1.6 Inner Ear

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1.6.1 Zoster Oticus

SALVATORE IURATO AND WOLFGANG ARNOLD

1.6.1.1 Synonyms

Herpes zoster oticus, herpes zoster cephalicus, Ramsay Hunt syndrome.

1.6.1.2 Definition

Disease characterized by the rapid onset of peripheral facial paralysis associated with severe otalgia and ipsilateral varicelliform vesicles involving the auricle and the external auditory canal.

1.6.1.3 Aetiology/Epidemiology

Caused by reactivation of the varicella zoster virus at the level of the ganglion cells of cranial nerves VII and VIII, herpes zoster oticus accounts for approximately 10–15% of acute facial palsy cases.

1.6.1.4 Symptoms

- One or 2 days of general malaise with fever.
- Burning earache followed by a vesicular eruption involving the aperture of the external auditory canal and the auricle. The bluish-red vesicles eventually form crusts within a few days.
- Facial palsy.
- Sensorineural hearing loss and vertigo (lesion of cranial nerve VIII).
- Pain in the pharynx (lesion of cranial nerves IX and X).
- Facial pain due to involvement of cranial nerve V.
- Headache, neck stiffness, photophobia.

1.6.1.5 Complications

Persistent complete hearing loss, persistent vertigo, postherpetic pain, persistent facial paralysis.

1.6.1.6 Diagnostic Procedures

- Inspection: varicella zoster vesicles on the lateral surface of the auricle, concha and entrance of the auditory canal (Fig. 1.6.1). Vesicles may occur over the face and neck and may involve the buccal mucosa. **Warning**: the vesicles may be small or may resolve before the patient is evaluated (pay attention to encrusted areas).
- Otomicroscopy: vesicles or vesicle remnants in the external auditory canal and on the tympanic membrane.
- Audiometric evaluation: sensorineural hearing loss.
- Vestibular tests: spontaneous nystagmus towards the affected side (at the beginning), later towards the opposite direction.
- Serological confirmation: rarely utilized.

1.6.1.7 Therapy

Conservative Treatment

- Local treatment of the vesicles with drying agents (e. g. 70% alcohol) and acyclovir cream.
- Antiviral treatment with acyclovir (800 mg five times a day orally). Alternatively, brivudin or famciclovir may be given.
- Prednisolone, 100 mg/day for 3–5 days together with proton pumps blockers. Alternatively, prednisone, 1 mg/kg of body weight daily orally for 7–10 days with tapering to zero over the following 10 days.

Surgical Treatment

In cases of persistent facial paralysis, gold weight implant in the upper eyelid, hypoglossal-facial anastomosis.

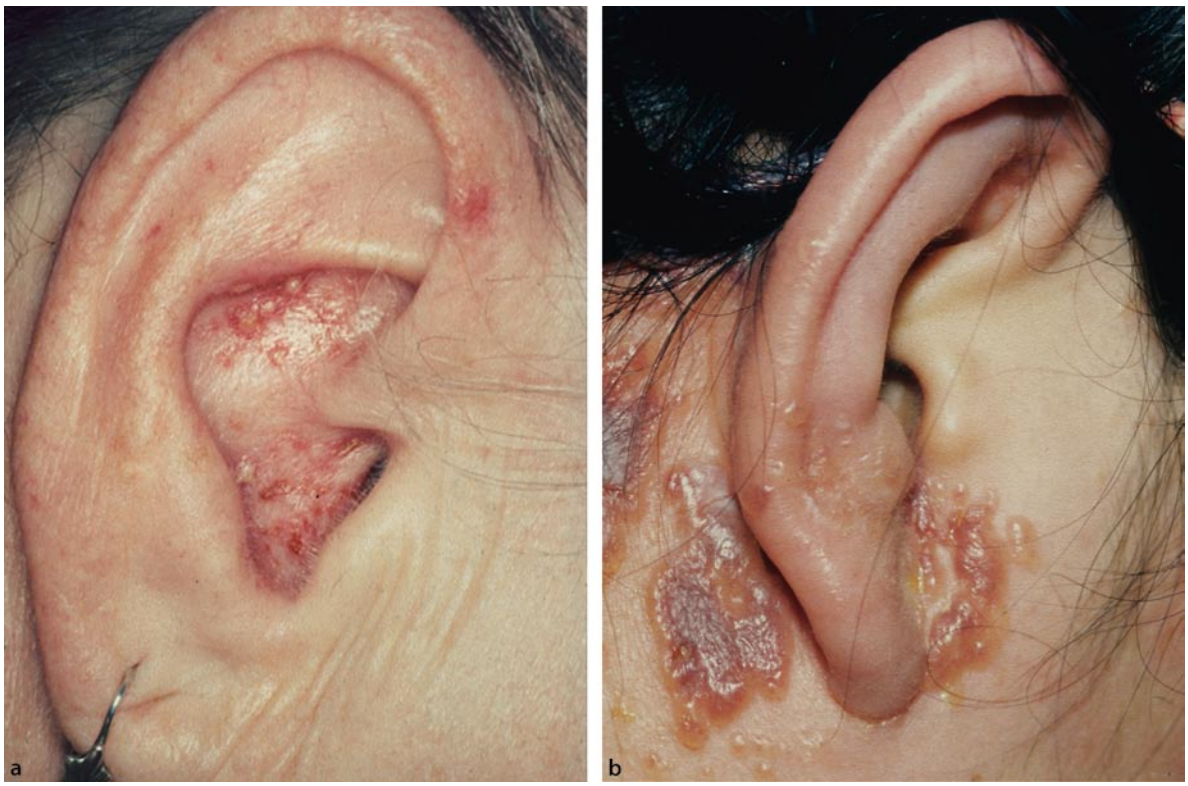

Fig. 1.6.1a,b Herpes zoster oticus. **a** Numerous vesicles (some of them crusted) are seen scattered throughout the concha. **b** Numerous vesicles in the concha, in front and behind the auricle

1.6.1.8 Differential Diagnosis

- Bell's palsy (idiopathic facial palsy): the characteristic cutaneous lesions and the high incidence of cochlear and vestibular symptoms differentiate zoster oticus from Bell's palsy.
- External otitis, perichondritis, erysipelas, neoplasm, labyrinthitis, Melkersson–Rosenthal syndrome.

1.6.1.9 Prognosis

- The prognosis for recovery of the facial function is less favourable than that of Bell's palsy. Incomplete recovery is frequent (only less than 50% of the patients recover satisfactorily). Prognosis is worse in elderly patients.
- Often persistent neuralgia (years).
- Complete hearing loss and complete vestibular areflexia are irreversible.

Suggested Reading

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2. Gross G, Doerr H-W (2006) Herpes zoster. Recent aspects of diagnosis and control. Karger, Basel
3. Schaitkin BM et al. (2000) Idiopathic (Bell's) palsy, herpes zoster cephalicus, and other facial nerve disorders of viral origin. In: May M, Schaitkin BM (eds) The facial nerve. Thieme, New York, pp 319–338

1.6.2 Labyrinthitis

WOLFGANG ARNOLD

1.6.2.1 Synonym

Acute or chronic inflammation of the labyrinth.

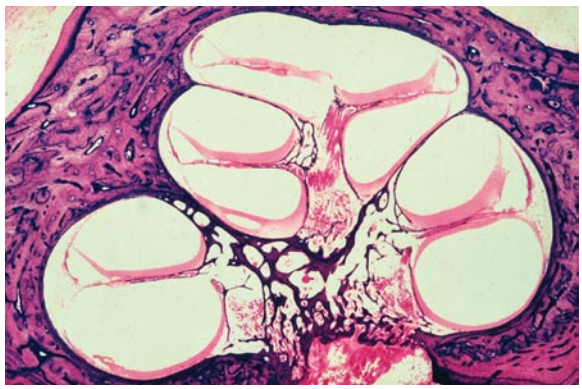

Fig. 1.6.2 Mild serous labyrinthitis in the course of lethal viral meningitis. Protein deposits can be seen in the perilymphatic spaces

1.6.2.2 Definition

Acute or chronic serous (Fig. 1.6.2) or purulent (Fig. 1.6.3) inflammatory reaction within the fluid spaces and membranes of the vestibulocochlear labyrinth caused by bacteria, viruses, spirochetes or fungi.

The route of infection may be *otogenic*, *meningogenic* or *haematogenic*.

1.6.2.3 Viral Labyrinthitis

The symptoms are:

- **Otogenic:** during the course of a viral infection of the upper respiratory tract including the middle ear (picornavirus, influenza virus, parainfluenza virus, respiratory syncytial virus, coronavirus, adenovirus). Serous effusion of the middle ear, vestibular disturbances, combined or pure sensorineural hearing loss, tinnitus. No earache.
- **Meningogenic:** during the course of mumps, measles or parainfluenza meningitis. Route of infection are the internal auditory canal and the cochlear aqueduct. Protein deposits can be seen in the perilymphatic spaces (Fig. 1.6.2). Clinical signs of meningitis are fatigue, vomiting, headache, stiff neck, fever, and unilateral or bilateral deafness.

1.6.2.4 Bacterial or Purulent Labyrinthitis

Aetiology/Epidemiology

Purulent (suppurative) bacterial labyrinthitis may be secondary to acute otitis media or purulent meningitis. In acute otitis media, bacteria may enter the inner ear through the oval and round windows (Fig. 1.6.3).

Purulent labyrinthitis is frequently followed by meningitis as the microorganisms gain access to the subarachnoid space through the cochlea aqueduct or internal auditory canal. Bacterial labyrinthitis can be a complication of cholesteatoma, spontaneous or acquired labyrinth fistula or may occur in malformations of the cochlea with enlarged perilymphatic spaces (Mondini dysplasia).

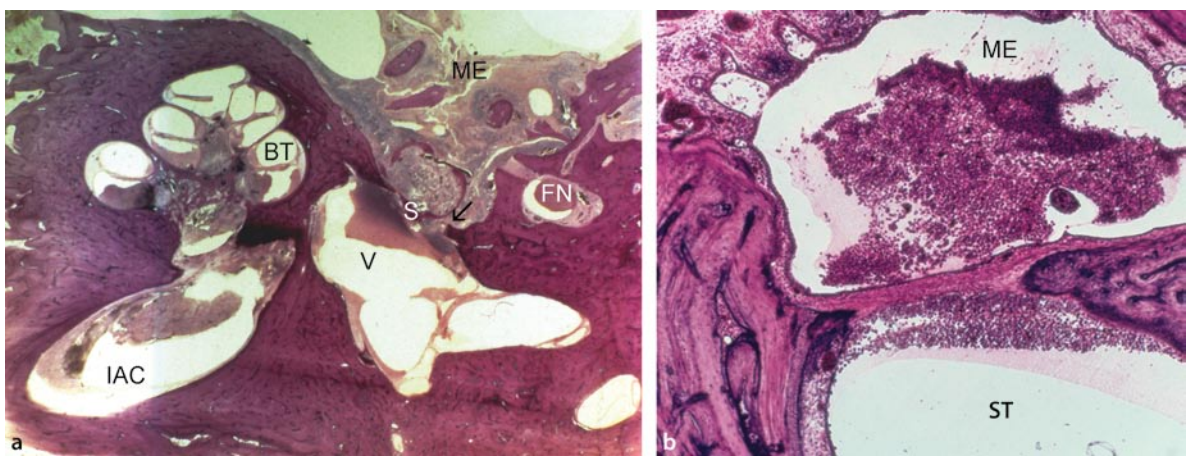

Fig. 1.6.3 **a** Purulent otogenic labyrinthitis and meningitis in a 4-year-old child, who died from the disease. The route of penetration is the oval window (*arrow*). **b** Acute otogenic labyrinthitis. The route of penetration is the thickened round window membrane. On both sides of the round window membrane purulent exudates can be seen. *IAC* internal auditory canal, *S* stapes, *FN* facial nerve, *ME* middle ear cavity, *BT* basal cochlear turn, *V* vestibule, *ST* scala tympani, basal turn

The most frequent bacteria causing otogenic labyrinthitis are pneumococci, *Haemophilus influenzae*, *Streptococcus* (A), *Escherichia coli* and *Klebsiella pneumoniae.*

The most frequent bacteria causing meningogenic labyrinthitis are meningococci, pneumococci and *Haemophilus influenzae* type B.

Symptoms

- **Otogenic labyrinthitis**: severe vertigo with nystagmus, vomiting, high fever. It invariably results in complete hearing loss and is often followed by facial paralysis.
- **Meningogenic labyrinthitis:** classic symptoms of meningitis, severe vertigo with nystagmus, vomiting, unilateral or bilateral, often fluctuating hearing loss or complete deafness. Postinflammatory rapid ossification of the cochlear fluid spaces.

Complications of Otogenic Bacterial Labyrinthitis

Meningitis, encephalitis, brain abscess, complete deafness, lethal outcome.

1.6.2.5 Haematogenic Labyrinthitis

- ***Treponema pallidum***: congenital or acquired syphilis (third stage). Fluctuating hearing loss, endolymphatic hydrops, dizziness. Hennebert sign positive. If untreated, it results in complete hearing loss.
- ***Mycobacterium tuberculosis***: formation of tuberculous granuloma along vascular spaces which spread into the labyrinth spaces. Progressive sensorineural hearing loss. In most cases it is associated with a systemic haematogenic spreading of tuberculosis.
- **Mucormycosis:** systemic, in many cases fatal mycotic sepsis (caused by, e.g., *Rhizopus*, *Absidia*) in immunodepressed patients (AIDS, leukaemia, diabetes): high temperature, meningitis, deafness, facial palsy, vertigo, purulent sinusitis.

1.6.2.6 Diagnostic Procedures

Recommended European Standard

- Otoscopy or ear microscopy: serous or purulent otitis media, pulsating tympanic membrane, cholesteatoma, bone fracture
- Hearing examination: tuning fork, audiogram (mixed or pure sensorineural hearing loss, deafness)
- Frenzel glasses: at the beginning nystagmus in the direction of the affected ear, later in the opposite direction
- High-resolution "emergency CT"

Microbiology

- Culture taken from the purulent ear secretion or from the nasopharynx
- Blood cell differentiation, blood sedimentation rate
- CSF diagnostic: cell count, protein and sugar elevation, culture taken from the CSF

Additional/Useful Diagnostic Procedures

- Serology: rubella virus, paramyxovirus, influenza virus, adenovirus, syphilis
- Bone scintigraphy (single photon emission scintigraphy)
- Counselling: neurology, neuropaediatrics, dermatology

1.6.2.7 Therapy

Conservative Treatment for Viral Labyrinthitis

- Glucocorticoids, initially 500 mg intravenously for 3 days followed by oral administration of prednisolone starting with 120 mg per day, then reducing the dosage by 10 mg per day
- Treatment of the rhinogenic infection (nasal spray, mucolytica)
- Vestibular suppressant medications and antiemetics
- Antibiotics intravenously to avoid bacterial superinfection (cephalosporine, aminopenicillin with or without a β-lactamase-inhibitor)

Conservative Treatment for Bacterial Labyrinthitis

- High doses of antibiotics, e.g. cephalosporines (third generation), chloramphenicole and aminoglycosides, according to the smear culture results
- Vestibular suppressant medications and antiemetics
- In lues III, penicillin G or tetracycline
- In tuberculosis, tuberculostatic therapy
- In mucormycosis, amphotericine B, control of diabetes
- Infusions with electrolytes; antipyretica

Surgical Therapy

Each otogenic labyrinthitis caused by bacteria needs surgical intervention.

Surgical Principles

- Myringotomy, insertion of ventilating tubes (Fig. 1.4.5)
- Mastoidectomy, radical ear surgery, labyrinthectomy

1.6.2.8 Prognosis

- Early onset of therapy (antibiotics, antimycotics) is obligatory in this life-threatening infection of the temporal bone.
- Deafness and long-lasting dizziness. Lethal outcome in most cases of generalized mucormycosis.
- In cases of bilateral complete deafness following bacterial meningitis, early cochlear implantation is recommended before ossification of the cochlea occurs (see Sect 1.6.12).

Suggested Reading

1. Arnold W, Bredberg G, Gstöttner W et al. (2002) Meningitis following cochlear implantation: pathomechanisms, clinical symptoms, conservative and surgical treatment. ORL J Otorhinolaryngol Relat Spec 64:382–289
2. Gulya AJ (1998) Infections of the labyrinth. In: Bailey BJ (1998) Head & neck surgery—otolaryngology, 2nd edn. Lippincott-Raven, chap 145
3. Schachern PA, Paparella MM, Hybertson R et al. (1922) Bacterial tympanogenic labyrinthitis, meningitis and sensorineural damage. Arch Otolaryngol Head Neck Surg 118:53–57
4. Schuknecht HF (1993) Infections of the ear. In: Pathology of the ear, 2nd edn. Lea and Febiger, Philadelphia

1.6.3 Contusio Labyrinthi

WOLFGANG ARNOLD

1.6.3.1 Synonym

Concussion of the labyrinth.

1.6.3.2 Definition

Microinjuries of the vestibulocochlear organ (bleeding, membrane ruptures, microfractures) caused by a blunt concussion of the skull, with or without fracture of the skull base or contusio cerebri.

1.6.3.3 Symptoms

Sensorineural hearing loss affecting all frequencies or mainly the frequencies above 3 kHz. In lateral (parietal) trauma, mainly the opposite ear is affected (contrecoup); when the blunt forces act from behind (occipital trauma) then both labyrinths can be damaged. Vertigo and tinnitus are accompanying symptoms.

1.6.3.4 Complications

- Luxation of the ossicles (e. g. incus), perilymphatic fistula, slowly progressive sensorineural hearing loss, deafness, long-lasting vertigo, subdural bleeding, posttraumatic endolymphatic hydrops.
- Postconcussion disequilibrium syndrome: pathomechanisms and symtoms are identical with those of benigne paroxysmal vertigo (see Sect. 1.6.15) and cupolithiasis [1].

1.6.3.5 Diagnostic Procedures

Recommended European Standard

- Detailed history (of forensic importance)
- Inspection of the skull, searching for skin injuries, haematoma
- Inspection of the oral, nasal and nasopharyngeal cavities
- Otoscopy or ear microscopy, searching for fracture signs, haemotympanum, rupture of the tympanic membrane
- Hearing examination: tuning forks, audiogram, tympanogram, stapedius reflexes
- Frenzel glasses: to exclude vestibular irritation or loss of function of one vestibular organ

Additional/Useful Diagnostic Procedures

- Schüller-X-ray, high-resolution CT: to exclude fractures
- Examination of the vestibular function (electronystagmography, video-oculography)
- Examination of the sense of smell to exclude rupture of the fila olfactoria

1.6.3.6 Therapy

Conservative Treatment

The recommended European standard is use of antioedematous principles (see Sect. 1.6.5).

Surgical Treatment

If there is an additional conductive hearing loss caused by trauma of the middle ear structures, reconstruction of the ossicular chain is recommended some weeks following the injury.

1.6.3.7 Prognosis

The prognosis is uncertain: in many cases complete restoration of the cochleovestibular deficit is possible; in some cases progressive hearing loss and/or long-lasting vertigo are possible.

References

1. Schuknecht HF, Davison RC (1956) Deafness and vertigo from head injury. Arch Otolaryngol 63:513–528

Suggested Reading

1. Schuknecht HF (1969) Mechanism of inner ear injury from blows to the head. Ann Otol Rhinol Laryngol 78:253–262
2. Von Schulthess G (1961) Innenohr und Trauma mit besonderer Berücksichtigung des Krankheitsverlaufes. Fortschr Hals-Nase-Ohrenheilk 7:1–102
3. Tuohimaa P (1978) Vestibular disturbances after acute mild head injury. Acta Otolaryngol Suppl 359:3–67

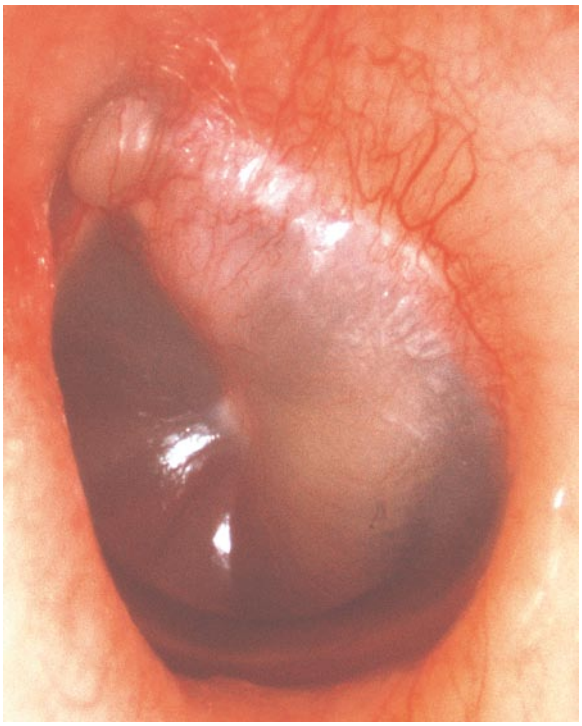

Fig. 1.6.4 This patient was operated on to remove a vestibular schwannoma with a leak along the posterior wall of the pharynx and a watery rhinorrhoea on bending her head forwards. The tympanic picture mimics a serotympanum

1.6.4 Otoliquorrhoea/Otorhinoliquorrhoea

SALVATORE IURATO

1.6.4.1 Definition

An abnormal communication between the subarachnoid space and the middle ear is a precondition:

- **Otoliquorrhoea**: outflow of CSF into the external auditory canal through a rupture of the tympanic membrane.
- **Otorhinoliquorrhoea**: if the tympanic membrane is intact, CSF drains from the middle ear into the nose through the Eustachian tube.

1.6.4.2 Aetiology/Epidemiology

- *Congenital* (*labyrinthine and perilabyrinthine abnormalities*): bone and meningeal defects in the tegmen tympani and tegmen antri areas, cochlear dysplasia, arachnoid granulations, defects in the Fallopian canals, enlarged and patent cochlear aqueduct (labyrinthine abnormalities), fissula ante fenestram, stapes malformations, cerebral herniations (meningoencephaloceles) in the middle ear and mastoid
- *Acquired* (more common): head trauma specially in cases of temporal bone fractures (see Sect. 1.5.3); in 29% of longitudinal fractures and in 44% of transverse fractures
- *Postoperative* CSF leak: e. g. complication of vestibular schwannoma surgery, middle and posterior fossa surgery, surgery for chronic ear disease (less frequently)

1.6.4.3 Symptoms

A watery pulsating secretion when there is a tympanic membrane perforation or rupture. When the tympanic membrane is intact, there is clear fluid behind the tympanic membrane simulating a serous otitis media (Fig. 1.6.4). Outflow of watery fluid from the nose (otorhinoliquorrhoea). Coughing at night. A sensation of salty fluid in the mouth.

1.6.4.4 Complications

Recurrent meningitis, meningoencephalitis.

1.6.4.5 Diagnostic Procedures

- Clinical history
- Otoscopy/ear microscopy (Waterlike fluid behind the tympanic membrane? Clear secretion through a perforation?)
- Inspection of the oral cavity (Any leak along the posterior wall of the pharynx?)
- Queckenstedt's sign (a rise in the CSF pressure compressing the veins in the neck)
- Hearing examination: tuning fork, audiogram, tympanometry
- High-resolution CT scan (axial and coronal scan)
- MRI to prove herniated brain tissue within the temporal bone
- Laboratory tests: glucose content examination (50% of blood sugar), protein content (maximum 2 g/l), β_2 transferrin
- In some cases, intrathecal fluoroscein (attention to complications such as seizures and transverse myelitis with paralysis!), CSF scintigraphy

1.6.4.6 Therapy

Conservative Treatment

- Rest in bed, with the head elevated. Avoid straining at stool.
- Never occlude the external auditory canal!
- Diuretics (acetazolamide).
- Antibiotic treatment to prevent ascending infections: cephalosporins, ciprofloxacin.
- Continuous lumbar CSF drainage (attention to the sterility and the volume and rate of flow!) if the above-mentioned measures do not work.

In vestibular schwannoma surgery careful sealing during surgical closure and a compressive bandage are necessary to prevent postauricular CSF leak.

Surgical Treatment

Surgical treatment is necessary when CSF fistulas do not heal spontaneously. The best approach for spontaneous and post-traumatic leaks is through the middle fossa (Fig. 1.5.9). For CSF fistulas following vestibular schwannoma surgery, revision of the surgical wound and insertion of plugs of periostium and abdominal fat should be performed.

1.6.4.7 Differential Diagnosis

- Serotympanum (Fig. 1.4.6a)
- Monolateral serous rhinitis
- Rhinorrhoea

Suggested Reading

1. Hoffman RA (1994) Cerebrospinal fluid leak of temporal bone origin. In: Jackler RK, Brackmann DE (eds) Neurotology. Mosby, St Louis, pp 919–928

1.6.5 Sudden Sensorineural Hearing Loss (Including Perilymphatic Fistula)

KERSTIN LAMM

1.6.5.1 Synonyms

Sudden deafness, sudden idiopathic sensorineural hearing loss.

1.6.5.2 Definition

Sudden sensorineural hearing loss is characterized by an *acute* in the majority of cases *unilateral hearing loss originating in the inner ear* of *unknown pathogenesis and origin*. Hearing loss may be slight, moderate, profound or complete and concerns the high, middle, deep or all frequencies. The disease is accompanied by tinnitus in about 85% of cases and dizziness in about 30% of cases.

1.6.5.3 Epidemiology

The incidence ranges between 20 cases per 100,000 residents per year in Austria and Germany, eight to 13 cases per 100,000 residents per year in Japan and about 11 cases per 100,000 residents per year in the USA.

Women are equally affected as men, most frequently at the age of 50±5 years. However, there is an increasing incidence in younger people, whereas children are rarely concerned.

1.6.5.4 Aetiology/Classification

The cause is still unknown. The following pathomechanisms are currently discussed:

- Impairment of cochlear blood flow due to vasomotor paralysis or vasospasm, endothelial oedema or other endothelial disorders resulting in microcirculatory rheological disturbances
- Impairment of cochlear blood flow due to systemic rheological disorders
- Malfunction of ion channels in hair cells resulting in cellular dysfunction
- Synaptic dysfunction due to insufficiency or toxicity of afferent neurotransmitters

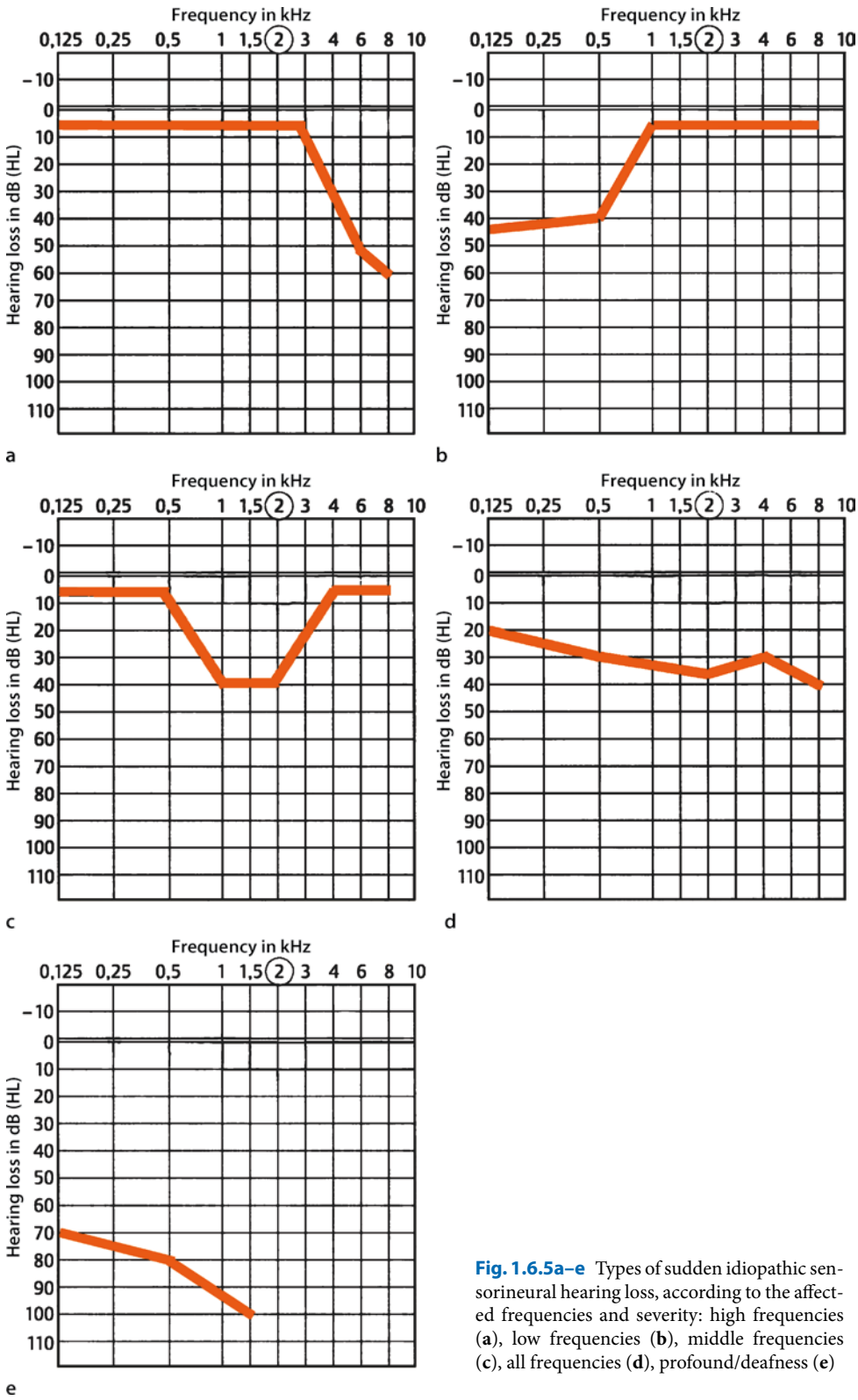

Fig. 1.6.5a–e Types of sudden idiopathic sensorineural hearing loss, according to the affected frequencies and severity: high frequencies (**a**), low frequencies (**b**), middle frequencies (**c**), all frequencies (**d**), profound/deafness (**e**)

- Malfunction of the cochlear efferent pathways
- Malfunction of ion channels in strial cells resulting in endolymphatic water–electrolyte imbalance, potentially even in endolymphatic hydrops
- Inflammatory tissue alterations, e. g. within the endolymphatic sac
- Alteration of other biochemical and physiological mechanisms involved in cochlear homeostasis

According to the affected frequencies and severity the following types of sudden idiopathic sensorineural hearing loss can be distinguished (Fig. 1.6.5):

- **High-frequency idiopathic sensorineural hearing loss**, which may be due to cellular malfunction of outer (below 50 dB HL) and inner (beyond 60 dB HL) hair cells (Fig. 1.6.5a).
- **Low-frequency idiopathic sensorineural hearing loss**, which may be due to endolymphatic water–electrolyte imbalance or even endolymphatic hydrops as a possible result of impaired blood flow within the vascular stria and subsequent hypoxic cellular damage (Fig. 1.6.5b).
- **Middle-frequency idiopathic sensorineural hearing loss**, which may be due to impaired blood flow within the spiral lamina and subsequent hypoxic cellular damage of the organ of Corti or, alternatively may be due to genetic defects (Fig. 1.6.5c).
- **All-frequency idiopathic sensorineural hearing loss**, which may be due to blood flow impairment in the spiral modiolar artery and/or upstream arteries resulting in hypoxic damage of cochlear tissues (Fig. 1.6.5d).
- **Profound idiopathic sensorineural hearing loss and deafness**, which may be due to thrombotic or embolic blood flow obstruction in the arteria cochlearis communis or spiral modiolar artery resulting in hypoxic damage of cochlear tissues. Perilymphatic fistula due to rupture of the round window membrane or lesion of the oval window which may result in acute profound hearing loss or even deafness is a disorder of a distinct origin and, therefore, cannot be referred to as idiopathic sensorineural hearing loss by definition (Fig. 1.6.5e).
- **Other types**, such as fluctuating hearing thresholds, progressive hearing loss in spite of current therapy etc.

1.6.5.5 Symptoms

Patients complain about symptoms in the following descending order of incidence:

- *Acute unilateral subjective hearing loss*, although this symptom may be not perceived in cases of slight hearing loss restricted to a few frequencies
- *Tinnitus* (in about 85% of cases)
- *Feeling of pressure in the ear*
- *Dizziness* (in about 30% of cases)
- *Distorted hearing*, *diplacusis*, *hyperacusis*
- *Periaural dysaesthesia*
- *Secondary psychoemotional ailments* such as anxiety, fear, distress, restlessness, agitation, resignation and sense of guilt

1.6.5.6 Complications

Complications such as fluctuating hearing threshold or progressive hearing loss in spite of current therapeutic intervention raise the question of a distinct cause such as beginning Ménière's disease, pressure variations of CSF, vestibular schwannoma and (auto)immune-mediated disease.

Another complication may concern psychoemotional and psychosocial problems in cases of persistent hearing loss and/or tinnitus (see Sect. 1.6.8).

1.6.5.7 Diagnosis

Recommended European Standard

1. Detailed case history
   - Side, onset and duration of hearing loss
   - Associated symptoms such as tinnitus, dizziness, feeling of pressure in the ear, periaural dysaesthesia, secondary psychoemotional ailments
   - Previous idiopathic sudden sensorineural hearing loss or previous hearing loss due to noise exposure or other diseases, previous pure tone audiograms
   - History of otorhinolaryngological diseases and surgery
   - History of head trauma and/or other accidents
   - History of internal, neurological, psychiatric or psychosomatic or orthopaedic diseases
   - Family status, profession, still working, claim for early retirement, retired
   - Recreational activities
2. Microscopy of the ear canal and ear drum
3. Rhinoscopy using 0°/30° endoscopes
4. Posterior rhinoscopy using a 70° endoscope
5. Sonography of the maxillary and frontal paranasal sinus
6. Pharyngoscopy
7. Hypopharyngoscopy and laryngoscopy using a 90° endoscope
8. Tympanometry and stapedial reflex measurements (ipsilateral and contralateral evoked reflex responses at 0.5, 1, 2 and 4 kHz)
9. Pure tone audiogram (0.125, 0.25, 0.5, 0.75, 1, 1.5, 2, 3, 4, 6 and 8 kHz)

10. Transitory evoked otoacoustic emissions
11. Vestibular tests using video-oculography
12. Auditory evoked brainstem responses (ABRs) or cerebral MRI in the case of equivocal ABR measurement results or pathological ABRs and/or video-oculography findings and/or in the case of a previous or suspected intracranial disease
13. Blood pressure measurement

Useful Additional Diagnostic Procedures

- Audiometric tinnitus measurement (frequency-matching, intensity, minimal masking level using broadband or narrowband noise or pure tones, residual inhibition) (Fig. 1.6.7)
- Speech audiometry
- Distortion products of otoacoustic emissions
- Auditory evoked cortical responses
- Electrocochleography
- Glycerol test according to Klockhoff
- Tympanoscopy in cases of profound hearing loss or deafness
- Blood cell count, haemoglobin, fibrinogen, lipids, creatinine, C-reactive protein
- Serologic tests on borrelia, lues, herpesvirus type 1, herpes zoster virus, HIV
- Doppler duplex sonography of extracranial vessels
- Depending on case history internal, neurological, psychosomatic, orthopaedic, genetic examination

1.6.5.8 Therapy

Sudden idiopathic sensorineural hearing loss must be managed with care and as soon as possible concerning the diagnostic procedures and the mode and beginning of therapy. According to the categories of the Oxford Centre for Evidence-Based Medicine (http://www.cebm.net), the evidence level of most clinical trials on therapy of sudden deafness is relatively low. In addition, the clinical trials considered in the Cochrane International Register of Controlled Clinical Trials, e. g. prospective, randomized, placebo-controlled and double-blind conducted studies, are of relatively high level of evidence; however, the underlying study protocols vary tremendously. Therefore, universally valid therapeutic options based on reproducible results are not available. Instead, therapeutic recommendations are merely formed empirically.

Spontaneous recovery rates range from 31 to 68%; however, these data were revealed from retrospective, non-randomized and mostly non-controlled trials on a few patients only. Furthermore, the term "recovery" was not exactly defined, e. g. partial remission was not distinguished from complete remission, and a persistent tinnitus was not taken into consideration. Patients would not regard themselves as fully recovered when their tinnitus is still persistent.

However, in well-informed patients presenting with a slight hearing loss without previous ipsilateral or contralateral hearing loss and without tinnitus and/or dizziness, one may await spontaneous recovery for a few days.

Conservative Therapy

Basic therapeutic interventions comprise normalization of systemic blood pressure, heart rate and haematocrit level (below 45). For patients complaining of moderate to severe psychoemotional ailments such as anxiety, fear, distress, restlessness, agitation, resignation and sense of guilt, a psychotherapist should be included.

Recommended European Standard

According to the aforementioned cause and pathogenesis possibly involved, therapeutic recommendations differentiate between the various types of sudden idiopathic sensorineural hearing loss.

- **High-frequency idiopathic sensorineural hearing loss.** A daily dose of 250–500 mg *prednisolone* intravenously on three consecutive days is recommended.
  In the case of partial or no remission, prednisolone treatment should be continued orally for 16 days, starting with 100, 80, 60, 40, 20, 10, 5 and 2.5 mg each dosage for two days.
  Prednisolone therapy should be accompanied by gastric proton pump inhibitors (40 mg omeprazole per day or 150 mg ranitidine per day or others).
  Prednisolone is a synthetic analogue of endogenous corticosteroid hormones classified as glucocorticoids. Besides anti-inflammatory effects, prednisolone possesses multiple other cellular effects. The rationale for administration of prednisolone is based on the consideration that inflammatory tissue alterations are also elicited by tissue ischaemia and hypoxia. In addition, prednisolone mobilizes amino acids for gluconeogenesis, alters glucose utilization and influences protein metabolism. Finally, prednisolone binds with equal affinity to both glucocorticoid and mineralocorticoid receptors widely distributed in cochlear tissues, thereby contributing to restoration of cellular osmolarity, electrochemical gradients, transmembrane ion flux and neuronal conduction.
  In patients presenting with *contraindications* (e. g. diabetes, severe chronic gastritis, gastric or duodenal ulcera) *for the treatment with prednisolone*, intravenous infusion of a hyperoncotic hydrophilic haemodilutive plasma-expanding agent, such as *hydroxyethyl starch or others* (250–500 ml/day for 5–10 days) is a good alternative option.

However, contraindications (e. g. arterial hypertension) should be noted, and potential side effects (e. g. temporary pruritus) should be considered. *In the case of contraindications for haemodilution*, another haemorheological active drug such as *pentoxifylline* (100 mg, equivalent to 5 ml/day dissolved in 100 ml 0.9% isotonic saline) may be intravenously infused for 5–10 days.

Additional administration of scavangers of toxic free oxygen radicals such *α-lipoic acid* (600 mg/day orally) during haemodilutive or haemorheological infusion therapy may be reasonable to prevent reperfusion injury within the cochlea.

- **Low-frequency and middle-frequency idiopathic sensorineural hearing loss**. A daily dose of 250–500 mg *prednisolone* intravenously on three consecutive days is recommended. In the case of partial or no remission, prednisolone treatment should be continued orally for 16 days together with gastric proton pump inhibitors as described above. Additional *osmotic infusion therapy* using *mannitol* (15 g in 100 ml solution) and *acetazolamide* (500 mg) intravenously on three consecutive days may be administered. Acetazolamide (250 mg/day) may be continued orally for 10 days.

  In patients presenting with *contraindications* (e. g. diabetes, severe chronic gastritis, gastric or duodenal ulcer) *for the treatment with prednisolone,* initial osmotic therapy is recommended.
- **All-frequency idiopathic sensorineural hearing loss**. A daily dose of 250–500 mg *prednisolone* intravenously on three consecutive days is recommended. In the case of partial or no remission, prednisolone treatment should be continued orally for 16 days together with gastric proton pump inhibitors and additional *haemodilutive/haemorheological infusion therapy* together with *α-lipoic acid* should be administered as described for high-frequency idiopathic sensorineural hearing loss.

  In patients suffering from an elevated fibrinogen level (above 300 mg/dl) *fibrinogen and low density lipoprotein apheresis* may be a good alternative to haemodilutive or haemorheological infusion therapy. However, the expense is relatively high and the long-term outcome on hearing gain was proved to be equally effective as with conventional therapy as described above.
- **Profound idiopathic sensorineural hearing loss and deafness**. A daily dose of 500 mg *prednisolone* intravenously on three consecutive days together with *haemodilutive/haemorheological infusion therapy* and *α-lipoic acid* should be administered as described for high-frequency idiopathic sensorineural hearing loss. Prednisolone treatment should be continued orally for 16 days together with gastric proton pump inhibitors as described, and haemodilutive/haemorheological infusion therapy and α-lipoic acid should be continued for another seven consecutive days.

  In patients suffering from an elevated fibrinogen level (above 300 mg/dl) *fibrinogen and low density lipoprotein apheresis* may be additionally tried (but see the comments for all-frequency idiopathic sensorineural hearing loss).

  In the case of minor or no remission at all, perilymphatic fistula due to rupture of the round window membrane or lesion of the oval window may be considered and *tympanoscopy* should be performed preferably within 5–10 days after onset of hearing loss.

Useful Additional Therapeutic Strategies

If initial therapy with prednisolone and/or haemodilutive or alternative haemorheological agents is only partially or not effective at all, and this is the case in over 25% of patients, *hyperbaric oxygenation therapy* (ten to 15 sessions on ten to 15 consecutive days) should be started as soon as possible, preferably within 3–6 weeks after the onset of idiopathic sensorineural hearing loss.

Obsolete Therapeutic Options

- Isobaric oxygenation therapy, e. g. breathing of oxygen under normal atmospheric pressure
- Ozone therapy
- Ultraviolet light irradiation
- Ultrasonic therapy
- Laser irradiation of any type or mode
- Electromagnetic stimulation
- Suggestive psychotherapy, hypnosis
- Acupuncture
- Autohaemotherapy, autologous blood transfusion
- Monotherapy with vasodilative agents which may induce intracochlear vascular steal effects resulting in an impaired cochlear blood flow

Surgery

In the case of progressive hearing loss in spite of current therapeutic intervention or in the case of minor or no remission of profound hearing loss, *tympanoscopy* should be performed to exclude or seal a rupture of the round window membrane or a lesion of the oval window (see Sect. 1.6.5.9).

1.6.5.9 Differential Diagnosis

Sudden sensorineural hearing loss may also be attributable to the following diseases:

- **Perilymphatic fistula**. Perilymphatic fistulae of the oval or round window are rare clinical findings. They

may occur during skull trauma, following stapes surgery, middle ear surgery, after lifting heavy weights or spontaneously.

Perilymphatic fistulae in the area of the annular ligament of the stapes footplate cause sensorineural hearing loss and dizziness of different extent. They can occur after stapes surgery with insufficient sealing of the prosthesis. Congenital perilymphatic fistulae of the stapes footplate occur in cases of malformation of the stapes and cause recurrent meningitis. They are usually detected during exploration of the oval window.

Perilymphatic fistulae of the round window membrane are rare findings in cases of sudden hearing loss. There are no clear clinical symptoms characteristic for round window membrane rupture. In some cases the history reveals strong physical exertion (explosion trauma after Goodhill).

All diagnosed perilymphatic fistulae should be completely sealed with connective tissue and fibrin glue to avoid labyrinthitis and/or meningitis. Surgical sealing of the ruptured round window membrane or annular ligament with connective tissue sometimes results in partial or complete, but not predictable, restoration of hearing.

- Haematological diseases (e. g. polycythaemia, polyglobulia, leukaemia, exsicosis).
- Cardiovascular diseases.
- Bacterial labyrinthitis (due to otitis media, borrelia, lues). See Sect. 1.6.2.
- Meningitis.
- Virus infection (adenovirus, herpesvirus type 1, herpes zoster virus, mumps virus, HIV).
- Encephalitis disseminata (multiple sclerosis).
- Autoimmune vasculitis (e. g. Cogan's syndrome).
- Intoxication (drugs, industrial pollution).
- Renal failure, dialysis.
- Intracerebral tumours (e. g. vestibular schwannoma).
- Barotrauma or decompression trauma of the inner ear. See Sect. 1.5.1.
- Acute noise induced hearing loss, acoustic trauma. See Sect. 1.6.9.3.
- Head trauma with labyrinthine contusion. See Sect. 1.6.3.
- Severe distortion trauma of the cervical spine.
- Pressure variations of CSF (e. g. after CSF puncture).
- Cochlear malformation.
- Genetic defects, hereditary sensorineural hearing loss.
- Genetic syndrome (e. g. Usher, Pendred).
- Psychogenic hearing loss.

1.6.5.10 Prognosis

The prognosis is most favourable in low-frequency and middle-frequency idiopathic sensorineural hearing loss without previous hearing loss. However, recurrence rates of such hearing losses range up to 30%.

Patients with slight to moderate threshold shifts recover better than patients presenting with moderate to profound hearing loss.

A relatively worse prognosis is expected in profound hearing loss or even sudden deafness.

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1.6.6 Ménière's Disease

SALVATORE IURATO AND WOLFGANG ARNOLD

1.6.6.1 Synonyms

Ménière's syndrome, idiopathic endolymphatic hydrops, morbus Ménière.

1.6.6.2 Definition

The syndrome is characterized by: recurrent spontenous attacks of vertigo, fluctuating hearing loss (at the early beginning mainly affecting low frequencies) tinnitus and aural fullness.

According to the American Academy of Otolaryngology—Head and Neck Surgery at least two attacks of objective rotational vertigo, each of 20-min duration or longer must occur to confirm the diagnosis of Ménière's disease. Unilateral sensorineural hearing loss

must be documented audiometrically on at least one occasion.

The diagnosis "Ménière's disease" always is a diagnosis "per exclusionem" since other diseases of the cochleovestibular system can mimic Ménière's symptoms.

1.6.6.3 Aetiology/Epidemiology

Temporal bone histopathology from Ménière's patients are usually showing dilatations (hydrops), distortions and/or ruptures (Fig. 1.6.6) of the delicate membranes separating the endolymphatic from the perilymphatic fluid compartements. Hydrops can develop simultaneously as well in the cochlear as in the vestibular compartments, but also isolated in only one of the compartments (this may explain why in Ménière the symptoms can only consist of attacks of vertigo or hearing loss with tinnitus). A malfunctioning spiral ligament and /or endolymphatic sac (disturbed resorption of the endolymph, immunologic factors causing "saccitis") seem to be involved in the pathophysiology of endolymphatic hydrops. Recently the homing of Herpes Type I viruses has been demonstrated as well within the endolymphatic sac as in ganglion cells of Scarpa's ganglion. Reactivation of these viruses seems to be triggered by (immunologic) stress factors clinically causing the typical symptoms of the disease.

There are many known causes of endolymphatic hydrops: viral labyrinthitis, autoimmune inner ear disease (e.g. Cogan's syndrome), otosclerosis, leukaemia, otosyphilis, surgical trauma to the inner ear, temporal bone trauma, etc. When the cause cannot be identified, the term "Ménière's disease" is used. The incidence is one in 8,000 individuals per year in Europe. Approximately 30% of patients will develop contralateral involvement over time (25% at 10-year follow-up).

1.6.6.4 Symptoms

Classic symptoms are:

- Recurrent attacks of rotational vertigo (the most disabling symptom for the patients) which last from several minutes to hours. Vertigo is often associated with nausea and vomiting.
- Fluctuating, unilateral low-frequency hearing loss of the sensorineural type (upsloping audiometric pattern). In the late stages of the disease, there is flat or downsloping non-fluctuating sensorineural hearing loss (hearing threshold of 50–60 dB, speech discrimination of 50–60%).
- During the attacks, subjective tinnitus and aural fullness/pressure are present. Between attacks patients are not dizzy but aural fullness and tinnitus may persist. Vertigo attacks can vary in frequency, intensity and duration. Generally the attacks increase in severity and frequency with progression of the disease. The frequency and intensity of the vertigo attacks decrease after approximately 5–10 years.

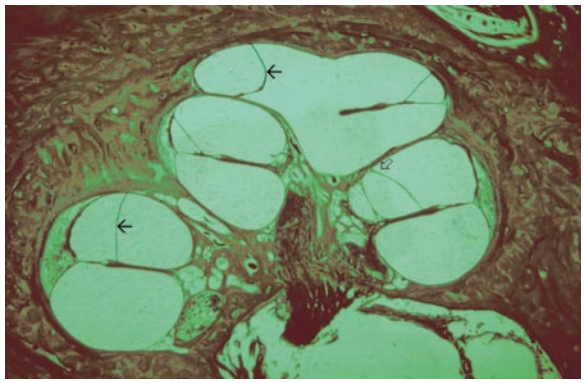

Fig. 1.6.6 Severe endolymphatic hydrops (←) with distension of Reissner's membrane, ruptures and adhesions (⇦) in a patient, age 58 who suffered from bilateral Menière's disease

1.6.6.5 Diagnostic Procedures

- Clinical history: extremely important.
- Physical examination: normal findings. Between the attacks patients usually display horizontal, spontaneous nystagmus beating away from the affected ear (paretic nystagmus).
- Electrocochleography: a significant enhancement of the summating potential to action potential amplitude ratio (more than 30%) occurs in 60% of patients with Ménière's disease.
- Auditory brainstem response.
- MRI with contrast medium plays an important role in excluding a retrocochlear lesion (vestibular schwannoma) in *any patient* with unilateral neurotologic symptoms!

1.6.6.6 Additional/Useful Diagnostic Procedures

- Blood sedimentation rate, antinuclear antibody test for autoimmune ear disorders (see Sect. 1.6.7); fluorescent treponemal antibody absorption test to rule out syphilis.
- Glucose tolerance and thyroid function tests are recommended as hypothyroidism and diabetes may be associated with the disease. Lipid profile.
- Caloric tests (with electronystagmography): reduced caloric response on the affected side.

- Glycerol test (1.2–1.5 ml/kg body weight of glycerol mixed with lemon juice): frequently used in the past, but much less nowadays because of its side effects (headache, nausea, vomiting).

Note: *the diagnosis is often by exclusion.*

1.6.6.7 Differential Diagnosis

- Vestibular schwannoma
- Sudden sensorineural hearing loss
- Autoimmune hearing loss
- Migraine

1.6.6.8 Therapy

Recommended European Standard

Conservative Treatment

1. In acute attacks bed rest, vestibular suppressant medication (diazepam) and antiemetics (transdermal scopolamine, thiethylperazine, prochlorperazine) are recommended
2. To prevent attacks, decrease the amount of fluid in the inner ear by diet and diuretics
   - Diet: low salt intake (less than 3 g per day) and reduced water intake
   - Diuretics: acetazolamide, chlortalidone, hydrochlorothiazide, furosemide. Replace potassium if needed
3. In addition
   - Vasoactive drugs (betahistidine) to improve blood flow in the inner ear
   - Steroids to suppress inflammatory and/or allergic tissue reactions within the inner ear (endolymphatic sac)
4. Avoidance of caffeine, alcohol, tobacco
5. Antiviral approach [1]: oral acyclovir (800 mg 3 times a day) for 3 weeks; if the patient feels better, reduce the dose to 800 mg 2 times a day for 1 month and then to 800 mg daily for another month before ending the treatment.

Semiconservative Treatment

- Insertion of a *transtympanic ventilating tube* (Montandon) (Fig. 1.4.5)
- Transtympanic unilateral *chemical labyrinthectomy with gentamicin*: good results on vertigo control, low to moderate risk of hearing deterioration
- For bilateral cases, intramuscular *streptomycin*
- Following insertion of a transtympanic ventilating tube (Fig. 1.4.5), self-administered treatment with the *Meniett device* (intermittent low-pressure pulses to stimulate the flow of the endolymph)
- Intratympanic dexamethasone injections (0.4–1.0 ml)
- Intratympanic application of ganciclovir 50 mg/ml per 10 days through a ventilating tube or a microwick inserted into the tympanic membrane [2].

Surgical Treatment

- Endolymphatic sac decompression: Sham operation or first-line procedure? Control of vertigo attacks in approximately 70% of patients.
- Vestibular neurectomy (middle fossa or retrosigmoid approach). This procedure has a 98% success rate in relieving vertigo, which is the most distressing symptom, but does not improve hearing.
- Labyrinthectomy, which implies the complete loss of hearing; high success rate in eliminating major vertigo attacks.
- Vestibulotomy: high success rate (control of vertigo 90%, reduction of tinnitus loudness 60%, risk of hearing loss).
- Tenotomy (section of the tensor tympani and stapedius muscle tendons): a new entry.

Note: *Ménière's disease does not have a currently known cause and this may explain the variety of therapeutic options and some geographical differences.*

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1.6.7 Autoimmune Inner Ear Disease

ROBERTO BOVO AND ALESSANDRO MARTINI

1.6.7.1 Synonyms

Autoimmune hearing loss, immune-mediated inner ear disease.

1.6.7.2 Aetiology/Epidemiology

- An underlying genetic predisposition results in autoimmune disease expression following immunoregulatory defects in immune response to unknown environmental pathogens.
- Today there is substantial evidence of autoimmune mechanisms in relapsing polychondritis (Fig. 1.3.2c), cochlear vasculitis (e. g. Cogan's syndrome), progressive sensorineural hearing loss of both sides and some types of sudden deafness.
- With regards to Ménière's disease, around 16% of bilateral cases and 6% of monolateral cases may be due to immune dysfunction.
- Autoimmune inner ear disease represents less than 1% of all cases of hearing impairment or dizziness; nevertheless, the diagnosis might be overlooked because of the lack of a specific diagnostic test. The disease seems to be more common in females than in males; the first onset of symptoms generally occurs between 20 and 50 years of age.
- Sympathetic cochleopathy: sudden sensorineural hearing loss in the last hearing ear (similar to sympathetic ophthalmopathy).

1.6.7.3 Symptoms

- Hearing loss: a rapidly progressive, often fluctuating, *bilateral* sensorineural hearing loss over a period of weeks to months. The progression of hearing loss is too rapid to be diagnosed as idiopathic progressive sensorineural *hearing loss* or presbyacusis and too slow to conclude a diagnosis of sudden sensorineural hearing loss.
- Tinnitus: 25–50% of patients also have tinnitus (ringing, hissing, roaring) and aural fullness, which can fluctuate.
- Vertigo and/or imbalance: generalized imbalance, ataxia, motion intolerance, positional vertigo and episodic vertigo may be present in up to 50% of patients.
- Occasionally only one ear is affected initially, but bilateral hearing loss occurs in most patients (80%), with symmetric or asymmetric audiometric thresholds.
- Systemic autoimmune disease coexists in up to 20% of patients (systemic lupus erythematosus, rheumatoid arthritis, disseminated vasculitis, Sjögren's syndrome, myasthenia gravis, Hashimoto's thyroiditis, Cogan's syndrome, Behçet's disease, sarcoidosis, Wegener's granulomatosis, colitis ulcerosa, relapsing polychondritis; Fig. 1.3.2c).

1.6.7.4 Diagnostic Procedure

- Currently, the diagnosis of autoimmune inner ear disease is based either on clinical criteria or on a positive response to steroids. There is seldom convincing evidence from broader laboratory tests indicating autoimmunity.
- Detailed history: Endocrine diseases? Recurrent fever?

Physical Examination

Otoscopy findings are usually normal; nevertheless external ear skin and/or cartilage inflammation and/or facial palsy may rarely occur (e. g. relapsing polychondritis), as well as tissue destruction at the level of the tympanic membrane, middle ear and mastoid (e. g. Wegener's granulomatosis).

Laboratory Studies

There are no antigen-specific tests (migration inhibition test, lymphocyte transformation test and western blot analysis) that are commercially available and proven to be useful for the diagnosis of systemic autoimmune diseases.

In clinical practice next to the indispensable blood sedimentation rate (BSR) a non-specific antigen screening test may be useful for evidence of systemic immunologic dysfunction; yet it does not strictly correlate with a diagnosis of immune-mediated inner ear disease.

Recommended tests are:

- Blood tests for autoimmune disorders: levels of circulating immune complexes, BSR, antinuclear antibodies, rheumatoid factor, complement C1Q, smooth muscle antibody, TSH and antimicrosomal antibodies, antigliadin antibodies (for celiac disease), HLA testing.
- Blood tests for conditions that resemble autoimmune disorders: fluorescent treponemal antibody absorption test (for syphilis), Lyme titre, HbA1c (for diabetes, which is often also autoimmune-mediated), HIV (HIV is associated with auditory neuropathy).

Note that a commercially available test, called "anti-68 kD (hsp-70) western blot" (OTOblot™) was reported to de-

tect a local autoimmune inner ear process in the absence of any systemic autoimmune process and to be correlated with steroid responsiveness. The test uses purified hsp-70 kDa antigen from a bovine kidney cell line and is based on the assumption that the 68-kDa protein is heat shock protein 70 (hsp-70). Unfortunately, this assumption has now been refuted: in fact, there is mounting evidence that the target antigen of the 68-kDa antibody is not hsp-70 (as was believed over the last 15 years), but the human choline transporter-like protein 2 (CTL2). Furthermore, the sensitivity and specificity of this test are very low.

1.6.7.5 Differential Diagnosis

- Bilateral Ménière's disease
- Luetic inner ear disease
- Lyme disease
- Toxoplasmosis
- Treatment with ototoxic drugs (gentamicin, cisplatin)
- Charcot–Marie–Tooth disease (hereditary neuropathy)
- Large vestibular aqueduct syndrome
- Endocranic hypertension

1.6.7.6 Therapy

- Prednisolone, 1 mg/kg per day for 4 weeks followed by a gradual tapering over several weeks to a maintenance dose of 10–20 mg per day or every other day. Shorter-term or lower-dose long-term therapy either has been ineffective or appears to increase the risk of relapse. Patients often learn the necessary maintenance dose to preserve their hearing, as the disease activity often waxes and vanishes. If hearing suddenly worsens or tinnitus reappears in one or both ears during the tapering period, repetition of the initial high-dose treatment is indicated.
- Oral as well as systemic steroid treatment over long period of time should always be accompanied by proton pump inhibitors to avoid a gastric ulcer.
- In patients with no response to steroids within 6–8 weeks, methotrexate and cyclophosphamide have been used over the long term. These agents are associated with considerable toxicity and the decision regarding when and how to use them should always be multidisciplinary. The normal oral dose of methotrexate is 7.5–20 mg weekly with folic acid. Cyclophosphamide in addition to steroids has been used with the following regimen: cyclophosphamide 5mg/kg per day intravenously for 2 weeks, followed by a rest period of 2 weeks, and then a final period of infusions for 2 weeks.

1.6.7.7 Prognosis

Autoimmune inner ear disease is analogous to rapidly progressive glomerulonephritis. If not treated, the inner ear inflammation progresses to severe irreversible damage within 3 months of onset (and often much more quickly). On the other hand, steroid responsiveness is high and with prompt treatment the hearing loss may be reversible. Nevertheless, several patients become steroid-dependent.

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1.6.8 Tinnitus

KERSTIN LAMM

1.6.8.1 Synonym

Ringing in the ears (from Latin *tinnire* meaning "ringing").

1.6.8.2 Definition/Symptoms

It is important to distinguish between objective and subjective tinnitus:

- Patients affected by an **objective tinnitus** notice a *real existent endogenous acoustic source* originating in the middle ear, Eustachian tube, soft palate or extracranial or intracranial vessels. Such acoustic phenomena may also be perceived by the non-affected fellow human being using a stethoscope; however, the incidence of this kind of tinnitus is relatively seldom. Depending on the underlying disease, the patients notice intermittent clicks or crackles due to spasm of the middle

ear muscles or myoclonus of soft palate muscles, respiratory noise and breath, respectively, due to an abnormally wide Eustachian tube, or a pulsatile noise due to intracranial hypertension, glomus tumour, angioma, aneurysm, arteriovenous fistula, stenosis or thrombosis of extracranial or intracranial vessels as well as due to systemic rheological diseases such as hyperglobulinaemia or anaemia.

- In contrast, a **subjective tinnitus** is exclusively perceived by the affected patient. In most cases it consists in an intermittent or continuous whistling or fizzling, in broadband or narrowband noise, hum, ping or ringing, or even in pure tones of various frequency, intensity and duration. Such auditory perceptions emerge from deficient neuronal plasticity within the central auditory system triggered by an auditory input failure. In this respect, a subjective tinnitus is *a symptom of any disease of the peripheral and/or central auditory system* associated with malfunction of hearing. In this particular context it should be emphasized that *tinnitus is not* a symptom of an organic or functional disease of the cervical spine, temporomandibular joint or any other orthopaedic or dental distress; likewise tinnitus is not caused by emotional, mental or physical distress, although the intensity and annoyance of tinnitus may be amplified by such problems.

1.6.8.3 Epidemiology

There are no epidemiological data available concerning the incidence and prevalence of an **objective tinnitus.**

A transient **subjective tinnitus** is perceived by about 35–45% of the population of industrial nations at least once in their life; 13–17% have perceived tinnitus over a longer period in their life, with an annual incidence of 0.33% in Germany.

The point-prevalence of a constant chronic tinnitus (perceived as longer than 6 months up to years) averages about 4% of the population of industrial nations, of which about 0.5–1% regard themselves as severely psychoemotionally affected.

Of those patients who perceive tinnitus over a longer period in their life, 35–37% *notice the tinnitus in a silent environment only (grade I)*; 44–51% perceive their tinnitus permanently, however it *may be masked by moderate environmental noise (grade II)*. In only 14–17% of cases is tinnitus *perceived even in a relatively loud environment (grade III)*.

According to a recent evaluation of about 12,000 members of the German Tinnitus Support Group (Deutsche Tinnitus-Liga, http://www.tinnitus-liga.de) who perceived tinnitus for more than 6 months up to years, 45% localized their tinnitus in both ears, while a further 24% localized their tinnitus perception in the middle of the head. Of the unilaterally affected, 29% perceived tinnitus in the left ear and 20% in the right ear.

The high incidence of bilateral tinnitus averaging about 46% and the slight preference of the left ear is also known from former evaluations among non-selected populations.

The prevalence of tinnitus is only somewhat higher (by 1–5%) in females than in males.

Respecting the age distribution, manifestation of tinnitus is most common between 40 and 60 years of age; however, there is an increasing incidence of noise-induced tinnitus in younger people owing to exposure to leisure noise such as noisy toys, amplified music, motorcycling and other loud recreational activities. Data from long-term studies concerning the incidence and prevalence of a chronic tinnitus (for more than 6 months up to years) in this population are not available so far.

1.6.8.4 Aetiology

Concerning an **objective tinnitus**, there are no data available regarding the incidence of the underlying diseases, as mentioned already in Sect. 1.6.8.2.

The **subjective tinnitus** is a symptom of any diseases of the peripheral and/or central auditory system associated with malfunction of hearing in the following descending order of incidence: in **32%** tinnitus is being caused by **noise-induced damage of the inner ear** due to single or repetitive exposure to industrial or leisure noise; in **12%** by **acute acoustic trauma of the inner ear** due to single or repetitive sound impulses from pistols, revolvers, military rifles, sport guns, firecrackers, fireworks and others; in **8–10%** by **idiopathic sensorineural hearing loss** (sudden deafness); in **8%** by **Ménière's disease** (morbus Ménière); in **7%** by **age-related sensorineural hearing loss** (presbyacusis); in **6%** by **toxic labyrinthitis** due to an acute serous or purulent otitis media; in **4%** by **chronic otitis media** inclusive of cholesteatoma; in **2–3%** by **otosclerosis**; and in **1%** by a **vestibular schwannoma** (acoustic neurinoma).

In the **remaining 20–23%** tinnitus may be caused by obliteration of the outer ear canal with wax, exostosis or others; myringitis, rupture or perforation of the ear drum; dysfunction of the Eustachian tube due to acute or chronic infections of the upper respiratory tract; barotrauma of the middle ear; tympanosclerosis; luxation of the incudomalleal or incudostapedial articulation; rupture of the round window membrane; perilymphatic fistula of the round or oval window; labyrinthine contusion or fracture of the temporal bone due to head trauma; meningitis or encephalitis; ototoxic medication (aminoglycosides, cisplatin, etc.); intoxication by alcohol or drugs; peridural

anaesthesia; lumbar puncture; multiple sclerosis; neurofibromatosis; neurolues; polyneuropathia; varicella zoster infection; systemic viral infections (rubella, measles, mumps, etc.); borreliosis; inner ear hearing loss associated with various syndromes (Alport, Addison, Cogan, Usher, etc.) or other autoimmune diseases (sympathic cochleopathia similar to sympathic ophtalmopathy, periarteritis nodosa, granulomatosis Wegener, lupus erythematodes, etc.); inner ear hearing loss associated with diabetes, arterial hypertension, renal failure etc.; and congenital inner ear hearing loss.

In summary, *there are almost 100 diseases of the peripheral and/or central auditory system which may cause tinnitus*. In this respect, an accurate otoneurological diagnostic procedure is of prime importance.

1.6.8.5 Diagnostic Procedures

Recommended European Standard

1. Detailed case history in the style of standardized tinnitus questionnaires, e. g. according to Goebel and Hiller (1998)
   - Type/character of tinnitus: unilateral, bilateral, in the head; intermittent or continuous; frequency; masking level by environmental noise—grade I–III (see Sect. 1.6.8.3); fluctuant or constant loudness
   - Visual analogue scale (range from 1 to 10) concerning loudness and annoyance
   - Onset and duration
   - Potential causal relationship and trigger mechanisms (exposure to noise of any kind, etc.)
   - Associated hearing problems, hyperacusis or phonophobia
   - Vestibular complaints
   - History of otorhinolaryngological diseases and surgery
   - History of head trauma and/or other accidents, actions under civil law
   - History of internal, neurological, psychiatric or psychosomatic or orthopaedic diseases
   - Alleviating or amplifying circumstances
   - Tinnitus-associated and/or other complaints (sleep disturbance, concentration and attention problems, psychoemotional and psychosocial problems, etc.)
   - Family status
   - Profession, still working, claim for early retirement, retired
   - Recreational activities
2. Microscopy of the ear canal and ear drum
3. Compression–decompression test using the Politzer balloon
4. Tympanometry and stapedial reflex measurements (ipsilateral and contralateral evoked reflex responses at 0.5, 1, 2 and 4 kHz)

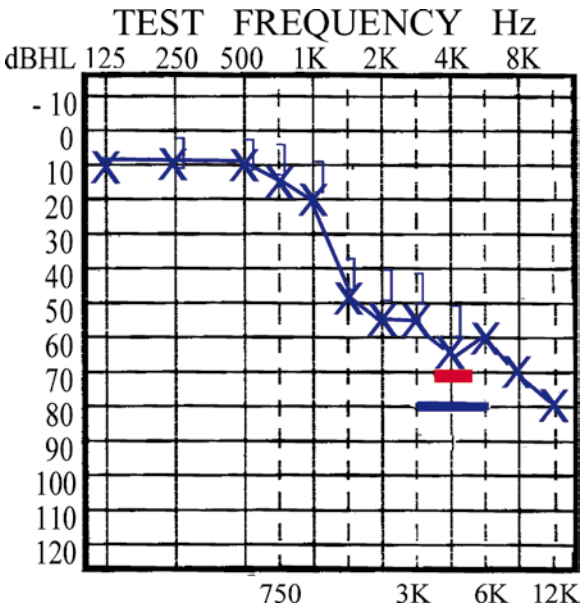

Fig. 1.6.7 Tinnitus analysis: the patient described his tinnitus as similar to a 4-kHz tone with an intensity of 70 dB. A broadband 80-dB noise masked the tinnitus which was heard again by the patient after 1 min, which means that a residual inhibition was not present. *Red line* tinnitus, *blue line* broadband noise

5. Pure tone audiogram (0.125, 0.25, 0.5, 0.75, 1, 1.5, 2, 3, 4, 6 and 8 kHz)
6. Audiometric tinnitus measurement [frequency-matching, intensity (Fig. 1.6.7), minimal masking level using broadband or narrowband noise or pure tones, residual inhibition]
7. Auditory evoked brainstem responses

Useful Additional Diagnostic Procedures

- Transitory evoked otoacoustic emissions
- Distortion products of otoacoustic emissions
- Auditory evoked cortical responses
- Speech audiometry
- Vestibular tests using video-oculography
- Electrocochleography
- Cerebral MRI in the case of pathological auditory evoked brainstem responses and/or video-oculography findings and/or in the case of a suspected intracranial disease
- CT scan of the temporal bone in the case of chronic otitis media, mastoiditis, cholesteatoma, head trauma, etc.
- Doppler duplex sonography of extracranial and intracranial vessels, if necessary angio-MRI
- Rhinoscopy using 0°/30° endoscopes

- Posterior rhinoscopy using a 70° endoscope
- Sonography of the maxillary and frontal paranasal sinuses
- Pharyngoscopy
- Hypopharyngoscopy and laryngoscopy using a 90° endoscope
- Exploratory vestibular tests using Frenzel glasses
- Depending on the case history, internal, neurological, psychosomatic or orthopaedic examination

1.6.8.6 Therapy

Objective Tinnitus

Treatment is directed to the underlying disease:

- In the case of spasms of the middle ear muscles, myringotomy and insertion of a grommet or transection of the tensor tympanic muscle or stapedial tendon may be helpful.
- In the case of a myoclonus of soft palate muscles, injection of botulinum toxin achieves relatively good results.
- If the patient perceives respiratory noise and breath owing to an abnormally wide Eustachian tube, an attempt to treat the tinnitus by application of an inert ointment onto the ear drum or myringotomy and insertion of a grommet may be worthwhile.
- A pulsatile tinnitus due to intracranial hypertension may require neurosurgery in selected cases. A pulsatile tinnitus due to angioma, aneurysm, arteriovenous fistula, stenosis, thrombosis of extracranial or intracranial vessels as well as due to systemic rheological diseases such as hyperglobulinaemia or anaemia should be treated by the angiologist and vascular surgeon, respectively.
- For surgical and/or radiation therapy of a glomus tumour, see Sect. 1.5.4.3.

Subjective Tinnitus

Subjective tinnitus cannot be treated directly. In fact, treatment is targeted to the underlying disease of the peripheral and/or central auditory system to achieve elimination of auditory input failure, thereby correcting pathological neuronal plasticity within the central auditory system. It is important to explain these issues to the patient in the acute stage, e. g. **first tinnitus counselling**.

1. **Acute stage (less than 3 months' duration)**. Acute tinnitus due to noise-induced damage or acoustic trauma of the inner ear, idiopathic sensorineural hearing loss (sudden deafness), acute attack of Ménière's disease, toxic labyrinthitis, rupture of the round window, perilymphatic fistula of the round or oval window, labyrinthine contusion or fractures of the temporal bone due to head trauma should be treated with a daily dose of 250–500 mg **prednisolone** intravenously on three consecutive days. In the case of partial or no remission, prednisolone treatment should be continued orally for 16 days, starting with 100, 80, 60, 40, 20, 10, 5 and 2.5 mg each on two consecutive days. Prednisolone therapy should be accompanied by gastric proton pump inhibitors (40 mg omeprazole per day or 150 mg ranitidine per day or others).
   In patients presenting with contraindications (e. g. diabetes, severe chronic gastritis, gastric or duodenal ulcer) for the treatment with corticosteroids classified as glucocorticoids such as prednisolone, infusion therapy as described below may be a good alternative option.
   In cases of severe hearing loss, *additional* intravenous infusion therapy using a hyperosmotic hydrophilic haemodilutive plasma-expanding agent, such as *hydroxyethyl starch* or others (250–500 ml/day for 5–10 days), would be reasonable to improve microcirculation. However, contraindications (e. g. arterial hypertension) should be noted, and potential side effects (e. g. temporary pruritus) should be considered. *In the case of contraindications for haemodilution*, another haemorheological active drug such as *pentoxiphylline* (100 mg, equivalent to 5 ml/day dissolved in 100 ml 0.9% isotonic saline) may be intravenously infused for 5–10 days.
   If this initial therapy with prednisolone and/or haemodilutive or alternative haemorheological agents is only partially effective or not effective at all, and this is the case in over 25% of patients, *hyperbaric oxygenation therapy* should be started as soon as possible (ten to 15 sessions on ten to 15 consecutive days). However, according to clinical trials therapeutic results on tinnitus are only available from patients suffering from acoustic trauma, noise-induced hearing loss and idiopathic sensory-neural hearing loss (sudden deafness); therefore, hyperbaric oxygenation therapy should be restricted to these three indications.
   For additional treatment of tinnitus due to an acute attack of Ménière's disease, toxic labyrinthitis, rupture of the round window, perilymphatic fistula of the round or oval window, labyrinthine contusion or fractures of the temporal bone due to head trauma, see the specific sections.
   Likewise, for treatment of all other diseases of the peripheral and/or central auditory system which may cause a subjective tinnitus, such as chronic otitis media, cholesteatoma, mastoiditis, otosclerosis and vestibular schwannoma (acoustic neurinoma), see the specific sections.
2. **Subacute stage (duration of more than 3 months up to 1 year) and chronic stage (duration of more than 1 year)**. In both, the subacute and the chronic stage a

second tinnitus counselling should be performed as follows:

- Tinnitus is an auditory phantom perception which emerges from deficient neuronal plasticity within the central auditory system triggered by an auditory input failure due to any disease of the peripheral and/or central auditory system.
- It is ensured that all aforementioned initial therapies and/or other causal treatment of the underlying disease of the peripheral and/or central auditory system have failed to ameliorate or eliminate tinnitus at the latest by the end of the subacute stage.
- The patient should be briefed that *the following* pharmaceuticals, natural remedies and other therapeutic strategies have been *proven to have no persistent effect on subacute and/or chronic tinnitus in placebo-controlled clinical trials:* intratympanal application of glucocorticoids, lidocaine, glutamate-receptor agonists or antagonists; systemic administration of antiarrhythmics such as lidocaine, anticonvulsive drugs such as lamotrigine or carbamazepine, antidepressives such as trimipramine, nortriptyline or amitriptyline, benzodiazepines such as diazepam or alprazolam, vasodilative drugs such as pentoxifylline, cyclandelate, prostaglandin E1, analogues of histamine such as betahistine, antagonists of histamine receptors such as cinnarizine, calcium antagonists such as flunarizine or trimetazidine, GABA agonists such as baclofen, antiphlogistic drugs such as azapropazone, diuretics such as dyazide, melatonin, zinc, vitamins, *Ginkgo biloba*, laser therapy of any kind with or without *Ginkgo biloba*, acupuncture of any kind, ultrasonic therapy and electromagnetic stimulation of any kind.
- Therefore, in the chronic stage correction of pathological neuronal plasticity within the central auditory system cannot be achieved directly by pharmaceuticals, natural remedies or other treatment strategies already mentioned, but can be achieved indirectly by compensation of the remaining hearing loss using hearing aids as soon as possible. However, in cases where hearing loss is less than 30 dB HL at three or fewer frequencies (2 kHz included), a broadband noise generator (formerly tinnitus masking device) may be helpful to defocus on tinnitus.
- In addition, active listening to music of someone's own choice and/or to audiobooks are reasonable therapeutic auditory training strategies which may be helpful to direct the patient's attention to external auditory stimuli again.
- Patients presenting with tinnitus-associated and/or other complaints, such as sleep disturbance, concentration and attention problems, psychoemotional and psychosocial problems, should be admitted to a cognitive tinnitus coping therapy including a multimodal behavioural treatment in a specialized psychotherapeutic–psychosomatic outpatient department, or in selected cases to a psychotherapeutic–psychosomatic inpatient clinic.
- A promising novel therapeutic innovation is neurofeedback training, which has been shown to effectively relieve stress-associated symptoms and thereby annoyance of tinnitus.

1.6.8.7 Prognosis

The outcome is dependent on the quality of the initial and the following medical attendance, e. g.:

- Briefing of the patient that tinnitus perceptions emerge from deficient neuronal plasticity within the central auditory system triggered by an auditory input failure.
- An accurate case history and otoneurological examination to diagnose the disease of the peripheral and/or central auditory system which has caused the tinnitus.
- Briefing of the patient that pharmaceutical and/or surgical treatment can be only targeted on the tinnitus-underlying disease of the peripheral and/or central auditory system to achieve elimination or at least reduction of the auditory input failure and malfunction of hearing, respectively.
- Including technical devices if necessary.
- Including psychotherapeutic–psychosomatic treatment strategies if necessary.

Suggested Reading

1. Bartels H, Staal MJ, Albers FW (2007) Tinnitus and neural plasticity of the brain. Otol Neurotol 28:178–184
2. Dohrmann K, Elbert T, Schlee W, Weisz N (2007) Tuning the tinnitus percept by modification of synchronous brain activity. Restor Neurol Neurosci 25:371–378
3. Dohrmann K, Weisz N, Schlee W, Hartmann T, Elbert T (2007) Neurofeedback for treating tinnitus. Prog Brain Res 166:473–485
4. Møller AR (2006) Neural plasticity in tinnitus. Prog Brain Res 157:365–372
5. Schenk S, Lamm K, Gündel H, Ladwig KH (2005) Neurofeedback-based EEG alpha and EEG beta training. Effectiveness in patients with chronically decompensated tinnitus. HNO 53:29–37

1.6.9 Noise-induced Hearing Loss

UWE GANZER AND ANDREAS ARNOLD

The disorder is defined as hearing loss caused by acute or chronic exposure to high-intensity sound.

1.6.9.1 Explosions

Definition

Impulse sound exposure with an intensity above 150 dB(A) SPL peak equivalent with a peak sound pressure duration greater than 3 ms.

Aetiology/Epidemiology

Explosion traumas are seen in explosives fabrication and the processing industry, in the military, in the gas, tyre and chemical industry as well as in motor vehicle accidents. Additionally, they can occur from a physical injury to the ear, such as a blow to the side of the head, diving head first into water or deployment of an airbag.

Symptoms/Findings

An explosion trauma of the ear generally leads to inner ear damage concomitant with a rupture of the eardrum and, most often, a disruption of the ossicular chain (Fig. 1.4.1b). This leads to a unilateral or bilateral, moderate to severe combined (conductive and sensorineural) hearing loss with otalgia, tinnitus and, frequently, vestibular symptoms. At the time of examination, a serous otitis media has commonly developed.

Complications

Complete hearing loss and tinnitus are feared. Further complications are long-lasting vertigo and rupture of the round window membrane.

Diagnostic Procedures

- Detailed patient history—important in the case of a later lawsuit and for expert reports.
- Complete ENT examination.
- Otomicroscopy, with special attention to perforations of the tympanic membrane, haemotympanum and pathologic secretion.
- Hearing test battery: tuning fork, audiogram, speech audiogram, testing of recruitment. Mostly, the findings are a single-sided or unilateral pronounced high-frequency hearing loss or a pancochlear sensorineural hearing loss up to deafness. Recruitment is positive. In the case of rupture of the ear drum and/or luxation of ossicles, an additional conductive hearing loss is present.
- Vestibular testing: exclusion of spontaneous and provoked nystagmus with Frenzel glasses. Positional and positioning tests and examination of vestibulospinal reflexes. Assessment of vibration nystagmus can be helpful.

Additional/Useful Diagnostic Procedures

- Video-oculography (VOG) or electronystagmography (ENG). An eventual caloric testing must be performed with cold and warm air insufflation in the case of ear drum injury.
- Tinnitus analysis with subjective loudness assessment and testing of masking possibility (Fig. 1.6.7).

Therapy

Conservative Treatment

- As in sudden hearing loss (see Sect. 1.6.5)
- Initiate hyperbaric oxygen therapy as soon as possible!

Surgical Treatment

- In the case of complete hearing loss and suspicion of rupture of a cochlear window: tympanotomy and covering of a perilymphatic fistula
- In case of ear drum rupture: myringoplasty, tympanoplasty
- The surgical treatment is no replacement for conservative therapy. If needed, it is carried out additionally

Differential Diagnosis

The circumstances surrounding the patient's accident and the clinical findings derived from the patient's medical history should allow consideration of other possible diagnoses:

- Cochlear window rupture
- Sudden hearing loss (see Sect. 1.6.5)
- Toxic inner ear damage
- Craniocerebral injury, distortion of the cervical spine (whiplash injury)

Prognosis

- If high-dose glucocorticoid and rheologic therapy, perhaps combined with a hyperbaric oxygen therapy, is administered within 12 h after an acute incident, recovery of hearing is expected in up to 75% of cases. A

minor high-frequency sensorineural hearing loss and a more or less disturbing tinnitus can persist in spite of rapid and correct therapy.
- The chances of spontaneous recovery without therapy is low. Sometimes untreated post-traumatic hearing loss is progressive.

Surgical Principles
- Myringoplasty (see Sect. 1.4.6)
- Tympanoplasty (see Sect. 1.4.6)

Special Remarks
With occupational explosion trauma, special compensation insurance must be addressed (Berufsgenossenschaft in Germany, SUVA in Switzerland, INAIL in Italy).

1.6.9.2 Impulsive Noise Trauma

Synonyms
Shooting trauma, muzzle blast trauma.

Definition
Impulse sound exposure with an intensity above 150 dB(A) SPL peak equivalent with a peak sound pressure duration shorter than 3 ms (normally 0.4–1.5 ms).

Aetiology/Epidemiology
The main cause of shooting trauma is inappropriate use of a handgun, e. g. in the military, by police or by hunters. However, also guns firing blanks and toy guns produce sound pressures greater 100 dB(A)! Additionally, fireworks and airbags (Fig. 1.6.8) can cause muzzle blast trauma.

Symptoms/Findings
Otomicroscopy shows no abnormalities. In audiometric testing, there is an acute, most often unilateral mild to moderate sensorineural hearing loss of around 4 kHz (c^5 notch) and a positive recruitment. Tinnitus in the damaged ear is common. Sometimes otalgia or vestibular disturbance is present. In the case of repeated muzzle blast trauma in a short period of time (toy gun), a steep high-frequency hearing loss above 2 kHz develops.

Complications
Persistent tinnitus. Complete deafness or persistent vestibular vertigo is very rare.

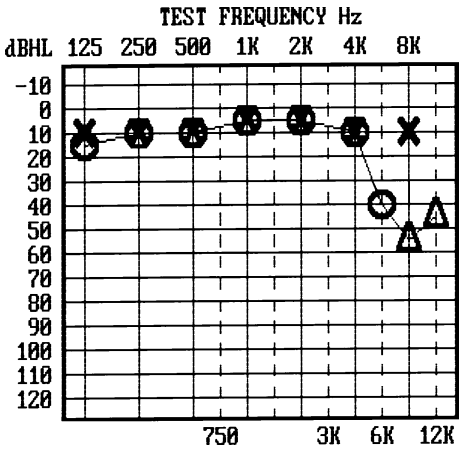

Fig. 1.6.8 Acoustic trauma at the right ear following airbag (full-size) explosion

Diagnostic Procedures
- Detailed patient history—important in the case of a later lawsuit and for expert reports
- Complete ENT examination
- Otomicroscopy, with special attention to perforations of the tympanic membrane
- Hearing test battery: tuning forks, audiogram (typical c^5 notch), speech audiogram, testing of recruitment
- Tinnitus-analysis with subjective loudness-assessment and testing of masking possibility

Additional/Useful Diagnostic Procedures
- Distortion products of otoacoustic emissions (DPOAE), otoacoustic emissions (OAE).
- Vestibular testing: exclusion of spontaneous and provoked nystagmus with Frenzel glasses. Positional and positioning tests and examination of vestibulospinal reflexes.
- Caloric vestibular testing with or without VOG/ENG.
- Brainstem evoked response audiometry (BERA): at the earliest 1 week after the incident, because of the additional noise exposure.

Therapy

Conservative Treatment
- As in sudden hearing loss (see Sect. 1.6.5).
- With more severe inner ear damage, initiate additional hyperbaric oxygen therapy as soon as possible.

Surgical Treatment
- In the case of complete hearing loss and suspicion of rupture of a cochlear window: tympanotomy and closure of a perilymphatic fistula (see Sect. 1.6.5).

- The surgical treatment is no replacement for conservative therapy. If needed, it is carried out additionally.

Differential Diagnosis

Derived from patient history, the event of accident and clinical findings any of the following other possible diagnoses should be considered:

- Cochlear window rupture
- Contusio labyrinthi
- Distortion of the cervical spine (whiplash injury)

Prognosis

- An initial good recovery of the high-frequency hearing loss occurs in approximately half of the cases. After some weeks, a remaining hearing loss or a persisting tinnitus must be considered as permanent damage. A post-traumatic increase of hearing loss above 2 kHz is possible. A spontaneous recovery of a high-frequency tinnitus is rare.
- If high-dose glucocorticoid, rheologic therapy and hyperbaric oxygen therapy is administered as soon as possible, recovery to normal hearing is frequently achievable.

Special Remarks

With occupational blast trauma any special compensation insurance must be addressed (Berufsgenossenschaft in Germany, SUVA in Switzerland, INAIL in Italy).

1.6.9.3 Acute Acoustic Trauma

Definition

Broadband sound exposure with an intensity above 100 dB(A) SPL peak equivalent for minutes to hours.

Aetiopathology

Exposure to non-impulse permanent sound causes metabolic as well as mechanical ultrastructurally visible damage at the level of the organ of Corti (outer hair cells, stereocilia). Excessive demand for oxygen and subsequent progressive ischaemia of the cochlea lead to production of free radicals, depletion of endogenous cellular antioxidants and, finally, apoptotic cell death.

Epidemiology

- Discotheques: averaged over 15 min, a music sound pressure level of 100–110 dB(A) with sound pressure peaks of up to 125–130 dB(A) SPL can be measured.
- Concerts: heavy metal, punk, hard rock reach sound pressure peaks of up to 120 dB(A), rock and pop up to 110 dB(A).
- Walkman: averaged music sound pressure level of 85–89 dB(A). In 3% of Walkman-users sound pressure levels reach up to 110 dB(A).
- Musical instruments: drums, percussion and wind instruments played indoors quickly reach sound intensities loud enough to cause hearing loss. Infants are most endangered (music school!).
- Toys: tool imitations, toy ambulances, military imitations, etc. produce an average sound pressure level of up to 100 dB(A) at a distance of 10 cm from the ear or 130–140 dB(A) directly at the ear.
- Motor sports, airplanes, do-it-yourself hobbies can reach sound intensities of up to 100 dB(A).
- Low-level flights: 50% have peak levels around 85–119 dB(A) and 2% even peak levels of around 120–125 dB(A) with impulselike, steep peak attacks. Such sounds can be encountered at or near airports.

Symptoms

Acute, most often bilateral, mild to moderate high-frequency sensorineural hearing loss (c^5 notch) with positive recruitment. Tinnitus is frequent, as is a temporary threshold shift. Otalgias or vestibular symptoms occur rarely.

Complications

Permanent hearing loss and/or tinnitus.

Diagnostic Procedures

- Detailed patient history—important in the case of a later lawsuit and for expert reports.
- Complete ENT examination.
- Otomicroscopy: without pathological findings.
- Hearing test battery: tuning fork, audiogram (typical c^5 notch), speech audiogram, testing of recruitment. Findings are most often a unilateral or more severe on one side c^5 notch or a pantonal sensorineural hearing loss of varying degree. The recruitment is always positive.
- Tinnitus analysis with subjective loudness assessment and testing of masking possibility.

Additional/Useful Diagnostic Procedures

- Vestibular testing: exclusion of spontaneous and provoked nystagmus with Frenzel glasses. Testing of vibration nystagmus. Positional and positioning tests and examination of vestibulospinal reflexes
- VOG/ENG with caloric vestibular testing

- DPOAE, OAE
- BERA: at the earliest 1 week after the incident, because of the additional noise exposure

Therapy

Conservative Treatment

As in sudden hearing loss (see Sect. 1.6.5).

Differential Diagnosis

Derived from patient history and examination:

- Explosion trauma (see Sect. 1.6.9.1)
- Shooting trauma (see Sect. 1.6.9.2)
- Sudden idiopathic hearing loss (see Sect. 1.6.5)
- Contusio labyrinthi (see Sect. 1.6.3)

Prognosis

The prognosis is unfavourable despite correct therapy; progress is possible.

1.6.9.4 Occupational Hearing Loss

Definition

Bilateral, mostly symmetric sensorineural hearing loss following intermittent exposure to broadband and/or impulse sound with an intensity above 80–85 dB(A) and a daily exposure of 6–8 h (work shift) over many years. Therefore, it is a matter of chronic noise damage.

Aetiopathology

Chronic noise exposure causes metabolic as well as mechanic ultrastructural visible damage at the level of the organ of Corti, initially causing a loss of outer hair cells, leading finally to neuronal degeneration. Typically, hearing loss initially occurs as a sensorineural high-frequency notch, normally at 4 kHz (c^5 notch). The middle frequencies, e. g. the main speech frequencies, are affected considerably later. The recruitment is always positive. In approximately 70% of cases, a bilateral tonal tinnitus exists. The extent and the progress of the hearing loss depend on the intensity, duration of exposure and frequency composition of the sound as well as the duration of recovery phases and the *individual noise susceptibility*. An individual noise susceptibility is suspected in genetically predamaged inner ears, after sudden hearing loss, after treatment with ototoxic medication and trauma. Moreover, humans with blond hair, fair complexion and lightly coloured eyes seem to be especially endangered owing to a lack of melanin.

Aetiology/Epidemiology

By definition, noise-induced hearing loss is an effect of working in noise and therefore occurs most frequently in metalworkers, mineworkers, airport workers, radio operators, disc jockeys and military personnel as well as construction workers and orchestra musicians, etc.

Symptoms

Because of extensive recruitment, this progressive binaural hearing loss leads to an important communication problem in noisy environments (conversation of multiple persons, theatre, restaurant). A pronounced hyperacusis is common. Tinnitus is found in 70% of patients.

Complications

Social isolation following hearing loss, psychiatric impairment secondary to the tinnitus.

Diagnostic Procedures

- Detailed patient history, including professional and recreational sound exposure. If necessary, inquire regarding professional sound exposure at employment or special insurance organizations (Berufsgenossenschaft in Germany, SUVA in Switzerland, INAIL in Italy).
- Complete ENT examination.
- Otomicroscopy: without pathological findings.
- Hearing test battery: tuning fork, audiogram, speech audiogram, tympanometry, stapedial reflexes, testing of recruitment.
- Tinnitus analysis (Fig. 1.6.7) with subjective loudness assessment and testing of masking possibility.

Additional/Useful Diagnostic Procedures

- OAE, DPOAE, transitory evoked otoacoustic emissions.
- BERA, auditory evoked cortical responses.
- VOG/ENG.
- Vestibular testing: exclusion of spontaneous and provoked nystagmus with Frenzel glasses. Positional and positioning tests and examination of vestibulospinal reflexes.

Therapy

Conservative Treatment

- A pharmacological therapy of the chronic noise-induced hearing loss is not possible. Glucocorticoids and rheologics (see Sect. 1.6.5) can be used in an attempt to treat the tinnitus.

- In most cases, the hearing loss can be compensated for with hearing aids. In selected cases, a tinnitus masker can be helpful.

Additional Useful Therapeutic Strategies

By avoidance of noise or use of effective ear protection when working in a noisy environment, the progress of the chronic noise induced hearing loss can be prevented.

Differential Diagnosis

- Presbyacusis (see Sect. 1.6.11), progressive idiopathic hearing loss, hereditary sensorineural hearing loss.
- Drug-induced or toxic sensorineural hearing loss.
- Post-traumatic sensorineural hearing loss following contusio or commotio labyrinthi, craniocerebral injury, distortion of cervical spine (whiplash injury).
- If evident asymmetry of hearing thresholds and/or pathologic findings in vestibular testing or BERA are present, the diagnosis of noise-induced hearing loss is unlikely and a search for a retrocochlear cause of hearing loss (tumour of the cerebellopontine angle, multiple sclerosis, etc.) is necessary.

Prognosis

- After cessation of activity in a noisy environment, noise-induced hearing loss is not progressive anymore. This is the reason why routine use of noise protectors or a change to a less noisy work environment can stop the progression of hearing loss.
- If a sensorineural hearing loss shows progression despite cessation of noise exposure, other or additional causes must be sought.

Special Remarks

1. To reliably distinguish professional noise-induced hearing loss from other types of hearing losses, the following requirements must be fulfilled:
   - An adequate noise exposure has to be confirmed.
   - The hearing loss must have developed during the time of noise exposure.
   - The hearing loss must be more or less symmetric.
2. The patient history must cover questions regarding earlier ear diseases and head injuries.
3. If a noise-induced hearing loss is suspected, any special compensation insurance (Berufsgenossenschaft in Germany, SUVA in Switzerland, INAIL in Italy) must be addressed.
4. For an assessment of noise-induced hearing loss, the recommendations of special insurance organizations (e. g. Berufsgenossenschaft in Germany, SUVA in Switzerland, INAIL in Italy) are important guidelines, which must be obeyed.

Suggested Reading

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2. Sataloff J, Sataloff RT, Menduke H, et al. (1983) Intermittent exposure to noise: effects on hearing. Ann Otol Rhinol Laryngol, 92:623–628
3. Spoendlin H (1971) Primary structural changes in the organ of Corti after acoustic overstimulation. Acta Otolaryngol, 71:166–176

1.6.10 Ototoxicity

SALVATORE IURATO

1.6.10.1 Definition

Adverse effect to the cochlear or vestibular portion of the inner ear caused by pharmaceutical agents.

1.6.10.2 Aetiology/Epidemiology

The most ototoxic compounds in clinical practice are aminoglycoside antibiotics (streptomycin, dihydrostreptomycin, neomycin—all routes of administration, kanamycin, gentamicin), loop diuretics, quinines and chemiotherapy agents (cisplatin) (Table 1.6.1).

The incidence of ototoxicity has not been accurately determined. Risk factors are a decreased renal function, increased daily doses, extended duration, concomitant administration with more than one ototoxic drug and prematurity.

1.6.10.3 Symptoms

The following symptoms may be temporary or permanent: high-pitched tinnitus (earliest sign of cochlear damage), hearing loss (with or without vertigo), nausea, dizziness. Initially the loss of hearing affects the high frequencies. As damage progresses, the lower frequencies become involved.

1.6.10.4 Audiometric Monitoring of Risk Patients on Ototoxic Drug Therapy

- Establish "baseline" hearing level (air-conduction thresholds at 0.5, 1, 2, 4, 6 and 8 kHz).
- Repeat the test during therapy (every 2 days, every week). Ototoxicity is defined as a shift from the

Table 1.6.1 Ototoxic compounds in clinical practice

Drugs	Main damage	
	Primarily ototoxic	Primarily vestibulotoxic
Streptomycin	+	+++
Dihydrostreptomycin	+++	+
Neomycin	+++	+
Kanamycin	+++	+
Gentamicin	+	+++
Ethacrynic acid	+	+
Salicylate	+	
Cisplatin	++	

baseline of 15 dB or more at both 6 and 8 kHz, either unilaterally or bilaterally as assessed 5–7 weeks after beginning of the treatment.
- Monitoring of the status of the cochlea with the acoustic emissions and high-frequency audiometry.

1.6.10.5 Prevention

- Discontinue or change the medication (if it is possible).
- Antioxidant therapy (iron chelators, vitamin E, ascorbic acid).
- Prophylactic treatment (aspirin).
- In a patient who has decreased renal function, dose schedules should be adjusted.

1.6.10.6 Rehabilitation

If the hearing is still serviceable, amplification with a hearing aid may be used.

1.6.10.7 Potential Ototoxicity of Topical Otic Preparations

Through a perforation of the tympanic membrane otic drops may enter into the round window niche, diffuse across the round window membrane and reach the membranous labyrinth. Ototoxic preparations include alcohol, povidone iodide, gentamicin, neomycin, polymixin B, chloramphenicol and hydrocortisone. Non-ototoxic preparations include amphotericin B, sulphacetamide, ciprofloxacin, triamcinolone and dexamethasone.

Suggested Reading

1. Bergstrom LV, Thompson PL (1984) Ototoxicity. In: Northern JL (ed) Hearing disorders. Little, Brown. Boston, pp 119–134
2. Riggs LC, Matz GJ, Rybak LP (1998) Ototoxicity. In: Bailey BJ (ed) Head neck surgery–otolaryngology, 2nd edn. Lippincott-Raven, Philadelphia
3. Slattery W, Brownlee R (1955) Otic preparations. In: Swarbick J, Boylan J (eds) Encyclopaedia of pharmaceutical technology, vol 11. Dekker, New York

1.6.11 Presbyacusis

UWE GANZER AND ANDREAS ARNOLD

1.6.11.1 Synonym

Age-related hearing loss.

1.6.11.2 Definition

Presbyacusis describes the progressive sensorineural hearing loss nearly every human experiences starting in the fifth decade of life. It is more or less symmetric and begins in the higher-frequency range with or without tinnitus.

1.6.11.3 Aetiopathology

Degeneration of hair cells, cochlear neurons, stria vascularis, cochlear nerve and components of the central auditory pathway, e. g. cochlear nuclei.

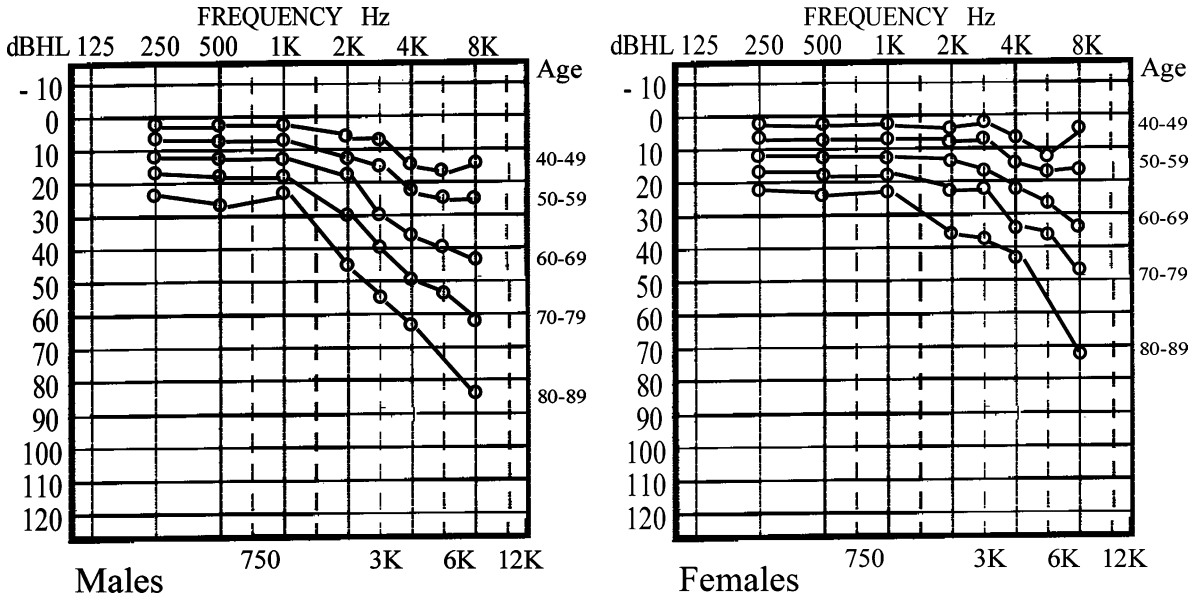

Fig. 1.6.9 Presbyacusis: average hearing loss in industrialized countries correlated with the age

1.6.11.4 Aetiology/Epidemiology

- The hearing loss is caused, on one hand, by the physiologic processes of aging based on individual genetic predisposition and, on the other hand, by exogenous degeneration of parts of the inner ear (supporting cells, basilar membrane, outer hair cells) and central auditory pathway components. The exogenous degeneration is essentially the consequence of environmental influences, nutritional habits, toxicity of legal drugs, professional and recreational noise exposure, ototoxic medications, medical and neurological problems, etc. A hereditary component of presbyacusis has been demonstrated.
- Worldwide, 400 million humans are affected. In the UK 92% of persons older than 60 years show a hearing loss of more than 25 dB. In Denmark, the percentage of hearing loss per decade is 3 dB up to the age of 55 years and 9 dB for persons older than 55 years.

1.6.11.5 Symptoms

Progressive hearing loss with the greatest threshold shift in the high frequencies (Fig. 1.6.9). Speech comprehension is reduced, mainly in ambient noise (party effect), loss of discrimination. Discomfort in noisy environments, during phone calls. Decline of directional hearing. Tinnitus is common.

1.6.11.6 Complications

Psychosocial isolation und suspiciousness of the environment are caused by the loss of ability to communicate, secondary to the hearing loss. Depressive crisis can be triggered by the tinnitus.

1.6.11.7 Diagnostic Procedures

- Detailed patient history, including professional and recreational sound exposure as well as family history.
- Complete ENT examination.
- Otomicroscopy: without pathological findings.
- Hearing test battery: tuning fork, audiogram, speech audiogram, tympanogram and stapedial reflex audiometry. Symmetric sensorineural hearing loss limited to the higher frequencies, pantonal or gently declining from 1 kHz. Poor discrimination in the speech audiogram. Mostly, a positive recruitment is seen. With pancochlear sensorineural hearing loss, recruitment can also be negative.
- Tinnitus analysis with subjective loudness assessment and testing of masking possibility (Fig. 1.6.6).

1.6.11.8 Additional/Useful Diagnostic Procedures

- Basic vestibular testing with Frenzel glasses: exclusion of spontaneous and provoked nystagmus, positional

and positioning tests and examination of vestibulospinal reflexes.
- Brainstem evoked response audiometry: in the case of a clear asymmetry of the hearing loss.
- Vestibular testing with video-oculography/electronystagmography including a caloric test.
- MRI, if the results of vestibular and/or brainstem evoked response audiometry examinations are pathologic.

1.6.11.9 Therapy

Conservative treatment includes the following:
- Binaural hearing aids should be prescribed as soon as possible, with additional hearing training and lip reading training in the case of severe hearing loss.
- Phone amplifier. Optical signal devices to help at home.
- Resocialization, possibly with concomitant psychotherapy.
- Medical therapy only in the case of rapid progress of presbyacusis or disturbing tinnitus. In such cases rheologic drugs, α-lipoic acid or antioxidants such as vitamin C or E can be used. In most cases, these measures remain unsuccessful.

1.6.11.10 Differential Diagnosis

- Hereditary sensorineural hearing loss, otosclerosis
- Symptomatic sensorineural hearing loss with medical or neurological disease or systemic autoimmune disease
- Noise-induced hearing loss, ototoxic medication, etc.
- Traumatic sensorineural hearing loss secondary to contusio or commotio labyrinthi, craniocerebral injury, whiplash injuries

1.6.11.11 Prognosis

Normally, unavoidably slowly progressive hearing loss.

1.6.11.12 Special Remarks

The diagnosis of "presbyacusis" requires the exclusion of all other possible causes. Not every hearing loss in the elderly is a presbyacusis!

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1.6.12 Hearing Impairment in Childhood

JAN KIEFER AND GERARD M. O'DONOGHUE

1.6.12.1 Synonym

Childhood deafness.

1.6.12.2 Definition

- **Hearing impairment in childhood** refers to any hearing loss, occurring from birth to late childhood, e. g. around the age of 16 years. It may be unilateral or bilateral.
- It may be present at birth, e. g. **congenital**, or **acquired** after birth, either during the perinatal period of life or later during lifetime. As hearing impairment affects the acquisition of speech, its occurrence is also classified in relation to the stages of speech development as *prelingual* (0–2 years of age), *perilingual* (2–4 years) and *postlingual* (more than 4 years).
- It can be classified into four categories of severity: **mild** (average hearing levels ranging from 10 to 39 dB), **moderate** (average hearing levels from 40 to 69 dB), **severe** (average levels ranging from 70 to 94 dB) and **profound** (hearing loss that has average hearing levels greater than 95 dB).
- In principle, hearing loss can be **conductive** (due to malfunction of the outer ear or the middle ear) or **sensorineural** (due to malfunction of the inner ear or the auditory nerve), or **mixed**.
- Its course may be temporarily, fluctuating, progressive or permanent (**permanent childhood hearing impairment, or PCHI**).

1.6.12.3 Aetiology/Epidemiology

1. By far the commonest cause of hearing loss during childhood is a **conductive hearing loss** caused by **otitis media with effusion** (OME) following episodes of acute otitis media (see Sect. 1.4.4). Fortunately, for most children, the hearing loss is only transient and, if

not, is readily correctible by myringotomy, placement of ventilation tubes and adenoidectomy if conservative treatment fails (see Sect. 1.4.4).

2. Other causes for **conductive hearing loss** may be chronic inflammatory middle ear disease such as chronic mesotympanic otitis media (see Sect. 1.4.6), cholesteatoma (see Sect. 1.4.7), or congenital malformations of the outer ear canal and middle ear (see Sect. 1.2).
3. **Sensorineural hearing loss** is primarily caused by malfunction of the organ of Corti (sensory hearing loss), rarely by malfunction of the auditory nerve (neural hearing loss) or the interface between sensory hair cells and spiral ganglion cells (auditory neuropathy).
4. **PCHI** has a prevalence of approximately one to two per 1,000 children in Europe; literature values range from one to 4.2 per 1,000 children in selected populations.
5. **Congenital hearing impairment:** a genetic cause may account for more than 50% of these cases. Approximately three quarters of these are non-syndromic; one quarter are associated with hereditary syndromes carrying other specific hereditary features in addition to deafness. Most frequently, the hereditary course is autosomal recessive. The commonest findings are mutations in the gene *GJB2*, coding for connexin 26, a gap junction protein in the inner ear that is necessary for the maintenance of the endocochlear potential. Other hereditary causes are autosomal dominant with various phenotypes or X-linked.

Further causes of congenital hearing loss are:

- Infections during pregnancy (rubella, cytomegalovirus, toxoplasmosis).
- Inner ear malformations due to developmental arrest in embryonic stages such as Mondini malformation or common cavity malformation. They may be related to syndromes that are associated with other symptoms or occur in isolated forms.

6. **Acquired hearing impairment:** hearing loss may be acquired in the perinatal period owing to hypoxaemia, severe infections, prolonged newborn icterus and others. Following the perinatal period, the commonest cause of acquired inner ear hearing loss in early childhood is bacterial meningitis, with permanent hearing loss complicating up to 30% of affected children, mostly mild or unilateral. However, about 2% of children affected by meningitis develop a bilateral permanent and profound hearing loss. Vaccination against *Haemophilus influenzae* type B and early vaccination against *Streptococcus pneumoniae* reduce the risk of developing meningitis as well as the risk for acquired deafness due to meningitis. Other causes include infections (e. g. measles, mumps, varicella), trauma, middle ear diseases and administration of ototoxic drugs.
7. **Progressive hearing impairment:** progressive inner ear hearing loss during childhood can be associated with genetic mutations causing syndromes such as Pendred syndrome (Fig. 1.1.16) and the genetically associated syndrome of a large vestibular aqueduct (Fig. 1.1.26), Usher syndrome (retinitis pigmentosa and progressive hearing loss) and Alport syndrome. Other forms of genetically caused hearing loss occur as progressive hearing loss starting during adolescence. Frequently, the cause of progressive hearing loss remains unknown.

1.6.12.4 Symptoms

The most important symptoms of PCHI are the absence of adequate reactions to environmental sounds and speech as well as the delay or absence of normal speech development, depending on the severity of the hearing impairment. Additional symptoms may be poor general communication skills and behavioural difficulties out of frustration in communicational attempts.

However, it is important to understand that these symptoms are not easy to recognize, even for professional child carers, and that diagnosis even of severe hearing impairments is often considerably delayed, even up to the age of 2–4 years, if based only on the recognition of these symptoms. In many cases, the parents' concern precedes professional diagnosis and should therefore be taken for serious and prompt further hearing assessments for the child.

Risk factors that are associated with PCHI such as a family history of hearing impairment, infections during pregnancy (e. g. rubella, toxoplasmosis, cytomegalovirus), prematurity, low birth weight, necessity of admission to intensive care unit, prolonged newborn icterus, administration of ototoxic drugs (e. g. aminoglycosides), craniofacial abnormalities and hereditary syndromes should draw attention to possible hearing impairment.

Deficits in speech development include receptive skills of speech understanding as well as speech production. Affected areas range from basic skills such as segmentation and analysis of phonemic structure, short-term auditory memory as well as vocabulary to higher levels of speech such as syntax and grammar.

It is important to monitor the speech development of children, since some children may have progressive hearing loss occurring during childhood.

1.6.12.5 Complications

- The early auditory system is particularly receptive to sounds and speech. Sufficient auditory input is necessary to induce the maturation of the auditory system. Failure to stimulate the auditory system during this period (referred to as the critical period) can have lifelong detrimental effects on the acquisition of spoken language. Untreated hearing loss can also compromise a

child's reading ability and educational attainment. This may limit access to further education, may restrict employment opportunity and lead to greater dependence on social services later in life. Thus, permanent untreated hearing impairment can have far-reaching consequences for the child, its family and for the wider community. Evidence suggests that early identification and treatment may significantly reduce the impact of PCHI. Fitting of hearing aids or cochlear implantation after the critical period of language development (from 0 to 4–6 years) will not be able to fully recover these effects.

- After meningitis, labyrinthitis may develop to fibrosis and/or neo-ossification of the cochlear duct, making cochlear implantation difficult and less successful. Early diagnosis via MRI or CT after meningitis (Fig. 1.1.25) is recommended to detect possible early signs and proceed to implantation.

1.6.12.6 Diagnostic Procedures

Recommended European Standard: Diagnostic Steps or Investigations in Neonatal Screening for Hearing Impairment

1. General neonatal screening for hearing impairment is recommended (European consensus conference 1998) since diagnosis of hearing impairment is often delayed and early intervention is of great importance. All newborns should be screened during their first days of life. Screening methods should have high sensitivity and specificity, and should be objective as well as time- and cost-efficient. The following methods are available:
   - Automated measurements of otoacoustic emissions (OAE), such as transitory evoked OAE (TEOAE) and distortion products of OAE (DPOAE)
   - Automated brainstem evoked response audiometry (BERA)
   - Automated measurements of amplitude modulation following response (AMFR)
2. If the child fails to pass the screening for hearing impairment, follow-up with eventual rescreening and more extensive auditory testing to confirm or exclude hearing loss is necessary.

History

1. Risk factors that are associated with PCHI such as a family history of hearing impairment, infections during pregnancy, prematurity, low birth weight, necessity of admission to an intensive care unit, prolonged newborn icterus and administration of ototoxic drugs should be investigated.
2. Ask about:
   - Reactions to sound and speech: Does the child startle at loud sounds, does it react to voice when the speaker is not visible, e. g. calming or smiling?
   - Language development: Does the child vocalize, does it imitate (mama, papa), what is the range of vocabulary, are there articulation problems, are there problems with syntax or grammar?
   - Behavioural abnormalities: e. g. aggression, low tolerance to frustration, communicative strategies.

Physical Examination

1. Inspection
   - General physical examination should pay attention to any signs of hereditary syndromes.
   - Look at the facial features of the child and its parents: craniofacial abnormalities, e. g. outer canthi of the eyelid slant downwards in Treacher Collins syndrome or upwards in the branchio-oculo-facial syndrome.
   - Ear anomalies such as hypoplasia or aplasia of the pinna, preauricular appendages and atresic ear canal draw attention to possible conductive hearing loss or associated inner ear malformations.
   - The neck should be evaluated for any branchial remnants (found in the branchio-oto-renal syndrome) or an enlarged thyroid gland (Pendred syndrome and associated enlarged vestibular aqueduct).
   - Blue sclerae are associated with osteogenesis imperfecta.
   - A white forelock and pigmental anomalies of the iris may indicate Waardenburg syndrome.
2. **Otoscopy**: Note anomalies of the pinna and external auditory ear canal, check for the presence of a normal tympanic membrane, possible anomalies of the handle of the malleus, signs of OME (retraction, fluid behind the eardrum) or chronic otitis media. Rarely, a whitish mass behind the eardrum may indicate congenital cholesteatoma (Fig. 1.4.14). Note that in sensorineural hearing losses, the tympanic membrane is normal.
3. Rhinoscopy: to exclude nasal stenosis, choanal atresia, nasal infections.
4. Pharyngoscopy: to exclude hyperplastic or infected adenoids or tonsils.

Audiological Testing

We distinguish between **subjective** and **objective** auditory tests. Subjective tests require some form of reaction of the subjects tested, whereas objective tests can be carried out without active feedback. Even in small children and babies, age-adequate subjective tests are possible (e. g. behavioural response audiometry, conditioned response audiometry); however, they require special expertise. Objective tests such as OAE and BERA can be performed at

any age, sometimes requiring sedation. Diagnosis should only be based on the combination of subjective and objective tests, and should be reevaluated at subsequent developmental stages of the child to reach a higher degree of exactitude, to distinguish between transitory, permanent or progressive problems and to account for maturation and developmental processes.

Subjective tests are:

- **Behavioural response audiometry** (age range 0–2 years): Spontaneous responses to sounds such as calming, blinking and startling are watched for by experienced examiners. In visual reinforcement audiometry, reactions of the child such as head turning are reinforced by attractive visual stimuli. Bilateral free-field testing is possible; thresholds found are generally 20–30 dB above real auditory thresholds.
- **Performance test and play audiometry** (age range 2–5 years): This test requires that a child can be actively involved in a task. It uses a conditioned response (e. g. stacking cubes) to evaluate auditory thresholds in bilateral free-field conditions.
- **Pure tone audiometry** (age range from 3.5–4 years upwards): testing side specific pure tone thresholds (see Sect. 1.1.5.2). The child must be able to wear headphones and cooperate in the task.
- **Speech audiometry**: Various speech tests with age-appropriate language material are available. Results are influenced by auditory thresholds, capacity of auditory speech analysis as well as general speech development.
- Tests for **central auditory processing disorders**: Special tests such as the dichotic listening test or hearing in noise are used to diagnose central auditory processing disorders such as in auditory attention deficit syndrome.

Objective tests are:

- **Screening tests**: They are designed to detect hearing losses greater than 30–40 dB, in general without giving detailed thresholds.
- **Tympanometry**: to detect middle ear problems (e. g. OME). Stapedial reflex measurements are useful to estimate thresholds (see Sect. 1.1.5.2).
- **OAE**: OAE reflect the normal activity of outer hair cells. They are generally present when hearing loss does not exceed 30–40 dB. However, they do not reflect the function of inner hair cells and the auditory nerve; therefore, they might be present in cases of auditory neuropathy or neural hearing loss. TEOAE are click-evoked and have a broad response spectrum over the whole frequency range. DPOAE are evoked by continuous two tones and are frequency-specific. By determining growth functions of DPOAE, one can obtain an approximation of auditory thresholds.
- **Auditory brainstem response**: The synchronized neural activity of the auditory pathway (spiral ganglion and cochlear nerve, cochlear nucleus, lateral lemniscus, superior olive and inferior colliculus) is recorded in response to click stimuli (broad frequency response) or tone bursts (limited frequency specificity). Auditory thresholds can be approximated. The method may require sedation in children.
- **AMFR**: Like for auditory brainstem response, the synchronized neural activity is measured but is elicited by sine waves that are amplitude-modulated. Higher frequency specificity can be achieved.

Assessment of Speech and Language Development

As one of the main symptom of PCHI is delayed speech and language development, it is important to include assessment of speech and language development by psychologists, speech and language therapists or teachers of the deaf in a multidisciplinary approach.

Neurological and Ophthalmological Examination

Many children with PCHI have additional neurological and vision deficits. Neurological examination and vision screening should be carried out if indicated.

Diagnostic Imaging

- High-resolution, thin-section CT is the modality of choice to visualize the bony structures of the outer ear, the middle ear, the mastoid, the inner ear and the internal acoustic meatus (Figs. 1.1.23, 1.1.24). Soft tissue masses or fluid in the middle ear or mastoid can also be detected. Special attention has to be paid to detect possible malformations of the ossicles, of the inner ear, e. g. Mondini malformation (Fig. 1.2.2), common cavity, labyrinthine malformations, enlarged vestibular aqueduct (Fig. 1.1.26), or in postmeningitis cases, neo-ossifications of the scala tympani, media or the labyrinth (Fig. 1.1.25).
- MRI is indicated to visualize the fluid content of the inner ear, the auditory and vestibular and facial nerves in the inner acoustic meatus, and to detect central nervous system abnormalities. It is of special importance to detect early signs of fibrosis and neo-ossification of the cochlea after meningitis.

Genetic Testing and Counselling

Genetic testing may be useful in syndromic as well as non-syndromic hearing loss for diagnostic purposes and counselling of patients and parents. Specific genes associated with syndromes such as Waardenburg, Pendred and Usher have been identified. In non-syndromic hearing

loss, the *GJB2* gene encodes for the connexin 26 molecule. It can be tested in many centres and may account for approximately 50% of cases of presumed non-syndromic genetic deafness. However, the number of gene mutations associated with deafness continues to increase; more then 100 mutations have been described. Therefore, negative findings in genetic testing do not preclude the genetic origin of a hearing loss.

1.6.12.7 Therapy

Conservative Therapy

- **Early intervention** is a key factor to prevent sequelae of hearing impairment. Even children as young as 3–6 months can be fitted with hearing aids; however, special expertise is needed to fit very small children.
- **Fitting of hearing aids**: The first step in therapy of PCHI is providing adequate amplification by means of hearing aids. They should be fitted on the basis of subjective and objective measures, and bilaterally, if the hearing loss is bilateral. Hearing aids have to be maintained and ear moulds have to be adjusted regularly to fit the ear canals, which typically enlarge with age. In conductive hearing loss, e. g. in ear malformations, that is not ready to be corrected surgically, bone-conduction hearing aids are the treatment of choice (Fig. 1.2.4).
- **Monitor the children's development:** Reactions to sound and speech as well as speech and language development of children fitted with hearing aids have to be monitored by the children's parents, paedaudiologists, teachers and therapists to make sure that amplification is adequate and optimal benefits are obtained. Training of communicative skills and counselling of parents is of great importance and should start as soon as possible.
- **Consider cochlear implantation**: If hearing capacities as well as speech and language development remain insufficient in patients with severe or profound hearing impairment, despite optimally fitted hearing aids, a cochlear implantation has to be considered.

Surgical Treatment

Conductive Hearing Loss

- **OME**: If conservative treatment fails, myringotomy and placement of ventilation tubes should be performed (see Sect. 1.4.4).
- **Chronic mesotympanic otitis media and cholesteatoma**: Surgical treatment is indicated (see Sects. 1.4.6, 1.4.7).
- **Malformations of the ear**: Reconstruction of the outer ear canal and ossicular chain, implantation of active middle ear implants and placement of bone-anchored hearing aids are possible surgical options (see Sect. 1.2).

Sensorineural Hearing Loss

Function of Cochlear Implants

Cochlear implants replace the function of the inner ear in transferring acoustic sounds into neural excitation patterns. Unlike hearing aids, which amplify sounds acoustically, cochlear implants convert the sounds into electrical stimulation patterns, which electrically stimulate fibres of the cochlear nerve and thus elicit hearing sensations (Fig. 1.6.10). A cochlear implant system consists of two parts: the external speech processor and the implant itself (Fig. 1.6.11). The speech processor picks up external sounds, analyses them for frequency and time content and generates instructions for stimulation. Together with the necessary energy, the information is sent to the implant via a short high-frequency radio connection. The sender is centred over the implant with a magnetic link. The implant receives the instructions and generates electric pulses. These are delivered by the intracochlear electrodes (currently between 12 and 22) that follow the tonotopic organization of the cochlea. Electrodes at the base (near the round window) elicit high-pitched auditory sensations; electrodes near the apex elicit low pitches.

Indications for Cochlear Implantation

- For profoundly deaf children (typically those with hearing losses greater than 100 dB) and those chil-

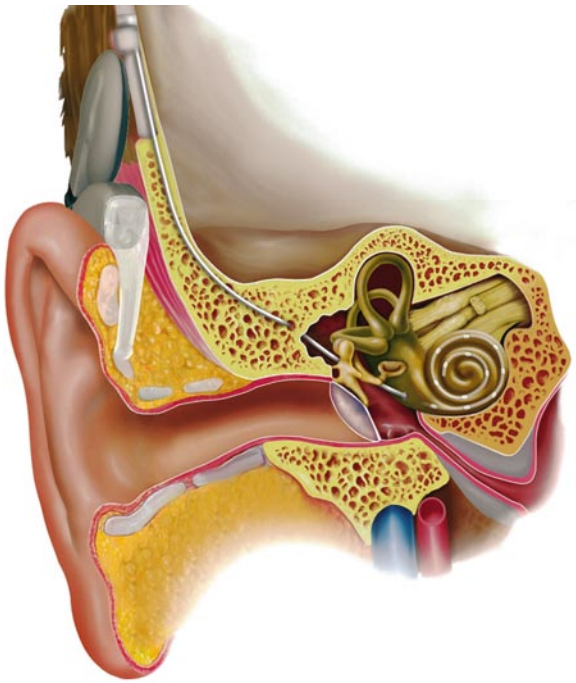

Fig. 1.6.10 Cochlear implant

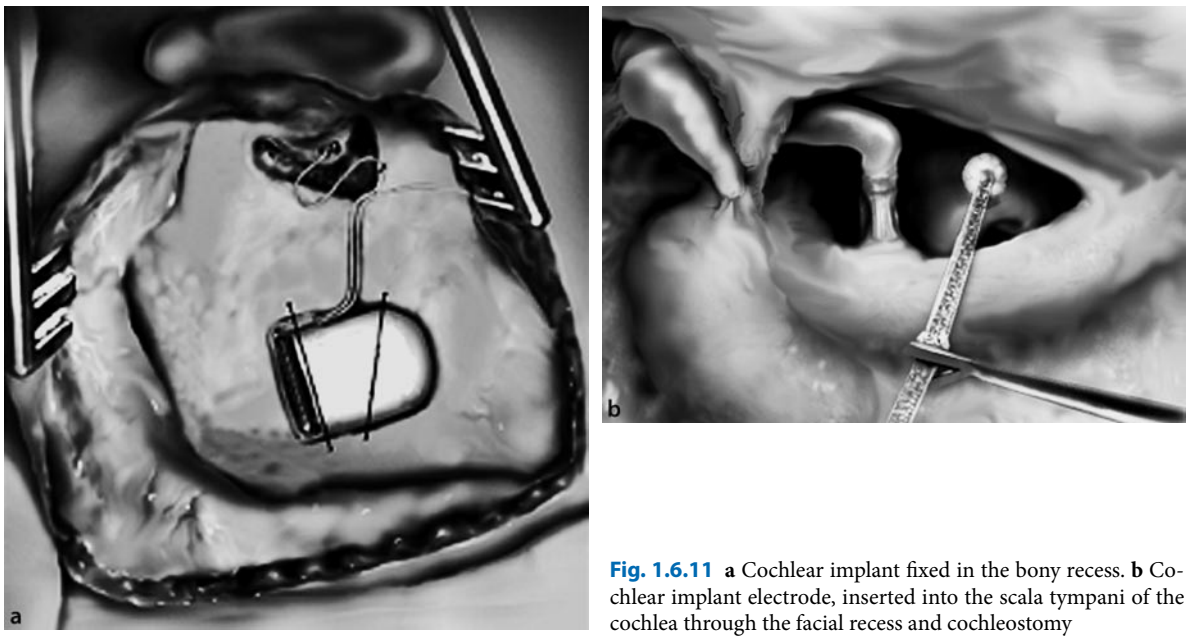

Fig. 1.6.11 **a** Cochlear implant fixed in the bony recess. **b** Cochlear implant electrode, inserted into the scala tympani of the cochlea through the facial recess and cochleostomy

dren with severe to profound hearing loss, who do not obtain sufficient benefit from powerful hearing aids to develop their speech and language skills, cochlear implants are extremely valuable in providing access to speech signal and sounds.

- Cochlear implantation can be performed as early as 8–12 months if indicated. Early intervention, e. g. implantation before the age of 2 years, is best to make use of critical periods in hearing as well as speech and language development. Therefore, early diagnosis and hearing aid trial periods are of importance.
- Bilateral implantation is possible and beneficial in adults and in children, allowing for better speech understanding under difficult listening conditions and partial development of spatial and directional hearing.
- Children with multiple handicaps in addition to profound hearing loss will obtain significant profit from cochlear implantation in most cases, even if receptive and expressive language development may not be expected owing to, e. g., intellectual handicaps.
- For malformations of the inner ear or neo-ossifications after meningitis, special surgical techniques have been developed.

Contraindications for Cochlear Implantation

- Cochlear implantation is not possible if the auditory nerve is absent. In these cases, brainstem implants may be an alternative approach.
- Cochlear implantation is contraindicated if sufficient rehabilitational and/or technical support for maintenance of the device function cannot be ensured.

Additional Therapeutic Strategies

- Additional speech and language therapy is necessary in most patients with severe and profound hearing impairment.
- General support, careful choice of educational settings and counselling of parents is of importance.
- Children with a cochlear implant require regular programming and control of the speech processor, which is best ensured in multidisciplinary cochlear implant rehabilitation programmes.

1.6.12.8 Differential Diagnosis

- Differential diagnosis between conductive and sensorineural hearing loss is an essential prerequisite.
- PCHI has to be differentiated from central auditory processing disorders that may be mistaken for peripheral hearing loss.
- Non-organic hearing loss (psychogenic) should not be missed, occurring most often in teenage children.
- Unilateral hearing loss is often overlooked in childhood.
- Progressive loss may pass unrecognized; the same is true for hearing loss affecting only part of the frequency range as the findings of the initial objective hearing screening can be normal.

1.6.12.9 Prognosis

Evidence confirms that early identification and treatment, coupled with sustained, appropriate habilitation and educational support can achieve excellent outcomes. Development of spoken language can proceed at rates similar to that for normal-hearing children even in profoundly deaf children, provided that early identification and cochlear implantation are achieved. They can achieve impressive competence with oral communication, and can often attend mainstream schools (with varying degrees of assistance), achieving their full educational potential.

Negative prognostic factors include late age at diagnosis, the presence of other cognitive disabilities, inappropriate communication strategies, inadequate educational and rehabilitational support and poor socioeconomic status.

1.6.12.10 Surgical Procedure: Cochlear Implantation

See Fig. 1.6.10.

The procedure involves the following:

- Monitoring of facial nerve function is recommended to avoid injury to the facial nerve.
- Extended retroauricular incision.
- Cortical mastoidectomy.
- Posterior tympanotomy with identification of chorda tympani, facial nerve, stapedial tendon, stapes and round window niche. The round window membrane may be identified for better localization of the scala tympani.
- Drilling of the bed for the implant housing and preparation of tie-down sutures (Fig. 1.6.11a).
- Cochleostomy of the scala tympani in front of the anterior/inferior aspect of the round window membrane, alternatively incision of the round window membrane (Fig. 1.6.11b).
- Insertion of the cochlear implant electrode until the point of first resistance, no forceful manoeuvres to avoid cochlear trauma, sealing of the cochleostomy (Fig. 1.6.11b).
- Fixation of implant housing with tie-down sutures or tightly sutured periosteum, depending on the type of implant and incision (Fig. 1.6.11a).
- Intraoperative tests to ensure correct function and placement (electrode impedance, implant function, neural responses to electric stimulation and measurement of electrically evoked stapedial reflexes).
- Fixation of the electrode, wound closure, sterile dressing.
- Postoperative radiological control of electrode placement and depth of insertion (transorbital or Stenvers view, Fig. 1.1.21).

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1.6.13 Hearing Aids

WALTER LIVI

1.6.13.1 Definition and Components

A hearing aid is a miniature electronic instrument that detects, amplifies, elaborates and transmits sound to the hearing impaired patient's ear. Its basic components are the *microphone* (input), the *amplifier* (elaborator) and the *receiver* (output).

1.6.13.2 Classification

Hearing aids can be classified into three groups depending on the technology used:

1. **Analogue**: The microphone converts sound waves to a continuous electrical signal that is similar to the stimulus in intensity, frequency and time. The amplifier then amplifies the electrical signal, which can be modified by manual controls (trimmers), and then transmitted to the receiver that reconverts the elaborated electrical signal to a sound wave.
2. **Digitally programmable analogue**: They represent an evolution of analogue hearing aids that differ only in the phase of amplification. The amplified electrical signal is not modified by a manual trimmer but is amplified electronically by the computer. The elaboration of the signal remains an analogue process.
3. **Fully digital**: The microphone converts sound waves into an analogue electrical signal. The analogue-to-digital converter transforms the continuous electrical signal into a series of binary numbers (0–1). The digital sound processor digitally elaborates the numerical signal according to algorithms contained in the program. The analogue-to-digital converter transforms

Fig. 1.6.12 From *left to right*, examples of completely inside the canal (CIC), in the concha (ITC), mainly in the external auditory canal (ITE) and behind the ear (BTE) hearing aids

the series of numbers into an electrical signal. The receiver then reconverts the electrical signal into sound waves.

Types of hearing aids are shown in Fig. 1.6.12. Depending on the place in which they are worn, hearing aids can be classified in the following way: *behind the ear* (BTE), *in the ear* (ITE), *body aid*, and *eyeglass aid* (*spectacles*). The choice is influenced by the type and entity of the hearing impairment and by the needs of each patient:

1. **BTE hearing aids**
   - Can be used for all types of hearing impairments.
   - Are composed of a plastic shell that contains the microphone, amplifier, receiver, volume control (or other manual controls or switches) and battery.
   - They are quite small and are placed BTE (pinna).
   - They are connected to the flexible tube of the ear mould by a plastic hook or by a fine wire to a receiver positioned directly in the ear canal (receiver in the ear, RITE) (Fig. 1.6.13). The ear mould is composed of biocompatible material that is made to measure for the ear canal of the patient.
2. **ITE hearing aids**
   - They can be placed completely inside the canal (CIC) or mainly in the external ear canal (ITE) or in the concha of the external ear canal (ITC) (Fig. 1.6.12).
   - They are made of biocompatible material and are well accepted by patients because they are small and practical.

Fig. 1.6.13 The application of modern design to advanced electronic technology. A hearing aid that implements the receiver in the ear (RITE) solution

   - They are not a good choice in cases of severe to profound hearing loss because they do not provide sufficient amplification.
   - They are not often prescribed to children, because of the limited size of the child's ear canal and its continuous change in size.
3. **Body worn pocket aids**: The plastic case of the aid contains all components except for the receiver, which is placed in the ear canal. They are no longer in use because they are not very practical.
4. **Eyeglasses/spectacles**: All components of the hearing aid are in the arm of the glasses. They can transmit sounds by:
   - **Air conduction**: Air-conduction spectacles are practically obsolete.
   - **Bone conduction**: Bone-conduction spectacles contain the vibrator at the end of the arm and transmit the vibrations to the mastoid. They may be prescribed in cases of mild to moderate conductive loss and of mixed loss up to 35 dB. Note that a much better type of bone-conduction aid is the bone-anchored-hearing aid.
5. **Implantable hearing devices**: A conventional hearing aid takes sound and makes it louder. The amplified sound is conducted to the ear canal either via an ear mould or directly via the hearing aid.
   - A device that is semi-implantable (RetroX) has been classified by the FDA as a transcutaneous air-conduction hearing aid system (TACHAS). This is a conventional hearing aid, where the sound-transmitting silicon tube is placed from behind through the skin and cartilage of the auricle to direct the sound into the outer ear canal.
   - In the Vibrant Soundbridge implantable hearing system a tiny magnet (floating mass transducer, FMT) is directly attached to the ossicular chain (during surgery) and amplifies the natural vibrations of the ossicles. Many patients report that "direct" coupling leads to improved hearing quality and improved speech understanding. The system consists of external and internal parts. The external part, called the audio processor, is worn underneath the hair and held in place with a magnet. It contains a microphone, a battery and electronics. The audio processor converts environmental sounds into signals that are transmitted to the implanted internal coil of the Soundbridge. The implanted part consists of the internal coil, magnet, conductor link and the FMT. The signal from the audio processor is transmitted across the skin to the internal coil, which relays the signal down the conductor link to the FMT. The FMT is attached either to the incus or to the round window membrane (Fig. 1.2.5). The FMT converts the signal into vibrations that directly drive and move the ossicles or via the round window the peri-

lymph and amplify their natural movement. These vibrations then conduct the sound to the basilar membrane and the organ of Corti. This system is designed for mild to severe sensorineural hearing loss (FMT attached to the incus) or for moderate or severe mixed hearing loss (FMT attached to the round window), e. g. cochlear otosclerosis, malformations of the ear. Another middle ear implant is the middle ear transducer, an implantable hearing device where the sound is transmitted by a similar vibrating driving system attached to the head of the malleus.

– A bone-anchored-hearing aid is a hearing aid fixed to a bone-anchored titanium screw, in which bone conduction is used to transmit sound directly via the skull into the cochlea (Fig. 1.2.4).
– In deaf patients cochlear implants are used (see Sect. 1.6.12). Sound is transformed by the so-called speech processor to electric signals which are sent to a retroauricular subcutaneously implanted receiver. The receiver is connected with a stimulating electrode which is inserted into the cochlea.

1.6.13.3 Recommendations

It is well known that hearing impairment can negatively influence interpersonal relations and create difficulties in everyday tasks. The fitting of a hearing aid is necessary for patients who cannot benefit from pharmacological therapy and/or surgical procedures, and in some cases may be of support to the latter.

Criteria

Patients with a bilateral hearing loss with a loss in the better ear of at least 30 dB for at least one of the frequencies examined (from 0.5–3.0 kHz) and when the speech discrimination for monosyllabic words in the better ear is 80%. In cases of monolateral hearing loss, the loss should be 30 dB or more at 2.0 kHz or at two frequencies between 0.5 and 3.0 kHz. Make certain that the patient can properly use the hearing aids after a period of training with the hearing healthcare professional. The patient must also be motivated to use the hearing aid all the time. When deciding on hearing aids, the professional and the patient must evaluate subjective, social, cognitive and lifestyle aspects.

Recommendations for Binaural Fittings in Cases of Bilateral Hearing Loss

Stereophonic hearing is necessary for good speech discrimination, especially in noisy surroundings. If both ears can benefit from amplification, the rule today is to fit binaurally to guarantee the best result for speech discrimination (interpersonal communication). The patient must be motivated to correctly use both hearing aids.

Procedures for Fitting Hearing Aids

Three phases should be respected for an optimal result in fitting a hearing aid:

1. *Prescription*: In this phase the medical specialist is involved and he/she must carry out the testing necessary and an otomicroscopic objective examination. The testing includes subjective and objective tests; that is pure tone audiometry using earphones and free field. Impedance testing with particular attention to the stapedial reflex and in some cases, especially with children, the study of evoked potentials (auditory evoked brainstem responses).
2. *Fitting*: This is carried out by the audiologist/dispenser on the basis of the results from the testing and diagnosis. In this phase the hearing aid is chosen along with any assistive listening devices (if necessary) to satisfy the individual needs of the patient.
3. *Follow-up*: To obtain the maximum benefit from hearing aids it is necessary that the patient and audiologist/dispenser work together closely. After approximately 2 months, the plastic processes are completed. In this phase the medical and paramedical competences (audiologist/dispenser, ENT specialist, speech therapist, psychologist) converge to obtain the best result.

Choosing a Hearing Aid

Although hearing aids from a technological point of view rely on extremely sophisticated technologies, they are "obsolete" from the cosmetic point of view and this is usually the reason why many patients refuse hearing aids. For this reason hearing aids today have been restyled and special attention is given to their "design". If a hearing aid is to be accepted it should be perceived as a modern assistive device for communication, an extension of the patient's body, eliminating the sense of shame that the patient feels by wearing a hearing aid. There is a kind of tabu that is linked to dentures, hearing aids, cosmetics for men and in the past to eyeglasses. But today these negative connotations are decreasing and that changes the perception of the abovementioned items. They are no longer disturbing; they may become a part of fashion trends!

According to the audiological classification of hearing loss there is a distinction on the basis of the average tone threshold into *mild* (threshold between 20 and 40 dB), *moderate* (40–70 dB), *severe* (70–90 dB) and *profound* (90–120 dB) hearing loss. The choice of the hearing aid with respect to the above classification takes into account the audiometric curve (flat, symmetrical, asymmetrical,

downward slope, upward slope) and can be categorized as follows:

1. *Sensorineural hearing loss moderate (40–70 dB) to severe (70–90 dB)*
   - *Moderate hearing loss:* (1) digital CIC or ITC; (2) digital BTE with made-to-measure ear mould.
   - *Mild and moderate hearing loss downward slope:* digital BTE with open fitting, traditional (with tube), or RITE.
   - *Severe hearing loss:* digital BTE with custom ear mould.
2. *Postlingual profound hearing loss (above 90 dB) with normal linguistic ability*
   - Digital BTE aids with made-to-measure ear mould.
   - Alternatively, cochlear implant.
3. *Conductive and mixed hearing loss with normal tympanic membrane and external auditory canal*
   - Digital air-conduction hearing aid if the bone-conduction threshold is above 40 dB for the middle to high frequencies (in some cases).
   - Bone-conduction spectacles or better bone-anchored hearing aid if the bone-conduction threshold is more than 35 dB for middle to high frequencies, including 2.0 kHz.
4. *Conductive and mixed hearing loss, with problems related to application to the ear canal: bilateral agenesis of the external auditory canal; chronic bilateral otitis media; after tympanoplasty open or closed, radical cavity*
   - Implantable bone-conduction aids (bone-anchored hearing aids) in adults in some cases and in children (over the age of 6 years).
   - Bone-conduction vibrators mounted on a headband for a child.
   - Bone-conduction spectacles for adults.
5. *Prelingual profound hearing loss in adults who have never used hearing aids*. All hearing aids, including cochlear implants give unsatisfactory results.
6. *Prelingual profound hearing loss in adults who have always used analogue hearing aids*. In many cases digital hearing aids with custom-made ear moulds can be recommended and when possible cochlear implants.

The most frequent complaints that arise from patients using traditional hearing aids are the hearing aid whistles (feedback), loud sounds are uncomfortable, unsatisfactory speech discrimination in noisy surroundings and the perception of the person's own voice altered owing to the occlusion effect of the external auditory canal. To solve these problems today there are digital ITE and BTE hearing aids with artificial intelligence that use technologies capable of improving speech discrimination in noise, eliminating feedback and with the "open fitting" system the problems connected to the occlusion effect are resolved.

Hearing Aid Fitting in Children

Fitting hearing aids in children is difficult both for the diagnosis and in the actual fitting. It is essential that children are fitted with hearing aids at a very early stage (within 6–12 months). To obtain the best results, the family must be actively involved in the process, the child must be placed in an adequate scholastic and social environment and followed closely by a speech therapist. To evaluate the results of the fitting, objective audiometric testing (impedance testing, auditory evoked brainstem response, electrocochleography, otoacoustic emissions) is essential. The testing and the evaluation of the hearing aid fitting must be carried out in a medical environment with the cooperation of the audiologist/dispenser. When the child is in Kindergarten it is important to use a personal FM system to eliminate any interference from background noise present in classrooms. Most hearing-impaired children suffer from moderate hearing loss and in those cases a hearing aid is an adequate solution. In cases of profound hearing impairment, after approximately 6 months of hearing aid use and an accurate evaluation of the entity of the hearing loss and after a careful psychological evaluation and speech evaluation, the possibility of a cochlear implant may be considered and must be carried out before the child is 18 months old.

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1.6.14 Vestibular Neuritis

KARL-FRIEDRICH HAMANN

1.6.14.1 Synonyms

Vestibular neuropathy, "vestibular neuronitis" (wrong term, because one neuron cannot be inflamed).

1.6.14.2 Definition

Acute unilateral, partial or complete loss of peripheral vestibular function, caused probably by a viral inflammation.

1.6.14.3 Aetiology/Epidemiology

Current findings point to a viral origin (herpes simplex virus) similar to facial palsy. Owing to immunologic deficiencies, herpes simplex viruses, which were already present in the patient as a result of an earlier infection, are reactivated and destroy vestibular sensory fibres. Vestibular neuritis is one of the most frequent peripheral vestibular disorders (about 25% of vertigo patients seen in an ENT vertigo unit suffer from this disease).

1.6.14.4 Symptoms

The symptoms are marked by the acute appearance of severe vertigo, mostly purely rotatory, sometimes accompanied by vomiting, nausea and ataxia. In the acute state a violent horizontal-rotatory nystagmus, beating towards the intact side, is always present. Hearing impairment or tinnitus do not belong to vestibular neuritis.

1.6.14.5 Complications

In the acute state a tendency to fall is obvious, so a prevention against falls is necessary, to avoid orthopaedic sequelae. Not seldom benign paroxysmal positioning vertigo follows a vestibular neuritis within a short delay, this is named "*Lindsay–Hemenway syndrome*".

1.6.14.6 Diagnostic Procedures

A careful questionnaire reveals the sudden onset of the vertiginous complaints, which decrease within a period of some days. The only objective sign is a strong horizontal-rotatory nystagmus, beating to the intact side. The caloric test proves the hypofunction of the lesioned side, which is in the beginning not compensated in the rotatory test. Vestibular spinal tests show a marked deviation and a tendency to falls directed to the lesioned side.

1.6.14.7 Additional Useful Diagnostic Procedures

A lesion in the auditory system, which does not belong to the vestibular neuritis, can be excluded by audiological tests. A vestibular schwannoma, which in very rare cases is mimicked by the same symptoms, can be excluded by MRI.

1.6.14.8 Therapy

Conservative Treatment

Treatment is exclusively conservative. In the early stage of the disease sedating drugs such as H1 antagonists (50 mg dimenhydrinate 2–3 times a day) are recommended, but only for a short time, not longer than 2 days. As soon as possible vestibular habituation training should be started to induce a rapid vestibular compensation.

Corticosteroid treatment is recommended during the first 14 days (start with 500 mg intravenously with decreasing doses to 0 mg within 10 days).

Additional Useful Therapeutic Options

To accelerate vestibular compensation active movements (sports), stimulating agents such as caffeine and avoidance of calming procedures such as bed rest seem useful therapeutic options.

1.6.14.9 Differential Diagnosis

Differential diagnosis of vestibular neuritis is simple, because the duration of vertigo for some days is very characteristic; therefore, benign paroxysmal positioning vertigo, vestibular paroxysmia, Ménière's disease or vestibular migraine can be excluded by a careful questionnaire. The fact that vestibular neuritis is monosymptomatic facilitates the differential diagnosis in patients with additional hearing problems. Although vestibular schwannoma only very seldom becomes apparent by vertigo complaints, MRI allows a clear differential diagnosis.

1.6.14.10 Prognosis

The prognosis is generally favourable. If there are additional factors which can inhibit vestibular compensation, such as sedating drugs, old age or additional abnormalities in the CNS, the complaints can continue for a long time.

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3. Hamann KF (1987) Training gegen Schwindel. Springer
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1.6.15 Benign Paroxysmal Positioning Vertigo

KARL-FRIEDRICH HAMANN

1.6.15.1 Synonym

Benign paroxysmal positional vertigo.

1.6.15.2 Definition

Benign paroxysmal positioning vertigo (BPPV) is a mechanically induced vertigo caused by a canalolithiasis or a cupulolithiasis.

1.6.15.3 Aetiology/Epidemiology

Normally otoliths are fixed in the otolithic membrane. By head traumatism, in old age or idiopathically, otoliths can be loosened and travel in one or some of the semicircular canals. BPPV is one of the most frequent kinds of vertigo, mainly in the elderly. The different semicircular canals are not affected equally. In 96% of cases the posterior vertical canal is concerned, the horizontal canal in 3% of cases and the anterior vertical semicircular canal only in 1% of cases.

1.6.15.4 Symptoms

The vertigo attacks have a short duration, only a few seconds, typically triggered by certain head movements, for example by head turning in the morning for a look at the alarm clock. BPPV never occurs when the head is not moved.

1.6.15.5 Complications

Complications in the real sense of the word do not exist. As for all forms of vertigo, falls can occur.

1.6.15.6 Diagnostic Procedures

The questionnaire reveals that vertigo appears only during head movements and lasts only for some seconds. Apart from the characteristic complaints, the diagnosis is made by nystagmus observation under Frenzel's glasses. By specific positioning of the head in the plane of one of the semicircular canals (Hallpike manoeuvre), one can prove a BPPV if a typical nystagmus appears. The involved semicircular canal can be identified by analysis of the nystagmus, because the pattern of eye movements for each semicircular canal is known. Other neurotological tests such as caloric or rotatory tests do not show pathological findings; the auditory system is not involved as well.

1.6.15.7 Additional Useful Diagnostic Procedures

Additional diagnostic procedures are not necessary. Imaging techniques are only suitable for exclusion of possible central abnormalities.

1.6.15.8 Therapy

Conservative Treatment

Owing to the mechanical pathophysiologic nature of BPPV only a mechanical treatment is reasonable. The goal of a rational treatment of BPPV is to liberate the semicircular canals from the dislocated otoliths. This can be carried out by liberatory manoeuvres of Semont [2] (Fig. 1.6.14) or Epley [1] (Fig. 1.6.15). Both have principally the same intention, namely to bring the dislocated otoliths by specific movements of the head to a "neutral point" in the vestibular apparatus nearby the utriculus. For the treatment of a canalolithiasis or a cupulolithiasis of the horizontal canal, a barbecue rotation or Brandt–Daroff exercises can be recommended as for a prophylaxis of BPPV.

Surgical Treatment

Only in extremely rare cases (less than 1%) surgical treatment can be indicated. Two procedures exist: (1) neurectomy of the posterior canal nerve, (2) plugging of the affected canal.

1.6.15.9 Differential Diagnosis

Because of the typical clinical signs (vertigo only in combination with head movements, duration of vertigo never more than 60 s, provocation of typical nystagmus by specific positioning), normally the differential diagnosis does not present a problem. One of the rare differential-

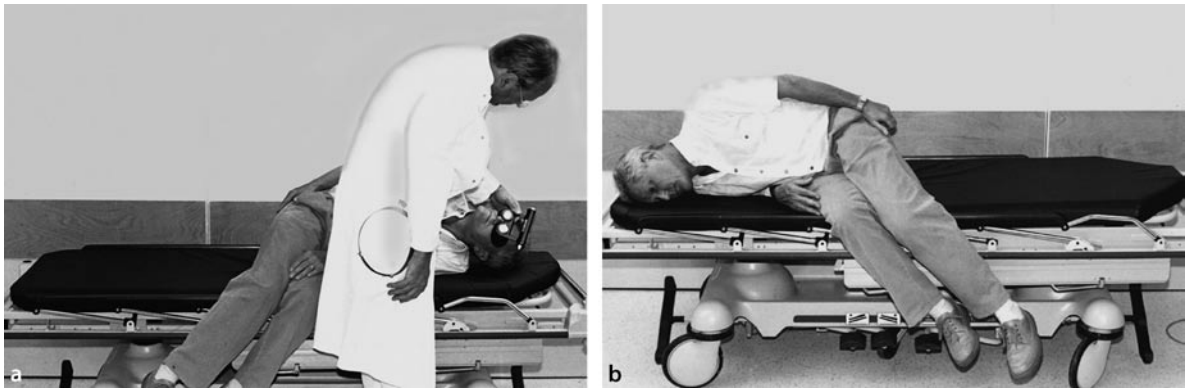

Fig. 1.6.14 **a** First movement of a liberatory manoeuvre (Semont) for treatment of a canalolithiasis of the left posterior canal: starting from a sitting position the patient is positioned to the left side, the head turned 45° to the unaffected right side. **b** Second movement of the liberatory manoeuvre (Semont): the head and trunk of the patient were thrown from the left side to the right side without changing the position of the head relative to the body

diagnostic possibilities is the vestibular paroxysmia, which is characterized by vertigo attacks of a few seconds. But these attacks are not correlated typically with certain head movements which trigger the BPPV.

Another differential diagnosis, but less frequent, is the possibility of a vestibular migraine, which, in contrast to BPPV, should be accompanied by headaches.

1.6.15.10 Prognosis

The prognosis of BPPV is very favourable. After one liberatory manoeuvre about 70% of patients no longer have complaints. In the remaining 30%, repetitive liberatory manoeuvres lead to a complete cure. If necessary, Brandt–Daroff exercises must be continued. The rate of recurrences is relatively high. In a period of 2 years after a liberatory manoeuvre, about 20% of patients complain about vertigo again; in a period of 8 years, the rate of recurrences reaches 55%. Only in extremely rare cases does a surgical treatment become necessary (see Sect. 1.6.15.8).

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1.6.16 Motion Sickness

KARL-FRIEDRICH HAMANN

1.6.16.1 Synonyms

Corresponding to the environment: car sickness, sea sickness, space sickness.

1.6.16.2 Definition

Motion sickness is a special kind of physiological vertigo, induced by an unusual stimulation of the multisensory system, which normally guarantees adequate orientation in space.

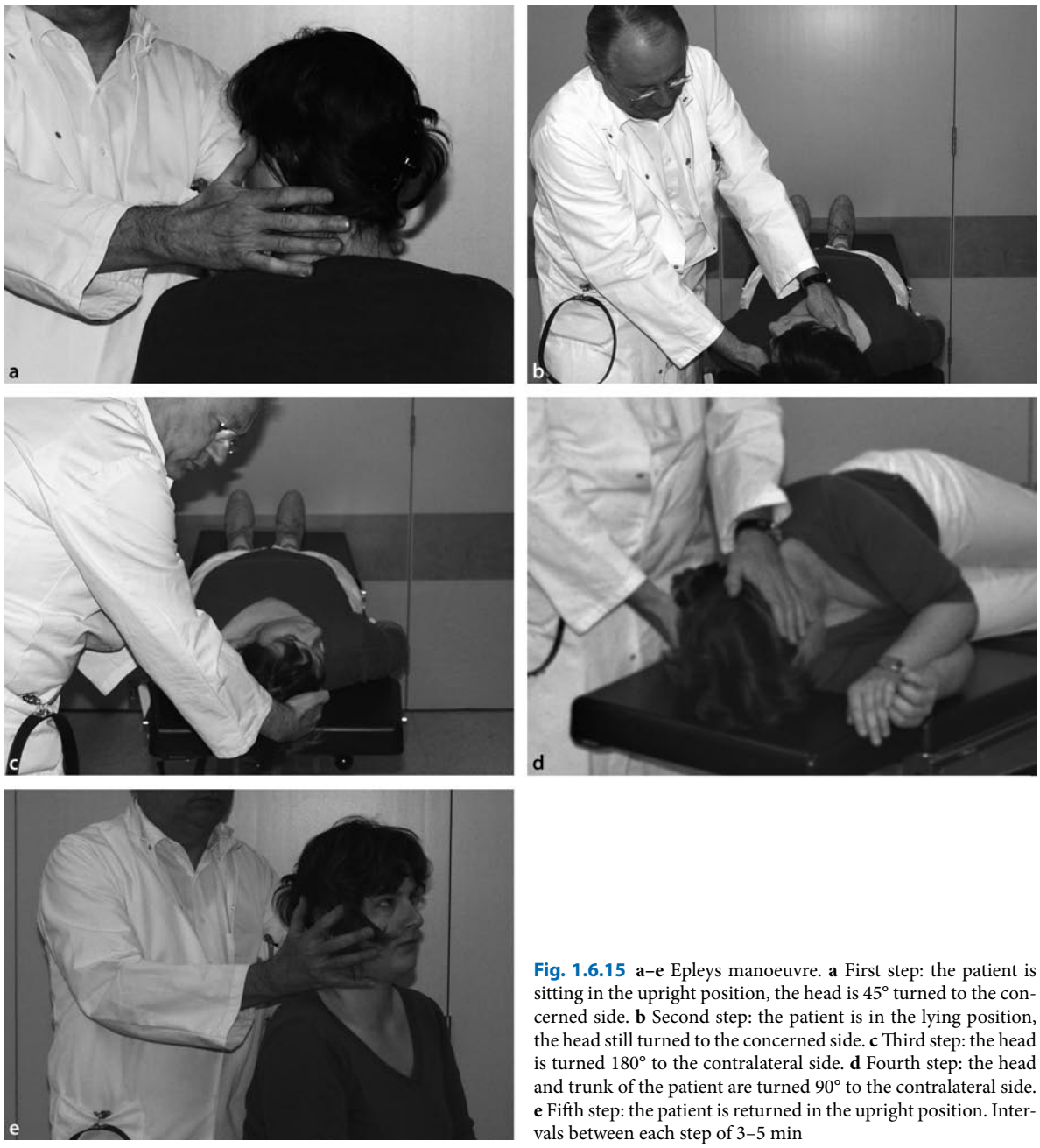

Fig. 1.6.15 **a–e** Epleys manoeuvre. **a** First step: the patient is sitting in the upright position, the head is 45° turned to the concerned side. **b** Second step: the patient is in the lying position, the head still turned to the concerned side. **c** Third step: the head is turned 180° to the contralateral side. **d** Fourth step: the head and trunk of the patient are turned 90° to the contralateral side. **e** Fifth step: the patient is returned in the upright position. Intervals between each step of 3–5 min

1.6.16.3 Aetiology/Epidemiology

It is generally accepted that motion sickness develops when a sensory conflict between the different sensory systems, responsible for the orientation in space, occurs. Both intrasensory mismatch (within the vestibular system) and intersensory mismatch (between the vestibular and visual system for example) can trigger the symptoms of motion sickness. The crucial condition is that different sensory receptors give different signals about the passive motion of an individual in space. So the perceived pieces of information do not correspond amongst themselves nor to the expected perception pattern, previously adapted by experience. Principally in all people with an intact sensory system motion, sickness can be induced, if certain conditions which can create a sensory conflict are fulfilled.

1.6.16.4 Symptoms

Motion sickness is clinically characterized by nausea, pallor, yawing, vomiting and mainly by a feeling of severe discomfort. These symptoms last not only for the time of conflict stimulation but also for a certain period afterwards, when motion stimulation had stopped.

1.6.16.5 Complications

Complications in the real sense of the word do not exist. Falls and aspiration caused by vomiting are sequelae of the symptoms themselves.

1.6.16.6 Diagnosis

Motion sickness is diagnosed very simply, because the coincidence of the inducing motion and the typical symptoms is pathognomonic. Further diagnostic procedures are not necessary.

1.6.16.7 Therapy

A prophylactic therapy can be useful if it is predictable that a motion sickness inducing sensory conflict will occur. The best prevention consists in a vestibular habituation training with the intention to prepare the orientation system for a conflict stimulation. As a medical prophylactic treatment, scopolamine used in the form of a transdermal skin patch can be recommended. When the symptoms of motion sickness appear, one can try to break free from the sensory conflict situation. In the case of seasickness the suffering person has to leave the cabin and should go on the ship's deck. Then he/she should fixate on an object not too far away. In this way there the correspondence between visual and vestibular information increases. The medical treatment consists in the uptake of an H1 histamine antagonist such as dimenhydrinate or meclozine. It must be pointed out that all H1 antagonists have sedative side effects. Interestingly, ginger root, given in a pulverized form, has significantly favourable effects on motion sickness symptoms.

1.6.16.8 Differential Diagnosis

A differential diagnosis of motion sickness does not exist. The only exception is that a real vestibular disease can be triggered also by a specific movement or a sensory conflict.

1.6.16.9 Prognosis

The prognosis of motion sickness is very favourable. At the latest in the moment when the inducing motion ceases, a rapid decrease of the uncomfortable symptoms begins. After some hours, the symptoms of motion sickness disappear.

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