1.1 Basics

SALVATORE IURATO

1.1.1 Clinical Anatomy

SALVATORE IURATO

1.1.1.1 External Ear

The external ear includes the auricle and the external auditory canal (Fig. 1.1.1):
- The shape of the auricle is quite complex and is determined by the shape of the aural cartilage.
- The external auditory canal (meatus) measures approximately 2.5 cm in length and is about 9 mm high by 6.5 mm wide. The lateral third consists of the elastic cartilage of the auricle (Fig. 1.1.1). Superiorly the cartilage is lacking between the tragus and the helical crus. This incisura terminalis is used by the surgeon when making an extracartilaginous endaural incision (see Sect. 1.4.6.8) without cutting into the cartilage. The fissures of Santorini in the cartilage of the external auditory canal (Fig. 1.1.1) are a potential route for the spreading of infection to the infratemporal fossa and

Fig. 1.1.1 Coronal view of the temporal bone (Modified from Brödel [2])
1.1 Basics

The medial third of the external auditory canal is osseous (Fig. 1.1.1).

The narrowest part of the external auditory canal (*isthmus*) is located between the fibrocartilaginous and the bony canal. The skin of the fibrocartilaginous canal is bound to the perichondrium without a subcutaneous layer. In the osseous part the skin is much thinner and closely adherent to the periosteum, and is devoid of hair follicles and ceruminous glands, whereas these are present in the cartilaginous part. Therefore, furuncles occur only in the cartilaginous meatus. Owing to its thinness, the skin of the osseous canal is easily traumatized during manipulations (e.g. wax removal with cotton tips).

The sensory innervation of the external ear comes from the greater auricular nerve (C³), lesser occipital nerves (C² and C³), the auricular branch (Arnold’s) of the vagus nerve (X) and the auriculotemporal nerve (V). For local anaesthesia, firstly infiltrate the skin, the soft tissues and the periosteum of the lateral surface of the mastoid, up to incisura terminalis, then the posterior, superior and inferior walls of the canal and, finally, the anterior wall.

1.1.1.2 Middle Ear and Mastoid

**Tympanic Membrane**

- Elliptical in shape, slightly conical like a loudspeaker, pale grey in colour, the tympanic membrane forms an acute angle with the inferior wall of the auditory canal (Fig. 1.1.1). This angle should be respected in myringoplastic procedures (blunting strongly reduces the vibratory capacity of the tympanic membrane).
- Landmarks visible at otoscopy (Fig. 1.1.2): the lateral or short process and the handle of the malleus, the pars flaccida (Shrapnell’s), and the pars tensa with the umbo and the triangular cone of light (*light reflex*). In the thin, transparent tympanic membrane: the chorda tympani, the long process of the incus with its lenticular process and its articulation with the head of the stapes (Figs. 1.1.1, 1.1.2).
- Histologically, the tympanic membrane is formed by an outer epidermal layer which is continuous with the skin of the external auditory canal, a fibrous middle layer (radial and circular fibres) and an inner mucosal layer which is continuous with the mucosal layer lining the middle ear cavity.
- The fibrous layer thickens peripherally to form the annulus (Fig. 1.1.2), which is inserted in a bony groove (*tympanic sulcus*). The fibrous layer is missing in the pars flaccida, which does not have the annulus.
- **Innervation:** of the anterior part, from the auriculotemporal branch of the V nerve; of the posterior part, from the auricular branch (Arnold’s) of the X
nerve; of the inner surface, from the tympanic branch (Jacobson’s) of the IX nerve.

**Surgical Landmarks**

1. On the lateral surface of the temporal bone
   - Suprameatal spine of Henle
   - Cribriform area
   - Posterior root of the zygomatic process and temporal line
   - Tympanomastoid suture
2. On the superior surface of the temporal bone (middle fossa approach)
   - Arcuate eminence (superior semicircular canal) (Fig.1.1.1).
   - Facial hiatus and greater petrosal nerve
   - Meatal plane
   - Superior petrosal sinus

**Middle Ear Ossicles**
- The average height of the stapes is 3.26 mm.
- The average size of the stapes footplate is 2.99 mm × 1.41 mm.
- The approximate distance (Fig. 1.1.3) between the stapes footplate and the utricle is 2.0–3.0 mm and the stapes footplate and the saccule is 1.0–1.5 mm.
- The weakest part of the ossicular chain is the long process of the incus at the level of its lenticular process, which articulates with the head of the stapes (Fig. 1.1.1).

**Middle Ear Muscles**
- Tensor tympani, innervated by a branch of the mandibular nerve (V)
- Stapedius muscle, innervated by a branch of the VII nerve

**Other Important Anatomical Features of the Middle Ear**
- Notch of Rivinus where the tympanic sulcus and the annulus tympanicus are both absent.
- Promontory (corresponding to the basal turn of the cochlea, Fig. 1.1.1) with the tympanic nerve (Jacobson’s; Fig. 1.4.8c), which arises from the inferior ganglion

![Fig. 1.1.3](image)  
*a* Average measurements in the middle and inner ear according to Anson and Donaldson [1]. Tympanic membrane – footplate = 8.0 mm; Tympanic membrane – facial nerve (FN) = 5.0 mm; Malleus handle – long process of incus = 2.0 mm; Footplate – utricle (U) = 3.0 mm; Footplate – saccule (S) = 2.0 mm; Stapes height = 3.26 mm; Footplate length = 2.99 mm; Footplate width = 1.41 mm.  
*b* Oval window surgical anatomy (right ear) in relation to stapedotomy calibrated hole 0.6 mm diameter
of the IX nerve and is joined by the caroticotympanic nerve.
- **Cochleariform process** (mind cholesteatoma matrix remnants!).
- **Oval window, ponticulus, sinus tympani, subiculum and round window** (Fig. 1.1.1).
- **Footplate of the stapes which articulates with the oval window by the annular ligament.**
- **Facial nerve in the Fallopian canal** (labyrinthine portion; first turn; tympanic portion with its relation with the bony lateral semicircular canal and the stapes; second turn with its relation with the short process of the incus; vertical or mastoid portion). The facial nerve is dehiscent in its tympanic course (oval window region) in about 30% of individuals.
- **Round window membrane** which plays an important role in the transmission of acoustic energy to the inner ear, and as a site through which toxic substances and therapeutic principles may enter the inner ear.
- **Chorda tympani** containing sensory (taste) fibres from the geniculate ganglion, directed to the anterior two thirds of the tongue.
- **Epitympanic space** (Fig. 1.1.1) containing the head of the malleus and the body of the incus. Bone defects in the roof of the tympanic cavity (regmen tympani; Fig. 1.1.1) are present in about 35% of individuals. These bony dehiscences are sometimes associated with meningoencephaloceles.
- **Floor of the tympanic cavity (hypotympanum)** overlying the jugular bulb (Fig. 1.1.1) (individual variations).
- **Anterior wall** (protympanum) with (1) the semicanal of the tensor tympani, (2) the ostium of the Eustachian tube (Figs. 1.1.1, 1.4.8d) and (3) the vertical segment of the petrous carotid artery.
- **Anterior epitympanic recess** (area to be explored as cholesteatoma may penetrate and be hidden).
- **Posterior wall** with (1) the pyramidal eminence and stapedial tendon (Fig. 1.4.9a), (2) the chordal eminence and (3) the facial recess between the pyramidal eminence and the chordal eminence.

**Mastoid**
- Normal mastoids may be fully pneumatized, diploic or sclerotic. Pneumatization may extend (beyond the mastoid, perilabyrinthine and petrous apex regions) into the roof of the zygomatic process and into the squamous part of the temporal bone.
- **Mastoid cavities** (the main cavity is called the “antrum”) are lined with a very thin mucosa and communicate with the epitympanic space of the middle ear via the aditus ad antrum. Mastoid cavities play a role in pressure regulation and gas exchanges.

**Eustachian Tube**
- The bony part (11–14 mm) opens into the protympanum (tympanic ostium), and the fibrocartilaginous part (20–25 mm) opens into the lateral wall of the rhinopharynx (Fig. 1.1.1).
- Lined with respiratory ciliated mucosa, the tube connects the rhinopharynx with the middle ear and permits ventilation of the pneumatized temporal bone spaces. It opens on swallowing.
- **Blockage of the Eustachian tube** is responsible for otitis media with effusion, middle ear atelectasis, retraction pockets and failures in middle ear surgery.
- The **patulous tube syndrome** seems to be caused by a defect in the mucosal valve located within the cartilaginous portion of the Eustachian tube.

1.1.1.3 Inner Ear

**The Bony Labyrinth**
- The osseous portion (otic capsule, the densest bone in the body) consists of the vestibule, the three semicircular canals, the cochlea and the two aqueducts: the vestibular aqueduct, which contains the endolymphatic duct, and the cochlear aqueduct connecting the scala tympani in the basal turn with the subarachnoid space. A patent cochlear aqueduct (or small defects in the fundus of the internal auditory canal) is responsible for profuse outflow of the perilymph/cerebrospinal fluid from the oval window (“gusher”) in stapes surgery. A large vestibular aqueduct is the most common inner ear anomaly, and is associated with a congenital hearing impairment.
- The cochlea makes 2.5 turns around the modiolus and contains the **cochlear duct** (scala media).
- The three semicircular canals lie at right angles to each other. The lateral (external) semicircular canal forms a 30° angle with the horizontal plane.

**The Membranous Labyrinth**
- The membranous labyrinth consists of the cochlear duct, the saccule, the utricle and the membranous semicircular canals. The **cochlear duct** communicates with the saccule via the ductus reuniens. The utricular duct and the saccular duct form the endolymphatic duct, which ends in the endolymphatic sac in the posterior fossa.
- The ampullae of the semicircular canals contain sense organs (crotae ampullares). The sense organs contained in the utricle and saccule are called **maculae**. The cristae ampullares and the maculae contain the vestibular hair cells and their supporting cells.
- The cochlear duct (scala media) is separated from the **scala tympani** by the basilar membrane, which con-
nects the bony spiral lamina with the spiral ligament. *Reissner's membrane* separates the scala media from the *scala vestibuli*. The *organ of Corti* (Fig. 1.1.4) is situated on the basilar membrane and consists of the sensory (hair) cells and of the supporting cells (pillars, Deiters', Hensen's, inner and outer supporting cells).

**Endolymph and Perilymph**
The scala media contains endolymph (with a high potassium and a low sodium concentration) and has a positive resting potential of 80 mV (endocochlear potential), maintained by the stria vascularis. The scala tympani and scala vestibuli contain perilymph (with a low potassium and a high sodium concentration similar to that of extracellular fluid).

**Inner and Outer Hair Cells**
- There are 3,500 *inner hair cells* (IHCs) and 12,000 *outer hair cells* (OHCs).
- The stereocilia of the OHCs are firmly embedded in the tectorial membrane, those of the IHCs are only loosely connected to the tectorial membrane or have no connection at all (Fig. 1.1.4).
- The OHCs contain contractile proteins. They are able to shorten and lengthen like muscle cells in response to neural signals. This active mechanism plays an important role in excitation of the IHCs.

**Innervation: Peripheral**

**Vestibular Sensory Organs**
- Superior vestibular nerve: n. utricularis + superior and lateral ampullary nerves + saccular branch (Voit's)
- Inferior vestibular nerve: n. saccularis + posterior (singular) ampullary nerve

**Cochlea**
- **Afferent innervation**: About 35,000 ascending first-order neurons convey signals from the cochlea to the CNS. They are components of the *spiral ganglion* (*cochlear ganglion*), which is located in Rosenthal’s canal, spirally arranged in the modiolus, the central support of the bony cochlea; 90–95% of them (bipolar cochlear type I neurons) innervate the IHCs (each IHC is innervated by ten to 15 type I neurons); 5–10% of them (pseudomonopolar cochlear type II neurons) innervate the OHCs (each type II neuron innervates about ten OHCs) (Fig. 1.1.5).
1.1 Basics

- **Efferent innervation**: A much smaller population (approximately 1,600) of descending neurons sends signals from the CNS to the cochlea. Efferent fibres synapse directly with the OHCs but not with the IHCs. Most of the fibres from the lateral olivocochlear bundle (uncrossed efferents) synapse in the inner spiral bundle with the afferent fibres associated with the IHCs. Most of the fibres from the medial olivocochlear bundle (crossed efferents) synapse directly with the OHCs.

- **In summary**: The 3,500 IHCs have a mainly afferent innervation. The 12,000 OHCs have a mainly efferent innervation.

- **Adrenergic innervation**: A perivascular adrenergic plexus supplies the branches of the modiolar artery and a second adrenergic plexus, independent of the blood vessels, supplies the habenula perforata.

**Innervation: Central**

There is an increasing order of complexity from the cochlea/auditory nerve to the central auditory nervous system (Fig. 1.1.6).

- The cochlear nerve fibres (**first-order neurons**) leave the temporal bone through the internal auditory canal, pass through the cerebellopontine angle and enter the brainstem at the [ventral cochlear nucleus](#) or the [dorsal cochlear nucleus](#), where they synapse with the second-order neurons.

- Some fibres from **second-order neurons** extend to the superior olivary complex on the same side, but a majority cross to the contralateral side via the trapezoid body. Some of the fibres that cross over synapse in the contralateral superior olivary complex and some of them ascend to the contralateral lateral lemniscus. The superior olivary complex receives information from both ears (bilateral representation). The acoustic stapedial reflex and the aural-palpebral reflex mediate at this level.

- **Third-order neurons** arise from the superior olivary complex and lateral lemniscus. They may synapse at the inferior colliculus or terminate at the medial geniculate body of the thalamus like the fibres originating from the inferior colliculus.

- The medial geniculate body of the thalamus is the subcortical station in the auditory pathway. Fibres from the medial geniculate ascend along the auditory radiations to the auditory cortex. **Tonotopic organization** is present at each level of the auditory system from the cochlea up to the cortex.

**References**


Suggested Reading


1.1.2 Physiology

**Salvatore Iurato**

The external ear (auricle and external auditory canal) and the middle ear (tympanic membrane and ossicular chain) are called the “conductive system” because their function is to conduct the sound signals from the air to the inner ear. The term “sensorineural apparatus” comprises the hair cells (sensory) and the cochlear nerve (neural).

1.1.2.1 Auricle and External Auditory Canal

The auricle and external auditory canal collect, amplify and convey sound to the tympanic membrane.

1.1.2.2 Tympanic Membrane and Ossicular Chain

These structures efficiently transfer sound energy from the air to the inner ear fluids:

- **Hydraulic effect**: the area of the tympanic membrane is much larger than that of the footplate (17:1).

- **Lever effect**: the malleus handle is longer than the long process of the incus (1.3:1).

- The approximate total gain is 27.5 dB.

1.1.2.3 Oval and Round Windows

Sound waves reach the two windows in different phases. The round window is protected by the intact tympanic membrane and by the margin of the round window niche. Large perforations of the tympanic membrane and those exposing the round window are often associated with a
high degree of hearing loss. In surgical procedures, the round window membrane must be protected from the direct impact of sound.

1.1.2.4 Basilar Membrane and Travelling Waves

The basilar membrane is narrower and stiffer in the basal turn than in the middle and apical turns. The mechanical stimulation of the cochlea results from a travelling wave. The maximum displacement of the basilar membrane is related to the frequency of the stimulus: near the base of the cochlea for high-frequency tones, near the apex for low-frequency tones. This tonotopic organization is preserved along the auditory pathway from the cochlea to the auditory cortex.

1.1.2.5 Inner and Outer Hair Cells

- There are shearing forces between the tectorial membrane and the stereocilia of the hair cells. The hair cells respond when their stereocilia are bent towards the basal body (towards the lateral wall of the cochlear duct). There are cross-links (filaments) between the
stereocilia. Pulling of the filaments opens a pore which permits ions to flow (activation); bending of the filaments in the opposite direction closes the pore (inhibition).

- The inner hair cells (IHCs) are the mechanoelectrical transducers of the inner ear. They synapse with dendrites of cochlear type I neurons. Glutamate is accepted to be their main neurotransmitter.
- The outer hair cells (OHCs), with cilia connected to the tectorial membrane, receive neural signals from the efferent olivocochlear bundle. The efferent innervation controls the contractile activity of the OHCs (cochlear amplifier), which enhances the movements of the basilar membrane (travelling waves) that stimulate the IHCs. The contractile activity of the OHCs is responsible for the ability of the cochlea to produce sounds (otoacoustic emissions). The nerve endings of the efferent olivocochlear bundle have vesicles containing the neurotransmitter acetylcholine. The ability to hear soft sounds and fine frequency distinctions is dependent on the integrity of the OHCs; without the OHCs, the hearing threshold is raised by 40–50 dB and frequency discrimination deteriorates.
- The stria vascularis provides the high concentration of potassium necessary to maintain the endocochlear potential (80 mV).

1.1.2.6 Semicircular Canals, Utricle and Saccule

- The semicircular canals with their ampullae provide information about angular acceleration.
- The receptor organ contained in each ampulla is called the “crista”. It contains the vestibular hair cells, the supporting cells and a gelatinous mass called the “cupula”. The stereocilia of the vestibular sensory cells extend into the cupula and are bent by deflection of the cupula. Deflection of the cupula is caused by the flow of endolymph.
- Like the cochlear hair cells, the vestibular hair cells are polarized: when the stereocilia are bent towards the kinocilium the response is excitatory; it is inhibitory when they bend away from the kinocilium. In the lateral semicircular canal, bending towards the utricle is excitatory, bending away is inhibitory. The opposing arrangement of the kinocilium in the two vertical canals is responsible for their opposite response (excitatory away from the utricle, inhibitory towards the utricle).
- The maculae of the utricle and saccule are covered by the otolithic membrane, on the top of which there are calcium carbonate crystals called “otoliths”. They provide information about linear acceleration and the position of the head in space. Spontaneous de-generative changes of the otoliths, particularly in geriatric patients, or their traumatic displacement, are responsible for their release into the endolymph (see Sect. 1.6.15).
- The endolymphatic sac has a pressure-regulation role because of its capacity to absorb water. Its content is markedly hyperosmolar in comparison to the osmolality of the endolymph contained in the other parts of the membranous labyrinth. In patients with the large vestibular aqueduct syndrome pressure changes may cause reflux of hyperosmolar endolymph from the endolymphatic sac into the cochlear duct, and damage to the cochlear hair cells. The epithelium of the endolymphatic sac is metabolically active and plays an important role in the immunological defence of the inner ear. It has properties similar to those of the epithelium of the mucosa-associated lymphatic tissue system. According to whether antigen stimulation is systemic or local (inner ear), there is a cellular reaction as well as specific local antibody production.

Suggested Reading

1.1.3 Ear-related Questions

SALVATORE IURATO AND WOLFGANG ARNOLD

No otologic symptom alone is diagnostic and therefore the otologic history must include questions about hearing loss, earache, otorrhoea, tinnitus and dizziness, plus a careful medical history. “It has been said that if you will listen to what a patient says, he will give you the diagnosis” [1].

1.1.3.1 Hearing Loss

- Bilateral or unilateral, which side?
- Recent or long-standing?
- Sudden? Stable? Progressive? Fluctuating?
- Continuous or intermittent?
- Is it associated with autophony?
- Is the speech discrimination poor?
- Is there a family history of hearing loss?
- Is the onset associated with a specific event (e.g. noise exposure, head trauma or disease)?
- What is the patient’s occupation? Is there occupational or recreational (e.g. hunting, discotheque, rock concert, fireworks) noise exposure?
1.1.3 Ear-related Questions

- Is there a history of diseases such as meningitis, parotitis, measles or syphilis, or of high fever of unknown origin? Is there a history of head trauma or unconsciousness?
- Is there a history of ototoxic drugs (e.g. streptomycin, gentamicin, diuretics, quinine, cytostatic drugs)? Diabetes, hypertension?
- Is there a history of previous ear surgery?

1.1.3.2 Earache (Otalgia, Ear Pain)

See also the algorithm for earache in Fig. 1.1.7.
1. Bilateral or unilateral, which side?
2. Continuous or intermittent?
3. Is it associated with a discharge and/or movement of the pinna, pressure on the tragus?
4. Is it associated with swimming or diving?
5. Is it associated with a recent infection of the upper respiratory airways?
6. In normal otoscopic findings (referred ear pain)
   - Dental problems, dentition difficulties
   - Irritation of the temporomandibular joint (temporomandibular joint syndrome)
   - Cervicofacial syndrome (patients with a history of cervical trauma or elderly patients with cervical arthritis)
   - Neuralgia of the auriculotemporal branch of the trigeminal nerve (diagnosis of exclusion), neuralgia of the auricular branch of the vagus nerve (Arnold’s) in acute tonsillitis or following tonsillectomy, neuralgia of the tympanic branch of the glossopharyngeal nerve (Jacobson’s) in oropharyngeal carcinoma
   - Aerodigestive malignancies (detectable with an accurate endoscopic examination in cases of mucosal lesions and with X-rays in cases of submucosal lesions), neoplasia of the infratemporal fossa (MRI with gadolinium)
   - Elongated styloid process syndrome (Eagle): irritation of the glossopharyngeal nerve

1.1.3.3 Otorrhoea (Aural Discharge)

See also the algorithm for otorrhoea in Fig. 1.1.8.
- Bilateral or unilateral, which side?
- Character (mucous, mucopurulent, purulent, bloody, foul, clear)

- Haematoma
- Trauma
- Laceration
- Furuncle
- Acute diffuse otitis externa (severe earache)
- Otomycosis
- Herpes zoster oticus
- External canal neoplasm
- Malignant external otitis
- Cerumen impaction
- Foreign body
- Bullous myringitis
- Traumatic perforation
- Aerotitis
- Haemotympanum
- Acute otitis media
- Serous otitis media (mild earache)
- Dental problems
- TMJ syndrome
- Acute tonsillitis
- Peritonsillar abscess
- Post-tonsillectomy pain
- Cervicofacial syndrome
- Myofascial pain dysfunction
- Mucosal aerodigestive malignancies
- Elongated styloid process syndrome (Eagle)
- Aerodigestive submucosal malignancies
- Intracranial malignancies
- Occult carcinoma of rhinopharynx
- Infrate mporal fossa malignancies
- Primary neuralgia (diagnosis of exclusion)

Fig. 1.1.7 Earache. Basic diagnostic procedures: case history, otoscopic examination, complete head and neck examination including endoscopy, dental and temporomandibular joint (TMJ) evaluation.
1.1 Basics

• Continuous or intermittent?
• Recent or long-standing?
• Is the onset associated with a specific event (e.g. swimming or diving or an episode of upper respiratory airways infection?)

1.1.3.4 Tinnitus (Head Noise)

See also the algorithm for tinnitus in Fig. 1.1.9.
• Bilateral or unilateral, which side?
• Time of onset (recent or chronic)
• Continuous or intermittent, pulsating?
• Character of the tinnitus (low-pitched like the noise of the sea, high-pitched like a whistle)
• Is the onset associated with a specific event or noise exposure? With ototoxic drugs (aspirin, aminoglycoside antibiotics, quinine)?

1.1.3.5 Dizziness/Vertigo

See also the algorithm for vertigo in Fig. 1.1.10.
Vertigo of otologic origin implies a sensation of motion (spinning), is usually accompanied by rhythmic eye movements (nystagmus) and may be due to peripheral or central vestibular disorders. If peripheral in origin, it causes episodic attacks with normal equilibrium between the spells. The episodic attacks may last from a few seconds, as in the case of benign paroxysmal vertigo, to some hours, as in the case of Ménière’s disease. Central non/vestibular dizziness begins insidiously and lasts longer. It is essential to collect a detailed clinical history:
• Is the dizziness continuous (often described as unsteadiness) or does the patient experience the walls of the room whirling around him/her only during the attacks?
• What is the interval between the spells?
• Does head motion incite the attacks of vertigo?
• Are there accompanying symptoms such as nausea and vomiting?
• Are there associated cochlear symptoms such as unilateral hearing loss, hearing fluctuation, ear fullness and/or tinnitus?
• Is vertigo and/or oscillopsia evoked by sounds of high intensity (Tullio’s phenomenon) or tragal compression (Hennebert’s sign) or the Valsalva manoeuvre?
• Are there ocular symptoms such as double vision, or neurological symptoms such as numbness of the face or extremities, altered consciousness, difficult swallowing, speech problems, loss of memory?
• Is there a history of chronic ear infection?
• Is there a history of previous ear operations?
• Is there a history of disorders such as diabetes, syphilis, arteriosclerosis, cardiac problems, head trauma, and flulike symptoms?

References

![Character of the aural discharge](image)

**Fig. 1.1.8** Otorrhoea. Basic diagnostic procedures: case history, otoscopic examination, aspiration, rough hearing assessment with tuning fork tests, pure-tone audiometry
1.1.3 Ear-related Questions

- Acute otitis media
- Cholesteatoma
- Labyrinthine fistula
- Otosclerosis
- Superior canal dehiscence syndrome
- Ménière's disease
- Sudden hearing loss
- Tumour
- Vascular loop
- MS
- Orthostatic dizziness
- Cardiac dizziness
- Central origin
- Endocrine origin
- Psychogenic origin
- Benign paroxysmal vertigo
- Central vestibular defect
- Suspected defect
- Migraine
- Palatal myoclonus (objective tinnitus)
- Benign intracranial hypertension (objective pulsatile tinnitus)
- High blood pressure

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Fig. 1.1.9 Tinnitus. Basic diagnostic procedures: case history, ENT examination, otomicroscopy, hearing evaluation (pure-tone audiogram, tympanometry, stapedial reflex), tinnitus measurement, masking tests of tinnitus, blood pressure. ABR auditory brainstem response

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Fig. 1.1.10 Vertigo. Basic diagnostic procedures: case history, ENT examination, hearing evaluation (pure-tone audiogram, speech perception test, tympanometry, stapedial reflex), vestibular examination (nystagmus spontaneous, after provocation, vestibulospinal reflex, fistula symptoms, caloric testing), functional testing of the cervical spine, blood pressure, and blood sugar
1.1 Basics

1.1.4 Principles of Clinical Examination

Salvatore Iurato

1.1.4.1 Inspection and Palpation

Gross inspection and digital palpation of the external ear and surrounding structures may reveal the presence of inflammatory processes, congenital malformations and neoplastic diseases.

Otoscopy

See also the algorithm for otoscopy in Fig. 1.1.11.

The *electric otoscope* (magnification ×2.5) is adequate for a routine otolaryngological examination (patient sitting) but when an otologic disease is suspected, examination at ×6–10 with the binocular *operation microscope* (patient supine) is recommended.

1. The largest speculum which can be inserted into the canal without pain should be used.
2. For a right-handed examiner using the microscope, the ear speculum is held between the thumb and forefinger of the left hand, with the ring finger in the concha, pulling the external ear posteriorly (conveniently straightening the external auditory canal) and the little finger on the patient's head to stabilize the speculum. The right hand is free to move the microscope and to manipulate instruments and exert suction.
3. The external auditory canal is inspected for wax, foreign bodies, mycosis, secretions, granulations, exostoses, growths and ulcerations. The quality of the skin should be noted.
4. Often only a portion of the tympanic membrane is visible at one time. The speculum and the microscope should be adjusted to make a mental reconstruction of the entire tympanic membrane. To facilitate orientation, the non-expert otologist should begin by identifying the short process of the malleus (Fig. 1.1.2).
5. Through a thin tympanic membrane, the long process of the incus may be seen posteriorly as a shadow (Fig. 1.1.2), as may the round window, although this is less common. Anteriorly, the opening of the Eustachian tube may be seen by transparency. Also the chorda tympani may be seen in the upper posterior part.
6. An intact pars tensa does not necessarily mean a healthy middle ear.
7. Systemic inspection must be made of the tympanic membrane both in the pars tensa and in the pars flaccida for:
   - Variations in shape (see light reflex), retraction pockets, atelectasis (the result of chronic dysfunction of the Eustachian tube). Distinguish between stable (self-cleaning) and unstable retraction pockets.
   - Perforations (central, marginal, of the pars flaccida).
   - Secretion: clear, purulent, mucopurulent, bloody, foul. Fetid secretion indicates cholesteatoma or osteitis, odourless secretion indicates chronic otitis media without cholesteatoma but odourless secretion turns fetid if retained.
   - Crusting: dry secretion masking perforations, retraction pockets, osteitis, cholesteatoma. The crusts can be removed with a small round ear hook to reveal the underlying disease.
   - Middle ear mucosa (in cases of perforation): normal, with granulations or polyps, with keratin.
   - Cholesteatoma: marginal perforations, bony erosions, attic perforations.
   - Oscillar disease through a perforation.
8. The 0°, 4 mm *tele-otoscope* enables the observer to photograph the external canal and the whole tympanic membrane with excellent optical resolution (Fig. 1.1.2). Good-quality TV pictures may also be obtained for documentation and/or teaching purposes.
9. The 30° and the 70°, 1.9 mm *tele-otoscopes* enable the observer to examine the retrotympanum through a perforation. Warning: take care not to damage the stapes while using the 70° otoscope!
10. Pneumatic *otoscopy*, increasing and decreasing the pressure in the external ear canal with the pneumatic Siegle speculum with plain glass to:
   - Evaluate the mobility of the tympanic membrane and malleus
   - Identify middle ear effusions
   - Identify small perforations
   - Distinguish between fixed and mobile retraction pockets
11. Paper patch test: Under microscopic control, place a wet paper patch over the perforation and repeat the audiogram. An improvement of hearing with closure of the air–bone gap means that tympanic membrane perforation is the only problem. If after application of the patch the hearing is the same or worse, the problem involves the ossicular chain.
12. Stapedial reflex: Mobility of the stapes can be evaluated directly under the microscope when the stapes is visible through a perforation, by stimulating the opposite ear with a Barany noise box (crossed stapedial reflex).
13. Tonsbee's manoeuvre: The patient is asked to swallow with the nose firmly closed: an inwards movement of the posterior half of the tympanic membrane (ob-
1.1.4 Principles of Clinical Examination

### 1.1.4.2 Fistula Test

A Politzer bag is used to apply pressure to the ear canal. In the case of a bony fistula, pressure changes are transmitted through the fistula to the labyrinthine fluids. A positive fistula test is associated with deviation of the eyes towards the contralateral ear, followed by nystagmus towards the involved ear if the pressure is held. If the pressure is released, the eyes return towards the midline. In fistula of the lateral semicircular canal (the most common), nystagmus is horizontal, whereas it is rotatory if the fistula is present in the superior canal and vertical if it is in the posterior canal. A positive response in the presence of an intact tympanic membrane (**Hennebert’s sign**), originally described in congenital syphilis, may be seen in perilymphatic fistula, in labyrinthine hydrops and in the superior semicircular canal dehiscence syndrome.

- **False-positive fistula test result:** owing to an abnormally mobile stapes.
- **False-negative fistula test result:** owing to loss of labyrinthine function.

### 1.1.4.3 Tube Inflation

Tube inflation can be carried out (1) by Valsalva manoeuvre (autoinflation) or (2) by politzerization with a Politzer bag or (3) by using a catheter to:

- Evaluate a retraction pocket (fixed or mobile?) or a tympanic membrane atelectasis
- Detect a tiny perforation
- Confirm the presence of a serous effusion in the middle ear

### 1.1.4.4 Patulous Eustachian Tube

Synchronous movements of the tympanic membrane with respiration may be observed under the microscope. Otoscopy should be carried out with the patient in the sitting position, during forced inspiration and expiration through the nose while occluding the contralateral nostril.

---

**Otoscopy**

**Interpretation of the otoscopic picture**

<table>
<thead>
<tr>
<th>Colour</th>
<th>Perforation/s</th>
<th>Other changes</th>
</tr>
</thead>
</table>
| • White chalky patches: Tympanosclerosis  
• Yellowish/golden: OME  
• Pink (Schwartz’s sign): Active otosclerotic focus  
• Whitish underneath: Congenital cholesteatoma  
• Blue: Haemotympanum  
Barotrauma  
Cholesterol granuloma  
High jugular bulb  
• Red: Glomus tumour  
Ectopic ICA | • Central “safe”: COM  
Trauma  
Previous AOM  
• Marginal “unsafe”: Pars tensa cholesteatoma  
Attic: Pars flacida cholesteatoma  
• Multiple: Recurrent otitis media  
Tuberculosis | • Haemorrhagic blisters: Acute bullous myringitis  
Herpes zoster oticus  
• Granular aspect: Chronic myringitis  
• Dry cast: Recent AOM  
• Vascular changes, redness: AOM (earliest sign)  
• Bulging, *Pis de vache*: AOM (stage of impending suppuration)  
• Retraction (partial or complete):  
Atelectasis  
Adhesive otitis media  
Retraction pockets  
Potential cholesteatoma  
• Polyp/s: Inflammatory (COM)  
Cholesteatoma  
Squamous cell carcinoma  
• Keratinising squamous epithelium  
Cholesteatoma  
• Air-fluid level/air bubbles:  
Serous otitis media  
Barotrauma  
• False fundus: postinflammatory EAC stenosis |

---

**Fig. 1.1.11** Otoscopy. **AOM** acute otitis media, **ICA** internal carotid artery, **COM** chronic otitis media, **OME** otitis media with effusion (serotympanum, glue ear), **EAC** external auditory canal

- **Baumann’s line:** serves with the microscope) means that the Eustachian tube opens regularly.

---
1.1 Basics

Comment: The tuning fork tests are not an optional but a standard part of the otologic evaluation. The results must be in agreement with those of the pure-tone audiometry.

Basic Audiologic Evaluation
See also the algorithms for hearing loss and steps in audiolologic evaluation in Figs. 1.1.12 and 1.1.13.

Pure-tone Audiometry
- Determination of the air-conduction and bone-conduction thresholds at frequencies from 125 to 8,000 Hz, beginning with 1,000 Hz.
- The patient responds by raising an index finger or lighting a signal light when she/he hears the tone.
- The bone-conduction vibrator should be applied with a force of 400 g to the midline of the forehead or to the mastoid. At low frequencies, more energy (10–15 dB) is needed in frontal placement to reach the threshold as compared with mastoid placement.
- The hearing thresholds obtained with an audiometer are reported in decibels and registered on a graph (audiogram).
- The air-conduction pure-tone average is the average of the hearing levels at the frequencies 500, 1,000 and 2,000 Hz.

Masking
- It is essential to avoid interference of the opposite ear. For air-conduction testing using standard earphones, masking should be applied to the opposite ear whenever the threshold of the test ear exceeds the threshold of the non-test ear by 40 dB (value of interaural attenuation for air conduction) or more. For bone-conduction testing, interaural attenuation practically does not exist; therefore, during bone-conduction testing, masking should always be applied to the non-test ear. There is no need to mask when no air-bone gap is present in either ear and when the Weber test is lateralized to the test ear. The amount of effective masking should be evaluated to avoid both undermasking and overmasking. The masking rules apply for both pure-tone and speech audiometry.

Interpretation
- Air conduction tests the entire hearing system. Bone conduction tests only the inner ear and auditory nerve, bypassing the conductive mechanism. Comparing the air-conduction with the bone-conduction thresholds allows us to differentiate conductive (problem located in the external auditory canal or middle ear), sensorineural (problem located in the cochlea or auditory nerve [retrocochlear lesion]) and mixed hearing loss. A difference between the air-conduction and air-conduction thresholds (A/B gap) implies that there is a problem with the conductive mechanism (Figs. 1.1.14, 1.1.15). When the sensorineural part accounts for the total amount of loss (bone conduction is

1.1.5 Technical Diagnostic Procedures

Salvatore Iurato

1.1.5.1 Subjective Tests
- Tuning fork tests
- Pure-tone audiometry
- Speech audiometry

1.1.5.2 Objective Tests
- Tympanometry
- Acoustic reflex
- Evoked response audiometry
- Otoacoustic emissions

Tuning Fork Tests
The 512-Hz (c2) aluminium tuning fork is struck on the examiner’s elbow or knee.

Weber Test
Firmly place the stem of the vibrating tuning fork in the middle of the forehead or on the teeth.
- Lateralization to the ear with a hearing loss or with the greater hearing loss (advise the patient that this apparently strange possibility exists): conductive loss.
- Lateralization to the better hearing ear or no lateralization suggests that the problem in the involved ear is sensorineural.

Rinne Test
Comparison of bone conduction (stem of the fork firmly pressed on the mastoid) and air conduction (2–3 cm lateral to the tragus with the tines oriented parallel to the frontal plane of the skull):
- If the tone is louder on bone (negative Rinne test finding), this indicates conductive hearing loss or greater conductive hearing loss.
- If the tone is louder by air (positive Rinne test finding), this indicates sensorineural hearing loss.
- Warning: To minimize the risk of operating on a non-hearing ear always confirm the negative Rinne test finding by masking the contralateral ear with a Barany noise box!

Bing Test
Firmly place the stem of the vibrating tuning fork on the mastoid and then occlude the meatus by pushing the tragus:
- In conductive hearing loss there is no change.
- In sensorineural loss the tuning fork is heard louder.

Comment: The tuning fork tests are not an optional but a standard part of the otologic evaluation. The results must be in agreement with those of the pure-tone audiometry.
equivalent to air conduction), it means that the whole loss is sensorineural and the conductive mechanism is normal (Figs. 1.1.16, 1.1.17). An example of a bilateral mixed hearing loss (conductive and sensorineural) is shown in Fig. 1.1.18: both the conductive mechanism and the cochlea are impaired.

**Speech Audiometry**

Lists of two-syllable words with equal stress on both syllables are administered by air conduction through earphones or in free-field to both ears. The intensity of the words is varied through the audiometer attenuator. The patient is asked to repeat the words and the results are plotted on a special graph (speech audiogram).

At the speech reception threshold the patient repeats correctly approximately 50% of the words. In conductive hearing loss there is a good agreement between the speech reception threshold and the pure-tone average obtained at the frequencies 500, 1,000 and 2,000 Hz. When the speech reception threshold is much better than the pure-tone average, the examination should be repeated (suspected *non-organic hearing loss*). Patients with a conductive hearing loss and normal cochlear function reach a discrimination of 100%. Patients with a moderate sensorineural loss generally also have a good speech discrimination. Speech discrimination is not so good in patients with a severe sensorineural loss and in patients with a high tone loss. In lesions of the cochlear nerve and in those of the auditory central pathways, the speech discrimination is often poorer than expected from the pure-tone average (*tone–speech dissociation*). In some of these patients, discrimination decreases when the lists of words are administered at a higher intensity (*helmet curve* in the speech audiogram): this indicates the possible presence of a retrocochlear lesion.

**Impedance Audiometry**

**Tympanometry**

Tympanometry is an objective method serving to evaluate the mobility of the tympanic membrane and the functional condition of the middle ear, as pressure is altered in the external auditory canal from +400 to –600 daPa. A tympanogram is its graphic representation (Fig. 1.1.19):
- **Type A** curve in patients with a normal tympanic membrane, good tube function (normal patients and patients with a sensorineural loss).
- **Type A** curve (Fig. 1.1.14) in patients with reduced compliance (associated with otosclerosis, tympanosclerosis, fixed malleus).

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**Fig. 1.1.12** Hearing loss. Basic diagnostic procedures: family history, case history, otoscopic examination, complete hearing evaluation (pure-tone audiometry, speech audiometry, tympanometry, acoustic reflex). *ABR* auditory brainstem response, *EVA* enlarged vestibular aqueduct.
Movements of the tympanic membrane due to a patulous Eustachian tube may be registered.

Stapedial Reflex (Acoustic Reflex)
The stapedial muscles contract bilaterally when one of the two ears is stimulated with a sufficiently loud sound.

- In the normal ear the contraction reflex occurs at an intensity of 70–100 dB SL (mean value 82.2 dB SL for pure tones and 65 dB SL for white noise). The lowest signal intensity capable of eliciting the reflex is recorded as the acoustic reflex threshold for the stimulated ear (and not for the probe ear).

- In cochlear sensorineural hearing loss the reflex threshold decreases (positive recruitment).

- In retrocochlear hearing loss (e.g. acoustic tumour) the acoustic reflex may be absent or decline (Fig. 1.1.17) under continued pure-tone stimulation (acoustic reflex decay).

**Fig. 1.1.13** Steps in audiologic evaluation. **AC** air conduction, **BC** bone conduction, **SRT** speech reception threshold, **PTA** pure-tone average, **OME** otitis media with effusion, **A/B gap** air/bone gap
In bilateral conductive hearing loss the reflex is absent bilaterally. In unilateral conductive hearing loss of 30 dB HL or more the reflex is also absent bilaterally. This occurs because when the sound is presented to the ear with a conductive loss of 30 dB or more, the loss itself prevents the sound from being heard loudly enough to elicit the reflex. When the sound is presented to the normal ear, with the probe in the ear with unilateral hearing loss, the disease prevents the change in compliance.

**Warning:** Avoid measuring the reflex in cases of acute sensorineural hearing loss or recent tinnitus, to avoid the risk of further damage.

---

**Evoked Response Audiometry**

The main applications of evoked potential audiometry are (1) to diagnose hearing impairment in non-cooperative patients (infants and handicapped adults) and (2) to identify retrocochlear disorders (site-of-lesion tests). The principle is that of algebraic summation of the electrical events following repeated stimulation, thus discriminating stimulus-related electrical potentials from spontaneous activity. Depending on their latencies in milliseconds, first (0–2 ms), early (2–10 ms), middle (10–50 ms), slow (50–300 ms) and late (300 ms or more) potentials have been described (Fig. 1.1.20).

**Electrocochleography**

The *cochlear microphonics, summing potentials and action potentials* are electrical potentials that occur before the auditory brainstem response (ABR) potentials. They are recorded with a needle electrode through the tympanic membrane (trans tympanic electrocochleography) or with distant electrodes and indicate the function of the inner ear and auditory nerve.

Electrocochleography is used in the evaluation of patients with suspected Ménière’s disease (a significant enhancement of the summing potential to action potential amplitude ratio occurs in 60% of patients with Mé-
nière's disease), patients with a suspected perilymphatic fistula and to exclude residual cochlear function before performing a cochlear implant.

**Auditory Brainstem Response**
1. Latency is between 1.5 and 10 ms. Peaks following wave I occur at approximately 1-ms intervals and are indicated as I–VII (Jewett). Wave I Jewett corresponds to wave N1 of electrocochleography (Fig. 1.1.20). The sources of the waves are:
   - I and II: eighth nerve
   - III: cochlear nucleus
   - IV: olivary complex
   - V: lateral lemniscus
   - VI: inferior colliculus
2. Latency measurements to differentiate retrocochlear from cochlear lesions are the interwave interval (usually measured between waves I and V), the interaural latency and the absolute latency (Fig. 1.1.17).
3. A I–V interval longer than 4.4 ms is considered abnormal (however, each centre should have its own normative data).
4. A difference greater than 0.4 ms between wave V latency in the two ears (interaural latency) is considered an indication of retrocochlear abnormality (Fig. 1.1.17). Latency is corrected by subtracting 0.10 ms for every 10 dB of hearing loss greater than 50 dB HL at 4 Hz.
5. ABR should be present at high-intensity levels if the pure-tone threshold at 4 kHz does not exceed 70 B HL.
6. Absent ABR in a patient with a mild sensorineural hearing loss is an indication of retrocochlear abnormality.

**Cortical Evoked Response Audiometry**
The late components (evoked response audiometry and vertex potentials of the old terminology) are not used routinely in clinical hearing evaluation, mainly because the results are very variable.

**Otoacoustic Emissions**
Small sounds from healthy outer hair cell activity (mobility) transmitted back into the external auditory canal (echoes) and picked up by the probe tip microphone.
- *Spontaneous otoacoustic emissions* are present in 50% of normal young individuals.
- *Evoked otoacoustic emissions* in response to acoustic stimulation are present if hearing loss is equal to or better than 30 dB HL, and are absent if the hearing loss is greater than 30 dB HL.
1.1.5 Technical Diagnostic Procedures

Conditioned Orientation Reflex
A visual reinforcement acts as a reward, increasing the chances that the child will continue to respond to subsequent sound presentations. It is used for infants in the 25–36 months age range, together with impedance audiometry.

Warning: Behavioural audiometry and conditioned orientation reflex are free-field tests, so they do not reveal the status of each ear.

Play Audiometry
The child is trained to answer with a motor response (game). Thresholds should be obtained before habituation occurs.

Examination of the Vestibular Organ
A complete clinical and medical history is essential in the evaluation of a patient complaining of dizziness.

- Otoacoustic emissions are frequently used in screening of newborns and to monitor the status of the cochlea during treatment with ototoxic drugs.
- Distortion product otoacoustic emissions. On simultaneous stimulation with two tones of different frequency, the cochlea generates another tone. Distortion product otoacoustic emissions are absent when there is a sensorineural hearing loss of about 50–60 dB HL or greater.

Assessment of Infants and Children
For details see Sect. 1.6.12. The hearing assessment of neonates and infants during the first 3–4 months of life is mainly based on physiological tests (otoacoustic emissions and ABR). After 5 months, the use of both behavioural and physiological tests is recommended.

Behavioural Audiometry
Behavioural audiometry involves watching the responses to sudden and intense stimulus sounds presented in the sound field. It is used for infants in the 5–24 months age range, in combination with impedance audiometry.

Conditioned Orientation Reflex
A visual reinforcement acts as a reward, increasing the chances that the child will continue to respond to subsequent sound presentations. It is used for infants in the 25–36 months age range, together with impedance audiometry.

Warning: Behavioural audiometry and conditioned orientation reflex are free-field tests, so they do not reveal the status of each ear.

Play Audiometry
The child is trained to answer with a motor response (game). Thresholds should be obtained before habituation occurs.

Examining of the Vestibular Organ
A complete clinical and medical history is essential in the evaluation of a patient complaining of dizziness.

---

Fig. 1.1.16 An example of bilateral sensorineural hearing loss (Pendred’s syndrome). The air-conduction and bone-conduction thresholds are equal and the acoustic reflex is present on both sides (From Arnold et al. [1])

<table>
<thead>
<tr>
<th>Test Frequency Hz</th>
<th>125</th>
<th>250</th>
<th>500</th>
<th>1K</th>
<th>2K</th>
<th>4KHz</th>
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<tr>
<td>CONTRA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Probe Right Bar</td>
<td>110</td>
<td>110</td>
<td>110</td>
<td>100</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Probe Left Bar</td>
<td>100</td>
<td>100</td>
<td>nr</td>
<td>nr</td>
<td>nr</td>
<td></td>
</tr>
</tbody>
</table>
Fig. 1.1.17 An example of unilateral sensorineural hearing loss in the left ear (with decay of the acoustic reflex and pathological ABR). The retrocochlear abnormality (vestibular schwannoma) was confirmed by MRI and at surgery (From Arnold et al. [1])
1.1.5 Technical Diagnostic Procedures

Fig. 1.1.18 An example of bilateral mixed hearing loss (bilateral chronic otitis media with cochlear impairment).

Fig. 1.1.19 Classification of tympanograms: type A, normal; type A\textsubscript{s}, reduced compliance (e.g. otosclerosis)—see also Fig. 1.1.14; type A\textsubscript{o}, flaccid /scarred tympanic membrane; type B, non-mobile tympanic membrane (e.g. OME)—see also Fig. 1.1.15; type C, poor Eustachian tube function—see also Fig. 1.1.15; type D, hypermobile tympanic membrane (e.g. ossicular disruption). (From Arnold et al. [1])
• Does the patient experience a spinning sensation (this means a disease of the vestibular system) or a sensation of unsteadiness (non-vestibular dizziness)?

• Is the vertigo episodic, with a normal equilibrium between the attacks? The spells last seconds in the case of benign paroxysmal vertigo and hours in the case of Ménière's disease. Short episodes associated with nausea and vomiting suggest a peripheral labyrinthine abnormality. Acute rotational vertigo lasting several days with slow recovery is characteristic of viral, traumatic or vascular labyrinthitis or vestibular neuritis.

• Does a rapid head movement, particularly when turning over in bed, incite vertigo? If so, benign paroxysmal vertigo is likely.

• Does vertigo occur with neck motion? If so, cervical vertigo is likely.

• Central non-vestibular disease begins insidiously and lasts days or months. The sensation of dysequilibrium is usually chronic.

• Are there associated cochlear symptoms? Hearing loss, fullness, tinnitus? These symptoms suggest a cochlear (Ménière) or a retrocochlear (vestibular schwannoma) abnormality.

• Are there other otologic symptoms, acute otitis media, cholesteatoma, perilymphatic fistula?

• Are there other disorders, including cardiovascular disease, cerebrovascular disease, neurological disorders, visual impairment and diabetes?

• Does the patient use alcohol, medications or other drugs (ototoxic antibiotics, antihypertensive, antidepressant, sedative-hypnotic drugs)?

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**Fig. 1.1.20** Otoacoustic emissions (OAE), electrocochleography (ECoG), ABR, cortical evoked response audiometry (ERA): topographical map
**Nystagmus**
This is rhythmic, involuntary movements of the eyes. The slow phase is vestibular, the quick phase central. The direction of the quick component, which is more evident at visual inspection, is used to define the direction of nystagmus.

**Spontaneous Nystagmus**
As visual fixation may suppress spontaneous nystagmus, movements of the eyes are observed without and with illuminated Frenzel glasses (20-diopter lenses that magnify the eyes and eliminate fixation). Spontaneous nystagmus may result from peripheral or central disorders (Table 1.1.1).

- **Peripheral**: attack of Ménière, labyrinthitis, temporal bone fracture, sudden unilateral loss of the vestibular function, benign paroxysmal vertigo. Horizontal or horizontal-rotatory, visually suppressed, often associated with auditory symptoms and vertigo, directed towards the irritated labyrinth, away from the paretic labyrinth. Degree I occurs only with the eyes pointing in the direction of the quick phase. It should be differentiated from physiological positional nystagmus. Degree II also occurs with the eyes looking straight ahead. Degree III also occurs with the eyes pointing in the direction of the slow phase.
- **Central**: head trauma, encephalitis, multiple sclerosis, brain tumour, drugs, alcohol. Any direction, often vertical, not visually suppressed, may or may not occur with vertigo, points towards the side of lesion, unusual auditory symptoms.

**Positional Nystagmus**
Patient supine with Frenzel glasses, slow changes of position.
- Head in mid position
- Head turned to the right
- Head turned to the left
- Head hanging over the edge of the bed

**Postural Test (Hallpike Manoeuvre)**
Patient seated (with Frenzel glasses) on the examining couch in such a way that when moved into the supine position, the head and neck will extend beyond the edge of the couch. The head is held by the physician, who rapidly moves the patient into the following nine positions (time of observation, 20–30 s per position):
1. Patient seated, head in mid position
2. Patient supine, head hanging
3. Patient seated, head in mid position
4. Patient seated, head to the right
5. Patient supine, head hanging turned to the right
6. Patient seated, head to the right
7. Patient seated, head to the left
8. Patient supine, head hanging turned to the left
9. Patient seated, head to the left

The postural test evaluated as follows:
- **Peripheral**: fixed direction of the nystagmus (beats in the same direction in different positions), occurs after a latent period, lasts less than 60 s, undergoes fatigue (weaker and shorter responses when repeating the manoeuvre). Classic finding of benign paroxysmal positioning vertigo.
- **Central**: changing direction of the nystagmus, no latency, lasts more than 60 s, does not undergo fatigue.

**Vestibulospinal Reflexes**
Abnormalities in the vestibulospinal pathways are revealed by the following tests:
- Finger-to-nose test (dysmetria)
- Heel-to-toe
- Dysdiadochokinesis (inability to rapidly turn the hand prone and supine)
- Pointing test
- Romberg test (standing with eyes closed, feet together and arms to the side)
- Posturography (objective Romberg test) with Luzern platform
- Unterberger test (stepping in place with eyes closed).

**Caloric Test**
The minimal caloric test is performed in the office setting with the patient sitting with head tilted backwards by 60°, and 5 ml of water injected in the ear at a temperature of 23°C. The opposite ear is tested in the same way. The length of the nystagmus is recorded. The ice-water caloric test is administered if no response is obtained with the 23°C test.

**Electronystagmography/Videonystagmography**
Nystagmus can be recorded electrically (electronystagmography), by means of an infrared photoelectric system, or by a video camera (videonystagmography).
- The patient is placed supine with the head raised by 30°, with the lateral semicircular canal placed in the optimal position for stimulation. A heat irrigator containing a pump system is used for irrigation (70–100 ml water per 30 s) with water at 7°C above and 7°C below body temperature. The caloric test sequence consists of right cold, left cold, left warm and right warm irrigation. The speed of the slow phase of nystagmus can be measured with a computer.
- Cold caloric stimulation means nystagmus beating away from the stimulated side.
- Warm caloric stimulation means nystagmus beating towards the stimulated side.
A “weaker” response on one side by 20% or greater is interpreted as a sign of a peripheral vestibular lesion (on that side).

**Vestibular Evoked Myogenic Potentials (VEMP)**
- **Definition:** Sound-induced vestibular evoked myogenic potentials are a valuable clinical tool for evaluation of vestibular function (functional status of the saccule and inferior vestibular nerve).
- **Principles:** To obtain information on its function, the saccule is acoustically stimulated by means of clicks at an intensity of 120 dB SPL and its response is registered at the level of the sternocleidomastoid muscle.
- **Indications:** In Ménière’s disease, in vestibular neuritis, vestibular schwannomas originating from the inferior vestibular nerve, in forensic medicine, in rare isolated functional disorders of the saccule and when a superior canal dehiscence is suspected (see Sect. 1.5.2.10).

**References**

**Suggested Reading**

<table>
<thead>
<tr>
<th><strong>Table 1.1.1</strong> Features in the history that help distinguish between peripheral and central causes of vertigo (from Kesser and Friedman [2])</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Peripheral</strong></td>
</tr>
<tr>
<td>Imbalance</td>
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<td>Nausea and vomiting</td>
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<tr>
<td>Auditory symptoms</td>
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<tr>
<td>Neurological symptoms</td>
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<tr>
<td>Compensation</td>
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</tbody>
</table>

**1.1.6 Imaging of the Temporal Bone**

**MARTIN DOBRITZ**

This section provides an overview of the principles of conventional radiography, computed tomography (CT) and magnetic resonance imaging (MRI) as well as details of their use in the practice of otology (Table 1.1.2). The physics of CT and MRI is beyond the scope of these guidelines.

**1.1.6.1 Conventional Radiography**

Although conventional radiography more and more has been replaced by CT, it is still worthwhile summarizing the principles and indications of some conventional techniques. The reason is that the Schüller and the transorbital projections are still useful to identify some preoperative and postoperative details, as well as for the recognition of large lesions in or around the temporal bone.

One of the most frequent indications for the Schüller projection is the preoperative assessment of the mastoid size and degree of pneumatization (Fig. 1.1.21a), the course of the sigmoid sinus, fracture lines and bony defects, e.g. caused by a cholesteatoma (Fig. 1.1.21b).

One of the most frequent indications for the transorbital projection is the postoperative demonstration of the correct position and integrity of the cochlear implant electrodes (Fig. 1.1.22). Note that CT techniques are not able to show the continuity of the electrodes in one image.

**Schüller Projection**
- **Projection:** Lateral view of the mastoid
- **Sagittal plane of the head parallel to the tabletop**
- **X-ray beam rotated 30°**

What can be visualized:
- Extent of pneumatization of the mastoid
1.1.6 Imaging of the Temporal Bone

- Distribution/degree of aeration of the air cells
- Status of trabecular pattern
- Position and course of the sigmoid sinus

Notes:
- The sinus plate casts a sharp radiodense vertical line (Fig. 1.1.21) behind the external auditory canal (EAC).
- The oblique line crossing the radiolucency of the EAC is the superior ridge of the petrous bone.
- The internal auditory canal (IAC) is superimposed to the EAC.
- The temporomandibular joint can be seen directly anterior to the EAC (Fig. 1.1.21).

Transorbital Projection

Projection:
- Occiput facing the film
- Head flexed on the chin (15°)
- Each side obtained separately
- Beam directed to orbit centre

What can be visualized:
- Petrous apex, which appears foreshortened
- IAC in its full length
- Lateral to the IAC the vestibule and the semicircular canals
- Apical and middle coils of the cochlea superimposed upon the IAC, the basal turn underneath the vestibule (Fig. 1.1.22)

1.1.6.2 Computed Tomography

Since its introduction in the 1970s, CT has developed to the widely accepted gold standard for evaluating most of the diseases of the temporal bone for the following reasons.

The high contrast between air and osseous structures in connection with the high spatial resolution leads to easier differentiation and excellent recognition of inflammatory or neoplastic changes of the bone as well as fractures. In addition to that, the ossicular chain and the status of pneumatized cells can be determined in an excellent way.

Multislice techniques produce isotropic 3D datasets to perform excellent multiplanar reformations. Besides re-

<table>
<thead>
<tr>
<th>Pathologic conditions</th>
<th>CT (bony structures)</th>
<th>MRI (soft tissues)</th>
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</thead>
<tbody>
<tr>
<td>Temporal bone fractures</td>
<td>+++</td>
<td>–</td>
</tr>
<tr>
<td>Exostosis, osteoma of external auditory canal</td>
<td>++</td>
<td>–</td>
</tr>
<tr>
<td>Congenital malformations of middle and inner ear</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>Inflammatory conditions (mastoiditis, chronic otitis media, malignant external otitis)</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Otosclerosis</td>
<td>+++</td>
<td>–</td>
</tr>
<tr>
<td>Cholesteatoma</td>
<td>+++</td>
<td>+</td>
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<tr>
<td>Petrous apex lesions</td>
<td>+++</td>
<td>+++</td>
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<tr>
<td>Cerebral abscess</td>
<td>++</td>
<td>+++</td>
</tr>
<tr>
<td>Labyrinthitis, meningitis</td>
<td>+</td>
<td>++</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>–</td>
<td>+++</td>
</tr>
<tr>
<td>Sinus thrombosis</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Endolymphatic sac tumour</td>
<td>++</td>
<td>+++</td>
</tr>
<tr>
<td>Arterial and venous variants</td>
<td>++</td>
<td>+</td>
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<tr>
<td>Tumour of cerebellopontine angle</td>
<td>–</td>
<td>+++</td>
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<tr>
<td>Paraganglioma</td>
<td>++</td>
<td>+++</td>
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<tr>
<td>Malignant neoplasms</td>
<td>++</td>
<td>+++</td>
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<tr>
<td>Postoperative imaging:</td>
<td></td>
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<tr>
<td>Cochlear implant, position of the prosthesis following stapedectomy, status of mastoid following “close” tympanoplasty</td>
<td>+++</td>
<td>–</td>
</tr>
<tr>
<td>Follow-up vestibular schwannoma surgery, skull base surgery</td>
<td>–</td>
<td>+++</td>
</tr>
</tbody>
</table>
Reducing radiation exposure for the patient, the secondary reconstruction helps to localize abnormalities in vertical and horizontal planes as well as to define the relationship to adjacent intracranial and extracranial lesions.

The following list gives the reader an update of the actual CT technique and its limitations:

- Slice thickness less than 1 mm, multiplanar reformations (axial slices, coronal plane, paracoronal plane), imaging of calcified structures
- Multislice spiral CT (64 slices)
- Isotropic datasets (primary axial data acquisition)
- Multiplanar reconstructions possible
- Bone reconstruction algorithm/high-resolution osseous structures.
- Window, width 4,000 HU, centre 500 HU

What can be visualized:
1. Anatomy of the temporal bone (Figs. 1.1.23, 1.1.24).
2. Tympanic membrane and tensor tympani muscle.
3. Auditory ossicles (Fig. 1.1.23; Fig. 9.3.1b). Note that the whole stapes is visible only in the axial projection (Figs. 1.1.23a, 1.5.5b).
4. Bony labyrinth
   - Cochlea, e.g. congenital malformations (Fig. 1.2.2)?
   - Ossification of the cochlear turns (Fig. 1.1.25) following meningitis, prior to cochlear implantation?
   - Cochlear otosclerosis?
   - Vestibule (air bubbles? Fig. 1.5.5a)
   - Semicircular canals (fistula?)
5. Round and oval windows (Fig. 9.3.1a)
6. Vestibular aqueduct (Fig. 1.1.23, normal; Fig. 1.1.26, enlarged).
7. Intratemporal facial nerve (labyrinthine, tympanic and mastoid segments).
8. Defects of the temporal bone (Fig. 1.4.17b).
9. Fractures (Figs. 1.5.5b, 1.5.8b, 1.7.4).

Notes:
- No contrast media.
- No useful information in cases of vestibular schwannoma.
- The reconstructed images are not always as satisfactory as the native sections in coronal or lateral projections, which, on the other hand, are sometimes difficult to obtain (children, traumatized patients, older people).

1.1.6.3 Magnetic Resonance Imaging

As CT is the gold standard for bony lesions, MRI is an excellent modality for imaging of the cranial nerves (Fig. 1.2.3), the vestibulocochlear system, soft tissue lesions of the temporal bone and the cerebellopontine angle (see Table 1.1.1). Otologists ordering MRI should be aware of the following basic principles:
- Compatibility with certain implanted medical devices; cave, a pacemaker and a cochlear implant are contraindications; titanium implants and modern stapes prosthesis are compatible.
- In MRI there is no involvement of ionizing radiation.
- After a high-frequency impulse, the resonance of the protons provides the signal used for the images.
- Visualization of water and soft tissue.
- Air and cortical bone do not have protons and therefore these components are not visible in any sequence. Consequently, MRI does not delineate bony anatomy (Fig. 1.2.3).
- Differences in relaxation properties among tissues cause typical contrast between different tissues. Fat appears as bright areas in T1-weighted images (Fig. 1.1.27a, 1.4.18a,b) and fluids (CSF, inner ear fluids) appear as bright areas in the T2-weighted images (Figs. 1.1.27b, Fig. 1.4.18c). Vessels containing flowing blood emit no or low signal (Fig. 1.5.6c) because the excited protons move out of the area before detection of the emitted signal (most sequences). See Sect. 1.5.5 for the differential diagnosis of congenital cholesteatoma, cholesterol granuloma and mucocele.
- A variety of techniques can be employed in MRI of lesions of the temporal bone area.
- 3D T2-weighted sequences (3D constructive interference in steady state and 3D fast spin echo) offer submillimetre isotropic high-resolution datasets.
- Intravenous contrast agents (Fig. 1.1.27a) can be employed to enhance MRI (most commonly gadolinium as a T1-positive contrast agent is used).
- Enhancement is commonly observed in neoplasms (Fig. 1.1.27a) and inflammatory conditions (Fig. 1.7.2).

Suggested Reading

Fig. 1.1.23a,b  Axial computed tomography (CT) slice, right ear. Tensor tympani muscle (†), stapes (⊙), cochlea (≈), facial nerve (●), vestibular aqueduct (○), internal auditory canal (▲), vestibule (⊙), lateral semicircular canal (●). Note the incudomalleal joint and the “ice-cream cone” (body of the incus with the malleus head).
Fig. 1.1.24a–c Coronal CT reformations, right ear. Cochlea (argent), styloid process (<d>), facial nerve (<r>), malleus head (<z>), superior semicircular canal (<y>), lateral semicircular canal (<i>), stapes (<y>), internal auditory canal (<A>)

Fig. 1.1.25 Coronal CT reformation right side. Labyrinthine ossification (<i>)
1.1 Basics

Fig. 1.1.26 Large endolymphatic sac anomaly with progressive deafness in a 1 year-old girl (large vestibular aqueduct syndrome). Axial CT: internal auditory canal (\(\triangle\)), vestibular aqueduct (\(\diamond\)), endolymphatic sac area (\(\dagger\)). Insert: magnetic resonance imaging (MRI), T2-weighted: cochlea (\(\approx\)), lateral semicircular canal (\(\uparrow\)), endolymphatic duct (\(\times\)), endolymphatic sac (\(\dagger\)), vestibule (\(\circ\))

Fig. 1.1.27a,b Vestibular schwannoma (\(\ast\)) left side. Internal auditory canal (\(\bigtriangledown\)). a MRI, T1-weighted contrast enhanced. b MRI, T2-weighted
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