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Vertigo and Dizziness

Common Complaints

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Preface

There are three convincing arguments why it is important to learn about the management of vertigo:

- After headache, it is the second most common complaint of patients, not only in neurology and ENT departments.
- Most syndromes of vertigo can be correctly diagnosed only by means of a careful medical history and physical examination of the patient.
- The majority of these cases have a benign cause, take a favourable natural course, and respond positively to therapy.

Vertigo and dizziness are not disease entities, but rather unspecific syndromes consisting of various disorders with different causes. For this reason, our clinically oriented book is for physicians of different specialisations who treat patients with vertigo and for medical students. To make the book easy to use, we have provided an overview of the most important syndromes of vertigo and dizziness, each with elucidating clinical descriptions and illustrations.

A general chapter deals with how the vestibular system functions, its disorders, the pathophysiological mechanisms involved, diagnostic signs, history taking, examination procedures, laboratory diagnostics and principles of therapy. The most important clinical syndromes of vertigo are treated in individual chapters organised as follows: patient medical history, clinical aspects and natural course, pathophysiology and principles of therapy, pragmatic therapy, ineffective treatments, as well as differential diagnosis and clinical problems. We have put special emphasis on the various drug, physical, operative or psychotherapeutic treatments available. The book is based on the common experience that we have accumulated over many years working in a multi-

regional referral center for dizziness outpatients. Many parts of the text, tables and figures are updated versions of those in a considerably more detailed monograph on the clinical and scientific aspects of vertigo (Brandt T. *Vertigo: its multisensory syndromes*, 2nd ed. Springer, London, 1999). The accompanying DVD presents typical case histories, results of examinations for the individual syndromes, physical examination techniques and laboratory diagnostics. The book is oriented to daily medical practice, and we hope that it will prove helpful by providing readily accessible information. The whole field of vertigo and dizziness, imbalance and eye movement disorders has been considered extremely difficult because of the variety of its manifestations and its resistance to compartmentalisation. We hope that we have succeeded in making these syndromes more understandable by using clear, anatomical categories and clinical classifications.

We would especially like to express our thanks to the neuro-orthoptists Miriam Glaser, Cornelia Karch and Nicole Rettinger for compiling the videos. Our appreciation also to Ms Judy Benson for copyediting the text and to Dr. Steven Russell for carefully reading the manuscript. We also thank Ms Sabine Eßer for designing the graphics and Ms Melissa Morton and Eva Senior of Springer-Verlag London for cooperating on the production of this book in such a pleasant and efficient manner. The German edition of the book, "Vertigo-Leitsymptom Schwindel" by T. Brandt, M. Dieterich and M. Strupp, was published by Steinkopff-Verlag in 2004.

Contents

1

Introductory Remarks

1.1 Vertigo and Dizziness: Multisensory Syndromes

Vertigo and dizziness are not unique disease entities. Sometimes vertigo is attributed to vestibular disorders, while dizziness is not (Neuhäuser and Lempert 2004). There is, however, no general agreement, and visual stimuli can cause vertigo (e.g., height vertigo or optokinetic vection), just as central vestibular or otolith disorders can cause dizziness. The two terms cover a number of multisensory and sensorimotor syndromes of various aetiologies and pathogeneses, which can be elucidated only within an interdisciplinary approach. After headache, vertigo and dizziness are among the most frequent presenting symptoms, not only in neurology. According to a survey of over 30,000 persons, the prevalence of vertigo as a function of age is around 17%; it rises to 39% in those over 80 years of age (Davis and Moorjani 2003). Whether caused by physiological stimulation (motion sickness, height vertigo) or a lesion (unilateral labyrinthine failure, central vestibular pathway lesions), the resulting vertigo syndrome characteristically exhibits similar signs and symptoms despite the different pathomechanisms—dizziness/vertigo, nausea, nystagmus and ataxia (Figure 1.1). Disorders of perception (dizziness/vertigo), gaze stabilisation (nystagmus), postural control (falling tendency, ataxia) and the vegetative system (nausea) are related to the main functions of the vestibular system, which are located in different sites in the brain.

The most important anatomical structure of the vestibular system is the vestibulo-ocular reflex (VOR). The VOR has three major planes of action:

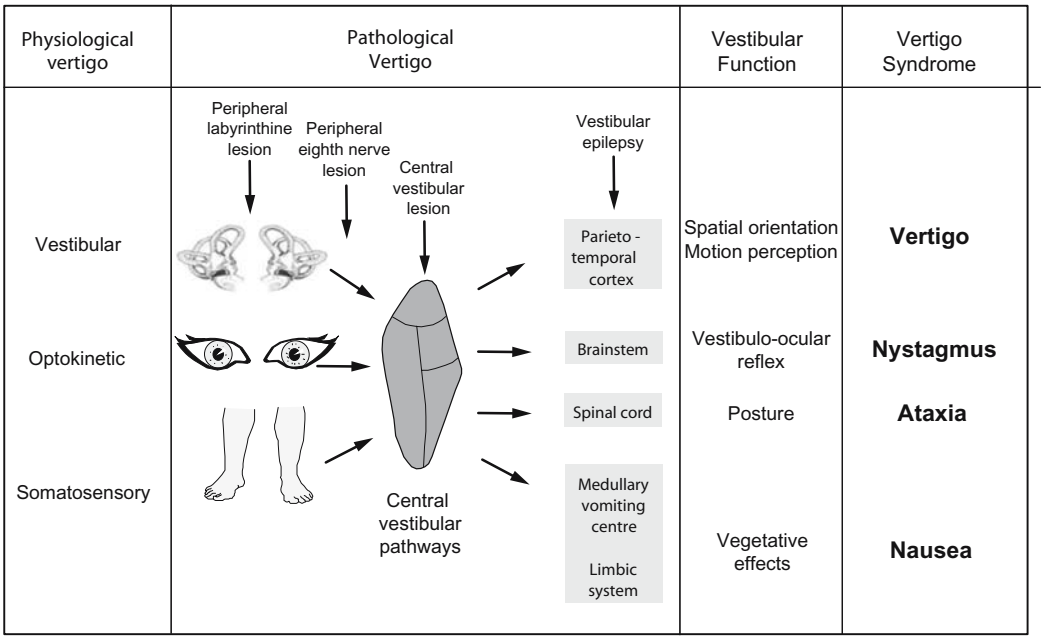


Figure 1.1. Physiological vertigo (motion stimulation) and pathological vertigo (induced by lesion or stimuli) are characterised by similar signs and symptoms that derive from the functions of the multisensory vestibular system (Brandt and Daroff 1980)

- horizontal head rotation about the vertical z-axis (yaw)
- head extension and flexion about the horizontal y-axis (pitch)
- lateral head tilt about the horizontal x-axis (roll).

These three planes represent the three-dimensional (3-D) space in which the vestibular and ocular motor systems responsible for spatial orientation, perception of self-movement, stabilisation of gaze and postural control operate. The neuronal circuitry of the horizontal and vertical semicircular canals as well as the otoliths is based on a sensory convergence that takes place within the VOR (Figure 1.2). The VOR connects a set of extraocular eye muscles that are aligned by their primary direction of pull with the same particular spatial plane of the horizontal, the anterior or the posterior canal. The canals of both labyrinths form functional pairs in the horizontal and vertical working planes. In other words, the canals are excited or inhibited pairwise: the horizontal right and left pair; the vertical anterior of one side along with the posterior canal of the opposite side. The vertical planes of "pitch" and "roll" are a result of the wiring connecting the two vertical canals that are diagonal to the sagittal plane in the head. The pairs of canals function as a gauge of rotatory acceleration and react to the rotational movements of the head in the corre-

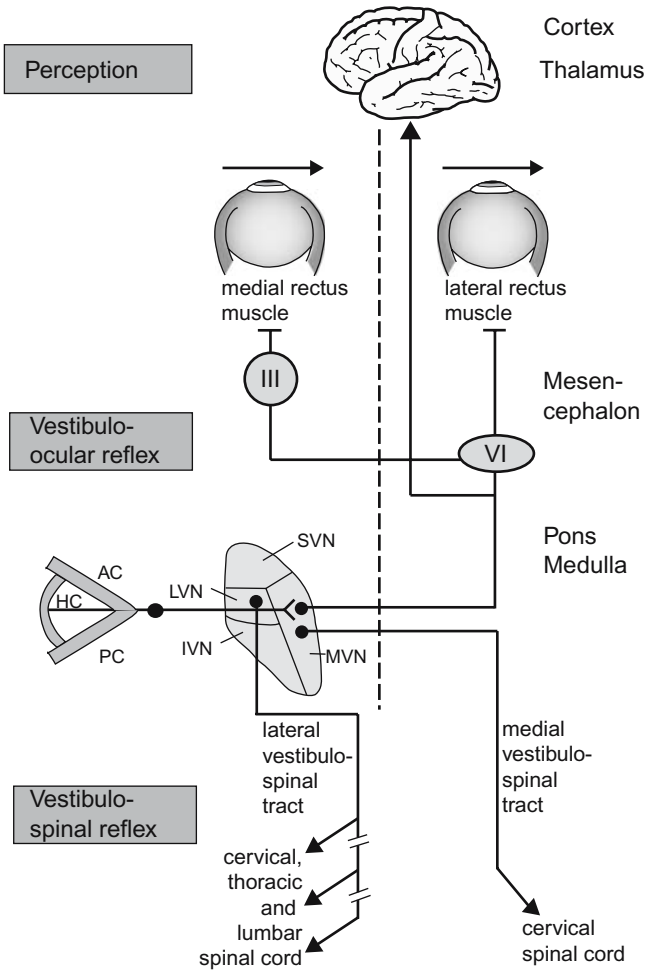


Figure 1.2. Schematic drawing of the horizontal vestibulo-ocular reflex (VOR). The VOR is a part of a complex sensorimotor system, which makes possible perception of head position and motion (connections via the thalamus to the vestibular cortex), gaze stability (three-neuron arc to the nuclei of the ocular muscles), as well as head and postural control (vestibulospinal reflexes). *AC*, *HC*, *PC* anterior, horizontal and posterior semicircular canals; *SVN*, *LVN*, *IVN*, *MVN* superior, lateral, inferior and medial vestibular nuclei; *III*, *VI*, oculomotor and abducens nuclei.

sponding plane. The otoliths function as a gauge of gravity and linear acceleration.

The most frequent forms of peripheral vestibular vertigo are benign paroxysmal positioning vertigo (BPPV), vestibular neuritis and Menière's disease. The neurovascular compression syndrome of the VIIIth cranial nerve (vestibular paroxysmia), bilateral vestibulopathy and perilymph fistulas occur more

rarely. Peripheral vestibular attacks, which affect semicircular canal function, are characterised by a strong rotatory vertigo and spontaneous nystagmus in one direction, a tendency to fall in the other direction, nausea and vomiting.

Central vestibular forms of vertigo arise from lesions at the neuronal circuitry between the vestibular nuclei and the vestibulocerebellum, as well as those between the vestibular nuclei, the vestibular and ocular motor structures of the brainstem, cerebellum, thalamus and vestibular cortex. On the one hand, these are clearly defined clinical syndromes of various aetiologies, for example, upbeat or downbeat nystagmus (the quick phase of nystagmus beats upward or downward). The occurrence of these typical ocular motor findings in only the central brainstem or cerebellar disorders allows their definitive localization. On the other hand, central vestibular vertigo can also be a part of a more complex infratentorial clinical syndrome. Then other signs and symptoms such as supranuclear or nuclear ocular motor disorders and/or other neurological brainstem signs (e.g., Wallenberg's syndrome) can also be observed. Central forms of vertigo can manifest as attacks lasting for seconds or minutes (basilar/vestibular migraine), for hours up to days (brainstem infarction), or as a permanent symptom (downbeat nystagmus in cases of Arnold–Chiari malformation).

Our first overview of the most frequent forms of vertigo/dizziness was made in an unselected group of in- and outpatients in the early 1980s. It showed that BPPV, vestibular neuritis, phobic postural vertigo and Menière's disease accounted for 85% of cases of vertigo; their respective frequency is according to the order cited. After the establishment of a multiregional neurological unit for vertigo and dizziness, a second overview was made for the years 1989 to 2003 (for 4,790 patients). It revealed that the whole spectrum as well as the relative frequency of the individual diagnoses had shifted (Table 1.1).

Benign paroxysmal positioning vertigo was still the most frequent cause, occurring in 18.3% of the patients. The second most frequent diagnosis was phobic postural vertigo (15.9%), followed by central forms of vestibular vertigo in vascular and inflammatory diseases (lacunar infarcts or multiple sclerosis [MS] plaques) of the brainstem or the cerebellum. Basilar/vestibular migraine has two frequency peaks: one in the second decade and another in the sixth decade, thus it is by no means a disease of only younger women. Now it is the fourth most frequent cause, followed by vestibular neuritis and Menière's disease.

It is difficult to compare the frequency data of various hospitals and medical specialisations, because the definitions of the concept "vertigo/dizziness" differ greatly. Some are broader, others more narrow. Vertigo/dizziness is seen either as a subjec-

Table 1.1. Relative frequency of different vertigo syndromes diagnosed in a dedicated neurological dizziness unit (n = 4,790 patients in 1989–2003)

Diagnosis	Frequency %
Benign paroxysmal positioning vertigo	18.3
Phobic postural vertigo (PPV)	15.9
Central vestibular vertigo	13.5
Vestibular migraine	9.6
Vestibular neuritis	7.9
Menière's disease	7.8
Bilateral vestibulopathy	3.6
Psychogenic vertigo (without PPV)	3.6
Vestibular paroxysmia	2.9
Perilymph fistula	0.4
Various other disorders	12.3
Unknown aetiology	4.2

tive symptom or as a functional vestibular disorder that can be objectified. Both tendencies are dissatisfying, as on the one hand, the symptom of dizziness or vertigo is observed in non-vestibular functional disturbances (orthostatic dysregulation) and, on the other hand, central vestibular functional disturbances (lateropulsion in Wallenberg's syndrome) also occur without any subjective dizziness.

1.2 Patient History

Vertigo is considered either an unpleasant disturbance of spatial orientation or the illusory perception of a movement of the body (spinning and wobbling) and/or of the surroundings. Care is necessary when taking the neuro-otological history of the patient (the usual pre-prepared vertigo questionnaire cannot replace it), especially because the patient's complaint of being "dizzy" is ambiguous. The important criteria for differentiating the various dizziness/vertigo syndromes, the basis for clinical classification, are as follows:

- **type of vertigo**: rotatory vertigo as experienced when riding a merry-go-round (e.g., vestibular neuritis) or postural imbalance, such as during boat trips (e.g., phobic postural vertigo) or numbness (e.g., medication, drug intoxications) (Table 1.2)
- **duration of vertigo**: attacks of vertigo that last for seconds to minutes (vestibular paroxysmia), over hours (e.g., Menière's disease, basilar/vestibular migraine; Table 1.3), persistent

Table 1.2. Numbness as a key symptom

Presyncopal dizziness
Orthostatic dysregulation
Vasovagal attacks
Neurocardiogenic (pre) syncope
Cardiac arrhythmia and other heart diseases
Psychiatric illnesses
Hyperventilation syndrome
Panic attacks
Agoraphobia
Acrophobia
Phobic postural vertigo
Metabolic disorders
Hypoglycaemia
Electrolyte disorders (hypercalcaemia, hyponatraemia)
Intoxication
Alcohol
Medications (see Table 6.3, p. 127)
Toxic substances (see Table 2.2, p. 79)

Table 1.3. Episodic vertigo, diseases with recurrent attacks of vertigo

Labyrinth/vestibulocochlear (VIIIth cranial) nerve
• Menière's disease
• Vestibular paroxysmia
• Perilymph fistula or superior canal dehiscence syndrome (induced by coughing, pressing, or loud sounds of a specific frequency, i.e., a Tullio phenomenon)
• Benign paroxysmal positioning vertigo (only during changes of head position relative to gravity)
• Cogan's syndrome
• Cysts or tumours of the cerebellopontine angle
Central vestibular system
• Transient vertebrobasilar ischaemia
Rare
• "Rotational vertebral artery occlusion syndrome"
• Vestibular epilepsy
• "Room-tilt illusion"
• Paroxysmal ataxia/dysarthrophonia (multiple sclerosis)
• Familial episodic ataxia types 1 and 2
• Paroxysmal "ocular tilt reaction"
Peripheral and/or central
• Basilar/vestibular migraine
• Benign paroxysmal vertigo of childhood
• Vertebrobasilar transient ischaemia (e.g., anterior inferior cerebellar artery)

Table 1.4. Persistent vertigo or dizziness

Infections
- *Viral*
  Vestibular neuritis
  Herpes zoster oticus
  Viral neurolabyrinthitis
- *Bacterial*
  Bacterial meningitis
  Tuberculous labyrinthitis
  Syphilitic labyrinthitis
  Chlamydial labyrinthitis
  Lyme borreliosis
  Otitis media (rarely)

Autoimmunological inner ear diseases (see Table 2.2, p. 79)

Tumours
Vestibular schwannoma
Meningeoma
Cholesteatoma
Epidermoid cyst
Glomus tumour
Metastasis
Meningeosis carcinomatosa

Vascular
Labyrinthine infarction (anterior inferior cerebellar artery)
Pontomedullary brainstem infarction
Vertebrobasilar ectasia
Hyperviscosity syndrome

Traumatic
Temporal bone fracture (transverse > longitudinal fracture)
Labyrinthine concussion
Post-traumatic otolith vertigo
Perilymph fistula
Brainstem concussion

Iatrogenic
Temporal bone surgery
Systemic or transtympanal treatment with aminoglycosides
Other ototoxic substances (see Table 2.2)

vertigo lasting for days to a few weeks (e.g., vestibular neuritis; Table 1.4), attacks of postural vertigo lasting from minutes to hours (e.g., transient ischaemic attack of the brainstem or cerebellar structures; Table 1.3)

- **trigger/exacerbation of vertigo**: no trigger (e.g., vestibular neuritis), walking (e.g., bilateral vestibulopathy), head turning (e.g., vestibular paroxysmia; Table 1.5), head positioning (e.g., BPPV), coughing, pressing, loud sounds of a certain frequency—as a Tullio phenomenon (e.g., perilymph fistula), or in certain social situations (e.g., phobic postural vertigo).

Table 1.5. Vertigo elicited by lateral head rotation

- Vestibular paroxysmia
- "Rotational vertebral artery occlusion syndrome"
- Compression of the VIIIth nerve due to cerebellopontine angle mass
- Carotid sinus syndrome

Further questions and investigations should try to identify other possible accompanying symptoms.

Combination of vestibular and audiological symptoms

- Menière's disease
- Perilymph fistula or superior canal dehiscence syndrome
- Vestibular paroxysmia
- Cerebellopontine angle tumour
- Cogan's syndrome or other inner ear autoimmune diseases
- Ear/head trauma
- Pontomedullary brainstem infarct
- Pontomedullary MS plaque
- Labyrinthine infarct (anterior inferior cerebellar artery, labyrinthine artery)
- Hyperviscosity syndrome
- Neurolabyrinthitis
- Zoster oticus
- Cholesteatoma
- Inner ear malformation
- Vestibular atelectasis
- Otosclerosis
- Vestibular epilepsy

Illusionary movements of the surroundings (oscillopsia)

Without head movements

- Spontaneous vestibular nystagmus (e.g., in vestibular neuritis)
- Congenital nystagmus (depending on direction of gaze)
- Downbeat nystagmus
- Upbeat nystagmus
- Acquired pendular nystagmus
- Periodic alternating nystagmus
- Opsoclonus
- Ocular flutter
- Vestibular paroxysmia
- Myokymia of the superior oblique muscle (monocular)
- Paroxysmal "ocular tilt reaction"
- Spasmus nutans (infants)
- Voluntary nystagmus

Only during head movements
- Bilateral vestibulopathy
- Disorders of the ocular motor system (peripheral or central)
- Vestibular paroxysmia (only in part)
- Benign paroxysmal positioning vertigo
- Central positional/positioning vertigo
- Vestibulocerebellar ataxia
- Perilymph fistulas, superior canal dehiscence syndrome
- Post-traumatic otolith vertigo
- "Rotational vertebral artery occlusion syndrome"
- Intoxication (e.g., anticonvulsants, alcohol)

Vertigo with additional brainstem/cerebellar symptoms
- Basilar/vestibular migraine
- Intoxication
- Craniocervical malformations (e.g., Arnold–Chiari malformation)
- Lacunar or territorial infarcts
- Haemorrhages (e.g., cavernoma)
- Inflammation (e.g., MS plaque)
- Brainstem encephalitis
- Head trauma
- Tumours of the cerebellopontine angle, brainstem or cerebellum
- Familial episodic ataxia type 2
- Creutzfeldt–Jakob disease

Vertigo with headache
- Migraine without aura ("motion sickness")
- Basilar/vestibular migraine
- Brainstem/cerebellar ischaemia
- Vertebrobasilar dissection
- Infratentorial haemorrhage
- Inner/middle ear infection
- Head trauma (especially transverse temporal bone fracture)
- Infratentorial tumour
- Zoster oticus

The analysis of postural and gait instability frequently allows differentiation between peripheral (Table 1.6) and central vestibular (Table 1.7) disorders.

Table 1.6. Disturbance of posture and gait control in peripheral vestibular disorders

Illness	Direction of deviation	Pathomechanism
• Vestibular neuritis	Ipsiversive	Vestibular tonus imbalance due to failure of the horizontal and anterior semicircular canal and utricle
• Benign paroxysmal positioning vertigo	Forward and ipsiversive	Ampullofugal stimulation of the posterior canal due to canalolithiasis that leads to endolymph flow
• Attacks of Menière's disease (Tumarkin's otolithic crisis)	Lateral ipsiversive or contraversive (sudden falls)	Variations of the endolymph pressure lead to an abnormal stimulation of the otoliths and sudden vestibulospinal tonus failure
• Tullio phenomenon	Backward, contraversive, diagonal	Stimulation of the otoliths by sounds of certain frequencies, e.g., in cases of perilymph fistulas
• Vestibular paroxysmia	Contraversive or in different directions	Neurovascular compression of the vestibulocochlear nerve and excitation (rarely inhibition) of the vestibular nerve
• Bilateral vestibulopathy	Different directions, especially forward and backward	Failure of vestibulospinal postural reflexes, exacerbated in the dark and on uneven ground

Table 1.7. Disturbance of posture and gait control in central vestibular disorders

Illness	Direction of deviation	Pathomechanism
• Vestibular epilepsy (rare)	Contraversive	Focal seizures due to epileptic discharges of the vestibular cortex
• Thalamic astasia (often overlooked)	Contraversive or ipsiversive	Vestibular tonus imbalance due to posterolateral lesions of the thalamus
• Ocular tilt reaction	Contraversive with mesencephalic lesions, ipsiversive with pontomedullary lesions	Tonus imbalance of the vestibulo-ocular reflex in the roll plane with lesions of the vertical canals or otolith pathways
• Paroxysmal "ocular tilt reaction"	Ipsiversive with mesencephalic excitation, contraversive with pontomedullary excitation or excitation of the vestibular nerve	Pathological excitation of the otolith or vertical canal pathways (VOR in the roll plane)
• Lateropulsion (Wallenberg's syndrome)	Ipsiversive, diagonal	Central vestibular tonus imbalance ("roll and yaw planes") with tilt of subjective vertical
• Downbeat nystagmus syndrome	Backward	Vestibular tonus imbalance in the "pitch plane"

1.3 Neuro-ophthalmological and Neuro-otological Examination

Besides the patient history, the neuro-ophthalmological and neuro-otological examinations are especially important. In the clinical examination, the physician should at first try to differentiate between peripheral vestibular and central vestibular forms of vertigo. Figures 1.3 to 1.18 and Table 1.8 (p. 25) illustrate the individual examination procedures, the essential findings and their interpretation. Additional laboratory and imaging examinations (see Section 1.4) are in many cases of less clinical significance.

Figure 1.3. Measurement of head tilt. An abnormal head posture to the right or left shoulder or a constant, abnormal tilt is especially observed in patients with paresis of the oblique eye muscles (e.g., in superior oblique palsy, the head is turned to the non-affected side to lessen diplopia), or in those with an ocular tilt reaction due to a tonus imbalance of the vestibulo-ocular reflex in roll. As a rule, the head is tilted to the side of the lower eye. Acute unilateral lower medullary lesions (e.g., in Wallenberg's syndrome) or acute unilateral peripheral vestibular lesions cause an ipsiversive head tilt, whereas mesencephalic lesions cause a contraversive head tilt.

Figure 1.4. Cover tests to detect misalignments of the visual axes. The prerequisite for all of these tests is foveal fixation. Fixation in primary eye position already reveals heterotropia (manifest strabismus), i.e., a misalignment of the visual axes when both eyes look at a single target. First the patient has to fixate either a near target (at a distance of 30–40 cm) or one 5–6 m away. Then the examiner covers one eye and looks for movements of the now uncovered eye (correction movements). If the uncovered eye moves from the inside outward, esotropia is present; if it moves from the outside inward, exotropia; if it moves from above downward, hypertropia; if it moves from below upward, hypotropia. The other eye is then examined. The unilateral cover test reveals heterophoria (latent strabismus), i.e., a misalignment of the visual axes when an object is fixed by only one eye. (Note that the cover test has to be performed first to exclude heterotropia.) At first only one eye is covered, then the cover is removed and its corrective movement is observed. If the eye moves to the outside, esophoria is present; if it moves to the inside, exophoria; if it moves downward, hyperphoria; if it moves upward, hypophoria. The alternating cover test reveals the maximal deviation of the eye axes, in cases of tropia or phoria. It is also useful for detecting skew deviation (part of the ocular tilt reaction), i.e., a vertical misalignment of the eyes that cannot be explained by an ocular muscle palsy or damage of the peripheral nerve. Vertical corrective movements are looked for, when the cover is switched from one eye to the other. In contrast to IVth cranial nerve palsy, the vertical misalignment changes little during different directions of gaze. Latent congenital nystagmus is a jerk nystagmus that is absent when both eyes fixate; it appears when one eye is covered and, depending on which eye is covered, often changes direction.

Figure 1.5. Clinical examination of the eyes in nine different positions to determine ocular alignment (a possible misalignment of the axes of the eyes), fixation deficits, nystagmus, range of movement, and disorders of gaze-holding ability. The examination can be performed with an object for fixation or a small rod-shaped flashlight. In the primary position, one should first look for periodic eye movements, such as nystagmus, e.g., horizontal–rotatory, suppressed by fixation as in peripheral vestibular dysfunction or vertically upward or downward (upbeat/downbeat nystagmus syndrome), or horizontal or torsional movements with only slight suppression (or increase) of intensity during fixation, as in a central vestibular dysfunction; as a rule, a congenital nystagmus beats horizontally at various frequencies and amplitudes and increases during fixation; so-called square-wave jerks (small saccades [0.5°–5°] that cause the eyes to oscillate around the primary position and increasingly occur in progressive supranuclear palsy or certain cerebellar syndromes); ocular flutter (intermittent rapid bursts of horizontal oscillations without an intersaccadic interval); or opsoclonus (combined horizontal, vertical and torsional oscillations, which occur in various disorders such as encephalitis, tumours of the brainstem or cerebellum, intoxication, or in paraneoplastic syndromes). After checking for possible eye movements in primary position and the misalignment of the axes of the eyes (see cover test), the examiner should then establish the range of eye movements monocularly and binocularly in the eight end-positions; deficits found here can indicate ocular muscle or nerve palsy. Gaze-holding deficits can also be determined by examining eccentric gaze position.

Figure 1.6. Clinical examination of eye positions or movements with an examination flashlight. Using a small rod-shaped flashlight has the advantage that the corneal reflex images can be observed and thus ocular misalignments can be easily detected. Note: it is important to observe the corneal reflex images from the direction of the illumination and to ensure that the patient attentively fixates the object. The flashlight also allows one to determine whether the patient can fixate with one or both eyes in the end-positions. This is important for detecting a defect of gaze holding. Gaze-evoked nystagmus can only be clearly identified when the patient fixates with both eyes. It is most often a side-effect of medication (e.g., anticonvulsants, benzodiazepines) or toxins (e.g., alcohol). Horizontal gaze-evoked nystagmus can indicate a structural lesion in the area of the brainstem or cerebellum (vestibular nucleus, nucleus prepositus hypoglossi, flocculus, i.e., the neural integrator). Vertical gaze-evoked nystagmus is observed in midbrain lesions involving the interstitial nucleus of Cajal. A dissociated horizontal gaze-evoked nystagmus (greater in the abducting than the adducting eye) in combination with an adduction deficit are the signs of internucle ophthalmoplegia due to a defect of the medial longitudinal fascicle, ipsilateral to the adduction deficit. Downbeat nystagmus usually increases in the eccentric gaze position and when looking down. To examine for a so-called rebound nystagmus, the patient should gaze for at least 15 s to one side and then return the eyes to the primary position; this can cause a transient nystagmus to appear with slow phases in the direction of the previous eye position. Rebound nystagmus generally indicates cerebellar dysfunction or damage of the cerebellar pathways.

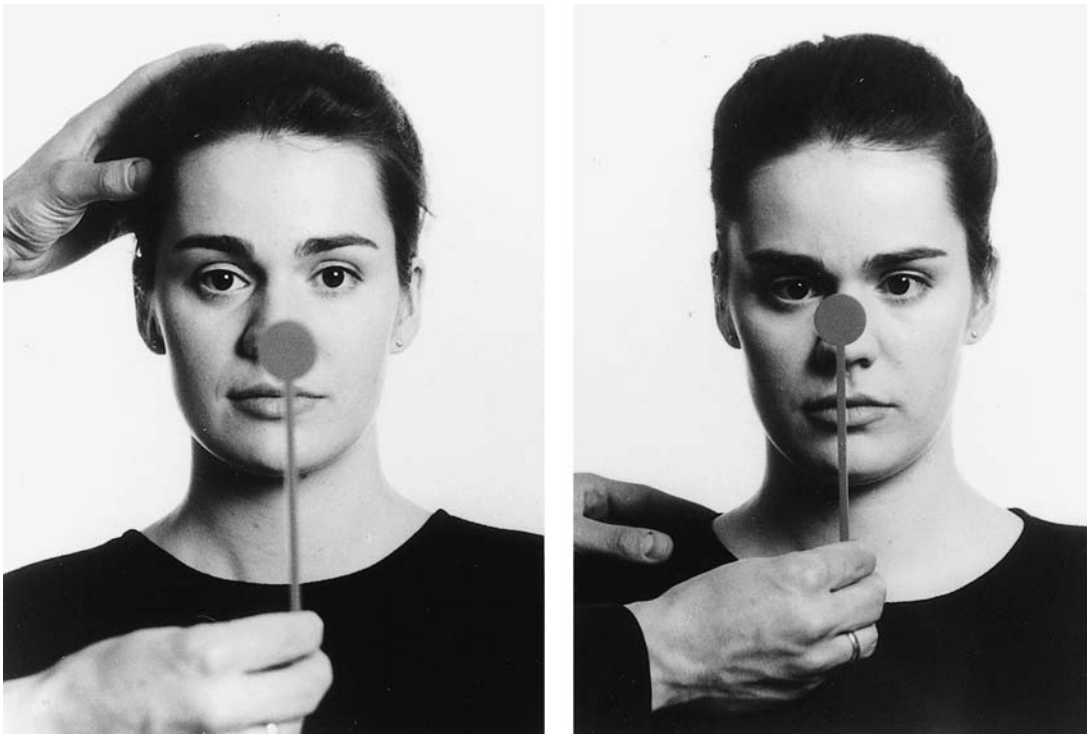


Figure 1.7. Vergence test and convergence reaction: a target is moved from a distance of about 50 cm toward the patient's eyes. This causes vergence, accommodation and miosis, i.e., a convergence reaction. Neurons important for the convergence reaction are in the area of the mesencephalic reticular formation and the oculomotor nucleus. This explains why the convergence reaction is disturbed in rostral midbrain lesions and tumours of the pineal region and thalamus, and why abnormalities of vertical gaze are often associated with these defects. In certain neurodegenerative disorders such as progressive supranuclear palsy, convergence may also be impaired. Inborn defects of accommodative-convergence synkinesis also accompany some forms of childhood strabismus. Convergence-retraction nystagmus can be induced by having the patient look at a moving optokinetic drum with its stripes going downward or by having him make upward saccades. Instead of making vertical saccades, the patient makes convergent eye movements that are associated with retractions of the eyeball. Animal experiments have shown that lesions of the posterior commissure can cause convergence-retraction nystagmus. Spasm of the near reflex is a voluntary convergence accompanied by pupillary constriction. This can mimic bilateral abducens palsy (occasionally it is psychogenic).

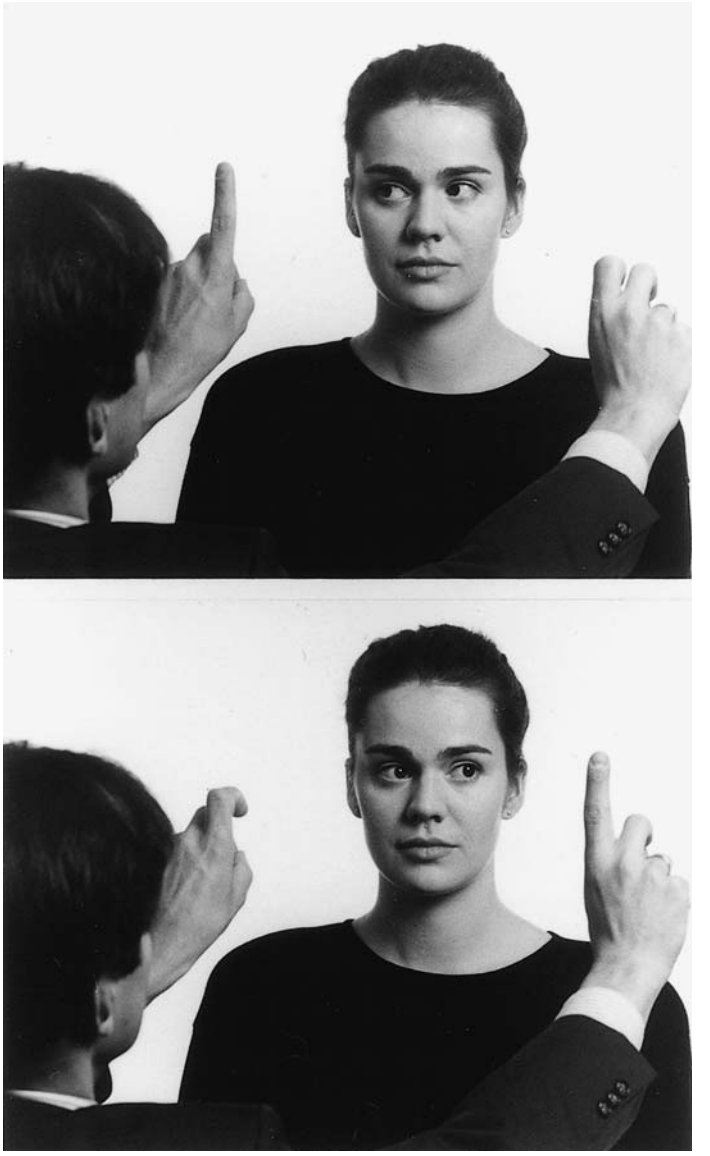


Figure 1.8. Clinical examination of saccades. First it is necessary to observe spontaneous saccades triggered by visual or auditory stimuli. Then the patient is asked to glance back and forth between two horizontal or two vertical targets. The velocity, accuracy and the conjugacy of the saccades should be noted. Normal individuals can immediately reach the target with a single fast movement or one small corrective saccade. Slowing of saccades—often accompanied by hypometric saccades—occurs for example with intoxication (medication, especially anticonvulsants or benzodiazepines) or in neurodegenerative disorders. Slowing of horizontal saccades is generally observed in brainstem lesions; there is most often a dysfunction of the ipsilateral paramedian pontine reticular formation. Slowing of vertical saccades indicates a midbrain lesion in which the rostral interstitial medial longitudinal fascicle (riMLF) is involved, not only in ischaemic or inflammatory diseases but also in neurodegenerative diseases, especially progressive supranuclear palsy. Hypermetric saccades, which can be identified by a corrective saccade back to the object, indicate lesions of the cerebellum (especially the vermis) or the cerebellar pathways. Patients with Wallenberg's syndrome make hypermetric saccades toward the side of the lesion due to a dysfunction of the inferior cerebellar peduncle; defects of the superior cerebellar peduncle, conversely, lead to contralateral hypermetric saccades. A slowing of the adducting saccade ipsilateral to a defective MLF is pathognomonic for internuclear ophthalmoplegia. Delayed-onset saccades are most often caused by supratentorial cortical dysfunction.

Figure 1.9. Clinical examination of smooth pursuit eye movements. The patient is asked to track visually an object moving slowly in horizontal and vertical directions (10–20°/s) while keeping the head stationary. Corrective (catch-up or back-up) saccades are looked for; they indicate a smooth pursuit gain (ratio of eye movement velocity and object velocity) that is too low or too high. Many anatomical structures (visual cortex, medial temporal area, medial superior temporal area, frontal eye fields, dorsolateral pontine nuclei, cerebellum, vestibular and ocular motor nuclei) are involved in smooth pursuit eye movements, which keep the image of a moving object stable on the fovea. These eye movements are also influenced by alertness, a number of drugs and age. Even healthy people exhibit a slightly saccadic smooth pursuit during vertical downward eye movements. For these reasons, as a rule, a saccadic smooth pursuit does not facilitate topographical localization or aetiological classification. Marked asymmetries of smooth pursuit, however, indicate a structural lesion; strongly impaired smooth pursuit is observed in intoxication (anticonvulsants, benzodiazepines or alcohol) as well as degenerative disorders involving the cerebellum or extrapyramidal system. A reversal of slow smooth pursuit eye movements is typical for congenital nystagmus.

Figure 1.10. Clinical examination with Frenzel's glasses. The magnifying lenses (+16 diopters) with light inside prevent visual fixation, which could suppress spontaneous nystagmus. Frenzel's glasses enable the clinician to observe spontaneous eye movements. Examination should include spontaneous and gaze-evoked nystagmus, head-shaking nystagmus (the patient is instructed to turn his head quickly to the right and to the left about 20 times; the eye movements are observed after head shaking), positioning and positional nystagmus, as well as hyperventilation-induced nystagmus. Spontaneous nystagmus indicates a tonus imbalance of the vestibulo-ocular reflex; if it is caused by a peripheral lesion—as in vestibular neuritis—the nystagmus is typically dampened by visual fixation. Head-shaking nystagmus shows a latent asymmetry of the so-called velocity storage, which can be due to peripheral and central vestibular disorders.

Figure 1.11. Clinical examination with Frenzel's glasses and a Politzer balloon. Changes in middle-ear pressure caused by applying pressure (positive or negative) to the tympanic membrane with a Politzer balloon, by tones of a certain frequency or loudness (Tullio phenomenon), or by tragal compression can induce nystagmus in patients with perilymph fistula or superior canal dehiscence syndrome. Nystagmus can also be induced by Valsalva's manoeuvre (which causes increased pressure in the middle ear), coughing, pressing, sneezing or swallowing.

Figure 1.12. The examination of one eye with an ophthalmoscope (the other eye is covered) is a sensitive method for detecting nystagmus while simultaneously checking for movements of the optic papilla or retinal vessels (Zee 1978), even with low, slow-phase velocities/frequencies or square-wave jerks (small saccades [0.5°–5°] that are often observed in progressive supranuclear palsy or certain cerebellar syndromes). Since the retina is behind the axis of rotation of the eyeball, the direction of any observed vertical or horizontal movement is opposite to that of the nystagmus, i.e., a downbeat nystagmus causes a rapid, upward movement of the optic papilla or retinal vessels.

Figure 1.13. Examination of eye movements with the optokinetic drum allows combined testing of smooth pursuit movements and saccades in horizontal and vertical directions. It is especially helpful with uncooperative or drowsy patients. One should look for asymmetries, e.g., between right and left (indicates a supratentorial cortical lesion), vertical worse than horizontal (indicative of a supranuclear gaze palsy due to a mesencephalic lesion), dissociation of the two eyes (a sign of diminished adduction in internuclear ophthalmoplegia) and reversal of pursuit (indicates congenital nystagmus).

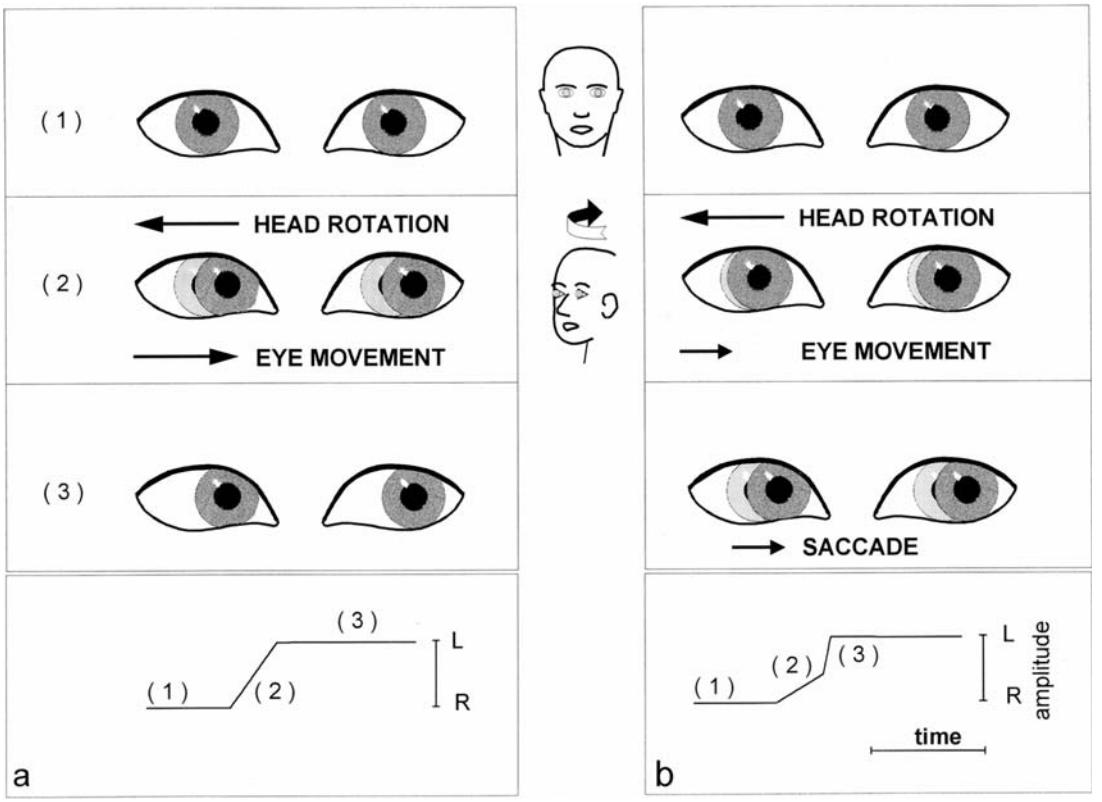


Figure 1.14. Clinical examination of the horizontal vestibulo-ocular reflex (VOR) by the head-impulse test (Halmagyi and Curthoys 1988). To test the horizontal VOR, the examiner holds the patient's head between both hands, asks him to fixate a target in front of his eyes, and rapidly turns the patient's head horizontally to the left and then to the right. This rotation of the head in a healthy subject causes rapid compensatory eye movements in the opposite direction (**a**). In cases of unilateral labyrinthine loss (exemplified by loss of the right horizontal canal) during head rotations toward the affected ear, the patient is not able to generate a fast contraversive eye movement and has to perform a corrective (catch-up) saccade to fixate the target (**b**). This catch-up saccade can be easily observed by the examiner. In **c** the examination situation is shown.

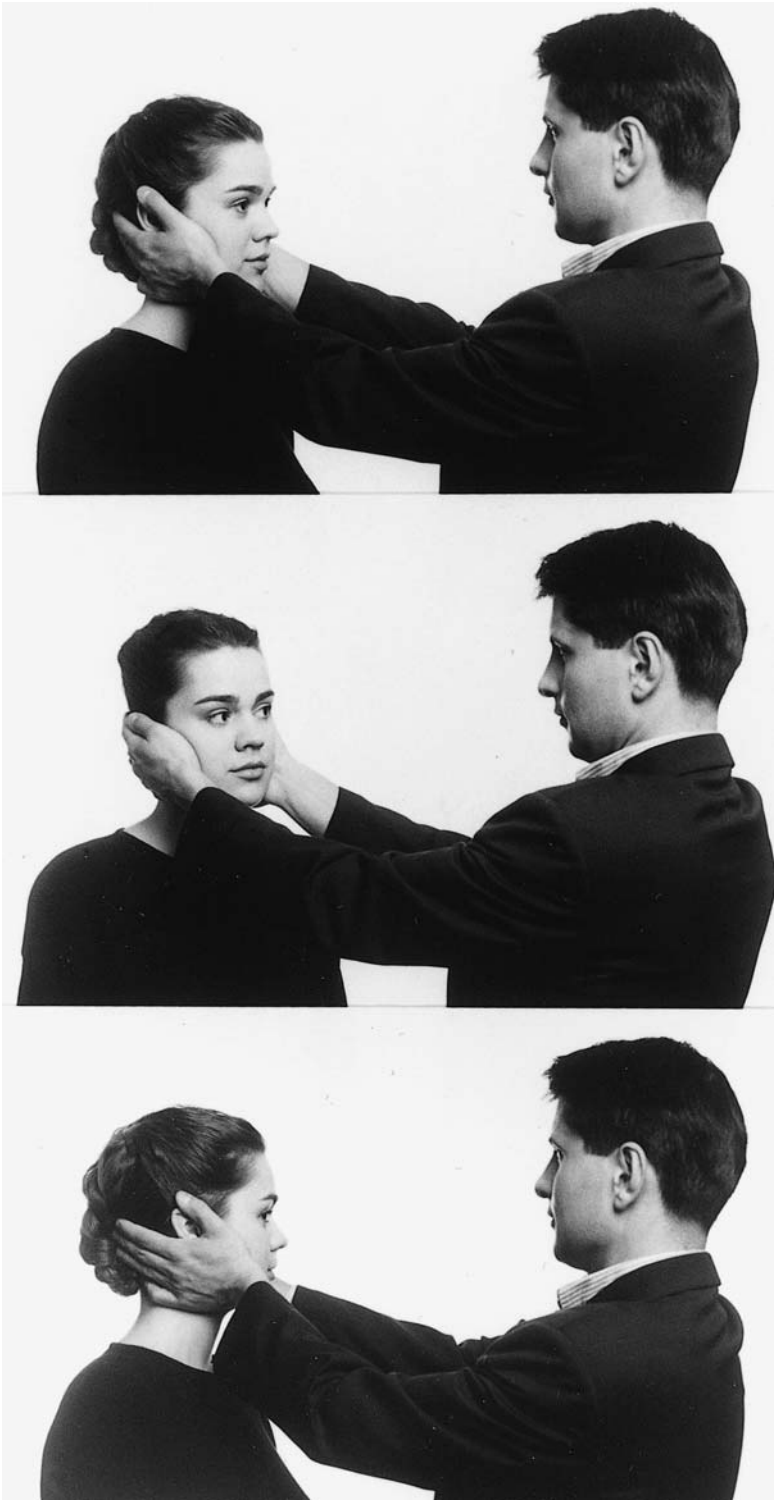


Figure 1.14. *Continued* **c**

Figure 1.15. Clinical testing of visual fixation suppression of the vestibulo-ocular reflex (VOR). Before performing the test, the examiner must be sure that the VOR is intact (see Fig. 1.14). The patient is asked to fixate a target in front of his eyes while moving his head as uniformly as possible, first horizontally and then vertically, back and forth at moderate speed. The examiner should watch for corrective saccades. A disorder of visual fixation suppression of the VOR (which as a rule occurs with smooth pursuit abnormalities, as these two functions are mediated by common neural pathways) is often observed in lesions of the cerebellum (flocculus or paraflocculus) or of the cerebellar pathways and in progressive supranuclear palsy. Anticonvulsants and sedatives can also impair visual fixation suppression of the VOR.

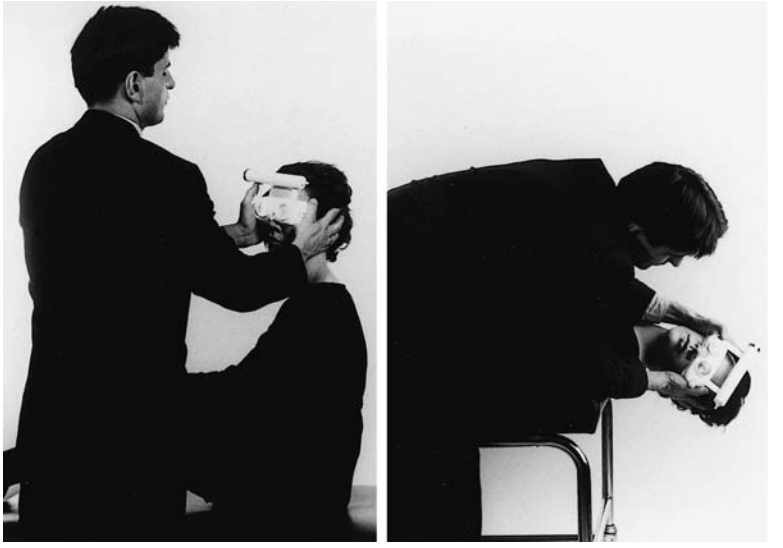


Figure 1.16. The so-called Dix–Hallpike manoeuvre is performed especially to determine whether benign paroxysmal positioning vertigo (BPPV) is present. While the patient is sitting, his head is turned by 45° to one side, and then he is rapidly put in a supine position with the head hanging over the end of the examination couch. If a BPPV of the left posterior semicircular canal, for example, is present, this manoeuvre will induce, with a certain latency, a crescendo–decrescendo-like nystagmus, which from the patient's viewpoint beats counterclockwise toward the left ear and to the forehead. When the patient is returned to a sitting position, the direction of nystagmus will change.

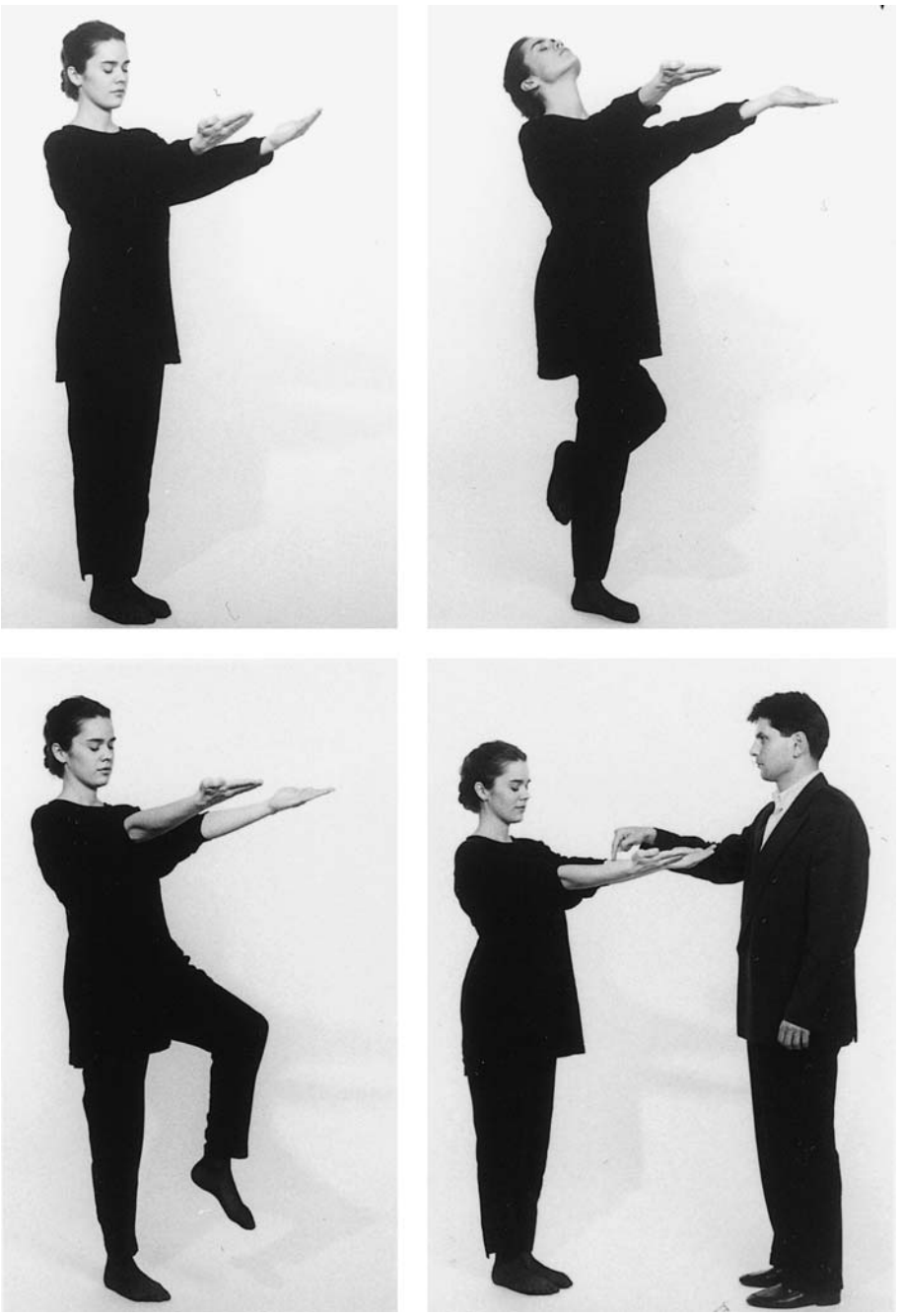


Figure 1.17. Clinical examination of balance under static conditions. There are different variations of the Romberg and of the one-leg stance test: feet next to each other with eyes first open and then closed (to eliminate visual cues, *upper left*); standing on one foot at a time with the head in a normal position (*lower left*) or with reclining head (the latter, *upper right*, creates extreme imbalance). If a psychogenic disorder is suspected, the examiner distracts the patient by writing numbers on his arm (lower right) or having him do maths mentally. If there is improvement under the last condition, the stance disorder has a psychogenic origin. Another variation is the Romberg test in tandem, during which the patient places one foot directly in front of the other (the toes of one foot touch the heel of the other, not shown). Excessive fore–aft, right–left, or diagonal sway should be looked for. A peripheral vestibular functional disorder typically causes ipsiversive falls; up- and downbeat nystagmus syndromes are typically associated with increased body sway forward and backward once the eyes are closed.

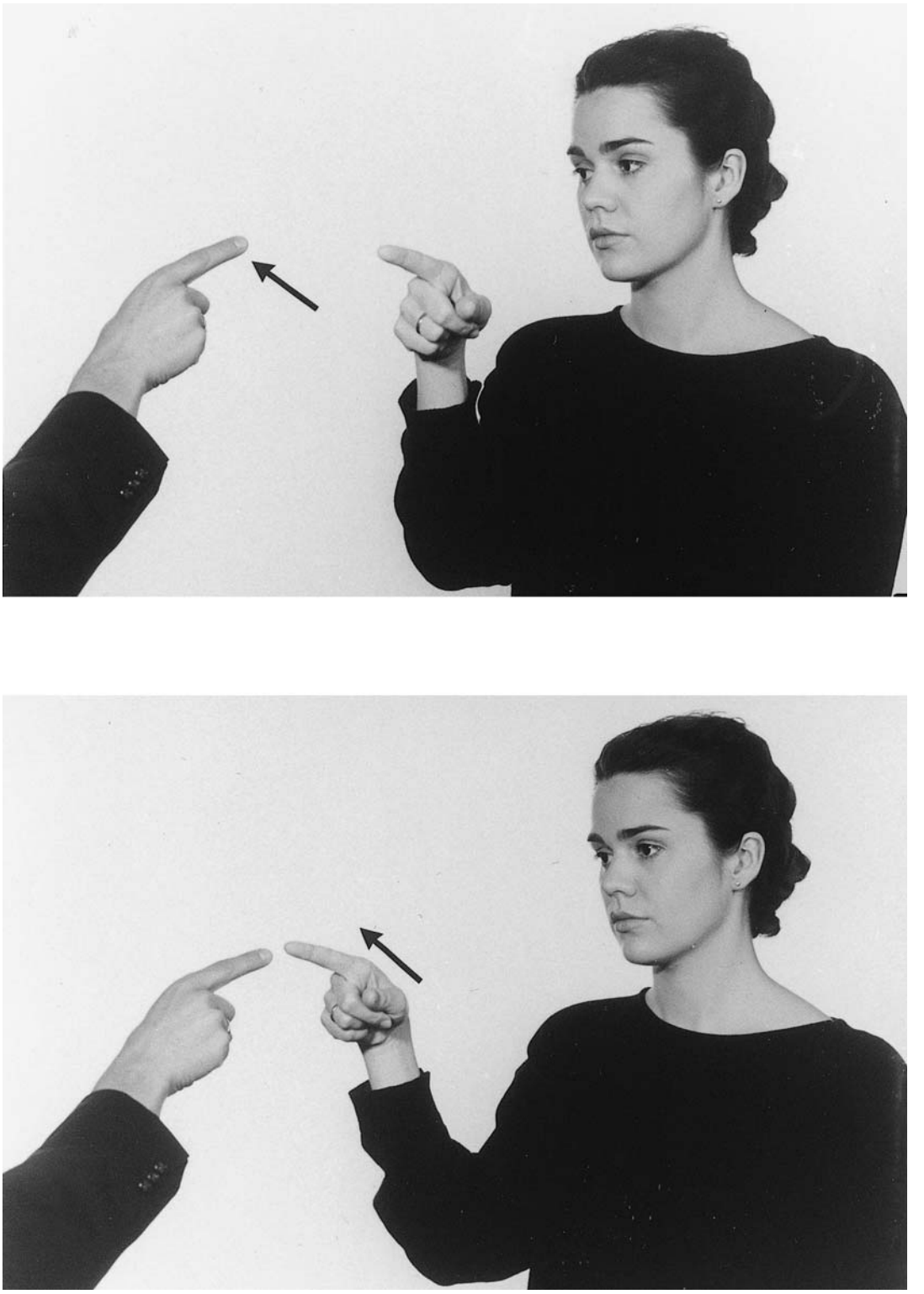


Figure 1.18. Finger-pointing test. The patient is instructed to follow the finger of the examiner by rapidly pointing toward each new position. Cerebellar ataxia is often indicated by hypermetric movements with an intention tremor. This test is more sensitive than the finger-to-nose test.

Table 1.8. Examination procedure for ocular motor and vestibular systems

Type of examination	Question
Inspection	
• Head/body posture	Tilt or turn of head/body
• Position of eyelids	Ptosis
Eye position/motility	
• Position of eyes during straight-ahead gaze	Misalignment in primary position, spontaneous or fixation nystagmus
• Cover test	Horizontal or vertical misalignment
• Examination of eyes in eight positions (binocular and monocular)	Determination of range of motility, gaze-evoked nystagmus, end-position nystagmus
Gaze-holding function	
• 10–40° in the horizontal or 10–20° in the vertical and back to 0°	Gaze-evoked nystagmus: horizontal and vertical, rebound nystagmus
Smooth pursuit movements	
• Horizontal and vertical	Smooth or saccadic
Saccades	
• Horizontal and vertical when looking around or at targets	Latency, velocity, accuracy, conjugacy
Optokinetic nystagmus (OKN)	
• Horizontal and vertical with OKN drum or tape	Inducible, direction, phase (reversal or monocularly diagonal)
Peripheral vestibular function	
• Clinical testing of the VOR (Halmagyi–Curthoys test): rapid turning of the head and fixation of a stationary target	Unilateral or bilateral peripheral vestibular deficit
Fixation suppression of the VOR	
• Turn of head and fixation of a target moving at same speed	Failure of fixation suppression
Examination with Frenzel's glasses	
• Straight-ahead gaze, to the right, to the left, downward and upward	Spontaneous nystagmus
• Head-shaking test	Provocation-induced nystagmus
Positional manoeuvre (with Frenzel's glasses)	
• To the right, left, head-hanging position, turning about the cephalo-caudal axis	Peripheral positional or positioning nystagmus, central positional nystagmus
Posture and balance control	
• Romberg's test and simple/difficult posture and gait tests • Open–closed eyes • With/without reclining the head	Instability, tendency to fall
• With/without distraction (writing numbers on the skin, doing maths mentally)	Psychogenic etiology

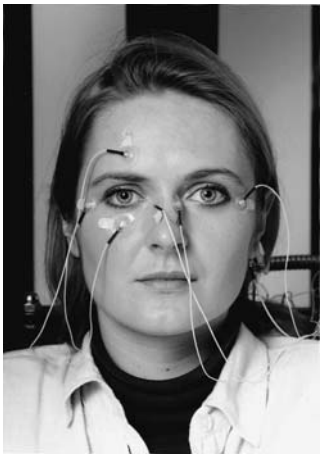


Figure 1.19. Electronystagmography (ENG). Placement of the electrodes for monocular recording of horizontal and vertical eye movements. The electrophysiological basis of the ENG is the corneo-retinal dipole (a potential difference of about 1 μV). The dipole is parallel to the longitudinal axis of the eye, with the retina having a negative potential. Changes in this dipole between the horizontal or vertical electrodes are DC-amplified. The ENG allows non-invasive horizontal recordings of ±40° with an accuracy of ca. 1° and vertical recordings of ±20°. Major disadvantages are susceptibility to eyeblink artefacts, electromyographic activity and unstable baseline; torsional eye movements cannot be recorded with the ENG.

1.4 Laboratory and Imaging Examinations

If the patient's history is taken accurately and the clinical examination is precise, laboratory examinations are in many cases only of secondary importance. Figures 1.19 to 1.26 and Table 1.9 (p. 33) summarise the essential neuro-ophthalmological procedures of examination, give typical findings, and indicate how to interpret them. The most important additional laboratory examinations allow the measurement of eye movements, e.g., electronystagmography (ENG), video-oculography, and the magnetic search-coil technique, the latter primarily for research.

1.4.1 Electronystagmography

To record eye movements quantitatively, two electrodes are placed horizontally and vertically on each eye so that the changes in the dipole between the retina and cornea, which occur with eye movements, can be recorded (Figure 1.19). Electronystagmography (ENG) also allows documentation of the findings (important for monitoring the course of the patient) and, for example, relatively exact measurements of saccade velocity and

Figure 1.20. Electronystagmography. Rotatory chair and rotatory drum (with vertical stripes) with an apparatus that projects a laser spot (above the patient). This setup allows recordings of eye movements under static conditions (e.g., test for spontaneous or gaze-evoked nystagmus, saccades, pursuit and optokinetic nystagmus) and under dynamic conditions (per- and postrotatory nystagmus, fixation suppression of the vestibulo-ocular reflex), as well as positional and positioning testing and caloric irrigation.

saccade accuracy. In addition, patients can be tested with a rotatory chair and rotatory drum (Figure 1.20). Irrigation of the external auditory canal with 30°C cool and 44°C warm water (caloric testing) can be used to identify any disturbance of labyrinthine function (horizontal canal) (Figure 1.21).

1.4.2 Video-oculography

Video-oculography is another non-invasive method that is now being used more frequently. The eyes are first filmed by one or two video cameras (i.e., monocular or binocular recording) inte-

Figure 1.21. Electronystagmography. By means of caloric testing, the excitability of the individual horizontal canals can be determined and thus whether or not they are functioning. After excluding the possibility of a lesion of the eardrum, the head of the patient is tilted 30° upward, so that the horizontal semicircular canals approach the vertical plane. This allows optimal caloric stimulation. The external auditory canals on each side are separately irrigated under standard conditions with 30°C cool and 44°C warm water. At the same time the horizontal and vertical eye movements are recorded by means of electronystagmography. The irrigation with 44°C warm water causes excitation of the hair cells of the horizontal canal along with slow contraversive eye movements; the 30°C cool water leads to an inhibition with the slow ipsiversive eye movements. The unit of measurement is the maximal velocity of the irrigation-induced eye movements (peak slow phase velocity, PSPV); PSPV values less than 5°/s are considered pathological. Since there is considerable interindividual variation of caloric excitability, the so-called "vestibular paresis formula" of Jongkees (Jongkees et al. 1962) is also used to compare both labyrinths:

$$(((R30°C + R44°C) - (L30°C + L44°C))/ (R30°C + R44°C + L30°C + L44°C)) \times 100,$$

where, for instance, R 30°C is the PSPV during caloric irrigation of the right ear with 30°C cool water. Values of >25% asymmetry between the affected and non-affected labyrinth are considered pathological and indicate, for example, a unilateral peripheral vestibular disorder.

grated in a mask attached to the head. Then a computer analysis of the image of movements of the pupils and light reflexes is performed to represent the eye movements in two dimensions. This method allows rapid and reliable recording of horizontal and vertical eye movements (without muscle artefacts or unstable baseline). Recording is only possible when the eyes are wide open, and the resolution is limited due to the image repeat frequency of the video camera (today generally limited to 100 Hz). There is a largely linear resolution in the range of ±30°. The use of 3-D representation of eye movements for research purposes (i.e., additional measurement of torsion) requires an extensive

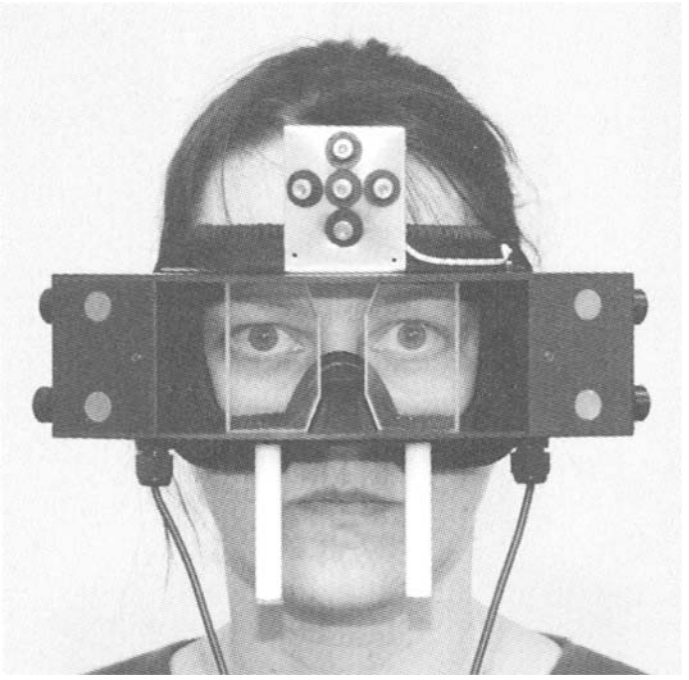


Figure 1.22. Video-oculography performed with a mask attached to the head in which a camera is integrated. An infrared headlight built into the mask also allows measurement of eye movements in complete darkness. The signal of the integrated camera is transmitted to a normal digital video camera and finally stored on a computer. The pictures are analysed offline by means of a video-oculography program that calculates the eye movements. A Cajal dot placed on the sclera alleviates the registration or analysis of eye movements in the roll plane.

analysis of the image of the iritic structures or of two additional marker dots applied to the sclera (see Figure 1.22).

1.4.3 Neuro-orthoptic and Psychophysical Procedures

Recent years have witnessed the growing importance of neuro-orthoptic and psychophysical examination procedures (e.g., measurement of vertical divergence of the eyes and the psychophysical determination of the subjective visual vertical) or of the "scanning laser ophthalmoscope" for measurement of ocular torsion (Figures 1.23 and 1.24). The topographical and diagnostic significance of these procedures is particularly evident when differentiating between peripheral and central vestibular or ocular motor lesions.

1.4.4 Audiometry

As a rule, an audiologist gives a hearing test. In connection with the main symptom of vertigo, audiometry is especially important

Figure 1.23. The subjective visual vertical (SVV). For determination of the SVV, the patient sits upright in front of a hemispheric dome (60 cm in diameter) and looks into it. The surface of the dome extends beyond the limits of the patient's visual field and is covered with a random pattern of dots. This prevents the patient from orienting himself spatially by fixed external structures. The hemispheric dome is connected axially to a motor, and can be rotated; a circular target disk (14° of visual field) with a straight line through the centre is 30 cm in front of the patient at eye level. The line is also connected with a DC motor and can be adjusted by the patient by means of a potentiometer until he has the subjective impression that the line is "vertical." The deviation of the line from the objective vertical axis is measured in degrees and registered on a PC. The mean of ten measurements equals the SVV. Under these conditions, the normal range (mean ± 2 SDs) of the SVV is 0° ± 2.5°. Measurements can be made under static (*left*, dome stationary) and dynamic (*right*, dome rotating) conditions.

for diagnosing Menière's disease, labyrinthitis, vestibular schwannoma, and other diseases affecting the vestibulocochlear nerve, as well as in bilateral vestibulopathy.

1.4.5 Additional Laboratory Examinations

To clarify the cause of disorders (differential diagnoses: ischaemia, haemorrhage, tumour or inflammation), additional imaging techniques are necessary (cranial magnetic resonance imaging (MRI) with precise sections of the brainstem and the cerebellopontine angle as well as computed tomography with high resolution images of the bony labyrinth), Doppler sonography, and in some cases also a spinal tap, as well as posturography (Figure 1.25, page 32) and evoked potentials (e.g., auditory-evoked potentials or vestibular-evoked myogenic potentials; Figure 1.26, page 34).

Figure 1.24. Measurement of the eye position in the roll plane. The scanning laser ophthalmoscope (SLO) can be used to make photographs of the fundus of the eye (examination is also possible with a fundus camera). The rolling of the eye or eye torsion can be measured in degrees on the fundus photographs as the angle between the horizontal and the so-called papillofoveal meridian. The patient sits upright, looks into the SLO and fixates a dot. (It is not necessary to administer a mydriatic drug; however, this is necessary if the measurement is made with traditional fundus photography.) Both eyes of healthy controls exhibit a slightly excyclotropic position in the roll plane, i.e., counterclockwise rotation of the right eye, clockwise rotation of the left eye (from the viewpoint of the examiner). The normal range (±2 SDs) is from –1 to 11.5°. Values outside this range are considered pathological (e.g., patients with a peripheral vestibular lesion show an ipsiversive torsion of both eyes with an excyclotropia of the ipsilateral eye and an incyclotropia of the contralateral eye).

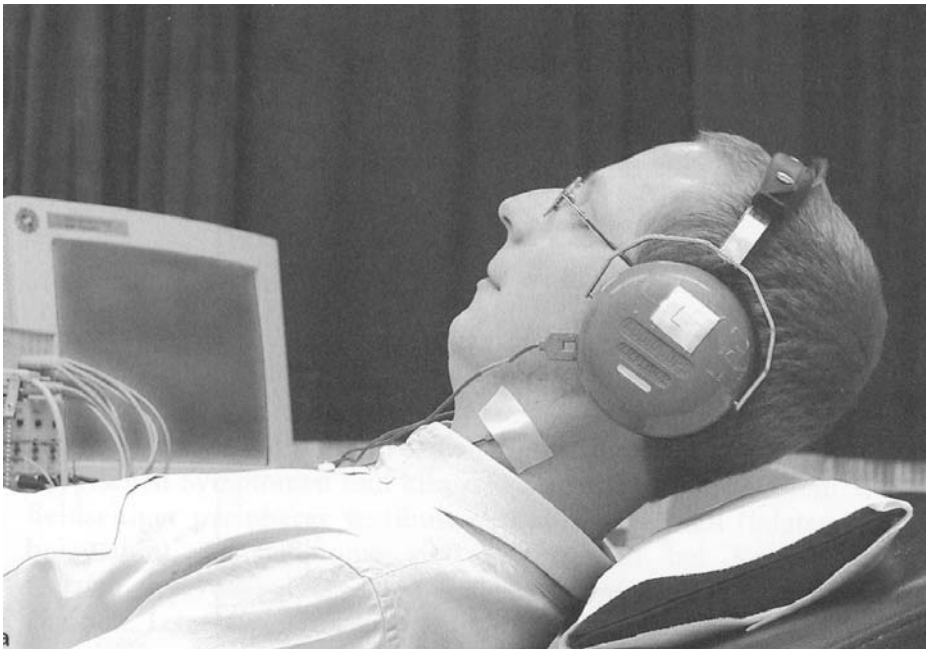
1.4.6 Imaging of the Petrous Bone, the Cerebellopontine Angle and the Brainstem with Computed Tomography and Magnetic Resonance Imaging

As a result of new developments with high-resolution MRI and computed tomography (CT) of the petrous bone, it is now possible to identify the following peripheral vestibular diseases: masses in the cerebellopontine angle, internal auditory canal (e.g., vestibular schwannoma) or middle ear (e.g., cholesteatoma); post-traumatic forms of vertigo due to petrous bone fractures; and "vestibular pseudo neuritis" due to fascicular lesions of the vestibular nerve at the entry zone of the brainstem (MS plaques or ischaemic lesions). Imaging is also important for the diagnosis of inflammatory (e.g., labyrinthitis,

Figure 1.25. Posturography. Posturography allows the examination of control of postural stability (a Kistler platform is used). The parameters include the original registrations of body sway to the right or left, forward or backward, upward or downward; the frequency analysis of the sway (Fourier analysis); and the so-called sway path values (SP, m/min). The SP is defined as the length of the path described by the centre of foot pressure during a given time. Healthy subjects also exhibit body sway as a result of inherent physiological instability when standing on a recording platform; SP is exacerbated in vestibular disorders. The SP values can be derived automatically with a computer for the anteroposterior, mediolateral and craniocaudal directions. These values are calculated as the distances between two consecutive sampling points (measured every 25 ms); the anteroposterior (sagittal = x) plane, i.e., sagittal sway (calculated as $\Sigma|\Delta x|$), the mediolateral (frontal = y) plane, i.e., frontal sway (calculated as $\Sigma|\Delta y|$) and the craniocaudal (transversal = z) plane, i.e., the transversal sway (calculated as $\Sigma|\Delta z|$) or for all three planes as the total SP.

Table 1.9. Neuro-ophthalmological examination and laboratory tests for vertigo/dizziness and eye movement disorders

Technique	Features	Advantages	Disadvantages
• Neuro-ophthal. examination	Total range of eye movements, horizontal, vertical (torsional)	No technical requirements, simple, resolution < 1°	No recording, eye movement velocity cannot be measured
• Orthoptic examination	Fundus photography, determination of eye misalignment and psychophysical determination of e.g., the subjective visual vertical	Precise measurement with documentation, non-invasive, well tolerated	Expensive apparatus (e.g., scanning laser ophthalmoscope)
• ENG	Measurement range ±40°, resolution of 1°	Non-invasive, well tolerated, even by children, caloric stimulation possible, widespread method	No measurement of torsional and poor measurement of vertical movements, eyelid artefacts, baseline drift
• Video-oculography	Measurement range ±40°, resolution of 0.1–1°	Non-invasive, well tolerated, possible to measure torsion	Measurement only possible with eyes open, 3-D analysis is still complicated and expensive
• Infrared system	Measurement range ±20°, resolution of 0.1°	High resolution, non-invasive	Measurements only possible with open eyes, relatively expensive, vertical measurements poor, torsional measurements not possible
• Magnetic-coil technique	Measurement range ±40°, resolution of 0.02°	Best resolution of horizontal, vertical and torsional movements (research)	Semi-invasive, unpleasant, expensive, only with cooperative patients, maximum of 30 min, local anaesthetic necessary
• Vestibular-evoked myogenic potentials	Examination of sacculus function	Non-invasive, well tolerated, simple to perform	Still moderate clinical experience has been made, some findings in part still contradictory



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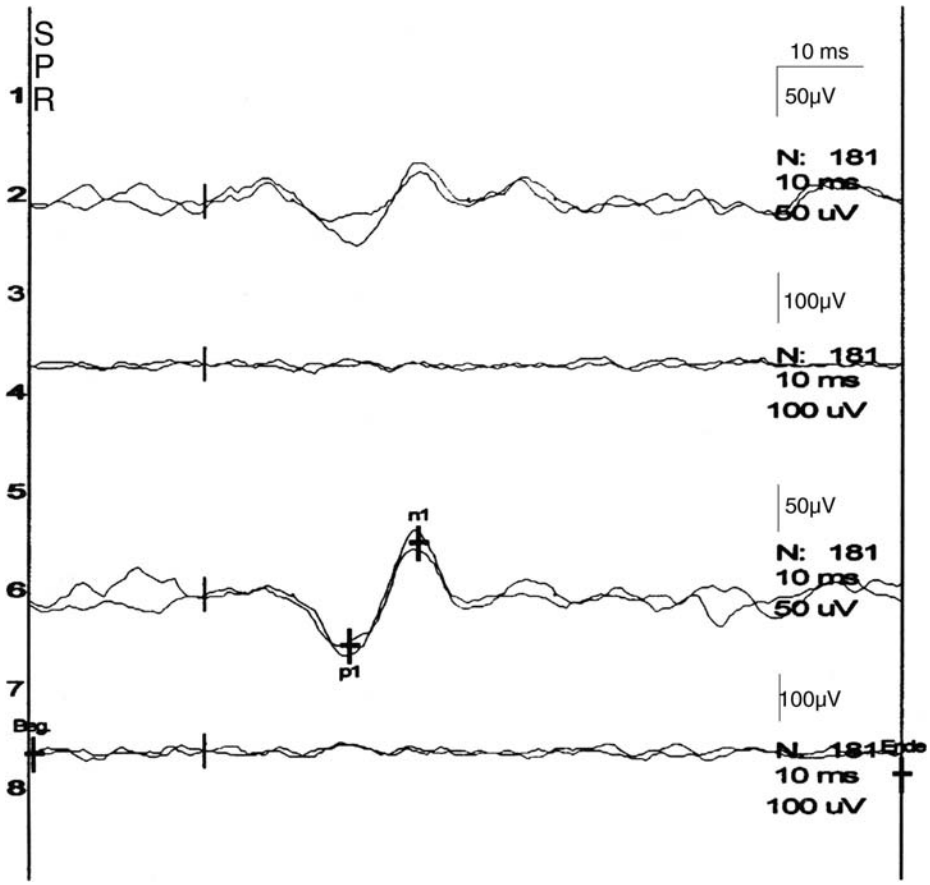

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Figure 1.26.

Figure 1.26. Vestibular-evoked myogenic potentials (VEMPs). The VEMP is used to test the reflex arc of the saccule, which extends over the vestibular nerves, vestibular nuclei, interneurons, and motor neurons to the neck musculature (sternocleidomastoid m.). It complements caloric testing, since the latter tests only the canal system and not the otolith function. The prerequisite for VEMP testing is an intact middle ear function; it is not necessary that hearing be preserved, since the "sensitivity to sound" of the saccule can be used in the VEMP. The reflex is triggered by a loud click. Surface EMG is used to record from both sternocleidomastoid muscles. **a**. Healthy subjects first show on the ipsilateral side a positive wave (about 14 s after the stimulus) as well as negative wave (about 21 ms; lines 1 and 3). **b**. The responses can as a rule not be recorded contralaterally (lines 2 and 4). Approximately 50–100 averagings are necessary for the recording. It is important that the musculature is tense; for this the test person can raise his head from the support surface. Evaluation criteria are the presence of the waves P14 and N21 as well as their amplitude. Both their absence as well as a clear reduction in amplitude are considered pathological; the relevance of changes in latency must still be determined. The findings for individual illnesses are as follows. Vestibular neuritis: the VEMP is preserved in two-thirds of the patients. This is due to the sparing of the pars inferior of the vestibular nerve, which supplies the saccule and posterior canal, among others. Tullio phenomenon in cases of superior canal dehiscence syndromes or perilymph fistula: here there is a clearly lowered stimulus threshold, i.e., a stimulus reaction occurs already at low dB values. Bilateral vestibulopathy: the VEMP is absent in only a portion of the patients; this should be interpreted as a sign of additional damage of the saccule function. Menière's disease: the VEMPs are frequently reduced or absent. The clinical relevance of VEMP for general vestibular testing must still be evaluated.

Cogan's syndrome), hereditary (e.g., Mondini–Alexander dysplasia) or neoplastic (e.g., meningeosis carcinomatosa) inner ear diseases, as well as vestibular paroxysmia (due to neurovascular cross-compression), perilymph fistula, non-idiopathic vestibular neuritis (e.g., due to herpes zoster) or labyrinthine concussion. So far, BPPV, Menière's disease and vestibular neuritis cannot be diagnosed by means of imaging techniques.

• ***High-resolution Computed Tomography of the Petrous Bone***
The new generation of CT machines provides a considerably refined image quality due to improved scanning geometry and thinner slices. The spiral mode of operation (with 0.6-mm slice thickness) allows examination of the petrous bone with a high spatial resolution of $0.4 \times 0.4 \times 0.4$ mm. The data are reconstructed for each side separately. The possibility of reconstructing up to four intermediary steps per mm guarantees both good 2-D imaging as well as detailed 3-D surface reconstruction or 3-D multiplanar reconstructions. Compared with slice thicknesses of 1.5–2 mm, this high-resolution technique allows, for example, a better delimitation of the fracture lines. Slices in the axial plane

are used for CT of the petrous bone. Depending on the area to be assessed, for example, the skull base or the facial nerve canal, an axial plane slice should be complemented by a coronal plane slice. A high-resolution spiral CT of the petrous bone allows evaluation of the bony labyrinth, the facial nerve canal and the skull base. CT is indicated for determining the presence of a fracture, the superior canal dehiscence syndrome, malformations (e.g., Mondini–Alexander dysplasia), ossifications of the labyrinth in chronic illnesses (e.g., otosclerosis or Cogan's syndrome), as well as benign and malignant growth processes (e.g., cholesteatoma, cholesterol cysts, jugular diverticula, vestibular schwannoma, rhabdomyosarcoma, basaloma or adenocarcinoma).

- ***High-Resolution Magnetic Resonance Imaging of the Petrous Bone***

The MRI examination of the petrous bone and the cerebellopontine angle is performed in a circularly polarised head coil. This technique is clearly superior to CT as regards the imaging of non-osseous structures or lesions, e.g., tumors and inflammatory growths. The numerous anatomical structures confined in a very small space within the petrous bone make high demands on the MRI technology. The examination protocol should include the following (or corresponding) sequences:

- a proton and T2-weighted fast spin-echo sequence with a double echo in a 4-mm thick slice and an interslice distance of less than 0.8 mm for evaluating the brainstem and cerebellum,
- a T1-weighted sequence (e.g., 2-D fast low angle shot, FLASH) with a 2-mm thick slice and a spatial resolution of ca. 0.55 mm, and
- a high-resolution T2-weighted sequence (e.g., 3-D constructive interference in steady state, CISS) of ca. 0.5 mm spatial resolution and 0.7 mm slice thickness.

The slices should be acquired axially for all sequences; after administration of intravenous MRI contrast medium (0.1–0.2 mmol/kg), it is advisable to perform the high-resolution T1-weighted sequence in axial and coronal projections. If a neurovascular cross-compression is suspected, further complementary examinations are recommended, a 3-D MRI angiography sequence (e.g., time of flight) and a CISS sequence. By using the maximum intensity projection (MIP) procedure, it is possible to represent the signal-intense structures of the inner ear three-dimensionally and in any orientation. The MRI procedure of multiplanar reconstruction (MPR) permits free selection of the orientation of the planes to be reconstructed. MIP and MPR procedures can, however, only be used with 3-D data records.

1.5 General Principles of Therapy

The various forms of vertigo are treated with pharmacological therapy, physical therapy, surgery and psychotherapeutic measures (Table 1.10). Before treatment begins, the patient should be told that the prognosis is generally good, for two reasons:

- many forms of vertigo have a favourable natural course (e.g., the peripheral vestibular function improves or central vestibular compensation of the vestibular tonus imbalance takes place) and
- most forms can be successfully treated.

1.5.1 Pharmacological Therapy

So-called antivertiginous drugs, such as dimenhydrinate (Dramamine), the belladonna alkaloid scopolamine (Transderm Scop) and benzodiazepine (Valium, Klonopin), are indicated for the symptomatic treatment of dizziness and nausea only in the following cases:

- acute peripheral vestibulopathy (for a maximum of 1–3 days)
- acute brainstem lesion near the vestibular nuclei
- frequent and severe attacks of vertigo accompanied by nausea and vomiting
- severe BPPV with nausea and vomiting (0.5 h before the liberatory manoeuvres)
- prevention of motion sickness
- central positional/positioning vertigo with vomiting.

None of these pharmaceuticals are suitable for long-term treatment, for example, in cases of chronic (central vestibular) vertigo, archicerebellar ataxia, or certain forms of positional vertigo. Besides antivertiginous drugs, other pharmaceuticals are increasingly being used effectively to treat other forms of vertigo, such as beta-receptor blockers for basilar/vestibular migraine, baclofen and 4-diaminopyridine for downbeat nystagmus and familial episodic ataxia type 2, and carbamazepine for vestibular paroxysmia. An overview of the drugs for the various forms of vertigo has been compiled in Tables 1.10 and 1.11.

1.5.2 Physical Therapy: Balance Training and Liberatory Manoeuvres

A specific form of balance training is performed to improve central vestibular compensation in cases of peripheral and central vestibular damage. It entails special exercises for the vestibular, somatosensory and ocular motor systems in order to, for

Table 1.10. Drug, physical and surgical therapies for vertigo

Therapy	Indication
Drug	
• Anticonvulsants	—vestibular epilepsy —vestibular paroxysmia (neurovascular compression) —other central vestibular paroxysmias —myokymia of the superior oblique muscle
• Antivertiginous substances	—for symptoms of nausea and vomiting in cases of acute labyrinthine lesion or lesions of the vestibular nerve/nuclei, central "positional vomiting", severe attacks with vomiting after liberatory manoeuvre in BPPV —prevention of motion sickness
• Beta-receptor blocker	—basilar/vestibular migraine
• Betahistine	—Menière's disease
• Corticosteroids	—vestibular neuritis
• Ototoxic antibiotics	—Menière's disease —vestibular drop attacks
• Baclofen, 3,4-diaminopyridine and 4-aminopyridine	—downbeat nystagmus (upbeat nystagmus)
• Acetazolamide and 4-aminopyridine	—familial episodic ataxia type 2
• Selective serotonin reuptake inhibitors	—phobic postural vertigo
Physical	
• Liberatory/positional manoeuvre	—benign paroxysmal positioning vertigo
• Vestibular exercises	—improvement of the central vestibular compensation of a vestibular tonus imbalance (e.g., unilateral labyrinthine loss) —habituation to prevent motion sickness
Surgical	
• Removal or resection of intracranial mass	—tumours or arachnoidal cysts of the posterior cranial fossa (vestibular schwannoma)
• Surgical patching	—perilymph fistulas
• Section of canal nerves or obliteration of canal	—benign paroxysmal positioning vertigo (ultima ratio)
• Labyrinthectomy or section of the vestibular nerve	—Menière's disease (ultima ratio)
• Neurovascular decompression	—vestibular paroxysmia (ultima ratio)

Table 1.11. Antivertiginous and antiemetic drugs

Drug	Dosage	Action
Anticholinergics		
• Scopolamine (Transderm Scop)	Transdermal 1.0 mg/72 h	Muscarine antagonist
Antihistamines		
• Dimenhydrinate (Dramamine)	Tabl. (50 mg) every 4–6 h Suppos. (100 mg) 1–2/day	Histamine (H_1) antagonist
• Meclizine (Antivert)	Tabl. (25 mg) every 4–6 h Suppos. (50 mg) 1–2/day	
Benzodiazepines		
Diazepam (Valium)	Tabl. (5 or 10 mg) every 4–6 h or injected solution of 10 mg i.m.	$GABA_A$ agonist
Clonazepam (Klonopin)	Tabl. (0.5 mg) every 4–6 h	

example, promote substitution by other systems for the missing vestibular information. The efficacy of this therapy has been proven in animal and clinical experiments, both for acute lesions (e.g., acute unilateral labyrinthine failure due to vestibular neuritis) as well as for chronic conditions (e.g., due to a vestibular schwannoma). The positioning exercises or liberatory manoeuvres used in BPPV lead within a few days to the absence of complaints in almost all cases.

1.5.3 Surgery

If vertigo is caused by a tumour or a cavernoma of the brainstem, for example, surgery (or gamma knife) is the primary treatment option. Otherwise, surgery is necessary only in very rare cases of Menière's disease or vestibular paroxysmia (recurrent severe attacks of rotatory vertigo of short duration caused by neurovascular cross-compression) when drug therapy fails. Surgery should also be considered in cases of perilymph fistulas. In most of the other forms of vertigo, surgery is rarely indicated.

1.5.4 Psychological/Psychiatric Treatment

Phobic postural vertigo is the second most frequent type of vertigo in our dizziness outpatient unit, and as such its treatment is of special importance. This consists of behavioural therapy involving desensitisation by self-exposure to the precipitating situations.

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2

Peripheral Vestibular Forms of Vertigo

Three forms of peripheral vestibular disorders, each with its typical symptoms and clinical signs, can be differentiated:

- **Bilateral peripheral loss of vestibular function (bilateral vestibulopathy).** The main symptoms are oscillopsia during head movements (failure of the vestibulo-ocular reflex) and instability of gait and posture. The latter two symptoms increase in darkness and on uneven ground (due to reduced or absent visual or somatosensory information).
- **Acute/subacute unilateral failure of vestibular function** (labyrinth and/or vestibular nerve), which causes a vestibular tonus imbalance. Main symptoms are rotatory vertigo (for a few days or weeks), nausea, oscillopsia and a tendency to fall in a certain direction.
- **Inadequate paroxysmal stimulation of the peripheral vestibular system** of the labyrinth (e.g., during benign paroxysmal positioning vertigo) or of the vestibular nerve (e.g., during vestibular paroxysmia due to ectopic discharges). The main symptoms are attacks of rotatory or postural vertigo.

2.1 Benign Paroxysmal Positioning Vertigo

2.1.1 Patient History

The main symptoms of benign paroxysmal positioning vertigo (BPPV) include brief, sometimes severe attacks of rotatory vertigo with and without nausea, which are caused by rapid changes in head position relative to gravity. Typical triggers

include lying down or sitting up in bed, turning around in bed, and also bending over to tie the shoelaces, or extending the head in order to look up or do something above the head. If BPPV is elicited while the patient is upright, he is in danger of falling. Attacks of vertigo frequently occur in the morning and are most pronounced during the first change in position after sleep; repeated changes in position cause a transient lessening of the symptoms. The complaints are so typical that a diagnosis can often be made solely on the basis of the patient history; occasionally even the affected ear can be identified ("rotatory vertigo only occurs when I lie on my right side").

2.1.2 Clinical Features and Course

Benign paroxysmal positioning vertigo is the most common cause of vertigo, not only in the elderly (Table 1.1, p. 5). It is so frequent that about one-third of all over 70-years old have experienced BPPV at least once. This condition is characterised by brief attacks of rotatory vertigo and simultaneous positioning rotatory-linear nystagmus toward the undermost ear. It can be accompanied by nausea. BPPV is elicited by extending the head or positioning the head or body toward the affected ear. Rotatory vertigo and nystagmus occur after such positioning with a short latency of seconds in the form of a crescendo/decrescendo course of maximally 30–60 seconds. The beating direction of the nystagmus depends on the direction of gaze; it is primarily rotating when gaze is to the undermost ear and mostly vertical (to the forehead) during gaze to the uppermost ear. The nystagmus corresponds to an (ampullofugal) excitation of the posterior canal of the undermost ear.

Benign paroxysmal positioning vertigo can appear at any time from childhood to senility, but the idiopathic form is typically a disease of old age, peaking in the sixth to seventh decades. More than 90% of all cases are classified as degenerative or idiopathic (women : men = 2 : 1), whereas the symptomatic cases (women : men = 1 : 1) are most frequently caused by head injury (17%) or vestibular neuritis (15%). BPPV also occurs strikingly often in cases of extensive bed rest in connection with other illnesses or after operations. About 10% of the spontaneous cases and 20% of the trauma cases show a bilateral, generally asymmetrically pronounced BPPV.

It is called benign because it generally resolves spontaneously within weeks to months; in some cases, however, it can last for years. We found that the history of the disorder until its diagnosis lasted more than 4 weeks in 50% of our patients and more than 6 months in 10%. If not treated, BPPV persisted in about 30%

Table 2.1. Clinical features differentiating a benign paroxysmal positioning vertigo (BPPV) from a central positioning vertigo/nystagmus (CPV)

Feature	BPPV	CPV
Latency following precipitating positioning manoeuvres	1–15 s (shorter in horizontal BPPV, hBPPV)	No latency or 1–5 s
Vertigo	Typical	Typical
Duration of attack	5–40 s (longer in hBPPV and in rare cupulolithiasis)	5–>60 s
Direction of nystagmus	Torsional-vertical with head positioning in the plane of the posterior (pBPPV) or the anterior (aBPPV) canal, horizontal with head positioning in the plane of the horizontal (hBPPV) canal	Purely vertical or torsional, combined torsional/linear, direction of nystagmus does not correspond with the plane of the canal stimulated by the head movement
Course of vertigo and nystagmus in the attack	Crescendo/decrescendo (with typical canalolithiasis)	Crescendo/decrescendo is possible
Nausea and vomiting	Rare with single head positioning manoeuvres (if so, then associated with intense positioning nystagmus); frequent with repeated manoeuvres	Frequent with single head positioning manoeuvres (not necessarily associated with intense nystagmus)
Natural course	Spontaneous recovery within days to months in 70%–80%	Dependent on aetiology, spontaneous recovery within weeks in most cases
Associated neurological signs and symptoms	None (in idiopathic BPPV)	Frequent cerebellar and ocular motor signs such as ataxia, saccadic pursuit, gaze-evoked nystagmus, downbeat nystagmus, impaired fixation suppression of the VOR
Brain imaging	Normal	—Lesions dorsolateral to the fourth ventricle and/or the dorsal vermis (tumour, haemorrhages, infarctions, or multiple sclerosis plaques) —Less specific lesions (cerebellar degeneration, paraneoplastic syndromes, encephalopathy, intoxication)

of our patients; another 20–30% had relapses within months or years (recurrence risk of 15% per year).

2.1.3 Pathophysiology and Therapeutic Principles

According to the histologically based cupulolithiasis model of Schuknecht (1969), heavy, anorganic particles (otoconia) of specific weight, which become detached as a result of trauma or spontaneous degeneration from the utricular otoliths of the cupula, settle in the underlying ampulla of the posterior canal. Whereas the cupula normally has the same specific weight as the endolymph, it is heavier with these particles, i.e., the canal is transformed from a sensor of rotatory acceleration into a transducer of linear acceleration. This hypothesis was generally accepted for many years, despite its inability to explain many of the typical criteria of nystagmus in cases of positioning vertigo.

In contrast, the canalolithiasis hypothesis, discussed by Parnes and McClure (1991) and Epley (1992) and proven by Brandt and Steddin (1993), can explain all symptoms of positioning nystagmus. According to this hypothesis, the particles float freely within the endolymph of the canal instead of being firmly attached to the cupula, and the "heavy conglomerate", which almost fills the canal, is assumed to be the cause of the positioning vertigo. The movement of the conglomerate causes either an ampullofugal or an ampullopetal deflection of the endolymph depending on the direction of the sedimentation. A valid model of the pathomechanism of BPPV must be able to predict the direction, latency, duration and fatigability of the typical nystagmus, as well as changes in these parameters due to other head manoeuvres (Figure 2.1).

- ***Latency***

Rotatory vertigo and nystagmus develop as soon as the particles in the canal precipitate due to gravity. This causes a deflection of the cupula, which exceeds the stimulus threshhold of the sensory epithelium after 1–5 seconds.

- ***Duration***

The particles move to and precipitate at the lowest point within the canal after the change in position. Depending on their size and composition, this requires about 10 seconds.

- ***Course of Attacks***

After the positioning, the particles precipitate away from the canal wall and are accelerated by the forces of gravity. The particles are accelerated from standstill, reach a maximal speed

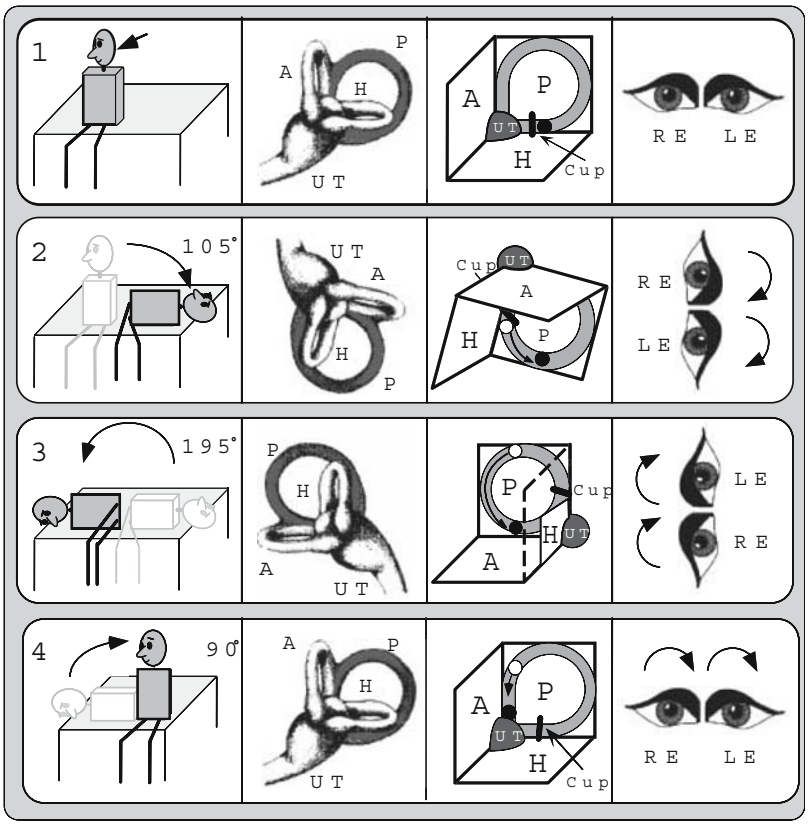

Figure 2.1. Schematic drawing of the Semont liberatory manoeuvre of a patient with typical benign paroxysmal positioning vertigo of the left ear. Panels from left to right: position of body and head, position of labyrinth in space, position and movement of the clot in the posterior canal (which causes cupula deflection) and the direction of the rotatory nystagmus. The clot is depicted as an *open circle* within the canal; a *black circle* represents the final resting position of the clot. (*1*) In the sitting position, the head is turned horizontally 45° to the unaffected ear. The clot, which is heavier than endolymph, settles at the base of the left posterior semicircular canal. (2) The patient is tilted approximately 105° to the left (affected) ear. The change in head position, relative to gravity, causes the clot to gravitate to the lowermost part of the canal and the cupula to deflect downward, inducing BPPV with rotatory nystagmus beating toward the undermost ear. The patient maintains this position for 1 min. (*3*) The patient is turned approximately 195° with the nose down, causing the clot to move toward the exit of the canal. The endolymphatic flow again deflects the cupula such that the nystagmus beats toward the left ear, now uppermost. The patient remains in this position for 1 min. (*4*) The patient is slowly moved into the sitting position; this causes the clot to enter the utricular cavity. (From Brandt et al. 1994.) *A*, *P*, *H* anterior, posterior, horizontal semicircular canals, *Cup* cupula, *UT* utricular cavity, *RE* right eye, *LE* left eye.

during their fall, and return to standstill at the lowest point in the canal. This explains the temporal crescendo–decrescendo-like course of the attacks; the cupula time constant increases the duration of the nystagmus and vertigo.

- ***Direction of Nystagmus***

The ampullofugal stimulation of the posterior canal causes eye movements around the axis of ocular rotation, which is perpendicular to the canal plane, by means of the vestibulo-ocular reflex (VOR). To the physician this will appear to be a combination of linear (toward the forehead and the undermost ear) and rotatory eye movements.

- ***Reversal of Nystagmus***

If the direction of the positioning movement is reversed when sitting up, the particles move in the opposite direction. Now the cupula is deflected in the opposite (ampullopetal) direction. This results in reversal of both the rotatory vertigo and the direction of nystagmus due to inhibition of the vestibular hair cells.

- ***Fatigability***

The loose particles form a plug or clump, which falls apart more and more during changes in the head position. Small particles cannot cause suction or pressure on the cupula independently of each other, as does a single clump, whose volume almost fills the canal. If the patient holds his head still for several hours (e.g., during sleep), the particles, which had fallen apart before, coalesce into a clump in the lowest place within the canal and again induce vertigo when the head position is changed.

- ***Liberatory Manoeuvre***

The efficacy of positioning (liberatory) manoeuvres of the head can only be explained by canalolithiasis, i.e., the clot moves freely within the canal. As a result of these manoeuvres, the plug is washed out of the canal and then cannot cause any positioning vertigo (Brandt and Steddin 1993; Brandt et al. 1994). Proceeding from the explanations of cupulolithiasis or canalolithiasis, Brandt and Daroff (1980) were the first to devise an effective exercise programme, which, by means of the simple physical measure of head positioning, loosens the heavy degenerative otolithic material and distributes it into other areas of the labyrinth, where it comes to rest and no longer impairs canal function. Originally the exercises were based on the concept of cupulolithiasis. Nowadays, in accordance with the modification of Semont et al. (1988), we recommend that the patient's position be changed from the inducing position by a tilt of 180° to the opposite side (Figure 2.1). In 1992, Epley proposed another liberatory manoeuvre that

involved turning the patient from a supine position into a head-hanging position (Figure 2.2). All these manoeuvres are effective (Hilton and Pinder 2002; Radke et al. 2004) and can be explained by the mechanism of canalolithiasis (Brandt et al. 1994). Only in very rare cases that are refractory to these manoeuvres should surgery, i.e., the obliteration of the canal, be considered (Parnes and McClure 1991).

2.1.4 Pragmatic Therapy

- ***Physical Liberatory Manoeuvres***

When correctly performed, all three physical liberatory manoeuvres (Semont or Epley liberatory manoeuvres, Brandt–Daroff exercises; Figures 2.1, 2.2 and 2.3) are successful in almost all patients (Herdman et al. 1993). We recommend Semont's liberatory manoeuvre depicted in Figure 2.1 as the therapy of first choice. The three positioning steps are performed quickly with the aid of a therapist as the patient lies on the examination couch and then by the patient himself at home. It is important that the head of the sitting patient is turned by 45° to the healthy ear, in order to put the responsible posterior canal into a position parallel to the plane of movement during the positioning. Relief is thus achieved in about 90% of cases within one week.

The positioning nystagmus toward the uppermost ear (Figure 2.1, panel 3) indicates that the plug has left the canal, i.e., the therapy was successful. Conversely, positioning nystagmus toward the undermost healthy ear indicates that the liberatory manoeuvre failed and must be repeated (Figure 2.4).

Epley's alternative liberatory manoeuvre requires that the patient's head and trunk be rotated after being tilted backward into a slightly head-hanging position (see the four positioning steps in Figure 2.2). This manoeuvre is as effective as Semont's. If, however, the plug is not dislodged during the outpatient visit, the patient can be quickly instructed how to proceed on his own at home. Series of these exercises should be performed five to ten times per day, preferably three times during the early morning and three times at noon. The manoeuvres seem to be most effective then, since the clot, which develops during rest, can be more easily removed from the canal than single otoconia. As a rule almost all patients are free of complaints after several days or sometimes a few weeks (Hilton and Pinder 2002; Levat et al. 2003). Despite successful liberatory manoeuvres, many patients complain of transient postural vertigo and dizziness afterwards. This can be explained by the partial repositioning of the otoconia toward the otolith organs (i.e., most likely an otolithic vertigo). Patients should be informed in advance about this side-effect of the manoeuvres, which goes away within a few days.

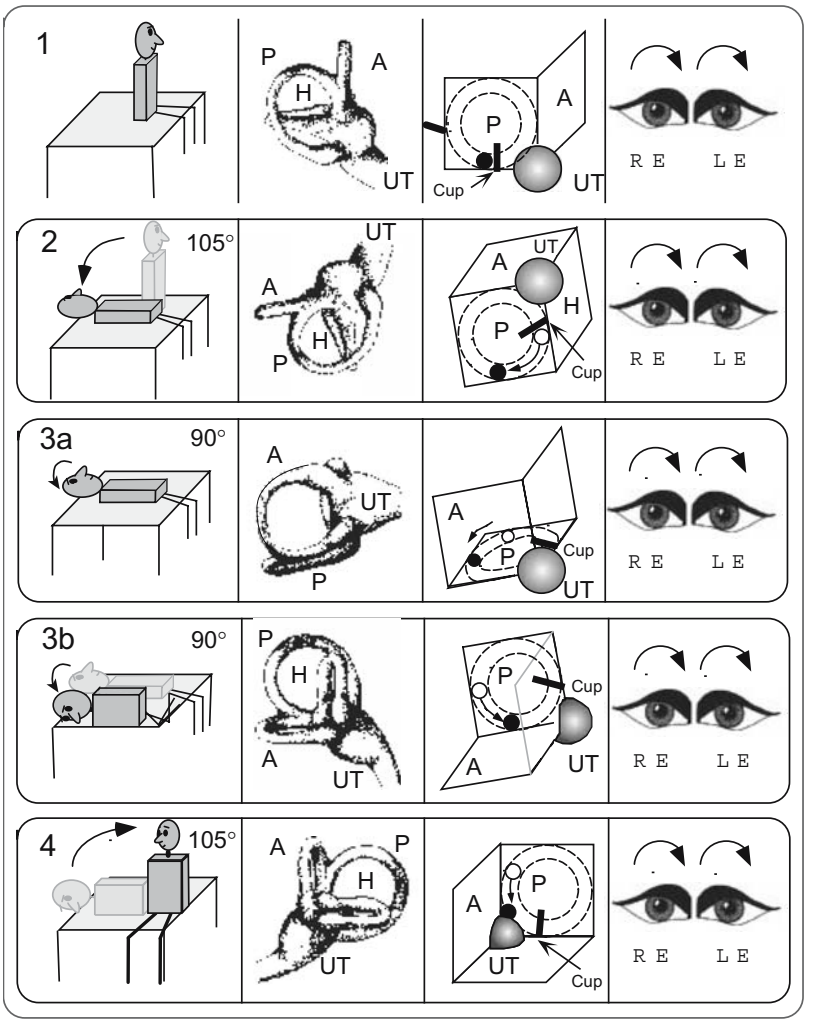

Figure 2.2. Schematic drawing of modified Epley liberatory manoeuvre. Patient characteristics and abbreviations are as in Figure 2.1. (*1*) In the sitting position, the head is turned horizontally 45° to the affected (left) ear. (*2*) The patient is tilted approximately 105° backward into a slightly head-hanging position, causing the clot to move in the canal, deflecting the cupula downward, and inducing the benign paroxysmal positioning vertigo. The patient remains in this position for 1 minute. (*3a*) The head is turned 90° to the unaffected ear, now undermost, and (*3b*) the head and trunk continue turning another 90° to the right, causing the clot to move toward the exit of the canal. The patient remains in this position for 1 min. The positioning nystagmus beating toward the affected (uppermost) ear in positions 3a and 3b indicates effective therapy. (*4*) The patient is moved into the sitting position. (From Brandt et al. 1994.)

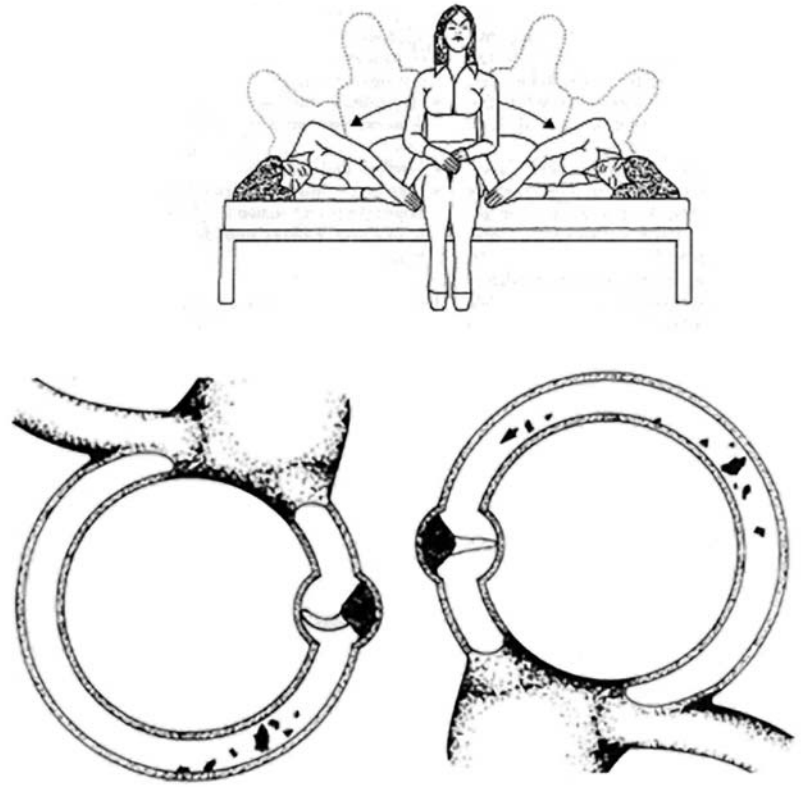

Figure 2.3. Schematic drawing of a positioning manoeuvre of a patient with benign paroxysmal positioning vertigo (after Brandt and Daroff 1980). *Above* are shown the initial sitting position and the side positioning with somewhat oblique head position; each position should be held for 20–30 seconds for physical therapy. These positionings are performed serially several times a day. *Below*: a schematic drawing of canalolithiasis.

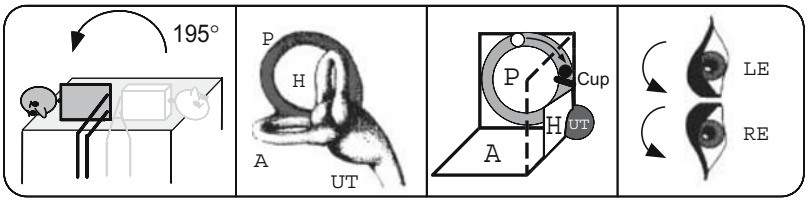

Figure 2.4. Schematic drawing of an ineffective liberatory manoeuvre (compare with Figure 2.1, panel 3). After the patient with left-sided benign paroxysmal positioning vertigo is tilted from the symptomatic position to the right, the particles do not leave the canal but sediment once again ampullopetally onto the cupula. This causes an ampullopetal cupula deflection with positioning nystagmus, which in this position beats downward toward the unaffected right ear. This indicates that the liberatory manoeuvre has failed and must be repeated (Brandt et al. 1994).

Following effective physical liberation, approximately 50% of patients will experience a recurrence of attacks; most often patients have positioning vertigo in the morning of the day after the manoeuvres. About 20% of such attacks occur in the first two weeks. The estimated recurrence rate per year is 15% and the recurrence rate of BPPV 40 months after treatment is about 50%. This high recurrence rate emphasizes the need for patient counseling. The recurrences are most likely due to reentry of the debris into the posterior canal from the utricular cavity. They should be treated with the same manoeuvre that induced resolution of the initial episode.

- ***Surgery***

In our experience with more than 2,000 BPPV patients, only one proved to be refractory to therapy and required operative sectioning of the posterior canal nerves. Such selective neurectomy (Gacek 1978) is difficult, and there is a risk of a permanent hearing loss. Neurectomy has now been replaced by plugging of the posterior canal (Parnes and McClure 1991; Pace-Balzan and Rutka 1991). It is evidently a safer and more effective measure than sectioning of the nerve; however, in our opinion it is used too often in some centres, i.e., before the possibilities of the simple, effective physical therapy have been exhausted.

- ***Ineffective Measures***

Because of the pathomechanism of BPPV, drug treatment with antivertiginous substances is neither possible, nor are drugs sufficiently effective against the symptoms in the long term. The exception is sensitive patients who have severe nausea after a single manoeuvre. In this case the administration of, for example, dimenhydrinate (100 mg) half an hour before performing the liberatory manoeuvres can make therapy easier.

2.1.5 Benign Paroxysmal Positioning Vertigo of the Horizontal Canal

Benign paroxysmal positioning vertigo of the horizontal canal (McClure 1985; Baloh et al. 1993) is less frequent than posterior BPPV but is still diagnosed too seldom. Its cardinal features differ from those of posterior BPPV:

- It can be induced by turning the head along the longitudinal axis of the supine body (either to the right or to the left) (Figure 2.5). This results in an ampullopetal deflection of the cupula (with more severe vertigo and nystagmus) when the head is turned to the side of the affected ear.
- The beating direction of nystagmus corresponds to the stimulation or inhibition of the horizontal canal, i.e., it beats linear and horizontal to the undermost ear.

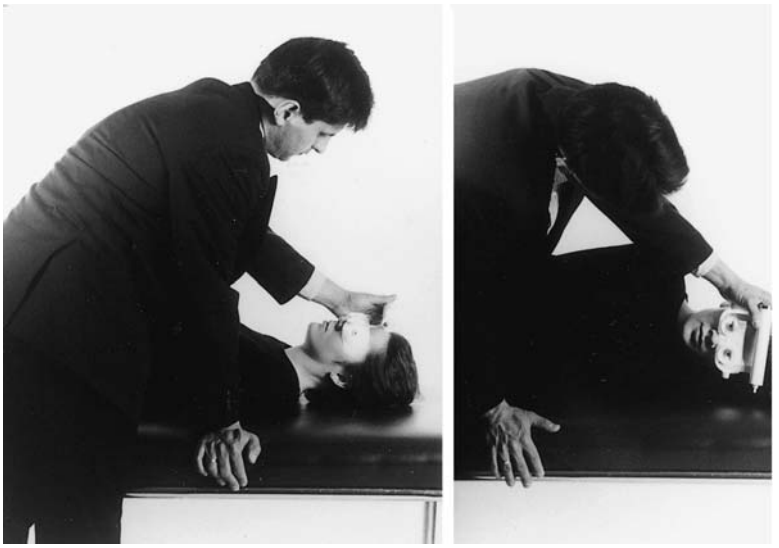

Figure 2.5. Precipitating positioning manoeuvres for benign paroxysmal positioning vertigo of the horizontal semicircular canal (hBPPV) by turning the head (to the right as well as to the left) along the longitudinal axis of the body while the subject is supine. When the head is turned to the side of the affected ear, there is an ampullopetal deflection of the cupula (and thus stimulation of the vestibular hair cells). This causes a more severe vertigo and nystagmus than when turned to the unaffected ear.

- Repeated positioning manoeuvres cause hardly any fatigue of the positioning nystagmus.
- The duration of the attacks and the nystagmus is longer because of the so-called central storage mechanism of velocity for the horizontal canal. Positioning nystagmus frequently shows a reversal of direction during the attacks; this corresponds to post-rotatory nystagmus (so-called PI and PII).

The typical case of horizontal BPPV can also only be explained by canalolithiasis (Strupp et al. 1995), although it has occasionally been observed that the mechanism switches from canalolithiasis to cupulolithiasis (Steddin and Brandt 1996). In the rare form of horizontal BPPV due to cupulolithiasis (characterised by nystagmus beating horizontally to the uppermost ear), the "zero point" of positional nystagmus (beyond which direction changes) can be determined by turning the patient's head 10–20° around the longitudinal axis while in the supine position; this is possible because the cupula of the ipsilateral horizontal canal is then parallel to the gravity vector (Bisdorff and Debatisse 2001). In this way one can also determine which side is affected by horizontal BPPV.

There is strong evidence that persistent horizontal BPPV occurs when there is a certain narrowness of the canal and the congealed clump cannot leave the canal, which narrows toward its exit in an ampullofugal direction, because of its size. Otherwise it could be assumed that the particles would independently and inevitably leave the canal with every accidental rotation around the longitudinal axis of the body (e.g., in bed). The striking feature of horizontal BPPV, i.e., it does not fatigue, agrees with this assumption, as does the general experience that horizontal BPPV is more difficult to treat than posterior BPPV.

Serial and alternating positionings according to the method of Brandt and Daroff (1980) are more likely to lead to success (Herdman et al. 1993), as the repeated bilateral head positionings evidently cause the disintegration of the conglomerate and wash out the particles from the canal. Lempert and Tiel-Wilck (1996) have also reported success with simple 270° rotations toward the unaffected ear around the longitudinal axis while the patient is supine. Bed rest with positioning of the head to the side of the unaffected ear (for 12 hours) is evidently more effective (Vannucchi et al. 1997). We recommend a combination of both methods.

2.1.6 Differential Diagnosis and Clinical Problems

The diagnosis of BPPV can in most cases be made on the basis of a typical patient history (brief rotatory vertigo when turning over or sitting up/lying down in bed) and the clinical findings. Especially in cases of therapy-refractory rotatory vertigo (despite correct positioning exercises), the following syndromes should be considered along with unilateral BPPV in the differential diagnosis:

- central positional nystagmus (infrequent, see below)
- bilateral BPPV, particularly post-traumatic (ca. 10%)
- BPPV of the horizontal canal (too rarely diagnosed, see above)
- vestibular paroxysmia (see Section 2.4)
- central infratentorial lesions that mimic BPPV (very rare, see Table 2.1).

2.1.7 Central Positional Vertigo/Nystagmus

Central positional nystagmus and central positional vertigo are caused by infratentorial lesions, which involve connections between the vestibular nuclei in the medulla oblongata and cerebellar structures close to the midline (vermis). It is important to distinguish between peripheral and central vestibular disorders, as the latter require further laboratory diagnostics. Four characteristic forms of central positional vertigo/nystagmus can be

distinguished, although the symptoms overlap and combinations occur:

- central downbeat nystagmus, typically in head-hanging position (with or without accompanying vertigo)
- central positional nystagmus (without vertigo)
- central paroxysmal positional/positioning vertigo with nystagmus
- "central positioning vomiting".

These central vestibular disorders occur much more seldom than the typical BPPV. However, it can be difficult to distinguish peripheral and central function disorders in the individual patient (Bertholon et al. 2002). The following clinical rules are important for diagnosing a central positional vertigo/nystagmus (Table 2.1) (Büttner et al. 1999):

- persisting positional nystagmus (slow-phase velocity >5°/s) without associated vertigo
- positioning-induced vomiting after single head movements without any substantial vertigo or nystagmus
- positional/positioning vertigo with nystagmus of purely torsional or vertical character (downbeat or upbeat directions); a purely horizontal direction of nystagmus is typical for hBPPV
- positional/positioning nystagmus that does not correspond to the plane of the semicircular canal stimulated by the head positioning (e.g., torsional nystagmus after stimulation of the horizontal canal). In practice this seems to be the most important feature by which a central positional nystagmus can be identified.

According to earlier rules, positional nystagmus beating toward the uppermost ear or lasting longer than 1 minute indicated a central pathology; this is no longer considered a reliable differentiating feature, as both occur with the cupulolithiasis variant of BPPV.

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2.2 *Vestibular Neuritis (Acute Partial Unilateral Vestibular Deficit)*

2.2.1 Patient History

The main symptoms of acute unilateral vestibular deficit are a sustained violent rotatory vertigo, oscillopsia, gait and postural imbalance with a tendency to fall, as well as nausea and vomiting. All of these symptoms have an acute or subacute onset and last for a few days or weeks. Hearing disorders or other neurological deficits are not present. There are no typical antecedent signs or triggers, except for occasional spells of vertigo a few days before in some patients. As the patients' complaints are exacerbated by any movements of the head, they intuitively seek peace and quiet.

2.2.2 Clinical Features and Course

Acute unilateral vestibular deficit with severe, persistent rotatory vertigo, nystagmus, a tendency to fall and vomiting lasting for days is the second most common cause of peripheral vestibular vertigo (after BPPV). The disease is analogous to sudden hearing loss and idiopathic facial paresis. The head-impulse (VOR) test (Halmagyi and Curthoys 1988) and caloric testing show hypo- or non-responsiveness of the ipsilateral horizontal semicircular canal. The difference in tonus between the neural signals from homologous receptors of both labyrinths causes a vestibular tonus imbalance which leads to a horizontal-rotatory spontaneous nystagmus, a rotatory vertigo to the healthy side, and an obvious tendency to fall to the lesioned side. The clinical syndrome of vestibular neuritis is accordingly characterised by (Figure 2.6):

- persistent rotatory vertigo (contraversive) with pathological tilting of the subjective visual vertical (ipsiversive)
- horizontal-rotatory spontaneous nystagmus (contraversive) with illusory movements of the surroundings (oscillopsia)
- gait deviation, tendency to fall, and past-pointing (ipsiversive)
- nausea and vomiting
- unilateral functional deficit of the horizontal canal, which can be detected by the Halmagyi–Curthoys head-impulse (VOR) test and caloric irrigation.

Vestibular neuritis affects adults most often between 30 and 60 years of age. Short attacks of rotatory vertigo occasionally precede the onset of manifest loss of function by a few days. Major complaints of vertigo, nausea, imbalance and a tendency to fall resolve slowly over a few weeks, and within 3–5 weeks

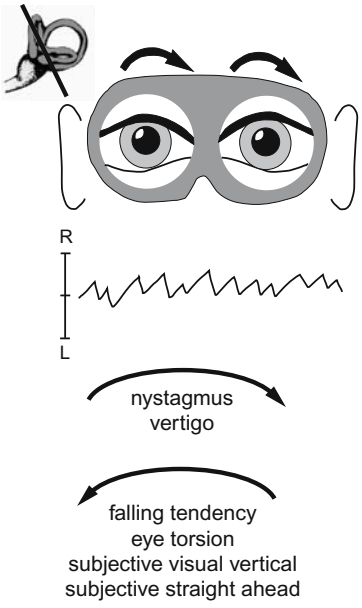

Figure 2.6. Symptoms and clinical signs of acute unilateral labyrinthine deficit. Spontaneous nystagmus (quick phase) and rotatory vertigo to the unaffected side, accompanied by a tendency to fall, ocular torsion, and deviation of the subjective visual vertical and the subjective straight-ahead to the affected side.

the patient is generally free of them at rest. Recovery is the result of a combination of:

- restoration of peripheral labyrinthine function (frequently incomplete),
- substitution of the functional loss by the contralateral vestibular system as well as by somatosensory (neck proprioception) and visual afferents, and
- central compensation of the peripheral vestibular tonus imbalance.

In the course of the illness only about 40% of patients have complete recovery of peripheral vestibular function after 24 months (Okinaka et al. 1993), in 20–30% there is only partial recovery, and the rest have a persisting unilateral defect. Even in cases in which the peripheral deficit remains, all "static" symptoms (without head movement) resolve, such as spontaneous nystagmus, vertigo and a tendency to fall. The remaining deficit, however, is still manifest in the form of a "dynamic" dysfunction: retinal slip of images of the visual scene with oscillopsia during rapid, high-frequency head movements and walking and running because of the insufficiency of the VOR (Halmagyi and Curthoys 1988).

2.2.3 Aetiology, Pathophysiology and Therapeutic Principles

Analogously to "idiopathic facial paresis", vestibular neuritis most likely has a viral aetiology, but this has not yet been proven

(Schuknecht and Kitamura 1981; Nadol 1995; Baloh et al. 1996; Gacek and Gacek 2002). Arguments supporting a viral aetiology are its endemic occurrence at certain seasons, autopsy studies (which have shown inflammatory degeneration of the vestibular nerves), proof of elevated protein levels in the cerebrospinal fluid, and the presence of herpes simplex virus DNA and RNA as well as the latency-associated transcript in the vestibular ganglia (Figure 2.7) (Arbusow et al. 1999; Theil et al. 2001, 2002).

Evidently vestibular neuritis tends to affect the superior portion of the vestibular nerves, which supplies the horizontal and anterior canals as well as the utricle and parts of the saccule. This means that vestibular neuritis is not equivalent to a total vestibular deficit (Büchele and Brandt 1988), as was previously suspected due to the co-occurrence of vestibular neuritis and BPPV in the same ear; this was later confirmed by Fetter and Dichgans (1996) with three-dimensional (3-D) analysis of the canal function. Rare variants have been described, e.g., "inferior vestibular neuritis" (here there is a selective deficit of the posterior canal combined with a sparing of the lateral and anterior canals) (Halmagyi et al. 2002) and a form in which a dysfunction of the posterior canal is combined with one of the cochlea. The latter probably does not have a viral but rather a vascular aetiology, since both structures have a common vascular supply. Despite the absence of vestibular-evoked myogenic potentials, normal galvanic-evoked myogenic potentials were found in

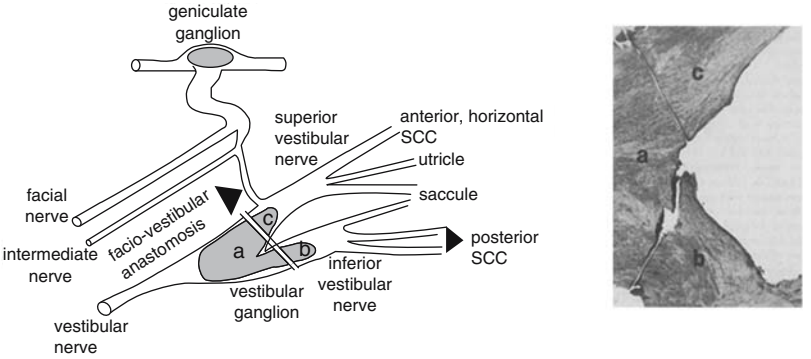

Figure 2.7. Schematic drawing of the vestibular and facial nerves, the facio-vestibular anastomosis, the geniculate ganglion and different sections of the vestibular ganglion (*a* stem, *b* inferior portion, *c* superior portion). *Right*: longitudinal cryosection of a human vestibular ganglion, in which the individual portions are separated. The presence of herpes simplex virus-1 DNA was confirmed by polymerase chain reaction in ca. 60% of all human vestibular ganglia. Moreover, the double innervation of the posterior canal, which may lead to the preservation of its function during vestibular neuritis, is visible (Arbusow et al. 1999). *SCC*, semicircular canal.

some patients with vestibular neuritis; this finding is compatible with the presence of labyrinthine lesions (Murofushi et al. 2003).

- ***Causal Therapy***

A recent prospective randomised study in 141 patients with placebo, methylprednisolone, valacyclovir, and methylprednisolone plus valacyclovir groups showed that monotherapy with corticosteroids significantly improved the peripheral vestibular function of patients with vestibular neuritis (follow-up time: 12 months). There was no evidence for synergy between methylprednisolone and valacyclovir (Strupp et al., 2004) despite the assumed viral etiology. It is conceivable that the replication of the HSV-1 in the vestibular ganglia had already occurred by the time that the antiviral agent was applied, i.e., within 3 days after symptom onset. This is supported by findings of studies on HSV-1 encephalitis, which showed that the most relevant prognostic factor is early acyclovir treatment, i.e., within 2 days after admission to the hospital. As in Bell's palsy, the benefit of corticosteroids might be explained by their anti-inflammatory effects, which reduce the swelling that causes a mechanical compression of the vestibular nerve within the temporal bone. Thus, steroids but not antiviral agents should be recommended as a treatment for acute vestibular neuritis, as they cause a significant functional improvement.

Glucocorticoids not only have an effect on the inflammatory process; they also promote central compensation of a unilateral labyrinthine deficit, at least in animal experiments (Yamanaka et al. 1995; Jerram et al. 1995).

- ***Symptomatic Therapy***

Antivertiginous drugs should only be administered during the first days and only in cases of severe nausea and vomiting, as they delay central compensation of the peripheral vestibular deficit (Zee 1985; Curthoys 2000).

- ***Improvement of Central Vestibular Compensation***

So far the most important principle of therapy is to promote central compensation by means of physical therapy. This so-called central compensation is not a uniform process. It involves various neural and structural mechanisms that operate in different locations (vestibulospinal or vestibulo-ocular structures) within different time courses, have various limited possibilities, and cause incomplete results, especially as regards head oscillations at high frequencies (Brandt et al. 1997). Central compensation of a unilateral labyrinthine lesion is enhanced and increased if movement stimuli trigger inadequate and incongruent afferent

signals that provoke a sensory mismatch. Vestibular exercises, first recommended by Cawthorne (1944) and later modified according to new knowledge of vestibular function (Hamann 1988; Brandt 1999; Strupp et al. 1998; Herdman 2000), include the following:

- voluntary eye movements and fixation to improve impaired gaze stabilisation
- active head movements to recalibrate the VOR
- balance training, goal-directed movements and gait exercises to improve the vestibulospinal regulation of posture and goal-directed motion.

Animal experiments have proven the efficacy of exercises to promote central compensation of spontaneous nystagmus and to counteract the tendency to fall after unilateral labyrinthine lesions (Igarashi 1986). A prospective, randomised controlled study (Strupp et al. 1998) has demonstrated that intensive physiotherapy significantly improved vestibulospinal compensation in patients with vestibular neuritis (Figure 2.8).

Pharmacological and metabolic studies in animals suggest that alcohol, phenobarbital, chlorpromazine, diazepam and ACTH antagonists retard central compensation, whereas caffeine, amphetamines and ACTH accelerate it (Zee 1985; Curthoys 2000). So far no clinical studies have been performed on this subject (overview in Strupp et al. 2001).

2.2.4 Pragmatic Therapy

- ***Symptomatic Therapy***

In the acute phase during the first 1–3 days, 100 mg dimenhydrinate (Dramamine, one to three suppositories per day) or other antivertiginous drugs (Table 1.11, p. 39) can be given to suppress nausea and vomiting. Drugs should be stopped as soon as the patient no longer vomits, as they prolong the time required to achieve central compensation.

- ***Causal Therapy***

Corticosteroids (methylprednisolone) should be given within 3 days after symptom onset and for 3 weeks (initially 100 mg/day and then tapered off by 20 mg every 3 days). The administration of valacyclovir or a combination of corticosteroids with valacyclovir does not cause any further improvement (Strupp et al. 2004).

- ***Improvement of Central Vestibular Compensation***

A gradual programme of physical exercise under the supervision of a physiotherapist improves the central vestibular compensa-

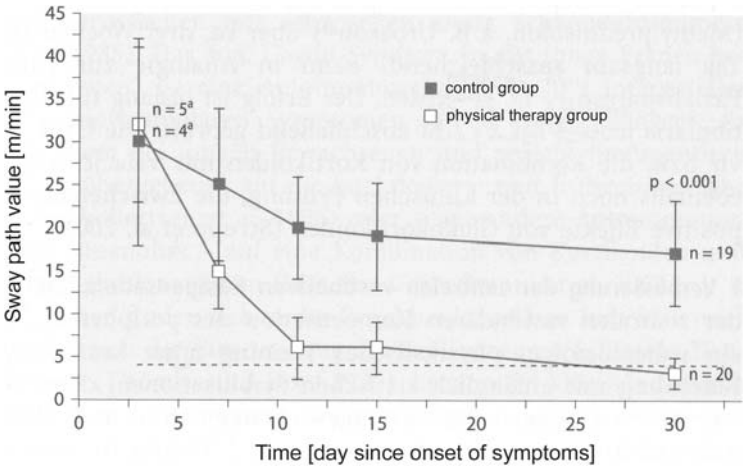

Figure 2.8. Time course of the changes in total sway path (SP) values for two patient groups, both after acute vestibular neuritis without recovery of the labyrinthine function. Whereas the initial values for SP (m/min, mean ± SD), measured with eyes closed and standing on a compliant foam rubber-padded posturography platform, were not significantly different in the two groups, there was a significantly faster normalisation of the SP values in the therapy group in the course of the study. On day 30 (statistical endpoint) there was a significant difference between the two groups (analysis of variance, $p < 0.001$). Thus, balance training improves the vestibulospinal compensation of an acute unilateral peripheral vestibular deficit. The dotted line indicates the normal range. *During the first days after onset of the illness some of the patients had such pronounced disturbances of postural control that they were unable to stand on the platform for the amount of time required to perform the measurements (>10 seconds) without falling (from Strupp et al. 1998).

tion of a peripheral deficit. First, static stabilisation is concentrated on, then dynamic exercises are done for balance control and gaze stabilisation during eye–head–body movements. It is important that exercises for equilibrium and balance are successively increased in degree of difficulty above normal levels, both with and without visual stabilisation. The efficacy of physiotherapy in improving central vestibulospinal compensation in patients with vestibular neuritis has been proven in a prospective, randomised controlled clinical study (Strupp et al. 1998).

- ***Ineffective Therapy***

Treatment with measures to improve circulation (vasodilators, low-molecular weight dextrans, hydroxyethyl starches, local anaesthetics or inhibitors of the ganglion stellatum) is ineffective.

2.2.5 *Differential Diagnosis and Clinical Problems*

Attacks of Menière's disease or basilar/vestibular migraine that last a maximum of 1 day, as well as labyrinthine or vestibular nerve lesions due to other causes (e.g., vestibular paroxysmia, most often lasting only seconds) must be considered in the differential diagnosis. Any of the specific accompanying symptoms (see below), the duration and the recurrence of complaints also prove useful. An initially burning pain and blisters, as well as hearing disorders and facial paresis, are typical of herpes zoster oticus (Ramsey–Hunt syndrome) (in such cases acyclovir or valacyclovir is indicated). Toxically serous types of labyrinthitis that accompany a middle-ear inflammation are usually painful (here antibiotic treatment is indicated); the acute suppurative form of labyrinthitis is also characterised by pain and frequently by hearing loss as well (this requires in addition operative decompression and drainage). Tuberculous labyrinthitis is more frequently a complication of tuberculous meningitis than of tuberculous middle-ear inflammation. The frequency of acquired syphilitic labyrinthitis peaks in the fifth to sixth decades. Borrelia infection only very rarely causes an acute vertigo syndrome (Rosenhall et al. 1988). Cogan's syndrome (relatively rare) is an autoimmune disease accompanied by interstitial keratitis and audiovestibular symptoms (hearing disorders are most prominent). It occurs most often in young adults and responds, in part only temporarily, to the very early administration of high doses of glucocorticoids (1,000 mg/day for 5 days, then slowly tapered off) (Vollertsen et al. 1986) or—like other autoimmune diseases of the inner ear—to a combination of steroids and cyclophosphamide (McCabe 1989; Orsoni et al. 2002).

Brainstem signs usually occur with lacunar infarctions or multiple sclerosis (MS) plaques in the entry zone of the VIIIth cranial nerve ("pseudo vestibular neuritis") (Thömke and Hopf 1999). The latter is clinically characterised by incomplete caloric hyporesponsiveness and in addition central ocular motor signs such as vertical saccadic smooth pursuit and/or a complete ocular tilt reaction. Cerebellar signs are found in infarctions of the cerebellum near the midline; they can also lead to a pseudo vestibular neuritis.

Acute unilateral disorders of labyrinthine function can also be caused by ischaemia as a result of labyrinthine infarctions with or without hearing loss (see above) (supply region of the A. labyrinthi or anterior inferior cerebellar artery, AICA) as well as, but very rarely, by venous obstruction due to a hyperviscosity syndrome. A reduction of blood viscosity is therapeutically effective for all symptoms of the hyperviscosity syndrome, including vertigo (Andrews et al. 1988).

Vestibular schwannomas, which arise in the myelin sheaths of the vestibular part of the VIIIth nerve, mainly cause vertigo, a tendency to fall and nystagmus when the pontomedullary brainstem and the flocculus are compressed and the increasing peripheral tonus difference can no longer be neutralised by central compensation. The main symptom is a slowly progressive unilateral reduction of hearing without any identifiable otological cause, which is combined with a caloric hypoexcitability or nonexcitability. Rarely, there is also a loss of hearing as well as acute vertigo in cases of a purely intracanalicular neuroma, which can be confirmed by magnetic resonance imaging (MRI) and treated early by microsurgery or with the "gamma knife". Other tumours that can cause vertigo are meningeomas of the cerebellopontine angle, epidermoid cysts, carcinomas or glomus tumours of the vagus or glossopharyngeal nerves.

In 10–15% of patients with vestibular neuritis, a typical BPPV develops in the affected ear within a few weeks. It is possible that the otoconia loosen during the additional inflammation of the labyrinth, and this eventually results in canalolithiasis. Patients should be warned about this possible complication, because there are therapeutic liberatory manoeuvres that can quickly free the patient of his complaints (see Section 2.1). The second important complication is that vestibular neuritis can develop into a somatoform phobic postural vertigo (see Section 5.1). The traumatic experience of a persisting organic rotatory vertigo leads to fearful introspection that results in a somatoform, fluctuating and persistent postural vertigo, which is reinforced in specific situations and culminates in a phobic behaviour of avoidance.

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2.3 Menière's Disease

2.3.1 Patient History

The main symptoms of an attack of Menière's disease include rotatory vertigo with spontaneous nystagmus and a tendency to fall in a certain direction, nausea and vomiting, and "ear symptoms" such as tinnitus, reduced hearing and a feeling of fullness in one ear. Single attacks usually last several hours and mostly have no antecedent signs or recognisable precipitating factors. They occur both in daytime and at night. One-third of patients, however, report that increased tinnitus and hearing loss, as well as a subjective feeling of fullness of the ear, precede the vertigo attack (aura). Monosymptomatic attacks that are purely cochlear or vestibular can occur, particularly at the beginning of Menière's disease. During the course of the disease most patients develop a persistent hypoacusis of the affected ear.

2.3.2 Clinical Syndrome and Course

Menière's disease is typically a combination of abruptly occurring attacks with vestibular and/or cochlear symptoms, fluctuating, slowly progressive hearing reduction, and in the course of time tinnitus. During the attack there is first a unilateral short vestibular excitation, then a temporary vestibular deficit with the following clinical findings:

- during the initial vestibular excitation: ipsiversive rotatory vertigo and ipsiversive nystagmus,
- during the vestibular deficit: contraversive rotatory vertigo and contraversive nystagmus,
- in addition there are cochlear symptoms in the form of tinnitus, reduced hearing of the affected ear, as well as pressure and a feeling of fullness in the ear, and
- deviation of gait and a tendency to fall.

The diagnosis of monosymptomatic forms is frequently difficult to determine or indeterminate (see below).

The American Academy of Ophthalmology and Otolaryngology, Head and Neck Surgery formulated the following diagnostic criteria in 1995:

Certain Menière's disease
- histopathological confirmation of endolymphatic hydrops
- symptoms as in "definite Menière's disease" criteria

Definite Menière's disease
- two or more attacks of vertigo, each lasting more than 20 min
- audiometrically documented hearing loss in at least one examination
- tinnitus or aural fullness in the affected ear
- other causes excluded

Probable Menière's disease
- at least one vertigo episode
- audiometrically documented hearing loss in at least one examination
- tinnitus or aural fullness in the affected ear
- other causes excluded

Possible Menière's disease
- episodic vertigo but without documented hearing loss
- sensorineural hearing loss, fluctuating or fixed, with disequilibrium, but without definite episodes of vertigo
- other causes excluded.

It seems to us that these recommendations require improvement in at least two respects: as regards the certainty of the diagnosis of Menière's disease and its distinction from other differential diagnoses, since there are several overlaps, e.g., with basilar/vestibular migraine, perilymph fistula and vestibular paroxysmia.

The onset of Menière's disease is usually between the fourth and sixth decades; it seldom occurs in childhood or after the eighth decade. Men are affected somewhat more often than women (Sade and Yaniv 1984; Stahle et al. 1978). The disease begins in one ear with very irregular attacks, which at first generally increase in frequency and then decrease in frequency in the course of several years. The patients are initially free of complaints in the attack-free interval, but then develop increasing deficits such as unilateral peripheral vestibular hypofunction, unilateral tinnitus and hearing disorder (usually low frequencies). The extent of fluctuation of these deficits is unusual when compared with other inner-ear illnesses.

While the onset of the disease is unilateral, the other ear can also become affected in time. The longer one follows a patient with Menière's disease, the more often one sees bilateral illnesses (Morrison 1986). In an early stage (up to 2 years' duration), about 15% of the cases are bilateral; 30–60% of these become bilateral

after one to two decades. In the meantime it has become generally acknowledged that the course of Menière's disease is on the whole relatively benign, having a remission rate (of the episodes but not of chronic hearing loss) of about 80% within 5–10 years (Friberg et al. 1984). The spontaneous remission of these attacks probably occurs when there is a permanent fistula of the membrane separating endolymph and perilymph, for this would allow the continual, asymptomatic drainage of excess endolymph.

2.3.3 Aetiology, Pathophysiology and Therapeutic Principles

Menière's disease develops from endolymphatic labyrinthine hydrops with periodical rupturing of the membrane separating the endolymph space from the perilymph space. This triggers attacks that generally last several hours. The overflow of the potassium-rich endolymph into the perilymph space (here ionic ratios are similar to those in the extracellular space) leads to a potassium-induced depolarisation of the vestibulocochlear nerve, which causes an initially temporary excitation and finally a block of conduction, as the fast sodium canal can no longer switch into an active state due to the stronger depolarisation. The cause of hydrops is an impaired resorption in the endolymphatic sac mainly due to perisaccular fibrosis or an obliteration of the endolymphatic duct, which interrupts the longitudinal endolymph flow.

Endolymphatic hydrops can also occur without symptoms and has either an inflammatory (labyrinthitis), traumatic (perilymph fistula), autoimmunological or idiopathic aetiology (Schuknecht and Gulya 1983). The attacks generally last several hours, during which the vertigo changes from an acute rotatory vertigo into a postural vertigo, which later turns into a gait instability. In rare cases the first phase of the acute attacks can also be characterised by a brief, severe rotatory vertigo or a sudden drop attack due to the hydrops and shifting of the otoliths (Tumarkin's otolithic crisis, see below).

Despite various indications that an inflammatory origin or an autoimmunological process is involved, so far prospective studies on immune-inhibiting medications are lacking. Reviews of the literature covering a large number of therapy studies have agreed that positive effects on the frequency of attacks have been confirmed for only betahistine and diuretics (Claes and van de Heyning 1997; James and Thorp 2001). For this reason the H1 agonist and H3 antagonist betahistine is currently recommended as the prophylactic therapy of first choice. It improves the microcirculation by acting on the precapillary sphincters of the stria vascularis and at the same time exercises a regulating influence by means of the H3 receptors on the vestibular nuclei (Van

Cauwenberge and de Moor 1997; Yabe et al. 1993). This possibly leads to reduced production and increased resorption of the endolymph. A placebo-controlled double-blind study has shown that it has a significant influence on the natural course of the disease, especially on the vestibular symptoms (Meyer 1985). The production of endolymph can also be reduced by the method suggested by Schuknecht, i.e., "switching off the inner ear" by the instillation and diffusion of ototoxic antibiotics (e.g., gentamycin), which may also lead to destruction of the hair cells. In the meantime this method has been so refined that it is possible to selectively affect the secretory epithelium while largely sparing the vestibular and cochlear sensory cells.

Besides avoiding any further attacks of Menière's disease and treating an acute attack, another important principle of treatment is to promote central compensation of the peripheral vestibular deficits by means of physical therapy (see Section 2.3).

2.3.4 Pragmatic Therapy

- ***Treatment of Attacks***

The acute attack is itself limited. Vertigo and nausea can be reduced by antivertiginous drugs used in other acute disorders of labyrinthine function, e.g. dimenhydrinate 100 mg as suppository or benzodiazepine.

- ***Prophylactic Therapy***

The goal of prophylactic treatment is to reduce the endolymphatic hydrops. Therefore in cases of repeating attacks of rotatory vertigo, possibly with fluctuating loss of inner-ear hearing, tinnitus or pressure in the ear, the following treatment is indicated:

- betahistine, 3 × 2 tablets/day containing 20–24 mg for 6–12 months; dose can be tapered depending on course (the patient should keep a vertigo diary, in order to document the effects of therapy)
- if improvement is insufficient, hydrochlorothiazide plus triamterene (a half to one tablet in the mornings) can be administered in addition to betahistine.

Otolaryngologists recommend administration of steroids or a salt-free diet; however, no studies have yet proven their efficacy.

In rare cases of frequent Menière's attacks (with or without inner-ear hearing loss) that are refractory to drug treatment:

- intratympanal instillations of ototoxic antibiotics are indicated (e.g., 1–2 ml gentamycin at concentrations of 20–40 mg/ml) at interims of 2 or, better, more weeks.

Instillations used to be done on a daily basis until Magnusson et al. (1991) observed that the onset of ototoxic effects was delayed. Nowadays only single instillations are recommended at interims of several weeks. However, there is still no consensus on the dose and the duration of the interims (Blakley 2000).

Endolymphatic sac operations used to be performed everywhere and were considered at first to be a type of shunt operation. However, it was realised that they had only a placebo effect (Thomson et al. 1981); nowadays they are no longer performed. Currently fewer than 1–3% of patients are considered for operative measures.

- ***Treatment of Vestibular Drop Attacks (Tumarkin's Otolithic Crisis)***

Recurrent vestibular drop attacks (Tumarkin's otolithic crisis) pose an extreme impairment to patients in their everyday life. Moreover, they are dangerous because of the high rate of injuries. Depending on clinical judgement as to the severity of the disorder, intratympanic gentamycin treatment can be administered with success (Ödkvist and Bergenius 1988). The prerequisite for such treatment is that the causative ear can be definitely identified.

- ***Ineffective Treatment***

Endolymphatic sac operations are nowadays no longer performed. The efficacy of a large number of conservative treatment programmes (e.g., circulation-promoting measures with vasodilators, low-molecular weight dextrans, hydroxyethyl starches) has not been proven.

2.3.5 Differential Diagnosis and Clinical Problems

The first attacks of Menière's disease must be differentiated from an acute unilateral vestibular deficit, e.g. in connection with vestibular neuritis (Section 2.2). Here the duration of the attacks is helpful: whereas in Menière's disease they generally last several hours and at most one day, in vestibular neuritis they last several days. The accompanying symptoms are also helpful for the diagnosis, for example, "ear symptoms" in Menière's disease and inflamed eye signs and hearing disturbances in Cogan's syndrome or hearing disorders and possibly central signs of infarctions of the AICA/labyrinthine artery. Central disorders of the ocular motor system or central vestibular function also occur after lacunar infarctions or MS plaques in the area of the entry zone of the VIIIth cranial nerve ("fascicular lesion"). Since a caloric hyporeactivity occurs in all of the above-named diseases, this cannot be used in the differential diagnosis.

Rare, sudden recurrent falls, so-called vestibular drop attacks (Tumarkin's otolithic crisis), which occur in the early or late

stages of Menière's disease without definite triggers, antecedent signs or disturbances of consciousness, are difficult to differentiate from drop attacks caused by vertebrobasilar ischaemia (Baloh et al. 1990). Such attacks apparently result from fluctuations in endolymphatic pressure caused by unilateral exacerbation of the otoliths and inadequate vestibulospinal postural reaction.

Another important differential diagnosis is basilar/vestibular migraine, which can manifest not only in the form of short attacks, but also as attacks lasting several hours (Section 3.2). Signs that the attack is a vestibular migraine are: (1) central ocular motor disorders during the attack-free interval, (2) the absence of progressive hearing loss despite many attacks, (3) the association with other neurological symptoms such as numbness of the face (basilar migraine), (4) head and neck pain, and (5) response to prophylactic treatment with beta-blockers. There are increasing indications in the recent literature of a link between Menière's disease and vestibular migraine (Radtke et al. 2002). Vestibular paroxysmia, which is caused by neurovascular compression, is also characterised by recurrent attacks with vertigo and/or occasionally other ear symptoms. These attacks, contrary to Menière's disease, typically last only seconds.

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2.4 *Vestibular Paroxysmia*

2.4.1 Patient History

The main symptoms are brief attacks of rotatory or postural vertigo lasting seconds or rarely a few minutes, with or without ear symptoms (tinnitus and hypoacusis). In many patients the attacks frequently depend on certain positions of the head; they can occasionally be induced by hyperventilation. Hearing loss and tinnitus can also be present during the attack-free intervals.

2.4.2 Clinical Aspects and Course

Vestibular paroxysmia is suspected if brief and frequent attacks of vertigo are accompanied by the following features (Brandt and Dieterich 1994):

- short attacks of rotatory or to-and-fro vertigo last for seconds to minutes with instability of posture and gait
- attacks may be triggered in some patients by particular head positions, hyperventilation, or the attack may be influenced by changing the head position
- unilateral hypoacusis or tinnitus occurs during the attack, occasionally or permanently
- in the course of the disease, measurable vestibular and/or cochlear deficits increase during the attack and are less pronounced during the attack-free interval (neurophysiological function tests used include audiogram, acoustic evoked potentials, caloric testing, test for subjective visual vertical)
- attacks are improved or lessened by administering carbamazepine (even low dosage effective)
- no central vestibular/ocular motor disorders or brainstem signs.

Conclusions can be drawn from the type of complaints—vestibular (originating from the canals or otolith organs) or cochlear symptoms—about the affected portion of the nerve. If there is a combination of symptoms of various nerves, the site of the lesion can be deduced. Thus, for example, simultaneously occurring symptoms of the VIIth and VIIIth cranial nerves (with contraction of the frontal muscle, vertigo and slightly staggered double images; Straube et al. 1994) indicate an irritation of both nerves in the internal acoustic meatus, where both lie in close proximity to each other.

There seem to be two peaks of frequency, one that begins early in cases of vertebrobasilar vascular anomalies and a second between the fifth and seventh decades with vascular elongation during old age. The course is generally chronic. Men are affected twice as often as women.

2.4.3 Aetiology, Pathophysiology and Therapeutic Principles

As in trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia or myokymia of the superior oblique muscle, it is assumed that a neurovascular cross-compression of the VIIIth cranial nerve is the cause of these short episodes of vertigo (Møller et al. 1986; Møller 1988; Brandt and Dieterich 1994). Aberrant, in part arteriosclerotically elongated and dilated, and consequently more pulsating, arteries in the cerebellopontine angle are thought to be the pathophysiological cause of a segmental pressure-induced lesion with demyelination of the central (oligodendroglia) myelin. Mostly a loop of the AICA seems to be involved. The symptoms are triggered by direct pulsatile compression and/or ephaptic discharges, i.e., pathologically paroxysmal interaxonal transmission between neighbouring and in part demyelinated axons. Another cause under discussion is central hyperactivity in the nucleus, which is induced and maintained by the compression. Finally, in addition to elongation and increased looping, a vascular malformation or arterial ectasia of the posterior fossa can also cause the nerve compression (Buettner et al. 1983; Yu et al. 1982).

This aetiology was previously connected with the so-called "disabling positional vertigo" (Jannetta 1975; Jannetta et al. 1984), a very heterogeneous syndrome of vertigo with a great number of symptoms of various duration (from seconds to days), various characteristic features (rotatory or positional vertigo, numbness or gait instability without vertigo), and varying accompanying symptoms. As these vague descriptions also applied to patients with benign positional vertigo (Section 2.1), Menière's disease or phobic somatoform postural vertigo (Section 5.1), the clinical

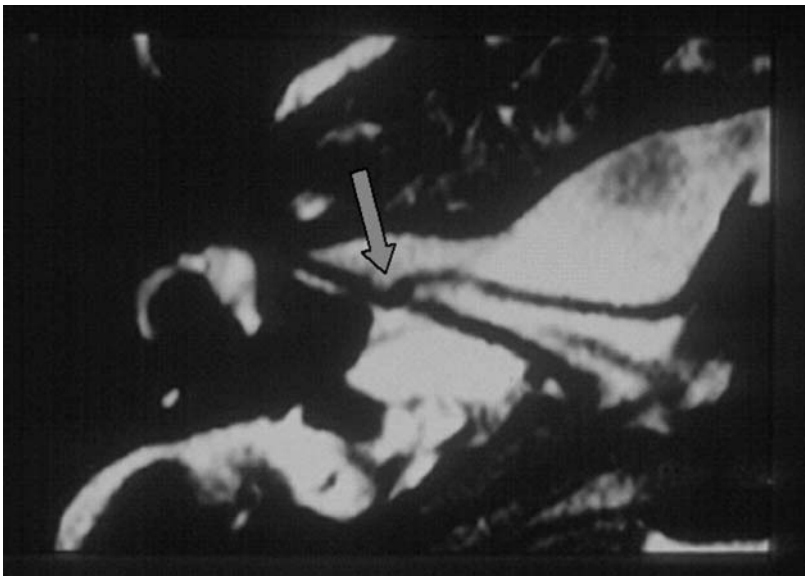

Figure 2.9. Neurovascular cross-compression of the vestibulocochlear nerve by a loop of the anterior inferior cerebellar artery (*arrow*) in a magnetic resonance image with three-dimensional constructive interference in steady-state sequence.

definition was subsequently made more precise (Brandt and Dieterich 1994).

Despite signs of an arterial compression of the VIIIth cranial nerve, which are visible in MRI (constructive interference in steady-state sequence; Figure 2.9), prospective clinical studies are still lacking as to how frequently such neurovascular contacts can be imaged. Such studies are necessary, as neurovascular cross-compression is also seen in healthy persons, thus raising the question of which region of the myelin sheath of the vestibulocochlear nerve is the most vulnerable (distance measured precisely in millimetres from the nerve exit zone out of the brainstem). In patients with myokymia of the trochlear nerve, there was a neurovascular contact 0–1 mm from the nerve exit zone, whereas a neurovascular contact was proven to occur in 14% of healthy subjects at a mean distance of 3.4 mm (Yousry et al. 2002a, b).

Occasionally, in addition to neurovascular contact, vertigo attacks lasting seconds and caused by head movements point to an arachnoid cyst that stretches the vestibulocochlear nerve (Figure 2.10) (Arbusow et al. 1998). This pathogenesis can result in a combination of longer-lasting conduction-block symptoms in one direction (hours to days) and paroxysmal symptoms of excitation (for seconds) in the opposite direction.

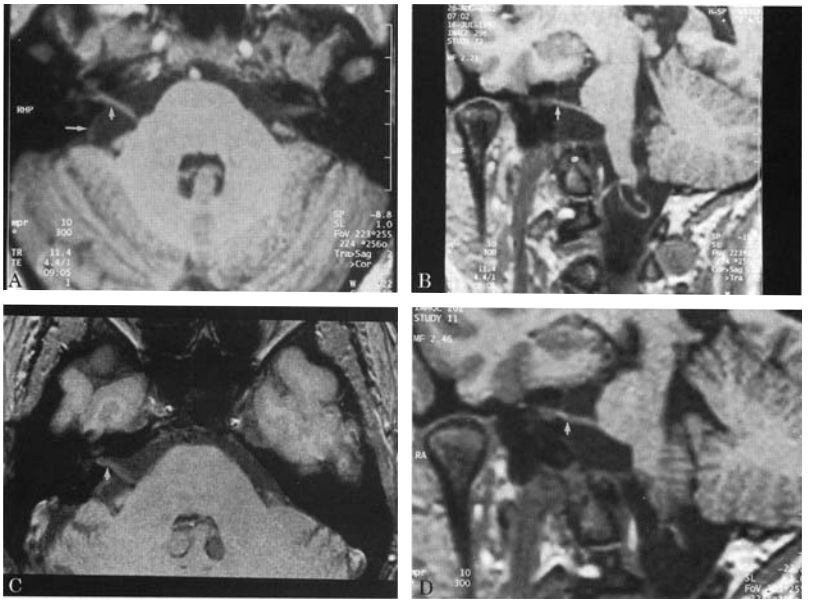

Figure 2.10. An arachnoid cyst in the region of the cerebellopontine angle. **A, B** T1-weighted magnetic resonance image before operation. The right vestibulocochlear nerve (*short arrow*) is displaced ventrally (**A**) and rostrally (**B**) by a cyst (*longer arrow* indicates its delimitation). **C, D** Postoperatively there is no longer any displacement and the patient is without complaints.

2.4.4 Pragmatic Therapy

Therapy with a low dosage of carbamazepine (200–600 mg/day) or oxcarbamazepine has an early therapeutic onset, and a positive response is diagnostic. In case of intolerance, gabapentine, valproic acid or phenytoin are possible alternatives. Despite the report of partial successes (Møller et al. 1986), operative microvascular decompression should be avoided for four reasons: (1) 5–10% risk of hearing loss; (2) efficacy in only 50–60%, even in well-selected patients; (3) difficulty to decide on which side to operate; and (4) rare stroke (symptomatic rate ~ 1%) from inadvertent sacrifice of small brainstem perforator (perhaps vasospasm) and/or cerebellar retraction, if retrotemporal bone approach used. However, if there are additional causes, such as the above-mentioned arachnoid cyst in the cerebellar pontine angle, the operation is recommended, as drug therapy only rarely leads to the absence of symptoms.

- ***Ineffective Treatment***

Treatments with circulation-promoting measures and antivertiginous drugs are ineffective.

2.4.5 Differential Diagnosis and Clinical Problems

Important differential diagnoses are:

- benign paroxysmal positioning vertigo
- paroxysmal brainstem attacks
- vestibular migraine
- phobic postural vertigo
- vertebral artery occlusion syndrome (dependent on head position)
- central positional/positioning nystagmus.

The differential diagnosis is generally straightforward, because of the characteristic brevity (seconds up to a few minutes) of the frequently recurring attacks of vertigo. Only paroxysmal brainstem attacks with vertigo and, for example, ataxia, can be difficult to distinguish, as they too respond to low dosages of carbamazepine. It is assumed that they are caused by a brainstem lesion (MS or infarction), which also leads to ephaptic activation of neighbouring fibres of the brainstem paths. In such cases the use of MRI with thin brainstem slices is expedient for establishing the diagnosis.

Benign paroxysmal positioning vertigo due to canalolithiasis can be diagnosed by the typical crescendo–decrescendo nystagmus caused by the positioning manoeuvre, which typically does not occur with vestibular paroxysmia and is not triggered as regularly by positioning.

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2.5 *Bilateral Vestibulopathy*

2.5.1 *Patient History*

Key symptoms of bilateral vestibulopathy are:

- oscillopsia with blurred vision during head movements and walking
- unsteadiness of gait, particularly in the dark and on uneven ground
- spatial memory and navigation disorders.

Patients mostly complain that their environment seems to move when they are walking or running, they can no longer read street signs, and they cannot definitely identify the faces of people approaching them. Patients with sequential or "idiopathic" bilateral vestibulopathy may report rotatory or postural vertigo in the initial phases, which persists for several minutes or even days.

2.5.2 *Clinical Aspects and Natural Course*

Bilateral vestibular failure is a rare disorder of the labyrinth and/or the vestibular nerves of various aetiologies, which is frequently not diagnosed (Rinne et al. 1995; Vibert et al. 1995; Brandt 1996). Clinical suspicion of a bilateral vestibulopathy is based on the above-mentioned key symptoms; the diagnosis is confirmed by testing the VOR. The simple, clinical bedside test of Halmagyi and Curthoys (1988), in which the head of the patient is quickly and passively turned, has proved its worth (see Figure 1.14, p. 20). When the head is quickly turned horizontally to the right and then to the left, refixation saccades are seen, a sign of a defective VOR. The VOR reading test is also abnormal. Caloric testing with recording of eye movements (see Figure 1.21, p. 28) serves to document and quantify the deficit, especially as regards differences between the two sides. Moreover, there are no abnormalities in the ocular motor system and the patient has no complaint as long as the head remains unmoved. Tests of balance with the eyes open are basically normal; only when the eyes are

closed is there increased body instability during the Romberg test; this becomes more obvious during tandem standing or walking toe-to-heel. In the latter two tests there is a danger of falling. Asymmetries of the vestibular function are observed when the patient walks straight ahead with closed eyes. The direction of gait deviation as a rule indicates the side most affected.

Although bilateral vestibulopathy is rare (see Table 1.1 for relative frequency, p. 5), it can become manifest at any age, and a sexual predominance has not been observed.

In the course of bilateral vestibulopathy, both labyrinths and/or vestibular nerves can be affected at the same time or sequentially; it can occur acutely or be slowly progressive, be complete or incomplete, with or without a difference in the side affected. Bilateral vestibulopathy can occur with or without associated hearing loss. The long-term prognosis has not been sufficiently studied. Recovery of vestibular deficit and hearing loss is possible in post-meningitis cases due to a serous non-suppurative labyrinthitis (Rinne et al. 1995). Partial recovery has been described in more than 50% of patients with simultaneous or sequential idiopathic bilateral vestibulopathy (Vibert et al. 1995).

2.5.3 Aetiology, Pathophysiology and Therapeutic Principles

The key symptoms of bilateral vestibulopathy can be explained by the loss of vestibulo-ocular and vestibulospinal functions.

- ***Oscillopsia and Blurred Vision***

During rapid head movements the VOR cannot maintain the target of gaze on the fovea, and thus there is an involuntary movement of the image on the retina, which is experienced as an illusory movement and reduces the visual acuity. Conversely, when head movements are slow, the smooth pursuit system is able to stabilise the gaze sufficiently in space, and illusory movements or blurriness do not occur.

- ***Unsteadiness of Posture and Gait***

Unsteadiness of posture and gait is increased in the dark and on uneven ground. Due to the redundant sensorimotor control of posture, the visual system can basically substitute for any defective regulation of postural control in light. The somatosensory system also contributes to the maintenance of balance, above all via the muscle spindle afferents and the mechanoreceptors of the skin. If the contribution of the visual system (in darkness or due to visual disorders) is reduced, gait imbalance increases with the risk of falls. This is further intensified when the patients walk in the dark over uneven or yielding ground. A sensory poly-

neuropathy primarily of the legs also reduces the somatosensory contribution to posture control and thereby increases instability in cases of bilateral vestibulopathy.

- ***Deficits of Spatial Memory and Navigation***

Intact vestibular function is important for spatial orientation and spatial memory (Smith 1997). Recently, significant deficits of spatial memory were demonstrated in patients with bilateral vestibulopathy (neurofibromatosis type 2) (Schautzer et al. 2003).

The most frequent causes of bilateral vestibulopathy are autoimmune diseases, such as Cogan's syndrome (MRI shows typical haemorrhages and enhancement by contrast medium in the labyrinth and/or cochlea; Figure 2.11), cerebellar degeneration, ototoxic substances, meningitis, tumours, neuropathies, bilateral Menière's disease, congenital malformations and familial vestibulopathies (Table 2.2). The cause remains unknown in 20–30% of cases; they are referred to as "idiopathic bilateral vestibulopathy".

Treatment of the various forms of bilateral vestibulopathy follows three lines of action (Brandt 1996):

- prophylaxis of progressive vestibular loss
- recovery of vestibular function
- promotion of the compensation and substitution of missing vestibular function by physical therapy.

2.5.4 Pragmatic Therapy

Prevention is most important for the group of patients with ototoxic labyrinthine damage, above all that due to aminoglycoside. Aminoglycoside therapy should be used only if strictly indicated and then only in a once-daily dose. Plasma levels should also be monitored. Patients with renal insufficiency, advanced age or familial susceptibility to aminoglycoside ototoxicity are at particular risk. Ototoxic antibiotics should not be combined with other ototoxic substances, such as loop diuretics, as this can greatly increase inner-ear damage. Careful monitoring of the hearing and vestibular function is necessary during treatment. However, the physician must remain vigilant, as the ototoxic effects of gentamycin have a delayed onset, often appearing only after days or weeks (Magnusson et al. 1991).

Recovery of vestibular function is possible in individual cases of the autoimmunologically induced forms of inner-ear disease, which are diagnosed too infrequently (Schüler et al. 2003). Although controlled prospective studies are lacking, immune treatment is theoretically expedient, only if there are clinical signs of a systemic autoimmune disease or if antibodies against inner-

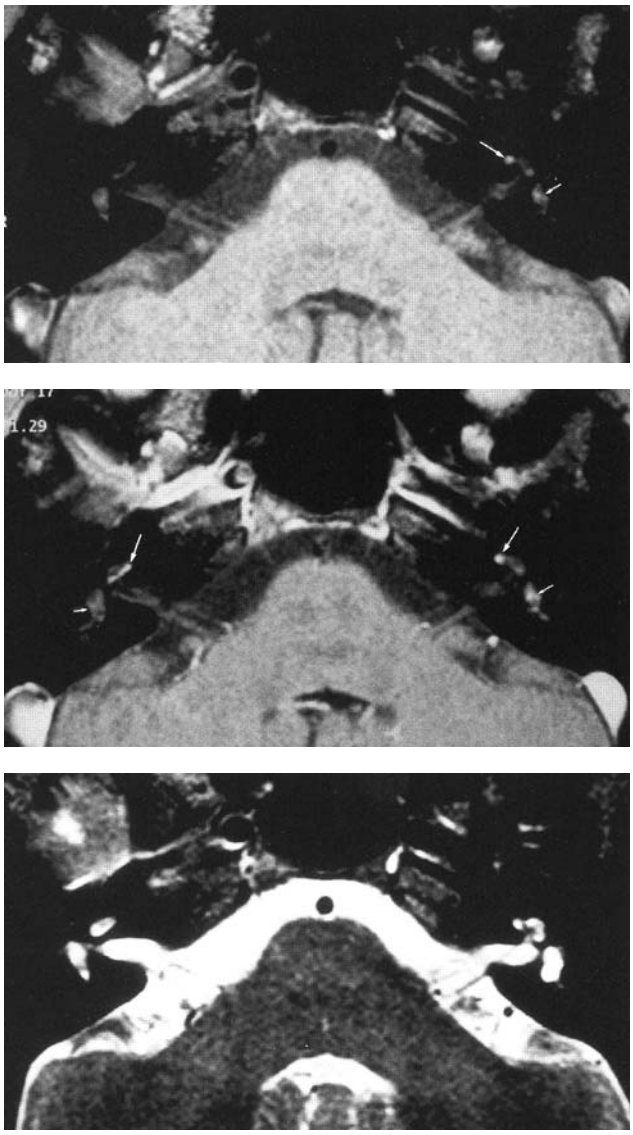

Figure 2.11. **a** Axial projection, T1-weighted, two-dimensional fast low angle shot (2D-FLASH), no contrast medium. Cogan's syndrome, subacute stage. Signs of subacute haemorrhages are the signal increases in the vestibule (*short arrow*) and in the cochlea (*long arrow*); **b** axial projection, T1-weighted 2D-FLASH after administration of Gd-DTPA in the same patient. Clear enhancement by contrast medium in the area of the haemorrhage, namely in the cochlea (*long arrow*) and in the vestibule (*short arrow*); **c** axial projection, T2-weighted turbo-spin-echo imaging, no contrast medium, in a patient with Cogan's syndrome. The canals cannot be delimited; this is a sign of obliteration from scarring.

Table 2.2. Causes of bilateral vestibulopathy

Relatively frequent	**Underlying cause**
Idiopathic (20–30%)	
Ototoxicity	Gentamycin and other antibiotics
	Anticancer chemotherapy
	Loop diuretics
	Aspirin
Cerebellar degeneration	Spinocerebellar degeneration
	Multi-system atrophy
Meningitis or labyrinthitis	e.g., *S*treptococci, Neisseria meningitis,
	Mycobacterium tuberculosis,
	HIV-associated infections
Tumours	Neurofibromatosis type II (bilateral vestibular schwannomas)
	Non-Hodgkin's lymphoma
	Meningeosis carcinomatosa
	Infiltration of the skull base
Autoimmune disorders	Cogan's syndrome
	Neurosarcoidosis
	Behçet's disease
	Cerebral vasculitis
	Systemic lupus erythematosus
	Polychondritis
	Rheumatoid arthritis
	Polyarteritis nodosa
	Wegener's granulomatosis
	Giant cell arteritis
	Primary antiphospholipid syndrome
Neuropathies	B_{12} deficiency
	B_6 deficiency
	Hereditary sensory and autonomic neuropathy (HSAN IV)
Bilateral sequential vestibular neuritis	Herpes simplex virus type I
Bilateral Menière's disease	Delayed endolymphatic hydrops
Relatively rare (or single cases)	
Congenital malformation	Usher's syndrome and other rare hereditary conditions
Rare or single case	Bilateral temporal bone fractures
	Paget's disease
	Macroglobulinaemia
	Vertebrobasilar dolichoectasia

ear structures are detected (reviewed in Brandt 1999; Schüler et al. 2003). Initially, corticosteroids can be tried (e.g. prednisolone in doses of 80 mg/day, tapered for ca. 3–4 weeks). In Cogan's syndrome, higher doses of steroids (1 g i.v. for 5 days, then tapered for several weeks) are recommended. If the response is inadequate or relapses occur, additional but temporary administration of azathioprine or cyclophosphamide is recommended (Orsoni et al.

2002). Besides this, treatment of the causative underlying disease (Table 2.2) is important and in individual cases also successful.

Patient response to physical therapy with gait and balance training is quite positive. This therapy alleviates the adaptation to loss of function by promoting visual and somatosensory substitution. However, a comparison of trained and untrained patients showed that there was no significant improvement of balance in the long term, although the patients felt subjectively more secure (Herdman 2000). It is important to inform the patients carefully about the type, mechanism and course of their illness. It is our experience that the diagnosis of a bilateral vestibulopathy is still established much too late, despite many visits to the physician, a fact that only intensifies the complaints of the patients. Frequently, simply informing the patient leads to a reduction of these subjective complaints.

2.5.5 Differential Diagnosis and Clinical Problems

Considerations for the differential diagnosis proceed along two lines. On the one hand, it is important to look for the causes listed in Table 2.2 if clinical signs of a bilateral vestibulopathy are present. On the other hand, it is necessary to differentiate the illness from other vestibular and non-vestibular diseases, which are also characterised by oscillopsia and/or instability of posture and gait (see Tables 1.6 and 1.7, pages 10 and 11). The following steps are important for the differential diagnosis:

- differentiate the various causes and mechanisms of bilateral vestibulopathy (see Table 2.2)
- differentiate from illnesses with similar main symptoms:
  - — cerebellar or eye movement disorders without bilateral vestibulopathy
  - — phobic postural vertigo
  - — intoxications
  - — vestibular paroxysmia
  - — perilymph fistulas
  - — orthostatic hypotension
  - — hyperventilation syndrome
  - — visual disorders
  - — unilateral vestibular deficit.

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2.6 Perilymph Fistulas

2.6.1 Patient History

The cardinal symptoms of perilymph fistula and superior canal dehiscence syndrome are attacks of rotatory or postural vertigo caused by changes in pressure, for example, by coughing, pressing, sneezing, lifting, or loud noises and accompanied by illusory movements of the environment (oscillopsia), and instability of posture and gait with or without hearing disturbances. The attacks, which can last seconds to days, can also occur during changes in the position of the head (e.g., when bending over) and when making significant changes in altitude (e.g., mountain tours, flights). When taking the patient history, it is important to ask about traumas that could cause or trigger such attacks, for example, barotrauma, head trauma, ear trauma (also operations of the ear) or excessive Valsalva manoeuvres due to lifting of heavy weights.

2.6.2 Clinical Aspects and Course

The clinical spectrum of complaints is characterised by a wide range of symptoms: episodic dizziness or rotatory vertigo of various intensity and duration (from seconds to days), oscillop-

sia, imbalance and hearing loss. This variability depends on the site of the fistula, so that either canal or otolith symptoms dominate. Consequently, it is correspondingly difficult to establish the diagnosis. Rotatory vertigo suggests more the canal type, whereas unsteadiness and gait ataxia suggest more the otolith type. Linear and rotatory nystagmus, oscillopsia and a tendency to fall in a certain direction can occur in both forms.

Provocation tests, which attempt to trigger attacks during observation (using Frenzel's glasses) or recording of the resulting eye movements, can help to establish a diagnosis. Such tests include the Valsalva manoeuvre, the tragal pressure test, examination with a Politzer balloon (see Figure 1.11, p. 18), and positional manoeuvres, such as head-hanging position. The affected side can also be identified with the pressure test by means of the Politzer balloon or the tragal pressure test. A feeling of increased pressure in the ear, tinnitus, reduced hearing or autophonia—all of which can indicate the affected ear. A fistula of the horizontal canal is indicated by a linear horizontal nystagmus, whereas a fistula of the vertical canal is characterised by a vertical rotatory nystagmus.

The imaging techniques of MRI and computed tomography (CT) (in the form of high-resolution thin-slice CT, see Section 1.4.6) can be helpful, especially to prove congenital labyrinth dysplasias and the presence of a dehiscence of the superior semicircular canal (Figure 2.12). Otolaryngologists use exploratory

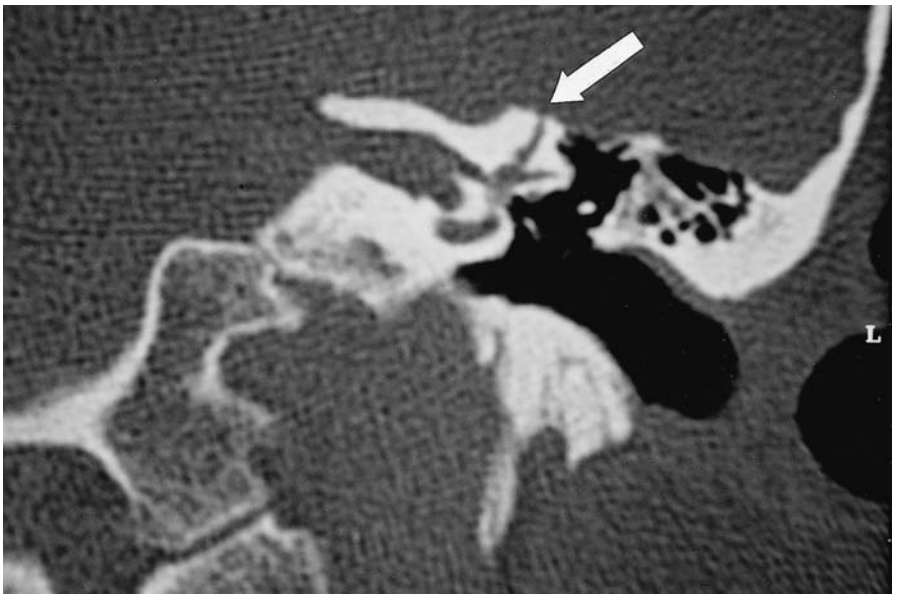

Figure 2.12. Superior canal dehiscence syndrome. High-resolution computed tomography scan of the petrous bone, coronal slice. There is a bony defect in the apical part of the superior canal extending toward the epidural space (*arrow*).

tympanoscopy when fistulas of the round and oval windows are strongly suspected. However, some specialists consider this examination rather insensitive and unspecific.

The Tullio phenomenon is characterised by the occurrence of vestibular otolith or canal symptoms caused by loud sounds. In this case, the attempt should be made to trigger vertigo and eye movements by exposing each of the patient's ears separately to loud sounds of different frequencies.

The incidence and prevalence of perilymph fistulas are not known because of the uncertain diagnosis (evidently they are relatively higher in childhood). However, perilymph fistulas can occur at any time during life and there is no obvious preference for either sex. The course of the illness varies; sometimes the attacks are rare and sometimes frequent; as a rule, they resolve spontaneously and there are symptom-free periods of various duration, which, however, can be reactivated by new traumas.

- ***Dehiscence of the Superior Semicircular Canal***

In 1998, Minor and colleagues described a new variant of a fistula without perilymph leakage, which is a cause of episodic vertigo. This form seems to be the most important one, because it is probably the most frequent and the most overlooked. The main symptoms are rotatory or postural vertigo with oscillopsia induced by changes of pressure and sometimes by loud sounds. In more than half of the patients, these complaints first occur after slight head concussion or barotrauma. Observance and recordings of the eye movements reveal rotating vertical nystagmus.

The diagnosis of dehiscence of the superior semicircular canal can be confirmed by thin-slice CT of the petrous bone (Figure 2.12), which shows a bony apical defect of the superior canal, and by 3-D analysis of eye movements induced by changes in pressure (Figures 2.13 and 2.14). The vestibular-evoked myogenic potentials (see Figure 1.26, p. 35) clearly show an electrophysiologically reduced sensory threshold in the affected ear (Watson et al. 2000).

2.6.3 Pathophysiology and Therapeutic Principles

The perilymphatic space and the endolymphatic space lie within the bony labyrinth. The border of tissue between the bony labyrinth and the middle ear forms the annular ligament of the base of the stapes and the membrane of the round window. These structures can rupture by stretching (blunt brain concussion), intracranial increase of pressure ("explosive pressure due to pressing", barotrauma) or pressure-wave-induced trauma ("implosion"). Perilymph fistulas can develop in patients with weak membranes (thin, bony apical cover of the superior canal) or a cholesteatoma after a bagatelle trauma, for example, sneezing or lifting heavy burdens, and not be noticed by them.

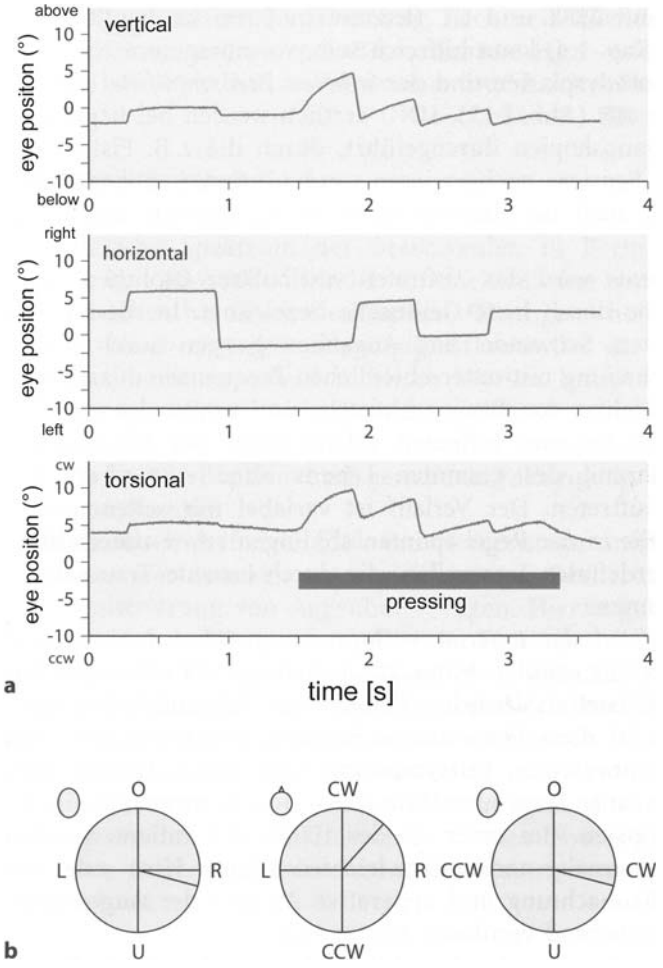

Figure 2.13. Dehiscence of left superior canal. **a** Original three-dimensional recording of vertical, horizontal and torsional eye movements with the search-coil technique before, during and after a Valsalva manoeuvre. Pressure-induced vertical (slow phase upward) and torsional (slow phase from viewpoint of the patient, counterclockwise) eye movements (*cw* clockwise, *ccw* counterclockwise), which are associated with rotatory vertigo and oscillopsia. **b** Eye rotation axes (calculated from vector analysis of the pressure-induced eye movements) from behind (*left*), from above onto the head (*middle*) and from the right side (*right*). (From Strupp et al. 2000.)

The underlying cause of all perilymph fistulas is pressure that is pathologically transmitted between the perilymphatic space and the middle ear or between the perilymphatic space and the intracranial space, which is caused by:

- a pathological motility of the membrane of the oval or round window or the ossicular chain with a hypermotility of the stapes footplate (Dieterich et al. 1989)

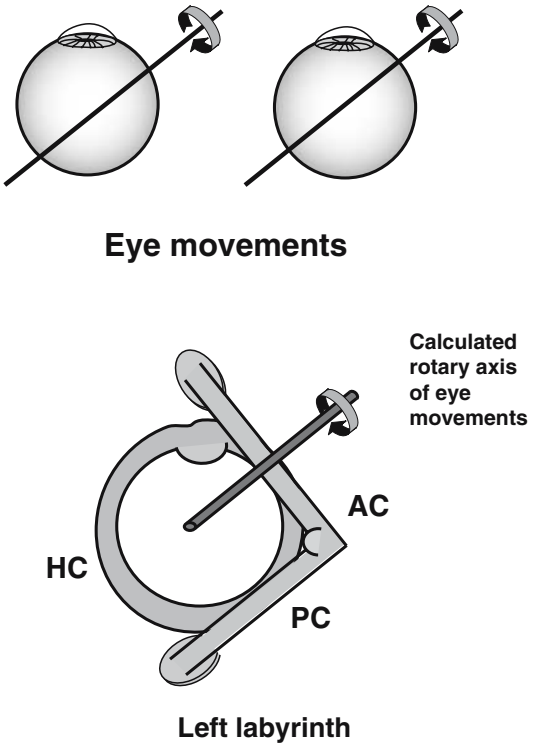

Figure 2.14. Schematic drawing of the slow eye movements triggered by stimulation of the left anterior canal (*top*). Eye movements result with a vertical upward and (from the viewpoint of the observer) torsional and counterclockwise direction. *Below*: the left labyrinth and the rotation axis of the eyes calculated from the three-dimensional vector analysis of the pressure-induced eye movements (see Fig. 2.13). The axis is perpendicular to the plane of the left anterior canal. *HC*, *AC*, *PC* horizontal, anterior, posterior canal.

- bony defects in the region of the lateral wall of the labyrinth (toward the middle ear) together with a partial collapse of the perilymphatic space ("floating labyrinth"; Nomura et al. 1992). Both lead to pathologically transmitted pressure from the middle ear to the labyrinth, for example, during a Valsalva manoeuvre. This determines the main clinical symptoms of fistulas (see above)
- a bony defect toward the epidural space in dehiscence of the superior semicircular canal. This bony defect leads to a third motile window (in addition to the round and oval windows in the middle ear), which induces a pathological transmission of intracranial pressure changes to the perilymphatic space of the superior canal and thus causes a stimulation/inhibition of the canal (due to ampullofugal or ampullopetal deviation of

the cupula). Pressure is transmitted in the opposite direction when sound triggers the attacks (e.g., Tullio phenomenon).

The therapeutic principles derive from the pathophysiology: conservative (spontaneous) or operative closure of the fistula.

2.6.4 Pragmatic Therapy

The therapy of first choice for "external fistulas" is conservative, as most fistulas close spontaneously.

- ***Conservative Therapy***

Conservative therapy consists of 1–3 weeks of bed rest with moderate elevation of the head, if necessary a mild sedative, the administration of laxatives (to avoid pressing during bowel movements), and several weeks of limited physical activity that avoids all heavy lifting, abdominal pressing, strong coughing or sneezing, even after improvement. This almost always leads to complete recovery (Singleton 1986). If conservative therapy fails and disturbing vestibular symptoms persist, exploratory tympanoscopy is indicated in order to examine the oval and round windows.

- ***Surgical Therapy***

Surgical therapy with operative closure of the fistula is successful in relieving vestibular vertigo in up to 70% of patients; the pre-existing hearing loss generally does not improve at all. The operative procedure involves the removal of the mucous membrane in the region of the fistula and its substitution with autologous material (prechondral tissue of the tragus or fascia by means of gel foam). Fistulas in the oval window adjacent to the stapes footplate require a stapedectomy and prosthesis. Even if the operation is successful, the postoperative sensitivity of the patients to extreme physical strain (abdominal pressing, barotrauma) is greater than that of healthy subjects.

It is possible that a part of the fistulas that were assumed to be in the middle ear were actually a dehiscence of the superior semicircular canal, as these can indirectly also lead to a pathological motility of the middle-ear window. This probably explains in part the low rate of improvement after the above-mentioned operations. The dehiscence of the superior semicircular canal can be treated neurosurgically by covering the bony defect or by occlusion (so-called plugging) of the canal (Minor et al. 2001; Strupp et al. 2000). Prospective studies to determine which procedure is more effective are, however, still lacking.

2.6.5 Differential Diagnosis and Clinical Problems

The differential diagnosis of perilymph fistulas includes the following illnesses:

- benign paroxysmal positioning vertigo
- positional vertigo of central origin
- Menière's disease
- vestibular paroxysmia
- phobic postural vertigo
- labyrinthine concussion
- bilateral vestibulopathy.

The presence of perilymph fistula should be considered particularly in children who present with episodic vertigo with or without hearing loss and in patients who complain of vertigo and/or hearing loss after trauma of the ear or skull, or barotrauma.

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3

Central Vestibular Forms of Vertigo

Introduction

Central vestibular forms of vertigo are caused by lesions along the vestibular pathways, which extend from the vestibular nuclei in the medulla oblongata to the ocular motor nuclei and integration centres in the rostral midbrain, and to the vestibulocerebellum, the thalamus, and multisensory vestibular cortex areas in the temporoparietal cortex (Brandt and Dieterich 1995). These forms of vertigo are often clearly defined clinical syndromes of various aetiologies, with typical ocular motor, perceptual and postural manifestations that permit precise (topographical) localization. The analysis of nystagmus can also be helpful for localising the lesion site (Büttner et al. 1995). This section discusses such typical findings in detail. Depending on the size of the lesion, central vestibular syndromes can occur in isolation or as part of a complex infratentorial syndrome. Additional symptoms of supranuclear or nuclear ocular motor disorders and/or other neurological brainstem deficits can also occur (e.g., Wallenberg's syndrome with ocular tilt reaction, as well as Horner's syndrome, sensory deficits, ataxia, dysarthria and dysphagia).

The most important structures for central vestibular forms of vertigo are the neuronal pathways for mediating the vestibulo-ocular reflex (VOR). They travel from the peripheral labyrinth over the vestibular nuclei in the medullary brainstem to the ocular motor nuclei (III, IV, VI) and the supranuclear integration centres in the pons and midbrain (interstitial nucleus of Cajal, INC; and rostral interstitial nucleus of the medial longitudinal fascicle, riMLF) (Brandt and Dieterich 1994, 1995; Figure 3.1). This

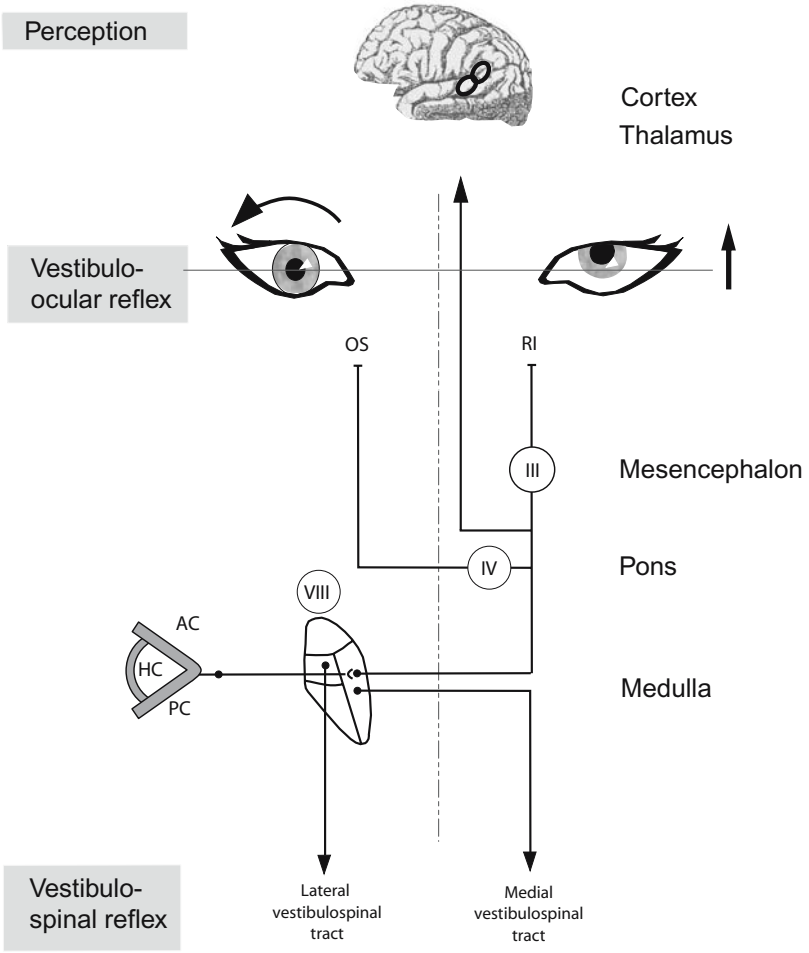


Figure 3.1. Schematic drawing representing the vestibulo-ocular reflex with its three-neuron reflex arc and its mediation of ocular motor, perceptual and postural functions; as well as an ocular tilt reaction with ocular torsion and vertical divergence of the eyes due to a tonus imbalance of the graviceptive pathways. *AC, HC, PC* anterior, horizontal and posterior semicircular canals.

three-neuron reflex arc makes compensatory eye movements possible during rapid head and body movements. It is thus crucially responsible for regulating the ocular motor system. Another branch of the VOR system runs over the posterolateral thalamus up to the vestibular areas in the parietotemporal cortex, such as the parietoinsular vestibular cortex (PIVC), area 7, and areas in the superior temporal gyrus, which are primarily responsible for perception of self-motion and orientation. Descending pathways lead from the vestibular nuclei along the medial and lateral vestibulospinal tract into the spinal cord to mediate pos-

tural control. In addition, there are pathways to the vestibulocerebellum and to the hippocampus.

Thus, disorders of the VOR are characterised not only by ocular motor deficits, but also by disorders of perception due to impaired vestibulocortical projections of the VOR and by disorders of postural control due to impaired vestibulospinal projections of the VOR (Figure 3.1).

Central vestibular syndromes are the result of lesions of these pathways caused by infarction, haemorrhage or tumour, by multiple sclerosis (MS) plaques, or more seldom by pathological excitations such as paroxysmal brainstem attacks (with ataxia and dysarthria, as occur in MS) or in vestibular epilepsy. Table 3.1 provides an overview of ischaemic lesions caused by lacunar or territorial infarctions in the region of the central vestibular system, along with the typical clinical syndromes and the arteries responsible.

3.1 Central Vestibular Syndromes

3.1.1 Clinical Aspects, Course of Disease, Pathophysiology and Therapeutic Principles

To differentiate central vestibular forms of vertigo from other forms, it is helpful to refer to the duration of the symptoms:

- Short rotatory or postural vertigo attacks lasting seconds to minutes or for a few hours are caused by transient ischaemic attacks within the vertebrobasilar territory, basilar/vestibular migraine, paroxysmal brainstem attacks with ataxia/dysarthria in MS, and the rare vestibular epilepsy.
- Persisting rotatory or postural vertigo attacks lasting hours to several days, generally with additional brainstem deficits, can be caused by an infarction, haemorrhage or MS plaque in the brainstem, seldom by a long-lasting basilar migraine attack.
- Several days to weeks of permanent postural vertigo (seldom permanent rotatory vertigo), combined with a tendency to fall, is usually caused by persisting damage to the brainstem or the cerebellum bilaterally, e.g., downbeat nystagmus syndrome due to Arnold–Chiari malformation or upbeat nystagmus syndrome due to paramedian pontomedullary or pontomesencephalic damage (infarction, haemorrhage, tumour).

3.1.2 Central Vestibular Syndromes in the Three Planes of Action of the Vestibulo-ocular Reflex

For a simple clinical overview, the central vestibular brainstem syndromes can be classified according to the three major planes of action of the VOR (Figure 3.2):

Table 3.1. Clinical syndrome and corresponding arterial territory affected by unilateral vascular lesions

Brain site	Clinical syndrome	Artery
Medulla oblongata	Wallenberg's syndrome with OTR and its features (head tilt, vertical divergence of the eyes, ocular torsion, deviation of the SVV) ipsiversive to the side of the lesion of the medial vestibular nuclei	Branches of the vertebral artery or PICA Rare: posterior spinal artery
	"Vestibular pseudo neuritis"	Branches of the vertebral artery or PICA
	OTR ipsiversive to the lesion of the superior vestibular nuclei	Anterior inferior cerebellar artery
Pons and midbrain	OTR or its components toward the opposite side of the lesion of the MLF	Paramedian arteries of the basilar artery
Rostral midbrain	OTR or its components contraversive to the lesion of the INC and riMLF	Paramedian midbrain arteries from the basilar artery
Paramedian thalamus	OTR contraversive to the lesion, only if rostral midbrain is affected (INC lesion)	50% of the paramedian midbrain arteries originate with the paramedian thalamus arteries from the basilar artery
Posterolateral thalamus	Tendency to fall to the side, SVV deviation, perhaps also astasia ipsiversive or contraversive	Thalamogeniculate arteries or perhaps branches of the posterior cerebral artery
Temporoparietal cortex	Tendency to fall to the side, SVV deviation, mainly contraversive	Branches of the middle cerebral artery

OTR ocular tilt reaction, *MLF* medial longitudinal fascicle, *riMLF* rostral interstitial nucleus of the MLF, *INC* interstitial nucleus of Cajal, *SVV* subjective visual vertical, *PICA* posterior inferior cerebellar artery

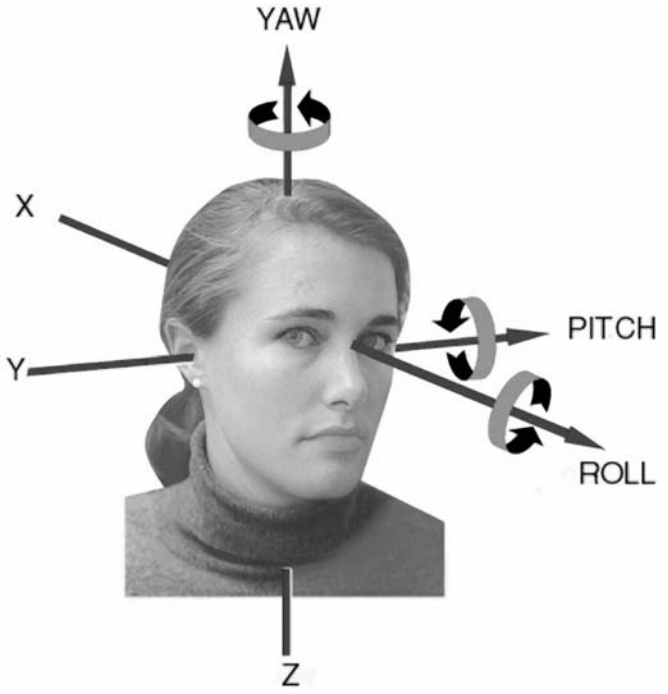


Figure 3.2. Orientation of the three major planes of action of the vestibulo-ocular reflex: yaw, pitch and roll.

Plane of action of the VOR	Clinical symptoms
Horizontal plane (yaw)	"Vestibular pseudo neuritis", spontaneous horizontal nystagmus Horizontal past-pointing to the right/left (subjective straight-ahead) Postural instability, falling tendency to one side, turning in the Unterberger step test
Sagittal plane (pitch)	Downbeat nystagmus, upbeat nystagmus Deviation of the subjective horizontal upwards or downwards Postural instability with a falling tendency forward or backward
Frontal plane (roll)	Ocular tilt reaction, skew deviation, ocular torsion, head tilt Deviation of the subjective visual vertical clockwise or counterclockwise Postural instability with a falling tendency to one side

• ***Vestibular Syndromes in the Horizontal (Yaw) Plane***

Vestibular syndromes in the horizontal (yaw) plane are rare, for example, horizontal benign paroxysmal positioning vertigo (BPPV) due to a canalolithiasis in the horizontal canal of the

labyrinth (Baloh et al. 1993). As far as we know, central syndromes in yaw are caused only by lesions in the area of the entry zone of the vestibular nerve in the medulla oblongata, the medial and/or superior vestibular nuclei, and the neighbouring integration centres for horizontal eye movements (nucleus praepositus hypoglossi and paramedian pontine reticular formation) (Figures 3.3 and 3.4). Other clinical signs are ipsilateral caloric hyporesponsiveness, horizontal gaze deviation, falling tendency to the affected side, and a past-pointing corresponding to a deviation of the "subjective straight-ahead". The clinical symptoms are similar to those of an acute peripheral vestibular lesion as occurs in vestibular neuritis and thus is also called "vestibular pseudo neuritis". In most cases there is a horizontal rotatory nystagmus. A purely central yaw plane syndrome is rare, because the area of a lesion that can theoretically cause a pure tonus imbalance in the yaw plane adjoins and in part overlaps with structures in the vestibular nuclei, which are also responsible for vestibular function in the roll plane. For this reason mixed patterns are found more frequently.

The most common causes include MS plaques or ischaemic infarctions within the vestibular nuclei or fascicles. If the lesion extends beyond the vestibular nuclei, other accompanying brainstem symptoms can be detected. Since mostly a unilateral medullary ischaemic or inflammatory brainstem lesion is present, the prognosis is favourable because central compensation takes place over the opposite side. The symptoms can be expected to resolve slowly within days to weeks. Thus, central compensation can be promoted by early balance training together with simultaneous treatment of the underlying illness.

- ***Vestibular Syndromes in the Sagittal (Pitch) Plane***

Vestibular syndromes in the sagittal (pitch) plane have so far been attributed to lesions in the following three places: paramedian bilaterally in the medullary and pontomedullary brainstem, the pontomesencephalic brainstem with the adjacent cerebellar peduncle, or the cerebellar flocculus bilaterally (Figure 3.3).

The **downbeat nystagmus syndrome** is characterised by a fixational nystagmus, frequently acquired, which beats downward in primary gaze position, is exacerbated on lateral gaze and in head-hanging position, may have a rotatory component, and is accompanied by a combination of visual and vestibulocerebellar ataxia with a tendency to fall backward and past-pointing upward (Baloh and Spooner 1981; Dieterich et al. 1998). The syndrome is frequently persistent. The individual components can differ, since there are probably other pathogeneses besides the vestibular one with imbalance in the graviceptive VOR (impairment in the projection of otolithic information), e.g., imbalance of

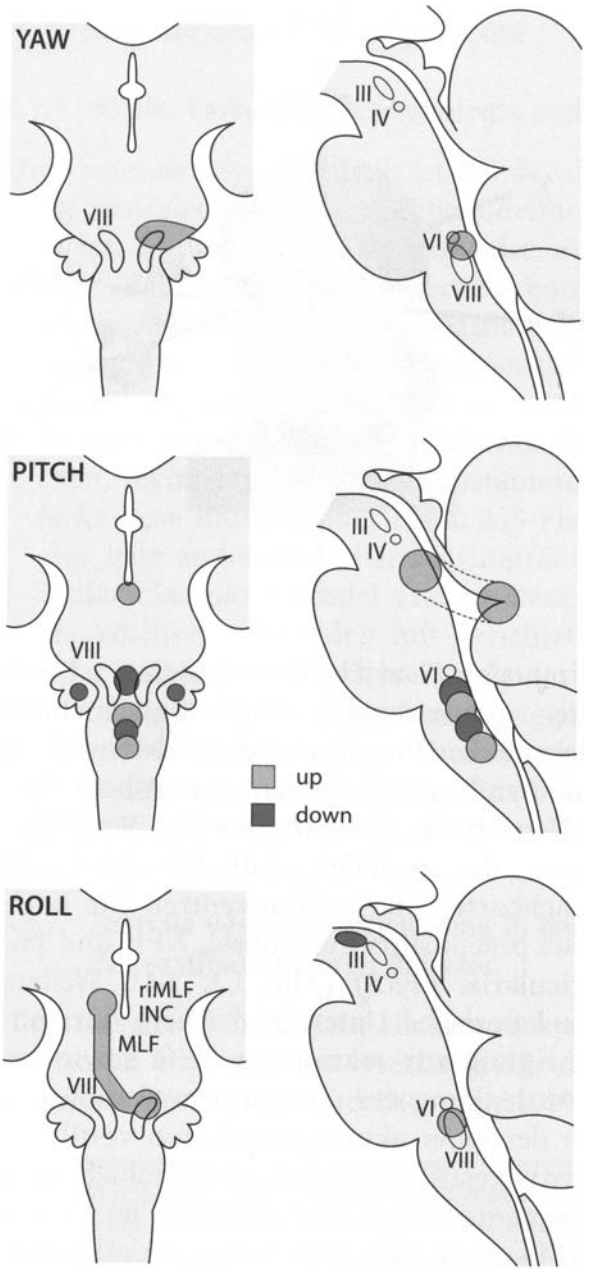


Figure 3.3. Schematic drawing of the brainstem and cerebellum with the typical sites of lesions that induce vestibular syndromes in the three planes of the vestibulo-ocular reflex. *III*, *IV*, *VI*, *VIII* cranial nerve nuclei, *MLF* medial longitudinal fascicle, *riMLF* rostral interstitial nucleus of the medial longitudinal fascicle, *INC* interstitial nucleus of Cajal.

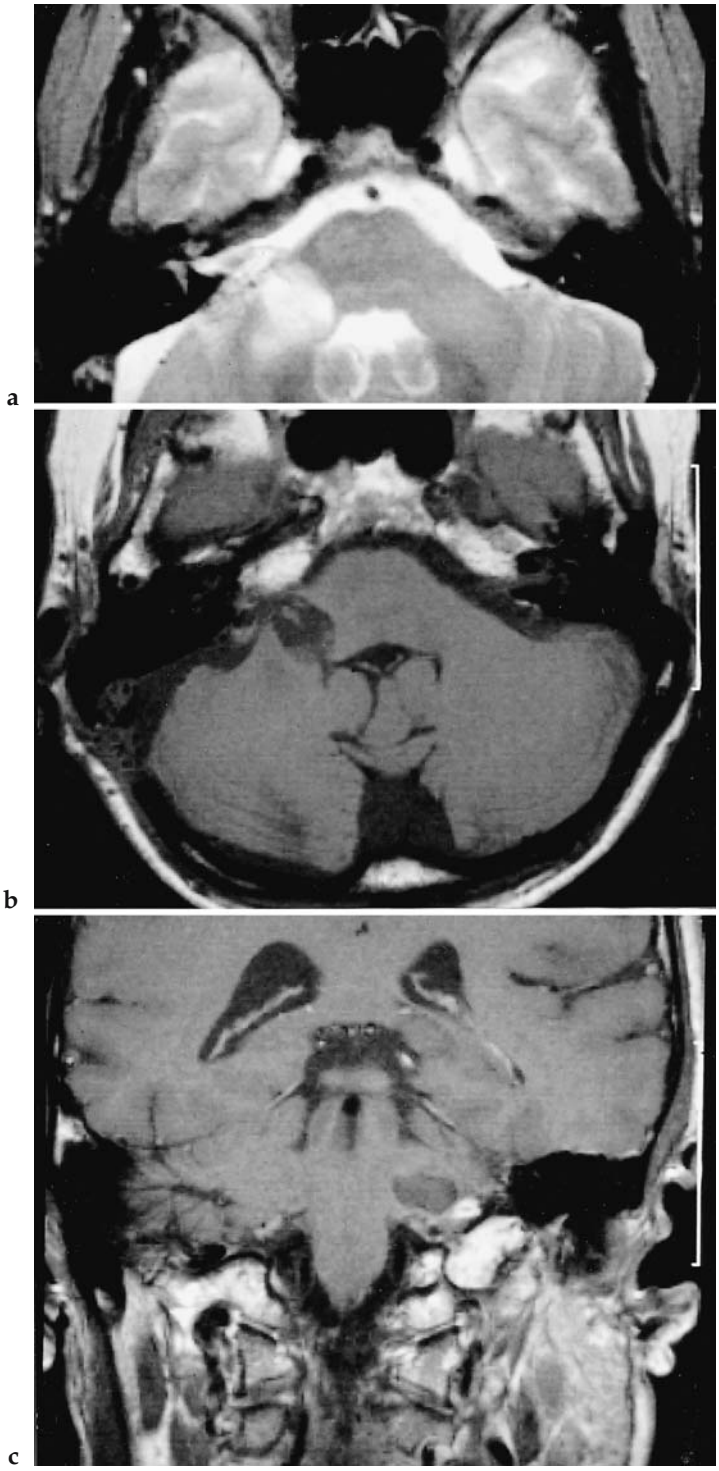


Figure 3.4. Magnetic resonance image of a patient with "vestibular pseudo neuritis", a disorder of the vestibulo-ocular reflex in the yaw plane. The **a** T2-weighted and **b, c** T1-weighted sequences show a pontomedullary brainstem infarction, which extends into the cerebellar peduncle and impairs the fascicle of the VIIIth cranial nerve as well as the region of the medial vestibular nucleus.

dysfunction of the neuronal ocular motor integrator and of the saccade-burst generator (Glasauer et al. 2003). Downbeat nystagmus is often the result of a bilateral lesion of the flocculus or the paraflocculus (e.g., intoxication due to anticonvulsant drugs) or caused by a lesion at the bottom of the fourth ventricle (Baloh and Spooner 1981; Zee et al. 1981). Accordingly it is mostly a drug-induced dysfunction or congenital: 25% of patients have craniocervical junction anomalies (Arnold–Chiari malformation), 20% have cerebellar degeneration, and more seldom lesions in MS. It can also be caused by a paramedian lesion of the medulla oblongata (Cox et al. 1981), and by MS, haemorrhage, infarction or tumour, for example. A downbeat nystagmus due to a lesion in the upper medulla at the level of the rostral nucleus praepositus hypoglossi has so far only been reported in monkeys, not in humans (DeJong et al. 1980).

Upbeat nystagmus is rarer than downbeat nystagmus. It is also a fixation-induced nystagmus that beats upward in primary gaze position, and is combined with a disorder of the vertical smooth-pursuit eye movements, a visual and vestibulospinal ataxia with a tendency to fall backward, and past-pointing downward (Janssen et al. 1998; Dieterich et al. 1998).

On the one hand, the pathoanatomic location of most acute lesions is paramedian in the medulla oblongata in neurons of the paramedian tract, close to the caudal part of the perihypoglossal nucleus (Janssen et al. 1998; Baloh and Yee 1989; Ranalli and Sharpe 1988), which is responsible for vertical gaze-holding (Büttner-Ennever et al. 1989). On the other hand, lesions have been reported paramedian in the tegmentum of the pontomesencephalic junction, the brachium conjunctivum, and probably in the anterior vermis (Nakada and Remler 1981; Kattah and Dagi 1990). The symptoms persist as a rule for several weeks but are not permanent. Because the eye movements generally have larger amplitudes, oscillopsia in upbeat nystagmus is very distressing and impairs vision. Upbeat nystagmus due to damage of the pontomesencephalic brainstem is frequently combined with a unilateral or bilateral internuclear ophthalmoplegia, indicating that the MLF is affected. The main aetiologies are bilateral lesions in MS, brainstem ischaemia or tumour (Figure 3.5), Wernicke's encephalopathy, cerebellar degeneration and dysfunction of the cerebellum due to intoxication.

The course and prognosis depend on the underlying illness. It is therapeutically expedient to attempt treatment of the symptoms of persisting downbeat or upbeat nystagmus syndrome by administering 4-aminopyridine (Strupp et al. 2003; Kalla et al. 2004), gabapentin (3×200 mg/day p.o.), baclofen (3×5–15 mg/day p.o.), or clonazepam (3×0.5 mg/day p.o.) (Dieterich et al. 1991; Averbuch-Heller et al. 1997).

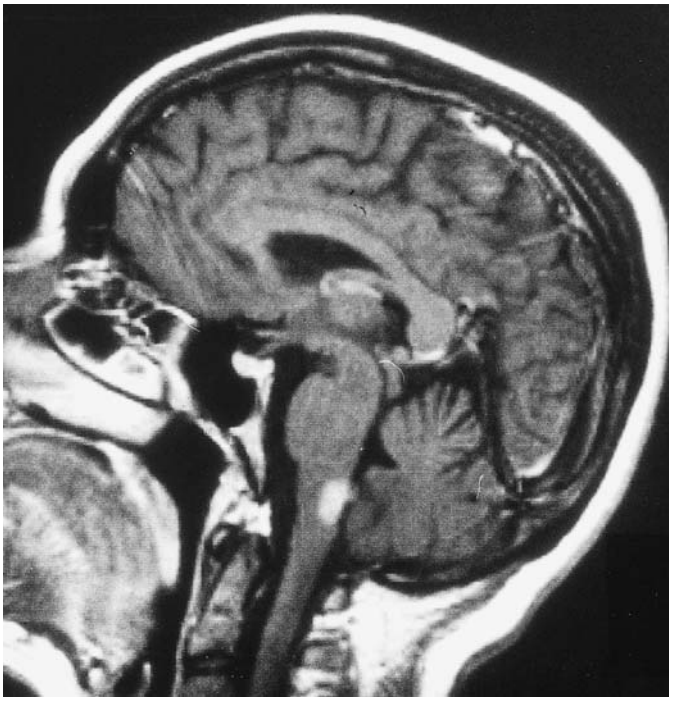
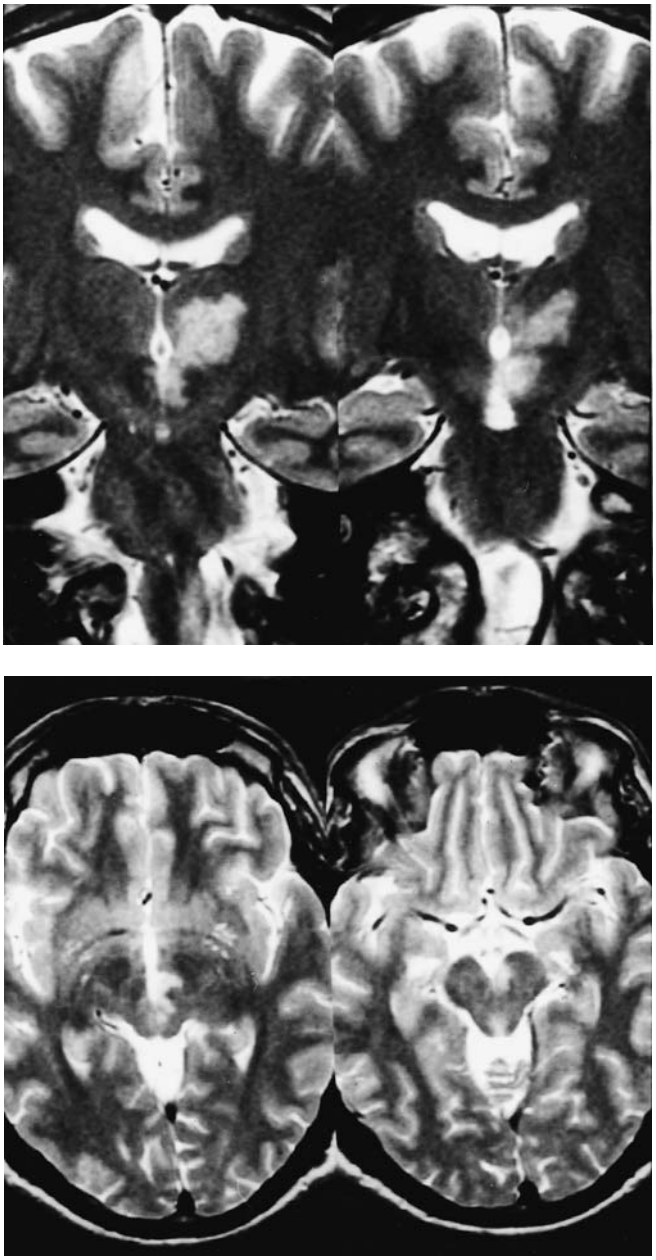


Figure 3.5. Magnetic resonance scan (with contrast medium) of a patient with an upbeat nystagmus syndrome that was induced by a contrast-enhancing tumour located near the median plane of the medulla oblongata.

- ***Vestibular Syndromes in the Vertical (Roll) Plane***
  - — These syndromes indicate an acute unilateral deficit of the "graviceptive" vestibular pathways, which run from the vertical canals and otoliths over the ipsilateral (medial and superior) vestibular nuclei and the contralateral MLF to the ocular motor nuclei and integration centres for vertical and torsional eye movements (INC and riMLF) in the rostral mid-brain (Brandt and Dieterich 1994; Dieterich and Brandt 1992, 1993a).
  - — More rostral to the midbrain, only the vestibular projection of the VOR for perception in the roll plane (determination of the subjective visual vertical, SVV) runs over the vestibular nuclei in the posterolateral thalamus (Dieterich and Brandt 1993b) to the parietoinsular vestibular cortex (PIVC) in the posterior insula (Brandt and Dieterich 1994; Brandt et al. 1994). The crossing of these pathways at pontine level is especially important for topographic diagnosis of the brainstem.
  - — All signs of lesions in the roll plane—single components or a complete ocular tilt reaction (i.e., head tilt, vertical diver-

gence of the eyes, ocular torsion, SVV deviation)—represent an ipsiversive tilt (ipsilateral eye lowermost) in both the unilateral peripheral vestibular lesion and the pontomedullary lesion (medial and superior vestibular nuclei) below the decussation in the brainstem.
— All signs in the roll plane—ocular motor, perceptual and postural—exhibit contraversive deviations (contralateral eye lowermost) for unilateral pontomesencephalic lesions of the brainstem above the decussation and indicate a deficit of the MLF or of the supranuclear center of the INC.
— Unilateral lesions of vestibular structures located rostral from the INC manifest with perceptual deficits only (deviation of the SVV) without accompanying ocular motor deficits or head tilt.
— Ocular tilt reaction in unilateral infarctions of the paramedian thalamus (in 50%) is caused by a simultaneous lesion in the paramedian rostral midbrain (INC).
— Unilateral lesions of the posterolateral thalamus can cause thalamic astasia with moderate ipsiversive or contraversive SVV tilts, which indicate involvement of the vestibular thalamic nuclei. This generally resolves within a matter of days or a few weeks.
— Unilateral lesions of the PIVC cause moderate, mostly contraversive SVV tilts lasting several days (Brandt et al. 1994).
— Perceptual deficits in the sense of pathological deviations of the SVV occur during unilateral deficits along the entire VOR projection and are one of the most sensitive signs of acute brainstem lesions (in ca. 90% of cases of acute unilateral infarctions) (Dieterich and Brandt 1993a).
— If instead of a functional deficit due to a lesion, there is an excitation of the VOR projection on one side, the same effects will be triggered, but in the opposite direction.
— If a torsional nystagmus occurs in the acute phase, the rapid nystagmus phase will be in the opposite direction of the tonic skew deviation and the ocular torsion (Helmchen et al. 1998).

Midbrain lesions occasionally induce complicated ocular motor syndromes in which a central vestibular deficit in the roll plane (e.g., due to the INC deficit) is combined with a nuclear or fascicular IIIrd nerve palsy (Figure 3.6). This leads to a "mixed pattern" that can nevertheless be clearly differentiated by determining the SVV, either binocularly or especially for each individual eye separately (monocular measurement), as well as by measuring the tonic ocular torsion by fundus photographs (Dichgans and Dieterich 1995). A central vestibular lesion induces in both eyes (with binocular vision) a contralateral SVV



a

b

Figure 3.6. Magnetic resonance image (T2-weighted) of a patient with a mixed pattern of ocular tilt reaction to the right and a lesion of the oculomotor nucleus and fascicle on the left due to an acute paramedian thalamus midbrain infarction, which was caused by a dissection of the left vertebral artery with embolism. The left-sided lesion encompasses in the upper midbrain the region of oculomotor nuclei with the fascicle as well as the supranuclear ocular motor and vestibular centres of the interstitial nucleus of Cajal and the rostral interstitial nucleus of the medial longitudinal fascicle. The deviation of the subjective visual vertical still amounted to +4–5° for the ipsilateral left eye, and +8–10° for the contralateral right eye 1 month after the infarction; fundus photography showed an incyclorotation of the left eye of 4° and an excyclorotation of the right eye of 7°. There was additional gaze palsy for upward and downward directions.

deviation of generally 10–20° in the same direction. If there is also a lesion of the IIIrd or IVth cranial nerve—mostly in one eye—the SVV deviation in the affected eye will be reduced or even reversed, with the result that the SVV deviation clearly differs for both eyes or points in totally opposite directions and the additionally affected eye looks in the ipsilateral direction (mixed pattern: ipsilateral eye to the ipsilateral side, contralateral eye to the contralateral side).

The measurement of the SVV for each eye separately also allows differentiation of the clinically similar peripheral infranuclear IIIrd and IVth nerve palsies from central vestibular syndromes, as infranuclear palsies cannot, of course, induce any SVV deviation or ocular torsion in both eyes (Dieterich and Brandt 1993c). Even the Bielschowsky head-tilt test does not allow the differentiation of a IVth nerve palsy and an ocular tilt reaction, as does determination of SVV.

The aetiology of these unilateral lesions is frequently an infarction of the brainstem or the paramedian thalamus, which extends into the rostral midbrain (Dieterich and Brandt 1993a,b). Course and prognosis depend also here on the aetiology of the underlying illness. One can count on a significant, generally complete recovery from the symptoms in the roll plane within days to weeks due to the central compensation over the opposite side (Dieterich and Brandt 1992, 1993b) (Figure 3.7).

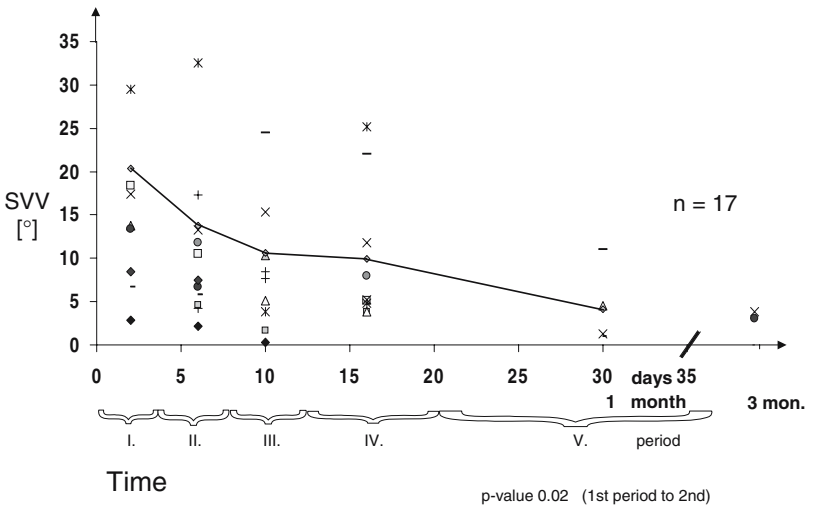


Figure 3.7. Course of the disorders in the roll plane represented by the deviation of the subjective visual vertical (SVV in °; normal range ± 2.5°) in 17 patients with an acute unilateral infarction of the lateral medulla oblongata. In most patients the deviation completely resolved within 4–6 weeks (due to central compensation of a unilateral central vestibular lesion).

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3.2 Basilar/Vestibular Migraine

3.2.1 Patient History

The main symptoms of basilar migraine are recurring attacks of various combinations of vertigo, ataxia of stance and gait, visual disorders and other brainstem symptoms accompanied or followed by occipitally located head pressure or pain, nausea and vomiting. As any movement increases the complaints, patients often have a need for rest.

3.2.2 Clinical Aspects and Course

If the attacks of vertigo are associated with other brainstem symptoms, more rarely also disturbances of consciousness, psychomotor deficits or changes of mood, they are called **basilar migraine**. The attacks can, however, be monosymptomatic, manifesting with only vertigo and perhaps also with a hearing disorder; these are called **vestibular migraine**. Monosymptomatic audiovestibular attacks predominate in ca. 75% of such cases (Dieterich and Brandt 1999). They are more difficult to recognise, especially if headache is missing (in ca. 30%; Dieterich and Brandt 1999). The diagnosis is simple if the attacks are usually or always followed by occipital pressure in the head or a headache and if there is a positive family or personal history of other types of migraine (ca. 50%; Dieterich and Brandt 1999). It is easier to establish the diagnosis if the following symptoms occur: light and

sound hypersensitivity, need for rest, tiredness after the attack and urge to urinate. The duration of the attacks of vertigo varies greatly and lasts either only seconds to minutes or several hours to days (Cutrer and Baloh 1992; Dieterich and Brandt 1999; Neuhauser et al. 2001). Contrary to other forms of migraine, more than 60% of persons with vestibular migraine also show slight central ocular motor disorders during attack-free intervals, e.g., a gaze-evoked nystagmus, saccadic smooth pursuit, a horizontal or vertical spontaneous nystagmus or central positional nystagmus (Dieterich and Brandt 1999). The patients are generally hypersensitive to movements and motion sickness, particularly during the migraine attack (Cutrer and Baloh 1992). This is similar to phonophobia and photophobia during migraine attacks, which is induced by a neuronal sensory overexcitability, for example of the inner-ear hair cells.

Originally, basilar migraine was described by Bickerstaff in 1961 as a typical illness of adolescence, which clearly predominated in females. Retrospective studies have, however, shown that basilar migraine with dizziness and vestibular migraine can develop throughout the patient's entire life, most often between the third and sixth decades (Dieterich and Brandt 1999; Neuhauser et al. 2001). The mean age of women at its first occurrence is ca. 38 years, that of men, around 42 years (Figure 3.8). The ratio of women and men affected is 1.5:1.

The frequency of basilar migraine with dizziness or vestibular migraine is 7–9% in dedicated outpatient clinics for dizziness (Dieterich and Brandt 1999; Neuhauser et al. 2001). The preva-

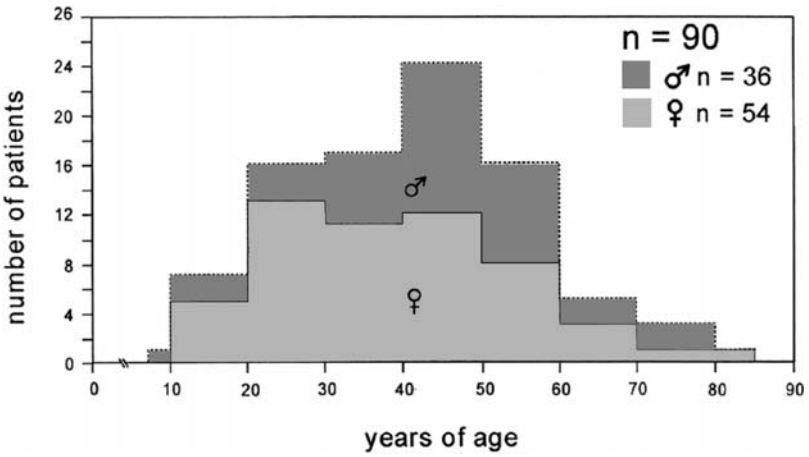

Figure 3.8. Age of 90 patients with basilar/vestibular migraine at the time of first manifestation of episodic dizziness (54 women, 36 men). Basilar/vestibular migraine can occur in women and men at any age, frequently between the ages of 20 and 60 years, and peaks between the ages of 40 and 50 years (Dieterich and Brandt 1999).

lence of basilar migraine with dizziness is not known; naturally that for migraine without aura is clearly higher and has a 1-year prevalence of 12–14% for women after puberty and 7–8% for men after puberty. Women are affected two to three times more often than men.

If the attacks take a monosymptomatic course without headache, they cannot be differentiated clinically from benign paroxysmal vertigo in childhood. The latter is considered equivalent to migraine with attacks that begin between the first and fourth year of life, last only seconds to minutes, and disappear spontaneously within a few years.

3.2.3 Pathophysiology and Therapeutic Principles

As regards the pathogenesis of basilar migraine, it is interesting that the rare episodic ataxia type 2 (due to a mutation of a gene on chromosome 19p in the calcium channel) occurs in several families in combination with hemiplegic migraine, which is also located on chromosome 19 (Ophoff et al. 1996). Moreover, the findings of central ocular motor disorders during the symptom-free interval, as in episodic ataxia, also indicate that patients with vestibular migraine may have a hereditary neuronal disorder in the brainstem nuclei (channelopathy?). Neuronal deficits in the brainstem are also discussed as factors in the pathophysiology of migraine without aura. In this primarily neurovascular headache syndrome, in which the trigeminovascular system, along with neurogenic inflammatory reactions, plays a central role, animal studies identified the nucleus locus coeruleus in the pontine brainstem as the modulator of the cerebral blood flow, the most important central nucleus of the noradrenergic system (Goadsby 2000). Furthermore, the serotonergic dorsal raphe nucleus in the midbrain seems to play an important role. Positron emission tomography studies have shown that this region and that of the dorsal pons with the nucleus coeruleus are also activated in patients during migraine attacks without aura (Weiller et al. 1995). However, these brainstem nuclei are also activated immediately after successful treatment of a migraine attack but not during the symptom-free interval. Drug therapy acts at various sites within the trigeminovascular system and the neurogenic inflammatory cascade.

3.2.4 Pragmatic Therapy

The same principles of therapy that have proven effective in migraine with aura are used, both for treating the attacks as well as for migraine prophylaxis, although the symptoms of dizziness and the accompanying headache can respond differently. To stop attacks lasting 45 minutes and longer, it is advisable to

administer an antiemetic (e.g., metoclopramide, domperidone) early, in combination with a non-steroidal anti-inflammatory agent (e.g., ibuprofen, diclofenac), an analgesic (acetylsalicylic acid as a soluble tablet or paracetamol as a suppository), or an ergotamine (ergotamine tartrate). The tryptanes are very effective against migraine attacks without aura and act at the 5-$HT_{1B/1D}$ receptors of the vascular walls. There is a relative contraindication for the treatment of migraine with aura because of the danger of a cerebral or cardiac infarction as a result of the vasoconstriction of the arteries. In individual cases, however, they have been reported to have positive effects in attacks of dizziness.

The treatment of first choice for migraine prophylaxis is the administration of the beta-receptor blocker metoprolol retard (ca. 100 mg/day) for about 6 months. Alternatives are valproic acid (600–1,200 mg/day) or lamotrigine (50–100 mg/day).

3.2.5 Ineffective Treatments

The sole use of opioids, antivertiginous drugs (dimenhydrinate) or anticonvulsants such as carbamazepine, diphenylhydantoin and primidone is ineffective.

3.2.6 Differential Diagnosis and Clinical Problems

Differentiating basilar migraine from transient ischaemic attacks, Menière's disease or vestibular paroxysmia can occasionally be difficult. In some cases the diagnosis can only be established on the basis of the response to a "specific" therapy. Transitional and mixed forms or pathophysiological combinations are being discussed, especially for Menière's disease and vestibular migraine. Currently there are no reliable data, in part because patients with primarily vestibular symptoms are more frequently misdiagnosed as having Menière's disease. This would explain the considerable difference in prevalence of migraine in patients with "classic" Menière's disease (22%) as opposed to "vestibular" Menière's disease (81%) (Rassekh and Harker 1992). A recent interview-based study involving 78 patients with certain unilateral or bilateral Menière's disease determined that the lifetime prevalence of migraine with and without aura is 56% compared with 25% in an age-matched control group (Radtke et al. 2002). This indicates that either there is a pathophysiological link between the two diseases or alternatively the current diagnostic criteria cannot yet differentiate between them.

As BPPV occurs three times more frequently in migraine patients than in trauma patients, according to a retrospective study (Ishiyama et al. 2000), it has been speculated that a relapsing functional deficit of the inner ear may be the underlying cause

of the vestibular migraine attacks (e.g., in the form of a vasospasm). The therapy for BPPV in migraine patients is similar to the liberatory manoeuvres used to treat idiopathic BPPV.

The rare episodic ataxia type 2 is also characterised by episodic attacks of dizziness with central ocular motor deficits, even during the attack-free interval (Griggs and Nutt 1995); here acetazolamide and 4-aminopyridine (Strupp et al. 2004) can be prescribed with success.

Transient ischaemic attacks in the vertebrobasilar system, basilar artery thrombosis, and brainstem/cerebellum haemorrhage can also accompany headache centred primarily in the nuchal region. These important conditions should be rapidly clarified in a differential diagnosis. Basilar artery thrombosis and brainstem haemorrhage usually develop quite rapidly, along with vigilance disorders, and can worsen to coma, increasing deficits of the cranial nerves and paresis, or sensory deficits in the extremities. A vertebral artery dissection can occur spontaneously or after head trauma or chiropractic manoeuvres. It is associated with occipital head and neck pain, neck pressure, dizziness and other brainstem symptoms. As brainstem ischaemia, which is induced in the context of various mechanisms, is part of various differential diagnoses with acute life-threatening significance, it is especially important to consider the possibility of this dangerous brainstem ischaemia during the appearance of the first or first three migraine attacks, in order to quickly introduce adequate diagnostic testing (MRI, CT scan, ultrasound).

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4

Traumatic Forms of Vertigo

Introduction

After head and neck pain, vertigo/dizziness is the most frequent complication of a head trauma or a whiplash injury. If a petrous bone fracture is not seen radiologically, and since it is not possible to clinically confirm a brainstem concussion, the first questions to ask are the following: Is this dizziness organic or psychogenic? What is the underlying mechanism of the dizziness (peripheral, central vestibular or cervical)?

The following post-traumatic forms of vertigo/dizziness are well known: benign paroxysmal positioning vertigo (BPPV, canalolithiasis), labyrinth dysfunction (e.g., petrous bone fracture), labyrinth concussion, perilymph fistula (perilymph leakage) or barotrauma-induced vertigo. Following a whiplash injury, cervicogenic vertigo is much too frequently diagnosed; however, the underlying pathomechanism is not clear. Traumatic dizziness is probably often a result of the loosening of otoconia (without canalolithiasis), which subsequently causes a post-traumatic otolith vertigo in the form of transient gait and movement instability as well as oscillopsia during any linear acceleration of the head.

4.1 Traumatic Peripheral Vestibular Vertigo/Dizziness

4.1.1 Benign Paroxysmal Positioning Vertigo

The most frequent peripheral labyrinthine form of vertigo is BPPV. It is characterised by short attacks of rotatory vertigo and

typical crescendo–decrescendo nystagmus (see Section 2.1), which resolves within seconds and is triggered by positioning the head toward the affected ear or tilting it backward. Rotatory vertigo and nystagmus occur after positioning with a short latency of seconds and cease temporarily after repeated positioning manoeuvres. BPPV occurs in approximately 17% of patients as post-traumatic positioning vertigo. It is frequently bilateral and asymmetrical (ca. 20%) and occasionally also occurs in children. Its pathophysiology and therapy correspond to that of idiopathic BPPV. Because it frequently occurs bilaterally, the therapy phase is occasionally longer and liberatory manoeuvres have to be repeated beginning with the treatment of the more-affected ear until the patient is symptom-free. Three different types of manoeuvre are successfully used for such an aetiology (Semont, Epley, and Brandt–Daroff [Herdman 1990]). All are suitable for treating canalolithiasis of the posterior canal. In cases of the more rarely affected horizontal canal, the simple 270° rotations toward the unaffected ear around the longitudinal axis while the patient is supine are used in combination with subsequent bed rest on the side of the unaffected ear for 12 hours (see Section 2.1.5).

4.1.2 Traumatic Labyrinthine Failure

A unilateral haemorrhage or petrous bone fracture can lead to direct injury of the vestibular nerve or of the labyrinth. This causes violent rotatory vertigo, which continues for days, horizontal rotatory nystagmus to the un-affected side, posture and gait instability and nausea and vomiting. The clinical symptoms are similar to those of vestibular neuritis (see Section 2.2). Two forms of petrous bone fractures can be differentiated: longitudinal and transverse. Longitudinal petrous bone fractures (Figure 4.1) account for about 80% of cases; they cause injury to the middle ear and bleeding from the ear. About 20% of cases are the result of a transverse petrous bone fracture with labyrinthine lesion and resulting rotatory vertigo and hearing loss, as well as possible injury to the facial nerve. Transverse petrous bone fractures cause vestibular and cochlear symptoms much more often than longitudinal fractures. If there is a direct trauma of the petrous bone and corresponding symptoms of rotatory vertigo and hearing loss, but the injury cannot be confirmed either macroscopically or by X-ray, one must consider a labyrinthine concussion.

The first phase of dysfunction is characterised by a strong feeling of illness with continuous rotatory vertigo, nausea and vomiting. These symptoms dissipate after 2–3 weeks. Bed rest and antivertiginous drugs (e.g., dimenhydrinate, benzodiazepines) should be prescribed only within the first days for severe nausea and vomiting. The same limited therapy is recom-

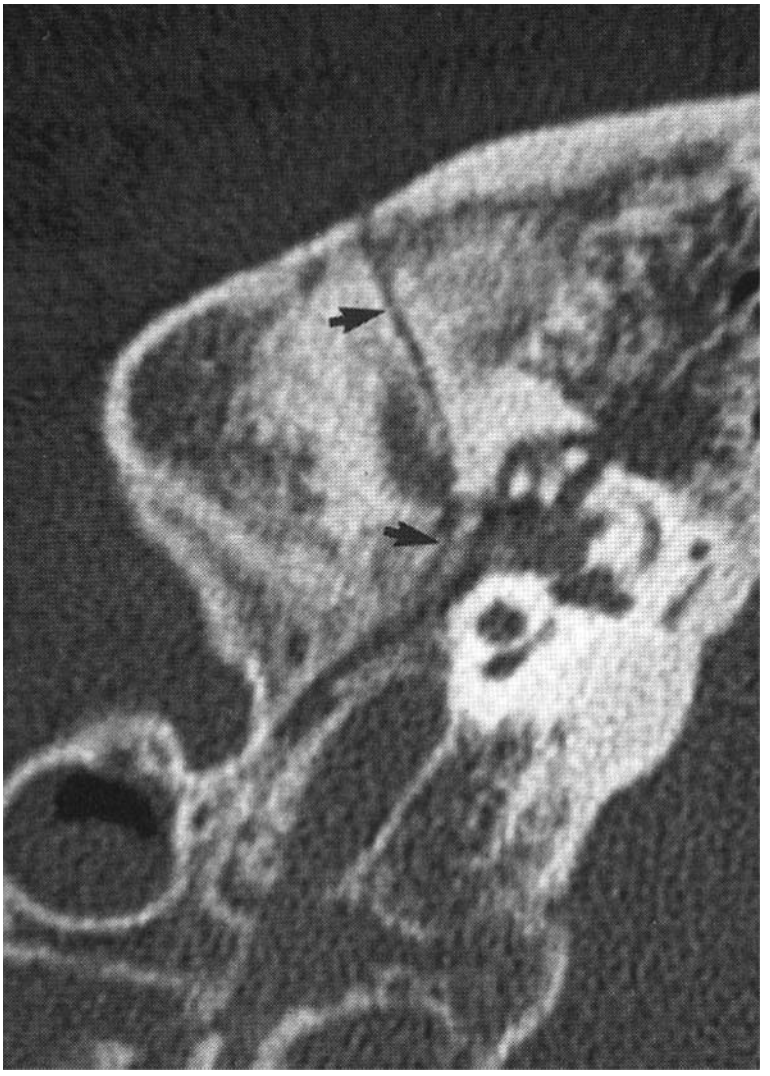

Figure 4.1. Longitudinal fracture of the petrous bone (*arrows*); axial projection, computed tomography. The fracture extends to the geniculate ganglion of the facial nerve and then follows the carotic canal.

mended for vestibular neuritis, as these drugs delay central compensation. The patient should begin a vestibular training programme as soon as possible to accelerate and improve central compensation (Strupp et al. 1998). Similarly, treatment with corticosteroids (methylprednisolone) is also indicated for several days, in most cases because of trauma-induced oedema.

4.1.3 Perilymph Fistula

Usually the air in the middle ear is of normal atmospheric pressure, as pressure is equalised via the Eustachian tube. During head trauma, extreme increases in pressure can occur in the

middle ear and cause a leakage of the perilymph at the round or oval windows; a luxation of the stapes footplate in the direction of the inner ear can also occur, but more rarely. Attacks of dizziness with fluctuating hearing loss, ear pressure and tinnitus follow. The patient's complaints frequently depend on the position of the head, on movement or on air pressure and can be exacerbated by pressing (Valsalva manoeuvre, lifting heavy objects or sneezing). This is also true in perilymph fistulas of other aetiologies (see Section 2.6).

Clinically, vertigo/dizziness can be classified as either a canal type with rotatory vertigo and nystagmus or an otolith type (in cases of fistulas of the oval window) with unsteadiness, gait ataxia and oscillopsia, especially during linear head acceleration (when standing up or walking). The otolith type of dizziness can also be caused by luxation of the stapes footplate without resulting in continuous perilymph leakage; this happens when the luxated stapes footplate mechanically stimulates the otoliths during the acoustically induced stapedial reflex (otolithic Tullio phenomenon). At the same time, sound induces paroxysmal otolithic symptoms (eye movements and head tilt, oscillopsia and tendency to fall).

In the majority of cases, the initially conservative therapy of several days of bed rest with the head elevated, perhaps mild sedation and the administration of laxatives, results in spontaneous recovery. If such conservative therapy fails and hearing reduction or vestibular symptoms increase, an exploratory tympanotomy is indicated.

4.1.4 Alternobaric Vertigo

Rapid changes of pressure in the middle ear—primarily during the decompression experienced by divers or by aircrews during flights—can cause a transient rotatory vertigo that is called alternobaric vertigo. At the very beginning of the rotatory vertigo and nystagmus, which spontaneously resolves after seconds or hours, there is a feeling of fullness in the ear. Acute rotatory vertigo indicates an inadequate stimulation of a semicircular canal, which is triggered by excessive acute pressure on the round and oval windows in the middle ear. The same mechanism is found in perilymph fistulas.

4.1.5 Otolith Vertigo

The incidence of vertigo following head traumas of various aetiologies varies according to the available data between 14% and 40–60%. Traumatic otolith vertigo probably occurs more often (Brandt and Daroff 1980). Immediately after head trauma, patients often describe a postural imbalance and oscillopsia that

are exacerbated by head movements and gait instability that is like walking on a water pillow. These are typical disorders of otolith function. The traumatic accelerations probably cause loosening of the otoconia, which leads to unequal otolith masses on the two sides. This has been shown in animal experiments. Because of the different otolith weights on the two sides, a temporary disturbance of spatial orientation can result. Within days or weeks, however, a central compensation corrects for the otolith imbalance and the postural instability during head movements, and gait ataxia resolves.

4.2 Traumatic Central Vestibular Forms of Vertigo

The various central vestibular syndromes are triggered by disorders of brainstem function in connection with a concussion or haemorrhage. In principle, all parts of the brainstem and cerebellum, from the midbrain across the pons to the medulla oblongata and cerebellum, can be affected, depending on the localisation of the haemorrhage. However, the mesencephalon is affected somewhat more often. The individual syndromes are described in Section 3.1.

4.3 Traumatic Cervical Vertigo

The question of whether there is a medical entity "cervicogenic vertigo" is still a subject of controversy (see Section 6.3). The neck afferents not only take part in coordinating eyes, head and body, but they are also involved in the orientation of the body in space and the control of posture. This means that in principle a stimulation or lesion of these structures can trigger a "cervicogenic vertigo". It has been shown in animal experiments in primates (*Macaca*) that a unilateral local anaesthesia or section of the superior cervical roots induces a falling tendency due to a temporary increase of muscle tonus on the ipsilateral side and a decrease on the contralateral side, as well as ipsiversive past-pointing. Positional nystagmus, however, is elicited only in certain species and to different degrees (most pronounced in rabbits, less in cats) but not at all in Rhesus monkeys (DeJong et al. 1977). This type of positional nystagmus that derives from an imbalance in tonus of the superior cervical roots cannot be proven in humans. Patients who had a blockade of the C2 roots (for cervicogenic headache) exhibited a slight instability of gait with minor ipsilateral deviation of gait and past-pointing without ocular motor disorders (Dieterich et al. 1993); this corresponds to the animal experiments with *Macaca*. One would expect similar symptoms such as instability of gait in "cervicogenic vertigo", always in connection with

cervico-vertebrogenic pain and movement restriction of the spinal cord. One would not expect rotatory vertigo or spontaneous, positional or provocation-induced nystagmus.

Unfortunately, there are still no useful tests to confirm "cervicogenic vertigo" (gait instability), as the tests performed, which use passive head turns while the trunk is fixed, trigger the same amount of nystagmus with the same frequency in healthy subjects (Holtmann et al. 1993). These tests are still in use in many places nowadays, but they fail to give meaningful results.

4.4 Post-traumatic Psychogenic Vertigo

If vertigo persists for a long time after a head trauma or a whiplash injury and deficient otoneurological or ocular motor findings cannot be determined, this can indicate a psychogenic vertigo (see also Chapter 5). The most frequent psychosomatic form of vertigo and the second most frequent cause of vertigo in neurological patients is the somatoform disorder phobic postural vertigo. It often occurs secondary to organic forms of vertigo. In cases of chronic, long-term complaints, the desire to retire ("secondary gain") must also be considered in the differential diagnosis.

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5

Psychogenic Forms of Vertigo and Dizziness

Introduction

Somatoform disorders play a causal or contributory role in a large portion of patients presenting with complex forms of dizziness. In the course of their illness, even after several years, about 70% of these patients with complex somatoform dizziness still show symptoms and are more impaired in their professional and daily activities than those with organic forms of dizziness (Furman and Jacob 1997; Yardley and Redfern 2001; Eckhardt-Henn et al. 2003). The most frequent underlying psychiatric disorders are anxiety and depression as well as dissociative somatoform (ICD-10:F45) disorders.

Somatoform dizziness first occurs without psychopathological symptoms. Most often this causes the patients to go to otolaryngologists, neurologists or internists. The patients describe experiencing frequent postural imbalance or a diffuse feeling of dizziness (a feeling of numbness, light-headedness, unsteadiness when walking, a feeling of toppling over) or very rarely rotatory vertigo with accompanying vegetative symptoms and nausea. Depending on the underlying psychiatric illness (see above), the following additional symptoms can be present: disorders of motivation and concentration, decline in performance, restrictions in professional and daily activities that are subjective, vegetative symptoms that accompany the dizziness (accelerated heart rate, nausea, sweats, apnoea, fear of suffocating, loss of appetite, weight loss), emotional and mood disorders, sleep disturbances and symptoms of anxiety. Typically all of these symptoms are

experienced. The patients believe that the symptoms are triggered and induced by the dizziness. Patients seldom spontaneously report conflict and stress situations that can function as triggers of vertigo/dizziness; often they are initially totally unaware of them. This makes it difficult to establish the diagnosis.

The treatment depends on the clinical picture. Psychotherapy should be begun. Psychodynamic and also behavioural therapy are advisable; the choice depends on the clinical finding and the underlying conflict or stress situation. Outpatient therapy focused on the leading symptom can be quite successful in cases of short-term dizziness that is not very pronounced. Long-term techniques should be selected depending on the underlying conflict situation (e.g., psychoanalysis). For subjects with a strongly pronounced disorder and considerable suffering, we recommend combination therapy with a psychoactive drug; the drugs of choice are preparations belonging to the serotonin-reuptake inhibitors (e.g., paroxetine, citalopram or sertraline). In very few patients it is necessary to initially supplement these drugs with an anxiolytic drug (e.g., lorazepam) for a short time.

In the following section, phobic postural vertigo, an important form of somatoform dizziness and the second most common form of vertigo (see Table 1.1, p. 5), is discussed.

5.1 Phobic Postural Vertigo

5.1.1 Patient History

The cardinal symptoms and features of phobic postural vertigo include the following (Brandt and Dieterich 1986; Huppert et al. 1995; Brandt 1996):

- Patients complain about postural dizziness and subjective postural and gait unsteadiness without this being visible to an observer.
- Dizziness is described as a numbness with varying degrees of unsteadiness of posture and gait, attack-like fear of falling without any real falls, in part also unintentional body swaying of short duration.
- The attacks often occur in typical situations known to be external triggers of other phobic syndromes (e.g., large crowds of people in a store or restaurant, bridges, driving a car, empty rooms).
- During the course of the illness, the patient begins to generalise the complaints and increasingly to avoid the triggering stimuli. During or shortly after the attacks (frequently mentioned only when asked), patients report anxiety and vegetative disturbances; most also report attacks of vertigo without anxiety.

- If asked, patients frequently report that the complaints improve after imbibing a little alcohol and during sports.
- Frequently at the beginning, there is an organic vestibular illness, e.g., resolved vestibular neuritis, benign paroxysmal positioning vertigo (Huppert et al. 1995) or psychosocial stress situations (Kapfhammer et al. 1997).
- Patients with phobic postural vertigo often exhibit obsessive–compulsive and perfectionistic personality traits and during the course of the disease reactive–depressive symptoms.

5.1.2 Clinical Aspects and Course of the Illness

The combination of postural vertigo with subjective instability of posture and gait in patients with normal neurological findings in vestibular and balance test results (otoneurological examination, electronystagmography including caloric irrigation, posturography) or disorders that cannot explain the complaints, and a compulsive personality structure, is characteristic. The monosymptomatic subjective disorder of balance is connected with standing or walking, manifests with attack-like worsening that occurs with or without recognisable triggers and with or without accompanying anxiety. The absence of recognisable triggers or vertigo without accompanying anxiety causes many patients and the doctor treating them to doubt the diagnosis of a somatoform disorder.

Patients with phobic postural vertigo generally have a compulsive primary personality (in the sense of "pronounced personality traits") and a tendency to intensified introspection and the need "to keep everything under control". They are more likely to be ambitious and place high demands on themselves, and are often easily irritated and fearful.

Such patients rarely go to a psychiatrist first; they tend to see the "specialist" for their symptom, especially as they feel themselves to be organically sick. However, as phobic postural vertigo is not yet part of the diagnostic repertoire of most neurologists and otolaryngologists, the illness often lasts quite a long time before a diagnosis is established (a mean of 3 years for 154 patients with phobic postural vertigo; Huppert et al. 1995). Diagnosis is established only after a number of visits to different specialists, superfluous laboratory examinations, and erroneous classifications such as "cervicogenic vertigo" or "recurrent vertebrobasilar ischaemia", with correspondingly unsuccessful treatment attempts. A psychiatric longitudinal study confirmed that phobic postural vertigo is a unique medical entity, which can be clearly differentiated from panic disorder with or without agoraphobia (Kapfhammer et al. 1997).

Phobic postural vertigo can manifest in adults of every age, most often in the second and fifth decades; it is the most common

form of vertigo in this age group (Strupp et al. 2003). There is no sexual predominance. If phobic postural vertigo remains untreated, the complaints are exacerbated, generalisation develops, and avoidance behaviour increases until the patient is unable to leave his own apartment without help.

5.1.3 Pathophysiology and Therapeutic Principles

We have tried to explain the illusory perception of postural vertigo and postural instability by hypothesising that there is a disturbance of space constancy, which results from a decoupling of the efference-copy signal for active head and body movements. Under normal conditions, we do not perceive such slight, self-generated body sway or involuntary head movements during upright stance as accelerations. The environment also appears to be stationary during active movements, although there are shifts of retinal images caused by these relative movements. Space constancy seems to be maintained by the simultaneous occurrence of a voluntary impulse to initiate a movement and the delivery of adequate information in parallel to identify self-motion (Figure 5.1). According to von Holst and Mittelstaedt (1950), this efference copy may provide a sensory pattern of expectation based on earlier experience, which by means of the movement-triggered actual sensory information is then so interpreted that self-motion can be differentiated from the motion of the environment. If this efference copy is missing, e.g., if we move the eyeball by a finger on the eyelid, illusory movements of the environment occur, so-called oscillopsia. The sensation of vertigo described by phobic patients (involving involuntary body sway and the occasional perception of individual head movements as disturbing external perturbations) can be explained by a transient decoupling of efference and efference copy, leading to a mismatch between anticipated and actual motion. Healthy persons can experience similar mild sensations of vertigo without simultaneous anxiety during a state of total exhaustion, when the difference between voluntary head movements and involuntary sway becomes blurred. In phobic patients, this partial decoupling may be caused by their constant preoccupation with anxious monitoring and checking of balance. This leads to the perception of sensorimotor adjustments that would otherwise occur unconsciously by means of learned (and reflex-like) muscle activation programmes called up to maintain upright posture.

Precise posturographic analyses show that these patients increase their postural sway during normal stance by co-contracting the flexor and extensor muscles of the foot. This is evidently an expression of an unnecessary fearful strategy to control stance. Healthy subjects use this strategy only when in

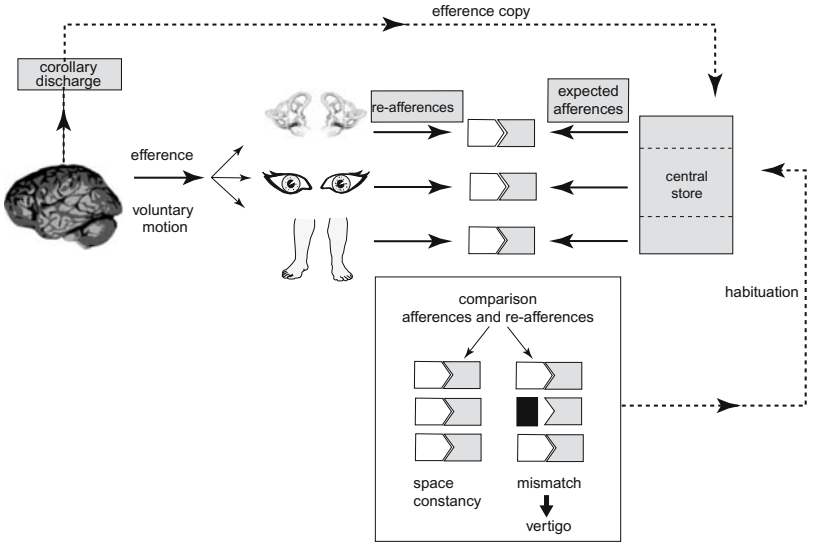

Figure 5.1. Schematic diagram of how dizziness develops as a result of an impairment of the space constancy mechanism during active movements. Intentional head movements cause sensory stimulation of the vestibular, visual and somatosensory organs. Their signals are compared with a multisensory pattern of expectation calibrated by earlier experience of movements. The pattern of expectation is prepared by the efference-copy signal, which is emitted parallel to and simultaneously with the voluntary movement impulse. If concurrent sensory stimulation and the pattern of expectation are in agreement, self-motion is perceived while "space constancy" is maintained. If there is a partial "decoupling" of the efference-copy signal and thus a sensorimotor mismatch between the input and the expected pattern, vertigo and imbalance develop. The patient no longer experiences an intentional self-generated head motion in a stationary environment, but rather an exogenic head perturbation and simultaneously an illusory movement of the environment.

real danger of falling. During difficult balancing tasks, such as tandem stance with closed eyes, the posturographic data of the patients do not differ from those of healthy subjects, i.e., the more difficult the demands of balance, the more "healthy" the balance performance of the patients with phobic postural vertigo (Querner et al. 2000). Patients with phobic postural vertigo often report a particularly increased unsteadiness when looking at moving visual scenes. However, when exposed to large-field visual motion stimulation in the roll plane, body sway does not exhibit any increased risk of falling (Querner et al. 2002). Vibratory stimulation showed that the patients were more sensitive to proprioceptive disturbances than healthy subjects, and less apt to use visual information to control upright stance (Holmberg et al. 2003).

A doctor–patient consultation that provides a detailed explanation of the mechanism of the disease and of the necessity of self-controlled desensitisation, i.e., the patient consciously confronts those situations that induce dizziness, is essential for the therapy to succeed.

5.1.4 Pragmatic Therapy

The treatment is based on three or four measures:

- thorough diagnosis and diagnosis of exclusion
- "psychoeducational" explanation
- desensitisation by self-exposure to triggers and regular exercise
- if complaints persist, behavioural therapy with or without accompanying pharmacotherapy (Brandt 1996).

In our experience, the most important therapeutic measure is to relieve the patient of his fear of having an organic illness by carefully examining him and explaining the psychogenic mechanism ("increased self-observation" in the context of the corresponding primary personality structure). Desensitisation by exposure to the causative situations should follow, i.e., the patient should not avoid such situations but, on the contrary, seek them out. At the same time, regular exercise has proven to be helpful to give the patient confidence in his own sense of balance. If the explanation and self-densensitisation do not result in sufficient improvement after weeks to months, behavioural therapy with or without drug therapy should be started, i.e., with a selective serotonin-reuptake inhibitor (e.g., paroxetine, 10–40 mg/day) or a tri-/tetracyclic antidepressive for 3–6 months. In rare cases of situation-dependent attacks, tranquillisers can also be administered to certain patients, although there is a danger of addiction.

In a follow-up study (0.5–5.5 years after the initial diagnosis) involving 78 patients, we showed that 72% of the patients were free of symptoms or exhibited a clear improvement after receiving therapy (Brandt et al. 1994). In a more recent long-term follow-up study (5–15 years) of 106 patients, the improvement rate was 75%; 27% reported a complete remission (Huppert et al. 2005). There was a negative correlation between the duration of the condition before assessment of the diagnosis and the improvement/regression rate. There was no indication in both follow-up studies that we had misdiagnosed anyone.

The readiness of most of the patients, who experience much stress as a result of their suffering, to understand the psychogenic mechanism and to overcome it by desensitisation is a positive experience for both the physician and the patient.

5.1.5 Differential Diagnosis and Clinical Problems

The differential diagnosis of phobic postural vertigo includes psychiatric–psychogenic syndromes as well as vestibular and non-vestibular organic syndromes.

The most important psychiatric syndromes include:

- panic disorder with or without agoraphobia
- space phobia (Marks 1981)
- visual vertigo (Bronstein 1995, 2004)
- mal de debarquement syndrome (Murphy 1993)
- depression.

The most important organic syndromes include:

- primary orthostatic tremor with a pathognomonic frequency peak of 14–16 Hz in electromyography and posturography (Yarrow et al. 2001)
- bilateral vestibulopathy (Section 2.5)
- vestibular paroxysmia (Section 2.4)
- perilymph fistula or superior canal dehiscence syndrome (Section 2.6)
- basilar/vestibular migraine (Section 3.2)
- episodic ataxia types 1 and 2
- neurodegenerative disorders (spinocerebellar ataxias, multisystem atrophy)
- central vestibular syndromes (Section 3.1)
- orthostatic dysregulation.

In contrast to the long list of possible differential diagnoses, the combination of traits connected with the complaints, normal physical findings and primary personality type is so characteristic that there is seldom any doubt as to the diagnosis after the first examination.

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6

Various Vertigo Syndromes

6.1 Vertigo/Dizziness in Childhood and Hereditary Vertigo Syndromes

Dizziness occurs less often as a major symptom in childhood than in adulthood. Most forms of dizziness and vestibular syndromes of adulthood, however, can also appear in childhood. For this reason we will limit ourselves in this chapter to the essential features of an indicative patient history. Episodic vertigo syndromes can manifest in childhood as an equivalent of migraine, an epileptic aura, or in the presence of perilymph fistulas; they seldom occur in the context of a familial episodic ataxia. Sustained rotatory vertigo can be the sequela of vestibular neuritis, a labyrinthitis or brain concussion. Benign paroxysmal positioning vertigo in children is also often caused by trauma. Oscillopsia during head movements and balance disorders that worsen in darkness are typical for bilateral vestibulopathy, which can develop in children after a case of bacterial meningitis, for example, or be caused by ototoxic antibiotics. Moreover, labyrinthine malformations may also cause congenital bilateral vestibulopathy (Table 6.1). The treatment of these various forms of vertigo corresponds to that in adults; however, a paediatrician should be closely consulted.

The following three main complaints (with or without accompanying clinical findings) are helpful for the differential diagnosis of childhood forms of vertigo.

Attacks of vertigo

- *Episodic vertigo without pathological findings in the attack-free interval:*

Table 6.1. Vertigo or loss of vestibular function in childhood

Labyrinth/nerve	Central vestibular origin
Hereditary/congenital	
• Labyrinthine malformation (see Table 6.2)	Familial episodic ataxia type 2
• Perilymph fistulas	Downbeat nystagmus
• Congenitally acquired malformations —rubella, cytomegalovirus —toxic agents	Upbeat nystagmus
• Various hereditary audiovestibular syndromes (see Table 6.2)	
• Familial vestibular areflexia	
• Syphilitic labyrinthitis (endolymphatic hydrops)	
Positive migraine family history Benign paroxysmal vertigo of childhood Basilar/vestibular migraine	
Acquired	
• Labyrinthitis with vestibulopathy (viral, bacterial, tuberculous)	—Infratentorial tumours (medulloblastoma, astrocytoma, epidermoid cysts, meningeoma)
• Perilymph fistulas	—Epileptic aura, vestibular epilepsy
• Trauma (temporal bone fracture, benign paroxysmal positioning vertigo)	—Trauma (brainstem or vestibulocerebellar concussion)
• Menière's disease (endolymphatic hydrops)	—Encephalitis
• Vestibular neuritis	—Toxic agents (e.g., upbeat/downbeat nystagmus when on anticonvulsants)
• Herpes zoster oticus	
• Cholesteatoma	
• Ototoxic drugs	
• Cogan's syndrome, other inner-ear autoimmune disorders	

benign paroxysmal vertigo of childhood/vestibular migraine, vestibular paroxysmia, epileptic aura or vestibular epilepsy, orthostatic dysregulation, psychogenic vertigo

- *Episodic vertigo with inner-ear hypoacusis:*
  perilymph fistula, Menière's disease, vestibular paroxysmia
- *Episodic vertigo with oculomotor abnormalities in the attack-free interval:*
  basilar/vestibular migraine, familial episodic ataxia type 2
- *Episodic vertigo followed in the course of the disease by oscillopsia during head movements and imbalance that worsens in darkness:*

Table 6.2. Syndromal hearing losses

	Vestibular	Other organs	Heredity
Usher type I	+	Eyes	AR
Usher type II	–	Eyes	AR
Usher type III	+/–	Eyes	AR
Alström	+	Eyes Diabetes Obesity	AR
Refsum	+	Eyes Nerves	AR
Waardenburg	?	Skin Eyes	AD
Alports	+	Kidney Eyes	AD, AR, X
Pendred	+/–	Thyroid gland	AR
Jervell–Lange	?	Heart	AR

+ vestibular hypofunction, – normal, +/– variable, ? unknown, *AD* autosomal dominant, *AR* autosomal recessive, *X* X-linked (Møller 2003).

development of bilateral vestibular failure (also with familial vestibulopathy)

Sustained vertigo

- *Sustained vertigo without hearing loss:*
  vestibular neuritis
- *Sustained vertigo with hearing loss:*
  labyrinthitis, inner-ear autoimmune disease
- *Post-traumatic sustained vertigo:*
  temporal bone fracture, labyrinthine concussion

Oscillopsia with stance and gait imbalance

- *Delayed stance and gait development with or without hearing loss:*
  congenital bilateral vestibulopathy
- *Oscillopsia during head movements and gait imbalance that worsens in darkness:*
  congenital or early acquired bilateral vestibulopathy, perilymph fistula, post-traumatic otolith vertigo
- *Slowly increasing oscillopsia during head movements, gait imbalance, and inner-ear hypoacusis or hearing loss:*
  various hereditary and congenital disorders causing progressive audio-vestibular loss
- *Progressive ataxia, balance and ocular motor disorders:*
  infratentorial tumours with lesions of the vestibulocerebellar and pontomedullary brainstem structures, spinocerebellar ataxias

We will now discuss in detail three forms of vestibular syndromes of childhood.

6.1.1 Benign Paroxysmal Vertigo of Childhood

Benign paroxysmal vertigo of childhood is a vestibular migraine with aura but without headache. Probably the most frequent form of episodic vertigo in childhood, it has a prevalence of 2.6% (Abu-Arafeh and Russel 1995) and is characterised by sudden brief attacks of vertigo associated with nystagmus. It normally begins between ages 1 and 4 years, and remits spontaneously within a few years. There are frequently transitions to other forms of migraine with and without aura (Lanzi et al. 1994). Basilar migraine also peaks in incidence during adolescence (Bickerstaff 1961). Another early manifestation of migraine is benign paroxysmal torticollis in infancy. It has recently been shown to be linked to the CACNA1A mutation (Giffin et al. 2002).

6.1.2 Familial Episodic Ataxia Types 1 and 2

The familial episodic ataxias are rare autosomal dominant diseases that occur in at least two well-defined groups: episodic ataxia type 1 without vertigo but with interictal myokymia (Brunt and van Weerden 1990) and episodic ataxia type 2 with vertigo, longer duration of the attacks and interictal nystagmus (Griggs and Nutt 1995). Like other autosomally dominant episodic diseases that react to acetazolamide (e.g., myotonias, dyskalemic periodic paralyses), episodic ataxia types 1 and 2 have been identified to be hereditary potassium (type 1) or calcium (type 2) channelopathies.

The drug therapy of choice for episodic ataxia type 1 is acetazolamide, administered in daily doses of 60–750 mg (Brunt and van Weerden 1990; Brandt and Strupp 1997). Phenytoin should be given to treat myokymia. Acetazolamide is most often effective in episodic ataxia type 2; the potassium channel blocker 4-aminopyridine was recently shown to also prevent these attacks (Strupp et al. 2004). Acetazolamide appears to be effective also as long-term therapy and may hinder the development of progressive ataxias (Griggs and Nutt 1995).

6.1.3 Motion Sickness

Before the age of 2 years, children are seldom affected by motion sickness. Later and until puberty, however, they exhibit a greater susceptibility to motion sickness when riding in a vehicle than do adults (see Section 6.4).

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6.2 *Drug-induced Vertigo*

Vertigo is frequently induced by medication, a fact that is generally underestimated (Table 6.3). The key to diagnosis of such cases is a carefully taken patient history. The temporal relationship between the onset of the respective drug treatment and the occurrence of symptoms is especially important (if suspected, try drug elimination). As the complaints and the clinical picture

Table 6.3. Drug groups that can cause vertigo as a side-effect

Nervous and musculoskeletal systems	**Heart, blood vessels, blood**
Anticonvulsants	Beta-receptor blockers
Analgesics	Anti-arrhythmics
Tranquillisers	Vasodilators/vasoconstrictors
Muscle relaxants	Anticoagulants
Hypnotics	**Kidneys and bladder**
Antiemetics	Diuretics
Antidepressants	Spasmolytics
Anticholinergics	**Respiratory organs**
Dopamine agonists, levodopa	Expectorants
Antiphlogistics	Antitussives
Local anaesthetics	Bronchospasm relaxants
Hormones	Mucus dissolvents
Corticosteroids	**Miscellaneous**
Antidiabetics	Antiallergics
Gonadal hormones	Prostaglandins
Oral contraceptives	X-ray contrast media
Inflammations	
Antibiotics	
Tuberculostatics	
Antihelmintics	
Antifungals	

differ greatly and the underlying mechanism of many medications that induce vertigo is unclear, there is still no satisfactory classification of drug-induced vertigo. On the one hand, certain drugs are known to have ototoxic effects, such as the aminoglycosides, which cause direct (practically selective) damage of the hair cells (see Section 2.5). On the other hand, drugs such as anticonvulsants (e.g., carbamazepine and diphenylhydantoin) cause pronounced central (dose-dependent) ocular motor disorders, although according to their principal mechanism, they affect all neurons of the central nervous system (overview in Rascol et al. 1995). Clinical neurological investigations frequently show that the latter mostly cause saccadic smooth pursuit and a gaze-holding deficit in all directions (see Section 1.3). Antihypertensives and diuretics are also relevant, as they can lead to orthostatic dysregulation and falls (overview in Tinetti 2002). Thus affected patients often experience a postural vertigo of short duration upon getting up. The diagnosis can be based on the Schellong test. Table 6.3 presents a selection of groups of drugs that elicit the undesired side-effect of vertigo.

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6.3 Cervicogenic Vertigo

Somatosensors in the muscles, joints and skin can induce sensations of self-motion and trigger nystagmus. Vision satisfactorily substitutes for sensory loss caused by disorders such as polyneuropathy or spinal cord diseases, and thus ensures spatial orientation and postural control; however, in darkness or under poor visual conditions, disturbed proprioception typically causes postural imbalance. There is, therefore, also a somatosensory vertigo.

The clinical picture of a cervicogenic vertigo, triggered only by a disorder of the neck afferents, is still controversial, although the important contribution of these receptors in spatial orientation, postural control and head–trunk coordination is well known.

The difficulty of clinical evaluation is based on:

- insufficient pathophysiological knowledge of function and multimodal interaction of the sensory signals from the neck afferents
- the existing conceptual confusion about so-called cervicogenic vertigo (Brandt and Bronstein 2001).

The neural connections between the neck receptors and the central vestibular system—the cervico-ocular reflex and the neck reflexes for postural control—have been investigated experimentally; however, so far they have remained clinically irrelevant. In humans a unilateral anaesthesia of the deep posterolateral neck region (e.g., C2 blockade for cervicogenic headache) causes a transient ataxia accompanied by ipsiversive deviation of gait and past-pointing without spontaneous nystagmus (Dieterich et al. 1993). It is difficult to apply these findings to patients with pain in the neck or back of the head, postural and gait imbalance, because the diagnosis cannot be confirmed at the present time. The recommended neck-turning test along with examination of the static cervico-ocular reflex or Romberg's test of stance while the patient leans his head backward are non-specific and insufficiently standardised (de Jong and Bles 1986). It was shown that during the neck-turning test, nystagmus was induced in patients no more often than in healthy controls (Holtmann et al. 1993). Optimistic and uncontrolled reports about the frequency of cervicogenic vertigo and the fantastic successes achieved by chiropractic treatment must be carefully evaluated.

Most of the controversial debate about the reality or fiction of cervicogenic vertigo resembles a "war between believers and doubters" that lacks any corresponding practical significance. As the cervical syndrome is treated by drugs or physical therapy, and once other causes are excluded, the hypothetical neurophysiological explanation mentioned above is still only of theoretical significance.

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6.4 Motion Sickness

6.4.1 Clinical Aspects and Pathogenesis

Acute motion sickness arises during passive transportation in motor vehicles; it resolves spontaneously within 1 day at the most after the disappearance of the inducing stimulus (Figure 6.1). The full picture of acute severe motion sickness develops

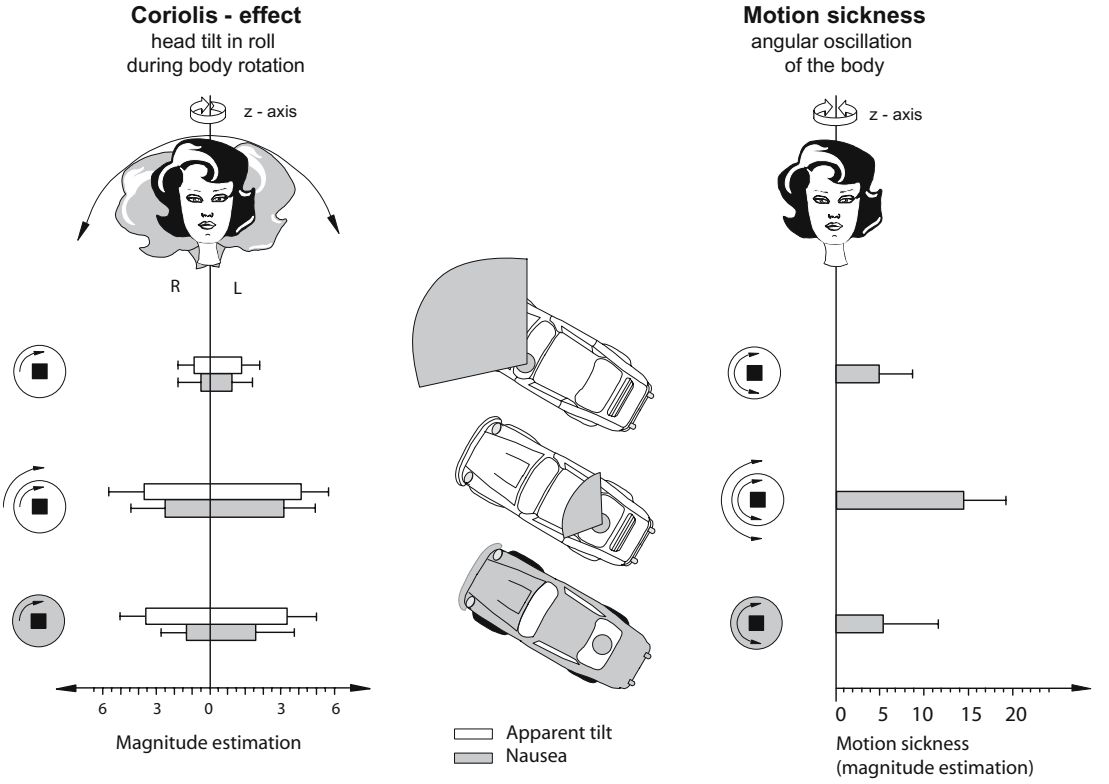

Figure 6.1. Visual influences during body acceleration in a combined drum and chair system and their effects on vertigo and motion sickness. *Left* Magnitude estimations of apparent tilt and nausea induced by Coriolis effects (sideward movement of the head during simultaneous chair and visual surround motion) at a constant angular velocity of 60°/s. *Right* Magnitude estimation of motion sickness induced by 15 min of sinusoidal angular oscillation of the body at 0.02 Hz and peak velocity of 100°/s. The three visual conditions were: eyes open (*top*), rotary chair and visual surroundings mechanically coupled (*middle*) and eyes open in complete darkness (*bottom*). Estimations of nausea indicate that the symptom is maximal with conflicting visual-vestibular stimuli (combined movement of chair and drum), when vestibular acceleration disagrees with the visual information of no movement (Brandt et al. 1976).

after initial symptoms of dizziness, physical discomfort, tiredness, periodic yawning and pallor, as well as slight vertigo with apparent surround-motion and self-motion. An increase in facial pallor is followed by cold sweats, increased salivation, hypersensitivity to smells, pains in the back of the head and feelings of pressure in the upper abdomen. Finally, the central symptoms of nausea, retching and vomiting develop with motor incoordination, loss of drive and concentration, apathy and fear of impending doom (Money 1970).

Motion sickness is not caused by vestibular "over-stimulation" during strong accelerations of the body, but by unfamiliar (i.e., non-adapted) motion stimuli and particularly by intersensory perceptual incongruences among the visual, vestibular and somatosensory systems. The most important concept explaining the pathogenesis of motion sickness is the so-called mismatch theory (Reason 1978; Dichgans and Brandt 1978). According to this theory, the decisive trigger is the incongruence of the signals of motion from various sensory channels or the incongruence between expected and actual sensory stimulation.

Well-known varieties of motion sickness are car sickness (visual-vestibular conflict of stimuli), seasickness (unfamiliar, complex linear and angular accelerations of low frequency, below 1 Hz), vehicle simulator sickness (optokinetic motion sickness) and space sickness (incongruent sensory stimuli of the otoliths, semicircular canals and the visual system during active head movements in microgravity).

6.4.2 Course and Therapy

Despite considerable interindividual variation in susceptibility, every healthy individual can experience motion sickness when exposed to extreme acceleration stimuli (e.g., cross-coupled acceleration like the Coriolis effect). Figures on the incidence of motion sickness in different motor vehicles vary between 1 and 90%. During the first days of an Atlantic crossing with moderate turbulence, about 25–30% of ship passengers experience motion sickness, whereas 80% of subjects on small life rafts or adrift with flotation vests become severely seasick. The latter's survival chances are reduced by the additional loss of water and electrolytes. Women are more susceptible than men, and children and young adults are more susceptible than the aged. Newborns and infants up to the age of 1 year are extremely resistant to motion sickness, apparently because they use the visual system for dynamic space orientation only once they have learned to stand alone and walk. Thus, they do not experience any optical-vestibular conflict of perception while being transported in a vehicle (Brandt et al. 1976). Loss of labyrinthine function causes complete resistance to motion sickness, whereas blindness does not.

Motion sickness is an acute clinical syndrome. Nausea and vomiting develop within minutes to hours, and the symptoms show spontaneous remission in hours to 1 day after the end of the stimulus. If the stimulus continues (ship or space travel), relief occurs via centrally mediated adaptation (habituation) within 3 days.

The mal-de-debarquement syndrome refers to a postural and gait unsteadiness with postural imbalance that develops on land after longer ship travel (Brown and Baloh 1987; Murphy 1993). Such complaints can occur at short notice in the form of sensorimotor after-effects even in healthy subjects following a persistent motion stimulus (e.g., seamen's legs). The syndrome lasts for months or years and recalls the development of a somatoform (psychogenic) vertigo similar to that of phobic postural vertigo (Chapter 5).

The most effective physical means of prevention is adaptation (habituation) by intermittent exposure to the stimulus. This adaptation, however, is only temporary and specific for each type of acceleration, i.e., resistance to seasickness does not protect from flight sickness.

If resistance by vestibular training is not indicated, the head should be kept still during the stimulus and additional accelerations that are complexly coupled with the vehicle motion should be avoided.

Motion sickness develops above all in closed vehicles or while reading on the back seat of a car, when the body is being accelerated but a stationary environment is being viewed, which contradicts the labyrinthine stimuli. By maintaining adequate visual control of the vehicle movement, one can significantly reduce motion sickness from that experienced under eyes-closed conditions. Conversely, susceptibility is significantly increased if primarily stationary contrasts fill the field of vision (Dichgans and Brandt 1973; Probst et al. 1982).

Antivertiginous drugs such as dimenhydrinate (Dramamine) or scopolamine (Transderm Scop) can inhibit the spontaneous activity of the neurons of the vestibular nuclei as well as the neuronal frequency modulation during body acceleration, thus reducing the susceptibility to motion sickness.

6.4.3 Pragmatic Therapy

The possibilities of physical and drug prevention are listed in Tables 6.4 and 6.5. Doubling the usual single doses (100 mg dimenhydrinate; 0.6 mg scopolamine) clearly increases the central sedating side-effects without causing any essential improvement in resistance to motion sickness (Wood et al. 1966). The effect of the individual substances can be intensified in severe cases by combining an antihistamine with a sympathicomimetic drug (25 mg promethazine and 25 mg amphetamine) (Wood and Graybiel 1970).

Table 6.4. Physical manoeuvres to prevent motion sickness

Measure	Goal
Before	
"Vestibular training" by repeated exposure to stimulus and active head movements or	Movement-specific central habituation
Vehicle simulator training	Utilisation of the visual-vestibular transfer of habituation
Acute	
Fixation of the head	Avoidance of additional accelerations, which are complexly coupled with vehicle acceleration (e.g., Coriolis effect)
Head position (toward the vector of gravitation) Ship: supine Car: supine with head in direction of movement Helicopter: sitting position	Utilisation of the head-axis specific difference in resistance to the acceleration; acceleration along the z-axis most favourable
Possible counter-regulation of the body motion induced by vehicle acceleration (e.g., inclining toward the curve)	
Visual control of the vehicle movement; if not possible, then close the eyes	Avoidance of a visual vestibular conflict of perception

Table 6.5. Drug prophylaxis for motion sickness

Drugs	Side-effects
Antihistamine 100 mg dimenhydrinate (Dramamine)	Sedation, reduced reactions and concentration, dryness of the mouth, blurred vision, numbness
Belladonna alkaloids 0.5 mg scopolamine as transdermal therapeutic system (Transderm Scop) 4–6 h before trip onset effective up to 72 hours	

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6.5 Height Vertigo

6.5.1 Syndromal Aspects and Pathogenesis

Physiological height vertigo is a visually induced destabilisation of posture and locomotion accompanied by individually varying amounts of strong anxiety and vegetative symptoms at the sight of ladders, buildings, a cliff or a mountain ridge.

Although height vertigo has long been considered a phobia, there is a physiological explanation for the postural instability and vertigo caused by optical stimuli at the sight of a free-standing building (Figure 6.2) (Bles et al. 1980; Brandt et al. 1980). Physiological height vertigo is instead a "distance vertigo" caused by visual destabilisation of upright postural balance, when the distance between the observer's eye and the nearest visible stationary contrasts within the field of vision becomes critically large. Head and body sway can then no longer be corrected by vision, as the movements are not registered by the sensors because of the sub-threshold smallness of the retinal slip. The vestibular and somatosensory signals of a shifting of the body's centre of gravity over the standing surface contradict the visual information of preserved body stability. Under such stimulus conditions, the body sway is increased, and above all the visual postural reflexes to the disturbing input are so impaired that there is real danger of an accident or fall. The critical stimulus parameters of the trigger and also practical pointers for prophylaxis can be derived from this physiological mechanism (Table 6.6).

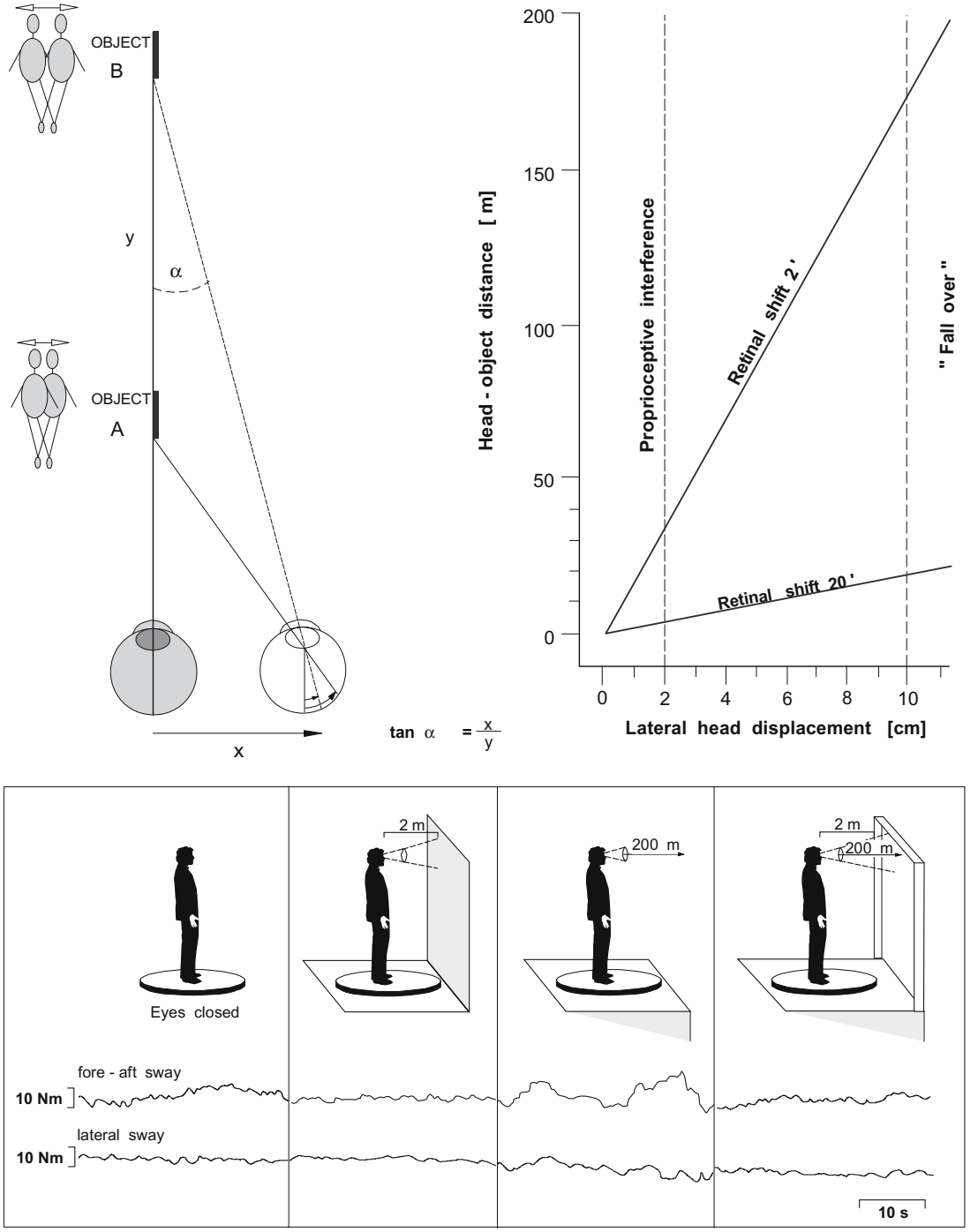

Figure 6.2. Mechanism of physiological height vertigo. Geometrical analysis indicates that, in order to be visually detected, body sway must increase with increasing distance between the eyes and the nearest stationary contrasts. Angular displacements α, on the retina, caused by a lateral head displacement, are smaller, the greater the distance y is to the object. The diagram shows the relationship between head–object distance, y, and lateral head displacement, x, for a given retinal displacement threshold of either 2 or 20 min of arc. However, because postural regulation involves the multiloop control, additional proprioceptive cues may alter sway amplitude as well. Fore–aft and lateral body sway (*original traces*) with eyes closed, eyes open in front of a wall, and eyes open on a high building with and without additional stationary contours in the peripheral visual field. Sway amplitudes increase under height vertigo conditions, especially in the low frequency range. Simultaneous, nearby stationary contrasts in the seen periphery stabilise "height vertigo sway" (from Brandt et al. 1980).

Table 6.6. Physiological height vertigo

Mechanism	Distance vertigo with visual destabilisation of body balance if the distance between eye and the nearest stationary objects exceeds 3 m (if physiological height vertigo induces a conditioned phobic reaction, then acrophobia develops)
Features	
Body posture:	Strongest when standing without support; weakest when lying; increases during extreme tilts of the head
Height:	Begins after ca. 3 m, maximal after ca. 20 m
Gradient:	Begins after 40–50°, maximal after 70–80° slope of the ground
Gaze direction:	Postural instability also occurs when gazing upward; decisive is the distance between eye and object
Prevention	
Body posture:	Improvement of posture stabilisation by leaning on something, holding tight to something or sitting down, particularly if there are additional disturbing stimuli such as wind; avoid extreme inclinations of the head in order to keep the otoliths in optimal operating position
Vision:	Look at near stationary contrasts; when looking into the abyss, keep near stationary objects in sight in the peripheral field of vision in order to maintain visual control of posture; avoid large-field motion stimuli that can lead to visually induced illusory motion; when in danger of falling, do not look through binoculars without some kind of support/ stabilisation

6.5.2 Course and Therapy

Many animal species as well as humans have to a large extent a genetically based fear and exhibit avoidance behaviour when visually approaching a step or an abyss (visual-cliff phenomenon; Walk et al. 1957). Height vertigo and height fear are accordingly physiological and must be differentiated from pathological acrophobia. Height vertigo develops within a matter of only seconds after a glance into an abyss, but once the inducing situation disappears, it rapidly resolves. Patients with disorders of labyrinthine function or balance and alcoholics are more susceptible. Repeated exposure to the stimulus can lead to a certain amount of adaptation.

Practical hints for reducing physiological height vertigo are given in Table 6.6. Acrophobia arises when physiological height

Table 6.7. Behavioural therapy for acrophobia

Systematic desensitisation	Presentation of graduated hierarchy of imagined anxiety-provoking visual scenes during muscle relaxation, which has been trained earlier
Environmental desensitisation	
—successive approximation:	Gradual approach to situations provoking anxiety with the help of instructions and reinforcement under conditions close to reality
—contact desensitisation:	The therapist subjects himself while in close physical contact with the patient to the same provoking situation and serves as a model
—flooding:	Direct confrontation as long as possible of the patient with the strongest provoking situations under real-life conditions

Drugs such as tranquillisers or antidepressants can also be used temporarily as support. Even without psychotherapy, most phobias improve spontaneously or resolve within a matter of years.

vertigo induces a conditioned phobic reaction characterised by a dissociation of the subjective and objective danger of falling. Although the acrophobic patient also recognises this discrepancy, he can typically and only with difficulty overcome the panic anxiety (Eckhardt-Henn et al. 2003; Furman and Jacob 1997; Jacob et al. 2004), vegetative symptoms and the inappropriate avoidance behaviour.

Psychotherapy for acrophobia and agoraphobia is dominated by behavioural therapy approaches, which can be classified as either systematic or environmental densensitisation (Table 6.7). The method of systematic desensitisation (Wolpe 1958) is based on the creation of a graduated hierarchy of visual scenes that cause anxiety and are then presented to the patients during a period of calm after they have "absolved a training phase of muscle relaxation". Desensitisation procedures, in which anxiety is supposed to be reduced during provoking situations that are close to life and not imagined, are, however, more effective. The stepwise approach (successive approximation) to the fear-inducing situation is supported by instructions and reinforcement. So-called contact densensitisation (Ritter 1969) stresses the advantages of the therapist's participation and physical nearness, serving as a model for the patient (participant modelling) during the graduated approach to situations triggering height vertigo. An alternative method is the confrontation by the patient of the strongest provoking situation for as long as possible, so-called flooding. Cumulative histories of anxiety neurosis patients with phobias show that even without psychotherapy, most childhood

phobias and 40–60% of adult phobias improve spontaneously or resolve after an interval of 5–6 years (Agras et al. 1972; Noyes et al. 1980, 1990; Keller et al. 1994).

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Index

Page numbers followed by f indicate figures; t, tables

List of Videos

Patient history

Benign paroxysmal positioning vertigo
Bilateral vestibulopathy
Cogan's syndrome
Familial episodic ataxia type 2
Superior semicircular canal (superior canal) dehiscence
Menière's disease
Vestibular neuritis
Phobic postural vertigo
Vestibular/basilar migraine
Vestibular paroxysmia
Vestibular paroxysmia (in childhood)

Neuro-ophthalmological and neuro-otological examinations

Unilateral cover test for exotropia (left eye)
Adducting saccades (slowing on both sides in cases of bilateral internuclear ophthalmoplegia)
Adducting saccades in cases of internuclear ophthalmoplegia (slowing on left)
Alternating cover test for esophoria (slight abducens palsy on the right side)
Benign paroxysmal positioning vertigo (left posterior canal)
Downbeat nystagmus (shown on scanning laser ophthalmoscope)
Fixation suppression of the vestibulo-ocular reflex (disturbed on left)
Superior semicircular canal dehiscence
Internuclear ophthalmoplegia (bilateral)
Congenital nystagmus
Convergence-retraction nystagmus
Positioning manoeuvre during benign paroxysmal positioning vertigo (canalolithiasis of the left posterior canal)