Question: 1

A 10-year-old girl has had a rash for 4 days without other symptoms. She is taking no medications. Physical examination reveals erythematous cheeks (Item Q1A) and a lacy, reticulated erythema involving the extremities (Item Q1B).

Of the following, the MOST likely diagnosis is

A. erythema infectiosum

B. phototoxic reaction

C. polymorphous light eruption

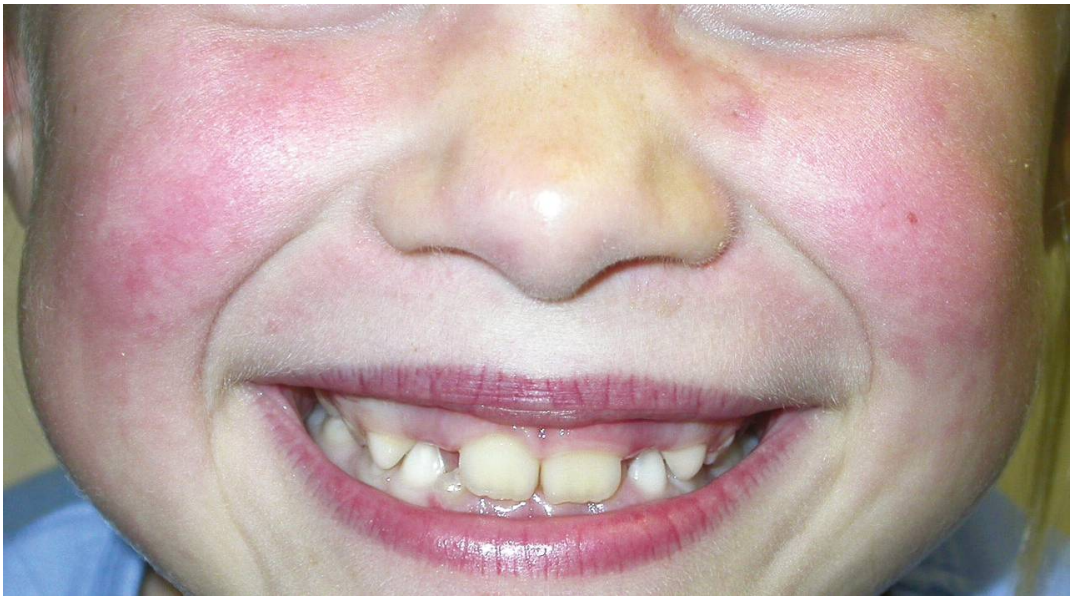
D. scarlet fever

E. systemic lupus erythematosus

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Question: 1



Courtesy of D. Krowchuk

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Question: 1



Courtesy of D. Krowchuk

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Critique: 1 Preferred Response: A

The girl described in the vignette has an eruption consistent with erythema infectiosum (fifth disease), the most common clinical expression of infection with human parvovirus B19. In some patients, fever, malaise, myalgia, or headache precede the eruption by 7 to 10 days. The rash begins as confluent erythema of both cheeks (the so-called "slapped-cheek" appearance) (Item C1A), followed by a lacy, reticulated, pink erythema of the extremities or trunk (Item C1B). After 3 to 5 days, the eruption fades, although it may return for up to 4 months following exercise, overheating, or sun exposure. Other syndromes associated with human parvovirus B19 infection include an atypical rash (eg, one that mimics rubella or a petechial eruption involving the hands and feet [papular purpuric “gloves and socks” syndrome]) (Item C1C), transient aplastic crisis in patients who have hemolytic anemias, and bone marrow failure in immunodeficient individuals. Infection during pregnancy, particularly during the first half of pregnancy, may cause fetal hydrops and death.

A number of disorders produce facial or widespread erythema, but these generally can be distinguished clinically from erythema infectiosum. In none of these disorders is the characteristic lacy, reticulated eruption of erythema infectiosum present. Erythema of sun-exposed skin may result from phototoxic drug reactions in which ultraviolet radiation activates a drug (eg, nonsteroidal anti-inflammatory drugs, tetracyclines, quinolones, phenothiazines). Nonimmunologic cellular damage produces sunburnlike redness that appears 2 to 6 hours after sun exposure. Polymorphous light eruption, a delayed hypersensitivity reaction to ultraviolet light, typically appears 1 to 2 days following sun exposure. Erythematous papules or vesicles develop on sun-exposed sites, such as the face, sides of the neck, and extremities (Item C1D). Children who have scarlet fever often manifest facial erythema and perioral pallor, but the rash is composed of fine, rough-feeling erythematous papules (Item C1E) that are concentrated in flexural areas. Associated symptoms and signs are common and include fever, sore throat, a “strawberry tongue,” and tonsillar exudates. As many as 60% of patients who have systemic lupus erythematosus exhibit a photosensitive malar rash. Unlike the "slapped-cheek" appearance of erythema infectiosum, this eruption may involve the bridge of the nose and cheeks and often is scaling (Item C1F).

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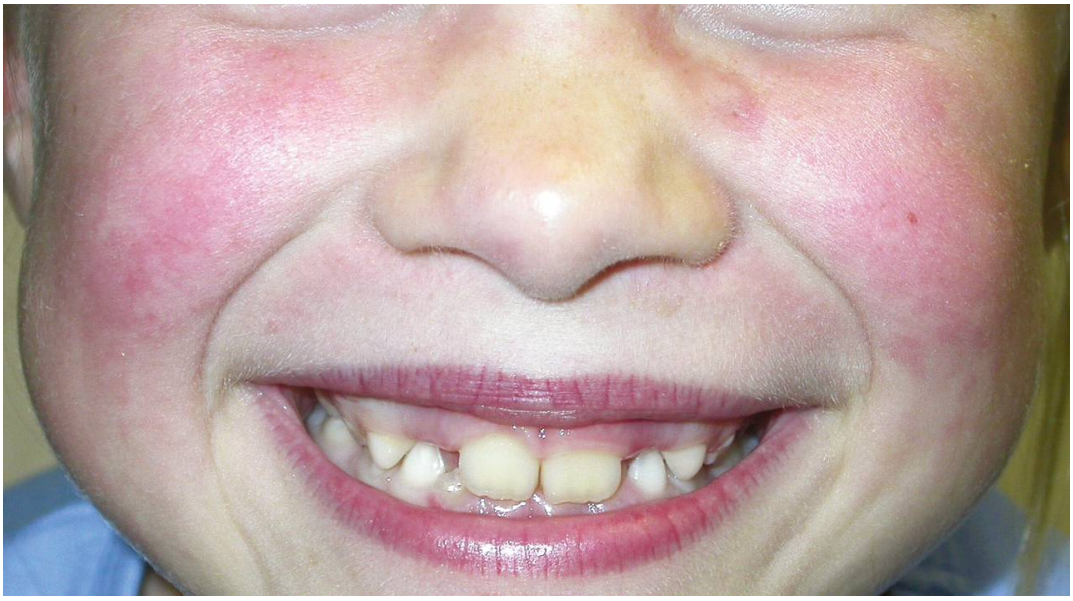
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Critique: 1



The rash of erythema infectiosum begins as confluent erythema of the cheeks, the so-called "slapped-cheek" appearance.

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Critique: 1



Erythema infectiosum is characterized by a lacy, reticulated erythema that appears on the extremities or trunk.

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Critique: 1



Papular purpuric "gloves and socks" syndrome is a unique manifestation of parvovirus B19 infection characterized by a petechial or purpuric eruption on the hands and feet.

Courtesy of dermatlas.org

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Critique: 1



In polymorphous light eruption, erythematous papules or vesicles appear in sun-exposed areas.

Courtesy of M. Rimsza

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Critique: 1



The eruption of scarlet fever is composed of tiny erythematous papules.

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Critique: 1



Patients who have systemic lupus erythematosus often have an erythematous, scaling malar eruption.

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Question: 2

An 8-month-old infant presents with the primary complaint of irritability. He has been exclusively breastfed since birth. His mother was not interested in providing any supplemental foods because her milk supply has been adequate. Physical examination reveals a fussy infant who has frontal bossing and whose weight and height are both at the 25th percentile. The infant becomes irritable with movement of the left arm. Arm radiography reveals a humeral fracture and bowing of both radii. Chest radiography demonstrates enlargement of the costochondral junctions.

Of the following, the MOST likely diagnosis is

A. congenital syphilis

B. osteogenesis imperfecta

C. vitamin D-deficient rickets

D. vitamin D-resistant rickets

E. vitamin E deficiency

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Critique: 2 Preferred Response: C

Older infants eating a restricted diet or receiving a single nutritional source may be at risk for micronutrient deficiencies. For example, children raised on vegan diets are at risk for vitamin B12 deficiency, and children fed with goat's milk are at risk for folate deficiency. The clinical presentation and radiographic findings (Item C2A) described for the exclusively breastfed infant in the vignette suggest vitamin D-deficient rickets. Congenital syphilis (Item C2B), vitamin D-resistant rickets, and osteogenesis imperfecta (Item C2C) all can present with skeletal abnormalities, but these conditions are less common than vitamin D-deficient rickets and typically have additional clinical findings. Vitamin E deficiency presents with neuropathy and hemolysis rather than skeletal abnormalities.

There are two primary forms of vitamin D: vitamin D2 is added to milk and multivitamin preparations, and vitamin D3 is synthesized endogenously in humans from 7-dehydrocholesterol via a photochemical reaction that requires exposure to the ultraviolet B radiation found in sunlight. Humans living at high latitudes produce very little vitamin D3 from November to March. Vitamin D is hydroxylated to 25-hydroxyvitamin D in the liver and converted to 1,25-dihydroxyvitamin D in the renal tubular epithelium. Vitamin D status usually is assessed by measurement of the 25-hydroxyvitamin D form; a concentration of less than 20 ng/mL (50 nmol/L) is considered vitamin D insufficiency, and less than 15 ng/mL (37 nmol/L) is considered deficiency. Biochemical complications of vitamin D deficiency include reduced calcium and phosphorus absorption, increased parathyroid hormone secretion, and phosphaturia. Initially, infants who have vitamin D-deficient rickets have normal serum calcium, low phosphorus, elevated alkaline phosphatase, and low 25-hydroxyvitamin D values.

Presenting clinical features of vitamin D-deficient rickets in infancy include fractures, irritability, and rarely, seizures from hypocalcemia. There is little vitamin D in human milk, and if an infant does not receive either vitamin D supplementation or adequate sun exposure, rickets may occur. African-American infants and children from higher latitudes are at increased risk for this condition. Therefore, the American Academy of Pediatrics Committee on Nutrition recommends the administration of 200 IU/d of supplementary vitamin D to infants older than 2 months of age until they are weaned to at least 500 mL/d of formula.

References:

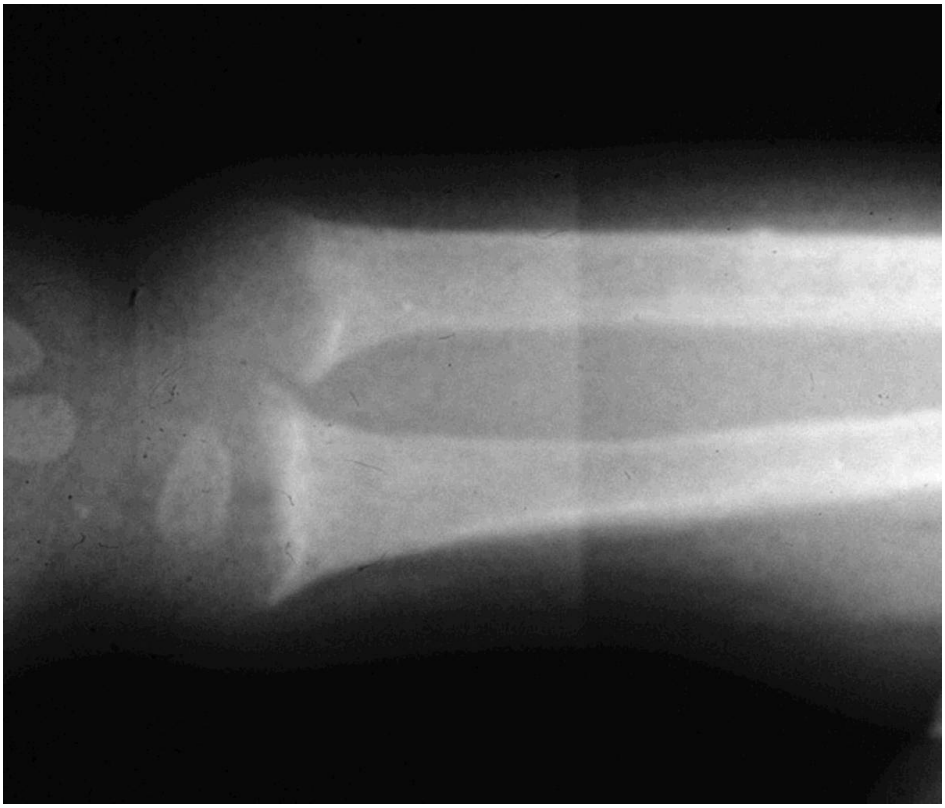
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Critique: 2

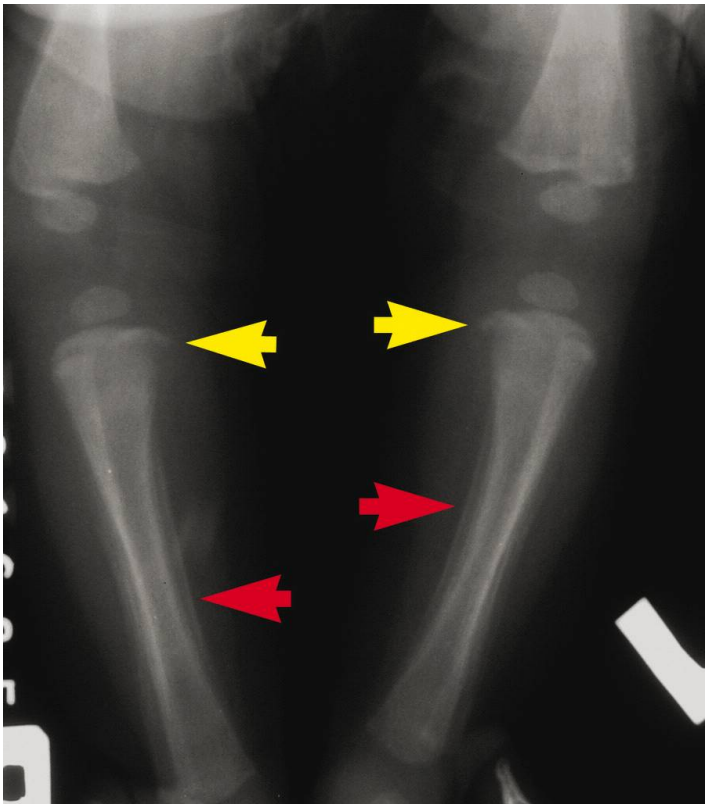


Rickets is characterized by metaphyseal cupping, fraying, and widening.

Courtesy of R. Schwartz

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Critique: 2

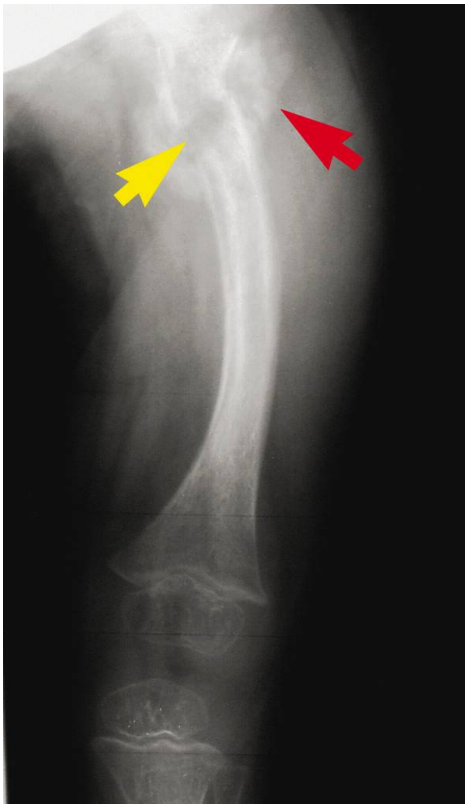


Anteroposterior radiograph of the lower extremities demonstrating two features of congenital syphilis: destruction of the medial aspects of the tibial metaphyses (yellow arrows) and subperiosteal new bone formation (red arrows).

Courtesy of D. Mulvihill

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Critique: 2



Anteroposterior radiograph of the femur in osteogenesis imperfecta showing osteopenia, bowing of the femur, and a proximal fracture (yellow arrow) with abundant callus formation (red arrow).

Courtesy of D. Mulvihill

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Question: 3

You are evaluating an 8-week-old infant whose birthweight was 1,000 g and who was delivered at 30 weeks' gestation. He experienced early respiratory distress and sepsis, but now these problems have resolved, and he recently progressed from parenteral nutrition to full enteral feedings.

Of the following, the feeding that will provide the BEST mineral content to ensure healthy bone development for this infant is

A. cow milk-based infant formula

B. human milk

C. premature formula

D. protein hydrolysate formula

E. soy protein-based formula

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Critique: 3 Preferred Response: C

Very low-birthweight (VLBW) preterm infants, such as the baby described in the vignette, are at risk for delayed bone mineralization due to constraints in delivering optimal nutrition to them while in the neonatal intensive care unit. A key component to bone health, mineralization, and overall nutritional well-being is the balance of calcium (Ca++) and phosphorous (P). Optimal Ca++ and P delivery that matches in utero accretion cannot be attained with total parenteral nutrition (TPN). Only after attaining full enteral nutrition goals can the desired delivery of Ca++ and P be provided and the infant's bone mineralization approach that of a healthy term infant.

Inadequate P delivery in the VLBW preterm infant results in demineralization of bone and metabolic bone disease (osteopenia, neonatal rickets). Such disease typically presents after 4 weeks of TPN and often is accompanied by normal serum P and Ca++ concentrations and elevated alkaline phosphatase activity. Excessive P delivery is uncommon in preterm infants, but may result in hypocalcemia, tetany, and seizure activity. Inadequate nutritional Ca++ delivery also can result in bone resorption as the body attempts to maintain normal serum Ca++ concentrations.

The optimal source of nutrition for the infant in the vignette should provide sufficient energy substrate (carbohydrate and lipid) and protein to facilitate growth and development as well as the necessary minerals and vitamins to help make up for delayed bone mineralization. Term infant cow milk-based formula lacks sufficient calories, protein, Ca++, P, and other trace minerals and vitamins, as does unsupplemented human milk. Term formulas and human milk require supplementation with a fortifier to meet these goals.

Formulas designed specifically for preterm infants contain higher caloric density; more readily absorbed lipids; greater protein content; and enriched Ca++, P, and other minerals and vitamins. They provide the best mineral content to ensure healthy bone development in VLBW preterm infants and generally do so by the time the infant attains a postconceptive age of 44 weeks. Protein hydrolysate formulas and soy protein-based formulas deliver suboptimal energy, protein, minerals, and vitamins to VLBW preterm infants and should be used only for a specific indication for a limited period of time.

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Question: 4

You are addressing a group of new mothers regarding infant feeding. One asks you when an infant can be switched from formula to whole cow milk.

Of the following, you are MOST likely to respond that whole cow milk

A. can be introduced at 6 months of age if an infant has significant gastroesophageal reflux

B. can be given at 9 months of age if the infant is also taking a wide variety of supplemental foods

C. may be given as a supplement at any age as long as the infant also receives human milk

D. should be avoided until 12 months of age because its iron content is absorbed poorly

E. should be avoided until 2 years of age because its caloric content is inadequate for optimal growth

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Critique: 4 Preferred Response: D

Iron-fortified formulas are the preferred nutrition for infants up to 12 months of age if a mother is unable or chooses not to breastfeed. These formulas contain 10 to 12 mg/L of iron, approximately 4% of which is absorbed by the infant. This amount of iron is sufficient to prevent iron deficiency in most term infants until 4 to 6 months of age. At this age, iron stores become depleted and supplemental foods, such as iron-fortified cereals, should be added.

The iron content of cow milk is approximately 0.5 mg/L, and although up to 10% of the iron is absorbed, it is inadequate to prevent iron deficiency, even if iron-fortified foods are added. In addition, cow milk may cause increased fecal blood loss in some infants, further exacerbating iron deficiency. Cow milk also has a higher content of protein and electrolytes, such as sodium and potassium, which results in a renal solute load that is too high for the infant kidney. For these reasons, cow milk is not recommended until 12 months of age. It is appropriate to switch to whole cow milk at this time because the caloric content (19 kcal/oz) is adequate for growth at this age, and the child's diet generally includes more iron-containing foods.

The introduction of cow milk does not prevent or treat gastroesophageal reflux and should not be recommended for this condition. Breastfeeding mothers wishing to provide additional nutrition because of inadequate milk supply or other reasons should be advised to use iron-fortified formulas for supplementation.

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Question: 5

An 18-month-old girl who has been adopted from Russia presents to your office for an initial health supervision visit. The mother notes that the girl limps, and she thinks one leg is longer than the other. There is no history of medical attention for this problem in the adoption documentation. Review of available medical history and the current physical examination reveal no signs of illness. The child appears to be otherwise growing and developing normally. On physical examination, you measure a leg length discrepancy of 2 cm, and the girl has a "waddling" type limp and difficulty in abducting her right thigh at the hip.

Of the following, the MOST likely diagnosis is

A. developmental dysplasia of the hip

B. femoral anteversion (internal femoral torsion)

C. Legg-Calvé-Perthes disease

D. slipped capital femoral epiphysis

E. vitamin D-dependent rickets

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Critique: 5 Preferred Response: A

Leg length discrepancy in a child may result from fracture, dislocation, bone infection, vascular malformation, or any process that may interfere with bone growth. It is seen most often in developmental dysplasia of the hip, which is found in 1 in 5,000 children by the age of 18 months.

Although abnormal hip abduction on physical examination in early infancy is the usual finding associated with hip dysplasia, older infants and children, especially those who walk, may present with waddling gait. One reason for the change in nomenclature from “congenital” to “developmental” hip dysplasia is that a subpopulation of children seems to present in later infancy or toddlerhood with hip dysplasia despite no findings on physical examination. Development of the femoral head and acetabulum are intimately related, and normal adult hip joints depend on further growth of these structures. Hip dysplasia may occur in utero, perinatally, or during infancy. Asymmetric thigh or gluteal folds (Item C5A), better observed when the child is prone; apparent limb length discrepancy, as described for the girl in the vignette; and restricted motion, especially abduction, are significant, albeit not pathognomonic signs.

Developmental dysplasia usually is identified with abnormal Ortolani (Item C5B) and Barlow (Item C5C) maneuvers in early infancy and abnormal abduction until about 3 to 4 months of age. Older infants and children may present with waddling gaits.

Leg length discrepancy should prompt ultrasonographic evaluation (Item C5D) of the hip in the infant younger than 12 weeks of age. Plain radiographs (Item C5E) with anteroposterior and frogleg views of the hip in the older infant and toddler can reveal hip dysplasia, healed fractures, ossification, and other abnormalities.

Failure to identify and treat developmental dysplasia of the hip may result in degenerative changes of the hip and arthritis.

A number of other disorders may cause abnormal gait. Rickets normally presents with bowing of the legs, along with a “waddling” gait, and may be accompanied by other findings, such as swelling of the wrists or ankles or a “rachitic rosary” (enlargement of the costochondral junctions). Legg-Calvé-Perthes disease (Item C5F) may cause pain and a limp, but typically occurs in children 2 to 12 years of age (mean age, 7 years). Slipped capital femoral epiphysis (Item C5G) is a disease of the older, heavier preadolescent and is not seen in the toddler/preschool age group. In contrast to these conditions, femoral anteversion (internal femoral torsion) causes intoeing, not a "waddling" gait or leg-length discrepancy. Typically, children present at 2 years of age or older and have increased internal rotation of the hips when examined in the prone position.

References:

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Scherl S. Common lower extremity problems in children. *Pediatr Rev*. 2004;25:52-62. Available at: http://pedsinreview.aappublications.org/cgi/content/full/25/2/52

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Critique: 5

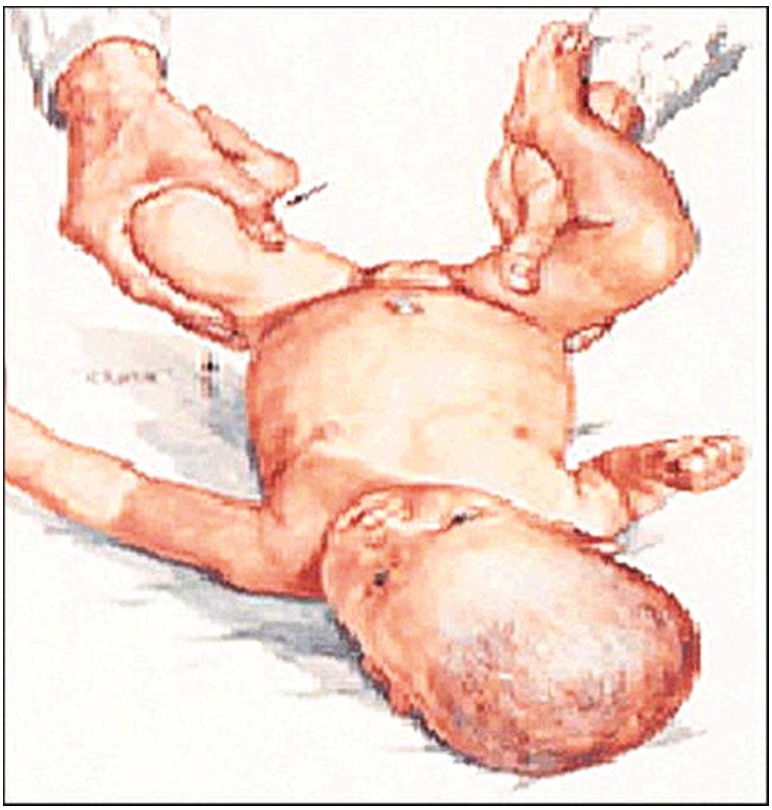


Asymmetry of the thigh folds in developmental dysplasia of the hip. There is an additional skin fold on the affected (right) side.

Courtesy of W. Accosti

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Critique: 5

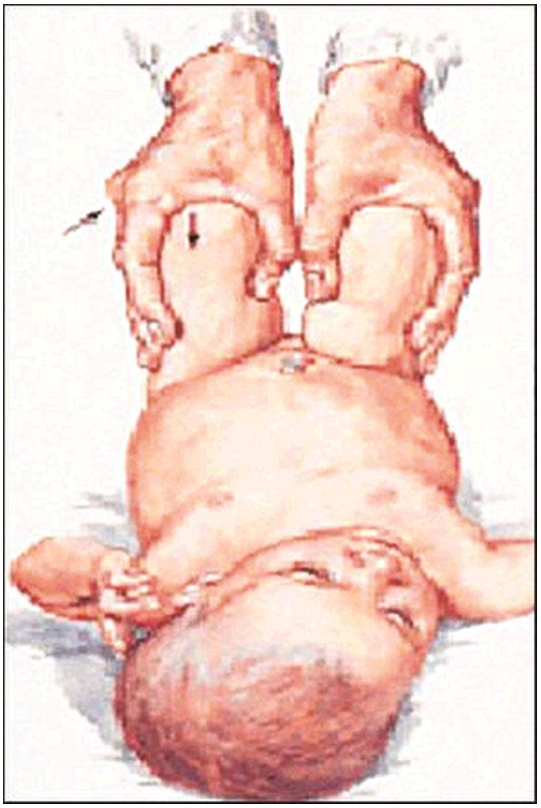


In the Ortolani test, the hip is flexed and abducted and the femoral head is lifted anteriorly into the acetabulum. The maneuver relocates a dislocated hip, resulting in a "clunk" that may be heard and felt.

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Critique: 5

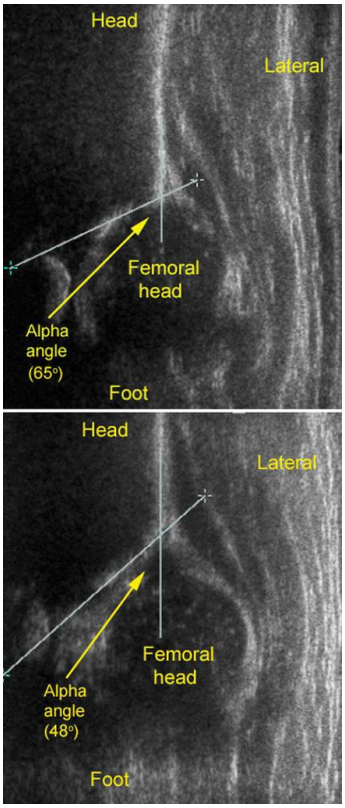


The Barlow test dislocates the hip. The hip is flexed and adducted. Applying a posterior force (pushing down) produces a "clunk" as the femoral head dislocates posteriorly.

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Critique: 5



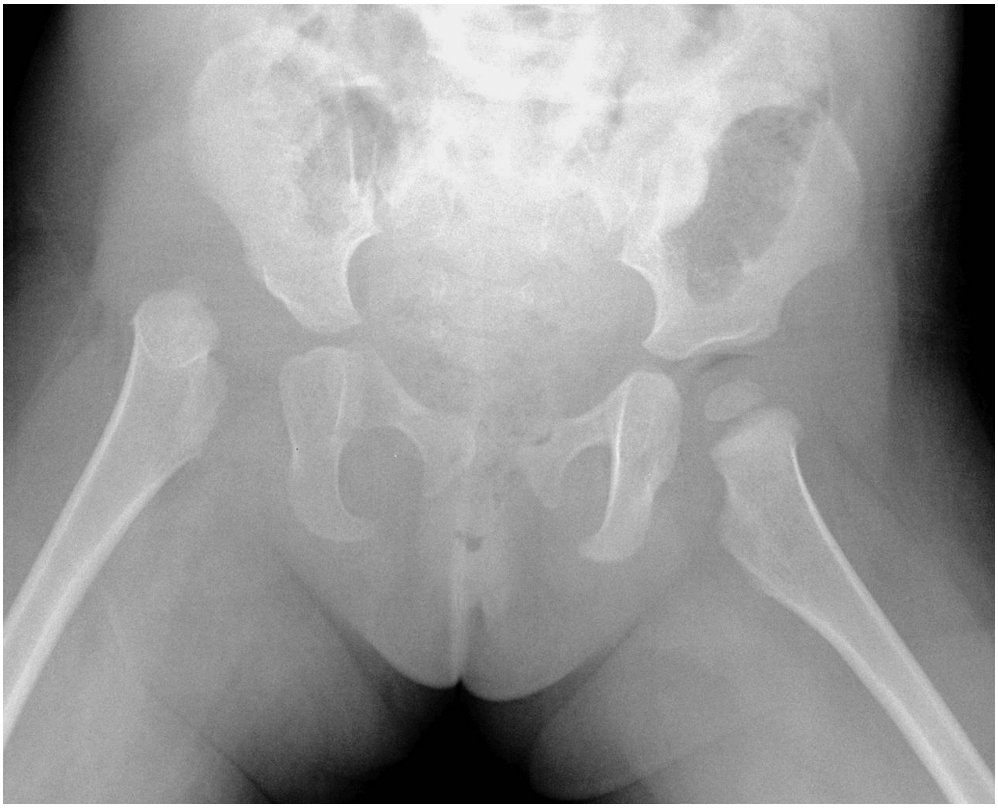
Ultrasonography of the normal hip (top) reveals that more than 50% of the femoral head is covered by the acetabulum. The acetabulum is deep, and the alpha angle is 60 degrees or greater.

Ultrasonography of the hip in developmental dysplasia (bottom) shows that the acetabulum is shallow, resulting in an alpha angle of 50 degrees or less (48 degrees in this patient).

Courtesy of D. Mulvihill

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Critique: 5

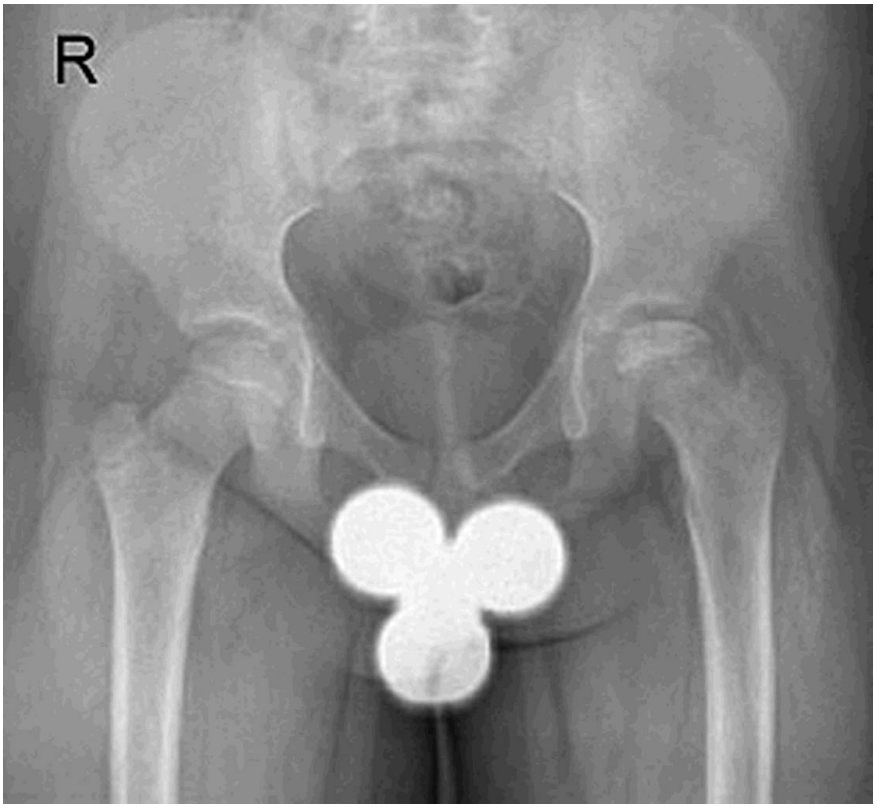


Developmental dysplasia of the hip. The right acetabulum is shallow and steep in orientation. The right hip is dislocated superolaterally. The right ossification center is smaller than the left.

Courtesy of D. Krowchuk

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Critique: 5

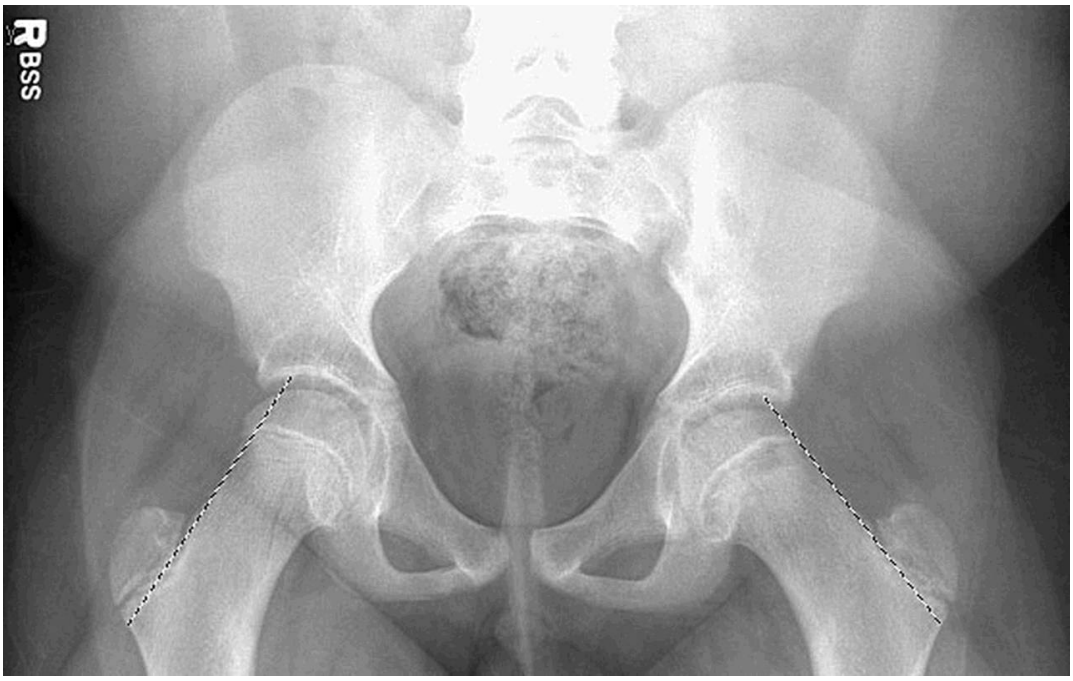


Legg-Calvé-Perthes disease: Anteroposterior view of the hips demonstrates sclerosis around the left femoral epiphysis with flattening of the femoral head. The left medial joint space is widened, and there is demineralization of the left proximal femur compared with the right.

Courtesy of D. Krowchuk

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Critique: 5



Slipped capital femoral epiphysis is characterized by an upward anterior movement of the femoral neck on the capital epiphysis (which becomes displaced posteriorly and inferiorly). In the normal hip (right), a line drawn along the superior margin of the femoral neck transects a portion of the ossified epiphysis. This does not occur on the affected side (left).

Courtesy of D. Krowchuk

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Question: 6

A 2-week-old infant presents to the emergency department with a 1-day history of decreased feeding, pallor, diaphoresis, and increasing somnolence. He was born at term, and the delivery was uncomplicated. On physical examination, his heart rate is 190 beats/min, his respiratory rate is 80 breaths/min, his blood pressure is 50/30 mm Hg, and his extremities are cool and pale with poor pulses. You place the infant on a cardiorespiratory monitor and begin your assessment and management.

Of the following, the MOST appropriate pair of tests to consider in this child is

A. complete blood count and bone scan

B. electrocardiography and echocardiography

C. electroencephalography and head ultrasonography

D. lumbar puncture and head computed tomography scan

E. serum electrolytes and chest computed tomography scan

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Critique: 6 Preferred Response: B

The child described in the vignette is in shock and has clinical signs and symptoms of diminished systemic perfusion. His heart rate is elevated to increase cardiac output (cardiac output = heart rate x stroke volume). Shock has several causes in the neonate, including hypovolemia, sepsis, and metabolic and cardiogenic dysfunction. Differentiating these causes upon presentation in the neonate can be difficult.

Although it is appropriate for the infant described in the vignette to receive empiric broad-spectrum antibiotics against the most likely infectious organisms in this age group, performing a lumbar puncture may compromise the infant's cardiorespiratory status further and is contraindicated. Because the findings in this infant are most consistent with cardiogenic shock, an electocardiogram and echocardiogram should be obtained as soon as possible. These tests can be performed at the bedsides of even the most critically ill infants. There is no reason to perform computed tomography scan, bone scan, or electrocephalography in this infant. Although a complete blood count and electrolytes should be measured, it is unlikely that these test will aid in the immediate management of the infant described in this vignette.

In the first few weeks after birth, cardiogenic shock often is due to cardiac anomalies. Congenital heart lesions that may lead to such a dramatic presentation include those that require ductal patency for the maintenance of systemic blood flow. Among these are critical aortic stenosis, aortic coarctation, interruption of the aortic arch, and hypoplastic left heart syndrome. In each of these left heart obstructive lesions, persistence of the fetal shunting pattern through the ductus arteriosus from the pulmonary artery to the aorta provides the necessary blood flow to the aorta to maintain end-organ and cellular perfusion. As the ductus arteriosus begins to close in the hours and days after delivery, the volume of blood delivered to the systemic circulation diminishes in patients who have severe left heart obstruction. Although the ductus arteriosus closes by the fourth postnatal day in approximately 90% of neonates, in some infants, closure does not occur for several weeks. Therefore, left heart obstructive lesions should be considered in any neonate who presents with congestive failure and shock, and administration of prostaglandin E to establish or maintain ductal patency should be considered.

Head ultrasonography may be important in determining this child's prognosis after stabilization, but it cannot aid in diagnosis and acute management.

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Question: 7

A 7-year-old boy presents for evaluation of attention-deficit/hyperactivity disorder, following the suggestion of his first-grade teacher. The child's academic and behavioral function were described as normal in preschool and kindergarten, but this year he constantly talks out of turn, does not stay in his chair, and has frequent emotional "melt-downs." His mother says his memory is poor and grades are declining. On physical examination, you note very brisk reflexes in the arms and legs, two beats of clonus at the ankles, and slow and clumsy fine-motor movements.

Of the following, the most appropriate initial diagnostic test is

A. brain magnetic resonance imaging

B. electroencephalography

C. electromyography

D. serum creatine kinase measurement

E. serum lactate measurement

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Critique: 7 Preferred Response: A

Although most children presenting for evaluation for attention deficit/hyperactivity disorder (ADHD) do not have a degenerative central nervous system (CNS) disease, the child described in the vignette has history and physical examination findings that should prompt a thorough diagnostic evaluation for a neurologic disease. There are several areas in which classic ADHD and degenerative diseases may overlap.

The tasks of sustaining attention and controlling behavioral impulses localize to the frontal lobes, the most complex area of the brain and the last region to mature fully. Cerebral maturation in children typically results in increasing efficiency at these tasks. ADHD is a diagnostic label applied to children whose abilities in these areas are developmentally inappropriate. Relative to their peers, children who have ADHD are less efficient at focusing on complex cognitive tasks and may be more physically hyperactive and behaviorally impulsive.

Most children whose cognitive and behavioral functions fit ADHD diagnostic criteria have an "idiopathic" problem. The biology of this condition involves abnormal "hard wiring" of brain circuits or abnormal neural transmission within brain circuits due to a complex array of genetic and environmental factors. Results of physical and neurologic examinations of children who have idiopathic ADHD generally are normal, with the possible exception of subtle neurologic signs involving fine or gross motor skills. No laboratory tests, neuroimaging, or electroencephalography (EEG) are required to confirm the diagnosis of idiopathic ADHD.

Although the boy described in the vignette has some of these problems, two of his features are atypical for idiopathic ADHD and mandate a medical diagnostic evaluation for potential CNS degenerative disease: 1) a clear decline in cognitive and behavioral function, and 2) abnormal reflexes and clonus. Although parental and teacher reports of decline in cognitive and behavioral function often are ambiguous and may be related to changes in the home or school environment, such a functional decline still requires evaluation. The findings of abnormal reflexes and clonus described for the boy, which presumably were not noted during prior health supervision visits, localize to "upper motor neurons" and can aid in making the appropriate decision about diagnostic testing. Magnetic resonance imaging (MRI) is the preferred diagnostic test because it provides information about the structure of both gray and white matter. Abnormalities identified in one or both tissues (Item C7) can guide further molecular diagnostic studies. For example, characteristic findings in white matter might lead to a search for peroxisomal disorders such as adrenoleukodystrophy.

EEG typically provides nonspecific information such as "slowing" and may show normal results in this case. In the absence of a history of epilepsy, its value is low in evaluating suspected CNS degenerative disease. Electromyography provides information about intrinsic pathology in muscle and signaling of the peripheral nervous system to muscle. Although the boy in the vignette could have a disease affecting muscles or nerves, neurologic examination findings do not support this possibility. For the same reason, measurement of serum creatine kinase, whose concentrations typically are elevated in muscle disease, likely would not be helpful. Serum lactate concentrations might be abnormally elevated, particularly in the presence of a mitochondrial disease, but the yield in this clinical setting is low. Such testing may be indicated based on findings from MRI.

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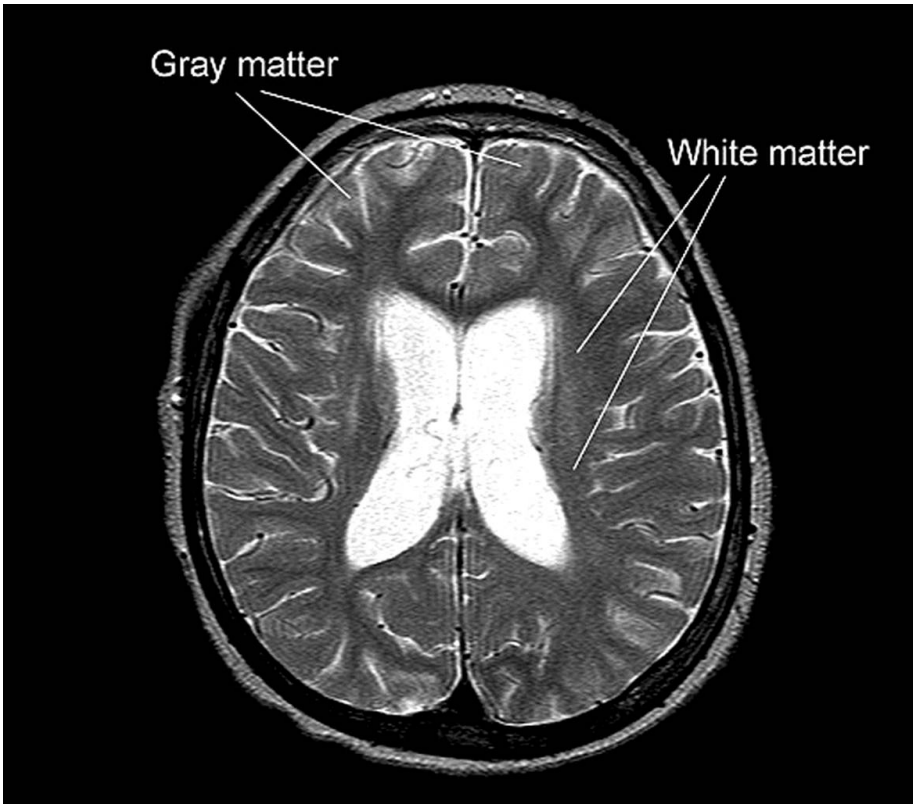
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Critique: 7



Axial magnetic resonance imaging of the brain showing ventricular enlargement and diffuse reduction in white matter.

Courtesy of D. Mulvihill

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Question: 8

You are urgently called to the newborn nursery to evaluate a 3-day-old term male infant who is lethargic. The baby was taking formula well for the first 2 days but vomited after his last 2 feedings and has become increasingly difficult to arouse. A review of the record reveals that he was born at 36 weeks' gestation to a 30-year-old gravida 2, now para 2 woman. The mother is Rh-negative and received Rh immune globulin during the pregnancy. Pregnancy, labor, and delivery were uneventful; Apgar scores were 8 and 9 at 1 and 5 minutes. Family history is noncontributory. On physical examination, the baby appears well developed, is very jaundiced, and has hypotonia and tachypnea. He responds with a grimace to sternal rubbing but does not arouse to voice or to touch. Findings on abdominal examination are normal. You order a series of laboratory tests, start intravenous fluids, and arrange for transfer to the neonatal intensive care unit. A laboratory technician subsequently notifies you of some critical laboratory values, including a platelet count of 35x103/mcL (35x109/L), serum carbon dioxide of 4 mEq/L (4 mmol/L), anion gap of 28 mEq/L (28 mmol/L), serum ammonia of 250 mcmol/L, and total bilirubin of 20 mg/dL (342 mcmol/L). There are large ketones in the urine.

Of the following, the MOST likely diagnosis is

A. bilirubin encephalopathy

B. citrullinemia

C. hypoxic-ischemic encephalopathy

D. propionic acidemia

E. transient hyperammonemia of the newborn

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Critique: 8 Preferred Response: D

The newborn who develops seizures, lethargy, or coma during the first days after birth poses an important diagnostic dilemma. Because these symptoms can be associated with sepsis and asphyxia, the infant should be treated with these diagnoses in mind. However, the possibility of an inborn error of metabolism must be considered early to allow laboratory evaluation and optimally effective treatment that can reduce morbidity and mortality. It is also important to remember that certain inborn errors predispose to sepsis (eg, galactosemia and propionic acidemia). Therefore, the diagnosis of sepsis does not exclude the possibility of an inborn error.

Laboratory results may provide important clues to the underlying diagnosis. Metabolic acidosis with an increased anion gap is suggestive of an organic acidemia. Infants who have organic acidemias typically have had uneventful deliveries and initially appear normal. Bone marrow suppression is seen in some organic acidemias, and the platelet count of 35x103/mcL (35x109/L) reported for the infant in the vignette also supports this possibility. Propionic acidemia is caused by defective activity of the enzyme propionyl CoA carboxylase that results in the inability to break down numerous amino and fatty acids and cholesterol, thereby compromising the citric acid cycle. Affected individuals have large ketones in the blood and urine, and the urea cycle also is affected negatively, resulting in hyperammonemia in some cases. Hyperbilirubinemia of the newborn may be exacerbated by dehydration associated with vomiting. Precise diagnosis of a suspected organic acid disorder is based on urine organic and amino acid and plasma amino acid test results.

Citrullinemia is an inborn error of urea cycle function caused by defective production or formation of the enzyme argininosuccinate synthetase that results in massive hyperammonemia, with concentrations often exceeding 560 mcg/dL (400 mcmol/L). Affected infants typically present in the first few days after birth with poor feeding, vomiting, lethargy, and coma. There may be hepatomegaly. In addition to elevated serum ammonia values, serum glutamic-oxaloacetic transaminase and glutamic-pyruvic transaminase concentrations often are elevated, and prothrombin time and partial thromboplastin time may be prolonged. Metabolic acidosis is not present; respiratory alkalosis is more typical. Precise diagnosis is based on the finding of very elevated citrulline values on plasma amino acid analysis.

Although no absolute concentration of serum total bilirubin (STB) is linked with certainty to neurotoxicity, it is highly unlikely that a term infant whose STB concentration is less than 30 mg/dL (513 mcmol/L) will suffer neurologic damage. Newborns who have bilirubin-induced encephalopathy present with poor suck, stupor, hypotonia, and seizures toward the end of the first week after birth, followed by hypertonia. Other than elevated serum bilirubin concentrations, there are no typical metabolic derangements.

Neonatal hypoxic-ischemic encephalopathy (HIE) is caused by diminished oxygen delivery to the brain due to any number of processes, including birth asphyxia, postnatal respiratory insufficiency, severe left-to-right shunt, cardiac insufficiency, and loss of cerebrovascular autoregulation. Affected infants are more likely to have had complications during labor and delivery than infants who have inborn errors of metabolism. Clinical features vary as time passes and include stupor or coma, seizures, and fluctuating tone. Metabolic derangements may include lactic acidosis with a normal anion gap, hypoglycemia, hypocalcemia, hyponatremia, and inappropriate secretion of antidiuretic hormone. Hyperammonemia also may be present, possibly due to increased protein catabolism. Hypertonia in the neonate who has altered sensorium often is associated with HIE.

Transient hyperammonemia of the newborn is a rare and poorly understood condition that is seen almost exclusively in preterm infants who have symptomatic pulmonary disease. Affected infants may become comatose and develop seizures in the first 24 hours after birth and before the introduction of dietary protein. As in other conditions, hyperammonemia should be treated aggressively. Despite such treatment, however, 20% to 35% of infants who have such transient hyperammonemia die.

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Question: 9

A 15-year-old girl comes to your office with the complaint of a vaginal discharge. She states that she is sexually active with a new partner and has noted a cream-colored, thin discharge. She also has had some burning with urination and vaginal itching. On pelvic examination, you note reddened labia majora and minora; a frothy, foul-smelling discharge in the vagina (Item Q9A); and a cervix that has small erosions and petechiae. Her pregnancy test results are negative, and microscopic evaluation of vaginal secretions shows motile organisms (Item Q9B).

Of the following, the MOST appropriate treatment is

A. azithromycin 1 g orally in a single dose

B. boric acid 600 mg capsule BID intravaginally for 14 days

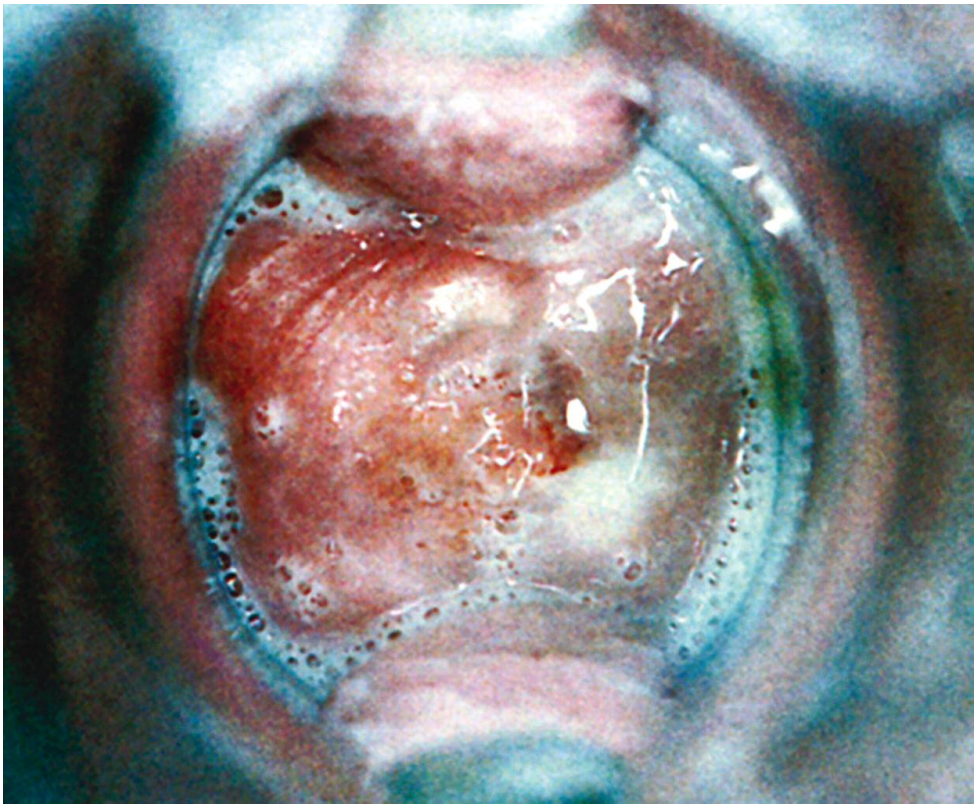
C. clindamycin 300 mg BID orally for 7 days

D. fluconazole 150 mg orally in a single dose

E. metronidazole 2 g orally in a single dose

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Question: 9



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Critique: 9 Preferred Response: E

Sexually active 15- to 19-year-olds have the highest rates of many sexually transmitted infections (STIs). Many STIs manifest as abnormal vaginal discharge in females. Trichomoniasis, caused by the protozoan *Trichomonas vaginalis*, is characterized by a diffuse, malodorous, frothy, yellow-green discharge (Item C9A). There also can be vulvar irritation. However, some young women have minimal or no symptoms.

Trichomoniasis is diagnosed most commonly by microscopic evaluation of vaginal secretions. As demonstrated for the young woman described in the vignette, trichomonads are seen on microscopy as motile bodies that have three to five anterior flagella and one posterior flagellum (Item C9B). When the wet preparation slide is evaluated immediately, microscopic sensitivity is approximately 60% to 70%. Other United States Food and Drug Administration-approved tests for trichomoniasis in women include immunochromatographic capillary flow dipstick technology and a nucleic acid probe, which have greater than 83% sensitivity and greater than 97% specificity. Culture is the most sensitive and specific method of diagnosis.

T vaginalis infects the vaginal epithelium and other sites, most commonly the urethra (82.5% of cases) and periurethral glands (98% of cases). The organism almost always is transmitted sexually, although it can be transmitted through fomites.

According to the Centers for Disease Control and Prevention *Sexually Transmitted Disease Treatment Guidelines, 2006*, the recommended treatment regimens for trichomoniasis are single oral doses of metronidazole 2 g or tinidazole 2 g. The recommended alternative regimen is metronidazole 500 mg orally twice a day for 7 days. Alcohol use should be avoided during treatment with both of these medications. Topically applied metronidazole gel is considerably less efficacious for the treatment of trichomoniasis. Follow-up is unnecessary for men and women who become asymptomatic after treatment or who are initially asymptomatic. Sexual partners should be treated as well.

Azithromycin, boric acid, clindamycin, and fluconazole are not appropriate treatments for trichomoniasis.

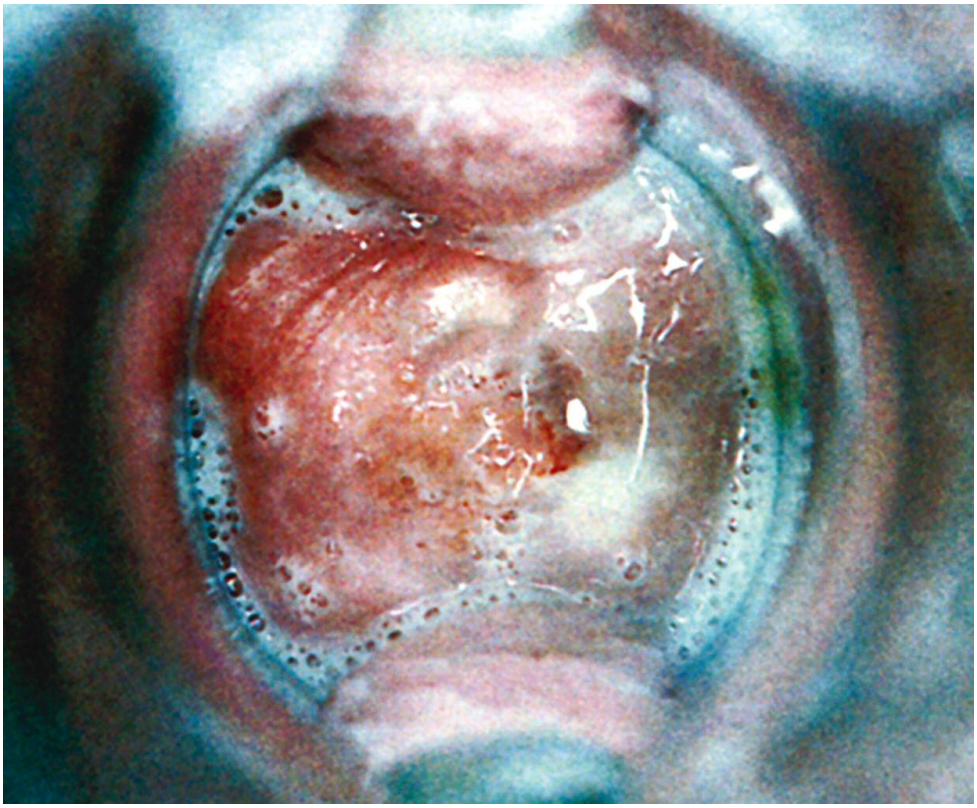
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Critique: 9



Vaginal discharge caused by infection with *Trichomonas vaginalis* often appears frothy.

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Question: 10

During the health supervision visit of a 5-year-old girl, you notice pubic hair (Sexual Maturity Rating 3). Her height is at the 75th percentile and weight is at the 95th percentile. She has no acne or clitoromegaly. Her mother tells you the girl developed an adult body odor around 8 months ago, and the mother noticed the pubic hair about 6 months ago. She adds that the pubic hair is a little more noticeable now than when she first saw it.

Of the following, the MOST important initial screening study is

A. bone age radiograph

B. measurement of dehydroepiandrosterone sulfate (DHEA-S)

C. measurement of 17-hydroxyprogesterone

D. measurement of testosterone

E. pelvic and abdominal ultrasonography

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Critique: 10 Preferred Response: A

Early adrenal puberty (adrenarche) is the usual reason for slow development of pubic and axillary hair without evidence of rapid masculinization (increased growth rate, clitoral enlargement, acne) in children older than 4 years of age. The growth rate is stable in such children, and the only signs of masculinization are usually adult body odor followed by increased pubic and axillary hair, as described for the girl in the vignette. The first step in evaluation of such children is to determine their bone age. The bone age rarely is more than 1 year advanced beyond chronologic age, and if bone radiographs document that fact, usually only clinical follow-up is necessary.

If measured, dehydroepiandrosterone sulfate (DHEA-S) concentrations are somewhat elevated in children experiencing early adrenal puberty, but this is not an abnormal finding in children older than age 4 years. The adrenarchal increase in DHEA-S commences between 4 and 6 years. Testosterone concentrations are low and are unmeasurable with present commercial assays. However, because many laboratory assays are unstable in the low ranges, a report of a low measurable testosterone value that is within the limit of error for the laboratory assay can be alarming.

If the bone age radiograph is advanced more than 1 year beyond chronologic age, there is a possibility that the patient may have late-onset congenital adrenal hyperplasia (CAH). This condition is caused most commonly by mild 21-hydroxylase deficiency and is associated with an elevated 17-hydroxyprogesterone value either at baseline or following adrenocorticotropic hormone (ACTH) stimulation. Because blood-drawing itself may provoke ACTH release and, therefore, adrenal stimulation, if an unstimulated 17-hydroxyprogesterone value is only modestly elevated, an ACTH stimulation test is needed to rule out CAH. Therefore, many endocrinologists prefer obtaining an ACTH test as the next study if bone age is advanced in children who have premature adrenarche. At this time, a serum testosterone measurement also may be obtained. Ultrasonographic studies of the adrenals and ovaries might reveal an androgen-producing tumor, but this usually is accompanied by more obvious signs of virilization than described for the child in the vignette and could include clitoromegaly, acne, and rapid growth. Ultrasonography of the adrenals is not useful in the diagnosis of CAH.

Premature adrenarche is more common in overweight children and may be associated with insulin resistance. It may be a harbinger of polycystic ovary syndrome in some adolescent girls.

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Question: 11

The parents of a 15-year-old boy previously diagnosed as having constitutional delay of growth and puberty are concerned that their son is being bullied by his 10th-grade classmates. His grades have declined from As to Bs, and he says he dislikes school. He has been in good health, and he eats and exercises in moderation. On physical examination, his height and weight are at the 10th percentile, and his genitalia are at Sexual Maturity Rating 1. When you speak to the boy in private, he becomes tearful, complaining that he is afraid to change for gym and that his friends no longer wish to be with him. He and his parents ask for your advice in this situation.

Of the following, the BEST response is to

A. offer reassurance and arrange for follow-up evaluation in your office in 6 months

B. recommend caloric supplements

C. refer the family for behavioral counseling

D. refer the boy for a psychoeducational evaluation

E. refer the boy to an endocrinologist for re-evaluation and possible hormonal therapy

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Critique: 11 Preferred Response: E

A teenager is considered to have delayed puberty if there is no evidence of Sexual Maturity Rating stage 1 genital development by 13 years in girls and 14 years in boys. Adolescents who present with evidence of delayed pubertal development, such as the boy described in the vignette, require medical evaluation and need to be screened for difficulties in psychosocial functioning. Individuals who have notable delays may have feelings of poor self-image and low self-esteem. They may be teased or bullied by their peers, leading to withdrawal from social activities as well as parental overprotection. Their school performance may decline, and they may wish to avoid attending school. Boys who look younger than their chronologic age have fewer opportunities to engage in activities with their peers and may feel unpopular. Girls experience less of an emotional impact from delays in puberty.

The amount of intervention in patients who have constitutional delay should be based, in part, on the amount of emotional stress expressed by the adolescent. If puberty is expected to occur and the individual has minimal emotional impact, simple reassurance may be enough. Adolescents who have significant emotional distress, such as the boy in the vignette, may benefit from hormonal therapy to initiate or accelerate puberty. Therefore, referral to an endocrinologist is indicated.

Additional caloric intake is not appropriate for an adolescent whose weight and height are normal; such an intervention will not accelerate his rate of pubertal development. A psychoeducational evaluation is not indicated for this boy because although his grades are declining, they still are above average. Behavioral counseling may help the boy cope with his current level of distress, but it will not address his delayed puberty.

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Question: 12

A 16-year-old girl presents to the clinic with a 6-day history of low-grade fever and cough. On physical examination, she has a temperature of 100.6°F (38.1°C) and widespread crackles throughout her lung fields. You believe she has a “walking pneumonia” caused by *Mycoplasma pneumoniae*.

Of the following, the MOST accurate method used to establish the diagnosis is

A. polymerase chain reaction

B. serum cold agglutinins

C. serum *Mycoplasma* titers

D. sputum culture

E. sputum Gram stain

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Critique: 12 Preferred Response: C

Diagnosing disease caused by *Mycoplasma pneumoniae* can be difficult. Although this organism can be grown in special enriched broth or agar media, it takes up to 21 days to recover. Polymerase chain reaction tests have been developed and appear to be sensitive and specific, but they have not yet been standardized. Serum cold agglutinins is a popular test, but it is only positive in about 50% of patients who have pneumonia caused by *M pneumoniae,* and false-positive results can occur due to cross-reactivity with other agents (eg, adenovirus, Epstein-Barr virus).

Commercially available kits for detecting specific *M pneumoniae* immunoglobulin (Ig)M and IgG antibodies currently are the most accurate diagnostic method. Although the presence of IgM antibody confirms a recent infection due to *M pneumoniae*, these antibodies persist in the serum for several months and may not indicate a current infection. Therefore, test results should be interpreted in conjunction with findings on the clinical history and physical examination. *M pneumoniae* is not detected on sputum Gram stain.

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Question: 13

You are evaluating a 12-year-old girl who has a 1-month history of daily fevers (up to 104ºF [40°C]), cervical adenopathy, severe malaise, headache, and lower back pain. She lives at home with her parents and two sisters, all of whom have been well. She has a 5-year-old cat and two birds for pets. Six months ago, she spent 2 weeks visiting relatives who live on a ranch in Mexico where she learned to milk the cows, feed the pigs and chickens, and ride horses. She also sampled the local cuisine. Physical examination reveals a febrile, tired-appearing girl who is having rigors. She has diffuse 1 x 1-cm nontender cervical adenopathy, with splenomegaly and tenderness to palpation of her lower back. Her white blood cell count is 4.9x103/mcL (4.9x109/L) with 31% polymorphonuclear leukocytes, 15% band forms, 48% lymphocytes, and 6% monocytes; erythrocyte sedimentation rate is 70 mm/hr; and C-reactive protein concentration is 6.8 mg/dL.

Of the following, the MOST likely diagnosis is

A. brucellosis

B. cat-scratch disease

C. Epstein-Barr virus mononucleosis

D. leptospirosis

E. toxocariasis

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Critique: 13 Preferred Response: A

Zoonoses are a group of diseases caused by a diversity of pathogenic microorganisms that ordinarily reside and cause disease in nonhuman animals. The defining criteria for a zoonotic disease are that they have a vertebrate reservoir exclusive of humans and that they are transmitted by an agent directly to people from products derived from the host animal or through an arthropod intermediate. Item 13 (Item C13) shows some common zoonoses that are acquired directly or indirectly from animals or arthropod vectors and their modes of transmission.

The findings described for the patient in the vignette are consistent with brucellosis, which she probably acquired from the animals with which she had contact on the ranch. Toxocariasis usually is characterized by cough, leukocytosis, and eosinophilia. Leptospirosis is an acute febrile illness associated with nonpurulent conjunctivitis, headache, myalgias of the calf and lumbar regions, aseptic meningitis, uveitis, muscle tenderness, and rash. Epstein-Barr virus mononucleosis usually involves exudative pharyngitis, lymphadenopathy, and atypical lymphocytosis. The patient has no history of being scratched or bitten by her cat, and cat-scratch disease is transmitted most commonly by kittens or young cats.

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Critique: 13

Item13: Common Zoonoses

Disease	Pathogen	Route of Transmission
Anthrax	*Bacillus anthracis*	*Contact, ingestion, respiratory
Brucellosis	*Brucella sp*	Contact, ingestion, respiratory
Campylobacteriosis	*Campylobacter jejuni*	Ingestion
Cat-scratch disease	*Bartonella henselae*	Contact
Leptospirosis	*Leptospira interrogans*	Contact, ingestion
Lyme disease	*Borrelia burgdorferi*	**Arthropod vector
Plague	*Yersinia pestis*	Arthropod, contact, respiratory
Rabies	*Rhabdovirus*	Contact
Rickettsiosis	*Rickettsia sp*	Arthropod
Arboviral disease	Different viruses	Arthropod
Giardiasis	*Giardia lamblia*	Ingestion
Cysticercosis	*Taenia solium*	Ingestion
Toxocariasis	*Toxocara canis, T cati*	Contact
Toxoplasmosis	*Toxoplasma gondii*	Ingestion

*Bites, scratches or water

**Mosquito, tick, flea, mite

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Question: 14

A male infant who is experiencing failure to thrive and hypernatremic dehydration is admitted to the hospital. After administration of intravenous fluids, euvolemia is restored and the serum sodium is normalized to 140 mEq/L (140 mmol/L). A water deprivation test results in a 5% loss of body weight over 4 hours and an increase in serum sodium concentration to 145 mEq/L (145 mmol/L) and serum osmolality to 310 mOsm/kg (310 mmol/kg). Simultaneous urine osmolality is 50 mOsm/kg (50 mmol/kg). Subcutaneous administration of desmopressin does not reduce urine output or increase urine osmolality. You decide to place the infant on a formula that possesses a low renal solute load.

Of the following, the statement that BEST describes the properties of infant formulas as they relate to renal solute load is that

A. human milk has a greater renal solute load than do cow milk-based formulas

B. the carbohydrate composition of the formula increases the renal solute load

C. the fat composition of infant formulas more greatly alters the renal solute load when the medium-chain triglycerides component is increased

D. the primary minerals involved in renal solute load are sodium, potassium, chloride, and phosphorus

E. the protein composition of the formula has no effect on renal solute load

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Critique: 14 Preferred Response: D

The inappropriately low urine osmolality in the setting of elevated serum osmolality for the boy in the vignette indicates a urinary concentrating defect suggestive of diabetes insipidus (DI). The DI could be due to insufficient antidiuretic hormone (ADH) secretion by the hypothalamic-pituitary axis (central) or insufficient ADH response by the cortical collecting duct in the distal nephron (nephrogenic). The reappearance of hypernatremia following a water deprivation test after euvolemia is the expected response for a child who has DI. The lack of response to pharmacologic doses of desmopressin confirms that the infant has nephrogenic diabetes insipidus (NDI). This disorder is seen primarily in males and is due to a defect in the gene responsible for the vasopressin V2 receptor, which is located on the X chromosome. This genetic defect accounts for approximately 95% of cases of NDI. The remaining 5% of cases result from a defect in the human aquaporin-2 gene (*AQP2*) responsible for the vasopressin-regulated aquaporin-2 water channel that has been localized to chromosome 12q13. Unlike the X-linked V2 receptor gene, the *AQP2* gene locus is autosomally inherited and, therefore, results in the autosomal recessive NDI that can be seen in females.

One method of reducing the polyuria and polydipsia that occurs in DI is to decrease the renal solute load. The renal solute load is affected substantially by the dietary intake of minerals (sodium, potassium, calcium, magnesium, chloride, phosphorus, and sulfate) and protein. The protein load must be metabolized to urea, which ultimately contributes significantly to the renal solute load. A patient who consumes 300 mOsm/day needs to excrete this osmolar load in the requisite urine volume. If the patient can concentrate urine to 1,000 mOsm/L, only 300 mL (300 mOsm divided by 1,000 mOsm/L) of urine is necessary to excrete the solute load. On the other hand, a patient who has NDI and can only concentrate urine to 50 mOsm/L must generate 6 L of urine to excrete this same solute load (300 mOsm divided by 50 mOsm/L). Hence, a low-solute formula can minimize the solute load and reduce the burden of polydipsia and polyuria.

It should be noted that human milk has a lower renal solute load than cow milk-based formulas, due to the lower protein content of human milk. Neither carbohydrate nor fat contribute significantly to the renal solute load.

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Question: 15

A 10-year-old African-American boy presents to your office complaining of a 12-month history of stomach pain, nausea, bloating, and diarrhea that occurs 45 to 60 minutes after eating dairy foods. He states that his symptoms occur only when he eats “too much.” He denies emesis, hematochezia, or pruritus associated with these episodes. On physical examination, the boy appears healthy and has normal vital signs. His abdomen is soft and has normal bowel sounds, and results of a stool guaiac test are negative.

Of the following, the MOST likely cause for this boy’s symptoms is

A. allergic eosinophilic gastroenteritis

B. lactose intolerance

C. milk protein allergy

D. milk protein enterocolitis

E. oral allergy syndrome

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Critique: 15 Preferred Response: B

Adverse food reactions occur in 20% to 40% of the pediatric population. A detailed history that includes the timing of symptoms after food ingestion and reaction patterns can help determine the causative food and the involved mechanism. The onset of only gastrointestinal symptoms, the tolerance of smaller milk servings, and symptoms not occurring immediately (ie, onset after 30 minutes), as described for the boy in the vignette, are consistent with lactose intolerance. Lactose intolerance is a relatively common condition that results from decreased lactase activity. In affected children, lactase activity often does not decline to clinically significant levels until after the age of 6 years.

In contrast, milk protein allergy is an immunoglobulin (Ig) E-mediated reaction that almost always develops within the first postnatal year. Typical symptoms include urticaria, angioedema, atopic dermatitis, and anaphylaxis. With IgE-mediated reactions, the quantity of milk required to result in a reaction can be minute, such as milk touching the face or a taste of ice cream.

Milk protein enterocolitis, also called gastrointestinal protein allergy, is a non-IgE-mediated reaction that presents as hematochezia within the first few postnatal months. Unfortunately, most infants who have milk protein enterocolitis also experience symptoms with soy-based formulas and must be switched to an elemental or amino acid-based formula. Allergic eosinophilic gastroenteritis is an increasingly recognized condition that may be IgE- or non-IgE-mediated. Common symptoms include reflux, heartburn, and dysphagia after food ingestion, but weight loss or failure to thrive also may occur. The diagnosis may be difficult to make because multiple biopsies often are required to find the hallmark pathologic presentation of 15 or more eosinophils per high-power field. Although this condition would be included in the differential diagnosis for the boy in the vignette, it is much less common than lactose intolerance, and the lack of weight loss, emesis, or dysphagia make eosinophilic gastroenteritis unlikely.

Oral allergy syndrome is a localized reaction that occurs in approximately 10% to 40% of individuals who have allergic rhinitis. Ingestion of certain foods, usually raw fruits or vegetables, results in immediate oral pruritus and swelling. The causative foods contain proteins that are similar to those found in cross-reacting pollen proteins (eg, cantaloupe and ragweed). Reactions usually are benign and self-limited, although severe reactions have been described.

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Question: 16

A 2-year-old boy is brought to the emergency department after his father found the boy in the garage gagging, coughing, and drooling profusely. His parents report that he had opened a number of containers, but they are unable to recall the names of the products. On physical examination, the boy is awake and crying, his heart rate is 160 beats/min, his respiratory rate is 24 breaths/min, he has clear lung sounds, and his blood pressure is 100/60 mm Hg. He has reactive, mid-sized pupils and white eschars on his tongue and soft palate. Shortly after his examination, he begins to vomit.

Of the following, the MOST likely cause of this patient's symptoms is

A. antifreeze

B. drain cleaner

C. gasoline

D. glyphosophate weed killer

E. organophosphate insecticide

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Critique: 16 Preferred Response: B

Most unintentional ingestions in young children occur in the home, where toxic household products are readily available. Many cleaning products, hydrocarbons, garden chemicals, and automotive solutions pose significant toxicity risks if ingested. Often, as described in the vignette, a toddler is brought to medical attention with the suspicion of a toxic exposure to an unknown product. Recognition of the typical symptoms related to individual exposures is critical to providing the appropriate intervention.

Ingestion of a corrosive alkali, such as drain cleaner, causes injury to mucosal and skin surfaces by liquefaction necrosis. Affected patients commonly present with drooling, dysphagia, odynophagia, and intraoral burns, as reported for the boy in the vignette. Additionally, they may have vomiting with hematemesis; respiratory distress with stridor or wheezing; and burns on the face, hands, or chest. Because the primary mode of injury is direct tissue corrosion, systemic symptoms are rare, and cardiovascular instability is not typical. Apart from agitation due to pain, mental status usually is normal. The major clinical concerns with a caustic ingestion are airway or esophageal injury. Early airway visualization and protection are indicated in any patient who presents with stridor or respiratory distress, and evaluation of the esophagus by upper endoscopy is indicated in patients who have intraoral burns or other symptoms related to the ingestion. The need for endoscopy in the asymptomatic patient in whom caustic ingestion is questionable is controversial.

Ethylene glycol, the toxic agent in antifreeze, causes an initial clinical syndrome typical of all alcohols. Abnormal mental status is the most common presenting symptom, with alterations ranging from lethargy to coma. Tachypnea is common due to the significant metabolic acidosis that results from the production of glycolic and oxalic acids during metabolism of ethylene glycol by alcohol dehydrogenase. Relative bradycardia and hypotension related to sedative effects also may be seen. Crystalluria caused by precipitation of calcium oxalate crystals in the urine is a late finding (Item C16).

Gasoline and other hydrocarbon ingestions primarily cause respiratory distress due to pulmonary aspiration. Acutely, patients present with coughing, gagging, and choking. Among the potential findings on physical examination are fever; tachypnea; cyanosis; and abnormal lung findings that can include crackles, rales, and wheezing. In addition, direct central nervous system (CNS) effects of the hydrocarbon or related hypoxemia may lead to CNS depression, seizures, or coma.

Organophosphate toxicity is marked by clinical signs and symptoms related to the overstimulation of cholinergic receptors by excess acetylcholine. The features of cholinergic crisis include salivation, lacrimation, urinary incontinence, diarrhea and abdominal cramping, bronchorrhea and bronchospasm, and muscle weakness. Among the vital sign abnormalities are tachypnea and bradycardia, and pupils are constricted.

Glyphosphate-containing weed killers are irritants that can cause eye redness, cough with inhalation, and vomiting after ingestion.

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Available at:
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Critique: 16



Calcium oxalate crystalluria: Microscopy of uncentrifuged urine under polarized light reveals numerous clumps of calcium oxalate crystals.

Courtesy of K. Waibel

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Question: 17

You are evaluating a 4-year-old girl who has a 2-day history of perineal pruritus and dysuria. There is no history of trauma or sexual abuse. Physical examination reveals Sexual Maturity Rating 1 genitalia, with erythema of the labia majora, labia minora, and vaginal introitus. The hymenal tissue appears normal, and there is no vaginal discharge.

Of the following, the MOST appropriate management is

A. administration of fluconazole orally

B. administration of mebendazole orally

C. examination under anesthesia for vaginal foreign body

D. report to child protective services for sexual abuse

E. sitz baths followed by the application of an emollient

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Critique: 17 Preferred Response: E

The perineal pruritus, dysuria, and vulvar erythema described for the girl in the vignette are consistent with a diagnosis of vulvitis. Vulvitis may occur alone or in combination with vaginitis (ie, vulvovaginitis); the latter condition is characterized by vaginal discharge and malodor. Vulvovaginitis may be nonspecific (possibly due to fecal aerobes or abnormal numbers of anaerobes) or due to an identified cause (eg, *Candida*, pinworms, foreign body). In children, factors that predispose to the development of vulvovaginitis include:

- Absence of labial fat pads and pubic hair (that act as physical barriers)
- Proximity of the anus to the vagina and poor toileting hygiene (that increase the likelihood of bacterial contamination of the vulva and vagina)
- Thin vulvar skin that is prone to inflammation caused by chemicals (eg, fragrances in soaps or bubble baths), the effects of occlusive clothing (eg, leotards or tights), or trauma resulting from scratching
- Conditions favorable to bacterial growth in the vagina (eg, neutral pH, atrophic vaginal mucosa due to a physiologically low estrogen concentration)

In adolescents, some of these factors may continue to play a role. However, the use of douches or tampons also may contribute, and specific infections assume a greater etiologic importance.

The treatment of nonspecific vulvitis or vulvovaginitis includes sitz baths followed by the application of a bland emollient. Proper toileting hygiene (wiping front-to-back), careful hand washing, avoidance of tight-fitting underwear or clothing, wearing of cotton underwear (rather than synthetic fabrics), and avoidance of chemical irritants also may be important.

For patients who have vulvovaginitis due to *Candida* sp, fluconazole may be prescribed. However, vaginal infection due to *Candida* sp is uncommon in prepubertal girls unless they recently have completed a course of antibiotics, have diabetes mellitus, are immunocompromised, or are in diapers. In addition, infection is characterized by a thick, adherent vaginal discharge and often a typical cutaneous rash.

Pinworm infestation usually causes perianal itching that is worse at night, but can produce an associated vaginitis with resultant vaginal discharge. If present, this infestation is treated with mebendazole.

The child described in the vignette does not have symptoms of a vaginal foreign body (ie, purulent, blood-tinged vaginal discharge or malodor) and, therefore, examination under anesthesia is not indicated. Finally, because the findings on history and physical examination are not concerning for child sexual abuse, filing a report with child protective services is unwarranted without supporting information.

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Question: 18

You are examining a 12-week-old infant who was born at 28 weeks' gestation, developed necrotizing enterocolitis 7 days after birth, and underwent a bowel resection of 20 cm of ileum. The baby has been dependent on parenteral nutrition since shortly after birth and has cholestasis due to the parenteral nutrition. Attempts to feed her with a cow milk-based formula have resulted in diarrhea. Stool analysis demonstrates 0.25% (trace) reducing substances and 3+ fecal fat.

Of the following, the intervention that is MOST likely to decrease the infant's diarrhea is

A. addition of a vitamin supplement to the formula

B. addition of ranitidine to the formula

C. increase in the protein content of the formula

D. use of a formula that has a high medium-chain triglyceride content

E. use of a formula that has a lower concentration of carbohydrates

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Critique: 18 Preferred Response: D

The infant described in the vignette has evidence of fat malabsorption, manifested by diarrhea and abnormally high fecal fat content. The absence of significant amounts of reducing substances in the stool (<0.5%) suggests that the infant's carbohydrate digestion is adequate. The infant also has had an ileal resection. The terminal ileum is essential for reabsorbing bile acids that are secreted into the proximal intestine by the liver (enterohepatic circulation). The loss of endogenous bile acids in this infant increases the likelihood of steatorrhea. Substitution of medium-chain triglycerides for long-chain fats in her formula should help improve the diarrhea because medium-chain fats are absorbed by a lipase- and bile acid-independent pathway. Vitamin supplementation, the addition of ranitidine, increased protein content, and reduced carbohydrate content of the formula will not address steatorrhea.

Ingested triglycerides in humans are digested initially by lingual, gastric, and pancreatic lipase to free fatty acids and 2-monoglycerides. The process of lipid digestion depends, in part, on the formation of lipid micelles in the duodenum. The formation of micelles, in turn, depends on bile acids. Once fats have been broken down, the free fatty acids are transported across the enterocyte by a collection of transport proteins, resynthesized into triglycerides, and transported as chylomicrons to the liver. The preterm infant has a reduced ability to digest fats because of a number of factors, including reduced pancreatic lipase activity and decreased enterohepatic circulation of bile acids. The lipase activity in preterm infants has been estimated to be one third that of school-age children and may limit the infant's ability to digest fat. In addition to its other benefits, human milk contains bile salt-stimulated lipase, which is activated in the infant's duodenum and aids in lipid digestion.

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Question: 19

A term infant is delivered by emergency cesarean section following the acute onset of maternal vaginal bleeding and profound fetal bradycardia. The Apgar scores are 1, 2, and 3 at 1, 5, and 10 minutes, respectively. Resuscitation includes intubation and assisted ventilation, chest compressions, and intravenous epinephrine. The infant is admitted to the neonatal intensive care unit and has seizures 6 hours after birth.

Of the following, a TRUE statement about other organ-system injury that may occur in the infant is that

A. cardiovascular injury is uncommon

B. hypoxic-ischemic encephalopathy usually is an isolated condition

C. liver injury may result in a coagulopathy

D. most infants who have seizures develop cerebral palsy

E. necrotizing enterocolitis does not occur in term infants

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Critique: 19 Preferred Response: C

The infant described in the vignette required advanced cardiopulmonary resuscitation following a period of fetal bradycardia. In the face of acute maternal vaginal bleeding, perhaps due to placental abruption, placenta previa, or vasa previa, normal placental function was interrupted and fetal hemodynamics altered. The fetal response to asphyxiation characteristically includes a redistribution of cardiac output toward vital organs (eg, brain, heart, adrenal glands) and away from nonvital organ beds (eg, gut, kidneys, skin, bone marrow). As a result, the clinical manifestations of intrapartum asphyxia include perturbations of multiple organ systems that result from both ischemia and hypoxia.

Intrapartum asphyxia can affect various systems:

- Cardiovascular: systemic hypotension, pulmonary hypertension, dilated cardiomyopathy, or myocardial ischemia
- Pulmonary: respiratory distress, surfactant depletion/disruption with capillary-alveolar leak, hypoxic respiratory failure with pulmonary hypertension, or apnea
- Renal: oliguria, acute tubular necrosis, or renal failure
- Gastrointestinal: impaired gastric motility, gastrointestinal hemorrhage, necrotizing enterocolitis (NEC), ischemic hepatitis, or hepatopathy
- Hematopoietic: anemia, thrombocytopenia, coagulopathy
- Metabolic: acidemia, hypoglycemia, hypocalcemia, hypomagnesemia
- Central nervous system: hypoxic-ischemic encephalopathy (HIE), apnea, irritability, jitteriness, abnormalities in neuromuscular tone, seizure, or coma

Following intrapartum asphyxia, it is very rare for HIE to occur as an isolated condition, without evidence of any of the previously noted organ system injuries.

Although seizures may be common in the face of hypocalcemia, hypoglycemia, hypoxia, or severe acidosis (pH <7.0), most infants who have a seizure do not develop cerebral palsy. The cardiovascular abnormalities noted previously are among the most common of organ system injuries in this setting. Intrapartum asphyxia is one of the few settings in which NEC occurs in term infants. The hepatic synthesis of coagulation proteins that may result from hypoxic-ischemic liver injury contribute to coagulopathy.

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Question: 20

A medical student rotating in your clinic tells you about a 5-month-old infant he has evaluated. He reports that the infant is fed goat milk exclusively and asks you if this is adequate nutrition at this age.

Of the following, the MOST likely deficiency in this infant is of

A. folate

B. iron

C. niacin

D. vitamin A

E. vitamin D

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Critique: 20 Preferred Response: A

Goat milk is used exclusively for infant nutrition in some countries and has been used occasionally in the United States for infants who have cow milk allergies, but its routine use in the United States is not recommended. Although its fat may be digested more easily than fat found in cow milk preparations, it is deficient in several important nutrients, such as iron, vitamin D, and especially folate. Folate is a cofactor required in nucleoprotein synthesis, and deficiency ultimately results in ineffective erythropoiesis and megaloblastic anemia. Macrocytosis and hypersegmented neutrophils are typical findings on complete blood count, and if the anemia is severe, pancytopenia also can occur. Mothers who feed their infants goat milk exclusively should be counseled to switch to cow milk formula to avoid these complications. Niacin and vitamin A are sufficiently present in goat milk to avoid deficiency of these nutrients.

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Question: 21

You are seeing a 4-year-old boy for a health supervision visit prior to enrollment in preschool. His mother expresses some concerns about his speech articulation, although he uses many words. Results of his physical examination, including pneumatic otoscopy, are normal, as are results of tympanometry. Routine screening audiometry in your office, despite a cooperative child and a quiet examination room, yields equivocal results.

Of the following, the BEST next step in the evaluation of this child's hearing is

A. assessment for hearing aid placement

B. reassurance that the child will "grow out of it"

C. referral to an audiologist

D. referral to an otolaryngologist

E. sedated auditory brainstem evoked potentials

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Critique: 21 Preferred Response: C

Screening audiometry is performed commonly in schools and physician's offices. Using earphones, a noise stimulus is delivered to the ear in a quiet environment. It requires that the child be able to follow instructions and respond appropriately by indicating with a raised hand in which ear he or she hears the sound. Screening audiometry is designed to measure normal hearing in the range of human speech frequencies and intensity. Its use is recommended in children beginning at age 3 to 5 years to detect hearing loss that may be acquired after the newborn hearing screen and that may affect language acquisition and educational progress. Hearing screening failure at any age should prompt immediate referral; a “wait and see approach” never is appropriate.

Children who fail or have equivocal results of audiometric testing or who lack the physical, cognitive, or behavioral requisites for cooperating with audiometry should be referred to an audiologist for more sophisticated testing. Types of audiologic testing procedures might include visually reinforced audiometry (for those whose developmental age is 6 months to 3 years), conditioned play audiometry (for children who have a developmental age of 3 to 5 years), and behavioral observation audiometry (for infants younger than 6 months). Auditory brainstem response testing may be indicated if results of the previously mentioned studies are abnormal. Referral to an otolaryngologist or for hearing aid evaluation would be considered only after documentation of hearing loss by audiologic evaluation.

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Question: 22

Yesterday, you received a call from the newborn nursery that they were referring to you a term infant who was being discharged at 4 days of age. The female newborn's birthweight was 3.3 kg and the delivery was by repeat cesarean section. Findings on physical examination at discharge, including heart rate, respiratory rate, and blood pressure, were normal. Her lungs were clear, and no murmurs were noted. She was breastfeeding without difficulty. Today, her mother calls to tell you that she is difficult to awaken, pale, and breathing much more rapidly than she was in the hospital nursery. She has had one wet diaper in the last 12 hours. When you meet them in the emergency department, you note that the infant has cool extremities, weak pulses, and lethargy.

Of the following, the MOST likely cause of this newborn's condition is

A. aortic coarctation

B. atrioventricular septal (canal) defect

C. tetralogy of Fallot

D. transposition of the great arteries

E. ventricular septal defect

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Critique: 22 Preferred Response: A

The newborn described in the vignette has had an acute change in behavior, with diminished feeding as well as clinical signs of diminished systemic perfusion and shock. Despite the normal birth history and findings at hospital discharge, congenital heart disease may be a cause of the shock. In fact, the presentation is most consistent with a left heart obstructive disorder, such as aortic stenosis, coarctation of the aorta, or hypoplastic left heart syndrome. In such patients, tachypnea is caused by pulmonary congestion that results from decreased filling of the failing left heart. A gallop rhythm (Item C22A) may be audible on auscultation, due to filling of the noncompliant left ventricle. The tachypnea exacerbates the poor feeding resulting from the infant's inability to generate a prolonged suck while maintaining nasal breathing. When coupled with increased losses of water through the respiratory system, such poor intake leads to dehydration and decreased urine output. The lethargy exhibited by the infant in the vignette may reflect decreased perfusion to the brain and may be exacerbated by the metabolic acidosis that results from inadequate tissue perfusion. Immediate management of this critically ill patient includes the administration of prostaglandin E to re-establish patency of the ductus arteriosus. Definitive therapy involves surgery to repair the coarctation (Item C22B).

Atrioventricular septal (canal) defects and ventricular septal defects are both left-to-right shunting lesions that become apparent after birth when the pulmonary vascular resistance normally begins to decrease. These shunting lesions are characterized by increased blood flow into the pulmonary artery because the direction of shunt is always from the high-resistance circuit to the low-resistance circuit (normally the systemic to pulmonary system) (Item C22C). If the shunt volume is large, signs and symptoms of congestive heart failure ensue, but these almost always manifest over the first weeks, if not months, after birth.

Tetralogy of Fallot is the most common of the cyanotic congenital heart diseases and typically presents with the murmur of pulmonary blood flow obstruction. Because the degree of obstruction is progressive, patients may present with normal oxygen saturation or cyanosis resulting from desaturated blood moving right to left at the ventricular septal defect (Item C22D). Transposition of the great arteries is a cyanotic heart disease that presents within hours of birth in most cases (Item C22E). Neither tetralogy of Fallot nor transposition presents with shock.

References:

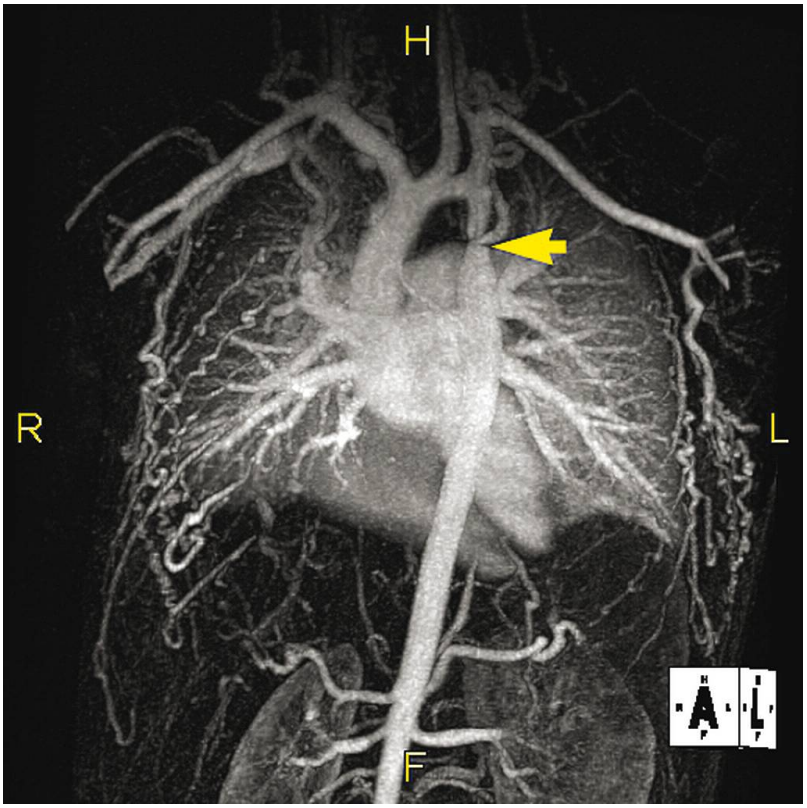
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Critique: 22

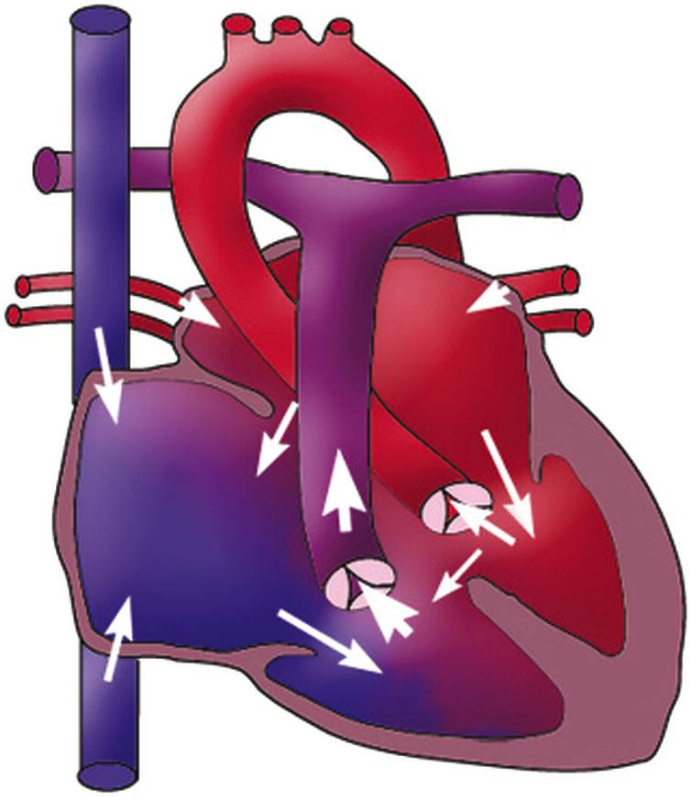


Anterolateral view from a three-dimensional magnetic resonance angiography reconstruction in a 6-year-old who has coarctation of the aorta (arrow) and numerous collateral vessels.

Courtesy of A. Cook

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Critique: 22

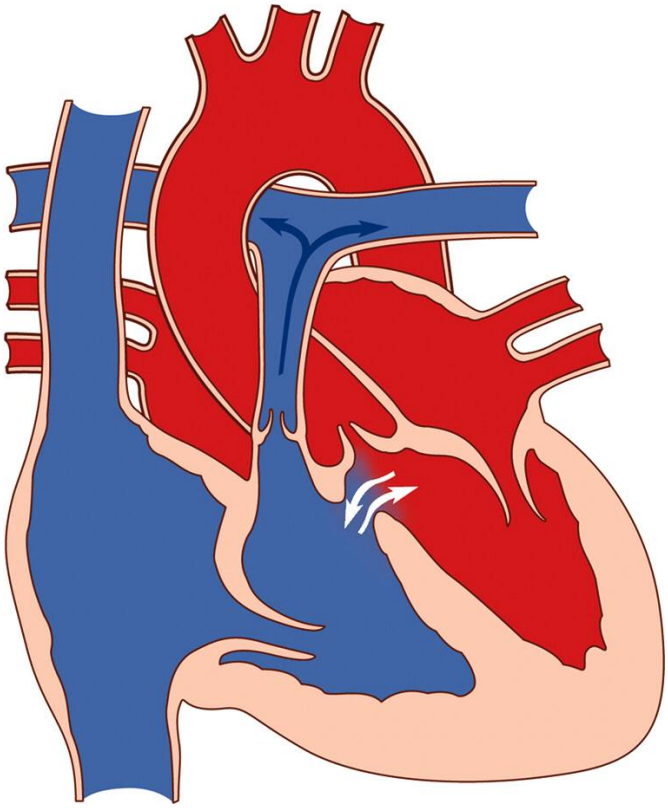


An atrioventricular canal defect results in shunting of blood from the systemic to pulmonary system. If the shunt volume is sufficient, congestive heart failure results.

Courtesy of the Media Lab at Doernbecher

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Critique: 22

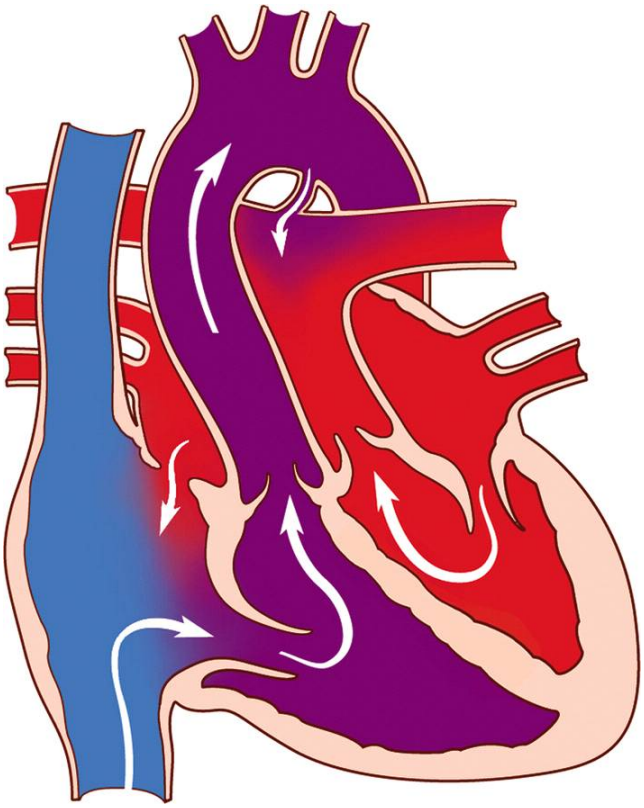


In tetralogy of Fallot, cyanosis is caused by right-to-left shunting at the ventricular septal defect as pulmonary stenosis worsens.

Courtesy of A. Johnson

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Critique: 22



In transposition of the great arteries, the left ventricle connects to the pulmonary artery and the right ventricle to the aorta.

Courtesy of A. Johnson

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Question: 23

A 14-year-old girl who has a 1-year history of migraine headaches presents to the emergency department with a severe headache that she calls “the worst headache of my life.” The headache occurred suddenly after she lifted a heavy box. Her mother says that the girl has been holding her head stiffly. On physical examination, she appears in severe pain and has meningismus. Other findings on the physical examination are normal.

Of the following, the MOST appropriate initial course of action is

A. emergent noncontrast head computed tomography scan

B. intravenous administration of ceftriaxone

C. intravenous administration of dihydroergotamine

D. lumbar puncture

E. oral administration of sumatriptan

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Critique: 23 Preferred Response: A

Although severe headaches in adolescents are often due to migraine, four key findings from the history provided by the girl described in the vignette indicate that she is experiencing a "symptomatic" headache (ie, headache due to an underlying disease) instead of a migraine headache. These findings are: 1) her description of this headache as “the worst ever,” 2) its abrupt onset, 3) the onset during a “Valsalva maneuver” (heavy lifting), and 4) the stiff head/neck position. These features are characteristic of a subarachnoid hemorrhage (SAH), likely a small hemorrhage at this point, because the girl is still conscious. In contrast, migraine headaches, which are common in children and adolescents, are recurrent headaches often associated with nausea, dizziness, and photo- or phonophobia. They represent an “idiopathic” chronic recurrent headache disorder, meaning that no space-occupying lesions (tumors, vascular malformations) are causing the pain. In “symptomatic” headaches, the pain is a symptom of an underlying disease process.

Head computed tomography (CT) scan is the preferred diagnostic modality for the girl in the vignette because it is fast, noninvasive, and highly sensitive to blood and structural lesions causing pain. Of note, CT findings are normal in patients whose headaches are caused by pseudotumor cerebri.

Although this girl has nuchal rigidity, which occurs in meningitis, she is afebrile, and her symptoms began abruptly while lifting, which should not be related to central nervous system infection. Accordingly, ceftriaxone treatment is not appropriate at this point. Intravenous administration of dihydroergotamine might be appropriate treatment for a severe migraine headache, not for the symptomatic headache described for the girl in the vignette. For the same reason, oral sumatriptan is not the appropriate initial therapy. Although a lumbar puncture eventually may be necessary, it should not be the initial diagnostic test because it is invasive and may be unsafe in the presence of a focal lesion. Because the history suggests SAH and the sensitivity of head CT in diagnosing this condition is at least 95%, CT should be the initial diagnostic test. If the suspicion for SAH is high and results of the CT are normal, lumbar puncture should be obtained to look for red blood cells.

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Question: 24

You care for a newborn who has Down syndrome due to an unbalanced 14;21 translocation. At the request of the cytogenetics laboratory director, you arranged for the parents' blood to be collected and karyotyped. You now are notified that the baby's mother has an unusual karyotype with a balanced 14;21 translocation. You plan to refer her for genetic counseling.

Of the following, the recurrence risk for having a baby who has Down syndrome in a future pregnancy for this woman is CLOSEST to

A. 1% added to her age-related risk

B. 15%

C. 30%

D. 50%

E. 100%

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Critique: 24 Preferred Response: B

Approximately 95% of individuals who have Down syndrome (DS) have an independent, extra copy of chromosome 21 due to nondisjunction (uneven chromosomal distribution caused by unequal cell division during gamete production or after conception). Another 4% have an unbalanced translocation between two chromosomes, resulting in an extra chromosome 21. The remaining 1% have a mosaic pattern, with the extra chromosome 21 present in some, but not all, cells.

When DS is due to nondisjunction, the recurrence risk to the parents of the affected child is an empiric 1% added to the maternal age-related risk for having a child with DS; this risk typically ranges from 1% to 4%, depending on the mother's age.

Almost all translocation DS involves a Robertsonian translocation. Robertsonian translocations affect the acrocentric chromosomes (the centromere is close to the tip of the chromosome, and there is no structural DNA beyond that point): numbers 13, 14, 15, 21, and 22. Chromosome analysis should be performed in children in whom DS is suspected to ensure the correct diagnosis and to determine if there is a chromosome rearrangement that could place the parents at increased risk for having a future affected child. When a child has translocation DS, blood should be requested from parents for chromosome analysis.

Approximately 25% of Robertsonian translocation DS cases are familial; 75% of such cases are de novo (happen for the first time in the affected individual). Empiric data show that when the translocation is de novo, the recurrence risk is less than 1%. If the father is the translocation carrier, the recurrence risk is about 1%. If the mother is the translocation carrier, as in the vignette, the likelihood of finding translocation DS at amniocentesis is 15%, and the risk for having a liveborn child who has translocation DS is about 10%.

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Question: 25

A 15-year-old girl comes to your office because she never has had a menstrual period. She has no chronic illnesses and is active playing softball once a week. Her mother and sister both had menarche at age 13 years. On physical examination, she is at the 15th percentile for height and weight and has no hirsutism or acne, no breast development, and Sexual Maturity Rating 3 pubic hair development.

Of the following, the MOST appropriate initial laboratory evaluations are

A. antiovarian antibody and antithyroid antibody concentrations

B. follicle-stimulating hormone concentration and karyotype

C. progesterone and 17-hydroxyprogesterone concentrations

D. testosterone and androstenedione concentrations

E. thyroid-stimulating hormone and thyroxine concentrations

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Critique: 25 Preferred Response: B

Among Americans, 95% to 97% of females reach menarche by 16 years of age and 98% by 18 years of age. About two thirds of young women reach menarche at Sexual Maturity Rating (SMR) 4, 5% attain menarche at SMR 2, 25% at SMR 3, and 10% at SMR 5. Primary amenorrhea is defined as having no menstrual period by the age of 16 years. In evaluating primary amenorrhea, the clinician needs to determine if there is a hypothalamic-pituitary-ovarian axis abnormality or a genital anomaly (eg, an imperforate hymen or agenesis of the vagina, cervix, or uterus).

The patient described in the vignette has primary amenorrhea and an absence of breast development, but she has pubic hair. These findings are consistent with a diagnosis of Turner syndrome, a condition characterized by short stature and ovarian dysgenesis in females who have a single X chromosome or absence of all or part of a second sex chromosome (X or Y). Turner syndrome is estimated to occur in 1 in 2,000 to 1 in 5,000 live female births. A young adolescent who has Turner syndrome has prepubertal female genitalia, streak gonads, and a normal uterus and vagina (Item C25). An older teen (15 to 16 years old) who has undiagnosed or untreated Turner syndrome usually has pubic and axillary hair but lacks breast development and estrogenization of the vaginal mucosa. Patients who have gonadal dysgenesis with a mosaic karyotype may show none or all of the classic physical characteristics of Turner syndrome.

Approximately 40% to 50% of those who have Turner syndrome have a mosaic karyotype (46,XX/45,X) or a structural abnormality of the second X chromosome consisting of a deletion of part of the short arm (p-) or long arm (q-) of the X chromosome, a ring chromosome, or an isochromosome. Normal ovarian function is based on having critical regions present on both the long and short arm of the X chromosome, and several deletions are associated with primary amenorrhea. Other deletions are associated with premature ovarian failure and secondary amenorrhea.

Because the patient's symptoms and physical findings suggest Turner syndrome, the most appropriate initial laboratory evaluations are a karyotype and measurement of follicle stimulating hormone (FSH) and luteinizing hormone. Although some girls who have mosaic karyotypes may not have increased FSH concentrations, most of those affected have elevated FSH values at birth, suppression of the hormone in early childhood, and increases to menopausal levels by 10 to 11 years in those who have gonadal failure. The young woman in the vignette likely has a mosaic karyotype, due to her low-normal height and lack of other Turner syndrome stigmata. The diagnosis of Turner syndrome should be excluded in any adolescent girl who has primary or secondary amenorrhea, especially if she is short.

Measurement of antiovarian and antithyroid antibodies can aid in the diagnosis of autoimmune ovarian failure. Although this disorder can be the cause of primary amenorrhea, it is much less common than Turner syndrome. In addition, signs of estrogenization, such as breast development, usually occur before complete ovarian failure. Measuring progesterone concentrations would not assist in diagnosing primary amenorrhea. Elevated 17-hydroxyprogesterone concentrations are associated with congenital adrenal hyperplasia (CAH). Although CAH may be a cause of primary amenorrhea, girls who have this condition usually exhibit signs of virilization (eg, clitoromegaly). Testosterone and androstenedione are produced by both ovaries and adrenals. Increased concentrations of these hormones may be seen in girls who have polycystic ovary syndrome or sex steroid-producing ovarian or adrenal tumors. Measurement of thyroid-stimulating hormone and thyroxine aid in the diagnosis of thyroid dysfunction, which might be associated with primary or secondary amenorrhea.

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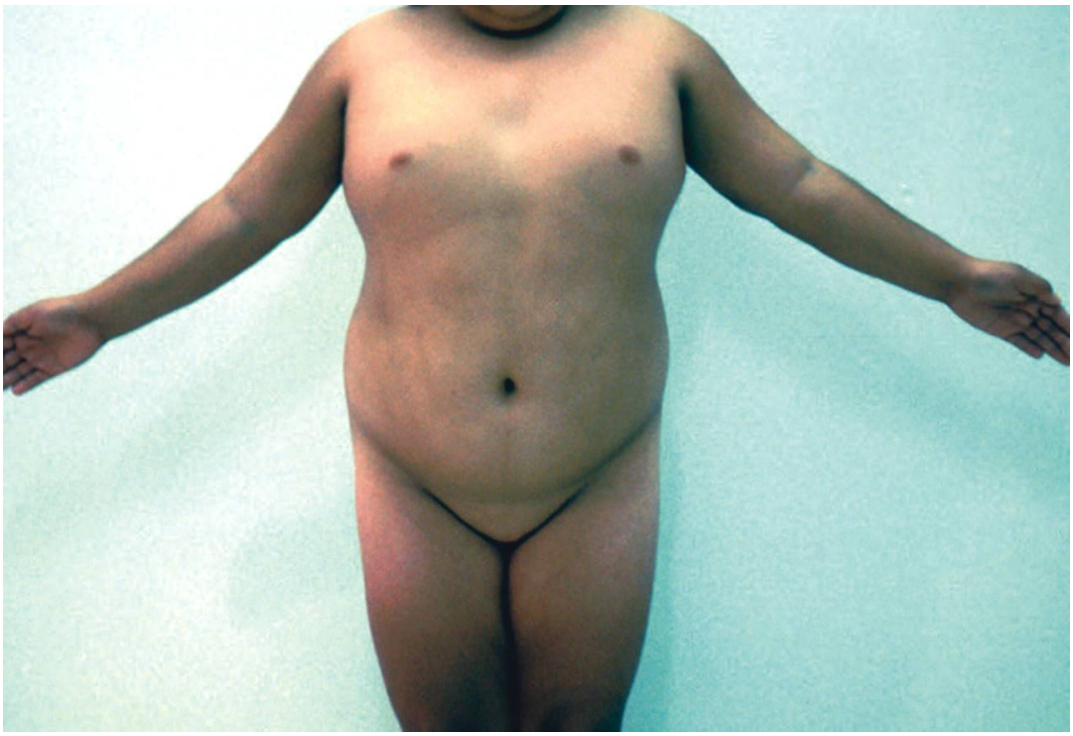
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Critique: 25



A girl who has Turner syndrome exhibits wide-spaced nipples, broad chest, and lack of secondary sexual development.

Courtesy of M. Rimsza

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Question: 26

You are seeing a 10-year-old girl for her yearly health supervision visit. On physical examination, you palpate a smooth and symmetric thyroid that seems twice normal size (Item Q26). There are no palpable nodules. Serum free thyroxine and thyroid-stimulating hormone (TSH) values are both normal. Serum thyroperoxidase antibody concentrations are elevated.

Of the following, the initial BEST approach to management is to

A. obtain a 123-I thyroid scan

B. obtain thyroid ultrasonography

C. recheck TSH concentration in 6 months

D. start treatment with triiodothyronine

E. start treatment with TSH

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Critique: 26 Preferred Response: C

Hashimoto thyroiditis or chronic lymphocytic thyroiditis is a common autoimmune disorder of the thyroid, affecting more than 1 in 600 children. It is more common in girls. The diagnostic criterion is the presence of antithyroid antibodies directed against the thyroid peroxidase (TPO) enzyme or against thyroglobulin. Pathologic evaluation of the thyroid would reveal the presence of lymphocytic infiltrates and lymphoid follicles within the thyroid gland. The spectrum of the disorder ranges from asymptomatic thyroid enlargement associated with lymphoid infiltration and, in most cases, positive serum antibodies to frank hypothyroidism with an enlarged or atrophic gland or, occasionally, transient hyperthyroidism. A child who is euthyroid but has positive antithyroid antibodies, such as the girl described in the vignette, should undergo thyroid function studies, including measurement of thyroid-stimulating hormone (TSH) and free thyroxine (fT4), at 6-month intervals or if symptoms of hypo- or hyperthyroidism are recognized. Most affected children eventually develop hypothyroidism, but this process may take many years and may not occur until adulthood.

TSH is available as a biosynthetic preparation and can be used as preparative therapy before radioactive iodine ablation or for evaluation for metastasis in individuals who have thyroid cancer, but it is not used to treat thyroiditis. Triiodothyronine (T3), the active form of thyroid hormone, has a relatively short half-life, and is produced as needed from T4 by most peripheral tissues. Therefore, treatment with T3 rarely is indicated. There is some evidence that treatment with T4 may reduce the size of the thyroid gland in a child who has chronic lymphocytic thyroiditis, even if the TSH value is normal, but this is still controversial. A thyroid scan using radioactive iodine may show a characteristic pattern of patchy uptake related to infiltration by lymphoid follicles in the child who has Hashimoto thyroiditis, but the study is not indicated if the patient is euthyroid and the gland is symmetric. Radioactive iodine scans should be reserved for the evaluation of thyrotoxicosis and, in rare circumstances, thyroid nodules. Thyroid ultrasonography would confirm the enlargement of the thyroid gland. There is a slightly higher risk of thyroid malignancy in patients who have thyroiditis, but if the gland is smooth and symmetric, there is no indication for this study.

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Question: 27

A 7-year-old girl is being treated with a gonadotropin-releasing hormone agonist by an endocrinologist for precocious puberty. On physical examination, she is at Sexual Maturity Rating 3. Her parents are pleased with the medical treatment, but are concerned that their daughter is having problems fitting in with the 7-year-old girls with whom she used to play, who are teasing her. She is now gravitating toward playing with older children in her neighborhood. The parents ask for guidance regarding their daughter's behavior.

Of the following, your BEST response to their concerns is to

A. explain that having older friends is beneficial to her self-esteem and should be encouraged

B. explain that their daughter should be treated more maturely because her body is maturing at a faster rate

C. recommend psychological counseling to help the girl deal with her feelings regarding the changes in her body

D. suggest that her parents call the girl's playmates and tell them to stop teasing her

E. suggest that the parents get a pet for their daughter so she will spend less time with the neighborhood children

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Critique: 27 Preferred Response: C

Precocious puberty is defined as the appearance of breast development prior to age 6 to 7 years in Caucasian girls and prior to 5 to 6 years in African-American or black girls, as is sexual development before 9 years in boys. The incidence of precocious puberty in the United States is estimated at 0.01% to 0.05% per year, with a four to ten times higher rate in girls than in boys.

Children who have precocious puberty tend to be shy and withdrawn with same-age peers and prefer to be with older individuals, as described for the girl in the vignette. Both parents and teachers should be told to expect age-appropriate behavior, even if the child appears older. Children who experience precocious puberty need to be monitored because they are at risk for sexual abuse and possible pregnancy.

In view of this child's difficulty with same-age peers, she should receive counseling to help her deal with her changing body image and guide her when interacting with peers. Her parents should treat her in an age-appropriate manner. They should encourage positive social interactions with her peers. A pet should not be obtained for the sole reason of trying to have her socialize less with the neighborhood children. Her parents should not directly contact her peers to avoid further embarrassment. They may wish to discuss the situation with school personnel, who may intervene in a less threatening manner. They should be wary of her gravitating toward older age peers, who may take advantage of her.

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Question: 28

A 6-year-old boy presents to the clinic with a 2-day history of fever and noisy breathing. His mother is concerned because she feels that her son is “going to die.” His past medical history is unremarkable, but he has not yet received his fifth diphtheria-tetanus-acellular pertussis (DTaP) or his second measles-mumps-rubella (MMR) vaccination. On physical examination, the boy appears scared and toxic and has labored respirations and a very harsh cough. He is not drooling and can lie flat while you examine him. His temperature is 103.5°F (39.7°C), respiratory rate is 35 breaths/min, heart rate is 168 beats/min, and blood pressure is 107/68 mm Hg. Although he has tachypnea, his lungs are clear to auscultation, he has no heart murmur, and findings on his abdominal examination are benign.

Of the following, the MOST likely diagnosis is

A. bacterial tracheitis

B. bronchitis

C. epiglottitis

D. foreign body aspiration

E. laryngotracheobronchitis

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Critique: 28 Preferred Response: A

The boy described in the vignette has bacterial tracheitis, a complication of a primary viral illness caused by a secondary bacterial infection of the upper airway (excluding the epiglottis). *Staphylococcus aureus*, *Moraxella catarrhalis*, nontypeable *Haemophilus influenzae*, and oral anaerobes all have been implicated as common causative agents. Mucosal swelling at the cricoid cartilage plus thick purulent secretions (Item C28) are responsible for the respiratory distress. The affected child typically has a high fever, appears toxic, and has a "brassy" cough. Treatment of bacterial tracheitis includes broad-spectrum antimicrobial agents that have antistaphylococcal coverage and may require placement of an artificial airway until the airway swelling and copious purulent secretions improve.

The patient who has epiglottitis usually cannot lay flat, drools, and has dysphagia. In parts of the world where the *H influenzae* type b vaccine is used, bacterial tracheitis occurs more frequently than epiglottitis. Most patients who have laryngotracheobronchitis improve with the administration of racemic epinephrine, are not highly febrile or toxic-appearing, and are younger than 3 years of age. Although acute bronchitis commonly is preceded by a viral upper respiratory tract infection, it is characterized by frequent dry hacking cough in an otherwise nontoxic-appearing patient. Similarly, patients who have aspirated foreign bodies usually are not febrile and toxic-appearing.

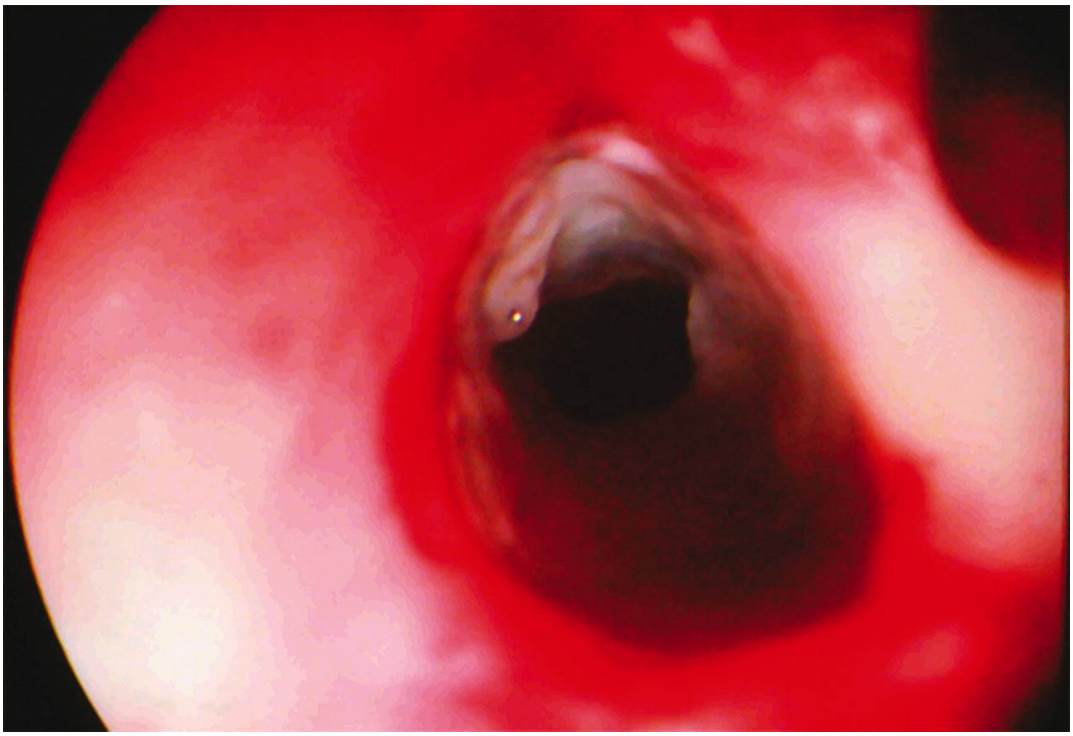
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Critique: 28



Bronchoscopic image in a patient who has bacterial tracheitis reveals thick secretions and pseudomembrane formation.

Courtesy of the Media Lab at Doernbecher

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Question: 29

You are speaking to the pediatric ward nurses regarding prevention of the nosocomial spread of rotavirus disease after three patients developed the disease while hospitalized. You stress the importance of good handwashing practices.

Of the following, the MOST appropriate additional statement regarding transmission-based precautions for rotavirus is that health-care personnel who have patient contact

A. do not need to take transmission-based precautions

B. need to put on gloves before entering patient rooms

C. need to put on gowns and gloves before entering patient rooms

D. need to put on gowns, gloves, and mask before entering patient rooms

E. need to put on gowns, gloves, mask, and shoe covers before entering patient rooms

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Critique: 29 Preferred Response: C

In the hospital setting, transmission-based precautions are designed for patients who are documented or suspected to be colonized or infected with pathogens spread by the airborne, droplet, or contact routes. Such patients require additional precautions beyond standard precautions (guidelines designed for the care of all patients regardless of their diagnosis or presumed infection status) to interrupt pathogen transmission or spread of disease. Airborne transmission occurs by dissemination of airborne droplet nuclei (small-particle residue of evaporated droplets containing microorganisms that remain suspended in the air for long periods), dust particles containing the infectious agent, or fungal spores. Microorganisms spread by the airborne route can be dispersed widely by air currents (eg, ventilation systems) and may be inhaled by or deposited on a susceptible host within the same room or a long distance from the source patient, depending on environmental factors. Therefore, special air handling and ventilation are required to prevent airborne transmission. Examples of microorganisms transmitted by airborne droplet nuclei are *Mycobacterium tuberculosis*, *Aspergillus* sp*,* measles virus, varicella virus, and disseminated zoster. Specific recommendations in airborne precautions include: the use of private rooms; negative air-pressure ventilation with externally exhausted or HEPA-filtered air, if recirculated; and wearing of respiratory protective devices in cases of suspected or proven tuberculosis.

Droplet transmission occurs when droplets containing microorganisms generated from the infected persons, primarily during coughing, sneezing, and talking, are propelled a short distance and deposited on the host's conjunctivae, nasal mucosa, or mouth. Because these relatively large droplets do not remain suspended in the air, special air handling and ventilation are not required to prevent transmission. Examples of microorganisms transmitted by droplets include: adenovirus, *Haemophilus influenzae* type b, influenza viruses, paramyxovirus (mumps), *Mycoplasma pneumoniae*, *Bordetella pertussis*, and group A beta-hemolytic streptococci (producing pharyngitis or scarlet fever, or pneumonia). Specific recommendations for droplet precautions include: providing the patient with a single-patient room (if available) or cohorting patients infected with the same organism with more than 3 feet between patients and the use of precautions between patients. Additionally, anyone entering the room or cubical space must wear a mask.

Contact transmission is the most frequent route of transmission of nosocomial infections and can occur by direct or indirect contact with the infected or colonized surface. Examples of microorganisms that are transmitted by contact include: *Clostridium difficile*, enteroviruses, *Escherichia coli* O157:H7 and other Shiga toxin-producing *E coli*, hepatitis A virus, herpes simplex virus, respiratory syncytial virus, rotavirus, scabies, and *S aureus*. Specific recommendations for contact precautions include: providing the patient with a single-patient room (if available) or cohorting patients infected with the same organism with more than 3 feet between patients and the use of precautions between patients. Personnel must wear gloves at all times and gowns for direct contact with a patient, environmental surfaces, or items in the patient room. For the rotavirus being discussed in the vignette, only contact precautions are needed, which entails the use of gowns and gloves before entering patient rooms.

Children living in institutional settings face special problems for the control of certain infections. The vaccine-preventable diseases that may spread rapidly in the institutional setting include measles, mumps, influenza, pertussis, hepatitis A and B, pneumococcal infections, and varicella. Other pathogens for which no immunizations are available that may spread in institutions are *Shigella* sp, *E coli* O157:H7, other enteric pathogens, *Streptococcus pyogenes*, *Staphylococcus aureus* (including community-acquired methicillin-resistant strains), respiratory tract viruses, cytomegalovirus, scabies, and lice. Ensuring that the residents of such institutions are appropriately immunized is important because of the risk of transmission within the facility and because the underlying conditions that led to institutionalization may increase the risk of complications from the diseases. All children entering a residential institution should have received recommended immunizations for their age; if not immunized appropriately, arrangements should be made to administer the immunizations as soon as possible.

Recommendations for treating children in institutional settings when exposed to a specific disease differ, depending on the disease (Item C29).

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Critique: 29

Item 29: Treatment of Infections in Institutionalized Children

Disease	Immunoglobulin (IG) or Other Chemoprophylaxis	Vaccine
Measles	Administer IG to immunocompromised children as soon as possible and within 6 days of exposure to all exposed susceptible children <1 year of age (will need measles-mumps-rubella [MMR] vaccine at 12 months of age or after, depending on age and dose of IG administered)	Administer MMR to all susceptible children ≥1 year of age within 72 hours of exposure if vaccine not contraindicated
Mumps	IG not effective; mumps-specific IG not available	Administer MMR to susceptible people to protect against future infection
Influenza	Appropriate use of antiviral chemoprophylaxis during influenza epidemics	Program of annual influenza immunization for residents and staff
Pertussis	Appropriate use of antibiotic treatment and chemoprophylaxis among infected people and their close contacts	Children who are not fully immunized should be vaccinated
Hepatitis A	IG for postexposure prophylaxis in susceptible residents and staff members in close personal contact with patients	Hepatitis A vaccine, in addition to IG, for postexposure prophylaxis for staff and children ≥1 year of age
Hepatitis B	After parenteral or sexual exposure, administer hepatitis B IG as postexposure prophylaxis to susceptible residents and staff	Immunize all susceptible residents and staff against hepatitis B virus at entry to facility; vaccinate unimmunized susceptible individual after parenteral or sexual exposure
Varicella	Recommended only for immunocompromised susceptible children at risk of serious complications or death from varicella	Administer to all healthy individuals ≥1 year of age who lack a reliable history of varicella disease or immunization

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Question: 30

You are asked to evaluate a 3,500-g term infant who was found to have unilateral renal agenesis on prenatal ultrasonography. Laboratory tests reveal a sodium concentration of 140 mEq/L (140 mmol/L), potassium of 4.1 mEq/L (4.1 mmol/L), chloride of 110 mEq/L (110 mmol/L), and bicarbonate of 19 mEq/L (19 mmol/L).

Of the following, the serum bicarbonate value can be explained BEST by

A. a normal value for age

B. an inborn error of metabolism

C. renal compensation for primary respiratory alkalosis

D. renal tubular acidosis

E. stool losses of bicarbonate resulting in metabolic acidosis

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Critique: 30 Preferred Response: A

Normal bicarbonate values vary with age in the pediatric patient. The greatest variation in values occurs in newborns and infants. Additional variation is seen between preterm and term infants, with normal values as low as 17 mEq/L (17 mmol/L) in preterm infants and 19 to 21 mEq/L (19 to 21 mmol/L) in term infants. The typical adult normal values of 24 to 26 mEq/L (24 to 26 mmol/L) occur in children older than 12 months of age. Thus, the value reported for the infant in the vignette is normal for age.

The age-related variation in bicarbonate values is believed to be caused by an altered renal threshold for bicarbonate reabsorption by the nephron. The proximal tubule accounts for approximately 90% of bicarbonate reabsorption; the remaining 10% occurs in the thick ascending limb of the loop of Henle and the outer medullary collecting tubule.

Inborn errors of metabolism, renal compensation for primary respiratory alkalosis, renal tubular acidosis (RTA), and bicarbonate stool losses resulting in metabolic acidosis depress serum bicarbonate concentrations. If concentrations are low, a blood gas (capillary or arterial) is essential to establish the presence of acidosis and discern whether it is metabolic or respiratory. Metabolic acidosis is present in inborn errors of metabolism and RTA and following stool bicarbonate losses, as seen in diarrhea. Inborn errors of metabolism classically are accompanied by an increased anion gap. Acidosis from RTA and diarrhea is associated with a normal anion gap.

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Question: 31

A 9-year-old boy who has a history of asthma and allergic rhinitis presents with coughing, wheezing, and chest tightness of 4 days' duration. He only has 1 week of school left before summer break, but his parents have kept him home because they are administering a beta-2 agonist inhaler every 4 hours to control his symptoms. They mention that "everyone was sick" in his class, and he developed clear rhinorrhea and a temperature of 99.0°F (37.3°C) at the start of his current illness.

Of the following, the MOST likely causative virus is

A. coronavirus

B. influenza virus

C. parainfluenza virus

D. respiratory syncytial virus

E. rhinovirus

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Critique: 31 Preferred Response: E

Wheezing and asthma exacerbations can be common during a respiratory infection in children who have asthma and atopy (eg, allergic rhinitis, atopic dermatitis). Although coronavirus, influenza virus, parainfluenza virus, respiratory syncytial virus, and rhinovirus all can cause both upper and lower airway inflammation and an asthma exacerbation, rhinovirus has been recognized as the most common viral cause of asthma exacerbations in older children. Previously believed to affect only the upper airway, rhinovirus has been shown to affect the lower airways directly and indirectly. Rhinovirus can be spread throughout the year, but the infection typically peaks in September and in late spring.

Human coronavirus is an RNA virus implicated in common colds in adults and children. Infections with this pathogen occur primarily from midwinter through early spring and can result in lower airway disease. Human coronavirus is less likely to be associated with asthma exacerbations than rhinovirus. Both influenza and parainfluenza viruses can cause upper or lower airway disease, but they are more prevalent in colder months. Also, parainfluenza typically affects most children by age 3 years; it is a less common trigger in older children.

Respiratory syncytial virus is the major pathogen associated with lower airway disease in infants and children younger than 2 years of age.

Regardless of age, most common respiratory viruses that reach the lower airways can cause transient airway hyperresponsiveness (AHR), which is characterized by airway narrowing and edema, mucus production, and altered ciliary function. Many individuals also experience a late-phase reaction 4 to 12 hours after the initial symptoms, which is characterized by AHR and an eosinophilic response. Other triggers that may result in similar early and late AHR in affected children include aeroallergens (eg, pollen, pet dander), smoke, and air pollutants (eg, sulfur, ozone).

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Question: 32

A 2-year-old boy is brought to the emergency department because of moderate respiratory distress. History reveals that he was born at 25 weeks' gestation and had bronchopulmonary dysplasia. He has had rhinorrhea and cough for the past 2 days, and this morning he developed retractions and wheezing that were unresponsive to albuterol. His usual medications include albuterol and oxygen at 1 L/min by nasal cannula to maintain his oxygen saturation at 93%. On physical examination, his heart rate is 160 beats/min, respiratory rate is 60 breaths/min, and oxygen saturation is 82%. You place him on 100% oxygen using a nonrebreather mask and obtain blood gases, which reveal a pH of 7.35, Pco2 of 70 mm Hg, Po2 of 226 mm Hg, and HCO3 of 35 mEq/L (35 mmol/L). As you are interpreting the blood gases, the nurse notifies you that the boy has developed apnea.

Of the following, the MOST likely explanation for his sudden deterioration is

A. acute pulmonary embolus with increased arterial-alveolar gradient

B. elimination of respiratory drive by correction of chronic hypoxemia

C. respiratory muscle fatigue due to acute metabolic acidosis

D. severe bronchospasm with acute respiratory acidosis

E. spontaneous pneumothorax with acute hypoxemia

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Critique: 32 Preferred Response: B

Chronic respiratory failure (CRF) is characterized by chronic hypoventilation that leads over time to chronic hypercarbia and hypoxemia. In the pediatric age group, CRF is seen most commonly in patients who have chronic lung disease, such as bronchopulmonary dysplasia and cystic fibrosis, or in the presence of neuromuscular disease, such as muscular dystrophy or anterior horn cell disease. Arterial blood gas findings in CRF include normal to near-normal pH, elevated Pco2, decreased Po2, and elevated serum bicarbonate concentrations, as reported for the boy in the vignette. Patients also may manifest polycythemia and pulmonary hypertension with right ventricular hypertrophy due to chronic hypoxemia.

Patients who have underlying CRF and hypoxemia may benefit from the administration of oxygen to maintain an oxygen saturation of 90% to 95% and limit the consequences of chronic hypoxemia or to treat acute respiratory decompensation. However, the respiratory drive of some patients who have CRF is dependent on hypoxemia (rather than on hypercarbia and acidosis, as would be the case in a patient who does not have chronic hypoventilation), and correction of hypoxia in such patients, such as the boy described in the vignette, could lead to respiratory arrest.

Acute pulmonary embolus can cause acute hypoxia and chest pain. In the pediatric population, pulmonary embolus is seen most commonly in patients who have hypercoagulopathies, indwelling vascular catheters, or critical illness. Depending on the extent of the ventilation-perfusion mismatch, oxygen administration may correct the hypoxemia, but would not be associated with respiratory arrest. Acute metabolic acidosis alone should not cause respiratory muscle fatigue or respiratory arrest, especially because acidosis is the primary determinant of respiratory drive. Arterial blood gas measurements in acute metabolic acidosis demonstrate a low pH and low serum bicarbonate concentration, with normal Pco2 and Po2 values. Patients who have severe acute respiratory distress may experience respiratory arrest if the underlying cause of the distress is not treated. Associated arterial blood gas measurements demonstrate evidence of acute respiratory acidosis, with a low pH, elevated Pco2, and low Po2. Acute hypoxemia alone due to spontaneous pneumothorax does not lead to respiratory arrest because hypoxemia supports the respiratory drive.

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Question: 33

A 4-year-old boy presents with a 3-month history of a recurring pruritic eruption on the abdomen. The parents report that each outbreak lasts 1 to 2 weeks. No one else in the home is similarly affected. Physical examination reveals clustered erythematous papules measuring 4 to 6 mm in diameter (Item Q33). A few papules have a central punctum.

Of the following, the MOST likely diagnosis is

A. folliculitis

B. Gianotti-Crosti disease

C. molluscum contagiosum

D. papular urticaria

E. scabies

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Question: 33



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Critique: 33 Preferred Response: D

Recurring crops of pruritic, clustered erythematous papules, some having a central punctum, suggests the diagnosis of papular urticaria (Item C33A). Lesions represent a delayed hypersensitivity reaction (not local immediate reaction) to stinging or biting arthropods. Dog or cat fleas usually are responsible. However, mosquitos; lice; or fowl, grain, or grass mites may be implicated. Typically, affected children are 18 months to 7 years of age and are the only members of their families to have symptoms. Each crop of papules lasts 2 to 10 days, and recurrences may be observed for 3 to 9 months. This history and the observation that some lesions lack puncta distinguish papular urticaria from immediate insect bite reactions.

The first step in management is to remove the offending agent. Animals should be examined for fleas or mites and treated appropriately. If fleas or mites are identified, treatment of the household environment by a pest control service is warranted. Established lesions may be treated with an appropriate topical corticosteroid and if pruritus is severe, an oral sedating antihistamine. If outdoor insects are implicated, patients may be advised to wear long sleeves and pants and to apply an insect repellent.

Folliculitis is an infection of hair follicles often caused by *Staphylococcus aureus*. Lesions are clustered small pustules that have surrounding erythema (Item C33B). Gianotti-Crosti disease is a unique childhood exanthem observed in association with viral or bacterial infections or with immunizations. Affected individuals develop erythematous papules on the face and extremities, with relative sparing of the trunk (Item C33C). The lesions of molluscum contagiosum are translucent or white papules; larger lesions often have a central umbilication (Item C33D). Children who have scabies develop erythematous papules or vesicles. In infants, the eruption is generalized (Item C33E); in older individuals, lesions are concentrated in flexural areas (eg, interdigital webs, wrist flexures), at the belt line, on the penis and scrotum in males, and on the areolae in women.

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Critique: 33

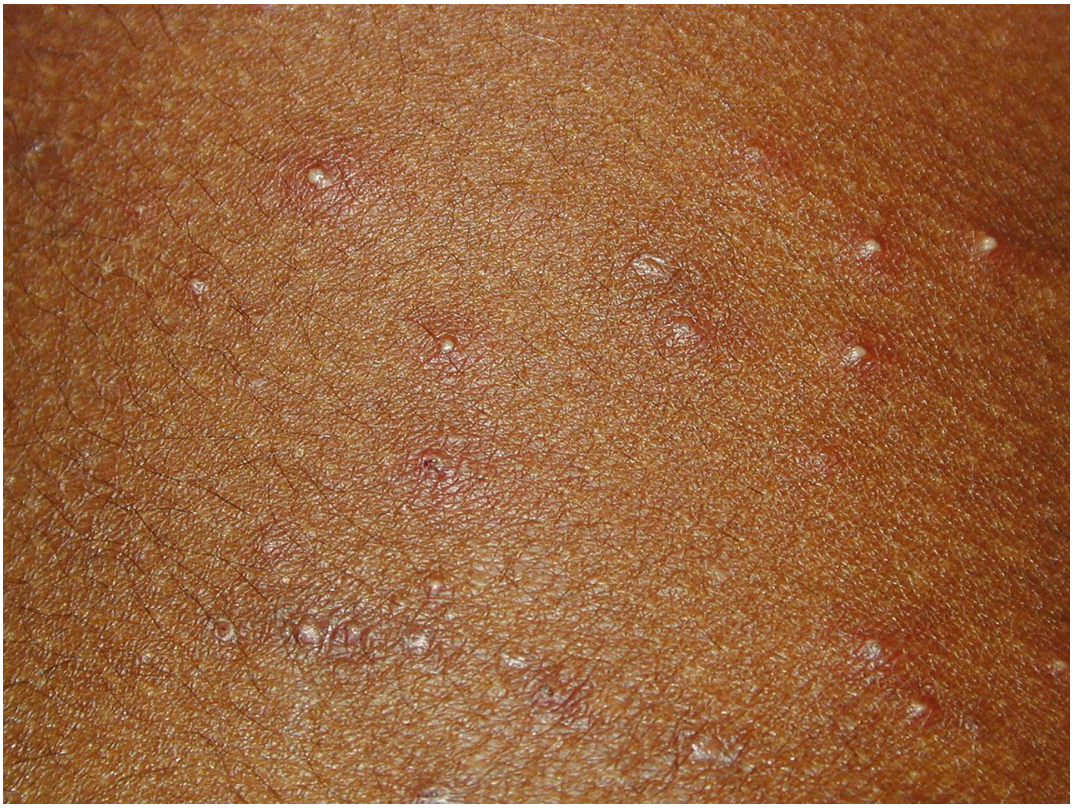


Papular urticaria is characterized by recurring crops of erythematous papules, some of which have a central punctum.

Courtesy of dermatlas.org

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Critique: 33



Folliculitis is characterized by small pustules with surrounding erythema.

Courtesy of D. Krowchuk

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Critique: 33



Erythematous papules that involve the face and extremities, with relative sparing of the trunk, are observed in Gianotti-Crosti disease.

Courtesy of W.W. Tunnessen, Jr

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Critique: 33



The lesions of molluscum contagiosum are translucent papules. Larger lesions have a central umbilication (arrow).

Courtesy of D. Krowchuk

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Critique: 33



In infants, scabies produces a generalized eruption composed of erythematous papules or vesicles.

Courtesy of D. Krowchuk

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Question: 34

A 10-week-old infant has undergone abdominal surgery for gastroschisis. After 6 weeks of parenteral nutrition (PN), cholestasis has developed.

Of the following, the intervention that is MOST likely to reduce the severity of cholestatic liver disease due to PN is

A. addition of 800 IU of alpha-tocopherol (vitamin E) to the daily PN

B. early introduction of hypocaloric (trophic) enteral feeding

C. elimination of intravenous long-chain triglyceride supplementation

D. reduction of the dextrose concentration of the PN to 15% (15 g/100 mL)

E. removal of branched-chain amino acids from the PN

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Critique: 34 Preferred Response: B

Parenteral nutrition (PN) is a lifesaving therapy in the neonate or infant who requires a prolonged period of fasting. Total parenteral nutrition (TPN) involves the administration of intravenous dextrose, free amino acids, lipids, and electrolytes into a central vein (typically the subclavian). Trace elements, including zinc, iron, and selenium, must be added if prolonged fasting is expected. For infants, prolonged TPN is used primarily in those who are critically ill and have bowel dysfunction due to gastrointestinal malformations (gastroschisis, omphalocele) or necrotizing enterocolitis. The four primary complications of this treatment are infection, thrombosis, electrolyte abnormalities, and cholestasis. Risk factors for cholestasis include severe necrotizing enterocolitis, prolonged bowel rest, bacterial overgrowth, and recurrent catheter-related sepsis.

Early enteral feeding, even in small volumes, is believed to treat PN cholestasis by stimulating bile flow and promoting intestinal motility. Although no randomized trials have proven definitely the concept that trophic feedings reduce cholestasis, animal studies suggest that they improve intestinal mucosal integrity and pancreaticobiliary function. Therefore, trophic feedings are recommended if patients can tolerate them. Additional interventions that may reduce PN cholestasis include: not giving excessive glucose, using ursodeoxycholic acid to increase bile flow, and treating bacterial overgrowth. Another strategy is to provide the patient's daily PN in a condensed time period ("cycling" the PN). A recent small case series also suggested that using an omega-3-based lipid emulsion (instead of the more commonly used omega-6-based emulsion) can treat cholestasis successfully. Reduction of dextrose, removal of branched-chain amino acids, addition of vitamin E, and elimination of the long-chain fat preparation have not been shown to be effective treatments for cholestasis.

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Question: 35

You are evaluating a 3-day-old preterm infant who was born at 26 weeks' gestation and weighed 800 g. Her blood pressure has dropped acutely, and she has developed seizures. Physical examination demonstrates equal mechanical breath sounds, no heart murmur, hypotonia, a bulging anterior fontanelle, and lethargy. Laboratory evaluation reveals anemia, metabolic acidemia, and hyperglycemia.

Of the following, the MOST likely explanation for these findings is

A. acute pneumothorax

B. intracranial hemorrhage

C. late-onset sepsis

D. patent ductus arteriosus

E. perinatal asphyxia

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Critique: 35 Preferred Response: B

The clinical presentation of intraventricular hemorrhage (IVH) in the preterm infant can vary from asymptomatic (in up to 75% of cases, usually with a mild-to-moderate grade I (Item C35A) to III (Item C35B) hemorrhage) to profound hemodynamic, metabolic, and acid-base abnormalities (in the more severe grade III and grade IV (Item C35C) parenchymal hemorrhages). The infant described in the vignette is in a high-risk category for experiencing an IVH due to her extreme prematurity and extremely low birthweight (incidence of 25% to 50%). Her acute instability, bulging fontanelle, lethargy, and seizure coupled with acidosis and anemia are explained best by a severe grade IV IVH. Most such hemorrhages occur in the first 96 hours of postnatal life. Laboratory data in IVH often document hyperglycemia (an indication of severe stress), anemia (acute decrease in hematocrit), thrombocytopenia (consumption), acidemia (metabolic, reflecting both tissue damage and hypovolemic shock), and hyponatremia indicating inappropriate secretion of antidiuretic hormone.

Managing IVH, or any other intracranial hemorrhage, requires initial stabilization of the airway, control of respiratory function (apnea, hypercarbia, and hypoxia are common), and support of the circulation (packed red blood cell transfusion) as well as correction of acidosis and hyperglycemia. Anticonvulsant therapy with phenobarbital generally is initiated in patients demonstrating seizures. The imaging study of choice for an unstable preterm infant is bedside cranial ultrasonography. For a more mature and stable patient or one for whom subarachnoid or subdural hemorrhage is the principal concern, computed tomography scan of the head is preferred. An investigation into any underlying contributing problems such as sepsis or coagulopathy is necessary. In the face of central nervous system injury, hypercarbia, hypoxia, and hypotension should be avoided.

Acute pneumothorax has been associated with IVH in preterm infants who have respiratory distress. The incidence has declined with the use of surfactant and improved ventilation strategies. The equal breath sounds reported for the infant in the vignette do not suggest a pneumothorax. By definition, late-onset sepsis occurs beyond 3 days after birth. Patent ductus arteriosus (PDA) does not manifest with anemia and a bulging fontanelle. The ligation of a PDA, accompanied by abrupt hemodynamic changes, has been an argued contributory cause to IVH, but this is less of a concern with pharmacologic treatment of the PDA using indomethacin. Perinatal asphyxia is associated with acidemia, seizures, and other problems in the first 24 hours after birth.

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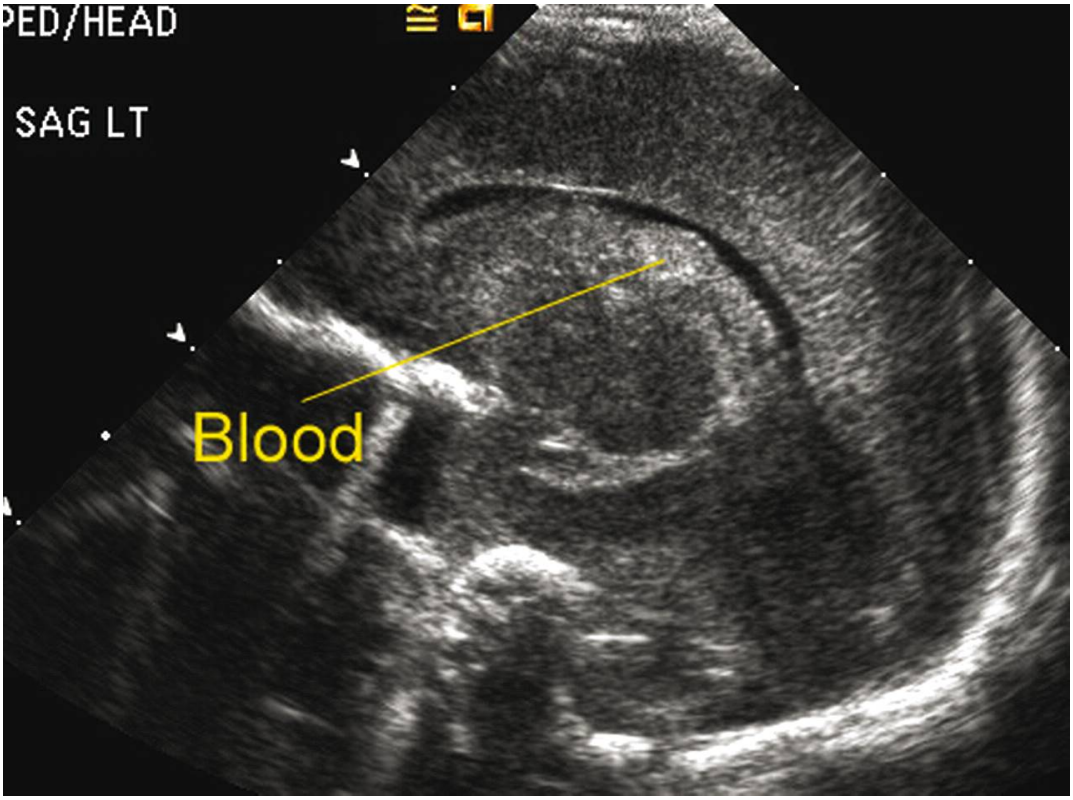
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Critique: 35

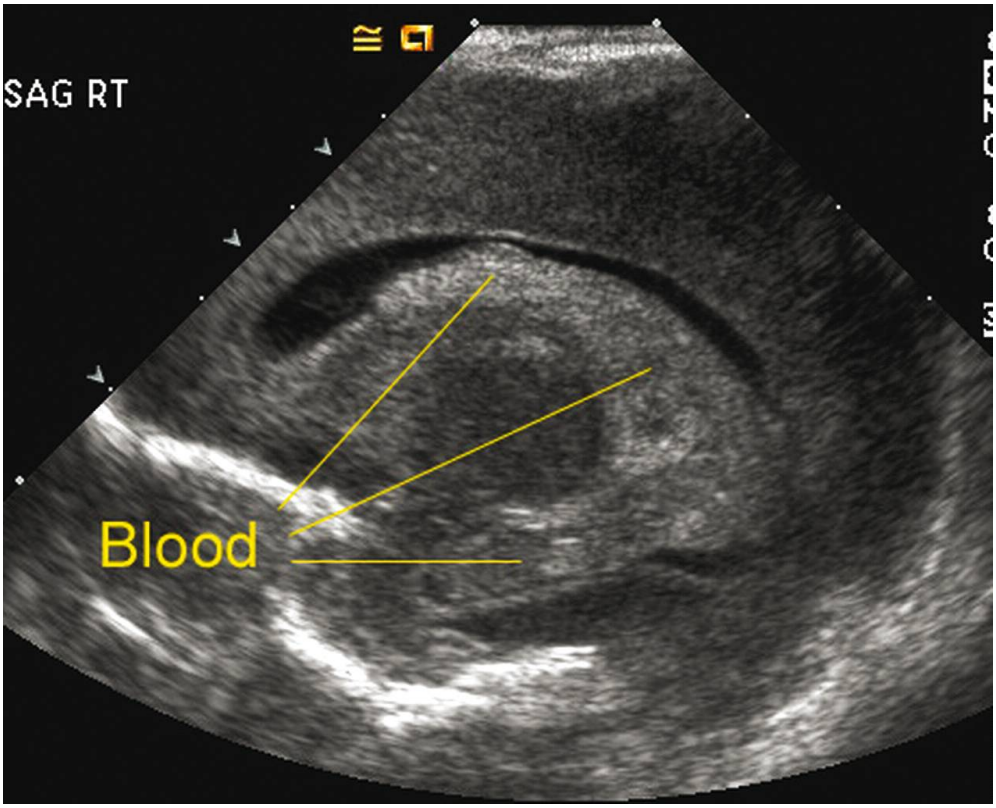


Grade I intraventricular hemorrhage: Sagittal ultrasonography of the head shows a small subependymal germinal matrix hemorrhage.

Courtesy of D. Mulvihill

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Critique: 35

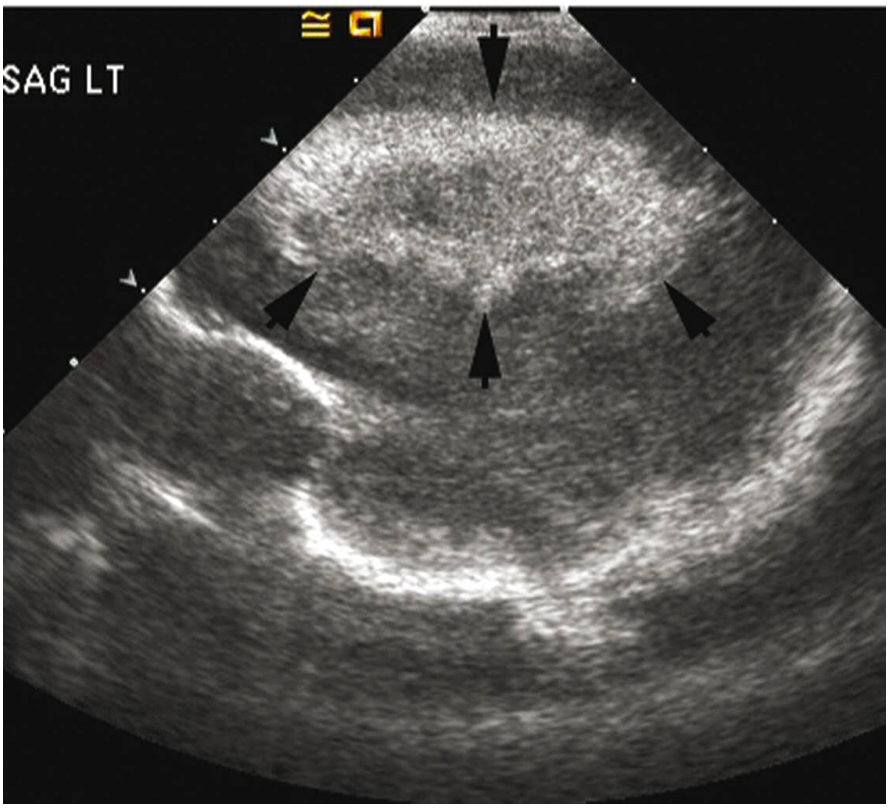


Grade III intraventricular hemorrhage: Sagittal ultrasonography of the head shows a dilated lateral ventricle filled with echogenic material representing blood.

Courtesy of D. Mulvihill

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Critique: 35



Grade IV intraventricular hemorrhage: Sagittal ultrasonography of the head shows a large parenchymal hemorrhage (defined by arrows) in the periventricular area.

Courtesy of D. Mulvihill

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Question: 36

During the prenatal visit with new parents, a mother expresses concern about regulating the temperature of the bath water for the new baby. You tell her that standards regarding hot water heaters have been determined.

Of the following, the temperature that has been determined to be appropriate for hot water heaters is CLOSEST to

A. 110ºF

B. 120ºF

C. 130ºF

D. 140ºF

E. 150ºF

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Critique: 36 Preferred Response: B

Burns are among the most common accidental injuries in infants and children, and scalding injuries occur most frequently. Children commonly are burned by hot liquids spilled from a table or stove, but approximately 3,800 injuries and 30 deaths each year are reported to be due to burns from tap water that is too hot.

The amount of contact time required to produce a third-degree burn is less than parents may realize.

Water Temperature	Time
150ºF	2 seconds
140ºF	6 seconds
130ºF	30 seconds
120ºF	5 minutes

Therefore, the Consumer Product Safety Commission has recommended that all water heaters be set to 120ºF. Parents should call their local electric or gas companies for instructions on adjusting the temperature. Parents who live in apartments should talk with their building managers about having the temperature lowered because the hot water heaters in most apartment buildings are set at higher temperatures to provide hot water to all tenants. In addition to setting hot water heaters to a lower temperature, parents should hand-test water before bathing children and infants, and young children never should be left unsupervised in the bathroom.

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Question: 37

A 7-year-old child who has autism presents with purulent, bloody discharge from the ear of 1 day's duration. His mother is unaware of any antecedent events. The child is unable to provide any history, but his mother explains that he is attending day camp. Examination of the affected ear reveals blood-tinged, purulent discharge that prohibits visualization of the tympanic membrane and apparent tenderness to touch of the pinna.

Of the following, the MOST appropriate next step is to

A. lavage the ear in an attempt to examine the tympanic membrane

B. prescribe oral trimethoprim-sulfamethoxazole

C. prescribe topical fluoroquinolone solution

D. prescribe topical tobramycin

E. refer the patient immediately to an otolaryngologist

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Critique: 37 Preferred Response: C

A child who has purulent ear drainage can present a diagnostic challenge. For the child who has no history of tympanostomy tube placement, potential diagnoses include otitis externa, a foreign body in the external meatus with associated irritation, or suppurative otitis media with perforation (Item C37) . For children who have a history of tympanostomy tube placement, the purulent drainage flows through a patent tube. In all cases, careful history and ear examination are required, although visualization of the tympanic membrane may be difficult or impossible due to the obscuring drainage, as described for the boy in the vignette.

Otitis externa usually is associated with swimming or other water activity in the absence of respiratory symptoms (eg, cough, rhinorrhea) or fever. Physical examination reveals tenderness of the pinna and tragus on manipulation. Otitis media with perforation typically is associated with a history of intense ear pain preceding the appearance of ear drainage, fever, and respiratory symptoms.

The initial treatment of otitis externa, otitis media with perforation, and drainage from tympanostomy tubes is the same. Topical treatment with fluoroquinolone drops (eg, ofloxacin, ciprofloxacin) has been found to be as effective as systemic therapy for perforated otitis media. If systemic (oral) therapy is employed, amoxicillin or a suitable alternative may be selected; trimethoprim-sulfamethoxazole is not indicated due to a high prevalence of resistance among causative organisms. There is no evidence of the superiority of combined topical and systemic therapy**,** and safety of this regimen has not been evaluated completely. Re-examination should be considered for children who do not improve after 3 to 5 days of topical therapy; waiting for completion of a full 10-day course may delay the diagnosis of persistent disease, foreign body, or other complications. Use of neomycin/polymyxin or aminoglycoside (eg, tobramycin) drops may cause ototoxicity and is best avoided.

Lavaging the ear is contraindicated if there is a chance that the tympanic membrane is perforated. Otolaryngology referral may be indicated for children who fail to respond to topical therapy, those in whom the inability to see the tympanic membrane persists, or those in whom the perforation does not heal over a period of weeks.

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Critique: 37

Item 37: Distinguishing Features of Three Types of Otitis Infections

	Otitis Externa (OE)	Acute Otitis Media (OM) With Perforation	Chronic Suppurative Otitis Media With Perforation
History			
Pain	Present; may be severe; also pruritis	Present; may note relief after perforation	Rare
Fever	Uncommon	May be present	Rare
Season	Summer	Winter/spring	Variable
Drainage	Mucopurulent; progresses with pain	Purulent; noted after perforation	Purulent; persists >2 wk
Associations	Swimming	Upper respiratory tract infection; history of OM	Perforation of tympanic membrane; recurrent OM
Physical Examination			
Ear canal	Edematous, erythematous, tender; tragus/pinna tender	Debris in canal; edema, erythema, tenderness depend on degree of secondary OE	Chronic inflammation; debris in canal
Tympanic Membrane (TM)	Usually normal	Perforation; red, opaque/purulent drainage behind TM	Perforation secondary to OM or tympanostomy tubes
Microbiology	*Pseudomonas aeruginosa; Staphylococcus aureus*	*Streptococcus pneumoniae; Haemophilus influenzae*	*Pseudomonas aeruginosa; Staphylococcus aureus*

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Question: 38

A 1-week-old infant presents to the emergency department with a 1-day history of poor feeding, pallor, diaphoresis, and increasing somnolence. She was born at term, and the delivery was uncomplicated. On physical examination, her heart rate is 180 beats/min, respiratory rate is 90 breaths/min, and blood pressure is 50/30 mm Hg. Her breath sounds are shallow, and cardiac evaluation reveals no murmurs but a gallop rhythm. Her liver is palpable at 3 cm below the costal margin. Her extremities are cool, pale, and mottled, and she has poor distal pulses. After you administer normal saline at 20 mL/kg, her heart rate is 194 beats/min.

Of the following, the MOST appropriate next step is

A. adenosine infusion at 50 mcg/kg

B. computed tomography scan of the head

C. dopamine infusion at 10 mcg/kg per minute

D. lumbar puncture followed by antibiotics

E. normal saline infusion at 20 mL/kg

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Critique: 38 Preferred Response: C

The newborn described in the vignette has the clinical signs and symptoms of diminished systemic perfusion and shock. Causes of shock in the neonate may include hypovolemia, sepsis, metabolic imbalance, and cardiogenic problems. The tachypnea described for the child likely results from pulmonary congestion caused by decreased filling of the failing left ventricle. The auscultatory corollary is the presence of the gallop rhythm caused by the abnormal filling of the noncompliant left ventricle. Abnormal filling of the left ventricle results from the chamber's inability to eject contents adequately, which may be caused by an acute afterload (left heart obstruction with a closing ductus arteriosus) or a cardiomyopathy (genetic, metabolic, or intrinsic). As filling of the left ventricle diminishes, pressure increases in the left atrium and subsequently the pulmonary veins, capillaries, and arteries. This pressure is transmitted back to the right ventricle, which becomes progressively hypertensive and may begin to fail. When right ventricular failure occurs, the systemic veins must drain at higher pressure into the failing right heart, which leads to hepatic congestion and enlargement, with the liver edge becoming palpable well below the costal margin, as described for the infant in the vignette.

The tachypnea exacerbates the poor feeding that results from the infant's inability to generate a prolonged suck while maintaining nasal breathing. When poor intake is coupled with increased water losses through the respiratory tree, hypovolemia, dehydration, and decreased urine output ensue. Lethargy may reflect decreased perfusion to the brain and may be exacerbated by the metabolic acidosis that results from inadequate tissue perfusion. The tachycardia is often compensatory to maintain cardiac output (cardiac output = heart rate x stroke volume). As the ability to maintain cardiac output fails, the blood pressure falls, as reported for this infant, and the compensated shock becomes uncompensated.

Administration of colloid or crystalloid often improves the cardiac output of patients who have shock due to hypovolemia or sepsis by allowing for increased stroke volume as the patient's "tank" becomes filled. The tachycardia may begin to resolve following initiation of this therapy. Conversely, those who suffer from cardiogenic causes of shock may not improve with the addition of volume, which may exacerbate further congestion in the pulmonary circuit and volume in the already dilated left ventricle. The Frank-Starling mechanism, which relates cardiac muscle fiber shortening to left ventricular end-diastolic volume, demonstrates that beyond a certain volume, when the heart is significantly dilated, the addition of volume may lead to diminished fiber shortening and reduced function and stroke volume. This can be characterized by an increase in tachycardia as attempts continue to maintain cardiac output in the face of the diminished stroke volume. The increase in heart rate after the administration of fluids reported for the patient in the vignette signals a cardiogenic cause for her shock.

Immediate management of this critically ill infant includes the administration of inotropic medications to improve cardiac muscle fiber shortening, thereby increasing stroke volume and cardiac output. Dopamine is an excellent drug for patients who have cardiogenic shock with hypotension. Dopamine has both inotropic effects on the heart and vasoconstrictive properties that can help to maintain or improve systemic blood pressure.

Adenosine, a fast-acting atrioventricular node blocker, is used to treat supraventricular tachycardia, commonly characterized in infants by heart rates greater than 240 beats/min. Administering more intravenous fluids can lead to progression of the cardiac failure. Transporting a patient such as the one described in the vignette for computed tomography scan or placing her in the positions necessary for lumbar puncture may lead to further hemodynamic compromise. Administering empiric antibiotics in such cases and obtaining cultures as the shock is being treated is appropriate.

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Question: 39

A 10-year-old boy presents with leg weakness that has progressed over 24 hours, bladder and bowel incontinence, and back pain. There is no history of trauma. On physical examination, leg reflexes are diminished, and there is numbness in the legs and lower trunk. Rectal examination demonstrates decreased tone. Sensory examination shows absent pinprick sensation below T6.

Of the following, the MOST appropriate initial diagnostic test is

A. brain magnetic resonance imaging with contrast

B. electromyography of the legs

C. lumbar puncture

D. nerve conduction velocities

E. spine magnetic resonance imaging with contrast

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Critique: 39 Preferred Response: E

The child described in the vignette has subacute rapidly progressive weakness and back pain, serious findings that require emergent diagnostic evaluation and management that includes imaging in an attempt to localize the problem to the brain, brainstem, spinal cord, nerve root, nerve, neuromuscular junction, or muscle. The reported sensory, motor, bowel and bladder symptoms are explained best by a lesion in the spinal cord. Such lesions can present in children abruptly, subacutely, or insidiously. Respiratory failure can occur due to high cervical spine lesions affecting the phrenic nerve to diaphragm function, but are not likely in this case. Autonomic difficulties other than bowel and bladder incontinence also are common in chronic spinal cord injuries, but again are unlikely in this setting. Failure to diagnose treatable spinal cord lesions can result in permanent neurologic deficits. Magnetic resonance imaging (MRI) of the thoracic spine with contrast has the highest diagnostic yield in the acute presentation (Item C39).

Subacute, bilateral progressive leg weakness could result from diseases in the brain such as hydrocephalus or a midline parasagittal lesion affecting cortical spinal tract fibers to the legs on both sides. Such conditions also could cause urinary incontinence. However, the absence of headache and alterations in consciousness, the back pain, and the reduced leg reflexes described for the boy in the vignette suggest that the cause of his leg weakness is not a brain lesion. Thus, brain magnetic resonance imaging is not the most appropriate initial diagnostic test.

Subacute progressive leg weakness also could result from peripheral nervous system disease, such as acute inflammatory demyelinating polyneuropathy (AIDP), also known as Guillain-Barré syndrome. Although the report of pain and the diminished leg reflexes for this patient are consistent with AIDP, the history of bowel and bladder incontinence and the sensory loss below the T6 spinal level do not support the diagnosis. Therefore, nerve conduction studies, which are used to diagnose nerve axonal or demyelinating disease, are not appropriate. Electromyography (which can aid in diagnosing intrinsic muscle disease and documenting denervation and reinnervation processes) and lumbar puncture (which is useful for diagnosing AIDP when the clinical findings suggest this diagnosis) are not indicated when a spinal cord lesion is suspected. AIDP can be associated with autonomic instability, loss of airway protection, and respiratory failure.

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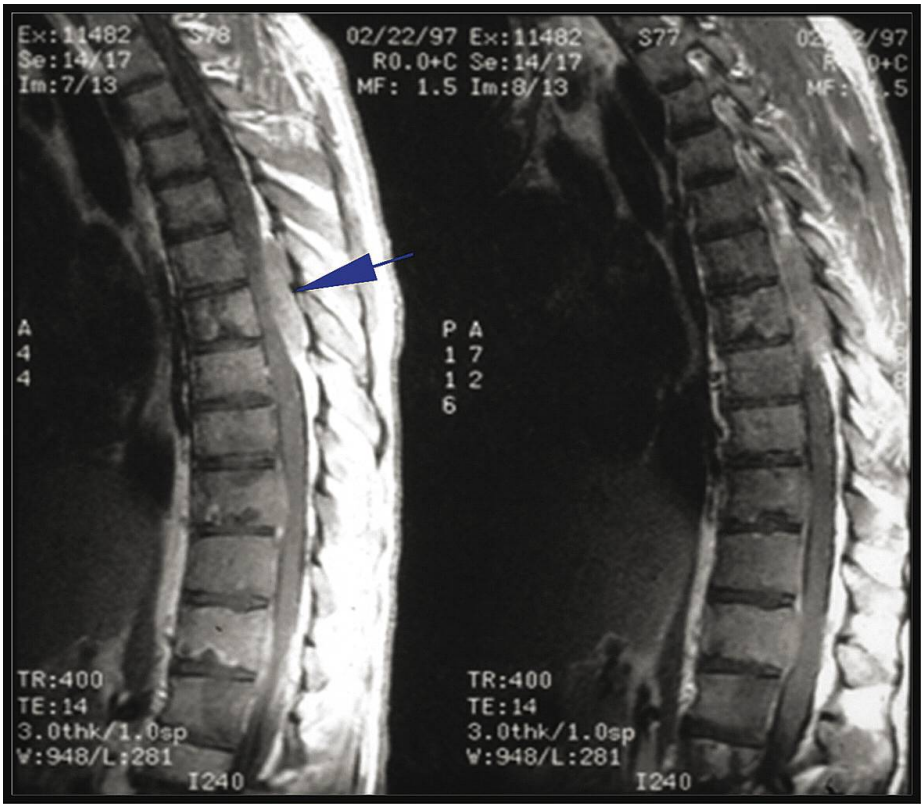
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Critique: 39



Mid-thoracic spinal cord compression caused by an extramedullary tumor (T1-weighted sagittal magnetic resonance imaging).

Courtesy of P. Fisher

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Question: 40

A mother who is new to your area brings her 6-year-old daughter to your office for evaluation due to poor school performance and behavioral concerns. The child is in the first grade and is struggling with math and reading. The mother says that the girl is “hyper” and does not play well with other children. Despite almost being hit by a car recently, she repeatedly runs into the road in front of their house. On physical examination, you note that the child’s height, weight, and head circumference are less than the 10th percentile. She has short palpebral fissures, a smooth philtrum, and a thin upper lip (Item Q40).

Of the following, the MOST likely diagnosis is

A. Down syndrome

B. fetal alcohol syndrome

C. hypochondroplasia

D. Russell-Silver syndrome

E. 47,XXX

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Question: 40



Short palpebral fissures and a thin upper lip, as described for the child in the vignette.

Courtesy of M. Rimsza

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Critique: 40 Preferred Response: B

The behavioral and physical features described for the girl in the vignette are most consistent with fetal alcohol syndrome (FAS). FAS is at one end of a spectrum of abnormalities associated with prenatal exposure to alcohol termed “fetal alcohol spectrum disorders” (FASD).

Discussion continues about the various methods used to diagnose FASD. The Institute of Medicine of the National Academy of Sciences established diagnostic criteria in 1996, which have been expounded upon further by Astley and Clarren at the University of Washington (Washington criteria) in 2000, the Centers for Disease Control and Prevention in 2004, and Hoyme and associates at Stanford in 2005. All of these maintain three key features for the diagnosis of FAS: prenatal-onset growth deficiency, characteristic facial appearance, and central nervous system damage/dysfunction.

Children who have FAS typically have height, weight, and occipitofrontal circumference (OFC) below the 10th percentile from birth and continue to grow slowly. Characteristic facial features include midface hypoplasia (including flat nasal bridge and epicanthal folds), short palpebral fissures, relatively smooth philtrum, and a narrow upper lip that has a poorly defined “cupid’s bow” (Item C40A). Psychomotor development is delayed, and there is a typical pattern of both cognitive and behavioral abnormalities, including poor impulse control and poor judgment.

An FASD diagnosis should be ascertained as early as possible to ensure the best possible outcome for affected children. Management issues include parent education, careful physical examination (with attention to vision, hearing, the heart, the kidneys, and the musculoskeletal system), appropriate referrals, neuropsychological testing with proper school placement, and anticipatory guidance.

Children who have Down syndrome (DS) also may have height, weight, and OFC below the 10th percentile, and their growth should be plotted on DS growth curves. Typically, children who have DS have greater motor delay than children who have FAS because of the presence of hypotonia. Although midface hypoplasia is seen in DS, the facial features are very distinct from those of FAS (Item C40B).

Hypochondroplasia is an autosomal dominant dwarfing condition. Affected children have relative or absolute macrocephaly (Item C40C), and growth may be normal until after 2 years of age, when velocity slows and height ultimately falls below the 10th percentile. Diagnosis is based on bone abnormalities noted on skeletal survey. Affected children are at increased risk for mental retardation, but many are cognitively normal.

Russell-Silver syndrome typically is a sporadic, prenatal-onset short stature syndrome in which head growth is spared. The face has a triangular shape, with a broad forehead and narrow chin (Item C40D). Affected children remain small throughout childhood. Intelligence is typically normal.

47,XXX is estimated to occur in 1 in 1,000 children. Affected girls are physically indistinguishable from girls who have a 46,XX chromosome complement. They may be slightly taller than their chromosomally normal female siblings. Intelligence usually is normal.

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Critique: 40



Facial features of fetal alcohol syndrome include epicanthal folds, short palpebral fissures, a relatively smooth philtrum, and a narrow upper lip.

Courtesy of M. Rimsza

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Critique: 40

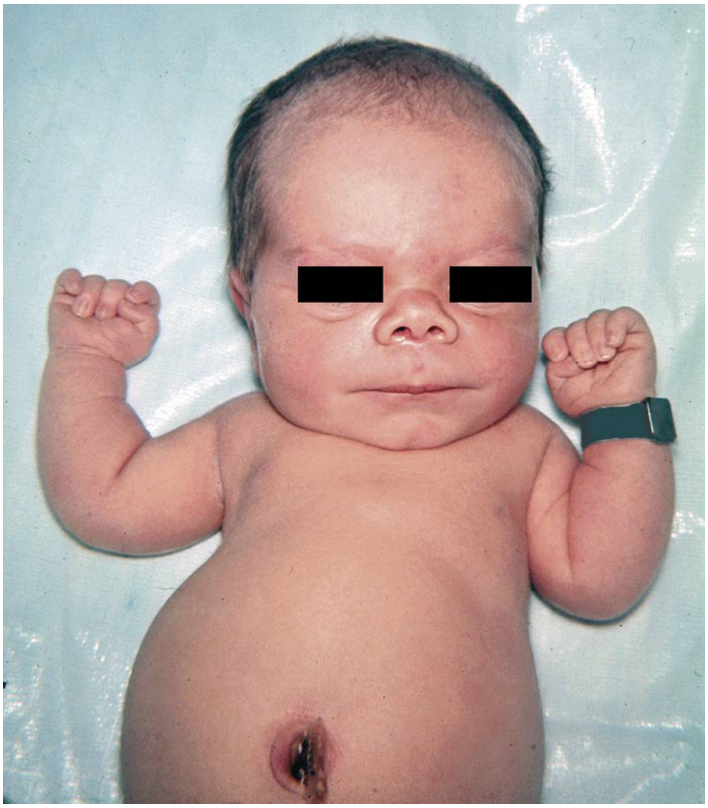


Children who have Down syndrome have epicanthal folds, upslanting palpebral fissures, and a flat nasal bridge.

Courtesy of M. Rimsza

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Critique: 40



Infants who have hypochondroplasia have findings similar to those who have achondroplasia (shown here): The head is large, the chest is small when compared with the abdomen, and there is proximal shortening of the upper extremities.

Courtesy of M. Rimsza

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Question: 41

A 16-year-old girl comes to your office with a 2-day history of moderate pelvic pain. She is sexually active and using no contraception. She has had a scant yellowish-white discharge from her vagina, but no vaginal itching. On physical examination, she is afebrile, has a heart rate of 95 beats/min and a blood pressure of 110/75 mm Hg. Bowel sounds are of normal pitch and frequency. She has no rebound or guarding on abdominal evaluation, but has moderate pelvic tenderness to palpation. She has no costovertebral angle tenderness. Findings on pelvic examination include scant yellow cervical discharge, a friable inflamed cervix, cervical motion tenderness, and mild uterine tenderness but no adnexal masses. Results of her pregnancy test are negative, and findings on urinalysis are normal.

Of the following, the MOST appropriate next step is

A. C-reactive protein measurement

B. endometrial biopsy

C. laparoscopy

D. pelvic computed tomography scan

E. testing for gonorrhea and chlamydia

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Critique: 41 Preferred Response: E

The clinical picture described for the girl in the vignette likely represents early acute pelvic inflammatory disease (PID). PID encompasses a spectrum of inflammatory disorders of the upper female genital tract and includes any combination of endometritis, salpingitis, tubo-ovarian abscess, and pelvic peritonitis. Although *Neisseria gonorrhoeae* and *Chlamydia trachomatis* are not the only causative organisms for PID, they are implicated in many cases. The Centers for Disease Control and Prevention (CDC) *Sexually Transmitted Disease Treatment Guidelines, 2006* state that "all women who are diagnosed with acute PID should be tested for *N. gonorrhoeae* and *C. trachomatis* and should be screened for HIV infection." Accordingly, the girl in the vignette should undergo nucleic acid amplification testing for gonorrhea and chlamydia. When PID is suspected and a gynecologic examination will be performed, such testing can be accomplished using a specimen obtained from the cervix. Urine-based testing is indicated for young women in whom a gynecologic examination may be unnecessary (eg, for those who are asymptomatic). Urine-based testing can also be used to screen male partners.

Acute PID may be difficult to diagnose because of the wide variations in symptoms. The CDC recommends empiric treatment in sexually active young women and other women at risk for sexually transmitted diseases who complain of pelvic or lower abdominal pain if no other cause for the illness can be identified and if pelvic examination reveals cervical motion, uterine, or adnexal tenderness. Laboratory documentation of cervical infection with *N gonorrhoeae* and *C trachomatis* can support the diagnosis and is recommended. Treatment choices should be aimed at both of these organisms because negative endocervical test results do not rule out upper reproductive tract infection.

Measurement of C-reactive protein, endometrial biopsy to show histopathologic evidence of endometritis, laparoscopy, and pelvic ultrasonography may be warranted if the diagnosis is in question or a complication of PID is suspected. Rarely, pelvic computed tomography scan may be useful.

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Question: 42

A 1-year-old boy presents with generalized seizures. His general physical examination findings are normal except for a prominently positive Chvostek response. Results of laboratory studies include total serum calcium of 4.5 mg/dL (1.1 mmol/L) and phosphorus of 8.2 mg/dL (2.73 mmol/L). Blood urea nitrogen and creatinine values are normal for age.

Of the following, the MOST likely diagnosis is

A. dietary calcium deficiency

B. hypoparathyroidism

C. hyperphosphatasia

D. vitamin D deficiency rickets

E. vitamin D-resistant rickets

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Critique: 42 Preferred Response: B

Parathyroid hormone (PTH) acts upon PTH receptors in bone and kidney, and deficiency of PTH leads to diminished release of calcium from bone and increased tubular reabsorption of phosphate. Therefore, laboratory findings in hypoparathyroidism include low serum calcium and elevated serum phosphate concentrations, as described for the baby in the vignette.

Dietary calcium deficiency is not associated with an elevated phosphate value because PTH values are increased in this condition. Hyperphosphatasia (juvenile Paget disease) is an autosomal recessive disorder characterized by long bone deformity, with widened diaphyses and kyphosis, and sometimes associated with a mutation in the gene encoding osteoprotegerin, a hormone that regulates osteoclast development. Vitamin D deficiency and vitamin D-resistant rickets are associated with low serum phosphate concentrations. PTH concentrations may be elevated to compensate for the failure of 1,25-hydroxyvitamin D to enhance calcium uptake from the gut. Elevated PTH concentrations enhance tubular excretion of phosphate, leading to low phosphate values. In the past, low calcium and elevated phosphate values in a baby might have been associated with feeding of high-phosphate cow milk, but this rarely is seen today. Renal failure is another possible cause of hypocalcemia with hyperphosphatemia but is unlikely in an infant who has normal creatinine values.

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Question: 43

The parents of a 12-year-old boy are concerned about his changing behavior. He was previously a straight A student, but now he is getting Cs in most of his classes. He no longer wants to be with his friends after school, and he recently quit the basketball team. On weekends, he sleeps much of the day, and when awake, he stays alone in his room. Findings on his physical examination are normal. He appears apathetic and speaks softly in a monotone.

Of the following, your MOST appropriate next step is to

A. evaluate him for attention-deficit/hyperactivity disorder

B. recommend that he rejoin his basketball team

C. refer him for educational evaluation

D. refer him for mental health evaluation

E. schedule a follow-up evaluation in 1 month

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Critique: 43 Preferred Response: D

The boy described in the vignette has behavioral manifestations of depression. It is not unusual for youth to have occasional feelings of sadness, but it is important to consider if the child is truly depressed and if there is impairment of daily functioning. The *Diagnostic and Statistical Manual of Mental Disorders* edition IV criteria for major depressive disorder (MDD) are depressed or irritable mood, decreased interest, or decreased pleasure that lasts for at least 2 weeks accompanied by changes in both cognitive and physical functioning. Dysthymic disorder is a less severe type of depression that involves chronic symptoms that last for at least 1 year. Approximately 2% of school-age children and 4% to 6% of adolescents have MDD at some period. Prior to puberty, there is equal prevalence in boys and girls. After puberty, there is a 2:1 ratio of boys to girls.

The following two questions are recommended for MDD screening by the United States Preventive Services Task Force: “Over the past 2 weeks have you ever felt down, depressed, or hopeless?” “Have you felt little interest or pleasure in doing things?” Depending on the answer to these questions, additional screening or diagnostic tools or referral to a mental health specialist may be necessary. Based on the history of changes in affect of the boy described in the vignette, referral for mental health evaluation is appropriate. It is important to note that suicide is the third leading cause of death among children and adolescents and that depression is an important risk factor for suicide. The goals of treatment for MDD are to ensure the child’s safety and establish good communication between the child and his or her parents.

The overt sadness exhibited by the boy in the vignette is not characteristic of attention-deficit/hyperactivity disorder. Having him rejoin the basketball team without addressing his mood would not treat the underlying problem. Although his grades have declined recently, this is not unusual in MDD, and other findings on the history are not supportive of a learning disability that requires educational evaluation. Having the boy return for follow-up in 1 month without any appropriate treatment places him at possible emotional risk.

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Question: 44

You are treating a 2-year-old girl who has suspected meningococcal bacteremia and meningitis. Over the past 2 hours, she has required multiple fluid boluses and inotropic support to help maintain her blood pressure. She has been intubated due to respiratory failure. Her temperature is 96°F (35.6°C), and she is covered in a petechial and purpuric rash (Item Q44). Her most recent laboratory results reveal a white blood cell count of 1.2x103/mcL (1.2x109/L) with 80% lymphocytes, 10% neutrophils, and 10% band forms and a platelet count of 32x103/mcL (32x109/L).

Of the following, the MOST important additional laboratory test is

A. erythrocyte sedimentation rate

B. measurement of creatine kinase

C. measurement of fibrinogen

D. measurement of lactic acid

E. review of peripheral blood smear

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Question: 44



Purpura, as described for the girl in the vignette.

Courtesy of G. Schutze

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Critique: 44 Preferred Response: C

Any patient presenting with hyperthermia/hypothermia and a petechial/purpuric rash (Item C44) should be considered as having a potentially life-threatening bacterial infection such as that caused by *Neisseria meningitidis*. The patient described in the vignette also demonstrates signs and symptoms of disseminated intravascular coagulation (DIC) associated with septic shock. DIC is characterized by the consumption of clotting factors, anticoagulant proteins, and platelets. This process leads to widespread intravascular deposition of fibrin, which results in tissue ischemia and necrosis, generalized hemorrhage, and hemolytic anemia. Venipuncture sites frequently bleed, and there may be a petechial, purpuric, or even ecchymotic rash. Tissue necrosis can involve virtually any organ.

Common laboratory abnormalities in DIC include thrombocytopenia and prolonged prothrombin and partial thromboplastin times. Degradation of fibrinogen results in low serum fibrinogen concentrations as well as the presence of fibrinogen degradation products (eg, D-dimers). Although the peripheral blood smear may reveal fragmented, burr, or helmet-shaped red blood cells that indicate a hemolytic process, serial measurement of fibrinogen is a very sensitive and specific test for DIC. Elevations of creatine kinase or lactic acid are common in patients who have septic shock, and an erythrocyte sedimentation rate may be elevated or depressed. Abnormal findings in these tests offer little helpful information in patients who have DIC.

The most important steps in treating DIC are to find and treat the cause and correct the shock, acidosis, and hypoxia that complicate DIC. If these problems can be controlled, the bleeding quickly stops. Blood components, such as platelets, cryoprecipitate, or fresh frozen plasma, also may be required to help stop the hemorrhage. Continuous intravenous infusion of heparin and administration of specific factor concentrates (eg, activated protein C) are not used routinely in children.

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Critique: 44



Purpura in a child who has meningococcemia.

Courtesy of M. Rimsza

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Question: 45

You are hiring a pediatrician as a hospitalist at a community hospital.

Of the following, the disease for which immune status MUST be documented at the time of employment is

A. diphtheria

B. hepatitis A

C. measles

D. meningococcal disease

E. tetanus

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Critique: 45 Preferred Response: C

Health-care personnel are in contact with patients who may have contagious, vaccine-preventable diseases and are at increased risk for contracting such diseases. Further, the employee who becomes infected is at risk for transmitting the disease to other patients who are susceptible to the disease. Therefore, all health-care personnel should protect themselves and susceptible patients by receiving appropriate immunizations. The vaccine-preventable infections that are of special concern to people involved in the health care of children include rubella, measles, mumps, hepatitis B, influenza, varicella, and tuberculosis. The immune status of the health-care worker against these diseases should be documented at the time of employment. Persons found to be susceptible should receive the appropriate vaccine or vaccine series for the disease to which they are susceptible. Diphtheria, hepatitis A, meningococcal disease, and tetanus are not commonly transmitted by patients in a health-care setting and do not require documentation of immune status.

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Question: 46

A 6-year-old child presents for a health supervision visit. On physical examination, his weight is 18 kg, height is 102 cm (<3rd percentile),pulse rate is 90 beats/min, respiratory rate is 18 breaths/min, and blood pressure is 134/88 mm Hg. Of note, he has pale conjunctivae and mild edema. Among the results of laboratory evaluation are:

• Hemoglobin, 7.5 g/dL (75 g/L)

• White blood cell count, 6.0 x103/mcL (6.0 x109/L)

• Platelet count, 275x103/mcL (275x109/L)

• Mean cell volume, 82 fL

• Reticulocyte count, 0.4% (0.004)

• Blood urea nitrogen, 94 mg/dL (33.6 mmol/L)

• Serum creatinine, 12.1 mg/dL (1,070 mcmol/L)

The stool is negative for occult blood.

Of the following, the MOST likely explanation for this patient's anemia is

A. chronic gastrointestinal blood loss

B. erythropoietin deficiency

C. folic acid deficiency

D. hemolysis

E. iron deficiency

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Critique: 46 Preferred Response: B

The patient described in the vignette exhibits poor growth, hypertension, normocytic anemia, and elevated blood urea nitrogen and creatinine values. Gastrointestinal blood loss is unlikely based on the negative stool test for occult blood. The additional finding of a low reticulocyte count can be explained best by reduced substrate for red blood cell synthesis (folate, vitamin B12, or iron), bone marrow failure, selective red cell aplasia, or erythropoietin deficiency. The normal mean cell volume (MCV) reported for the boy makes folic acid, vitamin B12, and iron deficiency unlikely. Hemolysis often is accompanied by reticulocytosis and a high MCV. Chronic blood loss results in an iron deficiency state with low MCV. Erythropoietin deficiency is expected in the presence of reduced nephron mass, as is seen in the renal failure exhibited by the child in the vignette. His renal failure most likely is chronic because of his poor growth.

Recombinant erythropoietin (rEpo) is used in patients who have severe renal dysfunction and anemia associated with decreased erythropoietin synthesis. Once the glomerular filtration rate falls below 25 mL/min per 1.73 m2 (normal, 100 to 120 mL/min per 1.73 m2), erythropoietin production usually is insufficient to maintain a normal hemoglobin concentration. Of note, rEpo also has been used to treat anemia associated with chronic inflammatory diseases. A study in adults demonstrated erythropoietin concentrations insufficiently low for the severity of anemia in the setting of inflammatory bowel disease. Treatment with erythropoietin and oral iron resulted in improved hemoglobin concentrations in most of the patients in the study.

References:

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Question: 47

You are working with a medical student in an outpatient pediatric clinic. His first case is a 4-month-old male infant who has a 4-day history of rhinorrhea, coughing, and tachypnea. On physical examination, the infant is fussy and has a temperature of 99.2°F (37.4°C), pulse rate of 110 beats/min, respiratory rate of 60 breaths/min, and pulse oximetry of 92% on room air. On auscultation, expiratory wheezes are audible in all lung fields. You discuss the differential diagnosis and ask the student what he knows about respiratory syncytial virus (RSV) bronchiolitis.

Of the following, the MOST accurate statement regarding RSV bronchiolitis is that

A. household pets can be colonized with RSV and may act as reservoirs for transmission to family members

B. most chest radiographs of infants hospitalized for RSV bronchiolitis appear normal

C. most infants who develop RSV bronchiolitis require hospitalization

D. recurrent wheezing may occur for several years after mild or severe RSV bronchiolitis

E. the initial RSV infection usually occurs in school-age children

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Critique: 47 Preferred Response: D

Respiratory syncytial virus (RSV) is the most common pathogen causing lower airway disease in infants and toddlers. This RNA virus of the Paramyxoviridae family occurs worldwide and causes yearly outbreaks. Transmission is via human-to-human contact, with attack rates ranging from 50% to 100% in susceptible individuals. Initial infection is almost universal by age 2 years, making a primary infection in a school-age child rare. Animals are not recognized vectors for virus transmission.

Most infants who develop RSV infection can be treated on an outpatient basis. Annual hospitalization rates are estimated to be 17 to 26 per 1,000 infants. Bronchiolitis and pneumonia are the primary manifestations of lower airway RSV disease. Chest radiographs of hospitalized infants appear normal in approximately 10% of infants, show peribronchial thickening or interstitial pneumonia in 50% to 80%, and document segmental consolidation in 10% to 25%.

Recurrent wheezing after RSV infection may occur for months to years in 33% to 50% of affected children. Males who wheeze during their RSV infection are at greater risk for prolonged airway hyperresponsiveness, recurrent wheezing, and a later diagnosis of asthma.

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Question: 48

A 2-year-old girl presents to your office with a 3-week history of a “barky” cough. According to her mother, the girl has had no fever or upper respiratory tract infection symptoms, but she has complained intermittently of a sore throat for the past 2 weeks. On physical examination, the child appears well and playful. Her throat is nonerythematous, and her lungs are clear to auscultation. Because of the persistence of her symptoms, you obtain chest radiograph (Item Q48).

Of the following, a TRUE statement about this patient’s condition is that

A. complications include bronchiectasis and pneumonia

B. immediate removal is recommended for foreign bodies retained >24 hours

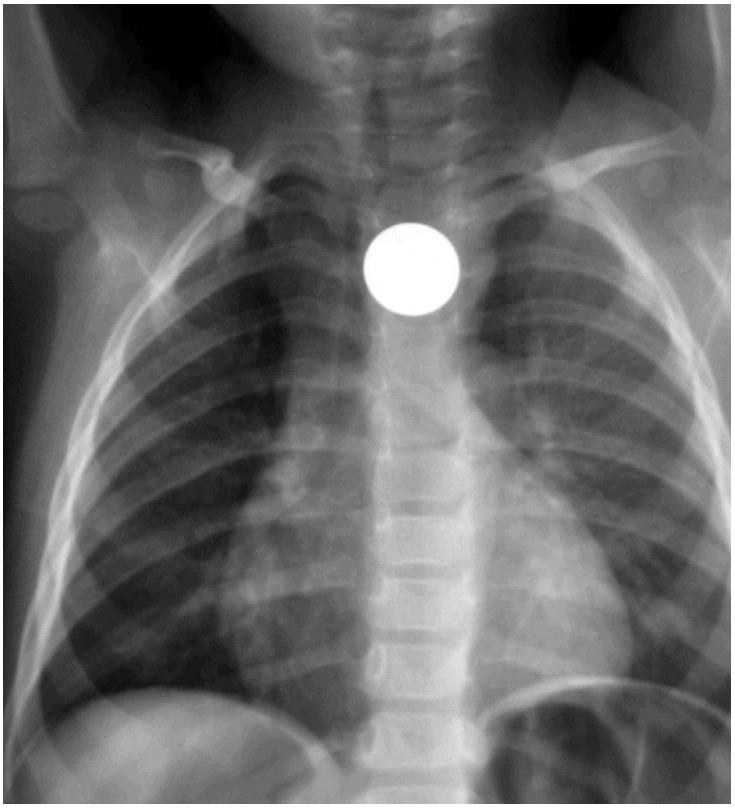
C. spontaneous passage of foreign bodies located at the thoracic inlet is likely to occur

D. the majority of patients who have retained foreign bodies are asymptomatic

E. the most likely location for a foreign body to lodge is at the gastroesophageal junction

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Question: 48



Courtesy of M. Wright

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Critique: 48 Preferred Response: B

Each year, more than 90,000 cases of ingested foreign bodies are reported to poison control centers in the United States. Most of the foreign bodies are coins, and most pass into the lower gastrointestinal tract without incident. Approximately 30% of ingested foreign bodies remain in the esophagus for hours to days, and many of these require removal. Although less than 1% of esophageal foreign bodies cause significant morbidity, complications have been reported, including esophageal erosion and perforation, esophageal stenosis, aortoesophageal or tracheoesophageal fistula, and death (Item C48A). Because most of the complications have been noted when foreign bodies are retained for more than 24 hours, current guidelines recommend removal of most foreign bodies within this time frame. Button batteries or sharp objects should be removed immediately, as should any foreign body in a significantly symptomatic patient (eg, respiratory distress, persistent retching, pain). Extra caution should be used in identifying and removing button batteries because of the potential for erosion due to corrosive electrolytic effects. In some cases, button batteries may be difficult to distinguish from coins.

Most children who have retained esophageal foreign bodies are symptomatic, although the symptoms often are nonspecific, and in 40% of cases, there is no history of ingestion. Common symptoms include drooling, chest pain, gagging, dysphagia, and refusal to eat as well as cough, stridor, or wheezing. A high clinical index of suspicion should prompt appropriate evaluation of such patients that includes anteroposterior and lateral radiographs (Item C48B) of the neck and chest and an upright abdominal film. The use of metal detectors for localizing metallic ingested foreign bodies has been reported and can be of use in identifying those that are below the diaphragm and, therefore, require no further intervention. Esophageal foreign bodies most commonly are retained at the thoracic inlet, at the level of the aortic arch, and at the lower esophageal junction. Of these three locations, the thoracic inlet is the site in more than 60% of cases.

Spontaneous passage has been reported in about one third of esophageal foreign bodies. Objects retained at the lower esophageal sphincter are most likely to pass within 24 hours of ingestion.

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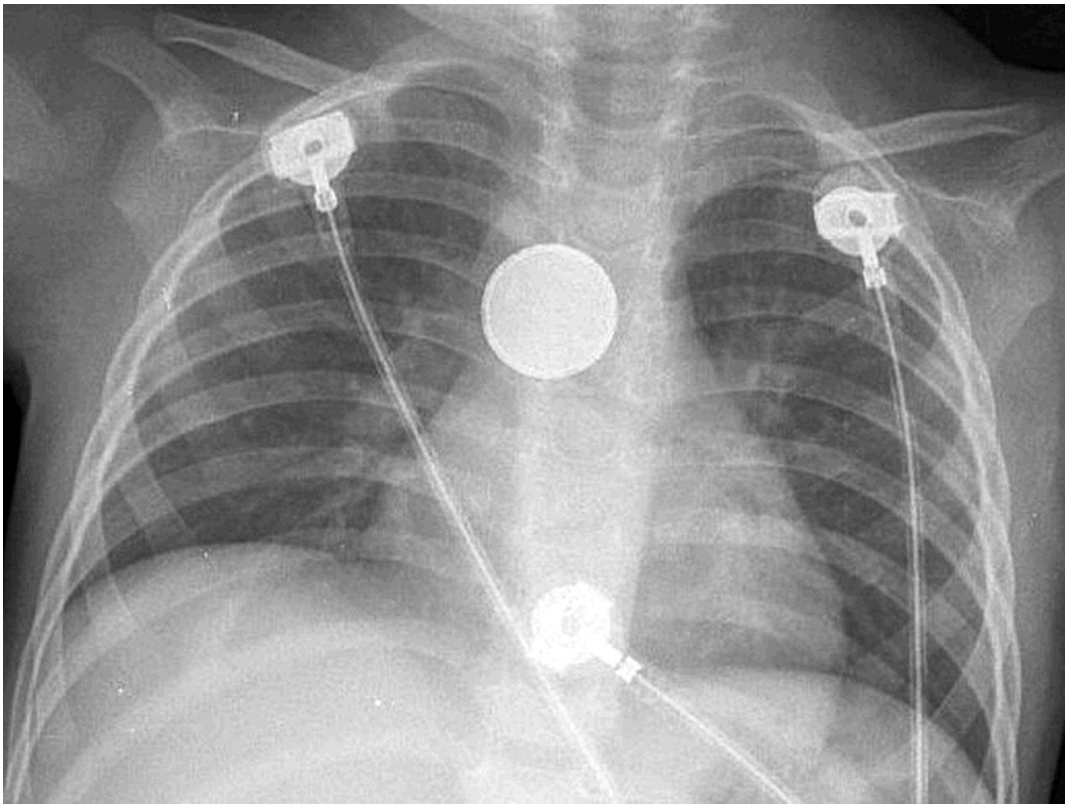
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Critique: 48

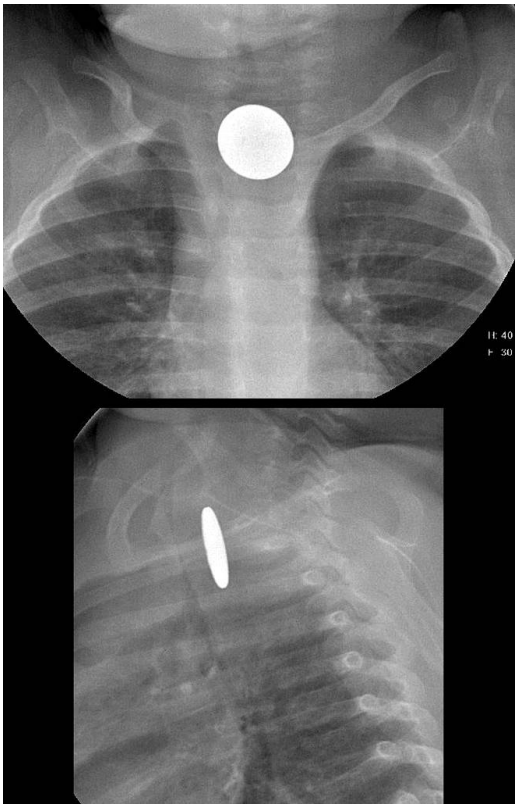


A toddler ingested a watch battery that eroded through the esophagus and aorta, resulting in death.

Courtesy of D. Mulvihill

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Critique: 48



Anteroposterior (top) and lateral (bottom) radiographs of the chest demonstrating a coin in the esophagus.

Courtesy of D. Mulvihill

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Question: 49

You are seeing an 8-month-old boy for the first time. His parents are concerned about a birth mark on his chest that has enlarged slowly over the past 5 to 6 months (Item Q49).

Of the following, the MOST appropriate management of this lesion is

A. interferon-alfa

B. intralesional corticosteroid

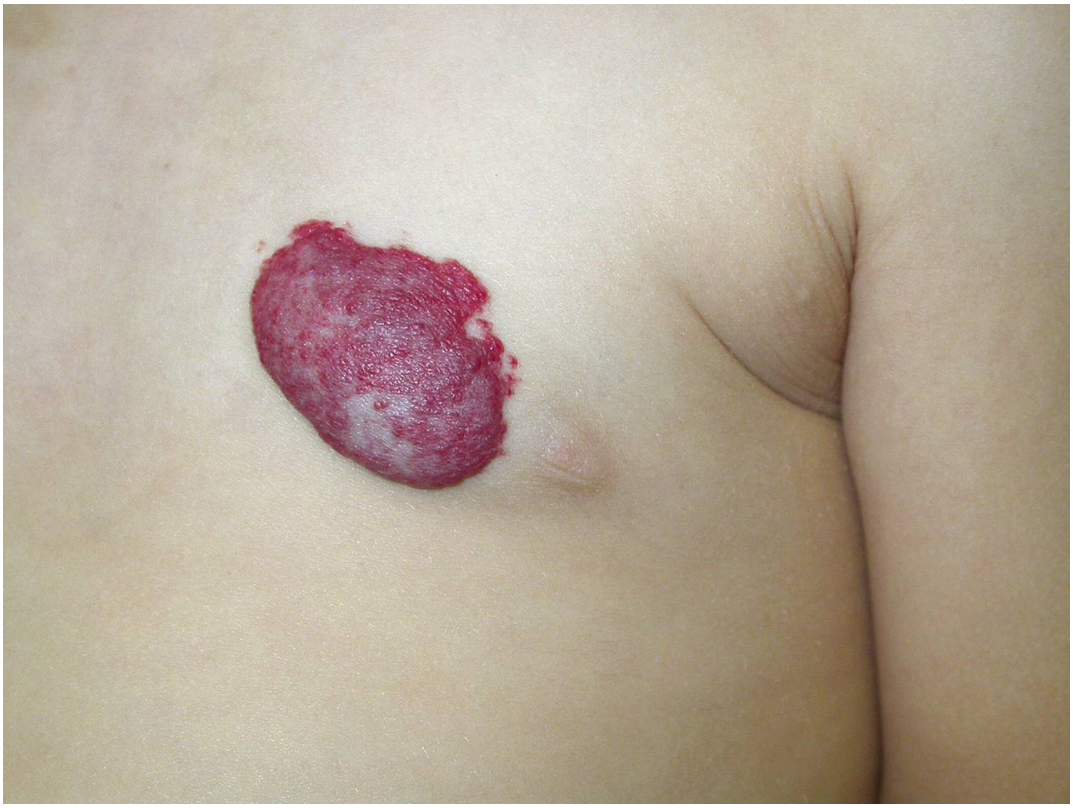
C. observation

D. pulsed dye laser

E. systemic corticosteroid

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Question: 49



Courtesy of D. Krowchuk

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Critique: 49 Preferred Response: C

Hemangiomas represent benign vascular tumors that are present in 1% of newborns and approximately 10% of 1-year-olds. They may be separated clinically into three types:

- Superficial: Bright red plaques or dome-shaped papules or nodules (Item C49A)
- Deep: Compressible nodules or tumors that have a bluish hue and often surface telangiectasias (Item C49B)
- Mixed: Lesions that have both superficial and deep features (Item C49C)

Most hemangiomas follow a benign course and do not threaten vital structures. Typically, they grow rapidly during the first 6 to 9 months after birth, stabilize, and subsequently begin to involute. The clinical hallmark of involution is the appearance of a gray or white discoloration on the lesion's surface (Item C49D). Complete involution is seen in 30% of children by 3 years of age, 50% by 5 years, 70% by 7 years, and 90% by 9 years. The infant described in the vignette has a superficial hemangioma that is exhibiting signs of involution. Therefore, no intervention is necessary, and the family should be counseled about the lesion and its natural history.

A variety of treatment options are available for problematic hemangiomas. Intralesional corticosteroids may be used for localized lesions, such as those involving the nasal tip (Item C49E) or ulcerated lesions in the diaper area (Item C49F). Proliferating lesions that threaten a vital structure (eg, the eye (Item C49G) or airway), are large, or are associated with systemic symptoms (eg, thrombocytopenia or disseminated intravascular coagulation as part of Kasabach-Merritt syndrome (Item C49H)) typically are treated with oral corticosteroids. Interferon-alfa is an inhibitor of angiogenesis that often is beneficial in the management of hemangiomas that fail to respond to systemic corticosteroid therapy. Hemangiomas that do not involute completely or ulcerated lesions that do not respond to local care may benefit from pulsed dye laser treatment.

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Critique: 49



A superficial hemangioma is a bright red or purple nodule or plaque.

Courtesy of D. Krowchuk

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Critique: 49



A deep hemangioma is a compressible nodule or tumor that is blue and has surface telangiectasias.

Courtesy of the Media Lab at Doernbecher

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Critique: 49

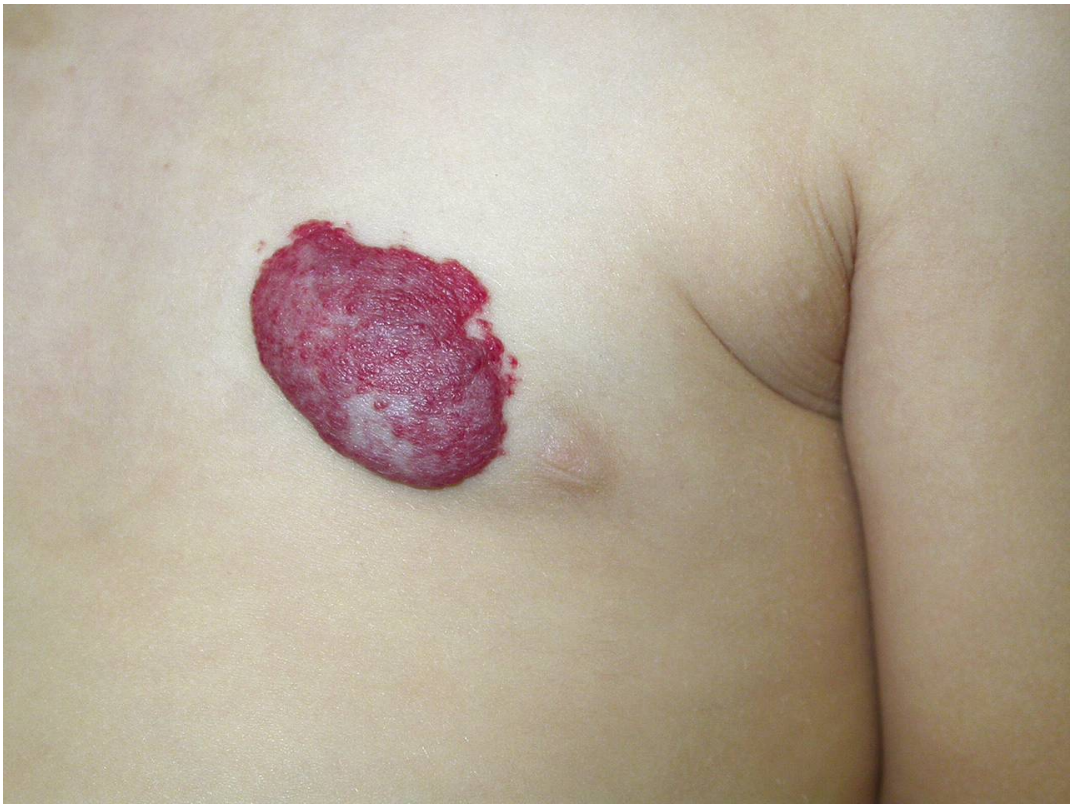


A mixed hemangioma has both superficial and deep features.

Courtesy of D. Krowchuk

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Critique: 49



The clinical hallmark of hemangioma involution is the appearance of a white or gray surface color.

Courtesy of D. Krowchuk

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Critique: 49



Hemangioma involving the nasal tip.

Courtesy of D. Krowchuk

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Critique: 49

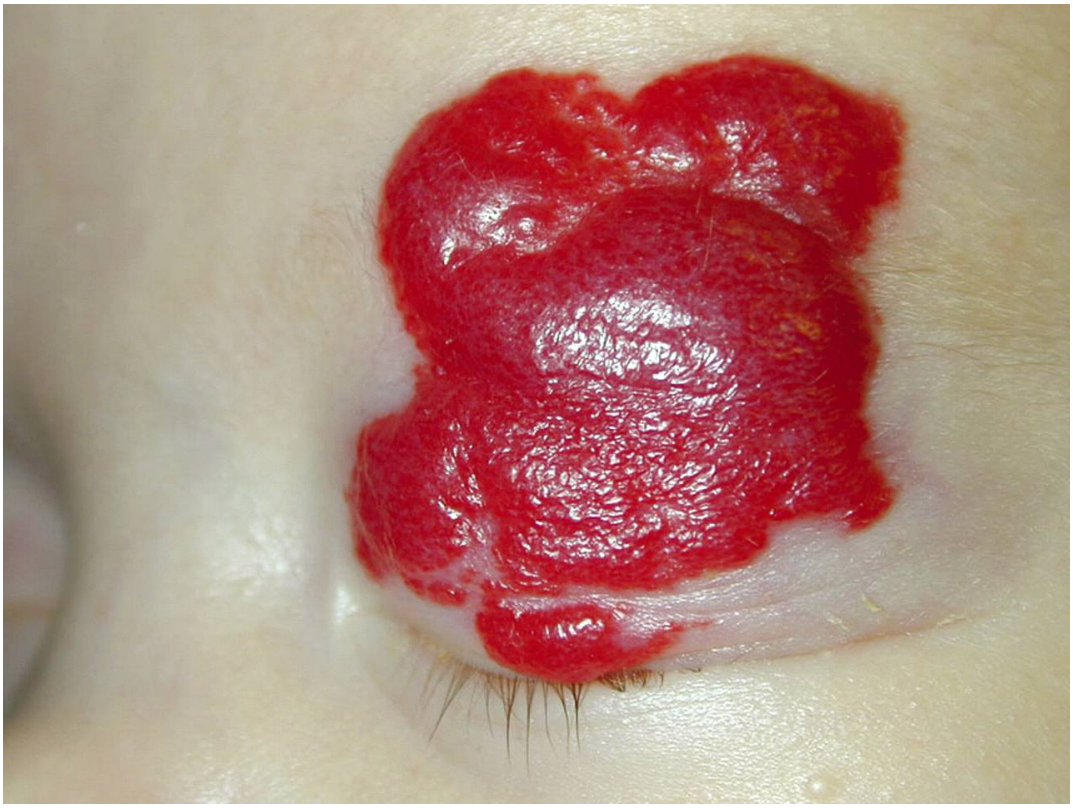


Ulcerated hemangioma in the diaper area.

Courtesy of D. Krowchuk

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Critique: 49



A proliferating hemangioma near the eye may require oral corticosteroid therapy.

Courtesy of D. Krowchuk

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Critique: 49



In Kasabach-Merritt syndrome, an atypical-appearing hemangioma (actually a hemangioendothelioma or tufted angioma) may cause thrombocytopenia or disseminated intravascular coagulation.

Courtesy of D. Krowchuk

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Question: 50

A mother brings in her 5-year-old son in for a health supervision visit. Family history reveals that the boy's father has had a soft-tissue sarcoma and a colectomy for "colon polyps." Results of the boy's physical examination are within normal limits. The mother asks if her son is at increased risk for polyps and cancer. You review the father's medical records, which indicate that his colectomy was performed at age 20 because of the discovery of 50 adenomas in the colon.

Of the following, the BEST recommendation for the son at this time is

A. a colonoscopy to survey for polyps

B. annual fecal occult blood testing

C. annual screening of serum alpha-fetoprotein

D. genetic testing to determine his risk

E. ultrasonography for testicular tumors

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Critique: 50 Preferred Response: D

The boy described in the vignette, whose father has familial adenomatous polyposis (FAP), has a 50% chance of inheriting this syndrome. This autosomal dominant condition is characterized by a mutation in the *APC* gene found on chromosome 5. Patients who have the mutation commonly develop colonic adenomas (small tumors ranging from 1 to 5 mm in size) (Item C50A) in their teenage years and have a 100% lifetime risk of developing colorectal neoplasia if the colon is not resected. Adenomas rarely develop before age 10; the mean age of adenoma development is 16 years. Other tumors associated with FAP include duodenal carcinoma, soft-tissue sarcoma, and mandibular osteomas. When FAP is associated with soft-tissue tumors such as desmoids and osteomas, it is referred to as Gardner syndrome. Approximately 1% of patients who have *APC* mutations present with hepatoblastoma in the first year after birth. Other inherited polyposis syndromes include juvenile polyposis coli, Peutz-Jegher syndrome (Item C50B), and Bannayan-Riley Ruvalcaba syndrome (Item C50C).

The availability of reliable and reproducible genetic screening has simplified the management of children born to parents who have FAP gene mutations. In this case, the father's and son's blood can be analyzed for mutations in the *APC* gene. If the son carries an FAP gene mutation, he should undergo colonoscopy annually beginning at age 10 years. If adenomas are identified, colectomy must be undertaken to prevent the development of cancer, although the exact timing of this procedure remains a topic of debate. In contrast, if results of genetic testing are negative, the patient can have colonoscopy postponed until 25 years of age.

Annual alpha-fetoprotein screening in children who have the FAP gene is recommended by some experts to screen for hepatoblastoma. However, this is not necessary if the patient does not carry the FAP gene. Similarly, ultrasonography, colonoscopy, and fecal occult blood testing are unnecessary at this time if the patient is not a gene carrier.

The indication for genetic testing is clear in this vignette because the father had colectomy findings consistent with FAP. However, many children who have polyposis syndromes present with rectal bleeding, but do not have a clear family history. The approach to evaluating a child in whom polyposis syndrome is suspected includes combining genotyping (as determined by genetic analysis) with phenotyping (as determined by upper endoscopy, colonoscopy, radiography, and video capsule endoscopy).

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Critique: 50

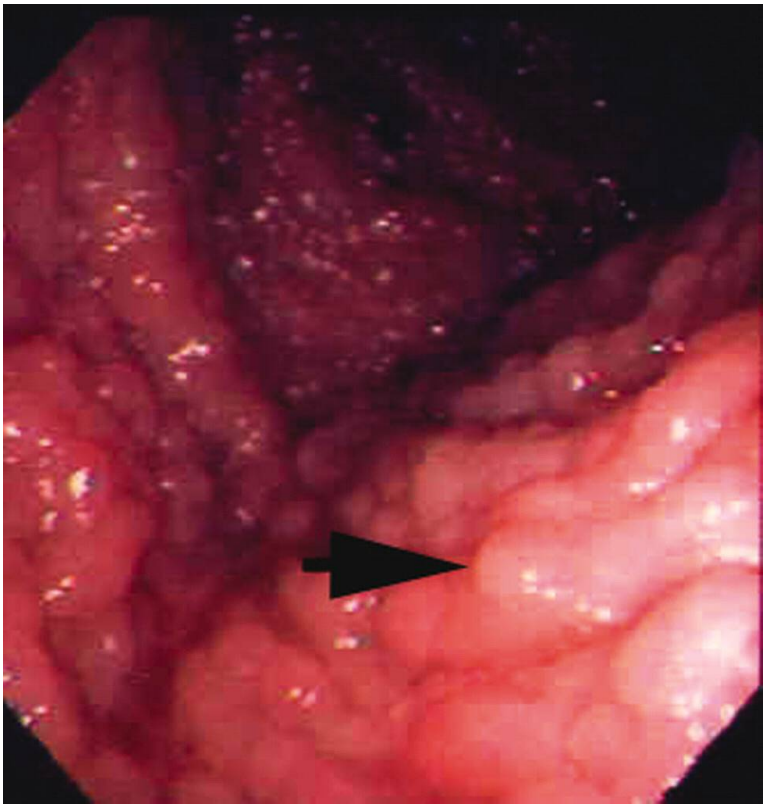


Image during colonoscopy of a patient who has familial adenomatous polyposis. Each of the nodules (arrow) represents an adenoma.

Courtesy of A. Bousvaros

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Critique: 50



Patients who have Peutz-Jegher syndrome have lentigines on the lips, oral mucosae, and elsewhere.

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Critique: 50

Item 50C: Some Familial Polyposis Syndromes Seen in Childhood

Syndrome	Inheritance	Mutated Gene	Type of Polyp	Associated Clinical Findings
Familial adenomatous polyposis	Autosomal dominant	FAP gene on chromosome 5	Multiple (usually between 100 and 1,000) small adenomas throughout the colon; small adenomas sometimes seen in periampullary region of the duodenum	• Congenital hypertrophy of the retinal pigment epithelium • Hepatoblastoma in infancy • Soft-tissue tumors • Mandibular osteomas • 100% risk of colon cancer in adulthood if colon not resected
Juvenile polyposis coli	Autosomal dominant	*PTEN* gene on chromosome 10 OR *SMAD4/DPC* gene on chromosome 18	Multiple (usually <50 at presentation) hamartomas in colon, small intestine, and sometimes stomach	
Peutz-Jegher syndrome	Autosomal dominant	*LBK1/STK11* gene on chromosome 19	Multiple hamartomatous polyps with branching smooth muscle in stomach, small intestine, and colon	• Blue freckling of the lips (Item 50B) and hands • Polyps cause intussusception • Increased risk of small bowel, colon and extraintestinal cancers
Bannayan-Riley-Ruvalcaba syndrome	Autosomal dominant	*PTEN* gene	Hamartomas	• Macrocephaly • Developmental delay • Penile macules
Cowden syndrome	Autosomal dominant	*PTEN* gene	Hamartomas	• Macrocephaly • Skin lesions (tricholemmomas) • Breast and thyroid cancer risk

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Question: 51

You are conducting prenatal counseling with a 30-year-old mother of two healthy children. She is 34 weeks pregnant and asks you about potential newborn problems for her soon-to-be delivered infant, reminding you that her last child was treated for early-onset group B streptococcal (GBS) infection.

Of the following, your MOST likely reply is that

A. antibiotic treatment during pregnancy will eradicate maternal GBS

B. both the mother and her newborn will require treatment

C. intrapartum chemoprophylaxis is indicated

D. previous GBS infection has no bearing on this pregnancy

E. urinary tract infection with GBS is uncommon

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Critique: 51 Preferred Response: C

Group B streptococcal (GBS) infection remains a concern for newborns around the world because from 10% to 30% of women of childbearing age are asymptomatic carriers of GBS. In the last decade, the incidence of early-onset GBS infection has been reduced with the use of antepartum screening strategies and intrapartum antibiotic prophylaxis (IAP). In addition to screening positive for GBS at 35 to 37 weeks' gestation, significant historical features in risk ascertainment include the occurrence of a GBS urinary tract infection during the present pregnancy or the birth of a previous infant who experienced early-onset GBS infection, as described for the mother in the vignette. Penicillin remains the drug of choice for IAP. Two or more doses should be administered intravenously (IV) at least 4 hours apart prior to delivering an appropriately identified at-risk woman to attain optimal penetration into the amniotic fluid and delivery to the fetus.

Although a 10- to 14-day course of ampicillin may reduce positive maternal GBS culture results, it does not eradicate the organism, and cultures return positive within days after ceasing antibiotics. When used appropriately, maternal IAP with two doses of IV penicillin at least 4 hours prior to delivery reduces the incidence of early-onset GBS infection, and treating the infant is not necessary. Urinary tract infection with GBS is not uncommon in GBS-positive women and may be associated with preterm labor.

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Question: 52

You are seeing 16-year-old twin brothers for health supervision visits. They tell you that they plan to spend most of the summer boating and fishing at their camp on the lake.

Of the following, the advice that is MOST likely to decrease their risk of a boating-related fatality is to

A. conduct regularly scheduled engine maintenance

B. have both boys take swimming lessons before the summer

C. install a carbon monoxide detector on the boat

D. post the phone number to the United States Coast Guard on the boat

E. wear life jackets at all times while on the boat

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Critique: 52 Preferred Response: E

Boating and the use of personal watercraft are popular recreational activities in the United States that are associated with an increased risk of death or injury. In 2004, 3,363 injuries and 676 deaths involving boating incidents were reported. The most common cause of death is drowning. Other deaths and injuries include carbon monoxide poisoning, hypothermia, vaginal lacerations (with personal watercraft), and other trauma, such as head injuries and fractures. In the 2004 report, alcohol was a factor in one third of deaths, and 90% of drowning victims were not wearing life jackets. Therefore, the best advice for people planning to use boats and personal watercraft is always to wear a life jacket or personal flotation device while on the boat or watercraft. Alcohol use also should be avoided while operating or riding in boats and watercraft.

Boating education courses are recommended to improve operator experience and reduce carelessness. Boaters can enroll their boats in the United States Coast Guard's Vessel Safety Check program, which checks safety equipment and gives information on safety procedures. Carbon monoxide poisoning can be prevented by installing detectors and ensuring sufficient ventilation on the boat. People who swim or stand near the swimming platform of houseboats are at particular risk for carbon monoxide poisoning.

Although conducting regularly scheduled maintenance visits, having passengers take swimming lessons, and posting the Coast Guard number on the boat are good ideas for boat safety, none of these actions prevents boating fatalities more than having all passengers wear life jackets while on board.

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Question: 53

A mother brings in her 18-month-old child in February with a “cold that won’t go away.” According to your notes in his medical record, you saw him 10 days ago because of the acute onset of fever to 103°F (39.5°C) with cough, coryza, listlessness, and decreased appetite. Rapid influenza testing at that office visit was positive for influenza A. The mother reports that the toddler seemed to improve for about 3 days, but physical examination today reveals purulent rhinorrhea, temperature of 102°F (39°C), and a faint rash (Item Q53) on his trunk. The patient’s 7-year-old brother recently complained of a sore throat and had fever; he improved without treatment after 3 days.

Of the following, the organism MOST likely to be responsible for this toddler’s illness is

A. *Bordetella pertussis*

B. group A beta-hemolytic *Streptococcus*

C. *Haemophilus influenzae* type b

D. influenza virus type A

E. respiratory syncytial virus

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Question: 53



Eruption composed of fine erythematous papules, as described for the child in the vignette.

Courtesy of D. Krowchuk

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Critique: 53 Preferred Response: B

The boy described in the vignette most likely has group A beta-hemolytic streptococcal (GABHS) infection. This infection requires a susceptible host and is facilitated by close contact. Close contact with a large number of individuals who potentially are infected with GABHS, such as in child care and elementary school, results in more frequent spread of illness. Classic signs of pharyngeal infection are uncommon in those younger than 3 years of age, possibly due to decreased attachment of GABHS to nasopharyngeal and oropharyngeal epithelial cells. Also, a preverbal child may be unable to communicate dysphagia, a prominent symptom of the infection. When infection occurs in toddlers, it most often involves the nasopharynx or skin (impetigo).

Toddlers (1 to 3 years of age) who have GABHS respiratory tract infection often present initially with serous rhinitis (Item C53) (as in any upper respiratory tract infection) and subsequently develop a protracted illness characterized by moderate fever, irritability, and anorexia (streptococcal fever), as reported for the boy in the vignette. Other symptoms and signs include purulent nasal discharge, impetiginous superinfection of the nares, and occasionally a scarlet fever rash. Differentiation of this condition from sinusitis can be difficult if these other hallmarks of streptococcal disease are not present.

Although pertussis and respiratory syncytial virus infection may present initially with coryza, the cough becomes prominent after the catarrhal phase**,** which typically lasts 3 to 5 days. Influenza usually presents with high fever, coryza, and cough, as initially experienced by the toddler in the vignette. However, influenza resolves within 5 to 7 days. *Haemophilus influenzae* type b disease is rare because of universal vaccination, and it usually causes invasive disease such as pneumonia or meningitis.

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Critique: 53



Protracted rhinitis is the most common clinical presentation of group A beta-hemolytic streptococcal infection in toddlers.

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Question: 54

You care for a 6-month-old boy who was born with pulmonary atresia and ventricular septal defect. He received a modified Blalock-Taussig (systemic-to-pulmonary artery) shunt 5 days after birth. His oxygen saturations have ranged between 70% and 84% at office visits over the past 2 months. During a health supervision visit, you record a hematocrit of 57% (0.57).

Of the following, this child's polycythemia puts him at INCREASED risk for

A. acute leukemia

B. bacteremia

C. cerebrovascular accident

D. congestive heart failure

E. necrotizing enterocolitis

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Critique: 54 Preferred Response: C

Cyanosis is the term given to the observation of a blue, maroon, or purple discoloration of the skin. A number of clinical conditions can lead to this finding, and cyanotic congenital heart disease is an important cause in the pediatric age group. Heart diseases that limit effective pulmonary blood flow or allow for shunting of desaturated blood from the right to left side result in desaturation and cyanosis. Until the underlying condition is corrected or palliated, the desaturation of arterial blood can cause increased erythropoietin secretion from specialized cells in the kidney, resulting in increased erythrocyte production and polycythemia.

Polycythemia leads to an increase in blood viscosity that has a direct relationship to vascular blood flow resistance. In addition, polycythemia increases oxygen-carrying capacity because of greater numbers of red blood cells (RBCs) and a greater concentration of hemoglobin. Because of the increased resistance to blood flow and oxygen-carrying capacity, blood flow at rest is reduced in those who have polycythemia.

Polycythemia is associated with significant clinical morbidity, particularly as the hematocrit increases to values well beyond normal. Complications may involve virtually any organ, but the infant, child, and adolescent typically present with headache, lethargy, irritability, joint pain, anorexia, and dyspnea. More serious complications can include thrombosis of the lungs, kidneys, or the brain. Thus, the balance of the benefit from the increased oxygen-carrying capacity and the risk from increased blood viscosity for the patient who has cyanotic heart disease is precarious. If the underlying congenital heart disease cannot be corrected, consideration should be given to partial volume exchange transfusion as the hematocrit exceeds 65% (0.65) or at lower values if signs and symptoms are present.

The patient described in the vignette is at risk for a cerebrovascular accident because he has developed polycythemia and its attendant hyperviscosity as a result of his pulmonary atresia with a ventricular septal defect (VSD). As he outgrows his Blalock-Taussig shunt, his oxygen saturations will decrease further because he will have decreased pulmonary blood flow and continued right-to-left shunting at the VSD. If not corrected, this can lead to more pronounced polycythemia and greater risk for its complications. Polycythemia does not increase the risk for acute leukemia, bacteremia, congestive heart failure, or necrotizing enterocolitis.

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Question: 55

During the health supervision visit of a 10-year-old boy, you note some wasting and weakness of his lower leg muscles, with diminished patellar and ankle reflexes. You examine his parents' legs and feet and notice that his mother has a bilateral foot drop and deformed feet (Item Q55).

Of the following, the MOST likely diagnosis is

A. Becker muscular dystrophy

B. celiac disease

C. chronic inflammatory demyelinating polyneuropathy

D. hereditary sensorimotor neuropathy

E. hydrocephalus

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Question: 55



Deformities of the feet (hammer toes), as exhibited by the mother of the boy described in the vignette.

Courtesy of D. Gilbert

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Critique: 55 Preferred Response: D

The child described in the vignette has physical findings suggestive of a chronic neuropathy. The distal weakness accompanied by reduced reflexes and loss of muscle bulk noted in the motor examination localize the problem to the peripheral nervous system. The presence of similar but more advanced symptoms in the mother (deformed foot with “hammer toes” — high-arched, flexed toes) supports a diagnosis of autosomal dominant hereditary sensorimotor neuropathy (eg, Charcot Marie Tooth disease). A large number of genes and loci have been identified for various forms of hereditary sensorimotor neuropathy. Referral to a neurologist, particularly one specializing in neuromuscular diseases, is recommended.

Neuropathy may occur as part of more than 400 heritable diseases affecting peripheral nerve axons, myelin, or both. The most common primary, heritable forms are the hereditary sensorimotor neuropathies. Diagnostic evaluation is based on history and clinical findings, associated neurologic and systemic signs and symptoms, and results of clinical neurophysiologic studies (electromyography/nerve conduction velocities) that may localize the problem more accurately. Neuropathy also can be acquired. Autoimmune diseases, heavy metals and other toxins, chemotherapeutic and other medications, vitamin deficiencies (B1, B6, B12, E), and endocrine disorders such as diabetes may damage peripheral nerves.

Becker muscular dystrophy results in muscle weakness, but it is not associated with reduced muscle bulk and hyporeflexia. Additionally, Becker muscular dystrophy is caused by mutations in the dystrophin gene, and the pattern of inheritance is X-linked recessive, so the mother would not be symptomatic.

Celiac disease can present with a variety of neuropathic problems, but both hands and feet are involved, pain in hands and feet often is prominent, and sensory ataxia might be present on physical examination. The prevalence in first-degree relatives is low.

Chronic inflammatory demyelinating polyneuropathy (CIDP) usually occurs as an adverse long-term outcome of Guillain-Barré syndrome, also known as acute inflammatory demyelinating polyneuropathy (AIDP). The absence of a prior history of AIDP and the apparent autosomal dominant inheritance through the mother in this case make CIDP unlikely.

Hydrocephalus can cause leg weakness due to compression of cortical spinal tract fibers to the legs, but the neurologic examination would document increased, not decreased reflexes.

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Question: 56

You are called to the nursery to evaluate a newborn who has respiratory distress. On observation, the baby appears pink while crying but becomes dusky when calm. You note a hypoplastic nose and depressed nasal bridge on physical examination. A review of her chest radiographs reveals stippling of the vertebrae.

Of the following, the MOST likely teratogen to cause these findings is

A. alcohol

B. hydantoin

C. retinoic acid

D. valproic acid

E. warfarin

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Critique: 56 Preferred Response: E

The hypoplastic nose, depressed nasal bridge, and stippling of the vertebrae described for the infant in the vignette are characteristic facial and skeletal features of warfarin embryopathy, also known as fetal warfarin syndrome and fetal coumarin syndrome. Coumarin derivatives, including warfarin, dicumarol, phenindione, acenocoumarol, diphenadione, phenprocoumon, and anisindione, are orally administered anticoagulants. Warfarin is the most frequently used of these agents and is less toxic than the others. Heparin, which is administered by injection, is considered to be safe for use during pregnancy.

The teratogenicity of coumarin is postulated to be due to its antagonism of vitamin K through the inhibition of vitamin K epoxide reductase, which results in abnormalities of cartilage and bone. The critical period of coumarin exposure in terms of facial abnormalities and bone stippling is between 6 and 9 weeks' gestation, and approximately one third of exposed fetuses are affected. Additional evidence suggests that prenatal exposure to coumarin after the first trimester increases fetal risk for central nervous system abnormalities, most likely due to hemorrhage.

Newborns who have warfarin embryopathy frequently have upper airway obstruction due to hypoplasia of the nasal passages, and it may be necessary to secure an airway. In severe cases, tracheostomy may be indicated. Even so, most affected children do well. The stippling is incorporated into calcifying cartilage and typically no longer is appreciated radiographically after 1 year of age.

The craniofacial features of fetal alcohol syndrome include midface hypoplasia with epicanthal folds and a flat nasal bridge. In addition, there are short palpebral fissures, a relatively smooth philtrum, and a narrow upper lip with a poorly defined "cupid's bow" (Item C56A). Affected infants do not typically have upper airway obstruction, and they do not have epiphyseal stippling.

Prenatal exposure to hydantoin results in an array of outcomes, ranging from normal to fetal hydantoin syndrome (FHS), also known as fetal Dilantin® syndrome. The characteristic craniofacial features of FHS are wide anterior fontanelle, prominent metopic ridge, broad and depressed nasal bridge, and cleft lip and palate (Item C56B). Affected children typically do not have upper airway obstruction or epiphyseal stippling.

Retinoic acid embryopathy (also called Accutane® embryopathy) occurs in approximately 35% of children born to women who take isotretinoin beyond 15 days after conception. Doses of 0.5 to 1.5 mg/kg of maternal body weight are believed to be teratogenic. Craniofacial features may include hypoplasia to absence of the external ear(s), depressed nasal bridge, ocular hypertelorism, and mottled dental enamel.

Fetal exposure to valproic acid is associated with a markedly increased risk for spina bifida, but it also may result in fetal valproate syndrome (FVS). Craniofacial features of FVS include narrow bifrontal diameter, tall forehead, midface hypoplasia, a relatively small mouth, and a small chin.

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Critique: 56



Short palpebral fissures, midface hypoplasia, and a relatively narrow upper lip are characteristic of fetal alcohol syndrome.

Courtesy of M. Rimsza

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Critique: 56



Arched eyebrows, wide-spaced eyes, and a short upturned nose are characteristic of infants who have been exposed to phenytoin during pregnancy.

Courtesy of M. Rimsza

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Question: 57

A 15-year-old boy comes to your office for a sports physical examination. Compared with when you saw him at his 10-year-old health supervision visit, he has progressed from Sexual Maturity Rating 1 to 4.

Of the following, the laboratory value that is MOST likely to have changed is

A. calcium

B. hemoglobin

C. platelet count

D. serum albumin

E. white blood cell count

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Critique: 57 Preferred Response: B

Biochemical changes that occur during puberty can reflect underlying physical growth. For example, hemoglobin and hematocrit values vary with pubertal stage and sex. Female adolescents have relatively stable values as they progress from Sexual Maturity Rating (SMR) 1 through 5. In contrast, male adolescents show a progressive increase in hemoglobin value as they progress to physical maturity at SMR 5, reaching a hemoglobin value of 14 to 18 g/dL (140 to 180 g/L). The increase in hemoglobin for males is due to the hematopoietic effects of testosterone. Values for serum albumin, calcium, platelet count, and white blood cell count remain stable across adolescent growth and development.

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Question: 58

A mother brings in her 1-year-old boy for the first time because she is concerned about his "bowed legs" (Item Q58A). The mother is 4 ft 10 in tall and says she needed to have surgery to straighten out her bowed legs when she was an adolescent, as did one of her brothers. Radiographs of the boy's long bones are obtained (Item Q58B).

Of the following, the MOST likely serum laboratory findings are

A. low calcium and high phosphorus

B. low calcium and normal phosphorus

C. low calcium and low phosphorus

D. normal calcium and high phosphorus

E. normal calcium and low phosphorus

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Question: 58

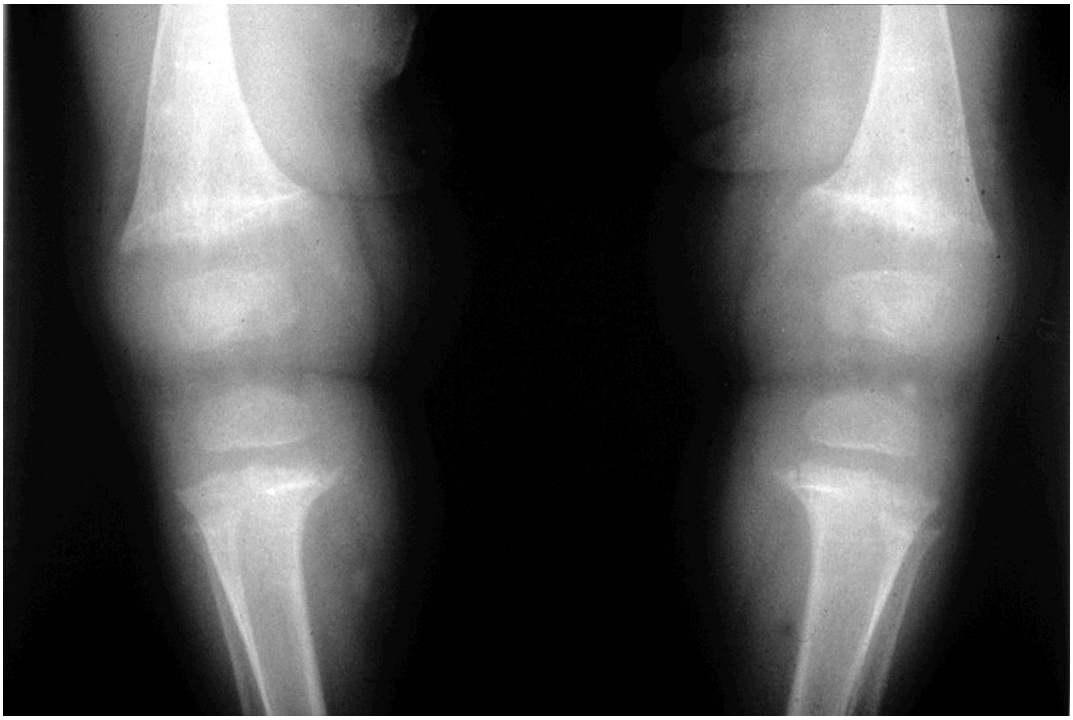


Bowing of the legs, as exhibited by the boy described in the vignette.

Courtesy of M. Rimsza

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Question: 58



Courtesy of R. Schwartz

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Critique: 58 Preferred Response: E

The child described in the vignette has clinical and radiologic evidence of rickets (Item C58) as well as a history compatible with a familial disorder. Accordingly, he most likely has familial hypophosphatemic rickets of either the autosomal dominant or sex-linked type. Mothers of boys who have the sex-linked disorder also have rachitic changes, although they might not be as severe as the changes in their sons. The typical laboratory findings in this disorder are normal serum calcium and low serum phosphate values. Affected children are said to have "phosphate wasting" because their renal tubular excretion of phosphate is very high. Sex-linked familial hypophosphatemic rickets is due to a mutation in the *PHEX* gene, which encodes a metalloprotease that must be important in the conversion of a prohormone that prevents phosphate wasting. Autosomal dominant familial hypophosphatemic rickets is due to a mutation in the fibroblast growth factor 23 gene.

Low calcium and phosphorus concentrations are associated with severe vitamin D deficiency rickets. Low calcium and high phosphorus values are the laboratory hallmark of phosphate overload, hypoparathyroidism, or pseudohypoparathyroidism. Low calcium and normal phosphorus concentrations may be found in the initial vitamin D repletion stage of healing vitamin D deficiency rickets. Normal calcium and high phosphorus values could be seen in conditions such as renal disease and growth hormone excess or in those ingesting a high-phosphorus diet. Because phosphorus concentrations in children are higher than those in adults, occasionally laboratories report "high phosphorus" values in normal children.

References:

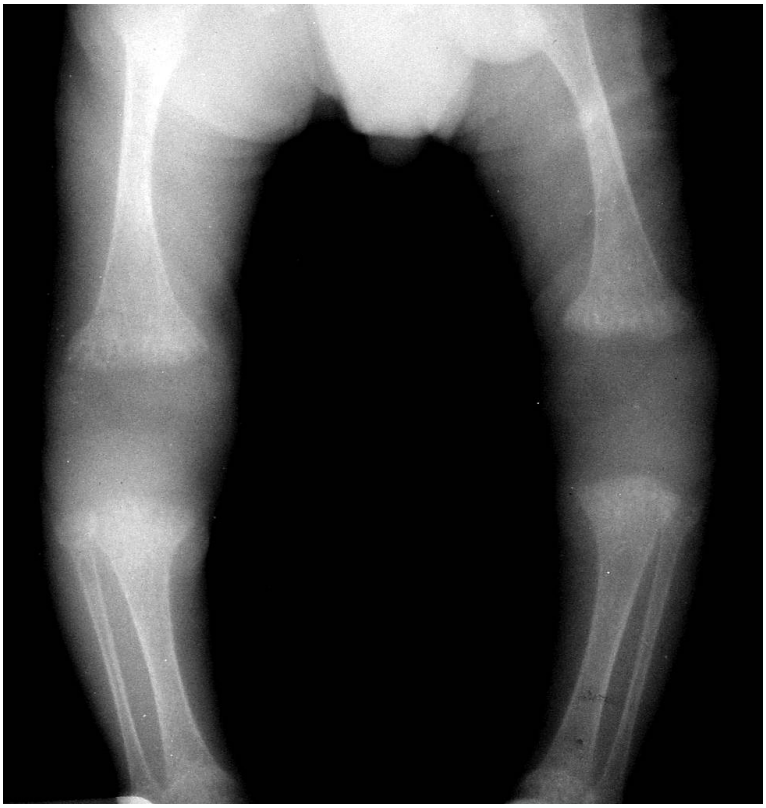
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Critique: 58



Anteroposterior radiograph of the lower extremities in rickets shows poor mineralization and widening and fraying of the metaphyses that is most marked at the knees.

Courtesy of D. Mulvihill

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Question: 59

A 12-year-old girl is receiving learning support because of difficulty with reading and language arts. She struggles to do well in the classroom but realizes she can never compete successfully with her older sister academically. She is well-coordinated and enjoys playing basketball in her yard. Her parents are concerned by her negative comments about herself and ask your advice on how they can help her improve her self-image.

Of the following, your BEST recommendation is to

A. evaluate the girl for attention-deficit/hyperactivity disorder

B. recommend extracurricular activities for the girl

C. repeat the psychoeducational evaluation

D. suggest that her older sister help with her homework

E. suggest the parents hire an educational advocate

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Critique: 59 Preferred Response: B

The term “learning disabilities” refers to a heterogeneous group of disorders manifested by significant difficulties in the acquisition and use of listening, speaking, reading, writing, reasoning, or mathematical abilities. Such disorders are intrinsic to the individual and presumed to be due to central nervous system dysfunction. Most definitions of learning disability include a discrepancy between ability as measured on an intelligence test and actual achievement in academic skills. The prevalence is estimated to be 3% to 5% in the school-age population.

A vital aspect of the appropriate management of learning disabilities is maintenance of the child’s self-esteem, which is affected adversely by repeated failure. Therefore, it is important to work closely with parents so they understand that the failure is not the child’s fault or purposeful.

Because the child described in the vignette displays feelings of poor self-worth, it is important for her to engage in activities that give her a sense of accomplishment, such as extracurricular activities. Having her sister help her with her homework may increase sibling rivalry. Although additional educational testing may provide further information about the child’s learning needs, it does not address her parents’ concerns regarding the girl’s insecurity and poor self-esteem. There is no indication in the vignette that the child is having problems paying attention or that her educational needs are not being met. Therefore an evaluation for attention-deficit/hyperactivity disorder or hiring an educational advocate are not indicated.

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Question: 60

A 12-year-old boy who has a history of Hodgkin disease presents to the emergency department with fever, headache, and malaise of 4 hours' duration. He has not received any cancer treatment in more than 1 year. His immunizations are up to date, including the 23-valent pneumococcal, *Haemophilus influenzae* type b, and polysaccharide meningococcal vaccines. He has had a splenectomy. He currently takes oral penicillin twice daily. Physical examination reveals a toxic-appearing boy who has a temperature of 104°F (40°C), heart rate of 110 beats/min, respiratory rate of 30 breaths/min, and a blood pressure of 118/76 mm Hg. No other findings on the physical examination suggest the source of his fever.

Of the following, the MOST likely cause of his fever is infection with

A. *Haemophilus influenzae* type b

B. *Pseudomonas aeruginosa*

C. *Staphylococcus aureus*

D. *Streptococcus pneumoniae*

E. *Streptococcus pyogenes*

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Critique: 60 Preferred Response: D

Patients who have Hodgkin disease and have undergone splenectomy, such as the boy described in the vignette, are at increased risk for sudden overwhelming infection. Encapsulated bacteria such as *Streptococcus pneumoniae*, *Haemophilus influenzae* type b, and *Neisseria meningitidis* account for approximately 80% of these infections, with *S pneumoniae* being the most common cause. Despite the use of the polysaccharide vaccine and penicillin prophylaxis, patients remain at risk. Splenectomized patients mount very poor polysaccharide antibody responses when vaccinated with the 23-valent polysaccharide pneumococcal vaccine, and the conjugate pneumococcal vaccine protects only against seven serotypes. Similarly, although penicillin prophylaxis has been demonstrated to reduce the risk of pneumococcal sepsis in patients who have hemoglobin SS, the data concerning other forms of asplenia are not clear. These factors, in combination with increased levels of penicillin resistance among pneumococcal isolates, must make clinicians think of *S pneumoniae* as the most probable agent causing sepsis in splenectomized patients. *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *S pyogenes* can cause bacterial sepsis, but these organisms are not encapsulated and are much less likely than *S pneumoniae* to be pathogens in this clinical situation. When surgical splenectomy is indicated, immunization against encapsulated organisms is recommended prior to surgery.

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Question: 61

You are evaluating a 6-year-old boy who has a 2-day history of fever (102ºF [38.9°C]), headache, vomiting, moderate crampy periumbilical pain, and watery diarrheal stools (six to eight a day) that have become blood-streaked in the last 12 hours. He just returned yesterday from a 3-week visit to his grandparents' farm in India. Physical examination reveals a tired boy who has a temperature of 102.5ºF (39.2°C), tacky mucous membranes, and dry lips. He has diffuse abdominal tenderness to palpation that is most pronounced in the periumbilical area and associated with some voluntary guarding but no rebound. Laboratory tests show a peripheral white blood cell count of 9.0 x103/mcL (9.0x109/L) with 60% polymorphonuclear leukocytes, 5% band forms, 30% lymphocytes, and 5% monocytes. A test for the presence of fecal leukocytes is positive. You suspect *Campylobacter* sp as the cause of this patient's condition.

Of the following, the laboratory condition that is REQUIRED to isolate this organism is

A. aerobic culture conditions

B. antibiotic-free culture media

C. blood agar media

D. incubation temperature of 35ºC

E. incubation temperature of 42ºC

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Critique: 61 Preferred Response: E

The microbiology laboratory needs to be notified of the clinician's suspicion of certain organisms because of specific criteria for the isolation of such pathogens. For example, certain organisms require specific growth requirements, such as incubation at 42°C and microaerophilic conditions for *Campylobacter* sp. In some cases, specific media may need to be inoculated, such as modified Thayer-Martin for gonococci, Bordet-Gengou or Regan-Lowe for *Bordetella pertussis*, or Loeffler or Tinsdale agar for *Corynebacterium diphtheriae*. The laboratory also needs to know if the culture must be held for a long period of time in the case of slow-growing organisms, such *Brucella* sp and *Mycobacterium tuberculosis,* and if special tests specific for a particular organism need to be performed.

The patient described in the vignette is suspected of having an infection caused by *Campylobacter* sp. Isolation of these organisms requires a microaerophilic environment, selective media containing antibiotics that reduce the growth of competing microorganisms, and incubation at 42°C. Aerobic culture conditions, antibiotic-free culture media, and blood agar media are not required.

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Question: 62

A 4-month-old girl presents with fever. Results of urinalysis include 50 to 100 white blood cells per high-power field and 3+ bacteria. Urine culture is positive for *Escherichia coli*. Ultrasonography reveals hydroureteral nephrosis of the left upper pole, and voiding cystourethrography shows a filling defect within the bladder (Item Q62).

Of the following, in addition to a urinary tract infection, the infant is MOST likely to have

A. bladder diverticulum

B. posterior urethral valves

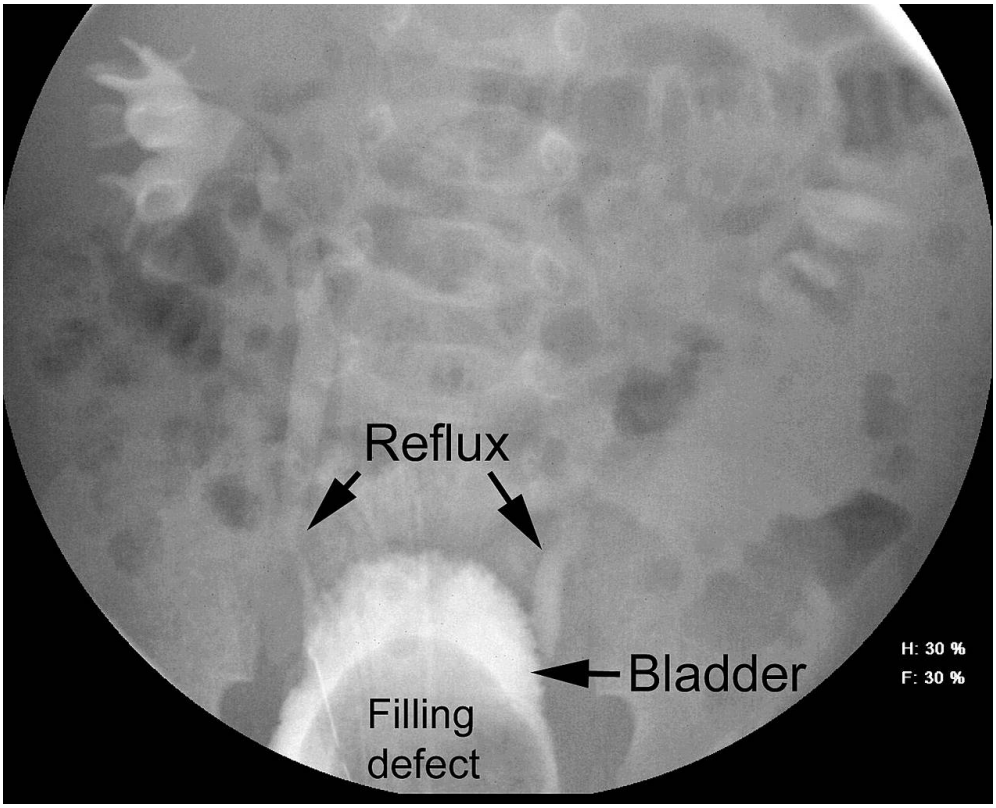
C. ureteral stone

D. ureteropelvic junction obstruction

E. ureterocele

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Question: 62



Fluoroscopic image from a voiding cystourethrogram showing a defect within the contrast-filled bladder, as described for the patient in the vignette. There is reflux into both lower pole collecting systems of duplex kidneys.

Courtesy of D. Mulvihill

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Critique: 62 Preferred Response: E

A ureterocele is a cystic dilation of the ureter where it inserts into the bladder. An intravesical ureterocele is contained entirely within the bladder. When a portion of the defect extends beyond the bladder (to the urethra or bladder neck), an extravesical ureterocele is present. Typically, the pelvocaliceal system draining into the ureterocele is obstructed. The incidence of ureteroceles in children is estimated between 1 in 5,000 and 1 in 10,000. Ureteroceles are associated with a duplex collecting system in 80% of children; the remainder are associated with a single collecting system. Ureteroceles are four times more common in females than males and occur almost exclusively in Caucasians.

A ureterocele is associated most commonly with a complete duplication of the renal collecting system (more common in the left kidney) where the involved ureter is linked to that draining the upper pole moiety. This lesion, which extends beyond the ureterovesical junction, results in ureteral obstruction, with hydroureteral nephrosis of the involved renal unit, usually the upper pole, as described for the infant in the vignette. A ureterocele often results in a mass lesion within the bladder that may be seen on bladder ultrasonography or indirectly as a filling defect of the bladder on voiding cystourethrography (Item C62A). The lower pole of the kidney of a duplex collecting system may drain into an orthotopic site and is associated with vesicoureteral reflux (VUR) in approximately 50% of cases. In addition, VUR is seen in approximately 25% of the kidneys contralateral to the duplex kidney that has a ureterocele.

Ureteroceles most commonly are associated with urinary tract infection in infants, although 25% are detected antenatally. Older children present with voiding symptoms or hematuria associated with minimal trauma. Surgical treatment is aimed at relieving the obstruction, preserving the functioning nephron mass, removing nonviable tissue that may result in infection, and treating VUR. Based on patient symptoms, treatments include upper pole nephroureterectomy, endoscopic incision of the ureterocele for relief of obstruction, and clinical observation.

Ureteropelvic junction obstruction (Item C62B), ureteral stones (Item C62C), and posterior urethral valves (Item C62D) cause hydronephrosis that involves the entire kidney, not just a portion, as is seen with a ureterocele. In addition, in posterior urethral valves, the hydronephrosis is bilateral. A bladder diverticulum (Item C62E) results in an outpouching of the bladder wall without a filling defect in the bladder.

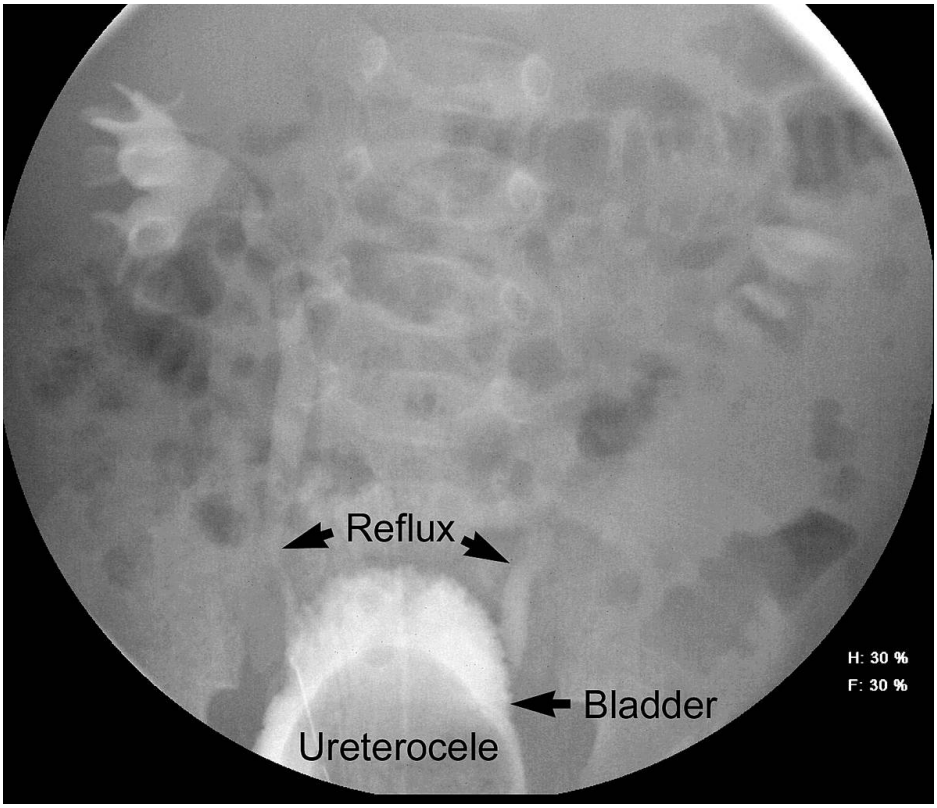
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Critique: 62

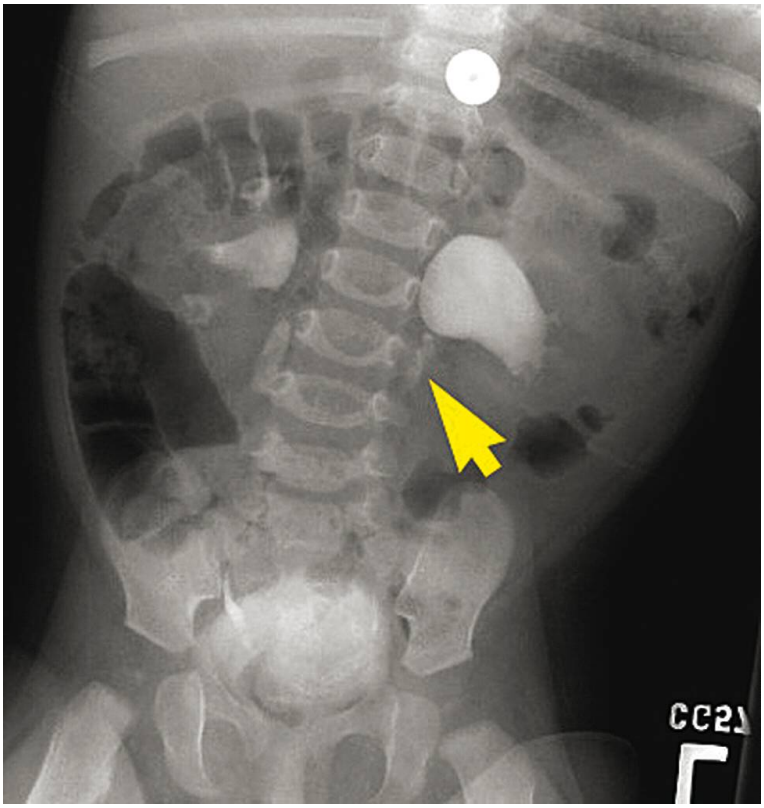


Fluoroscopic image from a voiding cystourethrogram showing a large mass within the contrast-filled bladder consistent with an ureterocele. There is reflux into both lower pole collecting systems of duplex kidneys.

Courtesy of D. Mulvihill

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Critique: 62

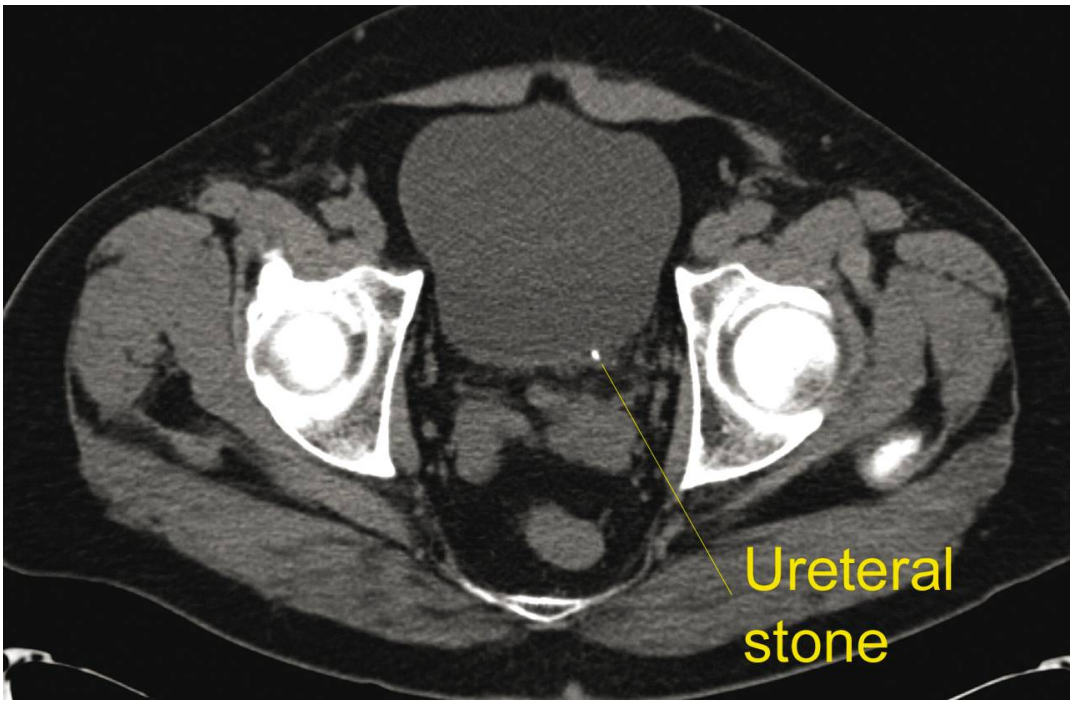


Intravenous pyelography of ureteropelvic junction obstruction shows a distended left renal pelvis with an abrupt change in caliber to a normal-sized ureter (arrow).

Courtesy of D. Mulvihill

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Critique: 62

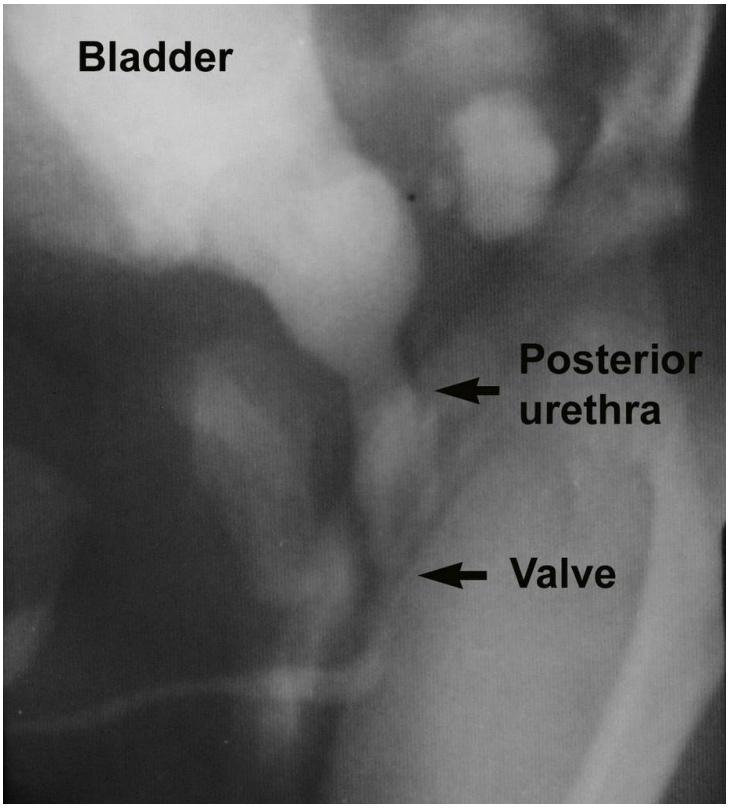


Noncontrast axial computed tomography (CT) scan of the pelvis shows a density at the ureterovesical junction consistent with a ureteral stone. CT is the preferred imaging method for the patient suspected of having a genitourinary stone.

Courtesy of D. Mulvihill

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Critique: 62

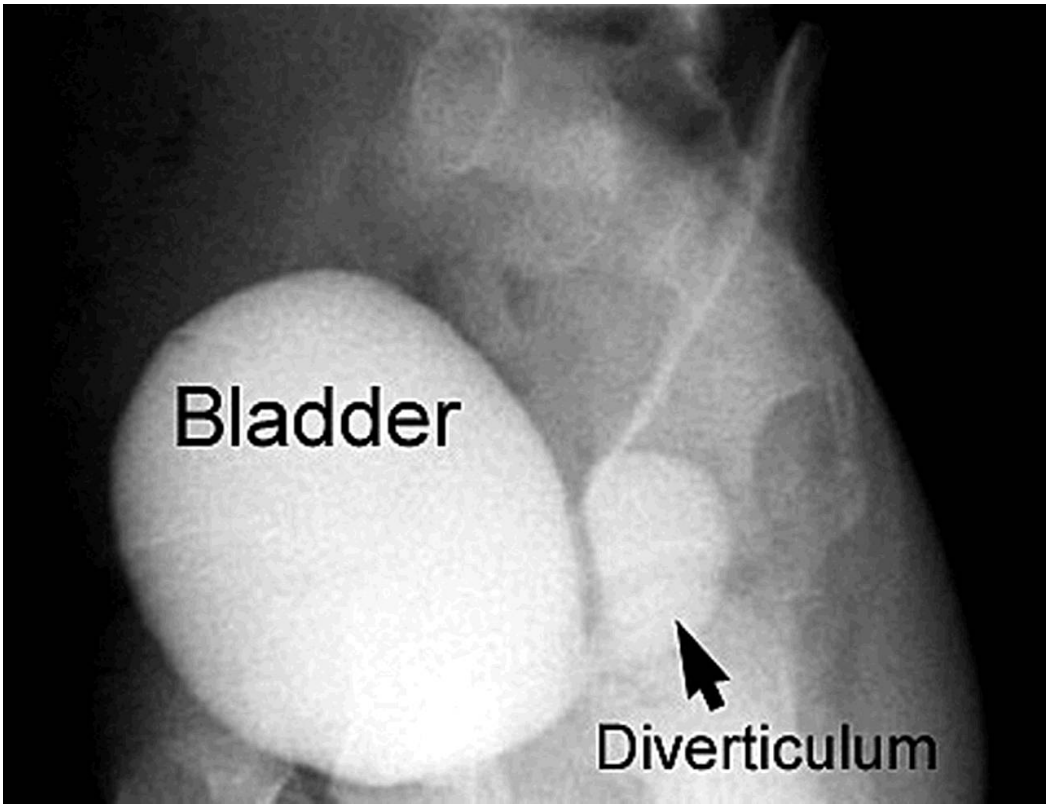


A fluoroscopic spot film from a voiding cystourethrogram shows a significant change in the caliber of the urethra at the level of the posterior urethral folds that is consistent with valves.

Courtesy of D. Mulvihill

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Critique: 62



Voiding cystourethrogram showing a filled bladder and a large left-sided diverticulum.

Courtesy of D. Mulvihill

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Question: 63

A 16-year-old girl who has moderate persistent asthma presents to the emergency department with coughing, wheezing, and increasing dyspnea. She states that she was feeling fine until she was exposed to cologne that one of her classmates was wearing. An ambulance was called after her symptoms did not improve following administration of two puffs of her beta-2 agonist inhaler. On physical examination, the teenager has a respiratory rate of 30 breaths/min, heart rate of 90 beats/min, and pulse oximetry of 98% on room air. She has difficulty completing a sentence and points to her neck, saying it is “hard to get air in.” Her lungs are clear to auscultation, and rhinolaryngoscopy demonstrates adduction of one of the vocal cords during inspiration with a posterior glottic “chink.” Pulmonary function testing shows a blunted inspiratory loop (Item Q63).

Of the following, the MOST likely cause for this patient’s symptoms is

A. asthma exacerbation

B. subglottic stenosis

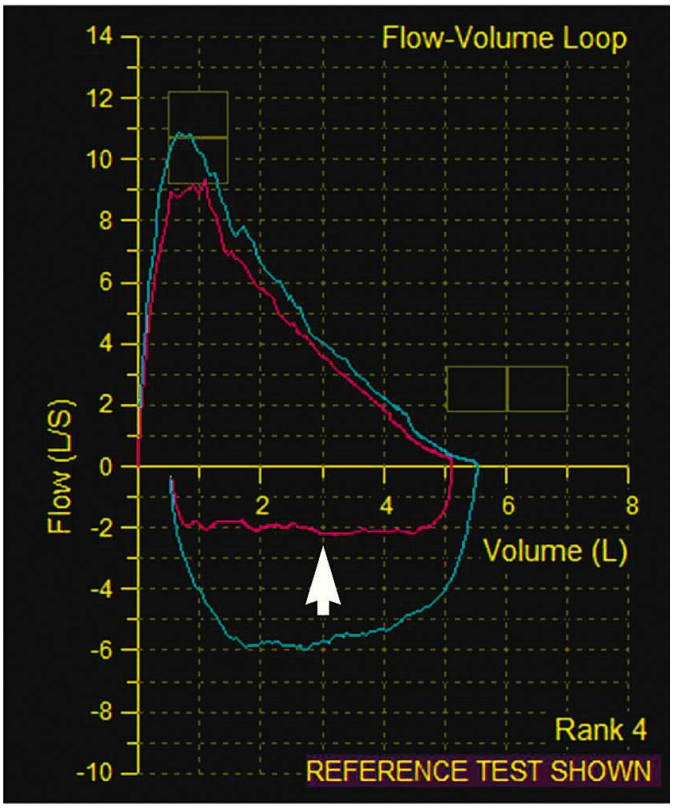
C. vocal cord dysfunction

D. vocal cord nodule

E. vocal cord paralysis

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Question: 63



Blunted inspiratory flow curve in pink (arrow), as exhibited by the patient in the vignette.

Courtesy of K. Waibel

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Critique: 63 Preferred Response: C

The teenager described in the vignette has signs and symptoms consistent with vocal cord dysfunction (VCD), a condition that can both mimic and coexist with asthma. In contrast to an asthma exacerbation, the key features that are consistent with VCD in this girl are normal room air pulse oximetry readings, failure to improve with the beta-2 agonist inhaler, clear lungs on examination, difficulty with inspiration instead of expiration, and a normal expiratory flow loop without signs of obstruction (Item C63A). The hallmark finding on rhinolaryngoscopy is paradoxic adduction of the vocal cords during inspiration. Endoscopy is the diagnostic tool of choice for laryngeal and vocal cord disorders.

Subglottic stenosis and vocal cord paralysis usually present with stridor and primarily affect infants and younger children. Subglottic stenosis can result from direct laryngotracheal trauma, intubation, or infection (eg, tuberculosis, diphtheria). Flexible or rigid laryngoscopy can be used to assess the degree of subglottic stenosis (Item C63B). Vocal cord paralysis (Item C63C) can occur in the context of congenital central nervous system lesions, surgical correction of congenital cardiac anomalies or tracheoesophageal fistula, or inflammatory processes of the larynx, or it may be idiopathic. Direct visualization of the vocal cords demonstrates the absence of vocal cord motion during inspiration and expiration.

Vocal cord nodules, also called singer's nodules (Item C63D), usually occur after repetitive vocal misuse or abuse (eg, excessive loudness, inappropriate pitch, excessive coughing, or throat clearing). On flexible laryngoscopy, nodules appear as superficial growths on the medial surface of the true vocal folds.

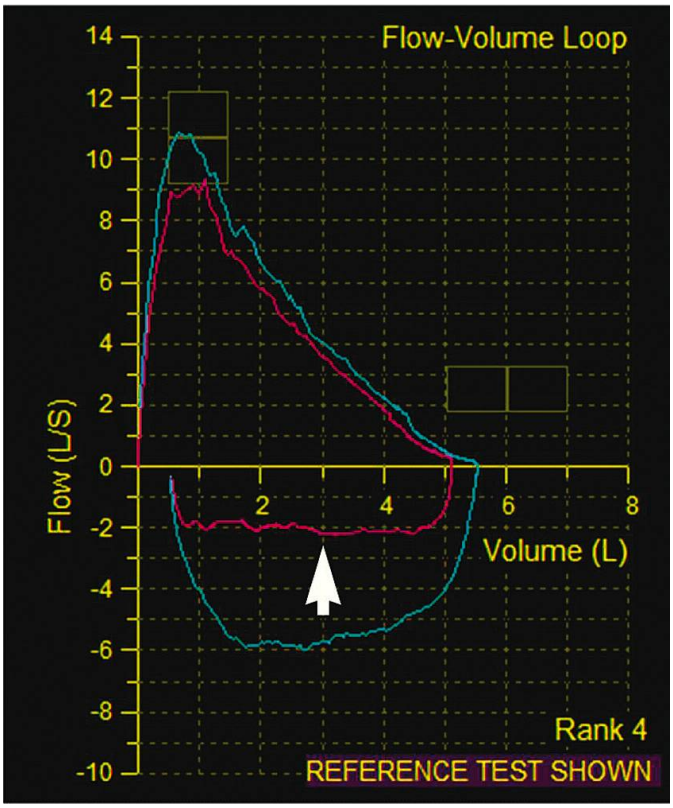
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Critique: 63

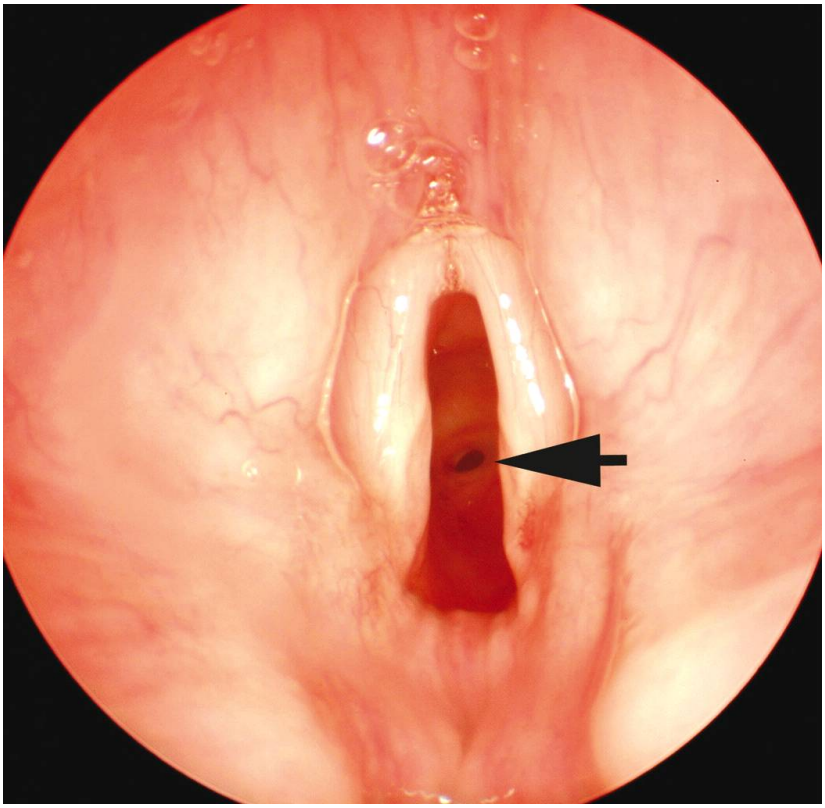


Expiratory (top) and inspiratory (bottom) flow loops demonstrate a normal flow-volume curve (blue) and a curve consistent with vocal cord dysfunction (pink). Symptomatic patients who have vocal cord dysfunction exhibit the characteristic blunted inspiratory loop (arrow) caused by paradoxic adduction of the vocal cords during inspiration.

Courtesy of K. Waibel

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Critique: 63

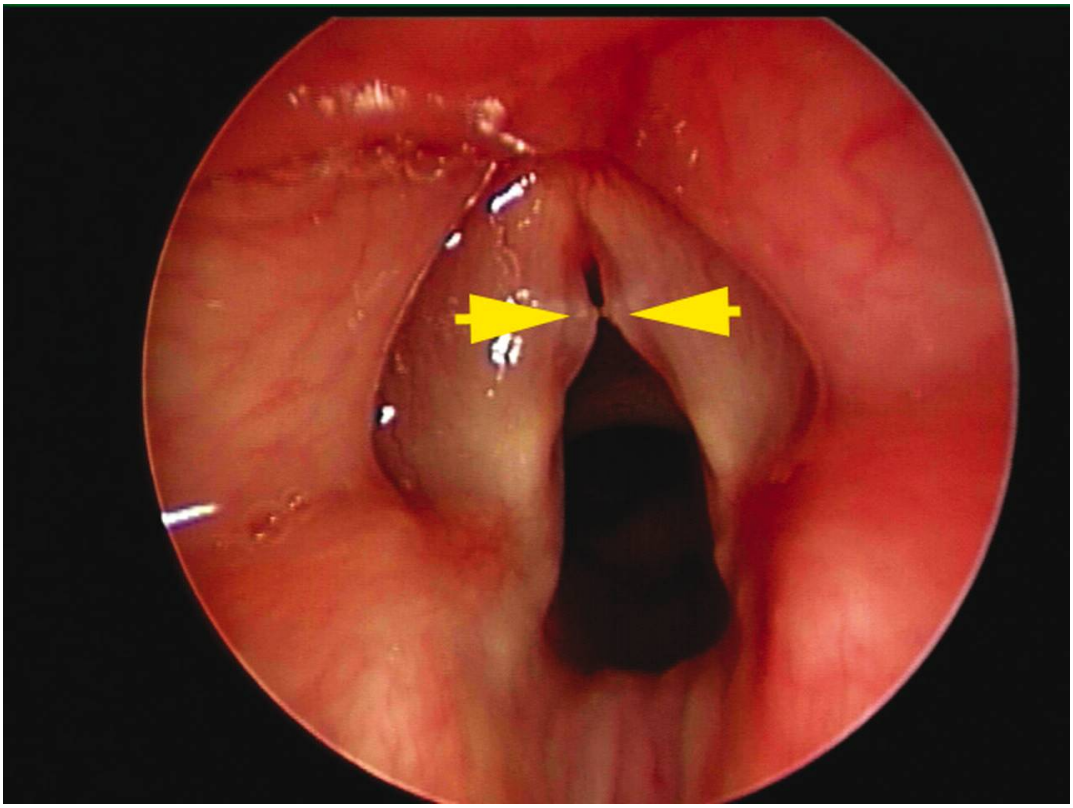


Acquired subglottic stenosis. Note the circumferential narrowing of the subglottic area (arrow).

Courtesy of D. Kirse

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Critique: 63



Vocal cord nodules appear as growths located anteriorly on the vocal cords.

Courtesy of D. Kirse

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Question: 64

A 1-year-old year boy who has chronic kidney disease from posterior urethral valves presents to your office because his breathing has been noisy for the past 2 hours. His usual medications include calcium carbonate and vitamin D. On physical examination, you note inspiratory stridor. He has not had upper respiratory tract symptoms or fever, and there is no history of choking. The mother reports that the boy was seen by the nephrologist 1 week ago, and because of worsening renal function, he was begun on sodium bicarbonate.

Of the following, the electrolyte abnormality that BEST explains his current symptoms is

A. hyperkalemia

B. hypermagnesemia

C. hypocalcemia

D. hyponatremia

E. hypophosphatemia

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Critique: 64 Preferred Response: C

Serum calcium and 1,25 dihydroxyvitamin D concentrations are decreased in children who have chronic kidney disease because of phosphate retention and impaired hydroxylation of 25-hydroxyvitamin D. Physiologic serum calcium concentrations are maintained largely through the use of vitamin D supplements and phosphate binders. As renal function worsens, hypocalcemia can result if vitamin D deficiency or hyperphosphatemia is not addressed or if the use of bicarbonate therapy to treat metabolic acidosis leads to metabolic alkalemia. Metabolic alkalemia increases the binding of calcium to albumin, which decreases the ionized calcium concentration. Decreased ionized calcium can lead to perioral or extremity paresthesias, carpopedal spasm, laryngospasm, and seizures. Children who have hypocalcemia from any cause may be diagnosed mistakenly with croup if they present with tetany-related laryngospasm.

Hyperkalemia may lead to muscle weakness and paresthesias and abnormalities in cardiac conduction. Clinical manifestations usually are preceded by conduction abnormalities, which may result in ventricular fibrillation and asystole. Hypermagnesemia generally is not associated with clinical symptoms. Severe hypophosphatemia can cause proximal muscle weakness, cardiac dysfunction, ataxia, seizures, and coma. Hyponatremia is a cause of nausea, vomiting, hypothermia, lethargy, agitation, headache, and seizures.

References:

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Question: 65

You are asked to evaluate a healthy 9-year-old boy who has a rash of several weeks' duration. Physical examination reveals several erythematous plaques (Item Q65) on the trunk surmounted by a thick, adherent scale. You observe pinpoint areas of hemorrhage on the surfaces of some lesions.

Of the following, the MOST likely diagnosis is

A. nummular eczema

B. pityriasis rosea

C. psoriasis

D. seborrheic dermatitis

E. tinea corporis

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Question: 65



Courtesy of D. Krowchuk

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Critique: 65 Preferred Response: C

The boy described in the vignette has findings typical of psoriasis: well-defined, round or oval plaques that have a thick, white, or silver scale. The scale is adherent, and when removed, pinpoint areas of hemorrhage may be observed (Auspitz sign) (Item C65A). Psoriasis is an inherited disorder that begins before age 16 in 25% to 45% of individuals. The disease has a predilection for the eyebrows, ears, extensor surfaces of the elbows and knees (Item C65B), umbilicus (Item C65C), and gluteal cleft. Most children exhibit scalp involvement with thick adherent scale (Item C65D). Pitting, yellowing, or thickening of the nails occurs in some patients (Item C65E). Lesions may appear in areas of trauma, such as scratches or abrasions (the Koebner phenomenon) (Item C65F).

A variant, called guttate (droplike) psoriasis, begins on the trunk as multiple erythematous macules that may mimic a viral exanthem. With time, the lesions become elevated and develop scale (Item C65G). Guttate psoriasis often is preceded by pharyngeal or perianal infection with group A beta-hemolytic *Streptococcus*, and patients who have the disorder should be evaluated for this possibility.

The morphology and distribution of psoriatic lesions generally permit their differentiation from lesions associated with other disorders. In particular, a psoriatic lesion typically has a thick, adherent scale that covers its entire surface. A unique form of eczema, termed nummular eczema (from the Latin word *nummulus*, meaning small coin), produces round, oozing, crusted erosions (Item C65H) or dry macules that have fine scale. The generalized eruption of pityriasis rosea may mimic guttate psoriasis. However, the lesions are small, oval, thin plaques that have fine scale located centrally, and they are oriented with their long axes parallel to lines of skin stress (Item C65I). Seborrheic dermatitis is characterized by erythematous macules or patches that have a greasy scale (Item C65J); the eyebrows, alar folds, or posterior auricular or presternal regions typically are involved, and there may be scaling of the scalp. Finally, tinea corporis causes one or several scaling annular lesions. Unlike the lesions of psoriasis, those of tinea corporis exhibit central clearing and peripheral scale (Item C65K).

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Critique: 65



When scale is removed from the surface of psoriatic lesions, pinpoint areas of bleeding (arrow) often are observed (ie, Auspitz sign).

Courtesy of D. Krowchuk

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Critique: 65



Psoriasis often involves the knees (shown here) or elbows.

Courtesy of D. Krowchuk

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Critique: 65



Psoriasis has a predilection for the umbilicus.

Courtesy of D. Krowchuk

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Critique: 65



Most children who have psoriasis exhibit scaling of the scalp.

Courtesy of D. Krowchuk

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Critique: 65



Pitting (shallow depressions) of the nails may occur in patients who have psoriasis.

Courtesy of D. Krowchuk

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Critique: 65



Lesions of psoriasis may occur in areas of trauma (ie, the Koebner phenomenon).

Courtesy of dermatlas.org

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Critique: 65



Guttate psoriasis is characterized by multiple erythematous scaling papules.

Courtesy of dermatlas.org

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Critique: 65



Nummular eczema produces round or oval crusted erosions.

Courtesy of D. Krowchuk

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Critique: 65



In pityriasis rosea, lesions are aligned with long axes parallel to lines of skin stress. On the back, this alignment may mimic the appearance of the branches of a fir tree.

Courtesy of D. Krowchuk

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Critique: 65



The lesions of seborrheic dermatitis are erythematous macules or patches that have a greasy scale.

Courtesy of D. Krowchuk

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Critique: 65



Tinea corporis produces annular lesions that have an elevated scaling border and central clearing.

Courtesy of D. Krowchuk

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Question: 66

During rounds with a group of medical students in the intensive care unit, you review the case of an 8-year-old boy who presented with gastrointestinal bleeding, received a blood transfusion, and underwent endoscopy. An ulcer was identified in the duodenal bulb (Item Q66). The boy now is being treated with intravenous pantoprazole.

Of the following, the BEST explanation of the mechanism of action of pantoprazole is that it

A. blocks the action of histamine by binding to the histamine receptor

B. eradicates gastric and duodenal *Helicobacter pylori*

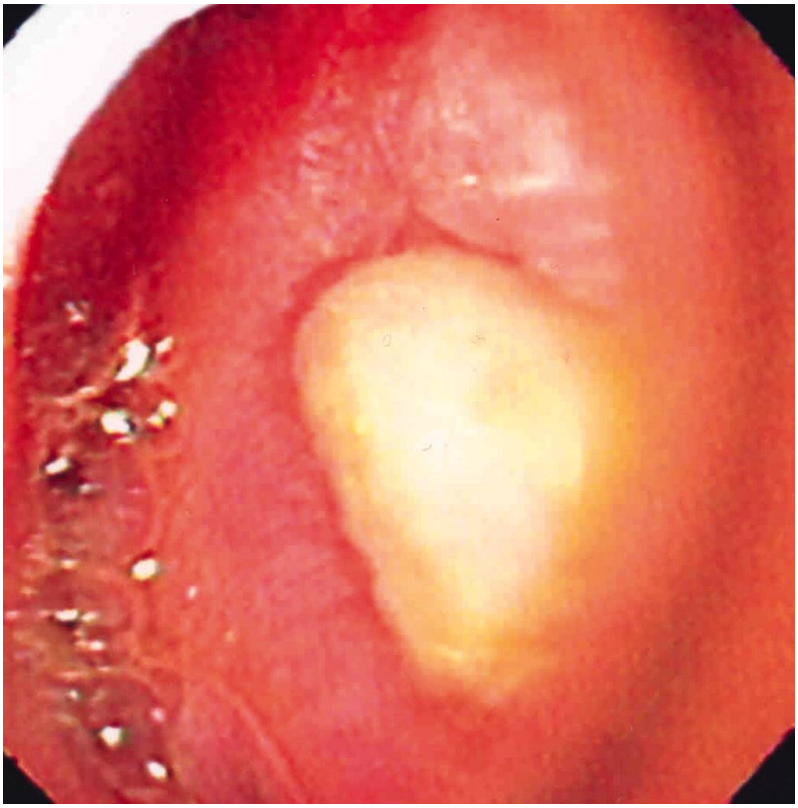
C. inactivates the hydrogen-potassium ATPase of the gastric parietal cells

D. inhibits histamine release by the enteric nervous system

E. stimulates prostaglandin and mucus release by the gastric epithelial cells

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Question: 66



Large ulcer in the duodenal bulb, as described for the boy in the vignette. The ulcer crater is covered by whitish exudate.

Courtesy of A. Bousvaros

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Critique: 66 Preferred Response: C

Medications that heal gastric inflammation, peptic ulcers, and reflux esophagitis most commonly work by decreasing gastric acid production. In the stomach, gastric acid is manufactured by parietal cells (Item C66), a group of polygonal cells located in the crypts of the gastric corpus. These cells possess a hydrogen-potassium ATPase on their luminal surface that pumps potassium intracellularly and hydrochloric acid into the lumen of the stomach. Parietal cell acid secretion is stimulated by gastrin, acetylcholine, and histamine. The histamine-2 blockers (eg, ranitidine, cimetidine, famotidine, nizatidine) work by blocking the effect of histamine on the histamine receptor of the parietal cell. In contrast, the proton pump inhibitors (PPIs), which include omeprazole, lansoprazole, pantoprazole, and rabeprazole, work by inactivating the hydrogen-potassium ATPase. PPIs may decrease the number of *Helicobacter pylori* bacteria in the stomach by creating a less favorable alkaline environment, but they do not eradicate the organism. Therefore, if *H pylori* eradication is necessary, the PPI should be combined with antibiotic therapy. Cromolyn, not PPIs, decreases histamine release by preventing mast cell degranulation. PPIs do not stimulate prostaglandin synthesis. Misoprostol, a synthetic prostaglandin, can protect the gastric mucosa of patients taking medications that deplete gastric prostaglandin, such as nonsteroidal anti-inflammatory drugs.

The potency and long half-life of PPIs make them highly effective medications that are used as first-line agents in treating bleeding ulcers or severe reflux esophagitis. However, their efficacy in reducing gastric acid production may eliminate an important host defense against bacterial overgrowth, and use of PPIs has been associated with pneumonia in the elderly. Therefore, long-term PPI therapy should be used only after considering the risk/benefit ratio for each patient.

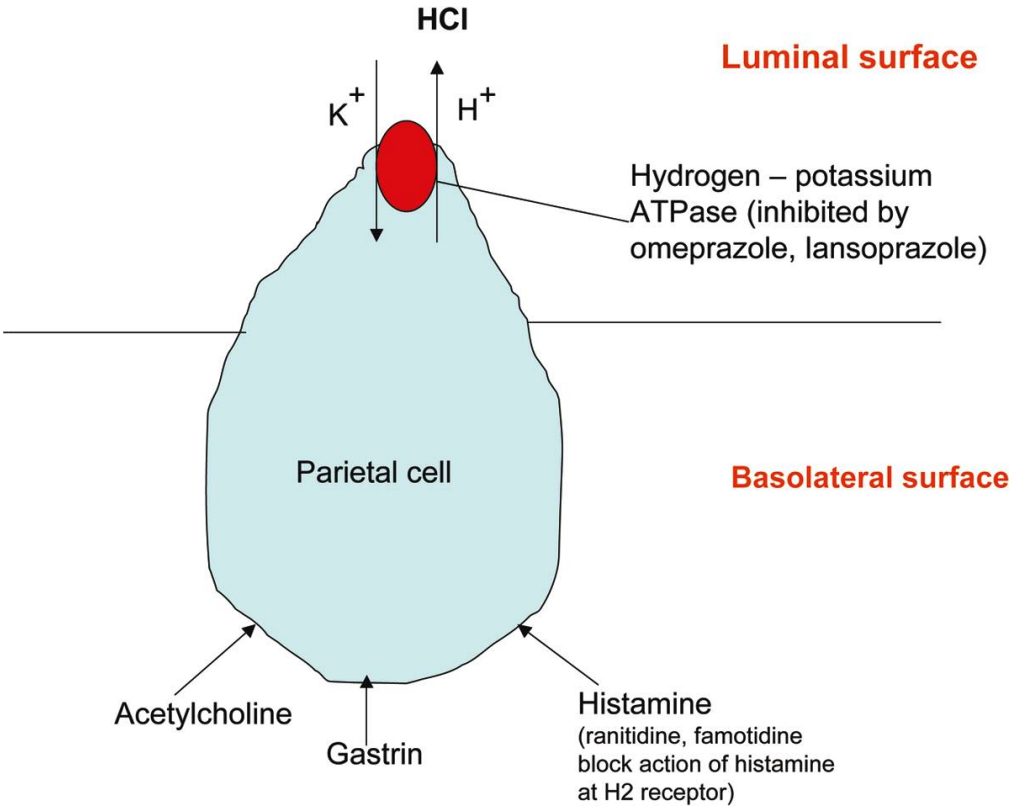
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Critique: 66



Drawing of a gastric parietal cell. Omeprazole and lansoprazole inhibit the hydrogen-potassium ATPase pump; ranitidine and famotidine block histamines effect by binding the histamine-2 receptor.

Courtesy of A. Bousvaros

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Question: 67

You are measuring serum electrolytes at 12 hours of age in a 4,500-g infant delivered by cesarean section at 36 weeks' gestation. The Apgar scores were 6 and 8 at 1 and 5 minutes, respectively. The infant is in no acute distress, breathing room air, and generally well-appearing, although he exhibits mild hypotonia. The laboratory results are:

• Sodium, 135 mEq/L (135 mmol/L)

• Potassium, 4 mEq/L (4 mmol/L)

• Chloride, 105 mEq/L (105 mmol/L)

• Carbon dioxide, 18 mEq/L (18 mmol/L)

• Calcium (total), 6.5 mg/dL (1.63 mmol/L)

• Phosphorus, 5.5 mg/dL (1.8 mmol/L)

• Magnesium 2 mg/dL (0.8 mmol/L)

The serum glucose value is 30 mg/dL (1.7 mmol/L).

Of the following, the MOST likely cause of neonatal hypocalcemia for this infant is

A. acute perinatal asphyxia

B. hypoglycemia

C. maternal diabetes mellitus

D. 22q11 deletion syndrome

E. vitamin D deficiency

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Critique: 67 Preferred Response: C

Hypocalcemia is one of the most common electrolyte disorders in the newborn period. It is defined by a serum total calcium [Ca2+] concentration of less than 7.0 mg/dL (1.75 mmol/L) or an ionized Ca2+ concentration of less than 4.0 mEq/L (1.0 mmol/L) and has two distinct presentations: early onset and late (Item C67). Early hypocalcemia may be prevented by adding elemental calcium as 10% calcium gluconate to maintenance intravenous fluids. Late hypocalcemia often has a specific cause that must be investigated and treated. Early hypocalcemia may be asymptomatic; clinical findings such as jitteriness, hypotonia, and twitching are inconsistent and do not correlate with serum Ca2+ concentrations. Late hypocalcemia may present with tetany.

The infant described in the vignette is large for gestational age (LGA), was born prematurely, and has early-onset hypocalcemia. Although no specific detail of maternal diabetes is noted, the findings of hypoglycemia, LGA status, and hypocalcemia strongly suggest that this is an infant of a diabetic mother.

No asphyxia is described or suggested by the Apgar scores and clinical condition. No heart murmur or cyanosis is noted to suggest conotruncal heart lesions associated with the 22q11 deletion syndrome. Hypoglycemia, although common in infants of diabetic mothers, does not cause hypocalcemia. Vitamin D deficiency results in late hypocalcemia.

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Critique: 67

Item 67: Causes of Neonatal Hypocalcemia

I. Early Hypocalcemia (first 3 days of postnatal life)

Prematurity (impaired transplacental mineral accretion)
Maternal diabetes mellitus (elevated calcitonin or hypomagnesemia)
Asphyxia (may involve elevated calcitonin)
Intrauterine growth restriction (impaired transplacental mineral accretion)
Maternal anticonvulsant therapy
Maternal hyperparathyroidism

II. Late Hypocalcemia (end of the first week or in the second week of postnatal life)

Elevated serum phosphate or low serum magnesium concentrations
Primary hypoparathyroidism: 22q11 deletion syndrome
Parathyroid hormone resistance
Vitamin D deficiency
Inherited disorders of vitamin D metabolism
Inherited disorders of calcium receptors
Intake of high-phosphate
Other:

- Alkalosis (shifts ionized Ca++ to protein-bound)
- Transfusion with citrate-containing preservative (chelates Ca++)
- Malabsorption or diarrhea

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Question: 68

A 2-year-old girl presents to the clinic with anal itching. Her perianal area is slightly reddened and shows mild excoriation. A tape test from the perianal area reveals the structures shown in Item 68 (Item Q68).

Of the following, the MOST appropriate treatment is

A. benznidazole in a single oral dose and repeated in 2 weeks

B. clotrimazole cream topically for 7 days

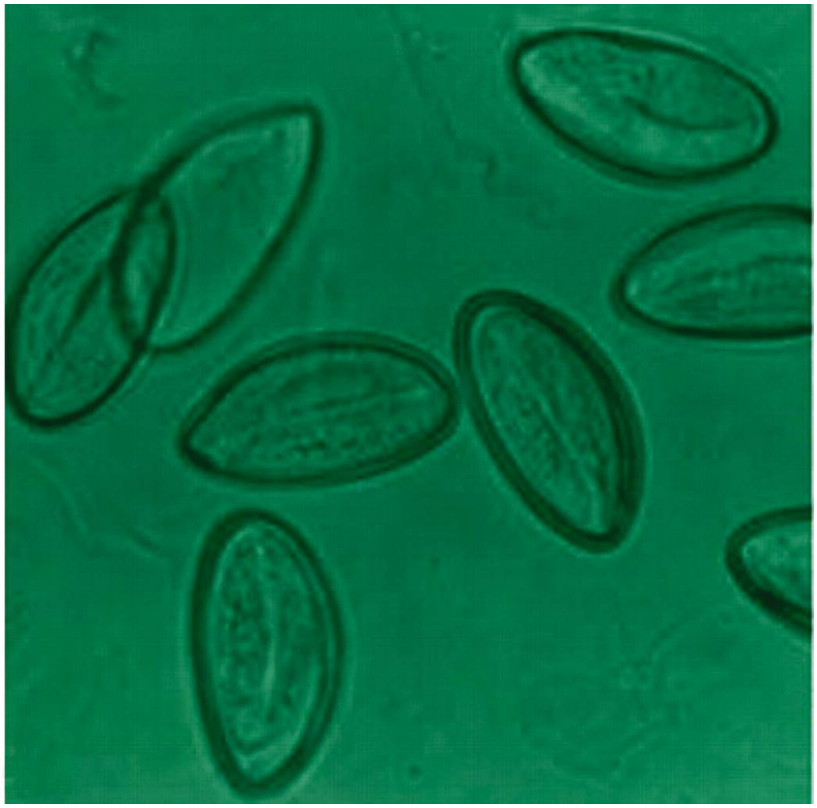
C. ketoconazole orally for 7 days

D. mebendazole in a single oral dose and repeated in 2 weeks

E. permethrin cream topically in a single dose

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Question: 68



Courtesy of *Red Book Online*

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Critique: 68 Preferred Response: D

Pinworm infection, which is caused by *Enterobius vermicularis,* is a common cause of anal itching in children. Transmission may occur via the fecal-oral route or by contact with contaminated hands or surfaces. The eggs are deposited on the perianal skin by the female pinworm, and reinfection via autoingestion can occur. The diagnosis is confirmed either by visualization of the adult worms on the perianal region or by applying transparent adhesive tape to the perianal skin to collect eggs (Item C68). Such specimens should be obtained immediately upon awakening in the morning.

Treatments for pinworm infection are mebendazole, pyrantel pamoate, or albendazole in a single dose, repeated in 2 weeks. These medications also are used to treat infections caused by roundworms, hookworms, and whipworms. Benznidazole is the treatment of choice for trypanosomiasis. Clotrimazole and ketoconazole are used in fungal infections, and permethrin is appropriate therapy for pediculosis and scabies.

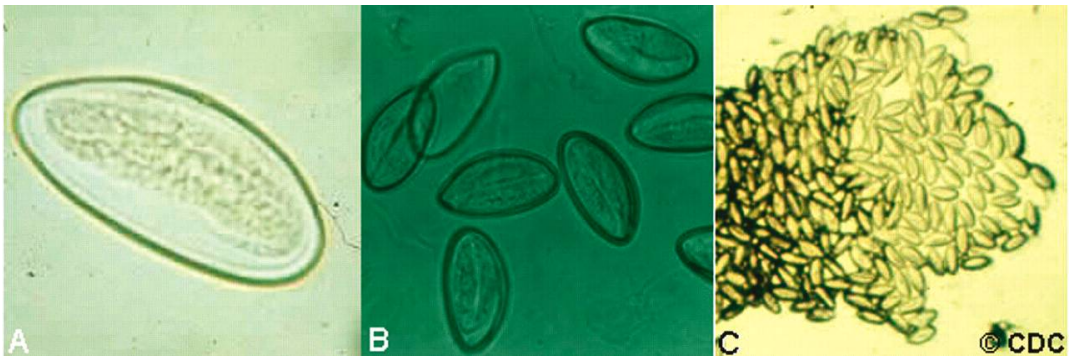
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Critique: 68



Pinworm infestation: Frames A and B are eggs, and Frame C is the appearance of eggs on cellulose tape test.

Courtesy of *Red Book Online* and the Centers for Disease Control and Prevention

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Question: 69

The father of a 9-year-old boy calls you because his son has been exposed to several children at school who have “strep throat.” Although the boy appears well and has no symptoms, the father notes that “all the other parents” are getting antibiotics for their children. He asks your advice.

Of the following, the BEST response is to

A. call in a prescription for amoxicillin

B. offer to see the boy if he develops a sore throat with fever

C. offer to see the boy immediately to examine him for signs of pharyngitis

D. perform a rapid streptococcal antigen test today

E. tell the father to bring the boy in immediately for an injection of benzathine penicillin

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Critique: 69 Preferred Response: B

Because the transmission rate of group A beta-hemolytic streptococcal disease has been estimated to be approximately 35% in schools and families, “prophylactic” treatment of asymptomatic individuals exposed to a patient in these settings is not indicated. Treatment also is not recommended because very early treatment of symptoms has been associated with suppression of the immune response to group A beta-hemolytic streptococci, thus increasing the risk of recurrent or persistent infection.

Treatment of the index case renders the patient noncontagious within 24 hours. After exposure, prompt treatment of children once symptoms are documented can reduce further spread in families and schools. Treatment of children who are symptomatic may be delayed as much as 9 days and still prevent rheumatic fever, the only important clinical outcome of streptococcal pharyngitis. If untreated, streptococcal pharyngitis resolves spontaneously in 2 to 5 days.

Accordingly, the best practice for asymptomatic children who have been exposed to an individual who has a documented group A beta-hemolytic streptococcal infection is to advise the parents to call the doctor if the child develops symptoms such as fever, pharyngitis, abdominal pain, and dysphagia. Because the child described in the vignette is asymptomatic, there is no reason to examine him immediately, perform testing for streptococcal infection, or treat him with an antibiotic.

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Question: 70

During a routine health supervision visit, the mother of one of your patients informs you that she is 28 weeks pregnant and that fetal echocardiography has revealed tetralogy of Fallot in an otherwise normal fetus. She asks you about the prognosis for children who have this condition.

Of the following, the MOST appropriate response is that

A. cardiac catheterization techniques may help avoid surgery

B. complete repair is associated with an excellent result, with less than 5% mortality

C. surgery most likely will be undertaken before the child is discharged after birth

D. tetralogy of Fallot rarely is associated with chromosomal abnormalities

E. the usual surgical approach is palliative and requires three stages

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Critique: 70 Preferred Response: B

Advances in the diagnosis, surgical technique, and perioperative care of congenital heart disease have led to dramatic improvements in patient outcomes. For example, the prognosis for tetralogy of Fallot (TOF), the most common of the cyanotic congenital heart diseases, now is excellent, with surgical survival of 95% and greater in many centers. More than 50 years ago, the first surgical procedure for TOF was performed with the placement of a systemic-to-pulmonary artery shunt (named the Blalock-Taussig shunt for the surgeon and cardiologist who conceived and subsequently performed the therapy). The shunting procedure supplied pulmonary blood flow to patients who previously would have died from progressive hypoxemia as pulmonary blood flow diminished to lethal levels. In the last several decades, a single operation has been used to repair TOF completely for newborns and infants who suffer from hypoxemia.

As surgical time has required briefer periods of cardiopulmonary bypass, perioperative management strategies have been developed to move children through postoperative care using evidence-based protocols. With improvements in surgical survival, studies to evaluate the long-term effects on neurodevelopment continue to shed new light on the effects of major congenital heart surgeries performed in the first weeks and months after birth. According to these data, nearly 95% of all children born with congenital heart disease necessitating surgical repair survive.

Despite advancing technique in the operating room and in postoperative units, morbidity from open heart surgery affects the quality of life for these patients. Risks for the infant undergoing repair of TOF include the development of progressive branch pulmonary artery stenosis, pulmonary valve insufficiency with resulting right heart dilation, right heart failure, and arrhythmia. In addition, a number (perhaps as high as 10%) of neonates requiring cardiopulmonary bypass surgery may have some element of neurodevelopmental delay or cognitive/school performance abnormality.

Cardiac catheterization is not performed routinely to assess TOF because the anatomic and hemodynamic details of the lesion are determined by echocardiography. As transcatheter balloon valvuloplasty became the therapy of choice for pulmonary stenosis in the 1980s, attempts were made to use this technique for the pulmonary outlet obstruction of TOF. Although relief was achieved in some patients, it was almost universally short-lived (days), and this is no longer a routine therapeutic approach for such patients. Surgical repair, which typically provides complete repair, is undertaken when patients begin to demonstrate desaturation from increasing right-to-left ventricular shunting, which results from the progressive obstruction of pulmonary blood flow. Most newborns who have TOF do not present with cyanosis, but rather with a murmur. As a result, surgical repair frequently is not required until later in infancy. It is now known that TOF can be associated with microdeletions in the q11 region of chromosome 22. Some reports suggest that such deletions occur in up to 20% of patients who have TOF.

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Question: 71

A 5-year-old girl presents with leg weakness of 12 hours' duration. She is afebrile and describes no pain in her back or elsewhere. Findings on physical examination include distal leg weakness and diminished reflexes in the legs. Finger-to-nose testing reveals slight dysmetria. Rectal tone is normal. You find an engorged tick on her occipital scalp.

Of the following, the MOST likely diagnosis is

A. botulism

B. Guillain-Barré syndrome

C. hydrocephalus

D. tick paralysis

E. transverse myelitis

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Critique: 71 Preferred Response: D

Acute and subacute progressive weakness requires an emergent neurologic evaluation that systematically considers possible sources of weakness in the brain, brainstem, spinal cord, nerve roots, nerves, neuromuscular junction, and muscle. However, a careful general physical examination also may lead to the proper diagnosis, as for the girl described in the vignette, on whom an engorged tick is identified. The findings of weakness, reduced reflexes, and mild ataxia (finger-to-nose dysmetria) are nonspecific, but in the presence of a tick, strongly suggest the diagnosis of tick paralysis.

The pathophysiology in botulism is at the neuromuscular junction. The botulinum toxin interferes with release of acetylcholine at the nerve terminal. Symptoms of botulism include pharyngeal and generalized weakness. Guillain-Barré syndrome, also known as acute inflammatory demyelinating polyneuropathy, can present with the findings described for the girl in the vignette, but affected patients usually complain of pain. Hydrocephalus, or an intracranial mass causing hydrocephalus, could present with leg weakness and ataxia, but the diminished leg reflexes and absence of headache for this girl make this diagnosis less likely. Transverse myelitis is an inflammatory disease of the spinal cord. Symptoms occur at and below the level of the lesion. The presence of dysmetria and the absence of bowel or bladder symptoms for this girl make this diagnosis unlikely.

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Question: 72

Parents who are new to your area bring their 4-year-old son to your office for evaluation of poor growth and developmental delays. They say that he was growing and developing normally until about 2 years of age. He now has a few single words. You review his records and note that his height was at the 50th percentile at age 2, and it is at the 10th percentile today. The occipitofrontal circumference is at the 97th percentile. On physical examination, the boy has dark and heavy eyebrows, full cheeks (Item Q72A), and full lips. His hands are broad, and his fingers are stubby and cannot be fully straightened (Item Q72B). There are flesh-colored nodules over the scapular regions and hepatosplenomegaly. A review of the family history reveals that the mother had two maternal uncles who died in mid- to late childhood of unknown causes.

Of the following, the condition that is MOST consistent with this child's presentation is

A. glycogen storage disease, type 1

B. Hunter syndrome

C. Hurler syndrome

D. I-cell disease

E. Sanfilippo syndrome

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Question: 72



Heavy eyebrows and full cheeks, as exhibited by the boy in the vignette.

Courtesy of M. Rimsza

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Question: 72



Inability to extend the fingers fully, as exhibited by the boy in the vignette.

Courtesy of M. Rimsza

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Critique: 72 Preferred Response: B

The mucopolysaccharidoses (MPSs) comprise a group of genetic disorders characterized by progressive physical deformation and, in some cases, cognitive limitation due to the abnormal production or function of certain lysosomal enzymes. These enzymes are required for the breakdown of glycosaminoglycans (GAGs), previously called mucopolysaccharides. When the enzymes are lacking, ongoing lysosomal storage of GAGs ultimately leads to destruction of the cell. The MPSs are chronic conditions that result in the progressive deterioration of multiple systems.

Classification of the MPSs is somewhat confusing because the disorders have eponymous names that were assigned before the genetics and variability of the conditions were known. Each condition has a range of manifestations. All of the MPSs are inherited in an autosomal recessive pattern, except for Hunter syndrome (MPSII), which is X-linked.

The severe form of Hunter syndrome typically presents between 2 and 4 years of age. In addition to the characteristic coarsening of the facial features common to most of the MPSs as well as dysostosis multiplex (skeletal radiographic findings include a flared rib cage, anterior vertebral wedging, and a J-shaped sella turcica), organomegaly, and progressive joint contractures, affected boys develop a “pebbly” appearance to the skin. Such skin findings commonly are appreciated over the scapulae, as described for the boy in the vignette, and often are self-limiting. The diagnosis is based on the finding of reduced iduronate sulfatase activity in white blood cells or skin fibroblasts.

Hurler syndrome, the severe form of MPSI, typically presents in the first 2 postnatal years with deceleration of growth and development, progressive facial coarsening, hirsutism, hepatosplenomegaly, cloudy corneas (Item C72), and findings consistent with dysostosis multiplex. The diagnosis is based on the finding of reduced alpha-L-iduronidase activity in white blood cells.

Sanfilippo syndrome (MPSIII) is unique among the MPSs in that affected individuals may have only minimal facial coarsening and may not have organomegaly or dysostosis multiplex. However, they have progressive reduction in cognitive function. Each of the four subtypes of Sanfilippo syndrome (A through D) has a distinct enzyme deficiency that can be diagnosed by analyzing lysosomal enzyme activity in white blood cells.

I-cell disease (also called “Leroy” I-cell disease or mucolipidosis II) is caused by deficiency of the enzyme N-acetylglucosaminyl-1-phosphotransferase, resulting in abnormal processing of lysosomal enzymes such that they cannot be recognized and taken up by the cell. Affected children present at birth with coarsening of the facial features, dislocated hips, clubfeet, and hernias. There is progressive contraction of the joints and severe mental retardation. The life expectancy is 2 to 10 years. Diagnosis is based on lysosomal enzyme assay in cultured fibroblasts.

Glycogen storage disease, type 1, or von Gierke disease, is caused by a deficiency of glucose-6-phosphatase, which results in glycogen storage in the liver, kidneys, pancreatic islet cells, and intestinal mucosa. Affected individuals may have hepatomegaly at birth, but the spleen is normal in size. Linear growth is retarded, and the children are at risk for recurrent hypoglycemia. Facial features are normal. Diagnosis is predicated on the finding of large amounts of glycogen in hepatocytes, followed by molecular genetic testing.

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Critique: 72



Patients who have Hurler syndrome may develop corneal clouding. This patient also has strabismus.

Courtesy of M. Rimsza

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Question: 73

A 16-year-old girl comes to your office complaining of irregular menstrual periods. She had menarche at 11 years of age and experiences menstrual periods every 2 weeks to 3 months. She has noticed increased acne, facial hair growth, and a 20-lb weight gain over the past year.

Of the following, the MOST likely diagnosis is

A. Cushing syndrome

B. hypothyroidism

C. Noonan syndrome

D. ovarian tumor

E. polycystic ovary syndrome

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Critique: 73 Preferred Response: E

Signs of androgen excess (especially hirsutism and acne) combined with oligomenorrhea or anovulatory bleeding, as described for the adolescent girl in the vignette, should alert the clinician to the possibility of polycystic ovary syndrome (PCOS). PCOS is the most common endocrinopathy in premenopausal women, but its clinical criteria for diagnosis, pathophysiology, and treatment remain controversial. Definite or probable criteria for diagnosis include laboratory or clinical hyperandrogenism, menstrual dysfunction, and exclusion of congenital adrenal hyperplasia. Other criteria often used to support a diagnosis include insulin resistance, perimenarcheal onset, elevated luteinizing hormone- to -follicle-stimulating hormone ratio, and ultrasonographic abnormalities. Abnormalities observed in patients who have PCOS occur in four key areas: 1) increase in luteinizing hormone secretion, 2) increase in adrenal androgen production, 3) increase in body mass, and 4) onset of adult patterns of insulin resistance. Some girls have a transient period of hyperandrogenism during the first 3 years after menarche; others have persistent PCOS symptoms. Patients who have PCOS are at increased risk for diabetes mellitus, obesity, insulin resistance, infertility, and impaired quality of life.

Cushing syndrome is less likely in the absence of other stigmata of the syndrome, including weakness, spontaneous ecchymoses, large purple striae (Item C73A), hypokalemia, and osteoporosis. A rapid onset of hirsutism with virilization is characteristic of ovarian tumors. Hypothyroidism can cause menstrual irregularities and weight gain, but is not associated with androgen excess. Patients who have Noonan syndrome (Item C73B) may present with delayed puberty and associated amenorrhea, but they do not exhibit androgen excess and weight gain.

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Critique: 73



Patients who have Cushing syndrome often exhibit large purple striae.

Courtesy of M. Rimsza

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Critique: 73



Noonan syndrome is characterized by hypertelorism, downward-slanted palpebral fissures, micrognathia, and webbing of the neck.

Courtesy of M. Rimsza

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Question: 74

An obstetrician calls to tell you she is caring for a woman who is 36 weeks pregnant and has required treatment with propylthiouracil during pregnancy for Graves disease. The mother is worried about the risk of neonatal thyrotoxicosis in the infant and wishes advice.

Of the following, a TRUE statement about infants born to mothers who have Graves disease is that

A. approximately 50% of infants have elevated concentrations of thyroid hormones, but only 20% require treatment

B. approximately 50% of infants have symptomatic thyrotoxicosis

C. concentrations of maternal thyroid-stimulating immunoglobulins do not correlate with fetal outcome

D. fewer than 10% of infants have symptomatic thyrotoxicosis

E. infants whose mother's disease is controlled adequately during pregnancy have a decreased risk of thyrotoxicosis

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Critique: 74 Preferred Response: D

Transplacental passage of maternal thyroid-stimulating immunoglobulins causes neonatal thyrotoxicosis, a condition that persists as long as sufficient stimulatory immunoglobulin remains in the blood, often for several months. Although up to 17% of infants of mothers who have thyrotoxicosis have laboratory evidence of high thyroid hormone concentrations, 10% or fewer are symptomatic. Symptoms in utero can include increased heart rate and a high output state. After birth, tachycardia, feeding problems, failure to gain weight, jitteriness, thyrotoxic stare, and persistent jaundice may be seen in severe cases. In general, maternal thyroid-stimulating antibody values correlate with the risk and severity of thyrotoxicosis. However, control of maternal thyrotoxicosis during pregnancy does not reduce the risk of neonatal thyrotoxicosis unless it is associated with decreases in thyroid-stimulating immunoglobulins. Neonatal thyrotoxicosis has been reported in infants born to women treated with ablative therapy for thyrotoxicosis years before the birth. It is important to remember that because of chronic suppression of thyroid-stimulating hormone in utero and after delivery, infants recovering from neonatal thyrotoxicosis may have prolonged and dangerous secondary hypothyroidism at a few months of age. Therefore, free thyroxine and thyroid-stimulating hormone (TSH) concentrations should be assessed every 3 to 4 weeks after recovery from thyrotoxicosis until TSH values rise to the normal range and free thyroxine values are persistently normal. Occasionally, treatment with thyroxine is necessary for some months if infants develop hypothyroidism.

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Question: 75

A 5-year-old girl recently was diagnosed with an autistic disorder and mental retardation. Her parents are upset by her lack of progress in her special education program and seek your guidance in treating her autism. At a parent support group, they were told about the use of complementary and alternative medical approaches to therapy. They ask whether they should pursue these interventions.

Of the following, your BEST response is to

A. explain to the parents that alternative treatments have been demonstrated to be ineffective

B. explain to the parents that they must consider the benefits, risks, and evidence regarding efficacy for each treatment

C. refer the parents for psychological counseling to deal with their guilt feelings

D. suggest the parents discuss the alternative treatments with the special education teachers

E. tell the parents that they should not expect much progress because their daughter has both mental retardation and autism

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Critique: 75 Preferred Response: B

Complementary and alternative medicine (CAM) is used frequently among children who have chronic illness or disability such as mental retardation or autism. Many parents become frustrated with biomedical therapies due to uncertainty of a cure and lack of an active role in the care plan. They also may be attracted to an approach that they perceive to be more "natural."

A number of CAM therapies have been advocated for children with developmental disabilities, including those who have autism, but to date no results from controlled trials support their efficacy. For example, there is no scientific evidence that vision therapy (eye exercise) or "patterning" (series of exercises promoted to enhance development) is effective in remediating pediatric developmental and neurologic conditions. Sensory integration therapy also lacks evidence-based research, although parents may report benefit for a child who has a high degree of sensory defensiveness. Other CAM therapies with no proven efficacy such as the use of hyperbaric oxygen or chelation may involve potential risks to the child as well.

The pediatrician is in a unique situation to help families evaluate CAM therapies and provide guidance regarding their benefits, risks, and evidence of efficacy. In particular, families should be informed about placebo effects and the need for controlled studies, as well as potential adverse effects. Discussion of CAM should not be referred to the child's special education teacher. Dismissal of CAM therapies as generally ineffective may be interpreted as a lack of sensitivity to the family's perspective. Referring the family to a counselor may help them discuss frustrations but would not address their question regarding the use of alternative therapies. Although the child has both mental retardation and autism, the clinician should be sensitive to the family's desire to have the child meet her cognitive potential.

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Question: 76

You receive a phone call at your office from a resident, who is making rounds in the hospital. She tells you that computed tomography scan of the 14-year-old girl you admitted last night with a history of fever and increasing headaches revealed a large brain abscess and asks your advice on choice of antimicrobial therapy.

Of the following, the BEST combination of antimicrobial agents to start for this patient is

A. cefazolin + metronidazole + gentamicin

B. clindamycin + ceftriaxone

C. nafcillin + metronidazole + cefuroxime

D. piperacillin-tazobactam + gentamicin

E. vancomycin + metronidazole + ceftriaxone

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Critique: 76 Preferred Response: E

Brain abscesses can occur at any age, but they are seen most commonly in children between 4 and 8 years of age. Patients who have right-to-left cardiac shunts, bacterial meningitis, chronic otitis media, sinusitis, orbital cellulitis, dental infections, and penetrating head wounds are at risk for the development of a brain abscess. The responsible bacteria vary, based on the mechanism of acquisition. A single organism is involved in approximately 70% of cases; the remainder of cases are polymicrobial. Abscesses that develop as a complication of chronic sinus disease also frequently contain anaerobes. In most situations, the early clinical manifestations of brain abscess are nonspecific complaints such as fever, headache, and lethargy. As the illness progresses, severe headache, vomiting, seizures, papilledema, focal neurologic signs, and coma may develop. Computed tomography and magnetic resonance imaging are the reliable methods of confirming the diagnosis (Item C76).

Because of the large number of potential pathogens involved in a brain abscess, the initial antimicrobial therapy should be broad, such as a combination regimen of vancomycin + metronidazole + ceftriaxone. Vancomycin provides coverage for methicillin-susceptible and -resistant *Staphylococcus aureus* as well as other aerobic gram-positive organisms (eg, streptococcal species). Metronidazole penetrates the blood-brain barrier well whether administered intravenously or orally and covers a large number of anaerobes. Ceftriaxone provides not only aerobic gram-positive coverage (eg, streptococcal species), but also addresses gram-negative organisms (eg, *Haemophilus influenzae*). The choice and number of agents can be altered according to culture results and the susceptibility of recovered organisms. The duration of antimicrobial therapy is usually 4 to 6 weeks.

Surgical management of brain abscesses has changed since the advent of neuroimaging. Current indications for surgical management include multiloculated abscess, abscess in the posterior fossa, fungal abscess, or the patient who is not improving or worsening with antimicrobial therapy.

Any combinations of antimicrobial agents that involve cefazolin or clindamycin for staphylococcal coverage are inferior to vancomyin due to their poor penetration of the central nervous system. Nafcillin penetrates the central nervous system well, but its use is limited because of the prevalence of methicillin-resistant *S aureus*. Combination penicillins (eg, piperacillin-tazobactam), cefuroxime, and gentamicin do not penetrate the central nervous system well.

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Critique: 76



Brain abscess: Computed tomography scan with contrast demonstrates a rim-enhancing lesion (arrow) extending to an area of bony destruction in an opacified frontal sinus. Vasogenic edema surrounding the abscess causes mass effect and shift of the midline to the right.

Courtesy of D. Krowchuk

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Question: 77

A 6-month-old male infant was treated for congenital syphilis at 2 weeks of age with 14 days of intravenous penicillin. At the time of his diagnosis, physical examination demonstrated only hepatosplenomegaly. Laboratory evaluation showed a rapid plasma reagin of 1:16, a negative cerebrospinal fluid VDRL result, and a positive fluorescent treponemal antibody absorption result.

Of the following, the MOST appropriate test to determine if the patient has been treated successfully is

A. blood culture

B. cerebrospinal fluid culture

C. cerebrospinal fluid VDRL

D. dark field analysis of nasal swab

E. rapid plasma reagin

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Critique: 77 Preferred Response: E

Of the congenital infections (eg toxoplasmosis, herpes, syphilis, human immunodeficiency virus infection, and cytomegalovirus infection), syphilis is the disease for which follow-up serologic testing is indicated to define disease activity and assess response to therapy. The nonspecific nontreponemal reaginic antibody tests for syphilis (Venereal Disease Research Laboratory [VDRL], rapid plasma reagin [RPR], automated reagin test [ART]) are performed rapidly and provide quantitative results that help to define disease activity and monitor response to therapy. The specific treponemal test antibody titers (fluorescent treponemal antibody absorption test [FTA-ABS], *Treponema pallidum* hemagglutination assay [TPHA], and microhemagglutination assay-*Treponema pallidum* [MHA-TP]) correlate poorly with disease activity and should not be used to assess response to therapy. Indeed, these specific treponemal test results may remain positive despite effective treatment.

All patients who have early or congenital syphilis should have repeat quantitative nontreponemal tests performed at 3, 6, and 12 months after therapy. An adequate response to therapy is indicated by a fourfold decrease in RPR titer or a titer that becomes nonreactive. Nontreponemal antibody titers should decrease by 3 months of age and be nonreactive by 6 months of age if the infant was infected and treated adequately or was not infected and initially seropositive because of transplacentally acquired maternal antibody. For infants treated after the neonatal period, the serologic response after therapy may be slower.

Blood culture and cerebrospinal fluid culture are not helpful in assessing response to therapy because *T pallidum* is not grown in culture. Repeat cerebrospinal VDRL is not helpful if the initial result is negative and the patient is asymptomatic. Dark field analysis may be helpful in initially diagnosing the patient if spirochetes (Item C77) are present, but it is not useful in assessing the response to therapy.

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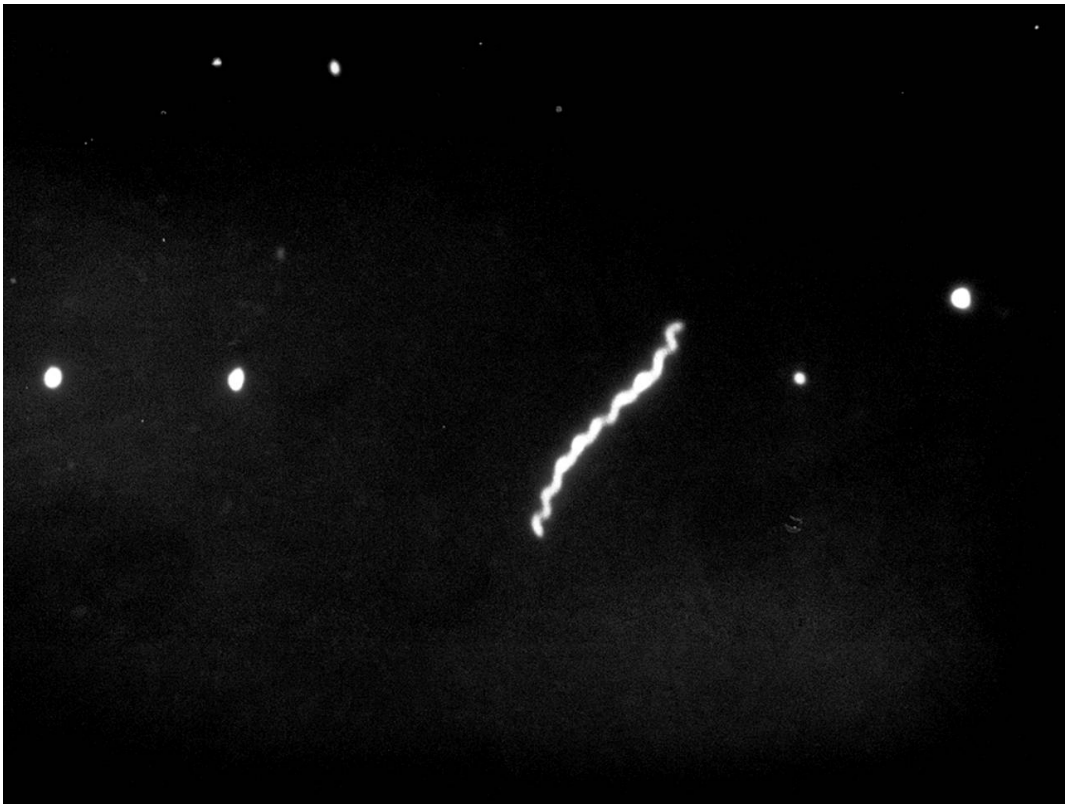
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Critique: 77



Photomicrograph of a *Treponema pallidum* bacterium (a form of spirochete) using darkfield microscopy (a method of illuminating organisms that are difficult to see using standard lighting techniques).

Courtesy of Centers for Disease Control and Prevention, Public Health Image Library

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Question: 78

During your evaluation of a 16-year-old boy at his health supervision visit, he reports that he had an episode of cola-colored urine associated with an upper respiratory tract infection 6 weeks ago. The only significant finding on his medical history is mild hearing loss. Today his blood pressure is 112/66 mm Hg. Urinalysis shows 3+ blood, 2+ protein, and 20 to 50 red blood cells per high-power field (RBC/HPF). Other laboratory findings include:

• Blood urea nitrogen, 14 mg/dL (5 mmol/L)

• Serum creatinine, 0.7 mg/dL (61.9 mcmol/L)

• Albumin, 4 g/dL (40 g/L)

Serum complement components (C3 and C4) are normal, and antinuclear antibody titers are negative. You discuss these results with his mother, who then tells you that she recalls being told she had blood in her urine. Urinalysis for the child's mother demonstrates 2+ blood with 10 to 20 RBC/HPF.

Of the following, a TRUE statement about this patient's disorder is that

A. boys who have this condition are more likely to develop chronic renal failure than are girls

B. immune complex deposition within the kidney is believed to be causative

C. it is classified as a small vessel vasculitis

D. the biologic defect in the kidney is believed to involve the glomerular epithelial cell

E. the condition typically has an autosomal dominant inheritance

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Critique: 78 Preferred Response: A

The adolescent male described in the vignette has hematuria and proteinuria documented on a health supervision visit and a history of cola-colored urine temporally associated with an upper respiratory tract illness 6 weeks earlier. Such findings suggest the presence of an underlying glomerulonephritis that can range from overt disease with gross hematuria when accompanied by an intercurrent illness to a subclinical state with persistent abnormal urinary findings.

The evaluation of a child who has glomerulonephritis should begin with careful measurement of blood pressure and renal function (serum creatinine) to assess the severity of the clinical situation, followed by a serologic evaluation to screen for an underlying cause. The normal complement components C3 and C4 described for this boy suggest three likely diagnostic possibilities: immunoglobulin A glomerulonephritis, pauci-immune vasculitis, or familial nephritis. The discovery of hematuria in the mother suggests familial nephritis.

Familial nephritis can be caused by either Alport syndrome (AS) or thin glomerular basement membrane (GBM) disease. Both disorders involve underlying defects of the GBM that result in a disruption of the glomerular capillary barrier. This barrier is comprised of the glomerular endothelial cell, the GBM, and the podocyte (visceral glomerular epithelial cell). Any disruption to this barrier permits access of restricted substances, such as red blood cells or protein, to the urinary space. AS is not caused by immune complex deposition, small vessel vasculitis, or defects in the glomerular epithelial cell.

AS is caused by a defect of type IV collagen, a component of the GBM. The cause in 80% of cases is an X-linked disease involving a gene defect of COL4A5 (which codes for the alpha 5 chain of type IV collagen). A common presentation for AS is a male child who has asymptomatic microscopic hematuria that may become overt hematuria in the presence of a respiratory infection. The renal lesion may progress to azotemia with nephrotic-range proteinuria in adolescence. End-stage renal disease has been reported in 50% to 90% of affected males by age 30 years, depending on the severity of the genetic defect. High-frequency sensorineural hearing loss occurs in approximately 50% of affected patients. A number of patients also have ophthalmologic findings, with anterior lenticonus, a defect of the lens bowing into the anterior chamber, being the most common.

Due to the X-linked inheritance of this disorder in most patients, males often present earlier than females and have more severe disease. As such, males are more likely than females to develop chronic renal failure. As in this case, the evaluation of a male child who has persistent asymptomatic hematuria should include a dipstick urinalysis of the mother to screen for hematuria. Based on the genetics of this disorder, 50% of male children of female carriers are affected. At present, there is no specific treatment for AS. Nephrologists focus on blood pressure control and reduction of proteinuria, often with the use of angiotensin-converting enzyme inhibitors.

Genetic defects of the alpha 3 or alpha 4 chain of type IV collagen result in an autosomal form of AS (autosomal recessive or dominant), which comprises the remaining 20% of cases of AS.

Thin GBM disease has overlapping characteristics with AS in that it runs in families, is caused by a defect of the GBM, and results in persistent microscopic hematuria. Recently, thin GBM disease has been traced to a heterozygous state for the gene defect of alpha 3 or alpha 4 of type IV collagen, which explains why it has a much more favorable prognosis than AS.

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Question: 79

A 17-year-old boy is applying for entry into military service and requires a complete history and physical examination. During the interview, he states that he is healthy, although he admits to being treated for three cases of pneumonia over the past 10 years. A chest radiograph performed during the last infection showed a left lower lobe pneumonia, and the patient states that the infection is “always on that side.” The only finding of note on the physical examination today is slightly diminished breath sounds over the left lower lobe.

Of the following, the MOST likely cause for this boy’s recurrent pneumonias is

A. bronchogenic cyst

B. congenital cystic adenomatoid malformation

C. congenital lobar emphysema

D. extrapulmonary sequestration

E. intrapulmonary sequestration

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Critique: 79 Preferred Response: E

The presentation of recurrent unilateral pneumonias should prompt the clinician to consider congenital malformations of the lung, specifically pulmonary sequestration. Pulmonary sequestrations can be classified as intrapulmonary or extrapulmonary. Intrapulmonary sequestrations account for 75% to 90% of all sequestrations. Patients usually present in adolescence or adulthood with cough, wheezing, fever, and recurrent pulmonary infections (Item C79A). Surgical lobectomy generally is curative. Although also located on the left side in most cases, extrapulmonary sequestrations usually present prior to 6 months of age and often occur in conjunction with other congenital anomalies such as colonic duplication, pulmonary hypoplasia, or vertebral anomalies. Extrapulmonary sequestration can present similarly to intrapulmonary sequestrations, with cough, dyspnea, and infection, but also can result in feeding difficulty and, in rare cases, congestive heart failure due to increased shunting.

Bronchogenic cysts are the most common cause of a cyst in the lung. Most commonly located near central airway structures, bronchogenic cysts may present with symptoms of airway compression or infection, but they frequently are asymptomatic and discovered incidentally on chest radiography (Item C79B).

Congenital cystic adenomatoid malformation (CCAM) is another common congenital lung anomaly that typically is identified on prenatal ultrasonography. Most CCAMs present in the newborn period with respiratory distress (Item C79C) and, depending on the type, may involve an entire lung, be associated with congenital anomalies, or result in fetal hydrops and pulmonary hypoplasia. Affected patients can present during childhood with recurrent pneumonia, but CCAM is less common than intrapulmonary sequestration at the age of the boy in the vignette.

Congenital lobar emphysema (CLE) is the most common neonatal cause of cystic malformation of the lung (Item C79D), and similar to CCAM, typically presents in the neonatal period with respiratory distress and airway obstruction.

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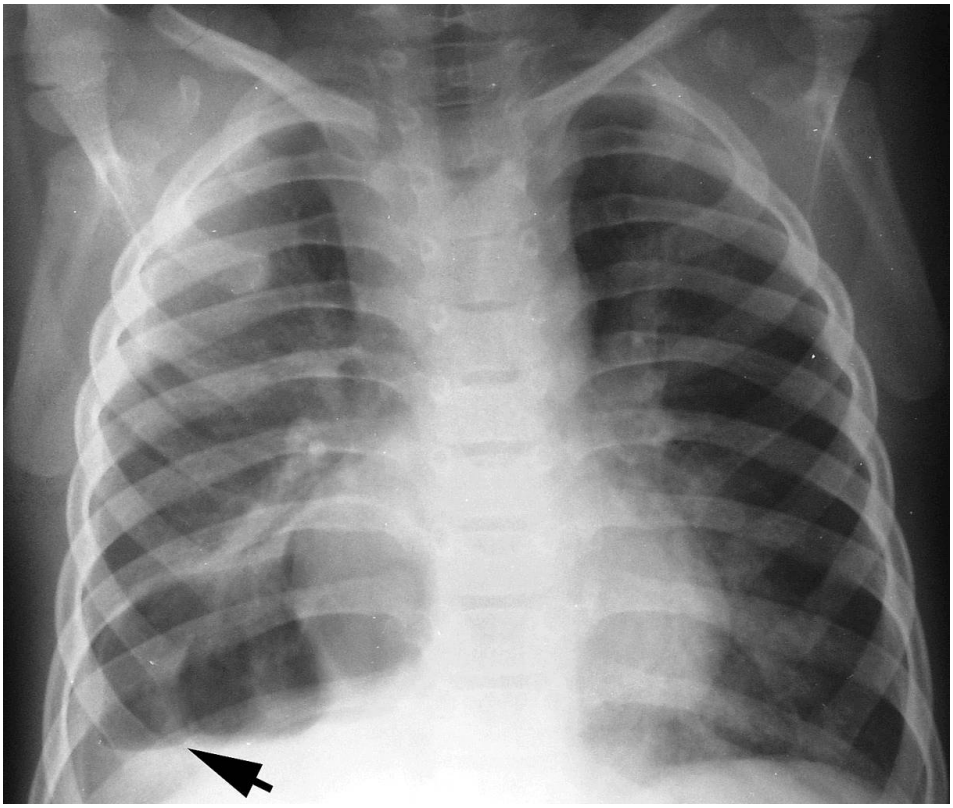
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Critique: 79

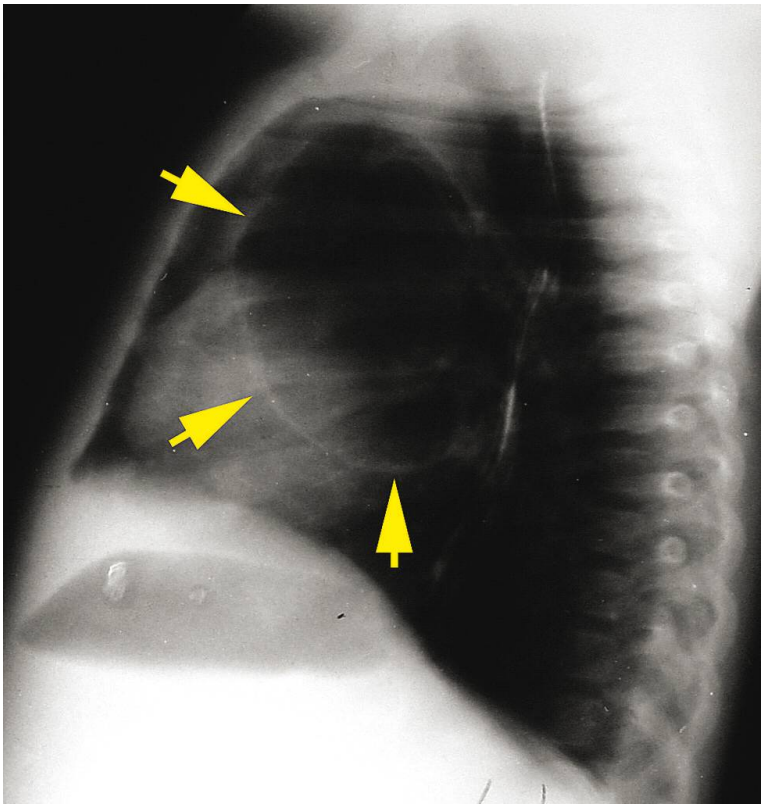


Anteroposterior radiograph of the chest shows multiple large cysts in the right lower lobe. Several of the cysts contain air fluid levels (arrow), likely due to superimposed infection, a common presentation for intralobar sequestration.

Courtesy of D. Mulvihill

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Critique: 79

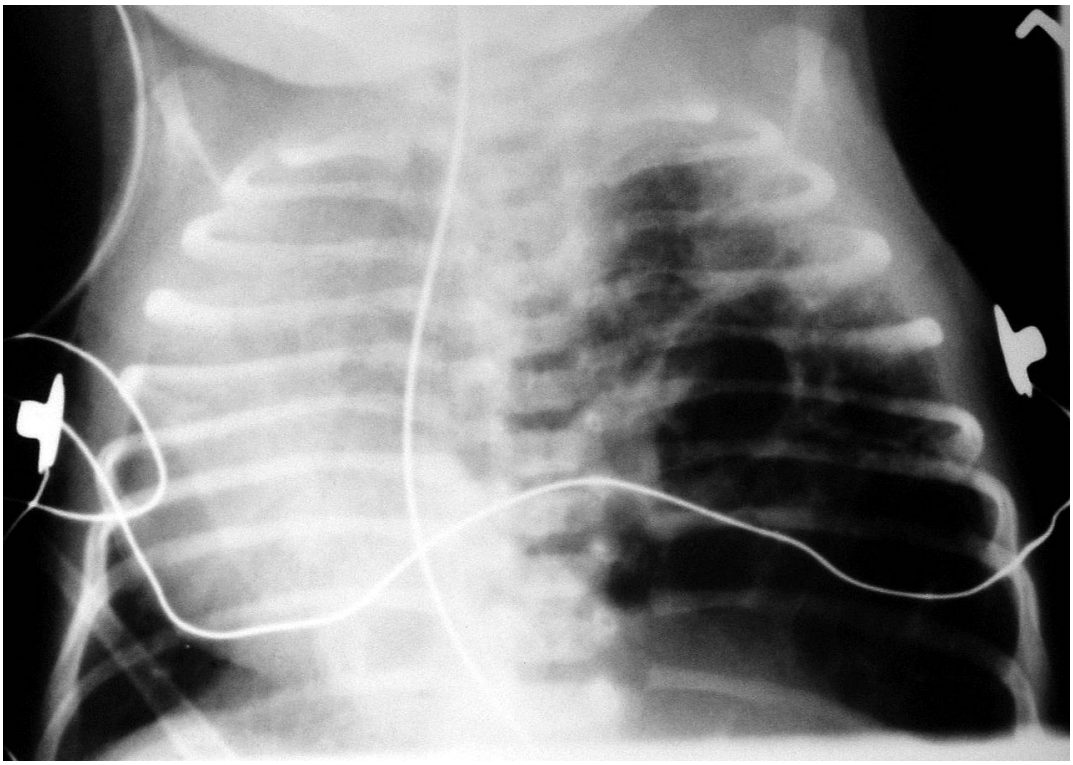


Lateral radiograph of the chest shows a single large bronchogenic cyst in the left lung without internal septations.

Courtesy of D. Mulvihill

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Critique: 79

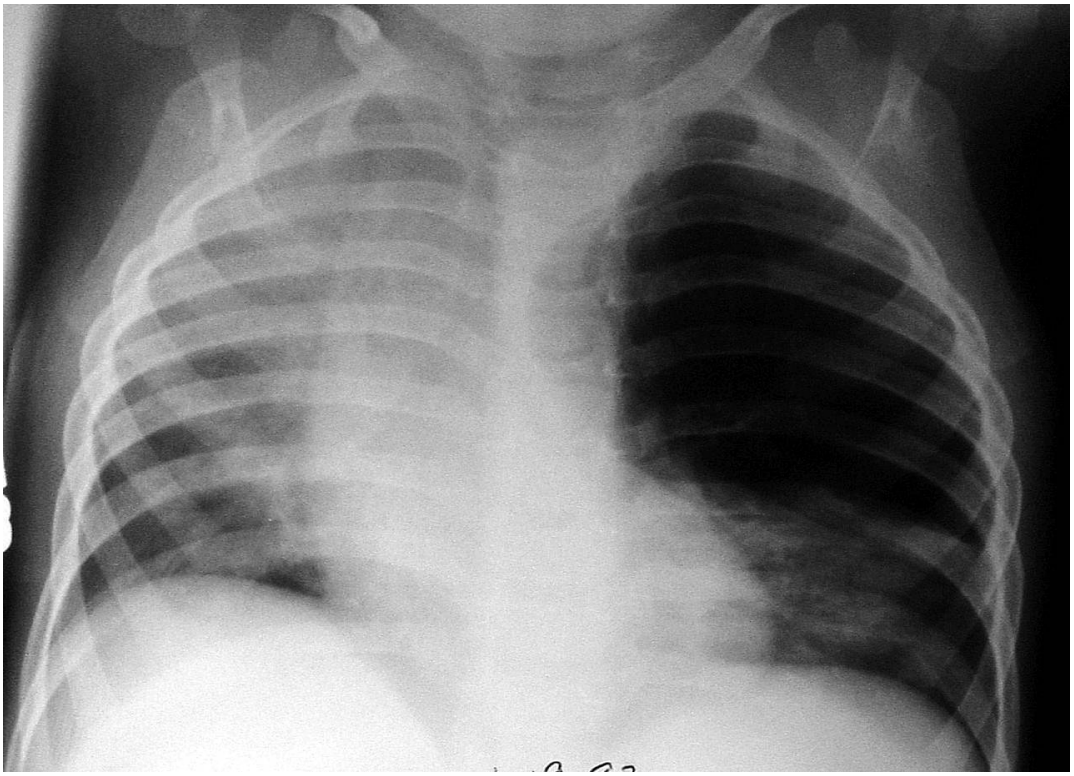


Anteroposterior chest radiograph in congenital cystic adenomatoid malformation reveals multiple cystic air collections of varying sizes involving the left lower lobe and causing displacement of the heart and mediastinum to the right.

Courtesy of D. Mulvihill

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Critique: 79



Anteroposterior radiograph of the chest in congenital lobar emphysema shows overinflation of the left upper lobe with shift of the mediastinum to the right. There is left lower lobe atelectasis.

Courtesy of D. Mulvihill

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Question: 80

A 14-year-old boy presents to the office with a 4-hour history of severe scrotal pain after running in a track meet. He is pale, nauseous, and in obvious pain. He denies trauma, dysuria, history of fever, or penile discharge. He is sexually active. On physical examination, his penis appears normal and without meatal discharge, but the scrotum is swollen bilaterally, and there is significant tenderness to palpation of both testicles. The testicles appear high in the scrotum, and the cremasteric reflexes are absent bilaterally.

Of the following, the next MOST appropriate step in the evaluation of this patient is

A. abdominal computed tomography scan

B. emergent surgical evaluation

C. polymerase chain reaction test for *Chlamydia*

D. radionuclide scan of the testicle

E. urinalysis

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Critique: 80 Preferred Response: B

Testicular torsion is a true urologic emergency; delay in restoration of testicular blood flow for more than 6 hours is associated with a high rate (>80%) of testicular loss. Therefore, it is critical to diagnose testicular torsion rapidly and refer the patient promptly to a urologist for surgical treatment.

Testicular torsion should be suspected in any male who presents with a swollen, painful scrotum. Peak ages for torsion are in the neonatal period and during adolescence. Patients typically present with sudden, acute onset of scrotal pain, often associated with nausea and vomiting, following vigorous activity or minor testicular trauma, as reported for the boy in the vignette. On physical examination, the scrotum is swollen and often erythematous (Item C80A), and the testicle is exquisitely tender to palpation. If the patient permits an adequate testicular exam, the testicle can be found high in the scrotum and may be positioned transversely. The cremasteric reflex rarely is present on the affected side. Although testicular torsion is usually unilateral, because the underlying anatomic abnormality that predisposes to testicular torsion (the “bell-clapper” deformity (Item C80B) in which the testicle lacks the normal attachment to the tunica vaginalis) is usually bilateral, testicular torsion may rarely present bilaterally.

The differential diagnosis of the acutely painful and/or swollen scrotum includes epididymitis, torsion of the testicular appendix, inguinal hernia, testicular trauma, and orchitis. If the clinical diagnosis of testicular torsion is in question, color Doppler ultrasonography, which can assess testicular blood flow as well as anatomy, can be a useful adjunct for delineating the cause of the symptoms, if it can be obtained without delay. Other imaging studies, including abdominal computed tomography scan or radionuclide testicular scan, cannot provide timely and adequate information to exclude testicular torsion as the diagnosis. Urinalysis or urethral swab for *Chlamydia* testing can be useful adjuncts to the evaluation if epididymitis is suspected, but they are not be helpful in excluding the diagnosis of testicular torsion.

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Critique: 80

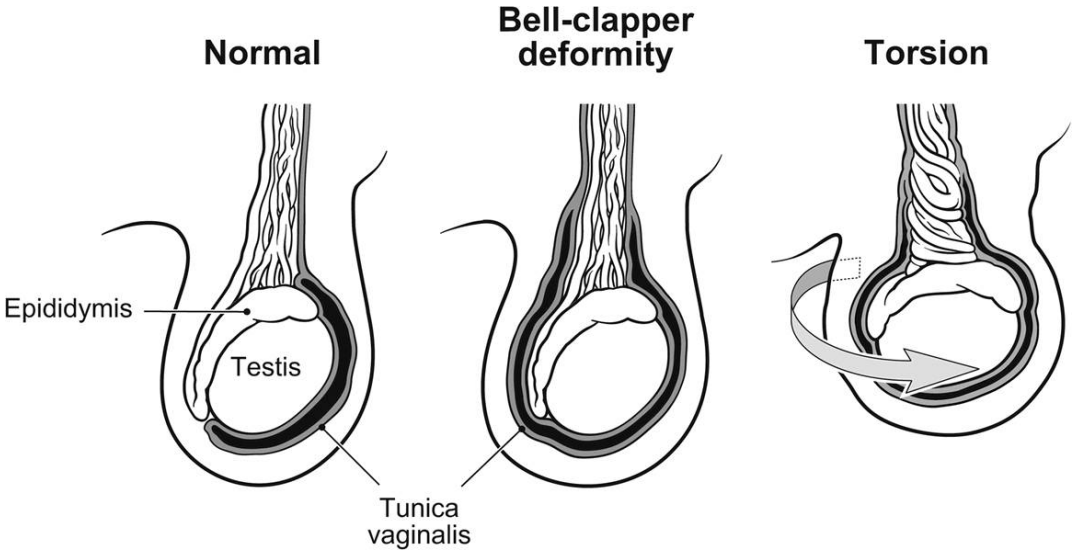


In testicular torsion, the scrotum is swollen and erythematous.

Courtesy of the Media Lab at Doernbecher

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Critique: 80



Normally (left), the tunica vaginalis attaches to the posterolateral aspect of the testis. In the bell-clapper deformity (center), the tunica vaginalis completely encircles the distal spermatic cord, epididymis, and testis, allowing the structures to swing and rotate within the tunica vaginalis (like a clapper in a bell). This abnormality predisposes to testicular torsion (right).

Courtesy of A. Johnson

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Question: 81

An 8-year-old boy presents with a 2-week history of an enlarging, tender lump on the scalp. The only notable findings on physical examination are alopecia overlying a boggy mass on the scalp and posterior cervical lymphadenopathy (Item Q81).

Of the following, the MOST appropriate treatment is

A. cefazolin intravenously

B. griseofulvin orally

C. incision and drainage

D. ketoconazole topically

E. mupirocin topically

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Question: 81



Courtesy of M. Rimsza

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Critique: 81 Preferred Response: B

The boy described in the vignette has a tender, boggy mass on the scalp that has surface pustules in addition to regional lymphadenopathy. These findings are consistent with the diagnosis of a kerion, one manifestation of an inflammatory response to a dermatophyte infection of the scalp (ie, tinea capitis). Although not present in this child, some patients who have a kerion develop fever.

Treatment of a kerion requires griseofulvin orally at a dose of approximately 20 mg/kg per day of the microsize preparation for 6 to 8 weeks. For resistant infections or when griseofulvin is not tolerated, terbinafine, itraconazole, or fluconazole may be used, although none of these agents is approved by the United States Food and Drug Administration for this indication in children. In those who have severe disease, adjunctive therapy with an oral corticosteroid (eg, prednisone at a dose of 0.5 to 1.0 mg/kg per day for 2 to 4 weeks) may be employed to reduce inflammation and pain. Oral antibiotic therapy for possible secondary *Staphylococcus aureus* infection is recommended by some, especially if there is widespread crusting of the scalp. Although most kerions resolve without complications, a minority of patients experience permanent alopecia that is the result of inflammation and scarring.

Topical antifungal agents (eg, ketoconazole) do not penetrate the follicle and will not eradicate tinea capitis and the resulting kerion. Although the patient described in the vignette has a lesion that is similar in appearance to a bacterial abscess, the presence of alopecia strongly suggests the diagnosis of a kerion. Accordingly, incision and drainage, intravenous cefazolin, and topical mupirocin are not indicated.

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Question: 82

A 14-year-old boy who recently emigrated from Vietnam is brought in by his mother because of intermittent abdominal pain and vomiting of 1 year's duration. He is a runner and has taken four doses of ibuprofen this month for chronic knee pain. In the past, he has been treated with ranitidine, which partially relieved the abdominal symptoms. You refer him to a gastroenterologist, who performs endoscopy and discovers a nodular antrum (Item Q82) and small duodenal ulcer.

Of the following, the MOST likely diagnosis for his symptoms and endoscopic findings is

A. celiac disease with lymphocytic gastritis

B. Crohn disease

C. eosinophilic gastritis

D. *Helicobacter pylori* infection

E. nonsteroidal anti-inflammatory drug-induced ulcer

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Question: 82



Courtesy of A. Bousvaros

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Critique: 82 Preferred Response: D

The endoscopic appearance of a nodular antrum and duodenal ulcer described for the boy in the vignette strongly suggest the presence of gastritis caused by *Helicobacter pylori* infection. The nodular antrum (Item C82A) is present because of lymphoid hyperplasia that occurs when the mucosal immune system is activated by *H pylori*. Upon careful examination of the gastric biopsy, spiral-shaped microorganisms (Item C82B) can be seen in the mucus layer overlying the epithelium in the gastric glands. Although *H pylori* is a common commensal organism in adults, it can cause ulcer disease. Strains that cause ulcers are more likely to possess a virulence protein called CagA (cytotoxin-associated gene A).

In the United States, the prevalence of *H pylori* infection is approximately 20% in Caucasians and 50% in African-Americans and Hispanic-Americans. The prevalence is lower in children, presumably because of improvements in hygiene and socioeconomic status over time. The gold standard for diagnosing *H pylori* is endoscopy and biopsy. The organism usually can be identified on histology, especially if silver stain is used; culture typically is not necessary. Noninvasive tests (including serum serology, fecal antigen, and urea breath testing) are useful in patients in whom the index of suspicion for *H pylori* infection is high. If the prevalence or likelihood of *H pylori* is low, serologic testing is likely to give a false-positive result. *H pylori* infection generally is treated with a 14-day course of therapy with a proton pump inhibitor plus two antibiotics (amoxicillin and clarithromycin, amoxicillin and metronidazole, tetracycline and metronidazole). If repeated courses of antibiotics do not eradicate the organism, culture and testing for antimicrobial resistance and the CagA virulence factor can be performed at research laboratories.

Other risk factors for ulcers in childhood include use of nonsteroidal anti-inflammatory drugs (NSAIDs), Crohn disease, viral infections, and physical stress (eg, intensive care hospitalization). Ulcers also may occur without any obvious predisposing cause. Celiac disease and eosinophilic gastroenteritis may cause gastric inflammation, but generally do not cause ulcers. In contrast, NSAID use and Crohn disease may cause ulcers, but not nodular gastritis.

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Critique: 82

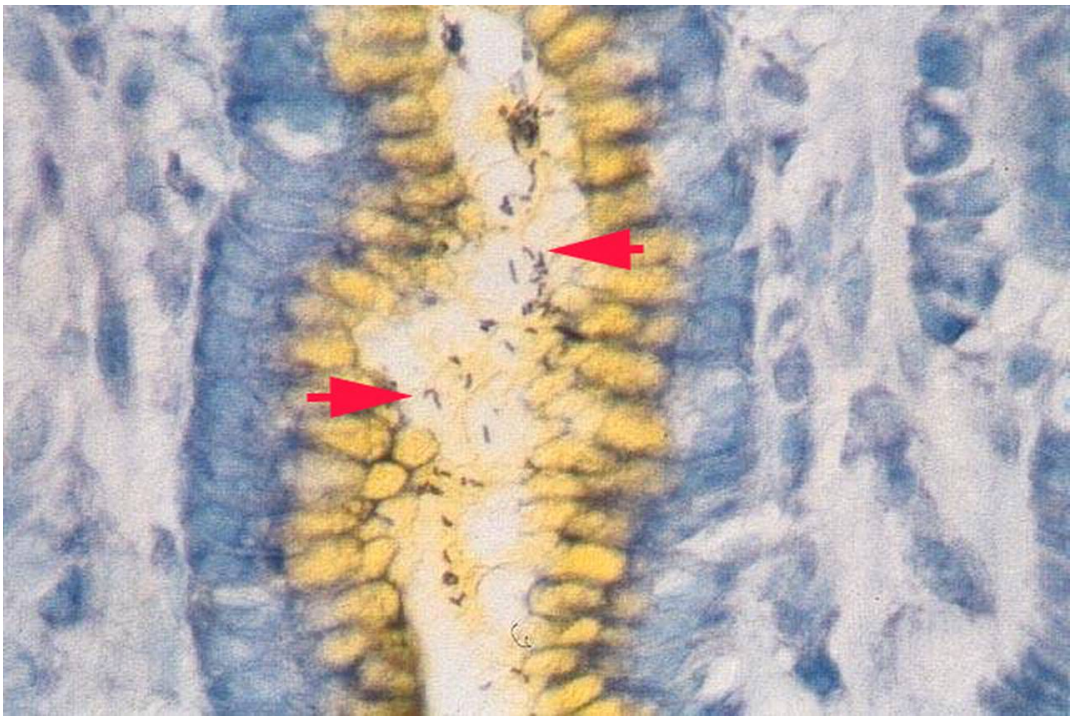


Endoscopic view of the stomach demonstrating nodular inflammation in the gastric antrum, a common feature of *Helicobacter pylori* gastritis.

Courtesy of A. Bousvaros

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Critique: 82



Silver stain of a gastric biopsy demonstrating curved-shaped *H pylori* bacteria in a gastric crypt (arrows).

Courtesy of J. Glickman

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Question: 83

A 5-day-old term infant presents to the emergency department with a history of bile-stained emesis. She is well nourished and hydrated and had an unremarkable course in the newborn nursery. She was discharged at 48 hours after birth and was breastfeeding, but her mother states the baby always has vomited. Physical examination reveals an afebrile infant who has normal vital signs, but no audible bowel sounds on abdominal evaluation. A flat-plate abdominal radiograph reveals a paucity of bowel gas (Item Q83).

Of the following, the MOST likely diagnosis is

A. anorectal atresia

B. cystic fibrosis

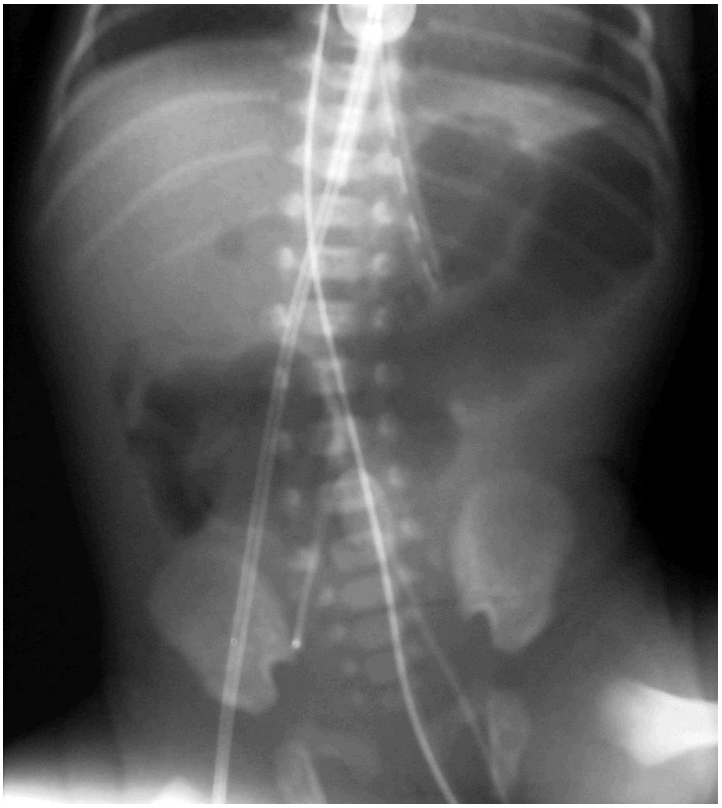
C. malrotation of the bowel

D. septic ileus

E. tracheoesophageal fistula

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Question: 83



Anteroposterior radiograph of the abdomen shows dilated loops of bowel in the upper abdomen but no gas distally.

Courtesy of D. Mulvihill

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Critique: 83 Preferred Response: C

The patient described in the vignette presents with bilious emesis in the first postnatal week. Bilious emesis always is a surgical emergency in the newborn. The differential diagnosis includes any form of anatomic or functional gastrointestinal obstruction, such as an ileus, that may be associated with sepsis. This infant is not systemically ill, febrile, dehydrated, or hemodynamically unstable. Although her abdomen is not distended, the absence of bowel sounds on auscultation and the paucity of bowel gas on abdominal radiograph (Item C83A) are concerning for malrotation of the bowel with a midgut volvulus. Early in this condition, findings on the physical examination may be as described, but they can change rapidly, depending on how much the mesenteric perfusion has been compromised. Later signs include rectal bleeding, hematemesis, palpable bowel loops, and an uncomfortably distended abdomen with respiratory embarrassment and hypovolemic shock. If not diagnosed and expeditiously addressed surgically, most of the small intestine may be lost.

Surgical exploration may need to precede any contrast gastrointestinal imaging (upper gastrointestinal radiographic series (Item C83B)) if the patient is unstable. Plain radiographs may demonstrate a normal, nonspecific bowel gas pattern; duodenal obstruction with the appearance of a “double bubble” (Item C83C); gastric distention with a paucity of distal intraluminal gas; or a generalized pattern of dilated small bowel loops.

Half of all cases of midgut volvulus occurring in the first postnatal year appear in the first week, another 25% appear in weeks 1 through 4, and the final 25% appear from 1 month to 1 year of age. These account for 90% of all cases of acute volvulus in pediatric patients.

Anorectal atresia is associated with delayed or absent passage of stool. Abdominal distention classically develops over the first 48 hours of postnatal life regardless of whether the infant is fed. This condition and tracheoesophageal fistula (TEF) may be part of a broader spectrum of associated malformations known as the VATER or VACTERL association (V=vertebral anomalies, A=anorectal atresia, C=cardiac malformations, TE=TEF, R=renal anomalies, L=limb anomalies). TEF typically presents with respiratory distress or poor handling of oropharyngeal secretions and may present with gastrointestinal obstruction in utero or postnatally. The clinician should evaluate the patient who has anorectal atresia or TEF carefully for other findings in the VACTERL spectrum.

Cystic fibrosis may be associated with meconium ileus and delayed passage of stool beyond 24 hours. Affected infants may have bilious emesis if fed, and plain abdominal radiography demonstrates dilated loops of bowel of varying caliber. If associated with meconium peritonitis or a pseudocyst, intraperitoneal calcification may be seen. A septic ileus is associated with systemic illness, abdominal distention, and a paucity of bowel gas or dilated loops of bowel on radiographs.

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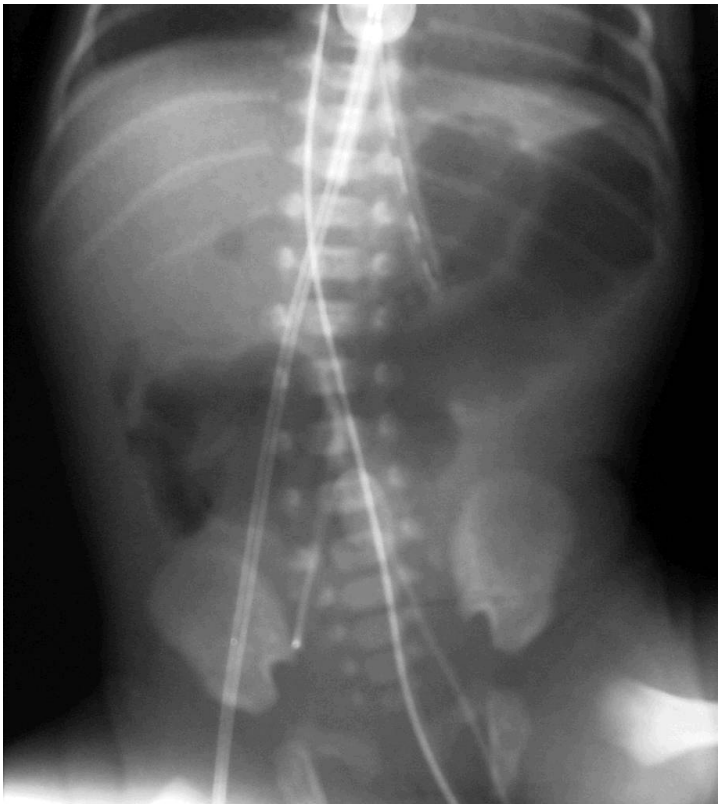
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Critique: 83

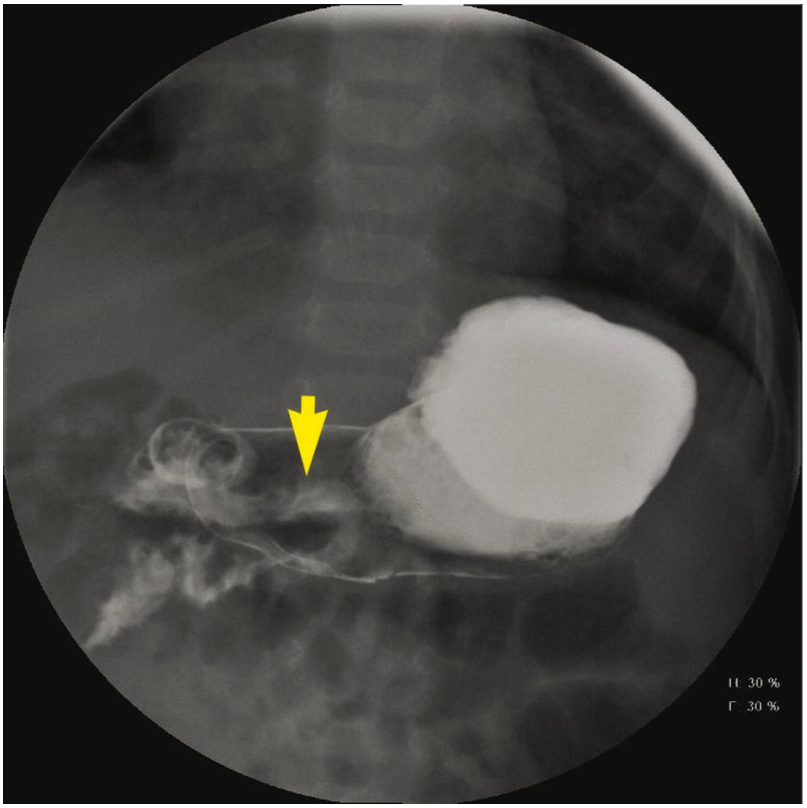


Anteroposterior radiograph of the abdomen shows dilated loops of bowel in the upper abdomen but no gas distally, findings that are consistent with malrotation and midgut volvulus.

Courtesy of D. Mulvihill

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Critique: 83

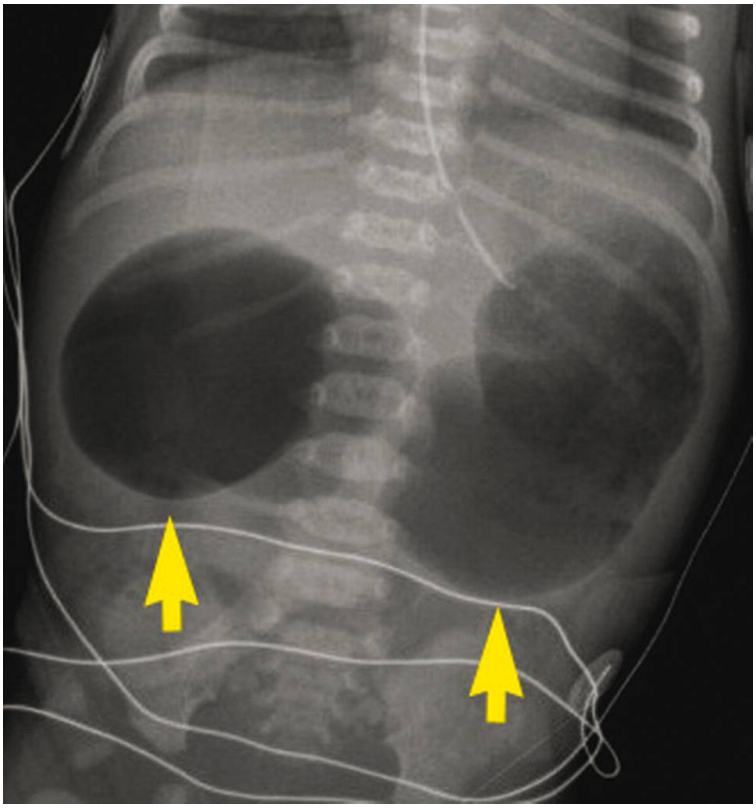


Upper gastrointestinal radiographic series demonstrating malrotation. The duodenal sweep (arrow) does not extend to the left of the midline before turning to the right, where most of the duodenum is positioned.

Courtesy of D. Mulvihill

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Critique: 83



Abdominal radiograph demonstrating the "double-bubble." In this case, the finding results from duodenal atresia. The appearance is due to a distended, gas-filled stomach (right arrow) and proximal duodenum (left arrow).

Courtesy of D. Mulvihill

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Question: 84

A worried mother brings her 18-month-old son to the emergency department because of a rash that developed today. She reports that he has had a runny nose and temperature to 104ºF (40°C) for the last 3 days, but the fever resolved yesterday. His immunizations are up to date. On physical examination, he appears well, is afebrile, and has diffuse 2- to 3-mm erythematous macules and papules over his trunk (Item Q84).

Of the following, the MOST likely cause of the rash is

A. adenovirus

B. Coxsackievirus

C. Epstein-Barr virus

D. human herpesvirus 6

E. parvovirus B19

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Question: 84



Courtesy of D. Krowchuk

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Critique: 84 Preferred Response: D

Roseola infantum is a common exanthem caused by human herpesvirus 6 or 7. The classic presentation is fever, which can be as high as 104° to 105.8°F (40º to 41ºC), for 3 to 5 days, followed by a period of defervescence and rash. The typical rash is comprised of 1- to 5-mm pink macules and papules and is more pronounced on the trunk, as described for the boy in the vignette (Item C84A). The fever and rash usually do not occur simultaneously, in contrast to other viral exanthems, such as rubeola, varicella, and parvovirus. Other clinical features of roseola infantum are febrile seizures during the first stage; cervical and occipital lymphadenopathy; bulging fontanelle and encephalopathy; and such nonspecific symptoms as vomiting, diarrhea, and nasal congestion.

Diagnostic tests to confirm the diagnosis are not widely available, so the diagnosis usually is made clinically. Treatment is supportive and consists of fluids and antipyretics.

Adenovirus causes symptoms in the upper respiratory tract (eg, rhinitis, pharyngitis), ear (otitis media), eyes (conjunctivitis), and gastrointestinal tract (diarrhea, vomiting). Rash is not a typical feature of the infection. Coxsackievirus is an enterovirus that usually causes herpangina, conjunctivitis, or hand-food-and-mouth syndrome (Item C84B). Children infected with Epstein-Barr virus either may experience a nonspecific febrile illness or develop signs and symptoms of infectious mononucleosis, including lymphadenopathy, hepatosplenomegaly, and fatigue. Rash may occur, usually developing after administration of amoxicillin or ampicillin (Item C84C). The skin findings associated with parvovirus B19 infection (Fifth disease) are erythema of the cheeks (slapped cheek appearance) followed by a maculopapular or reticular rash over the rest of the body (Item C84D).

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Critique: 84



The eruption of roseola is composed of pink-to-erythematous macules and papules.

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Critique: 84



In hand-foot-and-mouth disease, children develop oval vesicles surrounded by a rim of erythema. Lesions are concentrated on the hands and feet.

Courtesy of D. Krowchuk

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Critique: 84



Patients who have infectious mononucleosis may develop a morbilliform eruption if they receive amoxicillin or ampicillin.

Courtesy of D. Krowchuk

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Critique: 84



Erythema infectiosum is characterized by an erythematous, reticulated eruption located on the extremities or trunk.

Courtesy of D. Krowchuk

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Question: 85

A 10-year-old boy who has moved to your practice recently has a history of frequent sore throats for which he has received antibiotics. His mother is frustrated with the recurrences and wants his tonsils removed. The child reports a runny nose, mild cough, and abdominal pain. Findings on physical examination include a temperature of 100.3°F (38°C), conjunctival erythema, enlarged tonsils with some erythema and exudate, and vesicular lesions on the soft palate. There is no cervical adenopathy or rash. Results of a rapid streptococcal antigen test are negative.

Of the following, the MOST likely diagnosis is

A. adenovirus infection

B. Coxsackievirus infection

C. mononucleosis

D. sinusitis

E. streptococcal pharyngitis

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Critique: 85 Preferred Response: A

Exudative tonsillitis may be viral or bacterial. Important bacterial causes include group A beta-hemolytic *Streptococcus* (GABHS) (Item C85A) and *Staphylococcus aureus* (which is usually a superinfection rather than a primary infection). Viral infections represent 70% of exudative pharyngitis in children. Important pathogens include adenovirus, influenza A, Epstein-Barr virus, and herpes simplex viruses. In most settings, the preferred test for diagnosing GABHS pharyngeal infection in a child who has exudative pharyngitis is a rapid streptococcal antigen test. However, because the sensitivity of these tests varies from 70% to 90%, a negative rapid test result should be confirmed by throat culture.

Clinical criteria for diagnosing GABHS pharyngitis have been developed. For children who have four of the clinical Centor criteria for GABHS pharyngitis—history of fever, pharyngeal exudates, cervical lymphadenopathy, and absence of cough—90% of cultures for GABHS are positive.

Although the boy in the vignette has pharyngeal exudates, he has a cough, rhinorrhea, and low-grade fever, symptoms likely to be associated with a viral upper respiratory tract infection rather than GABHS pharyngitis. In addition, he has no lymphadenopathy, and results of the rapid streptococcal antigen test are negative**.**

Antibiotic treatment of streptococcal pharyngitis reduces the risk of glomerulonephritis and rheumatic fever and remains the standard of care. In developed countries, the risks of antibiotic complications and resistance may outweigh the benefits. Antibiotics also seem to reduce the duration of sore throat by about 16 hours. However, the empiric use of antibiotics for all cases of pharyngitis, even those that include exudates, without confirmatory laboratory testing for streptococcal antigen is not advised and may contribute to increasing antibiotic resistance.

The child described in the vignette has conjunctival erythema (Item C85B), which is classic for adenoviral infection. Although Coxsackievirus may cause vesicles on the palate (Item C85C), it generally does not produce conjunctivitis. Sinusitis typically presents with facial pain in the older child or protracted rhinorrhea in the younger child. Infectious mononucleosis may produce an exudative pharyngitis, but typically patients do not manifest cough, conjunctivitis, or vesicles on the palate.

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Critique: 85



Streptococcal pharyngitis is characterized by tonsillar enlargement and exudates.

Courtesy of M. Rimsza

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Critique: 85

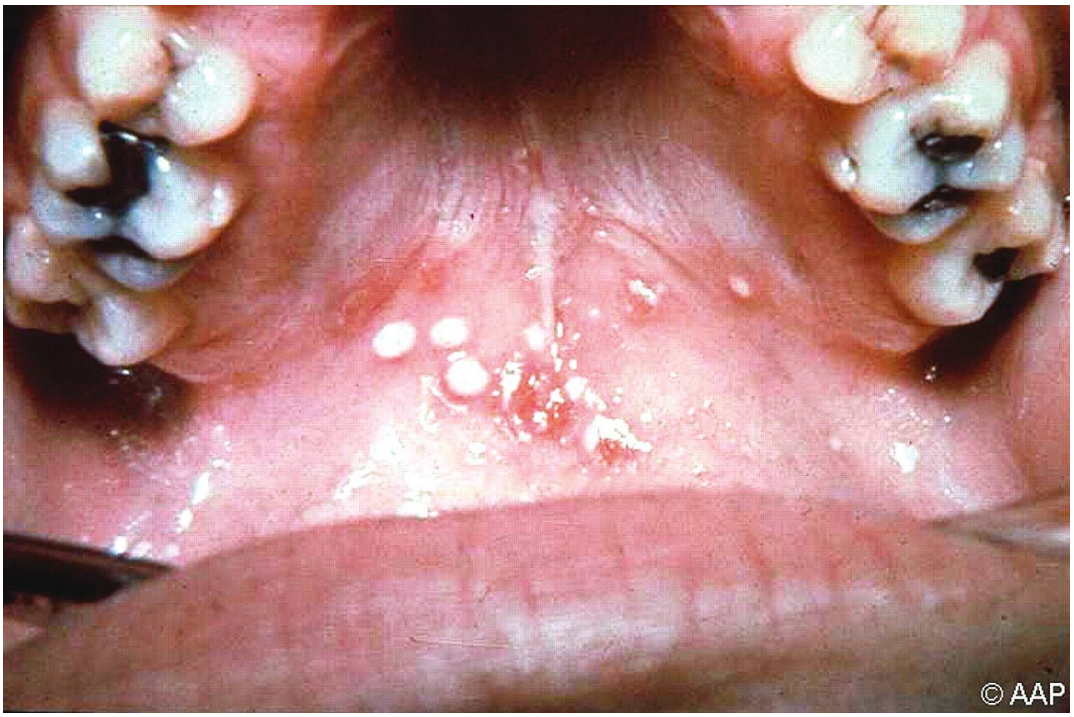


Adenovirus infection produces a follicular conjunctivitis.

Courtesy of *Red Book¨ Online*

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Critique: 85



In herpangina caused by Coxsackievirus infection, patients develop vesicles and ulcers in the posterior pharynx.

Courtesy of *Red Book¨ Online*

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Question: 86

A 5-day-old child is brought to the emergency department because he has been difficult to arouse over the last 6 hours. His parents report that he has not been interested in feeding today and that he has been breathing rapidly and with a grunting noise. On physical examination, the infant's heart rate is 185 beats/min, respiratory rate is 80 breaths/min, and blood pressure is 55/40 mm Hg. A pulse is palpable in the right brachial region, but not in the feet. All of his extremities are cool and mottled, with a capillary refill of more than 2 seconds.

Of the following, the MOST appropriate next step is to

A. arrange for echocardiography at the first appointment in the morning

B. initiate a furosemide infusion

C. initiate a prostaglandin infusion

D. obtain a computed tomography scan of the head

E. obtain a lumbar puncture

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Critique: 86 Preferred Response: C

The clinical presentation of the newborn described in the vignette suggests left heart obstruction with a closing ductus arteriosus. The discrepancy in pulses is consistent with a critical aortic coarctation.

Appreciation of the importance of the patent ductus arteriosus in maintaining systemic blood flow in the newborn who has severe left heart obstruction such as hypoplastic left heart syndrome (HLHS) and critical aortic coarctation requires a clear understanding of normal fetal shunting patterns. In the normal heart, the right atrium and right ventricle deliver desaturated blood to the organ of oxygenation. In the fetus, this organ is the placenta, and its fetal blood supply is via the umbilical artery that arises from the fetal descending aorta. The ductus arteriosus provides a fetal shunting pathway that allows the right side of the fetal heart to deliver desaturated blood to the organ of oxygenation by shunting this blood away from the high-resistance pulmonary arteries and into the descending aorta. This direction of flow occurs in part because fetal pulmonary vascular resistance (with fluid-filled developing lungs) is slightly higher than fetal systemic vascular resistance, and the placenta is a low-resistance circuit.

At birth, when the lungs expand with air and the placenta is removed from the circulation, pulmonary vascular resistance decreases and systemic vascular resistance increases. This leads to a reversal of flow across the ductus arteriosus (from the system into the pulmonary circuit). Over the subsequent hours and days, the ductus arteriosus begins the process of spontaneous closure.

The foramen ovale is an important fetal shunt that allows the relatively oxygenated blood returning from the placenta to cross from the right atrium into the left atrium prenatally. In so doing, the blood that has the highest oxygen content is directed to the coronary and cerebral circulations. Patency of the foramen in utero results from a slightly higher pressure in the right atrium than the left, because very little blood (~10% of the combined fetal cardiac output) returns to the left atrium from the lungs. The pressure difference “pushes” the flap of the foramen into the left atrium, creating the “hole” and allowing right-to-left blood flow. At birth, when the lungs expand and the entire cardiac output is directed into the lungs, pressure in the left atrium increases, rising slightly above that in the right atrium. When this occurs, the flap of the foramen is pushed back against the atrial septum and the “hole” is closed.

Left heart obstruction in the newborn often has a dramatic presentation that may include shock, cardiovascular collapse, and death if not recognized in a timely manner. HLHS and coarctation of the aorta are the two most common forms of left heart obstructive congenital heart disease that present in the first few postnatal days. HLHS is characterized by underdevelopment of the entire left side of the heart with a small or atretic mitral valve; a small, nonuseful left ventricle; and a small or atretic aortic valve. As a result, nearly all of the blood flow returning from the lungs is diverted to the right atrium through the foramen ovale because the left heart simply cannot accommodate it. The right heart then delivers blood to the lungs through the pulmonary arteries and to the system through right-to-left shunting across the ductus arteriosus. In severe aortic coarctation, the left heart is of sufficient size to handle the cardiac output returning from the lungs, but narrowing in the aortic arch leads to diminished flow to the distribution of the descending aorta and a progressive pressure load on the left ventricle. With patency of the ductus arteriosus, the right heart can provide blood flow to the descending aorta through right-to-left shunting across the ductus. In both HLHS and coarctation of the aorta, perfusion of the aorta with right ventricular output maintains systemic perfusion and minimizes the ischemia and subsequent metabolic acidosis that otherwise would ensue when the ductus arteriosus constricts.

In such cases, patency of the ductus arteriosus can be maintained by prostaglandins such as PGE1 administered as a continuous intravenous infusion. If the patency of the foramen ovale is not sufficient to maintain adequate decompression of the left atrium, a balloon atrial septostomy, a catheter procedure to enlarge the atrial communication, can be performed.

For the patient described in the vignette, the best management is to increase the systemic perfusion by maintaining the ductus arteriosus with the infusion of PGE1. Echocardiography is an important component of the diagnosis and management of patients who have suspected congenital heart disease, but its performance should not delay therapeutic options such as the

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initiation of the PGE1. Furosemide, a diuretic, has no beneficial role for the infant whose systemic perfusion is limited by a closing ductus arteriosus. Lumbar puncture and computed tomography scan of the head are not indicated as initial management.

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Question: 87

A 3-month-old infant presents to the emergency department with fussiness and decreased alertness. During triage, he experiences a seizure. Physical examination reveals somnolence and a bulging fontanelle. Emergent head computed tomography scan documents acute and chronic subdural hematomas (Item Q87).

Of the following, the procedure that is MOST likely to reveal the cause of these findings is

A. cerebral angiography

B. electroencephalography

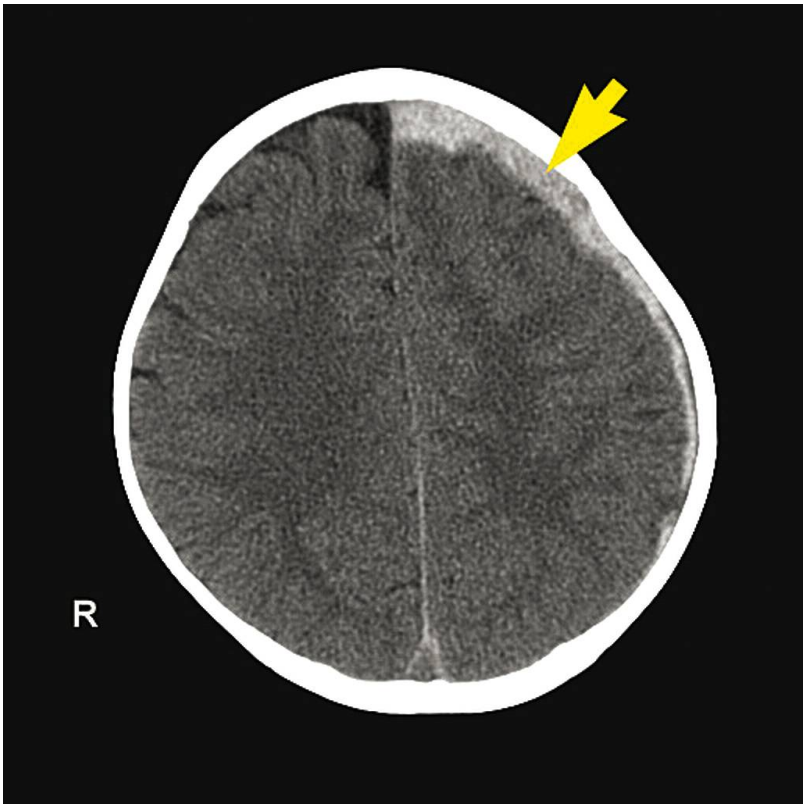
C. lumbar puncture

D. measurement of prothrombin time

E. retinal examination

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Question: 87



Computed tomography scan without contrast showing a subdural hematoma (arrow), as described for the child in the vignette.

Courtesy of D. Krowchuk

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Critique: 87 Preferred Response: E

Acute encephalopathy at any age typically indicates pathology in both cerebral hemispheres and mandates an emergent thorough examination. The irritability and somnolence reported for the infant in the vignette as well as the bulging fontanelle indicate increased intracranial pressure, likely due to a mass lesion. The noncontrast head computed tomography (CT) scan, the diagnostic modality of choice in the emergency department, shows chronic and acute subdural hematomas. Intracranial blood can trigger seizures, as reported for the infant. The most common cause of subdural hematoma in an infant is “shaken baby” or “shaken impact syndrome.” Retinal examination can identify retinal hemorrhages to support the diagnosis (Item C87), but the absence of retinal hemorrhages does not exclude shaken impact syndrome. Skeletal survey to look for fractures also is indicated in an infant when nonaccidental trauma is suspected.

Of note, although ultrasonography could be performed through the open fontanelle, head CT provides better spatial resolution and sensitivity to a wider variety of pathologic processes. Magnetic resonance imaging (MRI) also could be performed and may better delineate acute and chronic subdural hematomas and parenchymal hemorrhages in some instances, but it is not readily available in all emergency departments. In addition, in a medically unstable patient, the longer time required for MRI may increase risk.

Cerebral angiography involves imaging after injection of intravascular contrast into the arteries supplying the brain. Angiography and magnetic resonance angiography are useful in identifying vascular malformations, including aneurysms. However, subdural hemorrhages result from trauma to veins traversing the subdural space, making angiography unnecessary. Although lumbar puncture can be used to aid in the diagnosis of subarachnoid hemorrhage, which results from arterial blood, in this case, the subdural blood already is apparent on CT.

Measurement of the prothrombin time or other tests may be performed to evaluate for a clotting abnormality, a less common cause of intracranial hemorrhage. Babies who have clotting abnormalities are not likely to develop spontaneous subdural hemorrhages on multiple occasions. Electroencephalography (EEG) typically is not needed after an acute symptomatic seizure, although it is useful if there is ongoing concern about nonconvulsive status epilepticus contributing to the encephalopathy. EEG would not reveal the cause of the encephalopathy and subdural hemorrhages.

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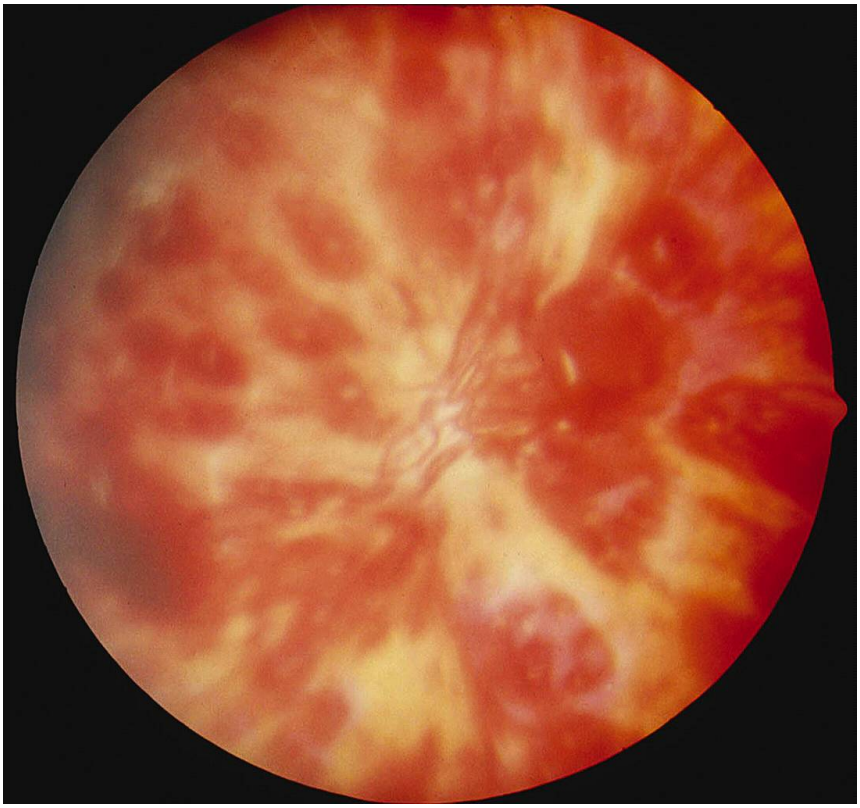
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Critique: 87



Retinal hemorrhages in an infant who was shaken.

Courtesy of R.G. Weaver, Jr

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Question: 88

You are treating a 16-year-old girl who has a history of generalized tonic-clonic seizures, the most recent of which occurred 6 months ago. She is followed by a neurologist who is treating her with phenylhydantoin with good results. During a routine physical examination, she confides that she is sexually active, and she requests a form of birth control. After some discussion, she asks to be started on oral contraceptive pills (OCPs).

Of the following, the MOST accurate statement about oral contraceptive use in this scenario is that

A. OCPs are contraindicated for use in women taking antiepileptics

B. only barrier methods should be recommended for this girl

C. the girl can be weaned off her antiepileptic medication at this time

D. the girl should be switched to a different antiepileptic before starting OCPs

E. the OCP estrogen dose should be higher than that usually prescribed

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Critique: 88 Preferred Response: E

It is important for physicians who care for women who have epilepsy to recognize factors that influence the choice of contraceptive method(s) in this population. Some antiepileptic drugs (AEDs) induce the cytochrome P450 hepatic enzyme pathway (phenobarbital, primidone, hydantoin, carbamazepine, oxcarbazepine). Such induction increases the metabolic rate of estrogen and the progestogens, potentially lowering serum concentrations of these hormones by 50% or more. Therefore, the use of hormonal contraceptives in women taking enzyme-inducing antiepileptic drugs (EIAEDs) must be considered carefully, and the dose of estrogen (and progestogen) should be increased. One recommendation is to prescribe only pills containing at least 35 mcg of ethinyl estradiol, although many women require a 50-mcg pill. Combined contraceptive patches are not suitable, and progestogen implants may not be appropriate because the dose of hormone they contain is insufficient for women receiving EIAEDs. The physician should research current best practices for prescribing hormonal contraceptives in women receiving AEDs because the practices are subject to change.

Of note, combined oral contraceptives reduce the concentration of lamotrigine by 40% to 60%, rendering this anticonvulsant ineffective if the problem is not anticipated and addressed.

It is critical to consider the potential impact of the choice of AED on the developing embryo and fetus. Women who require AEDs should discuss their choices with prescribing physicians prior to conception. Additionally, women taking AEDs should be made aware of preconceptional genetic counseling regarding recurrence risk of their seizure disorder to offspring, teratogenicity of AEDs, the importance of folic acid supplementation prior to and during pregnancy, and prenatal screening tests.

Although barrier methods of contraception are as effective for women who have epilepsy as for the general population, women who have epilepsy should not be confined to this choice.

If a patient has been seizure-free for at least 2 years and she does not have juvenile myoclonic epilepsy, consideration can be given to withdrawing the AED(s). However, this should only occur with the agreement and guidance of the prescribing physician. If this is not a viable option, the patient should be treated with the least teratogenic, lowest possible dose of AED that is efficacious and with monotherapy whenever possible.

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Question: 89

A 17-year-old boy comes to your office from a homeless shelter with complaints of a penile discharge. He denies pain with urination, skin rashes, or joint pain. He claims to use condoms most of the time. On physical examination, he is afebrile and has no skin rashes or testicular or epididymal tenderness or swelling, but he has copious purulent urethral discharge. Results of testing for sexually transmitted infections will not be available for 2 days.

Of the following, the MOST appropriate presumptive treatment is

A. acyclovir 400 mg orally TID for 7 to 10 days

B. azithromycin 1 g orally in a single dose plus cefixime 400 mg orally in a single dose

C. benzathine penicillin 2.4 million units intramuscularly in a single dose

D. doxycycline 100 mg orally BID for 7 days plus clindamycin 300 mg twice a day for 7 days

E. metronidazole 2 g orally in a single dose

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Critique: 89 Preferred Response: B

Urethritis is characterized by inflammation of the urethra and may be caused by infectious and noninfectious agents. Young men who have urethritis commonly are asymptomatic or they may complain of a mucopurulent or purulent discharge, dysuria, or urethral pruritus. *Neisseria gonorrhoeae* and *Chlamydia trachomatis* are common infectious causes of urethritis, and both may be present. The Centers for Disease Control and Prevention (CDC) *Sexually Transmitted Diseases Treatment Guidelines*, *2006* recommend that all patients who have confirmed or suspected urethritis be treated for both gonorrhea and chlamydia. Further testing to determine the specific causative pathogens is recommended because both gonorrhea and chlamydia are reportable infections, and a specific diagnosis might enhance partner notification as well as treatment adherence. Culture, nucleic acid hybridization tests, and nucleic acid amplification tests can detect both of the organisms, and amplification tests can be performed on urine specimens.

Nongonoccocal urethritis (NGU) is diagnosed when microscopy shows inflammation without the presence of gram-negative intracellular diplococci. *C trachomatis* is the etiologic agent in 15% to 55% of cases, but the cause of most cases of nonchlamydial NGU is unknown. Other infectious agents implicated in NGU include *Ureaplasma urealyticum*, *Mycoplasma genitalium*, *Trichomonas vaginalis*, herpes simplex virus, adenovirus, and enteric bacteria (that might be associated with penetrative anal sexual activity). Diagnostic testing and treatment for these organisms are reserved for when specific infections are suspected (such as with known partner contact) or when NGU is not responsive to therapy.

Recommended treatment for urethritis includes a single 1-g oral dose of azithromycin or doxycycline 100 mg orally twice a day for 7 days. Both are highly effective for chlamydial urethritis, but infections with *M genitalium* may respond better to azithromycin, and single-dose therapy is preferred both for adherence and for the ability to conduct direct observed treatment. The recommended treatment for *N gonorrhoeae* is ceftriaxone 125 mg intramuscularly or cefixime 400 mg orally as single-dose therapy. Because the young man in the vignette is homeless and diagnostic testing results will be unavailable for several days, empiric treatment of his urethritis is advisable. Because of increasing fluoroquinolone resistance in the United States and Canada, the CDC no longer recommends using these antimicrobials for the treatment of uncomplicated gonococcal urethritis and advises that all patients be treated with either cefixime or ceftriaxone, regardless of the patient's travel history or sexual behavior. Acyclovir, benzathine penicillin, clindamycin, or metronidazole is not recommended for empiric therapy.

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Question: 90

You are examining a newborn and discover that her genitalia appear abnormal. There is an enlarged clitoris (1.5 cm in length) (Item Q90) and a small amount of posterior labial fusion. Amniocentesis was performed during pregnancy, and the karyotype was 46,XX.

Of the following, the maternal history is MOST likely to reveal exposure to

A. danazol

B. ethinyl estradiol

C. fluorocarbons

D. organophosphate pesticides

E. progesterone in oil

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Question: 90



Clitoromegaly, as described for the child in the vignette.

Courtesy of M. Rimsza

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Critique: 90 Preferred Response: A

Danazol, used to treat endometriosis, is a modified progestogen that has an androgenic effect. It has produced virilization in female fetuses. Exposure after the first trimester can lead to clitoromegaly without midline fusion. Earlier exposure for more than 8 weeks after fertilization may cause labial fusion of varying degrees associated with clitoromegaly, as described for the newborn in the vignette.

Ethinyl estradiol is an estrogen commonly found in oral contraceptive agents. It has not been associated with masculinization of female fetuses. Fluorocarbons have been used to develop potential blood substitutes. Teratogenic effects have not been described. Progesterone in oil is the natural progesterone and has not been associated with masculinization; only substituted synthetic progestins have this effect. Organophosphate pesticides cause acute cholinesterase inhibition and have been responsible for fetal death but not fetal anomalies.

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Question: 91

You are seeing a 9-year-old boy who is new to your practice for a health supervision visit. He is a sociable child who talks to you about his new school. You notice that he has repeated facial movements, eye blinking, and throat clearing. When you mention this to his parents, they admit to noting these repetitive mannerisms for more than 1 year, which they have attributed to a nervous habit. They ask you what these mannerisms may represent.

Of the following, your BEST explanation is that his

A. eye blinking is most likely a transient tic

B. facial grimacing is probably due to an autistic spectrum disorder

C. repetitive mannerisms are most likely due to Sydenham chorea

D. throat clearing is most likely due to allergies

E. vocal and motor tics are most likely due to Tourette syndrome

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Critique: 91 Preferred Response: E

The boy described in the vignette has Tourette syndrome (TS). There is no confirmatory laboratory test for this condition; the diagnosis is based on a set of clinical diagnostic criteria from the *Diagnostic and Statistical Manual of Mental Disorders* edition IV (DMS-IV). The TS Classification Study Group criteria are as follows:

- Both multiple motor tics (Item C91A) and one or more phonic tics must be present at some time during the illness, although not necessarily concurrently
- Tics must occur many times a day, nearly every day, or intermittently throughout a period of more than 1 year
- Anatomic location, number, frequency, type, complexity, or severity of tics must change over time
- Onset of tics before the age of 21 years
- Involuntary movements and noises must not be explained by another medical condition
- Motor tics, phonic tics, or both must be witnessed by a reliable examiner at some point during the illness or be recorded by videotape or cinematography

The prevalence rate of TS is 1 in 2,000. It is three to four times more common in boys than girls. Some 40% of affected children meet criteria for obsessive-compulsive disorder, many have impulsive behavior, and studies report that most have attention-deficit/hyperactivity disorder. The cause is not known but may be due to genetic, neurobiological, psychological, and environmental factors. The Tourette Syndrome Association offers support to parents of children who have received a TS diagnosis.

A number of other disorders may produce tics or abnormal repetitive movements. Sydenham chorea is a self-limited poststreptococcal condition. In contrast to tics, which are patterned, repetitive, and decreased during purposeful movement, chorea is nonpatterned, continuous, and increased with purposeful movement (Item C91B). Stereotypies are associated with mental retardation, autism, Rett syndrome, and blindness, but they also occur in otherwise normal children (Item C91C). Examples of stereotypies are arm flapping and rocking. The features of tics, chorea, and stereotypies are summarized in Item C91D (Item C91D).

Tics that last for more than 1 year, as described for the boy in the vignette, are not categorized as transient. A simple tic would be the presence of one specific motor mannerism and would not change in location, complexity, or severity over time. Complex tics (eg, finger tapping, jumping) may include combinations of movements of multiple body parts and appear more "purposeful" than simple tics. In view of the presence of both motor tics as well as vocal tics (throat clearing) it is unlikely that allergies would be the cause of these behaviors.

Children who have autism may have tics, but tics are not part of the diagnostic criteria for this disorder. The DSM-IV criteria for an autistic disorder include qualitative impairment in social interactions, qualitative impairment in reciprocal social communication, and restricted areas of interest (including self-stimulatory mannerisms). Because the boy in the vignette is socially interactive, autism is unlikely.

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Critique: 91

Item 91D: Signs and Symptoms of Tics, Chorea, and Stereotypies

	Tics	Stereotypies	Chorea
Appearance	Patterned, simple, or complex	Patterned, complex	Random movement fragments
Awareness	Younger children not aware; older children typically are aware and feel an urge to perform some of them	Younger children not aware; older children typically are aware and may enjoy them	Aware, but no urge to perform them
Clinical Course	Onset after age 3 years; episodic, may change location over months to years	Onset usually before age 3 years; episodic, daily, remains relatively unchanged for years	Onset at any age, persistence depends on cause
Suppressibility	Some ability to suppress, at least briefly, is common	Older children who are socially aware will suppress	Cannot suppress but can mask
Exacerbating Factors	Stress, excitement	Occur during times of happy excitement, boredom, anxiety, or when socially overwhelmed	Stress, certain postures or movements
Relationship to Voluntary Movement	Rarely occur during purposeful voluntary activities (sports, music)	Parents always can interrupt the behavior, but child may return to it	Typically highly focused voluntary activity worsens movements
Impact on Daily Living	Usually none	Usually none	Almost always

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Question: 92

A 13-year-old boy presents to the emergency department with a 3-day history of severe sore throat and fever. He is having trouble swallowing due to pain. His past medical history is unremarkable. On physical examination, he has a temperature of 102.6°F (39.3°C) and a large left tonsil, with swelling of the left soft palate and deviation of the uvula to the right (Item Q92). You suspect a peritonsillar abscess.

Of the following, the MOST appropriate antimicrobial agent to start empirically is

A. ampicillin

B. ceftriaxone

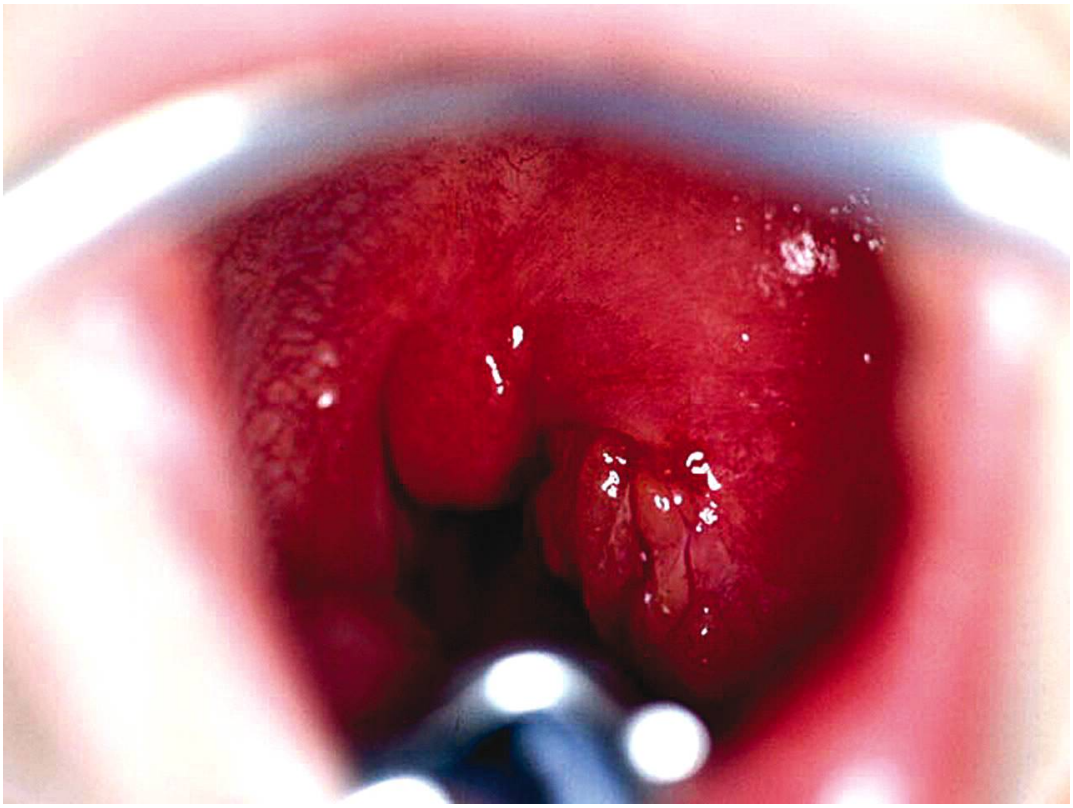
C. clindamycin

D. penicillin V

E. vancomycin

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Question: 92



Courtesy of the Media Lab at Doernbecher

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Critique: 92 Preferred Response: C

A peritonsillar abscess is caused by bacterial invasion through the tonsillar capsule into the surrounding tissue. The bacteria that are involved most commonly are group A *Streptococcus* and mixed oropharyngeal anaerobes. Physical examination usually reveals an asymmetric tonsillar bulge with a deviated uvula (Item C92), as described for the boy in the vignette. Treatment involves surgical drainage and antimicrobial therapy. Surgical drainage can be achieved through needle aspiration, incision and drainage, or tonsillectomy. Antimicrobial therapy must cover group A *Streptococcus* and oral anaerobes, and clindamycin is a good choice.

Although ampicillin, ceftriaxone, and vancomycin all cover group A *Streptococcus*, none of them covers anaerobes exceedingly well. Penicillin is an option, but the V variety is administered orally, and the initial therapy for peritonsillar abscess should be administered intravenously for a patient who has moderate systemic symptoms. After adequate surgical drainage, oral medications can be used. Combination penicillin (eg, ampicillin-sulbactam) also is a very good choice for empiric therapy.

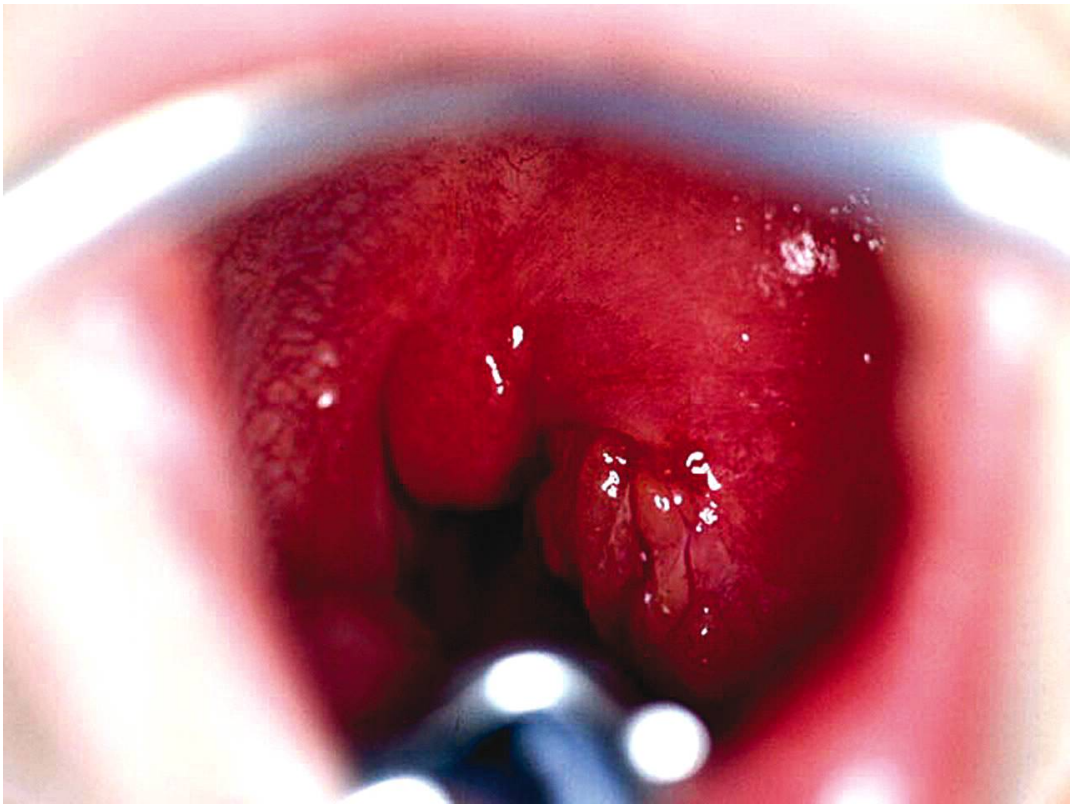
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Critique: 92



Peritonsillar abscess: A large left tonsil and swelling of the peritonsillar tissue that displaces the uvula to the right.

Courtesy of the Media Lab at Doernbecher

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Question: 93

You are evaluating a 5-month-old male adopted from a Somalian orphanage. According to his medical records, his biologic mother was treated for malaria during her last month of pregnancy and was suspected of having syphilis, based on a rapid plasma reagin (RPR) test result of 1:4, for which she received a course of treatment with an unknown oral antibiotic. Prior to arriving in the United States, the baby was found to have an RPR result of 1:2. Findings on physical examination of the infant today are completely normal.

Of the following, the MOST appropriate next step in management is to

A. obtain malaria thick and thin smears

B. perform polymerase chain reaction for treponemal DNA and repeat RPR

C. repeat RPR

D. repeat RPR and fluorescent treponemal antibody absorption test

E. treat the infant with intravenous penicillin

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Critique: 93 Preferred Response: D

The presumptive diagnosis of syphilis can be verified using two different types of serologic tests (nontreponemal and treponemal) that measure different types of antibody. To establish the diagnosis, the two types of serologic tests are used together.

The nonspecific nontreponemal reaginic antibody tests for syphilis include the Venereal Disease Research Laboratory (VDRL) slide test, the rapid plasma reagin (RPR) test, and the automated reagin test (ART). These tests measure antibody directed against lipoidal antigen from *Treponema pallidum*, antibody interaction with host tissues, or both. These inexpensive tests are performed rapidly and provide quantitative results that help to define disease activity and monitor response to therapy. They also are highly sensitive and, therefore, are the screening tests of choice.

Nontreponemal test results may be false-negative in patients who have early primary syphilis, latent acquired syphilis of long duration, or late congenital syphilis. Any reactive nontreponemal test result must be confirmed by one of the specific treponemal tests to exclude a false-positive result. False-positive results can be caused by certain viral infections (eg, Epstein-Barr virus, hepatitis, varicella, and measles), lymphoma, tuberculosis, malaria, endocarditis, connective tissue disease, pregnancy, abuse of injection drugs, laboratory error, or Wharton jelly contamination of cord blood specimens.

The specific treponemal tests measure antibodies against specific *T pallidum* antigens. The principal specific antitreponemal antibody tests in use are the fluorescent treponemal antibody absorption (FTA-ABS) and the *T pallidum* particle agglutination tests (*T pallidum* hemagglutination assay [TPHA] and microhemagglutination assay-*T pallidum* [MHA-TP]). These tests are used primarily to confirm a positive nontreponemal result. Once these tests yield a positive (reactive) result, the patient usually remains positive for life, even after successful therapy. The specific treponemal test antibody titers correlate poorly with disease activity and should not be used to assess response to therapy. These tests are not 100% specific for syphilis; positive reactions may be seen in patients who have other spirochetal diseases (eg, yaws, pinta, leptospirosis, rat-bite fever, relapsing fever, and Lyme disease).

The patient described in the vignette was born to a mother who had a positive nontreponemal test result that was not confirmed with a specific treponemal test. The infant has a positive nontreponemal test result of unclear etiology. The RPR should be repeated and a confirmatory specific treponemal test performed before instituting treatment with penicillin. The patient has no signs or symptoms consistent with malaria; accordingly, obtaining thick and thin blood smears is not indicated. Polymerase chain reaction for treponemal DNA currently is available only in a research setting.

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Question: 94

A 6-year-old boy presents with cola-colored urine. His mother reports that he had a sore throat 10 days ago. On physical examination, his blood pressure is 136/88 mm Hg, and he has mild swelling of the face and lower extremities.

Of the following, the MOST likely laboratory finding is

A. low C3 complement value

B. normal urinalysis results

C. positive antineutrophil cytoplasmic antibody titer

D. positive antinuclear antibody titer

E. positive urine culture

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Critique: 94 Preferred Response: A

The findings of cola-colored urine, swelling, and hypertension described for the boy in the vignette suggest the diagnosis of acute glomerulonephritis. The “sore throat” 10 days earlier makes acute poststreptococcal glomerulonephritis (APSGN) the most likely diagnosis. The initial assessment of a child in whom ASPGN is suspected must include measurement of blood pressure and serum creatinine to assess disease severity. Both severe hypertension and renal failure can occur as part of a rapidly progressive glomerulonephritis. After initial assessment, the most important diagnostic test is measurement of complement component 3 (C3) to confirm the presence of hypocomplementemia, which occurs in more than 90% of cases.

ASPGN is an immune complex-mediated glomerulonephritis that follows an infection by a nephritogenic strain of group A beta-hemolytic *Streptococcus* of the pharynx or skin. The interval between pharyngitis and the development of APSGN is approximately 1 to 2 weeks. In contrast, the latency period between a skin infection and ASPGN is 3 to 6 weeks. Most patients who have nephritis have a subclinical infection, which is estimated to occur four to five times more frequently than overt disease. APSGN in children is characterized by hematuria (100%), proteinuria (80%), edema (90%), hypertension (70%), and azotemia (33%). Thus, the urinalysis will not be normal. In addition, gross hematuria occurs in approximately 40% of children who have overt disease.

As noted previously, the characteristic laboratory feature of ASPGN is hypocomplementemia, which typically features depressed C3 and normal C4 values. The differential diagnosis of hypocomplementemic glomerulonephritis consists of membranoproliferative glomerulonephritis (MPGN) in a child who has disease limited to the kidney and systemic lupus erythematosus in a child who has multisystem disease. Rarer causes of hypocomplementemic glomerulonephritis include subacute bacterial endocarditis, shunt nephritis (in patients who have ventriculoatrial shunts), and essential mixed cryoglobulinemia.

Treatment of ASPGN is typically supportive and aims to reverse the sodium and fluid retention through the use of diuretics accompanied by restriction of sodium and fluid. Vasodilators also may be used for patients who have severe hypertension. Antibiotics can reduce the risk of transmission of the nephritogenic strain of streptococci to close contacts.

The key follow-up test is a repeat measurement of C3, which usually normalizes within 8 weeks. Patients in whom depression of C3 persists after 12 weeks may require a renal biopsy to rule out MPGN. The prognosis of ASPGN is excellent. Gross hematuria and hypertension usually resolve within a few weeks and proteinuria within a few months. Microscopic hematuria may persist for 1 to 3 years.

A normal urinalysis result is inconsistent with any form of glomerulonephritis, and would, therefore, be highly unlikely in a child who has hypertension and cola-colored urine. A positive urine culture would be unexpected in this clinical scenario. Patients who have hemorrhagic cystitis typically have bright red blood in the urine, often accompanied by clots. Other pertinent parts to the history that are absent in this scenario would be symptoms of dysuria, abdominal pain, frequency, urgency, and possibly fever.

Small vessel vasculitides such as Wegener granulomatosis, microscopic polyangiitis, and Churg Strauss disease are less common causes of glomerulonephritis and, therefore, anti-neutrophil cytoplasmic antibody testing is unlikely to be revealing. Another, less likely diagnostic possibility for the child in the vignette is lupus nephritis. However, anti-nuclear antibody should be measured in the setting of acute nephritis to exclude this possibility.

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Question: 95

A 5-month-old female infant presents with a 4-day history of increasing lethargy, drooling, constipation, and a weak cry. The parents report that the infant had been very healthy until this past week; she had no preceding sick contacts, viral symptoms, or fever. The infant is bottle fed and over the past month has been introduced to various home-pureed fresh fruits and vegetables, although the parents deny giving her any honey or homemade canned products. They currently have two dogs and a cat and live in the northwestern United States in a new subdivision that is still under construction. On physical examination, the infant is afebrile but has symmetrically decreased tone, absent deep tendon reflexes, poor muscle strength, and a weak cry.

Of the following, the MOST likely diagnosis is

A. botulism

B. diphtheria

C. lead poisoning

D. meningitis

E. tetanus

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Critique: 95 Preferred Response: A

The diffuse and progressive weakness described for the infant in the vignette is characteristic of infant botulism. Infant botulism usually occurs in infants younger than 6 months of age and is believed to be due to the inability of normal intestinal flora to prevent colonization by *Clostridium botulinum*. Although homemade canned food products and honey are common causes, spores found in soil and released by construction may be sufficient to result in clinical symptoms. Therapy is primarily supportive. Infants should be monitored closely because up to 50% can develop restrictive pulmonary disease, respiratory distress, or respiratory failure.

Weakness may follow diphtheria infection, but it typically is confined to the muscles of the pharynx and larynx. Diphtheria infection also may cause serosanguineous rhinitis, fever, and oropharyngeal edema, which were not present in the infant in the vignette.

Lead poisoning can cause a variety of clinical manifestations, including no symptoms, behavioral changes, gastrointestinal symptoms, lethargy, seizures, and death. The primary source in the United States remains lead-based paint, which would be unlikely in a newly constructed house.

Meningitis is an acute infection of the central nervous system. Symptoms vary, depending on the pathogen and host, but they typically include fever, vomiting, neck pain, or in more severe cases, obtundation, coma, seizures, or focal neurologic deficits. The lack of fever and the gradual progression of symptoms over 4 days described for the infant in the vignette make meningitis less likely.

In contrast to botulism, tetanus is a paralytic disease that results in stiffness, muscle spasm, and rigidity. One of the most common forms worldwide is neonatal tetanus, which typically is due to inappropriate or poor umbilical stump care.

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Question: 96

The mother of one of your patients calls you because she has found an engorged tick on her daughter's scalp. She wants to know how to remove it safely.

Of the following, the MOST appropriate method of tick removal is to

A. cover the tick with petroleum jelly

B. hold an ice cube to the tick

C. scrape across the skin surface with a credit card

D. touch the tick with a hot match tip

E. use tweezers to pull the tick from the skin surface

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Critique: 96 Preferred Response: E

The fundamental goals of tick removal are to: 1) Remove the tick as quickly as possible because prolonged duration of attachment is associated with increasing likelihood of disease transmission, 2) Remove all parts of the tick so that local irritation or foreign body reactions are prevented, and 3) Use a removal technique that does not encourage the tick to regurgitate gastric contents that may contain infectious material into the patient. Anecdotal reports in the literature espouse a wide variety of tick removal methods, and a randomized trial in an animal model (sheep) has compared a number of methods. Although the evidence in the literature supporting one method over another is weak, the consensus is that the safest and most effective method is to grasp the tick at the skin using angled mosquito forceps or splinter tweezers and remove it using gentle, steady traction (Item C96). Following removal, the area should be cleaned with soap and water. If mouth parts remain embedded, medical attention should be sought for removal.

Covering the tick with petroleum jelly or other occlusive materials to cause suffocation does not hasten detachment because a tick only breathes 3 to 15 times per hour.

Application of a hot match tip or an ice cube or subcutaneous injection of lidocaine to promote detachment through tick irritation also has been shown to be ineffective. In addition to putting the child at risk for a burn from the match, these methods may cause the tick to regurgitate.

Scraping across the skin with a credit card is a safe and effective method of removing the stinger after a hymenoptera sting, but it is likely to destroy the tick without removing the embedded mouthparts.

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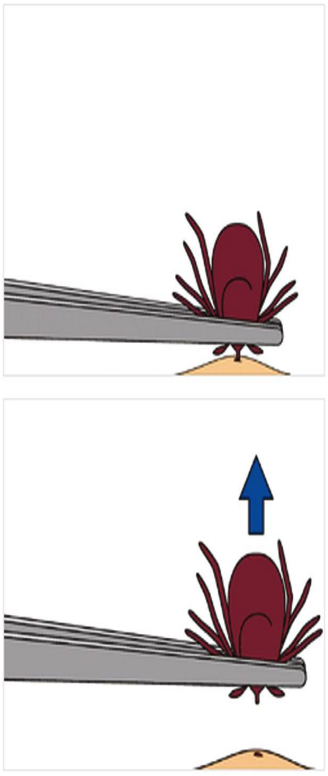
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Critique: 96



Proper tick removal: Using blunt forceps, grasp the tick as close to the skin as possible (top). Use traction perpendicular to the skin surface to remove the tick (bottom).

Courtesy of the Centers for Disease Control and Prevention

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Question: 97

A 14-year-old boy requests treatment for his acne. He is using no medications and has no known drug allergies. Physical examination of the face reveals a few small inflammatory papules and numerous blackheads and whiteheads; there is no scarring (Item Q97). No acne lesions are present on the chest and back.

Of the following, the MOST appropriate treatment is

A. benzoyl peroxide topically

B. benzoyl peroxide topically and tetracycline orally

C. benzoyl peroxide topically and tretinoin topically

D. clindamycin topically

E. tretinoin topically

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Question: 97



Courtesy of D. Krowchuk

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Critique: 97 Preferred Response: C

Acne affects 85% or more of adolescents and is the skin disorder most often treated by physicians. Multiple factors contribute to acne, including bacteria (that induce an inflammatory response, leading to erythematous papules and pustules (Item C97A)), androgens (that cause increased sebum production), and abnormal follicular keratinization (that causes blockage within pores, resulting in blackheads and whiteheads (Item C97B)). The treatment of acne depends on the types of lesions present (eg, blackheads/whiteheads, inflammatory, or both), the extent of disease (face versus face and trunk), and the severity of disease (more severe disease involves scarring and the presence of numerous and larger inflammatory lesions).

The patient described in the vignette has facial acne characterized by a few small inflammatory lesions and numerous obstructive lesions (ie, blackheads and whiteheads) (Item C97C). As a result, he requires treatment with two agents, one to control inflammatory lesions and one to control obstructive lesions. Because the inflammatory component of his acne is mild and limited to the face, it may be treated topically with benzoyl peroxide applied each morning. Alternatives might include a topical combination product (eg, benzoyl peroxide combined with either clindamycin or erythromycin) or possibly a topical antibiotic alone (although bacterial resistance to these agents when they are used without benzoyl peroxide is common).

Although benzoyl peroxide may be of some benefit in mild comedonal acne, the numerous lesions described for the patient indicate the need for a topical retinoid (eg, tretinoin or adapalene). Beyond this, many experts believe that topical retinoids are an essential part of any acne treatment program (regardless of lesion type observed) because they normalize the keratinization process and prevent new lesion formation. To avoid skin irritation, the patient is advised to apply the medication sparingly (a pea-sized amount is sufficient to cover the entire face) and use it every second or third night, progressing to nightly application as tolerated. Benzoyl peroxide inactivates tretinoin and, therefore, should not be applied simultaneously (ie, it is applied in the morning and tretinoin at night).

Systemic antibiotics such as tetracycline, erythromycin, doxycycline, minocycline, and others are indicated in the management of severe or widespread inflammatory acne or inflammatory disease that does not respond to appropriate topical therapy. Using topical clindamycin alone would not address the obstructive component of the patient's disease, just as tretinoin used alone may be insufficient to control inflammatory lesions.

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Critique: 97



Inflammatory lesions of acne are erythematous papules and pustules.

Courtesy of D. Krowchuk

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Critique: 97



Blackheads and whiteheads (skin-colored or white papules) are evidence of follicular obstruction in acne.

Courtesy of D. Krowchuk

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Critique: 97



Mixed facial acne that includes a few inflammatory lesions and multiple blackheads and whiteheads.

Courtesy of D. Krowchuk

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Question: 98

A 16-year-old girl who has juvenile idiopathic arthritis (juvenile rheumatoid arthritis) presents with a history of chronic anemia. Her current medications include ibuprofen, methotrexate, and folic acid. Physical examination demonstrates tenderness to palpation of the upper abdomen and a left knee effusion. The stool is positive for occult blood. Results of laboratory studies include a hematocrit of 28% (0.28), mean cell volume of 72 fL, reticulocyte count of 1%, white blood cell count of 4.2x103/mcL (4.2x109/L), serum iron concentration of 14 mcg/dL (2.5 mcmol/L), and total iron binding capacity of 400 mcg/dL (71.6 mcmol/L). Transaminases, bilirubin, amylase, and lipase values are normal. You initiate iron therapy.

Of the following, the MOST appropriate diagnostic test at this time is

A. abdominal ultrasonography

B. abdominal spiral computed tomography scan

C. mesenteric angiography

D. radionuclide tagged red cell study

E. upper endoscopy with biopsy

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Critique: 98 Preferred Response: E

The patient described in the vignette presents with iron deficiency anemia and guaiac-positive stool, which suggest the presence of inflammation and bleeding of the gastrointestinal tract. It is unlikely that she has an acute gastrointestinal hemorrhage because she is hemodynamically stable. The differential diagnosis of chronic gastrointestinal bleeding and anemia in this patient is broad and includes *Helicobacter pylori* infection, celiac disease, and Crohn disease. However, her chronic use of ibuprofen points toward gastritis and ulcer disease due to chronic use of nonsteroidal anti-inflammatory drugs (NSAIDs) (ibuprofen). Upper endoscopy is indicated to establish a diagnosis of NSAID gastropathy and to exclude other conditions.

Abdominal ultrasonography, abdominal spiral computed tomography scan, mesenteric angiography, and radionuclide tagged red cell study may be useful in the evaluation of large-volume gastrointestinal hemorrhages, but they lack the sensitivity to identify gastric or duodenal ulcers. Ideally, the ibuprofen should be held for 3 to 5 days prior to the upper endoscopy to reduce the risk of bleeding from biopsies. If ulcers are identified on upper endoscopy, the ibuprofen should be discontinued and the patient treated with a proton pump inhibitor (eg, omeprazole, lansoprazole).

NSAIDs are anti-inflammatory drugs that reduce the synthesis of prostaglandins. Nonselective NSAIDs (eg, aspirin, ibuprofen, and naproxen) work by inhibiting cyclooxygenase (COX) 1 and 2; COX2 inhibitors (eg, celecoxib) preferentially bind to COX2. The nonselective agents have a higher risk of gastrointestinal and renal toxicity, and the COX2 inhibitors have a higher risk of adverse cardiovascular events in adults. It is estimated that 1% to 4% of adults taking chronic NSAIDs develop ulcers annually. Peptic ulcer disease from NSAIDs most commonly presents with bleeding or anemia. If a patient requires chronic NSAID therapy and is at risk for gastrointestinal bleeding (eg, prior history of ulcer, taking corticosteroids or anticoagulants), medical options for ulcer prophylaxis include use of a proton pump inhibitor or the synthetic prostaglandin misoprostol.

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Question: 99

A 10-day-old infant who was born at 28 weeks' gestation has been weaned off of the ventilator and is receiving supplemental oxygen at an Fio2 of 0.30. He experiences episodic apnea 10 to 12 times daily and has corresponding bradycardia and oxygen desaturation, as measured by pulse oximetry (Item Q99). Some of the episodes require tactile stimulation and an increase in supplemental oxygen for resolution. Physical examination reveals no baseline tachypnea, tachycardia, or increased work of breathing. No heart murmur is appreciated, and he is normotensive. He appears acyanotic and well perfused, with a recent hematocrit of 35% (0.35). Results of head ultrasonography performed 3 days ago were normal.

Of the following, the BEST treatment for this infant is

A. continuous positive airway pressure

B. diaphragmatic (phrenic nerve) pacing

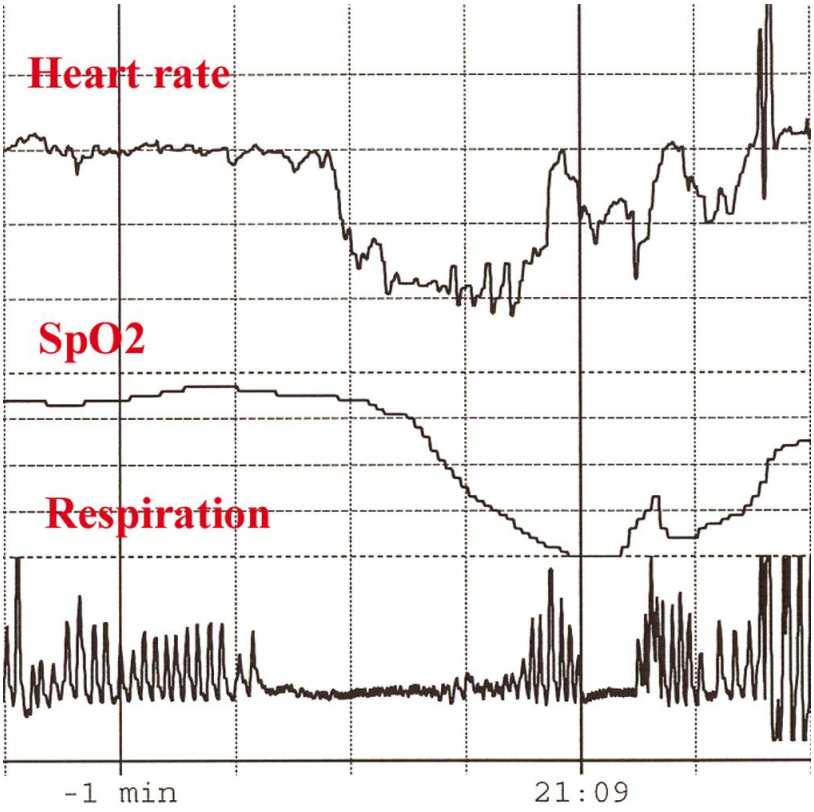
C. intravenous ampicillin

D. intravenous caffeine citrate

E. intubation and assisted ventilation

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Question: 99



Apnea, as described for the infant in the vignette: Respiration ceases for approximately 30 seconds and is associated with bradycardia and a decline in oxygen saturation.

Courtesy of B. Carter

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Critique: 99 Preferred Response: D

Apnea may be the most frequent respiratory problem in preterm infants not requiring assisted ventilation. Even in those treated with assisted ventilation and exogenous surfactant for respiratory distress syndrome, apnea may present as a clinical problem after weaning from the ventilator. The causes of apnea are protean (Item C99A) and must be evaluated individually in preterm patients. Immaturity of the respiratory control center is the most common underlying factor.

The preterm infant described in the vignette is extubated, experiencing apnea associated with bradycardia and arterial oxygen desaturation, and remains on supplemental oxygen. His cardiorespirogram tracing indicates a cessation of chest wall movement preceding bradycardia and desaturation (Item C99B). No history of gastroesophageal reflux is noted. There is no intracranial hemorrhage, profound anemia, or evidence for infection. These findings suggest the diagnosis of idiopathic recurrent apnea, which is treated best with intravenous caffeine citrate.

Continuous positive airway pressure may treat obstructive apnea effectively. Obstructive apnea is indicated by efforts of chest wall excursion on the tracing during the desaturation and bradycardia. Phrenic nerve pacing is invasive and reserved for patients who have congenital hypoventilation syndrome. Ampicillin is indicated for the treatment of sepsis, which is not seen in this patient. Apnea that is severe, associated with hypoxia or acidosis, and requiring resuscitation with bag-mask-valve positive pressure ventilation should be treated with intubation and assisted ventilation.

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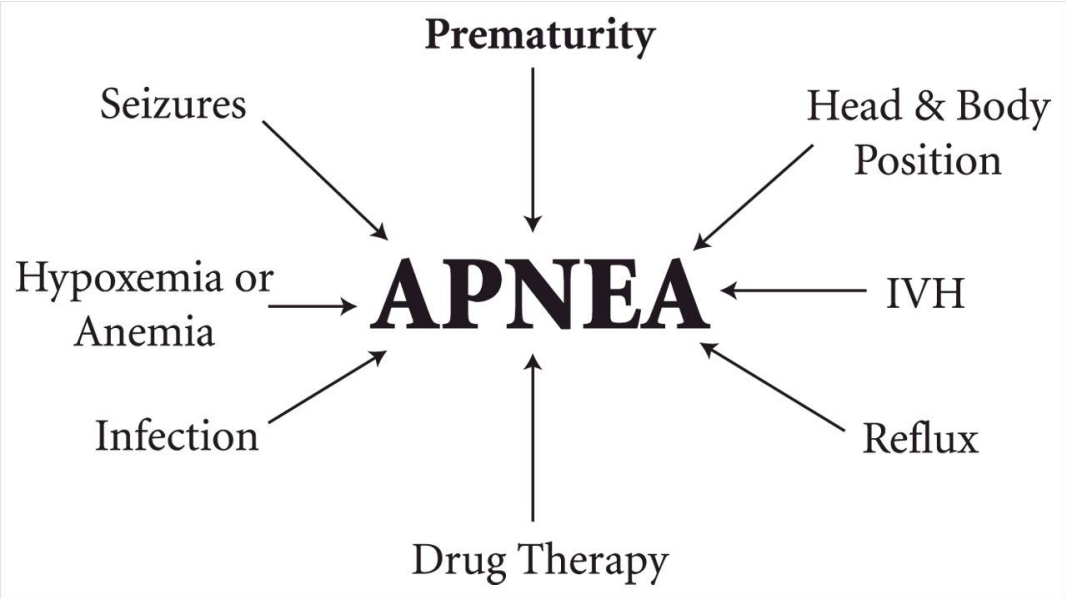
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Critique: 99

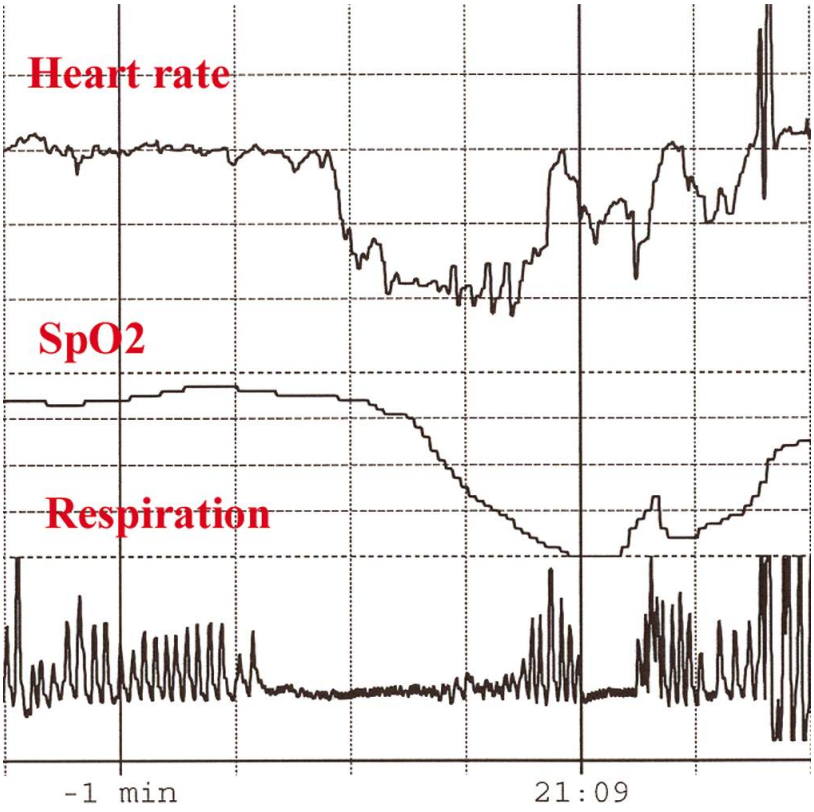


Multiple etiologic factors that may contribute to apnea in preterm infants.

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Critique: 99



Cardiorespirogram in central apnea: Cessation of respiration for approximately 30 seconds associated with bradycardia and a decline in oxygen saturation.

Courtesy of B. Carter

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Question: 100

You are evaluating a 10-year-old boy who just moved to the United States from Africa. He reports fever and abdominal pain of 2 weeks' duration. Physical examination reveals an ill-appearing boy who has a temperature of 103ºF (39.5°C), a respiratory rate of 40 breaths/min, hepatomegaly, and tenderness of the right upper quadrant. Abdominal ultrasonography shows a liver mass consistent with an abscess (Item Q100).

Of the following, the most likely etiologic agent is

A. *Ascaris lumbricoides*

B. *Entamoeba histolytica*

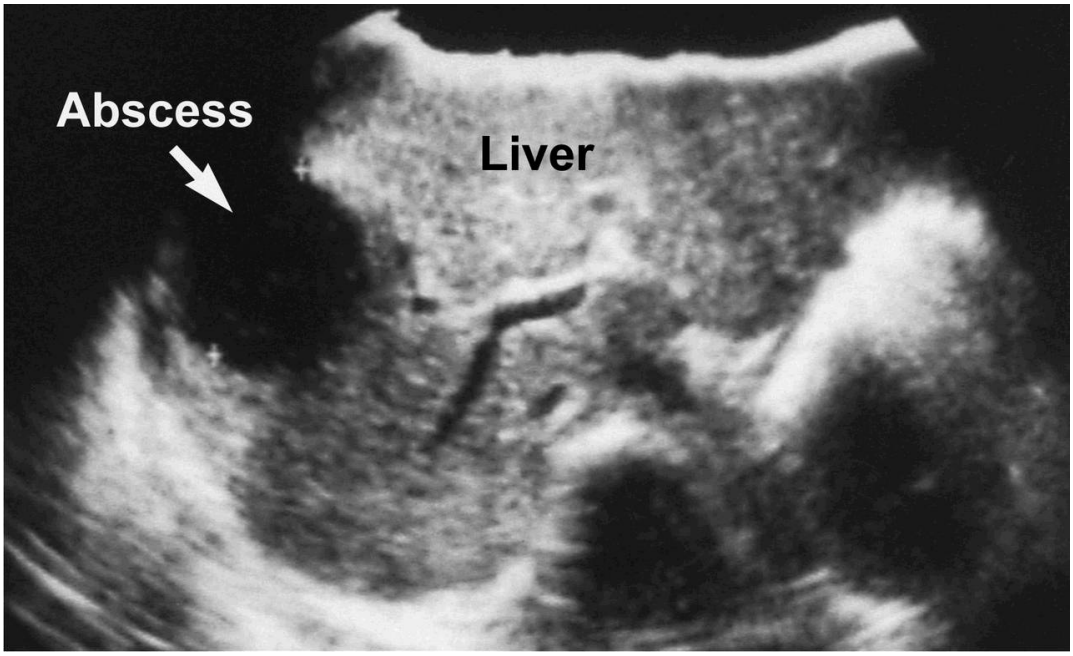
C. *Strongyloides stercoralis*

D. *Taenia solium*

E. *Wuchereria bancrofti*

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Question: 100



Transverse ultrasonographic image of the liver shows a large anechoic lesion in the right lobe consistent with an abscess.

Courtesy of D. Mulvihill

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Critique: 100 Preferred Response: B

Amebiasis, which is caused by infection with *Entamoeba histolytica,* can manifest as several clinical entities. Intestinal amebiasis causes weight loss, fever, and severe bloody diarrhea with lower abdominal pain, which may progress to fulminant colitis or toxic megacolon. Noninvasive intestinal infection results in vague, nonspecific abdominal complaints. Extraintestinal disease occurs most commonly as liver abscess, which causes fever, tachypnea, and tender hepatomegaly, as described for the boy in the vignette. Transmission occurs via ingestion of amebic cysts. The diagnosis is made by identification of cysts in stool or by serum antibody detection using indirect hemagglutination assay. Abdominal ultrasonography or computed tomography scan are used to diagnose liver abscess (Item C100A).

Ascariasis may cause pneumonitis and acute intestinal obstruction, but many patients are asymptomatic or only complain of mild nonspecific gastrointestinal symptoms. Common bile duct obstruction and cholangitis can occur, but liver abscess is atypical. *Strongyloides* infection can result in pneumonitis; gastrointestinal symptoms such as vomiting, diarrhea, and abdominal pain; and pruritic skin lesions. Infection with *Taenia solium* (tapeworm disease) is characterized by either a nonspecific gastrointestinal illness or neurocysticercosis (Item C100B), which may present with seizures. Filariasis, which may be caused by *Wuchereria bancrofti* or other nematodes, typically results in inguinal lymphadenitis, epididymitis, or orchitis and may progress to chronic lymphatic channel dysfunction (elephantiasis).

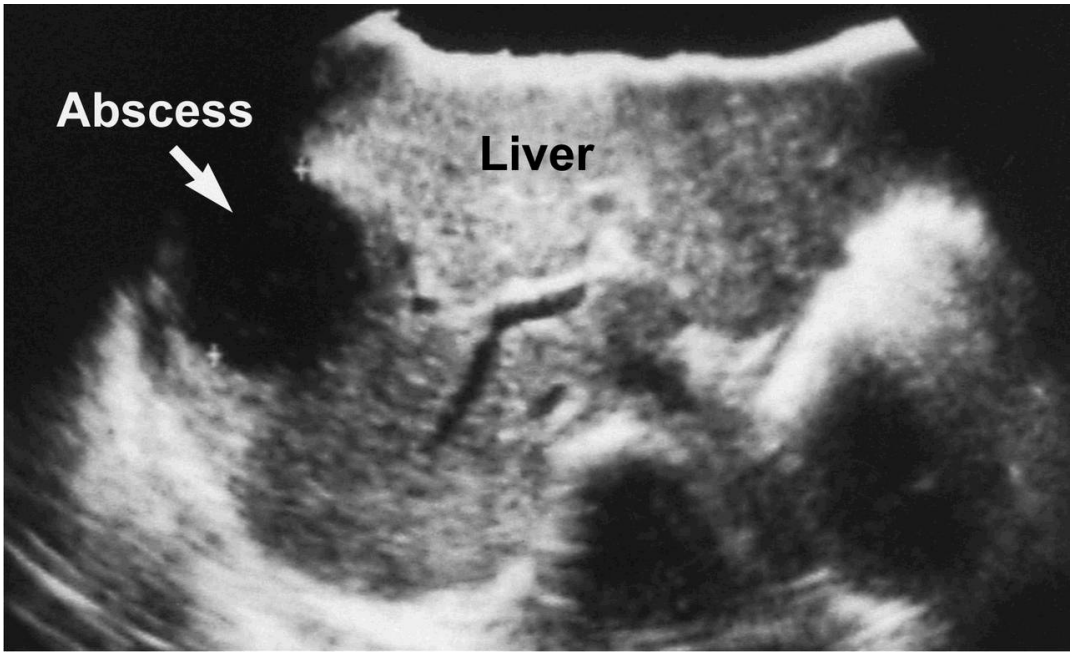
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Critique: 100

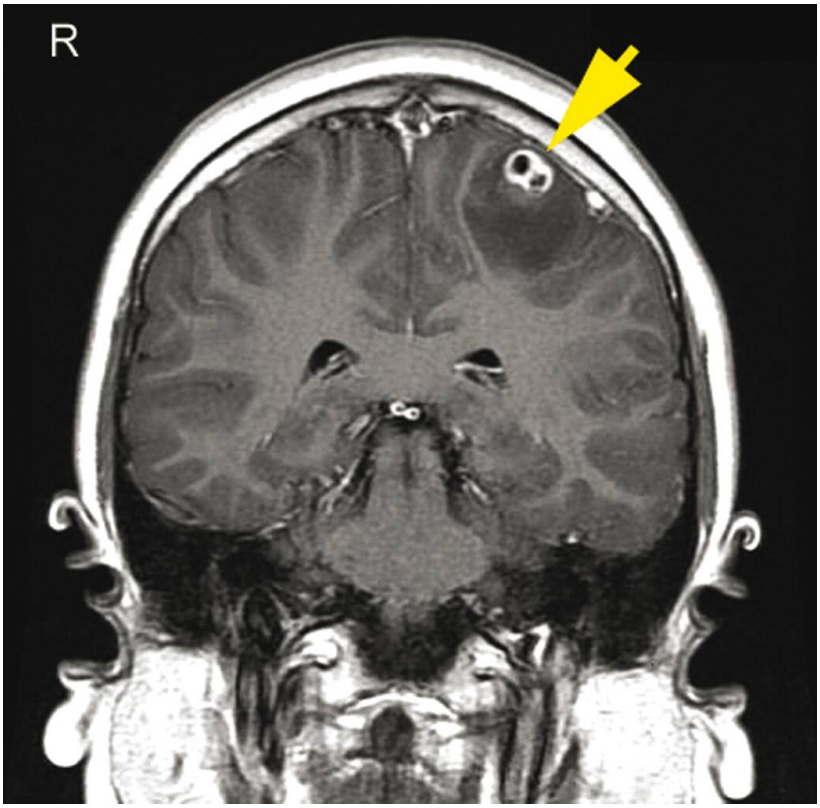


Transverse ultrasonographic image of the liver shows a large anechoic lesion in the right lobe consistent with an amebic abscess.

Courtesy of D. Mulvihill

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Critique: 100



Neurocysticercosis: Coronal T1-weighted magnetic resonance imaging following contrast demonstrates a 1-cm bilobed lesion in the parietal lobe that exhibits ring enhancement (arrow)t.

Courtesy of D. Gilbert

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Question: 101

A mother brings in her 6-month-old child for a health supervision visit. She explains that the family just moved from a rural area where they had obtained their water from a private well. In the past, she needed to give supplemental fluoride to her children beginning at age 6 months and wonders if this is necessary for this child.

Of the following, you are MOST likely to reply that

A. all children should receive fluoride supplementation starting at birth

B. children should not be supplemented if bottled drinking water is used

C. fluoride 0.25 mg/d supplementation should begin at age 6 months if there is less than 0.03 ppm in community water

D. once the baby's teeth erupt, fluoridated toothpaste will be sufficient

E. supplementation may stop when the child's deciduous teeth begin to fall out

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Critique: 101 Preferred Response: C

It is well established that the use of fluoride supplements in the diet and dental hygiene of children offers protection against the development of dental caries in both deciduous and permanent teeth. However, the challenge and controversy of providing fluoride in water without contributing to fluorosis and the difficulty with access to dental care for many children contribute to suboptimal fluoride use. Underuse of fluoride may contribute to the formation of dental caries in some children who may or may not have risk factors for nursing bottle caries and a family history of dental caries. Overuse of fluoride can result in “fluorosis,” (Item C101A) which includes white spots, pitting, or staining of teeth.

Decisions about fluoride use for children should be based on a child’s risk for caries. Individual and family risk factors include poverty, lack of access to dental care, non-white races, recent immigration, family history of dental caries, and residence in a community that does not have fluoridated water.

The Task Force on Community Preventive Service concluded that fluoridated water reduced tooth decay by 30% to 50% relative to what would be expected in those not consuming fluoridated water. Information about the fluoride content of the community water supply in the area to which the family in the vignette has moved should be available from the local health or water department. Fluoride supplementation may be prescribed accordingly (Item C101B).

Supplemental fluoride may be discontinued when the child reaches an age when fluoride varnish may be applied to the teeth (by a medical or dental professional) and when the child may use fluoridated toothpaste.

Guidelines from the American Academy of Pediatrics, published in 1995 and updated in 2002, lowered the recommended fluoride supplement dose for children younger than 6 years of age and eliminated supplementation for infants younger than 6 months of age. These changes were made in recognition of the risk of fluorosis related to excess fluoride exposure during permanent tooth development.

The period of highest susceptibility to fluorosis for permanent teeth is estimated to be between 15 and 24 months of age for boys and 21 and 30 months of age for girls. This is a good age for fluoride varnish application. Early toothbrushing (ie, before 2 to 3 years of age) with fluoride paste may lead to an unacceptable risk of fluorosis, especially in combination with the use of fluoridated water and fluoride varnish.

Children living in families who use exclusively bottled water for meal preparation and mixing formula and juice for their children should receive fluoride supplementation because most bottled water does not contain fluoride. However, the Centers for Disease Control and Prevention (CDC) have raised concern that the use of fluoridated water to prepare infant formula may be associated with an increased risk of fluorosis. The CDC has suggested that parents of infants fed primarily formula from concentrate or powder who are concerned about fluorosis consider the use of bottled water for formula preparation.

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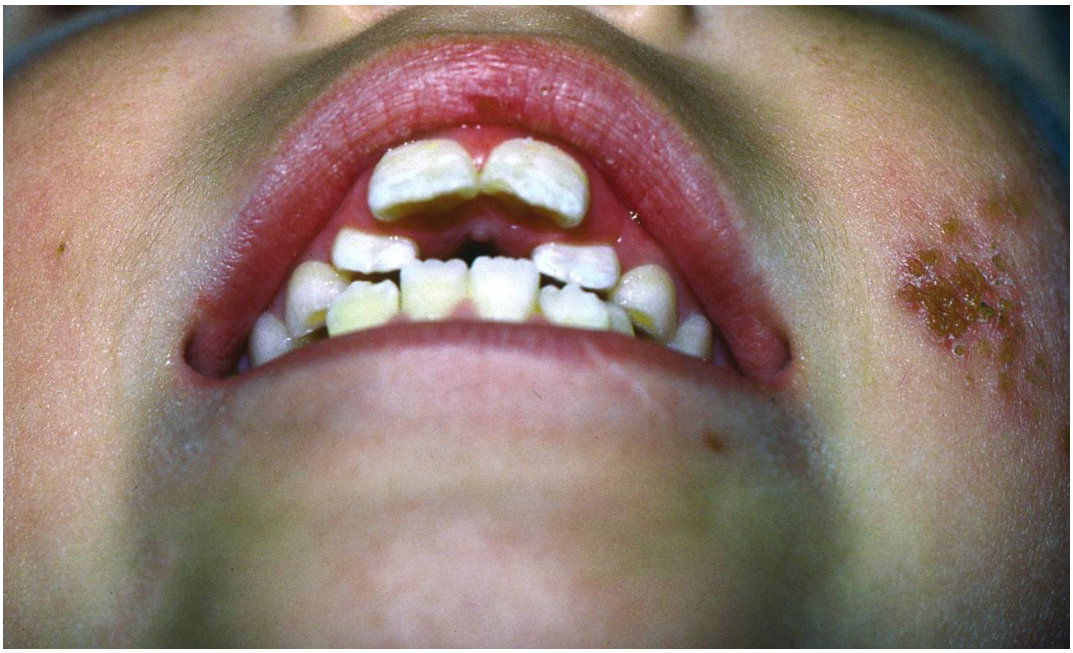
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Critique: 101



Fluorosis (mottled enamel) is characterized by white-to-brown patches on the teeth. In this child it is most evident on the maxillary central incisors.

Courtesy of M. Rimsza

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Critique: 101

Item 101B: AAP Current Fluoride Supplementation Schedule*

Age Range of Child	Drinking Water Fluoride Concentration		
	<0.3 ppm	0.3 to 0.6 ppm	>0.6 ppm
Birth to 6 mo	0	0	0
6 mo to 3 yr	0.25 mg	0	0
3 to 6 y	0.5 mg	0.25 mg	0
6 to 16 y	1.0 mg	0.5 mg	0

*Amount of supplemental fluoride recommended per day based on child's age and drinking water fluoride concentration.

Reprinted from Lewis CW, Milgrom P. Fluoride. *Pediatr Rev*. 2003;24:327-336

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Question: 102

A 6-month-old previously healthy girl is brought to your office because she has not been eating well today. The mother reports that the baby is interested in taking the bottle, but stops feeding within 1 minute and seems to have trouble breathing. She is irritable but consolable in her mother's arms. She is pale, afebrile, and has a respiratory rate of 70 breaths/min. Her heart rate is too fast to count, she has palpable pulses in all extremities, and her perfusion is fair, with a capillary refill time of 2 to 3 seconds.

Of the following, the MOST likely additional finding expected in this child is

A. crackles over the lungs

B. conjunctivitis

C. hepatomegaly

D. nuchal rigidity

E. rash on the extremities

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Critique: 102 Preferred Response: C

Arrhythmia encompasses any situation that leads to an abnormality in regular heart conduction and rate. Thus, the occurrence of an arrhythmia or cardiac dysrhythmia is relatively common in the pediatric population. At one end of the spectrum of dysrhythmias are the typically benign premature beats, which can occur from the ventricle (premature ventricular complex) or atria (premature atrial complex). When these occur in isolation, at rest, and with a single morphology, such events often are unnoticed and cause few, if any, symptoms. Arrhythmias that may be more serious can arise from either the atrial tissue or the ventricular muscle. Disorders of the former include supraventricular tachycardia, atrial ectopic tachycardia, atrial flutter, and atrial fibrillation; those of the latter include ventricular tachycardia and ventricular fibrillation.

Children and adolescents who experience atrial arrhythmias often complain of palpitation, with a sensation of a rapid, strong, or even irregular heart rate. If the rhythm persists, affected children may complain of shortness of breath, dizziness, light-headedness, or even syncope. In younger children and infants who may not be able to communicate effectively, such sensations may not become apparent until they lead to abnormalities of the hemodynamics that may result in pallor, diaphoresis, diminished perfusion, congestive cardiac failure, and even shock. Ventricular arrhythmias are less common in infants and children than they are in adolescents and adults and may be associated with inherited conditions such as the long QT syndrome, hypertrophic cardiomyopathy, and the Brugada syndrome. Ventricular arrhythmias are more likely to be malignant and to present with near-syncope, syncope, or even cardiac arrest.

The infant described in the vignette has a heart rate that is too fast to count and most likely suffers from an atrial arrhythmia such as supraventricular tachycardia. Because the heart rate in such disorders is typically greater than 240 beats/min, time for the ventricle to fill during diastole is greatly diminished. This leads to rising left atrial pressure, with subsequent pulmonary congestion as the pulmonary vessels face increased downstream pressure. The rising pressure in the pulmonary circuit leads to an increase in pressure on the right heart, which also faces the difficulty of diminished diastolic time with rising right atrial pressure. This is transmitted to the systemic veins and often is manifested on physical examination by distension of the jugular veins and hepatic congestion with hepatomegaly. The tachypnea that results from the pulmonary vascular congestion leads to difficulty feeding and may exacerbate the appearance of irritability. In addition, for children who have significant alteration of cardiac output resulting from the arrhythmia, irritability and lethargy may be the result of inadequate cerebral perfusion.

Children may manifest crackles over the lungs when in congestive heart failure, but this is not as typical a finding as it is in adults. One reason may be the relative inability of children to cooperate with deep and prolonged inspiration. Cardiac dysrhythmias would not present with conjunctivitis, nuchal rigidity, or rash, as might be expected with systemic or central nervous system infection.

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Question: 103

A 10-year-old boy in whom you diagnosed attention-deficit/hyperactivity disorder (ADHD) has been receiving stimulant medication daily since he was 7 years old. About 2 years ago, he developed persistent, repetitive throat clearing, and subsequently he has had waxing and waning motor and vocal tics that include eye rolling, grimacing, head bobbing, sniffing, and humming. For the past 2 months, his tics have worsened; he now has a loud squeak and head-jerking tic. His mother decided to stop his stimulant medication last week, fearing it might be causing his tics to worsen. Now he is much more hyperactive and is having more behavioral and attentional difficulties at school and home. His tics have not improved.

Of the following, the MOST appropriate treatment plan is to prescribe

A. carbamazepine to see if the movement disorders are seizures

B. daily penicillin to prevent tic exacerbations caused by streptococcal infections

C. haloperidol to treat the tics

D. methylphenidate again to treat the ADHD

E. sertraline to treat the tics

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Critique: 103 Preferred Response: D

The boy described in the vignette has a tic disorder characterized by multiple waxing and waning motor and vocal tics that have occurred for more than 1 year. Thus, he meets the diagnostic criteria for Tourette syndrome (Item C103). Most children who have Tourette syndrome also have at least one other diagnosis such as attention-deficit/hyperactivity disorder (ADHD) or an anxiety disorder such as obsessive compulsive disorder (OCD). This boy has ADHD and had been treated with psychostimulants with some benefit for 3 years. The decision to stop his stimulant medication, while understandable, was unwise because his symptoms have worsened since the medication was discontinued.

The severity of tics in Tourette syndrome tends to peak around age 10 years. Thus, the escalation in tics occurring in this boy may be part of the natural history of the disorder. The worsening of tics is not related to the use of psychostimulants, which he has taken for years. When parents and physicians discontinue stimulants in children who have Tourette syndrome and ADHD, the ADHD symptoms generally worsen, which leads to increased stress on the child. Tics do not improve and may worsen due to the increased stress.

Psychostimulants can precipitate or exacerbate tics in some individuals, and since the early 1980s, the United States Food and Drug Administration has mandated a warning statement that psychostimulants are contraindicated in Tourette syndrome. However, controlled clinical trials in the past 10 years have shown that psychostimulants improve ADHD symptoms in most children who have tics and ADHD and do not worsen tics. For the boy in the vignette, it is most appropriate to restart the psychostimulant because it was beneficial in the past and may significantly improve his ADHD symptoms.

Haloperidol reduces tics, but it is poorly tolerated. In one naturalistic study in Tourette syndrome, 80% of patients discontinued its use due to adverse effects. These adverse effects include weight gain, cognitive blunting, separation anxiety, and extrapyramidal symptoms, which can cause more problems than the tics. Alpha-adrenergic agonists such as guanfacine or clonidine are preferred first-line treatments for tic reduction, rather than typical neuroleptics (haloperidol, pimozide) or atypical antipsychotics (risperidone, olanzapine). Clonidine, given 3 to 4 times per day, has been shown specifically in a randomized, controlled trial in children who had ADHD and tics to reduce both ADHD and tic symptoms. However, sedation is a common adverse effect. Once the ADHD medication has been restarted, if the tic symptoms are causing substantial functional impairment, social impairment, or pain, medical treatment for tic suppression may be considered. The preferred approach is to add clonidine or guanfacine.

Sertraline and other selective serotonin reuptake inhibitors (SSRIs) do not directly diminish the presence of tics. If the child has untreated OCD or anxiety disorder, successful treatment with SSRIs or cognitive behavioral therapy may reduce stress and secondarily reduce tics.

In recent years, a large number of studies have explored a possible link between infections by group A beta-hemolytic streptococci (GABHS) and the explosive onset or exacerbation of tic or OCD symptoms. This has been termed Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS). The child described in the vignette did not have an explosive symptom onset or exacerbation and needs neither evaluation for GABHS exposure nor treatment with antibiotics.

There is no role for a trial of the antiseizure medication carbamazepine either to clarify a diagnosis of epilepsy or to reduce the frequency of tics. Tic disorder can be delineated from epilepsy accurately and reliably based on history.

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Question: 104

A 7-year-old patient who has Down syndrome is brought to the clinic by her mother, who is worried that the child has an increasingly abnormal gait and worsening clumsiness. At age 3 years, she was screened for cervical instability with flexion and extension cervical spine films, which showed normal results. On physical examination today, you note that she has an unsteady gait, and she has brisk deep tendon reflexes diffusely. These findings represent a significant change from 9 months ago when your neurologic examination showed only slightly diminished tone.

Of the following, the MOST likely cause of these symptoms and signs in a child who has Down syndrome is

A. cerebellar medulloblastoma

B. Chiari I malformation

C. leukemia involving the central nervous system

D. subluxation of the atlantoaxial joint

E. transverse myelitis of the cervical cord

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Critique: 104 Preferred Response: D

Atlantoaxial instability (AAI) is present in an estimated 15% of children (<21 years of age) who have Down syndrome (DS). Most affected individuals are asymptomatic, but an estimated 10% (1.5% of all who have DS) have symptoms relating to spinal cord compression.

The American Academy of Pediatrics (AAP) Committee on Genetics and the Down Syndrome Medical Interest Group recommend routine screening of all children who have DS between 3 and 5 years of age with lateral cervical radiographs in the neutral, flexed, and extended positions. Both groups note that screening may be a requirement for participation in the Special Olympics. The space between the posterior arch of C1 and the anterior segment of the odontoid process of C2 should be measured. Measurements of less than 5 mm are normal, 5 to 7 mm indicate instability, and more than 7 mm are markedly abnormal. However, this recommendation for routine screening for AAI remains controversial for a number of reasons.

In 1995, the Committee on Sports Medicine and Fitness of the AAP published a review of AAI in DS, highlighting several concerns regarding routine radiographic screening for AAI in children between 3 and 5 years of age. First, individuals who have DS and normal findings on screening may have abnormal radiographic results later in childhood. Conversely, some individuals whose study results initially are abnormal eventually have normalized results. Symptomatic AAI in DS is rare, raising the question of whether routine screening is necessary. Reproducibility of radiologic test results for AAI is poor in some studies. Asymptomatic AAI has not been shown to be a major risk factor for symptomatic AAI, and it is not clear that sports trauma is likely to precipitate symptomatic AAI. Finally, several publications report that individuals who have DS and AAI and symptoms or signs of cervical cord compression rarely present with rapid decompensation. Physical examination findings suggestive of spinal cord compression may be more predictive of significant spinal cord injury than plain radiographs, and it is wise for all individuals who have DS to undergo a careful neurologic examination prior to sports participation. At present, however, the recommendation for screening cervical spine radiographs stands, and further study is necessary before this recommendation is changed.

Cardiac health is the other major concern affecting participation in sports for individuals who have DS. Parents of children who have congenital heart disease should discuss this topic with their child's cardiologist.

The child described in the vignette has symptoms and signs suggestive of subluxation of the atlantoaxial joint with compression of the cervical spinal cord. Individuals who have symptomatic AAI may present with easy fatigability, difficulty walking, unusual gait, abnormal positioning of the neck, clumsiness, sensory deficits, and hyperreflexia or spasticity. Magnetic resonance imaging of the spinal cord is warranted in such instances.

The most likely presenting symptom of central nervous system leukemia is headache. Cerebellar medulloblastoma, Chiari malformation, and transverse myelitis are no more common in those who have DS than in the general population.

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Question: 105

A 17-year-old young woman comes to your clinic after having been diagnosed with pelvic inflammatory disease the preceding day. She was prescribed doxycycline 100 mg orally twice a day for 14 days and given ceftriaxone 250 mg intramuscularly in a single dose at the time of diagnosis. Since this visit, she vomited the doxycycline, has been unable to retain any fluids, has developed a fever, and has had worsening abdominal pain. External genital examination findings are normal.

Of the following, the MOST appropriate next step is to

A. administer a repeat dose of the oral doxycycline and send the patient home

B. administer benzathine penicillin G 2.4 million units intramuscularly in a single dose and send the patient home

C. change the oral medication to azithromycin 1 g given in a single dose and send the patient home

D. hospitalize the patient and begin intravenous cefotetan 2 g plus doxycycline 100 mg every 12 hours

E. hospitalize the patient for observation and add acyclovir 400 mg orally TID for 7 to 10 days

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Critique: 105 Preferred Response: D

Pelvic inflammatory disease (PID) treatment regimens must provide broad-spectrum coverage of likely pathogens, including *Neisseria gonorrhoeae* and *Chlamydia trachomatis*. Oral therapy can be used for women who have mild-to-moderately severe acute PID; the Centers for Disease Control and Prevention (CDC) report that clinical outcomes of women treated with oral therapy are similar to those seen in women treated with parenteral therapy. The young woman described in the vignette was treated appropriately as an outpatient, but subsequently was unable to tolerate an outpatient regimen and did not appear to respond clinically to an oral regimen (eg, doxycycline and azithromycin). In addition, a single dose of benzathine penicillin is not adequate therapy for PID. These are two of the criteria suggested by the CDC that indicate the need for hospitalization of women who have PID. Other criteria for hospitalization include when surgical emergencies (such as appendicitis) cannot be excluded; the patient is pregnant; the patient has a severe illness, nausea and vomiting, or high fever; and the patient has a tubo-ovarian abscess. Although many practitioners may prefer to hospitalize adolescents who have PID, no available evidence exists to support this strategy. Younger women who have mild-to-moderate acute PID have similar outcomes in response to outpatient or inpatient therapy, and clinical response to outpatient treatment is similar for older and younger women. The CDC states that the decision to hospitalize adolescents who have acute PID should be based on the same criteria, as stated above, used for older women.

Because the patient has no signs or symptoms of herpes simplex virus infection, oral acyclovir with hospitalization is not indicated.

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Question: 106

During the annual health supervision visit of a 9-year-old boy, you note that he has grown very little in the past year (Item Q106). He has been otherwise well. On physical examination, he has slightly increased abdominal fat and decreased muscle mass.

Of the following, the MOST likely diagnosis is

A. celiac disease

B. constitutional delay of maturation

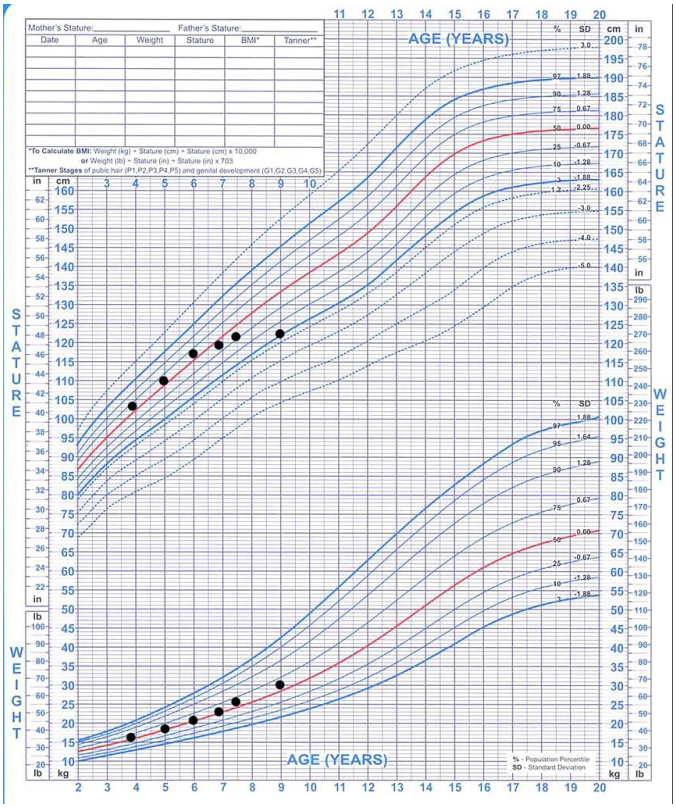
C. craniopharyngioma

D. hypochondroplasia

E. renal insufficiency

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Question: 106



Courtesy of L. Levitsky

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Critique: 106 Preferred Response: C

A hallmark of short stature due to endocrine disease is central adiposity and somewhat decreased muscle mass, as described for the boy in the vignette, which is found in growth hormone deficiency, hypothyroidism, and Cushing syndrome. Craniopharyngioma may present with endocrine deficiency disorders such as growth hormone and thyroid-stimulating hormone deficiency, leading to hypothyroidism. Celiac disease (Item C106) and renal insufficiency usually lead to weight loss when associated with slowing growth, but additional symptoms would be expected. Hypochondroplasia is associated with moderate short-limbed dwarfism that begins in early childhood. An activating mutation of the fibroblast growth factor 3 receptor, of less severity than in achondroplasia, is found in most children who have this autosomal dominant disorder. In constitutional delay of maturation growth attenuation begins 2 to 3 years before puberty, not as early as 7 years of age. The boy described in the vignette requires careful growth evaluation, which might reveal low insulin-like growth factor 1 (somatomedin C) and free thyroxine concentration with normal thyroid-stimulating hormone values. Children who have congenital growth hormone deficiency usually begin to manifest slowing growth by 6 months of age and soon develop a cherubic appearance. They may develop hypoglycemia. If hypopituitarism is present, there may be associated jaundice and, in boys, microphallus.

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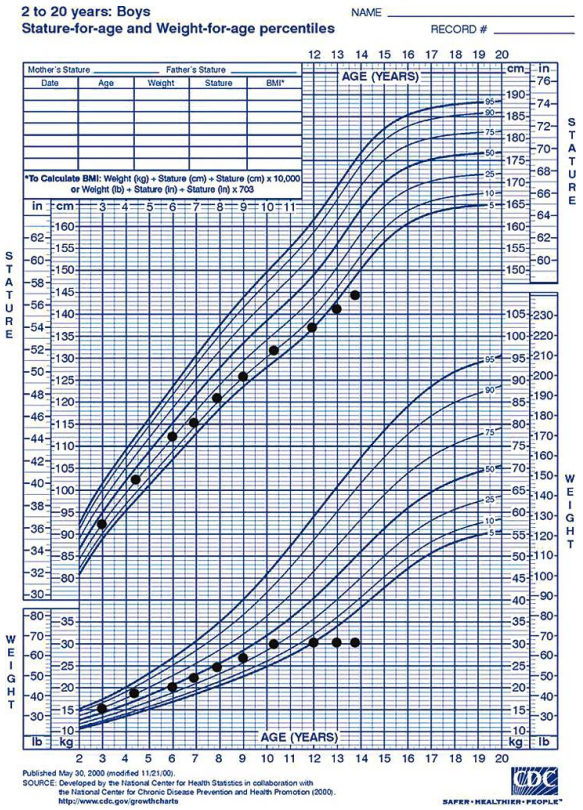
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Critique: 106



Growth curve in a child who has celiac disease documenting slowing of linear growth and weight loss.

Courtesy of L. Levitsky

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Question: 107

The parents of 9-month-old twins ask you if they should be concerned about the vast behavioral differences between the children. They explain that the boy whimpers when he is hungry, but the girl has a vigorous scream. The boy plays quietly while his diaper is changed, but the girl is constantly moving. Finally, the girl tends to display her emotions with strong intensity, while the boy is more easygoing.

Of the following, your BEST response is that

A. the behaviors the infants display are typical of twins

B. the behaviors the twins display are due to their different temperaments

C. the girl's behavior is indicative of a developmental disorder that should be monitored closely

D. the parents need behavioral counseling to improve their parenting skills

E. the parents should respond to both infants similarly

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Critique: 107 Preferred Response: B

Knowing an infant's temperamental characteristics helps to develop an optimal approach to early child care. A difficult child is described as one who has an inborn temperament that makes parenting a challenge. Approximately 15% of children are considered to be temperamentally difficult, and traits such as distractibility, negative mood, or stubbornness can be seen in both sexes. Studies of twins and adoption studies indicate that genetics play a role. These children have trouble with change and tend to be self-directed. If an infant has a difficult temperament or a child is slow to warm to others, the pediatrician can guide parents in gradually introducing the child to new events in advance of the actual situation. Some studies have suggested that temperamentally difficult children are most vulnerable to behavioral problems in early and middle childhood, but with parental counseling and other therapeutic measures, most children improved markedly by adolescence.

When assessing a child's temperament, it is important to examine "goodness of fit" in terms of how compatible the child is to his or her environment. The parents of a child who has a difficult temperament need to be educated that the child's behavior is not intentional but rather temperamentally driven. They need to use behavior management strategies to handle misbehavior. For example, the parents of an impulsive, easily distracted child should intervene early before the child's behavior escalates.

The behavior of the infants described in the vignette is not typical twin behavior; rather, it is indicative of differences in temperament between the two children. Therefore, the parents need to respond to each child according to his or her different temperamental traits. The parents need to be aware of the broad spectrum of normal temperaments in children. There is no indication from the vignette that the female child is displaying developmental issues. The parents of an extremely difficult child may benefit from referral to a mental health professional, but the vignette does not state that the parents find the twins' behaviors that challenging.

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Question: 108

You are taking your family on a safari in Africa. Everyone has received hepatitis A and typhoid immunization. Your husband is worried about “catching malaria” and asks how you are planning to prevent this.

Of the following, the BEST chemoprophylaxis to use for malaria is

A. atovaquone

B. chloroquine

C. dapsone

D. mefloquine

E. no prophylactic medications

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Critique: 108 Preferred Response: D

Malaria prevention consists of limiting or preventing mosquito bites and chemoprophylaxis. Travelers to endemic regions should understand that dusk to dawn is the time that transmission from the *Anopheles* mosquito is the highest. Therefore, they should remain in well-screened areas during this period and sleep under permethrin-treated mosquito netting. During the day, diethyltoluamide (DEET)-containing compounds should be used on areas of the skin that cannot be covered by clothing.

Chloroquine-resistant malaria (due to *Plasmodium falciparum*) is common throughout Africa, requiring the use of other medications, such as mefloquine, doxycycline, or atovaquone-proguanil, for chemoprophylaxis. Mefloquine is taken once weekly starting 1 week prior to travel, once weekly while traveling, and for 4 weeks after the travel has concluded. Although not approved for children weighing less than 5 kg or who are younger than 6 months of age, the Centers for Disease Control and Prevention suggest that mefloquine be used for all children if travel to a chloroquine-resistant area cannot be avoided. Alternatives to mefloquine should be used if patients have a known hypersensitivity to mefloquine, have cardiac conduction abnormalities, are taking beta blockers, have a history of psychiatric illness (eg, anxiety disorders, psychosis), or have a convulsive disorder. Mefloquine can affect fine motor coordination and spatial discrimination. Some patients, especially those who have underlying psychiatric illnesses, may develop disturbing nightmares and hallucinations.

Daily doxycycline starting 1 to 2 days prior to exposure and continuing for 4 weeks after departure from the malaria endemic region is another prophylactic option. Doxycycline use is limited because of the daily dosing requirement, potential photosensitivity, and limitation to children older than 8 years of age.

The combination of atovaquone-proguanil (not atovaquone alone) also can be used for the prevention of chloroquine-resistant malaria. The medication is taken daily starting 1 day prior to exposure and continued for 1 week after departure from the malaria-infected area. Atovaquone-proguanil has effects both in the erythrocytic and exoerythrocytic stages of the malaria life cycle. Therefore, it does not need to be taken for as long as mefloquine or doxycycline after a traveler has left the malaria endemic region. Atovaquone-proguanil is not approved for children weighing less than 11 kg. The most common adverse effects when used for chemoprophylaxis are abdominal pain, nausea, vomiting, and headache.

Chloroquine is an effective medication for chemoprophylaxis in chloroquine-susceptible regions and is given once weekly beginning 1 week prior to travel, during travel, and for 4 weeks after leaving the malaria endemic region. Adverse effects are uncommon when the agent is used for chemoprophylaxis. Minor adverse effects include headache, dizziness, pruritus, and blurred vision. Dapsone is ineffective and should not be used for malaria chemoprophylaxis.

Prevention of illness and trauma are important components to a successful trip for any traveler. Web sites from the Centers for Disease Control and Prevention (http://www.cdc.gov/travel/) and the World Health Organization (http://www.who.int/ith/en/) can provide up-to-date information to aid both travelers and their physicians.

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Question: 109

A 14-year-old previously healthy girl presents with a 3-day history of knee and ankle pain and a maculopapular lacy rash (Item Q109A) that is most prominent on her face (Item Q109B), trunk, and thighs. Ten days prior to the development of the rash and the arthralgias, she had a temperature of 102°F (38.9°C), malaise, and a headache that resolved after 2 days.

Of the following, the MOST appropriate method to confirm the diagnosis is

A. cell culture

B. nucleic acid hybridization

C. polymerase chain reaction

D. rapid antigen testing

E. serum immunoglobulin M titer

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Question: 109



Courtesy of D. Krowchuk

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Question: 109



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Critique: 109 Preferred Response: E

Culture-based detection of viruses requires seeing cytopathic changes in cell cultures. A drawback to conventional cell culture is the prolonged period of time that often is required for the development of cytopathic changes (up to 4 weeks for some viruses). Further, not all viral organisms can be isolated in culture because of undeveloped methodology, safety issues, or impracticality. For this reason, the detection of nonspecific or specific antibodies is useful diagnostically and epidemiologically. Antibody response to an infection varies, and distinction between active and past infection is difficult based on an elevated single antibody titer. Therefore, acute and convalescent titers should be obtained in most cases. Some common viruses that typically are diagnosed serologically through antibody titers are Epstein-Barr virus; human herpesvirus 6; parvovirus B19; hepatitis A, B, C, D, and E viruses; measles, mumps, and rubella viruses; arboviruses; and human immunodeficiency virus-1 and -2.

The patient described in the vignette has clinical findings that are consistent with a parvovirus B19 infection. Parvovirus B19 cannot be isolated by standard cell culture, and rapid antigen testing for this organism is not available. Nucleic acid hybridization and polymerase chain reaction may be used to detect parvovirus B19 in an immunocompromised patient who has chronic infection, but they are not sensitive tests for indicating acute disease.

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Question: 110

An 8-year-old boy presents with tingling in his lower extremities. He had been treated surgically for osteogenic sarcoma and remains on a chemotherapeutic regimen that includes cisplatin. He reports intermittent vomiting and loose stools during his chemotherapy treatment. Physical examination shows no other findings of note. Among the results of laboratory evaluation are a magnesium value of 0.9 mg/dL (0.37 mmol/L) and a potassium value of 2.7 mEq/L (2.7 mmol/L).

Of the following, the MOST likely explanation for this boy's electrolyte imbalance is

A. cellular shifting due to changes in pH

B. dietary deficiency

C. losses due to recurrent diarrhea

D. ongoing losses due to vomiting

E. urinary losses due to tubular damage

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Critique: 110 Preferred Response: E

The symptoms of tingling described for the boy in the vignette may be associated with hypomagnesemia and hypokalemia. Cisplatin is a chemotherapeutic agent that can cause acute losses of magnesium and potassium and chronic losses in some patients that necessitate chronic oral replacement of these electrolytes. Chemotherapeutic agents used to treat pediatric malignancies are associated with a number of untoward complications, including nephrotoxicity. The two classes of drugs that most commonly cause renal toxicity are the platinum drugs (cisplatin and carboplatin) and ifosfamide.

The platinum drugs (cisplatin more than carboplatin) cause injury to the late proximal tubule and collecting duct. More specifically, the proximal tubular damage can result in alterations of the brush border and tubular necrosis. The clinical manifestations of this damage are polyuria, electrolyte abnormalities, and azotemia. Inappropriate tubular losses of magnesium, sodium, and potassium are typical and result in the need for aggressive replacement. Prevention of cisplatin-associated nephrotoxicity has been aimed at optimizing hydration before, during, and after the infusion. Mannitol also may be used. Studies have shown that although many of the effects of cisplatin are temporary, altered renal function and magnesium wasting persist in many patients in long-term follow-up.

Ifosfamide, which is used to treat many pediatric solid tumors, has been associated with acute and chronic nephrotoxicity. This agent can result in the Fanconi syndrome due to damage to the early portion of the proximal tubule. As a result, glycosuria, phosphaturia, amino aciduria, proteinuria, and altered glomerular filtration rate can be seen. The risks of ifosfamide nephrotoxicity appear to be related to the age of the patient, coexisting nephrotoxins, and nephron mass (with worse outcomes in patients who have prior unilateral nephrectomy).

Serum potassium concentrations can be influenced by a number of factors, including dietary intake, intestinal absorption, and urinary excretion. Because potassium is primarily intracellular, in the setting of alkalosis, it shifts intracellularly, and the serum concentrations decrease. Like potassium, magnesium is primarily contained intracellularly. It is absorbed in the ileum, and its extracellular concentrations are regulated in the kidney.

When hypomagnesemia and hypokalemia occur together, renal causes should be strongly considered. A metabolic alkalosis may cause the serum potassium to fall, but should have minimal effect on magnesium. Moreover, the patient described in the vignette is not believed to be at risk for an alkalosis. In the setting of normal renal function without ongoing losses, magnesium and potassium concentrations would be expected to be normal, even in the setting of a dietary deficient in these minerals. The patient has only intermittent vomiting and loose stools, making losses from the gastrointestinal tract unlikely.

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Question: 111

A 4-year-old boy presents with a 2-year history of persistent bilateral nasal congestion. His parents are worried because at night he snores loudly and has had pauses in his breathing. His symptoms occur daily and have not improved with the administration of oral decongestants, nasal corticosteroids, oral antihistamines, or antibiotics. The boy denies ocular pruritus, sneezing, or rhinorrhea. On physical examination, a low-pitched inspiratory noise is audible, and there is “cobblestoning” of his posterior pharynx. Findings on the remainder of the physical examination, including the tonsils, nose (by nasal speculum examination), and neck, are unremarkable.

Of the following, the MOST likely diagnosis is

A. adenoidal hypertrophy

B. allergic rhinitis

C. choanal atresia

D. chronic sinusitis

E. juvenile nasopharyngeal angiofibroma

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Critique: 111 Preferred Response: A

Chronic nasal congestion that is bilateral and persistent in children 2 to 5 years of age should prompt evaluation for an obstructive cause, specifically adenoidal hypertrophy (Item C111A). Typical symptoms of adenoidal hypertrophy are chronic mouth breathing, snoring, nasal obstruction, and hyponasal speech; more severe symptoms include obstructive sleep apnea syndrome and recurrent infections (eg, sinusitis, otitis media). Because the adenoidal tissue is located at the posterior aspect of the nasopharynx, it cannot be seen in the usual nasal speculum examination; nasal rhinoscopy or a lateral neck radiograph is required (Item C111B). Further, although enlarged tonsillar tissue may indicate enlarged adenoid tissue, a direct correlation is not always true.

Allergic rhinitis may be seasonal or perennial and can produce symptoms of rhinorrhea and nasal obstruction. Allergic rhinitis usually does not occur until 5 to 10 years of age. The absence of sneezing or rhinorrhea, and the lack of improvement with usual allergy medications (eg, nasal corticosteroids, oral antihistamines) described for the boy in the vignette also make allergic rhinitis unlikely.

Choanal atresia (Item C111C) (Item C111D) and choanal stenosis (Item C111E) are rare congenital structural malformations of the nose that can result in nasal congestion and obstructive sleep apnea syndrome. Obstruction may be unilateral or bilateral and may not present until later in infancy if not complete or bilateral. However, symptoms often are noted at birth because of difficulty during feedings.

Chronic sinusitis can present at any age because the maxillary and ethmoid sinuses are present at birth. Typical symptoms may include chronic rhinorrhea (clear or discolored) and nasal obstruction. The lack of improvement with oral antibiotics for the boy in the vignette suggests that sinusitis is unlikely, but if the evaluation for adenoidal hypertrophy was negative, sinus imaging should be considered.

Juvenile nasopharyngeal angiofibroma has been described in children as young as 2 years, but typically it presents with profuse epistaxis and a nasal mass during puberty.

Other causes of obstructive sleep apnea syndrome include certain glycogen storage diseases, hypothyroidism, Down syndrome, achondroplasia, laryngomalacia, and Pierre Robin anomaly. In younger children and infants, central causes of apnea include myelomeningocele, hydrocephalus, and Arnold-Chiari malformation. Children who have central apnea lack the ability to sense hypercapnia. Finally, a mixed apnea pattern may be present that combines the aspects of central and obstructive causes.

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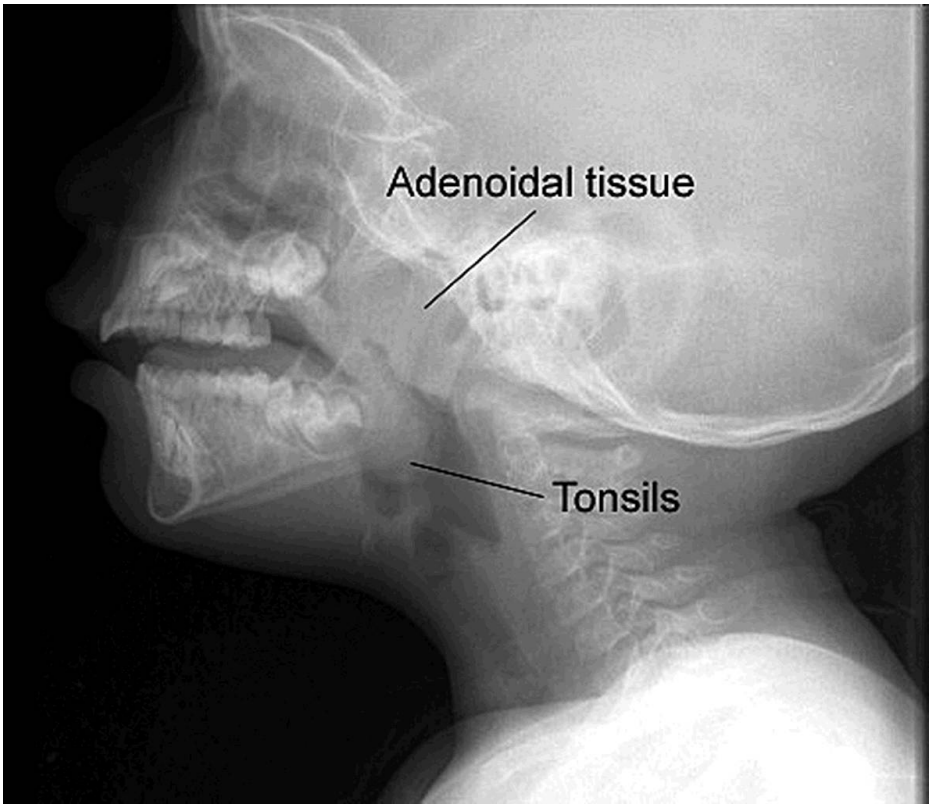
Critique: 111

Item 111A: Causes of Nasal Obstruction

- Adenoidal hypertrophy
- Choanal stenosis/atresia
- Hematoma
- Nasal polyps
- Allergic rhinitis
- Chronic sinusitis
- Foreign body
- Mass lesion

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Critique: 111

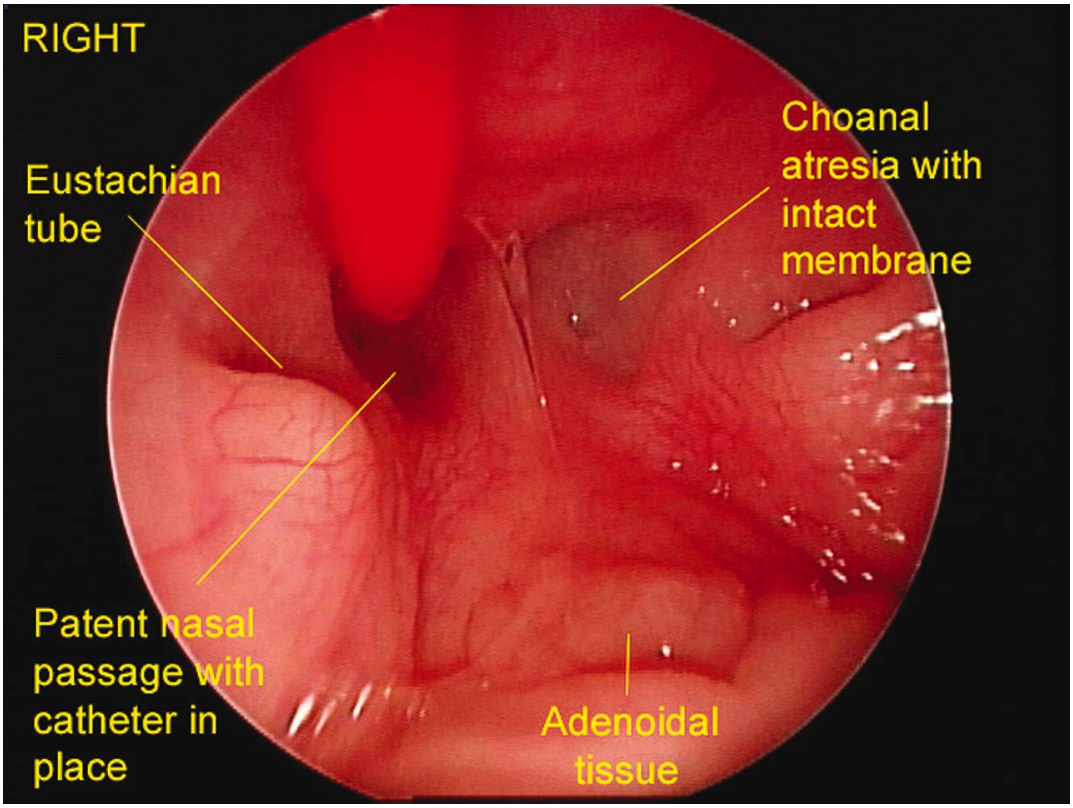


Enlarged adenoidal tissue impinging on the nasopharyngeal airway.

Courtesy of D. Mulvihill

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Critique: 111

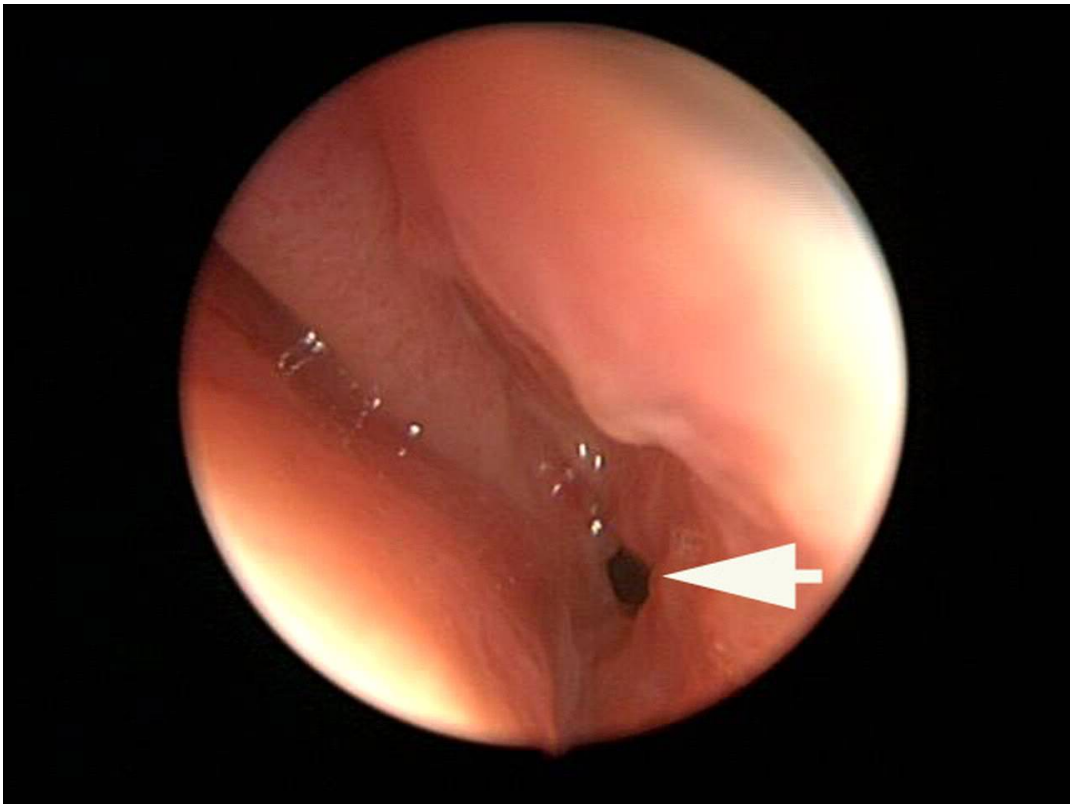


Choanal atresia, as observed from the pharynx.

Courtesy of D. Kirse

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Critique: 111



Choanal stenosis: Transnasal endoscopy demonstrates significant narrowing of the nasal passage (arrow).

Courtesy of M. Boston

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Question: 112

A 10-year-old soccer player sustained tibial and fibular fractures of his left leg yesterday during a soccer game and was placed in a long leg posterior splint in the emergency department. His mother calls you today because he is complaining of worsening pain in his lower leg over the last 4 hours that has not improved with acetaminophen with codeine and loosening of the splint. In addition, he says that he no longer can move the toes on his left foot, and the toes look white.

Of the following, a TRUE statement about this patient's condition is that

A. absence of pulses distal to the injury site is an early physical finding

B. anticoagulant therapy should be started immediately

C. elevation of the affected extremity will relieve the symptoms

D. lack of tenseness on palpation of the lower extremity rules out the diagnosis

E. measurement of the pressures in the muscle compartments of the lower leg is the diagnostic test of choice

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Critique: 112 Preferred Response: E

Worsening pain and neurovascular compromise following an extremity fracture suggest the possibility of compartment syndrome, a potentially limb-threatening complication. Compartment syndrome results when increased tissue pressures (typically related to muscle and soft-tissue edema) within closed osteofascial compartments (Item C112) cause decreased muscle perfusion and ischemia. Most commonly seen after extremity fractures, compartment syndrome also may complicate burns, snakebite, and crush injuries. The lower leg is affected most commonly, with tibial fractures accounting for 40% of fracture-related cases.

Compartment syndrome should be suspected in patients who have fractures and complain of worsening pain or pain out of proportion to the injury, such as the boy described in the vignette. If compartment syndrome is suspected, immediate surgical consultation is indicated, and compartment pressures should be measured. Normal muscle compartment pressure is typically less than 10 to 12 mm Hg. Subtracting the measured intracompartmental pressure from the diastolic blood pressure provides a measure of the compartmental perfusion pressure. A compartmental perfusion pressure of 30 mm Hg or less is indicative of ischemic compromise and should prompt consideration of surgical intervention to relieve the compartment pressure.

With the exception of pain, most of the classically described clinical features of compartment syndrome—"the 5 Ps" (pain, pallor, paresthesias, paralysis, pulselessness)—are late findings. It also is unusual for the muscle group to feel "tight" despite rising pressures. A high index of clinical suspicion should prompt the clinician to evaluate high-risk patients before neurovascular compromise is evident. Release of intracompartmental pressure within 6 to 12 hours of diagnosis is critical to a favorable outcome.

Initial treatment of compartment syndrome involves removal of constricting dressings, splints, or casts and maintenance of the extremity at "heart level" to maximize perfusion and minimize further dependent edema. Fasciotomies of the affected compartments are performed to release the intracompartmental pressure and restore muscle perfusion. Untreated compartment syndrome results in scarring of the affected muscle, with contracture formation and peripheral neuropathy of the local nerves leading to severe disability.

Anticoagulant therapy is indicated for treatment of deep venous thrombosis, which can cause pain in the affected extremity but does not lead to neurologic deficits or arterial compromise, as described in the vignette.

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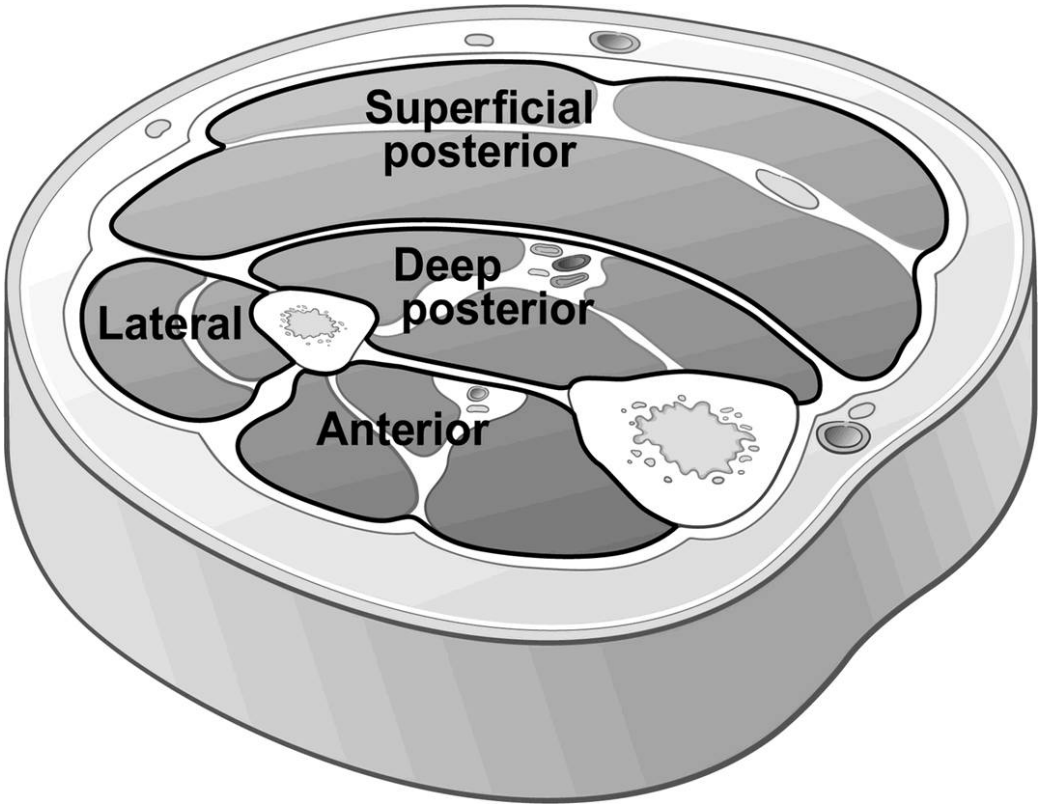
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Critique: 112



Cross-sectional drawing illustrating the muscle compartments of the leg. Increased pressure within these closed compartments may lead to decreased muscle perfusion and ischemia.

Courtesy of A. Johnson

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Question: 113

A 16-year-old girl requests treatment for acne. She has used a nonprescription medication containing benzoyl peroxide without significant benefit. Physical examination reveals inflammatory lesions and open and closed comedones on the face (Item Q113) and inflammatory lesions on the chest and back; there is no scarring. She has no known allergies to medications.

Of the following, the MOST appropriate treatment is

A. benzoyl peroxide topically and tretinoin topically

B. clindamycin topically

C. doxycycline orally and tretinoin topically

D. isotretinoin orally

E. tretinoin topically

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Question: 113



Courtesy of D. Krowchuk

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Critique: 113 Preferred Response: C

Acne affects 85% or more of adolescents and is the skin disorder most often treated by physicians. Multiple factors contribute to acne, including bacteria (that induce an inflammatory response, leading to erythematous papules and pustules (Item C113A)), androgens (that cause increased sebum production), and abnormal follicular keratinization (that causes blockage within pores, resulting in blackheads and whiteheads (Item C113B)). The treatment of acne depends on the types of lesions present (eg, blackheads/whiteheads, inflammatory, or both), the extent of disease (face versus face and trunk), and the severity of disease (more severe disease is characterized by scarring and the presence of numerous and larger inflammatory lesions).

The girl described in the vignette has widespread inflammatory acne involving the face and trunk and comedones on the face (Item C113C). Therefore, she requires therapy with an oral antibiotic (to control inflammatory papules and pustules) and a topical retinoid (to treat the blackheads and whiteheads). Because she has inflammatory lesions on the face, chest, and back, the use of a topical antimicrobial agent such as benzoyl peroxide or clindamycin is impractical. Using a topical retinoid as monotherapy would not have a significant impact on the inflammatory component of the girl's disease.

When inflammatory acne is severe (ie, associated with numerous or large lesions or when scarring is present), extensive, or unresponsive to topical antibiotics, an oral antibiotic should be employed. As with other acne therapies, 6 to 8 weeks may be required before a significant clinical effect is observed. Once the appearance of new lesions has ceased or been satisfactorily reduced, the dose may be tapered gradually or withdrawn.

Although many antibiotics have been used to treat acne, those for which data regarding efficacy are most compelling are tetracycline, erythromycin, doxycycline, and minocycline. Tetracycline and erythromycin often are used as first-line agents at a dose of 250 to 500 mg twice daily (the higher dose usually is favored). Although both agents are well-tolerated by most patients, clinicians should be aware of dosing requirements (eg, tetracycline must be taken without food) and adverse effects (eg, erythromycin may cause gastrointestinal upset) that may affect their effective use. An additional concern about erythromycin is the high prevalence of bacterial resistance.

For those who have more severe disease or who fail to respond to or cannot tolerate tetracycline or erythromycin, doxycycline or minocycline often are effective. Both are initiated at a dose of 50 to 100 mg twice daily (the higher dose usually is favored). Again, clinicians should be aware of potential adverse effects of each agent. For example, doxycycline may cause photosensitivity reactions, and both agents may produce gastrointestinal upset, esophagitis, or pseudotumor cerebri. Minocycline also may cause dizziness or uncommon but significant adverse effects that can include pigmentation of the skin, teeth, or mucosa or autoimmune syndromes (eg, a serum sickness-like reaction, a hypersensitivity syndrome, lupus erythematosus-like reaction, and hepatitis). For patients who have severe inflammatory acne that does not respond to oral antibiotics, oral isotretinoin should be considered.

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Critique: 113



Inflammatory lesions of acne are erythematous papules and pustules.

Courtesy of D. Krowchuk

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Critique: 113



Blackheads and whiteheads (skin-colored or white papules) are evidence of follicular obstruction in acne.

Courtesy of D. Krowchuk

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Critique: 113



Moderate mixed acne, including numerous inflammatory lesions, blackheads, and whiteheads.

Courtesy of D. Krowchuk

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Question: 114

A 5-year-old child presents to your office with a history of recurrent rectal prolapse that occurs at the time of bowel movements. Both the mother and child are very concerned when the rectal tissue protrudes from the anus, but the prolapse typically resolves without treatment.

Of the following, the MOST common cause of rectal prolapse in children is

A. celiac disease

B. cystic fibrosis

C. *Enterobius vermicularis* infestation (pinworms)

D. functional constipation

E. rectal polyps

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Critique: 114 Preferred Response: D

Rectal prolapse is the herniation of rectal tissue out of the anus. Prolapse typically occurs during defecation and often resolves spontaneously when a child relaxes after defecation. Nevertheless, prolapse usually is of major concern to a parent and child, and ongoing prolapse may be associated with rectal bleeding from tissue ulceration. The most common cause of rectal prolapse is chronic functional constipation, which accounts for 30% of cases. Other less common causes include rectal polyps, repaired anorectal malformations, constipation due to myelomeningocele, and cystic fibrosis. Although cystic fibrosis probably accounts for fewer than 10% of patients who have recurrent rectal prolapse, it still is recommended that any child who has recurrent rectal prolapse undergo a sweat test to exclude this condition. Temporary rectal prolapse also may be seen in children who have acute diarrheal disease, but should resolve after the illness. Celiac disease and pinworm infection are uncommon causes of rectal prolapse. Up to 20% of patients who have recurrent prolapse do not have an identifiable cause.

Treatment of rectal prolapse typically involves treating constipation with a combination of a lubricant (eg, mineral oil, polyethylene glycol) and a gentle stimulant (eg, senna). If there is significant rectal bleeding, colonoscopy should be considered to exclude lead points or ulcers. Patients who have persistent rectal prolapse despite medical therapy should be referred to a pediatric surgeon. Initial surgical therapy consists of sclerotherapy (injection of hypertonic saline or D50W into the rectal wall). For patients whose rectal prolapse persists despite sclerotherapy, rectopexy or anorectoplasty can be attempted. The prognosis is usually excellent.

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Question: 115

You are planning the discharge of an extremely low-birthweight infant from the neonatal intensive care unit (NICU) who requires home oxygen therapy for chronic lung disease. She also requires anticonvulsant therapy for a seizure disorder that is related to her having a grade III intraventricular hemorrhage with posthemorrhagic hydrocephalus and a ventriculoperitoneal shunt.

Of the following, a TRUE statement about the psychosocial effects of home care and equipment use by her parents is that

A. all parents are challenged by the stressors of home care and equipment use for infants who were in the NICU

B. mothers are more challenged than fathers about the provision of home care and equipment use

C. mothers who have visited regularly in the NICU have no problems with home medical equipment

D. parents who are health-care professionals can be expected to provide optimal care of their infant at home

E. the psychosocial stressors of home care and equipment use by parents require a psychiatric consultation

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Critique: 115 Preferred Response: A

The home care of technology-dependent or -assisted children has increased over recent decades in the United States. Commonly prescribed home medical equipment includes feeding tubes (nasogastric, nasojejunal, or gastrostomy) and pumps, oxygen delivery systems and suction apparatus, apnea and bradycardia monitors, intravenous cannulae, and surgical devices such as shunts. Home care typically is provided by the parents and a number of home-health personnel (respiratory therapists, physical therapists, nutritional and pharmacy services, and skilled or unskilled nurses or nursing assistants). The parents must balance the needs of their infant/child with daily routines that include meeting other family member needs, marital life, employment, and transportation. Hygienic practices need to become part of their care environment, and much of their home is changed to become a location for health-care delivery.

The infant described in the vignette has a number of needs for special home health care that, in addition to her history of extreme prematurity and prolonged stay in the neonatal intensive care unit (NICU), may increase the psychosocial stresses of her family. These challenges and stresses affect both parents. Studies show that mothers who visit the NICU frequently, who learn their infant's care needs, and who participate in providing this care early in the NICU course may cope better than those who do not, but they still may have logistical or psychosocial problems attendant to the use of home medical equipment. Parents who are health-care professionals do not have an advantage in providing optimal care of the infant at home, and expectations of such are unfounded and unrealistic; parents respond first and foremost to their infant as their child. Rarely does the stress of home care for the technology-dependent infant/child require psychiatric consultation, but pediatricians and primary care clinicians should be attentive to signs of maternal depression and refer parents for formal evaluation as indicated.

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Question: 116

The parents of a 6-month-old boy call you in the middle of the night because he is coughing and has nasal congestion. You had diagnosed a viral upper respiratory tract infection when you saw him earlier today. The parents are frustrated that the cough is persistent and request medicine for their boy so they can sleep.

Of the following, the MOST appropriate recommendation is

A. chlorpheniramine/pseudoephedrine combination

B. codeine

C. dextromethorphan

D. guaifenesin

E. saline nasal drops

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Critique: 116 Preferred Response: E

Viral upper respiratory tract infections are the most common type of infection in infants and young children, and cough is an important reflex that helps to clear excessive secretions from the airway. Parents frequently contact their pediatricians for help with cough, especially at night, when the cough may disrupt sleep. Many over-the-counter cough and cold preparations are available and marketed for the pediatric population, but parents should be advised as to the effectiveness and safety of these preparations.

Many of the studies on the efficacy of cough and cold preparations have been conducted in adults; only a few have been performed in children. Randomized, placebo-controlled, blinded trials have failed to show any benefit in children for medications such as antihistamines and decongestants (alone and in combination), antitussives, and narcotics. No studies have shown that the use of expectorants, such as guaifenesin, provides any benefit. Several adverse effects, including sedation, irritability, and tachycardia, may be seen with the use of these medications. In addition, a recent study attributed three infant deaths to cough and cold medications. Accordingly, the best choice for the parents described in the vignette is saline nasal drops, which are inexpensive and safe. Proper technique is essential for maximum improvement in congestion, and suctioning should be performed at times when it is most important to relieve congestion, such as before sleeping and eating. Parents also should be educated on the cause and expected duration of their child's illness as well as concerning symptoms that require further investigation, such as continuation of symptoms beyond 10 to 14 days and dehydration.

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Question: 117

One of your patients underwent enucleation of one eye for treatment of retinoblastoma. He now presents at age 5 years for his first preparticipation sports physical for t-ball.

Of the following, the MOST important advice for him and his parents is that

A. he must wear polycarbonate goggles to protect the remaining eye

B. he should be taught to slide head first into base so he can better see where he is going

C. he should not participate in any sports because he has only one eye

D. only children who have eye problems need to wear goggles when participating in sports

E. the parents should inquire as to the use of an age-appropriate type of ball

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Critique: 117 Preferred Response: A

Parents of children engaged in sporting activities usually are familiar with protection of major body parts from contusion during particular sports. Use of helmets, tooth guards, knee and elbow protection, and protective cups is common among child athletes. Parents are less aware of the need for eye protection during sports participation, despite the fact that eye injuries may be particularly devastating.

The leading cause of sports-related eye injuries in children in the United States is baseballs, and the highest incidence of injury occurs in children younger than 14 years of age. One third of baseball-related eye injuries result from being struck by a pitched ball. Therefore, children younger than 14 years should use batting helmets with polycarbonate face guards. These guards also protect against injuries to the teeth and facial bones. Functionally one-eyed athletes (best corrected vision in the worst eye of less than 20/50), such as the boy described in the vignette, must use these face guards, and they must protect their functional eye when fielding by wearing polycarbonate sports goggles (Item C117) prescribed and fitted by an optometrist.

The child who has compromised vision or monocular vision need not be excluded from participation in all sports. Clinicians should consult the American Academy of Pediatrics Committee on Sports Medicine and Fitness recommendations for medical conditions affecting sports participation. Clearly, involvement in athletic activities is beneficial for physical well-being, especially with today's prevalence of obesity, and the social and educational benefits are well described. However, all children are at risk for eye injury in sports such as baseball, hockey, golf, and other activities involving high-speed projectiles and should be equipped with appropriately fitting polycarbonate goggles.

Sliding head first into base is not appropriate for a child of this age. Younger children should play baseball with a ball specially designed to be softer than an official regulation baseball, but eye injuries still may result when the ball is thrown with force or from a ball machine.

Recreational activities such as paint-ball also carry a high risk of eye injuries, although they often are not considered "sports" by parents and involve little adult supervision. Safe participation in this activity requires the use of a mask that provides eye protection as well as protection against ingestion and inhalation of paint.

References:

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Critique: 117



An example of specially designed glasses with polycarbonate lenses that are recommended for athletes who have one functional eye.

Courtesy of D. Krowchuk

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Question: 118

You are seeing a 10-year-old boy for a health supervision visit. His brother and sister accompany him and his mother to the visit. The mother reports that her husband recently underwent a heart transplant for a “thick heart.” As you explore the family history in more detail, you learn that the boy’s paternal uncle and grandfather both have been diagnosed with hypertrophic cardiomyopathy. The boy has never had chest pain, palpitation, shortness of breath, dizziness, or syncope. He participates in sports and activities without any problems. Findings on physical examination are within normal limits.

Of the following, the MOST appropriate next step is

A. cardiology referral for this patient

B. cardiology referral for this patient and all of his siblings

C. cardiology referral for this patient and his male siblings

D. electrocardiography and echocardiography, followed by cardiology referral if results of either are abnormal

E. genetic testing for the entire family

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Critique: 118 Preferred Response: B

Among the inherited abnormalities of cardiovascular structure, function, and rhythm are the long QT syndrome, Marfan syndrome, and hypertrophic cardiomyopathy (HCM). The latter typically is inherited via one of several autosomal dominant genes that encode for the constituents of the sarcomere. There is significant heterogeneity of the genes responsible for the disorder, which may be one of the factors leading to the variability of severity and perhaps risk associated with the diagnosis. Interestingly, HCM may not become phenotypically apparent until adolescence and even young adulthood. Thus, repeat evaluation is necessary if a child has an affected first-degree relative, but no echocardiographic characteristics of the abnormality at the time of the first evaluation. Alternatively, genetic testing may be an option for some, but the heterogeneity of the gene and expense of testing may make this impractical in some cases. Accurate diagnosis of HCM in children and adolescents is essential because this condition is a known cause of morbidity and mortality. Thoughtful counseling is an important component of care for the child who has HCM; some sports and activities are believed to place the child at particularly high risk for the development of arrhythmia and even sudden death.

The family described in the vignette carries the diagnosis of HCM, and the children each have a 50% chance of having inherited the HCM gene from their father. Current recommendations suggest regular evaluation (perhaps as often as yearly) of such children, ideally by a cardiologist, involving detailed history, physical examination, and diagnostic testing that includes electrocardiography and echocardiography (Item C118). Because the HCM gene is inherited in an autosomal dominant pattern without gender preference, referring only the patient in the vignette or limiting evaluation to males in the family risks missing the diagnosis in some siblings.

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Maron BJ, Chaitman BR, Ackerman MJ, et al. Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. *Circulation*. 2004;109:2807-2816. Available at: http://circ.ahajournals.org/cgi/content/full/109/22/2807

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Question: 119

A 10-year-old boy presents to the emergency department with confusion. He is febrile. While you are examining him, his eyes glaze over and deviate to the right, he has automatic chewing movements, and he is completely unresponsive for 30 seconds, after which he is very sleepy. Emergent head computed tomography scan shows low density in the right temporal lobe.

Of the following, the MOST appropriate next step is

A. administration of intravenous acyclovir

B. administration of intravenous cefazolin

C. administration of intravenous dexamethasone

D. brain magnetic resonance imaging with contrast

E. emergent electroencephalography

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Critique: 119 Preferred Response: A

The signs and symptoms described for the child in the vignette are concerning for meningitis or encephalitis. Fever, focal seizure, and persistent confusion should immediately generate a strong suspicion for viral encephalitis. The low density in the temporal lobe documented by emergent head computed tomography (CT) scan supports a diagnosis of focal encephalitis. Unfortunately, early findings in the temporal lobe are obscured in some cases by artifact on CT. Because herpes simplex virus is the most likely and only treatable cause of viral encephalitis, treatment with acyclovir should be initiated immediately.

Herpes simplex encephalitis causes hemorrhagic necrosis (Item C119). Prior to the availability of acyclovir, some case series reported a 70% fatality rate. Early treatment with acyclovir is essential to reduce morbidity and mortality; delay in treatment can result in brain loss.

Administration of intravenous antibiotics to a child who has encephalopathy, fever, and a seizure is appropriate because bacterial meningitis is in the differential diagnosis. However, cefazolin does not have good cerebrospinal fluid (CSF) penetration and provides inadequate coverage for the common causes of bacterial meningitis. Intravenous dexamethasone reduces the elevated intracranial pressure that occurs due to vasogenic edema that develops around brain tumors. The low signal in the temporal lobe on head CT scan for this patient is due to cytotoxic edema, making dexamethasone treatment less valuable.

Further diagnostic testing should be deferred in favor of immediate treatment with acyclovir. If the CT scan does not show herniation, indicating that a lumbar puncture can be performed safely, this procedure should be undertaken and the CSF evaluated for cells, glucose, protein, and other diagnostic studies. The most accurate and rapid test for herpes encephalitis is polymerase chain reaction (PCR) for the herpes simplex DNA. Other PCR tests are available for the diagnosis of encephalitis caused by other viruses. Therefore, a CSF specimen should be saved for further evaluation in the event that herpes PCR tests results are negative.

Brain magnetic resonance imaging (MRI) is much more sensitive than head CT for early diagnosis of herpes encephalitis. Contrast enhancement provides additional diagnostic information about blood-brain barrier changes around the lesion. However, MRI is not readily available in all emergency departments, requires sedation, and limits monitoring of a medically unstable patient. Electroencephalography in herpes encephalitis may show a pattern called periodic lateralizing epileptiform discharges (PLEDs), which could provide confirmatory information in ambiguous cases, but such testing is not needed initially for medical decision making.

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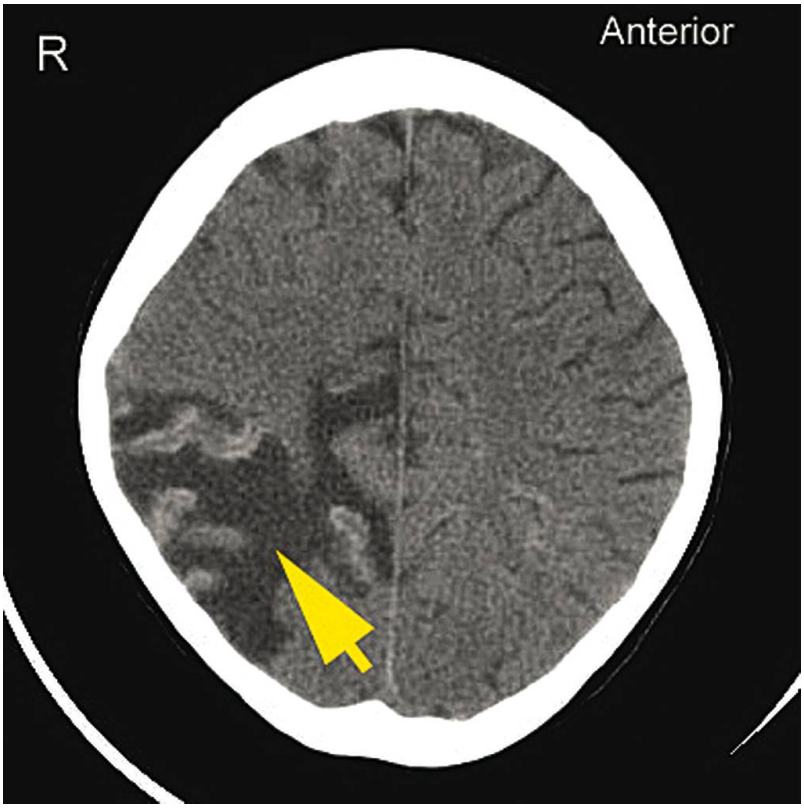
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Critique: 119



Herpes simplex virus encephalitis: Axial computed tomography scan performed 2 weeks after the onset of illness reveals necrosis in the right temporal lobe (arrow).

Courtesy of D. Gilbert

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Question: 120

The parents of a child in your practice call you for advice after learning that the husband's newborn niece has been diagnosed with a rare inborn error of metabolism. The mother of your patient is 16 weeks pregnant with their second child. They ask you whether their new baby will be affected. These parents are not related to one another, and the wife is not related to the mother of the affected newborn. You refer them for genetic counseling.

Of the following, the risk for this same metabolic condition to affect the unborn child is CLOSEST to

A. <1%

B. 5%

C. 10%

D. 25%

E. 50%

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Critique: 120 Preferred Response: A

Most inborn errors of metabolism are inherited in an autosomal recessive pattern. A few notable exceptions are Hunter syndrome (mucopolysaccharidosis type II) (X-linked), ornithine transcarbamylase (OTC) deficiency (X-linked), acute intermittent porphyria (autosomal dominant), and the mitochondrial myopathies (most of which are autosomal recessive, although some are due to mutations in the mitochondrial genome).

Assuming that the husband's niece in the vignette has an autosomal recessive condition, the clinician needs to calculate the "worst case scenario" risk for the parents who are calling for advice. Ideally, one would know the incidence of the disease in question, but the description "rare" suggests that an incidence of 1 in 10,000 to 1 in 50,000 is a reasonable estimate. Each parent of the affected child is a known carrier, and the likelihood that each of that parent's siblings (including the patient's father) is a carrier is 50% (½).

Because the pregnant mother has no family history of the condition in question and is not related to her husband, her risk of being a carrier is the same as the general population risk, which is 2 times the square root of the disease incidence (2 x the square root of 1/10,000), which is 1/50.

The likelihood that these two individuals would have an affected child is calculated as ½ x 1/50 x ¼ (the final fraction is the likelihood that two carriers for a recessive condition would have a child who has that condition), which equals 1/400 (<1%).

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Question: 121

A 16-year-old boy comes to your clinic with complaints of a penile discharge and dysuria. He reports no fever or scrotal tenderness. On physical examination, he is afebrile. He has a thick yellowish discharge at the penile meatus, but no genital rashes or lesions, no scrotal tenderness or swelling, and no inguinal adenopathy.

Of the following, the MOST likely causative organism for his symptoms is

A. herpes simplex 2

B. human papillomavirus

C. *Neisseria gonorrhoeae*

D. *Treponema pallidum*

E. *Trichomonas vaginalis*

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Critique: 121 Preferred Response: C

The symptoms of mucopurulent or purulent penile discharge with dysuria and urinary frequency described for the boy in the vignette are indicative of urethritis. Urethritis may be separated based on infectious etiology into gonococcal (due to *Neisseria gonorrhoeae*) and nongonococcal forms. Nongonococcal urethritis (NGU) can be caused by *Chlamydia trachomatis, Ureaplasma urealyticum*, *Mycoplasma genitalium*, *Trichomonas vaginalis*, herpes simplex virus, adenovirus, and enteric bacteria. Enteric bacteria (eg, *Salmonella*, *Shigella*) can be associated with penetrative anal sexual activity. Although no symptoms or signs reliably distinguish between gonococcal urethritis and NGU, infection with *N gonorrhoeae* usually is characterized by copious purulent discharge, as exhibited by the boy described in the vignette. In NGU, the urethral discharge typically is more scant and mucoid.

The absence of vesicles or ulcers makes herpes simplex infection less likely. NGU caused by *T vaginalis* accounts for approximately 2% to 5% of all NGUs. Balanitis and cutaneous lesions of the penis also are characteristic of *T vaginalis* infection. Primary infection with *Treponema pallidum* manifests as a chancre and tender inguinal adenopathy, and secondary stage infection is characterized by rash or condyloma lata. Exophytic lesions, not urethral discharge, signal infection with human papillomavirus.

References:

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Question: 122

A 7-year-old boy comes to your office for his annual health supervision visit. On physical examination, he has Sexual Maturity Rating 3 pubic hair, his penis is 5 cm in stretched length, his testes are 2 mL in volume, the scrotum is rugated, and there is no thinning of scrotal skin. His weight is at the 90th percentile, and his height is at the 50th percentile (Item Q122).

Of the following, the MOST likely diagnosis is

A. central precocious puberty

B. exposure to exogenous androgens

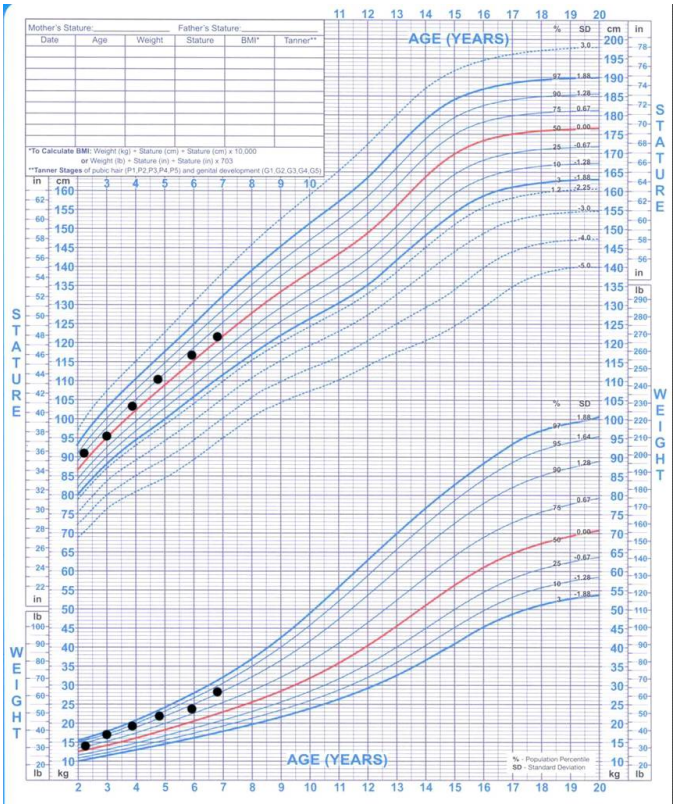
C. late-onset congenital adrenal hyperplasia

D. premature adrenarche

E. virilizing adrenal tumor

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Question: 122



Courtesy of L. Levitsky

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Critique: 122 Preferred Response: D

Precocious puberty in boys is defined as the appearance of secondary sexual characteristics before age 9 years. The only pubertal manifestation displayed by the boy in the vignette is pubic hair. He has no evidence of penile enlargement, his scrotum appears prepubertal, and his testes have not enlarged. Further, he has not had a growth spurt. The most likely diagnosis is early adrenal puberty (premature adrenarche). Expected laboratory findings include somewhat elevated concentrations of serum dehydroepiandrosterone and dehydroepiandrosterone-sulfate, with low concentrations of testosterone and no evidence of activation of the hypothalamic-pituitary-gonadal axis. This usually does not require treatment.

Testicular enlargement is the first sign of true puberty or central precocious puberty. Exposure to exogenous androgen, presence of a virilizing adrenal tumor, or late-onset congenital adrenal hyperplasia (mild 21-hydroxylase deficiency) leads to penile enlargement and pubic hair growth. The testes remain small, but a growth spurt occurs.

Precocious puberty in girls is defined as the appearance of secondary sexual characteristics before age 7 years. Sexual precocity may be manifested by breast development (thelarche) alone, pubic and/or axillary hair (adrenarche) alone, or a combination of the two. Thelarche alone can be due to exogenous estrogen, estrogen-secreting tumor, or early activation of the hypothalamic-pituitary axis. Adrenarche alone may be due to early adrenal puberty, exogenous androgen, androgen-secreting tumor, or congenital adrenal hyperplasia. Both adrenarche and thelarche may be seen in true central precocious puberty but also may be found if an ovarian or adrenal tumor secretes both androgen and estrogen.

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Question: 123

The mother of a 2-year-old girl is very concerned that her daughter is developmentally delayed. She explains that the girl speaks in two- to three-word phrases. She can feed herself with a spoon, but is unable to button her clothing. She can follow simple two-step commands and can climb stairs. However, she is not yet toilet trained. Findings on physical examination are unremarkable.

Of the following, you are MOST likely to

A. discuss the normal developmental milestones of a 2-year-old child

B. refer the child for a neurodevelopmental evaluation

C. refer the child for audiologic evaluation

D. refer the child for occupational therapy

E. schedule a 6-month follow-up evaluation to see if the child has reached the milestones

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Critique: 123 Preferred Response: A

According to the American Academy of Pediatrics, all infants and young children should be screened for developmental delays as part of health supervision visits. Specifically, a screening tool should be administered at 9-month, 18-month, and 24-month or 30-month visits and at other times, when the pediatrician has concerns about an individual child's delayed or disordered development. The use of standardized screening tools can help detect developmental and behavioral problems.

The child described in the vignette is meeting appropriate milestones for a 2-year-old child and has no evidence of developmental delays. Most children do not achieve day-time continence until 3 years of age. A 3-year-old child has the more refined finger and hand movements required to unbutton clothes and possibly place large buttons into button holes. Explaining the normal milestones expected for a 2-year-old child can help to reassure the mother. Providing developmental charts that describe appropriate milestones for her child will educate the mother about age-appropriate skills for the child. Because the child has age-appropriate development, there is no need to refer her for additional evaluations or occupational therapy or to schedule a follow-up evaluation.

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Question: 124

A 5-day-old infant who was found in a trash dumpster is brought to you for evaluation. Because you have no history on the baby, you decide to send serology specimens to test for syphilis, human immunodeficiency virus (HIV), hepatitis B, hepatitis C, and rubella. One week later, all of the test results are negative, with the exception of the presence of antibodies to rubella and HIV.

Of the following, the MOST appropriate next step in the evaluation and treatment of this infant is to

A. begin zidovudine

B. begin zidovudine, lamivudine, and saquinavir

C. observe the infant

D. order an HIV DNA polymerase chain reaction test

E. repeat the HIV serology in 3 months

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Critique: 124 Preferred Response: D

The presence of human immunodeficiency virus (HIV) antibody in a baby within the first 18 postnatal months is an indication of maternal infection. Immunoglobulin G (IgG) passes from mother to child starting at approximately 28 weeks of gestation and continues until delivery. Therefore, any test used for an infant that measures IgG has positive results if the mother had the disease or had been vaccinated against it. IgG to HIV is an especially problematic maternal antibody that can be detected for up to 18 months.

Any infant from birth to 18 months of age who has a positive HIV antibody test result requires further evaluation. The most appropriate next step for the infant described in the vignette is to obtain an HIV DNA polymerase chain reaction (PCR) test in an attempt to establish whether the infant is infected. Current recommendations state that infants born to HIV-infected women should be tested by HIV DNA PCR or HIV RNA PCR assay during the first 48 hours after birth, again from 2 weeks to 2 months of age, and a third time from 2 to 4 months of age. If an infant tests positive, testing should be repeated immediately on a second blood specimen to confirm the diagnosis. An infant is considered infected if two separate samples are positive using PCR testing. Infants who have two negative PCR test results (one performed at more than 4 months of age) should be considered not infected. Such a consideration can be verified by the loss of antibody to HIV on testing at 12 to 18 months of age.

Simple observation of the infant is not appropriate; an attempt should be made to establish whether the infant is infected. Similarly, because the maternal antibody survives for 18 months, simply repeating HIV serology in 3 months is not helpful.

The use of postpartum medications (eg, zidovudine for 6 weeks) to prevent mother-to-child transmission in addition to maternal treatment and intravenous treatment during labor is currently the standard of care. If the mother did not receive any therapy prior to delivery, most clinicians treat the baby for 6 weeks with zidovudine to decrease the chance of infant infection at delivery. However, treatment must be initiated within 6 to 12 hours after birth. By 48 hours after birth, approximately one third of infants who are infected have detectable virus by PCR detection, and this increases to almost 95% by 14 days of age. Therefore, most clinicians do not use chemoprophylaxis for infants who have positive antibody test results and are 72 hours of age or older, such as the infant described in the vignette.

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Question: 125

You are speaking to a group of medical students about the clinical symptoms, physical examination findings, and laboratory diagnosis of viral organisms that can cause acute encephalitis.

Of the following, the virus that can be identified MOST easily by culture techniques is

A. herpes simplex virus

B. Japanese encephalitis virus

C. LaCrosse encephalitis virus

D. St Louis encephalitis virus

E. West Nile virus

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Critique: 125 Preferred Response: A

Detection of viruses by culture is based on seeing cytopathic changes in cell cultures. Viral cultures should be obtained on fluid or tissue in cases of serious illnesses (eg, sepsis, encephalitis, meningitis, myocarditis) or fever of unknown origin, especially when all bacterial cultures are negative. Viral cultures also should be ordered whenever fluid or tissue is obtained for diagnostic purposes (eg, biopsy of brain, lung, or any tissue). However, not all viral organisms can be isolated in culture because of undeveloped methodology, safety issues, or impracticality. The most common viruses that can be isolated through culture are cytomegalovirus, Coxsackieviruses A and B, echoviruses, enteroviruses, herpes simplex virus, human immunodeficiency virus, influenza, measles, mumps, parainfluenza, polioviruses, rabies, respiratory syncytial virus, and rubella.

Japanese encephalitis, LaCrosse encephalitis, St Louis encephalitis, and West Nile viruses are associated with arboviral encephalitis and cannot be isolated in viral culture. Detection of these viruses is based on serology. Herpes simplex virus can be detected very easily by cell culture and is easy to grow. Alternatively, polymerase chain reaction can be performed to detect this virus.

References:

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Question: 126

An 18-month-old female presents with failure to thrive, polydipsia, and photophobia. Her weight is 8 kg and height is 70 cm (both <5th percentile). On physical examination, she appears pale and small for stated age, and she closes her eyes when you attempt to perform ophthalmoscopy. She has tacky mucous membranes and capillary refill of 2 to 3 seconds. Pertinent findings on laboratory evaluation include:

• Sodium, 135 mEq/L (135 mmol/L)

• Potassium, 2.3 mEq/L (2.3 mmol/L)

• Chloride, 109 mEq/L (109 mmol/L)

• Bicarbonate, 14 mEq/L (14 mmol/L)

• Blood urea nitrogen, 15 mg/dL (5.4 mmol/L)

• Creatinine, 0.3 mg/dL (26.5 mcmol/L)

• Calcium, 8.4 mg/dL (2.1 mmol/L)

• Phosphorus, 2.1 mg/dL (0.68 mmol/L)

• Magnesium, 1.4 mg/dL (0.56 mmol/L)

• Hemoglobin, 10.5 g/dL (105 g/L)

• Glucose, 102 mg/dL (5.7 mmol/L)

Of the following, the BEST test to establish the diagnosis is

A. a sweat chloride test

B. intact parathyroid hormone measurement

C. ophthalmologic examination

D. urine ammonia measurement

E. urine chloride measurement

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Critique: 126 Preferred Response: C

The child described in the vignette has failure to thrive; symptoms of polyuria and photophobia; signs of apparent mild dehydration; and laboratory findings that include hypokalemia, metabolic acidosis, and hypophosphatemia. Such electrolyte disturbances are characteristic of Fanconi syndrome, a proximal tubulopathy that results in urinary losses of sodium, potassium, bicarbonate, phosphate, amino acids, protein, and glucose.

Although the differential diagnosis for the causes of Fanconi syndrome in the pediatric patient is extensive, the condition often is due to a metabolic disturbance. Causes of Fanconi syndrome include inherited diseases such as glycogen storage disease, hereditary fructose intolerance, tyrosinemia, cytochrome c oxidase deficiency, galactosemia, Lowe syndrome, Wilson disease, Dent disease, and cystinosis. Acquired causes include heavy metal poisoning, ifosfamide, cisplatin, gentamicin, and ingestion of outdated tetracycline.

The most common cause of Fanconi syndrome in pediatrics is nephropathic cystinosis. Cystinosis is a lysosomal storage disorder that affects all cells in the body. Cystine normally is a product of protein degradation that is transported out of the lysosome. In cystinosis, cystine accumulates within lysosomes, resulting in cellular dysfunction. This autosomal recessive disorder has an estimated incidence of 1 in 100,000 to 200,000 live births, which translates into approximately 15 new cases diagnosed each year in the United States. The *CTNS* gene, which is located on chromosome 17p13, encodes for cystinosin, a transporter protein responsible for transporting cystine out of the lysosome.

Polyuria, polydipsia, growth failure, rickets, and electrolyte abnormalities manifest in the second half of the first postnatal year in approximately 95% of children who have cystinosis. Cystine accumulation within the proximal tubular cells may result in impaired energy generation and a subsequent defect in solute reabsorption. Cystine accumulation within the cornea results in intense photophobia, as described for the child in the vignette. Patients who have cystinosis also may develop hypothyroidism.

Cystinosis may be suspected when cystine crystals are visible within the cornea during slitlamp ophthalmologic examination. The diagnosis is confirmed by the finding of an elevated white blood cell cystine concentration. Treatment includes replacement of electrolyte losses and oral cysteamine therapy. Cysteamine is directed at the transport defect in cystinosis and has been shown to prolong renal survival from 10 years in untreated patients to 23 years when instituted prior to 3 years of age.

The practitioner evaluating a child who is failing to thrive should consider electrolyte measurement and renal function tests when no obvious nutritional cause is present. Polyuria, polydipsia, and nocturnal fluid intake (the child awakens from sleep to drink fluid) are suggestive of diabetes insipidus, but specific electrolyte abnormalities can indicate the possibility of Fanconi syndrome.

The patient in the vignette is exhibiting failure to thrive, but the electrolyte panel, which includes a normal chloride value and metabolic acidosis, is inconsistent with that of a patient who has cystic fibrosis (CF). Patients who have CF typically have excessive loss of chloride in their sweat that results in hypochloremia and metabolic alkalosis. Patients who have primary hyperparathyroidism have low phosphorus and slightly low bicarbonate values, but have hypercalcemia. This patient's normal calcium concentration and hypokalemia are inconsistent with an abnormality of parathyroid hormone secretion. Urine ammonia measurement can be useful in patients who have distal renal tubular acidosis, which is associated with normal anion gap metabolic acidosis, but few of the other electrolyte abnormalities reported for this patient. Urine chloride measurement is useful for those who have Bartter syndrome and Gitelman syndrome, which could explain the hypokalemia and hypomagnesemia, but these conditions are associated with hypochloremic metabolic alkalosis, which is not present in the patient in the vignette.

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Question: 127

A 14-year-old girl comes to the clinic with a temperature of 100.6°F (38.1°C), myalgias, arthralgias, and urticaria for the past 48 hours. She recently completed a 10-day course of amoxicillin for a sinus infection. Her symptoms developed shortly after completing the course of the antibiotic. On physical examination, she has a blanchable, erythematous rash on her hands (Item Q127) and feet. There are no target lesions, and the lips and oral mucosa are not involved. She denies rhinorrhea, cough, or recent sick contacts.

Of the following, the MOST likely diagnosis is

A. erythema multiforme

B. immunoglobulin E-mediated penicillin hypersensitivity

C. red man syndrome

D. serum sickness

E. toxic epidermal necrolysis

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Question: 127



Erythematous eruption, as described for the girl in the vignette.

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Critique: 127 Preferred Response: D

Adverse drug reactions can be characterized immunologically with the Gell and Coombs classification as immediate hypersensitivity (type I), cytotoxic antibody responses (type II), immune complex reactions (type III), and delayed hypersensitivity (type IV). The time of onset and type of symptoms described for the girl in the vignette strongly suggest serum sickness, a condition that previously was associated with the use of bovine serum but now is observed most commonly with penicillin. Serum sickness is a systemic type III immune complex reaction that involves antibody-antigen complex formation and complement activation. Symptoms generally begin 1 to 2 weeks after drug initiation and may include fever, rash (Item C127A), malaise, lymphadenopathy, arthralgia, and arthritis. In addition to drug discontinuation, supportive therapy is recommended with antihistamines, analgesics, and corticosteroids.

Erythema multiforme may be “minor” or “major.” Both forms typically present with targetoid cutaneous lesions (Item C127B). With the major form (Stevens-Johnson syndrome), the mucosal surfaces are involved. A skin biopsy may be helpful in establishing a diagnosis.

Toxic epidermal necrolysis now is considered distinct from Stevens-Johnson syndrome and is characterized by a severe blistering disease that affects more than 30% of the total surface body area (Item C127C). Lesions often demonstrate the hallmark Nikolsky sign in which skin separation occurs with slight pressure.

Red man syndrome classically is associated with vancomycin and presents as diffuse flushing during intravenous drug administration. The mechanism is believed to be nonspecific mast cell degranulation due to the rapid rate of infusion. Red man syndrome generally can be avoided by slowing the infusion.

Immunoglobulin (Ig) E-mediated penicillin hypersensitivity is a type I hypersensitivity reaction characterized by urticaria (Item C127D), flushing, pruritus, or angioedema developing within a few days of drug exposure. The lack of symptoms until after completion of the course of antibiotics in the vignette argues against an IgE-mediated drug reaction.

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Critique: 127



The rash of serum sickness or a serum sickness-like reaction often appears as urticarial plaques that have lilac or blue centers.

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Critique: 127



The lesions of erythema multiforme are erythematous papules or plaques that develop central violaceous discoloration.

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Critique: 127



In toxic epidermal necrolysis, blisters develop and rupture rapidly.

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Critique: 127



Urticaria is characterized by pink-to-red wheals of varying sizes and shapes.

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Question: 128

A 2-year-old boy presents with bloody drainage from the left ear. According to his mother, he has had upper respiratory tract infection symptoms for the past 3 days, and last night he was crying and holding his left ear. His pain seemed to improve after she cleaned out the ear with a cotton swab, but this morning there was blood on his pillow and around his left ear. On physical examination of his ears, you are unable to see the left tympanic membrane because of seropurulent fluid in the external auditory canal.

Of the following, the MOST likely cause of the bloody ear drainage is

A. basilar skull fracture

B. foreign body in the external auditory canal

C. otitis externa

D. otitis media with tympanic membrane perforation

E. traumatic tympanic membrane perforation

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Critique: 128 Preferred Response: D

Although there are many potential causes of otorrhea in childhood, it is due most often to acute or chronic otitis media with tympanic membrane perforation. The characteristics of the drainage and the history of other signs and symptoms associated with the condition can help to identify the cause and direct treatment.

Purulent or seropurulent drainage is most characteristic of acute otitis media with a tympanic membrane perforation. Purulent drainage also may be seen with otitis externa. An uncommon cause of purulent discharge from the external auditory canal in children is an infection in a first branchial cleft cyst.

Bloody ear drainage most often is due to trauma to the canal, the tympanic membrane, or temporal bone; chronic inflammation of the canal with granulation tissue or cholesteatoma; a foreign body in the external canal; or less commonly in children, neoplasm or hematologic abnormalities.

Clear drainage suggests leakage of serous fluid through a tympanic membrane perforation or cerebrospinal fluid otorrhea through a congenital anomaly or traumatic defect in the temporal bone or following mastoid or basilar skull surgery. Cerebrospinal fluid otorrhea is seen in 21% to 44% of temporal bone fractures.

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Question: 129

You are evaluating a 7-year-old girl who has a 2-day history of a rash without fever or other symptoms. The only notable findings on physical examination are round, erythematous, thin plaques, each of which has a central violaceous discoloration or blister (Item Q129). The lesions are concentrated on the extremities, including the hands and feet, with relative sparing of the trunk.

Of the following, the MOST likely diagnosis is

A. erythema migrans

B. erythema multiforme

C. Stevens-Johnson syndrome

D. toxic epidermal necrolysis

E. urticaria

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Question: 129



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Critique: 129 Preferred Response: B

The girl described in the vignette has round erythematous plaques that have a central violaceous discoloration (ie, target lesions) concentrated on acral surfaces and no mucosal involvement. These findings suggest a diagnosis of erythema multiforme (EM), previously called EM minor. EM is a hypersensitivity reaction that usually is precipitated by infection with herpes simplex virus (HSV) type 1. Although a recent history (within the previous 2 weeks) of herpes labialis is reported in 50% of cases, HSV DNA can be recovered from target lesions in 80% of patients. The eruption of EM begins abruptly without prodromal symptoms. Early lesions are round erythematous macules, wheals, or plaques that later develop a central violaceous discoloration, vesicle (Item C129A), or concentric rings. The lesions remain fixed in location until they resolve, typically in 7 days or more. Oral ulcers may be present in up to 50% of patients, but other mucosal sites are spared. EM resolves in 2 to 3 weeks, and treatment is supportive. For children who experience recurrences, given the association with HSV infection, prophylactic acyclovir therapy may be employed.

Stevens-Johnson syndrome (SJS) (previously termed EM major) and toxic epidermal necrolysis (TEN) represent hypersensitivity reactions to drugs and, less commonly, infections. They are believed to represent the same disorder, differing only in the extent of epidermal loss: SJS is diagnosed when less than 10% of the epidermis is lost, SJS/TEN overlap when the loss is 10% to 30%, and TEN is diagnosed when the loss is greater than 30%. Unlike in EM, patients who develop SJS or TEN manifest prodromal symptoms such as fever, malaise, cough, sore throat, or headache 1 to 14 days before the eruption appears. Target lesions are uncommon or absent; rather, individuals develop erythematous or violaceous macules or patches (Item C129B) that may develop blisters (Item C129C) that rupture, leaving denuded areas. Multiple mucosal sites are involved, including the conjunctivae (Item C129D), mouth (Item C129E), genitalia, urethra, and rectum.

During the early disseminated phase of Lyme disease, multiple erythema migrans lesions occasionally may be observed (Item C129F). Unlike the lesions of EM, however, those of erythema migrans generally lack the typical targetlike appearance. The appearance of the wheals of urticaria may mimic those of EM. However, individual lesions in urticaria disappear within 24 hours (usually within 2 to 3 hours), vary in size, often have unusual shapes (incomplete circles, large plaques with serpiginous borders), and lack the central epidermal change (eg, vesicle or crust) characteristic of the target lesions of EM (Item C129G).

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Critique: 129



Target lesions exhibit a central change: a violaceous discoloration, vesicle, or crust.

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Critique: 129



In Stevens-Johnson syndrome and toxic epidermal necrolysis, target lesions are uncommon. Rather, patients develop erythematous macules or patches.

Courtesy of D. Krowchuk

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Critique: 129



In Stevens-Johnson syndrome and toxic epidermal necrolysis, blisters develop (arrow) and subsequently rupture, leaving denuded areas.

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Critique: 129



Purulent conjunctivitis is a common finding in Stevens-Johnson syndrome and toxic epidermal necrolysis.

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Critique: 129



Swelling, ulceration, and crusting of the lips is a feature of Stevens-Johnson syndrome and toxic epidermal necrolysis.

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Critique: 129



Multiple erythema migrans lesions may be observed in the early disseminated phase of Lyme disease.

Courtesy of dermatlas.org

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Critique: 129



The wheals of urticaria vary in size and often have unusual shapes.

Courtesy of D. Krowchuk

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Question: 130

A 15-year-old girl who has Crohn disease has had poor appetite and chronic diarrhea despite her medical treatment. In evaluating her nutritional state, you document a low plasma zinc concentration and initiate zinc supplementation.

Of the following, a TRUE statement about zinc supplementation is that it

A. decreases serum alkaline phosphatase values

B. impairs wound healing

C. inhibits the absorption of dietary copper

D. should be given with dairy products

E. usually decreases appetite

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Critique: 130 Preferred Response: C

Zinc and copper are two micronutrients essential to human life. Zinc is an essential component of human enzymes involved in DNA replication, cellular receptors, and transcription proteins. Infants who have acrodermatitis enteropathica, a congenital defect in zinc transport, present with a severe oral and perianal skin rash (Item C130), intractable chronic diarrhea, and growth failure. This disease can be treated successfully with zinc supplementation. Even milder zinc deficiency, as seen in children who have malnutrition or chronic illness, is associated with diarrhea, skin lesions, dysgeusia (altered taste), and behavioral abnormalities.

Copper is another trace element essential to the function of human enzymes, especially those in the respiratory chain. The most severe form of copper deficiency, Menkes syndrome, is caused by mutation of copper ATPases that impairs copper absorption and transport. Infants who have this syndrome have developmental delay, seizures, and “steely hair.” Although copper deficiency is rare, it may occur in malnourished infants and children, especially preterm infants. The classic clinical features of copper deficiency include a hypochromic normocytic anemia, osteoporosis, and hypotonia. Excess copper accumulation is seen in Wilson disease, in which copper is absorbed normally but cannot be excreted from the liver into the bile because of a mutation in a hepatic copper transport protein.

Both copper and zinc are absorbed from the intestine. Zinc-containing foods include beef, cheese, and legumes (beans and nuts). Copper-containing foods include liver, fish, and legumes. Both zinc and iron can interfere with intestinal copper absorption. Therefore, if a zinc supplement is given to a patient who has chronic malnutrition, the patient should be monitored for potential copper deficiency. Zinc absorption can be impaired if zinc is given at the same time as dairy products. Patients who have zinc deficiency often have impaired wound healing, reduced alkaline phosphatase activity, and impaired taste that may affect appetite, all of which should improve after zinc supplementation.

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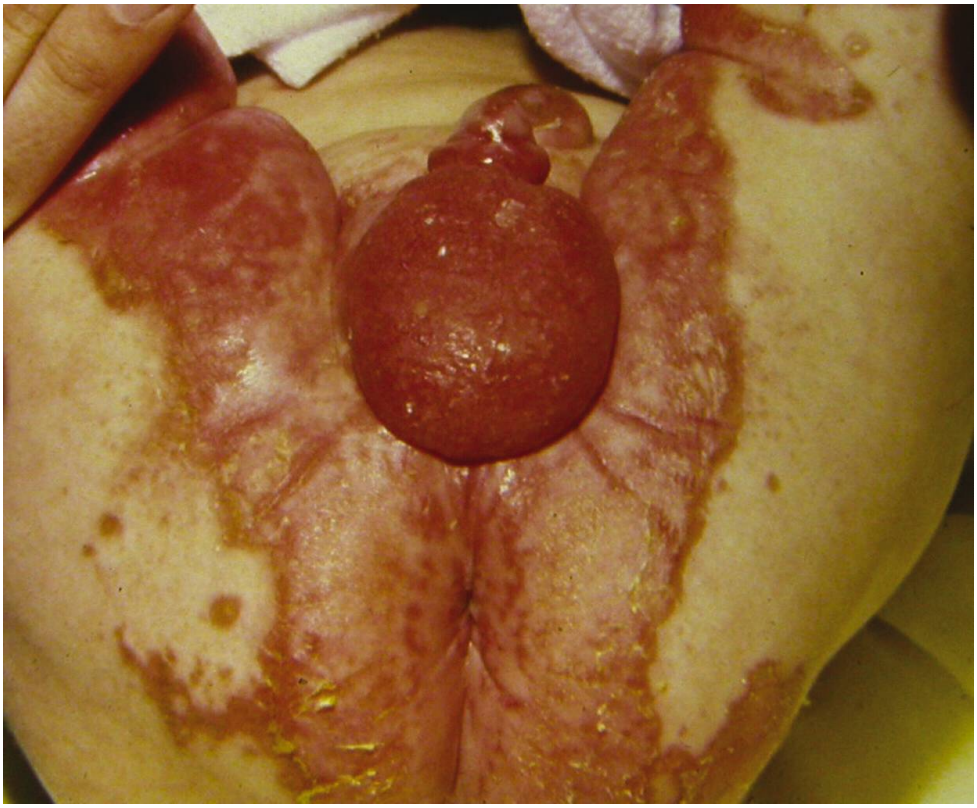
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Critique: 130



Acrodermatitis enteropathica is characterized by perianal and perioral erosions and crusting.

Courtesy of W.W. Tunnessen, Jr

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Question: 131

You admitted a 2,000-g term newborn to the neonatal intensive care unit. Her Apgar scores were 4 and 6 at 1 and 5 minutes, respectively. She exhibits seizure activity at 24 hours of age. Physical examination reveals microcephaly (Item Q131), respiratory distress requiring assisted ventilation, hepatosplenomegaly, cutaneous petechiae, and lethargy. Laboratory tests demonstrate thrombocytopenia, anemia, elevated liver transaminases, and hyperbilirubinemia.

Of the following, the viral agent MOST likely to cause these findings is

A. adenovirus

B. Coxsackievirus B

C. cytomegalovirus

D. hepatitis C virus

E. herpes simplex virus

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Question: 131



Microcephaly, as exhibited by the infant in the vignette.

Courtesy of B. Carter

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Critique: 131 Preferred Response: C

Cytomegalovirus (CMV) is the most prevalent viral pathogen affecting women of childbearing age. It is estimated that 1% of liveborn infants in the United States are infected with CMV. Most congenital CMV infections are asymptomatic; only 10% present as illness in the immediate newborn period. Among the 90% of asymptomatic newborns, up to 20% may develop deafness or mental retardation later in childhood.

The infant described in the vignette has findings consistent with symptomatic congenital CMV infection: intrauterine growth restriction; seizures, which are common in association with microcephaly (Item C131A); and intracranial calcifications. Respiratory distress may be due to pneumonitis, and hepatopathy may include elevated serum transaminase concentrations, coagulopathy, and persistent jaundice. Bone marrow suppression is not uncommon, with thrombocytopenia resulting in petechiae (Item C131B). If anemia is profound, extramedullary hematopoiesis may manifest with hepatomegaly, splenomegaly (Item C131C), and cutaneous nodules ("blueberry muffin" spots).

Adenovirus is an uncommon source of infection for the newborn that may be acquired from an infected mother in the perinatal period; it rarely causes more than upper respiratory tract symptoms in the newborn. Coxsackievirus B, an enterovirus, rarely affects the newborn, but infection may be lethal and is associated with myocarditis, hepatopathy, coagulopathy, and meningoencephalitis; neither microcephaly nor respiratory distress is characteristic. Hepatitis C virus is increasingly prevalent in women and may be transmitted to the newborn in up to 8% of cases. Although it causes no immediate neonatal pathology, follow-up for developing hepatitis later in infancy is warranted. Herpes simplex virus (HSV) is another common perinatal viral pathogen. It rarely is acquired in utero (5% of cases); most commonly, it is acquired at the time of parturition (85% of cases). It can be acquired after birth from infected persons who handle an infant. The typical presentation for HSV infection in the first days after birth is mucocutaneous, not systemic or neurologic.

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Critique: 131



Congenital cytomegalovirus infection often results in microcephaly.

Courtesy of B. Carter

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Critique: 131



Congenital cytomegalovirus infection may produce petechiae or purpuric macules, as seen on the face of this infant.

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Critique: 131



Infants who have congenital cytomegalovirus infection have hepatomegaly and splenomegaly (arrows show lines on the abdomen that denote the positions of the edges of the liver and spleen).

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Question: 132

A 2-year-old boy comes to the emergency department because of a barking cough. His mother reports that he has no fever or shortness of breath, but you note a barking, seal-like cough (Item Q132). His respiratory rate is 20 breaths/min, and there is no stridor. His lungs are clear, and other findings on the physical examination are normal.

Of the following, the MOST appropriate treatment is

A. cool mist therapy

B. helium/oxygen mixture

C. nebulized albuterol

D. nebulized racemic epinephrine

E. oral antibiotic

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Critique: 132 Preferred Response: A

The child described in the vignette has a barking cough but no other respiratory symptoms, which is most consistent with the diagnosis of laryngotracheobronchitis or croup. Croup may be caused by a number of respiratory viruses, including parainfluenza, influenza, respiratory syncytial virus, and adenovirus. Typical features are rhinorrhea and low-grade fever, followed by a barking cough and hoarseness. In severe cases, inspiratory stridor may be noted. Children who have croup generally appear well and tolerate oral intake well. Toxic appearance, drooling, and significant respiratory distress should alert the clinician to the possibility of a more serious airway infection, such as bacterial tracheitis or epiglottitis.

The mainstay of therapy for children who have simple viral croup is aerosolized cool mist therapy, which is administered best with the child seated on the parent's lap. The mist thins and moistens airway secretions to improve clearance. Although a recent review of clinical trials found little benefit of mist over no therapy in children who had acute croup, it is a safe and easily administered therapy that may be soothing to the inflamed mucosa. There is good evidence that administration of steroids, either systemic dexamethasone or nebulized budesonide, improves the clinical course by reducing laryngeal mucosa inflammation, so this could be added to the cool mist for maximum benefit.

Both helium/oxygen and racemic epinephrine have been shown to be beneficial in the treatment of moderate-to-severe croup, but the child in the vignette has no evidence of respiratory distress or stridor, so these therapies are not indicated unless the child's condition worsens. Nebulized albuterol may be helpful if wheezing was present and lower airway bronchospasm was suspected. Oral antibiotics have no role in the management of croup.

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Question: 133

A 15-year-old high school baseball player presents for his annual sports physical. As you complete the adolescent interview, he acknowledges using chewing tobacco “because all the other guys do it and the pros do it.”

Of the following, the BEST anticipatory guidance is to tell this teen that

A. chewing tobacco may cause halitosis

B. his risk for adenocarcinoma of the mouth and tongue is increased

C. his risk for lung cancer is less than with cigarettes

D. possession of tobacco in any form is illegal at his age

E. you plan to inform his parents and coach of his tobacco use

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Critique: 133 Preferred Response: A

Although the long-term sequelae of both smoked and smokeless tobacco are well known to most adolescents, rarely are these children aware of the immediate consequences. Counseling about increased risk for oral cancer, lung cancer, and cardiovascular/cerebrovascular disease rarely is helpful in the adolescent and young adult. Many teenagers have a sense of immortality and lack the abstract thinking necessary for future planning. It is well known that peer pressure has a greater impact on behavior in this age group than knowledge of potential future consequences.

There is no evidence that a tobacco cessation program employing “scare tactics” is effective in young people, but providing facts regarding risk-taking behavior may be helpful. The obvious hygiene issue with tobacco is that the odor of cigarettes lingers after smoking, and chewing tobacco causes halitosis, both of which may be socially unacceptable.

Young athletes who smoke complain of cough and shortness of breath more frequently than teammates who do not smoke. Further, although smokeless tobacco does not cause a difference in reaction time, it may cause diminished maximum force and force velocity in exercise. Nicotine also increases energy expenditure during exercise, which may result in earlier fatigue in distance runners and during other similar sustained activity. Increased catecholamine release during exercise in smokers may increase heart rate and vasoconstriction.

Many young athletes (especially baseball players for whom the “culture” introduces chewing tobacco) believe that nicotine may enhance sports performance, but multiple studies have failed to indicate any performance advantage associated with this drug. Such information might be incorporated into preparticipation sports physicals and annual health supervision visits.

Adolescents who are addicted to nicotine may report anxiety, tension, and irritability when they are denied access to tobacco products. This may be a talking point for cessation by pointing out that these symptoms may affect school performance and relationships with peers adversely.

Unfortunately, once addicted, smoking cessation can be difficult for the young person. Even in studies where objective measures were used to give feedback to adult smokers, such as exhaled carbon monoxide and spirometry, no evidence suggested any significant effect of such information on smoking cessation. Advising the teen that his risk for lung cancer is less with chewing tobacco than with cigarettes, although true, ignores the increased risk of oropharyngeal cancer. Discussion of the illegality of tobacco possession is unlikely to stop his use, and most teens already are aware of this fact. Disclosing his tobacco use to his parents would violate the confidentiality of the physician-patient relationship and likely dissuade him from seeking needed medical attention in the future.

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Question: 134

You are evaluating a 3-year-old boy in the urgent care clinic for fever. His mother tells you that he had been well until yesterday, when he had a temperature that she recorded by oral thermometer at 103°F (39.5°C). He has had a clear nasal discharge, cough, and one episode of emesis. At the time of your evaluation, the patient is eating ice chips from a cup that he is holding while he is sitting on the bed. He has a temperature of 102.7°F (39.3°C), a heart rate of 140 beats/min, a respiratory rate of 30 breaths/min, and a blood pressure of 110/66 mm Hg. He has coarse breath sounds with good air movement bilaterally. His pulses are strong throughout. His capillary refill time is between 3 and 4 seconds in his hands and 2 seconds in his feet.

Of the following, the BEST plan of management is

A. blood pressure measurement and pulse oximetry in the four extremities

B. echocardiography for coarctation of the aorta

C. empiric intravenous antibiotics for suspected bacteremia

D. inotropic therapy with dopamine for shock

E. repetition of the perfusion examination with the patient supine and hands warmed

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Critique: 134 Preferred Response: E

The principal function of the heart is to deliver oxygenated blood to the organs and cells of the body so that they may carry out their metabolic function. Inadequate perfusion of the cells and organs results in shock. A number of physical examination findings that correlate to diminished perfusion can be useful in assessment of the ill child. Perfusion of the extremities may be assessed by the quality of the distal pulses, the color and temperature of the skin, and the capillary refill of the tissues. Perfusion of the kidneys may be ascertained by following the urine output over time. Cerebral perfusion can be assessed by the appropriateness of child's behavior and level of consciousness.

Distal pulses should be equal in quality and timing to the central pulses (femoral, axillary, carotid). The upstroke should be brisk. It is always important to compare the right brachial or right radial pulse to the right femoral pulse to evaluate for the possibility of coarctation of the aorta. The extremities should be warm to the touch and pink. Cool, mottled, or cyanotic extremities imply diminished perfusion. Of course, assessment should be made at room temperature, and the patient should not have any external factors that are likely to affect the accuracy of the examination, such as the placement of ice to an injury, holding a popsicle, or having just been in a cold environment. Capillary refill is assessed by holding the extremity at the same level as the heart and gently pressing until there is blanching. Blood should return to the tissue within 2 seconds in the well-perfused extremity at room temperature being evaluated at the level of the heart.

The febrile child in the vignette has delayed capillary refill in his hands, but not in his feet, which can be explained by the ice chips that he is holding. His perfusion, and specifically his capillary refill, should be re-evaluated when his hands have been warmed. A febrile patient may have diminished perfusion, but a differential capillary refill is unexpected. All other aspects of his physical examination and behavior suggest fever without diminished perfusion, and neither empiric antibiotics nor inotropic therapy is warranted. Coarctation leads to differential blood pressure and pulse findings when extremities that receive their blood flow proximal to the area of coarctation are compared with those that are distal. Blood pressure measurement and pulse oximetry in all four extremities are redundant tests in a 3-year-old child and would not yield relevant clinical information.

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Question: 135

A 12-year-old girl in your practice had been born preterm and presented in early childhood with developmental delay. She was diagnosed with cerebral palsy and has been given physical, occupational, and speech therapy. Magnetic resonance imaging at age 3 years showed white matter volume loss, particularly adjacent to the ventricles. The parents are concerned their daughter's condition may be degenerating because she has fallen progressively further behind her peers, and she has become increasingly anxious and oppositional. She has not developed seizures. Her recent special education re-evaluation at school revealed verbal and performance intelligence quotients in the 70s, unchanged from 3 years ago. On physical examination, you note dolichocephaly, hyperreflexia at the knees, and two beats of clonus at each ankle.

Of the following, the MOST appropriate next step is

A. follow-up magnetic resonance imaging

B. measurement of urine organic acids

C. referral for behavioral therapy

D. referral to genetics

E. sleep-deprived electroencephalography

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Critique: 135 Preferred Response: C

The child described in the vignette has cerebral palsy, a static encephalopathy due to her preterm birth, and a typical constellation of motor, cognitive, and mood problems. Her physical examination findings are classic for cerebral palsy due to prematurity, with narrowing of the head due to brain volume loss and upper motor neuron signs affecting predominantly the legs.

Occasionally, new neurologic problems, such as seizures or dystonia, may emerge and superimpose on the stable brain lesions in cerebral palsy as the central nervous system (CNS) matures. Children who have stable, nonprogressive cognitive, motor, and behavioral disabilities may appear to their caregivers to be deteriorating due to the emergence of new mood problems such as anxiety or because academic achievement has leveled off at a time when the achievement of the child's peers is accelerating. The most important intervention at this point is referral for behavioral therapy. Both the parents and the child may need support as the child moves into adolescence, and the child may benefit from therapy to cope with anxiety.

Several features of this case make the presence of a degenerative CNS disorder unlikely. First, the school, as part of a standard re-evaluation, found no evidence of cognitive deterioration (the intelligence quotient was unchanged). Second, no loss of acquired skills is reported. Third, findings on neurologic examination appear to be stable and consistent with cerebral palsy due to prematurity.

In this setting, follow-up neuroimaging with magnetic resonance imaging has limited value. Measurement of urine organic acids is not indicated because the diagnosis of cerebral palsy has been established and in the absence of worsening symptoms or signs, an underlying metabolic disease is unlikely. For the same reasons, referral to genetics is unlikely to lead to a new diagnosis. Electroencephalography (EEG) to evaluate for developmental disabilities in the absence of a clinical history of epilepsy has low yield and generally does not provide useful information. EEG findings may be normal or nonspecific.

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Question: 136

A neighbor approaches you regarding her concerns about her newborn grandson. Her 23-year-old daughter, the mother of the baby, had an uneventful pregnancy, labor, and delivery, and "all of her prenatal tests were normal." The baby has been in the normal newborn nursery for 3 days, and he is feeding poorly. He "just can't seem to get the hang of sucking." She thinks he is probably just "lazy," and she describes him as feeling like a "rag doll" when she holds him. She has noticed that he has "a lot of skin" at the back of his neck, and his fifth fingers are "crooked" (Item Q136). She thinks his penis is small. You suggest that she alert the baby's physician to her concerns.

Of the following, the MOST likely diagnosis is

A. 22q11 deletion

B. achondroplasia

C. cri-du-chat syndrome

D. Down syndrome

E. Prader-Willi syndrome

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Question: 136



Deformity of the fifth fingers, as described for the infant in the vignette.

Courtesy of M. Rimsza

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Critique: 136 Preferred Response: D

The hypotonia, poor feeding, excess nuchal skin, incurving (clinodactyly) of the fifth fingers, and questionably small phallus described for the infant in the vignette are consistent with Down syndrome (DS). Hypotonia and excess nuchal skin are present in 80% of newborns who have DS, fifth finger clinodactyly is present in approximately 50% (Item C136A), and a relatively small penis with reduced testicular volume occurs frequently. Other common features in the newborn who has DS are: poor Moro reflex (85%), joint hyperextensibility (80%), flat facial profile (Item C136B) (90%), upslanting palpebral fissures (80%), and dysplastic ear pinnae (60%).

Babies who have Prader-Willi syndrome (PWS) also exhibit hypotonia, and there may be a small phallus and undescended testes in affected males. However, their facial features are distinct from those of DS; PWS is associated with almond-shaped eyes (Item C136C), bifrontal narrowing, and a narrow upper lip. The hands may have tapered fingers, and hands and feet tend to grow at a reduced rate, resulting in small hands and feet in later life.

Achondroplasia is associated with mild hypotonia, although affected newborns usually have an effective suck. Newborns typically are below the 10th percentile for length and have proximal shortening of arms and legs. There is typically macrocephaly, frontal bossing, and a depressed nasal bridge (Item C136D). The hands have a “trident” appearance due to a relatively large gap between the middle and distal phalanges of the third and fourth fingers (Item C136E).

22q11 deletion syndrome also is associated with hypotonia, and the distinctive facies includes a prominent nose with squared nasal root and pinched alae nasi, narrow palpebral fissures with hooded eyelids, and a small chin. Many affected individuals have cleft palate or submucous cleft palate.

Cri-du-chat syndrome (5p-) is characterized by hypotonia as well as downslanting palpebral fissures, ocular hypertelorism, and a catlike cry in the newborn period. Clinodactyly is not an associated feature, but a single transverse palmar crease is present in approximately 80% of affected individuals.

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Critique: 136



Clinodactyly (incurving) of the fifth fingers occurs in 50% of children who have Down syndrome.

Courtesy of M. Rimsza

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Critique: 136



Dysmorphic features of Down syndrome include a flat facial profile and upslanting palpebral fissures.

Courtesy of M. Rimsza

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Critique: 136

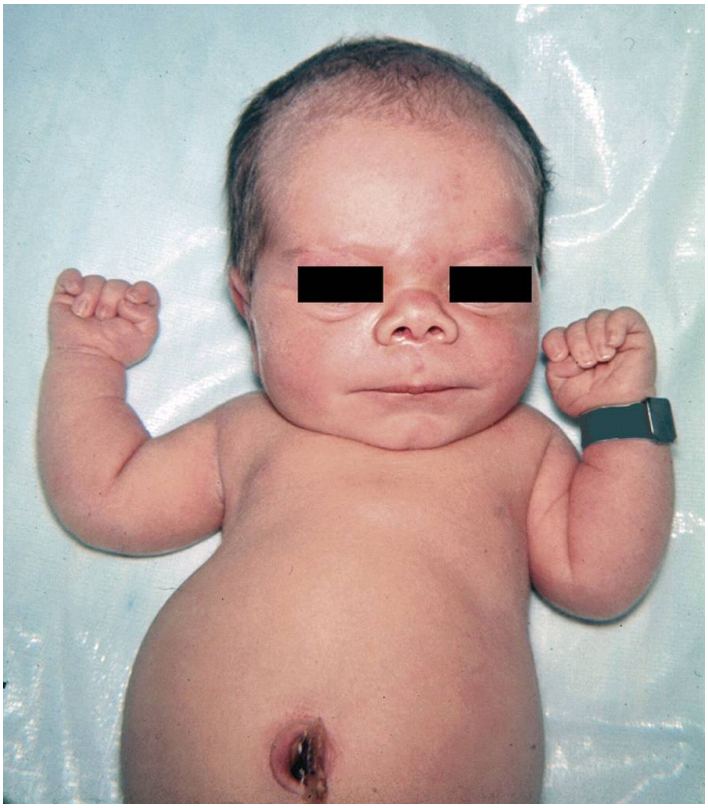


A 3-month-old who has Prader-Willi syndrome: Note the almond-shaped eyes and downturned mouth.

Reprinted with permission from Jonas JM, Demmer LA. Genetic syndromes determined by alterations in genomic imprinting pathways. *NeoReviews* 2007;8:e120-e126

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Critique: 136



Infants who have achondroplasia exhibit macrocephaly and a depressed nasal bridge. Another feature observed in this infant is a small chest compared with the abdomen.

Courtesy of M. Rimsza

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Critique: 136



For patients who have achondroplasia, tapered fingers and a relatively large gap between the third and fourth fingers may lead to a "trident" appearance of the hand.

Courtesy of T. Jewett

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Question: 137

During the health supervision visit for a 15-year-old girl, her mother states that her daughter is currently suspended from school for fighting and that she was previously caught carrying a knife to school. You decide to counsel them both about adolescent violence.

Of the following, a TRUE statement is that

A. effective violence intervention programs have a single primary focus

B. female sex is a risk factor for school violence

C. previous victimization is not associated with future perpetration of a violent act

D. risk factors for school violence include attendance at small rural schools

E. successful violence intervention programs stress conflict resolution in conjunction with mentoring

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Critique: 137 Preferred Response: E

Violence rates in the United States generally are declining, but rates for adolescent violence are increasing. Adolescents are increasingly the perpetrators of violence as well as victims. School-associated violence encompasses a wide variety of behaviors such as bullying, fighting, assault, and gang violence. Risk factors associated with perpetrating school violence are similar to those for school violence victimization. Large urban schools show higher rates of most types of school violence, including vandalism, fighting, larceny, and homicide. Although violent school deaths occur in all communities, the estimated rate of school homicide in urban school districts is nine times higher than that of rural school districts. Personal risk factors for school violence include male sex, alcohol or other drug use, poor impulse control, gang involvement, younger age (12 to 14 years of age), a history of carrying weapons, and a history of fighting and aggression.

Adolescent violence can beget further violence: both perpetrators and victims of violence are at risk of committing future violent acts, including interpersonal violence, date rape, gang violence, and homicide. Effective violence intervention programs have a multidisciplinary approach that involves families, physicians, mental health professionals, social services, and often the criminal justice system. The goals of these programs are to identify and modify characteristics associated with violence, promote self-esteem, promote anger management skills, treat alcohol and other drug disorders, and prevent future violent behavior. Strategies to prevent youth violence have been studied to develop best practice recommendations. Successful strategies to prevent youth violence stress nonviolent problem-solving techniques, conflict resolution skills (eg, negotiation, mediation), and mentoring. Other successful strategies include use of social-cognitive interventions to equip young people with skills to deal effectively with difficult social situations, home visiting, and parent- and family-based interventions designed to improve family relations.

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Question: 138

You are caring for a 12-year-old boy who has been receiving 125 mcg/d of thyroxine for treatment of congenital hypothyroidism. He has been growing well and doing well in school. His thyroxine and thyroid-stimulating hormone (TSH) values have been normal and stable for the past year. At this visit, you measure a free thyroxine (fT4) concentration of 2.5 ng/dL (32.3 pmol/L) (normal, 0.8 to 2.0 ng/dL [10.3 to 25.7 pmol/L]) and a TSH concentration of 12.6 mIU/L (12.6 mU/L) (normal, 0.5 to 5.0 mIU/L [0.5 to 5.0 mU/L]).

Of the following, the MOST likely reason for these abnormal laboratory study results is

A. patient nonadherance with therapy

B. pubertal thyroid hormone resistance

C. pubertal TSH resistance

D. resetting of normal TSH concentrations due to in utero hypothyroidism

E. substitution of generic thyroxine by pharmacy

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Critique: 138 Preferred Response: A

The child described in the vignette has an elevation in both his free thyroxine (fT4) and thyroid-stimulating hormone (TSH) values. The most common reason for these findings in an individual taking thyroid hormone is medication mismanagement. Although it is reasonable to take a double dose of medication if the thyroxine was forgotten the day before, both the TSH and the fT4 are likely to be elevated for up to 7 days because of discordance between thyroxine administration and TSH suppression. Because medication mismanagement is so common, TSH alone never should be used to monitor the need for changes in thyroxine dosage.

Thyroid hormone resistance is a rare genetic condition in which one of the thyroid hormone receptor genes is mutated and does not bind triiodothyronine appropriately so that the TSH and fT4 concentrations remain elevated to maintain cellular euthyroidism. This disorder is not acquired at puberty.

TSH resistance is a rare genetic disorder due to a mutation in the TSH receptor gene or an acquired disorder following blockade of the TSH receptor by nonfunctioning antibodies (blocking antibodies). Blocking antibodies may be passed transplacentally and cause transient hypothyroidism in a newborn. This disorder also is not acquired at puberty.

Prolonged hypothyroidism in the newborn period might "reset" the TSH response and make TSH harder to suppress, but this is rare today, probably because treatment of congenital hypothyroidism usually is initiated at a higher dose of thyroxine than in the past. This disorder would not be acquired after many years of adequate thyroxine replacement.

Although there is ongoing concern about generic substitution of thyroxine, recent data from the United States Food and Drug Administration have shown that most generic forms of thyroxine are equivalent to brand name products.

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Question: 139

The mother of one of your patients is in the process of getting a divorce and has just moved into a small apartment. Due to her new work schedule, she has not been able to unpack the boxes left in the kitchen. She brings her 4-year-old daughter to your office because the child cut herself with a knife trying to open one of the unpacked boxes. The mother is visibly upset. After you bandage the daughter's injured hand, you sit down to talk with the mother.

Of the following, you are MOST likely to

A. explore what support system the mother has to help her family settle into their new home

B. recommend that the mother place her child in time-out for playing with the knife

C. recommend that the mother have a psychiatric evaluation

D. refer the mother to parenting classes

E. tell the mother that you are obligated to report her to the child welfare agency

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Critique: 139 Preferred Response: A

Injuries are the leading cause of death and disability in children and young adults. Injuries often occur when parents are under stress and not observing the child directly. Situations that frequently are associated with accidents are hunger and fatigue in the hour prior to dinner, mother's pregnancy, illness or death in the family, changes in regular child care, tension between parents, and sudden changes in the environment. Divorce often leads to economic and emotional stress, and for many women, it means returning to work or moving to more affordable housing, which may cause additional stress on the child.

The woman described in the vignette is under substantial stress. The pediatrician can be most helpful by identifying support systems in the woman's family or community to assist her during this difficult time. This can promote her ability to provide a stable, nurturing home for her child. If necessary, the pediatrician also can provide age-appropriate anticipatory guidance regarding injury prevention.

Time-out is effective immediately after an inappropriate behavior, not at this later time. Because this mother does not display signs of psychopathology, she does not need a psychiatric evaluation. Parenting classes may be indicated if these problems persist, but advising the mother to attend parenting classes at this time may increase her stress. There is no evidence of child maltreatment or neglect that requires reporting of the mother for child abuse.

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Question: 140

During your morning nursery rounds, you find you have a new patient who was born to a mother infected with human immunodeficiency virus (HIV). You introduce yourself to the mother, and she asks you about any precautions she needs to take in the care of her newborn due to her HIV infection.

Of the following, you are MOST likely to tell the mother that she should

A. add a teaspoon of liquid bleach to the infant's bath water

B. avoid breastfeeding

C. avoid sharing eating utensils

D. take no specific action

E. wear gloves while changing diapers

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Critique: 140 Preferred Response: B

The risk of transmission of human immunodeficiency virus (HIV) to an infant from an infected mother without interventions is approximately 15% to 25%. Breastfeeding by an infected mother increases the risk by 5% to 20% to a total of approximately 20% to 45%. Therefore, in countries where safe alternatives to breastfeeding are readily acceptable, feasible, available, affordable, and sustainable, avoidance of all breastfeeding by HIV infected mothers is recommended. In countries where there are no safe alternatives to breastfeeding, the World Health Organization supports exclusive breastfeeding for the first 6 postnatal months due to the higher death rates for children who receive formula.

HIV is spread through contact with infected bodily fluids such as blood and seminal fluid. Routine activities of daily living are not associated with an increased risk, so others living in the house can share eating utensils, change diapers without gloves, and kiss infected individuals without a risk of infection. Sharing of razor blades, however, should be avoided. Additions to the bath water, such as bleach or special soaps, are not indicated.

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Question: 141

A 3-year-old patient who has acute lymphoblastic leukemia is admitted to the pediatric intensive care unit after developing severe sepsis due to *Pseudomonas aeruginosa*. She is intubated, ventilated, and requires intensive vasopressor support.

Of the following, the MOST appropriate antibiotic regimen for the treatment of this patient is an aminoglycoside plus

A. cefazolin

B. cefdinir

C. ceftazidime

D. ceftriaxone

E. cefuroxime

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Critique: 141 Preferred Response: C

Pseudomonas sp are gram-negative bacilli of relatively low virulence that are primarily waterborne and soilborne organisms. They are associated with a wide variety of infections. *P aeruginosa* is the most common pseudomonal species. It is classified as an opportunistic pathogen that infrequently causes disease in healthy hosts but is a major cause of infection in patients who have underlying conditions, such as the child described in the vignette.

Serious *P aeruginosa* infections, such as bacteremia, sepsis, and pneumonia, are more likely in patients who have underlying conditions, especially cystic fibrosis. Initial therapy usually consists of two antipseudomonal agents, such as a beta-lactam antibiotic (piperacillin, piperacillin-tazobactam, or ceftazidime) plus an aminoglycoside (gentamicin, tobramycin, or amikacin) until results of susceptibility tests are available. Imipenem and ciprofloxacin also may be used as single agents in such patients. The treatment of pneumonia usually includes the beta-lactam antibiotics ticarcillin, ticarcillin-clavulanic acid, piperacillin, piperacillin-tazobactam, or ceftazidime, usually in combination with an aminoglycoside. Combination therapy frequently is used to ensure treatment of resistant strains and to prevent selection of resistant mutants. The carbapenem class of antibiotics (imipenem or meropenem) and the monobactam antibiotic aztreonam generally are reserved for the treatment of serious infections caused by organisms resistant to the other beta-lactam antibiotics or for patients who have moderate renal disease and are at risk for aminoglycoside-related nephrotoxicity. These agents may be used as monotherapy or in combination therapy.

Cefazolin, cefdinir, ceftriaxone, and cefuroxime have no activity against pseudomonal organisms.

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Question: 142

You are seeing an 8-month-old infant for a health supervision visit. He was born at 28 weeks' gestation, weighed 1,200 g at birth, and has bronchopulmonary dysplasia. His only medication is furosemide, which he has been receiving for several months.

Of the following, the MOST likely expected abnormality in this infant is

A. hypercalciuria

B. hypermagnesemia

C. hypocalcemia

D. hyponatremic dehydration

E. metabolic acidosis

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Critique: 142 Preferred Response: A

Furosemide is a loop diuretic that acts at the ascending limb of the loop of Henle (LOH). Normally, approximately 20% to 25% of filtered sodium is reclaimed at this site, so blockade of sodium reabsorption results in a brisk diuresis, with urinary losses of sodium, potassium, and chloride. The blocked transporter is the Na+-K+-2Cl- channel.

In addition to direct effects on the electrolytes of this channel, loop diuretics influence calcium and magnesium transport. Calcium and magnesium are reabsorbed passively in the ascending limb of the LOH through paracellular transport as a result a gradient derived from normal sodium chloride transport. The passive reabsorption of calcium is believed to occur through a paracellular pathway that is facilitated by paracellin-1, a tight junction protein. Interruption of sodium chloride reabsorption results in impaired calcium reabsorption and consequent hypercalciuria and increased risk for nephrocalcinosis and kidney stones.

Furosemide also has an effect on the composition of endolymph in the inner ear because the Na+-K+-2Cl- transporter that is in the ascending limb of the LOH also can be found in the marginal cells of the cochlear duct of the inner ear. Furosemide induces changes in ion transport in these cells, reducing endocochlear potentials in the cells, which results in hearing loss. Ototoxicity is more likely when loop diuretics are used in high doses or are used in conjunction with other ototoxic agents such as aminoglycosides. Furosemide-induced ototoxicity usually is temporary, but can be permanent in some cases.

Patients receiving loop diuretics also may develop hypokalemia, metabolic alkalosis, and hypomagnesemia (from increased urinary losses). Abnormalities in serum calcium are not typical, despite hypercalciuria. Hypercalciuria can lead to nephrocalcinosis, which can be longstanding, even after discontinuation of furosemide. Patients receiving furosemide are at risk for dehydration, but this is typically hypernatremic dehydration; hyponatremic dehydration is more common with thiazide diuretics.

References:

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Question: 143

A 17-year-old girl presents for a health supervision visit before leaving for college. As you review her medical history, you note that she has marked “penicillin allergy” on the school health form. She remarks that her mother told her she had a rash after amoxicillin when she was 2 years old.

Of the following, the BEST statement regarding penicillin drug reactions is that

A. first-generation cephalosporins are less likely to cause a reaction in penicillin-allergic patients compared with third-generation cephalosporins

B. negative skin testing to major and minor determinants of penicillin can exclude almost all immunoglobulin (Ig) E-mediated reactions

C. nonpruritic maculopapular rash that occurs in patients who receive amoxicillin during mononucleosis is a contraindication for future penicillin therapy

D. serum sickness reactions due to penicillin usually are IgE-mediated

E. the incidence of IgE-mediated penicillin allergy among patients who have this history is greater than 20%

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Critique: 143 Preferred Response: B

As is often the case, the patient described in the vignette can only recall what her parents remembered about her drug reaction. Although the incidence of a true immunoglobulin (Ig) E-mediated penicillin allergy is 10% or less in this scenario, most clinicians continue to avoid this drug class in such patients. The administration of a penicillin during mononucleosis often results in a nonpruritic, maculopapular rash (Item C143) within a few days. The mechanism for the rash is unknown, but this reaction is not IgE-mediated and should not preclude future penicillin use. For patients who have experienced a suspected IgE-mediated penicillin reaction, the use of cephalosporins generally is endorsed for those whose previous reaction did not result in severe anaphylaxis. Further, the second- and third-generation cephalosporins are less likely to cross-react with penicillin than are first-generation cephalosporins. Overall, the risk for cross-reaction remains less than 10% for all cephalosporins.

Although not commercially available in the United States, skin testing to both major (penicilloyl) and minor (penicillin, penilloate, penicilloate) determinants can be very helpful. Negative skin test results to both major and minor determinants virtually rule out (>97% chance) that the patient is allergic to penicillins. On the other hand, positive skin test results are approximately 60% predictive for ruling in a penicillin allergy.

Serum sickness, erythema multiforme, fixed drug reactions, and toxic epidermal necrolysis are non-IgE-mediated penicillin reactions. Such reactions are not identified with skin testing because they are not IgE-mediated. When these reactions occur after penicillin administration, future penicillin use is contraindicated.

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Critique: 143



A generalized macular and papular eruption can follow the administration of a penicillin in those who have infectious mononucleosis.

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Question: 144

You are on the sidelines of a girl's high school lacrosse game when one of the players is struck in the mouth with the ball. She does not lose consciousness but runs off the field complaining of severe mouth pain. On inspection of her mouth, you observe profuse bleeding, lacerations of the upper alveolar mucosa, and avulsion of the right upper central incisor. Her teammate finds the missing tooth on the field and brings it to you. You apply direct pressure to the bleeding gums and make arrangements for the player to be transported to the emergency department.

Of the following, the MOST appropriate method for preparing the tooth for transport with the patient is to

A. dispose of the tooth because it cannot be saved

B. have the player hold the tooth inside her cheek

C. place the tooth on ice in a plastic bag

D. rub all debris from the tooth and place it in a dry plastic bag

E. wrap the tooth in a paper towel moistened with water

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Critique: 144 Preferred Response: B

Avulsion of a permanent tooth is a true dental emergency. The ultimate prognosis of the tooth and future need for dental restoration are correlated directly with the time taken to reimplant the avulsed tooth in the socket. A tooth that is reimplanted within 5 minutes of avulsion has an 85% to 97% chance of survival; a tooth reimplanted after 1 hour is unlikely to remain viable.

Reimplantation is a relatively simple procedure and can be performed by any available adult, including a parent, coach, teacher, or pediatrician. Holding the tooth by the crown, it is inserted into the socket with gentle pressure. The child is directed to hold the tooth in place manually during transport. The tooth should be rinsed gently before reimplantation, if possible, but never rubbed or scrubbed because vigorous manipulation can damage the periodontal ligament fibers. If a tooth cannot be reimplanted immediately because of significant damage to the alveolar tissues or inability of the child to cooperate, the tooth and the child should be transported to a facility that can perform reimplantation (eg, dentist's office or emergency department). The exception to this recommendation is avulsion of a primary tooth, which never should be reimplanted because of concern for further injury to the successor tooth.

An avulsed tooth that cannot be reimplanted immediately should be transported in a medium that maintains the viability of the periodontal ligament fibers, which can maximize the likelihood of tooth survival. Solutions that have been shown to support periodontal ligament survival in vitro for several hours include special cell culture media and Hank's balanced salt solution. Alternatives include milk, saliva (the cooperative child can hold the tooth in the buccal vestibule), or isotonic saline. If no other alternatives are available, tap water is preferred over transporting a dry tooth, although the low osmolality of the water causes periodontal cell death within minutes. Transporting the tooth in chilled media is preferred, but not directly on ice, which can freeze the ligamentous fibers.

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Question: 145

You are evaluating a newborn who has complete heart block and several 1.5-cm erythematous macules and annuli located on the forehead, behind the ears, and in the scalp (Item Q145).

Of the following, the test MOST likely to confirm the infant's diagnosis is

A. anti-Ro (SSA) antibodies

B. creatine kinase

C. hepatic function profile

D. platelet count

E. urinalysis

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Question: 145



Courtesy of D. Krowchuk

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Critique: 145 Preferred Response: A

The infant described in the vignette has complete heart block and erythematous annular plaques on sun-exposed areas, suggesting a diagnosis of neonatal lupus erythematosus (NLE). NLE is a rare disorder caused by transplacental passage of maternal autoantibodies. In the majority of cases (95%), the antibodies responsible are anti-Ro (SSA) antibodies alone or in association with anti-La (SSB) antibodies. A small proportion of patients and their mothers exhibit anti-U1RNP antibodies. Performance of these antibody studies on the infant and mother (if she has not been evaluated previously) help confirm the diagnosis.

Skin disease is the most common manifestation of NLE, being present in 50% of affected infants. Lesions usually begin at about 6 weeks of age but may be present at birth. Infants exhibit erythematous, scaling annular plaques in sun-exposed areas (Item C145). When present around the eyes, as occurs frequently, they may give the appearance of “raccoon eyes." Lesions resolve spontaneously, usually without scarring, and new lesions cease to appear after about 3 months of age. Treatment includes the application of a low-potency topical corticosteroid and sun avoidance.

NLE also may be associated with systemic complications involving the heart, liver, hematopoietic system, and rarely, the kidney. Congenital heart block resulting from inflammation and fibrosis of the sinoatrial node is observed in 15% to 30% of affected infants. Heart block may be asymptomatic or cause fetal bradycardia, intrauterine congestive heart failure, hydrops fetalis, or neonatal death. Infants who have complete heart block may require pacemaker insertion, but death rates as high as 15% have been reported despite this intervention.

Approximately 15% of infants who have NLE have hepatic involvement either alone or in association with cardiac, skin, or hematologic abnormalities. Typically, affected infants exhibit transient hepatomegaly, biochemical evidence of cholestasis, and possibly, transaminase elevations. Thrombocytopenia, occasionally accompanied by anemia or leukopenia, occurs in as many as 10% of affected infants. Platelet numbers generally normalize over the first weeks after birth, and treatment with high-dose corticosteroids only occasionally is required.

In view of the possibility of systemic involvement, infants who have NLE should undergo screening tests, including an electrocardiogram, complete blood count, platelet count, and hepatic function profile. Urinalysis generally is unnecessary because glomerulonephritis complicating NLE is rare and transient. Myopathy is not a feature of NLE and, therefore, measurement of a creatine kinase concentration is not indicated.

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Critique: 145



Lesions of neonatal lupus erythematosus are erythematous annular plaques located in sun-exposed areas.

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Question: 146

A 15-year-old girl presents with an episode of “feeling faint” and melena. On physical examination, you note a gallop rhythm and mild, nonspecific abdominal tenderness. Stool is guaiac-positive. Laboratory analysis demonstrates anemia, with a hematocrit of 18% (0.18). You administer fluid resuscitation and packed red blood cells, and the patient’s hemodynamic status stabilizes.

Of the following, the next MOST appropriate diagnostic test is

A. angiography

B. barium contrast upper gastrointestinal tract radiography

C. Doppler ultrasonography of portal and esophageal veins

D. upper gastrointestinal endoscopy

E. video capsule study

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Critique: 146 Preferred Response: D

Upper gastrointestinal bleeding (bleeding proximal to the ligament of Treitz) may be either acute (presenting with melena and hemodynamic instability) or chronic (presenting with anemia). Common causes include gastric or duodenal ulcers, chronic gastritis, esophageal or gastric varices, and reflux esophagitis. Vascular lesions such as arteriovenous malformations or telangiectasias (as seen in hereditary hemorrhagic telangiectasia [Osler-Weber-Rendu disease]) also can present with chronic gastrointestinal blood loss. Dieulafoy lesion is another cause of upper gastrointestinal hemorrhage that presents with massive and recurrent bleeding and is caused by an abnormally enlarged arteriole in the gastric cardia or duodenum that periodically bleeds into the gastric lumen.

Upper gastrointestinal endoscopy remains the initial diagnostic test of choice for most upper gastrointestinal hemorrhages because it is not only highly sensitive for mucosal lesions such as peptic ulcers and varices, but also allows the endoscopist to treat any bleeding lesions. Bleeding varices usually are treated by placing rubber bands around varices (variceal ablation by banding) or by injecting sclerosant into the varices (sclerotherapy). In contrast, bleeding gastric and duodenal ulcers typically are treated by electrocautery of the ulcer or by local injection of epinephrine. Video capsule endoscopy is a new technique in which a patient swallows a small "pill" containing a digital camera and transmitter. The camera can take thousands of pictures of the small intestine, which are "beamed" to a set of leads attached to the patient and downloaded onto a computer hard drive. For patients whose mucosal lesions of the upper gastrointestinal tract are not identified by routine endoscopy, video capsule endoscopy can identify bleeding lesions more distally throughout the jejunum and ileum (Item C146). Ultrasonography can identify varices, but is insensitive for ulcers. Angiography sometimes is useful in patients who have bleeding ulcers that cannot be identified or treated endoscopically. Barium studies are insensitive in evaluating gastrointestinal hemorrhages.

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Question: 147

A woman who has chorioamnionitis and had a positive group B streptococcal (GBS) screening culture at 36 weeks' gestation delivers an infant at term. The infant becomes ill in the first 4 hours after birth, demonstrating tachypnea, inability to maintain temperature, and poor perfusion. He is admitted to the intensive care nursery. On physical examination, there are retractions of the chest wall, coarse and shallow breath sounds, and delayed capillary refill. There is no heart murmur or cyanosis, but arterial oxygen saturation determined by pulse oximetry is only 80% on room air.

Of the following, the clinical manifestation that is observed MORE often in early-onset than late-onset GBS infection is

A. cellulitis

B. meningitis

C. osteomyelitis

D. pneumonia

E. septic arthritis

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Critique: 147 Preferred Response: D

Group B streptococcal (GBS) infection remains a concern for newborns around the world. Although the incidence of early-onset GBS infection has been reduced with the use antepartum screening strategies and intrapartum antibiotic prophylaxis (IAP), clinicians must be familiar with the spectrum of GBS illness.

Early-onset GBS typically presents in the first 24 hours after birth as a systemic sepsis syndrome that involves apnea, respiratory distress from pneumonia or pulmonary hypertension, shock, and occasionally meningitis (<10% of cases). Late-onset GBS infection presents after the first 7 days of postnatal life, most commonly in the third or fourth week. Meningitis is far more common in late-onset GBS infection, but other presentations include septic shock and focal infections such as cellulitis, arthritis, and osteomyelitis.

The infant described in the vignette has early-onset GBS infection, with tachypnea and temperature instability prompting an evaluation for sepsis. His mother, who is a GBS carrier, had chorioamnionitis at the time of delivery, which increases his risk for acquiring GBS. The respiratory distress may be due to pneumonia that is indistinguishable from hyaline membrane disease on chest radiography. In cases of GBS bacteremia, a GBS toxin also may mediate pulmonary vascular constriction and cause pulmonary hypertension, even when pneumonia is not present. With GBS bacteremia or septicemia, approximately 10% of infants have corresponding meningitis.

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Question: 148

An 18-month-old girl is brought to the emergency department by her mother, who reports that the girl has had a high fever and difficulty breathing for 1 day. She has had a barking cough for the last 3 days but had been afebrile and breathing comfortably until today. Her immunizations are up to date. Her temperature is 104ºF (40°C), her respiratory rate is 50 breaths/min, she exhibits inspiratory stridor and a brassy cough, and she appears ill. Lung examination reveals decreased breath sounds bilaterally with transmitted upper airway sounds. An anterior neck radiograph shows subglottic narrowing and a ragged tracheal air column.

Of the following, the MOST likely diagnosis is

A. acute viral laryngotracheitis

B. angioneurotic edema

C. bacterial tracheitis

D. epiglottitis

E. retropharyngeal abscess

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Critique: 148 Preferred Response: C

Bacterial tracheitis is a serious infection causing significant upper airway obstruction. It typically occurs as a secondary staphylococcal infection following viral laryngotracheobronchitis, but it also may develop as a primary infection without preceding symptoms of croup. Clinical features include a toxic appearance, with high fever, tachypnea, and brassy cough, as described for the girl in the vignette. Airway obstruction can be pronounced due to purulent airway secretions, and lower airway disease such as wheezes or rales also may be present. Neck radiographs often reveal a ragged air column or subglottic narrowing, and peripheral white blood cell counts frequently are elevated. Treatment consists of intravenous antibiotics and airway control. Endotracheal intubation frequently is required until the infection is treated and the severity of secretions improves.

Acute viral laryngotracheitis typically causes a barking cough with occasional inspiratory stridor, but affected children usually do not have high fevers or appear ill. Angioneurotic edema is a response to histamine release with an allergic reaction and often is accompanied by urticaria. Upper respiratory tract mucosae become edematous, so inspiratory stridor may be present, but fever is unlikely. Children who have epiglottitis appear toxic and have significant airway compromise and inspiratory stridor due to the swollen epiglottis. Cough is rare. Lateral neck radiograph reveals an enlarged epiglottis (“thumb sign”) (Item C148A) rather than a ragged air column. Further, epiglottitis is unlikely in a child whose immunizations are up to date. A retropharyngeal abscess can develop with pharyngitis, and inspiratory stridor and respiratory distress with high fever is typical. The lateral neck radiograph reveals widening of the prevertebral tissues (Item C148B), and computed tomography scan of the neck can confirm the presence of the abscess (Item C148C).

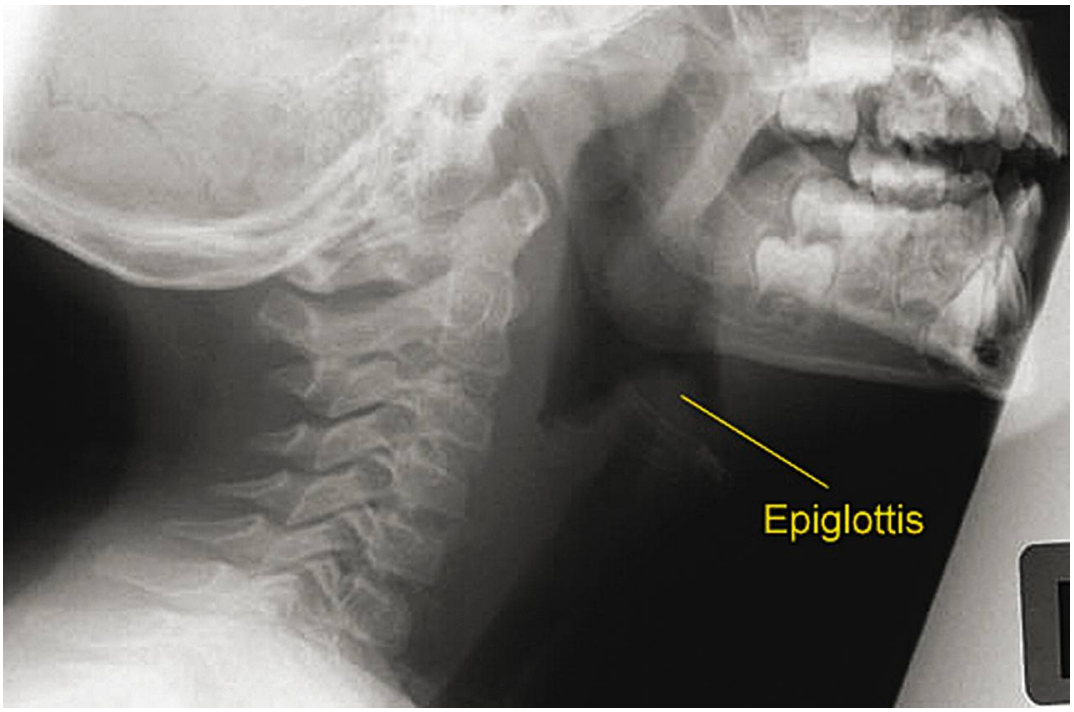
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Critique: 148

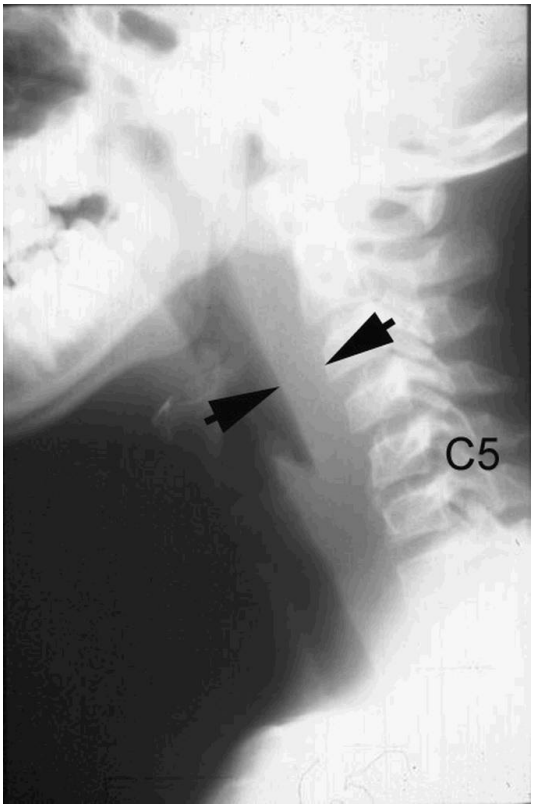


In epiglottitis, a lateral radiograph of the neck reveals a swollen epiglottis (the "thumb" sign).

Courtesy of the Media Lab at Doernbecher

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Critique: 148

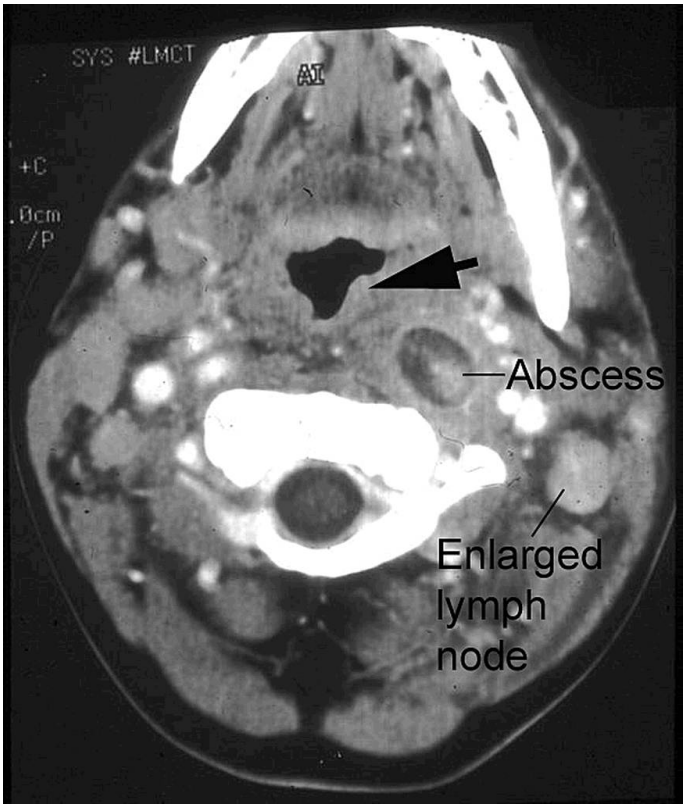


Retropharyngeal abscess: Lateral radiograph of the neck shows widening of the prevertebral soft-tissue space (defined by arrows). Normally, this space should measure no more than one half the width of the fifth cervical vertebral body.

Courtesy of B. Specter

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Critique: 148



Retropharyngeal abscess: Computed tomography scan of the neck at the level of the angle of the mandible shows soft-tissue swelling that impinges on the airway (arrow) and an abscess.

Courtesy of B. Specter

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Question: 149

A 17-year-old girl presents to your school-based clinic with her 3-day-old newborn for an early discharge follow-up visit. The young mother looks very tired and is tearful. She states that she is trying to breastfeed but has to supplement the feeding with a bottle because she “doesn’t have any milk.”

Of the following, the MOST appropriate course of action is to

A. call the infant’s grandmother immediately to come to the office

B. recommend that she stop breastfeeding

C. schedule a same-day visit with a lactation consultant in the office or hospital

D. schedule another follow-up visit in 1 month

E. suggest that she attend prenatal classes for her next pregnancy

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Critique: 149 Preferred Response: C

Although prenatal visits or classes are ideal for pregnant teens, they often are inaccessible and may not be reimbursed by insurers. Several types of prenatal visits with physicians or prenatal classes are directed toward the postnatal issues of infant care. Many parents who can afford to pay for such visits may seek a prenatal consultation, and some physicians do not charge for such visits in order to attract new patients to their practice. The goals of the visit are to establish rapport; gather basic information about the family, pregnancy, and plans for delivery and infant feeding; and disseminate information about the pediatric practice and the medical home. Additionally, providing support to the parents and identifying high-risk situations (both medical and social) are important.

Group prenatal visits, connected physically and temporally with childbirth preparation classes or given in community centers, community health entities, or even high schools, may be ideally suited for the adolescent mother. The goals of these visits are to discuss fears and concerns of the mothers; offer referrals for lactation support, child care, and other resources; teach basics of infant feeding and bathing; address early developmental issues such as colic and non-nutritive sucking; and reinforce the importance of immunizations and the concept of the medical home.

For the young mother described in the vignette, community resources such as referral to a lactation consultant at the hospital where she delivered or in the community, low-cost infant care classes, videotapes of infant care topics, and child care resources for resumption of school or work are important. Cessation of breastfeeding almost never is indicated. Calling the grandmother to help may be appropriate if there are signs of postpartum depression, as might rehospitalization or referral to a mental health professional, but such action is unlikely to empower the young mother. Recommending a prenatal visit for subsequent pregnancies does not help the mother with her current difficulties in caring for her infant. Delaying a follow-up visit for 1 month is inappropriate for a breastfed infant who has risk factors for poor weight gain and for this mother, who is at risk for postpartum depression.

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Question: 150

You are evaluating a 17-year-old boy whom you have known since early childhood. He is complaining of headaches over the past 2 weeks. He has a history of asthma, which has been well controlled, and he is an otherwise healthy member of the varsity football team at school. He has had a significant weight gain of 30 lb (13.5 kg) since his visit to you 1 year ago. He denies using illicit or prescription drugs. On physical examination, he appears very muscular and has a blood pressure of 180/120 mm Hg. You repeat the measurement using a leg cuff to ensure adequate cuff size and obtain the same result.

Of the following, the BEST management plan is

A. angiotensin-converting enzyme inhibition as an outpatient

B. beta blocker therapy as an outpatient

C. diuretic therapy as an inpatient

D. repeat blood pressure measurement in 1 to 2 weeks

E. vasodilator therapy as an inpatient

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Critique: 150 Preferred Response: E

Hypertension is a major cause of morbidity and mortality in adults, and growing data suggest that it is becoming a greater clinical problem in the pediatric population, particularly adolescents. Although yet to be defined clearly, the lifelong risks for the child who has hypertension or a prehypertensive state are likely to be substantial. Blood pressure is affected by height, weight, sex, and race. A complete medical history, particularly family history and medications (including over-the-counter supplements), and a thorough physical examination are essential to early and accurate diagnosis of hypertension and assessment of its secondary causes, comorbidities, and potential complications.

Measurement of the blood pressure is a salient component of the yearly health supervision visit for children beginning at 3 years of age. When the patient is calm and relaxed, blood pressure should be measured in the right arm with the patient seated and the arm resting at the level of the heart. The stethoscope should be placed about 2 cm superior to the cubital fossa, just over the brachial artery. It is extremely important to use the proper size cuff for each patient. The bladder of the cuff (not the cuff material) is the most important determinant of cuff size. The bladder width should cover 60% to 70% of the upper arm length. The cuff bladder length should cover 80% to 100% of the circumference of the arm to ensure complete compression of the brachial artery during cuff inflation. A cuff that is too small will result in a falsely elevated reading. A cuff that appears too large will not affect the measurement adversely. Most errors in blood pressure measurement occur in obese or highly muscularized patients when a cuff is used that is too small.

Severe hypertension and hypertensive crisis should be managed aggressively. The latter typically results from the ingestion of drugs that cause hypertension, injury, or disease of the kidney or previously unrecognized, progressive hypertension. Symptoms of severe hypertension may include headache, changes in vision, epistaxis, seizure, pulmonary edema with congestive heart failure, and those that may arise from renal failure.

The patient described in the vignette has a significantly elevated blood pressure that involves marked and reproducible systolic and diastolic hypertension. The best management plan is to monitor his blood pressure while the cause is ascertained and treatment begun, which involves admission to the hospital and initial treatment with an intravenous antihypertensive agent. The goal of such therapy is to reduce the blood pressure by 25% or less over the first 8 hours and gradually normalize it over the next 48 hours to avoid complications (eg, cerebrovascular accident).

The choice of chronic antihypertensive therapy depends, in part, on the cause of the hypertension, but for immediate short-term management, vasodilators (eg, calcium channel blockers, hydralazine, nitroprusside) are useful. These agents reduce the afterload against which the left ventricle pumps, thereby reducing its work and oxygen consumption. Alternatively, short-acting beta blockers could be used in the acute setting. When using beta blockers, however, the clinician must bear in mind their potential complications, including exacerbation of underlying asthma. Of importance, pharmacologic management of severe hypertension and hypertensive crisis should use medications that can be titrated to effect readily and have a fast onset of action. Diuretics, particularly the thiazide class, often are used as first-line antihypertensive agents for those who have mild or moderate hypertension that can be controlled on an outpatient basis. These may be used in combination with other agents, including but not limited to angiotensin-converting enzyme inhibitors or angiotensin receptor blockers, if adequate control is not obtained with a single agent. The significant hypertension reported for the boy in the vignette requires immediate action; repeating the blood pressure measurement in 1 to 2 weeks is not appropriate.

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Question: 151

A mother brings her 3-year-old boy to the emergency department. She explains that the boy suddenly stopped paying attention, stared, and had jerking of his arms and legs for about 1 minute. His lips turned blue, and he became incontinent of urine. After the episode, he appeared confused and became very sleepy. On physical examination, he has a temperature of 104°F (40°C). Following administration of acetaminophen, his temperature has decreased to 98.6°F (37°C). He is alert, interactive with his parents, and has normal findings on physical examination.

Of the following, the MOST appropriate next step is to

A. begin therapy with carbamazepine

B. obtain magnetic resonance imaging

C. obtain sleep-deprived electroencephalography

D. perform a lumbar puncture

E. provide the family with education

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Critique: 151 Preferred Response: E

A febrile seizure is the most common type of acute seizure in early childhood. Less commonly, other acute stresses to the central nervous system may result in a seizure. Examples include focal lesions affecting the brain (head trauma, intracerebral hemorrhage, stroke, tumor), infection involving the surface of the brain or surrounding tissues (meningitis, encephalitis, abscess), metabolic derangements (involving glucose, sodium, potassium, calcium, magnesium. or toxins), or intrinsic disturbances of neurotransmission (eg, the first seizure in a child who ultimately is diagnosed with epilepsy).

A seizure is an abrupt alteration of neurologic function. A generalized tonic-clonic seizure is characterized by classic motor symptoms (rhythmic jerking) and loss of awareness, followed by sleep. As is often the case, the parent did not know that the child described in the vignette was sick, and the first elevated temperature was detected after the seizure. The child experienced a simple febrile seizure. Simple febrile seizures usually occur in children between the age of 6 months and 6 years, last less than 5 minutes, are nonfocal, and occur at the beginning of the illness.

The family should be educated about febrile seizures. The key components of this education are: 1) seizure first aid, 2) seizure precautions, 3) risk of recurrence of seizures, and 4) prognosis.

First aid: If the child has another seizure, he should be placed on the floor on his side, away from furniture. Some families worry that the child may swallow his tongue and may want to place a spoon or other object in the mouth to prevent this. Parents should be told that tongue swallowing cannot and does not occur, and no object should be placed in the child's mouth. The parent should time the seizure to be able to report to the doctor its duration. If the seizure lasts 5 minutes, the family should call 911 for emergency assistance.

Precautions: Seizure precautions in children prior to driving age involve "wheels and water." This common sense advice includes the wearing of helmets when the child is "on wheels" and adult supervision whenever the child is in water, including bath water. The child can sleep in his or her own bed and does not need to sleep with a parent.

Recurrence risk: In an otherwise healthy child who experiences a single, simple febrile seizure, the recurrence risk for febrile seizures is about 33%.

Prognosis: There is no evidence that simple febrile seizures cause brain damage. The family should be reassured that the risk of epilepsy, ie, recurrent nonfebrile seizures, is less than 5%.

For a 3-year-old child who has a febrile seizure, the focus of the diagnostic evaluation is on the cause of the illness, not on the brain. Neuroimaging is not needed. In the absence of encephalopathy and at age 3 years, lumbar puncture is not recommended routinely. Lumbar puncture is recommended in children younger than 18 to 24 months of age and based on clinical judgment in other settings. There is no role for electroencephalography in the evaluation of a child who has a febrile seizure because it does not provide information that affects management.

Daily anticonvulsant medications such as carbamazepine are not prescribed after one or a few febrile seizures. Some physicians prescribe rectal diazepam or intranasal midazolam to all children after a single seizure to be used in the future should a prolonged seizure occur. This is especially important for a child who had a febrile seizure lasting more than 5 minutes, is medically fragile, or has limited access to medical facilities.

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Question: 152

You care for a 3-year-old girl who was born with spina bifida. The child is otherwise normally formed and is developmentally appropriate with respect to her social, fine motor, and language skills. During a routine visit, her mother tells you that she is contemplating another pregnancy. You mention the important role of folic acid in the prevention of neural tube defects, and you recommend that she discuss this with her obstetrician.

Of the following, the recommendation, implemented before and during pregnancy, that should have the GREATEST impact on reducing this woman's risk for having a future child affected with spina bifida is to

A. consume only grains and cereals that are folic acid-fortified

B. increase consumption of folic acid-rich leafy green vegetables

C. measure the woman's serum folic acid level and supplement accordingly

D. prescribe oral folic acid 0.4 mg daily

E. prescribe oral folic acid 4 mg daily

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Critique: 152 Preferred Response: E

In 1992, the United States Public Health Service issued the recommendation that all women capable of becoming pregnant consume 0.4 mg (400 mcg) of folic acid daily to reduce their risk for pregnancies affected by neural tube defects (NTDs). In 1996, the United States Food and Drug Administration authorized fortification of domestic grain products with folic acid, and by 1998, such fortification was required.

Studies performed by the Centers for Disease Control and Prevention (CDC) and others show that NTD rates have declined about 20% to 30% since the introduction of folic acid-fortified cereal grains. However, this falls short of the approximately 70% decrease in NTDs associated with daily doses of 0.4 mg of folic acid. Many reasons probably account for the discrepancy in outcomes between the two methods of folic acid supplementation, including socioeconomic variables and dietary preferences.

For women who have had a previous pregnancy affected by an NTD, the CDC recommends daily consumption of 4 mg (4,000 mcg) of folic acid daily from the time they begin trying to get pregnant through the first trimester of pregnancy. These same women should consume 0.4 mg of folic acid daily when not trying to become pregnant.

Because the 4-mg dose is ten times higher than that recommended for women at average risk, and because folic acid supplementation via fortified grains has been shown not to be as successful in reducing NTDs as folic acid tablets, the latter are recommended for the mother described in the vignette. Measuring a woman's serum folic acid concentration does not reliably determine her need over an entire pregnancy. Leafy green vegetables are rich in folic acid, and their consumption is encouraged, but the amount any woman consumes is uncertain.

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Question: 153

You are seeing a 15-year-old girl for her first health supervision visit to your practice. In explaining your practice's policies, you discuss confidentiality.

Of the following, you are MOST likely to state that

A. adolescents are more likely to seek health care for sensitive issues if they believe that their parents will be informed

B. billing policies of an outpatient or inpatient facility are always confidential in regard to sexually transmitted disease infection testing for adolescents

C. if an adolescent poses a threat to self or others, confidentiality can be broken

D. parents have access to all of an adolescent's health information through the Health Insurance Portability and Accountability Act of 1996 (HIPAA)

E. state laws mandate that adolescents in all states may receive confidential treatment for alcohol and other drug use disorders

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Critique: 153 Preferred Response: C

Respect for the privacy of individuals dictates that the information shared by adolescent patients with clinicians remain confidential unless there is a legal requirement to disclose information or the information reveals a serious threat to the adolescent's or another individual's health. Research has shown that when adolescents perceive that their health-care service is not confidential, they are less likely to seek care, especially for problems involving reproductive health and alcohol and other drug use. Research also indicates that adolescents are more likely to disclose sensitive information if clinicians make explicit statements about confidentiality.

It is a challenge to provide confidential services to adolescents, especially if they are covered by parental insurance, because billing policies (and electronic medical record systems) may not always have safeguards for confidentiality when billing for a specific test (such as sexually transmitted infection testing). State laws also vary in the extent to which they protect confidentiality of minors, and no law can cover every situation. State laws address alcohol and other drug abuse, but some specify only one or the other. Requirements of disclosure of drug and alcohol use to parents also vary among states. The Center for Adolescent Health and the Law recently published a compendium of state laws that addresses confidentiality and consent and allows clinicians to determine the specifics of the law in their own states of practice.

The Health Insurance Portability and Accountability Act of 1996 (HIPAA) protects confidentiality of adolescents who are considered minors under some circumstances. Parents and guardians have control over health information access for nonemancipated minor children, except in situations in which minors are able to consent to their own health care. The federal law of HIPAA defers to state laws that allow or prohibit disclosure of confidential information to parents. HIPAA allows a physician to disclose information to a parent if a state law requires the physician to do so. If the state law permits, but does not require, disclosure to a parent, HIPAA allows physician discretion for disclosure. If state law prohibits the disclosure of information to a parent, disclosure must not be made without the minor's permission. If there is no state law in place, the physician has discretion about parental disclosure.

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Question: 154

You are discussing treatment choices with the family of a 13-year-old girl in whom you have just diagnosed hyperthyroidism. You include antithyroid drug therapy, surgery, and radioactive iodine treatment in your discussion. They choose antithyroid drug therapy with methimazole for initial therapy.

Of the following, the MOST likely adverse effect of this therapy is

A. agranulocytosis

B. cholestatic jaundice

C. headache

D. hematuria

E. rash

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Critique: 154 Preferred Response: E

Treatment for hyperthyroidism includes thyroid ablative therapy using radioactive iodine, surgical thyroidectomy, or antithyroid drugs. In general, these options all are acceptable, and the decision regarding therapy should be reached jointly by the family and the physician.

Antithyroid drugs are used in the treatment of hyperthyroidism in the hope that the underlying immune disorder will remit spontaneously and long-term medication will not be required. Disease remission occurs in approximately 25% of individuals after each year of treatment. Clinical signs of potentially successful remission include a small thyroid gland and the need for minimal thionamide (eg, methimazole, propylthiouracil) therapy. The laboratory findings indicative of remission include a decrease in thyroid-stimulating immunoglobulin concentrations to the normal range. The most common adverse effects of the thionamide antithyroid drugs such as methimazole and propylthiouracil are rashes, arthritis, and arthralgias, which are considered minor complications. Headache is not a common adverse effect of these agents. More severe complications are relatively rare and include hepatitis. Liver involvement with methimazole is reversible, but propylthiouracil-induced hepatitis may result in liver failure. Neutropenia may be seen with either methimazole or propylthiouracil. It is important to obtain a baseline white blood cell count before starting therapy because thyrotoxicosis itself may be associated with neutropenia. Agranulocytosis is a rare complication, usually occurring in older individuals within the first few months of therapy, and sometimes is irreversible. It may be dose-dependent. Hematuria is an unusual complication of therapy with these drugs and results from systemic vasculitis. Vasculitis is more common with propylthiouracil and rarely reported with methimazole.

If a minor adverse effect (rash or myalgia) occurs, it may be possible to switch to the other antithyroid thionamide agent, but in most cases, a different treatment modality (either radioactive iodine or thyroidectomy) is suggested. These thyroid ablative treatments cause hypothyroidism, and there is an increased risk of recurrence if thyroid tissue is allowed to remain. The complications of radioactive iodine are primarily theoretical. Those of thyroidectomy are real and include the risk of hypoparathyroidism or laryngeal nerve damage. Such risks are lessened in the hands of a very experienced thyroid surgeon.

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Question: 155

One of your 2-year-old patients has prolonged crying and screaming episodes every time her parents deny her access to something she desires. The mother reports that the girl often throws herself on the floor, kicking and thrashing about for long periods of time. She asks you how she should handle her daughter's behavior.

Of the following, your BEST suggestion is that the parents should

A. consider giving in to the girl only when she is outside of the home to avoid a major tantrum

B. give the daughter 10 minutes of time-out for each temper tantrum

C. move the girl to a safe place if needed and ignore her when she has a tantrum

D. offer the child a treat if she calms down

E. physically restrain the child until the tantrum is over

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Critique: 155 Preferred Response: C

Almost every child has temper tantrums at some time. Tantrums occur when a child is frustrated beyond his or her ability to manage feelings. Studies show that up to 80% of toddlers have a weekly tantrum. The parents should be asked about the events that tend to lead to a tantrum, what behaviors occur during the tantrum, how they respond, and what the outcome is. An appropriate behavioral management plan may be developed if the parents understand the developmental issues that lead to a tantrum.

If a tantrum results from a child being frustrated by a task, distracting or redirecting the child to a task in which he or she can succeed may be helpful. If a tantrum occurs at a regular time, such as before meals or prior to going to bed, parents need to be advised that the child's physical state (hunger, fatigue) may be a factor. A routine and structured environment (eg, regular meals and bedtimes appropriate for age) may help decrease the child's tantrums. For tantrums resulting from the child being denied access to a desired object, as described for the child in the vignette, ignoring the behavior is the most appropriate response. Parents should be cautioned that the first response to this action may be escalating tantrum behavior. Such behavior tends to last a few days and eventually decreases and disappears.

Management of tantrums requires setting of limits, which should include positive reinforcement for appropriate behavior (time-in) and negative reinforcement or removal from a situation for undesired behavior. Parents should be consistent in their response to tantrums both inside and outside the home and not give in to the child or offer a treat, which serves as reinforcement of the tantrums. It is important to determine that the child is not in danger of hurting himself or herself during the tantrum. Children should be left alone in a safe place until they can calm down. Such a procedure should not entail a fixed amount of time; the purpose is to allow the child to self-regulate his or her behavior. As soon as the child calms down, the parent may engage the child in social interaction and provide praise for the calmer state.

The use of physical restraint may frustrate or possibly injure the child. Time-out is a management technique that is used to address specific behaviors that are under the child's control (eg, biting). Experts recommend that the time-out last 1 minute per year of the child's age. The child who has notable oppositional behavior that occurs in multiple settings with multiple caregivers and results in impairment of functioning may require referral for behavior management.

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Question: 156

A 3-year-old girl presents to the clinic with the complaint of a recurrent abscess on her buttock. The area has been indurated for 3 to 4 days, but the mother reports that some “nasty” drainage began last night. On physical examination, the girl is afebrile and has an erythematous, indurated area on her right buttock of approximately 2 cm in diameter. With pressure, it drains purulent material.

Of the following, in addition to incision and drainage, the BEST treatment is

A. amoxicillin/clavulanate

B. cephalexin

C. clindamycin

D. observation

E. trimethoprim-sulfamethoxazole

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Critique: 156 Preferred Response: D

Skin and soft-tissue infections due to *Staphylococcus aureus* (SA) and specifically to community-acquired methicillin-resistant *S aureus* (CA-MRSA) are on the rise. Group A *Streptococcus* (GAS) is a common cause of skin and soft-tissue infections, which are characterized by erythematous and painful skin. In contrast, infections due to SA or CA-MRSA usually produce purulent material. When treating carbuncles and furuncles due to CA-MRSA, most experts believe that if the lesion is less than 5 cm in diameter, as reported for the girl in the vignette, only incision and drainage are required. Clindamycin and trimethoprim-sulfamethoxazole are good antimicrobial choices for treatment of CA-MRSA, but these agents are not required for the child in the vignette due to the size of the lesion. In severe cases, a culture should be obtained to identify the organism and antimicrobial susceptibilities should be determined because a number of CA-MRSA isolates are clindamycin-resistant. Isolates of CA-MRSA are resistant to amoxicillin/clavulanate and cephalexin.

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Question: 157

You are talking to a group of medical students about the antibiotic vancomycin and its associated adverse effects. One of the students asks you to explain “red man syndrome.”

Of the following, the MOST appropriate response is that “red man syndrome” is

A. a manifestation of a life-threatening anaphylactic reaction

B. a manifestation of the development of ototoxicity

C. related to cytokine release from cells

D. related to endotoxin release from cells

E. related to histamine release from cells

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Critique: 157 Preferred Response: E

A reaction that is peculiar to vancomycin and occurs in 3.4% to 11.2% of infusions is referred to as “red man” or “red neck” syndrome. It consists of pruritus; an erythematous rash that involves the face, neck, and upper torso; and occasionally hypotension. The manifestations of the reaction are due to the nonimmunologically mediated release of histamine from basophils and mast cells. This complication can be avoided by slowing the infusion rate, decreasing the dose, and administering antihistamines prior to the infusion. Red man syndrome is not a manifestation of a life-threatening anaphylactic reaction or the development of ototoxicity.

Vancomycin is a tricyclic glycopeptide antibiotic that has broad activity against aerobic and anaerobic gram-positive organisms, including staphylococci (both methicillin-sensitive and methicillin-resistant *Staphylococcus aureus* and coagulase-negative staphylococci), streptococci (beta-hemolytic organisms, viridans streptococci), anaerobic cocci, corynebacteria, *Bacillus* sp, most strains of *Enterococcus faecalis,* and clostridia. It is bactericidal against most strains of staphylococci and nonenterococcal streptococci. It exerts its primary bactericidal effect by complexing with the D-alanyl-D-alanine portion of the peptide precursor units, which inhibits the biosynthesis of peptidoglycan in the bacterial cell wall. It is only bacteriostatic against most strains of enterococci, and *Listeria monocytogenes* usually is susceptible to this agent.

Vancomycin is the drug of choice for treatment of serious infections due to methicillin-resistant strains of *S aureus* and coagulase-negative staphylococci. It also is used to treat serious infections caused by methicillin-susceptible staphylococci in patients who cannot tolerate penicillins or cephalosporins, including therapy for endocarditis. Vancomycin is used extensively to treat infective endocarditis caused by streptococci, enterococci, and staphylococci. It also is the drug of choice for infections due to resistant corynebacteria, including *Corynebacterium jeikeium,* and multiresistant strains of *Streptococcus pneumoniae*. Vancomycin is recommended for the prevention of endocarditis in at-risk patients who are allergic to penicillin and are undergoing invasive genitourinary or gastrointestinal procedures associated with transient bacteremia. It also is used as prophylaxis in foreign body implants in orthopedic and cardiovascular surgery. Orally administered vancomycin generally is considered the drug of choice for treating seriously ill patients who have antibiotic-associated *Clostridium difficile* colitis or diarrhea.

Infusion-associated reactions are the most common adverse effects seen with vancomycin. Ototoxicity resulting in damage to the auditory nerve and consequent hearing loss is the most important potential adverse effect associated with the use of vancomycin. This adverse effect has been seen primarily in patients who have extremely high sustained serum concentrations (80 to 100 mcg/mL); it is rare when serum concentrations are maintained at 30 mcg/mL or lower. The risk of ototoxicity seems to be increased when vancomycin is administered in combination with an aminoglycoside. Chemical thrombophlebitis may be seen in up to 14% of patients when the drug is administered via a peripheral intravenous route. Reversible neutropenia occurs in about 2% of vancomycin-treated patients, with an onset around 15 to 40 days after initiation of therapy. Hypersensitivity maculopapular or urticarial drug eruptions and drug-induced fever occur rarely, and nephrotoxicity may occur but is rare.

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Question: 158

You are evaluating a 10-year-old girl for a health supervision visit. Her weight and height are at the 50th percentile for age, her blood pressure is 108/64 mm Hg, and there are no unusual findings on physical examination. A screening urinalysis shows a specific gravity of 1.030, pH of 6.5, 2+ blood, and no protein. Urine microscopy reveals 5 to 10 red blood cells/high-power field.

Of the following, the MOST appropriate next step is

A. abdominal computed tomography scan

B. antinuclear antibody and complement measurement

C. blood urea nitrogen and creatinine measurements

D. referral for cystoscopy

E. repeat urinalysis in 2 weeks

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Critique: 158 Preferred Response: E

The child who has asymptomatic, isolated microscopic hematuria is seen frequently in the ambulatory setting. Results of a urine dipstick test in patients who have hematuria are positive for blood, indicating the presence of hemoglobin or myoglobin. A microscopic evaluation that reveals more than 5 red blood cells/high-power field, as described for the girl in the vignette, confirms the presence of hematuria. Because isolated microscopic hematuria has been found in 4% of children on at least one of four tested samples, evaluation is not recommended unless hematuria is present on at least two of three urine samples. Accordingly, the girl in the vignette should undergo repeat urinalysis in 2 weeks. The prevalence of hematuria in two of three samples is 1% in females and 0.5% in males.

Red blood cells in the urine may arise from the kidney (glomerular or nonglomerular), ureter, bladder, or urethra. Persistent hematuria demands evaluation. It is customary to evaluate renal function by measuring blood urea nitrogen and creatinine and to obtain a complete blood count and platelet measurement (looking for thrombocytopenia as a possible cause) and an erythrocyte sedimentation rate (looking for an underlying inflammatory cause of hematuria). In addition, serologic tests, including complement components 3 and 4, antinuclear antibody, and anti-double-stranded DNA, are recommended to look for markers of immune complex-mediated glomerulonephritis, which can be seen with postinfectious nephritis, membranoproliferative glomerulonephritis, and lupus nephritis. Recommended urine studies consist of urinalysis with microscopy, urine protein and creatinine measurement, and urine culture. Measurement of a urine calcium-to-creatinine ratio is advocated by some but is not universally supported. Finally, abdominal computed tomography scan is not indicated in the evaluation of the patient who has isolated hematuria. This test may be helpful in identifying renal tumors, but these neoplasms usually can be diagnosed by ultrasonography.

The absence of symptoms in children who have isolated microscopic hematuria allows the practitioner to defer urgent evaluation because the likelihood of an underlying systemic disease or a disease limited to the urinary tract that warrants urgent attention is low. Similarly, patients who have microscopic hematuria unaccompanied by proteinuria are unlikely to have a significant disruption of the glomerular capillary barrier. On the other hand, if a child is asymptomatic but has hematuria and proteinuria, evaluation should proceed promptly, looking for an underlying renal parenchymal disorder such as glomerulonephritis. Cystoscopy rarely is indicated in pediatrics, and the decision to pursue this invasive evaluation should be made by a pediatric urologist.

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Question: 159

A 10-year-old boy presents for evaluation of hives that have occurred daily over the past 4 months. His parents are frustrated by the lack of change in their son's symptoms despite changing soap, fabric softener, and detergent. They would like to have their son seen by a specialist for more testing. They describe the hives as raised, erythematous, pruritic 1- to 2-cm lesions that involve the trunk and extremities. The hives resolve spontaneously within a few hours and seem to occur at any time of the day or night. The child is otherwise healthy and is only taking an over-the-counter antihistamine to help with itching.

Of the following, the MOST likely cause for this child's hives is

A. allergy to a food additive or preservative

B. allergy to dust mites

C. autoantibody to the immunoglobulin E receptor

D. autoimmune thyroid disease

E. systemic mastocytosis

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Critique: 159 Preferred Response: C

Chronic urticaria (CU) is defined as recurrent symptoms of pruritic eruptions (urticaria) for more than 6 weeks, as described for the boy in the vignette. Although the first step is to identify potential exacerbating triggers, most patients who have CU describe symptoms that occur regardless of the time of day, foods ingested, or activity level. A specific food or food additive/preservative may cause urticaria, but that should result in symptoms only shortly after food ingestion rather than throughout the day and night. Patients who have CU may have positive skin test results to dust mite and other allergens, but a positive allergy skin test in the context of CU rarely represents the primary reason for a patient's symptoms. Because of the unlikely association of CU with foods or aeroallergens, skin or blood testing for these is not recommended.

In recent years, up to 30% to 50% of both pediatric and adult cases of CU have been identified as autoimmune, specifically due to a circulating autoantibody directed against the high-affinity immunoglobulin (Ig) E receptor (FcεRI) located on mast cells and basophils. Activation of these cells by the autoantibody results in degranulation and histamine release. One diagnostic test that may help identify affected patients is the autologous serum skin test, which involves an intradermal injection of autologous serum with a positive and negative control.

Autoimmune thyroid diseases such as Hashimoto thyroiditis and Graves disease sometimes are associated with pruritus and urticaria. Evidence of thyroid autoantibodies is found less commonly in patients who have CU compared with the autoantibody to the FcεRI. Interestingly, patients who have thyroid autoantibodies usually are euthyroid, but sometimes can experience resolution of the urticaria with thyroid hormone replacement.

Systemic mastocytosis is a rare clonal disorder of mast cells marked by mast cell proliferation in the bone marrow and infiltration in extracutaneous organs. Symptoms and signs can include headache, flushing, dizziness, tachycardia, hypotension, syncope, anorexia, nausea, vomiting, abdominal pain, and diarrhea. Among the skin manifestations are urticaria pigmentosa (Item C159), blisters and bullae, papules, nodules, and diffuse induration.

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Critique: 159



Urticaria pigmentosa, a form of mastocytosis, is characterized by numerous hyperpigmented macules that exhibit an orange-peel texture.

Courtesy of D. Krowchuk

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Question: 160

A 15-year-old basketball player presents to your office with an acute shoulder injury. He reports that an opponent ran into his left, posteriorly outstretched arm as he was guarding him. He developed pain in his left shoulder and has been unable to move his arm since the injury. Physical examination reveals asymmetry of his shoulders, with increased shoulder slope on the left. The left arm is externally rotated and slightly abducted. Fullness is palpable inferior to the left mid-clavicle, and there is no pinprick sensation over the lateral deltoid. You place the patient's left arm in a sling, administer acetaminophen and codeine, and send him for radiographs.

Of the following, the MOST helpful radiographic view for diagnosing his injury is

A. anteroposterior view of the clavicle

B. anteroposterior view of the humerus

C. anteroposterior view of the left scapula

D. axillary view of the shoulder

E. lateral view of the humerus

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Critique: 160 Preferred Response: D

Shoulder dislocations are relatively uncommon injuries in children and adolescents because of the strength of the shoulder capsule and the glenohumeral ligaments. Most often seen in sports-related trauma, these injuries should be reduced urgently in an appropriate setting. Most shoulder dislocations involve anterior displacement of the humeral head, resulting from posteriorly directed force on an abducted, extended arm. The patient complains of significant shoulder pain and holds the arm slightly abducted in external rotation, as described for the boy in the vignette. The humeral head may be palpated inferiorly to the mid-clavicle on the affected side. The axillary nerve is the most commonly injured nerve following anterior shoulder dislocation, and its function should be assessed and documented. Axillary nerve injury results in numbness over the deltoid and the inability to abduct or extend the shoulder.

Initial management of shoulder dislocation involves sling immobilization of the affected arm with a pillow or blanket to fill the space between the arm and body. Analgesics should be provided. Although the diagnosis typically is based on clinical findings, radiographs are indicated for confirmation and to evaluate for any associated bony abnormalities, such as fractures of the humeral head or glenoid fossa (Item C160A). The initial radiologic evaluation should include anteroposterior, lateral, and axillary views of the shoulder. The axillary view is the most sensitive for seeing the relationship between the humeral head and the glenohumeral fossa. Humeral, scapular, or clavicular radiographs alone are not adequate to assess the location of the humeral head.

Reduction should be performed under procedural sedation or using intra-articular lidocaine for analgesia. A variety of reduction methods are likely to be successful, including traction-countertraction (Item C160B), scapular manipulation, or allowing the prone patient to hang the affected arm over the edge of the stretcher with 5- to 15-lb weights attached. Following successful reduction, the neurovascular integrity of the extremity should be redocumented, and the arm should be placed in a sling and swathed to the torso for 2 to 4 weeks before the patient resumes normal activity. Recurrence is common in young children, with an almost 100% likelihood if the proximal humeral growth plate is open. In older adolescents, the redislocation rate ranges from 55% to 95%.

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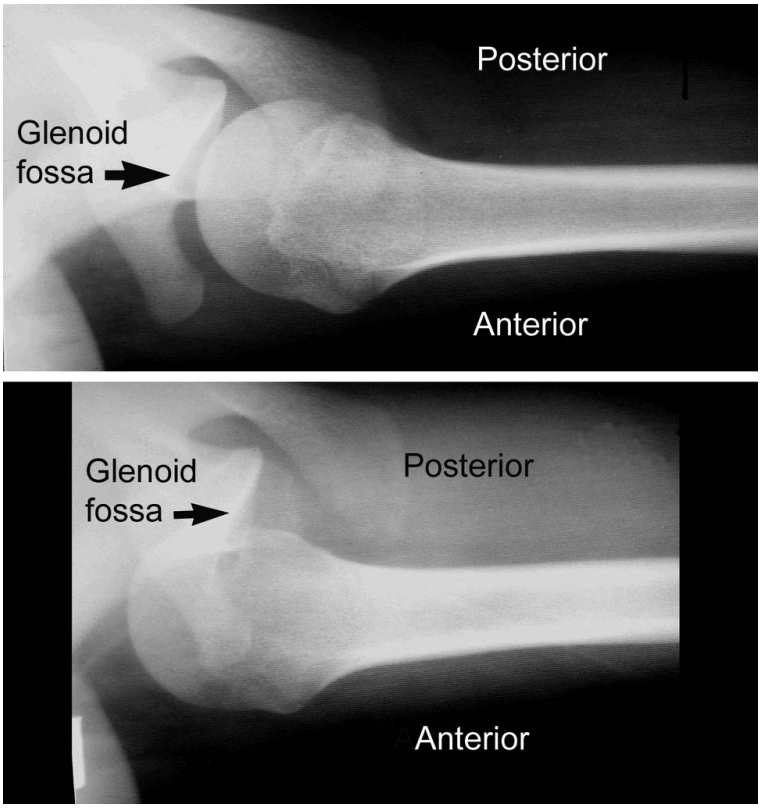
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Critique: 160



In the normal shoulder (top) the humeral head is adjacent to the glenoid fossa. In an anterior shoulder dislocation (bottom), the humeral head is positioned anterior to the glenoid fossa.

Courtesy of D. Mulvihill

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Question: 161

A 14-year-old girl presents for evaluation of areas of skin thickening, tightness, and discoloration that developed 2 months ago. Physical examination reveals shiny, hypopigmented patches with brown borders on the leg and ankle (Item Q161). The affected skin is immobile, firm, and has a "bound-down" feeling.

Of the following, the MOST likely diagnosis is

A. lichen sclerosus et atrophicus

B. linear scleroderma

C. pityriasis alba

D. progressive systemic sclerosis

E. vitiligo

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Question: 161



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Critique: 161 Preferred Response: B

Scleroderma is a rare connective tissue disease that is believed to have an autoimmune cause. It may be categorized as localized and systemic, with the localized form predominating. Localized scleroderma begins as areas of indurated skin that have violaceous borders. Over time, the violaceous color is lost, and the skin takes on a waxy, ivory appearance. As the disease remits, affected areas become atrophic and hypo- or hyperpigmented. Three clinical patterns of localized scleroderma exist: linear scleroderma, morphea, and generalized morphea. In linear scleroderma, as exhibited by the patient described in the vignette, lesions appear in a bandlike distribution, typically are unilateral, and usually involve the extremities (Item C161A). The abnormal tissue may span joints, resulting in diminished range of motion or deformity, and may extend to soft tissue, muscle, or bone. In morphea, one or two discrete areas are affected, often on the trunk. Generalized morphea is characterized by the presence of widespread or coalescent lesions.

The treatment of localized scleroderma is difficult and may include the application of potent topical corticosteroids or topical calcipotriene. For those whose disease is disfiguring or disabling, combination therapy with corticosteroids (orally or intravenously pulsed) and methotrexate (orally or intramuscularly) may be employed. In most instances, localized scleroderma is self-limited, lasting an average of 3 to 5 years. However, there may be considerable morbidity, particularly when the face is involved or joint function compromised. Fortunately, it is very rare for patients who have localized scleroderma to develop systemic involvement.

A number of dermatologic disorders are characterized by hypopigmentation, but do not manifest the thickening or sclerosis seen in scleroderma. Lichen sclerosus et atrophicus produces hypopigmented areas that are atrophic, not sclerotic (Item C161B). In children, the lesions often are located on the genitalia, where they may become eroded, causing pruritus, dysuria, or bleeding. Disorders characterized by inflammation, such as atopic dermatitis, may produce temporary hypopigmentation (often called pityriasis alba). Affected areas are flat, exhibit reduced pigment, and have indistinct borders (Item C161C). In contrast, the lesions of vitiligo lack any pigment and, therefore, are completely white and sharply demarcated (Item C161D).

Progressive systemic sclerosis is very rare in children. Most affected individuals experience tightening of the skin overlying the digits, dilated nail fold capillaries, and Raynaud phenomenon, not the focal cutaneous involvement characteristic of localized scleroderma. In addition, there may be involvement of multiple organ systems (eg, gastrointestinal, musculoskeletal, cardiac, pulmonary, or renal).

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Critique: 161



Linear scleroderma affecting the ankle: The skin has a waxy appearance, and there is central hypopigmentation and peripheral hyperpigmentation.

Courtesy of D. Krowchuk

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Critique: 161



Lichen sclerosus et atrophicus produces hypopigmentation and atrophy that usually affect the genitalia. (There is a suture at the site of a biopsy.)

Courtesy of D. Krowchuk

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Critique: 161



Postinflammatory hypopigmentation is characterized by indistinctly marginated hypopigmented macules or patches.

Courtesy of D. Krowchuk

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Critique: 161



Vitiligo causes depigmented macules or patches that have well-defined borders. In this patient, some areas have begun to repigment (arrow).

Courtesy of D. Krowchuk

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Question: 162

A 16-year-old girl presents with a 4-month history of right upper quadrant abdominal pain. The pain occurs at different times, but seems to strike primarily after meals, more frequently after she eats fatty foods. In your office, she complains of intermittent pain to deep palpation of the right upper quadrant. Complete blood count, alanine aminotransferase, alkaline phosphatase, serum bilirubin, amylase, and lipase findings are normal. Abdominal ultrasonography shows no evidence of stones or gallbladder thickening. Upper endoscopy and biopsy results are normal, with no evidence of ulcers or gastritis.

Of the following, the MOST appropriate next step is

A. abdominal computed tomography scan

B. endoscopic retrograde cholangiopancreatography (ERCP)

C. nuclear medicine gallbladder emptying scan with fatty meal

D. psychiatric consultation to rule out depression or anxiety

E. referral to an acupuncturist for chronic pain management

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Critique: 162 Preferred Response: C

The colicky abdominal pain in the right upper quadrant after the ingestion of fatty foods described for the patient in the vignette is strongly suggestive of gallbladder disease. However, results of laboratory studies and abdominal ultrasonography are within normal limits. Because of the symptom profile, the possibility of chronic acalculous cholecystitis with gallbladder dysmotility should be considered, and the test of choice to evaluate for this condition is a radionuclide gallbladder emptying scan. If the patient has markedly delayed gallbladder motility, consideration should be given to performing a cholecystectomy.

Classically, cholecystitis occurs when the gallbladder is inflamed and irritated by gallstones. Such gallstones typically are classified as cholesterol stones and pigment (bilirubin) stones. Risk factors for cholesterol stones include older age, female sex, pregnancy, and overweight. Risk factors for pigment stones include parenteral nutrition and hemolysis (as seen in children who have sickle cell disease). For patients who have stones, cholecystitis commonly presents with pain in the right upper quadrant, epigastrium, and back. Fever, jaundice, and abnormal liver enzyme values also may be present, especially if a gallstone is in the biliary tree (choledocholithiasis).

Children may have biliary symptoms without gallstones, a condition termed chronic acalculous cholecystitis or gallbladder dysmotility. Such patients have right upper quadrant pain with meals, but laboratory and ultrasonography results are normal. Hepatobiliary scintigraphy demonstrates delayed emptying of the gallbladder in response to a fatty meal. These children may experience significant symptomatic relief after cholecystectomy; some case series have reported improvement in up to 90% of patients after surgery. Abdominal computed tomography and endoscopic retrograde cholangiography should be considered if pancreatic disease is suspected, but these are not routine tests used to evaluate gallbladder dysmotility. Some patients who have chronic abdominal pain may benefit from psychiatric evaluation or acupuncture, but these measures are more supportive than diagnostic.

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Question: 163

An infant who was born at 26 weeks' gestation, weighing 700 g, is nearing 37 weeks corrected age. He is receiving nasal cannula oxygen (0.2 L/min) and being treated with diuretics for chronic lung disease. He has no intracranial hemorrhage and is growing well on enteral feedings via a nasogastric tube. Attempts at oral feeding have been unsuccessful because of frequent oxygen desaturation, bradycardia, and concerns over the inability to coordinate sucking-swallowing. The mother asks why you are continuing to feed her son via a feeding tube.

Of the following, the condition that BEST explains why her son has feeding problems is

A. apnea of prematurity

B. chronic lung disease

C. gastroesophageal reflux

D. necrotizing enterocolitis

E. tracheoesophageal fistula

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Critique: 163 Preferred Response: B

Long-term neuromotor problems complicate the course of many extremely low-birthweight (ELBW) (<1,000 g) infants. These may be related to intracranial hemorrhage, periventricular leukomalacia, or poor brain growth and function. The long-term outcome for ELBW infants also is affected by their nutrition and any chronic hypoxia. For preterm infants who have chronic lung disease (CLD), potential intermittent or chronic hypoxia and impaired feeding and caloric intake may have negative effects on their growth.

Many ELBW infants have an oral aversion following prolonged oral endotracheal intubation and delayed oromotor development and function. Swallowing dysfunction (dysphagia) may be ascertained clinically as feeding skills are assessed by experienced nurses, physicians, and speech pathologists, with monitoring of heart rate, breathing, and arterial oxygen saturation during feedings. Radiographic imaging using a fluoroscopic modified barium swallow study can reveal problems with hypopharyngeal pooling, laryngeal penetration, or frank aspiration of milk.

The infant described in the vignette has CLD and demonstrates problems in coordinating breathing with suckling and swallowing. Accordingly, he continues to be assisted with nasogastric tube feedings despite reaching a postconceptive age of 37 weeks. Apnea of prematurity is not present; the infant has passed the gestational age equivalent of prematurity. Gastroesophageal reflux does not occur during a feeding but typically presents with postprandial emesis. Necrotizing enterocolitis is a gastrointestinal disease associated with systemic decompensation and an acute abdomen. Tracheoesophageal fistula (TEF) is associated with respiratory distress and secretion control problems during intervals between feeding; the rare H-type fistula with an intact esophagus (accounting for <5% of all TEFs) typically does not present in the newborn period.

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Question: 164

You are asked to consult on a 9-month-old boy who has been hospitalized five times for wheezing. His history reveals occasional coughing with feedings, but results of a pH probe performed during his last admission were normal. His weight and height are at the 50th percentile. Except for scattered wheezes with good aeration bilaterally, results of his physical examination are normal.

Of the following, the test that is MOST likely to reveal the cause of his recurrent wheezing is

A. chest computed tomography scan

B. immunoglobulin panel

C. inspiratory and expiratory chest radiographs

D. pulmonary function testing

E. videofluoroscopic swallow study

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Critique: 164 Preferred Response: E

Recurrent wheezing can be caused by many diseases, including reactive airway disease, cystic fibrosis, extrinsic airway compression, and aspiration with and without gastroesophageal reflux. The history of coughing with feedings described for the boy in the vignette should alert the clinician to the possibility of swallowing dysfunction, with aspiration as the cause of his recurrent symptoms. Accordingly, a videofluoroscopic swallow study is the best diagnostic procedure to reveal the cause of his wheezing.

Gastroesophageal reflux is a common cause of recurrent aspiration, but swallowing dysfunction (Item C164A) without gastroesophageal reflux also can occur and cause significant recurrent respiratory symptoms. Several types of swallowing dysfunction are seen in infants. Laryngeal penetration (Item C164B) without aspiration describes the entry of food particles into the airway down to the level of the vocal cords. Aspiration is defined as the entry of food below the level of the vocal cords (Item C164C), and nasopharyngeal backflow or reflux is the entry of food posterior or superior to the soft palate (Item C164D). One study of infants referred for swallowing study due to recurrent respiratory difficulty showed that all had some degree of swallowing dysfunction and silent aspiration. The dysfunction resolved in all of the infants by age 9 months. Another study showed that of infants who had swallow studies, 50% showed laryngeal penetration, aspiration, or nasopharyngeal regurgitation. Most of these infants did not cough to clear their airway, which should remind the clinician that absence of cough with feedings does not eliminate the possibility of silent aspiration. If a fluoroscopic swallow study reveals swallowing dysfunction, thickening formula or human milk and feeding in the upright position may improve symptoms. In some cases, cessation of oral feedings and placement of a nasojejunal or gastrostomy tube may be indicated for a period of time.

Chest computed tomography scan may be indicated to rule out a structural anomaly if an infant has recurrent localized wheezing, but it probably would not be helpful for assessing recurrent diffuse wheezing. An immunoglobulin panel can aid in ruling out immunodeficiency, but in an infant who has no recurrent infections and is growing well, immunodeficiency is not likely. Inspiratory and expiratory chest radiographs and pulmonary function testing are technically difficult in infants and would not be of benefit in the evaluation of this child.

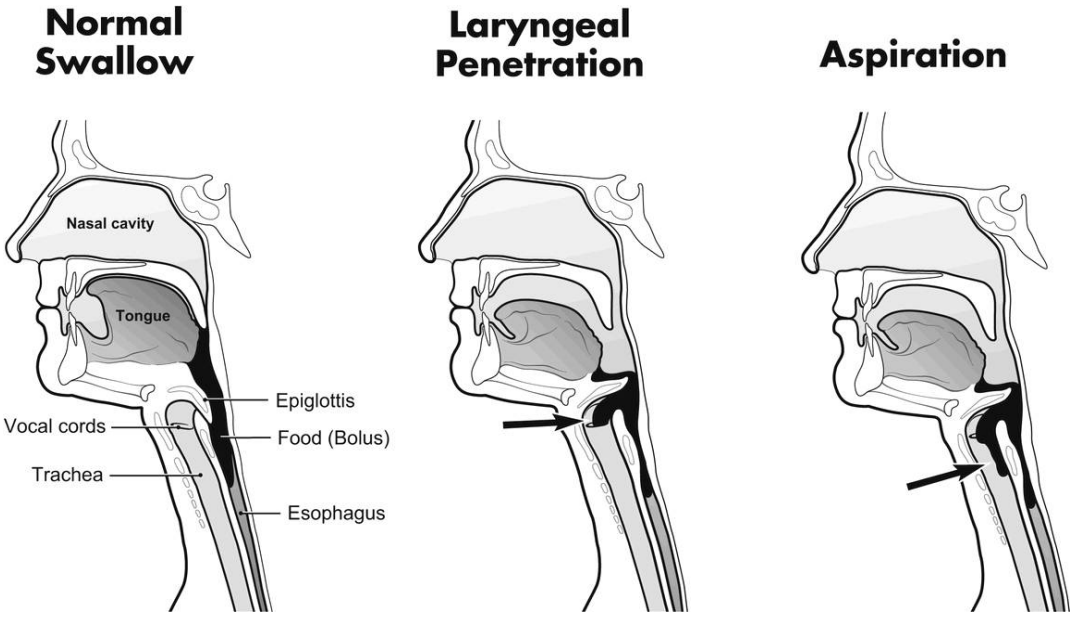
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Critique: 164



Normal and abnormal swallowing: In normal swallowing (left), the food bolus enters the esophagus. Laryngeal penetration occurs when food enters the airway above the vocal cords (center). In aspiration (right), food travels below the vocal cords into the trachea.

Courtesy of A. Johnson

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Question: 165

A 6-year-old boy has complained of periumbilical abdominal pain for the past 2 months. He has a history of mild constipation that his mother has managed successfully by increasing juice and fruits in his diet. The mother is frustrated because the boy has been sent home from school frequently due to complaints of pain. She reports that he is being evaluated for dyslexia. The pain occasionally occurs on weekends and school vacation breaks.

Of the following, the MOST likely diagnosis is

A. chronic constipation with fecal impaction

B. factitious abdominal pain

C. *Helicobacter pylori* gastritis

D. recurrent (functional) abdominal pain syndrome

E. school phobia

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Critique: 165 Preferred Response: D

Children often present to the pediatrician's office with recurrent pain syndromes, most notably abdominal pain and headache. Distinguishing significant pathology from minor functional or intercurrent illness is essential.

The child who presents with recurrent pain syndrome must have a psychosocial history taken because school is a significant stressor for many children and may be a cause of the symptoms. Additionally, school attendance and performance are good measures of the child's ability to perform in the face of recurring complaints. In addition to the pattern of pain and intestinal symptoms such as diarrhea, constipation, and dyspepsia, one of the diagnostic criteria for recurrent abdominal pain syndromes of childhood is the degree to which the symptoms interfere with school. Symptom exacerbation may be related to anxiety associated with academic or social school functioning.

In migraine and other recurrent headache syndromes, the photophobia, nausea, and other concomitant symptoms may preclude school attendance or impair performance, and therapy may be targeted at reducing the interference with activity. Conversely, children who have stress or tension headache may need to have their school performance assessed as a contributing factor to the illness. Finally, somatization disorders and school phobia often are accompanied by complaints similar to recurrent pain syndromes due to the anxiety that is associated with these disorders.

The symptoms exhibited by the child described in the vignette are most consistent with recurrent (functional) abdominal pain. The mother states that she has managed the child's mild constipation successfully, and there is no history of encopresis to suggest fecal impaction. School phobia is less likely because the pain also occurs on weekends and school holidays. Although *Helicobacter pylori* gastritis is a possibility in children who have epigastric abdominal pain, those affected often have associated nausea and vomiting.

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Question: 166

Included in your rounds today is a 36-hour-old boy who was born at term by normal, spontaneous vaginal delivery. His respiratory rate is 80 breaths/min and heart rate is 168 beats/min. He has easily palpable, bounding pulses in all four extremities, and his blood pressure is 72/30 mm Hg. Precordial examination reveals a lift and a 3/6 systolic ejection murmur at the upper left sternal border (Item Q166). You also note a murmur over the anterior fontanelle.

Of the following, the MOST likely diagnosis is

A. aortic coarctation with congestive heart failure

B. aortic insufficiency

C. large ventricular septal defect with congestive heart failure

D. left-to-right extracardiac shunting with congestive heart failure

E. right-to-left extracardiac shunting with right heart failure

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Critique: 166 Preferred Response: D

Systemic arteriovenous malformations constitute an extracardiac left-to-right shunt (system to venous). Blood flow always moves from high to low resistance when given the opportunity. When an arteriovenous communication occurs, as might be seen in the brain or in the liver, blood from the high-pressure, high-resistance systemic circulation can move directly into the low-pressure, low-resistance venous circulation, bypassing the capillary bed of the affected organ. In so doing, the volume of blood entering the venous system is increased, as is the oxygen content of the blood because the tissues have not been exposed to the oxygenated blood. With the passage of time and decreasing blood viscosity as physiologic anemia of the newborn ensues, the volume of blood "shunted" through the arteriovenous malformation becomes greater. Such excess blood flow "loads" the venous pool, which is delivered to the right atrium. As a result, the right atrium and right ventricle become dilated, and congestion may occur. If such congestion is significant, jugular venous distension and hepatomegaly may become evident on physical examination.

Other signs of the right heart volume overload include a prominent precordial lift on palpation and a systolic ejection murmur (Item C166) or relative pulmonary stenosis as the excess blood from the right heart makes its way across the pulmonary valve. The murmur is termed "relative" because the pulmonary valve annulus does not dilate despite dilation of the right ventricle. However, because more blood is ejected from the right ventricle, it must travel with greater velocity as it crosses the pulmonary valve, leading to turbulence, which produces the audible murmur on auscultation. The excess right heart output enters the pulmonary vascular bed, often leading to some level of congestion, with the resulting sign of tachypnea on examination. Similarly, because arterial blood has a "run-off" into the low-pressure veins, there is a pronounced pulse pressure with a typically low diastolic pressure that produces bounding pulses on examination. In some patients, a continuous murmur can be heard over the area of the arteriovenous malformation, such as the fontanelle, as in the patient described in the vignette, or the liver.

The newborn described in the vignette has physical findings and blood pressure that suggest a run-off lesion from the aorta, which could be significant aortic insufficiency, a large-volume ductus arteriosus, or an arteriovenous malformation. There is no diastolic murmur to suggest aortic insufficiency, and at 36 hours of age, a ductus arteriosus would not be expected to lead to symptoms. Similarly, a large ventricular septal defect might present with a holosystolic murmur and rarely leads to symptoms in the first few days after birth. Coarctation often leads to narrowed blood pressure and is associated with a pressure load on the left ventricle rather than a volume load, as in this patient. Right-to-left extracardiac shunting can occur only when pressure in the venous (right) vessel exceeds that in the arterial (left) vessel. This is a situation that does not exist.

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Question: 167

An 8-year-old girl is brought to the emergency department via ambulance. On the playground, she suddenly stopped playing, bent forward and fell to the ground, and had jerking of her arms and legs. She drooled excessively and was unresponsive. Afterwards, she was confused, her speech was slurred, and she was somewhat combative for about 30 minutes. In the emergency department, she is responding appropriately, is afebrile, and has normal findings on general and neurologic examinations. Her mother states that she has always been healthy and is an average student. Review of systems reveals no headaches or recent illness.

Of the following, the MOST appropriate next step prior to discharge from the emergency department is to

A. educate the family about prognosis and safety

B. obtain a stat electroencephalogram

C. obtain magnetic resonance imaging of the brain

D. order measurement of serum electrolytes

E. perform a lumbar puncture

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Critique: 167 Preferred Response: A

The child described in the vignette had a first unprovoked seizure. She was previously healthy and had no neurodevelopmental problems. There was no history of a focal neurologic lesion, the seizure did not begin focally or have focal paralysis afterward, and the return to normal was reasonably fast. In this setting, the risk of recurrence is about 33%. The standard of care after a first unprovoked seizure is not to treat with daily antiseizure medication. Additionally, no further tests are required in the emergency department.

The family should be educated about prognosis and safety after a first seizure. The key components of this education are: 1) seizure first aid, 2) seizure precautions, 3) risk of recurrence of seizures, and 4) prognosis.

First aid: If the child has another seizure, she should be placed on the floor on her side, away from furniture. Some families worry that the child may swallow her tongue and may want to place a spoon or other object in the mouth to prevent this. Parents should be told that tongue swallowing cannot and does not occur, and no object should be placed in the child's mouth. The parent should time the seizure to be able to report to the doctor its duration. If the seizure lasts 5 minutes, the family should call 911 for emergency assistance.

Precautions: Seizure precautions in children prior to driving age involve "wheels and water." This common sense advice includes the wearing of helmets when the child is "on wheels" and adult supervision whenever the child is in water, including bath water. The child can sleep in his or her own bed and does not need to sleep with a parent.

Recurrence risk: Most children who are developmentally normal and have a single seizure will not have a recurrence. In an otherwise healthy child who experiences a single nonfebrile seizure, the recurrence risk is about 33%.

Prognosis: There is no evidence that single seizures, or several seizures, can cause brain damage.

Blood testing for electrolyte disturbances is useful in selected clinical settings, such as after prolonged diarrhea, but generally is not helpful in well-nourished, otherwise healthy school-age children who experience a first seizure. A complete toxin screen also may be prudent in some clinical settings. The common practice in many emergency departments of sending blood tests for renal function, calcium, magnesium, phosphorous, and liver function in all children, irrespective of the history or examination, is not supported by evidence. A dextrose stick is recommended. In the absence of fever and encephalopathy, lumbar puncture is not necessary in children older than age 18 to 24 months of age.

Neuroimaging, usually noncontrast head computed tomography (CT) scan, commonly is obtained in emergency departments after a first unprovoked seizure. However, the probability of finding a focal treatable lesion at the time of first seizure in an otherwise healthy child is approximately 2%, and emergent treatment almost never is required in this setting. The risk of focal lesions is higher in teenagers than in children and is much higher in adults, in whom neuroimaging after a first unprovoked seizure is recommended. In the absence of encephalopathy and any persistent, new focal findings on history or physical examination in an otherwise healthy child, neuroimaging generally can be deferred and obtained as an outpatient. If the clinician judges that neuroimaging is indicated, outpatient magnetic resonance imaging has a higher diagnostic yield than a head CT scan. Experts generally recommend neuroimaging for first focal onset seizures.

Obtaining electroencephalography (EEG) in the emergency department after a first unprovoked seizure is not routine. Outpatient EEG has been recommended as a standard of care after a first afebrile seizure in childhood in the United States. However, the cost-effectiveness of EEG after a first unprovoked seizure versus after recurrent seizures (ie, epilepsy) has not been assessed. Some experts point out that EEG results after a first seizure would not routinely affect education or management, making selective, rather than routine, outpatient EEGs more appropriate. In Great Britain, the Pediatric First Seizure Guideline does not recommend that EEGs be obtained as a standard of care.

Some physicians prescribe rectal diazepam or intranasal midazolam to all children after a single seizure to be used in the future should a prolonged seizure occur. This is especially important for a child who had a seizure lasting more than 5 minutes, is medically fragile, or has

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limited access to medical facilities.

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Question: 168

A 12-year-old girl comes with her mother to your office for a health supervision visit. When you meet with the mother alone, she asks you about sexuality education for early adolescents.

Of the following, you are MOST likely to counsel the mother that

A. office-based intervention alone is the best approach to promoting sexual health among young people

B. parents have little influence on their children's sexual behaviors

C. schools have standardized, uniform curricula to teach about sexuality

D. talking about sexuality to children increases their risk of becoming sexually active

E. talking early and often with children about sexuality is an effective parental counseling strategy

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Critique: 168 Preferred Response: E

Although the amount of reported sexual activity among adolescents in the United States has decreased, initiation of sexual activity during adolescence remains the norm. Children and adolescents need accurate, timely, and comprehensive information about sexuality to practice safer and healthy sexual behavior. Sexual health includes both sexual development and reproductive health. Included in the development of healthy sexuality are the ability to appreciate one's own body; develop and maintain interpersonal relationships; avoid exploitative relationships; affirm one's own sexual orientation and respect the sexual orientation of others; and express affection, love, and intimacy in accord with personal values.

Parents have influence on their adolescents' sexual behaviors. Research clearly demonstrates that talking early and often with children about sexuality does not encourage them to become sexually active. Office-based education and intervention by pediatricians can provide young people with personalized information, confidential screening of risk status, and health promotion and counseling. School-based sexuality education programs vary tremendously. Abstinence-only programs have not shown successful outcomes with regard to delaying the onset of sexual activity or the use of safer sexual practices. Effective programs tend to provide practical skills, such as increasing communication and negotiating skills. Programs that discuss human immunodeficiency virus transmission prevention and contraception in addition to encouraging abstinence have been associated with delayed initiation of sexual activity and increased use of contraception among sexually active young people. Programs linking educational curricula with access to reproductive health services and comprehensive community-based interventions also have demonstrated reductions in pregnancy rates. Office-based interventions are most likely to contribute to reduced risks and promotion of sexual health if they occur in conjunction with other educational and support methods in schools, communities, and families.

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Question: 169

You are evaluating a 15-year-old girl who complains of malaise, fatigue, and occasional abdominal discomfort. You diagnosed hypothyroidism due to chronic lymphocytic thyroiditis (Hashimoto thyroiditis) 6 years ago. She has normal serum immunoglobulin A concentrations. A tissue transglutaminase antibody study was negative 1 month before this visit, and free thyroxine and thyroid-stimulating hormone (TSH) values were normal at that time. She has normal menses. She reports that she has been eating poorly and has lost 5 lb since you saw her at the beginning of the summer, but she obviously has had a good summer and has a tan.

Of the following, the MOST important laboratory studies to obtain at this time are

A. complete blood count and erythrocyte sedimentation rate

B. duodenal biopsy for cryptic celiac disease

C. measurement of cortisol and adrenocorticotropic hormone

D. measurement of free thyroxine and TSH

E. mononucleosis spot test and liver function study

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Critique: 169 Preferred Response: C

The malaise, fatigue, weight loss, abdominal discomfort, and tanned skin (Item C169) reported for the girl in the vignette are signs of Addison disease. Adrenal insufficiency causes an elevation in serum potassium concentrations, decrease in serum sodium concentrations, and shifts in muscle electrolyte concentrations that result in weakness, myalgias, and gastrointestinal symptoms. Skin pigmentation is increased by high concentrations of adrenocorticotropic hormone. The presence of both autoimmune hypothyroidism and suspected adrenal insufficiency in this girl suggests the diagnosis of autoimmune polyglandular syndrome type 2. The genetic defect in this disorder is not yet known, but the girl is at risk for other endocrine autoimmunities, including ovarian failure and diabetes.

A complete blood count and erythrocyte sedimentation rate probably will be normal, although adrenal insufficiency may increase the peripheral eosinophil count. Because the girl may be at risk for celiac disease (2% to 4% of those who have chronic lymphocytic thyroiditis may have celiac disease), routine screening for tissue transglutaminase (TTG) antibody was performed. The presence of a negative TTG study makes celiac disease unlikely. Many endocrinologists recommend screening for celiac disease (eg, TTG antibody test) at 2- to 3-year intervals in patients who have chronic lymphocytic thyroiditis. Because there is a greater risk of immunoglobulin A deficiency in individuals who have celiac disease and the TTG antibody is in the immunoglobulin A class, it was important to document normal immunoglobulin A values. Autoimmune hepatitis is rare in individuals who have autoimmune endocrine disorders; it is more common in association with diabetes. A test for mononucleosis is not an appropriate initial evaluation in the face of hyperpigmentation and the other complaints. Thyroid function studies were performed 1 month before, and if the patient has complied with therapy, such symptoms, including weight loss, which is unusual in hypothyroidism, would not be expected to result from thyroid hormone deficiency.

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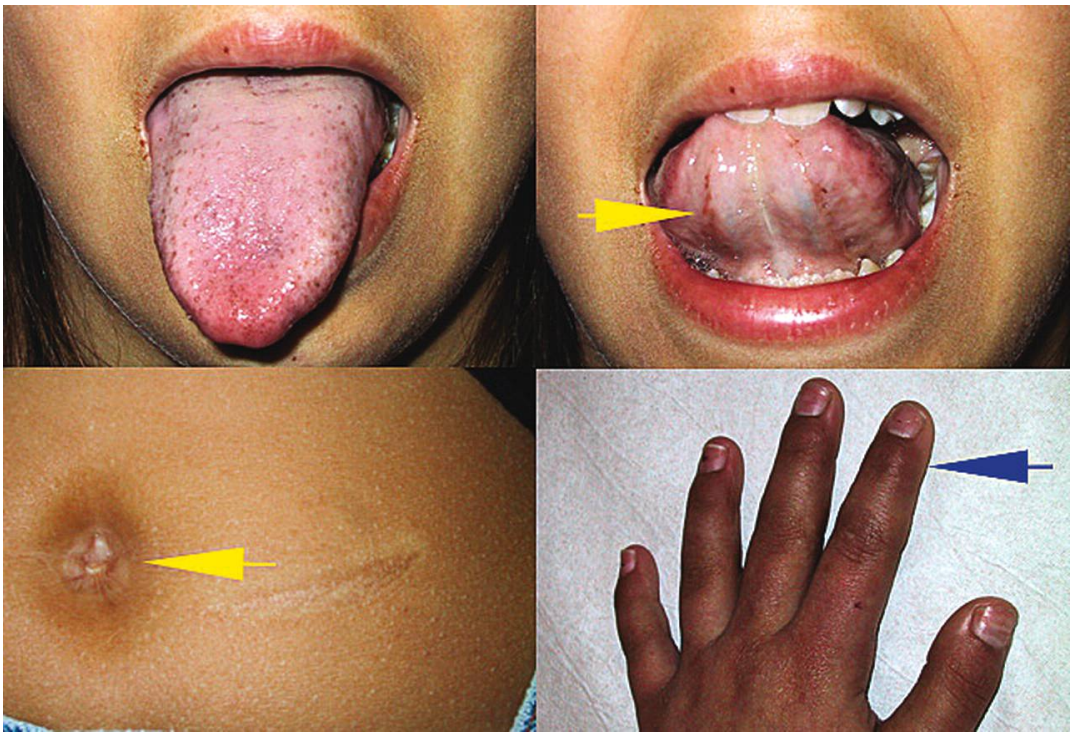
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Critique: 169



Patients who have Addison disease may have increased pigmentation that involves the skin and mucosa (arrow).

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Question: 170

A 3-year-old boy has a history of biting his parents' cheeks when he does not get his way, which they have always considered cute, calling it "love bites." His child care teacher has informed the parents that he is frequently biting other children. The boy's parents are concerned that he may be removed from his child care program and ask for your advice about how to stop this behavior.

Of the following, the BEST response is to

A. advise the parent to change child care centers

B. instruct the parents to set up a reward system for not biting

C. recommend treatment with a stimulant to help decrease the behavior quickly

D. take the child aside and explain that his behavior is not acceptable and may cause him to be removed from child care

E. tell the parents it is acceptable to gently bite the child or tap his backside to stop this behavior quickly

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Critique: 170 Preferred Response: B

Biting is a normal and often transient behavior that almost all children display during the first 3 years after birth. Biting occurs at certain developmental times. The initial stage is when teeth begin to erupt (4 to 6 months of age). The next phase is at 8 to 12 months of age, when the infant uses biting to express excitement. The natural reaction of jumping, being startled, or crying out accompanied by putting the infant down is effective in diminishing this behavior. Two-year-old children who are frustrated, especially when they have limited expressive language, may respond by biting. Toddlers or preschoolers may lack the self-control to express their anger peacefully. Instead, they naturally lash out, perhaps hitting or biting in frustration. In these cases, the parent needs to take control for the child and to help him or her develop judgment and self-discipline to express feelings more acceptably through words.

The child described in the vignette requires behavioral strategies to stop his biting. The initial intervention should involve watching the child and providing positive attention for appropriate behavior. He should be redirected when he gets frustrated or angry. When he bites, he should be told firmly “no” and removed from positive attention, such as placing him in a time-out situation. Prior to considering removal from the child care program, the parents should meet with the teacher to develop a consistent behavioral approach when the child bites. The parents should not bite or hit the child in response; this causes fear and anger in the child and leads to feelings of guilt in the parent. Stimulants are not indicated in the treatment of biting. The child likely will be unable to understand lengthy explanations about why he should not bite.

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Question: 171

A mother brings in her child because she found a tick on the girl's shoulder yesterday and is worried about Lyme disease. The mother found a site on the Internet that suggests her daughter needs an antibiotic called ceftriaxone. You assure the mother that only very few children who suffer tick bites actually develop a tick-associated disease.

Of the following, the manifestation of Lyme disease for which ceftriaxone administration is MOST appropriate is

A. acute arthritis

B. carditis

C. disseminated erythema migrans

D. isolated facial palsy

E. peripheral neuropathy

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Critique: 171 Preferred Response: B

Most patients who have Lyme disease can be treated with oral antimicrobial agents (Item C171). Doxycycline is the drug of choice for patients 8 years of age and older; amoxicillin should be used for those younger than 8 years of age. For patients allergic to penicillin, cefuroxime axetil is the recommended alternative agent. Erythromycin and azithromycin are less effective than doxycycline or amoxicillin.

Parenteral regimens rarely are indicated. Recurrent or persistent arthritis after treatment with an oral regimen, meningitis, and carditis should be treated with parenteral therapy (ceftriaxone or penicillin).

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Critique: 171

Item 171: Treatment Recommendations for Lyme Disease

Disease Category	Drug
Early localized disease	Oral regimen: Doxycycline or amoxicillin for 14 to 21 days Parenteral regimen: Cefuroxime for 14 to 21 days
Early disseminated and late disease	
Disseminated erythema migrans	Same oral regimen for 21 days
Isolated facial palsy	Same oral regimen for 21 to 28 days
Acute arthritis	Same oral regimen for 28 days
Peripheral neuropathy	Same oral regimen for 21 to 28 days
Persistent or recurrent arthritis	Same oral regimen for 14 to 21 days Parenteral regimen: Ceftriaxone or penicillin for 14 to 28 days
Carditis	Parenteral regimen: Ceftriaxone or penicillin for 14 to 28 days
Meningitis or encephalitis	Parenteral regimen: Ceftriaxone or penicillin for 14 to 28 days

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Question: 172

A 5-year-old boy who has neuroblastoma is admitted to the pediatric intensive care unit for treatment of fever, neutropenia, and severe hypotension due to *Klebsiella pneumoniae* sepsis. Over the last several months, he has received multiple courses of vancomycin and ceftazidime to treat fever and neutropenia. Antibiotic susceptibility testing of the pathogen shows it to be susceptible only to the carbapenem and aminoglycoside classes of antibiotics.

Of the following, the MOST likely mechanism of resistance in this organism is

A. alterations in penicillin-binding proteins

B. decreased affinity for ribosomal target binding sites

C. increased thickness of organism cell wall

D. production of an efflux pump

E. production of extended-spectrum beta-lactamases

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Critique: 172 Preferred Response: E

The incidence of broad-spectrum antibiotic resistance among nosocomial gram-negative organisms is a major problem that has made the treatment of many infections much more difficult. The rate at which this resistance has developed is due, in large part, to the extensive, indiscriminate, and inappropriate use of the extended-spectrum third-generation cephalosporin class of antibiotics. Resistance of gram-negative organisms to the third-generation cephalosporin antibiotics has been accelerated rapidly by the emergence of plasmid-mediated extended spectrum beta-lactamases (ESBLs) in these organisms, especially among *Klebsiella* sp, *Escherichia coli*, *Proteus mirabilis,* and *Pseudomonas aeruginosa*. Beta-lactamases are enzymes produced by gram-negative bacilli that hydrolyze the beta-lactam ring of penicillins and cephalosporins, thereby inactivating the antibiotic. ESBLs are produced by certain gram-negative bacilli and confer resistance not only to the extended-spectrum third-generation cephalosporins, but also potentially to the monobactam (aztreonam), aminoglycoside, and fluoroquinolone classes of antibiotics, all of which could be used to treat nosocomial gram-negative infections.

The mechanism of resistance for the *Klebsiella pneumoniae* strain described in the vignette most likely is the production of ESBLs. Although gram-negative organisms may possess the resistance mechanisms of altered penicillin-binding proteins, decreased affinity for ribosomal target binding sites, increased thickness of the organism cell wall, and production of an efflux pump, the production of ESBLs accounts for their resistance to a broad range of antibiotic agents.

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Question: 173

One of your patients is a 6-month-old boy who had unilateral hydronephrosis detected prenatally. Ultrasonography shows moderate hydronephrosis on the right and a normal left kidney, and voiding cystourethrography shows no reflux, with a normal bladder and urethra. During a MAG 3/furosemide renal scan, shortly after the furosemide is administered, the infant becomes extremely fussy and difficult to console.

Of the following, the MOST likely diagnosis is

A. duplex collecting system with a ureterocele

B. multicystic dysplastic kidney

C. nephrolithiasis

D. obstructive uropathy from posterior urethral valves

E. ureteropelvic junction obstruction

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Critique: 173 Preferred Response: E

The differential diagnosis for an infant who has hydronephrosis in the first postnatal year includes ureteropelvic junction (UPJ) obstruction, ureterovesical junction (UVJ) obstruction, single-system ureterocele, vesicoureteral reflux (VUR), and posterior urethral valves (usually causes bilateral hydronephrosis).

If hydronephrosis is found on renal/bladder ultrasonography, voiding cystourethrography (VCUG) is undertaken to rule out VUR and assess for a filling defect in the bladder caused by a ureterocele (Item C173A). Following VCUG, a technetium-99m mercaptoacetyltriglycerine (MAG-3) radioisotope scan with furosemide is performed to assess for UPJ obstruction. If UPJ obstruction is present, the radioisotope exhibits delayed washout. During the test, furosemide is infused and may precipitate an episode of renal colic, as reported for the infant in the vignette. If a UPJ obstruction is diagnosed, the practitioner should contact a pediatric nephrologist or urologist to help with further management.

A duplex collecting system with a ureterocele is detected by VCUG that reveals a filling defect in the bladder and VUR (in approximately 50% of cases). On ultrasonography, a multicystic dysplastic kidney (MCDK) (Item C173B) exhibits multiple large cysts that do not communicate and a rim of dysplastic renal parenchyma. An MCDK occurs unilaterally and has no inherent function. Obstructive uropathy from posterior urethral valves is characterized by narrowing of the posterior urethra (Item C173C) and is associated with bilateral hydronephrosis in nearly all cases and VUR in approximately 50% of cases. Nephrolithiasis is defined as a calculus within the renal parenchyma. Ultrasonography may reveal a calculus. Only if the calculus moves into the collecting system would hydronephrosis or renal colic following a diuretic renal scan occur. A calculus within the collecting system is known as urolithiasis.

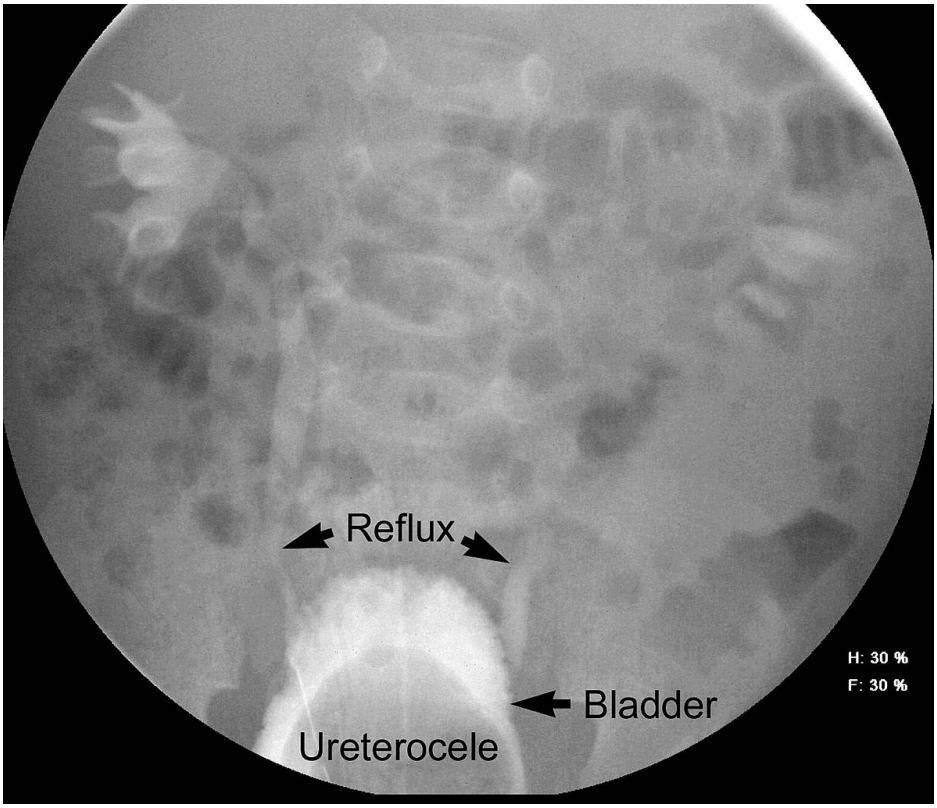
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Critique: 173

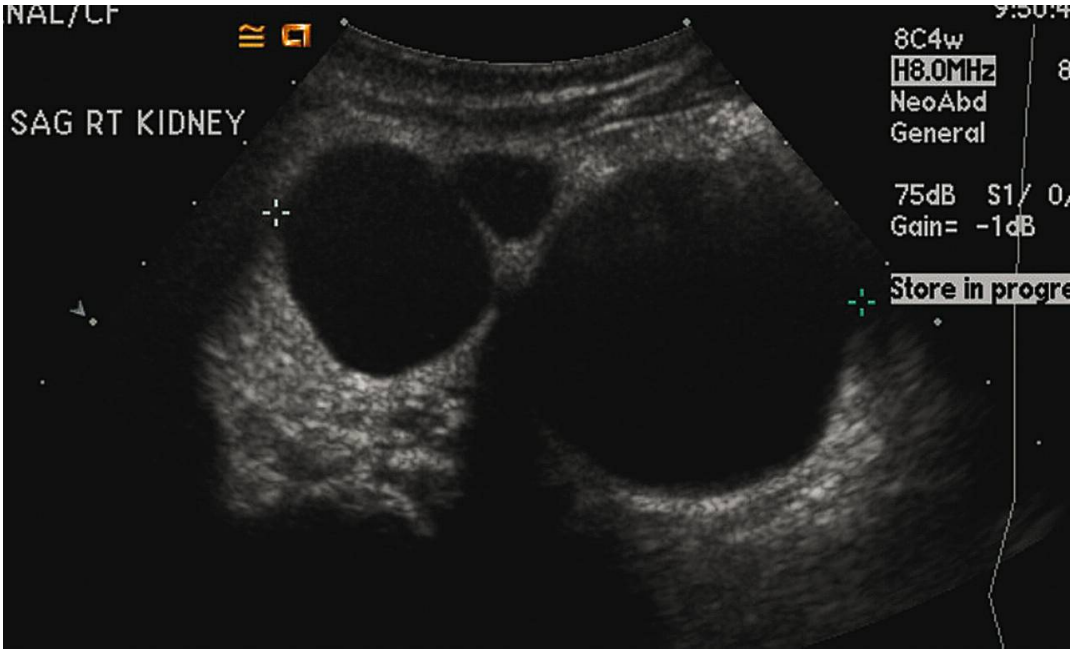


Fluoroscopic image from voiding cystourethrography showing a large mass within the contrast-filled bladder consistent with an ureterocele. There is reflux into both lower pole collecting systems of duplex kidneys.

Courtesy of D. Mulvihill

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Critique: 173

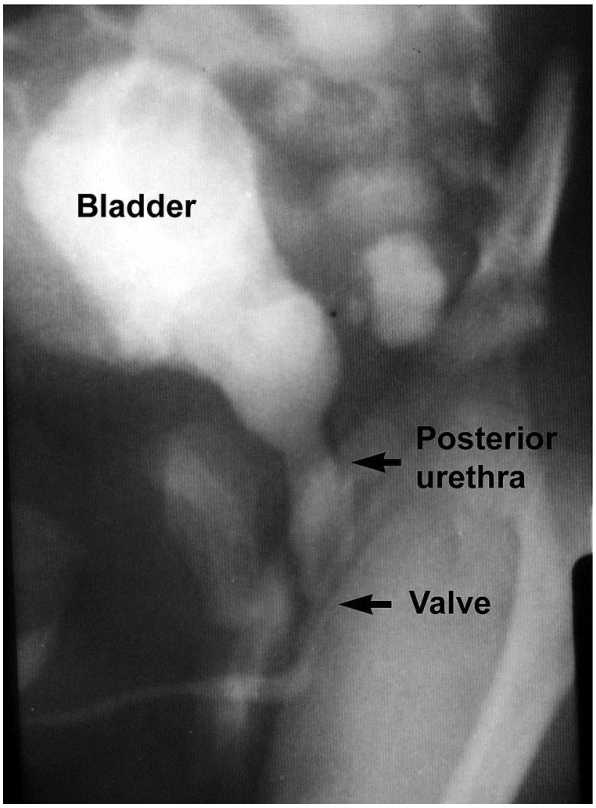


Sagittal ultrasonography in multicystic dysplastic kidneys shows only a collection of cysts of various sizes in the renal fossa without any parenchyma or a collecting system.

Courtesy of D. Mulvihill

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Critique: 173



A fluoroscopic spot film from voiding cystourethrography shows a significant change in urethral caliber at the level of the posterior urethral folds consistent with valves.

Courtesy of D. Mulvihill

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Question: 174

A 14-year-old boy presents to the emergency department with a severe asthma exacerbation and respiratory failure. Despite intubation and aggressive resuscitation, he develops severe acidosis, pulmonary edema, and hypoxic encephalopathy. His condition worsens over the next week, and the parents decide to withdraw care.

Of the following, the risk factor MOST associated with fatal asthma is

A. Caucasian race

B. high socioeconomic status

C. poor perception of symptoms

D. sensitivity to house dust mites

E. use of daily low-dose inhaled corticosteroids

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Critique: 174 Preferred Response: C

In the past 30 years, reported asthma-related case-fatality rates in industrialized countries have ranged from 1.5 to 8.5 per 100,000 persons. Overall, fatal asthma rates have increased worldwide, but some countries have experienced a decline in asthma mortality. Patients who have severe disease are believed to be at greatest risk, but those who have mild disease may have near-fatal or fatal asthma. Risk factors for near-fatal and fatal asthma include marked circadian variation in lung function, male sex, and poor perception of symptoms (Item C174). Neither daily inhaled corticosteroid use, Caucasian race, nor dust mite sensitivity has been linked to fatal asthma.

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Critique: 174

Item 174: Risk Factors for Asthma Deaths

- Past history of sudden severe exacerbations
- Prior intubation and mechanical ventilation for asthma
- Prior asthma admission to the intensive care unit
- Two or more asthma hospitalizations in the last year
- Three or more emergency department visits for asthma in the last year
- Hospitalization or emergency department visit in the last month
- Use of >2 canisters per month of short-acting bronchodilators
- Current use of systemic steroids or recent withdrawal from steroids
- Difficulty perceiving airflow obstruction and its severity
- Comorbidity that may affect cardiopulmonary status (eg, heart disease)
- Serious psychiatric disease or psychosocial problems
- Low socioeconomic status and urban residence
- Illicit drug use
- Sensitivity to *Alternaria*

Adapted from NHLBI, Expert Panel Report-2, 1997.
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Question: 175

A 16-year-old boy is brought to the emergency department because he had a seizure after returning home from being out with friends. His mother reports that he was agitated and aggressive when he first arrived home about 45 minutes ago and then became unresponsive and developed generalized extremity shaking that lasted for approximately 5 minutes. His friends reported that he “smoked weed,” but they denied any other drug or substance use. In the emergency department, he is diaphoretic and difficult to arouse, his heart rate is 140 beats/min, his respiratory rate is 30 breaths/min, his blood pressure is 135/95 mm Hg, and his pupils are mid-sized, equal, and sluggishly reactive. Nystagmus is noted.

Of the following, the MOST likely explanation for his symptoms is that

A. seizures are a common adverse effect of marijuana use

B. the marijuana was adulterated with phencyclidine

C. the patient has acute alcohol toxicity

D. the patient is exhibiting symptoms associated with Jimson weed exposure

E. the patient was inhaling gasoline in addition to smoking marijuana

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Critique: 175 Preferred Response: B

Drug use and abuse contribute significantly to the excess morbidity and mortality seen in adolescents. Although drug-related deaths are not common, drug use often contributes to adolescent deaths caused by motor vehicle crashes, homicide, other accidents, and suicide. Among adolescent patients who present for medical attention with drug-related symptoms, alcohol, marijuana, amphetamines, cocaine, and opiates are the agents most commonly involved, and often multiple ingestants are identified.

Emergency department visits for symptoms related to marijuana use occur more frequently than for those related to alcohol among adolescents. The most common reason cited for seeking medical care after marijuana exposure is the development of an unexpected reaction. Typical symptoms following marijuana exposure include somnolence and mild euphoria. Some patients may experience short-term memory impairment, decreased motor coordination and muscle strength, lethargy, slurred speech, ataxia, agitation, and panic reactions. Marijuana generally does not cause seizures, although rarely it may lower the seizure threshold in patients who have an underlying seizure disorder.

Marijuana commonly is adulterated with a variety of other drugs, one of which is phencyclidine. A powerful stimulant and dissociative agent, phencyclidine causes agitation, aggressive behavior, hypertension, tachycardia, and seizures. Rotary, vertical, and horizontal nystagmus (Item C175A) are seen in more than 50% of patients following phencyclidine exposure. Accordingly, the boy described in the vignette most likely smoked marijuana that was adulterated with phencyclidine.

Alcohol is a common coingestant in the adolescent age group. A sedative agent, alcohol typically produces somnolence, respiratory depression, bradycardia, and hypotension. Alcohol does not cause seizures, except in withdrawal states or in individuals who develop hypoglycemia, which is rare in the healthy adolescent.

Jimson weed *(Datura stramonium)* (Item C175B) is a plant that contains high concentrations of anticholinergic alkaloids and is found throughout the United States. The seeds of the plant are ingested or smoked for their hallucinogenic properties, but patients who use it also develop significant anticholinergic signs and symptoms. Ingestions are marked by bizarre behavior, tachycardia, hypertension, dry and flushed skin, and widely dilated pupils. Seizures may result.

The symptoms of inhalant exposure (eg, gasoline) typically are short-lived, resolving within 30 to 45 minutes of exposure. Symptoms generally are related to central nervous system stimulation or depression and include dizziness, excitation, lethargy, hallucinations, and ataxia. Seizures may occur acutely and often are related to acute hypoxia.

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Critique: 175



Jimson weed (*Datura stramonium*) contains high concentrations of anticholinergic alkaloids. The flowers are trumpet-shaped and white to purple. The leaves are large and have irregular teeth similar to oak leaves.

Courtesy of M. Rimsza

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Question: 176

A 5-year-old boy has been ill for 2 days with fever, decreased appetite, and a rash. On physical examination, you note ulcers on the tongue (Item Q176A) and soft palate, but the gingivae are spared. You also see oval vesicles with surrounding erythema on the hands (Item Q176B).

Of the following, the MOST likely diagnosis is

A. aphthae

B. hand-foot-and-mouth disease

C. herpangina

D. herpetic gingivostomatitis

E. thrush

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Question: 176



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Question: 176



Courtesy of D. Krowchuk

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Critique: 176 Preferred Response: B

Hand-foot-and-mouth disease is a distinctive syndrome usually caused by infection with Coxsackievirus A16 or enterovirus 71, although other Coxsackievirus types and echoviruses have been implicated. Hand-foot-and-mouth disease typically occurs during the summer and fall and is characterized by fever, sore throat, and rash. Oral involvement precedes the appearance of skin lesions. Shallow ulcers, measuring 1 to 5 mm, surrounded by a red border, are observed on the soft palate, uvula, tonsillar pillars, and tongue (Item C176A), as described for the boy in the vignette. Cutaneous lesions are erythematous papules or vesicles that typically are oval and surrounded by a rim of erythema. They are located acrally, involving the palms (Item C176B), soles, and digits, particularly the lateral aspects. The thighs and buttocks also may be affected, but the trunk and face generally are spared. Approximately two thirds of patients exhibit this typical presentation, 20% have only oral lesions, and 11% have acral lesions without mucosal involvement. Hand-foot-and-mouth disease generally is a mild illness and is self-limited, resolving in about 1 week.

Several disorders produce oral lesions that may mimic those of hand-foot-and-mouth disease, but the typical cutaneous lesions on acral surfaces are absent. Aphthae, also known as aphthous ulcers, are painful erosions that have a gray base and appear on the lips, gingivae, tongue, palate, and buccal mucosa (Item C176C). Individual lesions last 7 to 10 days and may occur in crops at irregular intervals. Oral ulcers are a feature of herpangina caused by Coxsackievirus A or B or echoviruses. Affected children may develop high fever, headache, and a characteristic enanthem composed of 1- to 2-mm vesicles and ulcers in the posterior pharynx, including the tonsillar pillars, soft palate, and uvula (Item C176D). Although the oral lesions of herpangina have a characteristic appearance, at times it may difficult to distinguish this disorder from incomplete forms of hand-foot-and-mouth disease (those in which cutaneous lesions are absent).

Gingivostomatitis due to herpes simplex virus infection occurs most often in children 6 months to 3 years of age. Symptoms include fever, irritability, and refusal to eat. Physical examination reveals vesicles and ulcers on the anterior soft palate, tongue, and gingivae. The gingivae are edematous, friable, and inflamed, and the breath is malodorous. Vesicles also may appear on the lips and chin (Item C176E); occasionally, autoinoculation results in lesions on the hands.

In contrast to the oral ulcers observed in patients who have hand-foot-and-mouth disease, thrush is characterized by white plaques, often on an erythematous base, located on the tongue and buccal mucosa (Item C176F).

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Critique: 176



In hand-foot-and-mouth disease, ulcers may appear on the tongue, soft palate, uvula, and tonsillar pillars.

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Critique: 176



Oval or round vesicles that have surrounding erythema occur on the palms and soles in hand-foot-and-mouth disease.

Courtesy of D. Krowchuk

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Critique: 176

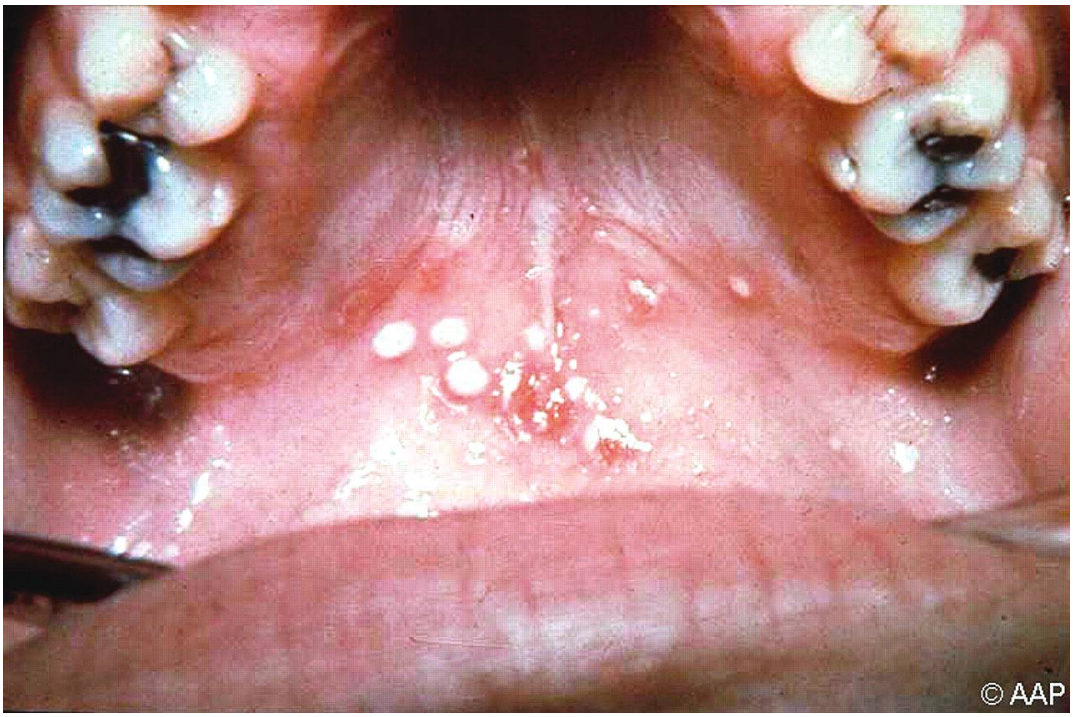


Aphthae are painful erosions or ulcers that appear on the lips, gingivae, tongue, palate, or buccal mucosa.

Courtesy of dermatlas.org

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Critique: 176



In herpangina, patients develop vesicles and ulcers in the posterior pharynx.

Courtesy of *Red Book¨ Online*

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Critique: 176



Vesicles and ulcers located on the soft palate, tongue, gingivae, lips, and chin are characteristic of herpetic gingivostomatitis.

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Critique: 176



Thrush produces white plaques, often on an erythematous base, located on the tongue and buccal mucosa.

Courtesy of D. Krowchuk

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Question: 177

A 13-month-old infant presents with a 1-month history of chronic diarrhea and weight loss. The baby tolerated cow milk formula well, but the diarrhea began around the time he was transitioned to whole milk. There is a family history of multiple food allergies. Physical examination demonstrates a thin infant whose weight is at the 10th percentile and height is at the 50th percentile. Stool cultures for enteric pathogens and viruses are negative. Results of complete blood count, chemistries, and serum immunoglobulin (Ig) A measurement are normal. Celiac serologies demonstrate a positive antigliadin IgG, negative antiendomysial antibodies, and negative tissue transglutaminase antibody. A small bowel biopsy demonstrates increased cellularity of the intestinal lamina propria and partial villous atrophy.

Of the following, a TRUE statement regarding the patient's small bowel biopsy is that the findings

A. are diagnostic for giardiasis

B. are nonspecific

C. are pathognomonic for rotavirus infection

D. exclude celiac disease

E. rule out milk protein allergy

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Critique: 177 Preferred Response: B

The patient described in the vignette underwent a small bowel biopsy to evaluate chronic diarrhea and weight loss. Although the patient has a positive antigliadin immunoglobulin G, the antibodies that are most sensitive and specific for celiac disease (anti-tissue transglutaminase and anti-endomysial) are both negative. The biopsy findings described in the vignette suggest intestinal injury, but are nonspecific. They do not distinguish between celiac disease, allergy, and infectious enteritis.

Intestinal small bowel biopsy of the duodenum or jejunum remains one of the most useful tests in the evaluation of a child who has chronic diarrhea or suspected malabsorption. Usually intestinal biopsy is performed by passing an endoscope into the duodenum and taking multiple tissue samples with a biopsy forceps. The samples can be evaluated by the pathologist to assess mucosal architecture and to look for specific infectious pathogens (eg, *Giardia lamblia*, *Cryptosporidium*). Biopsy findings associated with celiac disease typically show varying degrees of villous atrophy and increased intraepithelial lymphocytes. In contrast, increased eosinophils in the intestinal lamina propria can be seen in children who have allergic enteritis.

If necessary, a more detailed evaluation of the intestinal biopsy can be performed with special studies. Immunostaining can identify populations of inflammatory cells, thereby further characterizing autoimmune disease, immunodeficiency syndromes, or neoplastic cells. Electron microscopy can look for small pathogens such as microsporidia and identify ultrastructural abnormalities such as microvillous inclusion disease.

In spite of its usefulness, intestinal small bowel biopsy has many limitations. Many intestinal lesions are “patchy,” meaning some biopsy specimens may be normal while others are inflamed. For this reason, multiple (two to five) duodenal biopsies are preferred to minimize sampling error. The biopsy should be interpreted by an experienced gastrointestinal pathologist because minor degrees of inflammation are common in “normal” biopsies. Many findings, such as the increased cellularity and villous atrophy described in the vignette, are nonspecific indications of inflammation and can be seen in many different diseases (including celiac disease (Item C177), allergic enteropathy [eg, milk protein allergy], autoimmune enteropathy, and viral infection [eg, rotavirus infection]). Thus, although small bowel biopsy is an excellent tool to determine whether there is any intestinal mucosal damage, it does not always provide a specific diagnosis and must be interpreted in the context of the clinical and laboratory findings.

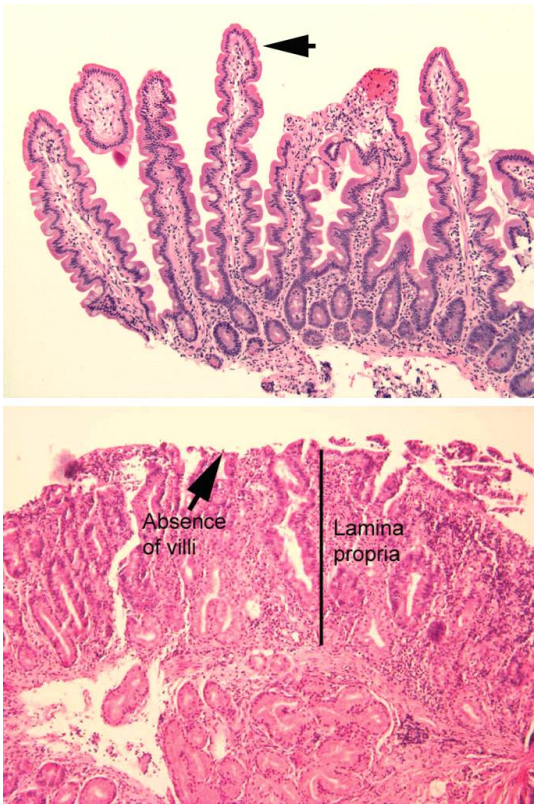
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Critique: 177



Top: Normal duodenum, with the villi (long fingerlike projections) (arrow) appearing normal.

Courtesy of J. Goldsmith

Bottom: Duodenal biopsy from a patient who has celiac disease shows atrophy of the villi (arrow) and increased cellular infiltrate in the lamina propria.

Courtesy of A. Bousvaros

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Question: 178

A 9-month-old infant who had been born at 25 weeks' gestation is receiving daily diuretics and nasal cannula oxygen with a baseline of 0.1 L/min flow. His mother called this morning, reporting that he had a temperature of 100.5°F (38.1°C), nasal congestion, increased work of breathing with a rapid respiratory rate, and "wheezing" cough. You instructed her to increase the oxygen flow rate to 0.5 L/min and advised her to bring him to your office. On arrival at the clinic, pulse oximetry reveals an oxygen saturation of 85% at rest. On physical examination, you note intercostal and subcostal retractions, a respiratory rate of 80 breaths/min, a heart rate of 140 beats/min, and a prolonged expiratory phase with audible wheezing. A copious clear nasal discharge is present. There is no heart murmur or gallop.

Of the following, the BEST explanation for this child's presenting signs of respiratory distress is

A. acute cor pulmonale

B. acute sinusitis

C. gastroesophageal reflux

D. lower respiratory tract infection

E. upper respiratory tract infection

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Critique: 178 Preferred Response: D

The preterm infant who has chronic lung disease (CLD) with an oxygen requirement at the time of discharge from the neonatal intensive care unit is at risk for a number of complications in the first 12 to 24 months at home. Chief among these are infections that may compromise pulmonary function. The infant described in the vignette was an extremely low-birthweight (ELBW) (<1,000 g) preterm infant whose CLD is being managed with diuretics and oxygen. The sudden onset of a low-grade fever, upper respiratory tract signs of congestion, lower respiratory tract signs of small airways constriction (wheezing and a prolonged expiratory phase), increased work of breathing, and increased oxygen requirement are best explained by a lower respiratory tract infection. Common viral pathogens include respiratory syncytial virus, parainfluenza virus, and human metapneumovirus. Lower respiratory tract processes may involve bronchiolitis or pneumonia. Chest radiographs may demonstrate hyperinflation with air-trapping, atelectasis, and patchy interstitial infiltrates (Item C178A).

Cor pulmonale may complicate CLD and presents with progressive increased work of breathing and oxygen requirement over weeks to months, symptoms not exhibited by the infant in the vignette. In addition, cardiomegaly may be observed on chest radiography, right ventricular hypertrophy and strain on electrocardiography, and likely findings of right ventricular dilation and pulmonary hypertension on echocardiography. Many ELBW infants who have CLD do not survive the development of secondary pulmonary hypertension and cor pulmonale, even in the postsurfactant era (Item C178B). Treatment of such infants with inhaled nitric oxide and phosphodiesterase inhibitors such as sildenafil is experimental at this time.

Acute sinusitis is uncommon in infancy and does not explain the associated hypoxemia reported for the infant in the vignette. Gastroesophageal reflux may be associated with wheezing or with frank aspiration of milk and pneumonitis, but there is no clear rhinorrhea. Upper respiratory tract infections may be characterized by low-grade fever and even corresponding wheezing, but the degree of hypoxemia and respiratory distress in the infant described in the vignette is explained best by a lower respiratory tract infection.

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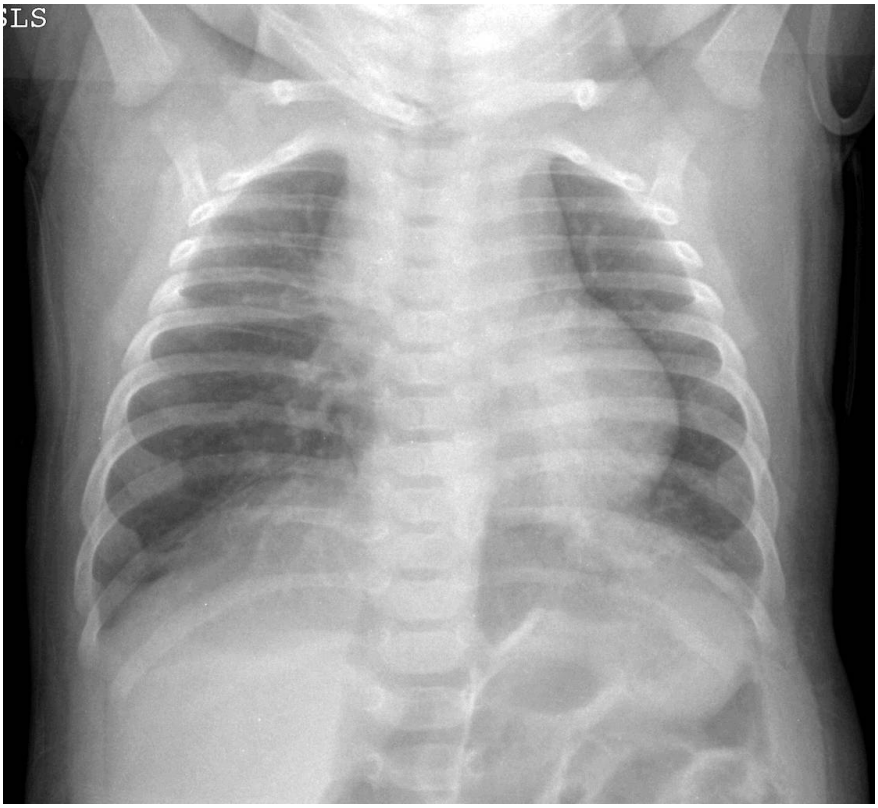
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Critique: 178

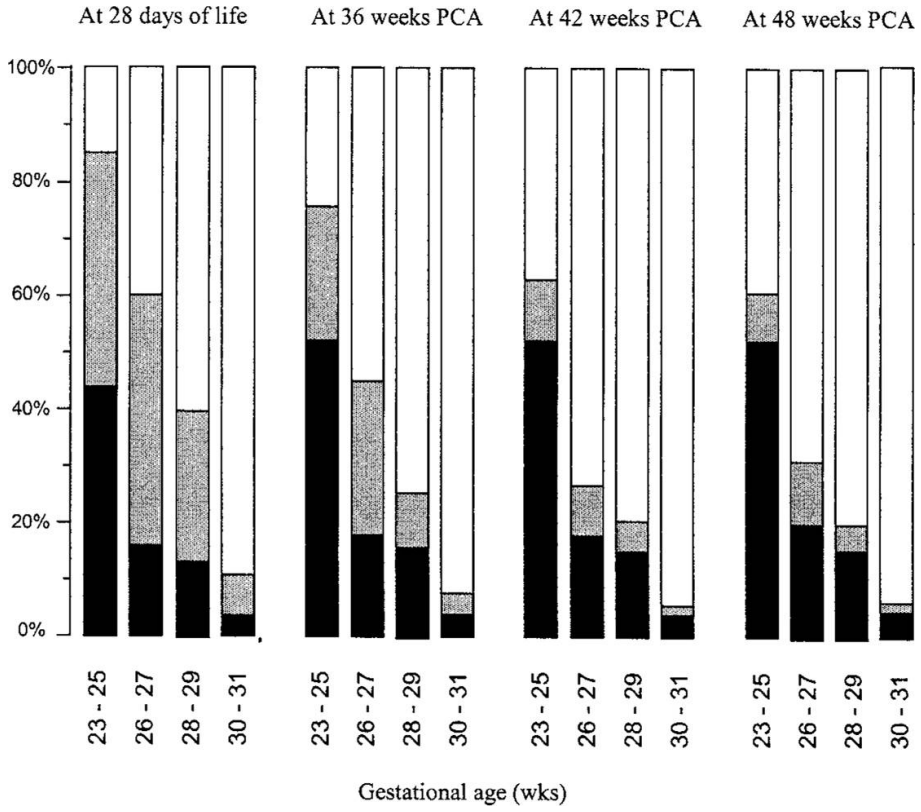


Respiratory syncytial virus bronchiolitis in which there is hyperinflation and bibasilar and right upper lobe atelectasis.

Courtesy of D. Krowchuk

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Critique: 178



Mortality and chronic oxygen dependency according to gestational age at 28 days after birth (days of life) and 36, 42, and 48 weeks' postconceptional age. For each band of gestational age, black bars indicate deceased patients, shaded bars indicate survivors who have chronic oxygen dependency (COD), and open bars indicate survivors who do not have COD.

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Question: 179

An 8-week-old breastfed boy is brought to the clinic for his health supervision visit. His mother thinks he may be more pale than her other children, but he has otherwise been healthy. Findings on physical examination and vital signs are normal. He does not appear pale to you. A complete blood count reveals hemoglobin of 9 g/dL (90 g/L) and a mean cell volume of 85 fL. The remainder of the complete blood count is normal.

Of the following, the MOST appropriate recommendation for this infant is to

A. administer a daily multivitamin and folic acid

B. admit the infant for a packed red blood cell transfusion

C. measure serum iron and transferrin concentrations

D. reassure the mother that the anemia will resolve

E. switch feeding to an iron-fortified infant formula

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Critique: 179 Preferred Response: D

Physiologic anemia of infancy occurs in term infants from 6 weeks to 3 months of age and is due to a number of factors. Fetal hemoglobin, which has a high affinity for oxygen, is replaced by an increasing amount of adult hemoglobin, which has a lower affinity for oxygen. This lower affinity, along with higher blood oxygen content, causes increased delivery of oxygen to the tissues, which downregulates production of erythropoietin. The result is decreased hemoglobin concentration, although the concentration rarely drops below 9 g/dL (90 g/L). The decrease may be more pronounced (down to 7 g/dL [70 g/L]) and occur earlier (3 to 6 weeks) in preterm infants. The anemia is normocytic, and other cell lines are unaffected. Once the tissue oxygen requirement exceeds oxygen delivery, erythropoietin is produced, and hemoglobin concentrations return to normal.

Normal mean cell volume values also vary by age. A normal value in a newborn is high, up to 110 fL at birth, decreasing to 70 to 74 fL at ages 6 months to 6 years. The mean cell volume provides valuable information in determining the cause of anemia in infants and children.

Because the infant described in the vignette is asymptomatic and has evidence of a mild normocytic anemia, the most likely diagnosis is physiologic anemia, and the best management is reassurance that the anemia will resolve by age 3 to 4 months. Adding a daily multivitamin with folic acid and switching to an iron-fortified infant formula are not indicated because the folate and iron contents of human milk at this age are appropriate for term infants. Further laboratory evaluation is not necessary, and a blood transfusion is not appropriate because most physiologic anemia resolves without hemodynamic compromise.

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Question: 180

A 6-month-old infant is hospitalized for recurrent apneic events. Notes in the history indicate multiple hospitalizations and emergency department visits, although no apnea alarms have been recorded in the presence of medical personnel. The alarms have occurred only when the mother is alone with the baby. The respiratory therapist now reports that she walked into the room as the alarm was going off and found the mother holding her hands over the baby's mouth and nose.

Of the following, the MOST important immediate step is to

A. arrange for video camera surveillance of the child's hospital room to document any repeat incidents

B. confront the mother after safely removing the infant from her care

C. consult child psychiatry on behalf of the family

D. consult social services to investigate the case

E. notify the child's father by phone of the incident

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Critique: 180 Preferred Response: B

Munchausen syndrome by proxy refers to the production of signs or symptoms of factitious illness in the child by the caregiver. The morbidity and mortality rate is high for this type of child abuse. For this reason, when the physician is faced with a possibility that symptoms and signs of illness in a child may be induced or fabricated by a caregiver, the primary concern is the safety of the patient.

One of the first steps in assessment for Munchausen syndrome by proxy is to observe the relationship between the occurrence of symptoms or signs and the presence or absence of the caregiver. In some cases, such as suspected airway compromise, acute life-threatening events, repeated poisoning, or suspected failure to give medication, the use of covert video surveillance has been proven to be useful, although its use is controversial. When Munchausen syndrome by proxy is proven or strongly suspected, as described for the infant in the vignette, the infant should be removed to a safe environment, after which clinicians should notify child welfare authorities, local law enforcement, and hospital security, as applicable. It is neither safe nor effective to confront the parent in the child's presence without ensuring the child's safety and the safety of the medical team. Because the event was witnessed, video surveillance is unnecessary and may place the infant at risk for further harm. Psychiatry consultation and social services referral both are indicated and helpful, but should be deferred until the child's safety is assured. Similarly, contacting the child's father may be deferred for a brief period of time.

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Question: 181

You are evaluating a 2-year-old girl for fever and fatigue. Her parents report that she has had a fever for 3 days, a progressive degree of fatigue, loss of appetite, and irritability. On examination, she has a temperature of 102.3°F (39.1°C), a heart rate of 160 beats/min, a respiratory rate of 40 breaths/min, and a blood pressure of 90/60 mm Hg. She has dry mucous membranes, mild intercostal retractions, and a 3/6 holosystolic murmur (Item Q181) at the cardiac apex. Her liver is palpable 3 cm below the costal margin. Her pulses are weak but palpable in all extremities.

Of the following, the MOST likely cause of this patient's clinical presentation is

A. dehydration from viral illness

B. Kawasaki disease

C. meningitis

D. myocarditis

E. rheumatic fever

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Critique: 181 Preferred Response: D

Myocarditis involves inflammation of the myocardium that causes some degree of tissue damage or necrosis. Causes may be infectious, the result of toxic exposures, or associated with connective tissue diseases. Numerous viral and bacterial agents have been associated with myocarditis, but Coxsackievirus B is the most common causative viral pathogen. Infants and young children are affected more commonly than older children, and there may be a seasonal distribution of cases, with occurrence in the spring and summer being more common.

Myocarditis is suspected when there is a new murmur, sudden cardiac failure, arrhythmia, or some combination of these following a flulike illness. Symptoms may be subtle, with subclinical changes in the myocardial performance, or severe, with cardiovascular collapse and shock. Most affected infants have the signs and symptoms of congestive heart failure, the typical result of significant myocardial inflammation. Young children may appear pale, diaphoretic, and irritable. With increasingly diminished cardiac output, there can be somnolence and lethargy. Findings on physical examination in infants typically include tachycardia and tachypnea as well as a holosystolic (Item C181) murmur of mitral regurgitation on auscultation, which is believed to result from distortion of the mitral valve supporting structure as the left ventricle dilates. Pulses and perfusion may be diminished, depending on the degree of myocardial dysfunction. Hepatomegaly may result if there is increased filling pressure in the atria.

The child described in the vignette has fatigue, anorexia, irritability, and a fever. Her clinical examination findings are most consistent with moderate congestive heart failure, and her new murmur of mitral regurgitation suggests the diagnosis of myocarditis. Dehydration and meningitis are not associated with hepatomegaly or a new cardiac murmur. Kawasaki disease and rheumatic fever may be associated with myocarditis, but both are systemic diseases that require the presence of other findings for diagnosis.

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Question: 182

An 18-month-old developmentally delayed child presents to your office for follow-up after an emergency department visit the previous day for a first seizure. The seizure began with jerking of the left arm, followed quickly by loss of responsiveness and jerking of the entire body. It lasted about 2 minutes and was followed by sleepiness for 4 hours. In the emergency department, the child was difficult to arouse. Head computed tomography scan showed no acute changes, and findings on lumbar puncture were normal. The child was afebrile. The examining physician diagnosed an ear infection and advised the family to follow up with you today. The child was born at 26 weeks' gestation. At present, he can sit but is not yet standing. Physical examination reveals a head circumference of 43 cm, and the head shape is dolichocephalic. Other remarkable findings on physical examination include hyperreflexia with crossed adduction at both knees and ankle clonus.

Of the following, you are MOST likely to advise the parents that

A. electroencephalography should be performed to confirm that the incident was a seizure

B. recurrent seizures in their child most likely will cause brain damage

C. the risk of seizure recurrence for their child is about one in three

D. the risk of seizure recurrence in their child is increased because of his developmental delay and hyperreflexia

E. their child has epilepsy and needs to begin antiseizure medication

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Critique: 182 Preferred Response: D

The child described in the vignette probably had an unprovoked seizure; there is no history of a fever to support a diagnosis of febrile seizure. For afebrile seizures, the neurodevelopmental history and the neurologic examination are critical for assessing the likelihood of seizure recurrence. Approximately 1 in 3 otherwise healthy children have a seizure recurrence in the first 12 months after their first afebrile seizure. However, because the child described in the vignette has neurologic abnormalities and developmental delay, the parents should be advised that his recurrence risk is higher. In several prospective studies of children who had first unprovoked seizures, the recurrence risk in children who had prior developmental delay, mental retardation, cerebral palsy, or other known central nervous system problems was 70% over the subsequent 5 years.

A careful history is more accurate than an electroencephalogram (EEG) for determining that a prior event was a seizure. The classic EEG finding in epilepsy, epileptiform discharges, has a sensitivity of 50% to 60% at best. In addition, epileptiform discharges occur in 2% to 5% of neurologically normal school children and in a much higher percentage of children who have neurologic abnormalities. For the child in the vignette, the predictive value of EEG for seizure recurrence is negligible.

A seizure in a child is extremely frightening for many parents, and an important role for the primary physician is to provide accurate information. Many parents fear seizures will cause brain damage or death. The current accepted view is that recurrent seizures in children rarely cause long-term brain damage. Parents who have unrealistic fears about death or brain damage from future seizures should be reassured that this is extremely unlikely.

Most experts do not recommend treatment with daily antiseizure medication after a single seizure, even in cases in which the recurrence risk is high. The child in the vignette does not meet diagnostic criteria for epilepsy. The diagnosis of epilepsy is clinical, based on a history of two or more unprovoked seizures, not based on EEG findings.

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Question: 183

You are seeing a 14-year-old boy for a physical examination, which he needs to have completed to attend summer camp. In screening him for safety issues, you find that he does not use a seat belt.

Of the following, a TRUE statement about seat belt use by adolescents is that

A. alcohol consumption is not significantly related to low seatbelt use in teens

B. male teens are more likely to use seatbelts than female teens

C. most high school students report that they rarely or never wear a seatbelt when riding as a passenger in a car

D. most youth who died as occupants in passenger vehicle crashes were not wearing seatbelts

E. motor vehicle collisions rank behind cancer and suicide as the leading cause of death for teenagers

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Critique: 183 Preferred Response: D

Motor vehicle crashes are the leading cause of death for teenagers. Seat belt use is the most effective countermeasure to prevent injuries and fatalities in motor vehicle crashes. The failure of adolescents to use seatbelts contributes greatly to mortality risk; in 2005, 56.8% of youth 16 to 20 years of age and 54.8% of youth 10 to 15 years of age who died as passenger car occupants were not wearing seatbelts. Fortunately, the percentage of high school students who reported rarely or never wearing a seatbelt when riding in a car driven by someone else decreased from 25.9% in 1991 to 10.2% in 2005, as measured by the National Youth Risk Behavior Survey. Teens who have been drinking are less likely to use seatbelts, and male teens are less likely to use a seatbelt than female teens.

Pediatricians should address the wearing of seatbelts throughout adolescence and support legislation setting more rigorous safety belt and child restraint laws that specify primary enforcement and mandatory use by all occupants. Pediatricians also should advise parents to set a good example of requiring all occupants to use seatbelts, not drink and drive, and not speed.

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Question: 184

A 17-year-old boy has had poorly controlled type 1 diabetes (DM1) since the age of 2 years. When you saw him 2 months ago, his hemoglobin A1c result was 11.3%. He complains of burning and tingling in his feet, which his diabetologist has told him is sensory diabetic neuropathy. He asks you about treatment for the burning.

Of the following, you are MOST likely to explain that neuropathic pain will be improved by

A. angiotensin-converting enzyme inhibitor therapy

B. coenzyme Q10 therapy

C. high-dose vitamin E therapy

D. intensive physical therapy

E. normalization of blood glucose

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Critique: 184 Preferred Response: E

The long-term complications of diabetes include retinopathy, nephropathy, neuropathy, and early microvascular and macrovascular disease. Delay or prevention of the complications of diabetes depends on glycemic control, blood pressure control, and smoking cessation. The Diabetes Control and Complications Trial proved conclusively that glycemic control could delay the onset of such complications. Even many years after the end of this trial in the early 1990s, individuals who were enrolled in the tight control group fared better in terms of severity of disease complications. The cause of neuropathy probably is multifactorial, involving metabolic changes as well as shifts in vascular supply to the nerves. Small studies demonstrate that the only treatment that offers long-term improvement in neuropathy is improvement in diabetes control. Unfortunately, following the institution of improved glycemic control, regrowth of small pain fibers may intensify neuropathic pain for a period of time until improvement occurs. Other therapies, including physical therapy, vitamin therapy, treatment with coenzyme Q10, and treatment to normalize proteinuria and lower blood pressure with an angiotensin-converting enzyme inhibitor have no objective effect on neuropathic pain. Tricyclic antidepressants, gabapentin, and locally applied capsaicin have been used with variable success.

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Question: 185

An 11-year-old boy was caught by his parents setting fire to paper in his bedroom wastebasket. He denied ever doing this before, but his parents found a used box of matches under his bed. He also is rough with the family pet, often pulling out the dog's fur. He once grabbed the dog by his ears and lifted him off the ground. His parents ask your advice on how to handle the boy's behavior.

Of the following, you are MOST likely to

A. reassure the parents that this is typical preadolescent behavior

B. recommend a medication trial with long-acting methylphenidate

C. refer the boy for an educational evaluation

D. refer the boy for a psychiatric evaluation

E. suggest that the parents restrict the boy's extracurricular activities if he misbehaves again

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Critique: 185 Preferred Response: D

Children who have a conduct disorder have a constellation of behavioral issues that make it difficult for them to behave in a socially acceptable manner. A child meets the criteria for conduct disorder established in the *Diagnostic and Statistical Manual of Mental Disorders* edition IV-TR by displaying some of the following behaviors: aggression to people and animals, destruction of property, deceitfulness, lying or stealing, or serious violations of rules. Conduct disorder generally is diagnosed at a younger age in boys (10 to 12 years) than in girls (14 to 16 years). Affected children may express their aggressive behaviors in the form of bullying and cruel behavior toward peers. Children may be verbally abusive and defiant. Persistent lying, frequent truancy, and vandalism are common in severe cases.

The fire-setting and abuse of the family pet described for the boy in the vignette is evidence of conduct disorder or another serious psychiatric disturbance for which evaluation and treatment should be coordinated by a psychiatrist. He is not exhibiting typical preadolescent behavior. The removal of privileges would not be effective in view of his mental health issues. The use of methylphenidate is indicated for treatment of attention-deficit/hyperactivity disorder, which is characterized by behaviors indicative of lack of attention and hyperactivity/impulsivity. An educational evaluation is indicated if there is evidence of academic difficulties.

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Question: 186

You evaluated a child in your clinic 3 days ago for a protracted course of diarrhea. Findings on your initial evaluation were unremarkable, but you did send stool samples for culture. Today the laboratory reports that the stool samples are growing *Yersinia enterocolitica*. When you tell the mother of these results, she is convinced that her daughter contracted the organism from contaminated food. She asks what types of foods transmit this organism.

Of the following, infections due to *Y enterocolitica* MOST likely are associated with the consumption of

A. beef

B. chicken

C. fish

D. pork

E. shrimp

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Critique: 186 Preferred Response: D

Human infections due to *Yersinia enterocolitica* result from ingestion of contaminated foods, unpasteurized milk, or water. The handling of raw pork or pork intestine is a major risk factor for the acquisition of this organism. Outbreaks of infection due to other pathogens such as *Escherichia coli* 0157:H7 (beef, spinach), *Salmonella* sp (peanut butter, poultry), or *Campylobacter jejuni* (poultry) are not associated with pork. Fish and shrimp usually are not associated with any specific bacterial agents.

Infections due to *Y enterocolitica* most commonly present as enterocolitis (fever and diarrhea) in children younger than 5 years of age. The stools may contain blood, mucus, and fecal leukocytes. In older children and adults, pseudoappendicitis is a common presentation. Patients have signs and symptoms consistent with appendicitis (abdominal pain, tenderness of the right lower quadrant, leukocytosis), but at the time of surgery, the appendix appears normal, and mesenteric adenitis may be found. Bacteremia and focal infections (eg, hepatic, splenic abscess) are uncommon with *Y enterocolitica* and usually are encountered in children who have predisposing conditions such as iron storage disorders and immunosuppressive states. Postinfectious sequelae such as erythema nodosum and reactive arthritis are seen in older children and adults, particularly those who are positive for the HLA-B27 antigen.

Y enterocolitica can be recovered from normally sterile sites and the stool, but because special media is required for recovery, the laboratory should be notified when a *Yersinia* infection is suspected. Patients in whom the organism is recovered from sites other than the stool or patients who are immunocompromised should receive antimicrobial therapy. *Y enterocolitica* usually is resistant to first-generation cephalosporins (eg, cephalexin) and penicillins but is susceptible to trimethoprim-sulfamethoxazole, aminoglycosides, cefotaxime, fluoroquinolones (for patients 18 years of age and older), and doxycycline. Other than decreasing the duration of fecal excretion of *Y enterocolitica*, the benefit of antimicrobial therapy for enterocolitis, pseudoappendicitis, or mesenteric adenitis has not been established.

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Question: 187

You are called by the director of a preschool regarding an outbreak of gastroenteritis that occurred in the 2- and 3-year-old toddler classes. She states that 8 of 20 children developed vomiting and diarrhea associated with fever several days after the school picnic. All the children recovered from their illness after about 1 week.

Of the following, the MOST likely causative organism was

A. adenovirus

B. *Bacillus cereus*

C. Coxsackievirus

D. rotavirus

E. *Staphylococcus aureus*

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Critique: 187 Preferred Response: A

Diarrheal diseases in children younger than 5 years of age remain one of the leading causes of morbidity and mortality in developing countries and are an important cause of morbidity in developed countries. In the United States, children in this age range have 35 to 40 million episodes of diarrhea annually, resulting in about 170,000 hospitalizations and 300 deaths due to complications from diarrheal disease. Item 187 (Item C187) lists the most common viral and bacterial diarrheal pathogens in children by age group.

Most of the infectious organisms that cause diarrhea are spread via the fecal-oral route or through contaminated food and water. Enteric adenovirus disease primarily affects children younger than 4 years of age, although infection may occur at any age. Noroviruses (formerly Norwalk virus or Norwalk-like virus) may be associated with the consumption of raw seafood, ice, salads, and cookies, usually contaminated by infected food handlers. Noroviruses are common causes of sporadic illness in children younger than 4 years of age, but outbreaks of gastroenteritis may occur in all age groups. Outbreaks tend to occur in closed populations, such as child care centers and cruise ships, and there is a high attack rate. Water also has been documented as a vehicle of transmission of *Giardia lamblia*, *Campylobacter*, *Cryptosporidium*, and noroviruses. Child care centers serve as important reservoirs for transmission of diarrheal agents, with the peak incidence of diarrhea occurring in children younger than 3 years of age who are not toilet-trained.

The children described in the vignette most likely developed diarrhea after ingesting contaminated food or water at the school picnic. Given the timing of the development of the diarrhea, the children most likely have adenoviral gastroenteritis. Most children have been infected with rotavirus by the age of 3 years, and rotavirus gastroenteritis typically is not associated with spread from contaminated food and water. Coxsackievirus infection commonly manifests as gastrointestinal symptoms when there is systemic disease, but such symptoms rarely are the major complaint. The children in the vignette demonstrate no other findings consistent with systemic Coxsackievirus infection. *Staphylococcus aureus* and *Bacillus cereus* both are causes of gastroenteritis, but the illness associated with both organisms usually develops within 1 to 8 hours after ingestion of the contaminated food item. Further, both illnesses are self-limited and usually resolve within 48 hours of onset.

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Critique: 187

Item187: Common Diarrheal Pathogens

Infants and Toddlers	Children 5 to 12 Years	Adolescents
Rotavirus	Noroviruses (formerly Norwalk and Norwalk-like virus)	Noroviruses (formerly Norwalk and Norwalk-like virus)
Enteric adenovirus	*Giardia*	*Campylobacter*
Salmonella	*Escherichia coli* – enteropathogenic (EPEC)	*Escherichia coli* – enterotoxigenic (ETEC)
Shigella	*Escherichia coli* – enterohemorrhagic (EHEC)	*Escherichia coli* – enterohemorrhagic (EHEC)
Campylobacter	*Escherichia coli* – enterotoxigenic (ETEC)	*Salmonella*
Yersinia	*Salmonella*	*Shigella*
Giardia	*Campylobacter*	

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Question: 188

During the routine examination of a 1-day-old term infant, you palpate an abdominal mass. His growth parameters and blood pressure are within normal limits.

Of the following, the MOST likely explanation for the mass is

A. bowel duplication

B. multicystic dysplastic kidney

C. neuroblastoma

D. renal vein thrombosis

E. Wilms tumor

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Critique: 188 Preferred Response: B

A palpable abdominal mass in the newborn usually is related to the kidney. The most common lesions responsible are multicystic dysplastic kidney (MCDK) and hydronephrosis due to ureteropelvic junction (UPJ) obstruction.

Anomalies such as MCDK are detected either on in utero ultrasonography or postnatally as an abdominal mass. On ultrasonography, the lesion is associated with large renal cysts that are noncommunicating (Item C188). An MCDK is a nonfunctioning nephron unit; a patient who has a unilateral MCDK and a normal contralateral kidney is considered to have a single kidney. The incidence of MCDK is estimated at 1 in 4,000 live births. An additional important aspect of MCDK is that vesicoureteral reflux (VUR) occurs in approximately 30% of cases into the functioning, contralateral kidney. Also, approximately 15% of contralateral kidneys may have a component of obstruction. The standard approach to an infant in whom a unilateral MCDK is diagnosed begins with the institution of prophylactic antibiotics (based on the 30% risk of VUR in the contralateral kidney). Voiding cystourethrography and a diuretic renal scan (DRS) should be obtained. The DRS generally demonstrates normal uptake by the "healthy" contralateral kidney and absence of uptake in the region of the nonfunctioning MCDK.

If not diagnosed by in utero ultrasonography, hydronephrosis may present as an abdominal mass in the newborn period. The most common cause is UPJ obstruction. More rarely, it is caused by ureterovesical junction (UVJ) obstruction, single-system ureterocele, or VUR. The approach to a patient in whom hydronephrosis is suspected begins with renal/bladder ultrasonography to confirm the hydronephrosis and screen for other lesions while assessing the contralateral kidney. Prophylactic antibiotics are recommended, followed by voiding cystourethrography to look for VUR and a technetium-99m mercaptoacetyltriglycerine (MAG-3) radioisotope scan with furosemide to screen for UPJ or UVJ obstruction.

Less common causes of an abdominal mass in a newborn include gastrointestinal abnormalities (eg, bowel duplication), neuroblastoma, renal vein thrombosis, and Wilms tumor (rare in this age group).

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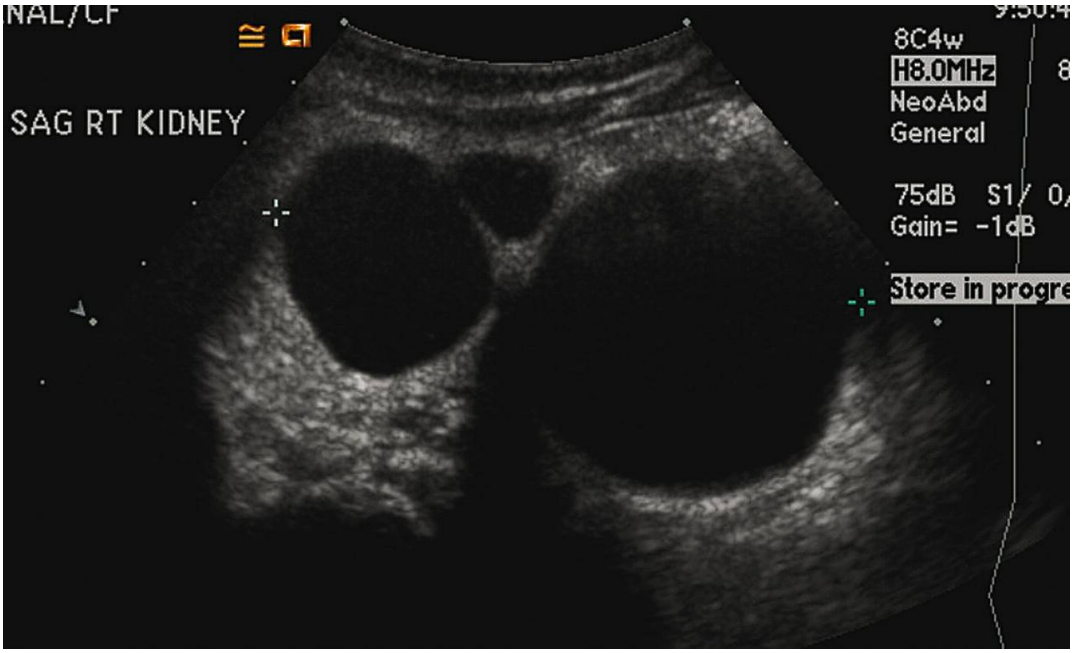
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Critique: 188



Sagittal ultrasonography of multicystic dysplastic kidneys shows only a collection of cysts of various sizes in the renal fossa without any parenchyma or a collecting system.

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Question: 189

An 11-year-old girl presents with a 6-month history of coughing, wheezing, and chest tightness. She usually has these symptoms three times a week during the day, but also wakes up at night once a month with the same symptoms. The symptoms have improved when she has used her mother's beta-2 agonist inhaler, but her parents are worried that she sometimes misses school because of her difficulty breathing. You suspect asthma.

Based on the frequency of her symptoms, the BEST categorization of this girl's asthma severity is

A. exercise-induced asthma

B. mild intermittent asthma

C. mild persistent asthma

D. moderate persistent asthma

E. severe persistent asthma

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Critique: 189 Preferred Response: C

After establishing the diagnosis of asthma, it is helpful to classify disease severity according to the National Asthma Education and Prevention Program (NAEPP) guidelines (Item C189). Current asthma guidelines use four clinical features to establish asthma severity: daytime symptoms, nighttime symptoms, lung function, and peak flow variability. An individual's level of severity is assigned based on the most severe of the four features. The most severe clinical feature described for the girl in the vignette is daytime symptoms, which occur more than two times a week but less than daily and categorize her disease as mild persistent asthma.

Although helpful, guidelines have limitations. For example, it is well known that overall asthma severity can change over time. Also, individuals can experience mild, moderate, and severe exacerbations regardless of baseline asthma severity. Guidelines provide an excellent basis for discussing therapeutic options, symptom control, and asthma education, but clinicians should be flexible in approaching an individual patient's medication and symptoms over time.

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Critique: 189

Item 189: Classification of Asthma Severity

	Mild intermittent	Mild persistent	Moderate persistent	Severe persistent
Daytime symptoms	≤2 times/wk	>2 times/wk	Daily symptoms	Continual symptoms
Nighttime symptoms	≤2 times/mo	>2 times/mo	>1 time/wk	Frequent
Lung function FEV_1	≥80%	≥80%	>60% to <80%	≤60%
Peak flow variability	<20%	20% to 30%	>30%	>30%

FEV_1=forced expiratory volume in 1 second. Adapted from Guill MF. *Pediatr Rev*. 2004;10:335-344.

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Question: 190

During a busy morning, a mother runs into your office with her 4-month-old infant, who is limp and cyanotic. She tells you that the baby was having difficulty breathing overnight, and on the way to the office this morning, he stopped breathing in the car. On physical examination, the infant has apnea and no pulses. You begin bag-valve-mask ventilation with 100% oxygen, and your nurse begins chest compressions.

Of the following, the MOST appropriate approach to cardiopulmonary resuscitation for this patient is

A. a compression rate of 80/min

B. a compression:ventilation rate of 30:2

C. application of an automatic external defibrillator

D. cessation of compressions when the heart rate is greater than 40 beats/min

E. use of the two thumb-encircling hands technique

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Critique: 190 Preferred Response: E

The appropriate hand position for infant cardiopulmonary resuscitation (CPR) when performed by clinicians is the two thumb-encircling hands technique (Item C190). The rescuer places his or her hands around the infant's thorax with the thumbs together over the lower half of the sternum above the xiphoid. Compressions are delivered with the thumbs while the fingers squeeze the thorax and provide counterpressure. This technique has been demonstrated to produce higher coronary artery perfusion pressure and more consistent compression depth than the two fingers on sternum technique.

When performing chest compressions during CPR, the rescuer should "push hard" to depress the chest approximately one third to one half the anteroposterior diameter and "push fast" at a rate of 100 compressions/min to generate adequate cardiac output to maintain cerebral and cardiac perfusion. In addition, rescuers should allow the chest to recoil at the end of each compression to enable cardiac refilling.

When two rescuers are performing CPR, the recommended compression-to-ventilation rate is 15 compressions to 2 breaths. If a definitive airway has not been secured through endotracheal intubation, the individual providing the compressions should pause after 15 compressions so that two breaths can be administered either via mouth-to-mouth or a bag-valve mask device. A single rescuer should administer 30 compressions, then pause to give two breaths. Once a definitive airway has been established, ventilations should be delivered at a rate of 8 to 10/min independently of compressions, which should be performed without pause at a rate of 100/min.

If available, the use of an automatic external defibrillator is recommended in children older than 1 year of age who suffer a sudden, witnessed arrest. This device analyzes the patient's cardiac rhythm and defibrillates the patient if ventricular fibrillation or ventricular tachycardia is identified. Although such dysrhythmias are not common in young children, they are increasingly recognized as causes of arrest in older children, and rapid defibrillation can be lifesaving.

Chest compressions should be discontinued in infants and children when the pulse rate is greater than 60 beats/min and perfusion is adequate. Bradycardia in infants and children (<60 beats/min) rarely is adequate to maintain perfusion and should be augmented by chest compressions. Conversely, adequacy of perfusion is not dependent on heart rate alone. In the patient whose heart rate is greater than 60 beats/min but who has evidence of poor perfusion (eg, delayed capillary refill, weak pulses, cool extremities), compressions should be continued until perfusion is deemed to be adequate.

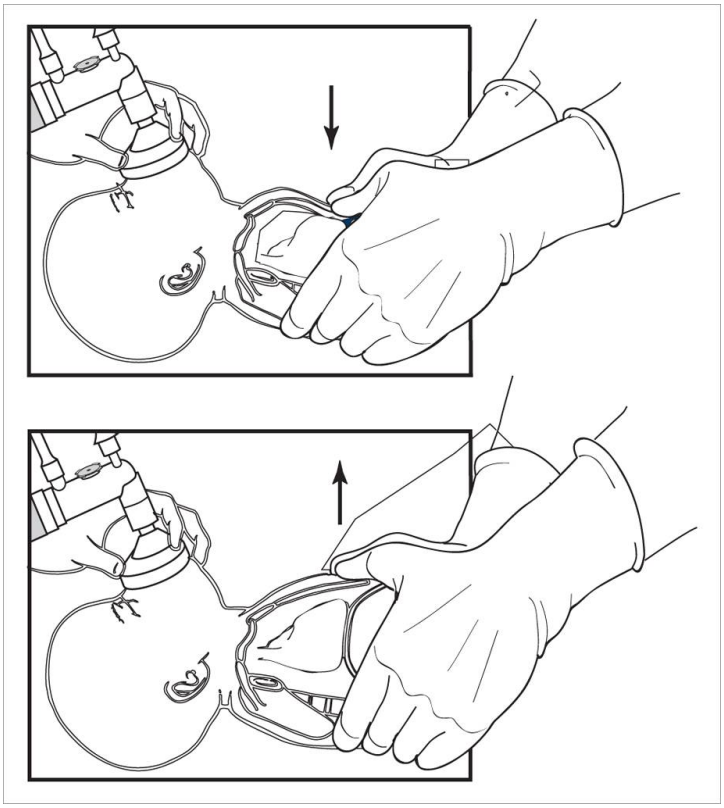
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Critique: 190



Compression (top) and release (bottom) phases of chest compression using the two-thumb encircling hands technique.

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Question: 191

During the examination of a 2-year-old child, you observe an abnormality of the labia minora (Item Q191). The patient's mother reports that the girl has had several episodes of perineal discomfort and pruritus during the past 6 months. The child's past medical history is unremarkable, and she is otherwise well.

Of the following, the MOST appropriate management is

A. application of a potent corticosteroid cream

B. application of an estrogen cream

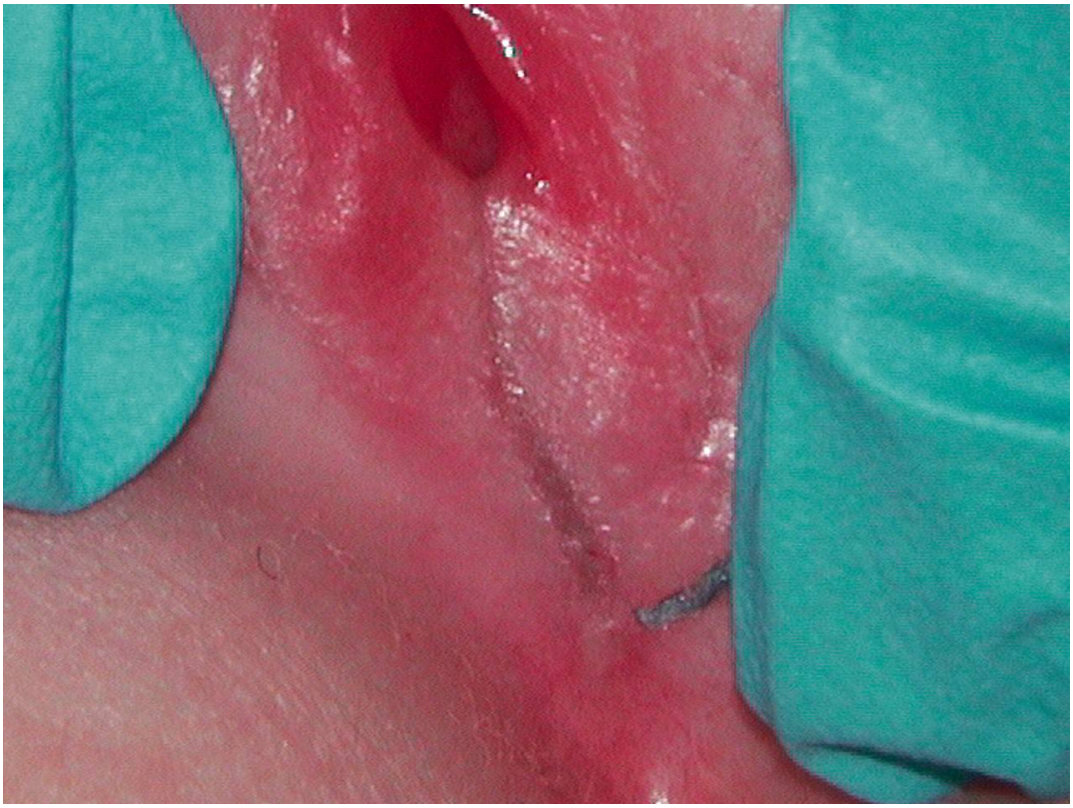
C. genital examination under anesthesia

D. manual lysis of adhesions

E. surgical lysis of adhesions

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Question: 191



Courtesy of S. Sinal

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Critique: 191 Preferred Response: B

The infant described in the vignette has posterior labial adhesions (Item C191). Labial adhesions are common in children younger than 6 years of age and are believed to result from inflammation due to local irritation and the physiologic hypoestrogenic state. Although often asymptomatic, labial adhesions may result in pooling of urine in the vagina and recurrent vulvovaginitis, factors that may cause dysuria, perineal discomfort, and possibly, recurrent urinary tract infections. In many patients, labial adhesions resolve spontaneously within 1 to 2 years; in nearly all, they disappear by puberty, when estrogen levels rise.

In an asymptomatic patient, the necessity for treatment is controversial. However, for those who have symptoms, such as the child described in the vignette, the application of estrogen cream is recommended. It should be applied sparingly and only to the area of labial fusion once or twice daily. Adhesions that are thin or filmy often respond within 7 to 10 days; those that are thicker may require longer treatment (6 to 8 weeks). Topical estrogen treatment is successful in 50% to 88% of patients. Although considered safe, prolonged or excessive application may result in vulvar hyperpigmentation, breast hypertrophy, or vaginal bleeding. Once separation has occurred, zinc oxide paste or petrolatum should be applied once or twice daily for 1 to 2 months in an attempt to prevent recurrent adhesions.

Topical corticosteroid creams are useful in the management of lichen sclerosus et atrophicus that may result in labial fusion, but are not indicated for management of typical adhesions. Examination under anesthesia is unnecessary to diagnose labial adhesions, but may be warranted in the evaluation of a child who has persistent unexplained vaginal discharge that may be the result of a foreign body. For the minority of patients who fail to respond to topical estrogen therapy and who have thick, fibrous adhesions, surgical lysis under anesthesia may be considered. Manual lysis performed in the office can be effective and obviates the need for general anesthesia, but despite the use of topical anesthetics, the procedure may be associated with moderate-to-severe discomfort.

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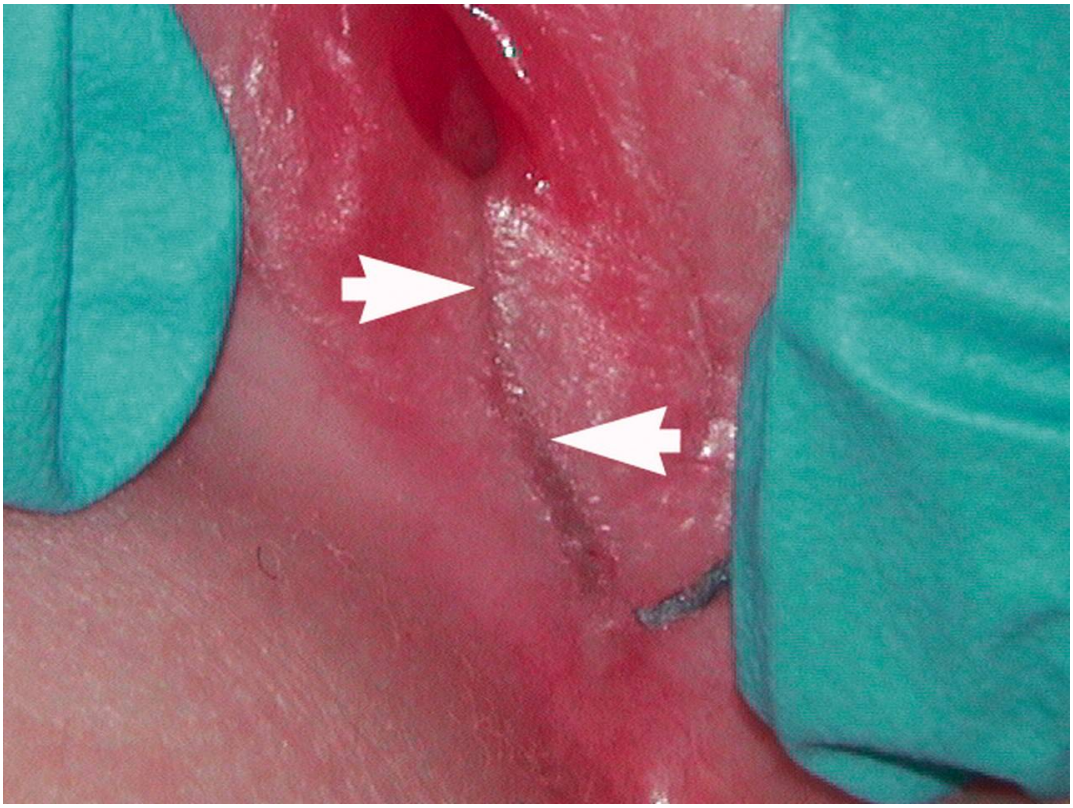
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Critique: 191



Labial adhesions (arrows) may result from inflammation due to local irritation.

Courtesy of S. Sinal

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Question: 192

You are evaluating a 1-month-old term infant who has persistent jaundice. The parents explain that his stools were green 2 weeks ago and now are pale yellow. Physical examination findings are unremarkable, except for a liver that is palpable 2 cm below the costal margin. The infant's total bilirubin is 6.1 mg/dL (104.3 mcmol/L) and direct bilirubin is 4.2 mg/dL (71.8 mcmol/L). Alanine aminotransferase is 240 U/L, and aspartate aminotransferase is 160 U/L. A hepatobiliary iminodiacetic (HIDA) nuclear medicine scan demonstrates absence of excretion of tracer into the bowel (Item Q192).

Of the following, the MOST definitive diagnostic test to establish the diagnosis is

A. intraoperative cholangiography

B. magnetic resonance cholangiopancreatography

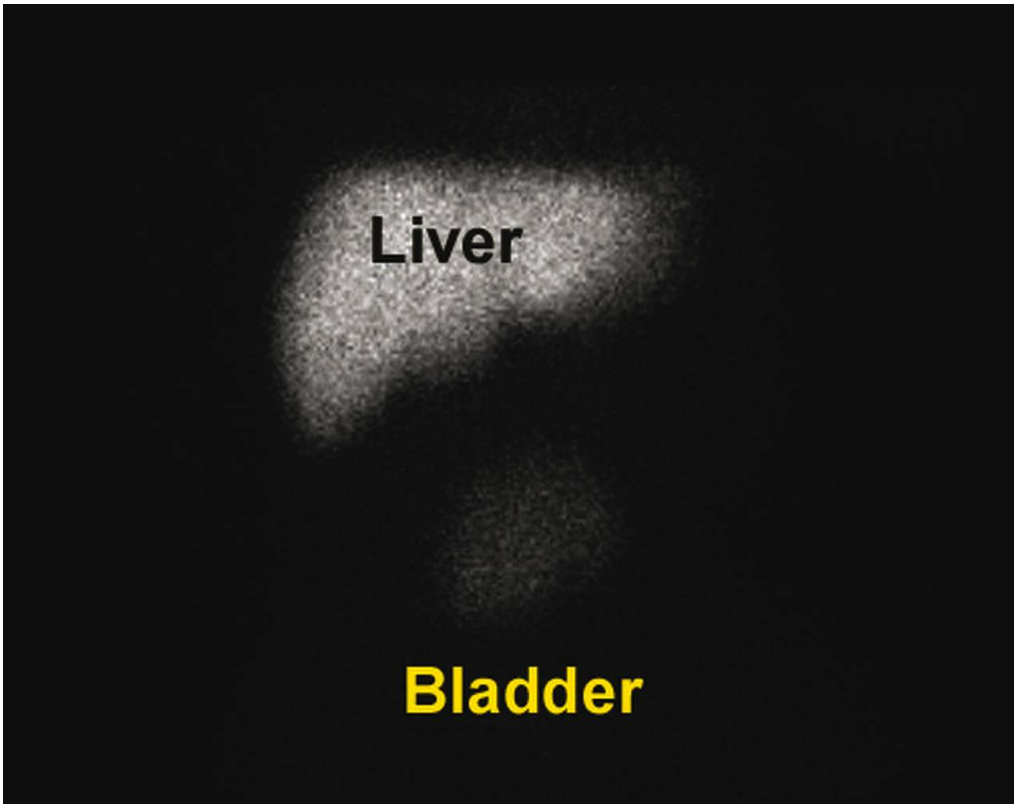
C. measurement of serum alpha-1-antitrypsin

D. sweat chloride test

E. ultrasonography of the biliary tree

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Question: 192



Anteroposterior view of a hepatobiliary iminodiacetic acid (HIDA) scan shows intense accumulation of tracer in the liver but not in the bowel. A small amount of tracer is seen in the bladder.

Courtesy of D. Mulvihill

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Critique: 192 Preferred Response: A

The initial evaluations of an infant who has a history of persistent jaundice, such as the one described in the vignette, are review of the perinatal history for any sign of blood group incompatibility and measurement of total and direct bilirubin and complete blood count. In this case, laboratory studies demonstrate a direct hyperbilirubinemia consistent with a neonatal cholestatic syndrome. For infants who have neonatal cholestasis, the pediatrician must exclude biliary atresia. If a radionuclide hepatobiliary scan does not show excretion into the bowel (Item C192), the most reliable test to exclude or diagnose biliary atresia is intraoperative cholangiography. Simultaneously, the surgeon can perform a wedge liver biopsy to evaluate the hepatic histology more carefully. Some gastroenterologists obtain a percutaneous needle liver biopsy before referral to a surgeon for intraoperative cholangiography. Although ultrasonography and magnetic resonance imaging studies can be helpful, at this time they are not sufficiently sensitive to exclude biliary atresia definitively. Sweat test and measurement of alpha-1-antitrypsin concentrations are useful when evaluating an infant who has cholestasis, but they cannot rule out biliary atresia.

Cholestasis is present if the conjugated (direct) bilirubin is greater than 2 mg/dL (34.2 mcmol/L) or if the conjugated fraction exceeds 20% of the total serum bilirubin. The evaluation of the infant who has neonatal cholestasis is complex and involves distinguishing among anatomic, infectious, and metabolic causes. The evaluation also depends greatly on the clinical history. For example, biliary atresia is very likely in a healthy term infant presenting with direct hyperbilirubinemia and acholic stool at 4 weeks of age. In contrast, biliary atresia is less likely for a preterm infant who has been receiving parenteral nutrition for 6 weeks and gradually develops cholestasis; that infant most likely has parenteral nutrition-associated liver disease. Physical examination may identify congenital cataracts (seen in rubella and galactosemia), neurologic abnormalities, or a heart murmur (seen in Alagille syndrome). The Alagille syndrome consists of: intrahepatic cholestasis, posterior embryotoxon of the iris, vertebral anomalies, peripheral pulmonic stenosis, and typical facies (prominent forehead, pointed chin, and hypertelorism).

Laboratory evaluation of a child who has cholestasis includes determination of liver chemistries (aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, gamma glutamyl transpeptidase, and bilirubin) as well as assessment of hepatic synthetic function (albumin, prothrombin time, partial thromboplastin time). Anatomy is assessed by ultrasonography, hepatobiliary scintigraphy, and if necessary, liver biopsy and cholangiography. If infection is suspected, evaluation for cytomegalovirus, rubella, hepatitis viruses, toxoplasmosis, syphilis, and human immunodeficiency virus can be performed. Metabolic testing may include sweat test, serum alpha-1-antitrypsin measurement, evaluation for galactosemia and tyrosinemia, serum bile acid analysis, and genetic testing for hereditary cholestatic syndromes.

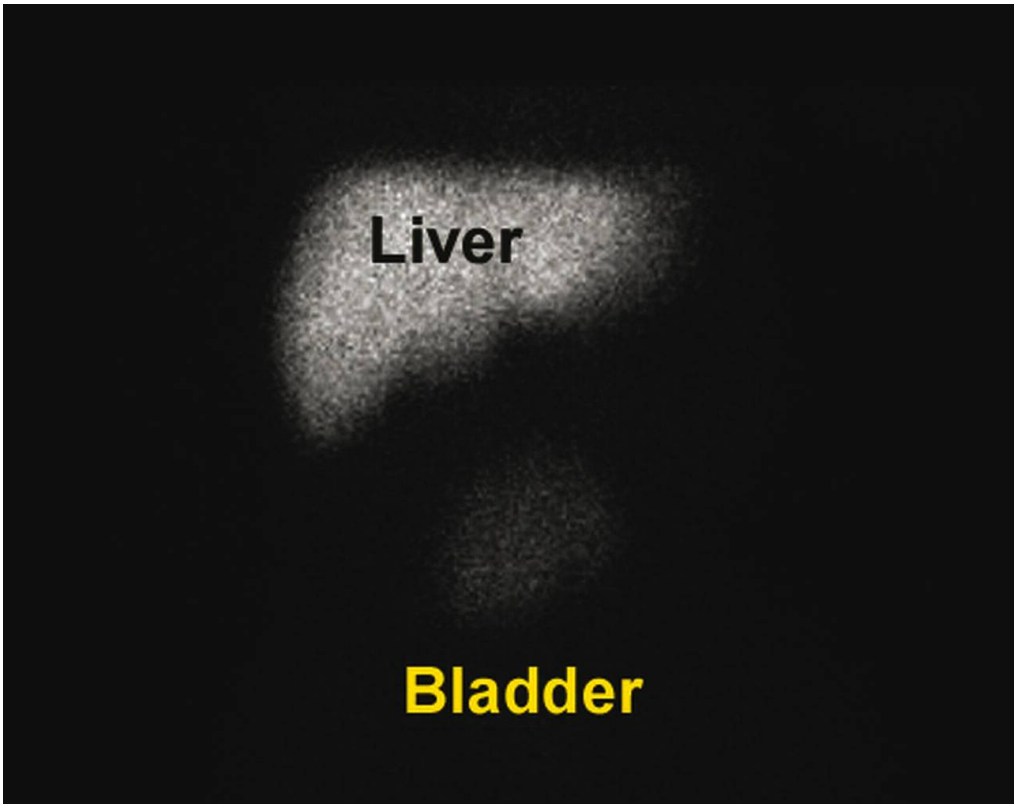
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Critique: 192



Anteroposterior views of a hepatobiliary iminodiacetic acid (HIDA) scan. In a normal study (top), there is tracer activity in the liver and in the bowel, indicating an intact biliary system. In biliary atresia (bottom), there is intense accumulation of tracer in the liver but not in the bowel. A small amount of tracer is seen in the bladder.

Courtesy of D. Mulvihill

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Question: 193

You are seeing a term 4,500-g large-for-gestational age (LGA) infant in the nursery at 2 hours of age. His delivery was complicated by a difficult vaginal extraction with forceps assistance, and he had a shoulder dystocia. Physical examination reveals a large infant who is well perfused and in no respiratory distress. There is no crepitus along the clavicles. You elicit an asymmetric Moro reflex with inability to raise the right arm at the shoulder. The infant holds his right arm in adduction and internal rotation, with pronation of the forearm (Item Q193). His left hand displays a normal grasp.

Of the following, a TRUE statement about this infant's condition and prognosis is that

A. brachial plexus injury with palsies affecting the lower arm and hand have a poorer prognosis than those with isolated upper arm palsy

B. brachial plexus injury with palsies affecting the lower arm and hand is due most commonly to in utero nerve damage acquired in infants of diabetic mothers and will be permanent

C. clavicular fracture often complicates brachial plexus injury in LGA infants and results in a long-term shoulder drop

D. complete avulsion of the brachial plexus is the most common injury and requires microsurgical repair by 3 months of age

E. Erb palsy is the least common form of brachial plexus injury, involves the lower arm and hand, and requires several months to heal

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Question: 193



Courtesy of B. Carter

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Critique: 193 Preferred Response: A

Injury to the brachial plexus, which occurs in approximately 1 per 1,000 births, often, but not always, is associated with shoulder dystocia and is seen most commonly in infants weighing greater than 4,000 g. The medical literature notes some brachial plexus palsies (BPP) following cesarean delivery.

The mechanism of injury typically is either a stretching of the cervical nerve roots that occurs with traction on the neck during delivery or nerve avulsion. Difficult deliveries that involve a prolonged second stage associated with shoulder dystocia, breech position, or operative assistance (vacuum, forceps) also present a risk. When the upper arm is affected (Erb-Duchenne palsy), damage has occurred to the 5th and 6th cervical nerve roots. This is the most common form of BPP. Damage to the 7th and 8th cervical and 1st thoracic nerves occurs less often and is referred to as Klumpke palsy, affecting only the lower arm. Klumpke palsy, which affects the lower arm and hand, has a poorer prognosis than Erb palsy. If all the brachial plexus nerve roots are damaged, the entire arm is paralyzed, and the prognosis for recovery is poor.

In 75% to 90% of cases, the mechanism of injury is stretching, rather than avulsion, of the nerve root, and recovery of function may be anticipated over the first several weeks of postnatal life. Permanent injury occurs in approximately 1 in 10,000 births.

The diagnosis of brachial plexus injury is made clinically and based on the presence of unilateral arm weakness. The Moro reflex is asymmetric, with the infant unable to raise the arm at the shoulder on the affected side. In Erb-Duchenne palsy, the affected arm is limp and held in internal rotation with flexion of the hand at the metacarpal phalangeal joints (“waiter’s tip” position) (Item C193). In an effort to differentiate between Erb-Duchenne palsy and total arm paralysis, examination of hand and finger extension and flexion is important. Finger extension, elicited by stroking the back of the hand, and finger flexion as part of the grasp reflex suggest Erb-Duchenne palsy rather than total arm paralysis.

The nerve roots contributing to the phrenic nerve also can be injured in BPP, and diaphragmatic function may be impaired. This is typically evidenced by tachypnea and an oxygen requirement or may be suggested when there is asymmetric chest motion during respiration and decreased breath sounds on the affected side. In such cases, the diagnosis is supported by a chest radiograph demonstrating an elevated hemidiaphragm and confirmed by ultrasonographic assessment revealing a lack of, or paradoxic, diaphragmatic movement. Clavicular fracture, which may occur in up to 0.5% of all livebirths, is associated with BPP in approximately 1 in 10 cases. Rarely, fractures of the humerus or shoulder dislocation can occur with BPP; shoulder drop is associated with damage to the spinal accessory nerve and trapezius muscle. Finally, the sympathetic nervous system outflow via nerve root T1 can be damaged, resulting in Horner syndrome, which presents as miosis, ptosis, and enophthalmos on the affected side.

Most patients who have BPP require only brief physical or occupational therapy once initial nerve swelling has subsided (typically after the first 7 to 14 days). Such therapies generally are not needed after 3 months of age. For the minority of children who do not demonstrate satisfactory improvement, surgical repair is required, but the optimal timing of surgery is debated.

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Critique: 193



In Erb palsy, the Moro reflex is absent on the affected (right) side. Additionally, there is adduction and internal rotation of the arm and pronation of the forearm.

Courtesy of B. Carter

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Question: 194

An African-American mother brings her 4-week-old daughter to the emergency department because of progressive shortness of breath and pallor. She had mild anemia and jaundice shortly after birth, which was believed to be due to ABO incompatibility (mother is type O-, infant is type A-). She required phototherapy for 1 day and was discharged without further complications. Her respiratory rate today is 65 breaths/min and heart rate is 170 beats/min. She appears pale and mildly icteric and has mild-to-moderate respiratory distress. Complete blood count reveals a hemoglobin of 5 mg/dL (50 g/L) and a mean cell volume of 95 fL.

Of the following, the MOST likely cause of this child's severe anemia is

A. ABO incompatibility

B. Diamond-Blackfan anemia

C. hemoglobin SS disease

D. iron deficiency

E. physiologic anemia of infancy

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Critique: 194 Preferred Response: A

The infant described in the vignette has anemia and jaundice, which indicates some type of hemolytic disease. Hemolytic disease of the newborn usually is caused by immune-mediated Rh and ABO incompatibility. Although ABO incompatibility can occur in the first pregnancy, Rh incompatibility usually does not occur until an Rh-negative mother who is carrying a Rh-positive (D antigen-positive) fetus becomes sensitized. Sensitization of the mother to the D antigen in the fetal blood may occur due to fetomaternal hemorrhage or obstetric procedures during the pregnancy. After sensitization, in subsequent pregnancies, maternal anti-D immunoglobulin G (IgG) crosses the placenta, causing significant fetal hemolysis if the infant is Rh-positive. Such hemolysis can result in only mild hyperbilirubinemia without anemia, but in many cases, hydrops fetalis or severe anemia and hyperbilirubinemia develop in neonates. The hemolysis and hyperbilirubinemia can persist for weeks due to prolonged maternal IgG half-life.

A similar mechanism is seen in ABO incompatibility, but clinical symptoms typically are less severe. ABO incompatibility usually occurs when type O mothers give birth to infants who have type A or B blood, such as the mother and infant in the vignette. Maternal anti-A or anti-B IgM and IgG develop in response to the A or B antigens on fetal red blood cells, and IgG crosses the placenta, causing hemolysis. This hemolysis typically is mild and resolves in a few days without treatment, although it can persist for weeks, as in the infant in the vignette. Treatment consists of supportive care with phototherapy; severe cases may necessitate exchange or simple red blood cell transfusion.

Diamond-Blackfan anemia is a pure red blood cell aplasia. Because hemolysis does not occur, icterus is not a feature. Affected infants typically present in early infancy with severe anemia, and dysmorphic features, including triphalangeal thumbs and facial defects, may be seen.

Hemoglobin SS disease is unlikely for the infant in the vignette because fetal hemoglobin concentrations are still high at 1 month of age, which prevents sickling with subsequent hemolysis and anemia. In addition, most infants are screened for this disease at birth.

Iron-deficiency anemia is rare at this age, and because it is caused by decreased production of red blood cells, jaundice is not a typical feature.

Physiologic anemia of infancy occurs at 6 weeks to 3 months of age due to decreased hematopoiesis, but hemoglobin usually does not fall below 9 mg/dL (90 g/L) in term infants, and jaundice is not present.

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Question: 195

A 4-year-old boy is brought in for evaluation by his mother. He has had “intoeing” all of his life, and his grandmother took him to a doctor who prescribed expensive corrective shoes, but he has not improved. Physical examination reveals no metatarsus adductus or tibial torsion. You note that he likes to sit on his knees with his legs behind him in a “W” shape.

Of the following, you are MOST likely to find on completely unclothed examination

A. bowed legs

B. equinovarus deformity

C. femoral anteversion

D. pain over the tibial tuberosity

E. waddling gait

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Critique: 195 Preferred Response: C

Children who present with “intoeing” usually have one of three problems. In early infancy, the condition may result from metatarsus adductus (Item C195A); in toddlerhood, tibial torsion may be evident; and in early childhood, femoral anteversion is common.

The history and physical findings for the child described in the vignette are consistent with a diagnosis of femoral anteversion (internal femoral torsion). Most children who have femoral anteversion are between 3 and 5 years of age. To assess for femoral anteversion, the child is placed in the prone position and the femurs rotated internally and externally with the knees flexed (Item C195B). Normally, children have 45 degrees of internal and 45 degrees of external rotation; in those who have femoral anteversion, internal rotation is increased (with a reduction in external rotation). Of note, all infants have physiologic femoral anteversion. Children who have femoral anteversion often sit in the “W” position with the knees flexed and legs behind them (Item C195C). Sitting in this position may aggravate the femoral anteversion, and some orthopedic surgeons discourage it. The natural course of femoral anteversion is spontaneous regression between the ages of 8 and 10 years.

Osgood-Schlatter disease (Item C195D) occurs in adolescents, produces pain over the tibial tuberosities, and is not associated with intoeing. Limp may result if the patient is experiencing pain. Bowed legs, which may be physiologic or pathologic (eg, due to Blount disease (Item C195E)), do not necessarily cause intoeing. Equinovarus deformity (Item C195F) generally is identified in infancy, presents with a limp, and typically is unilateral. Waddling gait generally results from hip instability or proximal limb girdle muscle weakness; limp is not a common association.

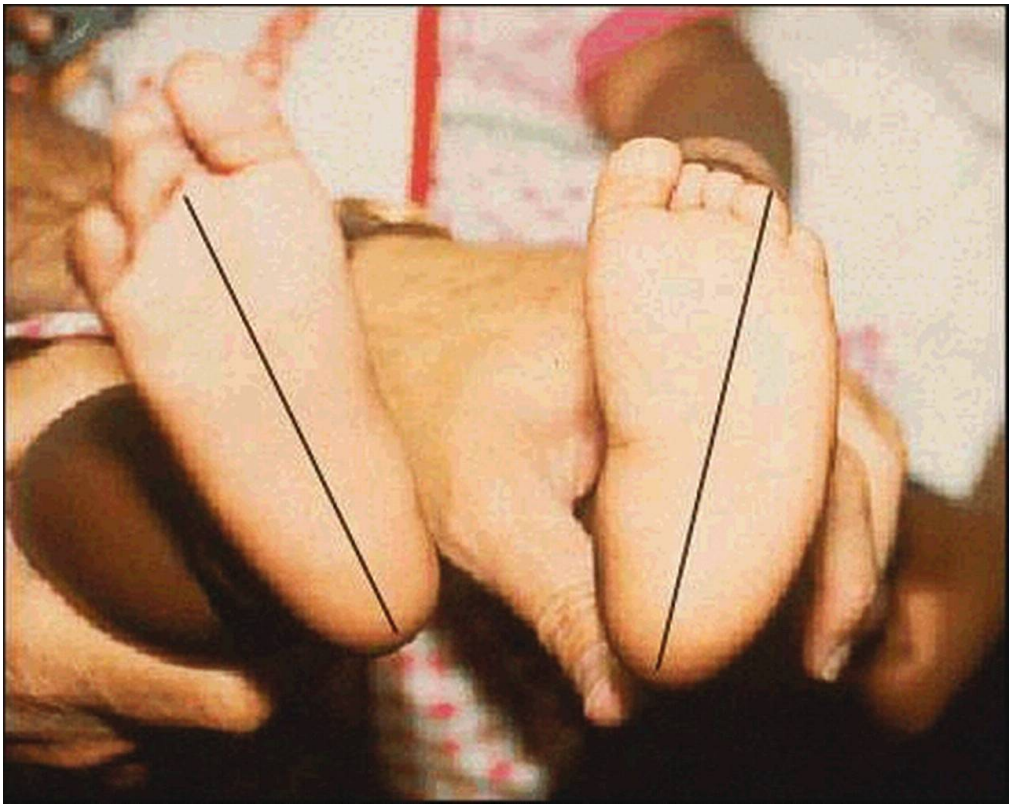
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Critique: 195

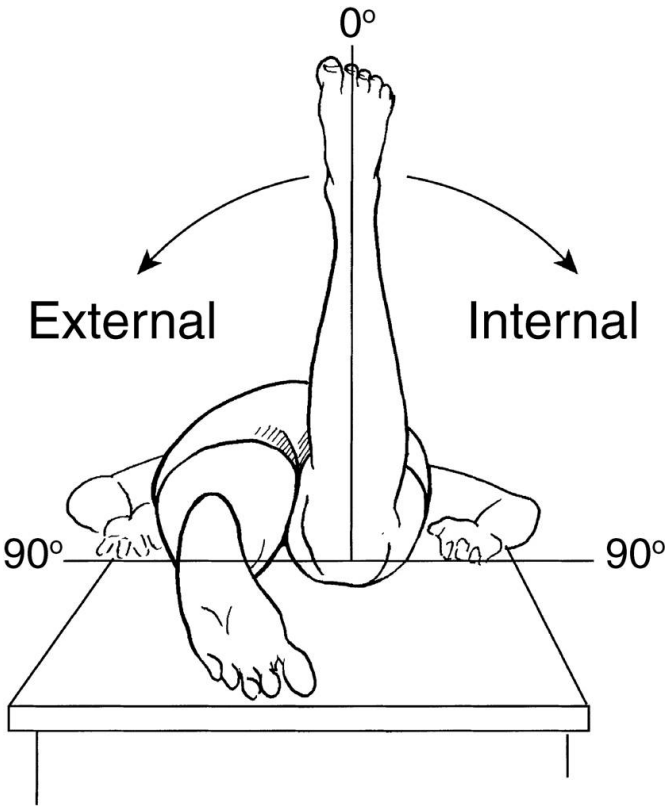


Metatarsus adductus represents an incurving of the forefoot. Normally, a line bisecting the heel passes between the second and third toes (left). In metatarsus adductus, the line bisecting the heel passes more laterally (right).

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Critique: 195



To measure hip rotation, the patient is placed in the prone position with the knee flexed. The hip then is rotated internally and externally.

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Critique: 195



The "W" sitting position.

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Critique: 195



Osgood-Schlatter disease is characterized by swelling and tenderness to palpation of the tibial tuberosity.

Courtesy of D. Krowchuk

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Critique: 195



In Blount disease, there is fragmentation and downsloping of the medial metaphyses of the proximal tibiae.

Courtesy of E. Anthony

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Critique: 195



Bilateral talipes equinovarus deformity in an infant who has arthrogryposis.

Courtesy of M. Rimsza

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Question: 196

You are conducting a preparticipation evaluation of a 14-year-old girl who is trying out for her school volleyball program. She is athletic and has never had any health problems. On physical examination, her height is at the 90th percentile and weight is at the 50th percentile for her age. Her lungs are clear, and cardiac examination reveals a systolic click and an apical systolic murmur that is late systolic and graded at 2/6 with radiation to the left axilla (Item Q196).

Of the following, the MOST likely diagnosis is

A. anemia with high-output state

B. aortic valve stenosis

C. atrial septal defect

D. mitral valve prolapse

E. small midmuscular ventricular septal defect

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Critique: 196 Preferred Response: D

Normally, the mitral valve closes very early in systole as pressure in the ventricle increases with the onset of contraction. The anterior and posterior leaflet of the valve come together to provide complete and straight apposition, thereby protecting the left atrium from the pressure and content of the left ventricle. In mitral valve prolapse (MVP), the mitral valve moves backwards (prolapses) into the left atrium during systole. The anterior or posterior leaflet does not achieve straight apposition; rather, one or both of the leaflets billows into the space of the left atrium as pressure in the left ventricle increases. If there is retrograde leakage of left ventricular blood into the left atrium, the MVP is associated with mitral regurgitation. The prolapse may result from either abnormality of one or both of the mitral valve leaflets (eg, redundancy, "floppy", myxomatous) or of the supporting apparatus (chordae tendineae, papillary muscles). MVP may occur primarily as an intrinsic abnormality of the mitral valve or its apparatus or it can occur secondarily from acquired disease such as rheumatic heart disease, myocarditis, or cardiomyopathy. Primary MVP is more common in females than males (2:1), and the disorder may be diagnosed in any age group.

The diagnosis of MVP is based on physical findings; patients can present with auscultatory findings that include a mid-systolic click (Item C196A) that is believed to occur from the snapping of the mitral valve in the closed position during ventricular systole similarly to a sail catching wind. This timing of the click during systole may change, depending on patient position and the relative volume of the left ventricle during the examination. Squatting fills the left ventricle and may make the click occur later; standing reduces the left ventricular volume and may move the click to an earlier portion of systole. If there is regurgitation across the mitral valve, a late systolic murmur (Item C196B) may be audible at the cardiac apex, with radiation to the left axilla.

The patient described in the vignette has physical findings indicative of MVP. The murmur associated with anemia typically is ejection, located along the left sternal boarder with radiation to the base, and not associated with a click. Aortic stenosis is associated with an ejection click that does not change with position, and the accompanying murmur (Item C196C) is best heard at the upper right sternal boarder with radiation into the neck. An atrial septal defect typically creates a murmur of relative pulmonary stenosis (Item C196D) as the left-to-right atrial shunt leads to increased right ventricular volume that subsequently must cross the pulmonary valve. The murmur is heard best at the upper left sternal border. Because there is no structural abnormality of the valve, no click is appreciated in patients who have atrial septal defects. The murmur of a small muscular ventricular septal defect (Item C196E) is high-pitched, heard along the sternal border, and not associated with a systolic click.

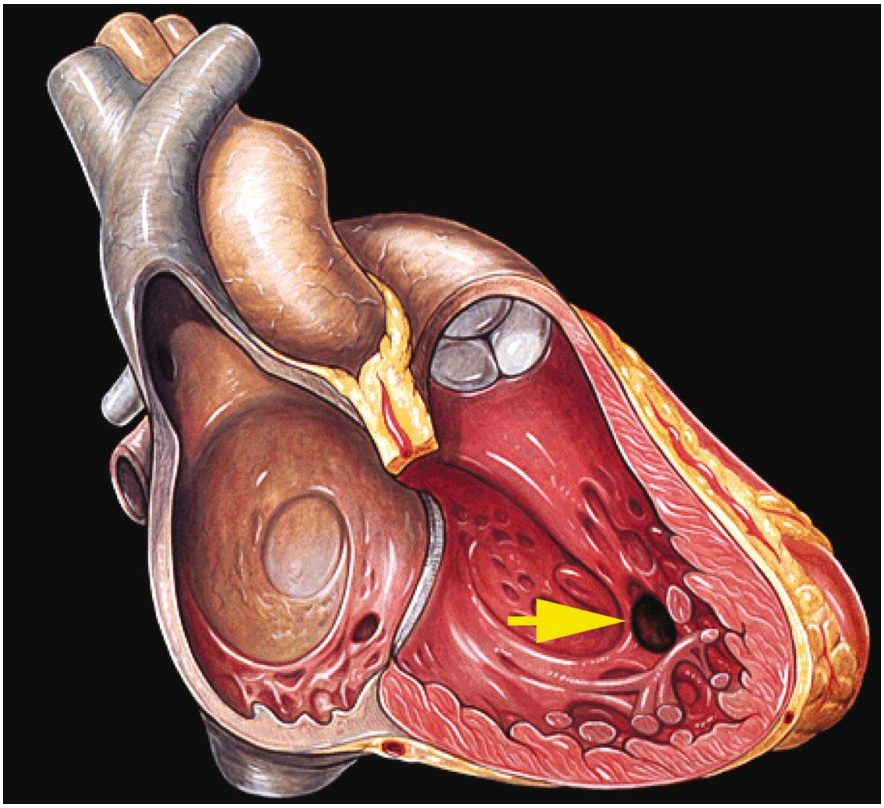
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Critique: 196



When the septum is viewed from the right side of the heart, a small ventricular septal defect is apparent in the muscular, trabeculized portion of the septum near the apex.

Courtesy of P. Lynch

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Question: 197

At 8 am, your nurse urgently calls you to see a child in the waiting room. You come out and observe a 5-year-old boy whose eyes are glassy and staring off to the right. He is making chewing movements and has urinated. He is not responding to his mother's calls or touch. He then blinks several times and begins to respond, but is clearly confused. His mother explains that her son has been diagnosed with epilepsy, but she ran out of medication 2 days ago.

Of the following, the MOST appropriate maintenance antiseizure medication for this child is

A. carbamazepine

B. diazepam

C. ethosuximide

D. phenobarbital

E. phenytoin

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Critique: 197 Preferred Response: A

The child described in the vignette has had a classic complex partial seizure. The seizure onset is likely in a nonmotor area, such as a temporal lobe, which caused the staring and nonresponsiveness. The chewing movements are automatisms. The confusion is typical for a postictal period. The cause of the seizure is the child's epilepsy and probably the parent's medical mismanagement. The reason for "running out" of medication needs to be explored with this parent so that she understands that abruptly discontinuing seizure medication places her child at risk of harm.

Selection of medications for the treatment of epilepsy depends on the type of epilepsy. The most basic distinction is whether the epilepsy is partial or generalized. Clinical data from a careful history are critical. For example, if the seizure begins with an aura (a smell, a particular sensation), a stare, or focal motor movements, it is likely to be a partial seizure. Partial seizure also may be characterized by asymmetric findings on the neurologic examination or the presence of focal weakness after the seizure. If a staring seizure is followed by immediate return to normal awareness, focal onset is much less likely and the child may have generalized onset, primary absence epilepsy. If the clinical history is unavailable or inconclusive as to the seizure type, but the clinician judges prior events to be highly likely to be seizures, electroencephalography may be helpful in defining epilepsy type and choosing appropriate medication.

Carbamazepine is effective for partial seizures, as experienced by the child in the vignette, and has a superior safety profile to phenobarbital or phenytoin (Item C197). Diazepam is used primarily for cluster or prolonged breakthrough seizures. Ethosuximide is the drug of choice for primary absence epilepsy.

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Critique: 197

Item 197: Use of Anticonvulsants for Epilepsy

Medication	Clinical Use	Comments
Carbamazepine	Partial epilepsy	May precipitate more seizures in generalized epilepsy, otherwise generally well tolerated
Diazepam	Status epilepticus, cluster seizures	For epilepsy, primarily used as a rectal preparation for cluster and prolonged breakthrough seizures
Ethosuximide	Primary absence epilepsy	Drug of choice for this epilepsy syndrome
Phenobarbital	Neonatal seizures, status epilepticus	Rarely used for maintenance treatment past the neonatal period due to cognitive and behavioral adverse effects
Phenytoin	Partial or generalized epilepsy, status epilepticus	Used intravenously as phenytoin or fosphenytoin for status epilepticus, but seldom used as a first choice for maintenance treatment in children due to erratic pharmacokinetics and adverse effects such as gingival hyperplasia
Valproic acid	Partial or generalized epilepsies	Broad-spectrum antiseizure medication, risk of hepatic failure in children younger than 2 years old

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Question: 198

A 13-year-old boy comes to your office for a preparticipation sports physical examination. He is interested in starting power lifting of heavy weights to prepare himself to play football in a community recreational league. On his physical examination, you find he is at Sexual Maturity Rating 2.

Of the following, you are MOST likely to explain to the boy that

A. adolescents should avoid power lifting and body building until they reach physical and skeletal maturity

B. resistance training should be used alone for the best general health benefits

C. strength training programs for adolescents should not include warm-up and cool-down periods

D. strength training programs have been proven to prevent sports-related musculoskeletal injuries in adolescents

E. strength training will reduce the incidence of catastrophic sports-related injuries

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Critique: 198 Preferred Response: A

Pediatricians very often are involved in the preparticipation sports physical examinations for young people. Strength training, often known as resistance training, is a common component of sports and physical fitness programs for youth, both as a means to improve sports performance and to enhance appearance. Such training programs may include the use of free weights, weight machines, elastic tubing, or body weight. Power lifting involves the use of maximum lifting and can be a competitive sport.

The American Academy of Pediatrics Committee on Sports Medicine and Fitness notes that strength training for preadolescents and adolescents can be safe and effective, but proper training techniques and safety precautions must be followed to avoid injury to an immature skeleton. A weight training program can be started with middle school-aged children (10 to 12 years old) but should be well supervised, use small free weights with high repetitions (15 to 20), have proper technique demonstrated, and avoid shorter sets of lifts that use heavier weights and maximum lifts. As an individual ages, weight training may continue, and as physical maturity (Sexual Maturity Rating 5) is reached, a program involving longer sets of lifts, with heavier weights and fewer repetitions (including power lifting), may be undertaken safely, while continuing to stress the use of proper technique.

Resistance training is best coupled with aerobic conditioning for optimal general health benefits. Strength training should include warm-up and cool-down periods. Although strength training may be beneficial to adolescent athletes' performance in weight lifting, there is no evidence that such training will prevent sports-related musculoskeletal injuries or decrease the incidence of catastrophic sports-related injuries.

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Question: 199

You are reviewing the growth chart (Item Q199) of a 10-year-old boy who has had type 1 diabetes for 4 years and whose hemoglobin A1c is 8.1% (normal, 3.8% to 6.4%) during his annual health supervision visit. His mother tells you that he has been eating poorly, and she thinks he may have lost some weight in the past 6 months. On physical examination, he appears well but somewhat tired, he has no abdominal masses, and his liver is palpable 1 cm below the right costal margin.

Of the following, the MOST likely new diagnosis is

A. adrenal insufficiency

B. anorexia nervosa

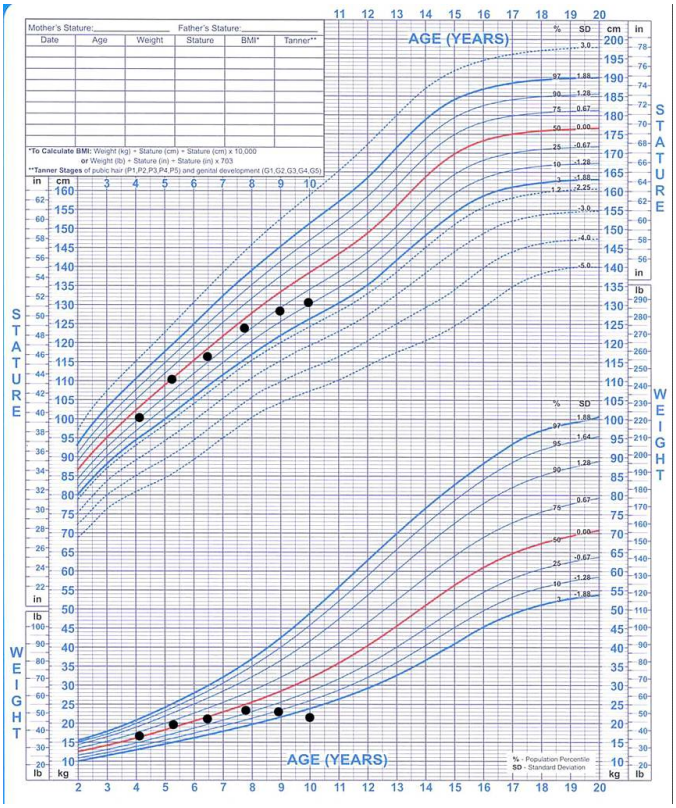
C. celiac disease

D. hypothyroidism

E. poor growth because of poor glycemic control

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Question: 199



Courtesy of L. Levitsky

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Critique: 199 Preferred Response: C

Patients who have type 1 diabetes are at increased risk for the development of various autoimmune disorders. Celiac disease occurs in 5% to 6% of all children who have type 1 diabetes and is more common in those who develop type 1 diabetes before the age of 10 years. The weight loss and poor growth for the boy described in the vignette, who does not have perfect glycemic control but has an adequate hemoglobin A1c value for age, suggests the diagnostic possibility of celiac disease. Adrenal insufficiency because of autoimmune adrenal failure is another possibility, but it is much less common than celiac disease (<1% of children who have diabetes).

Anorexia nervosa is unusual in boys and very unusual in children younger than 10 years of age. Hypothyroidism due to chronic lymphocytic thyroiditis may be found in 5% to 10% of individuals who have type 1 diabetes, but usually occurs after the first decade and typically is associated with weight gain, not weight loss. Poor growth because of poor glycemic control, which in its most flagrant form is referred to as Mauriac syndrome, is associated with hepatomegaly and very high blood glucose concentrations. The hemoglobin A1c value reported for the boy in the vignette is not in the 12% to 14% range associated with Mauriac syndrome.

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Question: 200

An 8-month-old infant often falls asleep while his mother is feeding him. He tends to sleep longest during the day and wakes frequently during the night. The parents are sleep-deprived and ask for your assistance in getting the infant to sleep more during the night.

Of the following, your BEST suggestion is to

A. instruct the parents to feed the infant promptly when he awakes at night

B. instruct the parents to keep the child awake more during the day

C. prescribe diphenhydramine for the infant at night to help him sleep

D. reassure the parents that this is a phase that will pass and recommend a follow-up evaluation in 2 months

E. recommend that the infant sleep with the parents to minimize nighttime disturbances

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Critique: 200 Preferred Response: B

At birth, infants typically sleep about 18 hours per day, with sleep split evenly between the day and night. By approximately 6 to 15 months of age, most children sleep about 10 to 12 hours at night and take two daytime naps. Spontaneous awakenings are normal and occur often during periods of rapid eye movement sleep. The ability of infants to settle (return themselves to sleep) usually develops around 3 to 4 months of age. Nighttime feedings and prolonged attention at night may prolong nighttime awakenings. To encourage night settling, day sleeping should be limited to 3 to 4 consecutive hours. Infants should be placed in their own cribs to fall asleep and be allowed to calm themselves. Nighttime feedings should be short, with little stimulation.

The infant described in the vignette needs to be stimulated during the day to encourage him to stay awake for longer periods of time. Telling the parents that this is just a phase will not help the baby learn to sleep for longer periods at night. The parents should be instructed to decrease caloric intake during the night and not to feed the infant immediately when he wakes at night. Giving diphenhydramine at night will not aid the infant in developing an inherent circadian rhythm, and its safety has not been established in infants. Suggesting that an infant sleep with the parents may increase the risk for infant suffocation or strangulation. However, for parents who choose to share a bed with their child, certain guidelines should be followed to minimize risk: use a firm mattress, never use alcohol or drugs, never use cigarettes (babies of mothers who smoke have a higher risk of dying from sudden infant death syndrome), do not place bed against the wall, and do not use heavy and bulky blankets.

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Question: 201

As you are examining a 2-year-old boy who is new to your practice, you note a swollen area on the right side of his neck. You ask the mother about the swelling, and she starts to cry. She states that this area has been swollen for 2 months despite courses of cephalexin, clindamycin, and trimethoprim-sulfamethoxazole. Upon further questioning, you learn that the family recently moved to “town” after living in a rural area for the past 10 years. She denies a history of animal or tick exposure. Because her husband is highly allergic to cats, they won’t even let their son pet one. The boy is afebrile, and other than a 3 x 3 cm swelling in the submandibular region of the right neck, results of the physical examination are normal. The swelling moves freely but is slightly tender to palpation.

Of the following, the organism MOST likely to cause the swelling is

A. *Bartonella henselae*

B. *Francisella tularensis*

C. *Mycobacterium avium-intracellulare*

D. *Staphylococcus aureus*

E. *Streptococcus pyogenes*

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Critique: 201 Preferred Response: C

Chronic cervical lymphadenopathy is defined as nodal swelling that has lasted for 3 weeks or longer. Most parents are worried about a malignancy, but the cause usually is infectious for patients who have a single, freely movable lymph node and no other findings on physical examination (eg, hepatosplenomegaly). Infectious agents that commonly cause chronic lymph node swelling include *Bartonella henselae*, *Francisella tularensis*, *Mycobacterium avium-intracellulare*, and *Mycobacterium tuberculosis* (MTB). *Staphylococcus aureus* and *Streptococcus pyogenes* are common agents causing acute adenitis, but they rarely cause chronic problems. The boy described in the vignette has no cat, tick, or rabbit exposure, so *B henselae* and *F tularensis* are much less likely. Therefore, the chronic lymphadenopathy most likely is due to an infection from *M avium-intracellulare*.

Nontuberculous mycobacteria (NTM) can cause disease in healthy children. The most common manifestation is chronic lymphadenopathy. Many people equate *M avium-intracellulare* to an opportunistic infection for patients infected with the human immunodeficiency virus (HIV), but it was known to cause chronic lymphadenitis in healthy children prior to the emergence of HIV. The many other species of NTM (Item C201) are not encountered commonly among children.

The NTM are found in soil, food, water, and animals. Definitive diagnosis requires isolation of the organism. NTM are relatively resistant to many medications, including standard antituberculous agents. Therapy, therefore, is based on the following: 1) the species of *Mycobacterium* recovered, 2) drug susceptibility testing results, 3) site of the infection, 4) the patient's underlying medical condition, and 5) the need to treat for MTB while awaiting culture results.

For patients such as the boy described in the vignette, complete surgical excision almost always is curative, but many experts suggest a course of clarithromycin or azithromycin in combination with ethambutol or rifabutin prior to surgery.

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Critique: 201

Item 201: Species of Nontuberculous *Mycobacterium*

Clinical Infection	Common *Mycobacterium* Species
Skin	*Mycobacterium chelonae*, *M fortuitum*, *M abscessus*, *M marinum*
Otologic	*M abscessus*
Pulmonary (mostly patients who have cystic fibrosis)	*M avium-intracellulare*, *M kansasii*, *M abscessus*
Catheter-related	*M chelonae*, *M fortuitum*
Skeletal	*M avium-intracellulare*, *M kansasii*, *M fortuitum*
Disseminated (immunocompromised patients)	*M avium-intracellulare*

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Question: 202

A 26-week-old preterm infant has developed line-associated *Staphylococcus epidermidis* sepsis.

Of the following, the MOST effective antibiotic regimen for this infant is

A. ampicillin

B. ampicillin + gentamicin

C. ceftriaxone

D. oxacillin + gentamicin

E. vancomycin + rifampin

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Critique: 202 Preferred Response: E

Staphylococci are catalase-positive organisms that are grown easily on blood agar and are differentiated by their ability to produce the enzyme coagulase, which congeals rabbit plasma. Fifteen different species of coagulase-negative staphylococci are indigenous to humans and of these, *Staphylococcus epidermidis* and *S saprophyticus* have been identified as pathogens in humans. Coagulase-negative staphylococci are natural inhabitants of human skin, with *S epidermidis* being the most prevalent and persistent.

Coagulase-negative staphylococci are frequent causative agents of nosocomial infections, primarily in immunocompromised patients who have indwelling medical devices that are inserted through the skin. They are also causes of native and prosthetic valve endocarditis, nosocomial bacteremia (especially in neonatal intensive care units), cerebrospinal fluid shunt infections, peritoneal dialysis catheter-associated peritonitis, urinary tract infections, infections of prosthetic joints, osteomyelitis, endophthalmitis, and infections of vascular grafts.

Coagulase-negative staphylococci typically are resistant to multiple antibiotics; more than 80% are resistant to methicillin and other beta-lactam agents. More than 50% are resistant to erythromycin, clindamycin, chloramphenicol, and tetracycline, and a growing proportion have developed resistance to gentamicin and trimethoprim. Antibiotics to which most coagulase-negative staphylococci are susceptible include vancomycin, rifampin, and ciprofloxacin.

The antibiotic regimen to which the pathogen identified in the vignette is susceptible is vancomycin and rifampin. *S epidermidis* has significant resistance to ampicillin, ceftriaxone, and oxacillin.

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Question: 203

Voiding cystourethrography in a 9-month-old boy who has new-onset febrile urinary tract infection reveals grade II vesicoureteral reflux (VUR). The parents ask you about their son's prognosis.

Of the following, you are MOST likely to explain that

A. approximately 80% of children who have newly diagnosed febrile urinary tract infections have VUR when tested

B. once VUR is established, no follow-up radiologic testing is indicated

C. males have a worse prognosis than females

D. referral to urology for ureteral reimplantation is warranted

E. unilateral grade II reflux has a high likelihood of resolution within 5 years of the diagnosis

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Critique: 203 Preferred Response: E

A child who has a febrile urinary tract infection (UTI) has a 30% to 50% likelihood of having underlying vesicoureteral reflux (VUR). VUR is the reflux of urine from the bladder into the ureter and possibly kidney across the ureterovesical junction (UVJ). It may be caused by anatomic abnormalities of the UVJ or bladder (eg neurogenic bladder) or bladder outlet dysfunction (eg posterior urethral valves). VUR is estimated to occur 10 times more frequently in Caucasians than in African-Americans. Males and females are nearly equally affected, and their prognosis is similar.

There appears to be a strong familial association for VUR, with approximately 30% of siblings of an index case also having VUR when studied by voiding cystourethrography (VCUG). Despite this association, screening of asymptomatic siblings of affected children is controversial because VCUG is an invasive procedure and the benefit of identifying and treating (with prophylactic antibiotics) a child who is well and lacks symptoms is uncertain. At present, there is no consensus of opinion, although the trend is not to study asymptomatic older siblings who are toilet trained; some recommend that asymptomatic siblings younger than 1 year of age undergo VCUG.

The American Academy of Pediatrics recommends performing ultrasonography and VCUG in all children after their first febrile UTI. The present standard of care for patients who have VUR is to receive prophylactic antibiotics until the reflux has resolved. Patients typically undergo a follow-up VCUG every 12 to 18 months; the time between VCUG studies is somewhat dependent on the age of the patient and the severity of reflux.

An international classification system for VUR has grades ranging from mild (grade I) to severe (grade V). A nonsurgical approach is recommended for children who have grades I to III reflux; spontaneous resolution occurred in 80% of cases within 5 years of the diagnosis in one study. Grade IV reflux also often is managed nonsurgically. Grade V reflux traditionally has been managed surgically. A newer technique that involves endoscopic subureteral injection of dextranomer/hyaluronic acid may offer an alternative to conventional ureteral reimplantation surgery. Long-term data for this technique are not yet available.

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Question: 204

An 8-year-old girl is admitted to the intensive care unit for a severe asthma exacerbation. As part of her management, she is placed on continuous albuterol nebulization at 15 mg/h.

Of the following, the MOST likely electrolyte abnormality to expect in this girl is

A. hypercalcemia

B. hypermagnesemia

C. hypernatremia

D. hypoglycemia

E. hypokalemia

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Critique: 204 Preferred Response: E

The most common adverse reactions to beta2-adrenergic agonists are tremor, tachycardia, and palpitations. Administration of therapeutic or multiple doses of a beta2-adrenergic also may cause transient decreases in PaO2, prolonged QTc interval, arrhythmias, and electrolyte abnormalities.

Continuous administration of a beta2-adrenergic agonist, as described in the vignette, can result in hyperglycemia due to glycogenolysis, hypomagnesemia, and hypokalemia. Hypokalemia results from stimulation of the Na+-K+ pump and may be characterized by a decrease in serum potassium concentrations by 0.4 to 0.9 mEq/L (0.4 to 0.9 mmol/L). Patients who have preexisting hypoglycemia or relative hypoglycemia due to diuretic therapy may be at higher risk for arrhythmias during continuous beta2-adrenergic agonist therapy.

Hypoglycemia and hypocalcemia have been reported in isolated cases, but hypermagnesemia, hypercalcemia, hyponatremia, and hypernatremia have not been reported during single or continuous beta2-adrenergic agonist administration.

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Question: 205

A 2-year-old girl has a choking episode in the waiting room of your office while eating peanuts. When you evaluate her in an examination room, she appears well, the coughing has resolved, her respiratory rate is 24 breaths/min, and her oxygen saturation is 97%. Auscultation of her lungs reveals coarse rhonchi at both lung bases with an end-expiratory wheeze in the right mid-lung field. Posteroanterior, lateral, and decubitus chest radiographs appear normal.

Of the following, the next MOST appropriate step in the evaluation of this patient is

A. computed tomography scan of the chest

B. flexible laryngoscopy

C. fluoroscopy of the chest

D. no further evaluation

E. rigid bronchoscopy

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Critique: 205 Preferred Response: E

Aspiration of foreign bodies is responsible for 5% of unintentional deaths in children younger than 4 years of age. Most episodes occur in children younger than 4 years of age, with the peak incidence seen between 1 and 2 years. Infants and toddlers commonly aspirate food, with peanuts accounting for most food-related aspirations. Toy balloons are the objects most commonly involved in fatal childhood foreign body aspiration (FBA).

Most patients who have a FBA present to medical attention with a history of a sudden choking episode that may be followed by the development of respiratory signs or symptoms. Commonly associated physical findings include cough, wheezing, unilaterally decreased breath sounds, tachypnea, or retractions. It is not unusual, however, for patients to be asymptomatic. In several studies, a history of choking had 76% to 95% sensitivity for the diagnosis, while the classic signs and symptoms of wheezing, cough, and diminished breath sounds were found in only 57% of patients in one series after a choking event. In contrast, a patient who has a history of choking and one or more signs or symptoms has a 90% likelihood of having a bronchial foreign body.

Although most aspirated foreign bodies are radiolucent, initial evaluation should be plain chest radiographs. Even if the object cannot be seen, indirect signs of air trapping may be evident, including unilateral hyperinflation, atelectasis, or mediastinal shift. Inspiratory/expiratory (Item C205) or decubitus films or fluoroscopy may be useful in accentuating any evidence of air trapping if the diagnosis is still in question following initial plain films. Normal plain radiographic studies do not rule out the diagnosis because they may appear normal in 27% to 65% of cases. Computed tomography scanning of the chest is no more sensitive than plain films in the acute setting. Flexible laryngoscopy is not useful because a bronchial foreign body cannot be seen.

Whenever the diagnosis of FBA is certain or the clinical index of suspicion is high, as in this vignette, rigid bronchoscopy should be performed emergently. Rigid bronchoscopy allows the patient to be ventilated throughout the procedure and provides a large aperture through which to extract the object. Flexible bronchoscopy has been used as a diagnostic procedure when the diagnosis is uncertain, but the object typically is removed after diagnosis with a rigid bronchoscope. A clinical index of suspicion when there is a history of choking should prompt evaluation by bronchoscopy because complications of missed bronchial foreign bodies can include lung abscess, recurrent pneumonia, bronchiectasis, and bronchial tree perforation and fistula formation.

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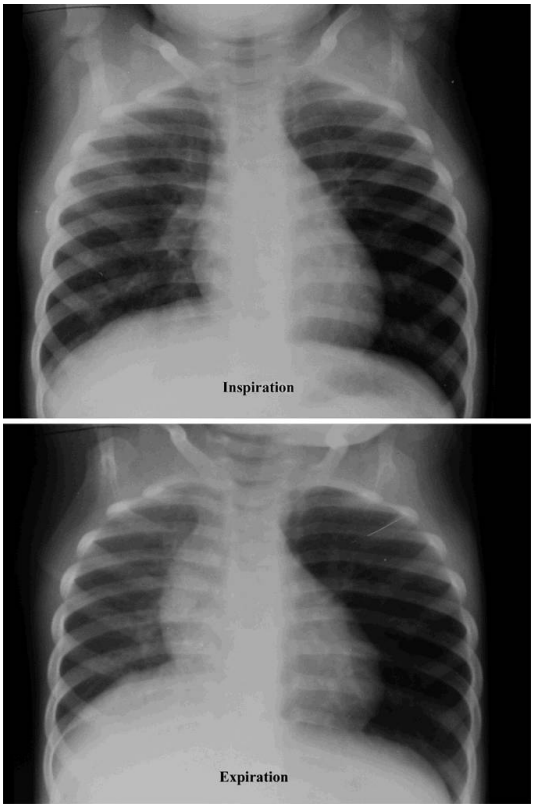
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Critique: 205



Foreign body aspiration: Chest radiograph during inspiration (top) reveals only slightly greater expansion of the left lung compared with the right. During expiration (bottom), the affected left lung remains expanded due to air trapping. The right hemidiaphragm has moved upward, and the heart and mediastinum have shifted to the unaffected right side.

Courtesy of D. Mulvihill

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Question: 206

A 16-year-old boy is concerned about “bumps” on his penis that have been present for several weeks (Item Q206).

Of the following, the MOST appropriate management for these lesions is

A. observation

B. penicillin parenterally

C. permethrin topically

D. podofilox topically

E. surgical excision

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Question: 206



Courtesy of D. Krowchuk

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Critique: 206 Preferred Response: D

The papules exhibited by the patient described in the vignette have a rough, somewhat pointed surface that is characteristic of genital warts (ie, condylomata acuminata) (Item C206A). Genital warts result from infection with human papillomavirus (97% of lesions are caused by types 6 and 11) and are present in approximately 1% of sexually active individuals. Lesions often have a cauliflowerlike appearance, and they may remain small or form large plaques. In men, genital warts usually occur on the foreskin (in those not circumcised) or penile shaft; in women, they may be located on the vulva (Item C206B), in the vagina, or on the cervix. Involvement of the urethral meatus and perianal region occurs in both women and men.

Without intervention, genital warts may resolve spontaneously, remain unchanged, enlarge, or spread to other sites. Nearly all patients request therapy, with the goal being to remove symptomatic lesions. Treatment may reduce infectivity but does not invariably eradicate the virus, and as a result, recurrences are possible. Several therapeutic options exist: patient-applied treatments include podofilox or imiquimod; clinician-administered options are cryotherapy or the application of podophyllin or bi- or trichloroacetic acid. Surgical excision and laser ablation are reserved for genital warts that are recalcitrant to other modalities.

Pearly penile papules occasionally may be mistaken for genital warts, although their characteristic appearance and distribution usually permit differentiation. Lesions are small, skin-colored papules that are distributed symmetrically on the glans, usually the corona (Item C206C). No therapy is necessary. In males, scabies infestation can result in papules or nodules on the penis and scrotum (Item C206D). The observation that the papules are not verrucous and that lesions are present elsewhere helps to distinguish scabies from genital warts. For those who are infested, application of permethrin 5% cream is curative. The lesion of primary syphilis is a chancre, a painless ulcer that has an indurated border (Item C206E), not a papule. Treatment is benzathine penicillin G administered intramuscularly.

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Critique: 206

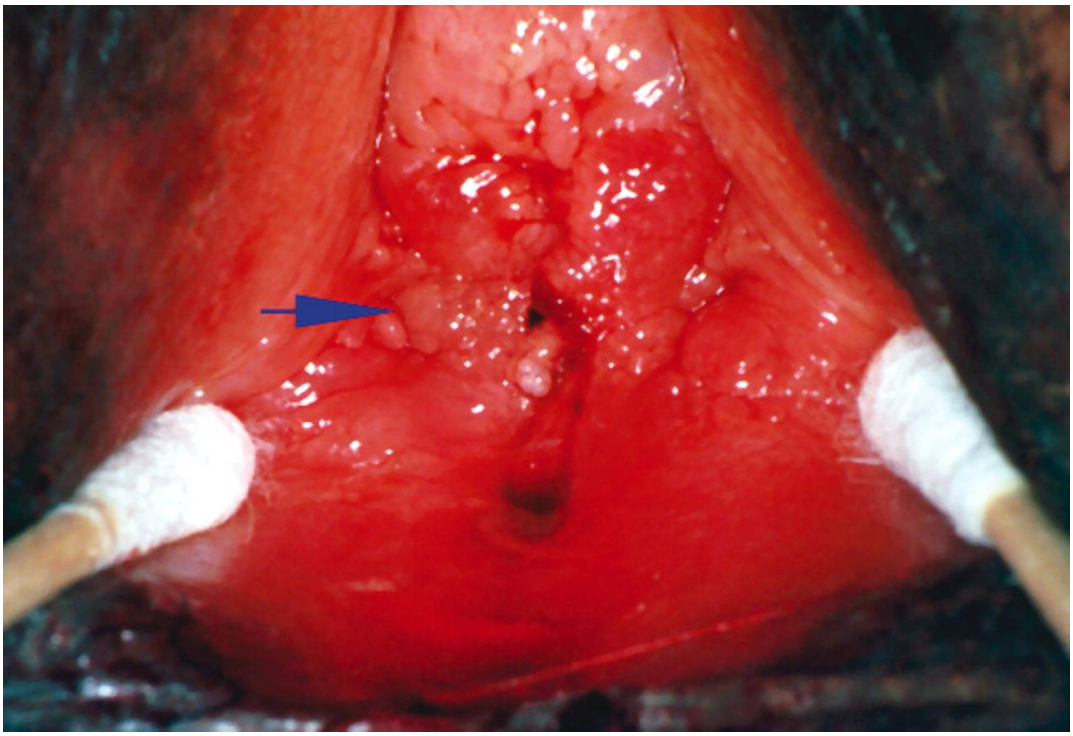


In males, genital warts often are located on the shaft of the penis, appearing as papules that have a pointed surface.

Courtesy of D. Krowchuk

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Critique: 206

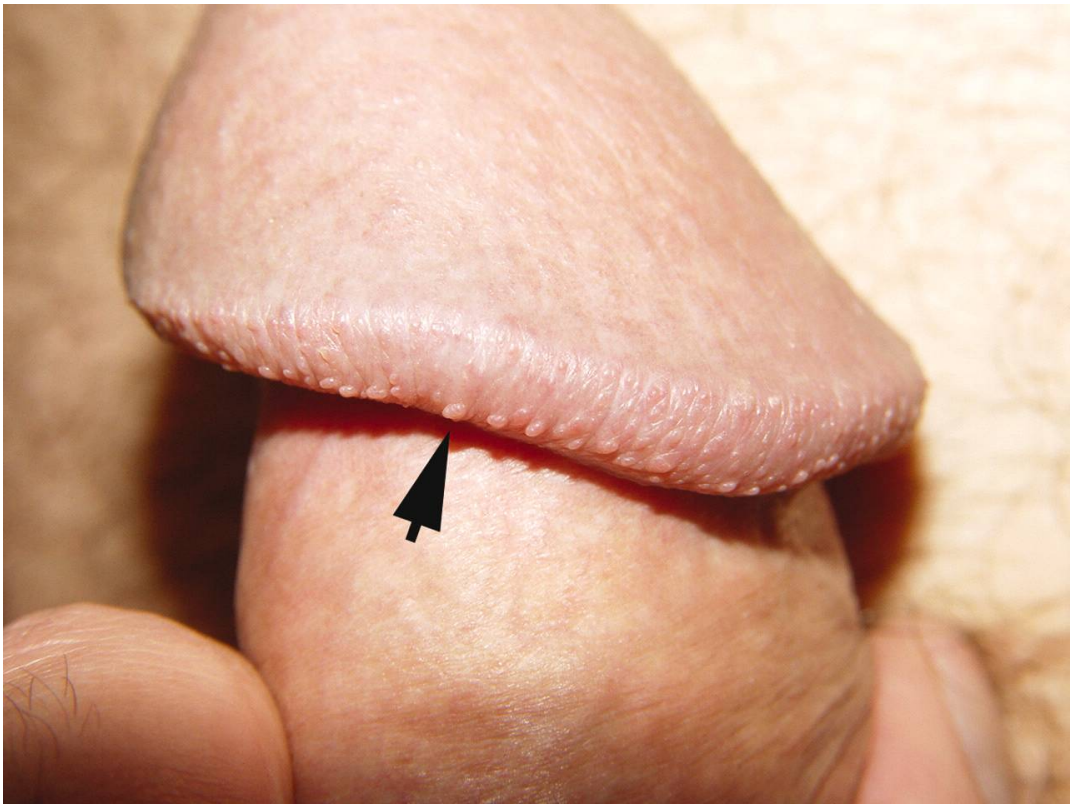


On mucosal surfaces, genital warts may appear as pedunculated growths that have multiple finger-like projections (arrow).

Courtesy of M. Rimsza

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Critique: 206



Pearly pink penile papules (arrow) are skin-colored papules that are symmetrically distributed on the glans, usually the corona.

Courtesy of dermatlas.org

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Critique: 206

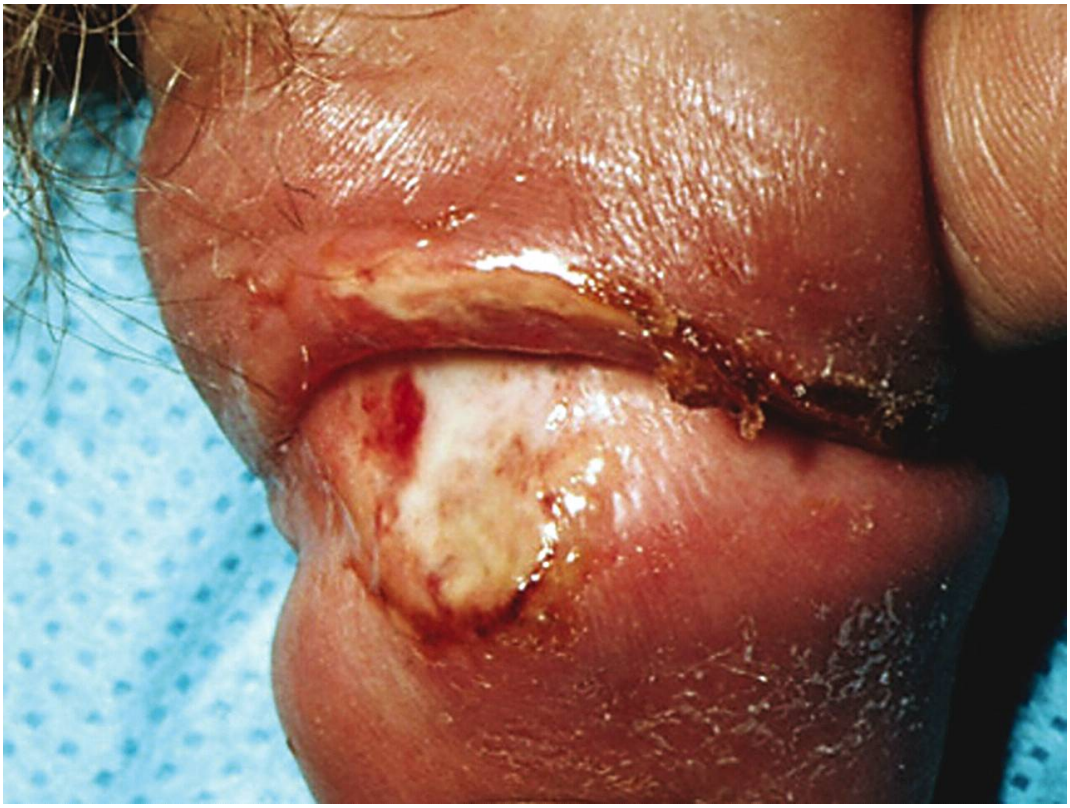


In males, scabies can affect the penis and scrotum.

Courtesy of D. Krowchuk

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Critique: 206



The chancre of primary syphilis is a painless ulcer that has an indurated border.

Courtesy of A.B. Fleischer, Jr

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Question: 207

A 3-month-old infant who was born at term has had persistent cholestasis since birth. At 6 weeks of age, he had acholic stools. At 2 months of age, intraoperative cholangiography demonstrated patent bile ducts. Liver biopsy demonstrated globules in the hepatocytes on a periodic acid-Schiff stain. Currently, his stools are pale yellow, and he is growing at the 10th percentile for both weight and height. His parents are healthy, but his sister was diagnosed with liver cirrhosis at 10 years of age. Physical examination reveals a healthy-appearing infant whose lungs are clear. Cardiac evaluation documents a normal heart rate and rhythm and no murmurs or gallops. The liver is mildly enlarged, with a span of 6 cm (2 cm below the costal margin), and the spleen tip is palpable. The patient's bilirubin is 5.4 mg/dL (92.3 mcmol/L), alanine aminotransferase is 124 U/L, gamma-glutamyltranspeptidase is 300 U/L, prothrombin time is 12 seconds, and partial thromboplastin time is 29 seconds.

Of the following, this patient's underlying condition places him at GREATEST risk for the development of

A. choreoathetosis

B. cryoglobulinemia

C. diabetes mellitus

D. emphysema

E. pancreatitis

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Critique: 207 Preferred Response: D

The patient described in the vignette presents with neonatal cholestasis and has a sister who has early-onset cirrhosis, suggesting a familial liver disease. Biliary atresia has been excluded by cholangiography, and liver biopsy demonstrates globules in the hepatocytes. The infant appears healthy, and aside from a mild elevation in liver enzymes, liver function appears good. This presentation is most consistent with alpha-1-antitrypsin deficiency, placing the child at risk for the premature development of emphysema.

Patients who have cystic fibrosis are at increased risk for the development of diabetes and pancreatitis and may present with neonatal cholestasis, but the absence of lung disease and failure to thrive and the findings on liver biopsy make cystic fibrosis highly unlikely in this infant. Cryoglobulinemia is a complication of hepatitis C in older children and adults. Choreoathetosis is seen in Wilson disease, a condition that does not present in infancy.

Alpha-1-antitrypsin is 394-amino acid protein that is a member of the serine protease inhibitor (serpin) family of proteins that includes antithrombin, antiplasmin, and protein C inhibitor. Most serpins inhibit other proteases in the body. For example, antithrombin prevents excessive clotting by inhibiting thrombin activity, and antitrypsin prevents tissue damage by inhibiting trypsin and elastase. In a manner analogous to hemoglobin, an individual inherits two antitrypsin alleles: one from the mother and one from the father. For hemoglobin, the normal allele pair is AA; sickle cell heterozygotes have AS; and homozygous sickle cell patients have hemoglobin SS. For alpha-1-antitrypsin, the normal pair is MM, heterozygotes have MZ, and recessive homozygotes (alpha-1-antitrypsin-deficient patients) have ZZ. Many different mutations in alpha-1-antitrypsin have been described, but the Z allele is the most common mutation that results in clinical symptoms.

Humans who make the normal alpha-1-antitrypsin (MM) secrete the protein normally into the blood. The protein is distributed via serum throughout the body, where its principal site of action is the lung. Alpha-1-antitrypsin protects the lung from injury by clearing elastase produced by pulmonary leukocytes from lung tissues. In contrast, recessive patients (ZZ type) have a single amino acid mutation in their alpha-1-antitrypsin (substitution of Glu by Lys in position 342). This mutation results in the accumulation of alpha-1-antitrypsin protein in the endoplasmic reticulum of the hepatocytes. Such accumulation may cause liver damage, which may lead to cirrhosis. In addition, low concentrations of antitrypsin in the serum predispose people who have alpha-1-antitrypsin deficiency to emphysema. Only a small proportion (approximately 10% of infants) of those who have alpha-1-antitrypsin deficiency present with cholestasis in infancy. The remainder of patients may have undetected disease in infancy, but can present with pulmonary or liver disease in later life.

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Question: 208

You are preparing to attend the vaginal delivery of an infant at 40 weeks' gestation. Artificial rupture of the fetal membranes 8 hours ago revealed meconium-stained amniotic fluid. The pediatric resident asks what she should plan to do.

Of the following, you are MOST likely to tell her to

A. always ask the delivering physician to suction the mouth and hypopharynx upon delivery of the head

B. be prepared to suction the mouth and hypopharynx upon receipt of the infant on the radiant warmer bed

C. intubate the trachea immediately and suction any meconium in all infants born through meconium-stained fluid

D. provide bag-and-mask positive-pressure ventilation upon receipt of the infant on the radiant warmer bed

E. provide tactile stimulation of the infant to initiate spontaneous respirations

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Critique: 208 Preferred Response: B

Resuscitation of the newborn delivered through meconium-stained amniotic fluid requires attention to airway patency and the possibility of meconium or mucus obstructing the airway. Upon receiving the infant from the delivering clinician, the clinician managing the infant should be prepared to position the infant's head, examine his or her oropharynx (mouth) and hypopharynx, and suction any meconium or mucus to provide a clear airway. If the infant exhibits depressed neuromotor tone, apnea, or bradycardia, the trachea should be examined and suctioned immediately. If the infant exhibits spontaneous respiration or is crying, tracheal suctioning is not necessary.

Routine suctioning of the mouth and hypopharynx by the delivering clinician following delivery of the head no longer is required. Nonetheless, the delivering clinician or the receiving clinician managing the infant may suction the mouth and hypopharynx immediately, when clinical judgment so warrants. Immediate tracheal intubation and suctioning is not required in all infants born through meconium-stained fluid and could result in soft-tissue injury or other trauma. Until the airway is examined or suctioned, bag-and-mask positive-pressure ventilation is not recommended because particulate meconium or mucus may be directed into the airway and aspirated. Tactile stimulation is ineffective for a depressed, apneic infant who does not exhibit spontaneous respirations.

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Question: 209

You are evaluating a 5-year-old girl who has a urinary tract infection. She has had four lower urinary tract infections in the last 2 years, all of which resolved completely with oral antibiotics. She denies symptoms of urgency and frequency. The only significant finding on her medical history is constipation. Results of renal ultrasonography and voiding cystourethrography are normal. Her growth parameters and physical examination findings are normal. You prescribe oral trimethoprim-sulfamethoxazole.

Of the following, the MOST appropriate additional step to help reduce the incidence of further urinary tract infection is to

A. begin an evaluation for immunodeficiency

B. perform renal scintigraphy

C. prescribe a stool softener and regular bowel routine

D. prescribe oral oxybutynin

E. refer her to a pediatric nephrologist

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Critique: 209 Preferred Response: C

Constipation is defined by infrequent or difficult passage of large or hard fecal material. It occurs commonly in the pediatric population, and its association with urinary tract dysfunction has been well described. Constipation can cause detrusor instability, which can lead to urinary incontinence, large bladder capacity, and dyscoordinated voiding. Urinary retention is common, either from dyscoordinated voiding or from outflow tract obstruction caused by large rectal fecal masses. All of these types of urinary dysfunction can lead to recurrent urinary tract infections. One study of children who had urinary retention found that 13% had functional constipation as the cause of their retention. Another found that approximately 30% of chronically constipated children complained of urinary incontinence, and 11% had urinary tract infection. Treatment of the underlying constipation improved urinary incontinence and prevented recurrent urinary tract infections.

The girl described in the vignette is otherwise healthy and growing well, and she has no anatomic urinary tract abnormalities. Accordingly, treatment of her underlying constipation is the next best step in the prevention of recurrent infections. Fecal disimpaction, stool softeners, and regular bowel evacuation using timed toilet sitting are the mainstays of treatment.

An evaluation for immunodeficiency is not warranted at this time, but could be considered if her growth parameters were abnormal or if she had additional infections outside of the urinary tract. Renal scintigraphy is used to assess renal anatomy and function and may be warranted in a child who has recurrent urinary tract infections, but it is not helpful in preventing future infections. Anticholinergic therapy, such as oxybutynin, can be helpful if overactive or unstable bladder is suspected, but the girl in the vignette has no symptoms of these conditions. Referral to a pediatric nephrologist may be indicated if evidence of renal dysfunction is present, but treatment of underlying risk factors, such as constipation, should be initiated first.

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Question: 210

A 12-year-old boy presents with an itchy rash that you diagnose as scabies. As he leaves the examination room, you note that he is limping. He is overweight, and his mother states he has been playing football to get some exercise. She believes he is limping because he was injured during football practice several weeks ago and has been complaining of left knee pain. Findings on physical examination of the knee are normal, but he complains of pain with hip motion.

Of the following, the radiographic study MOST likely to yield a diagnosis is

A. anteroposterior, lateral, and sunrise radiographs of the knee

B. bilateral anteroposterior and frog leg radiographs of the hips

C. magnetic resonance imaging of the knee

D. ultrasonography of the hip

E. ultrasonography of the knee

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Critique: 210 Preferred Response: B

Slipped capital femoral epiphysis (SCFE) occurs when the epiphysis of the femoral head "slips" in relation to the shaft of the femur. SCFE occurs most commonly in obese preteens and adolescents, although 30% of affected patients are not overweight. In its early stages, SCFE presents as pain, typically referred via the obturator nerve to the knee or distal thigh, as reported for the boy in the vignette. Such pain can prompt mistaken imaging of the knee. Clinically, many affected children walk with a limp or complain of pain with ambulation or sports participation. The clinician may observe external rotation of the hip when the patient walks and pain with and limitation of internal rotation of the hip during physical examination.

Bilateral anteroposterior and frogleg view radiographs (Item C210) of the hips are recommended to diagnose SCFE because the condition often is or ultimately becomes bilateral. Ultrasonography of the hip is unlikely to be helpful in the evaluation of an adolescent who has chronic pain and limp. Although the boy described in the vignette is complaining of knee pain, physical examination of the knee yields normal results, and he has pain with hip motion. Thus, it is unlikely that the knee is the source of the pain, and radiographs or magnetic resonance imaging of this area are not indicated. Ultrasonography is not a helpful study in evaluating the knee.

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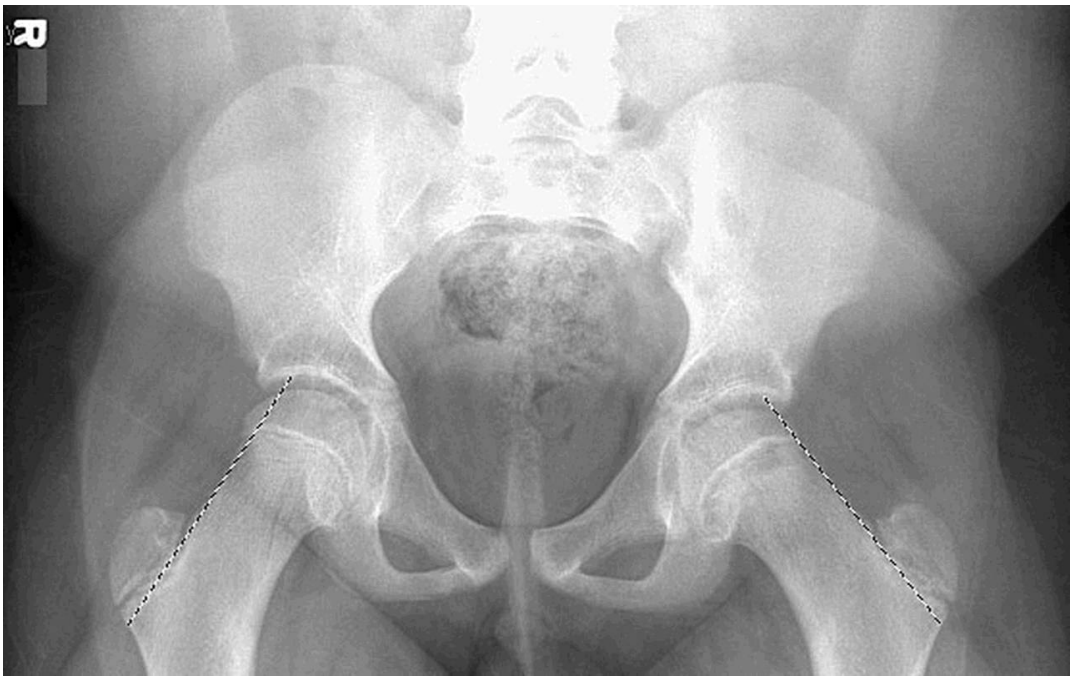
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Critique: 210



Slipped capital femoral epiphysis is characterized by an upward anterior movement of the femoral neck on the capital epiphysis (which becomes displaced posteriorly and inferiorly). In the normal hip (right), a line drawn along the superior margin of the femoral neck transects a portion of the ossified epiphysis. This does not occur on the affected side (left).

Courtesy of D. Krowchuk

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Question: 211

You are evaluating a 12-year-old boy as part of his annual health supervision visit. He has been in good health. His heart rate is 75 beats/min and blood pressure is 132/82 mm Hg using the appropriate-sized cuff. His weight is above the 95th percentile, and his height is at the 50th percentile. He has strong pulses at the right brachial and right femoral regions.

Of the following, the MOST appropriate diagnostic evaluation to pursue at this time is

A. blood urea nitrogen, creatinine, and electrolytes

B. echocardiography

C. radionuclide imaging of the kidneys

D. renal artery Doppler studies

E. urinary drug screen

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Critique: 211 Preferred Response: A

Hypertension is a major cause of morbidity and mortality in adults, and growing data suggest that it is becoming a greater clinical problem in the pediatric population, particularly adolescents. Although yet to be defined clearly, the lifelong risks for the child who has hypertension or a prehypertensive state are likely to be substantial. Blood pressure is affected by height, weight, sex, and race. A complete medical history, particularly family history and medications (including over-the-counter supplements), and a thorough physical examination are essential to early and accurate diagnosis of hypertension and assessment of secondary causes, comorbidities, and potential complications.

Specific questions in the history should seek to identify clinical findings that might suggest an underlying systemic disorder, including the presence of gross hematuria, swelling or edema, shortness of breath, or rashes. The past medical history should focus on prior hospitalizations, previous trauma, and urinary tract infections. The family history should evaluate for hypertension, diabetes, obesity, stroke, and renal disease. In addition, the history should explore the possibility of medications or drugs (prescribed, illicit, or over-the-counter) that can be associated with hypertension. An example of the latter may be the use of pseudoephedrine as a nasal decongestant. Additionally, some families and adolescents use dietary supplements, vitamins, herbal remedies, and homeopathic preparations. Some of these “supplements” may be associated with sympathomimetic or hypertensive effects.

The laboratory and diagnostic evaluation of the child who has hypertension should be guided by findings on the history and physical examination. For the pediatric patient who has confirmed hypertension, a screening panel of electrolytes, blood urea nitrogen, and creatinine as well as urinalysis and culture are indicated. Such blood tests are obtained to rule out renal disease.

A urinary drug screen is indicated if the history or patient behavior suggests a possible contribution by illicit substances, drugs, or supplements that can be associated with hypertension. The child described in the vignette is overweight, but he has no historical or physical examination findings suggestive of the use of illicit substances, drugs, or supplements. Echocardiography also is not necessary at this stage of diagnosis because there is no evidence of congenital heart disease such as coarctation or end-organ involvement. Pediatric patients who have comorbid factors such as diabetes or systemic lupus erythematosus should undergo echocardiography as part of their hypertension evaluations. Radionuclide imaging of the kidneys involves exposure to ionizing radiation and typically is reserved for children of this age whose blood pressures are more than 5 mm Hg greater than the 99th percentile. Although renal vascular ultrasonography does not involve ionizing radiation, it also generally is reserved for young children whose blood pressures are beyond the 95th percentile and older children and adolescents whose blood pressures exceed the 99th percentile.

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Question: 212

An 8-year-old boy presents to the emergency department following 3 days of progressive difficulty walking. He says that his back hurts. Physical examination shows no abnormalities. On neurologic examination, he is alert, makes good eye contact, and responds to questions appropriately, but seems distressed. Cranial nerve examination results are normal. On motor examination, strength is 4/5 in the arms and legs. Reflexes are absent. He cannot rise from the floor unassisted. Sensory examination findings are normal.

Of the following, the MOST important initial procedure is

A. brain magnetic resonance imaging

B. electroencephalography

C. electromyography

D. forced vital capacity

E. lumbar puncture

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Critique: 212 Preferred Response: D

The subacute progressive weakness and back pain described for the boy in the vignette are serious findings that require emergent diagnostic evaluation and management. Progressive weakness generally requires emergent evaluation that initially focuses on localization of the problem to the brain, brainstem, spinal cord, root, nerve, neuromuscular junction, or muscle.

The normal consciousness and normal findings on cranial nerve evaluation for this boy make a brain or brainstem cause of the general weakness unlikely. Bilateral upper and lower limb weakness with normal sensation typically does not localize to the spinal cord. The absent arm and leg reflexes and back pain make muscle and neuromuscular junction less likely sources of the problem. The clinical findings suggest a polyneuropathy or polyradiculopathy, most likely demyelinating, as in acute inflammatory demyelinating polyneuropathy (AIDP), also known as Guillain-Barré syndrome.

Subacute generalized weakness can progress rapidly to involve airway muscles or cause respiratory insufficiency and death. Therefore, assessment of respiratory function is paramount, and the preferred initial procedure is measurement of forced vital capacity or of negative inspiratory force. At the bedside, a rough estimate may be obtained by having the child inhale maximally and count continuously and clearly to as high a number as possible until expiration is complete. The final number is multiplied by 100 mL (eg, counting to 20 indicates 2,000 mL). Forced vital capacity should exceed 20 mL/kg. A measurement of less than 15 mL/kg is an indication for intensive care unit admission and ventilatory support. Pulse oximetry is not sensitive to neuromuscular respiratory insufficiency; the readings may be normal right up to the point of respiratory crisis. Therefore, after the patient is admitted, frequent forced vital capacity or negative inspiratory force evaluations are required, which are handled best in the intensive care unit.

Once respiratory (airway, respiratory sufficiency) and autonomic (rarer — cardiac arrhythmias, hyper- or hypotension, urinary retention) systems have been assessed and managed, making the proper diagnosis is critical. Based on the clinical presentation of the boy described in the vignette, tests of central nervous system function (electroencephalography) and structure (magnetic resonance imaging) are of low diagnostic yield. With a presentation that is consistent with AIDP, a lumbar puncture for the classic finding of elevated protein concentrations but normal white blood cell count (albuminocytologic dissociation) is important. However, early in the disease process, the cerebrospinal fluid protein concentration may be normal. Nerve conduction studies generally support the diagnosis and clarify whether the condition is demyelinating (more common) or axonal (less common, worse prognosis). Electromyography may not show intrinsic muscle disease but may reveal acute denervation.

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Question: 213

A 16-year-old girl is brought by ambulance to the emergency department with complaints of vomiting, abdominal cramping, and diarrhea over the past day. She complains of both hot and cold flashes and muscle aches, and she has had difficulty sleeping for the past several nights. She was in a drug rehabilitation program 2 years ago, but she denies the use of alcohol or other drugs now. On physical examination, her temperature is 101.3°F (38.5°C), heart rate is 120 beats/min, respiratory rate is 24 breaths/min, and blood pressure is 145/90 mm Hg. She is alert but anxious, her pupils are dilated, and she has a slight tremor. Cardiovascular examination results are normal except for tachycardia. Her lungs are clear to auscultation, and results of her abdominal examination are within normal limits.

Of the following, the MOST likely diagnosis is

A. acute intoxication with a barbiturate

B. acute intoxication with amphetamines

C. withdrawal from alcohol

D. withdrawal from chronic marijuana use

E. withdrawal from heroin

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Critique: 213 Preferred Response: E

Alcohol, tobacco, and other drug use continues to be problematic for young people in the United States. Data from the 2006 *Monitoring the Future Study*, which surveys nationally representative samples of about 50,000 8th-, 10th-, and 12th-grade students per year in about 400 private and public secondary schools in the United States, show that the use of any illicit drug by all of these age groups is down from recent peaks in 1996 and 1997. Use still continues at relatively high rates, however, with 48% of young people having tried an illicit drug by the time they finished high school. Marijuana remains the most commonly used illicit drug; 12% of 8th graders, 25% of 10th graders, and 32% of 12th graders reported use of the substance within the last year.

Although use of other drugs is less, recent concerns have included heroin and other narcotics, which have not shown decreases in use similar to marijuana. Heroin continues to be used both with and without a needle in a relatively small number of young people: 0.8% to 0.9% annually in 2006 for 8th-, 10th-, and 12th-grade students. Two specific narcotics, oxycodone and hydrocodone bitartrate acetaminophen (Vicodin®), have raised concern. Oxycodone use by 12th-graders in the past 12 months was reported at 4.3% in 2006, an almost 40% increase since its use was first measured in 2002. Vicodin use has remained relatively stable since first measured in 2002 at an annual prevalence of 9.5% in 12th graders. Both of these drugs have high addictive potential.

The young woman described in the vignette is at higher risk of drug use because she previously experienced problems with use. Her symptoms are classic for heroin withdrawal. Acute intoxication with a barbiturate presents with respiratory depression, hypotension, delirium, and psychosis. The acute effects of amphetamines reflect their stimulatory potential, including euphoria, agitation, hypertension, and possible arrhythmia and psychosis. Lower doses of marijuana may induce euphoria, impaired concentration, and auditory and visual enhancement; higher doses may induce mood fluctuations, depersonalization, and hallucinations. Anxiety, panic, delusions, hallucinations, paranoia, and psychosis may result from toxic reactions to marijuana. Withdrawal from chronic use is characterized by irritability, sleep disturbances, tremor, nystagmus, anorexia, nausea, vomiting, or diarrhea, but these reactions usually are relatively mild.

Mild withdrawal from alcohol involves headache, mild tremulousness, and nausea and vomiting. Severe withdrawal, which is very rare in adolescents, may have the effects of dilated pupils, tremulousness, seizures, tachycardia and hypertension, nausea and vomiting, hallucinations, confusion, anxiety, and agitation.

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Question: 214

A 5-year-old boy who has nephrotic syndrome required 3 months of prednisone therapy (2 mg/kg per day) to induce remission. He now has been weaned off prednisone for 1 week.

Of the following, the symptom or sign that is MOST indicative of adrenal insufficiency is

A. headache

B. hyperbilirubinemia

C. nausea

D. pruritus

E. weight gain

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Critique: 214 Preferred Response: C

The symptoms of acute adrenal insufficiency following withdrawal of glucocorticoid are those of adrenocorticotropic hormone and glucocorticoid (cortisol) deficiency. They include nausea, loss of appetite, myalgia and muscle weakness, and malaise. Signs might include hypotension, decreased pulse pressure, pallor, and rapid heart rate. Headache, hyperbilirubinemia, and pruritus are not prominent symptoms of glucocorticoid withdrawal. Anorexia rather than weight gain is the normal response.

Because mineralocorticoid (aldosterone) secretion usually is preserved, electrolyte imbalance that involves hyperkalemia and hyponatremia is unlikely. However, very ill children who have isolated cortisol deficiency may develop hyponatremia because of the compensatory release of vasopressin in response to decreased intravascular volume.

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Question: 215

The parents of a 17-year-old boy bring him to you because he has told his parents that he has a "computer in his head" that tells him what to do. He refuses to attend school or go outside the house because of the "scary faces" he sees at times. In your office, the boy is expressionless but begins to giggle for no apparent reason when you speak to him. Results of his urine drug screen are negative.

Of the following, the MOST likely diagnosis is

A. Asperger disorder

B. bipolar disorder

C. conduct disorder

D. depression

E. schizophrenia

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Critique: 215 Preferred Response: E

The boy described in the vignette is exhibiting behaviors indicative of schizophrenia. There is no pathognomonic sign or symptom for schizophrenia; every sign or symptom seen in the condition occurs in other psychiatric and neurologic disorders. The onset of schizophrenia can occur rarely even in childhood, but the frequency increases rapidly after the age of 13 years. Disease onset prior to age 18 years is considered early onset and prior to 13 years is considered very early onset. The diagnostic criteria are the same as in adults. The *Diagnostic and Statistical Manual of Mental Disorders* edition IV diagnostic criteria require the presence of at least two of the following symptoms for a significant period of time during a 1-month period: delusions, hallucinations, disorganized speech or catatonic behavior, negative symptoms, flat affect, and paucity of thought or speech. When there are bizarre delusions or hallucinations that consist of a voice commenting on how the person is behaving or thinking or two voices talking to each other, only one criterion is necessary. Continuous signs of disturbance must last for at least 6 months, and there must be at least 1 month of symptoms (active phase). Boys tend to become symptomatic at a younger age than girls. An earlier onset and chronic course of schizophrenia may indicate a poorer prognosis.

In the typical premorbid history of schizophrenia, patients who have schizoid or schizotypal personalities are characterized as quiet, passive, and introverted. They have few childhood friends. Preschizophrenic adolescents may have no close friends and may avoid team sports.

Schizophrenia occurs in several phases, with some degree of variability. In the prodrome phase, some adolescent patients may have a sudden onset of obsessive-compulsive behavior. Additional prodromal signs include markedly peculiar behavior, abnormal affect, unusual speech, bizarre ideas, and strange perceptual experiences, as described for the boy in the vignette. There may be a decrease in school functioning and poor personal hygiene. The acute phase is characterized by psychotic symptoms and decrease in function. Positive symptoms are those that are added on to normal experience and include delusions, hallucinations, and disorganized speech and behavior. Negative symptoms are those that are taken away from normal experience and include affective blunting, poverty of speech or speech content, poor grooming, lack of motivation, and social withdrawal. Despite treatment, some psychotic symptoms, as well as poor mood and confusion, may remain. Positive symptoms tend to respond better to treatments. Intoxication or withdrawal from drugs can present with the same symptoms as psychiatric disorders. Accordingly, a urine drug screening, as ordered for the boy in the vignette, is indicated to diagnosis substance-induced psychosis.

A teen who has Asperger disorder would have a long-standing history of impaired reciprocal social interactions. Those who have bipolar disorder demonstrate more elation and merriment than described for the boy in the vignette. Although an individual who has depression may have poor mood, delusions or hallucinations are not typical. The diagnostic criteria for conduct disorder include aggression to people and animals, destruction of property, and theft.

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Question: 216

An 8-year-old girl presents with a 4-day history of fever, headache, and abdominal pain. Her mother states that they live in a rural area and have multiple pets, including dogs, cats, horses, cows, and a pet raccoon. There is no history of tick bites. On physical examination, the girl appears mildly toxic, has a temperature of 102.2°F (39°C), and has a grade II/VI systolic ejection murmur best heard on the left side of the sternal border. Her right upper quadrant is tender to palpation, but there is no hepatosplenomegaly. Findings on her skin and extremity examination are normal. A complete blood count reveals a white blood cell count of 1.2x103/mcL (1.2x109/L) with 90% neutrophils and 10% lymphocytes. Her hemoglobin is 10 g/dL (100 g/L), and her platelet count is 50x103/mcL (50x109/L). Her alanine aminotransferase is 600 U/L, and her aspartate aminotransferase is 450 U/L. Her amylase and lipase values are normal. Serum sodium is 133 mEq/L (133 mmol/L), but the remainder of her electrolyte values are normal.

Of the following, the MOST likely diagnosis is

A. human monocytic ehrlichiosis

B. Lyme disease

C. Rocky Mountain spotted fever

D. tularemia

E. typhus

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Critique: 216 Preferred Response: A

Human monocytic ehrlichiosis (HME) is a rickettsial disease caused by *Ehrlichia chaffeensis,* which is transmitted to humans by the bite of a tick. Common complaints of HME include fever, headache, nausea, abdominal pain, and myalgias. Patients may have normal findings on physical examination, but hepatosplenomegaly, cardiac murmurs, lymphadenopathy, and rash have been reported in some patients. A rash is described in approximately two thirds of children and one third of adults and starts as maculopapular but may progress into petechial/purpuric. HME is clinically indistinguishable from Rocky Mountain spotted fever (RMSF). Laboratory abnormalities common to both infections include thrombocytopenia and hyponatremia, but patients who have HME are more likely to have elevated liver function test results and leukopenia with lymphopenia. Approximately 50% to 75% of patients have no history of a tick bite.

Human granulocytic ehrlichiosis (HGE) is caused by *Anaplasma phagocytophila* and also is transmitted by the bite of a tick. The tick vectors are different for HGE and HME. HME is transmitted by the bite of the wood tick (*Dermacentor andersoni*) or the Lone Star tick (*Amblyomma americanum*), while the deer tick (*Ixodes scapularis*) is most likely to transmit HGE. Patients infected with HGE may have fever and a headache, but findings on physical examination usually are normal. They have similar laboratory abnormalities to those who have HME except that they have neutropenia instead of lymphopenia, and the intracytoplasmic inclusions called morulae are more likely to be found. Doxycycline is used to treat both of these rickettsial diseases.

Patients who have Lyme disease typically do not appear toxic or have the laboratory abnormalities described in the vignette. Although there is a typhoidal form of illness due to *Francisella tularensis* (tularemia), it is extremely rare, and most affected children present with glandular or ulceroglandular disease. Typhus can be endemic or epidemic. Epidemic typhus is due to the bite of the human louse, and endemic typhus is caused by a mite bite. Although both of these rickettsial diseases can present with fever and a headache, patients usually are not toxic and do not have laboratory abnormalities such as those reported for the girl in the vignette.

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Question: 217

You are evaluating a 1-hour-old term infant for respiratory distress. The infant was born by spontaneous vaginal delivery to a 24-year-old para 1 gravida 1 mother who had premature rupture of membranes 19 hours prior to delivery. The mother had a temperature of 101.5ºF (38.6°C) at the time of delivery and received one dose of ampicillin. Her group B streptococcal culture at 34 weeks of pregnancy was negative. You suspect group B streptococcal infection in the infant.

Of the following, the MOST rapid method for diagnosis in the infant is

A. amniotic fluid analysis

B. antigen detection

C. C-reactive protein

D. culture

E. erythrocyte sedimentation rate

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Critique: 217 Preferred Response: B

Group B streptococci (GBS) are gram-positive diplococcal organisms that produce beta-hemolysis when plated on blood agar media (complete clearing of the erythrocytes). Colonies are 3 to 4 mm in diameter and are flat, grayish-white, and somewhat mucoid. Diagnosis of GBS infection is confirmed by isolation of the pathogen from a normally sterile body site such as blood, cerebrospinal fluid (CSF), bone aspirate or joint fluid, or soft tissue. Laboratory methods used to confirm the identity of the organism include: testing for resistance to bacitracin or trimethoprim-sulfamethoxazole, hydrolysis of sodium hippurate broth, failure to hydrolyze bile esculin, and CAMP testing (synergistic hemolysis on sheep blood agar when GBS are grown in the presence of the beta-toxin of *Staphylococcus aureus)*. Selective enrichment broths, such as Todd-Hewitt broth supplemented with gentamicin and nalidixic acid or colistin and nalidixic acid, may be used to enhance recovery of GBS from genital tract specimens.

Latex agglutination is a rapid and fairly sensitive test for detecting GBS antigens. The optimal fluid for latex agglutination testing is CSF; urine, sera, and other body fluids also may be tested, but nonspecific agglutination may occur, which may cause false-positive results. Depending on the commercial assay used, sensitivity ranges from 70% to 100% and specificity is close to 100%.

GBS remains uniformly susceptible to penicillin G and to other beta-lactam antibiotics, the cephalosporins, vancomycin, and the carbapenems. GBS resistance to the macrolide class of antibiotics has increased, with resistance rates as high as 18% to erythromycin and up to 15% to clindamycin. Almost 90% of GBS are resistant to tetracycline, and GBS are uniformly resistant to bacitracin, nalidixic acid, trimethoprim-sulfamethoxazole, and metronidazole.

The most rapid method for diagnosing GBS infection for the infant described in the vignette is antigen detection. Amniotic fluid analysis and culture require 48 to 72 hours for results, but if positive, they can confirm the diagnosis. C-reactive protein and erythrocyte sedimentation rate are both nonspecific tests that indicate the presence of inflammation or infection but are not specific for a particular pathogen.

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Question: 218

A mother brings in her 3-year-old daughter because of daytime urinary incontinence and abdominal pain. The mother explained that the girl was toilet trained at 2 years of age. On physical examination, growth parameters and vital signs are normal, although the girl has mild suprapubic tenderness without associated costovertebral angle tenderness or sacral dimples. Urinalysis shows a urine specific gravity of 1.025, pH of 6.5, 2+ blood, 1+ protein, 3+ leukocyte esterase, and positive nitrite. Urine microscopy demonstrates 5 to 10 red blood cells/high-power field, 20 to 50 white blood cells/high-power field, and 3+ bacteria.

Of the following, the MOST likely etiologic agent is

A. *Enterococcus faecalis*

B. *Escherichia coli*

C. *Klebsiella pneumoniae*

D. *Proteus mirabilis*

E. *Staphylococcus saprophyticus*

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Critique: 218 Preferred Response: B

Escherichia coli is the causative organism in 80% to 90% of first-time urinary tract infections (UTIs) in children. Other pathogens include *Klebsiella pneumonia*, *Proteus* sp, *Enterococcus* sp, and *Staphyloccus saprophyticus*. *Pseudomonas* also can be a pathogen in immunocompromised patients or those who have received repeated courses of antibiotics for recurrent infections. The clinician must assess the patient for a UTI based on signs, symptoms, and urinalysis findings.

Organisms such as *E coli, K pneumoniae*, and *Proteus* sp can reduce dietary nitrate to nitrite, so a positive urine dipstick test for nitrite, as reported for the girl in the vignette, is virtually diagnostic of gram-negative bacteruria. If the test result is negative in an older child in whom a UTI is suspected, the infection may be caused by a gram-positive organism such as *Enterococcus* sp or *S saprophyticus*. Of note, the nitrite test is much less helpful in infants. Conversion of nitrate to nitrite may take up to 4 hours. Because infants and young children have small bladder volumes and urinate frequently, there may be insufficient time for nitrites to be formed and, therefore, the nitrite test may be negative even in the presence of a UTI caused by a gram-negative organism.

Urine pH also may be useful in diagnosing UTIs. Urease-producing organisms (eg, *Proteus mirabilis*, some strains of *S saprophyticus*) degrade urea into ammonia, resulting in an elevated urine pH (8.0 to 8.5).

The girl described in the vignette has symptoms of a lower UTI. Options for therapy include trimethoprim-sulfamethoxazole (if local resistance patterns indicate low levels of *E coli* resistance) or a third-generation cephalosporin (eg, cefixime, cefdinir). These antibiotics also may be used for outpatient management of acute pyelonephritis. For hospitalized patients, a third-generation cephalosporin such as ceftriaxone or cefotaxime provides adequate coverage. An alternative regimen is ampicillin plus gentamicin.

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Question: 219

You have recently diagnosed mild persistent asthma in a 10-year-old boy and decide to initiate low-dose inhaled corticosteroid therapy. During your discussion with the parents, you review the potential benefits and adverse effects of inhaled corticosteroids.

Of the following, the MOST likely adverse effect is

A. adrenal suppression

B. cataracts

C. decreased adult height

D. dysphonia

E. increased bronchial hyperresponsiveness

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Critique: 219 Preferred Response: D

Despite inhaled corticosteroids remaining the cornerstone therapy for persistent asthma, many parents are reluctant to have their children begin such medications. Addressing parents' concerns and outlining the potential adverse effects may improve understanding and compliance significantly. Local oropharyngeal symptoms such as dysphonia, oral candidiasis, and cough are the most commonly encountered adverse effects. Local symptoms appear to be dose-related and can be lessened by using a "spacer" and rinsing the mouth after use. A number of studies have shown that transient growth suppression can occur during the first year of inhaled corticosteroid therapy, but a daily cumulative dose up to 400 mcg of inhaled budesonide has been shown not to affect predicted adult height.

Other, less common adverse effects of inhaled steroids are acne, mood swings, weight gain, decreased serum immunoglobulin G concentrations, and rarely, posterior subcapsular cataracts and adrenal suppression.

Inhaled steroids are used for persistent asthma to help halt lung remodeling by decreasing bronchial inflammation, reducing inflammatory mediators, and decreasing bronchial hyperresponsiveness.

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Question: 220

A 5-year-old boy is brought to the office 4 hours after falling off his bicycle. His mother reports that he was not wearing a helmet, and bystanders said that he did not lose consciousness. When his friends brought him home, he was tearful and sleepy but was answering questions appropriately. The mother noted an abrasion on the left side of his head and applied ice to a small area of swelling on the left temple. Over the last several hours, however, he has become increasingly confused and has had multiple episodes of vomiting. On physical examination, he is difficult to arouse, and his right pupil is larger than his left.

Of the following, a TRUE statement regarding this patient's likely diagnosis is that

A. improved survival is associated with prompt neurosurgical intervention in symptomatic patients

B. intracranial injury is unlikely because there was no loss of consciousness

C. mannitol is the initial treatment of choice

D. the gradual onset of symptoms is most consistent with a concussion

E. the injury is caused by laceration of the veins that bridge the dural sinuses and the brain

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Critique: 220 Preferred Response: A

Epidural hematoma should be suspected in pediatric patients who sustain seemingly minor head injuries but then exhibit progressive development of symptoms such as lethargy, worsening headache, vomiting, and focal neurologic signs, as described for the boy in the vignette. The absence of a history of loss of consciousness or signs of skull fracture is not a reliable negative predictor of an epidural hematoma. As many as 50% of affected children do not have changes in consciousness, and skull fractures may be absent in up to 40%. Epidural hematomas typically result from lacerations to meningeal blood vessels (Item C220); shearing of the bridging veins between the dural sinuses and the brain most often causes subdural hematomas.

Epidural hematomas are seen in 2% to 3% of all children admitted to the hospital following head trauma. When diagnosed and treated before patients deteriorate, the associated mortality is nearly 0%. The treatment of an epidural hematoma that is large, in the temporal or posterior fossae, causing focal neurologic deficits, or associated with alteration of consciousness is neurosurgical drainage. Selected patients who have small hemorrhages that produce minimal or no symptoms may be considered for nonoperative management.

The initial treatment for any patient in whom an intracranial injury is presumed is maintenance of adequate oxygenation, ventilation, and perfusion. Neurosurgical intervention may be indicated for patients who have space-occupying lesions. The use of mannitol as an adjunct for the treatment of cerebral edema and increased intracranial pressure no longer is recommended as an initial intervention. It has been demonstrated that survival is improved by maintaining the brain in a euvolemic or slightly hypervolemic state through the use of isotonic and hypertonic saline solutions. Mannitol may be employed to decrease intracranial pressure and improve cerebral blood flow in the intensive care unit for a patient who has an intracranial pressure monitor in place.

Concussions typically are associated with the rapid onset of neurologic impairment at the time of a direct blow to the head or a blow to the body that impacts the head with an “impulsive” force. Signs and symptoms of a concussion may include confusion, amnesia, loss of consciousness, headache, nausea/vomiting, visual disturbances, concentration difficulty, ataxia, and dizziness. Affected patients do not have focal motor deficits and abnormal neuroimaging findings. The concussive symptoms resolve spontaneously over time, with most patients returning to baseline within 7 to 10 days.

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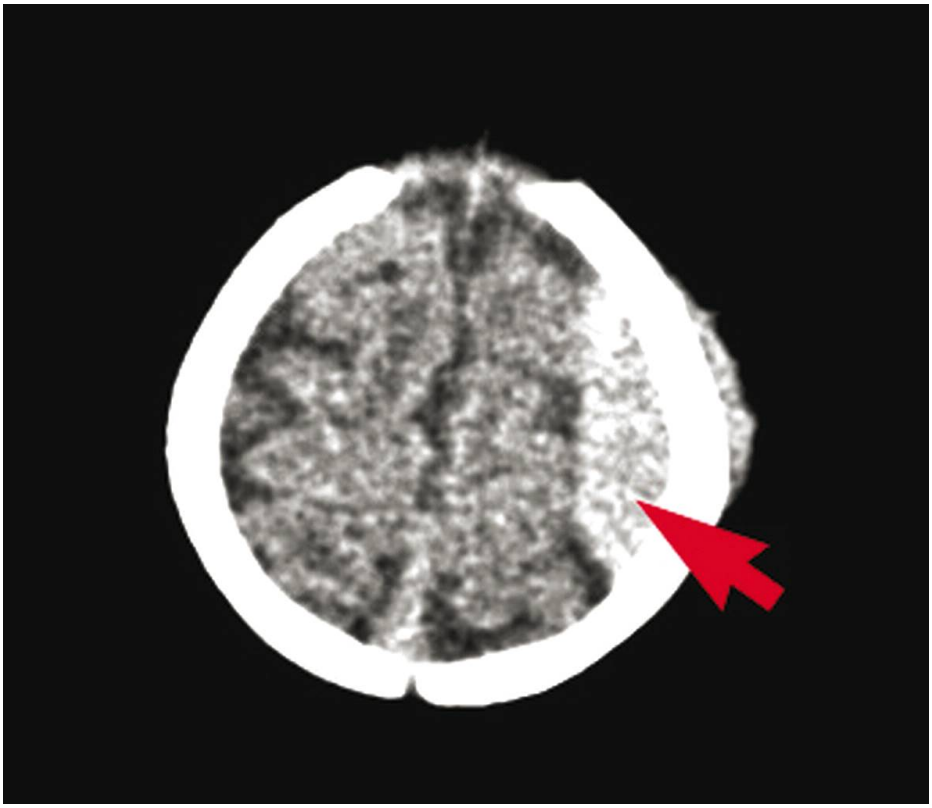
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Critique: 220



Axial computed tomography scan (brain window) shows an area of increased attenuation (arrow) with convex borders characteristic of an epidural hematoma.

Courtesy of D. Mulvihill

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Question: 221

An 11-year-old boy presents for evaluation of recurrent oral ulcers (Item Q221), joint pain, and weight loss for 3 months.

Of the following, the MOST likely explanation for these findings is

A. Crohn disease

B. cyclic neutropenia

C. hand-foot-and-mouth disease

D. herpangina

E. herpetic gingivostomatitis

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Question: 221



Aphthae, as observed in the boy described in the vignette.

Courtesy of dermatlas.org

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Critique: 221 Preferred Response: A

The clinical triad of recurrent oral ulcers (aphthae), joint pain, and weight loss exhibited by the boy described in the vignette suggests a diagnosis of Crohn disease. Extraintestinal manifestations occur in 25% to 35% of patients who have inflammatory bowel disease. Almost any system can be involved, but the skin and mucosa, joints, liver, eye, and bone are affected most often.

Oral aphthae occur in as many as 20% of patients who have Crohn disease. They are painful ulcers or erosions on the lips, gingivae, tongue, palate, or buccal mucosa (Item C221A). Individual lesions last 7 to 10 days and may occur in crops at irregular intervals. Aphthae parallel disease activity, but may precede the appearance of intestinal symptoms.

Oral ulcers also are a feature of cyclic neutropenia, a rare disorder in which periodic fluctuations in circulating neutrophil numbers are associated with fever, pharyngitis, and lymphadenopathy. Symptoms occur at regular intervals and are associated with a nadir in bone marrow function and consequent neutropenia.

A number of viral infections produce oral ulcers or erosions, but they may be distinguished from Crohn disease because they are self-limited, may have distinctive extraoral findings, and are not associated with chronic weight loss or joint pain. Hand-foot-and-mouth disease produces shallow ulcers that involve the soft palate, uvula, tonsillar pillars, and tongue (Item C221B). Most affected patients also exhibit erythematous papules or oval vesicles that have erythematous borders and are located on the palms (Item C221C), soles, and digits. Small (1- to 2-mm) vesicles and ulcers that involve the posterior oropharynx, including the tonsillar pillars, soft palate, and uvula, are characteristic of herpangina (Item C221D). Children who have herpetic gingivostomatitis exhibit fever, irritability, and refusal to eat. Vesicles and ulcers are observed on the anterior soft palate, tongue, and gingivae. The gingivae are edematous, friable, and inflamed. Vesicles and erosions also may appear on the lips and chin (Item C221E).

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Critique: 221



Aphthae are painful ulcers or erosions that appear on the lips, gingivae, tongue, palate, or buccal mucosa.

Courtesy of dermatlas.org

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Critique: 221



In hand-foot-and-mouth disease, ulcers may appear on the tongue, soft palate, uvula, and tonsillar pillars.

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Critique: 221

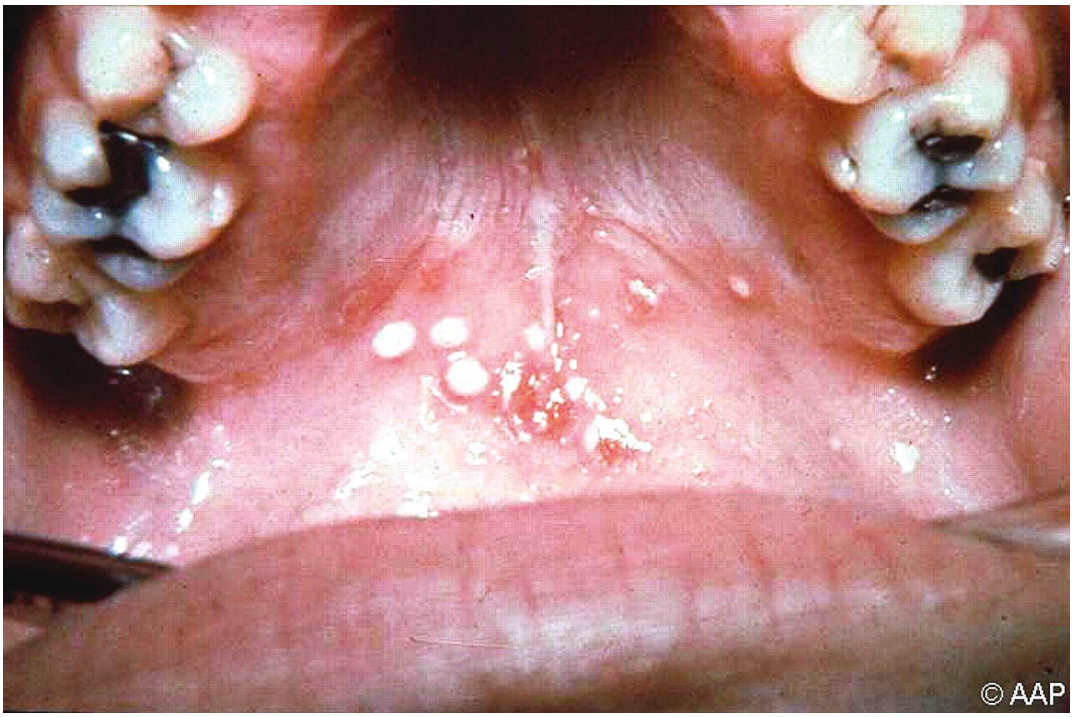


Oval or round vesicles with surrounding erythema occur on the palms and soles in hand-foot-and-mouth disease.

Courtesy of D. Krowchuk

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Critique: 221



In herpangina, patients develop vesicles and ulcers in the posterior pharynx.

Courtesy of *Red Book¨ Online*

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Critique: 221



Vesicles and ulcers located on the soft palate, tongue, gingivae, lips, and chin are characteristic of herpetic gingivostomatitis.

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Question: 222

You are following an 11-year-old girl who has Crohn disease involving the stomach, ileum, and colon. Her maintenance medications are mesalamine and 6-mercaptopurine. Over the past year, she has received four courses of corticosteroid treatment, but continues to have intermittent abdominal pain and diarrhea. Upon review of her growth curve, you note that her height has been the same over the past 12 months. You suspect that the combination of Crohn disease and corticosteroid therapy has resulted in growth arrest. You discuss your concerns with her gastroenterologist.

Of the following, the MOST appropriate medication to control this patient's disease and reduce her dependence on corticosteroids is

A. cyclophosphamide

B. infliximab

C. mycophenolate mofetil

D. tacrolimus

E. thalidomide

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Critique: 222 Preferred Response: B

Crohn disease (CD) and ulcerative colitis (UC) are serious illnesses mediated by the immune system that cause intestinal inflammation. The inflammation in UC is limited to the mucosal layer of the large bowel. Thus, affected patients most commonly present with pain associated with defecation, diarrhea, and rectal bleeding. In contrast, the inflammation in CD is transmural (involving the entire thickness of the bowel) and can involve any region of the gastrointestinal tract (most commonly the ileum and cecum). Therefore, patients who have CD present with a wide variety of symptoms, including diarrhea, rectal bleeding, anorexia, growth failure, anemia, abdominal pain, and perianal disease. The diagnosis of inflammatory bowel disease is established by contrast radiography, upper endoscopy, and colonoscopy.

Therapy for inflammatory bowel disease involves a combination of medical and nutritional therapy. The administration of a liquid diet (elemental or polymeric formula) for 6 to 8 weeks, combined with the cessation of eating during this period, may induce a remission in children who have CD, but this approach is not effective in UC. In addition, it is difficult to maintain such a diet indefinitely. For this reason, medications are essential in the long-term management of this condition. Most commonly, corticosteroids are used to induce remission in moderate-to-severe CD or UC. Salicylates (eg, mesalamine) are useful in maintaining remission in UC, but are less effective in CD. Immunomodulators (6-mercaptopurine, azathioprine, methotrexate) are used to maintain remission in patients who have UC and fail salicylate therapy and in those who have CD. Patients who have CD and fail salicylate and immunomodulator therapy, such as the girl described in the vignette, benefit from the addition of infliximab, an antibody to tumor necrosis factor-alpha. Infliximab has been approved by the United States Food and Drug Administration for use in children who have CD. Although this medication is highly effective treatment, patients have an increased risk of opportunistic infections (especially tuberculosis) and lymphoma. Therefore, a physician and patient considering infliximab need to weigh the risks and benefits carefully. Tacrolimus and thalidomide sometimes are used in the treatment of CD if patients failed or are intolerant to infliximab. Cyclophosphamide and mycophenolate rarely are used to treat inflammatory bowel disease.

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Question: 223

A 4-hour-old newborn who weighs 1,890 g and was born at 39 weeks' gestation has a serum glucose concentration of 25 mg/dL (1.4 mmol/L), resulting in tremors and jitteriness. The child appears to have intrauterine growth restriction and is small for gestational age (SGA), but is not ill and exhibits no dysmorphisms. You admit the infant to the special care nursery and order an intravenous dextrose 10% in water bolus and infusion and some additional laboratory tests. The complete blood count reveals a hemoglobin of 23 g/dL (230 g/L), hematocrit of 68% (0.68), platelet count of 150x103/mcL (150x109/L), and white blood cell count of 7x103/mcL (7x109/L) with a normal differential count.

Of the following, the MOST likely complication for this infant is

A. hyperbilirubinemia

B. hypercalcemia

C. hypertension

D. hypokalemia

E. hyponatremia

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Critique: 223 Preferred Response: A

Polycythemia is defined as a hematocrit (Hct) of greater than 65% (0.65) and indicates a relative increase in the red blood cell (RBC) mass over the plasma volume of whole blood. An Hct above 65% (0.65) may be associated with hyperviscosity of the blood. Neonatal polycythemia often is referred to as hyperviscosity syndrome. Increases in blood viscosity reduce the flow of blood through the microcirculation, which may result in hypoglycemia, respiratory distress, jitteriness, hypotonia, and feeding problems. A late phenomenon is hyperbilirubinemia as the increased RBC mass breaks down and hemoglobin degradation results in increased bilirubin load for hepatic glucuronidation and excretion.

The infant described in the vignette has two significant risk factors for polycythemia: growth restriction and low birthweight at a term gestation. In utero growth restriction may result from various causes, but maternal or placental disease states that reduce oxygen delivery to the fetus incite release of fetal erythropoietin and result in an increased RBC mass to ensure oxygen-carrying capacity and delivery to fetal tissues. Growth restriction or polycythemia itself may be associated with neonatal hypoglycemia. Because many clinical signs of the ill neonate are nonspecific, some signs of hypoglycemia clearly overlap with those of polycythemia and warrant urgent intervention. Hypoglycemia must be treated to preserve central nervous system function. If, in the face of polycythemia, hypoglycemia does not improve with intravenous glucose administration, consideration should be given to conducting a partial exchange transfusion with intravenous normal saline to reduce the RBC mass and facilitate improved circulatory flow in the microcirculation. Partial exchange transfusion also may be appropriate to treat symptomatic polycythemia.

Hypercalcemia and hypokalemia are not associated with polycythemia. Hypertension is uncommon in newborns and not related to polycythemia. Hyponatremia is uncommon in polycythemia.

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Question: 224

An 8-year-old boy is brought to your office after falling from a swing at school. He complains of right leg pain. Physical examination findings are normal, except he walks with a limp. A radiograph shows no fracture, but a bone cyst is noted.

Of the following, a TRUE statement about bone cysts is that

A. all bone cysts, regardless of type, are considered premalignant conditions

B. aneurysmal bone cysts occur most commonly in the flat bones

C. aneurysmal bone cysts typically do not cause pain

D. fractures are an uncommon complication of unicameral bone cysts

E. unicameral bone cysts occur most commonly in the humerus and femur

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Critique: 224 Preferred Response: E

Two primary types of bone cysts can be seen in the pediatric population. Unicameral bone cysts, or simple bone cysts, are most common before skeletal maturity. These fluid-filled cysts are found adjacent to the epiphyseal plate, usually at the proximal humerus and the proximal femur (Item C224A). Pathologic fractures are common, often after minor trauma, which is usually when they are diagnosed because most children are asymptomatic before the trauma. Simple bone cysts typically resolve after skeletal maturity. However, many experts advocate treating them, especially large cysts in the high-stress area of the proximal femur, to prevent further pathologic fractures. The treatment consists of allowing the first pathologic fracture to heal if present, followed by intralesional injection of steroids or, less commonly, autogenous bone marrow. These lesions are not considered precancerous.

Aneurysmal bone cysts usually are seen in the teenage years. These cavitary lesions arise within the metaphysis, and an associated periosteal reaction may be seen (Item C224B). They occur primarily in long bones, especially the femur and tibia, but spine lesions also are common. Most affected children complain of pain with or without swelling. The lesions can grow rapidly, and spinal lesions (which are more common in the posterior vertebrae than in the vertebral body) can spread to adjacent vertebrae and cause significant cord or nerve root compression. Because aneurysmal bone cysts may be associated with underlying bone tumors, referral to orthopedics for further evaluation and management is indicated. Management might include curettage with bone grafting.

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Critique: 224



Unicameral (simple) bone cyst: An anteroposterior radiograph of the tibia shows a metaphyseal bony defect with minimal expansion of the bony contours and few septations.

Courtesy of D. Mulvihill

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Critique: 224



Lateral radiograph of the humerus shows a lytic, expansile lesion with thin internal strands and a thin continuous rim of bone. These features are characteristic of an aneurysmal bone cyst.

Courtesy of D. Mulvihill

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Question: 225

An 11-year-old boy has been participating all summer on a swim team. He presents for his third visit in 2 months because of ear pain and bloody, crusty drainage from the ear. He is afebrile, and physical examination reveals normal tympanic membranes bilaterally, debris in the ear canals, and pain during otoscopy. You prescribe fluoroquinolone ear drops.

Of the following, the BEST subsequent course of action is

A. prohibition of swimming for the rest of the summer

B. prophylaxis with amoxicillin for the remainder of the season

C. prophylaxis with daily antibiotic ear drops

D. use of acetic acid ear drops after swimming

E. use of specially manufactured ear plugs when swimming

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Critique: 225 Preferred Response: D

Otitis externa is common in children who swim frequently, especially during the summer months. The pathogenesis is frequent dampness and maceration of the ear canal that causes a disruption in normal bacterial flora.

Children who develop otitis externa do not need to stop swimming once an infection has resolved; a variety of preventive actions are available. Acidification of the ear canal using boric acid solutions sold over the counter or 2% acetic acid solutions before and after swimming may be beneficial. Additionally, the use of earplugs during swimming has been reported to be helpful, but specially manufactured devices are not necessary; the easiest and most effective earplug is a cotton ball coated with petroleum jelly. Drying the ear canal after water recreation with a hair dryer on a low setting also may be helpful.

If prevention fails, mild otitis externa may be treated with a variety of topical agents designed to dry the ear, decrease inflammation, and eradicate organisms that cause infection at an alkaline pH. In general, a very inflamed ear canal with tenderness on movement of the pinna should be treated with a suspension that has a pH of about 5 rather than a solution that has a more acidic pH. The topical agents may contain a mixture of steroids and antibiotics, but those containing polymyxin or a fluoroquinolone cover both problematic *Pseudomonas* and *Staphylococcus* sp, two of the most common pathogens**.** Severe otitis externa, characterized by fever or regional lymphadenopathy, may require treatment with oral or parenteral antibiotics. The use of antibiotics, either systemic or topical, for prevention of otitis externa is not indicated.

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Question: 226

You receive a telephone call from the mother of one of your patients, who tells you that she is 27 weeks pregnant and that her obstetrician has diagnosed a fetal arrhythmia. In discussion with the obstetrician, you learn that the fetal heart rate is 240 beats/min and that there is a 1:1 relationship between the atrial and ventricular contraction.

Of the following, a TRUE statement about this clinical situation is that

A. atrial fibrillation is the most likely cause of the arrhythmia

B. fetal therapy will require umbilical vessel catheterization to deliver medication to the fetus

C. maternal testing for systemic lupus erythematosus should be undertaken

D. preterm delivery should be planned to begin antiarrhythmia therapy

E. the development of fetal hydrops would suggest fetal congestive heart failure

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Critique: 226 Preferred Response: E

Fetal arrhythmias are common even in healthy fetuses, but can be the source of much parental and physician anxiety. In fact, irregular heart rhythms are detected in about 1% of fetuses, most of which are due to extrasystoles and generally are of little clinical significance. Sustained fetal arrhythmias can be defined as bradycardic or tachycardic. The most common arrhythmia leading to fetal bradycardia is complete heart block, which may result from abnormalities of the fetal conduction system (especially the atrioventricular junction) in certain types of heterotaxy syndrome, atrioventricular-ventriculoarterial discordance (formerly referred to as L-transposition), or exposure to maternal antibodies to SS-A/Ro and SS-B/La antigens. The latter are seen most often in maternal autoimmune disorders such as lupus erythematosus. The atrial tachycardias, such as supraventricular tachycardia, atrial flutter, and atrial fibrillation, may result in cardiac failure that, in the fetus, often manifests as fetal hydrops. Such a nonimmune hydrops likely results from the myopathy that can occur following incessant tachycardia, can be severe, and can lead to fetal demise. When supraventricular tachycardia, the most common of the atrial tachycardias, occurs in the fetus, it frequently is at a rate of 240 beats/min or higher, and there is a direct one-to-one relationship between the atria and the ventricle in terms of conduction. Atrial fibrillation leads to an inconsistent relationship between the atria and ventricle and is decidedly rare in the fetus.

Fetal antiarrhythmic therapy should be undertaken only with an understanding of the underlying electrophysiology of the arrhythmia, fetal-maternal pharmacology, and the pharmacokinetics of the drugs being administered. All antiarrhythmic drugs can cause significant toxicity to both mother and fetus, so the risks and benefits of fetal treatment must be considered carefully before any treatment is begun. When therapy is required to control a fetal tachycardia, such as supraventricular tachycardia, it often can be accomplished with administration of the antiarrhythmic agent to the mother, which is followed by transplacental transfer to the fetus. This can be advantageous compared with other drug delivery options that involve umbilical catheterization or preterm delivery, both of which carry a risk of morbidity in addition to that associated with the arrhythmia. The fetal heart rate and atrial-ventricular relationship reported for the infant in the vignette is consistent with supraventricular tachycardia that requires close fetal monitoring and serious consideration of maternal administration of antiarrhythmic medication to control the fetal rate and rhythm and avoid fetal congestive heart failure and hydrops.

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Question: 227

A mother brings in her 4-year-old boy because she is concerned about his increasing clumsiness. He has been previously healthy and achieved developmental milestones on time. His growth parameters are normal. On physical examination, his mental status is normal, as are results of cranial nerve and sensory examinations and reflexes. However, he cannot rise from the floor without using his hands, and his running looks clumsy.

Of the following, the MOST appropriate next test to assess the cause of this child's symptoms is

A. electromyography

B. lumbar puncture

C. measurement of serum creatine kinase

D. muscle biopsy

E. spine magnetic resonance imaging

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Critique: 227 Preferred Response: C

The chronic progressive weakness described for the boy in the vignette requires a thorough, but not emergent, diagnostic evaluation to localize the problem to the brain, brainstem, spinal cord, root, nerve, neuromuscular junction, or muscle. The history of achieving developmental milestones on time, the normal growth parameters (including head circumference), and findings on examination of appropriate mental status and absence of hyperreflexia make a central nervous system problem unlikely. The normal sensory examination findings and reflexes make neuropathy less likely. The inability to arise from the floor without using his hands (Gower sign) indicate weakness of proximal muscles and suggests muscle disease.

Measurement of serum creatine kinase is an easy initial test for chronic progressive weakness that can confirm the impression of muscle disease. If this is elevated, referral to a neurologist, particularly a neuromuscular specialist, is the next recommended step for determination as to whether electromyography (EMG), muscle biopsy, or molecular diagnostic testing is warranted. A number of conditions are associated with elevated creatine kinase concentrations (Item C227).

Because the boy in the vignette has had no back pain, sensory change, or report of bowel or bladder incontinence, spine magnetic resonance imaging is not indicated. EMG may be helpful for defining intrinsic muscle disease, but because this is a painful and expensive procedure, localizing the problem more accurately to the muscle with serum creatine kinase measurement is preferred initially. Similarly, muscle biopsy ultimately may provide specific diagnostic information, but it is not preferred as the initial study. Lumbar puncture would not provide revealing diagnostic information for this clinical presentation.

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Critique: 227

Item 227B: Selected Diseases Associated With Elevated Creatine Kinase (CK)

Hereditary

- Duchenne and Becker dystrophy
- Limb girdle dystrophies (recessive and dominant)
- Acid maltase deficiency

Chronic acquired disorders

- Inflammatory myopathies
- Hypothyroidism

Acute muscle damage (Highest CK)

- Rhabdomyolysis
- Trauma
- Injection (especially phenothiazines)

Modified From: http://www.neuro.wustl.edu/neuromuscular/maltbrain.html

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Question: 228

An 18-year-old adolescent comes to your office for her precollege health supervision visit. She tells you that she is especially concerned about dating safety issues and sexual assault because a friend recently was assaulted.

Of the following, you are MOST likely to counsel her that

A. alcohol use is commonly associated with date rape

B. forceful verbal resistance is ineffective in avoiding rape

C. most assailants are not known to their victims

D. most sexual assaults take place outdoors in public places

E. sexual assailants are easy to recognize

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Critique: 228 Preferred Response: A

Dating violence is defined as “the threat or perpetration of an act of violence by at least one member of an unmarried couple on the other member in the context of dating” (McIntosh 2005). This broad definition of partner violence includes sexual assault, physical assault, and emotional abuse. By the late teen years, it is estimated that 30% to 60% of adolescents, both male and female, may have been subjected to dating violence, with as many as 25% experiencing extreme violence, such as rape or assault with a weapon. Personal risk factors for adolescent dating violence include the use of alcohol or other drugs; a previous history of physical abuse, interpersonal violence, or forced sex; minority race; history of pregnancy; partner of the same sex; and poor academic performance. A history of family violence or family sexual abuse, aggressive personality, and lack of adult supervision also are risk factors. Risk factors associated with the perpetration of dating violence are similar to risk factors for victimization by dating violence. The most significant risk factor is previous experience with violence.

Sexual assault and rape are two extreme forms of sexual violence and may present both psychological and medical emergencies. Research has shown that two thirds to three quarters of all adolescent rapes and sexual assaults are perpetrated by an acquaintance or relative of the adolescent victim. Older adolescents are more likely to become victims during social encounters (eg, a date), and younger adolescents are more likely to be victimized by a member of their extended family. It is estimated that 10% to 30% of offenders assaulting children are strangers, and about one third to one half are family members. Such intrafamilial sexual abuse may come to light in adolescence when a victim realizes that these are abnormal occurrences and seeks help. Adolescent victims are more likely than adult victims to have used alcohol or drugs but are less likely than adults to press charges and seek medical care.

There are many myths about rape and sexual assault. Some facts that may be helpful in counseling adolescents to avoid rape and sexual assault are that most sexual assault perpetrators appear normal and are young and married; they are not easy to recognize. Most rapes are planned, and the majority take place in the victim’s or perpetrator’s house. Rape occurs at all ages and with both sexes. Rape is a violent crime, with most perpetrators carrying a weapon or threatening violence. Forceful verbal resistance, physical resistance, and fleeing are associated with rape avoidance; nonforceful verbal resistance and no resistance are associated with rape. Injury rates have not been shown to be higher in women who used forceful resistance. Pediatricians should counsel both male and female adolescents regarding sexual assault prevention and help them identify high-risk situations to avoid. Screening of adolescents for sexual victimization should be part of a regular health history.

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Question: 229

A 9-month-old exclusively breastfed baby presents with a seizure. A chest radiograph obtained to rule out aspiration pneumonia reveals rachitic changes of the ribs.

Of the following, the MOST likely serum laboratory findings are

A. low calcium and elevated phosphorus

B. low calcium and low phosphorus

C. low calcium and normal phosphorus

D. normal calcium and elevated phosphorus

E. normal calcium and low phosphorus

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Critique: 229 Preferred Response: B

Exclusive breastfeeding without vitamin D supplementation after the age of 6 months is associated with vitamin D deficiency, because little of this vitamin is transferred in human milk. Mothers who are vitamin D-deficient during pregnancy may have infants who are symptomatically vitamin D-deficient before 6 months of age. Vitamin D deficiency prevents the uptake of calcium from the gut. Therefore, infants cannot absorb sufficient calcium and must release calcium from bone to maintain normal circulating calcium concentrations. Normal calcification of growing bone does not occur, and there is overgrowth of uncalcified osteoid, leading to radiologic rachitic changes, as described for the baby in the vignette. Bone resorption is enhanced by release of parathyroid hormone (PTH). Hypophosphatemia results from renal tubular phosphate losses because of elevated parathyroid hormone concentrations. If the child is exposed to a small amount of vitamin D without adequate calcium supplementation, phosphorus in the form of phosphate and calcium are driven into bone, and acute hypocalcemia with hypocalcemic seizures may develop. This disorder once was recognized in northern climates as "spring tetany." Children would manifest symptoms of hypocalcemia after their first brief exposures to sunlight in the spring of the year.

Low calcium and high phosphorus concentrations are expected in phosphate overload, hypoparathyroidism, or pseudohypoparathyroidism, but rachitic bone changes are not associated with these conditions. Normal serum calcium and low phosphorus values can be found in rickets, but this is not associated with seizure activity. Hypophosphatemia induces muscle weakness. Normal calcium and elevated phosphorus concentrations can be seen during healing rickets and in familial hyperphosphatasia or juvenile Paget disease, which is associated with bony overgrowth abnormalities that are distinctly different from rickets. Low calcium and normal phosphorus values may be seen during the healing phase of rickets but would be unusual at the initial presentation of a tetanic seizure. A low total serum calcium value also may be a sign of low total protein or albumin concentrations, and an ionized calcium value might be normal.

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Question: 230

During the health supervision visit of an infant, you place her prone on the examination table. She is able to track your penlight, following it 180 degrees by lifting her head and shoulders off the table.

Of the following, these developmental milestones are MOST typical for an infant who is

A. newborn

B. 1 month of age

C. 2 months of age

D. 4 months of age

E. 6 months of age

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Critique: 230 Preferred Response: C

To determine if an infant has delays in achieving motor milestones, the clinician must consider history, neurodevelopmental milestones, and the presence or absence of primitive reflexes and postural responses. A motor quotient (MQ) can be determined by:

MQ = motor age/chronological age x 100

An MQ greater than 70 is considered normal. An MQ between 50 and 70 is suspicious and requires monitoring. An MQ less than 50 is abnormal and requires referral to a neurodevelomental subspecialist.

The motor milestones described in the vignette are appropriate for a 2-month-old child. Appropriate milestones for a newborn include turning the head from side to side and fixating on a light in the line of vision. A 1-month-old child may lift his or her head momentarily and follow a moving object. At 2 months of age, a healthy infant in the prone position can lift his or her head and shoulders off the examination table as well as regard and follow an object 180 degrees (Item C230). A 4-month-old can lift up on his or her hands when in a prone position and may be able to roll front to back. Six-month-old children can use their hands for support in sitting, roll from back to front, and put weight on their legs while standing with assistance.

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Question: 231

You are called to the nursery to examine a newborn who has a large head. Your review of the maternal records reveals that the mother had negative findings on rapid plasma reagin, human immunodeficiency virus, and hepatitis B surface antigen testing and was rubella immune. The finding of interest in the mother's history is that she has a "bunch of cats" living with her. The male newborn has a birthweight of 2.2 kg (<5th percentile), a length of 47.5 cm (25th percentile), and a head circumference of 39 cm (97th percentile). Physical examination reveals hepatosplenomegaly and generalized adenopathy. The platelet count is 45x103/mcL (45x109/L). You suspect congenital toxoplasmosis.

Of the following, the MOST appropriate method to establish the diagnosis is

A. acute and convalescent serum immunoglobulin (Ig) G titers for *Toxoplasma gondii*

B. blood culture

C. serum IgE titers for *T gondii*

D. serum IgM titers for *T gondii*

E. urine culture

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Critique: 231 Preferred Response: D

The most appropriate method for establishing the diagnosis of congenital toxoplasmosis for the baby described in the vignette is the measurement of serum immunoglobulin M (IgM) titers for *Toxoplasma gondii*. Drawing acute and convalescent serum IgG titers against *T gondii* requires 1 to 2 months between sets of titers, which could delay the initiation of therapy. *T gondii*-specific IgA and IgE antibody tests also are very useful in establishing the diagnosis of congenital infections and infections in pregnant women, but they are not widely available at this time. Blood and urine cultures currently have no role in the diagnosis of toxoplasmosis.

Patients who have congenital toxoplasmosis are asymptomatic at birth in up to 90% of cases, but 80% to 90% of infected newborns demonstrate eye or neurologic disease or both by adulthood. The clinical manifestations of congenital toxoplasmosis in a symptomatic newborn can be indistinguishable from those of congenital cytomegalovirus (CMV) infections. The classic triad of congenital disease is unilateral or bilateral chorioretinitis (Item C231A), widespread intracerebral calcifications, and hydrocephalus. The classic manifestations of congenital CMV include unilateral or bilateral chorioretinitis (Item C231B), periventricular intracerebral calcifications, and microcephaly. Other signs and symptoms of congenital toxoplasmosis include anemia, thrombocytopenia, eosinophilia, jaundice, hepatosplenomegaly, lymphadenopathy, and seizures.

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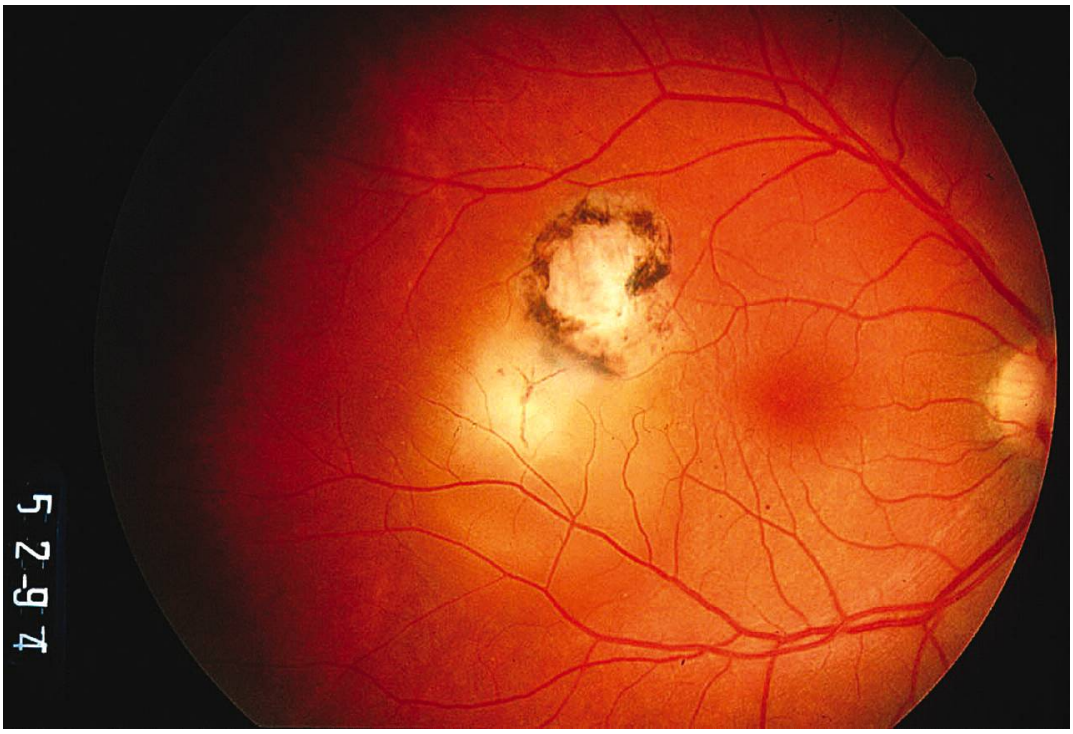
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Critique: 231



Retinitis due to toxoplasmosis: Superiorly there is an old lesion. The yellow-white area represents inflammation and edema and is surrounded by an area of hyperpigmentation. A new lesion is seen inferiorly, with inflammation and edema but no hyperpigmentation.

Courtesy of R.G. Weaver, Jr

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Critique: 231



Cytomegalovirus retinitis: White areas, often arranged along the veins, represent edema due to inflammation. Areas of early scarring (hyperpigmentation) are present.

Courtesy of R.G. Weaver, Jr

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Question: 232

You are evaluating a 4-year-old girl who has had a temperature of 102ºF (38.9°C), abdominal pain, and pain on urination. On physical examination, the child appears apprehensive and has a temperature of 101.8ºF (38.8°C), mild abdominal pain in the suprapubic area, and an erythematous urethral opening. Laboratory tests demonstrate a peripheral white blood cell count of 10.0 x103/mcL (10.0 x109/L) with 55% polymorphonuclear leukocytes, 40% lymphocytes, and 5% monocytes. Urinalysis is positive for blood and leukocyte esterase, and microscopic examination shows 5 to 10 red blood cells per high-power field (hpf) and 10 to 30 white blood cells/hpf. You send urine for culture and start therapy with oral cefuroxime axetil. Two days later, her mother telephones to report that the patient continues to be febrile and to complain of pain with urination. The urine culture is growing gram-positive cocci in chains.

Of the following, the MOST likely pathogen is

A. *Enterococcus faecalis*

B. *Escherichia coli*

C. *Pseudomonas aeruginosa*

D. *Streptococcus pneumoniae*

E. *Streptococcus pyogenes*

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Critique: 232 Preferred Response: A

Enterococci are gram-positive ovoid-shaped bacteria that are important causes of both nosocomial and community-acquired infections in children and adults. These organisms are part of the normal bowel flora of humans and many animals and are isolated frequently from environmental sources. Enterococci have low virulence, but they have emerged as an important cause of neonatal infections (eg, omphalitis, focal skin and soft-tissue infections, brain abscess, and conjunctivitis), bacteremia (especially catheter-related bacteremia), urinary tract infections, endocarditis, intra-abdominal infections (due to fecal contamination from intestinal perforation), pelvic infections, and polymicrobial wound infections. Among the less common manifestations of enterococcal infection are meningitis, septic arthritis, and primary cellulitis.

The urine culture of the patient described in the vignette is growing gram-positive cocci in chains, which suggests *E faecalis*. *Escherichia coli* and *Pseudomonas aeruginosa* are etiologic agents of urinary tract infections, but they are both gram-negative bacilli. *Streptococcus pneumoniae* and *S pyogenes* are gram-positive cocci, but they rarely cause urinary tract infections.

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Question: 233

A 4-year-old female presents with fever, chills, and vomiting. She has had abdominal pain and dysuria for 3 days. Her temperature is 104.2°F (40.1°C), and she has left-sided costovertebral angle tenderness. Laboratory evaluation reveals a white blood cell count of 18.7 x 103/mcL (18.7 x 109/L) with 85% neutrophils, 5% bands, 7% lymphocytes, and 3% monocytes. On urinalysis, the urine specific gravity is 1.025 and pH is 6.5, and there is 2+ blood, 1+ protein, 3+ leukocyte esterase, and positive nitrite. Urine microscopy demonstrates 5 to 10 red blood cells/high-power field, 50 to 100 white blood cells/high-power field, and 3+ bacteria. Findings on renal/bladder ultrasonography are normal. After a 3-day hospitalization for administration of intravenous antibiotics, discharge with a prescription for oral antibiotics is planned.

Of the following, the MOST appropriate study to complete this child's evaluation is

A. abdominal computed tomography scan

B. cystoscopy

C. intravenous pyelography

D. MAG-3 renal scan with furosemide

E. voiding cystourethrography

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Critique: 233 Preferred Response: E

The fever, chills, vomiting, abdominal pain, dysuria, and costovertebral angle tenderness described for the girl in the vignette suggest acute pyelonephritis. Laboratory evaluation demonstrates leukocytosis and abnormal urinalysis findings of hematuria, proteinuria, positive leukocyte esterase, positive nitrite, pyuria, and bacteriuria.

The initial imaging technique in children who have urinary tract infections (UTIs) is renal/bladder ultrasonography to identify anatomic abnormalities, such as hydronephrosis, renal cysts, nephrolithiasis, urolithiasis, ureteral dilatation, duplex collecting system, bladder wall thickening, and ureteroceles.

Following renal/bladder ultrasonography, a child who has a first febrile UTI should undergo voiding cystourethrography (VCUG) to screen for the presence of vesicoureteral reflux (VUR), which typically is present in approximately 30% of patients who experience their first UTI. There are two forms of cystography: fluoroscopic VCUG and radionuclide cystography (RNC). The fluoroscopic VCUG provides more detailed assessment of the reflux severity and reveals much more anatomic detail of the bladder and urethra. Therefore, males who have a first-time UTI must undergo fluoroscopic VCUG initially to rule out posterior urethral valves. Females for whom there are concerns of possible dysfunctional voiding also should undergo a fluoroscopic VCUG to assess bladder capacity and bladder emptying. Other patients and those previously found to have normal bladder and urethral anatomy by fluoroscopic VCUG may be evaluated by RNC, which is associated with reduced radiation exposure.

Abdominal computed tomography has a limited role in evaluating patients who have acute pyelonephritis. It is most useful when the child is not improving despite appropriate antibiotic treatment and concern exists for a renal abscess.

Cystoscopy (useful for stone removal or incisional treatment of a ureterocele), mercaptoacetyltriglycine (MAG-3)-Tc99 nuclear medicine renal imaging with furosemide (which may detect ureteropelvic junction obstruction), and intravenous pyelography have no role in the evaluation of a child who has a history of acute pyelonephritis. Another nuclear medicine scan, dimercaptosuccinic acid (DMSA)-Tc99, can identify regions of hypoperfusion within the renal parenchyma that occur in acute pyelonephritis or as a result of chronic renal scarring. However, its use generally is limited to those patients in whom uncertainty about the diagnosis of acute pyelonephritis exists.

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Question: 234

A 5-year-old child experiences acute hives, angioedema, and wheezing approximately 15 minutes after receiving his measles, mumps, and rubella (MMR) vaccination. He recovers completely after prompt treatment with intramuscular epinephrine, intravenous antihistamines, and corticosteroids. His parents ask what caused this reaction.

Of the following, the MOST likely cause for this child's reaction is an allergy to

A. egg

B. gelatin

C. latex

D. streptomycin

E. thimerosal

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Critique: 234 Preferred Response: B

Foods, drugs, insect stings, and vaccines are common causes of acute urticaria, angioedema, or anaphylaxis. The rapid onset of symptoms reported for the boy in the vignette is consistent with anaphylaxis to the measles, mumps, and rubella (MMR) vaccine. Most immediate vaccine reactions have been attributed to the components for the vaccine, which may include animal proteins introduced during production, antibiotics, and stabilizers.

In retrospective case-control studies, 25% of children experiencing an immediate reaction to the MMR vaccine had specific immunoglobulin (Ig) E to gelatin on skin or blood testing. Egg allergy can increase the risk for reactions to the influenza and yellow fever vaccines, but the egg content in the MMR vaccine is insignificant. Latex allergy always should be considered as a possible trigger during vaccination (eg, latex rubber in some vial stoppers, clinicians wearing latex gloves), but it is not a likely allergen in a previously healthy child.

Neither streptomycin nor thimerosal is found in the MMR vaccine. Thimerosal is used as a preservative in some multidose vaccine vials, although is not required in single-dose vials. With the exception of some influenza vaccines, thimerosal has been removed from all routine childhood vaccines. Vaccine reactions due to thimerosal usually are limited to erythema and swelling at the vaccination site or mild delayed hypersensitivity reactions.

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Question: 235

A 4-year-old child who was struck by a car is unconscious and has a shallow respiratory rate of 6 breaths/min on arrival at the emergency department. You begin bag-valve-mask ventilation and prepare to intubate the child. The respiratory therapist asks what size endotracheal tube you would like.

Of the following the CORRECT endotracheal tube is a

A. 4.5-mm cuffed

B. 5.0-mm cuffed

C. 5.5-mm cuffed

D. 5.0-mm uncuffed

E. 5.5-mm uncuffed

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Critique: 235 Preferred Response: D

As with most equipment in pediatric resuscitation, appropriate endotracheal tube (ETT) size depends on the size of the child. For infants and children, ETT size can be estimated in a number of ways. The diameter of the child's fifth finger can approximate the internal diameter (ID) of the correct ETT, although this method has been shown to be inconsistently reliable. The use of a length-based resuscitation tape can be helpful in estimating ETT size in children weighing up to 35 kg. The formula of age (yrs)/4 + 4 reliably estimates the ID of an uncuffed ETT in children up to 10 years of age; the formula of age (yrs)/4 + 3 approximates the cuffed ETT size. The appropriate ETT for the 4-year-old child described in the vignette is either a 5.0-mm uncuffed or a 4.0-mm cuffed. These sizes represent estimates, and the physician should prepare ETTs one size larger and one size smaller in case the estimate is not accurate.

The use of cuffed ETTs in infants and children has been controversial in the past because of concern that high cuff pressures could cause ischemic tracheal necrosis. Recent evidence suggests that in the hospital (pediatric intensive care unit, operating room), where cuff pressure can be measured and maintained at less than 20 mm Hg, the use of cuffed ETTs after the neonatal period is safe. In some clinical situations, such as the patient who has poor lung compliance or who requires high peak airway pressures to achieve adequate oxygenation and ventilation (eg, status asthmaticus, acute respiratory distress syndrome), cuffed ETTs must be used to avoid air leaks around the tube. In emergency and out-of-hospital settings, where cuff pressure monitoring is not feasible, the use of uncuffed tubes is preferred.

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Question: 236

You are discussing contact dermatitis due to plants (*Rhus* dermatitis) with a resident whom you are supervising.

Of the following, the statement that you are MOST likely to make is that

A. desensitization is effective for patients who have frequent recurrences

B. it is safe to handle poison ivy plants during winter months

C. patients who have severe or extensive disease should be treated with a 2-day course of prednisone

D. the rash of poison ivy may be spread by fluid from within vesicles

E. washing the skin immediately after contact with poison ivy may lessen the severity of the eruption

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Critique: 236 Preferred Response: E

A rash caused by poison ivy, oak, or sumac (*Rhus* dermatitis) is one of the most common forms of allergic contact dermatitis. Damaging any part of the plant (Item C236) liberates sap that contains urushiol, a potent antigen that penetrates the epidermis and sensitizes T lymphocytes. Re-exposure to the antigen results in a rash within 12 to 24 hours. If a person is aware that he or she has contacted poison ivy, oak, or sumac, washing the skin promptly (ideally within 10 to 15 minutes) may prevent or attenuate the rash.

Several myths surround the acquisition and spread of *Rhus* dermatitis. Although it often is stated that poison ivy may be handled safely during winter months, the sap retains its activity and can induce a rash. Many believe that fluid from vesicles and bullae can spread the rash. This may be based on the observation of new lesions continuing to appear for several days following a single exposure. However, this phenomenon occurs because areas that received the greatest concentration of resin erupt first, while those that have a lesser exposure develop the rash later. Thus, the eruption only occurs at sites that have come into contact with plant sap; once it has been removed by bathing and clothing has been changed, the rash cannot be spread.

When poison ivy dermatitis is extensive or severe or affects critical areas, treatment with an oral corticosteroid for 14 to 21 days is indicated. Discontinuing therapy earlier may result in a flare of symptoms. Although *Rhus* dermatitis can be prevented by avoiding offending plants or by the use of barrier preparations, desensitization is not recommended due to lack of efficacy, need for prolonged treatment, and potential adverse reactions.

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Critique: 236



Poison ivy typically grows in clusters of three leaflets, hence the saying "leaves of three, let them be."

Courtesy of D. Krowchuk

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Question: 237

A 3-year-old child presents with a history of intermittent painless rectal bleeding. Approximately once or twice a week, she passes a formed stool that contains up to “a teaspoon” of blood. Physical examination demonstrates no fissures or hemorrhoids. Hematocrit measurement and results of coagulation studies are normal. The bleeding persists despite stool softeners.

Of the following, the test that is MOST likely to establish a diagnosis is

A. colonoscopy

B. computed tomography scan of the abdomen

C. Meckel scan (radionuclide technetium scan)

D. magnetic resonance angiography

E. stool culture

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Critique: 237 Preferred Response: A

The patient described in the vignette has small-volume, painless rectal bleeding that persists despite stool softeners. There is no fever or signs of systemic illness to suggest an infection. The clinical presentation is more consistent with a colonic polyp (Item C237A) than with infection or Meckel diverticulum. Of the choices offered, colonoscopy is most likely to identify the polyp. A radionuclide scan (Item C237B) can help identify a Meckel diverticulum, but usually Meckel diverticulum presents with large-volume rectal bleeding. The absence of fever or cramping argues against *Salmonella* infection, which would require stool culture for diagnosis. Abdominal computed tomography scan and magnetic resonance angiography sometimes are useful in identifying bleeding gastrointestinal lesions, but they are not indicated until a polyp has been ruled out.

Rectal bleeding in a child can either be visible or occult (not seen, but detected by stool guaiac testing). Occult blood may occur or may result from causes anywhere in the gastrointestinal tract, including the esophagus (esophagitis), stomach (gastritis), small intestine, or colon. In contrast, visible maroon or bright red blood usually arises from the distal small bowel or colon.

Conditions causing lower gastrointestinal bleeding can be divided into two basic categories: those that cause bleeding in association with pain and those that result in painless rectal bleeding. Although constipation probably is the most common cause of rectal bleeding, patients who have constipation typically produce hard stools with small amounts of blood (less than 1 mL) on the surface of the stool. Hemorrhoidal bleeding usually results in blood on the toilet paper, but not on the stool. In contrast, patients who have colonic inflammation (colitis) generally have significant abdominal pain, especially around the time of defecation. The most common causes of colitis are infectious organisms (including enteric bacterial pathogens, *Clostridium difficile*, and amebae), inflammatory bowel disease, and Henoch-Schönlein purpura. In the infant, necrotizing enterocolitis, Hirschsprung disease, and allergic colitis can cause colonic inflammation.

Painless rectal bleeding generally is caused by anatomic rather than inflammatory lesions. Meckel diverticulum is an extra piece of intestine, typically located in the distal ileum, that can ulcerate and cause large-volume painless rectal bleeding. In toddlers, excessive numbers of lymph nodes in the colon (lymphoid nodular hyperplasia) sometimes may present with rectal bleeding. Colonic polyps may be either single or multiple and can be removed at colonoscopy (Item C237C). If more than one polyp is identified at the time of colonoscopy or if the histology is not typical for a juvenile polyp, the patient may need further evaluation for a hereditary polyposis syndrome.

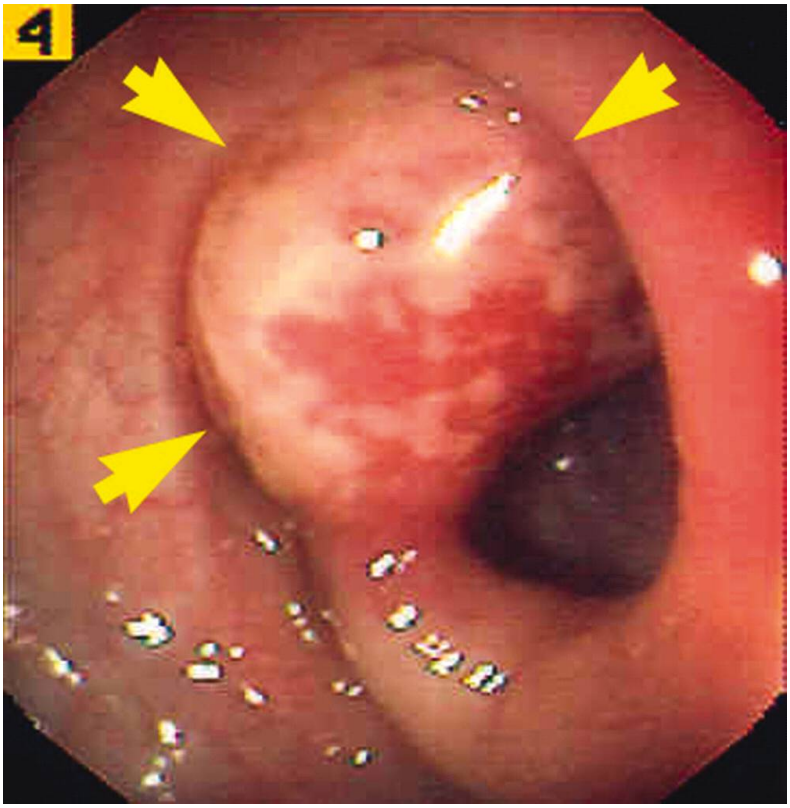
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Critique: 237

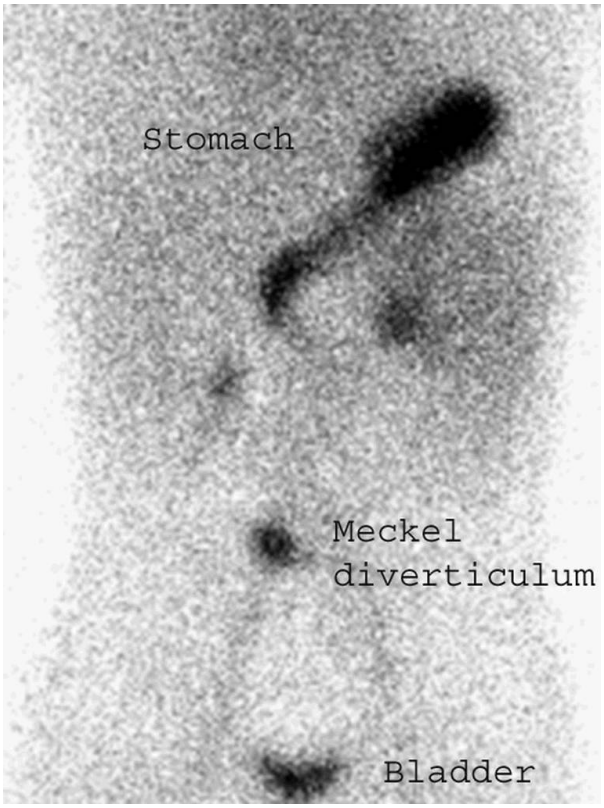


A juvenile polyp (defined by arrows) may cause rectal bleeding and can be identified on colonoscopy.

Courtesy of A. Bousvaros

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Critique: 237

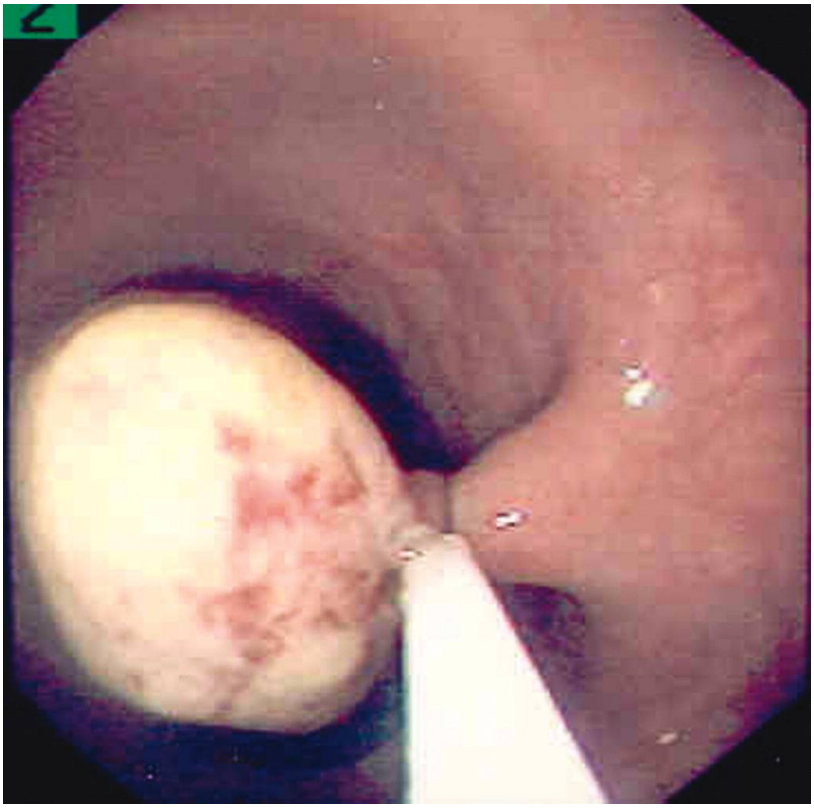


A 99m-Tc pertechnetate scan (Meckel scan) demonstrates uptake in a Meckel diverticulum.

Courtesy of L. Connelly

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Critique: 237



A juvenile polyp has been snared during endoscopy and is about to be cauterized.

Courtesy of A. Bousvaros

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Question: 238

You admit to the intensive care nursery a male infant who was born at 32 weeks' gestation and has respiratory distress. His mother did not receive antenatal steroids and did not have chorioamnionitis or premature rupture of membranes. Her group B streptococcal colonization status is unknown. On physical examination, the infant has tachypnea, retractions, and grunting respirations. There is no heart murmur. Under hood oxygen at an Fio2 of 0.50, his oxygen saturation by pulse oximetry is 85%. You obtain a chest radiograph.

Of the following, the MOST likely findings on this infant's chest radiograph are

A. air bronchograms, diffusely hazy lung fields, and low lung volume

B. cardiomegaly, hazy lung fields, and pulmonary vascular engorgement

C. fluid density in the horizontal fissure, hazy lung fields with central vascular prominence, and normal lung volume

D. gas-filled loops of bowel in the left hemithorax and opacification of the right lung field

E. patchy areas of diffuse atelectasis, focal areas of air trapping, and increased lung volumes

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Critique: 238 Preferred Response: A

The infant described in the vignette is a preterm male who is admitted to the neonatal intensive care unit with respiratory distress and a moderate supplemental oxygen requirement. His mother did not receive antepartum steroids. Features of the chest radiograph can help determine the likely underlying condition for the sick newborn and complete the cardiopulmonary examination. For this infant, the history is as important as the radiographic findings. The chest radiograph would be expected to reveal low lung volumes because the overall establishment of a functional pulmonary volume has yet to occur. Such low volumes are associated with diffuse microatelectasis that confers a diffusely hazy or "ground glass" density to the lung fields. Superimposed upon these diffuse areas of microatelectasis are air bronchograms, prominent air-filled conducting airways that extend into the periphery (Item C238A).

Cardiomegaly is not the most likely finding because congenital heart disease only occurs in 1% of all births. Pulmonary vascular engorgement is typical of pulmonary overcirculation, such as in transposition of the great vessels (Item C238B), truncus arteriosus, anomalous pulmonary venous return, or left-sided outflow tract obstructive lesions.

Fluid densities in the horizontal fissure in an adequately expanded lung that has perihilar central vascular prominence are characteristic of transient tachypnea of the newborn (Item C238C).

Gas-filled loops of bowel in the left hemithorax and opacification of the right lung field indicate a congenital diaphragmatic hernia (Item C238D). Such anomalies occur in 1 to 2 per 10,000 live births, most often in males. Ninety percent occur on the left side and pass through the foramen of Bochdalek.

Patchy areas of diffuse atelectasis, focal areas of air trapping, and increased lung volumes are characteristic of meconium aspiration (Item C238E). Meconium rarely is passed by the fetus before 34 weeks' gestation.

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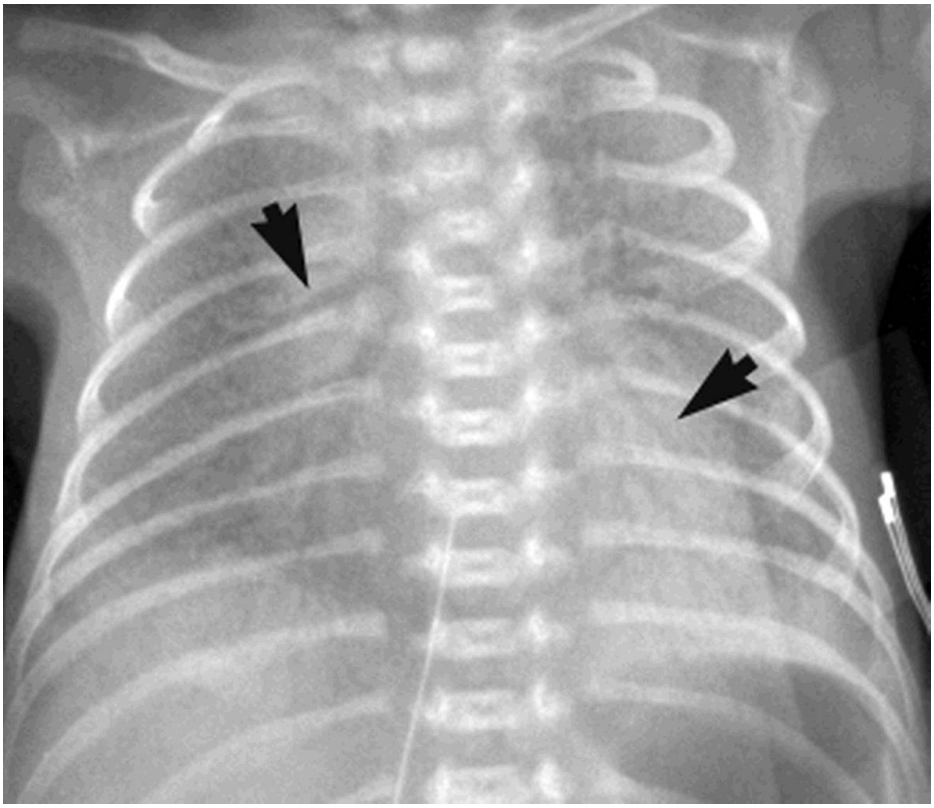
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Critique: 238

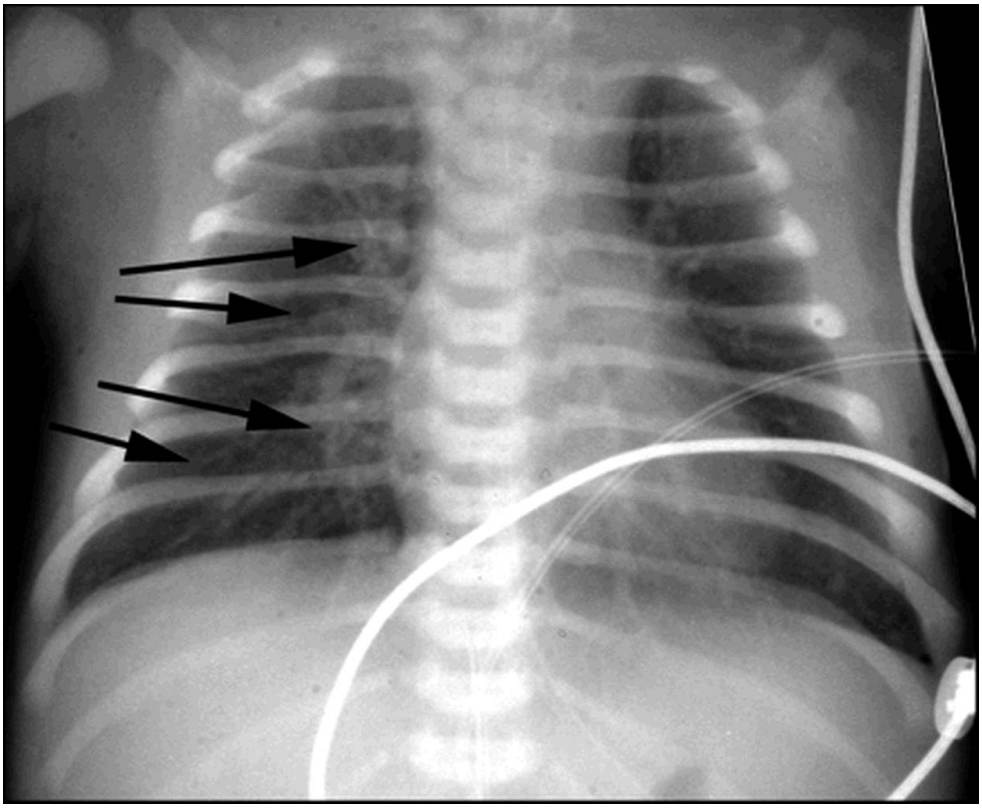


Underinflation, a "ground glass" appearance, and air bronchograms (arrows) are characteristic of respiratory distress syndrome.

Courtesy of B. Carter

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Critique: 238

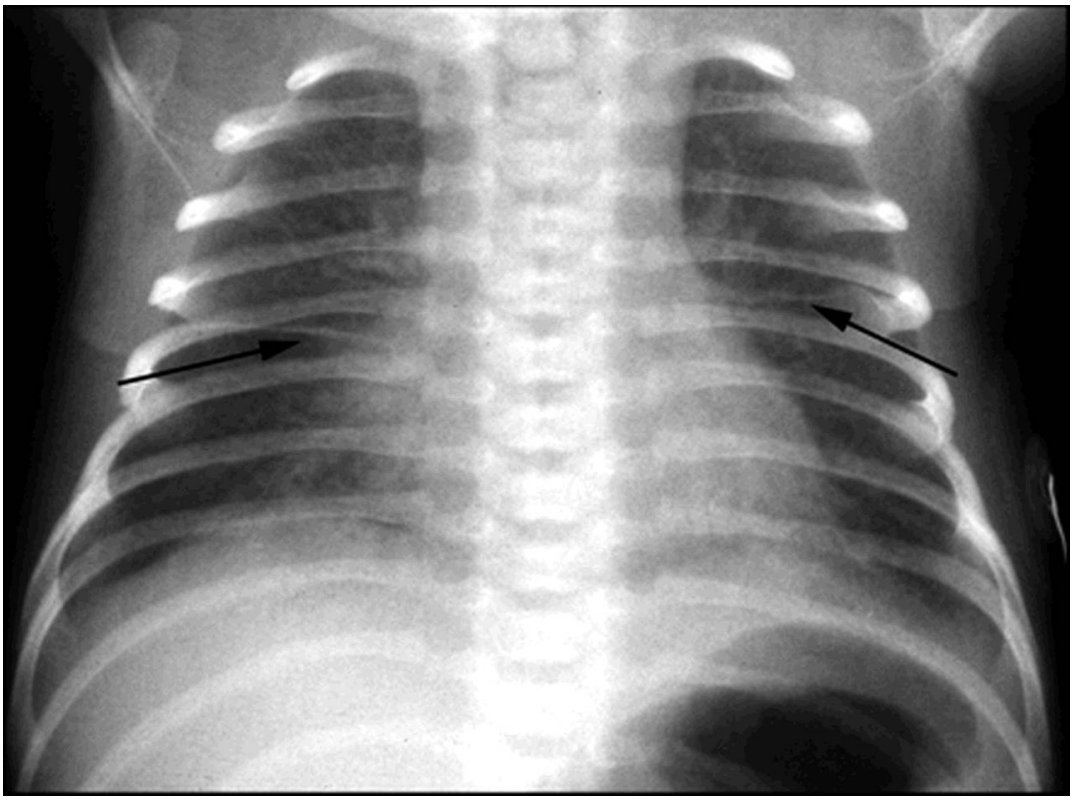


Chest radiograph in an infant who has transposition of the great vessels: Prominent pulmonary vessels (arrows) suggest pulmonary overcirculation.

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Critique: 238

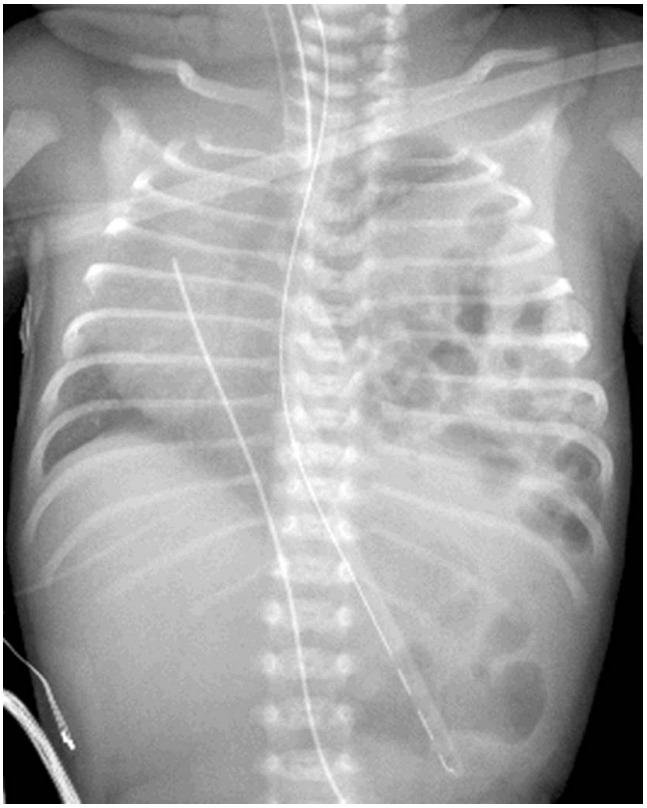


Chest radiograph in transient tachypnea of the newborn shows increased pulmonary interstitial markings and fluid in the interlobar fissures (arrows).

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Critique: 238

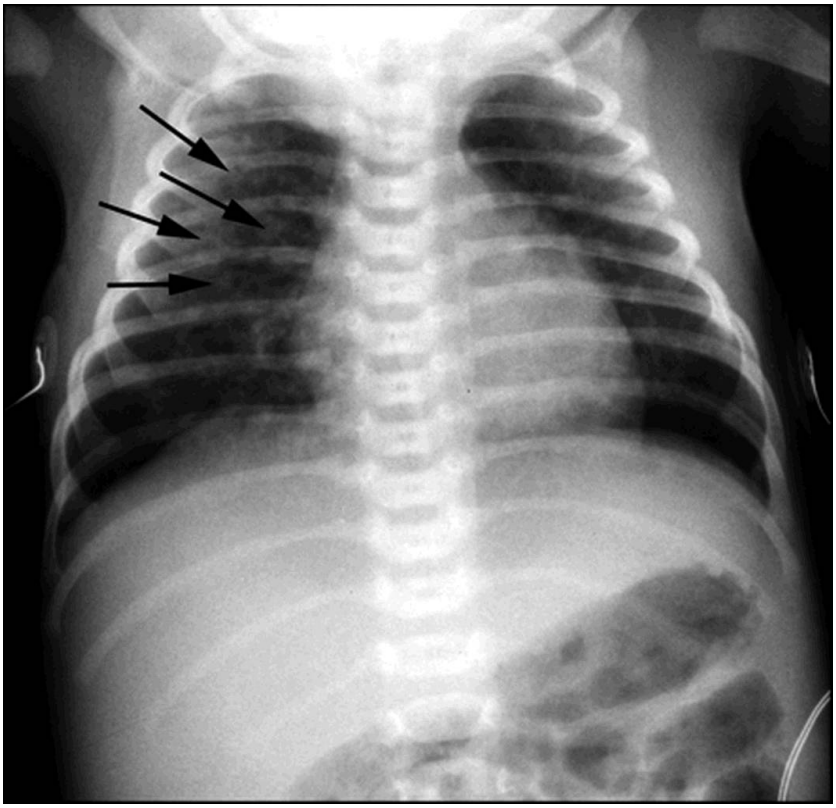


Plain radiograph of the chest and abdomen in a patient who has congenital diaphragmatic hernia. There is bowel in the left chest, with displacement of the heart to the right.

Courtesy of B. Carter

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Critique: 238



In meconium aspiration, there are areas of atelectasis (arrows) and hyperinflation (seen best at the bases).

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Question: 239

You are talking to a group of pediatric residents about the use of specific radiologic modalities in pediatric practice. You tell them that ultrasonography is used commonly because it is noninvasive and does not require sedation.

Of the following, ultrasonography is the PREFERRED diagnostic procedure for

A. Blount disease

B. Brodie abscess

C. developmental dysplasia of the hip

D. Legg-Calvé-Perthes disease

E. slipped capital femoral epiphysis

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Critique: 239 Preferred Response: C

Developmental dysplasia of the hip (DDH) occurs when the femoral head and acetabulum have an abnormal relationship. The spectrum of the abnormality includes frank dislocation, subluxation (partial dislocation), and capsular instability. DDH is more common in girls, infants who have a family history of DDH, infants born in the breech presentation, and infants born with other conditions seen with intrauterine crowding, such as congenital torticollis and clubfeet.

The diagnosis commonly is made at birth but may be delayed until after the child is walking. A thorough examination of the hips at birth, using the Ortolani and Barlow maneuvers, is critical for early diagnosis and treatment. The diagnostic procedure of choice is the physical examination (Ortolani and Barlow maneuvers). If signs are inconclusive and radiologic evaluation is considered, the best test for infants younger than 4 months of age is ultrasonography of the hips. The entire hip, including the cartilage and acetabulum, can be seen with ultrasonography, and hip stability can be assessed (Item C239A). Ultrasonography allows dynamic evaluation (Item C239B) of the hip, is noninvasive, and does not require sedation. The ability to diagnose DDH via ultrasonography improves with the experience of the technician. After 4 to 6 months of age, plain radiographs (Item C239C) of the hips are more reliable for diagnosis. Treatment consists of placing the child in a Pavlik harness to hold the hips in a position of abduction, flexion, and external rotation.

Blount disease is abnormal growth of the medial portion of the proximal tibial epiphysis and is a cause of bowing of the legs in children. Brodie abscess is a circumscribed area of osteomyelitis. Legg-Calvé-Perthes disease is idiopathic avascular necrosis of the femoral head (Item C239D), and slipped capital femoral epiphysis is displacement of the femoral head from the femoral neck (Item C239E). All of these conditions are diagnosed by plain radiographs rather than ultrasonography.

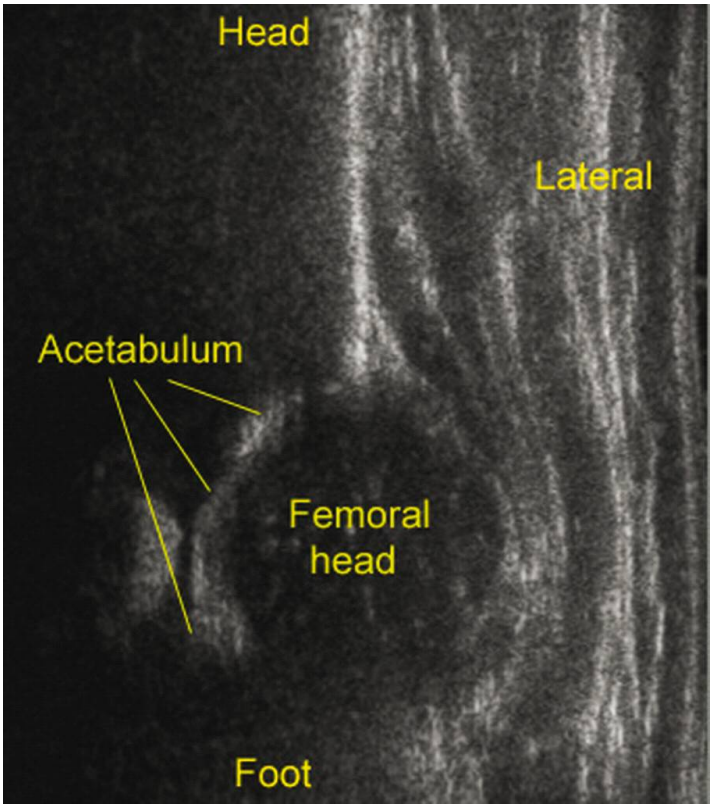
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Critique: 239

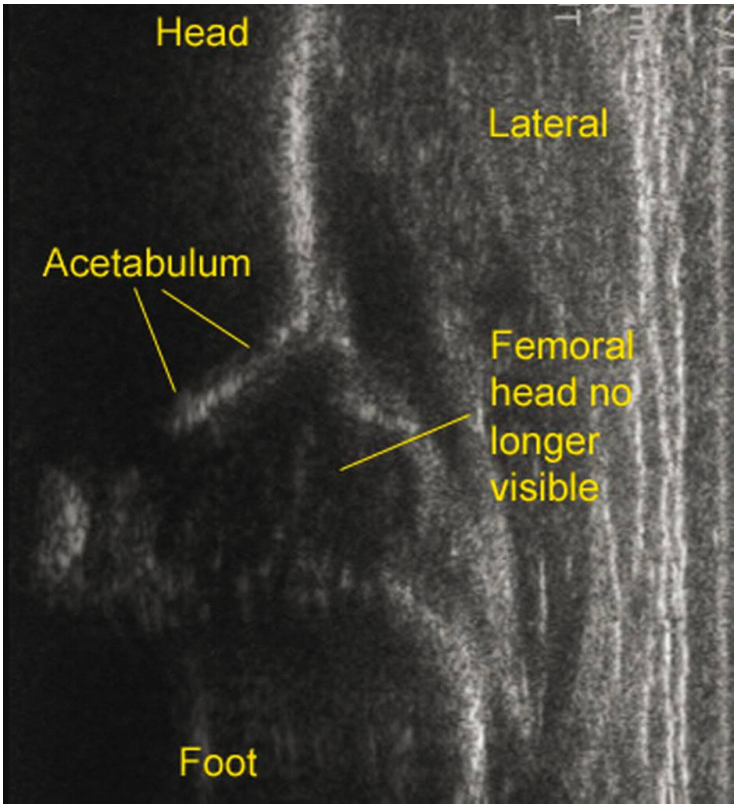


Normal hip ultrasonography findings in an infant. With the infant supine, the ultrasound transducer is placed over the lateral hip and directed medially, creating a coronal view. Normally, the femoral head is at least 50% covered by the acetabulum and cannot be moved out of the acetabulum with stress.

Courtesy of D. Mulvihill

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Critique: 239

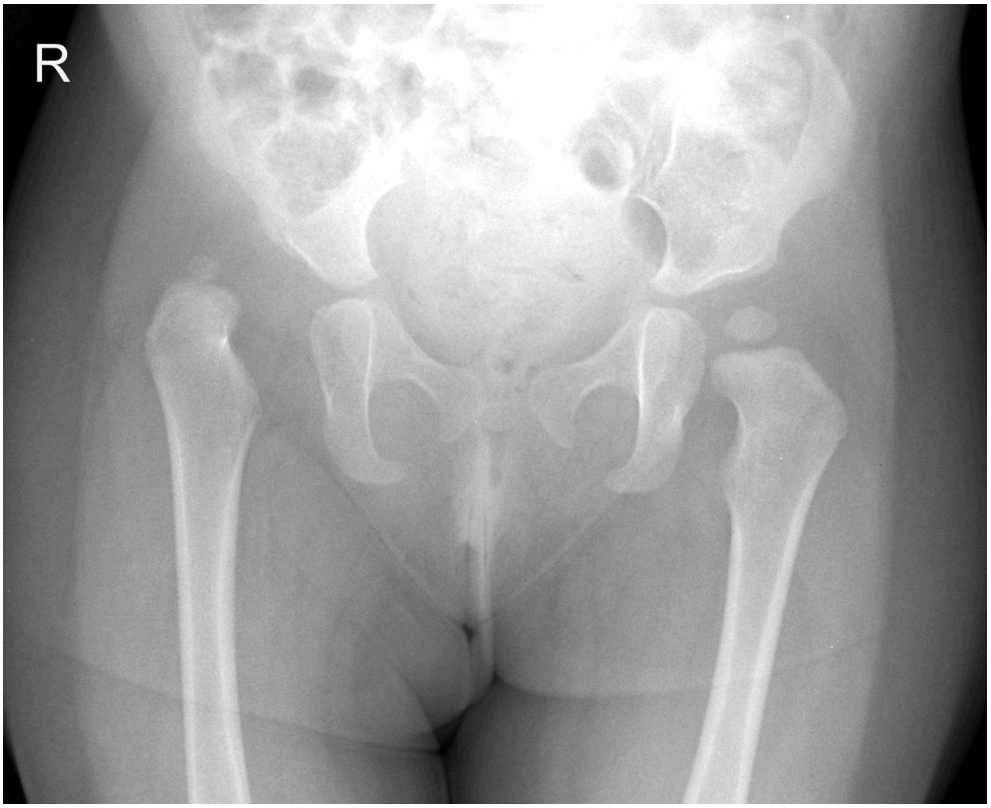


Ultrasonography of the hip in developmental dysplasia of the hip. With the knee and hip flexed, the femur is moved posteriorly (ie, Barlow maneuver). In the dislocatable hip, the femoral head disappears from ultrasonographic view.

Courtesy of D. Mulvihill

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Critique: 239

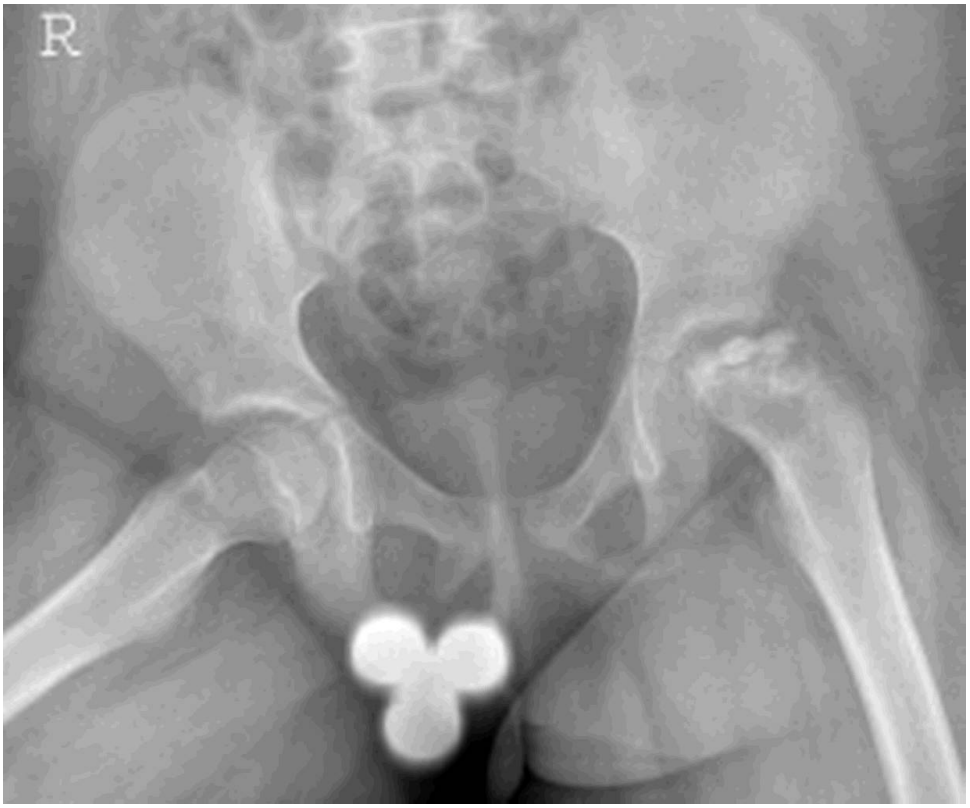


Developmental dysplasia of the hip in a 12-month-old who presented with a limp. The right acetabulum (affected side) is shallow and steep. The right hip is dislocated superolaterally.

Courtesy of D. Krowchuk

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Critique: 239

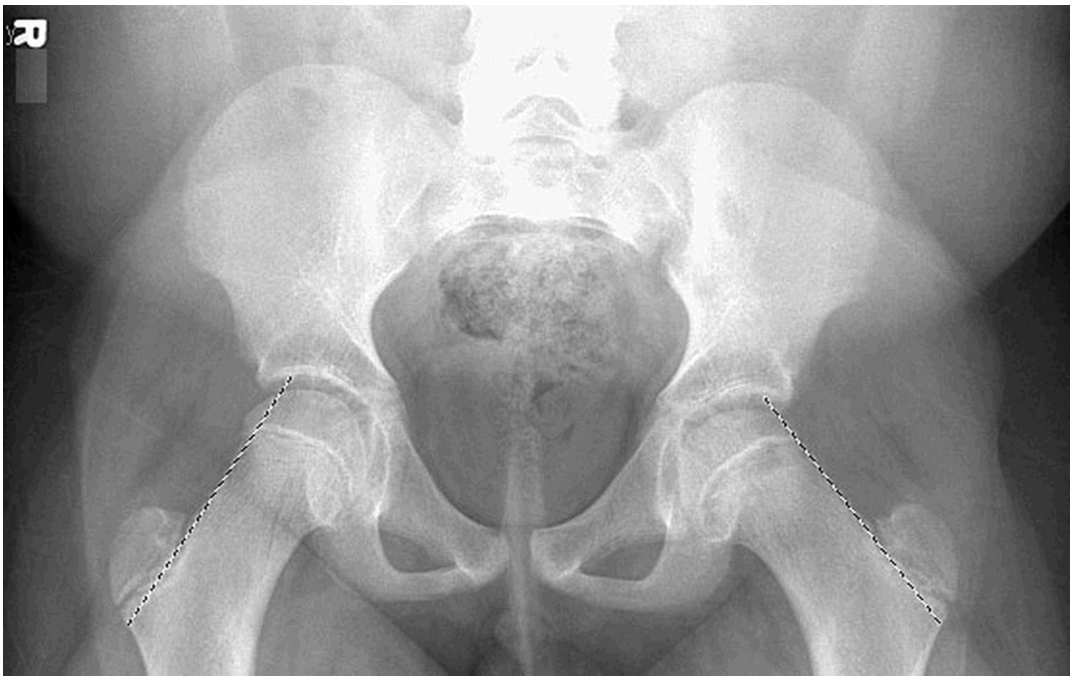


Legg-Calvé-Perthes disease: A lateral view of the hips demonstrates sclerosis around the left femoral epiphysis and flattening of the femoral head.

Courtesy of D. Krowchuk

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Critique: 239



Slipped capital femoral epiphysis consists of an upward anterior movement of the femoral neck on the capital epiphysis (which becomes displaced posteriorly and inferiorly). In the normal hip (right), a line along the superior margin of the femoral neck transects a portion of the ossified epiphysis. This does not occur on the affected side (left).

Courtesy of D. Krowchuk

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Question: 240

A 9-month-old child who had three episodes of otitis media during the winter presents for his health supervision visit. On physical examination, you note bilaterally transparent tympanic membranes with good landmarks but see bubbles behind the membranes and note no mobility on pneumatic otoscopy. The child is otherwise healthy, and results of developmental screening are appropriate for his age.

Of the following, the next BEST step is to

A. obtain auditory evoked responses

B. perform tympanocentesis

C. insert tympanostomy tubes

D. prescribe high-dose amoxicillin

E. reassess in 1 month

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Critique: 240 Preferred Response: E

Otitis media with effusion refers to the nonsuppurative presence of fluid in the middle ear, either chronically or as the subacute sequela of acute otitis media. Otitis media with effusion is diagnosed with pneumatic otoscopy; typical findings are a fluid level or bubbles behind or retraction of the tympanic membrane.

Management options for otitis media with effusion have fluctuated over the last few years, ranging from early referral for tympanostomy tube placement after more than four infections in a year to watchful waiting for spontaneous resolution. Treatment continues to be dictated by the risk factors for infection in the individual child and the risk of hearing loss and speech delay for that child.

Current evidence suggests that watchful waiting with follow-up pneumatic otoscopy over several months probably is the best approach for most children, including the child described in the vignette. Children who have underlying medical problems or increased risk for middle ear disease (eg, Down syndrome, cleft palate, immunodeficiency) may benefit from short courses of prophylactic antibiotics, although this strategy must be weighed against the risks of increased bacterial resistance.

Current evidence does not support the use of tympanostomy tubes to preserve hearing or speech development in the long term, although hearing may improve in the short term. Clearly, those children who have pre-existing speech and language difficulties may need more careful observation and audiologic assessment, such as auditory evoked response testing, and occasionally may deserve more aggressive medical or surgical attention, especially if effusions persist.

Evidence supporting therapies such as antihistamines and oral or nasal steroids is lacking. Tympanocentesis may be indicated for acute otitis media, but not otitis media with effusion (because the tympanic membrane heals rapidly and the effusion reaccumulates).

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Question: 241

You are evaluating a 15-year-old girl in your office for her annual health supervision visit. She is doing well in school and has no complaints about her health, although she would like to lose weight. She has joined the cross-country running team at school. She is not receiving any prescription medications but occasionally uses over-the-counter (OTC) cold remedies and vitamins. On physical examination, she appears thin but is in no distress. Both her height and weight are at the 25th percentile for her age, although her weight has decreased from the 50th percentile 1 year ago. Her heart rate is 100 beats/min and blood pressure is 145/95 mm Hg. Other findings on physical examination are normal.

Of the following, the MOST appropriate next step is to

A. evaluate for pheochromcytoma with blood and urine testing

B. order computed tomography scan of the abdomen

C. refer the girl for evaluation of anorexia nervosa

D. review the lists of OTC medications she has used

E. screen for use of anabolic steroids

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Critique: 241 Preferred Response: D

Hypertension may be caused by medications, including over-the-counter preparations, dietary supplements, and illicit drugs. Essential, or primary, hypertension has no identifiable cause, but secondary hypertension results from an underlying condition, disorder, drug, or other stimulus. Among the agents that can cause hypertension are corticosteroids, estrogens such as those in contraceptive therapy, migraine medications, nasal decongestants, and cyclosporine. Over-the-counter preparations that can be associated with hypertension include the many medications used for relief of cough, cold, and runny nose. Substances of abuse that can cause hypertension include alcohol, amphetamines, cocaine, and Ecstasy (MDMA and its derivatives).

The patient described in the vignette has tachycardia and hypertension, with both the systolic and diastolic pressures being higher than the 95th percentile. Other important findings in the history include a relative weight loss from the 50th percentile at age 14 to the 25th percentile (3.5 kg/7 lb) at age 15 years. It is essential to obtain more detailed information regarding the family history, past medical conditions, hospitalizations, and medication use. The patient confirms the use of over-the-counter cold remedies, some of which are known to be associated with hypertension as well as vitamins. She should be asked for further information about these substances.

Because this is her first documented episode of hypertension, neither an evaluation for pheochromocytoma nor computed tomography scan of the abdomen is warranted. Without further information regarding her dietary habits, there is no indication that she has anorexia nervosa. Similarly, findings on the history are not suggestive of the muscularization or other physical changes that can be associated with anabolic steroid use.

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Question: 242

A 16-year-old girl presents to the emergency department with the complaint of weakness for 3 weeks. She is having difficulty walking up stairs, particularly in the evening, and she has had double vision intermittently. She has no pain. On physical examination, this slim adolescent female appears sad and uninterested, with droopy eyelids. Cranial nerve examination shows slight limitation of abduction of the right eye, with complaints of double vision on the right, facial weakness, and a nasal-sounding voice. Motor examination documents 4/5 strength in the hands, shoulders, and hips. Reflexes are 1+ in arms and legs. Sensory examination results are normal.

Of the following, the MOST important initial test is

A. edrophonium chloride (Tensilon®) test

B. forced vital capacity

C. lumbar puncture

D. magnetic resonance imaging of the brain

E. measurement of serum creatine kinase

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Critique: 242 Preferred Response: B

The subacute progressive weakness and diplopia reported by the adolescent in the vignette require emergent diagnostic evaluation and management. Because subacute generalized weakness can progress rapidly to involve airway muscles or cause respiratory insufficiency and death, assessment of respiratory function is paramount, and the preferred initial procedure is measurement of forced vital capacity or negative inspiratory force. At the bedside, a rough estimate may be obtained by having the child inhale maximally and count continuously and clearly to as high a number as possible until expiration is complete. The final number is multiplied by 100 mL (eg, counting to 20 results in 2,000 mL). Forced vital capacity should exceed 20 mL/kg. A value of less than 15 mL/kg is an indication for intensive care unit admission for ventilatory support. Because pulse oximetry is not sensitive for monitoring neuromuscular respiratory insufficiency, the patient requires frequent forced vital capacity or negative inspiratory force evaluation, which is accomplished best in the intensive care unit.

The diagnostic evaluation of subacute progressive weakness should focus initially on localization of the problem to the brain, brainstem, spinal cord, root, nerve, neuromuscular junction, or muscle. The normal consciousness reported for the patient in the vignette makes a brain lesion an unlikely cause for her bilateral general weakness. The diplopia and pharyngeal weakness suggest the possibility of brainstem involvement, but fluctuating bilateral ptosis with preserved consciousness, bilateral limb weakness, and normal sensation make that less likely. Localization to the spinal cord is not probable because of the cranial nerve findings. Neuropathy is possible, although the fluctuation of symptoms, fatigability, and persistence of elicitable reflexes makes this localization unlikely. The difficulty walking up steps could point to a muscular problem. However, the fluctuations, the progression over 3 weeks without any muscle pain, the ptosis, and subtle extraocular muscle findings make muscle disease unlikely. Although adolescents may present with psychogenic weakness, this girl's physical examination findings make this diagnosis unlikely. Thus, the neuromuscular junction is the most likely localization. This clinical presentation is consistent with childhood-onset myasthenia gravis.

After respiratory issues have been assessed and managed, making the proper diagnosis is critical. Based on the clinical presentation, tests of the neuromuscular junction for myasthenia gravis, such as the Tensilon® test, performed under rigorously controlled conditions with atropine at the bedside, are important. Tensilon (edrophonium) inhibits acetylcholinesterase. Cholinergic adverse effects of edrophonium include bradycardia, which is reversed by atropine. Other adverse effects could include increased salivation and lacrimation, mild sweating, flushing, urgency, and perioral fasciculations.

Tests of central nervous system structure (brain magnetic resonance imaging) are not useful. Similarly, the yield of lumbar puncture and serum creatine kinase measurement is low.

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Question: 243

An 18-year-old young man comes to your office for his precollege health supervision visit. In screening for psychosocial risk factors, you find that he has ridden in a car with a drunk driver and driven himself after having four to five drinks.

Of the following, you are MOST likely to counsel him that

A. a blood alcohol level of 0.05% is not considered illegal for driving among those younger than 21 years of age

B. alcohol involvement in crashes peaks during daylight and nonholiday periods

C. at any blood alcohol concentration, the risk of being involved in a motor vehicle crash is higher for teen drivers than older drivers

D. teen drivers are more likely to drive after drinking than are older drivers

E. the minimum alcohol purchasing age of 21 years in all states has not been successful in reducing alcohol-related crashes among teenagers

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Critique: 243 Preferred Response: C

Alcohol use remains a significant factor in motor vehicle crashes among teenagers, both from drinking and driving themselves and riding with a driver who has been drinking. In the 2005 *National Youth Risk Behavior Survey*, 9.9% of high school youth (grades 9 through 12) reported that they drove a car or other vehicle one or more times in the 30 days preceding the survey after having consumed alcohol. In the same survey, 28.5% of high school students reported that they had ridden in a car or other vehicle in the 30 days prior to the survey that was driven by someone who had been drinking alcohol. As reported by the Insurance Institute for Highway Safety, young drivers are less likely than adults to drive after drinking alcohol, but their crash risk is higher at all levels of blood alcohol concentrations (BACs) when they do so. The elevated risk of crashes is especially valid at low and moderate BACs (ie, less than 0.08%). This increased risk is believed to be due to teenagers' relative inexperience with both drinking alcohol and driving.

All 50 states and the District of Columbia now have zero BAC thresholds for teenage drivers that were prompted by federal legislation that took effect October 1, 1998. Most states also have graduated licensing that, although differing by state, places restrictions on beginning drivers, including setting higher ages for initial licensure, requiring supervised driving, and restricting night driving and passengers in the car. Alcohol involvement in crashes peaks at nighttime and over holiday periods, making the nighttime restrictions especially appropriate. Since July 1988, all 50 states and the District of Columbia have set laws that require a minimum age of 21 years to purchase alcohol. Minimum purchasing age laws and zero tolerance laws both have been shown to be effective in reducing drinking and driving among teenagers, as evidenced by fewer nighttime fatal crashes. Research on graduated licensing also has shown a reduction in crash involvement rates for teens after programs were implemented.

Pediatricians should emphasize repeatedly to adolescents the importance of safe driving behaviors and ensure understanding of zero tolerance laws. Pediatricians also should screen adolescents routinely for risky use of alcohol and other drugs, which can cause similar impairments in driving behavior. The precollege health supervision visit is an ideal time to discuss risks of college drinking and reinforce the risks of drinking and driving as well as other drug use risks.

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Question: 244

A 3,200-g term infant has an abnormal finding on newborn screening that was obtained at 36 hours of age. The thyroxine (T4) value was 3.5 mcg/dL (45.2 nmol/L) (normal, >7 mcg/dL [90.3 nmol/L]). The thyroid-stimulating hormone (TSH) value was reported by the screening laboratory as within the normal range.

Of the following, the MOST likely reason for the low T4 value is

A. congenital hypothalamic hypothyroidism

B. congenital primary hypothyroidism

C. delayed TSH surge

D. euthyroid sick syndrome

E. thyroxine-binding globulin deficiency

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Critique: 244 Preferred Response: E

Approximately 1 in 8,000 individuals has congenital deficiency of thyroxine-binding globulin (TBG). This sex-linked disorder is expressed more completely in males. Females who are carriers have slightly lower concentrations of TBG, but they may not be identified as having a low thyroxine value on newborn thyroid screening. Low thyroxine values coupled with normal thyroid-stimulating hormone (TSH) findings on newborn screening, as reported for the infant in the vignette, represent either TBG deficiency (1:8,000) or, rarely (1:60,000), hypothalamic pituitary hypothyroidism due to deficiency of thyrotropin-releasing hormone or pituitary TSH deficiency. It is important to identify individuals who have TBG deficiency because they require no further evaluation or treatment. TBG deficiency can be diagnosed by obtaining a normal free thyroxine value, a low TBG value, or both.

Primary hypothyroidism is a disorder of thyroid gland maldevelopment or functional failure and is identified by low total thyroxine and elevated TSH values. Euthyroid sick syndrome is characterized by similar thyroxine and TSH values but is associated with severe illness or starvation in the child. TSH values usually rise in response to cold about 20 minutes after delivery, followed by a slow rise in thyroxine values. In preterm infants, the TSH surge may be somewhat delayed, but the infant in the vignette is a term healthy infant.

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Question: 245

You are examining a young boy during a health supervision visit. His mother reports that he says “mama,” “dada,” ” bye,” “up,” and “ball.” Following the examination, he sits on the floor in front of his mother while playing with a toy car. When he sees a jack-in-the-box on a shelf, he points to it. After being instructed to do so by his mother, he brings the jack-in-the-box to her.

Of the following, these developmental milestones suggest that the child is CLOSEST to

A. 12 months of age

B. 15 months of age

C. 18 months of age

D. 21 months of age

E. 24 months of age

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Critique: 245 Preferred Response: B

The milestones described in the vignette are most appropriate for a 15-month-old child. A healthy 15-month-old can say four to six words spontaneously and correctly, point to major body parts, and follow simple commands. Such children use jargon and are not distressed that no one seems to understand them. They tend to use primarily nonverbal communication to express their wants and needs. Typically developing infants of 12 months of age can understand that a particular set of sounds represents a certain object or action and may just be beginning to say their first words. An 18-month-old child may speak 10 to 15 words. A 21-month-old has a vocabulary of 30 to 50 words. At 24 months of age, a toddler may speak 100 words and begin to speak in two- to three-word phrases. A 24-month-old child can follow two-step commands.

When evaluating a child's language, cognitive, and behavior development, it is important to assess if the language development is appropriate to the cognitive development and if there are any atypical social behaviors. A developmental quotient (DQ) for language may be obtained by the equation LQ = language age/chronologic age x 100. Children with a language DQ of less than 70 should be referred for a speech language evaluation.

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Question: 246

A 2-week-old girl presents to the clinic with a history of drainage from her right eye. Her mother states that she noticed “the white part of her eye” turning red yesterday, but today the eye was redder and swollen with some drainage. The infant was born at term via normal spontaneous vaginal delivery without complications and is the mother’s second child. The mother denies any illness or sexually transmitted disease during her pregnancy, but states that she did smoke one quarter pack of cigarettes per day. Physical examination reveals an afebrile, healthy-appearing female whose only abnormality involves her right eye. The upper and lower eyelids are slightly swollen, and her conjunctiva is erythematous, with a nonpurulent-appearing discharge. You suspect she has neonatal conjunctivitis due to *Chlamydia trachomatis*.

Of the following, the MOST sensitive method for establishing the diagnosis is

A. conjunctival culture

B. detection of eosinophilia on a complete blood count

C. Giemsa staining of conjunctival scrapings

D. nucleic acid amplification test of conjunctival cells

E. serum immunoglobulin G testing for *C trachomatis*

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Critique: 246 Preferred Response: D

Chlamydia should be considered for all infants who are 30 days of age and younger and have conjunctivitis. Because *Chlamydia trachomatis* is an obligate intracellular organism, recovery by culture can be difficult. Nucleic acid amplification tests, such as polymerase chain reaction, are more sensitive than cell culture or serum immunoglobulin G testing. Laboratory governing bodies monitor clinical laboratories using these techniques closely because most nonculture tests are not approved by the United States Food and Drug Administration for the detection of chlamydia from conjunctival swabs. A correctly obtained conjunctival specimen must contain cells, not exudate alone. Specimens should be obtained from the everted eyelid using a Dacron®-tipped swab or the swab specified by the manufacturer's test kit.

Patients who have chlamydia conjunctivitis should be treated with oral erythromycin, which not only treats the conjunctival disease, but helps to prevent later chlamydial pneumonitis. Eosinophilia can be found in up to 20% of patients who have chlamydial pneumonia, but the finding is not helpful in chlamydial conjunctival disease. Giemsa stains do not aid in diagnosing chlamydial conjunctivitis.

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Question: 247

You are evaluating a previously healthy 3-year-old girl for white spots in her mouth and a worsening rash in her vaginal area. Her mother states that except for decreased oral intake and complaints of itchiness in her vaginal area, the child has had no fever or other systemic symptoms. She has received three different 10-day courses of oral antibiotics in the last 2 months for a throat infection and otitis media. She completed her last antibiotic course yesterday. Physical examination shows two small areas of whitish plaques (Item Q247) on her tongue and right buccal mucosa that cannot be removed easily with a tongue blade. She has no abdominal tenderness. Her vaginal area is erythematous, with several areas of excoriation; her hymenal tissue and urethral opening appear normal. The remainder of her physical examination findings are normal.

Of the following, the MOST likely cause of this patient's condition is

A. human immunodeficiency virus

B. repeated courses of antibiotics

C. sexual abuse

D. undiagnosed immunodeficiency

E. viral infection

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Question: 247



White plaques, as described for the girl in the vignette.

Courtesy of D. Krowchuk

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Critique: 247 Preferred Response: B

Candida albicans is a normal commensal organism in humans that commonly is found on skin, throughout the entire gastrointestinal (GI) tract, in the female genital tract, and in the urine of patients who have indwelling Foley catheters. It also is found in the environment (eg, soil, animals, hospitals, inanimate objects, and food). An interruption in normal host defense mechanisms is necessary for this human commensal organism to become a pathogen. This may occur either naturally, in association with underlying disease (eg, diabetes mellitus), or iatrogenically. Widespread and repeated use of antibiotics that suppress normal bacterial flora can allow *Candida* organisms to proliferate, especially in the GI tract. Certain classes of antibiotics potentiate proliferation of the organism by decreasing neutrophil *Candida* intracellular killing (sulfonamides) or by decreasing neutrophil phagocytosis (tetracyclines, aminoglycosides). Additionally, intravenous drug abuse and the widespread use of indwelling intravenous catheters for any purpose (eg, hyperalimentation, dialysis, or pressure monitoring) can provide a route for *Candida* to enter from the environment into the vascular system of susceptible patients. Implantation of prosthetic materials, especially cardiac valves, can interrupt normal defense mechanisms. Finally, clinical situations associated with general immune suppression (eg, multiple abdominal surgeries, transplantation, neoplastic diseases, use of steroids, and severe burns) can allow transformation of the candidal organisms.

The repeated courses of antibiotic described for the girl in the vignette most likely have created the conditions for *Candida* organisms to become pathogens. The findings on physical examination are not consistent with a viral infection or sexual abuse. Her history of being previously healthy makes a diagnosis of human immunodeficiency virus infection and an undiagnosed immunodeficiency unlikely.

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Question: 248

A mother brings in her 4-year-old son because his eyelids are swollen. On physical examination, the boy has normal growth parameters, normal blood pressure, bilateral periorbital edema (Item Q248), and pitting pretibial edema. Laboratory findings include normal electrolyte concentrations, blood urea nitrogen of 14 mg/dL (5 mmol/L), creatinine of 0.3 mg/dL (26.5 mcmol/L), albumin of 1.9 g/dL (19 g/L), and normal C3 and C4 complement values. Urinalysis reveals a specific gravity of 1.030, pH of 6.5, 4+ protein, and 1+ blood, and microscopy demonstrates 5 to 10 red blood cells/high-power field. Antinuclear antibody test results are negative, and serologic tests are negative for hepatitis B surface antigen, negative for hepatitis C, and nonreactive for human immunodeficiency virus. A purified protein derivative test is nonreactive after 48 hours.

Of the following, the MOST appropriate treatment for this patient is:

A. diphenhydramine

B. furosemide

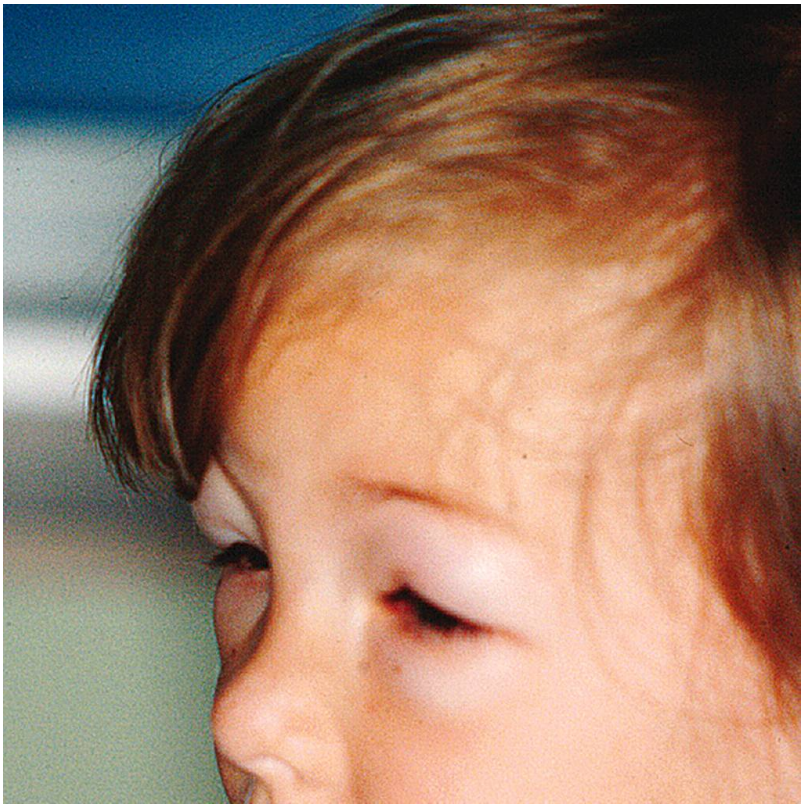
C. low-sodium diet

D. prednisone

E. protein-rich diet

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Question: 248



Periorbital edema, as described for the boy in the vignette.

Courtesy of M. Rimsza

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Critique: 248 Preferred Response: D

The boy described in the vignette has new-onset nephrotic syndrome (NS). The first important step is to establish the diagnosis based on the presence of severe proteinuria, hypoalbuminemia, and edema. Next, the practitioner must note the child's blood pressure (BP) and assess renal function by measuring a serum creatinine because mild degrees of BP elevation and mild azotemia are consistent with NS, but marked elevations of BP or creatinine are not. In addition, approximately 20% of children presenting with new-onset NS have microscopic hematuria; gross hematuria should not be present.

The next phase of the evaluation involves screening for secondary causes of NS. Typically, a serologic evaluation includes measurement of complement components (C3 and C4), antinuclear antibody, anti-double-stranded DNA, hepatitis B surface antigen and core antibody, hepatitis C antibody, and human immunodeficiency virus antibody. A complete blood count is obtained to look for hematologic abnormalities associated with NS, such as leukemia/lymphoma or sickle cell disease. A purified protein derivative test is placed to look for occult tuberculosis infection prior to starting treatment. If no secondary cause of the NS is identified by these tests, the treatment of choice is oral prednisone beginning at 60 mg/m2 per day in divided doses (maximum dose, 40 mg twice per day). The duration of daily therapy is 4 to 6 weeks, after which time the dose is reduced to 40 mg/m2 (maximum daily dose, 60 mg) on alternate days for 4 to 6 weeks. Recent studies suggest that a longer course (6 weeks daily followed by 6 weeks alternate-day) results in a higher sustained remission rate than a shorter course (4 weeks daily followed by 4 weeks alternate-day).

Although a low-sodium diet is recommended for children who have new-onset NS, it is adjunctive, not definitive therapy. Diuretics such as furosemide are not recommended in patients who have NS due to an increased risk of thrombosis. A protein-rich diet has no role in the treatment of NS because the hypoalbuminemia is due to glomerular losses, not malnutrition. Although allergies can cause periorbital swelling and may be treated with an antihistamine such as diphenydramine, patients typically exhibit other symptoms, such as rhinitis, sneezing, or conjunctival injection. If uncertainty exists as to the cause of periorbital swelling and there is concern about possible NS, performance of a urinalysis to detect proteinuria may be useful.

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Question: 249

You are evaluating a 14-year-old girl for seasonal allergic rhinitis. Despite a regimen of multiple allergy medications, she continues to have significant sneezing, rhinorrhea, and nasal congestion. You decide to consult an allergist for further evaluation, specifically aeroallergen skin testing and consideration for allergy shots.

Of the following, the MOST likely medication to alter the results of aeroallergen skin testing is a(n)

A. corticosteroid nasal spray

B. inhaled beta-2 agonist

C. low-dose inhaled corticosteroid

D. oral histamine-1 antihistamine

E. oral leukotriene antagonist

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Critique: 249 Preferred Response: D

When determining the impact of indoor and outdoor aeroallergens as triggers for allergic rhinitis, one of two diagnostic tools generally is used: skin testing or blood testing. Aeroallergen skin testing involves the application of specific allergens (eg, oak, Bermuda grass, cat, ragweed) on the skin using a prick or puncture method. Allergy skin testing is tolerated by most children and adolescents extremely well. The advantage of skin testing is that a broad array of allergens can be tested, and results are immediately evident to the patient. One disadvantage is that the wheal-and-flare response seen in positive responses can be suppressed with certain medications, particularly oral histamine-1 receptor antihistamines. Therefore, patients usually are asked to stop this medication class 1 week prior to skin testing. Other medications that can suppress immunoglobulin (Ig) E-mediated skin tests include tricyclic antidepressants, chronic oral steroids, histamine-2 antihistamines, topical corticosteroid creams, and epinephrine. Corticosteroid nasal spray, inhaled beta-2 agonists, low-dose inhaled corticosteroids, and oral leukotriene antagonists have not been shown to suppress aeroallergen IgE-mediated skin test responses.

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Question: 250

A mother brings her 2-year-old son to the office 30 minutes after spilling a cup of hot coffee onto his arm and chest. Physical examination reveals a 2x3-cm ruptured blister on his chest that has an erythematous, tender base and a 3x5-cm area of erythema on his right upper arm.

Of the following, a TRUE statement regarding the management of the burns is that

A. the burns should be cleaned with soap and water after debridement of the blistered area

B. the patient should be given a 5-day course of prophylactic cephalexin

C. the patient should be referred to a burn center because of the extent of his burns

D. the patient will require skin grafting for the burn on his chest

E. the burn on his upper arm should be dressed with bacitracin ointment and gauze

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Critique: 250 Preferred Response: A

Most burn injuries in children are minor and can be managed on an outpatient basis. Decisions regarding appropriateness of outpatient treatment are based on several criteria, including the depth (or degree) of the burn, the extent of the body surface area (BSA) involved, the causative mechanism, and the location of the injury. According to guidelines developed by the American Burn Association, pediatric patients should be referred to a burn center for specialized care for partial-thickness burns covering greater than 10% of the total BSA; burns that involve the face, hands, feet, genitalia, perineum, and major joints; full-thickness burns; and burns caused by chemicals or electricity.

Superficial (first-degree) burns involve injury to the epidermis only and are characterized by intact, erythematous, and painful skin. Partial-thickness (second-degree) burns (Item C250) injure both the epidermis and the dermis and often are characterized further as superficial partial thickness if they involve only superficial layers of the dermis or deep partial thickness when deeper dermal layers are damaged. Superficial partial-thickness burns are erythematous and painful, with blister formation. When the blisters rupture, the base of the burn is moist and bright red. Deep partial-thickness burns also are painful and erythematous, but the blister base may be pale and relatively less painful (but not anesthetic) than in a superficial partial-thickness burn. Full-thickness (third-degree) burns injure the epidermis and dermis and are dry, leathery, and insensate in the central portions of the burn where sensory nerves have been destroyed.

The percentage of BSA involvement can be estimated using the “rule of nines” modified for infants and children. This system assigns BSA percentages for various body parts and accounts for the relative BSA changes that occur during growth from infancy to adulthood. Measuring the burn area with the patient’s palm (including the fingers), which is approximately 1% of the BSA, also can be used to estimate the burn extent.

Treatment of superficial burns, such as that on the arm of the boy described in the vignette, involves analgesia and local cleansing. Topical antibacterial therapy is not indicated for these injuries because the intact epidermis still provides an adequate barrier. Partial-thickness burns, as described on the chest of the boy in the vignette, should be cleaned with mild soap and water after broken blisters are debrided, then covered with an antibacterial ointment such as bacitracin or silver sulfadiazine and a nonadherent dressing. A physician who has expertise in burn care should evaluate full-thickness burns, which often require excision and grafting of the wound in addition to topical therapy, as for partial-thickness burns. All burns should be evaluated daily for the first 48 hours because the ultimate depth and extent may not be evident until this time. The use of prophylactic systemic antibiotics is not indicated. The patient’s tetanus immunization status should be assessed and updated if indicated.

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Critique: 250



Partial-thickness burns, such as that shown here caused by hot water, involve the epidermis and dermis.

Courtesy of M. Rimsza

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