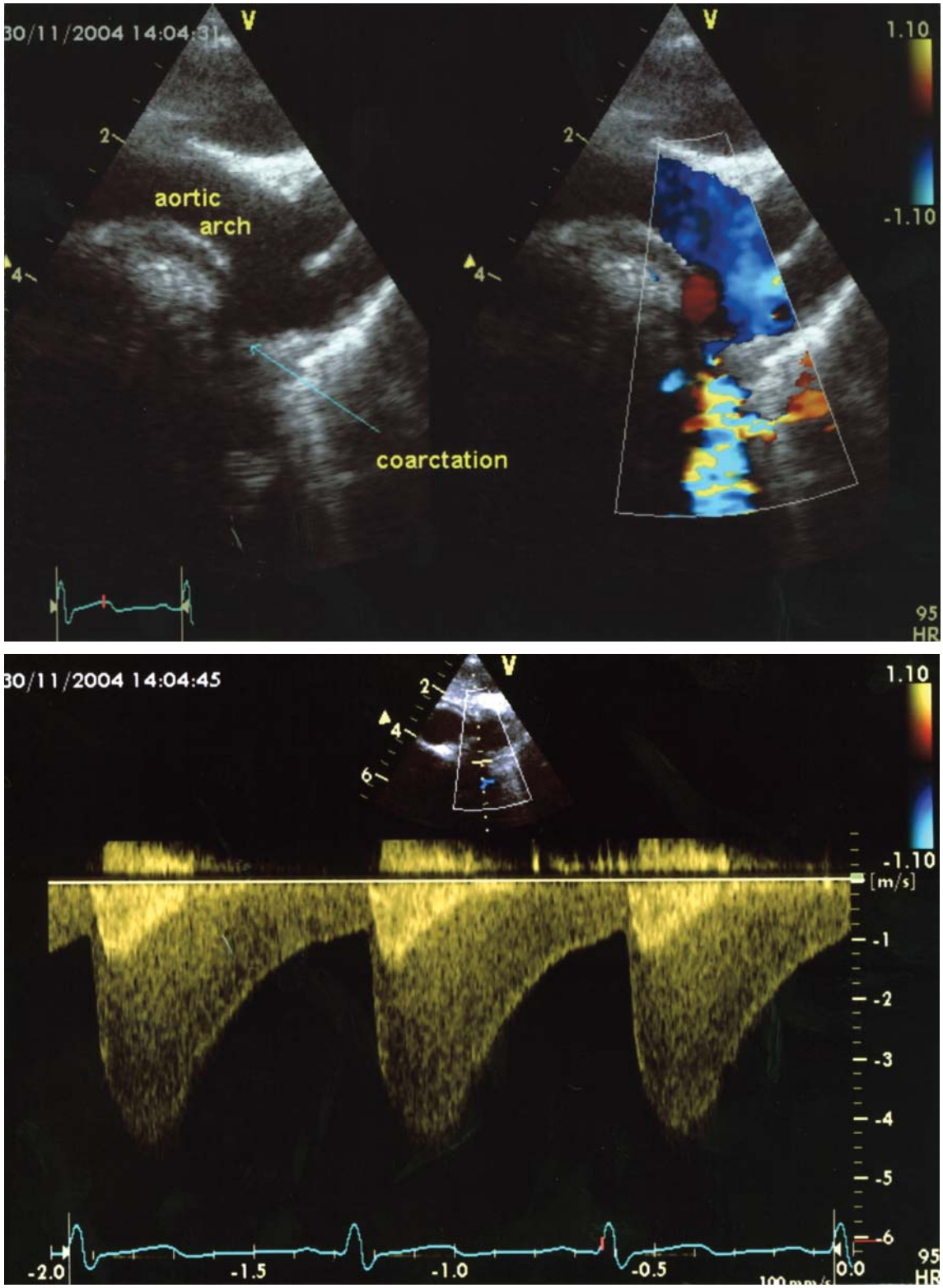

30/11/2004 14:04:31
aortic
arch
coarctation
1.10
-1.10
95
HR
30/11/2004 14:04:45
1.10
-1.10
[m/s]
-1
-2
-3
-4
-5
-6
-2.0
-1.5
-1.0
-0.5
0.0
95
HR

Plate 4

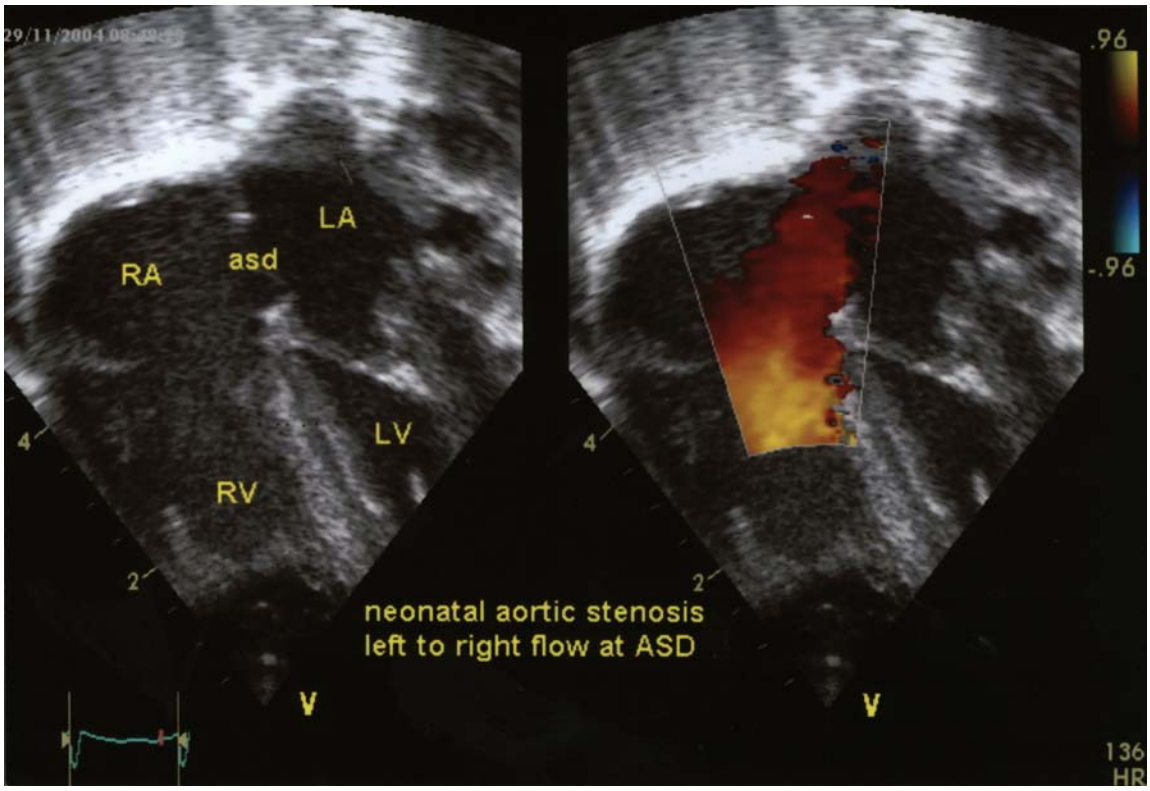

LA
RA
asd
LV
RV
4
2
neonatal aortic stenosis
left to right flow at ASD
V
.96
-.96
136
HR

Plate 5

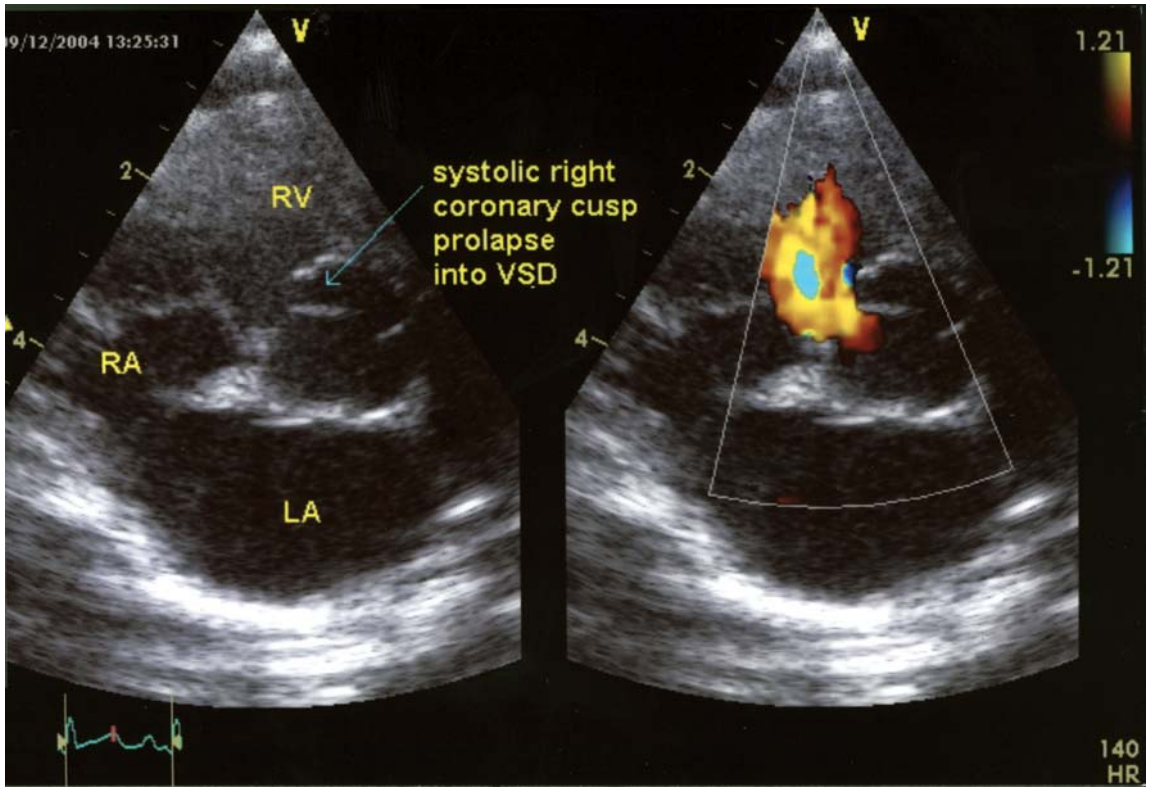

9/12/2004 13:25:31
V
2
RV
systolic right
coronary cusp
prolapse
into VSD
4
RA
LA
V
1.21
-1.21
2
4
140
HR

Plate 6

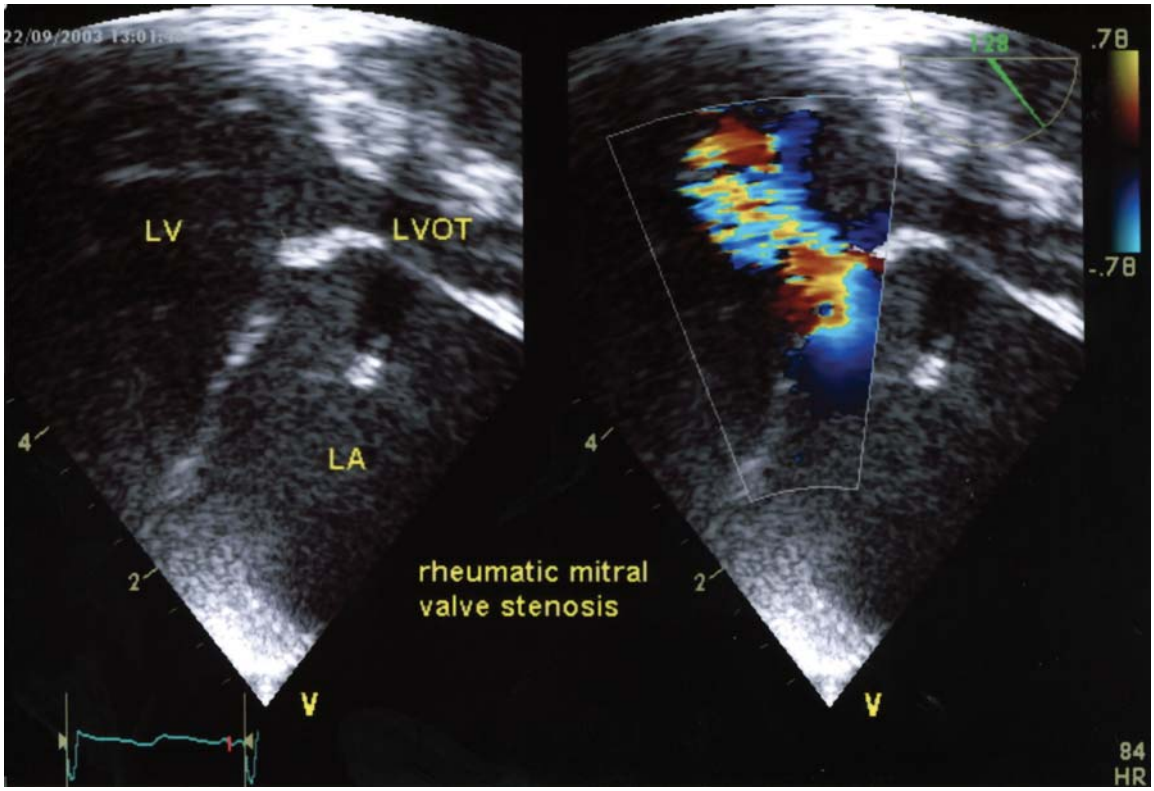


Plate 7

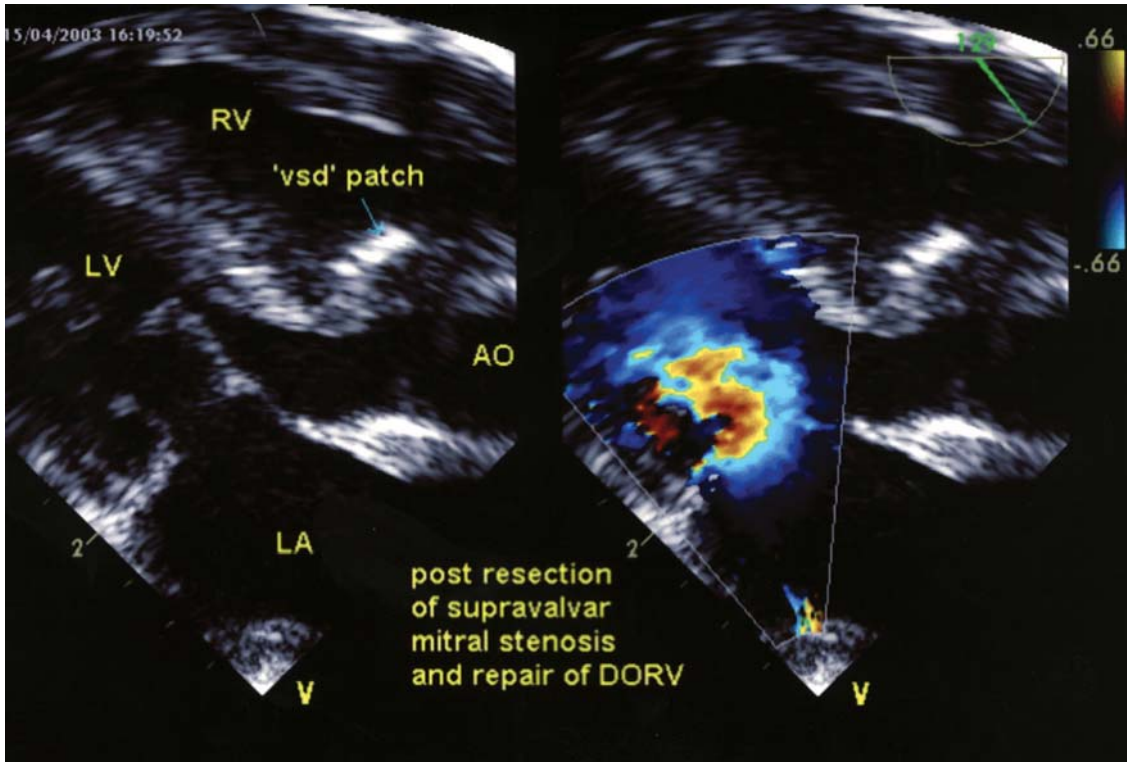

15/04/2003 16:19:52
RV
'vsd' patch
LV
AO
LA
2
V
post resection
of supravalvar
mitral stenosis
and repair of DORV
.66
-.66

Plate 8

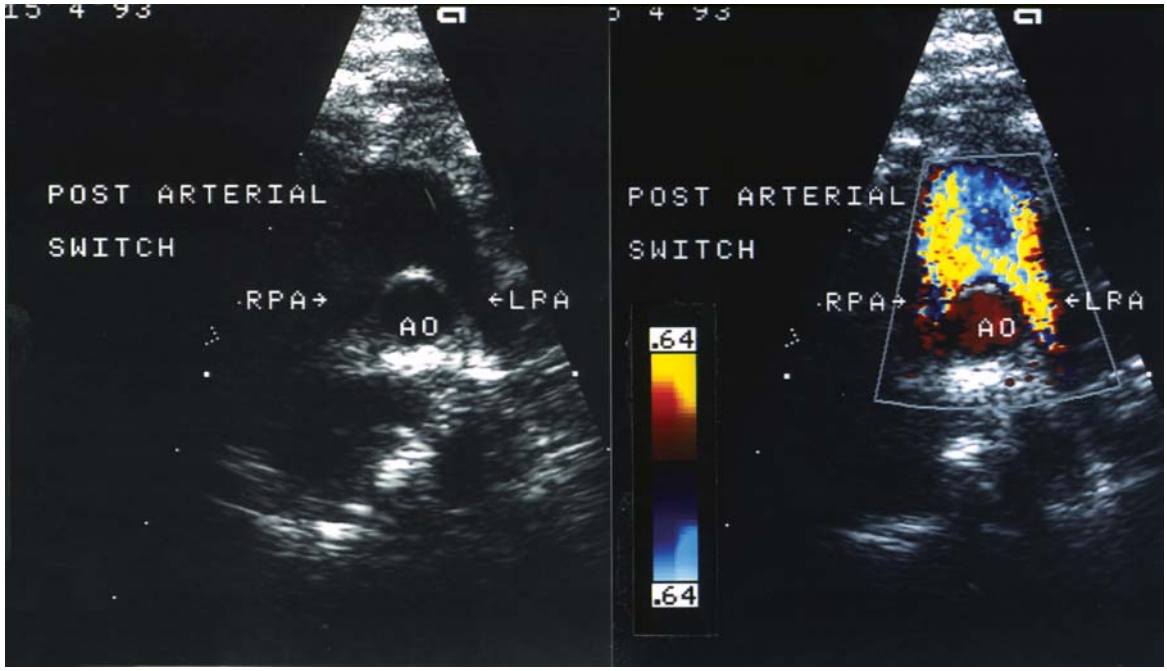
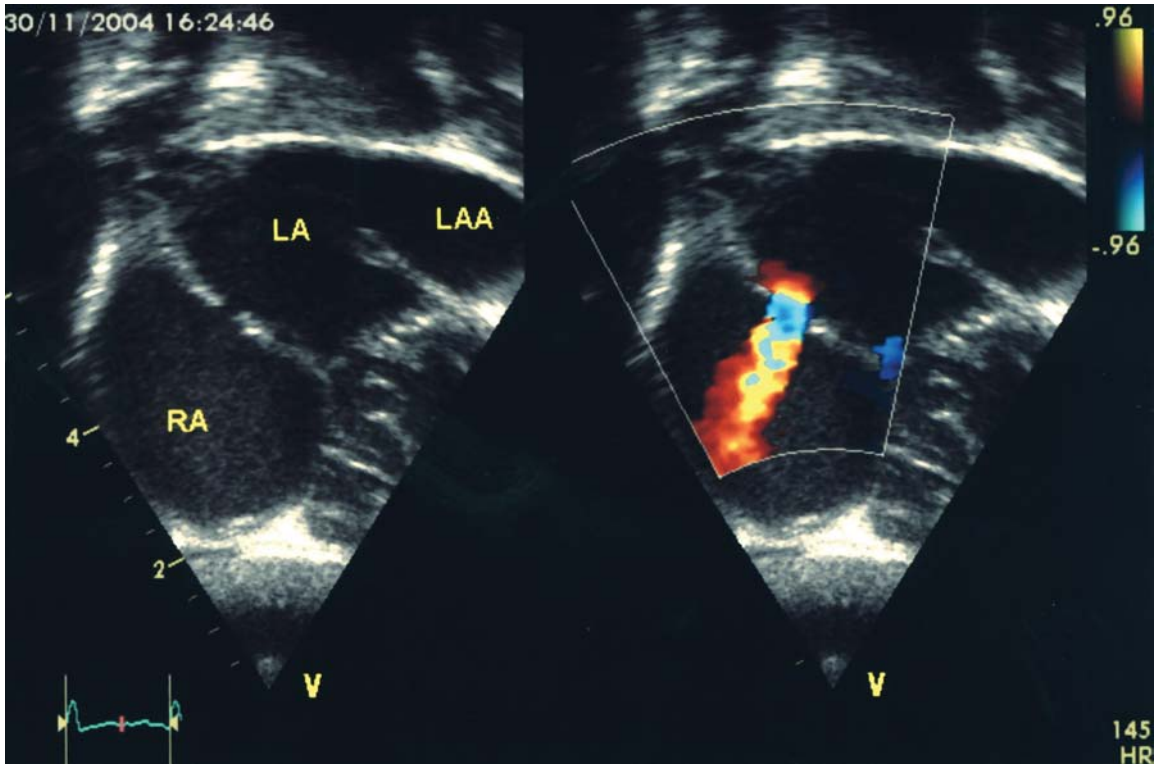

POST ARTERIAL
SWITCH
RPA→
←LPA
AO
POST ARTERIAL
SWITCH
RPA→
←LPA
AO
.64
.64

Plate 9


30/11/2004 16:24:46
LA
LAA
RA
.96
-.96
4
2
V
V
145
HR

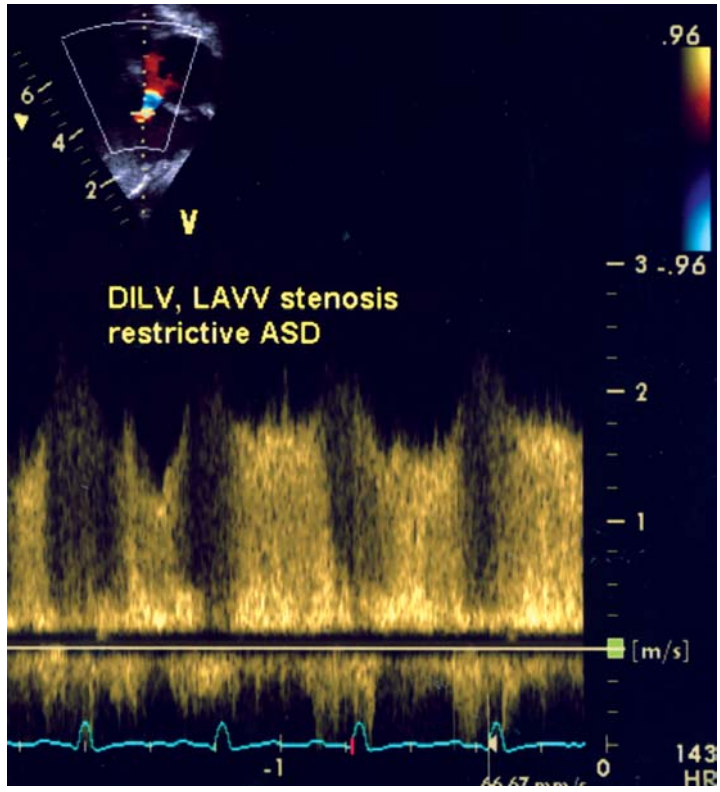

.96
6
4
2
V
DILV, LAVV stenosis
restrictive ASD
3
-.96
2
1
[m/s]
-1
0
143
HR

Plate 10