

Diseases of EAR, NOSE AND THROAT

& HEAD and NECK SURGERY

SEVENTH EDITION

PL Dhingra | Shruti Dhingra

Assisted by Dheeksha Dhingra



*Diseases of Ear,
Nose and Throat
& Head and Neck Surgery*

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Diseases of Ear, Nose and Throat & Head and Neck Surgery, **7th Edition, PL Dhingra, Shruti Dhingra and Deeksha Dhingra**

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*Dedicated to all my students: past, present and future who are the
inspiring force behind this work.*

*I reproduce below the invocation from our great ancient scripture—the
Kathopanishad which shows the relationship between the teacher and the taught.*

ॐ सह नौ अवतु। सह नौ भुनक्तु।
सह वीर्यम् करावावहै। तेजस्वि नौ अधीतम् अस्तु।
मा विद्विषावहै। ॐ शान्तिः शान्तिः शान्तिः

*“O God, the almighty, bless us both (the teacher and the student)
together, develop us both together, give us strength together. Let the
knowledge acquired by us be bright and illuminant, and second to none.
Let both of us live together with love, affection and harmony. O God, let
there be physical, mental and spiritual peace.”*


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Preface

It is a matter of pride and pleasure to bring out the *silver jubilee edition* of our book “*Diseases of Ear, Nose and Throat & Head and Neck Surgery.*” The book was first published in 1992 and has been well accepted and appreciated by the students and teachers all over the country as well as in adjoining South Asian countries. During this period of 25 years, six editions and several reprints were brought out. This was the result of growth of the speciality, innovations in technology and surgical techniques but behind it was students’ burning desire to know the subject and quest for knowledge. They freely interacted through emails, letters and other types of social media to clarify, and send suggestions, omissions, commissions, and their interest in the subject to further add the topics of their need. We complied practically with all of them as far as possible.

Our basic aim in writing this book has been to build concepts in disorders of ENT, superstructured with the students’ earlier knowledge of anatomy and physiology learnt in previous professionals. Since Otolaryngology, commonly called ENT, is a full-fledged subject in MBBS examination and is so recognized in various universities in India and Medical Council of India, we did not lose sight of the fact that students have to clear the exams too. The book covers disorders of ENT, surgical instruments,

imaging techniques, operative surgery, recent and newer modalities of treatment in a concise, lucid and student-friendly manner. The chapter on “Nuggets for Rapid Review” covers most of the questions that are often set in post-graduate entrance and Diplomate of National Board (DNB) examinations.

The present edition is revised, updated and expanded. Several new clinical photographs, diagrams, tables and flowcharts have been added to make the subject clear. A unique feature of this edition is white board lectures and videos () depicting through animations, the surgical procedures.

It is hoped that the present edition would continue to serve the needs of MBBS students, residents and practitioners. The postgraduate students of the DLO, MS and DNB will find it useful as a foundation book before taking recourse to comprehensive volumes on the subject.

The students of allied subjects, Audiology and Speech Therapy, Physiotherapy and those studying alternative medicine (Ayurveda, Sidha, Unani and Homeopathy) will also find it useful to learn basics and concepts of ENT.

The authors will gratefully accept any suggestions and comments from the learned teachers and students at pldhingra@gmail.com or shrudoc@hotmail.com or indiacontact@elsevier.com.

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Chapter 1

Anatomy of Ear

The ear is divided into:

1. External ear
2. Middle ear
3. Internal ear or the labyrinth

THE EXTERNAL EAR

The external ear consists of the (i) auricle or pinna, (ii) external acoustic canal and (iii) tympanic membrane (Figure 1.1A).

A. AURICLE OR PINNA

The entire pinna except its lobule and the outer part of external acoustic canal are made up of a framework of a single piece of yellow elastic cartilage covered with skin. The latter is closely adherent to the perichondrium on its lateral surface while it is slightly loose on the medial (cranial) surface. The various elevations and depressions seen on the lateral surface of pinna are shown in Figure 1.1B.

There is no cartilage between the tragus and crus of the helix, and this area is called *incisura terminalis* (Figure 1.1C). An incision made in this area will not cut through the cartilage and is used for endaural approach in surgery of the external auditory canal or the mastoid. Pinna is also the source of several graft materials for the surgeon. Cartilage from the tragus, perichondrium from the tragus or concha and fat from the lobule are frequently used for reconstructive surgery of the middle ear. The conchal cartilage has also been used to correct the depressed nasal bridge while the composite grafts of the skin and cartilage from the pinna are sometimes used for repair of defects of nasal ala.

B. EXTERNAL ACOUSTIC (AUDITORY) CANAL

It extends from the bottom of the concha to the tympanic membrane and measures about 24 mm along its posterior wall. It is not a straight tube; its outer part is directed upwards, backwards and medially while its inner part is directed downwards, forwards and medially. Therefore, to see the tympanic membrane, the pinna has to be pulled upwards, backwards and laterally so as to bring the two parts in alignment.

The canal is divided into two parts: (i) cartilaginous and (ii) bony.

1. Cartilaginous Part

It forms outer one-third (8 mm) of the canal. Cartilage is a continuation of the cartilage which forms the framework

of the pinna. It has two deficiencies—the “*fissures of Santorini*” in this part of the cartilage and through them the parotid or superficial mastoid infections can appear in the canal or vice versa. The skin covering the cartilaginous canal is thick and contains ceruminous and pilosebaceous glands which secrete wax. Hair is only confined to the outer canal and therefore furuncles (staphylococcal infection of hair follicles) are seen only in the outer one-third of the canal.

2. Bony Part

It forms inner two-thirds (16 mm). Skin lining the bony canal is thin and continuous over the tympanic membrane. It is devoid of hair and ceruminous glands. About 6 mm lateral to tympanic membrane, the bony meatus presents a narrowing called *isthmus*. Foreign bodies, lodged medial to the isthmus, get impacted, and are difficult to remove. Anteroinferior part of the deep meatus, beyond the isthmus, presents a recess called *anterior recess*, which acts as a cesspool for discharge and debris in cases of external and middle ear infections (Figure 1.2). Anteroinferior part of the bony canal may present a deficiency (*foramen of Huschke*) in children up to the age of four or sometimes even in adults, permitting infections to and from the parotid.

C. TYMPANIC MEMBRANE OR THE DRUMHEAD

It forms the partition between the external acoustic canal and the middle ear. It is obliquely set and as a result, its posterosuperior part is more lateral than its anteroinferior part. It is 9–10 mm tall, 8–9 mm wide and 0.1 mm thick. Tympanic membrane can be divided into two parts:

1. Pars Tensa

It forms most of tympanic membrane. Its periphery is thickened to form a fibrocartilaginous ring called *annulus tympanicus*, which fits in the tympanic sulcus. The central part of pars tensa is tented inwards at the level of the tip of malleus and is called *umbo*. A bright cone of light can be seen radiating from the tip of malleus to the periphery in the anteroinferior quadrant (Figure 1.3).

2. Pars Flaccida (Shrapnell's Membrane)

This is situated above the lateral process of malleus between the notch of Rivinus and the anterior and posterior malleal folds (earlier called malleolar folds). It is not so taut and may appear slightly pinkish. Various landmarks seen on the lateral surface of tympanic membrane are shown in Figure 1.4.

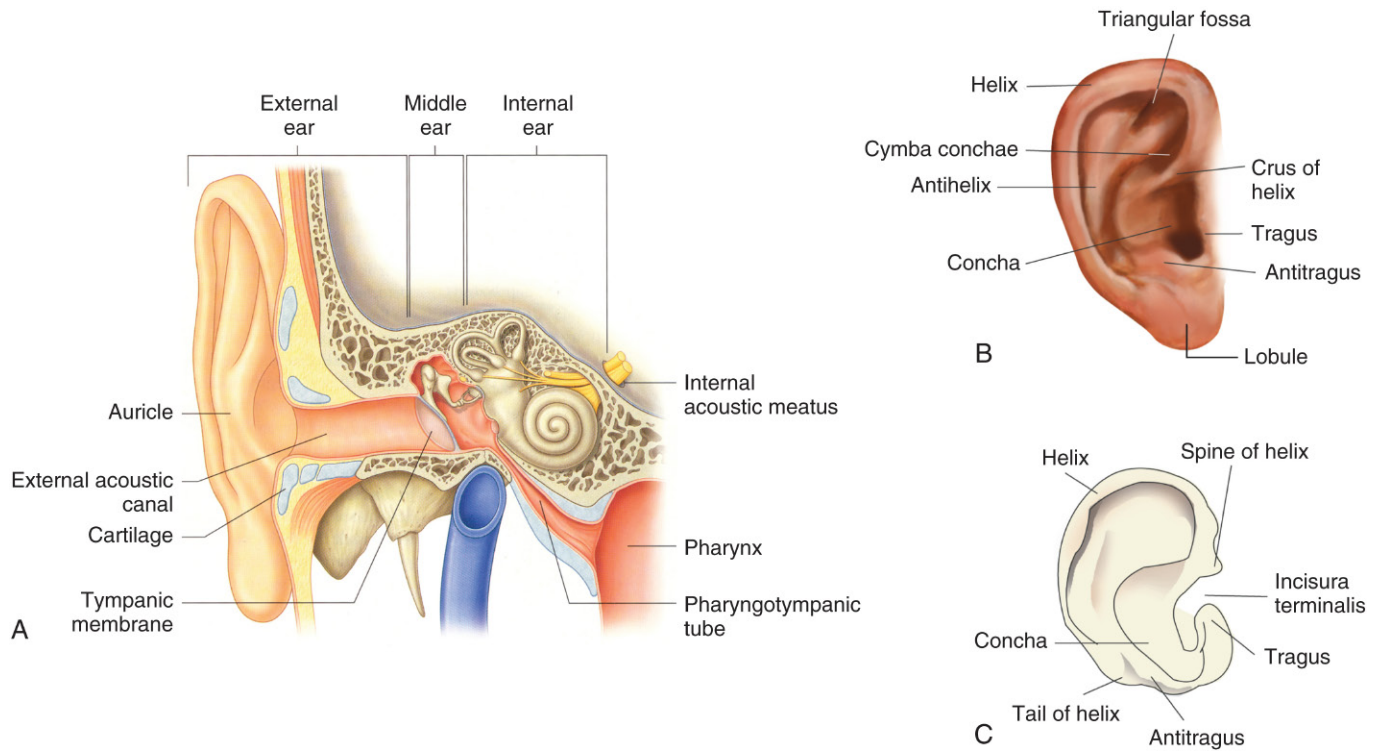


Figure 1.1. (A) The ear and its divisions. (B) The elevations and depressions on the lateral surface of pinna. (C) The auricular cartilage.

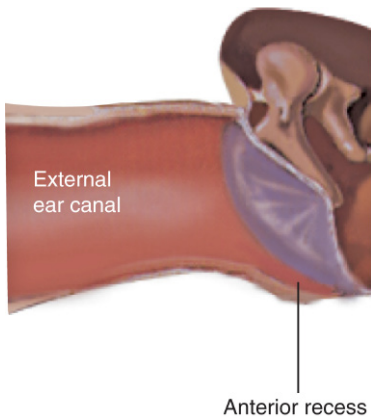


Figure 1.2. Anterior recess of the meatus. It is important to clean discharge and debris from this area.

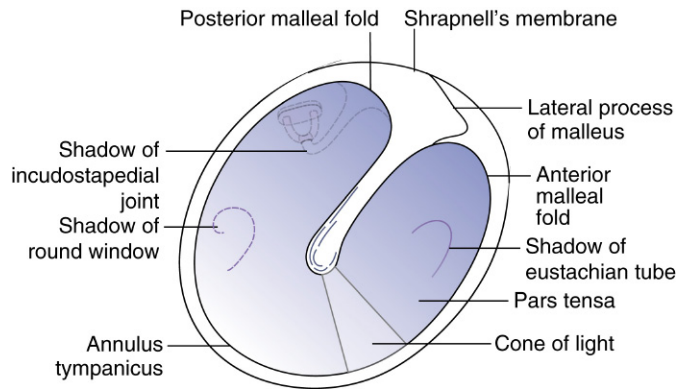


Figure 1.4. Landmarks of a normal tympanic membrane of right side.

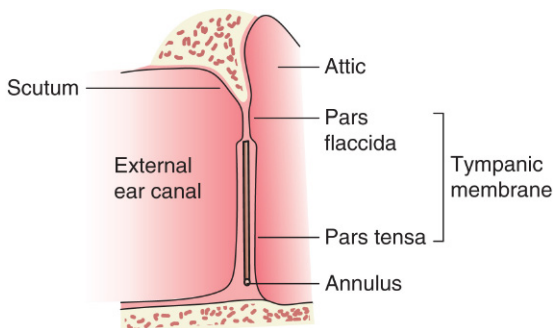


Figure 1.3. Coronal section through tympanic membrane and external ear canal showing structures of pars tensa and pars flaccida of tympanic membrane. Scutum forms a part of lateral attic wall.

Layers of Tympanic Membrane

Tympanic membrane consists of three layers:

- Outer epithelial layer, which is continuous with the skin lining the meatus (Figure 1.3).
- Inner mucosal layer, which is continuous with the mucosa of the middle ear.
- Middle fibrous layer, which encloses the handle of malleus and has three types of fibres—the radial, circular and parabolic (Figure 1.5).

Fibrous layer in the pars flaccida is thin and not organized into various fibres as in pars tensa.

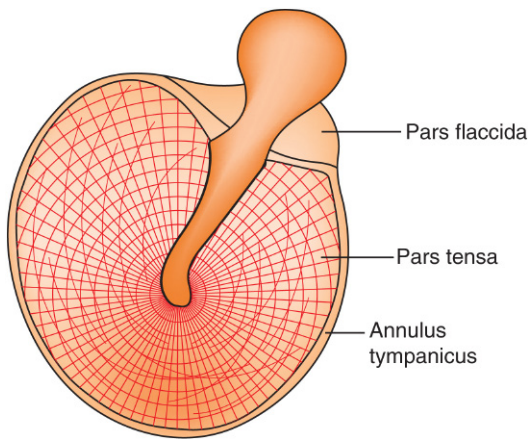


Figure 1.5. Radial, circular and parabolic fibres of pars tensa of tympanic membrane.

RELATIONS OF EXTERNAL ACOUSTIC MEATUS

- Superiorly: Middle cranial fossa
- Posteriorly: Mastoid air cells and the facial nerve
- Inferiorly: Parotid gland
- Anteriorly: Temporomandibular joint

Posterosuperior part of deeper canal near the tympanic membrane is related to the mastoid antrum. “Sagging” of this area may be noticed in acute mastoiditis.

NERVE SUPPLY OF THE EXTERNAL EAR

Pinna

1. Greater auricular nerve ($C_{2,3}$) supplies most of the medial surface of pinna and only posterior part of the lateral surface (Figure 1.6).
2. Lesser occipital (C_2) supplies upper part of medial surface.
3. Auriculotemporal (V_3) supplies tragus, crus of helix and the adjacent part of the helix.
4. Auricular branch of vagus (CN X), also called Arnold’s nerve, supplies the concha and corresponding eminence on the medial surface.
5. Facial nerve, which is distributed with fibres of auricular branch of vagus, supplies the concha and retroauricular groove.

External Auditory Canal

1. Anterior wall and roof: auriculotemporal (V_3).
2. Posterior wall and floor: auricular branch of vagus (CN X).
3. Posterior wall of the auditory canal also receives sensory fibres of CN VII through auricular branch of vagus (see Hitzelberger’s sign on p. 125).

In herpes zoster oticus, lesions are seen in the distribution of facial nerve, i.e. concha, posterior part of tympanic membrane and postauricular region.

Tympanic Membrane

1. Anterior half of lateral surface: auriculotemporal (V_3).
2. Posterior half of lateral surface: auricular branch of vagus (CN X).
3. Medial surface: tympanic branch of CN IX (Jacobson’s nerve).

THE MIDDLE EAR

The middle ear together with the eustachian tube, aditus, antrum and mastoid air cells is called *middle ear cleft* (Figure 1.7). It is lined by mucous membrane and filled with air.

The middle ear extends much beyond the limits of tympanic membrane which forms its lateral boundary and is sometimes divided into: (i) *mesotympanum* (lying opposite the pars tensa), (ii) *epitympanum* or the attic (lying above the pars tensa but medial to Shrapnell’s membrane and the bony lateral attic wall) and (iii) *hypotympanum* (lying below the level of pars tensa) (Figure 1.8). The portion of middle ear around the tympanic orifice of the eustachian tube is sometimes called *protympanum*.

Middle ear can be likened to a six-sided box with a roof, a floor, medial, lateral, anterior and posterior walls (Figure 1.9).

The **roof** is formed by a thin plate of bone called *tegmen tympani*. It also extends posteriorly to form the roof of the aditus and antrum. It separates tympanic cavity from the middle cranial fossa.

The **floor** is also a thin plate of bone, which separates tympanic cavity from the jugular bulb. Sometimes, it is congenitally deficient and the jugular bulb may then

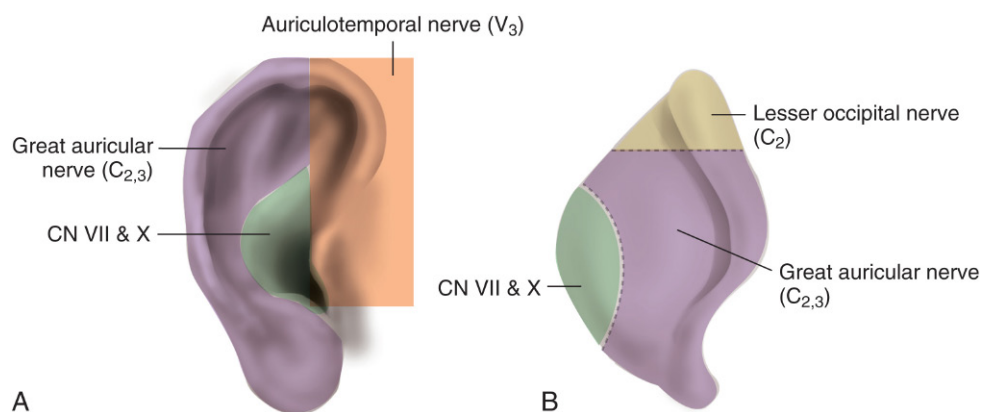


Figure 1.6. Nerve supply of pinna. (A) Lateral surface of pinna. (B) Medial or cranial surface of pinna.

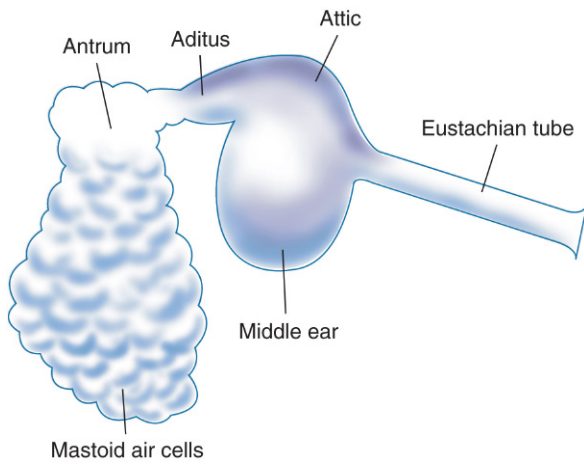


Figure 1.7. Middle ear cleft.

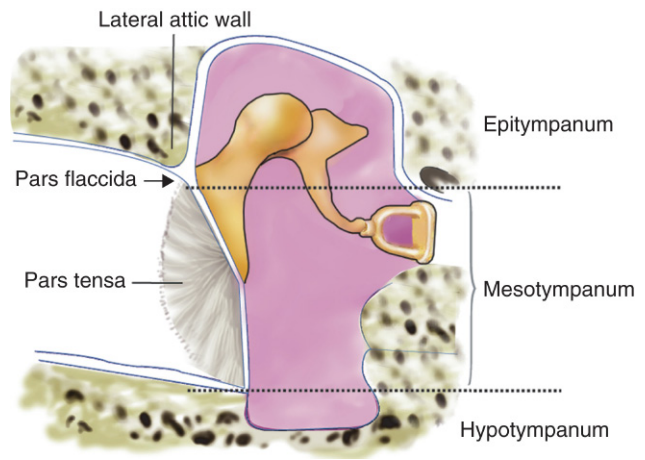


Figure 1.8. Divisions of middle ear into epi-, meso- and hypotympanum.

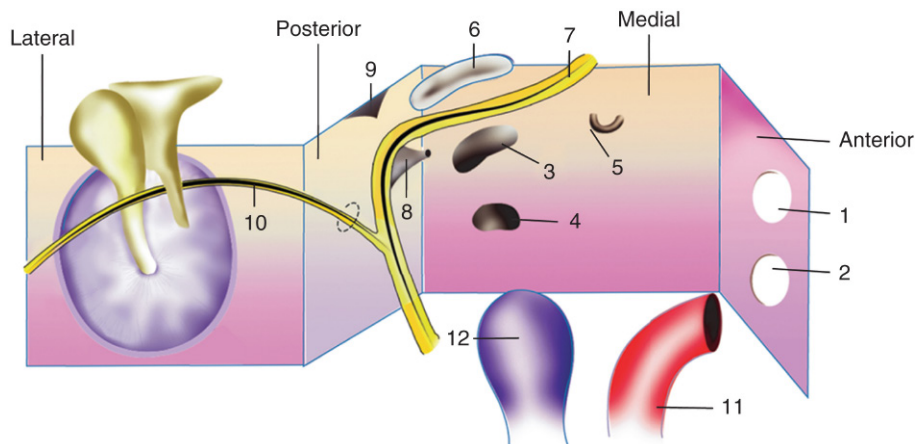


Figure 1.9. Walls of middle ear and the structures related to them. (1) Canal for tensor tympani, (2) Opening of eustachian tube, (3) Oval window, (4) Round window, (5) Processus cochleariformis, (6) Horizontal canal, (7) Facial nerve, (8) Pyramid, (9) Aditus, (10) Chorda tympani, (11) Carotid artery, (12) Jugular bulb.

project into the middle ear; separated from the cavity only by the mucosa.

The **anterior wall** has a thin plate of bone, which separates the cavity from internal carotid artery. It also has two openings; the lower one for the eustachian tube and the upper one for the canal of tensor tympani muscle.

The **posterior wall** lies close to the mastoid air cells. It presents a bony projection called *pyramid* through the summit of which appears the tendon of the stapedius muscle to get attachment to the neck of stapes. *Aditus*, an opening through which attic communicates with the antrum, lies above the pyramid. Facial nerve runs in the posterior wall just behind the pyramid. *Facial recess* or the *posterior sinus* is a depression in the posterior wall lateral to the pyramid. It is bounded medially by the vertical part of VIIth nerve, laterally by the chorda tympani and above, by the fossa incudis (Figure 1.10). Surgically, facial recess is important, as direct access can be made through this into the middle ear without disturbing posterior canal wall (intact canal wall technique, see p. 80).

The **medial wall** (Figure 1.11) is formed by the labyrinth. It presents a bulge called *promontory* which is due to the basal coil of cochlea; *oval window* into which is fixed the footplate of stapes; *round window* or the *fenestra cochleae* which is covered by the secondary tympanic membrane. Above the oval window is the *canal for facial nerve*. Its bony covering may sometimes be congenitally dehiscent and the nerve may lie exposed making it very vulnerable to injuries or infection. Above the canal for facial nerve is the prominence of lateral semicircular canal. Just anterior to the oval window, the medial wall presents a hook-like projection called *processus cochleariformis*. The tendon of tensor tympani takes a turn here to get attachment to the neck of malleus. The cochleariform process also marks the level of the first genu of the facial nerve which is an important landmark for surgery of the facial nerve. Medial to the pyramid is a deep recess called *sinus tympani*, which is bounded by the *subiculum* below and the *ponticulus* above (Figure 1.10).

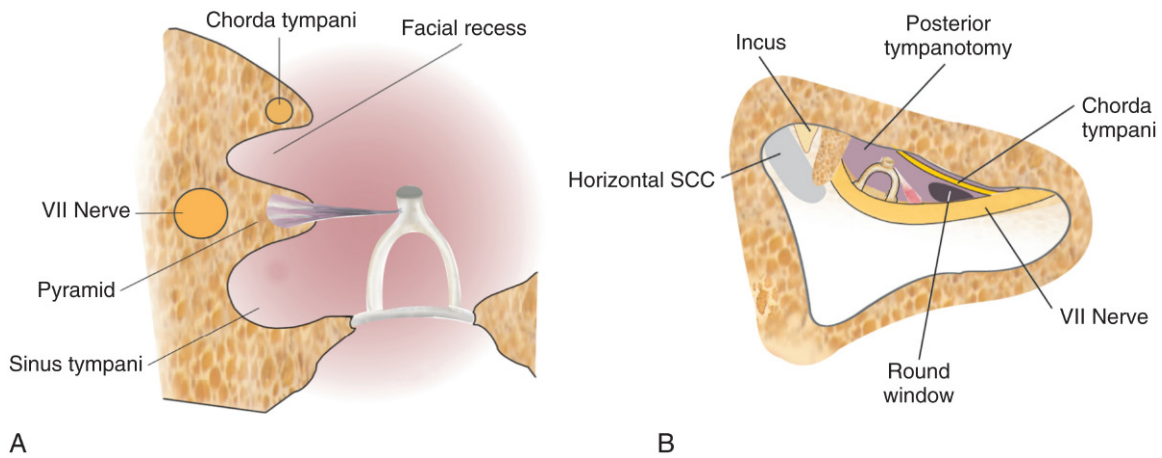


Figure 1.10. (A) Facial recess lies lateral and sinus tympani medial to the pyramidal eminence and vertical part of the facial nerve. (B) Exposure of facial recess through posterior tympanotomy as seen at mastoid surgery. SCC, semicircular canal.

The **lateral wall** is formed largely by the tympanic membrane and to a lesser extent by the bony outer attic wall called *scutum* (Figure 1.3). The tympanic membrane is semi-transparent and forms a “window” into the middle ear. It is possible to see some structures of the middle ear through the normal tympanic membrane, e.g. the long process of incus, incudostapedial joint and the round window.

MASTOID ANTRUM

It is a large, air-containing space in the upper part of mastoid and communicates with the attic through the aditus. Its roof is formed by *tegmen antri*, which is a continuation of the tegmen tympani and separates it from the middle cranial fossa. The lateral wall of antrum is formed by a plate of bone which is on an average 1.5 cm thick in the adult. It is marked externally on the surface of mastoid by *suprameatal (MacEwen's) triangle* (Figure 1.12).

ADITUS AD ANTRUM

Aditus is an opening through which the attic communicates with the antrum. The bony prominence of the hori-

zontal canal lies on its medial side while the fossa incudis, to which is attached the short process of incus, lies laterally. Facial nerve courses just below the aditus.

THE MASTOID AND ITS AIR CELL SYSTEM (FIGURE 1.13)

The mastoid consists of bony cortex with a “honeycomb” of air cells underneath. Depending on development of air cell, three types of mastoid have been described.

1. **Well-pneumatized or cellular.** Mastoid cells are well-developed and intervening septa are thin.
2. **Diploetic.** Mastoid consists of marrow spaces and a few air cells.
3. **Sclerotic or acellular.** There are no cells or marrow spaces.

With any type of mastoid pneumatization, antrum is always present. In sclerotic mastoids, antrum is usually small and the sigmoid sinus is anteposed.

Depending on the location, mastoid air cells are divided into:

1. Zygomatic cells (in the root of zygoma).
2. Tegmen cells (extending into the tegmen tympani).
3. Perisinus cells (overlying the sinus plate).
4. Retrofacial cells (round the facial nerve).

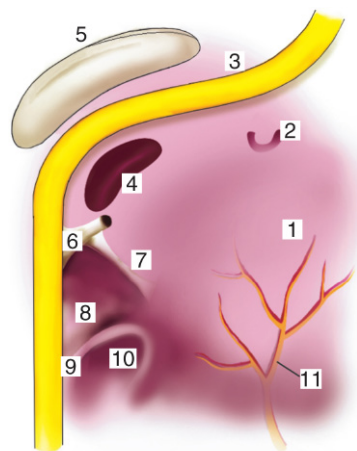


Figure 1.11. Medial wall of middle ear. (1) Promontory, (2) Process cochleariformis, (3) CN VII, (4) Oval window, (5) Horizontal canal, (6) Pyramid, (7) Ponticulus, (8) Sinus tympani, (9) Subiculum, (10) Round window, (11) Tympanic plexus.

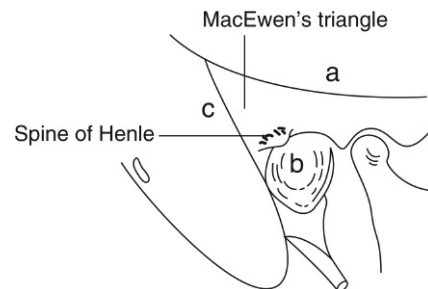


Figure 1.12. MacEwen's (suprameatal) triangle. It is bounded by temporal line (a), posterosuperior segment of bony external auditory canal (b) and the line drawn as a tangent to the external canal (c). It is an important landmark to locate the mastoid antrum in mastoid surgery.

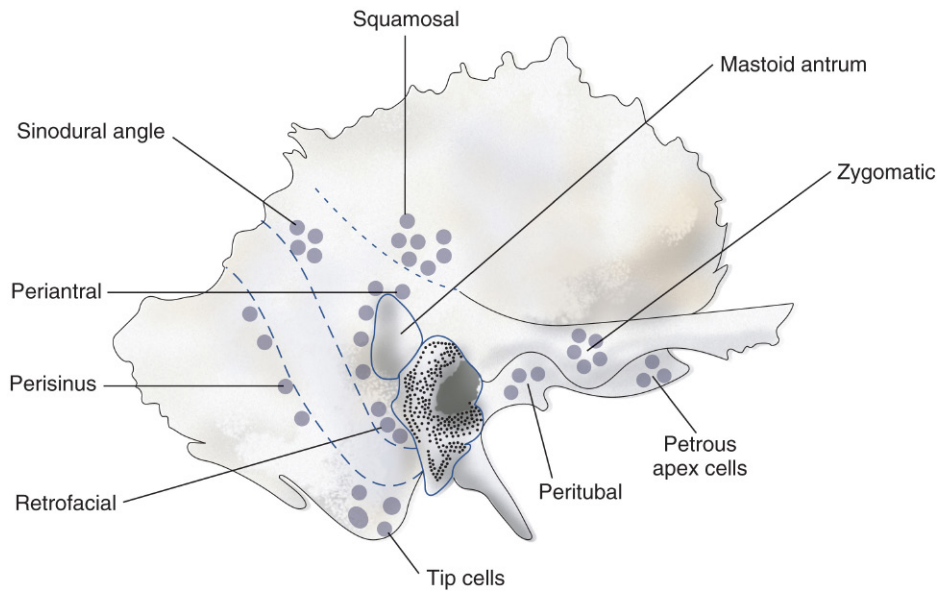


Figure 1.13. Air cells in the temporal bone.

5. Perilabyrinthine cells (located above, below and behind the labyrinth, some of them pass through the arch of superior semicircular canal. These cells may communicate with the petrous apex).
6. Peritubal (around the eustachian tube. Along with hypotympanic cells they also communicate with the petrous apex).
7. Tip cells (which are quite large and lie medial and lateral to the digastric ridge in the tip of mastoid).
8. Marginal cells (lying behind the sinus plate and may extend into the occipital bone).
9. Squamosal cells (lying in the squamous part of temporal bones).

Abscesses may form in relation to these air cells and may sometimes be located far from the mastoid region.

Development of Mastoid

Mastoid develops from the squamous and petrous bones. The petrosquamosal suture may persist as a bony plate—the *Korner's septum*, separating superficial squamosal cells from the deep petrosal cells. Korner's septum is surgically

important as it may cause difficulty in locating the antrum and the deeper cells; and thus may lead to incomplete removal of disease at mastoidectomy (Figure 1.14). Mastoid antrum cannot be reached unless the Korner's septum has been removed.

Petrous apex and its cell system

The petrous apex lies anterior and medial to the labyrinth. It may be pneumatized in 30% of individuals, with cell tracts running either from the mastoid or hypotympanum (Figure 1.15). They run inferior, superior or anterior to the bony capsule of the labyrinth and cochlea. Thus various surgical approaches have been used to drain the inflammatory or cystic lesions of the petrous apex.

1. **Inferior route.** This is the most common route. Two approaches are used:
 - a. Infralabyrinthine. Access is through mastoid, and cell tracts run below the labyrinth.
 - b. Infracochlear. Access is through the ear canal, and tract runs from the hypotympanum to the bony cochlea to petrous apex.

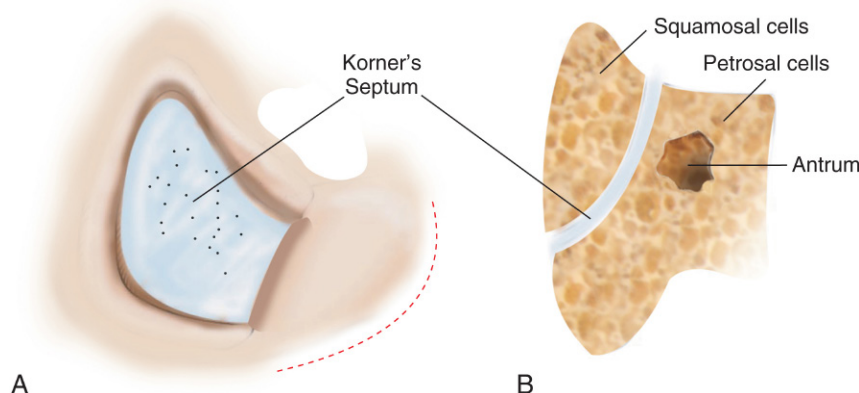


Figure 1.14. Korner's septum (A) as seen on mastoid exploration, (B) in coronal section of mastoid; in its presence there is difficulty in locating the antrum which lies deep to it.

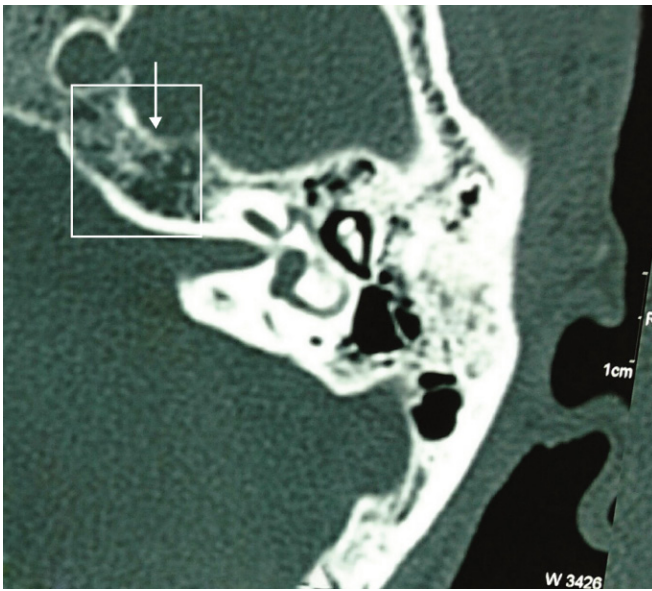


Figure 1.15. A pneumatized petrous apex.

- Superior route.** Various approaches are used. They are from the middle cranial fossa; through the arch of superior canal; through the attic region or the root of zygoma.
- Anterior route.** Anterior cell tract runs from the hypotympanum, anterior to the cochlea towards the petrous apex. Various approaches have earned the eponyms of Lempert, Ramadier or Eagleton approaches.

Another approach to petrous apex is the *translabyrinthine*, where the labyrinth is also removed. This results in total sensorineural loss and is used when useful hearing is already non-existent.

OSSICLES OF THE MIDDLE EAR

There are three ossicles in the middle ear—the malleus, incus and stapes (Figure 1.16).

The **malleus** has head, neck, handle (manubrium), a lateral and an anterior process. Head and neck of malleus lie in the attic. Manubrium is embedded in the fibrous layer of the tympanic membrane. The lateral process forms a knob-like projection on the outer surface of the tympanic membrane and gives attachment to the anterior and posterior malleal (malleolar) folds.

The **incus** has a body and a short process, both of which lie in the attic, and a long process which hangs vertically and attaches to the head of stapes.

The **stapes** has a head, neck, anterior and posterior crura, and a footplate. The footplate is held in the oval window by annular ligament.

The ossicles conduct sound energy from the tympanic membrane to the oval window and then to the inner ear fluid.

INTRATYMPANIC MUSCLES

There are two muscles—*tensor tympani* and the *stapedius*; the former attaches to the neck of malleus and tenses the tympanic membrane while the latter attaches to the neck of stapes and helps to dampen very loud

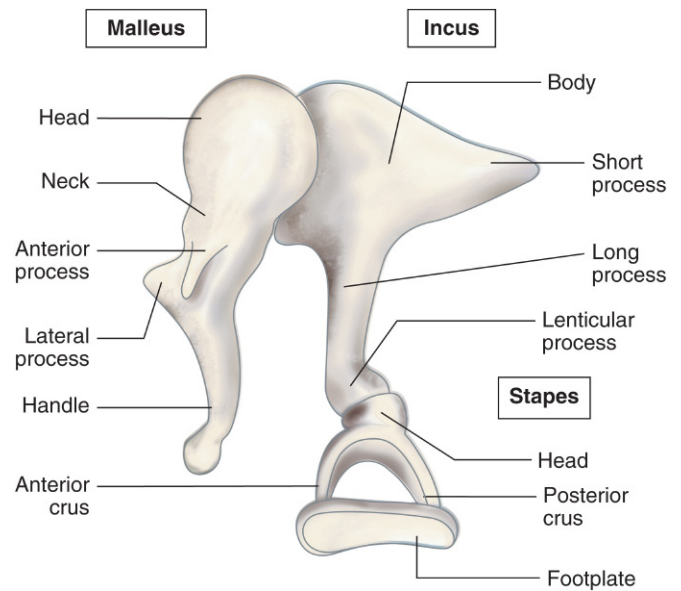


Figure 1.16. Ear ossicles and their parts.

sounds thus preventing noise trauma to the inner ear. Stapedius is a second arch muscle and is supplied by a branch of CN VII while tensor tympani develops from the first arch and is supplied by a branch of mandibular nerve (V_3).

TYMPANIC PLEXUS

It lies on the promontory and is formed by (i) tympanic branch of glossopharyngeal and (ii) sympathetic fibres from the plexus round the internal carotid artery. Tympanic plexus supplies innervation to the medial surface of the tympanic membrane, tympanic cavity, mastoid air cells and the bony eustachian tube. It also carries secretomotor fibres for the parotid gland. Section of tympanic branch of glossopharyngeal nerve can be carried out in the middle ear in cases of Frey's syndrome.

Course of secretomotor fibres to the parotid:

Inferior salivary nucleus → CN IX → Tympanic branch → Tympanic plexus → Lesser petrosal nerve → Otic ganglion → Auriculotemporal nerve → Parotid gland.

CHORDA TYMPANI NERVE

It is a branch of the facial nerve which enters the middle ear through posterior canaliculus, and runs on the medial surface of the tympanic membrane between the handle of malleus and long process of incus, above the attachment of tendon of tensor tympani. It carries taste from anterior two-thirds of tongue and supplies secretomotor fibres to the submaxillary and sublingual salivary glands (Figure 14.12, p.107).

LINING OF THE MIDDLE EAR CLEFT

Mucous membrane of the nasopharynx is continuous with that of the middle ear, aditus, antrum and the mastoid air cells. It wraps the middle ear structures—the ossicles, muscles, ligaments and nerves—like peritoneum

TABLE 1.1 LYMPHATIC DRAINAGE OF EAR

Area	Nodes
Concha, tragus, fossa triangularis and external cartilaginous canal	Preauricular and parotid nodes
Lobule and antitragus	Infra-auricular nodes
Helix and antihelix	Postauricular nodes, deep jugular and spinal accessory nodes
Middle ear and eustachian tube	Retropharyngeal nodes → upper jugular chain
Inner ear	No lymphatics

wraps various viscera in the abdomen—raising several folds and dividing the middle ear into various compartments. Middle ear contains nothing but the air; all the structures lie outside the mucous membrane.

Histologically, the eustachian tube is lined by ciliated epithelium, which is pseudostratified columnar in the cartilaginous part, columnar in the bony part with several mucous glands in the submucosa. Tympanic cavity is lined by ciliated columnar epithelium in its anterior and inferior part which changes to cuboidal type in the posterior part. Epitympanum and mastoid air cells are lined by flat, nonciliated epithelium.

BLOOD SUPPLY OF MIDDLE EAR

Middle ear is supplied by six arteries, out of which two are the main, i.e.

1. Anterior tympanic branch of maxillary artery which supplies tympanic membrane.
2. Stylomastoid branch of posterior auricular artery which supplies middle ear and mastoid air cells.

Four minor vessels are:

1. Petrosal branch of middle meningeal artery (runs along greater petrosal nerve).
2. Superior tympanic branch of middle meningeal artery traversing along the canal for tensor tympani muscle.
3. Branch of artery of pterygoid canal (runs along eustachian tube).
4. Tympanic branch of internal carotid.

Veins drain into pterygoid venous plexus and superior petrosal sinus.

LYMPHATIC DRAINAGE OF EAR

Lymphatic drainage of the ear is shown in Table 1.1. The inner ear doesn't have any lymphatics.

THE INTERNAL EAR

The internal ear or the labyrinth is an important organ of hearing and balance. It consists of a bony and a membranous labyrinth. The membranous labyrinth is filled with a clear fluid called *endolymph* while the space between membranous and bony labyrinths is filled with perilymph.

BONY LABYRINTH (FIGURE 1.17A)

It consists of three parts: the vestibule, the semicircular canals and the cochlea.

1. VESTIBULE. It is the central chamber of the labyrinth. In its lateral wall lies the oval window. The inside of its medial wall presents two recesses, a *spherical recess*, which lodges the saccule, and an *elliptical recess*, which lodges the utricle. Below the elliptical recess is the opening of aqueduct of vestibule through which passes the endolymphatic duct. In the posterosuperior part of vestibule are the five openings of semicircular canals (Figure 1.17C).

2. SEMICIRCULAR CANALS. They are three in number, the lateral, posterior and superior, and lie in planes at right angles to one another. Each canal has an ampullated end which opens independently into the vestibule and a nonampullated end. The nonampullated ends of posterior and superior canals unite to form a common channel called *crus commune*. Thus, the three canals open into the vestibule by five openings.

3. COCHLEA. The bony cochlea is a coiled tube making 2.5 to 2.75 turns round a central pyramid of bone called *modiolus*. The base of modiolus is directed towards internal acoustic meatus and transmits vessels and nerves to the cochlea. Around the modiolus and winding spirally

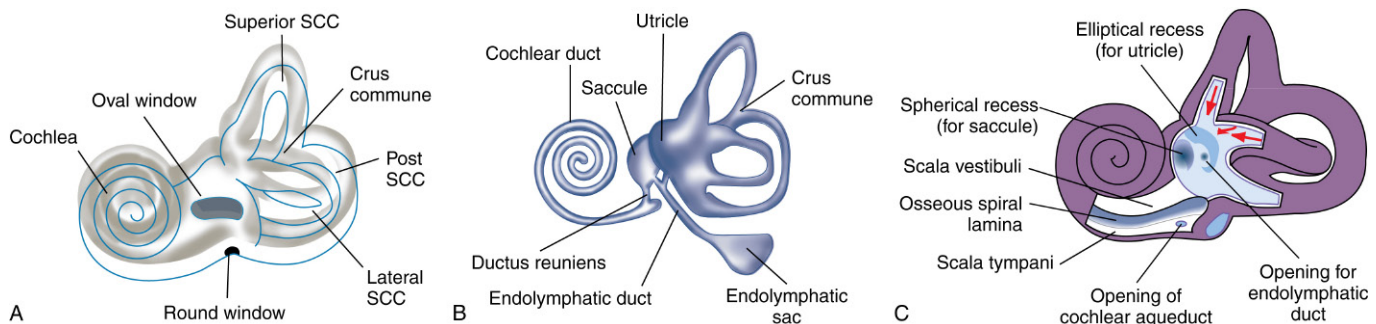


Figure 1.17. (A) Left bony labyrinth. (B) Left membranous labyrinth. (C) A cut section of the bony labyrinth.

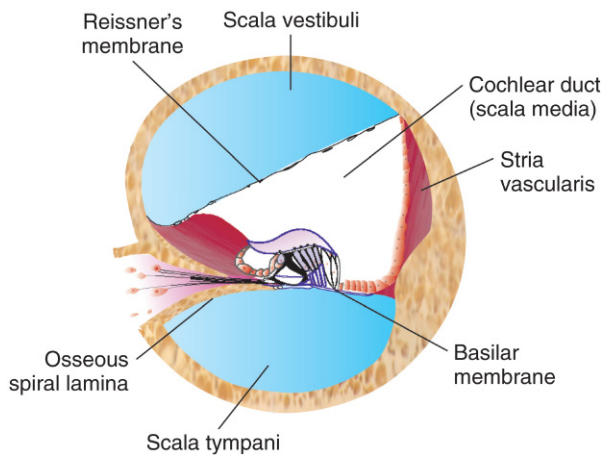


Figure 1.18. A section through the cochlea to show scala media (cochlear duct), scala vestibuli and scala tympani.

like the thread of a screw, is a thin plate of bone called *osseous spiral lamina*. It divides the bony cochlea incompletely and gives attachment to the basilar membrane. The bony bulge in the medial wall of middle ear, the promontory, is due to the basal coil of the cochlea. The bony cochlea contains three compartments:

- Scala vestibuli,
- Scala tympani,
- Scala media or the membranous cochlea (Figure 1.18).

The scala vestibuli and scala tympani are filled with perilymph and communicate with each other at the apex of cochlea through an opening called *helicotrema*. Scala vestibuli is closed by the footplate of stapes which separates it from the air-filled middle ear. The scala tympani is closed by secondary tympanic membrane; it is also connected with the subarachnoid space through the *aqueduct of cochlea* (Figure 1.19).

MEMBRANOUS LABYRINTH (FIGURE 1.17B)

It consists of the cochlear duct, the utricle and saccule, the three semicircular ducts, and the endolymphatic duct and sac.

1. COCHLEAR DUCT (FIGURE 1.18). Also called membranous cochlea or the scala media. It is a blind coiled tube.

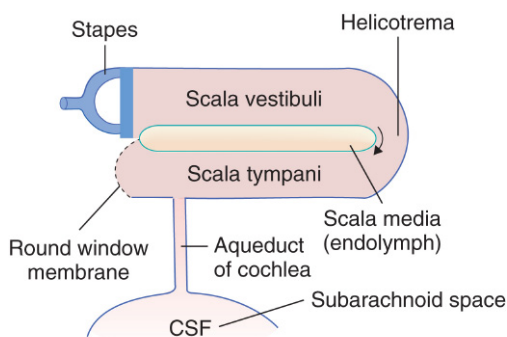


Figure 1.19. A diagrammatic representation of the perilymphatic system. CSF passes into the scala tympani through the aqueduct of the cochlea.

It appears triangular on cross-section and its three walls are formed by:

- the basilar membrane, which supports the organ of Corti;
- the Reissner's membrane, which separates it from the scala vestibule; and
- the stria vascularis, which contains vascular epithelium and is concerned with secretion of endolymph.

Cochlear duct is connected to the saccule by *ductus reuniens* (Figure 1.17B). The length of basilar membrane increases as we proceed from the basal coil to the apical coil. It is for this reason that higher frequencies of sound are heard at the basal coil while lower ones are heard at the apical coil.

2. UTRICLE AND SACCULE. The utricle lies in the posterior part of bony vestibule. It receives the five openings of the three semicircular ducts. It is also connected to the saccule through utriculosaccular duct. The sensory epithelium of the utricle is called *macula* and is concerned with linear acceleration and deceleration. The saccule also lies in the bony vestibule, anterior to the utricle and opposite the stapes footplate. Its sensory epithelium is also called *macula*. Its exact function is not known. It probably also responds to linear acceleration and deceleration. In Ménière's disease, the distended saccule lies against the stapes footplate and can be surgically decompressed by perforating the footplate.

3. SEMICIRCULAR DUCTS. They are three in number and correspond exactly to the three bony canals. They open in the utricle. The ampullated end of each duct contains a thickened ridge of neuroepithelium called *crista ampullaris*.

4. ENDOLYMPHATIC DUCT AND SAC. Endolymphatic duct is formed by the union of two ducts, one each from the saccule and the utricle. It passes through the vestibular aqueduct. Its terminal part is dilated to form endolymphatic sac, which lies between the two layers of dura on the posterior surface of the petrous bone.

Endolymphatic sac is surgically important. It is exposed for drainage or shunt operation in Ménière's disease.

INNER EAR FLUIDS AND THEIR CIRCULATION

There are two main fluids in the inner ear: perilymph and endolymph.

1. PERILYMPH. It resembles extracellular fluid and is rich in Na ions. It fills the space between the bony and the membranous labyrinth. It communicates with CSF through the aqueduct of cochlea which opens into the scala tympani near the round window. In fact this duct is not a direct communication but contains connective tissue resembling arachnoid through which perilymph percolates. There are two views regarding the formation of perilymph: (i) It is a filtrate of blood serum and is formed by capillaries of the spiral ligament and (ii) it is a direct continuation of CSF and reaches the labyrinth via aqueduct of cochlea.

TABLE 1.2 COMPOSITION OF INNER EAR FLUIDS

	Endolymph	Perilymph	CSF
Na ⁺ (mEq/L)	5	140	152
K ⁺ (mEq/L)	144	10	4
Protein (mg/dL)	126	200–400	20–50
Glucose (mg/dL)	10–40	85	70

Values are average and may differ slightly according to the site of collection of endolymph (cochlea, utricle, sac) and perilymph (scala tympani or scala vestibuli).

2. ENDOLYMPH. It fills the entire membranous labyrinth and resembles intracellular fluid, being rich in K ions. It is secreted by the secretory cells of the stria vascularis of the cochlea and by the *dark cells* (present in the utricle and also near the ampullated ends of semicircular ducts). There are two views regarding its flow: (i) longitudinal, i.e. endolymph from the cochlea reaches saccule, utricle and endolymphatic duct and gets absorbed through endolymphatic sac, which lies in the subdural space and (ii) radial, i.e. endolymph is secreted by stria vascularis and also gets absorbed by the stria vascularis. This view presumes that endolymphatic sac is a vestigial structure in man and plays no part in endolymph absorption. Composition of endolymph, perilymph and CSF is given in Table 1.2.

BLOOD SUPPLY OF LABYRINTH

The entire labyrinth receives its arterial supply through labyrinthine artery, which is a branch of anterior-inferior cerebellar artery but sometimes from the basilar. In the internal auditory canal it divides in the manner shown in Figures 1.20 and 1.21.

Venous drainage is through three veins, namely internal auditory vein, vein of cochlear aqueduct and vein of vestibular aqueduct, which ultimately drain into inferior petrosal sinus and lateral venous sinus.

It is to be noted that:

1. Blood supply to the inner ear is independent of blood supply to middle ear and bony otic capsule, and there is no cross circulation between the two.

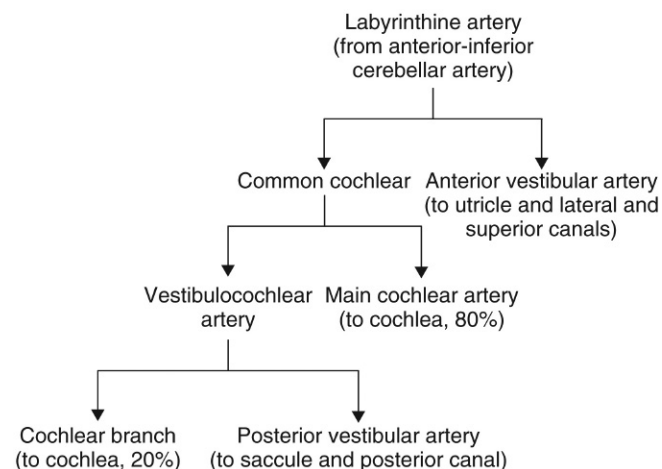


Figure 1.20. Divisions of the labyrinthine artery which supply blood to various parts of the labyrinth.

2. Blood supply to cochlea and vestibular labyrinth is segmental, therefore, independent ischaemic damage can occur to these organs causing either cochlear or vestibular symptoms.

DEVELOPMENT OF EAR

AURICLE. First branchial cleft is the precursor of external auditory canal. Around the 6th week of embryonic life, a series of six tubercles appear around the first branchial cleft. They progressively coalesce to form the auricle (Figure 1.22). Tragus develops from the tubercle of the first arch while the rest of the pinna develops from the remaining five tubercles of the second arch. Faulty fusion between the first and the second arch tubercles causes preauricular sinus or cyst, which is commonly seen between the tragus and crus of helix. By the 20th week, pinna achieves adult shape. Initially, the pinna is located low on the side of the neck and then moves on to a more lateral and cranial position.

EXTERNAL AUDITORY MEATUS. It develops from the first branchial cleft. By about the 16th embryonic week, cells proliferate from the bottom of ectodermal cleft and form a meatal plug. Recanalization of this plug forms the epithelial lining of the bony meatus. Recanalization begins from the deeper part near the tympanic membrane and progresses outwards, and that explains why deeper meatus is sometimes developed while there is atresia of canal in the outer part. External ear canal is fully formed by the 28th week of gestation.

TYMPANIC MEMBRANE. It develops from all the three germinal layers. Outer epithelial layer is formed by the ectoderm, inner mucosal layer by the endoderm and the middle fibrous layer by the mesoderm.

MIDDLE EAR CLEFT. The eustachian tube, tympanic cavity, attic, antrum and mastoid air cells develop from the endoderm of tubotympanic recess which arises from the first and partly from the second pharyngeal pouches (Figure 1.23).

Malleus and incus are derived from mesoderm of the first arch while the stapes develop from the second arch except its footplate and annular ligament which are derived from the otic capsule.

MEMBRANOUS INNER EAR. Development of the inner ear starts in the 3rd week of fetal life and is complete by the

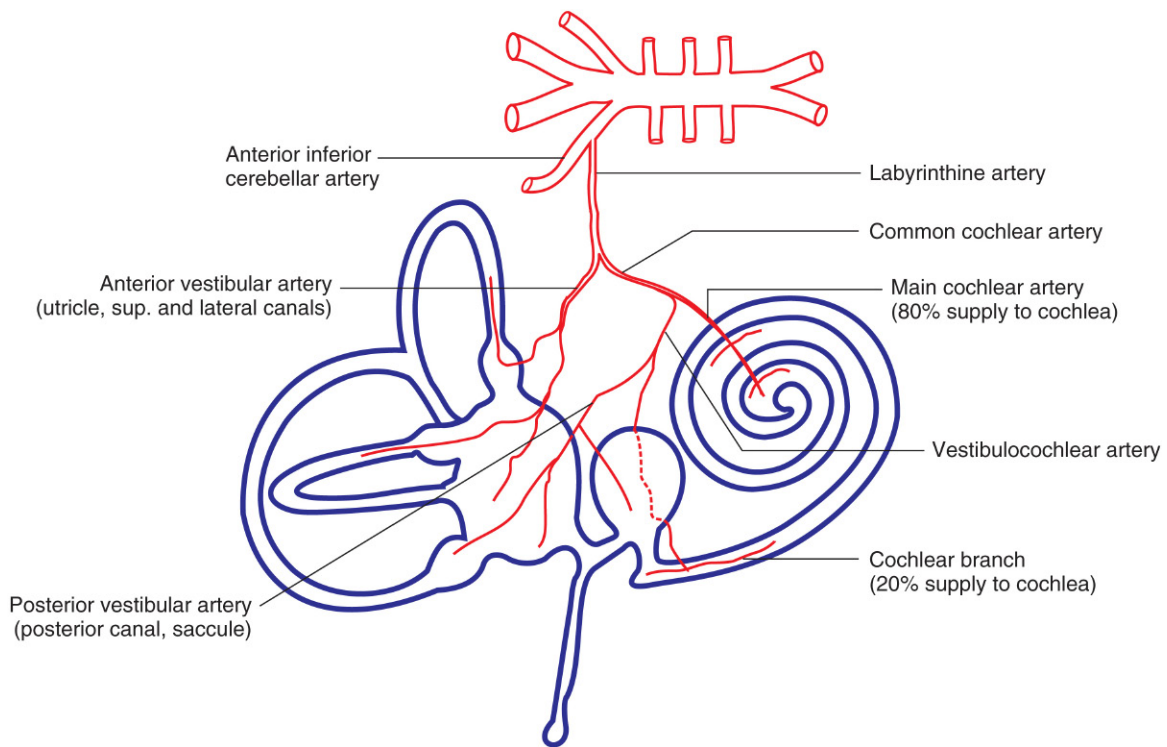


Figure 1.21. Blood supply of the labyrinth.

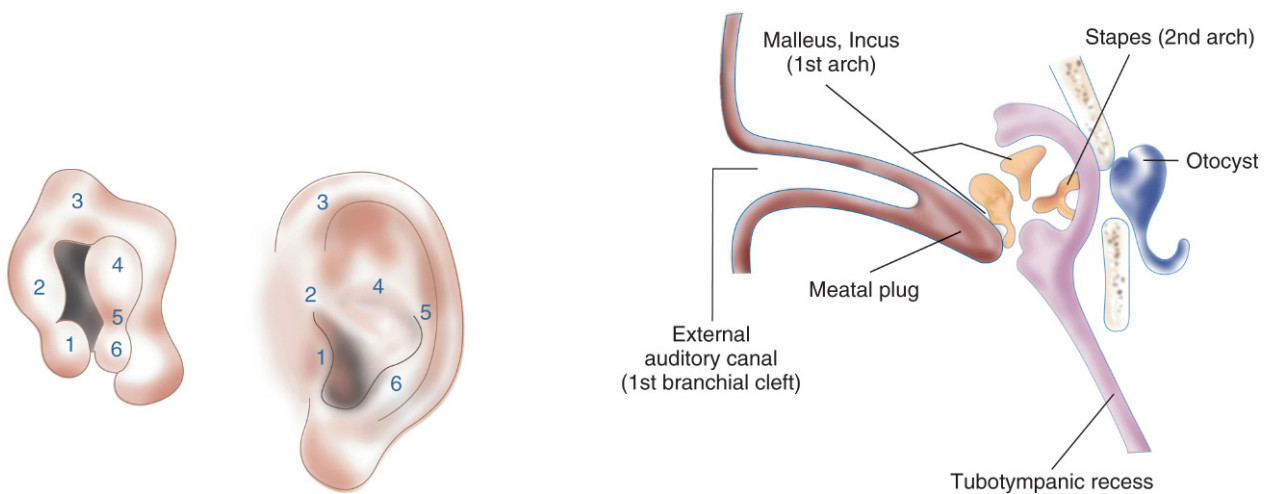


Figure 1.22. Development of pinna. Six hillocks of His around the first branchial cleft and the corresponding parts of pinna which develop from them.

Figure 1.23. Development of the external auditory canal and middle ear.

16th week. Ectoderm in the region of hindbrain thickens to form an *auditory placode*, which is invaginated to form auditory vesicle or the *otocyst*. The latter then differentiates into the endolymphatic duct and sac; the utricle, the semicircular ducts; and saccule and the cochlea. Development of phylogenetically older part of labyrinth—*pars superior* (semicircular canals and utricle) takes place earlier than *pars inferior* (saccule and cochlea).

The embryologic source and the time of development of external and middle ears are quite independent of the

development of the inner ear. It is therefore not unusual to see malformed and nonfunctional inner ear in the presence of normal external and middle ears, and vice versa.

The cochlea is developed sufficiently by 20 weeks of gestation (Table 1.3) and the fetus can hear in the womb of the mother. This probably explains how Abhimanyu, while still unborn, could have heard the conversation between his mother and father (Arjuna) in the legend given in the Great Indian epic of Mahabharata written thousands of years ago.

TABLE 1.3 TIMING OF DEVELOPMENT OF THE EAR IN THE WEEK OF GESTATION^a

Development	Pinna	Meatus	Middle ear	Vestibular labyrinth	Cochlea
Begins	6th	8th	3rd	3rd	3rd
Completes	20th	28th	30th	20th	20th

^aSource: Gulya AJ. Developmental Anatomy of the Ear. In: Glasscock and Shambaugh, editors. *Surgery of the Ear*. Philadelphia: W.B. Saunders Company, 1990.

Chapter 2

Peripheral Receptors and Physiology of Auditory and Vestibular Systems

AUDITORY SYSTEM

ORGAN OF CORTI (FIGURE 2.1)

Organ of Corti is the sense organ of hearing and is situated on the basilar membrane. Important components of the organ of Corti are:

1. TUNNEL OF CORTI. It is formed by the inner and outer rods. It contains a fluid called *cortilymph*. The exact function of the rods and cortilymph is not known.

2. HAIR CELLS. They are important receptor cells of hearing and transduce sound energy into electrical energy. Inner hair cells form a single row while outer hair cells are arranged in three or four rows. Inner hair cells are richly supplied by afferent cochlear fibres and are probably more important in the transmission of auditory impulses. Outer hair cells mainly receive efferent innervation from the olivary complex and are concerned with modulating the function of inner hair cells. Differences between inner and outer hair cells are given in Table 2.1.

3. SUPPORTING CELL. Deiters' cells are situated between the outer hair cells and provide support to the latter. Cells of Hensen lie outside the Deiters' cells.

4. TECTORIAL MEMBRANE. It consists of gelatinous matrix with delicate fibres. It overlies the organ of Corti. The shearing force between the hair cells and tectorial membrane produces the stimulus to hair cells.

NERVE SUPPLY OF HAIR CELLS

Ninety-five per cent of afferent fibres of spiral ganglion supply the inner hair cells while only five per cent supply the outer hair cells. Efferent fibres to the hair cells come from the olivocochlear bundle. Their cell bodies are situated in superior olivary complex. Each cochlea sends innervation to both sides of the brain.

AUDITORY NEURAL PATHWAYS AND THEIR NUCLEI (FIGURE 2.2)

Hair cells are innervated by dendrites of bipolar cells of spiral ganglion which is situated in Rosenthal's canal (canal running along the osseous spiral lamina). Axons of these bipolar cells form the cochlear division of CN VIII and end in the cochlear nuclei, the dorsal and ventral, on each side of the medulla. Further course of auditory

pathways is complex. From cochlear nuclei, the main nuclei in the ascending auditory pathways, sequentially, from below upwards are:

1. Superior olivary complex
2. Nucleus of lateral lemniscus
3. Inferior colliculus
4. Medial geniculate body
5. Auditory cortex

The auditory fibres travel via the ipsilateral and contralateral routes and have multiple decussation points. Thus each ear is represented in both cerebral hemispheres. The area of cortex, concerned with hearing is situated in the superior temporal gyrus (Brodmann's area 41). For auditory pathways, remember the mnemonic E.COLI-MA: Eighth nerve, Cochlear nuclei, Olivary complex, Lateral lemniscus, Inferior colliculus, Medial geniculate body and Auditory cortex.

PHYSIOLOGY OF HEARING

Any vibrating object causes waves of compression and rarefaction and is capable of producing sound. In the air, at 20°C and at sea level, sound travels at a speed of 344 m (1120 ft) per second. It travels faster in liquids and solids than in the air. Also, when sound energy has to pass from air to liquid medium, most of it is reflected because of the impedance offered by the liquid.

MECHANISM OF HEARING

A sound signal in the environment is collected by the pinna, passes through external auditory canal and strikes the tympanic membrane. Vibrations of the tympanic membrane are transmitted to stapes footplate through a chain of ossicles coupled to the tympanic membrane. Movements of stapes footplate cause pressure changes in the labyrinthine fluids, which move the basilar membrane. This stimulates the hair cells of the organ of Corti. It is these hair cells which act as transducers and convert the mechanical energy into electrical impulses, which travel along the auditory nerve. Thus, the mechanism of hearing can be broadly divided into:

1. Mechanical conduction of sound (conductive apparatus).
2. Transduction of mechanical energy to electrical impulses (sensory system of cochlea).
3. Conduction of electrical impulses to the brain (neural pathways).

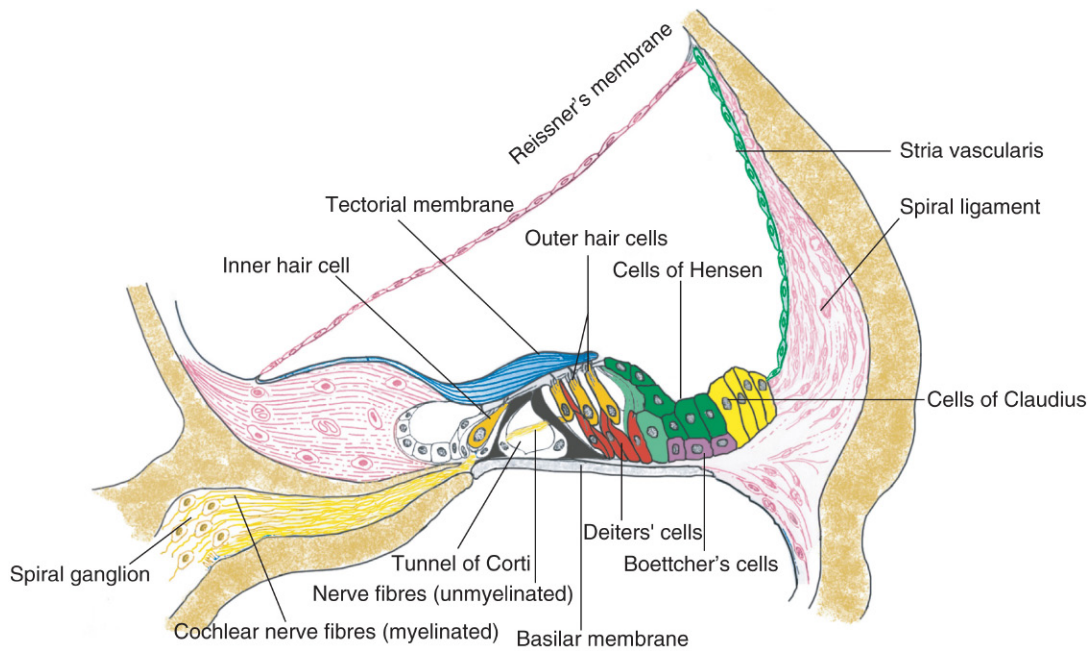


Figure 2.1. Structure of organ of Corti.

TABLE 2.1 DIFFERENCES BETWEEN INNER AND OUTER HAIR CELLS

	Inner hair cells	Outer hair cells
Total no.	3500	12,000
Rows	One row	Three or four rows
Shape	Flask shaped	Cylindrical
Nerve supply	Primarily afferent fibres and very few efferent	Mainly efferent fibres and very few afferent
Development	Develop earlier	Develop late
Function	Transmit auditory stimuli	Modulate function of inner hair cells
Vulnerability	More resistant	Easily damaged by ototoxic drugs and high intensity noise

1. Conduction of Sound

A person under water cannot hear any sound made in the air because 99.9% of the sound energy is reflected away from the surface of water because of the impedance offered by it. A similar situation exists in the ear when air-conducted sound has to travel to cochlear fluids. Nature has compensated for this loss of sound energy by interposing the middle ear which converts sound of greater amplitude but lesser force, to that of lesser amplitude but greater force. This function of the middle ear is called *impedance matching mechanism* or the *transformer action*.

It is accomplished by:

- (a) *Lever action of the ossicles.* Handle of malleus is 1.3 times longer than long process of the incus, providing a mechanical advantage of 1.3.
- (b) *Hydraulic action of tympanic membrane.* The area of tympanic membrane is much larger than the area of stapes footplate, the average ratio between the two being 21:1. As the effective vibratory area of tympanic membrane is only two-thirds, the effective areal ratio is reduced to 14:1, and this is the mechanical advantage provided by the tympanic membrane (Figure 2.3).

The product of areal ratio and lever action of ossicles is 18:1.

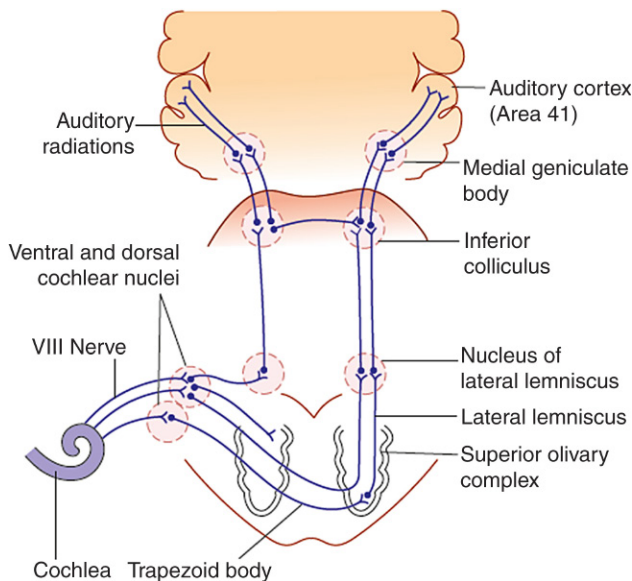


Figure 2.2. Auditory pathways from the right cochlea. Note bilateral route through brainstem and bilateral cortical representation.

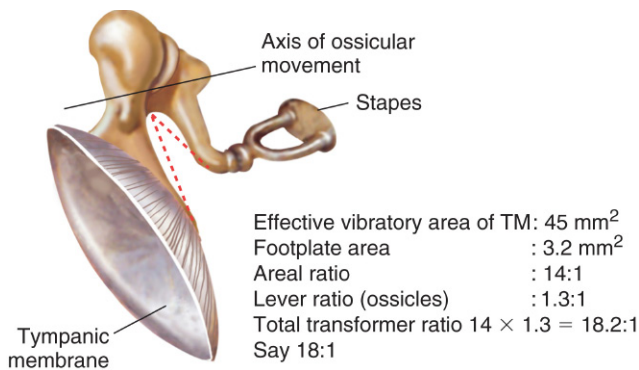


Figure 2.3. Transformer action of the middle ear. Hydraulic effect of tympanic membrane and lever action of ossicles combine to compensate the sound energy lost during its transmission from air to liquid medium.

According to some workers (Wever and Lawrence) out of a total of 90 mm² area of human tympanic membrane, only 55 mm² is functional and given the area of stapes footplate (3.2 mm²), the areal ratio is 17:1 and total transformer ratio (17 × 1.3) is 22.1.

- (c) *Curved membrane effect.* Movements of tympanic membrane are more at the periphery than at the centre where malleus handle is attached. This too provides some leverage.

PHASE DIFFERENTIAL BETWEEN OVAL AND ROUND WINDOWS. Sound waves striking the tympanic membrane do not reach the oval and round windows simultaneously. There is a preferential pathway to the oval window because of the ossicular chain. Thus, when oval window is receiving wave of compression, the round window is at the phase of rarefaction. If the sound waves were to strike both the windows simultaneously, they would cancel each other's effect with no movement of the perilymph and no hearing. This acoustic separation of windows is achieved by the presence of intact tympanic membrane and a cushion of air in the middle ear around the round window. Phase differential between the windows contributes 4 dB when tympanic membrane is intact.

NATURAL RESONANCE OF EXTERNAL AND MIDDLE EAR. Inherent anatomic and physiologic properties of the external and middle ear allow certain frequencies of sound to pass more easily to the inner ear due to their natural resonances. Natural resonance of external ear canal is 3000 Hz and that of middle ear 800 Hz. Frequencies most efficiently transmitted by ossicular chain are between 500 and 2000 Hz while that by tympanic membrane is 800–1600 Hz. Thus greatest sensitivity of the sound transmission is between 500 and 3000 Hz and these are the frequencies most important to man in day-to-day conversation (Table 2.2).

TABLE 2.2 NATURAL RESONANCE AND EFFICIENCY OF AUDITORY APPARATUS

External auditory canal	3000 Hz
Tympanic membrane	800–1600 Hz
Middle ear	800 Hz
Ossicular chain	500–2000 Hz

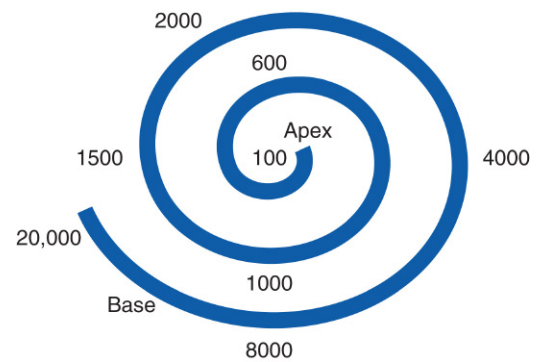


Figure 2.4. Frequency localization in the cochlea. Higher frequencies are localized in the basal turn and then progressively decrease towards the apex.

2. Transduction of Mechanical Energy to Electrical Impulses

Movements of the stapes footplate, transmitted to the cochlear fluids, move the basilar membrane and set up shearing force between the tectorial membrane and the hair cells. The distortion of hair cells gives rise to cochlear microphonics, which trigger the nerve impulse.

A sound wave, depending on its frequency, reaches maximum amplitude on a particular place on the basilar membrane and stimulates that segment (*travelling wave theory of von Bekesy*). Higher frequencies are represented in the basal turn of the cochlea and the progressively lower ones towards the apex (Figure 2.4).

3. Neural Pathways

Hair cells get innervation from the bipolar cells of spiral ganglion. Central axons of these cells collect to form the cochlear nerve which goes to the ventral and dorsal cochlear nuclei. From there, both crossed and uncrossed fibres travel to the superior olivary nucleus, lateral lemniscus, inferior colliculus, medial geniculate body and finally reach the auditory cortex of the temporal lobe (*Brodman's area 41 situated at the superior aspect of the temporal lobe along the floor of the lateral cerebral fissure*).

ELECTRICAL POTENTIALS OF COCHLEA AND CN VIII

Four types of potentials have been recorded; three from the cochlea and one from CN VIII fibres. They are:

- | | | |
|------------------------------|---|-------------------|
| 1. Endocochlear potential | } | from cochlea |
| 2. Cochlear microphonic | | |
| 3. Summating potential | | |
| 4. Compound action potential | — | from nerve fibres |

1. ENDOCOCHEAR POTENTIAL. It is a direct current (DC) potential recorded from scala media. It is +80 mV and is generated from the stria vascularis by Na⁺/K⁺-ATPase pump and provides source of energy for cochlear transduction (Figure 2.5). It is present at rest and does not require sound stimulus. This potential provides a sort of "battery" to drive the current through hair cells when they move in response to a sound stimulus.

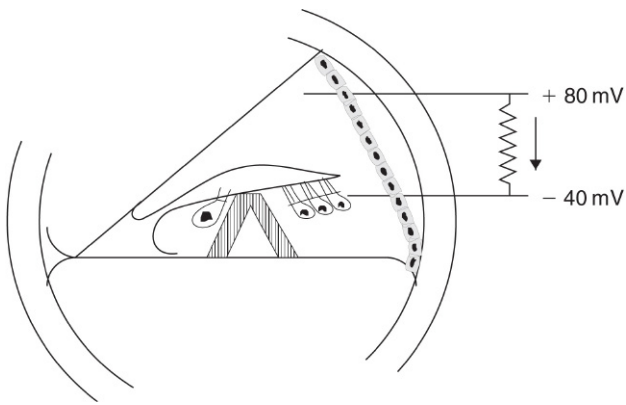


Figure 2.5. Davis' battery model of cochlear transduction. Scala media has a DC potential of 180 mV. Stimulation of hair cells produces intracellular potential of 240 mV. This provides flow of current of 120 mV through the top of hair cells.

2. COCHLEAR MICROPHONIC (CM). When basilar membrane moves in response to sound stimulus, electrical resistance at the tips of hair cells changes allowing flow of K^+ through hair cells and produces voltage fluctuations called cochlear microphonic. It is an alternating current (AC) potential.

3. SUMMATING POTENTIAL (SP). It is a DC potential and follows "envelope" of stimulating sound. It is produced by hair cells. It may be negative or positive. SP has been used in diagnosis of Ménière's disease. It is superimposed on VIII nerve action potential.

Both CM and SP are receptor potentials as seen in other sensory end-organs. They differ from action potentials in that: (i) they are graded rather than all or none phenomenon, (ii) have no latency, (iii) are not propagated and (iv) have no postresponse refractory period.

4. COMPOUND ACTION POTENTIAL. It is an all or none response of auditory nerve fibres.

VESTIBULAR SYSTEM

PERIPHERAL RECEPTORS

They are of two types:

1. Cristae

They are located in the ampullated ends of the three semicircular ducts. These receptors respond to angular acceleration.

2. Maculae

They are located in otolith organs (i.e. utricle and saccule). Macula of the utricle lies in its floor in a horizontal plane. Macula of the saccule lies in its medial wall in a vertical plane. They sense position of head in response to gravity and linear acceleration.

(A) STRUCTURE OF A CRISTA (FIGURE 2.6). It is a crest-like mound of connective tissues on which lie the sensory epithelial cells. The cilia of the sensory hair cells project into the cupula, which is a gelatinous mass extending

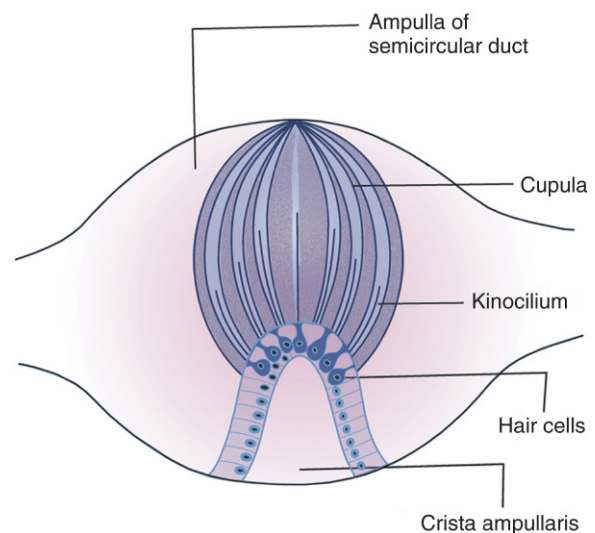


Figure 2.6. Structure of ampullary end of semicircular duct. Over the crista lie sensory hair cells interspersed with supporting cells. Hair from sensory cells project into the gelatinous substance of cupula.

from the surface of crista to the ceiling of the ampulla and forms a water tight partition, only to be displaced to one or the other side like a swing door, with movements of endolymph. The gelatinous mass of cupula consists of polysaccharide and contains canals into which project the cilia of sensory cells.

Hair cells are of two types (Figure 2.7). *Type I* cells are flask-shaped with a single large cup-like nerve terminal surrounding the base. *Type II* cells are cylindrical with multiple nerve terminals at the base. From the upper surface of each cell, project a single hair, the kinocilium and a number of other cilia, the stereocilia. The kinocilium is thicker and is located on the edge of the cell. Sensory cells are surrounded by supporting cells which show microvilli on their upper ends.

(B) STRUCTURE OF A MACULA. A macula consists mainly of two parts: (i) a sensory neuroepithelium, made up of type I and type II cells, similar to those in the crista; (ii) an otolithic membrane, which is made up of a gelatinous mass and on the top, the crystals of calcium carbonate called *otoliths* or *otoconia* (Figure 2.8). The cilia of hair cells

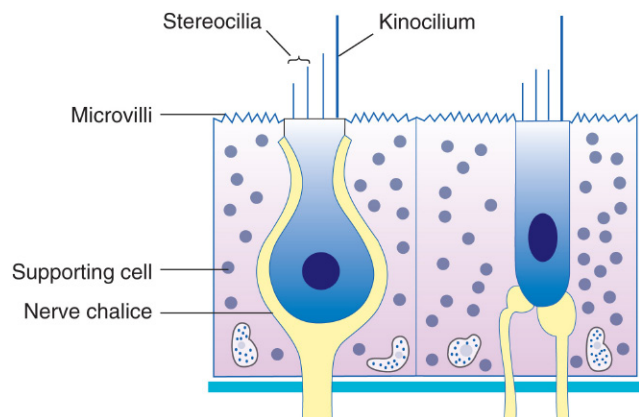


Figure 2.7. Sensory hair cells of the vestibular organs. Type I (left) and Type II (right).

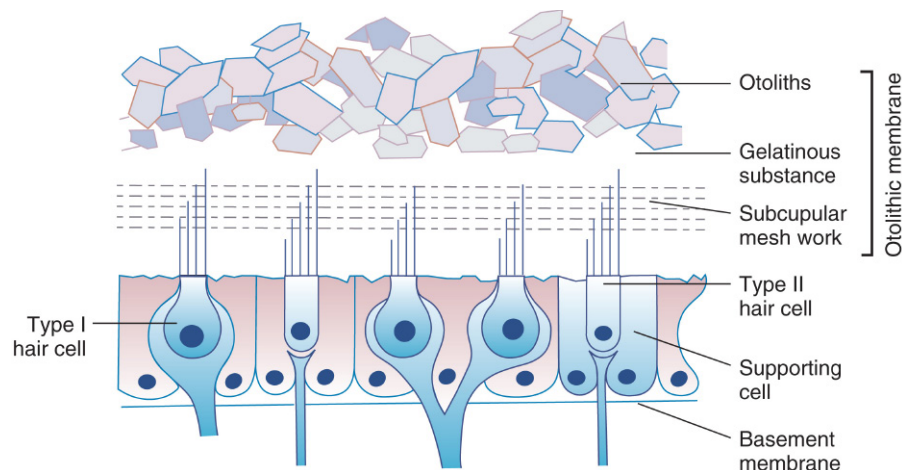


Figure 2.8. Structure of macula, the sensory end organ of the utricle and the saccule.

project into the gelatinous layer. The linear, gravitational and head tilt movements cause displacement of otolithic membrane and thus stimulate the hair cells which lie in different planes.

VESTIBULAR NERVE

Vestibular or Scarpa's ganglion is situated in the lateral part of the internal acoustic meatus. It contains bipolar cells. The distal processes of bipolar cells innervate the sensory epithelium of the labyrinth while its central processes aggregate to form the vestibular nerve.

CENTRAL VESTIBULAR CONNECTIONS

The fibres of vestibular nerve end in vestibular nuclei and some go to the cerebellum directly.

Vestibular nuclei are four in number, the superior, medial, lateral and descending. *Afferents* to these nuclei come from:

1. Peripheral vestibular receptors (semicircular canals, utricle and saccule)
2. Cerebellum
3. Reticular formation
4. Spinal cord
5. Contralateral vestibular nuclei

Thus, information received from the labyrinthine receptors is integrated with information from other somatosensory systems.

Efferents from vestibular nuclei go to:

1. Nuclei of CN III, IV, VI via medial longitudinal bundle. It is the pathway for vestibulo-ocular reflexes and this explains the genesis of nystagmus.
2. Motor part of spinal cord (vestibulospinal fibres). This coordinates the movements of head, neck and body in the maintenance of balance.
3. Cerebellum (vestibulocerebellar fibres). It helps to coordinate input information to maintain the body balance.
4. Autonomic nervous system. This explains nausea, vomiting, palpitation, sweating and pallor seen in vestibular disorders (e.g. Ménière's disease).

5. Vestibular nuclei of the opposite side.
6. Cerebral cortex (temporal lobe). This is responsible for subjective awareness of motion.

PHYSIOLOGY OF VESTIBULAR SYSTEM

Vestibular system is conveniently divided into:

1. *Peripheral*, which is made up of membranous labyrinth (semicircular ducts, utricle and saccule) and vestibular nerve.
2. *Central*, which is made up of nuclei and fibre tracts in the central nervous system to integrate vestibular impulses with other systems to maintain body balance.

SEMICIRCULAR CANALS

They respond to angular acceleration and deceleration. The three canals lie at right angles to each other but the one which lies at right angles to the axis of rotation is stimulated the most. Thus horizontal canal will respond maximum to rotation on the vertical axis and so on. Due to this arrangement of the three canals in three different planes, any change in position of head can be detected. Stimulation of semicircular canals produces nystagmus and the direction of nystagmus is determined by the plane of the canal being stimulated. Thus, nystagmus is horizontal from horizontal canal, rotatory from the superior canal and vertical from the posterior canal.

The stimulus to semicircular canal is flow of endolymph which displaces the cupula. The flow may be towards the cupula (ampullopetal) or away from it (ampullofugal), better called utriculopetal and utriculofugal. Ampullopetal flow is more effective than ampullofugal for the horizontal canal. The quick component of nystagmus is always opposite to the direction of flow of endolymph. Thus, if a person is rotated to the right for sometime and then abruptly stopped, the endolymph continues to move to the right due to inertia (i.e. ampullopetal for left canal), the nystagmus will be horizontal and directed to the left (Figure 2.9). Remember nystagmus is in the direction opposite to the direction of flow of endolymph.

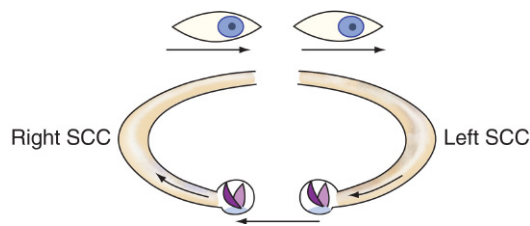


Figure 2.9. Rotation test. At the end of rotation to the right, semicircular canals stop but endolymph continues to move to the right, i.e. towards the left ampulla but away from the right, causing nystagmus to the left.

UTRICLE AND SACCULE

Utricle is stimulated by linear acceleration and deceleration or gravitational pull during the head tilts. The sensory hair cells of the macula lie in different planes and are stimulated by displacement of otolithic membrane during the head tilts.

The function of saccule is similar to that of utricle as the structure of maculae in the two organs is similar but experimentally, the saccule is also seen to respond to sound vibrations.

The vestibular system thus registers changes in the head position, linear or angular acceleration and deceleration, and gravitational effects. This information is sent to the central nervous system where information from other systems—visual, auditory, somatosensory (muscles, joints, tendons, skin)—is also received. All this information is integrated and used in the regulation of equilibrium and body posture.

Cerebellum, which is also connected to vestibular end organs, further coordinates muscle movements in their rate, range, force and duration and thus helps in the maintenance of balance.

MAINTENANCE OF BODY EQUILIBRIUM

A useful clinical approach to understand the physiology of equilibrium is to imagine that the balance system (vestibular, visual and somatosensory) is a two-sided push

and pull system. In static neutral position, each side contributes equal sensory information, i.e. push and pull system of one side is equal to that of the other side. If one side pulls more than the other, balance of the body is disturbed. During movement, i.e. turning or tilt, there is a temporary change in the push and pull system, which is corrected by appropriate reflexes and motor outputs to the eyes (vestibulo-ocular reflex), neck (vestibulocervical reflex), and trunk and limbs (vestibulospinal reflex) to maintain new position of head and body, but if any component of push and pull system of one side is disturbed for a longer time due to disease, vertigo and ataxia will develop.

VERTIGO AND DIZZINESS

Disorientation in space causes vertigo or dizziness and can arise from disorders of any of the three systems: vestibular, visual or somatosensory. Normally, the impulses reaching the brain from the three systems are equal and opposite. If any component on one side is inhibited or stimulated, the information reaching the cortex is mismatched, resulting in disorientation and vertigo. The vestibular inhibition on one side (e.g. acute vestibular failure, labyrinthectomy, Ménière's disease, VIIIth nerve section) causes vertigo. Similarly, stimulation of labyrinth by thermal or rotational stimulus causes vertigo. Dizziness can similarly result from the ocular causes, e.g. high errors of refraction or acute extraocular muscle paralysis with diplopia.

Vertigo and its causes are discussed in detail in [Chapter 7](#).

MOTION SICKNESS

It is characterized by nausea, vomiting, pallor and sweating during sea, air, bus or car travel in certain susceptible individuals. It can be induced by both real and apparent motion and is thought to arise from the mismatch of information reaching the vestibular nuclei and cerebellum from the visual, labyrinthine and somatosensory systems. It can be controlled by the usual labyrinthine sedatives.

Chapter 3

Audiology and Acoustics

This section aims to introduce certain terms which are frequently used in audiology and acoustics.

SOUND. It is a form of energy produced by a vibrating object. A sound wave consists of compression and rarefaction of molecules of the medium (air, liquid or solid) in which it travels. Velocity of sound is different in different media. In the air, at 20 °C, at sea level, sound travels 344 m (1120 ft) per second, and is faster in liquid and still faster in a solid medium.

FREQUENCY. It is the number of cycles per second. The unit of frequency is Hertz (Hz) named after the German scientist Heinrich Rudolf Hertz. A sound of 1000 Hz means 1000 cycles per second.

PURE TONE. A single frequency sound is called a pure tone, e.g. a sound of 250, 500 or 1000 Hz. In pure tone audiometry, we measure the threshold of hearing in decibels for various pure tones from 125 to 8000 Hz.

COMPLEX SOUND. Sound with more than one frequency is called a complex sound. Human voice is a complex sound.

PITCH. It is a subjective sensation produced by frequency of sound. Higher the frequency, greater is the pitch.

OVERTONES. A complex sound has a fundamental frequency, i.e. the lowest frequency at which a source vibrates. All frequencies above that tone are called the overtones. The latter determine the quality or the timbre of sound.

INTENSITY. It is the strength of sound which determines its loudness. It is usually measured in decibels. At a distance of 1 m, intensity of

Whisper	=	30 dB
Normal conversation	=	60 dB
Shout	=	90 dB
Discomfort of the ear	=	120 dB
Pain in the ear	=	130 dB

LOUDNESS. It is the subjective sensation produced by intensity. More the intensity of sound, greater the loudness.

DECIBEL (dB). It is 1/10th of a bel and is named after Alexander Graham Bell, the inventor of telephone. It is not an absolute figure but represents a logarithmic ratio between two sounds, namely the sound being described and the reference sound. Sound can be measured as

power, i.e. watts/cm² or as pressure, i.e. dynes/cm². In audiology, sound is measured as sound pressure level (SPL). It is compared with the reference sound which has an SPL of 0.0002 dynes/cm² or 20 μPa (micropascals), which roughly corresponds to the threshold of hearing in normal subjects at 1000 Hz. Decibel notation was introduced in audiology to avoid dealing with large figures of sound pressure level (0.0002 dynes/cm² at normal threshold of hearing to 200 dynes/cm² which causes pain in the ear. The latter is 1,000,000 times the former).

Formula for decibel is

$$\text{Sound in dB} = 10 \log \frac{\text{Power of } S_1}{\text{Power of } S_0}$$

S_1 = sound being described

S_0 = reference sound

$$\text{or } 10 \log \frac{(\text{SPL of } S_1)^2}{(\text{SPL of } S_0)^2}$$

(because power of sound is proportional to square of SPL)

$$\text{or } 20 \log \frac{\text{SPL of } S_1}{\text{SPL of } S_0}$$

If a sound has an SPL of 1000, i.e. (10³) times the reference sound, it is expressed as 20 × 3 = 60 dB. Similarly, a sound of 1,000,000, i.e. (10⁶) times the reference sound SPL is expressed simply as 120 dB and so on.

NOISE. It is defined as an aperiodic complex sound. There are three types of noise:

- White noise.** It contains all frequencies in audible spectrum and is comparable to the white light which contains all the colours of the visible spectrum. It is a broad-band noise and is used for masking.
- Narrow band noise.** It is white noise with certain frequencies, above and below the given noise, filtered out. Thus, it has a frequency range smaller than the broad-band white noise. It is used to mask the test frequency in pure tone audiometry.
- Speech noise.** It is a noise having frequencies in the speech range (300–3000 Hz). All other frequencies are filtered out.

MASKING. It is a phenomenon to produce inaudibility of one sound by the presentation of another. In clinical audiometry, one ear is kept busy by a sound while the other is being tested. Masking of nontest ear is essential in all bone conduction tests, but for air conduction tests, it is required only when difference of hearing between two ears exceeds 40 dB.

SOUND PRESSURE LEVEL. The SPL of a sound in decibels is 20 times the logarithm to the base 10, of the pressure of a sound to the reference pressure. The reference pressure is taken as $0.0002 \text{ dynes/cm}^2$ or $20 \text{ } \mu\text{Pa}$ (micropascals) for a frequency of 1000 Hz and represents the threshold of hearing in normally hearing young adults.

FREQUENCY RANGE IN NORMAL HEARING. A normal person can hear frequencies of 20–20,000 Hz but in routine audiometric testing only 125–8000 Hz are evaluated.

SPEECH FREQUENCIES. Frequencies of 500, 1000 and 2000 Hz are called *speech frequencies* as most of human voice falls within this range. PTA (pure tone average) is the average threshold of hearing in these three speech frequencies. It roughly corresponds to the speech reception threshold.

AUDIOMETRIC ZERO. Threshold of hearing, i.e. the faintest intensity which a normal healthy person can hear will vary from person to person. The International Standards Organization (ISO) adopted a standard for this, which is represented as the zero level on the audiometer. *According to ISO, audiometric zero is the mean value of minimal audible intensity in a group of normally hearing healthy young adults.*

HEARING LEVEL (HL). It is the sound pressure level produced by an audiometer at a specific frequency. It is measured in decibels with reference to audiometric zero. If an audiometer delivers a sound at 70 dB, it is represented as 70 dB HL.

SENSATION LEVEL (SL). It refers to the level of sound above the threshold of hearing for an individual. If someone is tested at 40dB SL, it means he was tested at 40 dB

above his threshold. For a normal person, this would be a sound of $0 + 40$, i.e. 40 dB HL, but for one with a hearing loss of say 30 dB, it would be $30 + 40$, i.e. 70 dB HL. In other words, *sensation level refers to the sound which will produce the same sensation, as in normally hearing person.* In speech audiometry, discrimination scores are tested at 30–40 dB SL. Stapedial reflex is elicited with a sound of 70–100 dB SL.

MOST COMFORTABLE LEVEL (MCL). It is the intensity level of sound that is most comfortable for the person.

LOUDNESS DISCOMFORT LEVEL. It is the level of sound which produces discomfort in the ear. Usually, it is 90–105 dB SL. It is important to find the loudness discomfort level of a person when prescribing a hearing aid.

DYNAMIC RANGE. It is the difference between the most comfortable level and the loudness discomfort level. The dynamic range is reduced in patients with positive recruitment phenomenon, as is the case in cochlear type of hearing loss.

SOUND LEVEL METER. It is an instrument to measure level of noise and other sounds. Sound level meters have different weighting networks (e.g. A, B or C) for different sensitivities at different frequencies. When describing a sound measured by a sound level meter, the weighting network must be indicated.

Noise levels are often expressed as dB(A) which refers to sound pressure level measured with 'A' network where the low and extremely high frequencies are given much less weightage compared to those in the middle range which are more important and are responsible for noise-induced hearing loss.

Chapter 4

Assessment of Hearing

Hearing loss can be of three types:

1. CONDUCTIVE HEARING LOSS. It is caused by any disease process interfering with the conduction of sound from the external ear to the stapediovestibular joint. Thus the cause may lie in the external ear (obstructions), tympanic membrane (perforation), middle ear (fluid), ossicles (fixation or disruption) or the eustachian tube (obstruction).

2. SENSORINEURAL (SN) HEARING LOSS. It results from lesions of the cochlea (sensory type) or VIIIth nerve and its central connections (neural type). The term *retrocochlear* is used when hearing loss is due to lesions of VIIIth nerve, and *central deafness*, when it is due to lesions of central auditory connections.

3. MIXED HEARING LOSS. In this type, elements of both conductive and sensorineural deafness are present in the same ear. There is air-bone gap indicating conductive element, and impairment of bone conduction indicating sensorineural loss. Mixed hearing loss is seen in some cases of otosclerosis and chronic suppurative otitis media.

While assessing the auditory function it is important to find out:

- Type of hearing loss* (conductive, sensorineural or mixed).
- Degree of hearing loss* (mild, moderate, moderately severe, severe, profound or total).
- Site of lesion.* If conductive, the lesion may be at external ear, tympanic membrane, middle ear, ossicles or eustachian tube. Clinical examination and tympanometry can be helpful to find the site of such lesions. If sensorineural, find out whether the lesion is cochlear, retrocochlear or central. Special tests of hearing will be required to differentiate these types.
- Cause of hearing loss.* The cause may be congenital, traumatic, infective, neoplastic, degenerative, metabolic, ototoxic, vascular or autoimmune process. Detailed history and laboratory investigations are required.

ASSESSMENT OF HEARING

Hearing of an individual can be tested by clinical and audiometric tests.

A. CLINICAL TESTS OF HEARING

1. Finger friction test
2. Watch test
3. Speech tests
4. Tuning fork tests

1. Finger Friction Test

It is a rough but quick method of screening and consists of rubbing or snapping the thumb and a finger close to patient's ear.

2. Watch Test

A clicking watch is brought close to the ear and the distance at which it is heard is measured. It had been popular as a screening test before the audiometric era but is practically obsolete now. Clicking watches are also obsolete.

3. Speech (Voice) Tests

Normally, a person hears conversational voice at 12 m (40 ft) and whisper (with residual air after normal expiration) at 6 m (20 ft) but for purposes of test, 6 m is taken as normal for both conversation and whisper.


The test is conducted in reasonably quiet surroundings. The patient stands with his test ear towards the examiner at a distance of 6 m. His eyes are shielded to prevent lip reading and the nontest ear is blocked by intermittent pressure on the tragus by an assistant. The examiner uses spondee words (e.g. black-night, football, daydream) or numbers with letters (X3B, 2AZ, M6D) and gradually walks towards the patient.

The distance at which conversational voice and the whispered voice are heard is measured. The disadvantage of speech tests is lack of standardization in intensity and pitch of voice used for testing and the ambient noise of the testing place.

4. Tuning Fork Tests

These tests are performed with tuning forks of different frequencies such as 128, 256, 512, 1024, 2048 and 4096 Hz, but for routine clinical practice, tuning fork of 512 Hz is ideal. Forks of lower frequencies produce sense of bone vibration while those of higher frequencies have a shorter decay time and are thus not routinely preferred.

A tuning fork is activated by striking it gently against the examiner's elbow, heel of hand or the rubber heel of the shoe.

To test air conduction (AC) (Figure 4.1 ) , a vibrating fork is placed vertically in line with the meatus, about 2 cm away from the opening of external auditory canal. The sound waves are transmitted through the tympanic membrane, middle ear and ossicles to the inner ear. Thus, by the air conduction test, the function of both the conducting mechanism and the cochlea are tested. Normally, hearing through air conduction is louder and heard twice as long as through the bone conduction route.

To test bone conduction (BC), the footplate of vibrating tuning fork is placed firmly on the mastoid bone. Cochlea is stimulated directly by vibrations conducted through

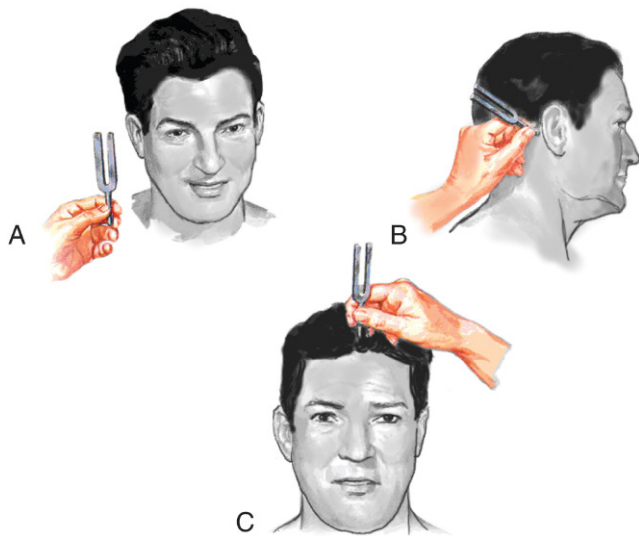


Figure 4.1. Tuning fork tests. (A) Testing for air conduction. (B) Testing for bone conduction. (C) Weber test.

 Scan to play Hearing Tests and Tuning Fork Test.

the skull bones. Thus, BC is a measure of the cochlear function only.

The clinically useful tuning fork tests include:

(A) RINNE TEST. In this test air conduction of the ear is compared with its bone conduction. A vibrating tuning fork is placed on the patient's mastoid and when he stops hearing, it is brought beside the meatus. If he still hears, AC is more than BC. Alternatively, the patient is asked to compare the loudness of sound heard through air and bone conduction. Rinne test is called positive when AC is longer or louder than BC. It is seen in normal persons or those having sensorineural deafness. A negative Rinne (BC > AC) is seen in conductive deafness. A negative Rinne indicates a minimum air-bone gap of 15–20 dB.

A prediction of air-bone gap can be made if tuning forks of 256, 512 and 1024 Hz are used.

- A Rinne test equal or negative for 256 Hz but positive for 512 Hz indicates air-bone gap of 20–30 dB.
- A Rinne test negative for 256 and 512 Hz but positive for 1024 Hz indicates air-bone gap of 30–45 dB.
- A Rinne negative for all the three tuning forks of 256, 512 and 1024 Hz indicates air-bone gap of 45–60 dB.

Remember that a negative Rinne for 256, 512 and 1024 Hz indicates a minimum AB gap of 15, 30, 45 dB, respectively.

FALSE NEGATIVE RINNE. It is seen in severe unilateral sensorineural hearing loss. Patient does not perceive any sound of tuning fork by air conduction but responds to bone conduction testing. This response to bone conduction is, in reality, from the opposite ear because of

transcranial transmission of sound. In such cases, correct diagnosis can be made by masking the nontest ear with Barany's noise box while testing for bone conduction. Weber test will further help as it gets lateralized to the better ear.

(B) WEBER TEST. In this test, a vibrating tuning fork is placed in the middle of the forehead or the vertex and the patient is asked in which ear the sound is heard. Normally, it is heard equally in both ears. It is lateralized to the worse ear in conductive deafness and to the better ear in sensorineural deafness. In weber test, sound travels directly to the cochlea via bone. Lateralization of sound in weber test with a tuning fork of 512 Hz implies a conductive loss of 15–25 dB in ipsilateral ear or a sensorineural loss in the contralateral ear.

(C) ABSOLUTE BONE CONDUCTION (ABC) TEST. Bone conduction is a measure of cochlear function. In ABC test, patient's bone conduction is compared with that of the examiner (presuming that the examiner has normal hearing). External auditory meatus of both the patient and examiner should be *occluded* (by pressing the tragus inwards) to prevent ambient noise entering through AC route. In conductive deafness, the patient and the examiner hear the fork for the same duration of time. In sensorineural deafness, the patient hears the fork for a shorter duration.

(D) SCHWABACH'S TEST. Here again BC of patient is compared with that of the normal hearing person (examiner) but meatus is *not occluded*. It has the same significance as absolute bone conduction test. Schwabach is reduced in sensorineural deafness and lengthened in conductive deafness.

Table 4.1 summarizes the interpretation of tuning fork tests.

(E) BING TEST. It is a test of bone conduction and examines the effect of *occlusion of ear canal* on the hearing. A vibrating tuning fork is placed on the mastoid while the examiner alternately closes and opens the ear canal by pressing on the tragus inwards. A normal person or one with sensorineural hearing loss hears louder when ear canal is occluded and softer when the canal is open (Bing positive). A patient with conductive hearing loss will appreciate no change (Bing negative).

(F) GELLE'S TEST. It is also a test of bone conduction and examines the effect of *increased air pressure* in ear canal on the hearing. Normally, when air pressure is increased in the ear canal by Siegel's speculum, it pushes the tympanic membrane and ossicles inwards, raises the intralabyrinthine pressure and causes immobility of basilar membrane and decreased hearing, but no change in hearing

TABLE 4.1 TUNING FORK TESTS AND THEIR INTERPRETATION

Test	Normal	Conductive deafness	SN deafness
Rinne	AC > BC (Rinne positive)	BC > AC (Rinne negative)	AC > BC
Weber	Not lateralized	Lateralized to poorer ear	Lateralized to better ear
ABC	Same as examiner's	Same as examiner's	Reduced
Schwabach	Equal	Lengthened	Shortened



Figure 4.2. Two-room audiometry setup. Audiometrician watches responses of the patient sitting across a glass partition.

is observed when ossicular chain is fixed or disconnected. Gelle's test is performed by placing a vibrating fork on the mastoid while changes in air pressure in the ear canal are brought about by Siegel's speculum. Gelle's test is positive in normal persons and in those with sensorineural hearing loss. It is negative when ossicular chain is fixed or disconnected. It was a popular test to find out stapes fixation in otosclerosis but has now been superseded by tympanometry.

B. AUDIOMETRIC TESTS

1. Pure Tone Audiometry

An audiometer is an electronic device which produces pure tones, the intensity of which can be increased or decreased in 5 dB steps (Figure 4.2). Usually air conduction thresholds are measured for tones of 125, 250, 500, 1000, 2000, 4000 and 8000 Hz and bone conduction thresholds for 250, 500, 1000, 2000 and 4000 Hz. The amount of intensity that has to be raised above the normal level is a measure of the degree of hearing impairment at that frequency. It is charted in the form of a graph called *audiogram*. The threshold of bone conduction is a measure of cochlear function. The difference in the thresholds of air and bone conduction (A-B gap) is a measure of the degree of conductive deafness. It may be noted that audiometer is so calibrated that the hearing of a normal person, both for air and bone conduction, is at 0 dB and there is no A-B gap, while tuning fork tests normally show AC > BC.

When difference between the two ears is 40 dB or above in air conduction thresholds, the better ear is masked to avoid getting a shadow curve from the nontest better ear. Similarly, masking is essential in all bone conduction studies. Masking is done by employing narrow-band noise to the nontest ear.

USES OF PURE TONE AUDIOGRAM

- (a) It is a measure of threshold of hearing by air and bone conduction and thus the degree and type of hearing loss.
- (b) A record can be kept for future reference.
- (c) Audiogram is essential for prescription of hearing aid.
- (d) Helps to find degree of handicap for medicolegal purposes.

(e) Helps to predict speech reception threshold.

2. Speech Audiometry

In this test, the patient's ability to hear and understand speech is measured. Two parameters are studied: (i) speech reception threshold and (ii) discrimination score.

(A) SPEECH RECEPTION THRESHOLD (SRT). It is the minimum intensity at which 50% of the words are repeated correctly by the patient. A set of spondee words (two syllable words with equal stress on each syllable, e.g. baseball, sunlight, daydream, etc.) is delivered to each ear through the headphone of an audiometer. The word lists are delivered in the form of recorded tapes or monitored voice and their intensity varied in 5 dB steps till half of them are correctly heard. Normally, SRT is within 10 dB of the average of pure tone threshold of three speech frequencies (500, 1000 and 2000 Hz). An SRT better than pure tone average by more than 10 dB suggests a functional hearing loss.

(B) SPEECH DISCRIMINATION SCORE. Also called *speech recognition* or *word recognition score*. It is a measure of patient's ability to understand speech. Here, a list of phonetically balanced (PB) words (single syllable words, e.g. pin, sin, day, bus, etc.) is delivered to the patient's each ear separately at 30–40 dB above his SRT and the percentage of words correctly heard by the patient is recorded. In normal persons and those with conductive hearing loss a high score of 90–100% can be obtained (Figure 4.3A, B and Table 4.2).

PERFORMANCE INTENSITY FUNCTION FOR PB WORDS

PB MAX. Instead of using a single suprathreshold intensity of 30–40 dB above SRT as described above, it is

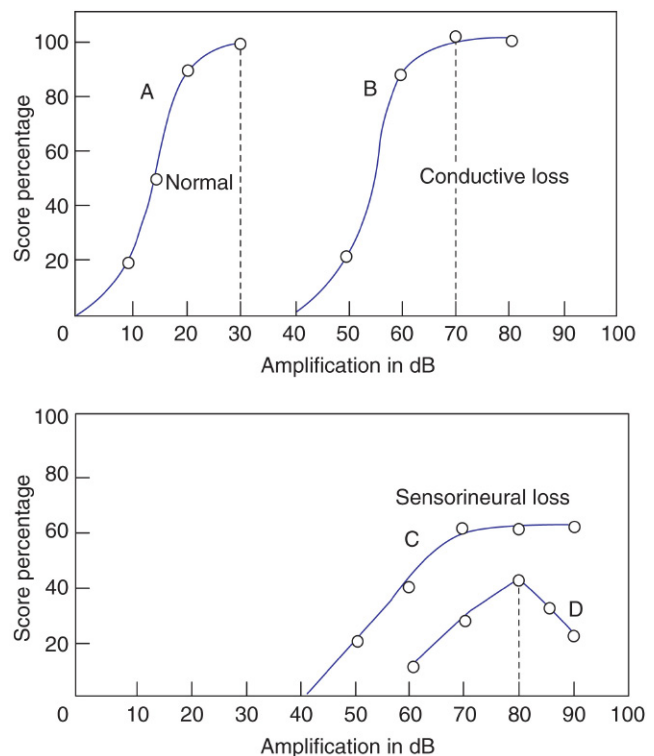


Figure 4.3. Speech audiogram.

TABLE 4.2 ABILITY TO UNDERSTAND SPEECH AND ITS RELATION TO SPEECH DISCRIMINATION (SD) SCORE A LIST OF 50 PB WORDS IS PRESENTED AND THE NUMBER CORRECTLY HEARD IS MULTIPLIED BY 2

SD score	Ability to understand speech
90–100%	Normal
76–88%	Slight difficulty
60–74%	Moderate difficulty
40–58%	Poor
<40%	Very poor

better to chart PB scores against several levels of speech intensity and find the maximum score (PB max) a person can attain. Also note the intensity of sound at which PB max is attained. It is a useful test clinically to set the volume of hearing aid (Figure 4.3C). Maximum volume of hearing aid should not be set above PB max.

ROLL OVER PHENOMENON. It is seen in retrocochlear hearing loss. With increase in speech intensity above a particular level, the PB word score falls rather than maintain a plateau as in cochlear type of sensorineural hearing loss (Figure 4.3D).

Thus speech audiometry is useful in several ways:

- (i) To find speech reception threshold which correlates well with average of three speech frequencies of pure tone audiogram.
- (ii) To differentiate organic from nonorganic (functional) hearing loss.
- (iii) To find the intensity at which discrimination score is best. This is helpful for fitting a hearing aid and setting its volume for maximum discrimination.
- (iv) To differentiate a cochlear from a retrocochlear sensorineural hearing loss.

3. Bekesy Audiometry

It is a self-recording audiometry where various pure tone frequencies automatically move from low to high while the patient controls the intensity through a button. Two tracings, one with continuous and the other with pulsed tone, are obtained. The tracings help to differentiate a cochlear from a retrocochlear and an organic from a functional hearing loss.

Various types of tracings obtained are:

- | | |
|----------|--|
| Type I | Continuous and pulsed tracings overlap. Seen in normal hearing or <i>conductive hearing loss</i> . |
| Type II | Continuous and pulsed tracings overlap up to 1000 Hz and then continuous tracing falls. Seen in <i>cochlear loss</i> . |
| Type III | Continuous tracing falls below pulsed tracing at 100–500 Hz even up to 40–50 dB. Seen in <i>retrocochlear/neural lesion</i> . |
| Type IV | Continuous tracing falls below pulsed lesion at frequencies up to 1000 Hz by more than 25 dB. Seen in <i>retrocochlear/neural lesion</i> . |
| Type V | Continuous tracing is above pulsed one. Seen in <i>nonorganic hearing loss</i> . |

Bekesy audiometry is seldom performed these days.

4. Impedance Audiometry (Figure 4.4)

It is an objective test, widely used in clinical practice and is particularly useful in children. It consists of:

- (a) Tympanometry
- (b) Acoustic reflex measurements

(A) TYMPANOMETRY. It is based on a simple principle, i.e. when a sound strikes tympanic membrane, some of the sound energy is absorbed while the rest is reflected. A stiffer tympanic membrane would reflect more of sound energy than a compliant one. By changing the pressures in a sealed external auditory canal and then measuring the reflected sound energy, it is possible to find the compliance or stiffness of the tympano-ossicular system and thus find the healthy or diseased status of the middle ear.

Essentially, the equipment consists of a probe which snugly fits into the external auditory canal and has three channels: (i) to deliver a tone of 220 Hz, (ii) to pick up the reflected sound through a microphone and (iii) to bring about changes in air pressure in the ear canal from positive to normal and then negative (Figure 4.5). By charting the compliance of tympano-ossicular system against various pressure changes, different types of graphs called



Figure 4.4. (A) Impedance audiometry in progress. (B) Impedance audiometer.

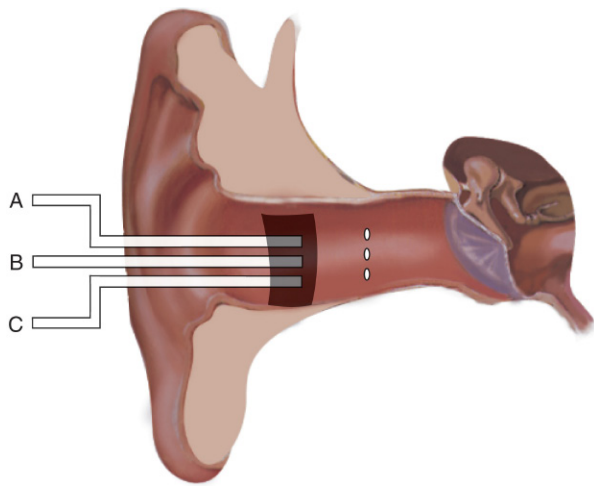


Figure 4.5. Principle of impedance audiometry. (A) Oscillator to produce a tone of 220 Hz. (B) Air pump to increase or decrease air pressure in the air canal. (C) Microphone to pick up and measure sound pressure level reflected from the tympanic membrane.

tympanograms are obtained which are diagnostic of certain middle ear pathologies.

Types of tympanograms (Figure 4.6)

Type A	Normal tympanogram.
Type As	Compliance is lower at or near ambient air pressure. Seen in fixation of ossicles, e.g. otosclerosis or malleus fixation.
Type Ad	High compliance at or near ambient pressure. Seen in ossicular discontinuity or thin and lax tympanic membrane.
Type B	A flat or dome-shaped graph. No change in compliance with pressure changes. Seen in middle ear fluid or thick tympanic membrane.
Type C	Maximum compliance occurs with negative pressure in excess of 100 mm H ₂ O. Seen in retracted tympanic membrane and may show some fluid in middle ear.

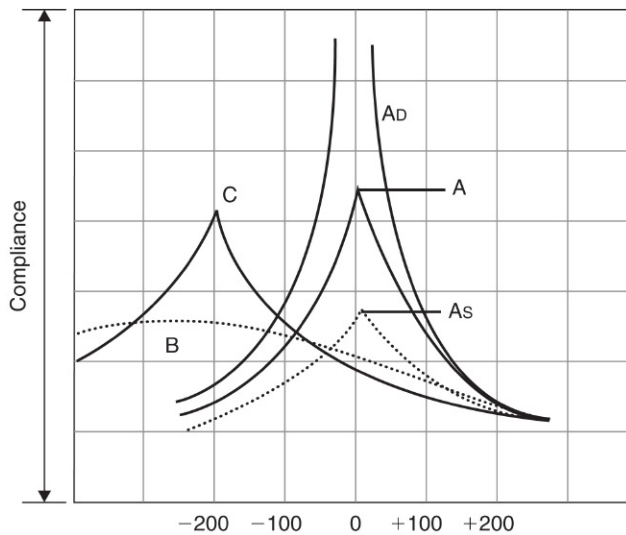


Figure 4.6. Types of tympanograms.

TESTING FUNCTION OF EUSTACHIAN TUBE. Tympanometry has also been used to find function of eustachian tube in cases of intact or perforated tympanic membrane. A negative or a positive pressure (–200 or +200 mm H₂O) is created in the middle ear and the person is asked to swallow five times in 20 s. The ability to equilibrate the pressure indicates normal tubal function. The test can also be used to find the patency of the grommet placed in the tympanic membrane in cases of serous otitis media.

(B) ACOUSTIC REFLEX. It is based on the fact that a loud sound, 70–100 dB above the threshold of hearing of a particular ear, causes bilateral contraction of the stapedial muscles which can be detected by tympanometry. Tone can be delivered to one ear and the reflex picked from the same or the contralateral ear. The reflex arc involved is:

Ipsilateral: CN VIII → ventral cochlear nucleus → CN VII nucleus ipsilateral stapedius muscle.

Contralateral: CN VIII → ventral cochlear nucleus → contralateral medial superior olivary nucleus → contralateral CN VII nucleus → contralateral stapedius muscle (Figure 4.7).

This test is useful in several ways:

- (i) To test the hearing in infants and young children. It is an objective method.
- (ii) To find malingerers. A person who feigns total deafness and does not give any response on pure tone audiometry but shows a positive stapedial reflex is a malingerer.
- (iii) To detect cochlear pathology. Presence of stapedial reflex at lower intensities, e.g. 40–60 dB than the usual 70 dB indicates recruitment and thus a cochlear type of hearing loss.
- (iv) To detect VIIIth nerve lesion. If a sustained tone of 500 or 1000 Hz, delivered 10 dB above acoustic reflex threshold, for a period of 10 s, brings the reflex amplitude to 50%, it shows abnormal adaptation and is indicative of VIIIth nerve lesion (stapedial reflex decay).
- (v) Lesions of facial nerve. Absence of stapedial reflex when hearing is normal indicates lesion of the facial nerve, proximal to the nerve to stapedius. The reflex can also be used to find prognosis of facial paralysis as the appearance of reflex, after it was absent, indicates return of function and a favourable prognosis.
- (vi) Lesion of brainstem. If ipsilateral reflex is present but the contralateral reflex is absent, lesion is in the area of crossed pathways in the brainstem.

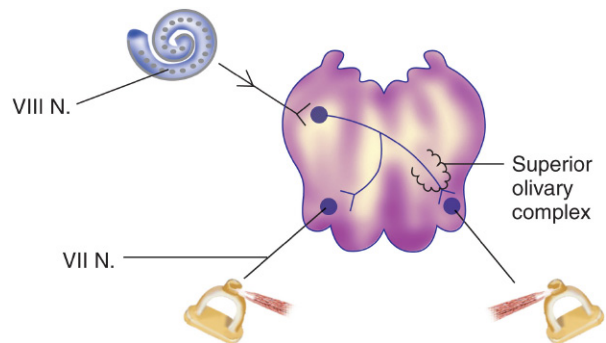


Figure 4.7. Acoustic reflex.

PHYSICAL VOLUME OF EAR CANAL. Acoustic immittance can also measure the physical volume of air between the probe tip and tympanic membrane. Normally it is up to 1.0 mL in children and 2 mL in adults. Any increase in volume, >2 mL in children and >2.5 mL in adults, indicates perforation of the tympanic membrane (because middle ear volume is added up to the volume of external ear canal). This has also been used to find patency of the ventilation tube.

C. SPECIAL TESTS OF HEARING

1. Recruitment

It is a phenomenon of abnormal growth of loudness. The ear which does not hear low intensity sound begins to hear greater intensity sounds as loud or even louder than normal hearing ear. Thus, a loud sound which is tolerable in normal ear may grow to abnormal levels of loudness in the recruiting ear and thus becomes intolerable. The patients with recruitment are poor candidates for hearing aids. Recruitment is typically seen in lesions of the cochlea (e.g. Ménière’s disease and presbycusis) and thus helps to differentiate a cochlear from a retrocochlear sensorineural hearing loss.

Alternate binaural loudness balance test is used to detect recruitment in unilateral cases. A tone, say of 1000 Hz, is played alternately to the normal and the affected ear and the intensity in the affected ear is adjusted to match the loudness in normal ear. The test is started at 20 dB above the threshold of deaf ear and then repeated at every 20 dB rise until the loudness is matched or the limits of audiometer reached. In conductive and neural deafness, the initial difference is maintained throughout while in cochlear lesions, partial, complete or over-recruitment may be seen (Figure 4.8).

2. Short Increment Sensitivity Index (SISI Test)

Patients with cochlear lesions distinguish smaller changes in intensity of pure tone better than normal persons and those with conductive or retrocochlear pathology. SISI test is thus used to differentiate a cochlear from a retrocochlear lesion.

In this test, a continuous tone is presented 20 dB above the threshold and sustained for about 2 min. Every 5 s, the tone is increased by 1 dB and 20 such blips are presented. Patient indicates the blips heard. In conductive deafness,

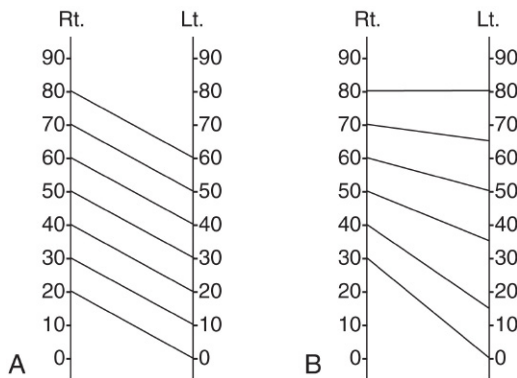


Figure 4.8. Alternate binaural loudness balance test.

SISI score is seldom more than 15%; it is 70–100% in cochlear deafness and 0–20% in nerve deafness.

3. Threshold Tone Decay Test

It is a measure of nerve fatigue and is used to detect retrocochlear lesions. Normally, a person can hear a tone continuously for 60 s. In nerve fatigue, he stops hearing earlier. The threshold tone decay test is simple and is performed in the following manner:

A tone of 4000 Hz is presented at 5 dB above the patient’s threshold of hearing, continuously for a period of 60 s. If patient stops hearing earlier, intensity is increased by another 5 dB. The procedure is continued till patient can hear the tone continuously for 60 s, or no level exists above the threshold where tone is audible for full 60 s. The result is expressed as number of dB of decay. A decay more than 25 dB is diagnostic of a retrocochlear lesion.

4. Evoked Response Audiometry

It is an objective test which measures electrical activity in the auditory pathways in response to auditory stimuli. It requires special equipment with an averaging computer. There are several components of evoked electric response but only two have gained clinical acceptance. They are:

- (a) **Electrocochleography (EcoG).** It measures electrical potentials arising in the cochlea and CN VIII in response to auditory stimuli within first 5 ms. The response is in the form of three phenomena: cochlear microphonics, summing potentials and the action potential of VIIIth nerve. The recording electrode is usually a thin needle passed through the tympanic membrane onto the promontory. In adults, it can be done under local anaesthesia but in children or anxious persons sedation or general anaesthesia is required. Sedation does not interfere in these responses. EcoG is useful (i) to find threshold of hearing in young infants and children

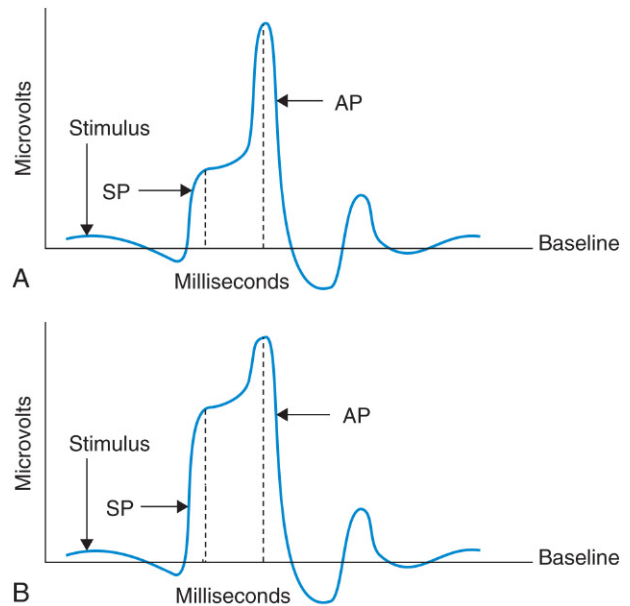


Figure 4.9. Electrocochleography. (A) Normal ear. (B) Ear with Ménière’s disease. Voltage of summing potential (SP) is compared with that of action potential (AP). Normally SP is 30% of AP. This ratio is enhanced in Ménière’s disease.

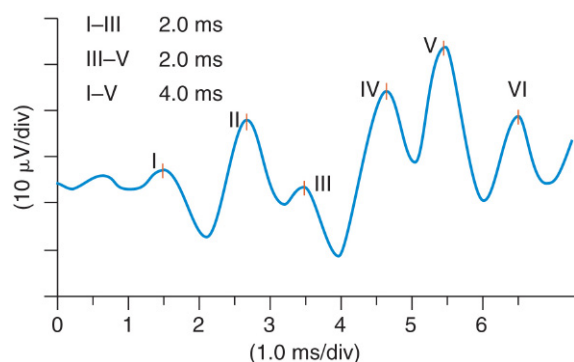


Figure 4.10. Brainstem auditory evoked potentials.

within 5–10 dB and (ii) to differentiate lesions of cochlea from those of the VIIIth nerve (Figure 4.9).

- (b) **Auditory brainstem response (ABR).** Also called BAER or BAEP (brainstem auditory evoked response or potential) or BERA (brainstem evoked response audiometry) is to elicit brainstem responses to auditory stimulation by clicks or tone bursts. It is a noninvasive technique to find the integrity of central auditory pathways through the VIIIth nerve, pons and midbrain. In this method, electrical potentials are generated in response to several click stimuli or tone bursts and picked up from the vertex by surface electrodes. It measures hearing sensitivity in the range of 1000–4000 Hz. In a normal person, seven waves are produced in the first 10 ms. The first, third and fifth waves are most stable and are used in measurements. The waves are studied for absolute *latency*, *interwave latency* (usually between wave I and V) and the *amplitude* (Figure 4.10).

The exact anatomic site of neural generators for various waves is disputed but the latest studies indicate the following sites:

Wave I	Distal part of CN VIII
Wave II	Proximal part of CN VIII near the brainstem
Wave III	Cochlear nucleus
Wave IV	Superior olivary complex
Wave V	Lateral lemniscus
Waves VI and VII	Inferior colliculus

As an aide memorie remember the mnemonic EE COLI (eight, eight, cochlear nucleus, olivary complex, lateral lemniscus, inferior colliculus) *compare* E COLI-MA in pathways of hearing.

ABR is used:

- (i) As a screening procedure for infants.
- (ii) To determine the threshold of hearing in infants; also in children and adults who do not cooperate and in malingersers.
- (iii) To diagnose retrocochlear pathology particularly acoustic neuroma.
- (iv) To diagnose brainstem pathology, e.g. multiple sclerosis or pontine tumours.
- (v) To monitor CN VIII intraoperatively in surgery of acoustic neuromas to preserve the function of cochlear nerve.

5. Auditory Steady State Response (ASSR)

Though ABR, conducted with tone bursts of various frequencies, can produce frequency-specific thresholds of hearing in infants to fit hearing aid at a young age, it has limitations. It cannot test hearing losses above 80 dB. It cannot detect hearing sensitivity in severe to profoundly deaf infants. ASSR is useful in such situations. It is, like ABR, an electrophysiological test which uses steady state pure tone signals instead of transient signals of tone bursts or clicks used in ABR. The steady state signals are also modulated rapidly in amplitude and frequency and thus gives a frequency-specific audiogram. Hearing losses exceeding 80 dB can be detected. It can help in selection of children for cochlear implantation at an early age.

6. Otoacoustic Emissions (OAEs)

They are low-intensity sounds produced by outer hair cells of a normal cochlea and can be elicited by a very sensitive microphone placed in the external ear canal and analyzed by a computer. Sound produced by outer hair cells travels in a reverse direction: outer hair cells → basilar membrane → perilymph → oval window → ossicles → tympanic membrane → ear canal. OAEs are present when outer hair cells are healthy and are absent when they are damaged and thus help to test the function of cochlea. They do not disappear in VIIIth nerve pathology as cochlear hair cells are normal.

TYPES OF OAEs. Broadly OAEs are of two types: spontaneous and evoked. The latter are elicited by a sound stimulus.

- (a) *Spontaneous OAEs.* They are present in healthy normal hearing persons where hearing loss does not exceed 30 dB. They may be absent in 50% of normal persons.
- (b) *Evoked OAEs.* They are further divided into two types depending on the sound stimulus used to elicit them.
 - (i) *Transient evoked OAEs (TEOAEs).* Evoked by clicks. A series of click stimuli are presented at 80–85 dB SPL (sound pressure level) and response recorded.
 - (ii) *Distortion product OAEs (DPOAEs).* Two tones are simultaneously presented to the cochlea to produce distortion products. They have been used to test hearing in the range of 1000–8000 Hz.

USES

- (a) OAEs are used as a screening test of hearing in neonates and to test hearing in uncooperative or mentally challenged individuals after sedation. Sedation does not interfere with OAEs.
- (b) They help to distinguish cochlear from retrocochlear hearing loss. OAEs are absent in cochlear lesions, e.g. ototoxic sensorineural hearing loss. They detect ototoxic effects earlier than pure tone audiometry.
- (c) OAEs are also useful to diagnose retrocochlear pathology, especially auditory neuropathy. Auditory neuropathy is a neurologic disorder of CN VIII. Audiometric tests, e.g. SNHL for pure tones, impaired speech discrimination score, absent or abnormal auditory brainstem response, show a retrocochlear type of lesion but OAEs are normal.

OAEs are absent in 50% of normal individuals, lesions of cochlea, middle ear disorders (as sound travelling in reverse direction cannot be picked up) and when hearing loss exceeds 30 dB.

7. Central Auditory Tests

Patients with central auditory disorders have difficulty in hearing in noisy surroundings or when the speech is distorted and not clearly spoken. Three different types of speech discrimination tests are used.

- (a) *Monotic test*. It is presented with speech message which is distorted. Patients with lesions of brain and cortex have difficulty to understand the message.
- (b) *Dichotic test*. Two different speech messages are presented simultaneously, one to each ear and patient is

asked to identify both. Staggered spondaic word test is the one more often used. Pairs of spondaic words along with digits or nonsense words are simultaneously presented to the ears. Patients with temporal lobe lesions will have difficulty identifying these words when presented to the ear opposite to that of the side of lesion.

- (c) *Binaural tests*. They are used to identify integration of information from both ears. Such tests are normal in cortical lesions but affected in lesions of brainstem and thus help to localize the site of lesion. Most common test used is binaural masking level difference test.

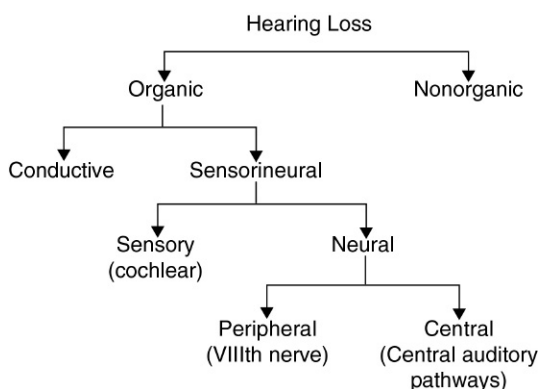
Central auditory tests are not used routinely.

8. Hearing Assessment in Infants and Children (see p. 132)

Chapter 5

Hearing Loss

CLASSIFICATION



Scan to play Types of Hearing Loss.

CONDUCTIVE HEARING LOSS AND ITS MANAGEMENT

Any disease process which interferes with the conduction of sound to reach cochlea causes conductive hearing loss. The lesion may lie in the external ear and tympanic membrane, middle ear or ossicles up to stapediovestibular joint.

The characteristics of conductive hearing loss are:

1. Negative Rinne test, i.e. BC > AC.
2. Weber lateralized to poorer ear.
3. Normal absolute bone conduction.
4. Low frequencies affected more.
5. Audiometry shows bone conduction better than air conduction with air-bone gap. Greater the air-bone gap, more is the conductive loss (Figure 5.1).
6. Loss is not more than 60 dB.
7. Speech discrimination is good.

AETIOLOGY

The cause may be congenital (Table 5.1) or acquired (Table 5.2).

AVERAGE HEARING LOSS SEEN IN DIFFERENT LESIONS OF CONDUCTIVE APPARATUS

1. Complete obstruction of ear canal:	30 dB
2. Perforation of tympanic membrane (It varies and is directly proportional to the size of perforation):	10–40 dB
3. Ossicular interruption with intact drum:	54 dB

4. Ossicular interruption with perforation:	38 dB
5. Malleus fixation:	10–25 dB
6. Closure of oval window:	60 dB

Note here that ossicular interruption with intact drum causes more loss than ossicular interruption with perforated drum.

MANAGEMENT

Most cases of conductive hearing loss can be managed by medical or surgical means. Treatment of these conditions is discussed in respective sections. Briefly, it consists of:

1. **Removal of canal obstructions**, e.g. impacted wax, foreign body, osteoma or exostosis, keratotic mass, benign or malignant tumours, or meatal atresia.
2. **Removal of fluid**. Myringotomy with or without grommet insertion.
3. **Removal of mass from middle ear**. Tympanotomy and removal of small middle ear tumours or cholesteatoma behind intact tympanic membrane.
4. **Stapedectomy**, as in otosclerotic fixation of stapes footplate.
5. **Tympanoplasty**. Repair of perforation, ossicular chain or both.
6. **Hearing aid**. In cases, where surgery is not possible, refused or has failed.

Tympanoplasty

It is an operation to (i) eradicate disease in the middle ear and (ii) to reconstruct hearing mechanism. It may be combined with mastoidectomy if disease process so demands. Type of middle ear reconstruction depends on the damage present in the ear. The procedure may be limited only to repair of tympanic membrane (*myringoplasty*), or to reconstruction of ossicular chain (*ossiculoplasty*), or both (*tympanoplasty*). Reconstructive surgery of the ear has been greatly facilitated by development of operating microscope, microsurgical instruments and biocompatible implant materials.

From the physiology of hearing mechanism, the following principles can be deduced to restore hearing surgically:

- (a) *An intact tympanic membrane*, to provide large hydraulic ratio between the tympanic membrane and stapes footplate.
- (b) *Ossicular chain*, to conduct sound from tympanic membrane to the oval window.
- (c) *Two functioning windows*, one on the scala vestibuli (to receive sound vibrations) and the other on the scala

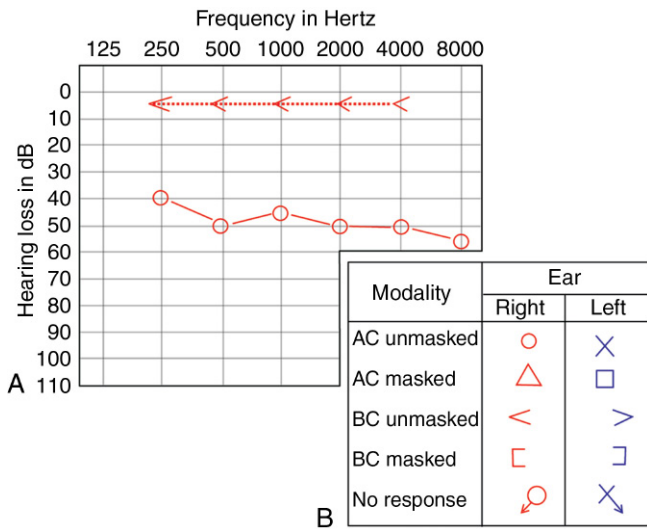


Figure 5.1. (A) Audiogram of right ear showing conductive hearing loss with A-B gap. (B) Symbols used in audiogram charting.

TABLE 5.1 CONGENITAL CAUSES OF CONDUCTIVE HEARING LOSS

- Meatal atresia
- Fixation of stapes footplate
- Fixation of malleus head
- Ossicular discontinuity
- Congenital cholesteatoma

TABLE 5.2 ACQUIRED CAUSES OF CONDUCTIVE HEARING LOSS

External ear	Any obstruction in the ear canal, e.g. wax, foreign body, furuncle, acute inflammatory swelling, benign or malignant tumour or atresia of canal.
Middle ear	(a) Perforation of tympanic membrane, traumatic or infective (b) Fluid in the middle ear, e.g. acute otitis media, serous otitis media or haemotympanum (c) Mass in middle ear, e.g. benign or malignant tumour (d) Disruption of ossicles, e.g. trauma to ossicular chain, chronic suppurative otitis media, cholesteatoma (e) Fixation of ossicles, e.g. otosclerosis, tympanosclerosis, adhesive otitis media (f) Eustachian tube blockage, e.g. retracted tympanic membrane, serous otitis media

tympani (to act as a relief window). If it is only one window, as in stapes fixation or closure of round window, there will be no movement of cochlear fluids resulting in conductive hearing loss.

- (d) *Acoustic separation of two windows*, so that sound does not reach both the windows simultaneously. It can be achieved by providing an intact tympanic membrane, preferential pathway to one window (usually the oval) by providing ossicular chain and by the presence of air in the middle ear.

- (e) *Functioning eustachian tube*, to provide aeration to the middle ear.
(f) *A functioning sensorineural apparatus*, i.e. the cochlea and VIIIth nerve.

TYPES OF TYMPANOPLASTY. Wullstein classified tympanoplasty into five types (Figure 5.2).

- Type I** Defect is perforation of tympanic membrane which is repaired with a graft. It is also called myringoplasty.
- Type II** Defect is perforation of tympanic membrane with erosion of malleus. Graft is placed on the incus or remnant of malleus.
- Type III** Malleus and incus are absent. Graft is placed directly on the stapes head. It is also called *myringostapediopexy* or *columella tympanoplasty*.
- Type IV** Only the footplate of stapes is present. It is exposed to the external ear, and graft is placed between the oval and round windows. A narrow middle ear (cavum minor) is thus created to have an air pocket around the round window. A mucosa-lined space extends from the eustachian tube to the round window. Sound waves in this case act directly on the footplate while the round window has been shielded.
- Type V** Stapes footplate is fixed but round window is functioning. In such cases, another window is created on horizontal semicircular canal and covered with a graft. Also called *fenestration operation*.

Several modifications have appeared in the above classification and they mainly pertain to the types of ossicular reconstruction.

MYRINGOPLASTY. It is repair of tympanic membrane. Graft materials of choice are temporalis fascia or the perichondrium taken from the patient. Sometimes, homografts such as dura, vein, fascia or cadaver tympanic membrane are also used. Repair can be done by two techniques—the underlay or the overlay. In the *underlay technique*, margins of perforation are freshened and the graft placed medial to perforation or tympanic annulus, if large, and is supported by gelfoam in the middle ear (Figure 5.3A). In the *overlay technique*, the graft is placed lateral to fibrous layer of the tympanic membrane after carefully removing all squamous epithelium from the lateral surface of tympanic membrane remnant (Figure 5.3B and Chapter 83).

OSSICULAR RECONSTRUCTION. Ossicles are essential for transmission of sound from tympanic membrane to labyrinth. Several types of prosthesis are available to replace ossicles depending on the ossicular defects (Table 5.3). Autograft ossicles can be sculptured to bridge the gap. Homograft preserved ossicles with or without tympanic membrane have been used but are difficult to procure and have danger of transmission of disease (Figure 5.4).

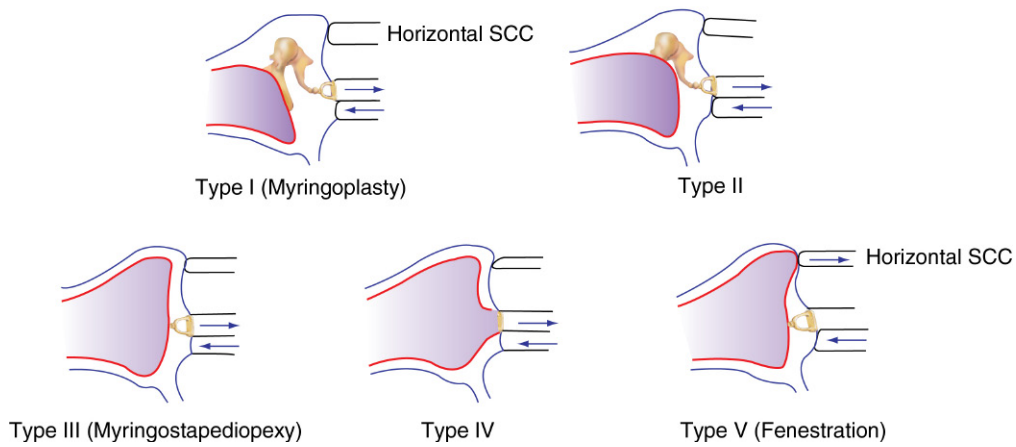


Figure 5.2. Types of tympanoplasty. The graft is progressively in contact with malleus (type I), incus (type II), stapes (type III), stapes footplate (type IV), or fenestra in horizontal semicircular canal (type V). In classical type IV, the graft was attached to promontory, this provides sound protection for round window while footplate was directly exposed.

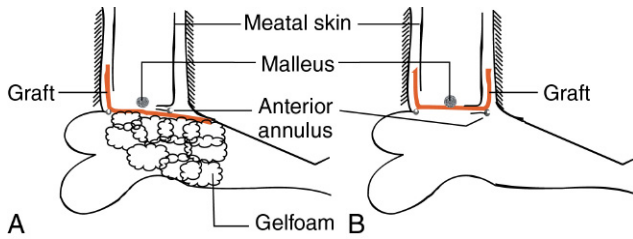


Figure 5.3. Myringoplasty. (A) Underlay technique—fascia graft is under the anterior annulus. It is supported by gelfoam in the middle ear to prevent medial displacement. (B) Overlay technique—fascia graft lies lateral to anterior annulus onto the anterior bony canal wall. It is placed medial to malleus handle to prevent lateralization.

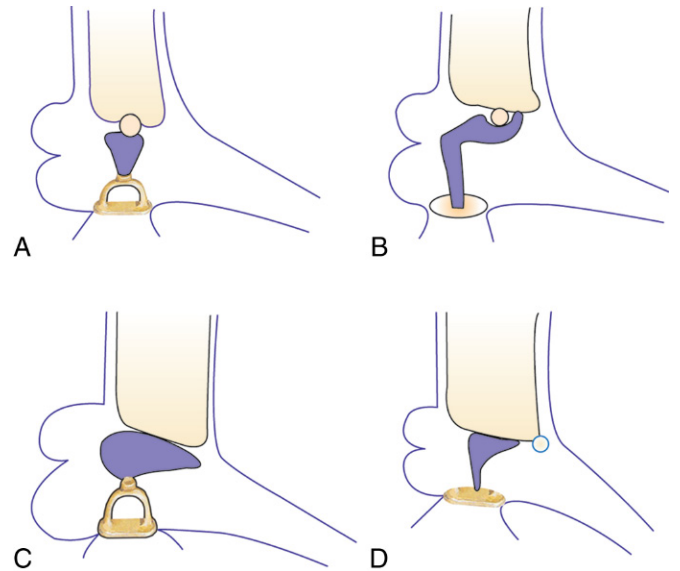


Figure 5.4. Ossicular reconstruction. Sculptured autograft or homograft ossicles have been used. (A) Malleus—Stapes assembly. Modified incus graft connecting malleus handle with stapes head. (B) Malleus—Footplate assembly. Modified malleus connecting malleus handle with stapes footplate. (C) Modified incus connecting tympanic membrane (TM) to stapes head. Malleus is missing. (D) Modified incus connecting TM to stapes footplate.

TABLE 5.3 MATERIALS USED FOR OSSICULAR RECONSTRUCTION

Type of graft	Material	Remarks
Autograft	<ul style="list-style-type: none"> Incus, head of malleus Cortical bone from mastoid 	<ul style="list-style-type: none"> Risk of harbouring disease Low cost
Allograft	<ul style="list-style-type: none"> Plastipore (polyethylene sponge) Hydroxyapatite (HA) implants Titanium implants Glass isomer Teflon prosthesis HA HA (50%) + Titanium (50%) HA + Silicon (flex-HA) HA + Polyethylene (HAPEX) 	<ul style="list-style-type: none"> Easily available Readymade Easy to store and use Costly Likely to be extruded
Homograft	<ul style="list-style-type: none"> Preserved ossicles only Ossicles with tympanic membrane 	<ul style="list-style-type: none"> Difficult to procure Risk of disease transmission

At the time of ossicular reconstruction in chronic otitis media, one should ensure:

- Middle ear is healthy and free of mucosal disease and cholesteatoma.
- Eustachian tube function is good. Atelectatic middle ear shows poor eustachian tube function.

In cases of canal wall-up mastoidectomy done for cholesteatoma or active mucosal disease, the procedure is delayed for about 6 months to ensure ear is free of disease.

Primary ossicular reconstruction can be performed in:

- Traumatic ossicular disruption
- Fixation of ossicles
- Canal wall down procedures when there is no mucosal disease or cholesteatoma

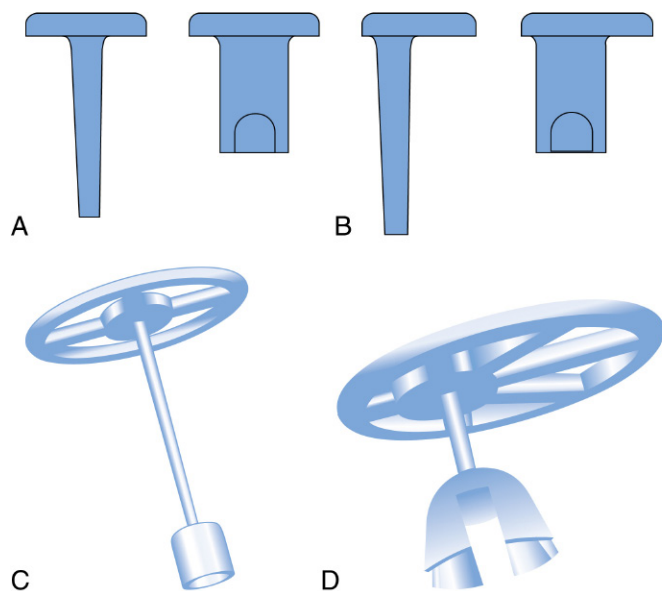


Figure 5.5. Hydroxyapatite TORP and PORP. (A) Centred and (B) off-set types. (C) Titanium TORP. (D) Titanium PORP.

Types of prosthesis (Figure 5.5)

1. **Incus prosthesis.** Used when incus is missing but handle of malleus and stapes with superstructure are present and functional.
2. **Incus–stapes prosthesis.** Used when incus and stapes superstructure are missing. Malleus and stapes footplate are functional.
3. **Partial ossicular replacement prosthesis (PORP).** Used when malleus and incus are absent. Stapes is present and mobile. PORP is placed between tympanic membrane and stapes head.
4. **Total ossicular replacement prosthesis (TORP).** Used when malleus, incus and stapes superstructure are absent. Only the stapes footplate is present and is mobile.

SENSORINEURAL HEARING LOSS AND ITS MANAGEMENT

Sensorineural hearing loss (SNHL) results from lesions of the cochlea, VIIIth nerve or central auditory pathways. It may be present at birth (congenital) or start later in life (acquired).

The characteristics of sensorineural hearing loss are:

1. A positive Rinne test, i.e. AC > BC.
2. Weber lateralized to better ear.
3. Bone conduction reduced on Schwabach and absolute bone conduction tests.
4. More often involving high frequencies.
5. No gap between air and bone conduction curve on audiometry (Figure 5.6).
6. Loss may exceed 60 dB.
7. Speech discrimination is poor.
8. There is difficulty in hearing in the presence of noise.

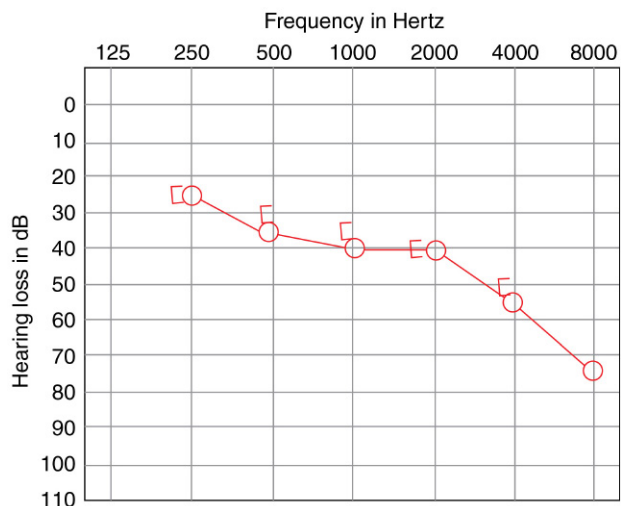


Figure 5.6. Audiogram of right ear showing sensorineural loss with no A–B gap.

AETIOLOGY

Congenital

It is present at birth and is the result of anomalies of the inner ear or damage to the hearing apparatus by prenatal or perinatal factors (see p. 129).

Acquired

It appears later in life. The cause may be genetic or non-genetic. The genetic hearing loss may manifest late (delayed onset) and may affect only the hearing, or be a part of a larger syndrome affecting other systems of the body as well (syndromal). Common causes of acquired SNHL include:

1. Infections of labyrinth—viral, bacterial or spirochaetal
2. Trauma to labyrinth or VIIIth nerve, e.g. fractures of temporal bone or concussion of the labyrinth or the ear surgery
3. Noise-induced hearing loss
4. Ototoxic drugs
5. Presbycusis
6. Ménière's disease
7. Acoustic neuroma
8. Sudden hearing loss
9. Familial progressive SNHL
10. Systemic disorders, e.g. diabetes, hypothyroidism, kidney disease, autoimmune disorders, multiple sclerosis, blood dyscrasias.

DIAGNOSIS

1. HISTORY. It is important to know whether disease is congenital or acquired, stationary or progressive, associated with other syndromes or not, involvement of other members of the family and possible aetiological factors.

2. SEVERITY OF DEAFNESS (MILD, MODERATE, MODERATELY SEVERE, SEVERE, PROFOUND OR TOTAL). This can be found out on audiometry.

3. TYPE OF AUDIOGRAM. Whether loss is high frequency, low frequency, mid-frequency or flat type.

4. SITE OF LESION. i.e. cochlear, retrocochlear or central.

5. LABORATORY TESTS. They depend on the aetiology suspected, e.g. X-rays or CT scan of temporal bone for evidence of bone destruction (congenital cholesteatoma, glomus tumour, middle ear malignancy or acoustic neuroma), blood counts (leukaemia), blood sugar (diabetes), serology for syphilis, thyroid functions (hypothyroidism), kidney function tests, etc.

MANAGEMENT

Early detection of SNHL is important as measures can be taken to stop its progress, reverse it or to start an early rehabilitation programme, so essential for communication.

Syphilis of the inner ear is treatable with high doses of penicillin and steroids with improvement in hearing. Hearing loss of *hypothyroidism* can be reversed with replacement therapy. *Serous labyrinthitis* can be reversed by attention to middle ear infection. Early management of *Ménière's disease* can prevent further episodes of vertigo and hearing loss. SNHL due to *perilymph fistula* can be corrected surgically by sealing the fistula in the oval or round window with fat.

Ototoxic drugs should be used with care and discontinued if causing hearing loss. In many such cases, it may be possible to regain hearing, total or partial, if the drug is stopped. *Noise-induced hearing loss* can be prevented from further deterioration if the person is removed from the noisy surroundings.

Rehabilitation of hearing impaired with hearing aids and other devices is discussed in [Chapter 20](#).

SPECIFIC FORMS OF HEARING LOSS

A. INFLAMMATIONS OF LABYRINTH

It may be viral, bacterial or syphilitic.

1. VIRAL LABYRINTHITIS. Viruses usually reach the inner ear by blood stream affecting stria vascularis and then the endolymph and organ of Corti. Measles, mumps and cytomegaloviruses are well-documented to cause labyrinthitis. Several other viruses, e.g. rubella, herpes zoster, herpes simplex, influenza and Epstein-Barr are clinically known to cause deafness but direct proof of their invasion of labyrinth is lacking.

2. BACTERIAL. Bacterial infections reach labyrinth through the middle ear (tympanogenic) or through CSF (meningogenic). Labyrinthitis as a complication of middle ear infection is discussed on page 45. Sensorineural hearing loss following meningitis is a well-known clinical entity. Bacteria can invade the labyrinth along nerves, vessels, cochlear aqueduct or the endolymphatic sac. Membranous labyrinth is totally destroyed.

3. SYPHILITIC. Sensorineural hearing loss is caused both by congenital and acquired syphilis. Congenital syphilis is of two types: the *early form*, manifesting at the age

of 2 or the *late form*, manifesting at the age of 8–20 years. Syphilitic involvement of the inner ear can cause:

- Sudden sensorineural hearing loss, which may be unilateral or bilateral. The latter is usually symmetrical in high frequencies or is a flat type.
- Ménière's syndrome with episodic vertigo, fluctuating hearing loss, tinnitus and aural fullness—a picture simulating Ménière's disease.
- Hennebert's sign. A positive fistula sign in the absence of a fistula. This is due to fibrous adhesions between the stapes footplate and the membranous labyrinth.
- Tullio phenomenon in which loud sounds produce vertigo.

Diagnosis of otosyphilis can be made by other clinical evidence of late acquired or congenital syphilis (interstitial keratitis, Hutchinson's teeth, saddle nose, nasal septal perforation and frontal bossing) and the laboratory tests. Fluorescent treponema-absorption test (FTA-ABS) and venereal disease research laboratory (VDRL) or rapid plasma reagin (RPR) tests from CSF are useful to establish the diagnosis.

Treatment of otosyphilis includes i.v. penicillin and steroids.

B. FAMILIAL PROGRESSIVE SENSORINEURAL HEARING LOSS

It is a genetic disorder in which there is progressive degeneration of the cochlea starting in late childhood or early adult life. Hearing loss is bilateral with flat or basin-shaped audiogram but an excellent speech discrimination.

C. OTOTOXICITY

Various drugs and chemicals can damage the inner ear and cause sensorineural hearing loss, tinnitus and sometimes vertigo ([Table 5.4](#)).

1. AMINOGLYCOSIDE ANTIBIOTICS. Streptomycin, gentamicin and tobramycin are primarily vestibulotoxic. They selectively destroy type I hair cells of the crista ampullaris but, administered in large doses, can also damage the cochlea.

Neomycin, kanamycin, amikacin, sisomicin and dihydrostreptomycin are cochleotoxic. They cause selective destruction of outer hair cells, starting at the basal coil and progressing onto the apex of cochlea.

Patients particularly at risk are those:

- having impaired renal function,
- elderly people above the age of 65,
- concomitantly receiving other ototoxic drugs,
- who have already received aminoglycoside antibiotics,
- who are receiving high doses of ototoxic drugs with high serum level of drug, and
- who have genetic susceptibility to aminoglycosides. Here the antibiotic binds to the ribosome and interferes with protein synthesis, thus causing death of the cochlear cells.

Symptoms of ototoxicity, hearing loss, tinnitus and/or giddiness may manifest during treatment or after completion of the treatment (delayed toxicity).

TABLE 5.4 OTOTOXIC DRUGS

<p>A. Aminoglycoside antibiotics</p> <ul style="list-style-type: none"> • Streptomycin • Dihydrostreptomycin • Gentamicin • Tobramycin • Neomycin • Kanamycin • Amikacin • Netilmycin • Sisomycin <p>B. Diuretics</p> <ul style="list-style-type: none"> • Furosemide • Ethacrynic acid • Bumetanide <p>C. Antimalarials</p> <ul style="list-style-type: none"> • Quinine • Chloroquine • Hydroxychloroquine <p>D. Cytotoxic drugs</p> <ul style="list-style-type: none"> • Nitrogen mustard (Methotrexate) • Cisplatin • Carboplatin 	<p>E. Analgesics</p> <ul style="list-style-type: none"> • Salicylates • Indomethacin • Phenylbutazone • Ibuprofen <p>F. Chemicals</p> <ul style="list-style-type: none"> • Alcohol • Tobacco • Marijuana • Carbon monoxide poisoning <p>G. Miscellaneous</p> <ul style="list-style-type: none"> • Erythromycin • Ampicillin • Propranolol • Propylthiouracil • Deferoxamine
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2. DIURETICS. Furosemide, bumetanide and ethacrynic acid are called *loop diuretics* as they block transport of sodium and chloride ions in the ascending loop of Henle. They are known to cause oedema and cystic changes in the stria vascularis of the cochlear duct. In most cases, the effect is *reversible* but permanent damage may occur. Hearing loss may be bilateral and symmetrical or sometime sudden in onset.

3. SALICYLATES. Symptoms of salicylate ototoxicity are tinnitus and bilateral sensorineural hearing loss particularly affecting higher frequencies. Site of lesion testing indicates cochlear involvement, but light and electron microscopy have failed to show any morphologic changes in the hair cells. Possibly they interfere at enzymatic level. Hearing loss due to salicylates is *reversible* after the drug is discontinued. SNHL has also been noted with other NSAIDs, e.g. naproxen, piroxicam and ketorolac but is reversible.

4. QUININE. Ototoxic symptoms due to quinine are tinnitus and sensorineural hearing loss, both of which are *reversible*. Higher doses may cause permanent loss. The symptoms generally appear with prolonged medication but may occur with smaller doses in those who are susceptible. Congenital deafness and hypoplasia of cochlea have been reported in children whose mothers received this drug during the first trimester of pregnancy. Ototoxic effects of quinine are due to vasoconstriction in the small vessels of the cochlea and stria vascularis.

5. CHLOROQUINE AND HYDROXYCHLOROQUINE. Effect is similar to that of quinine and cause reversible SNHL. Sometimes permanent deafness can result.

6. CYTOTOXIC DRUGS. Nitrogen mustard, cisplatin and carboplatin can cause cochlear damage. They affect the outer hair cells of the cochlea.

7. DEFEROXAMINE (DEFERRIOXAMINE). It is an iron-chelating substance used in the treatment of thalassaemic patients who receive repeated blood transfusions and in turn have high iron load. Like cisplatin and aminoglycosides, deferoxamine also causes high-frequency sensorineural hearing loss. Onset of hearing loss is sudden or delayed. It is permanent but in some cases it can be reversible when the drug is discontinued. It causes toxicity to nerves; children are affected more.

8. MISCELLANEOUS. Isolated cases of deafness have been reported with erythromycin, ampicillin and chloramphenicol, indomethacin, phenylbutazone, ibuprofen, tetanus antitoxin, propranolol and propylthiouracil.

Alcohol, tobacco and marijuana also cause damage to the inner ear.

9. TOPICAL EAR DROPS. Topical use of drugs in the middle ear can also cause damage to the cochlea by absorption through oval and round windows. Deafness has occurred with the use of chlorhexidine which was used in the preparation of ear canal before surgery or use of ear drops containing aminoglycoside antibiotics, e.g. neomycin, framycetin and gentamicin. Ototoxic potential is also present in ear drops containing polymyxin B, propylene glycol and antifungal agents. Use only approved ototopical drops for middle ear infection.

D. NOISE TRAUMA

Hearing loss associated with exposure to noise has been well-known in boiler makers, iron- and coppersmiths, and artillery men. Lately, noise trauma has assumed greater significance because of its being an occupational hazard; the compensations asked for and the responsibilities thrust upon the employer and the employee to conserve hearing. Hearing loss caused by excessive noise can be divided into two groups:

1. ACOUSTIC TRAUMA. Permanent damage to hearing can be caused by a *single brief exposure* to very intense sound without this being preceded by a temporary threshold shift. Also called impulse noise, such noise can arise from an explosion, gun fire or a powerful cracker and may reach or cross 140 dB. Noise level of a gun or rifle may reach 140–170 dB SPL (sound pressure level). Such brief and loud noises mechanically damage organ of Corti, tear Reissner's membrane, rupture hair cells and allowing mixing of perilymph and endolymph. A severe blast, in addition, may concomitantly damage the tympanic membrane and disrupt ossicles further adding conductive loss. Impulse noise may be as brief as 0.2 ms. No impulse noise more than 140 dB (A) is permitted.

2. NOISE-INDUCED HEARING LOSS (NIHL). Hearing loss, in this case, follows *chronic exposure* to less intense sounds than seen in acoustic trauma and is mainly a hazard of noisy occupations.

(a) *Temporary threshold shift (TTS).* The hearing is impaired immediately after exposure to noise but recovers after an interval of a few minutes to a few hours even up to 2 weeks. Amount of TTS depends on the noise—its intensity, frequency and duration.

TABLE 5.5 PERMISSIBLE EXPOSURE IN CASES OF CONTINUOUS NOISE OR A NUMBER OF SHORT-TERM EXPOSURES [GOVERNMENT OF INDIA, MINISTRY OF LABOUR, MODEL RULES UNDER FACTORIES ACT 1948 (CORRECTED UP TO 31.3.87)]

Noise level ^a (dBA)	Permitted daily exposure (h)
90	8.0
92	6.0
95	4.0
97	3.0
100	2.0
102	11/2
105	1.0
110	1/2
115	1/4

^a5 dB rule of time intensity states that “any rise of 5 dB noise level will reduce the permitted noise exposure time to half.”

(b) *Permanent threshold shift (PTS)*. The hearing impairment is permanent and does not recover at all.

The damage caused by noise trauma depends on several factors:

- (i) *Frequency of noise*. A frequency of 2000–3000 Hz causes more damage than lower or higher frequencies.
- (ii) *Intensity and duration of noise*. As the intensity increases, permissible time for exposure is reduced. Table 5.5 gives the permissible limits of time for various intensity levels for the safety of ear.
- (iii) *Continuous vs interrupted noise*. Continuous noise is more harmful.
- (iv) *Susceptibility of the individual*. Degree of TTS and PTS varies in different individuals.
- (v) *Pre-existing ear disease*.

A noise of 90 dB (A) SPL, 8 h a day for 5 days per week is the maximum safe limit as recommended by Ministry of Labour, Govt. of India, Model Rules under Factories Act (Table 5.5). No exposure in excess of 115 dB (A) is to be permitted. No impulse noise of intensity greater than 140 dB (A) is permitted.

The Noise Pollution (Regulation and Control) Rules 2000, Ministry of Environment and Forest, Govt. of India has defined permissible limits of noise for various zones or areas (Table 5.6). According to which silence zone is 100 m around the premises of hospitals, nursing homes, educational institutions and courts. Also manufacture, sale and use of fire crackers generating sound level above 125 dB (AI) or 145 dB (C) pk from 4 m distance from the point of bursting are not permitted (Environment Protection Rules 2006) [dB (AI) = A-weighted impulse sound pressure level in decibels; dB (C) pk = C-weighted peak sound pressure in decibels].

The audiogram in NIHL shows a typical notch, at 4 kHz, both for air and bone conduction (Figure 5.7). It is usually symmetrical on both sides. At this stage, patient complains of high-pitched tinnitus and difficulty in hearing in noisy surroundings but no difficulty in day-to-day hearing. As the duration of noise exposure increases, the notch deepens and also widens to involve lower and higher frequencies. Hearing impairment becomes clinically apparent to the patient when the frequencies of

TABLE 5.6 PERMISSIBLE LIMITS OF NOISE AS PER THE NOISE POLLUTION (REGULATION AND CONTROL) RULES 2000, MINISTRY OF ENVIRONMENT AND FOREST, GOVT. OF INDIA

Zone/Area	Day (6 am to 10 pm) Limits in dB(A) Leq ^a	Night (10 pm to 6 am) Limits in dB(A) Leq
Industrial	75	70
Commercial	65	55
Residential	55	45
Silence	50	40

^aLeq = Energy mean of noise level over a specified period.

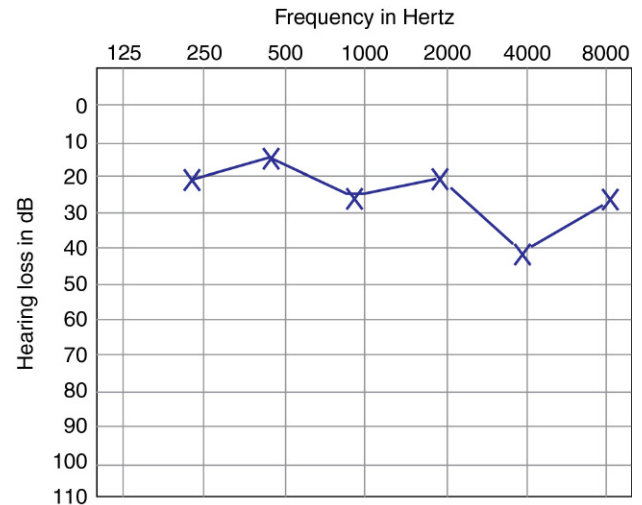


Figure 5.7. Early case of noise-induced hearing loss. Note dip at 4000 Hz.

TABLE 5.7 HEARING ATTENUATION PROVIDED BY DIFFERENT DEVICES

Cotton wool	5 dB
Ear plug	15–30 dB (mostly in range of 3–5 kHz)
Ear muffs	30–40 dB (500–1 kHz)
Ear plugs + muffs	More than 40 dB

Note: Hearing protectors provide more attenuation in higher frequencies, 25–40 dB for 1000–8000 Hz while only 10–30 dB attenuation for lower frequencies less than 500 Hz.

500, 1000 and 2000 Hz (the speech frequencies) are also affected.

NIHL causes damage to hair cells, starting in the basal turn of cochlea. Outer hair cells are affected before the inner hair cells.

Noise-induced hearing loss is preventable. Persons who have to work at places where noise is above 85 dB (A) should have pre-employment and then annual audiograms for early detection. Ear protectors (ear plugs or ear muffs) should be used where noise levels exceed 85 dB (A). They provide protection up to 35 dB (see Table 5.7). If hearing impairment has already occurred, rehabilitation is similar to that employed for other sensorineural hearing losses.

3. NONAUDITORY EFFECTS OF NOISE. Apart from hearing loss, noise can affect other systems of the body. It interferes with rest and sleep causing *chronic fatigue* and

stress. Through activation of the autonomic nervous system and pituitary–adrenal axis, it causes annoyance and irritability. Hypertension and peptic ulcer have also been attributed to it. It also adversely affects task performance where communication through speech is required. Laryngeal problems have been noticed in workers who have to speak loudly in persistently noisy surroundings.

E. AUTOIMMUNE (IMMUNE-MEDIATED) INNER EAR DISEASE

Immune-mediated inner ear disease (Syn. autoimmune SNHL) causes progressive bilateral sensorineural hearing loss. It occurs between 40 and 50 years with equal incidence in both sexes. Nearly 50% of patients also experience vestibular symptoms like disequilibrium, motion intolerance, positional or episodic vertigo. About 15% of patients have evidence of other autoimmune disorder such as ulcerative colitis, systemic lupus, rheumatoid arthritis or multiple sclerosis. Moscicki et al. defined the condition as:

‘Bilateral SNHL \geq 30 dB at any frequency and evidence of progression in at least one ear on two serial audiograms that are done at equal to or less than 3 months apart. Progression is defined as threshold shift of \geq 15 dB at one frequency or 10 dB at two or more consecutive frequencies or significant change in speech discrimination’.

Investigations

1. **Audiogram.** To establish above criteria, repeated audiograms can be taken at one month intervals. Audiogram may show loss at high and low frequencies.
2. **Speech audiogram.** Speech discrimination is affected though threshold of pure tones remains the same.
3. **Evoked response audiometry.** To exclude acoustic neuroma or multiple sclerosis.
4. **Contrast-enhanced MRI.**
5. **Blood tests** to exclude systemic autoimmune disorders. Total and differential counts, ESR, rheumatoid factor, antinuclear antibodies, C3 and C4 complement levels, Raji cell assay for circulating immune complexes.
6. **Western blot essay for anti-Hsp 70 (anti-heat shock protein 70) antibodies.** Antigen used in this test is crude protein extract from bovine renal cells. It is not a specific test for diagnosis but correlates to both active disease and steroid responsiveness.

Treatment

Prednisolone 1 mg/kg/day up to a total of 60 mg/day (for adults) for 4 weeks. Sometimes response is late. If no response is seen in 4 weeks, steroid is tapered off in 12 days. Responders continue till a plateau is reached and then continue on maintenance dose of 10–20 mg every other day for about 6 months. Side effects and risks of long-term steroid therapy should be kept in mind.

Those who cannot take steroids can be given methotrexate 15 mg/week for 6–8 weeks and if the patient responds, continue it for 6 months. If no response is obtained for 6–8 weeks trial, drug is discontinued.

Alternative to methotrexate is cyclophosphamide but it is more toxic.

Other treatments include intratympanic steroid injection, systemic IgG injection and plasmapheresis.

F. SUDDEN HEARING LOSS

Sudden SNHL is defined as 30 dB or more of SNHL over at least three contiguous frequencies occurring within a period of 3 days or less. Mostly it is unilateral. It may be accompanied by tinnitus or temporary spell of vertigo.

Aetiology

Most often the cause of sudden deafness remains obscure, in which case it is called the idiopathic variety. In such cases, three aetiological factors are considered—**viral, vascular** or the **rupture of cochlear membranes**. Spontaneous perilymph fistulae may form in the oval or round window. Other aetiological factors which cause sudden deafness and must be excluded are listed below. Remember the mnemonic “In The Very Ear Too No Major Pathology.”

1. **Infections.** Mumps, herpes zoster, meningitis, encephalitis, syphilis, otitis media.
2. **Trauma.** Head injury, ear operations, noise trauma, barotrauma, spontaneous rupture of cochlear membranes.
3. **Vascular.** Haemorrhage (leukaemia), embolism or thrombosis of labyrinthine or cochlear artery or their vasospasm. They may be associated with diabetes, hypertension, polycythaemia, macroglobinaemia or sickle cell trait.
4. **Ear (otologic).** Ménière’s disease, Cogan’s syndrome, large vestibular aqueduct.
5. **Toxic.** Ototoxic drugs, insecticides.
6. **Neoplastic.** Acoustic neuroma. Metastases in cerebellopontine angle, carcinomatous neuropathy.
7. **Miscellaneous.** Multiple sclerosis, hypothyroidism, sarcoidosis.
8. **Psychogenic.**

Management

As far as possible, the aetiology of sudden hearing loss should be discovered by detailed history, physical examination and laboratory investigations. The investigations may include audiometry, vestibular tests, imaging studies of temporal bones, sedimentation rate, tests for syphilis, diabetes, hypothyroidism, blood disorders and lipid profiles. Some cases may require exploratory tympanotomy where perilymph fistula is strongly suspected. Where the cause still remains obscure, treatment is empirical and consists of:

1. **Bed rest.**
2. **Steroid therapy.** Prednisolone 40–60 mg in a single morning dose for 1 week and then tapered off in a period of 3 weeks. Steroids are anti-inflammatory and relieve oedema. They have been found useful in idiopathic sudden hearing loss of moderate degree.
3. **Inhalation of carbogen** (5% CO₂ + 95% O₂). It increases cochlear blood flow and improves oxygenation.
4. **Vasodilator drugs.**
5. **Low molecular weight dextran.** It decreases blood viscosity. It is contraindicated in cardiac failure and bleeding disorders.
6. **Hyperbaric oxygen therapy.** Available only in selected centres, hyperbaric oxygen raises concentration of oxygen in labyrinthine fluids and improves cochlear function (*see p. 405*).

TABLE 5.8 PROGNOSTIC FACTORS IN SUDDEN SNHL

Good prognosis	Bad prognosis
Mild loss	Severe loss
Low and medium frequency loss	High frequency loss
Recovery starting in 2 weeks	Recovery does not start in 2 weeks
No history of vertigo	History of vertigo
Younger patients	Older patients
Early treatment	Late treatment

7. **Low-salt diet and a diuretic.** It is empirical and has same benefit as in cases of Ménière's disease.
8. **Intratympanic steroids therapy.** It raises the local concentration of steroids in cochlear fluids, thus avoiding side effects of systemic therapy.

Treatment

Many treatment protocols have been suggested for idiopathic sensorineural sudden hearing loss but none has shown significant benefit over the benefit of spontaneous recovery which occurs in 50–60% cases within first 2 weeks. None of the drugs, dextran 40, vasodilators, carbogen inhalation (5% CO₂ with 95% O₂), diatrizoate meglumine, have shown significant benefit.

Generally prescribed medicines include:

1. Steroids.
2. Inhalation of carbogen.
3. Low-salt diet and a diuretic.
4. Hyperbaric oxygen.

Prognosis

Fortunately, about half the patients of idiopathic sensorineural hearing loss recover spontaneously within 15 days. Chances of recovery are poor after 1 month. Severe hearing loss and that associated with vertigo have poor prognosis. Younger patients below 40 and those with moderate losses have better prognosis (see Table 5.8).

G. PRESBYCUSIS

Sensorineural hearing loss associated with physiological aging process in the ear is called presbycusis. It usually manifests at the age of 65 years but may do so early if there is hereditary predisposition, chronic noise exposure or generalized vascular disease.

Four pathological types of presbycusis have been identified.

1. **SENSORY.** This is characterized by degeneration of the organ of Corti, starting at the basal coil and progressing gradually to the apex. Higher frequencies are affected but speech discrimination remains good.
2. **NEURAL.** This is characterized by degeneration of the cells of spiral ganglion, starting at the basal coil and progressing to the apex. Neurons of higher auditory pathways may also be affected. This manifests with high tone loss but speech discrimination is poor and out of proportion to the pure tone loss.

3. **STRIAL OR METABOLIC.** This is characterized by atrophy of stria vascularis in all turns of cochlea. In this, the physical and chemical processes of energy production are affected. It runs in families. Audiogram is flat but speech discrimination is good.

4. **COCHLEAR CONDUCTIVE.** This is due to stiffening of the basilar membrane thus affecting its movements. Audiogram is sloping type.

Patients of presbycusis have great difficulty in hearing in the presence of background noise though they may hear well in quiet surroundings. They may complain of speech being heard but not understood. Recruitment phenomenon is positive and all the sounds suddenly become intolerable when volume is raised. Tinnitus is another bothersome problem and in some it is the only complaint.

Patients of presbycusis can be helped by a hearing aid. They should also have lessons in speech reading through visual cues. Curtailment of smoking and stimulants like tea and coffee may help to decrease tinnitus.

NONORGANIC HEARING LOSS (NOHL)

In this type of hearing loss, there is no organic lesion. It is either due to malingering or is psychogenic. In the former, usually there is a motive to claim some compensation for being exposed to industrial noises, head injury or ototoxic medication. Patient may present with any of the three clinical situations:

(i) Total hearing loss in both ears, (ii) total loss in only one ear or (iii) exaggerated loss in one or both ears. The responsibility of the physician is to find out: Is the patient malingering? If so, what is his actual threshold of hearing? This is accomplished by:

1. **HIGH INDEX OF SUSPICION.** Suspicion further rises when the patient makes exaggerated efforts to hear, frequently making requests to repeat the question or placing a cupped hand to the ear.

2. **INCONSISTENT RESULTS ON REPEAT PURE TONE AND SPEECH AUDIOMETRY TESTS.** Normally, the results of repeat tests are within ± 5 dB. A variation greater than 15 dB is diagnostic of NOHL.

3. **ABSENCE OF SHADOW CURVE.** Normally, a shadow curve can be obtained while testing bone conduction, if the healthy ear is not masked. This is due to transcranial transmission of sound to the healthy ear. Absence of this curve in a patient complaining of unilateral deafness is diagnostic of NOHL.

4. **INCONSISTENCY IN PTA AND SRT.** Normally, pure tone average (PTA) of three speech frequencies (500, 1000 and 2000 Hz) is within 10 dB of speech reception threshold (SRT). An SRT better than PTA by more than 10 dB points to NOHL.

5. **STENGER TEST.** It can be done with a pair of identical tuning forks or a double-channel audiometer. Principle involved is that, if a tone of two intensities, one greater than the other, is delivered to two ears simultaneously, only the ear which receives tone of greater intensity will hear it. To

do this test, take two tuning forks of equal frequency, strike and keep them say 25 cm from each ear. Patient will claim to hear it in the normal ear. Now bring the tuning fork on the side of feigned deafness to within 8 cm, keeping the tuning fork on the normal side at the same distance. The patient will deny hearing anything even though tuning fork on normal side is where it could be heard earlier. A person with true deafness should continue to hear on the normal side. Patient should be blindfolded during this test.

This same test can be performed with a two-channel audiometer using pure tone or speech signals.

6. ACOUSTIC REFLEX THRESHOLD. Normally, stapedial reflex is elicited at 70–100 dB SL. If patient claims total deafness but the reflex can be elicited, it indicates NOHL.

7. ELECTRIC RESPONSE AUDIOMETRY (ERA). It is very useful in NOHL and can establish hearing acuity of the person to within 5–10 dB of actual thresholds.

SOCIAL AND LEGAL ASPECTS OF HEARING LOSS

HEARING LOSS AND DEAFNESS

Hearing loss is impairment of hearing and its severity may vary from mild to severe or profound, while the term deafness is used, when there is little or no hearing at all. In some countries, this rigid differentiation is not made. They use the term deafness to denote any degree of hearing loss irrespective of its severity. In 1980, WHO recommended that the term “deaf” should be applied only to those individuals whose hearing impairment is so severe that they are unable to benefit from any type of amplification. A similar definition is used in India while extending benefits to the hearing handicapped.

DEFINITION OF DEAF

(Ministry of Social Welfare, Government of India—Scheme of Assistance to Hearing Handicap).

“The deaf are those in whom the sense of hearing is nonfunctional for ordinary purposes of life.” They do not hear/understand sounds at all even with amplified speech. The cases included in the category will be those having hearing loss more than 90 dB in the better ear (profound impairment) or total loss of hearing in both ears.

The partially hearing are defined as those falling under any one of the following categories:

Category	Hearing acuity
Mild impairment	More than 30 but not more than 45 dB in better ear
Serious impairment	More than 45 but not more than 60 dB in better ear
Severe impairment	More than 60 but not more than 90 dB in better ear

DEGREE OF HEARING LOSS (WHO CLASSIFICATION)

WHO (1980) recommended the following classification on the basis of pure tone audiogram taking the average

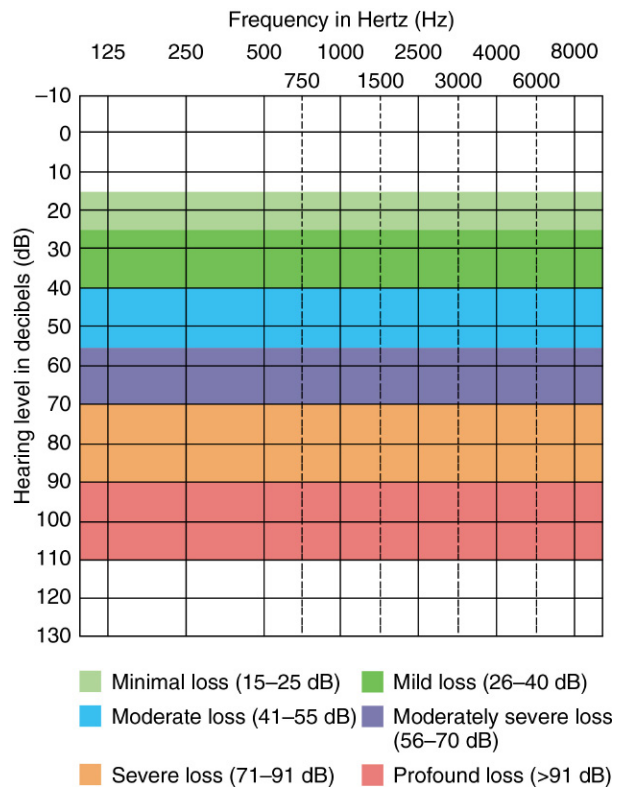


Figure 5.8. Classification of hearing loss. Ninety-five per cent of population has thresholds between 210 and 110 dB HL.

of the thresholds of hearing for frequencies of 500, 1000 and 2000 Hz with reference to ISO: R. 389–1970 (International Calibration of Audiometers).

Degree of hearing loss (Figure 5.8)

1.	Mild	26–40 dB
2.	Moderate	41–55 dB
3.	Moderately severe	56–70 dB
4.	Severe	71–91 dB
5.	Profound	More than 91 dB
6.	Total	

From this it is implied that there is no apparent impairment of hearing from 0 to 25 dB.

The disability to understand speech with different degrees of hearing loss is given in Table 5.9.

IMPAIRMENT, DISABILITY AND HANDICAP

When a disease process strikes an organ or a system it causes an impairment either in structure or function, but this impairment may or may not become clinically manifested. When impairment affects the ability to perform certain functions in the range considered normal for that individual it is called disability. The disability further restricts the duties and roles expected from an individual by society and is called a handicap.

To exemplify, injury (disease) to the ear may result in hearing impairment which, depending on its severity, will affect the individual’s ability to hear and perform certain activities (disability) and will be termed handicap by the society:

Disease → Impairment → Disability → Handicap.

TABLE 5.9 HEARING LOSS AND DIFFICULTY IN HEARING SPEECH

Hearing threshold in better ear (average of 500, 1000, 2000 Hz)	Degree of impairment (WHO classification)	Ability to understand speech
0–25	Not significant	No significant difficulty with faint speech
26–40	Mild	Difficulty with faint speech
41–55	Moderate	Frequent difficulty with normal speech
56–70	Moderately severe	Frequent difficulty even with loud speech
71–90	Severe	Can understand only shouted or amplified speech
91 above	Profound	Usually cannot understand even amplified speech

DEGREE OF HANDICAP

Sometimes it is desired to express the impairment and handicap in terms of percentage for the purposes of compensation. Different countries and professional bodies have adopted their own system to calculate this percentage.

One of the methods to find hearing handicap is given below:

- (i) Take an audiogram and calculate the average of thresholds of hearing for frequencies of 500, 1000 and 2000 Hz say = A.
- (ii) Deduct from it 25 dB (as there is no impairment up to 25 dB), i.e. $A - 25$.
- (iii) Multiply it by 1.5, i.e. $(A - 25) \times 1.5$.

This is the percentage of hearing impairment for that ear. Similarly calculate the percentage of hearing impairment for the other ear.

Total percentage handicap of an individual

$$= \frac{(\text{better ear}\% \times 5) + \text{worse ear}\%}{6}$$

Example:

	500 Hz	1000 Hz	2000 Hz	Average
Right ear	60	75	90	75 dB
Left ear	30	45	60	45 dB
Impairment Right ear:	$75 - 25 = 50$			$50 \times 1.5 = 75\%$
Impairment Left ear:	$45 - 25 = 20$			$20 \times 1.5 = 30\%$

$$\begin{aligned} \text{Total handicap} &= \frac{(30 \times 5) + 75}{6} = \frac{225}{6} \\ &= 37.5\% \\ &= 38\% \text{ (rounded off)} \end{aligned}$$

In the above calculation only three speech frequencies (500, 1000 and 2000 Hz) are taken into account but it is felt that frequency of 3000 Hz is important for hearing in the presence of noise and should also be taken into account. American Academy of Ophthalmology and Otolaryngology recommends and takes into account the average of four frequencies 500, 1000, 2000 and 3000 Hz when calculating the handicap.

Government of India reserved certain percentage of vacancies in Group C and D in favour of the physically handicapped and has extended certain other benefits. It has also recommended the classification based on percentage of impairment and the test required to be performed (see Table 5.10). (Brochure on Reservations and Concessions for Physically Handicapped in Central Govt. Services published by Ministry of Personnel, Public Grievances and Pensions, Dept. of Personnel and Training.)

UNILATERAL HEARING LOSS

Unilateral loss of hearing, even though total, does not produce a serious handicap or affect speech but it impairs localization of the sound source, difficulty in discrimination of speech in the presence of background noise and some difficulty at a meeting or in classroom when the speaker is on the side of affected ear. It should also alert the individual that he does not have a "spare or reserve ear" and has to take all precautions for the safety of the only hearing ear; also the surgeon should be careful when he is called upon to operate on this only hearing ear. Bone-anchored hearing aids are the treatment of choice for management of single-sided deafness (see p. 137).