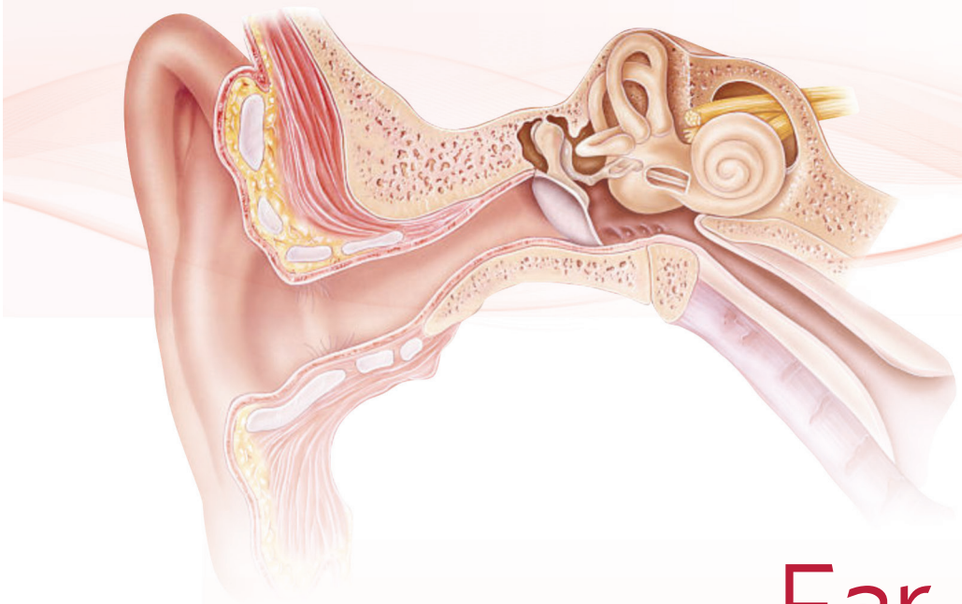


Section

1

Ear

1. Anatomy of Ear
2. Physiology of Hearing
3. Audiology
4. Vestibular System
5. Vertigo: Evaluation and Common Disorders
6. Diseases of the External Ear
7. Eustachian Tube and Disorders
8. Ototoxicity
9. Otitis Media
10. Otitis Media with Effusion
11. Chronic Otitis Media
12. Complications of Otitis Media
13. Presbycusis and Sudden Sensorineural Hearing Loss
14. Otosclerosis
15. Facial Nerve
16. Ménière's Disease
17. Hearing Impairment in Children
18. Cochlear Implant
19. Otalgia
20. Tinnitus
21. Paragangliomas of Temporal Bone
22. Tuberculosis of the Temporal Bone
23. Vestibular Schwannoma
24. Noise Induced Hearing Loss (NIHL) and Noise Pollution
25. Hearing Aids
26. National Programme for Control and Prevention of Deafness (NPPCD)

Chapter

1



Anatomy of Ear

EN1.1: Describe the Anatomy of ear.
AN40.2: Describe and demonstrate the boundaries, contents, relations and functional anatomy of middle ear and auditory tube.
AN40.3: Describe the features of internal ear.

DEVELOPMENT OF EAR

The development of the pinna or the auricle starts on the 38th day of intrauterine life. The pinna develops from the first and second branchial arches. These arches give rise to 6 nodular mesenchymal condensations called **auricular hillocks of His** (Fig. 1.1), which later fuse together to form the auricle. The anatomical development of the auricle is complete by the 20th week of intrauterine life. The tragus and part of the anterior crus of the helix is contributed by the first arch and the rest is contributed by the second arch.

The auricle is initially placed at the level of the lower neck and with development of the mandible it ascends to the side of head at the level of the eyes. The external auditory canal develops from the dorsal portion of the first pharyngeal cleft (Fig. 1.1).

On the 22nd day of intrauterine life, ectodermal thickening appears on the sides of the rhombencephalon, which are called the ***otic placodes*** (Fig. 1.2). These rapidly invaginate to form the **otic vesicle or otocysts**. The ventral part of the otocyst gives rise to the saccule and cochlear duct and the dorsal part of the otocyst forms the utricle, semicircular canal and endolymphatic duct (Fig. 1.3).

The tympanic cavity is derived from the first pharyngeal pouch. On lateral expansion, the pouch comes in contact with the floor of the first pharyngeal cleft (Fig. 1.3). The proximal part of the pouch gives rise to the Eustachian tube and the distal part to the tubotympanic recess. The malleus and incus (from *Meckel's cartilage*) are derived from the first pharyngeal arch. The stapes (from Reichert's cartilage) is derived from the second pharyngeal arch. The stapes footplate is derived from the otic capsule. The ossicles ossify by the 4th month of intrauterine life.* The mastoid antrum** and mastoid air cell system is derived as an extension from the tympanic cavity.

ANATOMY OF EAR

For the purpose of description, the ear is anatomically divided into (Fig. 1.4):
1. External ear
2. Middle ear cleft
3. Inner ear.

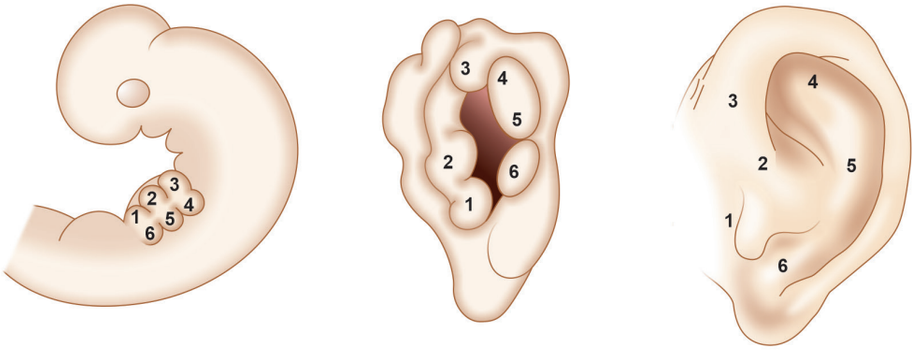
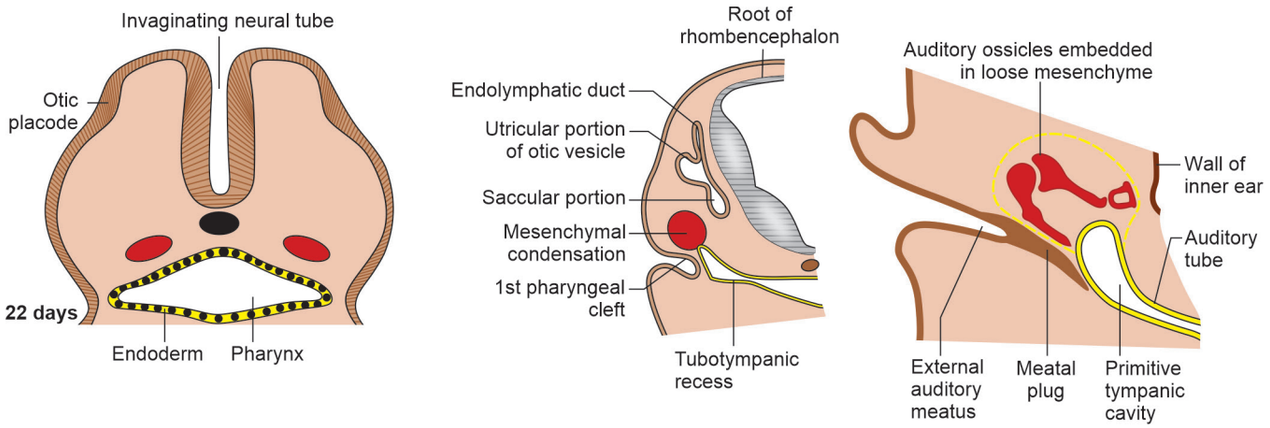


Fig. 1.1: Development of external ear

*Ossicles are the first bones to ossify in human body
**The antrum reaches adult size by birth.



Figs 1.2 and 1.3: Development of middle and inner ear

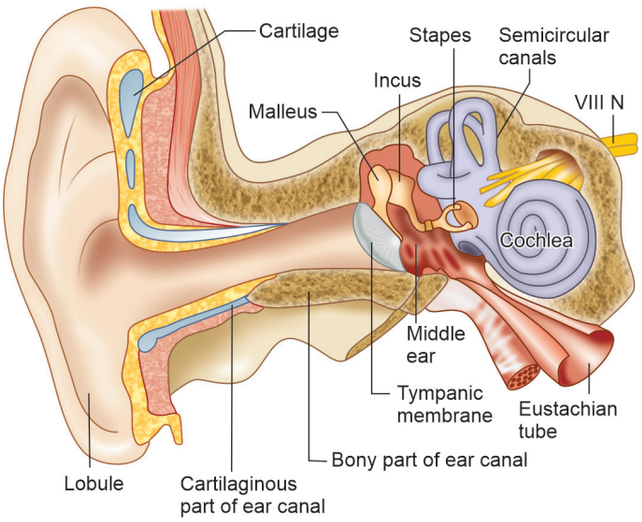


Fig. 1.4: Parts of ear

EXTERNAL EAR

External ear consists of:

1. Pinna or auricle
2. External auditory canal (EAC)
3. Outer layer of tympanic membrane.

AURICLE (PINNA)

Structure: The pinna is the outward projecting part of the external ear, which functions to collect, concentrate the sounds and direct them to the external auditory canal. It is made up of a single piece yellow elastic cartilage framework which is covered with skin. It has several prominences and depressions which are specific for an individual.

The outermost curved rim of pinna is called the **helix**, which might have a prominence called the *Darwin's tubercle*. Parallel to the helix, there is another elevation which is called an **antihelix**. Antihelix superiorly divides into two crura and form the **triangular fossa**. The **scaphoid fossa** lies superior to these crura.

The antihelix surrounds the depression called the **concha**. The concha is divided by the descending limb of the superior portion of the helix into two parts called the **cymba concha** and the **cavum concha**.

Note: The cymba concha anatomically overlies the suprameatal triangle of the temporal bone.

A triangular prominence called the **tragus** lies below the crus of the helix. **Antitragus** lies opposite to the tragus on the antihelix. Below the antitragus lies the **lobule**, which is composed of fibrous and fatty tissue (Fig. 1.5).



Fig. 1.5: Parts of pinna. 1. helix, 2. triangular fossa, 3. antihelix, 4. tragus, 5. cymba concha, 6. cavum concha, 7. antitragus, 8. lobule, 9. supratragal notch, 10. scaphoid fossa

Ligaments: Two extrinsic ligaments connect the auricle to the temporal bone. The anterior extrinsic ligament, runs from the tragus and crus of the helix to the root of the zygomatic arch. The posterior extrinsic ligament runs from the medial surface of the pinna to the mastoid bone. The intrinsic ligaments connect various parts of cartilaginous pinna.

Muscles: Various intrinsic and extrinsic muscles are attached to the perichondrium of the pinna cartilage. There are three extrinsic muscles namely auricularis anterior, superior and posterior. Noise evokes some postauricular myogenic response in these muscles. They are supplied by the posterior auricular branch of the facial nerve. There are six intrinsic muscles which are smaller in size and without much function.

Nerve Supply of Pinna (Fig. 1.6)

Nerve	*Area of pinna supplied*
Greater auricular (C2, C3)	Medial surface and posterior portion of lateral surface
Lesser occipital (C2, C3)	Superior part of medial surface
Auricular branch of X nerve	Concha and antihelix
Auriculotemporal branch of V3	Tragus, crus of helix
Facial nerve	Root of concha

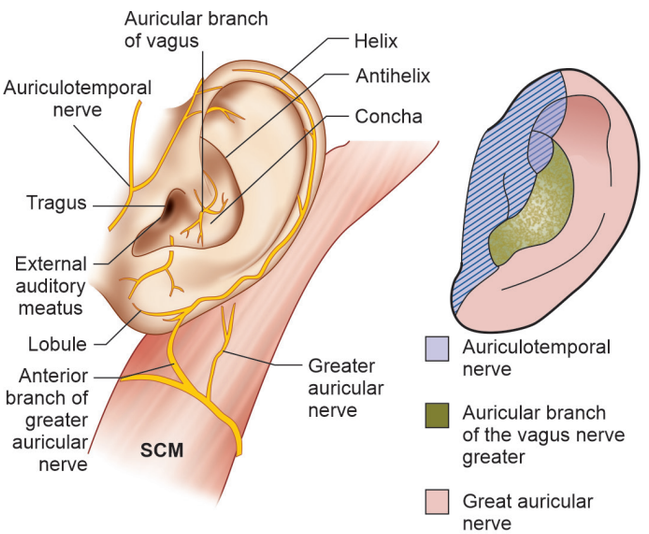


Fig. 1.6: Nerve supply of pinna (*SCM: Sternocleidomastoid muscle*)

Blood supply: The posterior auricular artery is the major artery supplying the medial surface of the pinna. The anterior auricular branch of the superficial temporal artery supplies the lateral surface of pinna, which has the helix, antihelix, triangular fossa, tragus and lobule. The superficial temporal artery is assisted by a small auricular branch from the occipital artery (Fig. 1.7).

Lymphatic drainage: The upper part of the anterior surface of pinna drains into the preauricular nodes and the rest of it drains to the upper deep cervical nodes. The posterior surface and lower part of pinna drains into lymph nodes at the mastoid tip.

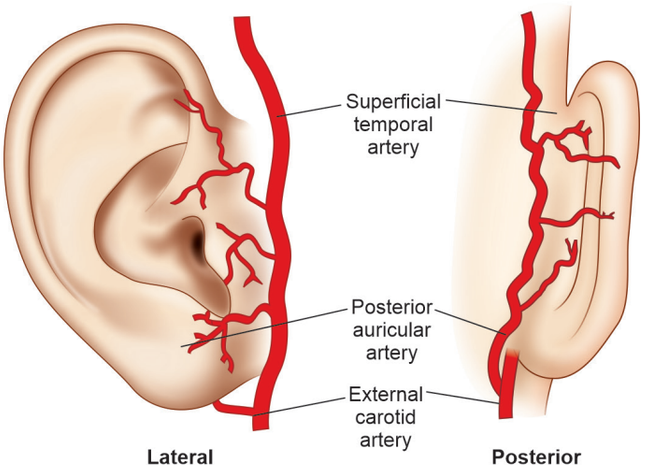


Fig. 1.7: Blood supply of pinna

Surgical considerations

1. The **incisura terminalis** (between the crus of helix and tragus) is an area where the pinna lacks cartilage. The vertical limb of Lempert's incision is made in this area during an endaural approach (Fig. 1.8).
2. The **'conchal stitch'** is placed through the concha while performing the endoscopic ear surgeries. It helps to straighten the external auditory canal, thereby leading to better visualization of tympanic membrane and middle ear.
3. The conchal cartilage is a good graft for attic reconstruction following an atticotomy or as a graft for revision tympanoplasty.
4. The tragal cartilage and perichondrium are ideal grafts for permeatal tympanoplasty.
5. Ear lobule is devoid of any cartilage and hence it is an ideal site for ear piercing. Remember the cosmetic ear piercing through the pinna cartilage is not recommended because it predisposes to keloid formation.

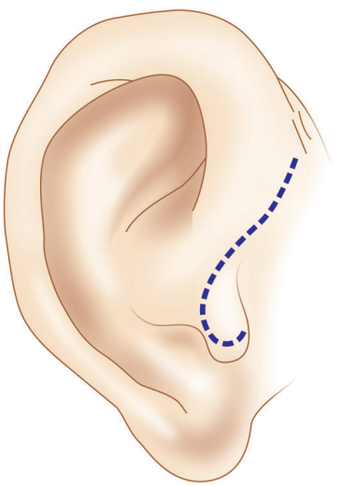


Fig. 1.8: Blue line shows the inter-cartilaginous limb of endaural incision (Lempert's incision)

EXTERNAL AUDITORY CANAL (EAC)

External auditory canal extends from the concha to the tympanic membrane. Its length is 24 mm, of which lateral 8 mm (1/3rd) is cartilaginous and medial 16 mm (2/3rd) is bony. The direction of the cartilaginous part in adults is downward, inward and forwards. Therefore, on pulling the pinna *upwards and backwards* the canal can be straightened for visualization of the tympanic membrane. In neonates the bony EAC is not developed, therefore it is more horizontal. Hence, gentle *downward and backward* movement of the pinna is required to visualise the tympanic membrane.

The bony cartilaginous junction of EAC has natural openings called **fissures of Santorini**, which form a communication for the infections and tumours to spread to and from the parotid gland.

The bony EAC has two suture lines, namely the **tympanosquamous suture** anteriorly and **tympanomastoid suture** posteriorly. These are difficult areas for tympanomeatal flap elevation during surgery because the periosteum is strongly adhered at the suture lines.

There are two constrictions in EAC: First constriction is at the bony-cartilaginous junction and the second constriction at a point 5 mm from the tympanic membrane, which is formed due to the prominence of the anterior canal wall, and is called the **isthmus**. This is the narrowest portion of the EAC. Medial to the isthmus is the **anterior recess** which is a difficult area to access clinically and is a common site for foreign body impaction.

The EAC is lined by keratinizing squamous epithelium. The skin of the cartilage shows ceruminous and sebaceous glands. The ceruminous glands are modified apocrine glands which produce watery white secretions in the roots of the hair follicles. The sebaceous glands produce oily secretion called *sebum*.

Wax is the combined secretions of ceruminous glands and sebaceous glands along with desquamated epithelial cells. It is either of dry or wet type, which is decided by Mendelian traits. The wet wax gene being the dominant gene phenotypically. The wet wax is yellowish brown in colour, sticky and wet and on the other hand the dry wax is yellowish or grey in colour, dry and flaky. Wax contains lysozymes, amino acids, fatty acids and immunoglobulins, which have some bactericidal effects.

The **arterial supply** to the EAC is via the auricular branch of the superficial temporal artery and posterior auricular artery. Deep auricular branches of the first part of the maxillary artery also supply the EAC.

The **nerve supply** of EAC is from the trigeminal, facial, glossopharyngeal and vagus nerves (Fig. 1.9). The floor of the EAC is supplied by auricular branch of the vagus nerve which is also called the **Arnold's nerve**. Stimulation of this nerve while cleaning the EAC can produce a cough reflex. Small part of the posterosuperior canal wall is supplied by the sensory branch of the facial nerve*. The anterior part of the EAC is supplied by the auriculotemporal branch of the mandibular division of the trigeminal nerve.

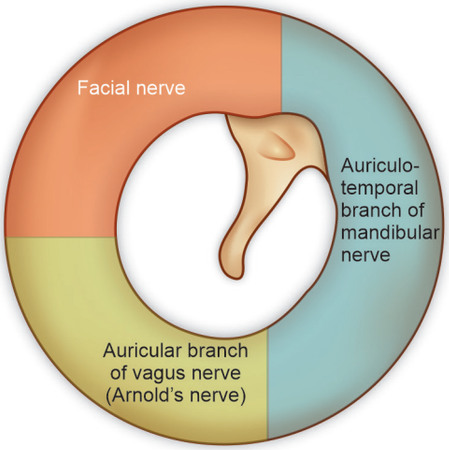


Fig. 1.9: Nerve supply of the external auditory canal

*__Note:__ In cases of vestibular schwannoma there occurs anaesthesia of the posterosuperior canal wall due to involvement of facial nerve at internal acoustic meatus and is called ***Hitselberger sign.***

MIDDLE EAR CLEFT

It consists of the tympanic cavity or middle ear space, Eustachian tube and the mastoid air cell system.

The middle ear is connected anteriorly to the Eustachian tube and posteriorly to the mastoid air cell system. It is laterally bounded by the tympanic membrane and medially by the osseous labyrinth.

EN4.4: Demonstrate the correct technique to hold visualize and assess the mobility of the tympanic membrane and interpret and diagrammatically represent the findings.

TYMPANIC MEMBRANE

It lies at the medial end of the external auditory canal. It is oval in shape and positioned at an angle of 55 degree with the floor of the external auditory canal. The handle of the malleus draws the tympanic membrane slightly inward, producing a flattened cone shape. Its longest diameter is from posterosuperior to anteroinferior and is 9–10 mm, while the diameter perpendicular to this and is 8–9 mm.

The peripheral part of the tympanic membrane is thick and fibrocartilaginous in nature and is called **tympanic annulus**. The annulus sits in the groove in the tympanic bone, which is called the tympanic sulcus. This sulcus is deficient superiorly where it forms the **notch of Rivinus**. The anterior and posterior malleal folds arise from the tympanic sulcus at the junction of the notch of Rivinus and are attached to the lateral process of the malleus.

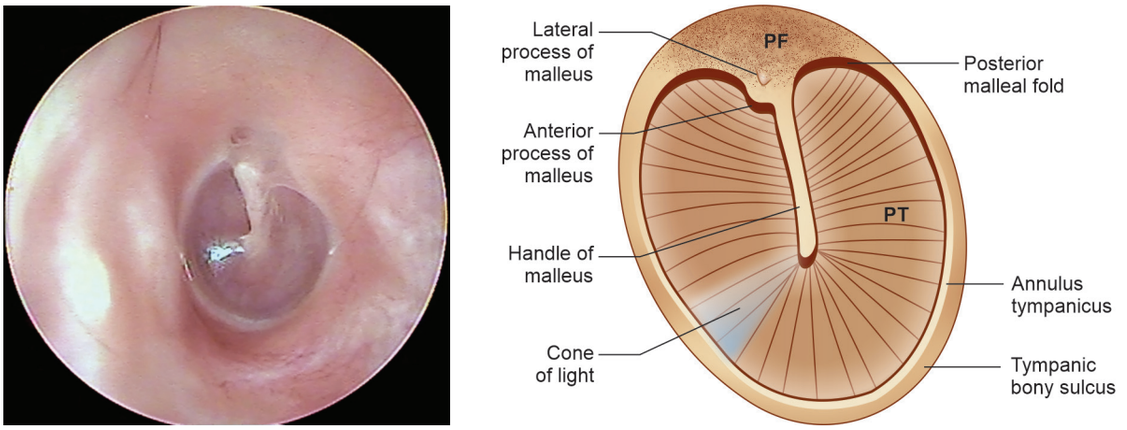


Fig. 1.10: Structure of tympanic membrane

The tympanic membrane can be divided into two parts, **pars tensa** and **pars flaccida**. The area between malleolar folds and the notch of Rivinus, is the pars flaccida. The rest of the tympanic membrane is called the pars tensa. The tip of the malleus attaches to the centre of the membrane, which is called the *umbo* (Fig. 1.10). The tympanic membrane has three layers: outer epithelial, middle fibrous (lamina propria) and inner mucosal. In pars tensa, the fibrous layer has radially oriented outer fibres, circular, parabolic and transversely arranged deeper fibres. These well-arranged fibres give rigidity to this part of the tympanic membrane and therefore is called pars tensa. In case of pars flaccida, the fibrous layer is less marked and the arrangement of the fibres is also random, hence the term pars flaccida.

Blood supply

- Epidermal surface of the tympanic membrane is supplied by the deep auricular branch of the maxillary artery.
- Mucosal surface of the membrane is supplied by the anterior tympanic branch of maxillary and stylomastoid branch of the posterior auricular artery.

Nerve supply: It is supplied by the branches of auriculotemporal nerve (V3), auricular branch of vagus and tympanic branches of IX nerve.

MIDDLE EAR SPACE

The tympanic cavity or the middle ear space is classically divided into 3 parts but with the advent of the ear endoscopes it has now been divided into 5 parts (Fig. 1.11).

The parts of the tympanic cavity are:

1. Epitympanum
2. Mesotympanum
3. Hypotympanum
4. Retrotympanum
5. Protympanum.

- The epitympanum or attic is the portion of the middle ear above the level of malleal folds.

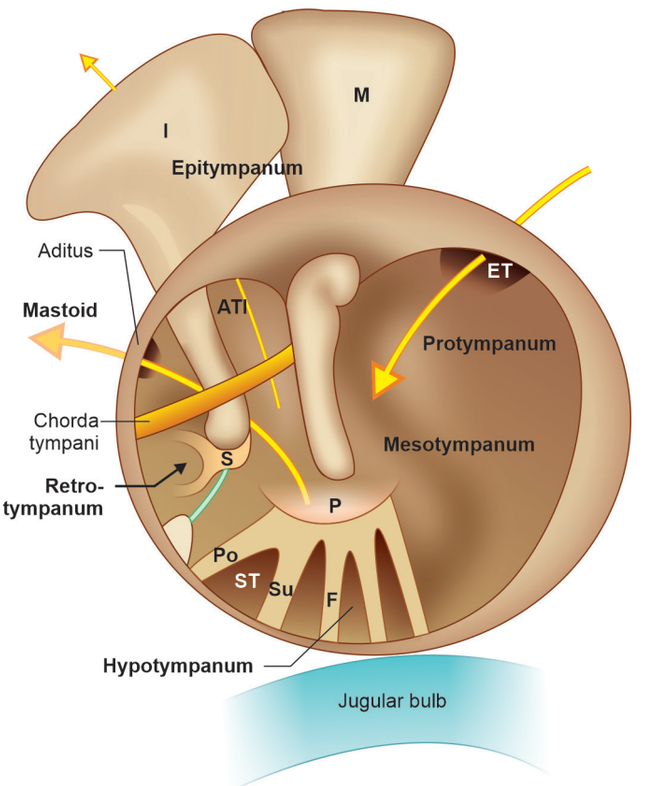


Fig. 1.11: Parts of the middle ear cavity and the ventilation pathways of middle ear cleft (*ATI: Anterior tympanic isthmus; I: Incus, M: Malleus; S: Stapes; ET: Eustachian tube; C: Chorda tympani; ST: Sinus tympani; P: Promontory; F: Funiculus; Su: Subiculum; Po: Ponticulus; J: Jugular bulb*)

- The mesotympanum lies below the level of malleal folds and is separated from the epitympanum by a series of mucosal folds and membranes. It is part of the tympanic cavity which corresponds to the pars tensa of the tympanic membrane. The mesotympanum is part of the tympanic cavity which can be seen via the external auditory canal by otoscope or microscope.
- The hypotympanum is the portion of the middle ear cavity below the level of tympanic sulcus.

- The retrotympanum includes the posterior wall and posteromedial portion of the tympanic cavity which can be seen only by using angled endoscopes via the large perforation or after elevation of tympanomeatal flap. This part of the middle ear cavity can sometimes be visualised through a large pars tensa perforation).
- The protympanum is the part anterior to the promontory which joins with the tympanic portion of the Eustachian tube. Recent studies, physiologically consider protympanum to be lateral most part of the Eustachian tube.

WALLS OF THE TYMPANIC CAVITY

The tympanic cavity resembles a cube with six sides.

1. **Lateral wall:** It is formed by the tympanic membrane and outer attic/epitympanic wall (scutum). The scutum erosion is a sign squamosal type of chronic otitis media.
2. **Roof:** The roof of the epitympanum is formed by the bony plate called the *tegmen tympani*. This separates the middle ear from the middle cranial fossa. **Cog or spur** is a bony projection from the tegmen which divides the epitympanum into a smaller anterior epitympanic space (AES) and the larger posterior epitympanic space (PES). The anterior epitympanic space may be the site for residual cholesteatoma (one of the *hidden areas* of microscopic ear surgery) (Fig. 1.12).
3. **Floor:** The floor separates the hypotympanum from the dome of the jugular bulb. It may sometimes be absent leading to dehiscent jugular bulb, which can increase risk of jugular bulb injury at the time of middle ear surgery (Fig. 1.13).
4. **Anterior wall:** The anterior wall is narrow because of convergence of the medial and the lateral walls anteriorly. The lower 1/3rd of the anterior wall is separated from the carotid artery by a thin plate. The

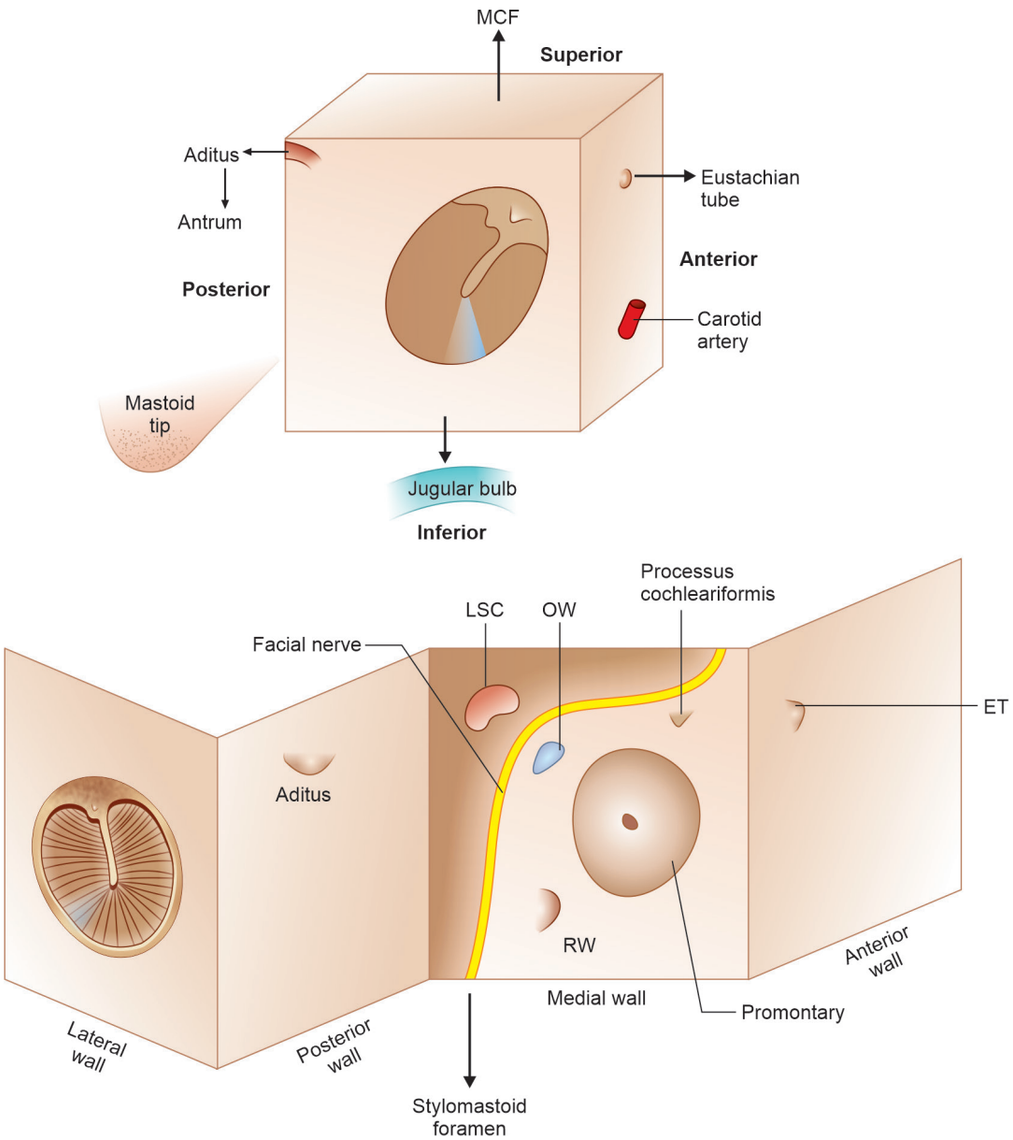


Fig. 1.12: Relation of middle ear with surrounding important structures

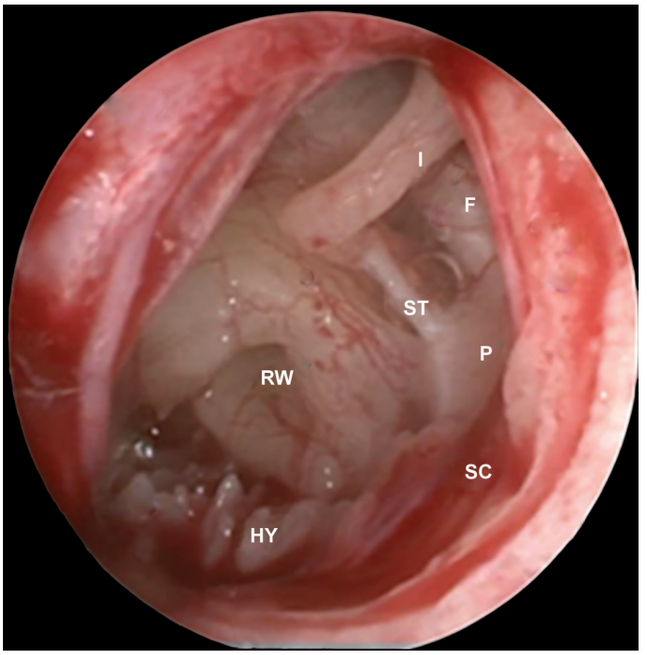


Fig. 1.13: Endoscopic view of hypotympanic cells and floor
I: Incus; ST: Stapedial tendon; F: Facial nerve; P: Pyramid; SC: Styloid complex; HY: Hypotympanic cells; RW: Round window

middle part of the anterior wall has the Eustachian tube orifice. The canal for the tensor tympani muscle lies above the Eustachian tube orifice.

5. **Medial wall:** It shows a rounded bulge of the basal turn of the cochlea called **promontory**. It has numerous grooves for the nerves of the tympanic plexus derived from the glossopharyngeal nerve and the sympathetic fibers along the carotid artery.

   Above and behind the promontory is the **oval window** niche which has a kidney shape. It is closed by the footplate of the stapes. Its average size is 3.25 mm in length × 1.75 mm in width.

   A triangular niche called the **round window** lies inferior and posterior to the oval window. **Subiculum** is a bony ridge, which extends posteriorly from the round window, outward to the styloid eminence region. Another bony ridge called the **ponticulus** extends from the pyramidal process towards the promontory. Ponticulus lies above the subiculum and area between the two is called the **sinus tympani**. **Funiculus** ('meaning borderline') is a bony ridge extending from the promontory to the jugular dome, which separates the retrotympanum from the hypotympanum (Fig. 1.14).

   The bony canal of facial nerve which is also called as **fallopian canal** runs above the promontory and oval window from anterior to posterior direction. Anteroinferior to the first genu of the facial nerve, the medial wall of the middle ear is marked by a bony projection called as **processus cochleariformis**. The tendon of the tensor tympani muscle takes a turn here

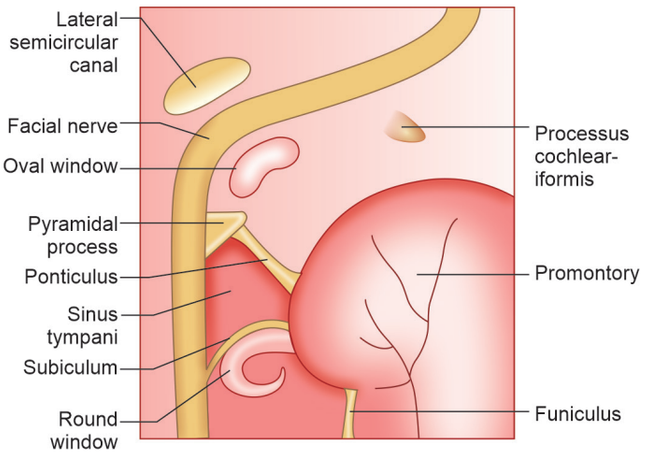


Fig. 1.14: Structures over the medial wall

and gets inserted on the medial aspect of the neck of the malleus. Posteriorly, behind the oval window the nerve turns inferiorly and progresses in the posterior wall of the tympanic cavity.

6. **Posterior wall:** The superior aspect of the posterior wall of the middle ear cavity communicates with the mastoid air cell system through an opening called the **aditus.** Just below the aditus lies the **fossa incudis**, which lodges the short process of incus. Just below the fossa incudis, the posterior wall has a bony projection called **pyramid**. It gives rise to the tendon of the stapedius muscle.

Facial recess is a triangular space bounded by the facial nerve medially, chorda tympani laterally and fossa incudis superiorly. This recess is utilized for the **posterior tympanotomy approach** to the middle ear especially retrotympanum (Fig. 1.15).

Sinus tympani is the posterior extension of the mesotympanum into the posterior canal wall. It is bounded by ponticulus superiorly and subiculum inferiorly. It lies medial to the facial nerve and can be deep up to 9 mm. It is one of the hidden areas and a common sites for the residual disease in cholesteatoma surgery. It can be best visualised by the use of angled endoscopes.

Contents

Ossicles

1. **Malleus** is the most lateral and the largest ossicle. It resembles a 'hammer'. It has a head, neck and handle or manubrium. The head of malleus articulates with the body of the incus in the epitympanum. Below the neck there are two processes called the anterior and lateral process. The anterior and posterior malleal folds which arise from the annulus tympanicus, are attached to the lateral process of malleus. The upper part of the manubrium receives the tendon of tensor tympani muscle (Fig. 1.16).

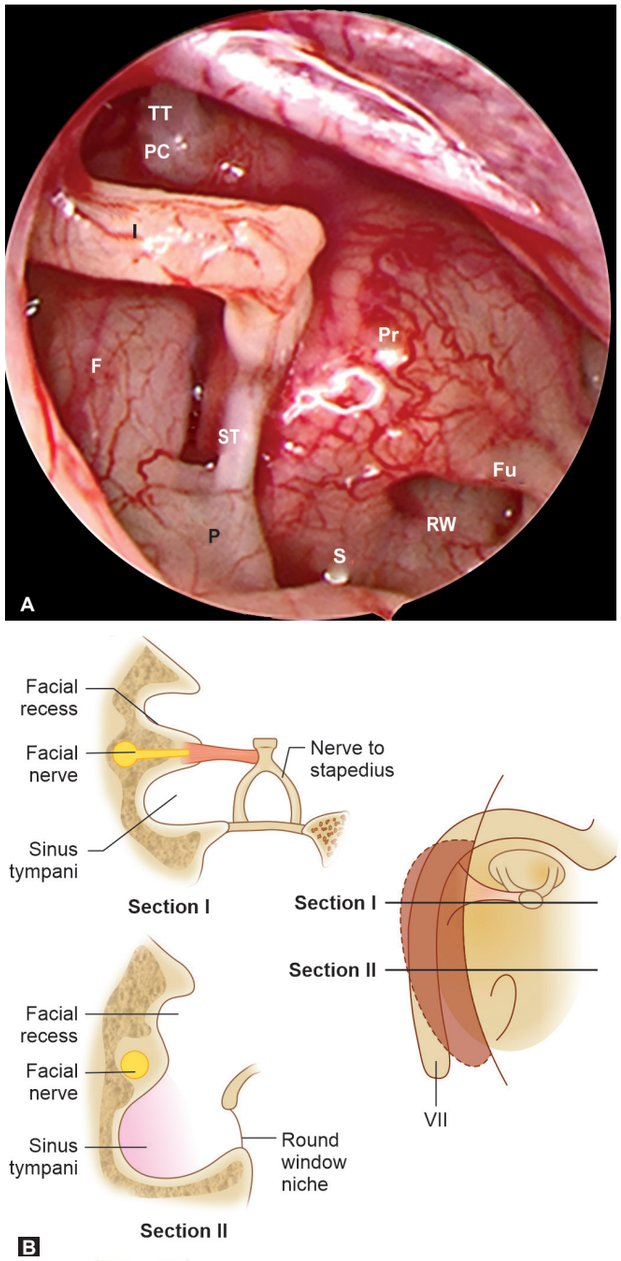


Fig. 1.15: Relationship of facial recess, facial nerve and sinus tympani. (A) Endoscopic view; (B) Diagrammatic representation
F: Facial nerve; P: Pyramid; RW: Round window; PR: Promontory; I: Incus; ST: Sinus tympani; PC: Processus cochleariformis; FI: Funiculus; S: Subiculum

2. **Incus** resembles an 'anvil'. It has a body, short process and long process. The long process has a slender lenticular process at its end, to which articulates the stapes head. The lenticular process is sometimes called the *fourth ossicle* or a sesamoid bone (Fig. 1.17).
3. **Stapes** resemble a 'stirrup'. It has a head, neck, anterior and posterior crura and the footplate. The stapedial tendon is attached to the posterior part of the neck and posterior part of posterior crura. The dimensions of the foot plate are 3 mm × 1.4 mm. The footplate is surrounded by the annular ligament (Figs 1.18 and 1.19).

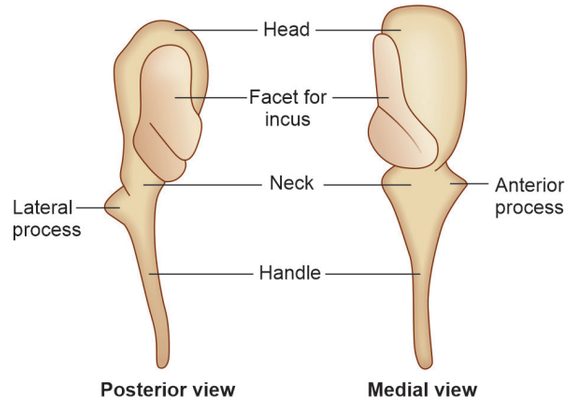


Fig. 1.16: Parts of malleus

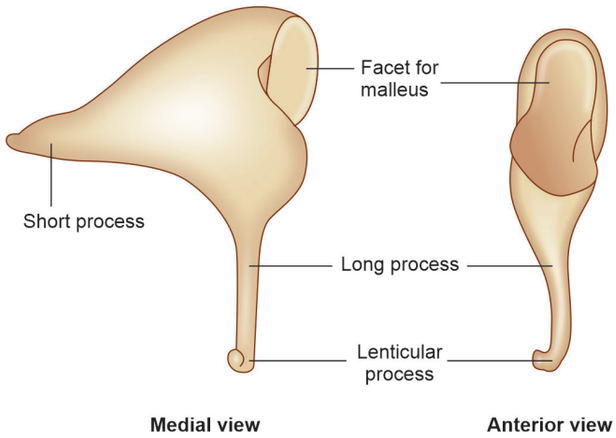


Fig. 1.17: Parts of incus

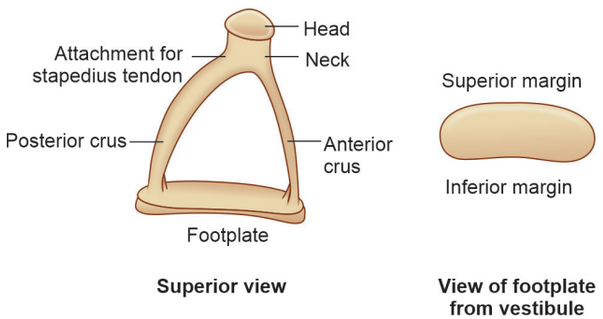


Fig. 1.18: Parts of stapes

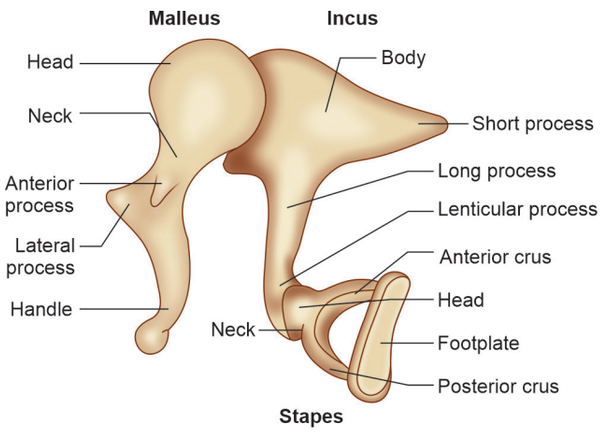


Fig. 1.19: Ossicular chain

MASTOID AIR CELL SYSTEM

The mastoid air cell system causes the pneumatization of the bony cortex of the mastoid bone. The pneumatization of mastoid is variable and is of three types:

1. *Cellular type or well pneumatized*: It has well-developed mastoid air cells with thin intervening septi (Fig. 1.20).
2. *Diploic*: There are few air cells with thick septa in-between.
3. *Sclerotic or acellular*: There is lack of pneumatization; therefore, no cells are seen apart from mastoid antrum. The mastoid antrum is always present though it may be contracted in size. A **'contracted antrum'** usually associated with low-lying dura and forwardly placed sigmoid sinus (Fig. 1.21).

There are various groups of air cells found in the mastoid and have been named according to their location:

Mastoid antrum*: It is the largest mastoid air cell and communicates with the middle air directly via the aditus. It is present at the depth of 1.5 mm from the mastoid cortex externally. It is a surface marked by the **suprameatal triangle (McEwen's triangle).** The volume of the antrum is approximately 2 ml. Its roof is formed by the *tegmen antri* (which separates it from the middle cranial fossa) and medial wall relates to lateral and posterior semicircular canals. While performing mastoidectomy, sometimes Korner's septum is encountered which gives a false impression of reaching the antrum but the actual antrum is still deeper and is found only after its removal.*

Boundaries of the Supramental Triangle (Fig. 1.22)

Anteriorly: The posterosuperior border of the bony external auditory canal along with the suprameatal spine of Henle.

Superiorly: The supramastoid crest or superior temporal line.

Posteriorly: A vertical line drawn tangent to the posterior margin of the external auditory canal.

Various Cells in the Mastoid Air Cell System

1. Periantral cell
2. Tegmen cells
3. Perisinus cells
4. Retrofacial cells
5. Perilabrynthine cells
6. Peritubal cells
7. Tip cells
8. Marginal cells
9. Zygomatic cells
10. Squamosal cells.

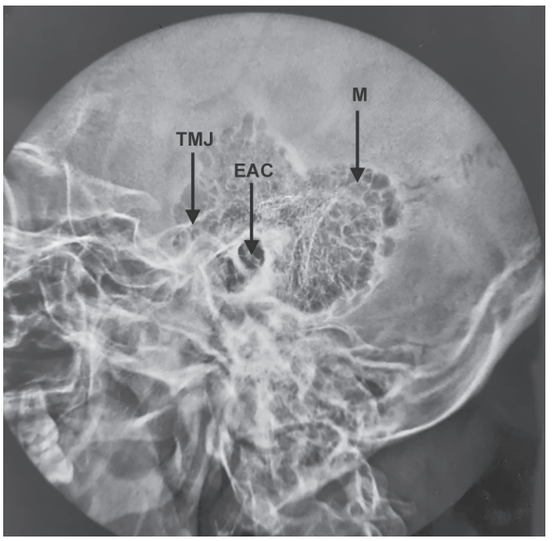


Fig. 1.20: X-ray mastoid—well pneumatized mastoid

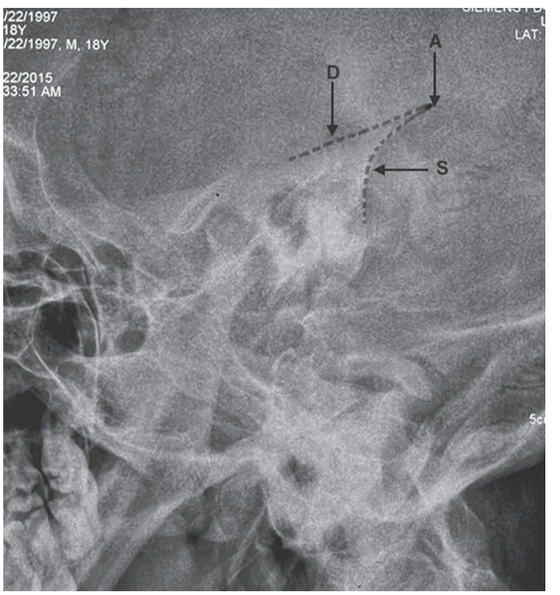


Fig. 1.21: X-ray mastoid—sclerosed mastoid
(EAC: External auditory canal; M: Mastoid; TMJ: Temporomandibular joint; D: Dural plate; S: Sinus plate; A: Sinodural angle)

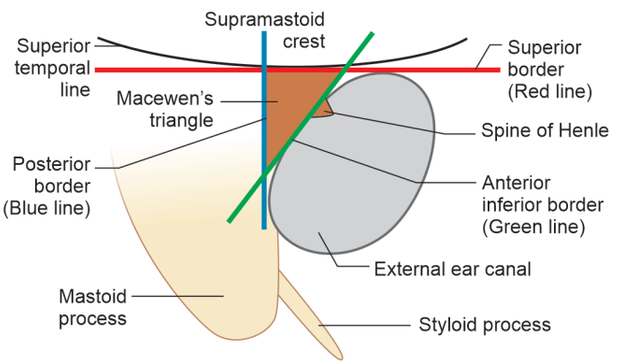


Fig. 1.22: Boundaries of the suprameatal triangle

**Korner's septum* is formed due to the persistence of petrosquamosal lamina. This bony lamina is seen during embryological stages at the junction of saccus superior (mastoid pneumatization) with saccus medius (petrous pneumatization). The persistence of this bony lamina forms the Korner's septum.

Mucosa of the Middle Ear Cleft

The middle ear cavity is lined by ciliated columnar epithelium in the anterior, inferior parts and cuboidal epithelium in the posterior part. The epitympanum and the mastoid air cell system are lined by flat non-ciliated epithelium without goblet cells and mucous glands. The lining of the nasopharynx is continuous with the middle ear cleft. The middle ear mucosa also covers the ossicles, muscles, nerves and ligaments in a manner similar to the peritoneum lining the abdominal viscera. Thus it forms various mucosal folds connecting the middle ear structures. The **epitympanic diaphragm** separates the epitympanum from mesotympanum. The ventilation of the epitympanum comes from the mesotympanum is via two openings called—anterior tympanic isthmus (ATI) and posterior tympanic isthmus (PTI).

The Eustachian tube is lined by ciliated epithelium near the nasopharyngeal end, pseudostratified columnar in the middle and columnar with mucous glands near the middle ear end.

Tympanic Plexus

The tympanic plexus is formed by the tympanic branch of the glossopharyngeal nerve **(Jacobson's nerve)** and the branches from the sympathetic plexus around the internal carotid artery. The plexus provides branches to the tympanic membrane and mucosal lining of the middle ear cleft.

Blood Supply of Middle Ear Cleft

The main blood supply of the middle ear cleft is derived from branches of the maxillary artery with some contributions from the internal carotid artery.

Lymphatic Drainage of Middle Ear

Middle ear and Eustachian tube drain into the retropharyngeal and level V lymph nodes.

> ***Note:*** The inner ear does not have lymphatics.

INNER EAR (LABYRINTH)

It has mainly two parts, namely the bony and membranous labyrinth. The bony labyrinth houses the membranous labyrinth containing the sensory portion of cochlea and vestibular structures.

Bony Labyrinth

It is the dense bony shell surrounding the vestibule, the semicircular canals and the cochlea.

The **vestibule** is the central portion of the bony labyrinth, which lies between the middle ear and the internal auditory meatus. It is 5 mm long, 5 mm high and 3 mm deep. **Fenestra vestibuli** is an opening on the lateral wall of vestibule, which is closed by the footplate of stapes.

The posterior wall of the vestibule has the five openings of the semicircular canals. The anterior wall of the vestibule contains the elliptical opening for the scala vestibuli of the cochlea. On the medial wall, anteriorly is the spherical recess for **saccule** from where starts the inferior vestibular nerve. Behind the saccule is the ridge called vestibular crest, lower end of which has the cochlear nerve fibres. Above and behind the crest is the elliptical recess for the **utricle**. The vestibular aqueduct opens below the elliptical recess. The aqueduct passes through the temporal bone to open into the posterior cranial fossa.

The semicircular canals (SCCs) are three in number—superior, posterior and lateral SCCs. Situated behind and above the vestibule, each SCC forms two-third turns of a circle with a diameter 0.8 mm. One end of each canal is dilated to form the **ampulla**. Each ampulla contains the vestibular sensory epithelium. Each ampullated end opens independently into the vestibule.

The non-ampullated ends of the superior and posterior SCC join to form the **crus commune**. The non-ampullary end of the lateral SCC opens into the vestibule independently below the crus commune. The lateral SCC bulges into the posteromedial wall of epitympanum. The apex of superior SCC lies close to the floor of the middle cranial fossa and the petrous bone over it is called the *arcuate eminence* and is important landmark during middle cranial fossa surgeries.

The cochlea is placed anterior to the vestibule and appears like a snail shell having approximately *two and half turns*. The cochlea coils about a central cone called the *modiolus*, tapering from the base to the apex.

The apex of the cochlea points laterally and forwards positioned at the upper part of the medial wall of the tympanic cavity. The basal turn of the cochlea forms the bulge of **promontory**. A thin bone arises from a modiolus spiral upwards in the lumen of cochlea as the spiral bony lamina. The membranous spiral lamina arises from this thin bone and divides the cochlea into *scala vestibuli*, *scala tympani* and *scala media* as part of membranous cochlea (Fig. 1.23).

The scala tympani is a blind tube having **fenestra cochleae** at its base, which is called *round window* and is closed by secondary tympanic membrane. It is also connected with the subarachnoid space through the *aqueduct of cochlea*. The scala vestibuli is closed at the oval window by the footplate of the stapes. The scala tympani and vestibule are both filled with perilymph and have communication with each other at the apex of cochlea called the *helicotrema*.

Membranous Labyrinth

It consists of a series of sacs filled with endolymph. It consists of utricle, saccule, cochlear duct (scala media) and semicircular canals.

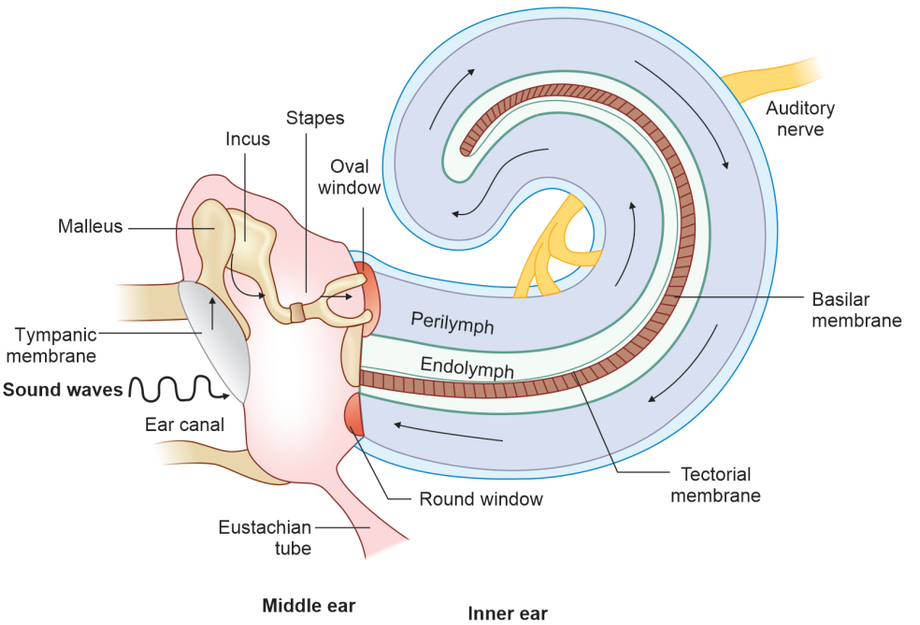


Fig. 1.23: Structure of cochlea

Scala Media (Cochlear Duct)

The cochlear duct is a tube within the bony cochlea. Its average length is around 34 mm. It is triangular in section and has three walls:

1. Basilar membrane which supports the organ of Corti.
2. Reissner's membrane which separates it from scala vestibuli.
3. Stria vascularis contains the vascular epithelium which contributes to the secretion of the endolymph.

Organ of Corti

On the floor of the cochlear duct, two ridges, the upper ridge supports the tectorial membrane and the lower ridge giving rise to membranous spiral lamina with fibres of acoustic nerve going to the organ of Corti.

The membranous spiral lamina has three layers:

1. Flattened epithelium of scala tympani (below).
2. Middle fibrous layer
3. Upper surface of the organ of Corti.

The organ of Corti is a complex arrangement of auditory sensory cells and supporting cells sitting on the basilar membrane. The parts of the organ of Corti are (Fig. 1.24):

1. **Sensory hair cells:** These cells have clusters of fine filaments projecting from the upper surface, called stereocilia. They are distinctively of two types:

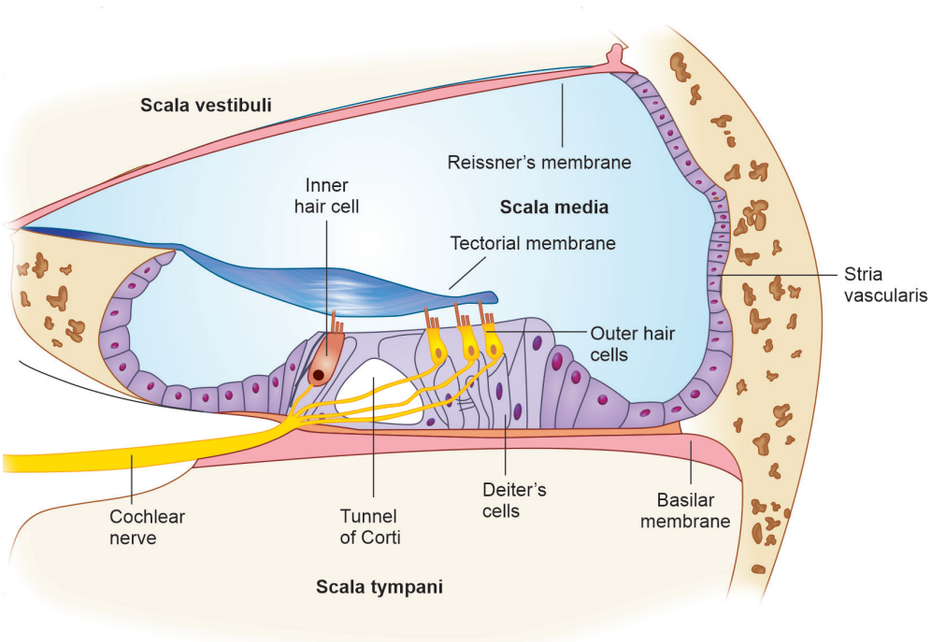


Fig. 1.24: Structure of organ of Corti

The ***inner hair cells (IHC)*** are a single row of cells on the inner or medial side of the spiral. The inner hair cells are flask shaped with a small apex and large cell body. They are richly supplied by afferent cochlear nerve fibres.

The ***outer hair cells (OHC)*** are cylindrical in shape and are arranged in 3–4 rows. They are present on the outer or lateral side of the spiral. The outer hair cells modulate the function of the inner hair cells.

2. **Extracellular spaces:** These are spaces within the organ of Corti, namely **spaces of Nuel** (around the OHC) and **tunnel of Corti** (between the OHC and IHC).
3. **Tectorial membrane:** It is an a highly hydrated extracellular matrix which lies over the mechanosensory hair cells in the cochlea. It has three main constituents: water (97%), glycosaminoglycans (GAG), collagenous and noncollagenous proteins. Recent studies show that mutations in genes controlling the constituent proteins lead to significant hearing losses. It is extremely sensitive to distortion and shrinkage. The tips of the longest stereocilia of OHC are embedded or attached to the tectorial membrane. The relative movement of hair cells and tectorial membrane generate stimulus for the hair cells.
4. **Supporting cells:** These are highly specialised and differentiated cells which provide support to the outer hair cells. These are the Pillar cells, Deiter's cells and Hensen's cells.

Nerve Supply of Cochlea

The nerve supply to the cochlea is from the brainstem via the afferent and efferent fibres.

Afferent nerve fibres that innervate the OHC and IHC are of mainly two types. Majority are type 1 myelinated neurons that innervate IHCs. The remaining 5–10% fibres are unmyelinated type 2 neurons which innervate the OHCs. All fibres collect in the spiral ganglion enclosed within the *Rosenthal canal*. The efferent neurons which innervate the cochlea have their cell bodies in the mid-brain regions.

Labyrinthine Fluids

1. **Perilymph:** It fills the space between the membranous and bony labyrinth. Its exact source is not known. Though it has been suggested to be a filtrate of blood serum from capillaries of spiral ligament or due to direct communication from CSF via the aqueduct of cochlea. It resembles plasma, interstitial fluid and CSF in composition (high in Na^+ and low in K^+ ions).
2. **Endolymph:** It fills the membranous labyrinth and it resembles the intracellular fluids. It is rich in K^+ ions and low in Na^+ ions. It is secreted by the stria vascularis of cochlea and the dark cells of the utricle and ampullated ends of SCC.

Blood Supply of Labyrinth

The main blood supply of the inner ear is from the labyrinthine artery which is a branch of the anterior inferior cerebellar artery (AICA). Labyrinthine arteries may sometimes arise directly from the basilar artery or even vertebral artery (Fig. 1.25).

The venous drainage is via anterior, posterior spiral vein and vestibular vein. These finally form the vein of cochlea which drains into the jugular bulb.

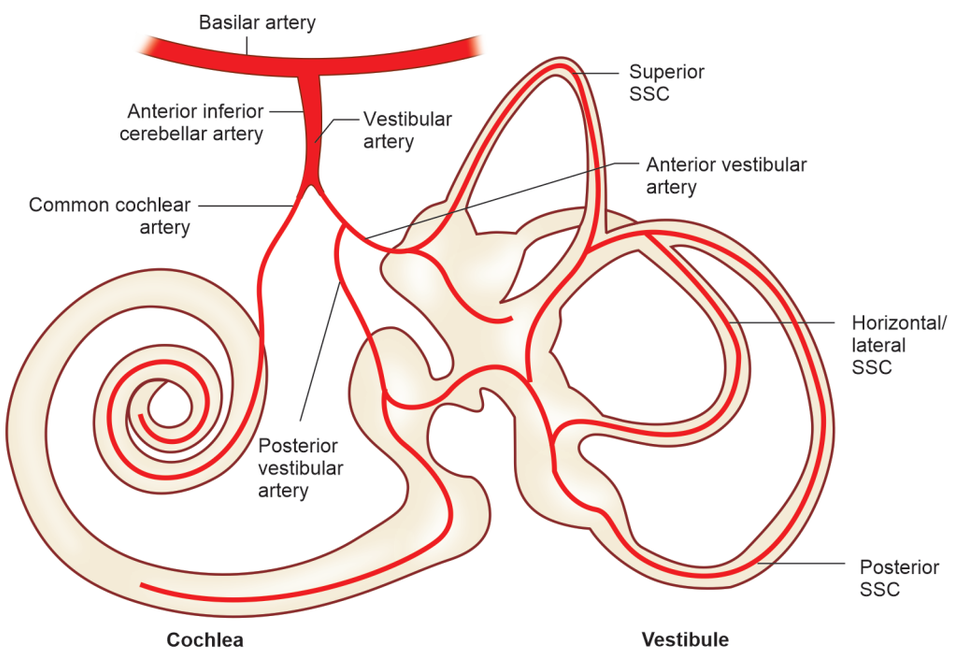


Fig. 1.25: Blood supply of membranous labyrinth

Chapter

2



Physiology of Hearing

EN1.1: Describe the physiology of ear.
PY10.15: Describe and discuss functional anatomy of ear and auditory pathways and physiology of hearing.

The sound requires a physical medium to travel. The sound generated by a vibrating object produce waves of compression and rarefaction in the conducting medium. It should also be understood that there is no actual movement of particles of the medium, but they only vibrate about their positions and transfer the vibrations forward. Hence, the propagation of sound is longitudinal in nature. A sound wave velocity is different in different media. In air at 20° celsius, at sea level, sound travels at the speed of 344 m/s. This speed increases in denser mediums. The speed of sound in water is around 1500 m/s. The vibratory energy which arrives at the ear, gives rise to sound sensation or hearing.

The two key properties of sound are its frequency and loudness. The frequency is linked to pitch and loudness is linked to intensity of sound.

The **Frequency** is measured in Hertz (Hz). It refers to the rate of vibration of the sound travelling through the air. The frequency range of human ear is between 20 to 20000 Hz.

The **Intensity** of sound is the strength of sound which determines its loudness. It is measured in decibels (dB). A decibel (dB) is not an absolute measure of intensity. It is a ratio. It complies that the decibel is the logarithm of intensity of sound divided by some agreed reference intensity.

dB = 10 Log 10 Intensity measured upon the intensity reference.

Hence on the decibel scale,

- 0 dB is the smallest audible sound.
- A sound 10 times more powerful is 10 dB.
- A sound 100 times more powerful than near total silence is 20 dB.
- A sound 1,000 times more powerful than near total silence is 30 dB and so on.

Whisper has a sound intensity of 20 dB, normal speech of 60 dB, shouting is of 90 dB, loud discomforting sounds is at 110 dB. The ear pain is produced by the sounds of intensity 120 dB and above (Table 2.1).

Table 2.1: Sound intensity levels

Source of sound	*Intensity level in dB*
Threshold of hearing	0
Rustle of leaves	10
Whisper	20
Ordinary conversation	65
Busy street traffic	70
Auto interior moving at 90 kph	75
Loud indoor rock concert	100
Threshold of pain	120

Loudness is the subjective sensation of an individual to an intensity of sound.

Acoustic impedance is the stiffness of the medium to the propagation of sound. The denser medium has a higher sound propagation velocity but tightly packed atomic structure provides resistance to molecular movement velocity. This resistance results in reduction of acoustic pressure transmission. This is especially relevant in our ear because the sound reaches our ear via air and finally is transmitted to fluid-filled cochlea, which would provide acoustic resistance. The acoustic resistance is overcome by outer and middle ear impedance matching mechanism, which leads to effective acoustic energy transmission into the cochlea.

APPLIED PHYSIOLOGY OF HEARING

The human ear can hear sounds between the frequency range of 20 to 20,000 Hz. The sound transmission from the environment to the auditory cortex in the superior temporal gyrus can be broadly divided into four parts:

1. Pinna and external auditory canal conduction pathway: Capture of sound by pinna and its conduction to the tympanic membrane by the external acoustic meatus.
2. Transformer action of the middle ear.
3. Transduction of the mechanical energy to electric impulses in the cochlea.
4. Retrocochlear signal transmission to the auditory cortex.

1. Pinna and EAC

Pinna: The sound vibrations are picked up by the pinna. The pinna is vestigial in humans because of the non-functional auricular muscles. In animals though, it helps to concentrate acoustic signals and transfer to the external auditory canal thereby helping to detect soft sounds and along with spatial resolution.

There are numerous anatomical ridges and convolutions in the human pinna. These are effective in transmission of horizontal and vertical, directional sounds to the external auditory canal. Pinna sound concentrating mechanism is most effective at above 1 kHz.

Clinical Importance

The anatomical abnormalities of pinna have been seen to closely relate to the external auditory canal abnormalities and sometimes with inner ear disorders.

> ***Note:*** Pinna and EAC impression is an important step in the placement of hearing aids. The concha and the antihelix are used to anchor the hearing aids.

External auditory canal (EAC) is considered like a resonating tube. The amplification of sound is in the range of 10–20 dB and is best useful for sounds with frequencies ranging from 2 to 7 kHz.

The EAC and bilateral pinna unit provide monaural and binaural clues for the localization of the sound.

2. Transformer Action of Tympanic Membrane and Middle Ear

The middle ear along with the tympanic membrane functions as an efficient dual apparatus to overcome the impedance of the acoustic signal at the air to fluid interface of middle ear with cochlea. There are various mechanisms involved in overcoming this impedance.

1. *Hydraulic amplification* (*Area ratio amplification*): The transfer of acoustic energy from the relatively large area of the tympanic membrane, which is 55 mm^2 (effective vibratory area) to that of much smaller area of the stapes footplate, which is 3.2 mm^2. The area ratio of the two is 17:1. The pressure gain is around 26 sound pressure level (SPL). The tympanic membrane works at a range of 1–4 kHz. The hydraulic amplification contributes to **17 times magnification** of sound energy.

> ***Note:*** In defects of the tympanic membrane or diseases like tympanosclerosis, the effective vibratory capacity is lowered leading to hearing loss.

2. *Lever action of the ossicles*: The length of the malleus handle is 1.3 times longer than the incus long process. This results in increased force transmission at the incus end. It mainly contributes to the pressure gain between 0.25 and 2 kHz. This mechanism leads to **magnification of sound by 1.3 times** (Fig. 2.1).

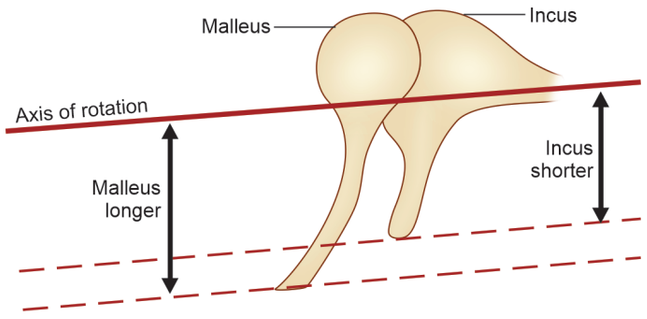


Fig. 2.1: Lever action of ossicles

3. *Curvature of the tympanic membrane* (***Catenary lever action***)-Helmholz 1868: The movement of the tympanic membrane is more at the periphery than at the centre where the malleus handle is attached. This leads to buckling of the membrane and this provides for additional pressure gain. **(It magnifies sound by 2 times, and produces 6 dB of hearing gain)** (Fig. 2.2).

The **net mechanical gain** due to the above mechanisms is $17 \times 1.3 \times 2 =$ **44.7 times**. In dB scale 20 (log 44.7) = 33 dB. Without the transformer action of the middle ear, 99% of the incident energy would have been reflected back. Additional 4 dB gain is provided by the *phase difference* of the oval window and the round window, which is explained below.

Middle Ear Windows

The middle ear communicates with the inner ear via two natural windows. The pressure difference between the oval and the round windows is the basis for the cochlear wave formation. In normal conditions the sound waves do not reach the two windows simultaneously because of preferential transmission of sound waves to the oval window via the ossicles. The time lag or *phase difference* leads to the cochlear fluid movement. In case this phase difference is disturbed, the both windows will try to produce compression waves into the cochlea, eventually leading to non-movement of fluid. This will lead to hearing losses. This acoustic difference is achieved by intact tympanic membrane and ossicular chain assembly.

Rarely there may be a virtual third window, thereby affecting preferential stapes-oval window sound distribution. The large vestibular aqueduct is one such virtual window which might lead to hearing loss. Another example is the dehiscence of the superior semicircular canal (SSCD).

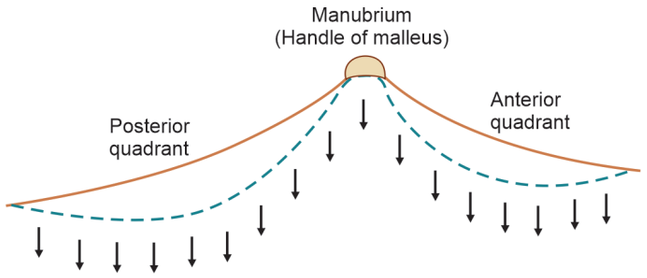


Fig. 2.2: Catenary lever action

Bone Conduction

Apart from the sound transmission via the oval window, the cochlea may also be stimulated by the bony skull composed of canal, ossicles and bony cochlea. These methods include:

1. Vibration induced compression and distortion of the cochlear space, thereby leading to distortion of the basilar membrane.
2. Vibration induced inertia of the ossicles transferring sound to the bony labyrinth.
3. Vibrations of sound may directly stimulate TM and ossicles which transmits sounds to the cochlea.

Middle Ear Muscles

The two middle ear muscles, stapedius and tensor tympani, participate to prevent damage to the delicate sensory epithelium of the cochlea by reducing the conduction of high intensity sounds.

The **stapedius muscle** is supplied by the facial nerve and it takes 100–200 ms to fully contract. It moves about 50 microns and stiffens the ossicular chain and helps to decrease the suprathreshold sounds of above 80 dB. Its contraction produces a reduction in the sound intensity by 30 dB. Due to lag in contraction of a few ms, the high intensity sudden sounds escape the stapedial reflex and might lead to cochlear damage.

The **tensor tympani muscle** is supplied by the Vth nerve. It attaches to the malleus manubrium and pulls it anteriorly. It becomes active whenever there is tactile stimulation of EAC, and face, swallowing, head movement, pressure over eyeballs and forced closing of eyelids.

4. Conversion of Mechanical Energy to Electrical Wave in Cochlea or Cochlear Mechanics

The mechanical movement of the footplate on the oval window induces the movements of the cochlear fluids, thereby generating a travelling wave (1st described by Von Bekesy). The wave impinges on the base of the cochlea first and then travels all the way up to the apex. The high frequency sounds produce maximum displacement along the basilar membrane towards the base of the cochlea and this movement response shifts towards the lower frequency at the apex (Fig. 2.3).

Sound transmitted to cochlea undergoes two significant processing:

1. Biological and active compression and amplification by the outer hair cells (OHCs).
2. Electrochemical transduction by the inner hair cells (IHCs).

The final neural output is a coded and gated signal carried by the auditory nerve via the spiral ganglion cells to the brainstem.

Hence the cochlea is responsible for the **frequency selection/analysis, loudness discrimination, temporal resolution, and spectral analysis** of the incoming sound.

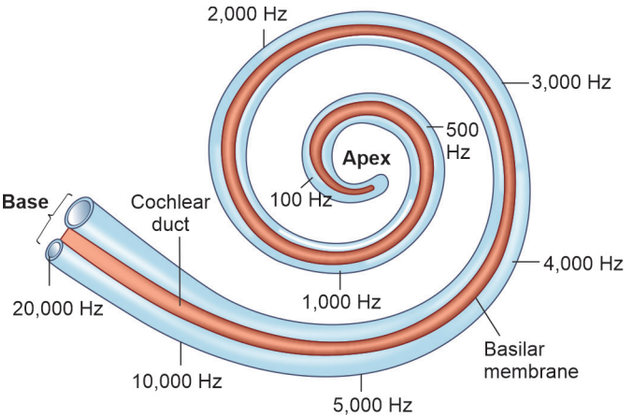


Fig. 2.3: Frequency of sound perception in cochlea

The dynamic range of the human ear is up to 120 dB, therefore high signal intensity must be modified and compressed. This is done via the outer hair cells (OHCs). The dynamic range is significantly lost and reduced in cochlear pathologies.

5. Auditory Neural Pathway

There are approximately 30,000 auditory nerve fibres which are innervating the IHCs via the bipolar neurons of the spiral ganglion in the Rosenthal's canal. These end up in the dorsal and ventral cochlear nuclei in the brainstem (Fig. 2.4).

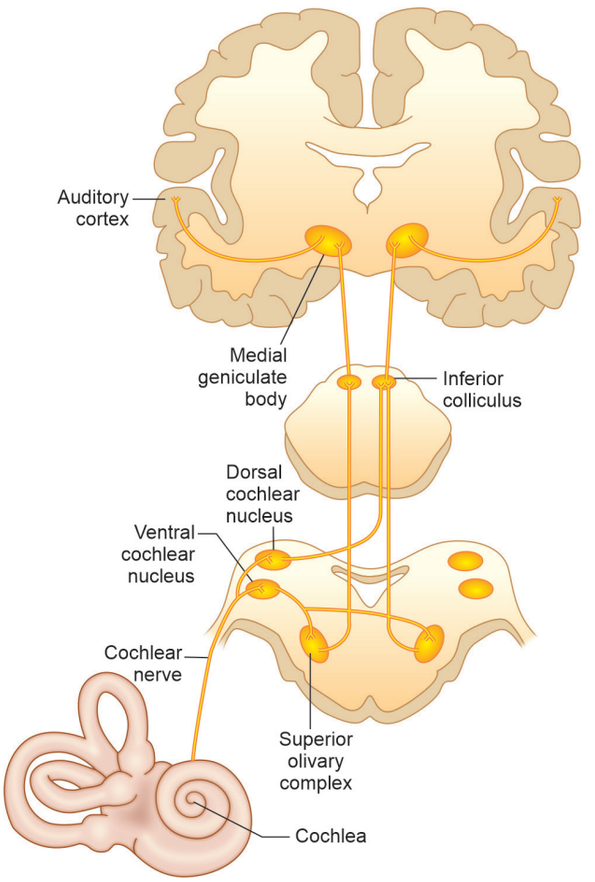


Fig. 2.4: Central auditory pathway

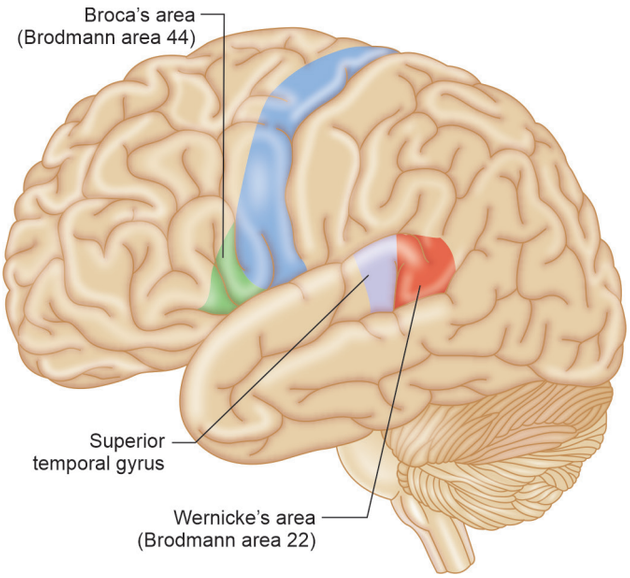
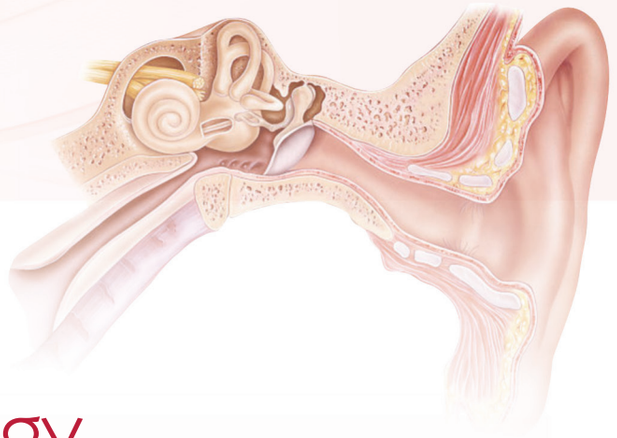


Fig. 2.5: Auditory cortex–Brodmann Area 41, 42

From the cochlear nuclei, there are serial connections with central ascending tracts on the ipsilateral and contralateral side. The cochlear nuclei are connected to the ipsilateral and contralateral superior olivary complex, lateral lemniscus and then to inferior colliculus. Inferior colliculus connects to the medial geniculate body and which finally provides auditory information to the auditory cortex in superior temporal gyrus (Fig. 2.5).

Chapter

3



Audiology

EN2.4: Demonstrate the correct technique of performance and interpret tuning fork tests.
EN2.8: Demonstrate the correct technique to perform and interpret pure tone audiogram and impedance audiogram.
EN4.16: Observe and describe the indications for and steps involved in the performance of pure tone audiometry.
EN4.17: Enumerate the indications and interpret the results of an audiogram.
PY10.16, PY10.20

Before we start the clinical testing of hearing, we should be knowing about the types of hearing loss (Fig. 3.1).

Hearing loss is a condition of partial or complete inability to hear from one or both ears.

TYPES OF HEARING LOSS

Conductive hearing loss: It is caused by the disease affecting the transmission of sound from the external ear to the oval window. The various causes may be infective, obstructive, fluid in middle ear, perforation of tympanic membrane, damage to ossicles, etc.

Sensorineural hearing loss: It results from lesions of the cochlea (sensory causes) or the pathologies of VIII nerve and central auditory pathway. The VIII nerve causes (peripheral causes) along with central auditory pathologies (central auditory causes) are combined called as retrocochlear pathologies.

Mixed hearing loss: It occurs in situations of combined conductive and sensorineural elements causing deafness.

Non-organic hearing loss (NOHL): This is a kind of hearing loss where there is disparity between the subjective and objective assessment of hearing loss thresholds without any pathology in the auditory system.

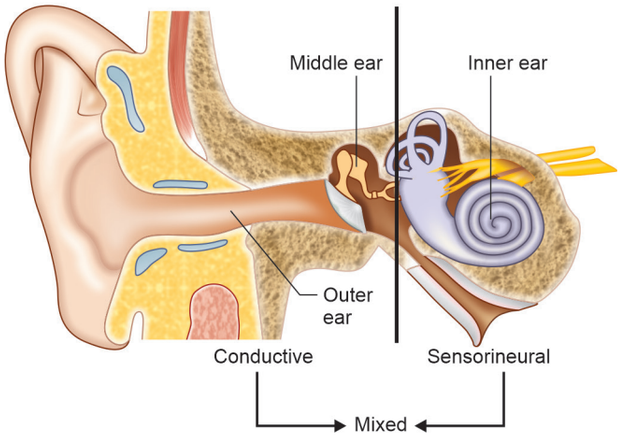


Fig. 3.1: Hearing pathway with type of hearing losses

Conductive Hearing Loss

The conductive loss is due to interference in the progression of sound from the outer ear to the cochlea. Hence, the pathology lies in the external auditory canal, tympanic membrane, ossicles or middle ear cavity. The feature of the conductive loss are:

1. Rinne test is negative (BC>AC).
2. Weber is lateralized to the affected ear.
3. Absolute bone conduction test is normal.
4. Pure tone audiometry will show an air-bone gap (AB gap).
5. Speech discrimination is normal.
6. Conductive loss is less than 60 dB.

The conductive hearing loss is either congenital or acquired (Table 3.1).

Table 3.1: Causes of conductive hearing loss

Congenital	*Acquired*
1. Congenital cholesteatoma 2. Fixation of malleus head 3. Meatal atresia 4. Fixation of footplate of stapes	**A. External ear causes:** Wax, foreign body, otitis externa, benign or malignant tumours, canal atresia **B. Middle ear causes:** • Perforation of tympanic membrane • Fluid in middle ear (SOM and AOM) • Fixation of ossicular chain • Disruption of ossicular chain • Mass in middle ear • Eustachian tube dysfunction

Sensorineural Hearing Loss

Sensorineural hearing loss (SNHL) results from pathologies of cochlea, VIII nerve and central auditory pathways.

The features of sensorineural hearing loss are:

1. Rinne test is positive (AC>BC).
2. Weber is lateralized to the better ear.
3. Absolute bone conduction test shows reduced bone conduction.
4. Air bone gap is not present.
5. Hearing loss may exceed 60 dB.
6. Speech discrimination is poor.

The SNHL may be acquired or congenital.

A. The congenital causes are discussed in Chapter 17: Hearing Impairment in Children

B. Acquired causes:

1. Infections of the labyrinth—viral (measles, mumps), bacterial or spirochaetal
2. Noise induced hearing loss
3. Trauma to the temporal bone
4. Ototoxic drugs
5. Presbycusis
6. Inner ear ischemia
7. Meneire's disease
8. Sudden sensorineural hearing loss
9. Cholesteatoma
10. Acoustic neuroma
11. Familial progressive SNHL
12. Systemic conditions causing SNHL: Diabetes, hypothyroidism, autoimmune disorders (Cogan syndrome, polyarteritis nodosa, relapsing polychondritis), hypothyroidism.

Objectives of Hearing Assessment

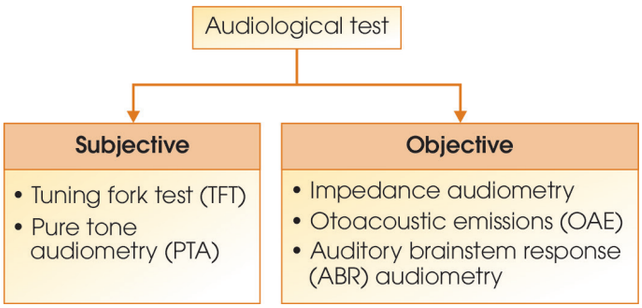
- To determines the type of hearing loss: Conductive, sensorineural or mixed
- To find the degree of hearing loss: Mild, moderate, moderately severe, profound or total
- To find the probable site of lesion for hearing loss
- To establish a baseline for management of the cases requiring treatment and monitoring the treatment.
- To calculate the degree of hearing disability
- For medicolegal purposes.

Tests for Hearing

Various nonstandardized tests are used by the ENT clinicians in their day-to-day practice to assess the hearing of the patients. These informal hearing tests include *Finger friction test* or *Rubbing the paper test*, *Voice test* and *Watch test*. These are used to a good effect for preliminary assessment of the patient but are not reliable.

The audiological tests have been mainly classified as subjective and objective tests (Flowchart 3.1).

Flowchart 3.1: Audiological tests



TUNING FORK TEST

The tuning fork test remains a reliable clinical test for preliminary hearing estimation. The tuning fork test is done by tuning forks of frequencies 256, 512, and 1024 Hz (Fig. 3.2). The tuning fork of frequency less than 256 Hz is not used because it causes stimulation of the vibrotactile pathway and not hearing, while tuning forks of above 1024 Hz, have a short decay time and are not suitable for hearing tests. Ideal tuning fork should have a decay time of more than 60 seconds.

Tuning fork: It is a metallic instrument which produces *pure tones* or single frequency sound. It has two prongs, which are connected at the neck. The neck rests on the stem, which is supported at the base by a foot piece.

How to use: The tuning fork is ideally struck at the junction of upper one third and lower two third of a prong, by a gentle, single blow on the examiners elbow or rubber heel of shoe. Do not strike the tuning fork on hard surfaces because this will produce overtones. If the tuning fork has been struck once, then its vibrations need to be dampened before the next use, to prevent production of overtones.

Tests with Tuning Fork

Before we start to look at the various tuning fork tests, it is necessary to know air conduction and bone conduction tests (Fig. 3.3A)

A. **Air conduction (AC):** After the tuning fork has been struck, it is placed 2 cm vertically, near the external

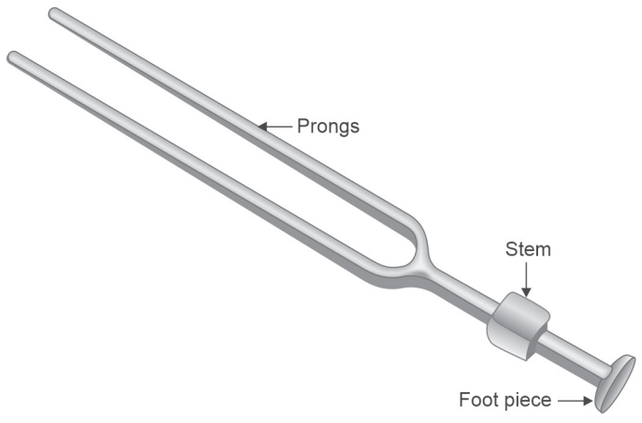


Fig. 3.2: Parts of the tuning fork

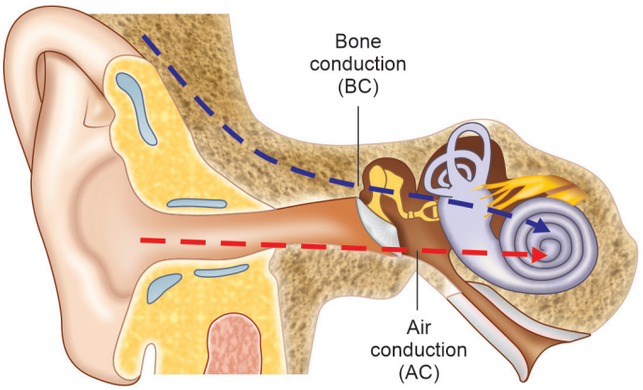


Fig. 3.3A: Air and Bone conduction pathway
Blue arrow–Bone conduction, Red arrow–Air conduction





Fig. 3.3B and C: (B) Air conduction (AC); (C) Bone conduction (BC)

auditory canal, with the prongs of the fork parallel to the EAC. The sound waves travel across the ear hearing pathway of tympanic membrane, ossicles and cochlea. Therefore, both the conductive and sensorineural pathways are simultaneously tested (Fig. 3.3B).

B. **Bone conduction (BC):** Here the tuning fork is vibrated and placed at the tip of the mastoid process with firm contact of the foot piece with the bone. The sound is directly conducted to the cochlea through the skull bones. Therefore, it directly measures the cochlear function (Fig. 3.3C).

Remember: Air conduction is more than bone conduction in normal hearing ear.

1. Rinne Test

The Rinne test is done by two methods:

a. **Loudness comparison method:** Once the tuning fork is struck, the air conduction and bone conduction are done alternatively (two seconds each) and the patient is asked to appreciate the loudness of the two sounds. Normally the air conduction is louder than the bone conduction and is interpreted as Rinne's positive (AC>BC).

b. **Threshold comparisons method:** In this test the tuning fork is struck and bone conduction is done. The patient is asked to tell when the bone conducting sounds are not heard and then the same tuning fork is used to do air conduction. Normally the patient will still hear the air conducting sounds because the air conduction is always more than bone conduction. (AC>BC), and the Rinne test is said to be positive. If the patient is not able to hear during air conduction test (BC>AC) then Rinne is said to be negative.

Quantitative assessment of hearing loss can be done with the Rinne test. All the three frequencies of tuning fork are tested (256, 512, 1024 Hz) (Table 3.2).

False-negative Rinne is seen in the patients with unilateral severe sensorineural hearing loss. This happens while performing bone conduction in patients with severe sensorineural hearing loss. The non-test ear will perceive sound because of the transcranial sound conduction. The false-negative Rinne can be avoided by masking the non-test ear with the **Barany's noise box**.

2. Weber Test

In this test a vibrating tuning fork of 512 Hz is placed firmly over the middle of the forehead or on the vertex

Table 3.2: Quantitative hearing loss assessment with Rinne test

Frequency of tuning fork (Hz)	*Hearing loss (dB)*
Positive with 512 and 1024, Negative with 256	20–30 dB
Positive with 1024, negative with 256 and 512	30–45 dB
Negative with all three frequencies	>45 dB

and the patient is asked about the lateralization of sound, i.e. in which ear it is heard better. In the test the sound travels directly to the cochlea through bone conduction.

1. In case of normal bilateral hearing, the patient hears sound equally in both ears.
2. In case of conductive hearing loss, the sound is lateralized to the worse ear. This is so because the air conduction normally interferes with bone conduction but in conductive losses the air conduction being less, the bone conduction perception increases.
3. In case of sensorineural hearing loss, the sound is lateralized to the better ear.

The Weber test lateralisation occurs when the hearing threshold difference between the two ears is at least 5 dB (Fig. 3.4).

3. Absolute Bone Conduction Test

In this test the cochlear function of the patient is compared with that of the examiner, presuming the examiner has normal hearing. Here the bone conduction is tested with the tuning fork of 512 Hz frequency, placed over the mastoid process firmly with the external auditory canal blocked. The EAC is blocked to prevent the AC affecting the BC. The patient is asked to inform when he stops hearing the sound. Then the same vibrating tuning fork is placed over the mastoid tip of the examiner (Fig. 3.5).

1. The examiner will not be able to hear the sound of tuning fork after the patient has stopped hearing in normal hearing or patient with conductive hearing loss (Interpretation is **same as examiner)**.
2. If the examiner is able to hear after the patient stops hearing, then the patient has sensorineural hearing loss (Interpretation is **reduced ABC)**.

4. Schwabach's Test

This is similar to the absolute bone conduction test but the external auditory canal is not occluded.



Fig. 3.4: Weber test



Fig. 3.5: Absolute bone conduction test

a. Normally both the examiner and patient have **equal** sound perception.
b. In case of sensorineural hearing loss the schwabach test is **reduced**.
c. In case of conductive hearing loss it is **lengthened**.

SPECIAL TUNING FORK TEST

1. Bing Test

In this test the bone conduction is alternatively done (512 Hz tuning fork) with and without external auditory canal occlusion. In case of a normal patient, occlusion of EAC leads to better perception of tuning fork sound via BC, because when EAC is occluded than AC no longer will interfere with BC perception (**Bing test is positive).** In cases of conductive hearing loss occlusion of EAC does not cause any change of tuning fork sound perception (**Bing test is negative)**.

2. Gelle's Test

The air pressure inside the external auditory canal can be increased by using a **Siegel's speculum**. It causes stiffening of the tympanic membrane and ossicular chain. This leads to the reduction of air conduction (AC).

In this test, the bone conduction is done by placing the tuning fork on the mastoid process, alternatively with or without EAC pressure increase. In case of increased pressure by Siegel's speculum, the AC is decreased, leading to better perception of BC in normal patients or sensorineural hearing loss (**Gelle's test positive**).

In cases of ossicular chain fixity or dislocation, no change is perceived (**Gelle's test negative**). Hence, in patients of otosclerosis, the Gelle's test is negative.

PURE TONE AUDIOMETRY

The pure tone audiometry (PTA) does qualitative and quantitative assessment of hearing loss. This is done using an electronic device called the **audiometer**. This



Fig. 3.6: Pure tone audiometry (PTA)

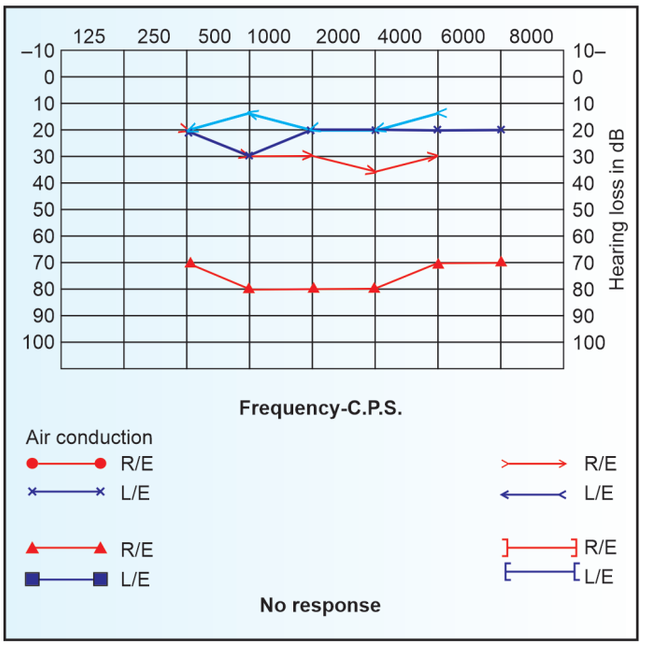


Fig. 3.7A: Audiogram

Legends	Right	Left
Air conduction	○	X
Air conduction with masking	△	□
Bone conduction	<	>
Bone conduction with masking	[	]
No response	↙	↘

Fig. 3.7B: Legends used in audiogram

is done to measure the air conduction from 250 to 8000 Hz using *headphones*, and bone conduction at 500 to 4000 Hz using the *bone vibrator*. Some audiometers include options of high frequency hearing assessment up to 16000–20000 Hz, which is used for monitoring the ototoxic effects of drugs. Patients usually below 5 years of age are not cooperative enough for this test (Fig. 3.6).

The audiometer is calibrated at audiometric zero individually for air and bone conduction. The **audiometric zero** is the minimal sound heard at a particular frequency. According to Indian standard organisation (ISO), the audiometric zero is the mean of the minimal audible intensity in a group of normally hearing healthy adults between 16 and 25 years of age. The levels of audiometric –10 dB to +20 dB is taken as normal. The calibration of the audiometer should be done at least once a year.

The air conduction (AC) and bone conduction (BC) are measured for the above said frequencies and further air-bone gap is calculated (AC–BC) at speech frequencies (500–4000 Hz). The recording to the AC and BC is marked as a graph on paper called the **audiogram** (Fig. 3.7).

Masking

Masking of the non-test is important because it can take part while testing the hearing for the test ear and hence lead to false results. In cases where air conduction is worse than bone conduction by more than 40 dB, masking is essential for testing air conduction. In case of testing bone conduction, masking is always essential.

Interpretation of Audiogram

(Fig. 3.8)

Read the air conduction thresholds above 25 dB, i.e AC>25 dB.

Read the bone conduction thresholds above 20 dB, i.e. BC > 20 dB.

Calculate the airborne gap (AB gap)/**Fletcher index** at 500 Hz, 1000 Hz, and 2000 Hz: Frequencies. AB gap > 25 dB is abnormal.

Uses of Pure Tone Audiometry

1. Determines the type and degree of hearing loss
2. Establishes a baseline for management
3. Degree of handicap calculation for medicolegal cases
4. For prescription of hearing aid.

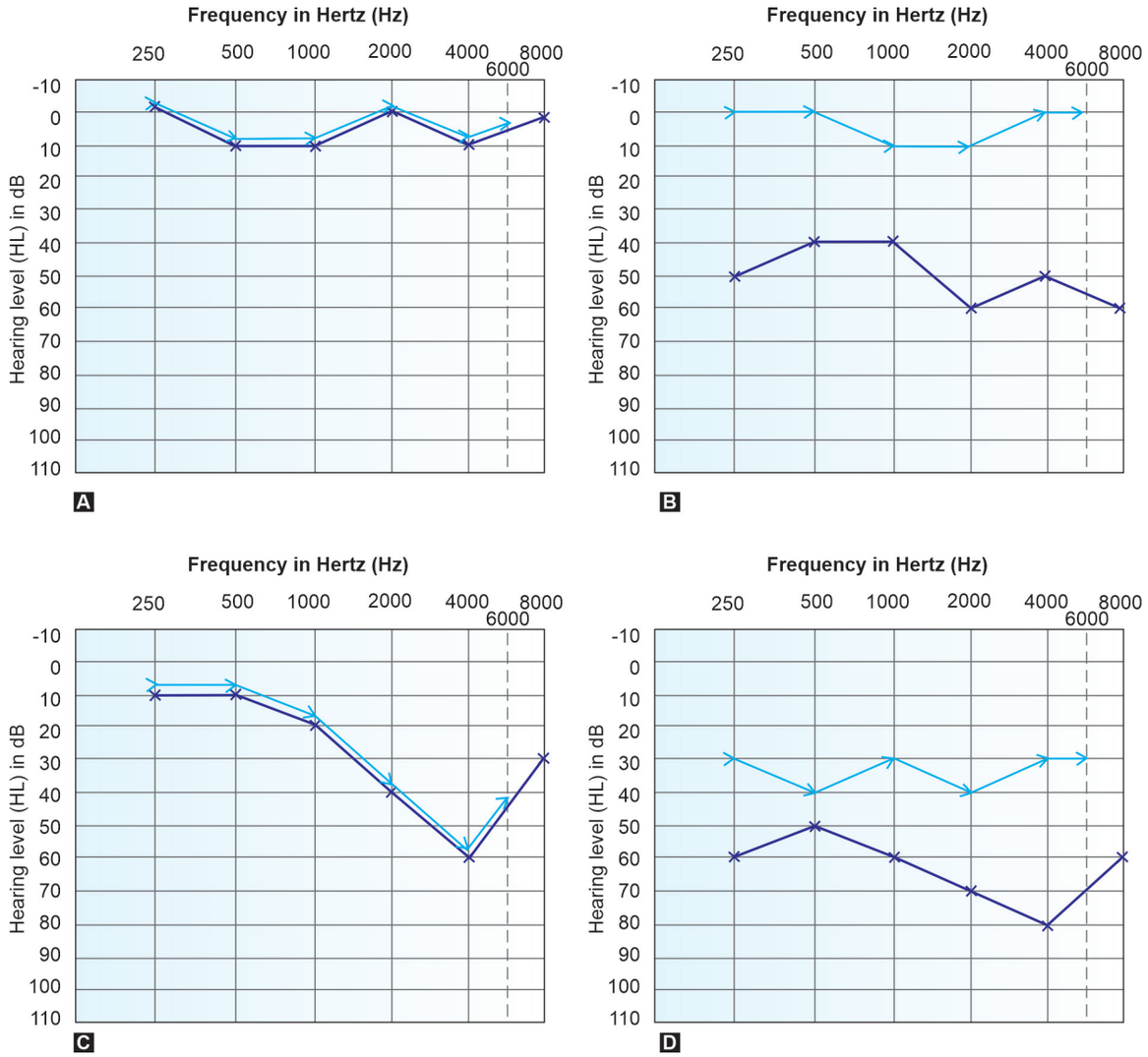


Fig. 3.8: (A) Normal hearing; (B) Conductive hearing loss; (C) Sensorineural hearing loss; (D) Mixed Hearing loss

SPEECH RECEPTION THRESHOLD (SRT)

In speech reception threshold test, a set of spondee words (like "star track", "football") are used instead of pure tones of the audiometer. The speech tests are delivered through a pre-recorded material on a CD or computer played through the audiometer to allow the presentation of the sound control. The threshold is taken as a sound intensity at which 50% of the words are repeated by the patient correctly. In normal patients the speech perception is within 10 dB of the pure tone audiogram. As the speech perception is being measured therefore only 500–2000 Hz frequency is tested.

SPEECH DISCRIMINATION SCORE (SDS)

Here a set of 25 phonetically balanced words are delivered to the patient. A graph is plotted of the correct responses at various frequencies. The normal speech audiogram is a sigmoid curve with the maximum speech discrimination (100%) at 40 dB and half the score is at 25 dB, above the speech reception threshold.

Simplified speech discrimination can be done by delivering words at 40–45 dB above the speech perception threshold (Fig. 3.9). A score above 95–100% is seen in normal and patients with conductive hearing loss. In cochlear pathologies maximum perception is at

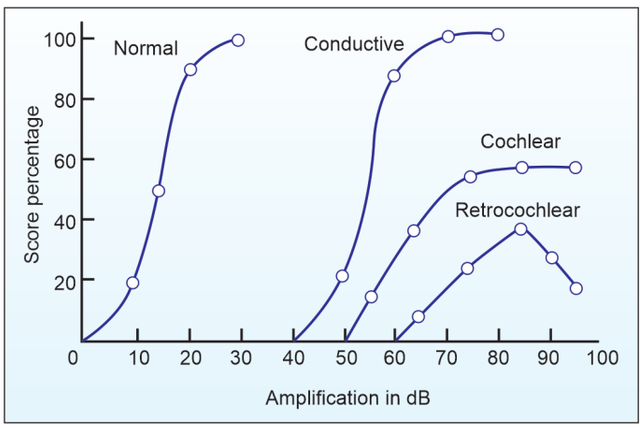


Fig. 3.9: Speech discrimination audiogram

70 dB above SRT but further it plateaus. In retrocochlear pathologies maximum perception is at 80 dB but further it rolls over with further increase of sound intensity (Fig. 3.9).

SPECIAL AUDIOMETRY TEST

1. Short Increment Sensitivity Index (SISI)

In this test a continuous sound 20 dB above the hearing threshold of the patient is presented. This is followed by an in-between short increment of 1 dB in tones for 0.3s, each at an interval of every 5 seconds. These are presented 20 times to the patient. The patient is to tell each such increase in tones and the number of responses are counted.

A normal patient has a score of 20–30%. A score of >60% indicates cochlear lesion (**due to recruitment**), and a score of <15% is seen with retrocochlear lesion.

2. Tone Decay Test

In this test a continuous tone of 5 db above the hearing threshold of the patient is presented at 4000 Hz. A normal subject should hear this sound for continuous 60 seconds. If the sound is not heard for continuous 60s then the tone intensity is increased by 5 db and the test is repeated. This increase in tone intensity is done till the subject hears the sound for continuous 60s or till a maximum of three minutes of the entire test.

A tone decay of >30 dB (the sound being increased to this level for continuous 60 s perception) is suggestive of retrocochlear pathology. The test is an indicator of nerve fatigability. A result of 0–5 dB is normal and 10–25 dB is seen in cochlear pathology.

3. Alternate Binaural Loudness Balance Test (ABLB)–Fowler's Test

Recruitment is a phenomenon of abnormal growth of loudness perception. The loudness perception increases greater than the same sound presented to the normal, so much so that the affected ear which had more difficulty hearing softer sound, will have intolerable loudness at slightly higher intensity. This is seen in cochlear lesions and this makes them poor candidates for hearing aids, as seen in Meniere's disease and sensory presbycusis.

In ABLB, both the ears are tested with prior requisite of one normal ear. A sound of 1000 Hz is presented to both ears at 20 dB over the hearing threshold of each ear. The test is repeated with an increase of 20 dB until the loudness of both ears match or the limit of the audiometer is reached.

In case of normal ear or sensorineural hearing loss, the loudness perception difference between ears is maintained but in cochlear pathologies the loudness perception for both ears becomes same or the difference is decreased due to recruitment (Fig. 3.10).

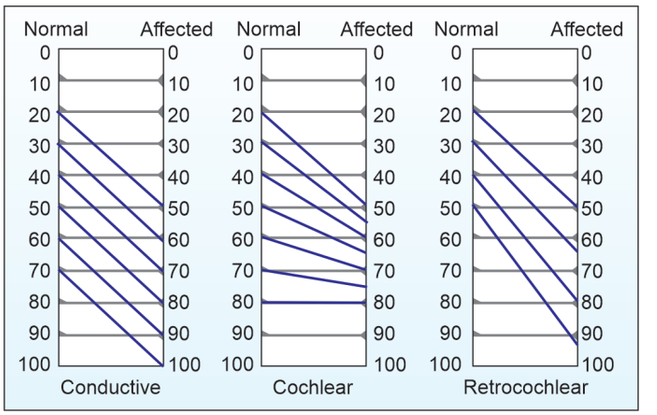


Fig. 3.10: Alternate binaural balance test (ABLB)

OBJECTIVE TESTS

1. Impedance Audiometry (Tympanometry)

The pressure changes in the sealed external auditory canal results in tympanic membrane compliance changes. An increased pressure causes tympanic membrane and ossicular chain stiffness. The maximum compliance or mobility of the tympanic membrane is when the pressure in EAC is same as that of the middle ear. The compliance is the ease with which the acoustic energy can be transmitted through the tympanic membrane and middle ear to the inner ear. It is the same as *admittance*. The opposite of compliance is *impedance* which means the mass and stiffness of the tympanic membrane and middle ear impedes the transfer of acoustic energy from EAC to inner ear via middle ear. When the pressure exceeds or decreases below the middle ear pressure, the compliance decreases and impedance increases. We can measure the compliance by tympanometry.

Tympanometry probe is snugly fitted in the ear canal. It has three channels. The first channel delivers a tone at 220 Hz. The second channel in the probe, picks up the reflected sound. The pressure changes can be made via the third channel in the probe. The compliance of the tympanic membrane is recorded and graph is generated.

In a tympanogram the middle ear pressure is marked on the x axis and the unit used is decapascals (daPa). The "compliance" or the "admittance'' of the tympanic membrane is marked on y axis and is measured in cubic centimeters.

Interpretation

- The maximum compliance is seen at the range of 0.39–1.30 ml in normal individuals between-100 mm H_2O and + mm H_2O (**type A curve**).
- In conditions like serous otitis media or tympanosclerosis or thick tympanic membrane, shows no change of compliance with pressure changes. A flat tympanogram **type B** is seen.
- In Eustachian tube dysfunction, the maximum compliance is below-100 daPa (**type C curve**).

- In case of ossicular discontinuity compliance is greatly increased and the peak may be open at the top. Compliance is greater than 3.5 ml. The graph is **type Ad**. It is seen in ossicular discontinuity or hypermobile tympanic membrane.
- In case of ossicular fixity the tympanic membrane has low compliance as seen in otosclerosis or malleus fixation. The curve is **type As** (Fig. 3.11A and B).



Fig. 3.11A: Impedance audiometry being carried out using a portable impedance audiometer

Acoustic Reflex (Stapedial Reflex)

When a sound of more than 70 dB (70–80 dB) above the hearing threshold is presented to the ear, there is bilateral contraction of the stapedial muscle, which can be picked up by the tympanometry as changes in middle ear compliance.

Reflex Arch

The acoustic reflex occurs via VIII nerve, cochlear nucleus and internuclear brainstem connections to the ipsilateral and contralateral facial nuclei and finally the stapedial nerve (branch of facial nerve).

Uses of stapedial reflex

1. *Detection of non-organic hearing loss* (*NOHL*): It is one of the tests to detect the malingering of hearing loss in patients especially in medicolegal cases or psychiatric disorders.
2. *Detection of VII nerve injury site*: In case the stapedial reflex is absent, this indicates the site of lesion of the facial nerve is proximal to the origin of the nerve to stapedius (second genu of facial nerve).
3. *Detection of VIII nerve lesion*: If a tone of 1000 Hz at 10 dB above threshold is given for more than 10s, then in case of VIII nerve lesion acoustic reflex amplitude is decreased by 50%.
4. *Detection of lesion of brainstem*: If the ipsilateral stapedial reflex is present but not the contralateral, then it might be suggestive of intra-axial brainstem

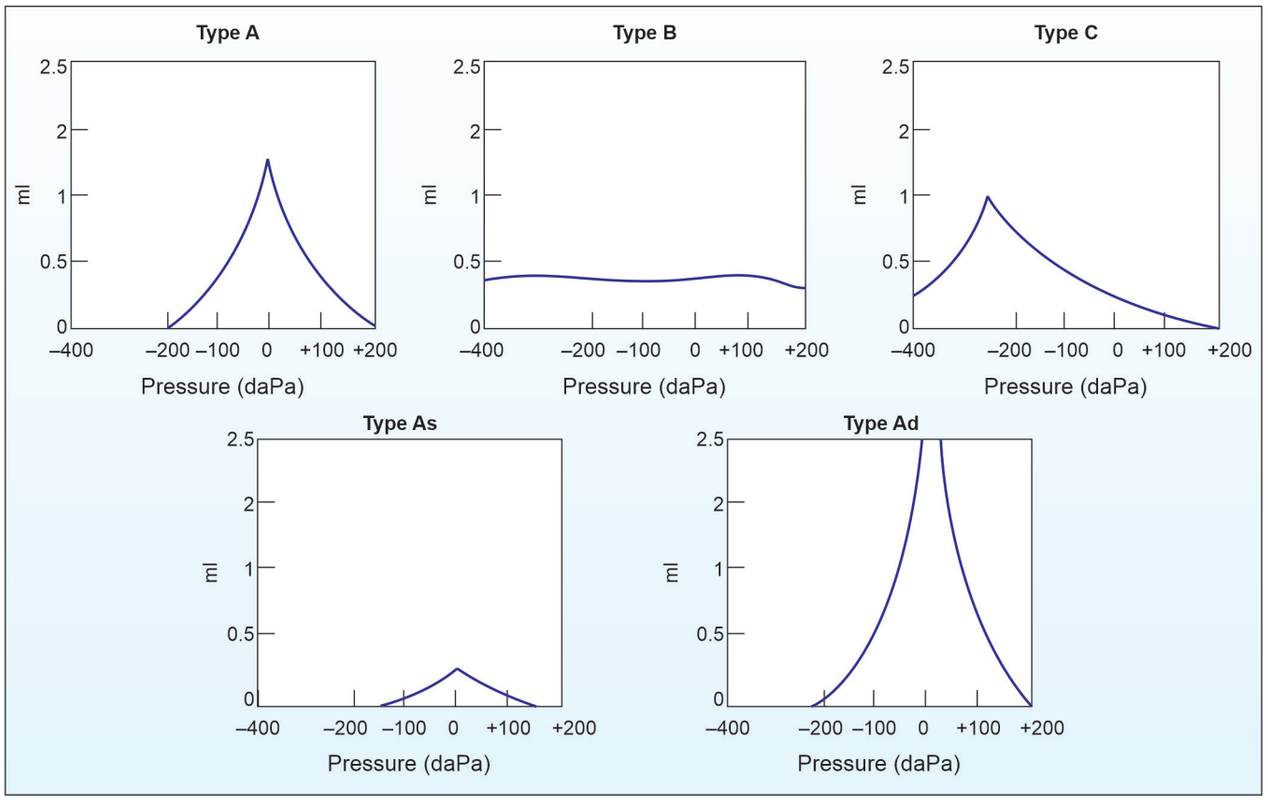


Fig. 3.11B: Various types of tympanograms

lesion. If the ipsilateral and contralateral reflexes are absent on the affected side but ipsilateral being present on the normal side, then ipsilateral VII nerve loss is present.

5. *Detection of cochlear pathology*: If the stapedial reflex is present at values less than 60 dB above the hearing threshold then cochlear pathology causing recruitment may be present.

2. Otoacoustic Emissions

With the discovery of the otoacoustic emissions (OAE) it has become possible to check the functional status of the inner ear. These are audio frequency signals which can be recorded by using sensitive microphones and signal coding. The otoacoustic emissions are thought to be due to the cochlear outer hair cell motility. The acoustic by-products of the outer hair cells are often called **cochlear echoes or Kemp echoes** after the name of the person who had first described it. It is important to note that the OAE are not electrical signals but actually acoustic energy.

The acoustic emissions can be divided into two types: Spontaneous OAE, which are produced without acoustic stimulation and the second type is the Evoked OAE, which are produced in response to the acoustic stimuli. Spontaneous OAE are present in around 50% of the individual.

Evoked OAE can be of two types:

1. *Transient evoked OAE* (*TEOAE*): These are produced in response to click or brief tone burst.
2. *Distortion product OAE* (*DPOAE*): These are produced in response to pure tones separated by a specific frequency difference.

The test uses a miniature microphone placed in the ear canal to produce sound signals, which lead to production of OAE to the sound. Compared to evoked potential tests like ABR, the evoked otoacoustic emission is a simple test where no elaborate patient preparation is required. It is an easy test which can be done bedside and does not take more than a minute hence it is a good hearing screening test for newborns.

Uses of OAE

1. OAE is being used for screening of all newborn babies and high risk babies.
2. It is used to check hearing in a mentally challenged person.
3. It helps to differentiate between retrocochlear and cochlear lesions. The OAE are absent in cochlear lesions, e.g. ototoxic hearing loss.

3. Auditory Brainstem Response

Auditory brainstem response (ABR) is a neurophysiological test of auditory brainstem function in response to auditory (click) stimuli. It is also sometimes called brainstem evoked response audiometry (BERA). The stimulus leads to the production of the waves from the inner ear till the central auditory pathway. It is a set of seven positive waves recorded during the first 10 milliseconds after a click stimuli to the inner ear. They are labelled as I – VII (Table 3.3).

It is important to identify waves properly to read BERA. Firstly the wave V identified, which is the most persistent wave. Waves I, and III are the other important waves (Figs 3.12 and 3.13).

Parameters which are seen

1. Raised threshold for wave formation.
2. Absolute latencies of wave I, III, V peaks and peak V latency in retrocochlear pathology.
3. Inter peak intervals of I–III, III–V and I–V – seen in conditions like hyperbilirubinemia.
4. Absolute latency interaural difference of wave V-prolonged in retrocochlear pathology.

Table 3.3: Origin of BERA waves

Wave	*Origin of waves*
I	Distal part of cochlear nerve
II	Proximal part of cochlear nerve
III	Dorsal and ventral cochlear nuclei
IV	Superior olivary complex and lateral lemniscus
V	Inferior colliculus
VI and VII	Medial geniculate body and lateral lemniscus

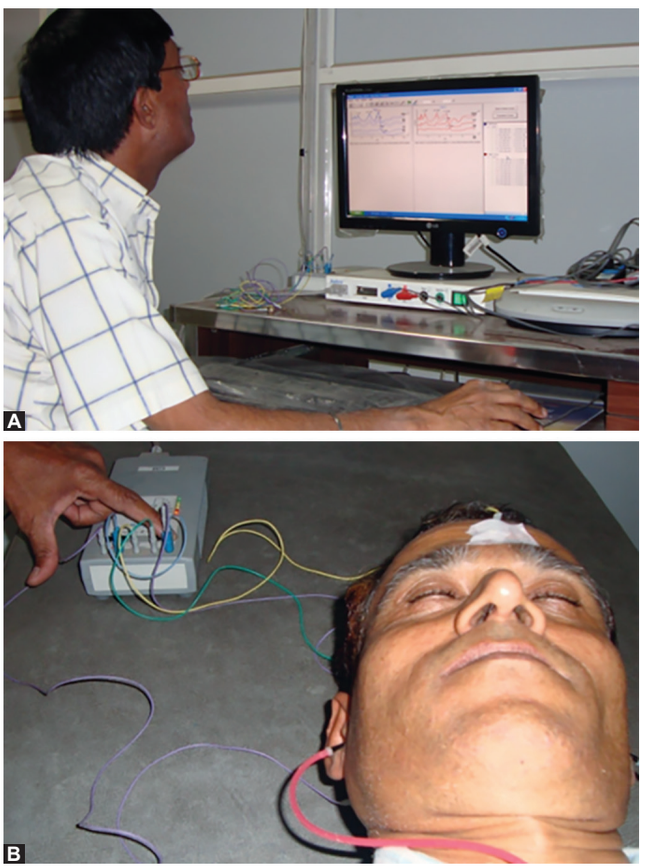


Fig. 3.12: Procedure of BERA

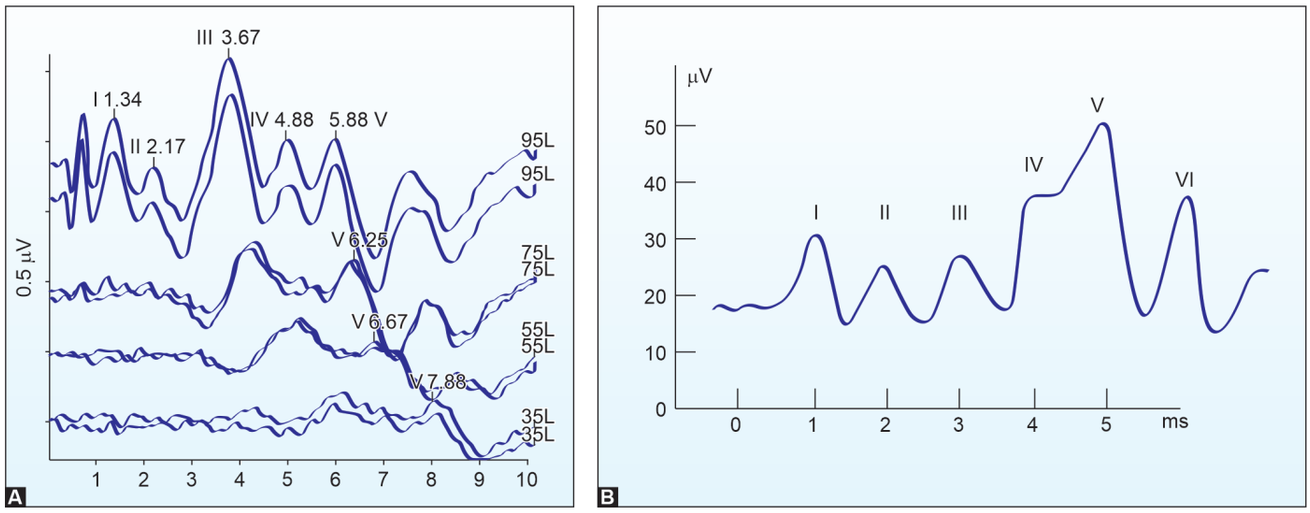


Fig. 3.13: Waves of BERA

Uses of ABR

1. To confirm hearing loss in newborn children who have failed OAE screening.
2. To measure the hearing threshold in a newborn and child.
3. It is an effective screening tool for retrocochlear pathology.
4. Useful in screening of newborns, especially high risk babies.
5. It helps us in diagnosing suspected demyelination disorders.
6. It helps to detect malingering.

Limitations

1. A normal ABR virtually rules out acoustic neuroma but not all intrinsic brainstem lesions and non-acoustic tumors of the cerebellopontine angle.
2. It is not useful to diagnose auditory cortex lesions.

NONORGANIC HEARING LOSS

Nonorganic hearing loss is a decrease in hearing that is unexplained by anatomic or physiologic abnormalities, or both. The term is synonymous with functional hearing loss and pseudohypacusis. It is the intentional production or feigning of hearing loss symptoms or disabilities often motivated by external stress or motives. In psychogenic hearing loss there are associated psychological conditions and the patient is not aware of deafness. Various tests have been designed to check for malingering.

1. **Erhardt's test:** This test helps in the detection of feigned total unilateral hearing loss. Occlusion of the ear will cause sound attenuation of 30 dB. The test is performed with the patient's eyes closed. Sound is given to the proclaimed normal ear. In a normal person even with a covered ear the spoken words are heard with 30 dB dampening. If the patient denies hearing spoken words, then malingering is positive.
2. **Lombard's test:** It is based on the principle that a subject would raise voice when speaking in a noisy environment. While performing this test, the patient is made to read. Noise is introduced in the room. A patient with normal hearing will gradually increase or raise the voice or will stop reading.
3. **Chimani Mooss test:** It is a modification of Weber's test. Normally in Weber's test the patient hears the best in the occluded ear. In malingering the patient will not accept hearing better with an occluded ear.
4. **Stenger's test:** Two tuning forks of 512 Hz are used to check for malingering. Both are struck and placed 25 cm from each ear. Malingering patients will say that he is hearing only from the normal ear. Then the tuning fork of the malingering ear is brought close to that ear. The increased intensity perceived in the malingering ear will cause the loss of perception in the normal ear. In normal circumstances the bringing near of the tuning fork to the deaf ear should not cause any decreased perception in the normal ear.
5. **Stapedial reflex:** In normal individuals the stapedial reflex is elicited at 70–100 dB. If this reflex can be elicited and the patient says he is totally deaf, then it is suggestive of malingering.
6. **Variable response to PTA:** The malingerer's responses on PTA are non-uniform and highly variable. Also, the results of PTA and speech audiometry are not comparable.
7. **BERA:** This is very useful in NOHL as it is an objective test and can correctly estimate the hearing thresholds.

Chapter

4



Vestibular System

The vestibular system helps to maintain balance as it provides information to the CNS about movement and position of the head and body in space. The peripheral vestibular system has five receptor organs in the inner ear. These are the utricle, saccule, and three semicircular canals.

PERIPHERAL VESTIBULAR RECEPTORS

There are two types of peripheral receptors (sensory epithelium), namely maculae and cristae. These are present inside the membranous labyrinth. The sensory cells are surrounded by the supporting cells.

Maculae

These sensory receptors are located in the utricle and the saccule. There are about 45,000–60,000 sensory hair cells in the two maculae of each ear. The macula of the utricle is located on the floor and is comma shaped. The macula of the saccule is elliptical in shape and is located on its anterior wall in the vertical plane. Maculae are responsive to linear acceleration and to the gravity gradient (head tilt).

Cristae

These are located in the ampullated ends of the three semicircular ducts at right angles to the longitudinal axis of the canal. There are about 23,000 sensory cells in the three cristae of each ear. These are responsive to the angular acceleration in various planes.

Structure of Receptors Cells

The vestibular hair cells (macula or crista) have two types of hair cells. The type I cells are flask shaped and type II is cylindrical (Table 4.1). They consist of sensory epithelial cells which have 50–100 cilia. These cilia are called **stereocilia**. One of these cilia is large, which is called the **kinocilium**. The glycocalyx and extracellular filaments connect the kinocilium with the other stereocilia (Fig. 4.1).

Table 4.1. Differences between the hair cells of vestibular sensory cells

Type I hair cells	*Type II hair cells*
1. Flask-shaped body	1. Cylindrical-shaped body
2. Has a calyx of single afferent nerve	2. Multiple bouton type afferent nerve endings

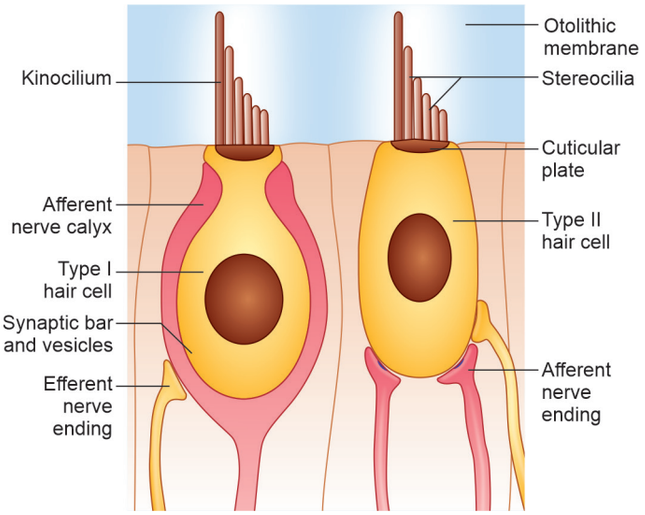


Fig. 4.1: Receptor hair cells: type I and II

STRUCTURE OF OTOLITHIC ORGANS OF UTRICLE AND SACCULE

The receptor hair cells (macula) of the utricle and saccule are covered by a gelatinous layer called the *otolithic membrane*. The cilia of the hair cells project into this gelatinous layer (Fig. 4.2).

On top of the gelatinous layer is the *otoconial layer* containing small calcium carbonate crystals. The otoconia show a very slow turnover rate. They are produced by the supporting cells of sensory epithelium. The bases and sides of the hair cells make synapses with the neurons of the vestibular nerve. The movement of the head leads to distortion of the stereocilia and kinocilia which bring about the change in rate of generation of the neural action potentials.

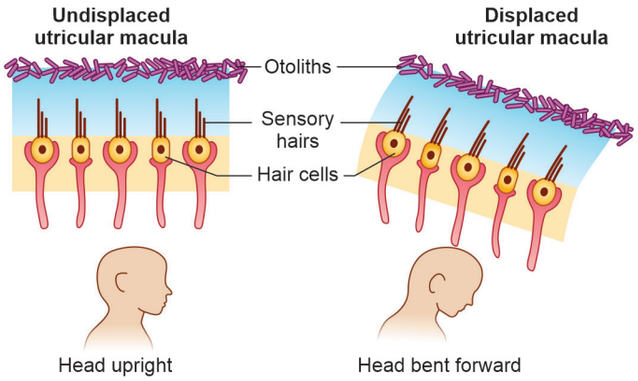


Fig. 4.2: Structure of otolithic organs

STRUCTURE OF AMPULLA OF SEMICIRCULAR CANALS

The sensory cells of crista are surrounded on top by a bulbous, gelatinous mass called the **cupula**. The cupula has the same specific gravity as the endolymph but because of a different constitution it is not soluble with it. The cilia of the sensory cells of the cristae project into the cupula (Fig. 4.3). The movement of the endolymph distorts the cupula which in turn deviates the cilia, especially the kinocilium. This generates an action potential in the nerve ending by opening up of the K^+ ion channels.

PHYSIOLOGY OF BALANCE

Semicircular Canals

Angular acceleration and rotation of the head in various planes are sensed by the three semicircular ducts that are oriented at right angles to one another. Rotation of the head leads to flow of the endolymph, which leads to deflection of stereocilia and kinocilium of cristae. This displaces the cupula and generates the neural impulses. According to the *Edward's law*:

1. Head and eye movements always take place in the plane of the canal being stimulated.
2. In the horizontal canal, ampullopetal endolymph flow causes greater response than ampullofugal flow.
3. In vertical canals ampullofugal endolymph flow causes a greater response than ampullopetal flow.

The quick component of the nystagmus is always opposite to the direction of the flow of endolymph.

The angular acceleration occurs in three planes. The **yaw** head movement is in the horizontal motion about the vertical plane, **pitch** is the flexion and extension motion about a vertical plane and lastly the **roll** movement due to lateral head tilt about a horizontal axis (Fig. 4.4).

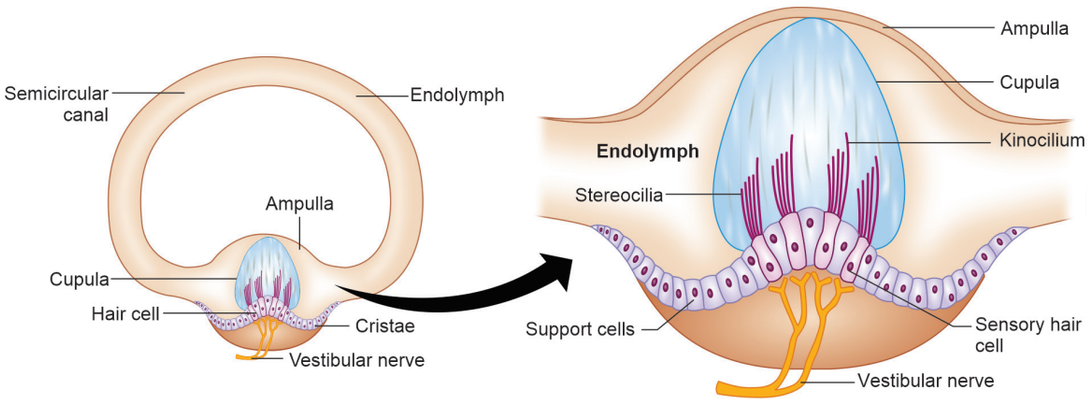
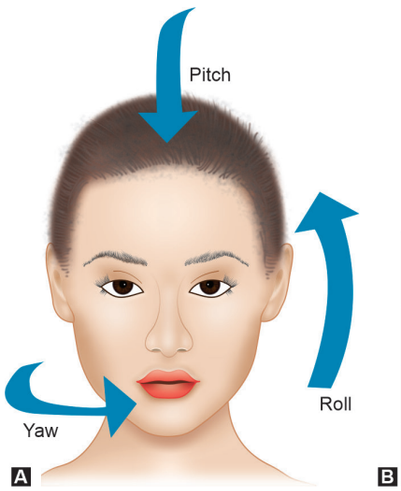


Fig. 4.3: Structure of ampulla



B

Head movement	Semicircular canal stimulated
Yaw	Lateral
Pitch	Posterior + Superior
Roll	Superior + Posterior

Fig. 4.4: Three planes of head movement producing angular acceleration and semicircular canals stimulation

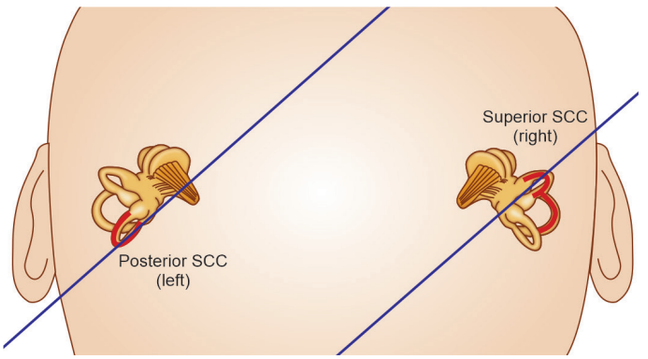


Fig. 4.5: Physiological pair of semicircular canals (SSC)

The six semicircular canals work in pairs, i.e. a pair of both horizontal canals work together, as do the posterior canal of one side with the superior canal of the other. This is so because these pairs are in the same plane (Fig. 4.5).

Utricle and Saccule

Displacements and linear accelerations of the head, such as those induced by tilting or translational movements are detected by the two otolith organs, the saccule and utricle. Macula of the utricle is responsive to horizontal linear acceleration and that of the saccule is responsive to vertical linear acceleration.

On stimulation of the hair cells the nerve impulses are generated. At rest also, there is a resting firing tone of nerve stimulation. This tone increases on head extension and decreases on head flexion. Similarly, on the side where linear acceleration occurs the tone decreases and tone increases on the other side. These messages are interpreted by the brain to decipher the body movements (Fig. 4.6).

CENTRAL VESTIBULAR SYSTEM

1. A plexus of unmyelinated nerve fibres is formed at the base of the sensory epithelium. From here arise myelinated fibres **(Primary vestibular neurons)** which pass to the bipolar cells in the **Scarpa's ganglion**.
2. The ascending afferent fibres of the neurons synapse with vestibular nuclei in the cerebellum **(Secondary vestibular neurons)**. The four vestibular nuclei which receive the afferent sensory inputs are superior, lateral (L), medial (M), and descending (D) nuclei (Fig. 4.7).
3. The central vestibular system allows for processing and interpretation of afferent signals and the output of efferent signals and production of vestibular responses. Three types of vestibular responses/ reflexes which are generated include:

Vestibular Responses

a. *Vestibulo-ocular reflexes*: These are generated by impulses from cristae and macula but the responses

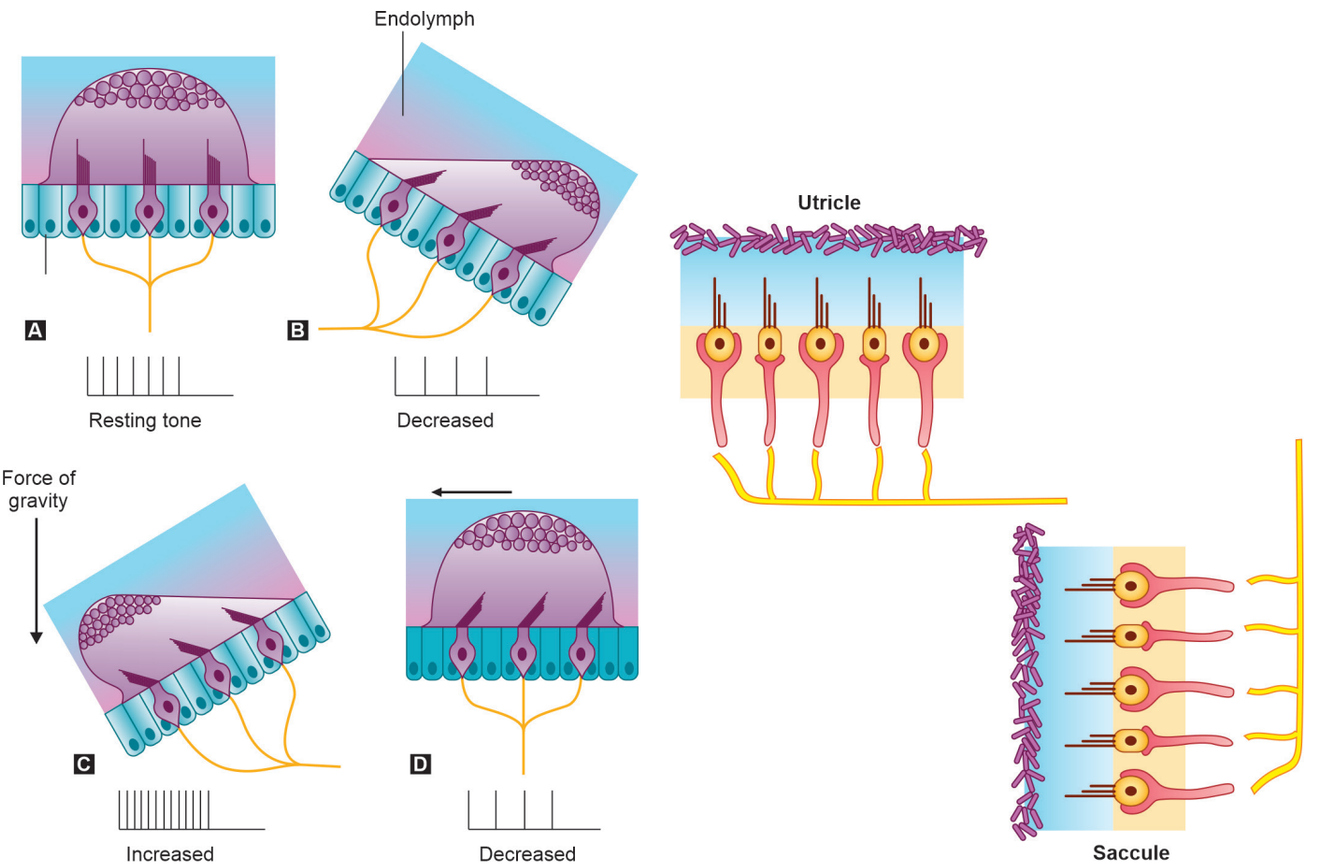


Fig. 4.6: Nerve potential generation due to head positions and linear acceleration

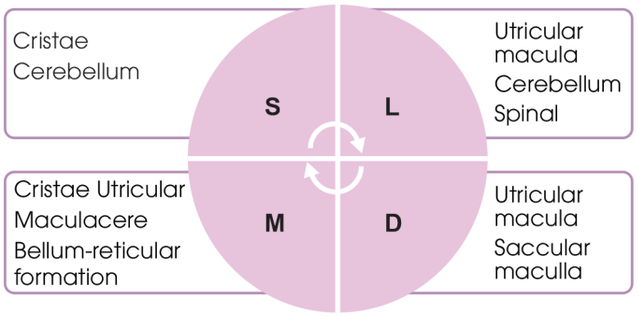


Fig. 4.7: Afferent supply to the four vestibular nuclei

of semicircular canals are predominant. In production of contradictory signals from the maculae and semicircular canals, the responses of later predominate.

b. *Vestibulospinal reflexes*: The vestibular responses affect the spinal anterior horn cells by three efferent pathways, i.e. the lateral and medial vestibulospinal tract and reticulospinal tract. The vestibular apparatus exerts the influence on the control of posture by the way of myotatic reflex which controls the tone of trunk and extremity skeletal muscles.

c. *Vestibulocollic reflexes*: Its role is uncertain.

The further pathway is a complex and interrelated system, which produces the desired body adjustments for balance (Fig. 4.8).

Mechanism for Maintenance of Balance

Balance is achieved and maintained by a coordinated control system that includes sensory input from vision, proprioception (touch input from muscles and joints), and the vestibular system (motion, equilibrium, spatial orientation). This sensory input is integrated in the central nervous system and motor output to the body muscles and eyes is sent for correction of the body posture and head orientation to maintain body balance (Fig. 4.9).

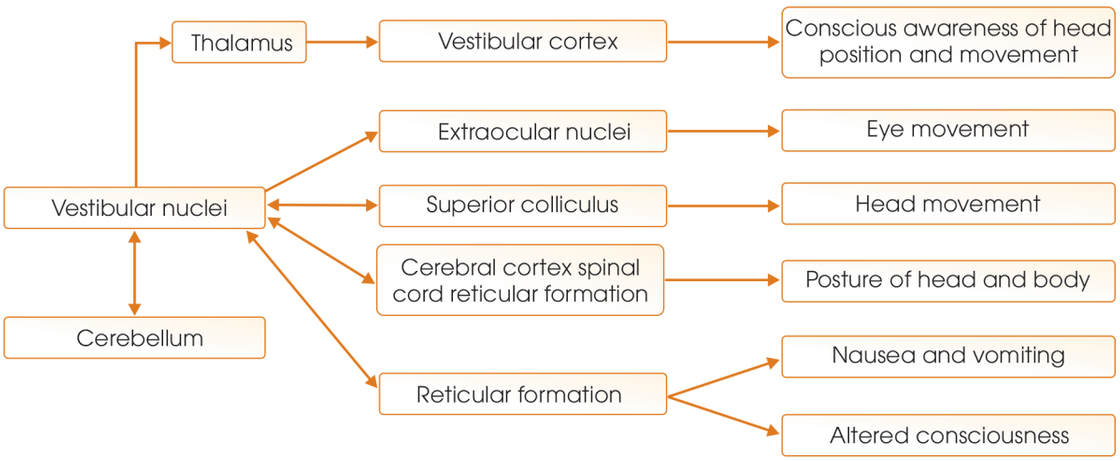


Fig. 4.8: Central vestibular and balance pathways

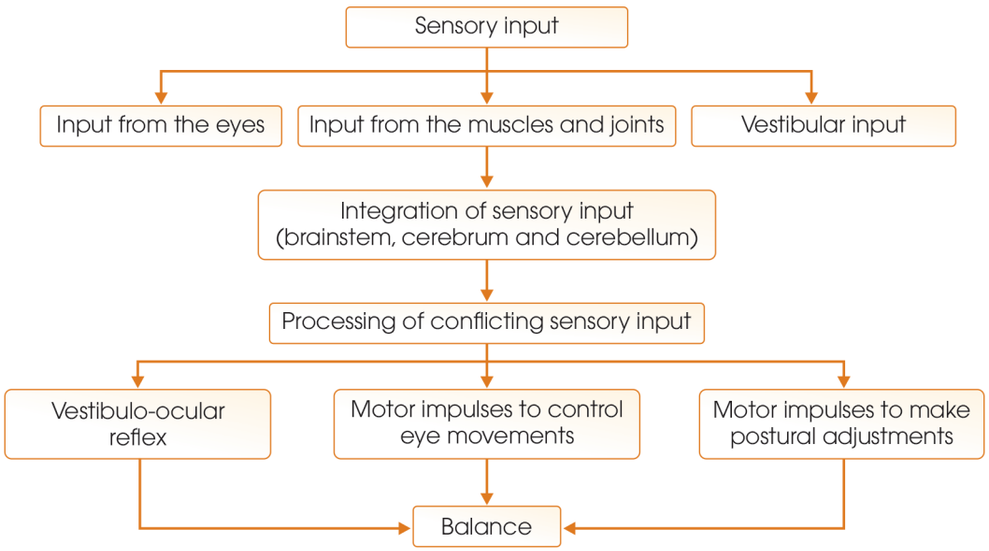
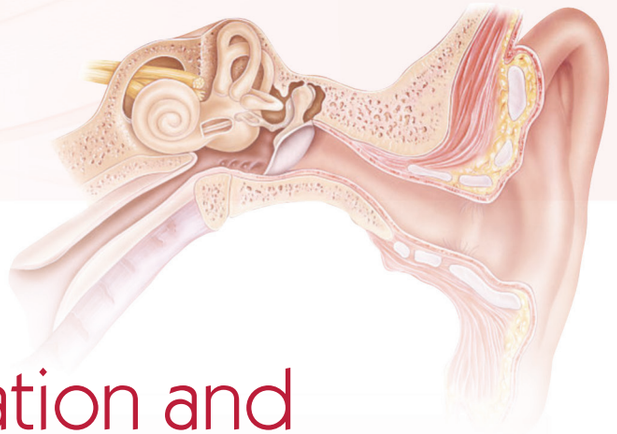


Fig. 4.9: Maintenance of balance

Chapter

5



Vertigo: Evaluation and Common Disorders

EN4.19: Describe the clinical features, investigations and principles of management of vertigo.

The clinical evaluation of the patient with vertigo includes thorough history and neuro-otological examination of the patient followed by laboratory tests when required. The clinical examination includes the nystagmus examination, fistula test, gait, Dix–Hallpike manoeuvre, Head impulse test (HIT) and cerebellar function test. Imaging has a limited role in the diagnosis except to rule out space-occupying lesions in the brain.

1. Nystagmus

Nystagmus is involuntary, rhythmical, oscillatory movement of the eyes. These involuntary eye movements can occur from side to side (horizontal), up and down (vertical), or in a circular pattern (rotatory). It is a combination of alternating slow and fast saccadic components in opposite directions. *Clinically, the direction of the fast component is taken as the direction of nystagmus.*

The nystagmus can be pathological or physiological. The *physiological nystagmus* is elicited by extremes of eye deviation (>30° laterally from the primary position) and is also called **gaze nystagmus**. It can also be induced by rotation, caloric and visual stimulation. The *pathological nystagmus* can be congenital or acquired and is due to some pathology in the vestibular system or organs responsible for maintaining balance. Vestibular nystagmus, the most common type of nystagmus, is caused by dysfunction of the vestibular part of the inner ear, the vestibular nerve, the vestibular nucleus within the brainstem, or parts of the cerebellum that transmit signals to the vestibular nucleus.

The vestibular nystagmus can be of peripheral or central type (Table 5.1).

A. Peripheral nystagmus: Peripheral vertigo may occur as a result of problems in the peripheral vestibular system from the inner ear to the vestibular division of the VIII cranial nerve. Peripheral vertigo accounts for over 90% of all causes of vertigo. The nystagmus of peripheral origin can be suppressed by visual fixation and enhanced in dark or by use of Frenzel glasses, which abolish visual fixation. It can be of two types:

a. Irritative nystagmus (fast component to the side of lesion)
b. Paretic nystagmus (fast component to the opposite of lesion).

Table 5.1: Differences between peripheral and central nystagmus

Features	*Peripheral*	*Central*
Latency	2–20 seconds	No latency
Duration	Less than 1 min	>1 min
Directions	Fixed direction	Direction changing
Fatigability	Fatigable	Non-fatigable
Other symptoms	Severe vertigo	Usually absent or mild

The **gradation** of peripheral nystagmus (also few central nystagmus) is by presence of nystagmus when seen in different directions. This follows **Alexander's law** which states that the spontaneous peripheral nystagmus is maximum when the position of gaze is in the direction of the fast component and least in the direction of slow component. Therefore the:

1. First degree nystagmus—is the presence of nystagmus when a patient looks in the direction of the fast component.
2. Second degree nystagmus—is the presence of nystagmus in primary position or looking straight ahead.
3. Third degree nystagmus—is the presence of nystagmus when the patient looks in the direction of the slow component.

B. Central nystagmus most commonly occurs as a result of ischemia of the central vestibular structures in the cerebellum, brainstem, or vestibular nuclei especially in the elderly with vascular risk factors. Central nystagmus cannot be suppressed by visual fixation.

Various Types of Central Nystagmus

1. *Downbeat nystagmus* is seen in association with Arnold–Chiari malformations and cerebellar disease.
2. *Rebound nystagmus* is seen in cerebellar dysfunction.

3. *Upbeat nystagmus* is seen with intra-axial lesions of the brainstem, tegmentum of pontomedullary junction.
4. *Pendular nystagmus* is seen in multiple sclerosis lesions.
5. *Sea-saw nystagmus* in midbrain lesions.
6. *Purely torsional* in syringomyelia.

Clinical examination for nystagmus: The patient is seated in front of the examiner or is made to lie down supine. The examiner places his finger 30 cm away and in the midline. Patient is then made to follow the finger (without head movement), which is moved side to side gently to the maximum of 30 degrees from the centre to avoid gaze nystagmus.

2. Fistula Test

This test stimulates the labyrinth by producing pressure changes in the external auditory canal. The pressure changes can be produced by means of either intermittent application of the pressure over the tragus or by using Siegel's speculum. The response appears after a latency of 2–3 seconds. In case there is nystagmus and vertigo, it is termed as *fistula test positive*. It is also a sign that the tested labyrinth is functional. Absence of nystagmus, is *fistula test negative*.

It is classically seen in labyrinthine fistula, where there is erosion of bony covering of the semicircular canal (commonly horizontal semicircular canal) by disease like cholesteatoma. It can occur following a surgical procedure (as a complication of mastoidectomy or fenestration operation). Such a fistula can be stimulated by the pressure changes, which produces nystagmus and vertigo (Table 5.2).

The fistula test can be *false-negative*, i.e. fistula is present but the fistula test is negative. This is seen in cases where cholesteatoma covers the fistula. The *false-positive fistula test* **(Hennebert's sign)** may be seen with congenital syphilis and Meniere's disease. The cause behind the false positive test in congenital syphilis is the vestibulofibrosis between the membranous labyrinth and stapes footplate or loosening of the stapedial ligament (which leads to abnormal stapes mobility). The same mechanism of adhesions between stapes footplate and labyrinth causes false-positive results in Meniere's disease.

> ***Note:*** Hennebert's sign is also seen in perilymph fistula.

3. Dix–Hallpike Manoeuvre (Positional Nystagmus)

In this test the patient is critically positioned so as to induce vertigo and nystagmus. It is a very valuable test for diagnosing benign paroxysmal positional vertigo (BPPV). The patient is seated comfortably on the examining table. The examiner holds the head and rotates it by 45 degrees to either right or left. The patient is then made to quickly lie supine with his head relaxed in the examiner's hands, hanging 30 degrees below the horizontal level at the edge of the table. Now the eyes of the patient are observed for nystagmus. The nystagmus is observed for latency, duration, direction and fatigability. The test is repeated for the other side.

Table 5.2: Fistula test-interpretation

Negative fistula test	*Normal*
Positive fistula test	1. Erosion of lateral semicircular canals
	2. Post-stapedectomy fistula
	3. Rupture of round window
	4. Fenestration operation
False negative fistula test	1. Dead labyrinth
	2. Cholesteatoma covering the fistula
False positive fistula test	1. Congenital syphilis
	2. Meniere's disease

In BPPV, there is a latency of 2–20s followed by the development of rotational nystagmus towards the under-most ear, along with vertigo. The vertigo and nystagmus both resolve after 50–60 seconds. On sitting up the vertigo reappears and finally resolves. On repetition of this test, nystagmus is elicited only for a shorter duration (Fig. 5.1).

In central lesions the nystagmus is produced immediately after the patient is put in the critical position without any fatigability.

4. Head Impulse Test (HIT)

It is a sensitive and specific test which detects the hypofunction of the peripheral vestibular system. This method tests the vestibular ocular reflex in the patient. The eyes of the patient have to be fixed on a target, usually the examiner's nose. The head is turned rapidly from the midline positions by 10–15 degrees briskly by the examiner with the velocity of several hundred degrees per second in either direction, while watching the eye for corrective movements (saccades). On the normal side, the functional vestibular system identifies all movements or changes in head position and rapidly corrects eye movement accordingly so that the vision remains on the target. When the head is turned towards

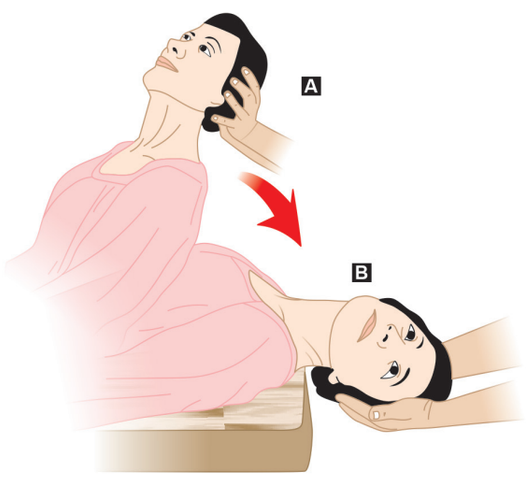


Fig. 5.1: (A) 45-degree side head rotation; (B) 30-degree head down position

the affected side, the vestibular ocular reflex fails to quickly correct the eye position on the target and it makes a *corrective saccade* to re-fixate on the visual target.

A corrective saccade indicates a *positive HIT test*. The positive test indicates peripheral vestibular lesion as opposed to brainstem disease (Fig. 5.2).

> ***Note:*** This test should be used carefully in patients with neck pathology since it involves rapid repositioning of the head.

5. Cerebellar Function Test

The cerebellum coordinates regulation of balance, muscle tone, and voluntary movements. Therefore, the clinical signs which should be looked for in a patient with suspected cerebellar disease are:

a. Finger to nose test and finger to finger test (*Asynergia* and *dysmetria*)
b. Rapid alternating movements (*Dysdiadochokinesia*)
c. Rebound phenomenon
d. Heel to shin test
e. Hypotonia—Pendular knee jerk
f. Gait change—Wide based staggering gait
g. Scanning speech
f. Nystagmus towards the affected side.

6. Romberg Test

The patient is asked to remove his shoes and stand with his two feet together. The arms are held next to the body. The clinician first asks the patient to first stand quietly with eyes open, and subsequently with eyes closed. The examiner watches for any instability and sway as the patient tries to maintain his balance. In patients with peripheral vestibular lesions, the patient sways to the side of the lesion. In central vestibular lesions, the patient has instability.

In the *sharpened Romberg test*, the patient stands with one heel in front of the toes of the other foot. Any inability to maintain balance is suggestive of vestibular impairment.

7. Unterburger Test

The patient is asked to close his eyes and is made to walk on the spot. The patient will turn towards the hypoactive side in case of peripheral lesions, and would be unstable in a case of central lesions.

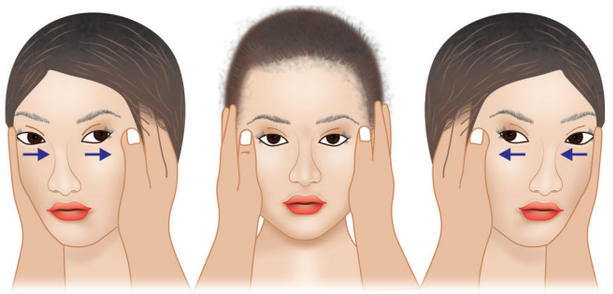


Fig. 5.2: Head impulse test (HIT)

A. LABORATORY TEST FOR VESTIBULAR FUNCTION

1. Caloric Test

Caloric testing is a useful clinical tool that can assess and quantify the functional status of the individual vestibular systems. The test utilizes the mechanics of vestibular–ocular reflex by using thermal gradient across the horizontal canals of both the labyrinths. It is contraindicated in cases of active external or middle ear disease.

Position: The patient is made to lie in the supine position with the head tilted 30 degrees upwards, so that the horizontal canal is placed into the vertical plane. This is the position of maximum sensitivity to the thermal gradient. The test can also be done with sitting position and head tilted backwards.

Response: Hot irrigation causes nystagmus towards the irrigated ear, while cold irrigation causes the nystagmus towards the opposite ear. *The mnemonic for this is 'COWS'—cold opposite and warm same*. The hot irrigation causes an ampullopetal flow of endolymph and nystagmus directed toward the stimulated ear and cold causes ampullofugal flow of endolymph with nystagmus directed toward the opposite ear.

Modified Kobrak test: In this test the ear is irrigated with ice cold water for a maximum of 60 seconds. Initially irrigation is done for 5 sec and then 10 sec with 20 ml and 40 ml cold water. In normal patients the nystagmus is elicited with 5 ml of ice water irrigation. If increased quantities of water are required to elicit response, it is suggestive of hypoactive labyrinth. If no response is seen with water irrigation up to 40 ml, it indicates a dead labyrinth.

Fitzgerald–Hallpike test (Bithermal caloric test): In this test both the ears are alternatively tested by using, both the warm water (44°C) and cold water (30°C). The irrigation is done for 40 seconds. The eyes are observed for nystagmus.

The order of irrigation is: Right ear-warm → left ear warm → right ear cold → left cold.

The interval between two stimulations should be at least 5 minutes. If no response is seen with bithermal stimulation then cold-water stimulation is done before the labyrinth is labelled dead. Alternatively, *warm-air caloric testing* can be done which has a sensitivity of around 87%.

Two distinct patterns are seen:

$$\textbf{Canal paresis (\%)} = \frac{(L30° + L44°) - (R\ 30° + R\ 44°)}{(L30° + L44° + R\ 30° + R\ 44°)} \times 100$$

$$\textbf{Directional preponderance} = \frac{(L\ 30° + R\ 44°) - (R\ 30° + L\ 44°)}{(L\ 30° + L\ 44° + R\ 30° + R\ 44°)} \times 100$$

Conditions:

a. If the duration of nystagmus is seen 30% more than the opposite side, it indicates same side central vestibular nystagmus and opposite side peripheral nystagmus.

b. If the same side canal paresis is seen with same sided directional preponderance, it points towards acoustic neuroma.
c. If the same side canal paresis is seen with opposite directional preponderance, it points towards Meniere's disease.

Cold–air caloric test: It is done in case of perforation of the tympanic membrane because the irrigation of the ear is contraindicated. **Dundas–Grant tube**, which is a coiled copper tube wrapped in a cloth. The ethyl chloride is poured on the apparatus and the cooled air is made to flow into the ear tested (Fig. 5.3).

2. Rotational Test

In the rotational test, the patient is seated in a **Barany's revolving chair** with the head tilted 30 degrees forward. It is rotated at the speed of 10 turns per 30 seconds. It is then stopped to look for nystagmus. Rotation test has advantages over other vestibular function tests in patients with bilateral caloric weakness, in young children, or in patients where VOR (visual–ocular reflexes) changes have to be assessed over time. The disadvantage of the test is that both the ears are tested simultaneously.

Limitations of caloric test and rotational test

1. Both cannot distinguish between pathology of vestibule and VIII nerve.
2. The utricle and saccule cannot be tested by caloric test.
3. It is difficult to detect the presence of pathology in the vertical canals.

3. Galvanic Testing

It is performed by applying small electrical currents to the vestibular labyrinth using electrodes while recording either eye movements or postural sway.

This test helps to differentiate between labyrinthine and retrolabyrinthine pathology. The test is done by placing saline pads either to the tragus, external auditory canal or over the mastoid and the other over the sternum. A current of 1–2 mA is used, sway and nystagmus is seen. The cessation of current produces an opposite response. The thresholds are raised in vestibular neuritis and Meniere's disease.

4. Posturography

This is used for quantitative assessment of the Romberg test. It exposes the standing patient to 6 sensory conditions of increasing difficulty. The purpose of testing is to isolate each of the three principal balance senses (visual, vestibular and somatosensory) and determine function of each and their individual performances. Although it does not provide aetiological information, it does provide a functional measure helping to predict the outcomes of therapeutic intervention with physical therapy.

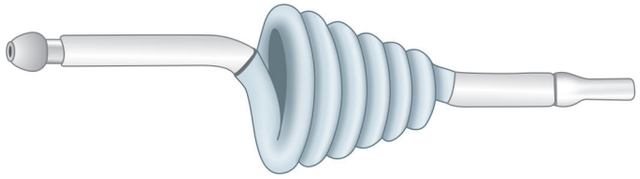


Fig. 5.3: Dunda Grant tube

DISORDERS OF VESTIBULAR SYSTEM

The disorders of the vestibular system are broadly divided into peripheral and central disorders. The peripheral disorders are due to lesions of vestibular end organs and the vestibular nerve. The central cause is due to the pathologies of the brainstem and central vestibular pathways (Table 5.3).

Benign Paroxysmal Positional Vertigo (BPPV)

This cause for peripheral vertigo was first described by Barany in 1921 and Dix and Hallpike in 1952. Otoconia are calcium carbonate crystals embedded in the otolithic organs of the utricle and saccule. In BPPV, degenerated otoconia from the utricle are thought to collect in the semicircular canals, making them abnormally gravity sensitive. There are two theories for BPPV. In **cupulolithiasis theory**, degenerated otoconia sticks to the cupula of ampulla and makes it gravity sensitive. In **canalolithiasis theory**, the degenerated otoconia debris floats in the endolymph of the semicircular canal. On exposure to change in gravity vector, the otoconia falls to the lowest part of the canal, causing a change in endolymph pressure with resultant displacement of cupula. This leads to the symptoms of BPPV (Fig. 5.4).

Causes

1. Most commonly BPPV is idiopathic
2. Head trauma can cause BPPV.

Table 5.3: Causes of vestibular disorders

Peripheral causes	*Central causes*
1. Benign paroxysmal positional vertigo	1. Vertebrobasilar insufficiency
2. Meniere's disease	2. Posterior inferior cerebellar artery syndrome
3. Vestibular neuritis	3. Basilar migraine
4. Superior semicircular canal dehiscence	4. Cerebellar disease
5. Labyrinthitis	5. Cervical vertigo
6. Vestibular migraine	6. Multiple sclerosis
7. Vestibulotoxic drugs	7. Epilepsy
8. Head injury	
9. Perilymph fistula	
10. Acoustic neuroma	
11. Syphilis	

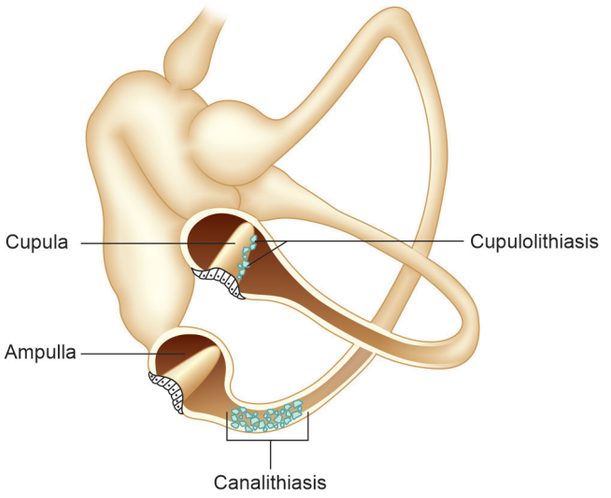


Fig. 5.4: Theories of BPPV

3. Otosclerosis is associated with BPPV in 50% of cases.
4. BPPV is seen in 10–15% patients following vestibular neuritis.
5. Meniere's disease
6. Osteopenia

Idiopathic BPPV is more seen in elderly population, which is due to age-related degeneration of otoconia.

Symptoms

BPPV most commonly arises from the posterior semicircular canal and less frequently from horizontal SCC and almost never from anterior (superior SCC). The posterior canal (80–90%) is the most commonly involved canal because it is the gravity dependent part of the vestibular labyrinth.

It is characterised by vertigo lasting seconds with or without nausea and imbalance on lying down or sitting up from lying or rolling over in bed. In between the episodes of attacks the patients are either absolutely symptom free or sensation of imbalance persists. There is no associated hearing loss.

Diagnosis

The diagnosis is made on performing the Dix Hallpike test. However, this test is specific for posterior SCC. For horizontal canal BPPV, supine roll test has to be done. The Dix–Hallpike manoeuvre should be avoided in a patient with cervical instability, vascular problems like vertebrobasilar insufficiency, acute neck trauma and cervical disc prolapse.

The BPPV has to be differentiated from other causes of vertigo like Meniere's disease, vestibular neuritis, superior canal dehiscence, post-traumatic vertigo, vestibular migraine and vertebrobasilar insufficiency.

Treatment

The treatment recommended is the **Epley's particle repositioning manoeuvre** (PRM), which is a canalith repositioning manoeuvre (Fig. 5.5).

1. The patient is seated on the couch facing away from the examiner and is brought down with head turned by 45 degrees (head is held by examiner) to the affected side and extended over the edge of the table (similar to Dix–Hallpike manoeuvre). Let the induced vertigo and nystagmus settle before the next step.

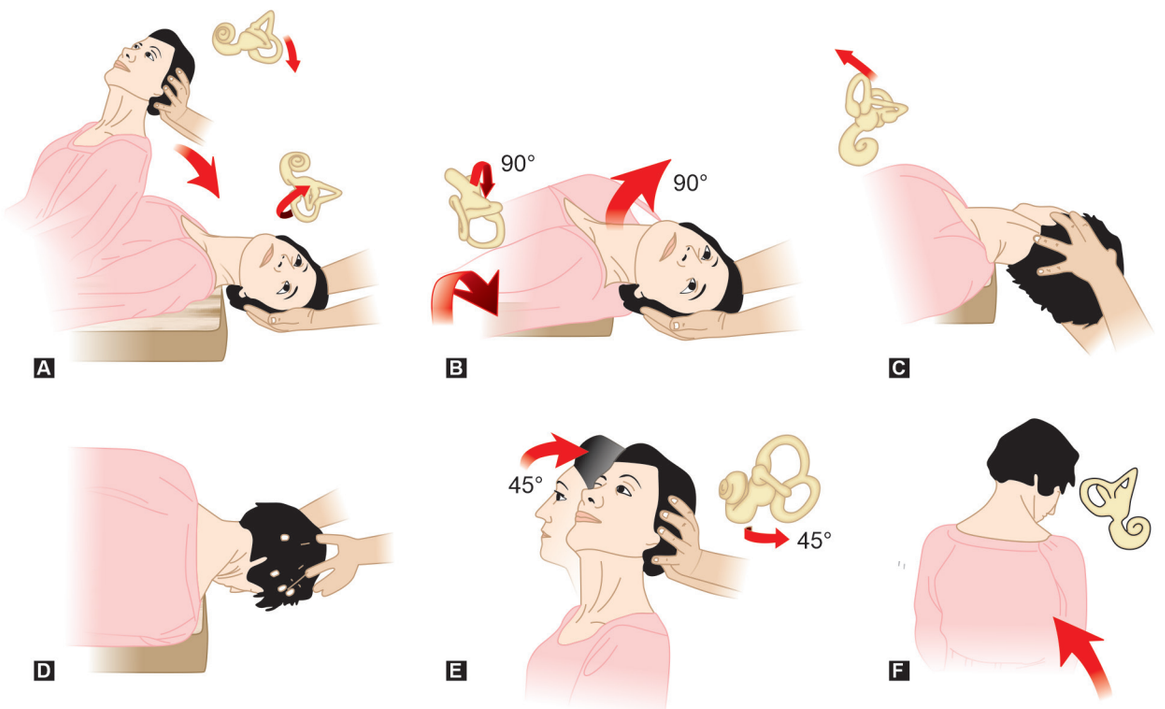


Fig. 5.5: Epley manoeuvre steps

2. The head is now turned by 90 degrees to the other side and the patient is now looking obliquely downwards.
3. The patient is made to roll over to the unaffected side by 90 degrees. Now the patient is looking towards the floor.
4. Next the patient is made to sit up with head 45 degrees towards the unaffected side.
5. The final step is to turn the head forward by 20 degrees.

The patient is made to lie for 10–20 minutes after the procedure to avoid vertigo. Patient is advised to sleep in a semirecumbent position for the same night and use two pillows for one week after that. The patient should sleep on the unaffected side for 2–3 weeks. The patients are told to avoid neck jerks for 1–2 weeks. The movements which require chin up positions should be avoided like beard shaving. The success rate of the procedure is around 90% if performed correctly and adequate precautions are taken.

Patients who have recurrent BPPV problems, should be educated about *Self Epley manoeuvre*. This is a self-application of Epley positions at home with the use of a pillow as the extension device. It is preferably done at night to avoid post positional vertigo.

For horizontal canal BPPV, *barrel roll* or barbeque roll manoeuvre is used for particle positioning and for Superior canal BPPV, reverse Epley or Yacovino manoeuvre is used.

Vestibular Neuritis

Acute severe dizziness, nausea and postural imbalance are the characteristic features of the disease. It occurs equally in both sexes, with typical age of presentation at 40–50 years of age. It is thought to be due to reactivation of latent viruses like the HSV type 1. The infection leads to inflammatory changes of the vestibular nerve with diffuse perivascular lymphocytic infiltrates.

Symptoms

These are sudden in onset with signs of unilateral vestibular loss. Severe symptoms include persistent vertigo associated with nausea, vomiting, pallor and sweating. The severe symptoms usually resolve within a few days. There are no associated cochlear symptoms of hearing loss or tinnitus.

On Examination

1. There is a spontaneous nystagmus which disappears or decreases on optical fixation.
2. The **head impulse test** (HIT) will show positive corrective saccades which are indicative of vestibular dysfunction.
3. The Dix–Hallpike test is done to rule out BPPV.
4. **Romberg test** shows increased sway towards the affected side and **gait test** shows deviations towards the affected side.
5. The **caloric test** is the gold standard test for unilateral peripheral dysfunction, which shows hypofunction.
6. For bilateral vestibular neuritis, **rotatory chair test** is the gold standard test.

Management

The treatment is required for the acute phase. It includes vestibular sedatives like prochlorperazine, antiemetics, antihistaminics and anticholinergics. Majority of patients improve in the next 1–6 months due to compensatory mechanisms. In cases of persistent symptoms, vestibular rehabilitation is started in terms of **Cawthorne–Cooksey exercises** (*vestibular rehabilitation*) and gait retraining.

Superior Semicircular Canal Dehiscence Syndrome (SSCDS)

There is a dehiscence of the bone over the dome of the superior semicircular canal, which results in formation of a third window. It is an abnormal connection between the inner ear and the middle cranial fossa. Any raise in intracranial pressure or any increased pressure at oval window/round window from loud sounds or straining leads to alteration in the inner fluid dynamics (Fig. 5.6).

Symptoms: There is a transient momentary vertigo episode. Patients also complain of autophony, tinnitus and hearing loss. Autophony of SSCDS has to be differentiated from the patulous Eustachian tube. SSCDS will have autophony with speech only but the patulous Eustachian tube will have autophony on speech and breathing both.

On examination we see an upward eye movement with ipsilateral intorsion or downward eye movements with ipsilateral extorsion.

Diagnosis: The HRCT temporal bone is the gold standard for identification of SSCDS. The CT imaging is done in a reformatted plane of the superior semicircular canal. This view is called **Poschl view** (Fig. 5.7).

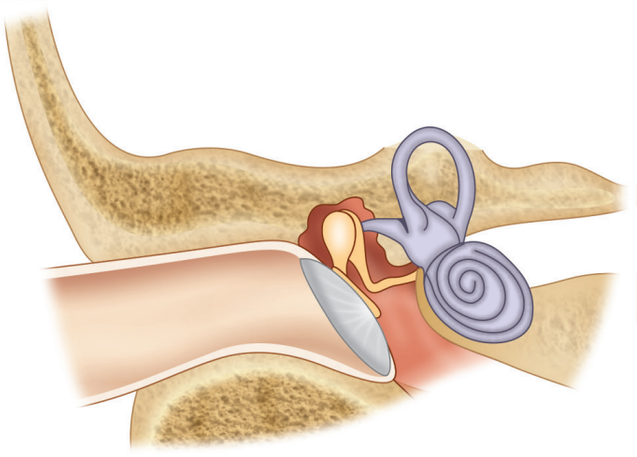


Fig. 5.6: Superior semicircular canal dehiscence syndrome



Fig. 5.7: HRCT temporal bone showing superior semicircular canal dehiscence (white arrow)

Treatment: The mild cases that are not troublesome require no active treatment. Ventilation tubes are beneficial in patients with pressure symptoms. If the symptoms are affecting day to day activities, then surgical management in terms of superior semicircular canal plugging, capping and resurfacing via transmastoid or middle cranial fossa approach is required.



Chapter

6

Diseases of the External Ear

EN2.13: Identify, resuscitate and manage ENT emergencies in a simulated environment (including tracheostomy, anterior nasal packing, removal of foreign bodies in ear, nose, throat and upper respiratory tract).
EN4.2: Elicit document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of diseases of the external ear.
AN40.4: Explain anatomical basis of otitis externa.
AN40.1: Describe and identify the parts, blood supply and nerve supply of external ear.

The auricle shows many **variations** in its development and these are not always pathological.

1. **Mozart's ear:** Great musician Mozart and his father both showed this dominant inheritance, where the antihelix is fused with helix.
2. **Wildermuth's ear:** The ear has a prominence of antihelix but the helix is underdeveloped. It has association with syndromes having sensorineural hearing loss.
3. **Darwin's tubercle:** It presents as a small elevation over the posterosuperior part of the helix.
4. Failure of lobule to separate from the side of the face is commonly seen in females and less so in males. It may affect all females of a family.
5. **Hypertrichosis pinnae auris:** It is a condition of abnormal excessive presence of hairs over the pinna. It is generally accepted to be genetically transmitted, probably Y-linked inheritance (Fig. 6.1).



Fig. 6.1: Hypertrichosis pinnae auris

CONGENITAL ABNORMALITIES OF AURICLE AND EXTERNAL AUDITORY CANAL

1. **Microtia and anotia:** This condition occurs due to arrest or maldevelopment of the mesenchymal nodules. In microtia, the pinna is rudimentary and malformed. It may be placed anterior and lower than the normal position. In anotia there is no development of pinna. These are usually associated with atresia of the external auditory canal. These children may require bone-anchored hearing aids (BAHA) and cochlear implantation for rehabilitation (Fig. 6.2).
2. **Bat ear:** The concha is large and there is poorly developed antihelix. It can have two variations: *Cup ear* and *lop ear* (Fig. 6.3). In the cup ear variant, most of the antihelix is underdeveloped and the lop ear variant has only crus of antihelix poorly developed. The treatment is surgical at 6–7 years of age (otoplasty).
3. **Macrotia:** These are oversized pinna which are large compared to the head. Normally the pinna size is 58–62 mm in females and 62–66 mm in males.
4. **Auricular appendages:** These appear as small elevations of skin with small amounts of elastic cartilage tissue. These are usually multiple and are found along the line joining the tragus and angle of mouth. The treatment is surgical.
5. **Atresia/stenosis of external auditory canal:** This is usually seen in association with pinna deformity, like microtia. It is taken as a sign of associated abnormalities of the middle ear (Fig. 6.4).





Fig. 6.2: Microtia



Fig. 6.3: Bat ear



Fig. 6.4: Stenosis of external auditory canal

CONGENITAL FISTULAE AND SINUSES

1. **Collaural fistula:** It is an abnormality of the first branchial arch. These can be superficial (type 1) which are only ectodermal. The type 2 has ectodermal and mesodermal origin contents and the fistula is usually between the floor of the external auditory canal opening anterior to the sternocleidomastoid muscle behind the angle of jaw. The tract is intimately related to the facial nerve. The treatment is surgical after adequate imaging modality to find the course of tract and relation to the facial nerve.
2. **Preauricular sinus:** This is formed due to the malfusion of the six *hillocks of His*. The opening of the sinus is formed in front of the crus of helix, which leads to a blind squamous lining track. The terminal portion of the tract may be adherent to the cartilage of the helix.

   The surgical excision of the sinus is required if there is history of repeated infections causing discharge and preauricular abscess formation (Fig. 6.5).

The preauricular sinuses may occur in association with syndromes. *The branchio-oto-renal syndrome* is one such common syndrome with renal abnormalities and associated external ear malformations.

PSEUDOCYST OF PINNA

Pseudocyst of the pinna is a rare disease, where a cystic swelling appears over the upper half of the anterior part of the auricle. Cyst contains a straw-coloured fluid and has no lining, hence a pseudocyst. It is more commonly seen in the Chinese population and is associated with



Fig. 6.5: Right preauricular sinus

vigorous rubbing of pinna or use of a hard pillow. The treatment is by *Button surgery*, where the entire cystic fluid is aspirated and large sized buttons are sutured on anterior and posterior side of the pinna across the cyst.

TRAUMA TO PINNA

1. Haematoma of Auricle

It is caused by trauma to pinna especially in closed contact sports like wrestling and boxing. There occurs a collection of blood between the perichondrium and cartilage. If left untreated it leads to permanent thickening resulting in **cauliflower ear/wrestler's ear** (Fig. 6.6).

The treatment is aspiration of the blood by a thick bore needle and pressure dressing. In case the patient has presented late, incision and drainage is done to evacuate the clots. The blood or clot removal should be done aseptically to prevent infection.

2. Avulsion and Laceration of Pinna

A deep laceration of pinna can be repaired by thin sutures. A small defect or loss of the superior part of the pinna can be treated with Burrow's triangles.

The re-implantation of the avulsed pinna should be done within 24 hours. This is done either by microvascular anastomosis or pinna being implanted as a composite graft. In case of loss of pinna, its reconstruction is done in a staged manner by using rib cartilage (Fig. 6.7).



Fig. 6.7: Avulsion of pinna



Fig. 6.6: (A) Hematoma of pinna (post traumatic); (B and C) Cauliflower ear

3. Frostbite

The higher surface area to its mass makes the pinna vulnerable to the extremes of temperature. The frostbite occurs in several stages:

i. **Frostnip:** Frostnip is seen in initial stages or in mild cases frostbite. Prolonged cold exposure leads to numbness of the pinna. As the skin warms, the patient may feel pain and tingling sensation. There is no permanent damage to skin on recovery.

ii. **Superficial frostbite:** Initially the skin is reddened that turns pale over time. On rewarming at this stage, the surface of skin appears mottled and stinging, burning and swelling is seen. A fluid-filled blister may appear 12–36 hours after rewarming the skin.

iii. **Deep/severe frostbite:** As frostbite progresses, all layers of the skin are affected and it turns white or bluish grey with numbness, losing all sensation of cold, pain or discomfort. Large blisters are formed 24–48 hours after rewarming, the area then turns black and hard, and there is tissue loss.

Current methods used to treat cases of frostbite include gradual rewarming, anti-inflammatory drugs, hyperbaric oxygen, vasodilators and surgical debridement. Systemic antibiotics may be added for deep infections.

4. Keloid

The keloid of pinna is formed most commonly by ear piercing for ornaments. It is commonly seen on piercing of the pinna cartilage (in high ear piercing). The treatment is either intralesional application of triamcinolone or surgical removal. It is recommended to inject steroids after surgical removal, to prevent recurrences (Fig. 6.8).



Fig. 6.8: Keloid

Pinna Cyst

The most common cyst of pinna is a sebaceous cyst and the treatment is simple excision (Fig. 6.9).

INFLAMMATIONS OF EAR CANAL AND PINNA

1. Perichondritis

It occurs due to bacterial infection of the surgical incision, haematoma or laceration. Initially pinna is red and tender (Fig. 6.10). This followed by swelling and eventually subperichondrial abscess between cartilage and perichondrium. *Pseudomonas* is the most common organism involved. Initial stages require antibiotics but if subperichondrial collection occurs it requires drainage to prevent avascular necrosis of pinna (Fig. 6.10).

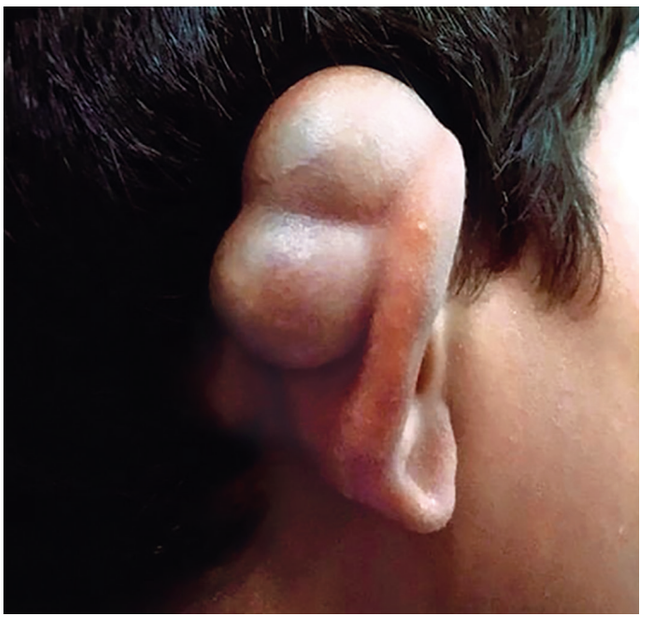


Fig. 6.9: Sebaceous cyst of pinna



Fig. 6.10: Perichondritis

2. Relapsing Polychondritis

It is an autoimmune disorder which is characterised by episodic inflammatory disease of the connective tissue and various cartilages of the head and neck region. It can involve the pinna cartilage and also nasal cartilage. The treatment is systemic steroids.

3. Chondrodermatitis Nodularis Chronica Helicis

This is a painful, persistent, erythematous nodular lesion of the pinna. It commonly appears on the helical rim of the pinna in men over 40 years of age. It is caused by trauma like frostbite, sun, wind and extreme temperature. Ageing is also a factor. The effective treatment is local excision. Alternatively, CO_2 laser can be used to vaporise the lesion.

4. Tophi

This is asymptomatic subperichondrial deposition of sodium biurate crystals in helix and antihelix of gout patients.

TUMOURS OF THE AURICLE

These can be benign or malignant. The benign tumours are papilloma, fibroma and chondroma. Rarely, haemangiomas may be seen.

The malignant tumours of the pinna are the squamous cell carcinoma, basal cell carcinoma and malignant melanoma.

CONDITIONS OF THE EXTERNAL AUDITORY MEATUS

1. Otitis Externa

Otitis externa is the inflammation of the external auditory canal skin. The predisposing factors include excessive wax production, narrow ear canal, heat and humidity, habit of itching the canal with matchsticks and hairgrips.

1. **Furunculosis:** It is the localised infection of the ear canal caused by gram-positive bacteria usually *Staphylococcus aureus*. It is a painful condition and may progressively increase in size to occlude the ear canal and obstruct the visualization of the tympanic membrane. It may also cause pain in the jaw movement. The infection of the posterior wall of the canal can spread to the postauricular sulcus. Large furuncles usually rupture and discharge pus and thereafter rapidly resolve. This condition is sometimes confused with acute mastoiditis (Table 6.1).

   Treatment:

   1. Cotton wick soaked in 10% ichthammol glycerol is effective in reduction of pain and infection. Glycerol is hygroscopic and decreases oedema of the canal and ichthammol is antiseptic.
   2. Systemic oral antibiotics with anti-inflammatory and analgesic drugs.
   3. Topical antibiotics are recommended for application to the ear and nasal vestibules (infection spread via fingernails to ear) in cases of recurrent infections, especially in diabetics.

Table 6.1: Differences between furunculosis and acute mastoiditis

Sign	*Furuncle*	*Acute mastoiditis*
1. Postauricular tenderness	Diffuse	Maximum over the antrum area
2. Pinna position	Forwards	Forwards and downwards
3. Lymphadenopathy	Present	Absent
4. Tragal tenderness	Present	Absent

2. **Diffuse otitis externa:** It is also called the *tropical ear* or *Singapore ear* because it is more common in hot and humid conditions. Other causes include vigorous ear cleaning, unskilled syringing of ear canal, and local trauma to the canal. It may also occur due to continuous discharge in active chronic otitis media (Fig. 6.11).

   Pseudomonas aeruginosa, *Bacillus proteus* and *Staphylococcus aureus* are the common pathogens involved.

   Acute stage begins with discomfort in the ear, which rapidly progresses to pain in and around the ear. There is swelling of the soft tissues and the pinna is displaced. There is a collection of debris and pus in the ear canal. The postauricular lymphadenopathy is generally seen. The tympanic membrane is dull in appearance.

   Treatment of acute stage:

   1. Cleaning of the ear canal is essential, especially the meatal recess.
   2. Antibiotic smeared (Mupirocin, neomycin) wicks packing in the ear canal.



Fig. 6.11: Diffuse otitis externa

3. Topical corticosteroid (ointment) may be used in cases of diffuse oedema and unresponsive cases.
4. Broad spectrum antibiotics are given especially in cases with lymphadenitis and gross oedema.
5. Anti-inflammatory and analgesic drugs are also combined.
6. Prevention of water to go inside the ear.

In the chronic stage, the chief symptom is irritation and discharge. There is no tenderness but the meatal skin is thickened. Pus and debris are present in the meatus. Few granulations may be found on tympanic membrane.

Treatment:

1. Cleaning of the ear canal is essential, especially the meatal recess.
2. Antibiotic and steroid smeared wicks are kept in meatus. Steroids help in reducing oedema and itching.

Failure to respond to medication points to either underlying chronic otitis media or fungal infection.

2. Otomycosis

Fungal infection of the meatus occurs commonly in hot and humid conditions. It can occur commonly in patients of mastoidectomy with open cavity and patients who use hearing aids. The common organisms involved are *Aspergillus niger*, *Candida albicans*, *dermatophytes* and *Actinomyces* (Fig. 6.12).

The patient usually complains of itching, discomfort in ear, watery discharge and ear fullness. The discharge is musty in odour. With deeper tissue inflammation the ear becomes painful. On examination, greyish white debris resembling a wet filter paper filling the meatus are seen. The conidiophores of *Aspergillus niger* appear black specs and those of *Candida* appear white or creamy.

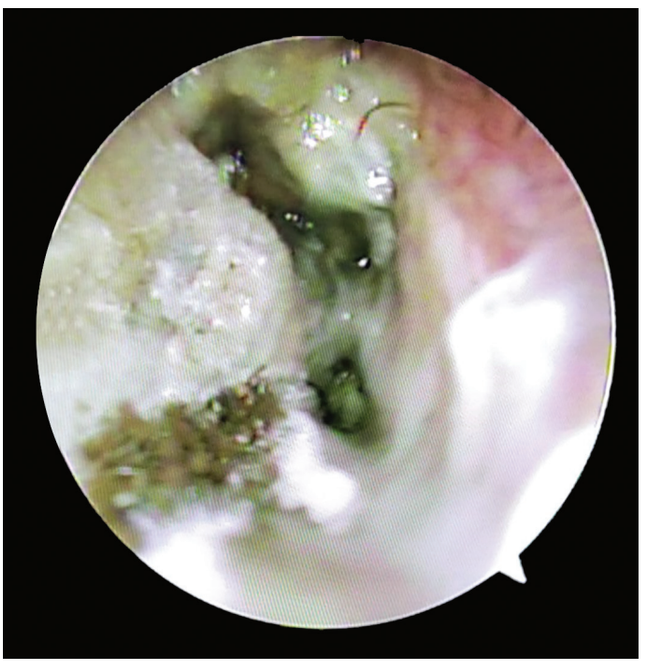


Fig. 6.12: Otomycosis (*Aspergillus niger*)

Treatment

1. Ear cleaning with suction or if required irrigation is vital to remove debris.
2. Application of antifungal agents 1% *Clotrimazole cream* or drops.
3. Gentian violet is an useful antifungal.
4. 2% salicylic acid in alcohol is an effective antifungal agent.

3. Otitis Externa Haemorrhagica (Bullous Myringitis)

It is characterised by the formation of purple blebs on the tympanic membrane and the deep meatus. The aetiology is uncertain, probably due to viral infection. It has been seen in influenza epidemics more commonly. Its symptoms are pain and serosanguineous discharge (blood tinged). The condition is self-resolving. Antibiotics may be given to prevent superadded infections. Analgesics are given for pain relief.

4. Malignant Otitis Externa

The malignant otitis externa or the necrotizing otitis externa is an infection involving the temporal bones, surrounding soft tissues and skull base. It is called malignant due to high mortality rate in earlier times and necrotizing because of the destructive nature of this infection. Its predisposing factors include old age, poorly controlled diabetics, immunosuppression by chemotherapy, IgA deficiency, acute monocytic leukaemia, neutropenia and bone marrow transplantation.

Pathogens

The most common infective organism is *Pseudomonas aeruginosa*, rarely it can be due to fungal infections (especially immunocompromised patients) of which *Aspergillus fumigatus* is the most common.

Spread

The infections start with cellulitis of external auditory meatus and progresses to form granulomas in the floor of deep meatus. *Pseudomonas* produces exotoxins and enzymes like the elastase which causes the necrotising vasculitis which aids the organism in spread. Eventually the disease spreads through naturally occurring fissures to the parotid gland, temporomandibular joint and the soft tissues of the skull base. The spread to the mastoid tip and stylomastoid foramen leads to facial palsy. Further the spread leads to thrombosis of lateral sinus. The infection spread to the petrous apex can spread to middle cranial fossa. Along with palsy of facial nerve, palsies of IX, X, XI cranial nerves at jugular foramen and XII nerve at hypoglossal canal are frequently seen.

Clinical Features

The most common symptom is *excruciating pain*. The pain is nocturnal, deep, lanciating and disproportionate to the clinical signs. Pain is often unresponsive to analgesics. The second most common symptom is purulent discharge. On examination, there are granulations in the deep part of meatus obscuring the view of the tympanic membrane. There might be hearing loss due to granulations and discharge. There can be accompanying facial palsy and lower cranial nerve palsies (Fig. 6.13).

Diagnosis is made on exclusion. The granulations in EAC are to be sent for histopathology to rule out malignancy. High resolution CT scan of the temporal bone and skull base is essential to diagnose bone erosion in the skull base. *Technetium-99m* methylene diphosphonate (MDP) is used for detection of temporal bone osteomyelitis. *Gallium-67-citrate* scanning is an ideal for monitoring the resolution of condition while on treatment.

Treatment: It includes control of underlying comorbidities like diabetes. The patient is admitted and started on insulin. The ear toilet is done to clean discharge and debris. Long-term use of systemic antibiotics like fluoroquinolones for 4–6 weeks is indicated. Monotherapy with ciprofloxacin is the preferred initial treatment (400 mg IV × 8 hourly for 4–6 weeks). Alternative regime with cephalosporins like ceftazidime and cefepime in combination with aminoglycosides are used in resistant cases. Use of hyperbaric oxygen has shown additional benefits. There is not much role in any surgery or facial nerve decompression.

5. Seborrheic Otitis Externa

It is associated with dandruff or scurf of the scalp. There is often scaling in the external auditory canal, postauricular sulcus and below the lobule. The aetiology is unknown. Scalp should be routinely washed with

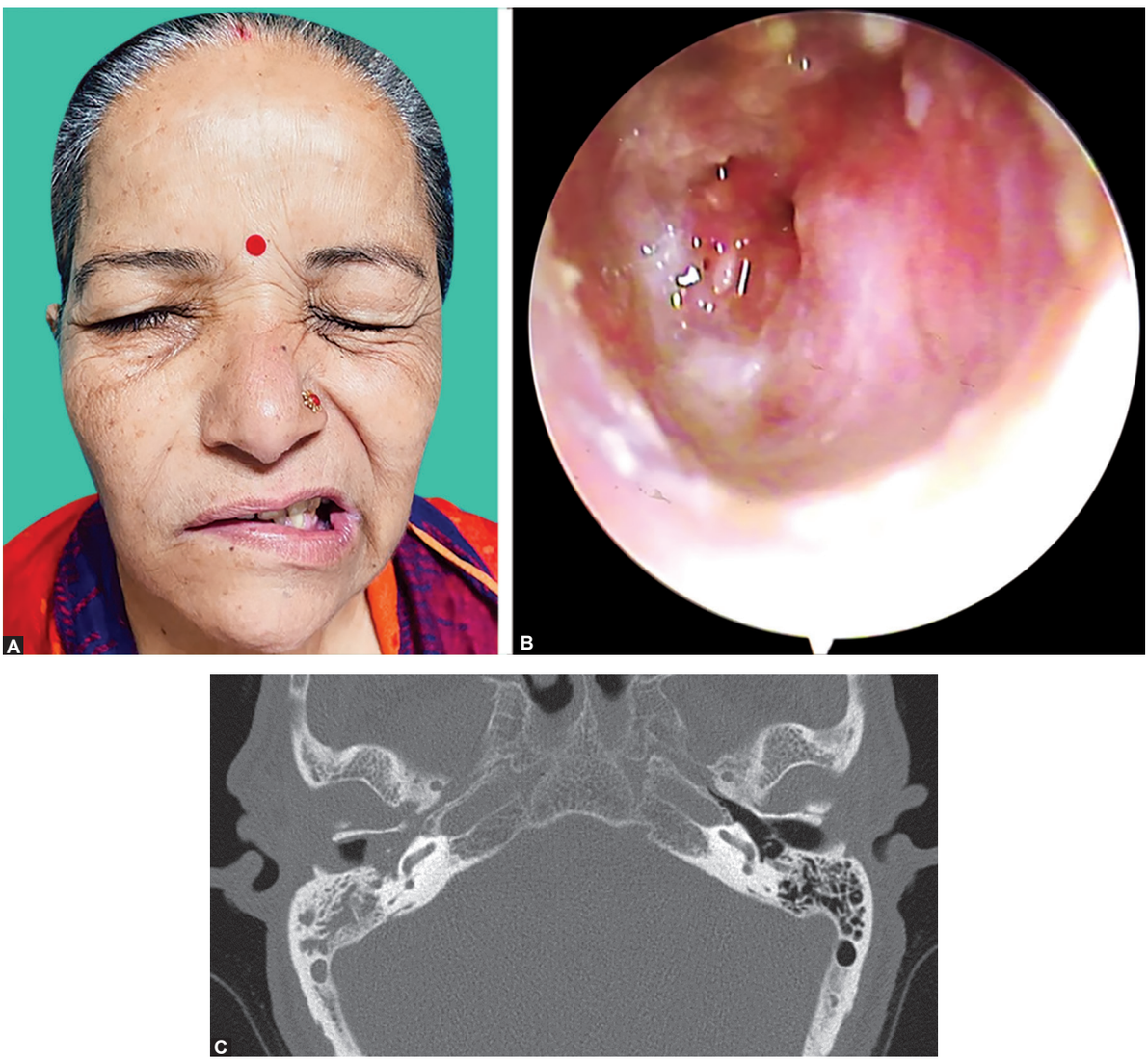


Fig. 6.13: (A) Right facial palsy seen in malignant otitis externa (*Courtesy*: Dr Ishwar Singh, Director Professor, ENT, MAMC). (B) Granulations in EAC extending into the middle ear; (C) HRCT temporal bone scan axial cut showing bony canal erosion on the left side in a case of malignant otitis externa (*Courtesy*: Dr Jyoti Kumar, Director Professor, Radiology, MAMC)



Fig. 6.14: Seborrheic dermatitis of pinna and EAC

keratolytic shampoo like cetrimide. Prevention of water entering the ear is recommended. Refrain from attempting to remove wax debris (Fig. 6.14).

6. Herpes Otitis Externa

It has been discussed on the topic of facial nerve.

7. Eczema (Atopic Dermatitis)

It is sensitization of the meatus by either infections or allergic material leading to eczematous reactions of the skin. It is commonly seen with neomycin ear drops. Vesicles are formed due to the eczematous reaction. These vesicles burst to produce serous discharge and intense itching. The treatment is to clean the meatus and application of antibiotic steroid ointment.

8. Neurodermatitis

It occurs due to psychosomatic disorder, with the tendency to compulsively itch the meatus. Initially the examination is normal. But with constant itching there is lichenification of the meatal skin with secondary infections sometimes. The secondary infections should be treated and psychological problems to be taken care of.

9. Keratosis Obturans

It is the presence of a keratotic plug of desquamating squamous epithelium in the bony part of the external auditory meatus. The aetiology is unknown but may be due to faulty migration of the squamous epithelial cells. This condition is commonly seen in younger age group. The condition is frequently associated with bronchiectasis and sinusitis. Gram-negative organisms and *Staphylococcus aureus* have been frequently isolated.

Clinical features: The common complaints are conductive hearing loss and otorrhoea. On examination there is a pearly white mass which fills the meatus tightly and is difficult to remove on cleaning (Fig. 6.15A). The tympanic membrane is not visualised.

Treatment consists of removal of the keratotic mass under local anaesthesia (Fig. 6.15B). Syringing is usually avoided as it may push the mass posteriorly and increase discomfort. The removal of mass reveals underlying bone meatus widening with associated few granulations. The external auditory canal is then packed with antibiotic and steroid ointment pack. Small subsets of patients show recurrences which require repeated keratotic plug removal.

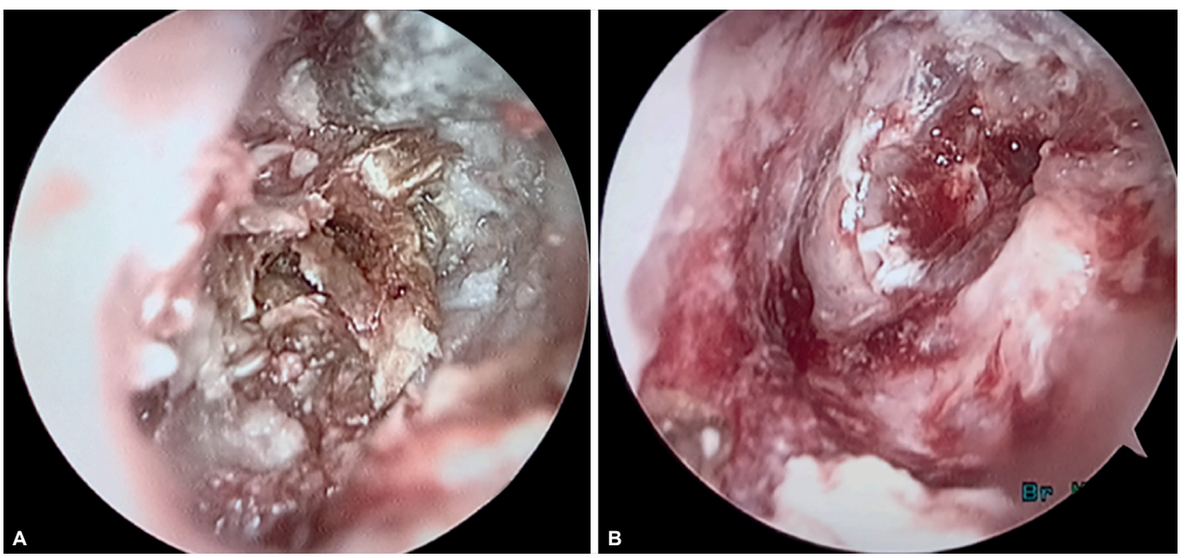


Fig. 6.15: (A) Keratotic plug occluding the canal; (B) Widened EAC after removal of plug

EN3.4: Observe and describe the indications for and steps involved in the removal of foreign bodies from ear, nose and throat.

10. Foreign Body Ear

Foreign bodies of the ear are commonly encountered in ENT practice. These can be living foreign bodies or non-living foreign bodies.

The *living foreign bodies* are flies, beetles, maggots, ants or other insects. Attempts to visualise and catch them live is difficult, therefore they are first killed by instilling oil in the meatus or by use of chloroform or spirit. Once these are killed, they are removed either by forceps or suction under microscope preferably (Fig. 6.16).

Non-living foreign bodies include paper, grains, chalk pieces, beads or metallic balls (Fig. 6.17). The methods to remove them include forceps removal, syringing, Suction removal and for impacted or sharp penetrating foreign bodies are removed by postauricular incision. Metallic balls may also be difficult foreign bodies and may require general anaesthesia for removal. The vegetative foreign bodies are not removed by syringing, because they swell on water contact and break up and make removal more difficult.

11. Impacted Wax (Cerumen)

Normally the wax produced dries up and is slowly expelled from the meatus by epithelial migration and jaw movement.

Aetiology and symptoms: Due to factors like narrow and tortuous meatus, obstructive lesions like exostosis, or excessive wax production, a mass of wax may dry up and form the impacted wax (Fig. 6.18). This causes hearing loss, tinnitus, reflex cough due to Arnold's nerve stimulation, earache and vertigo. It is also quite troublesome for the usage of hearing aids. Long-standing hard impacted wax may lead to the formation of granuloma (*wax granuloma*).

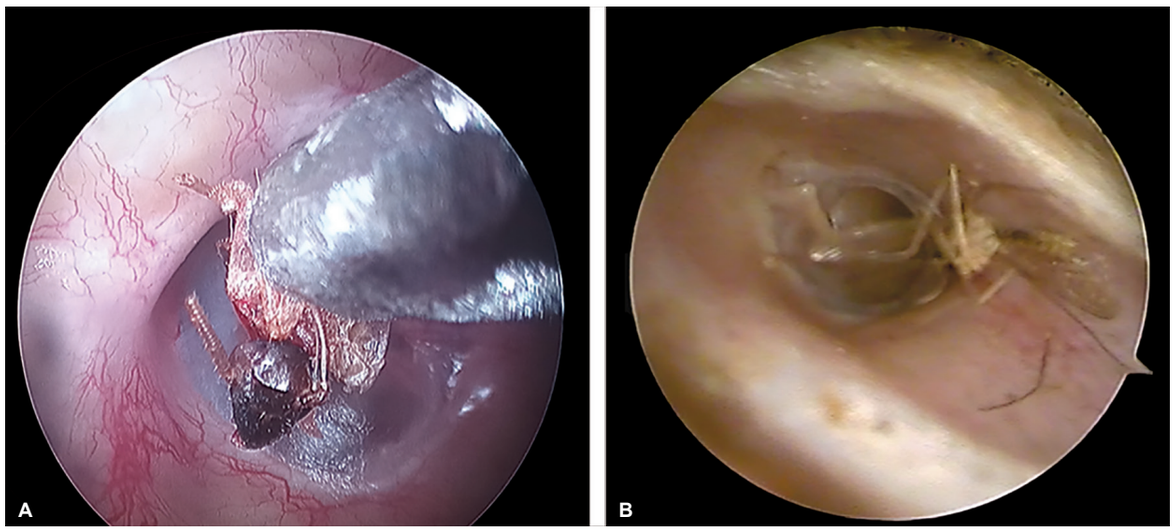


Fig. 6.16: Insects in EAC

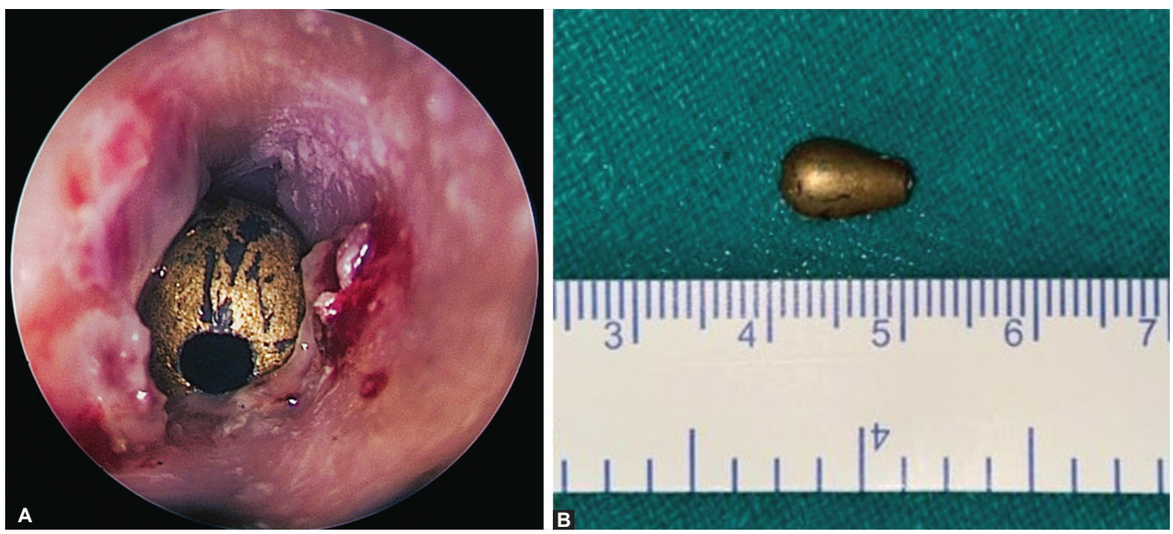


Fig. 6.17: (A) Plastic bead in the EAC with trauma to the canal; (B) Bead after removal from the canal



Fig. 6.18: Impacted wax in EAC

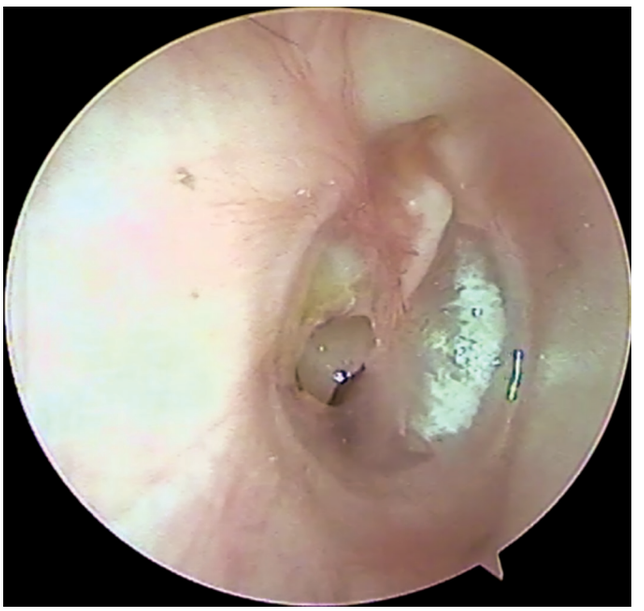


Fig. 6.19: Traumatic perforation of right ear following slap.

Treatment

1. It is advisable to soften the hard wax by use of wax softeners like commercially available 2% paradichlorobenzene, Hydrogen peroxide, liquid paraffin or olive oil and 5% soda bicarb with glycerine in equal amounts.
2. Wax is removed either by gentle probing using Jobson–Horne probe or by syringing.

TRAUMATIC PERFORATION OF TYMPANIC MEMBRANE

The tympanic membrane may be ruptured due to sudden air pressure changes in EAC, by fluids and sharp objects inserted in EAC (Fig. 6.19).

- Air pressure change trauma: A sudden blow to ear with sealing of meatus, can result in tympanic membrane rupture. This occurs due to trauma by a 'ball' or an open hand. The anteroinferior quadrant is the most likely area of trauma. The tendency to rupture of tympanic membrane increases with age.
- Fluid injury may occur while doing forceful syringing or performing water sports.
- Matchsticks and hairpins used to itch ears may result in trauma to the tympanic membrane.

Treatment: The healing of the perforation is dependent on secondary infection in EAC and the middle ear and on the amount of tympanic membrane tissue loss. Small perforations which are not infected have high chances (almost 90%) of spontaneous closure and regain normal function. In case there is a loss of a large part of the tympanic membrane as seen in blast injury the tympanic membrane is unlikely to heal spontaneously and will require some surgical intervention.

1. Cases without infection of EAC or middle ear, 'do nothing' and wait for spontaneous closure of the perforation. The prevention of secondary infection is needed. Especially prevent water from entering the EAC.
2. Antibiotic eardrops are contraindicated because they may introduce infection from the unsterile part of the lateral EAC.
3. If a patient presents with EAC or middle ear infection, systemic antibiotics are prescribed.
4. If the perforations do not heal in 3–6 months following trauma, myringoplasty is indicated.

EN4.9: Demonstrate the correct technique for syringing wax from the ear in a simulated environment.

SYRINGING PROCEDURE

The procedure is indicated for removal of impacted wax or non-vegetative foreign body from the EAC. Syringing is contraindicated in case of active infection of the external auditory canal like otitis externa and in perforation of the tympanic membrane.

The patient is seated in front of the examiner with the concerned ear towards the examiner. The examiner should wear a face shield to prevent water splash exposure. Once the procedure has been explained to the patient, the examiner wears a headlight or headmirror and focusses the light on the ear. The ear is inspected and pinna pulled upwards and backwards. A 50 ml syringe with cannula is introduced in the meatus, aiming in posterosuperior direction, water is then slowly injected into the canal. The water for irrigation should be at around 37 degree C The procedure might require to be repeated for more impacted wax. Expelled water and wax debris or a foreign body is collected in a kidney tray held below the ear snugly close to face. Once the wax has been removed, the ear canal is mopped dry (Fig. 6.20).

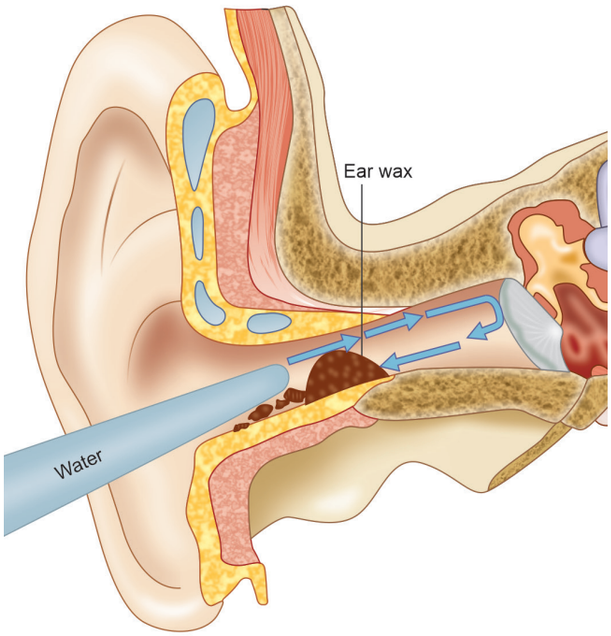


Fig. 6.20A: Method of syringing



Fig. 6.20B: Syringing procedure

Rarely forced high volume syringing may result in traumatic perforation especially in patients with atrophic tympanic membrane. These perforations are to be treated like other traumatic perforations.

BENIGN TUMOURS OF THE MEATUS

1. Papilloma

These are viral in origin and occur in the outer part of the ear canal. Usual mode of infection is from the infected fingers with similar lesions. These should be excised with electrocautery or Laser under anaesthesia. The base of papilloma should be cauterized to prevent recurrences. The diffuse papillomas are seen rarely, which can extend deep into the meatus. These can be removed by electrocautery or laser, but recurrence is not uncommon.

2. Hyperostosis

These are of mainly two types: Exostosis or osteomas. Both are due to the growth of the cortical bone.

Exostosis

They present as multiple bilateral, smooth sessile elevations in the deep part of the meatus. These are more common in cold water bathers. These have been seen in more than 50% of the surfers (*surfer's ear*). Small exostosis does not require any treatment. The large swellings causing hearing loss require surgical removal.

Osteomas

These are seen as single rounded pedunculated masses in the outer meatus (Fig. 6.21). These are usually unilateral. These are surgically drilled out if they cause significant obstruction to sound conduction to the tympanic membrane or accumulation of wax and debris behind them.

3. Adenomas

There are mainly two types of adenomas arising from the glands in the meatus.

1. The sebaceous adenoma arises from sebaceous glands. They can easily be treated by local wide excision.
2. The ceruminoma arises from ceruminous glands and is seen in the cartilaginous portion of EAC. It is treated by wide local excision.

MALIGNANT TUMOURS OF EAR

1. Squamous cell carcinoma
2. Ceruminous adenocarcinoma
3. Meningioma

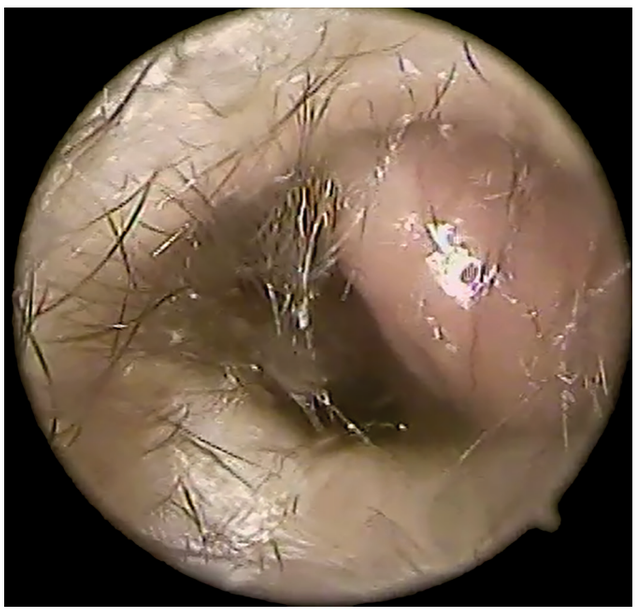


Fig. 6.21: Osteoma of left external auditory canal

4. Acoustic neuroma
5. Glomus tumours (paragangliomas)
6. Endolymphatic sac tumour (low grade papillary epithelial neoplasm).

According to the WHO classification of 2017, otosclerosis and cholesteatoma are also included in the list of low-grade tumours of the ear.

Squamous Cell Carcinoma of Ear

It is a relatively rare tumour, with an incidence of 1 in a million. They can arise from the external auditory canal or the middle ear. Squamous cell carcinoma is the most common tumour type of the temporal bone. They account for less than 0.5% of all cancers of the head and neck region. The tumour is most commonly seen in the seventh decade.

Aetiology

The most important risk factor for squamous cell carcinoma is the chronic otitis media. The other risk factors are radiation exposure (latency period of 5–30 years), exposure to ultraviolet light and chlorinated disinfectants.

Tumour Spread

Tumour commonly arises from external auditory canal.

1. The *foramen of Huschke* and *fissures of Santorini* in EAC provide minimal resistance to tumour spread to the parotid gland and temporomandibular joint.
2. The tumour spreads medially to the tympanic cavity. Here, it can cause involvement of the facial nerve. The middle ear mucosal involvement leads to serosanguinous discharge.
3. From the middle ear the tumour can spread to the middle cranial fossa by erosion of tegmen tympani.
4. Tumour spreads posteriorly to the mastoid cavity and can further spread to posterior cranial fossa.
5. The petrous apex may get further involved with medial spread and encasement of internal carotid artery may occur with anterior spread.
6. The last four cranial nerves can be involved by the inferior extension of the tumour to the jugular fossa.
7. Metastasis to lymph nodes is seen in around 10% of patients. The first echelon nodes are the intraparotid and postauricular lymph nodes.
8. Distant metastasis may be seen in the liver, lung and bones.

Presenting Symptoms

1. Otalgia is the commonest complaint.
2. Bloodstained otorrhea which is foul-smelling.
3. Hearing loss.
4. Cranial palsies: VII, IX, X, XI, and XII nerves.

On Examination

1. Exophytic mass, subdermal swelling or ulceroproliferative growth in external auditory canal (Fig. 6.22A).
2. Facial nerve along with other lower cranial nerves involvement, leading to facial asymmetry, dysphonia and dysphagia (Fig. 6.22B).
3. Neck examination especially for level II and III lymph nodes may be enlarged.

Diagnosis

1. Transcanal biopsy of the growth.
2. High resolution computed tomography and magnetic resonance imaging are vital to map tumour extensions especially intracranial involvement.

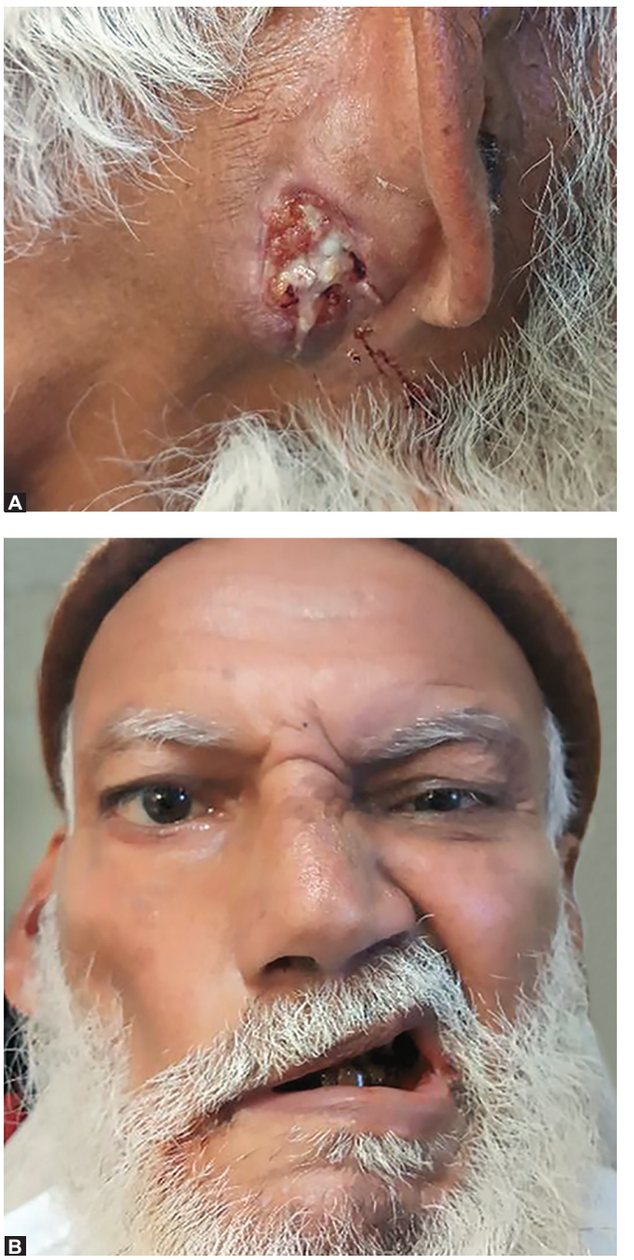


Fig. 6.22: Carcinoma of middle ear with mastoid involvement and facial palsy

Staging

TNM

T Classification

T1: Limited to the EAC without bony erosion or evidence of soft tissue involvement.

T2: Limited to the EAC with bone erosion (not full thickness) or limited soft tissue involvement (<0.5 cm).

T3: Erosion through the osseous EAC with limited soft tissue involvement (<0.5 cm), or tumour involvement in the middle ear and or mastoid.

T4: Erosion of the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura; with extensive soft tissue involvement (>0.5 cm, such as involvement of the TMJ or styloid process) or evidence of facial paresis.

N Classification

N0: No regional nodes involved

N1: Single metastatic regional node <3 cm in size

N2a: Single ipsilateral metastatic node 3–6 cm in size

N2b: Multiple ipsilateral metastatic lymph nodes

N2c: Contralateral metastatic lymph node

N3: Metastatic lymph node >6 cm in size.

Overall stage

I: T1N0

II: T2N0

III: T3N0

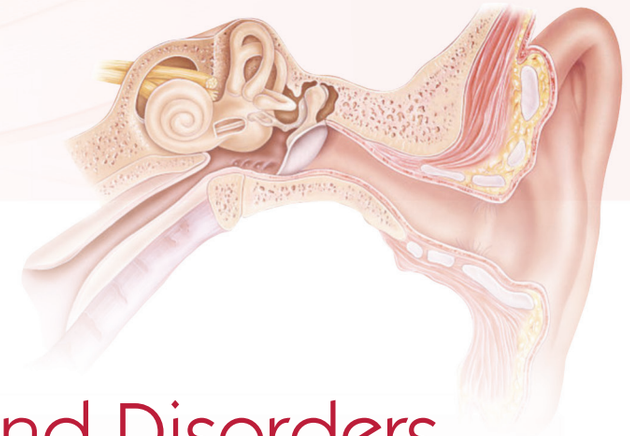
IV: T4N0 and T1-4N1-3.

Treatment

1. *Lateral temporal bone resection*: Tumours lateral to the tympanic membrane (T1 and T2) can be adequately excised by a lateral temporal bone resection. Extended cortical mastoidectomy is performed. Superior limit of dissection is the tegmen plate, anterior limit is the zygoma and the inferior limit is the stylomastoid foramen. The facial nerve is skeletonised and is preserved. Entire mucosa of the middle ear cleft is removed and the mastoid cavity is obliterated with muscle, along with Eustachian tube opening. Pinna is preserved if EAC tumour resection margin is >5 mm from away from it.
2. *Extended temporal bone resection*: This procedure is required for all T3 and T4 tumours. It is a more radical procedure requiring removal of the head of the mandible, middle ear, mastoid contents and the inner ear structures. Facial nerve is sacrificed. The middle cranial fossa dura is the superior limit of removal, jugular bulb inferiorly, ICA anteriorly, posterior cranial fossa dura posteriorly and petrous apex medially.
3. Radical neck dissection is done in cases of positive metastatic neck node.
4. Post operative radiotherapy is done in cases with positive surgical margins or perineural invasion.

Chapter

7



Eustachian Tube and Disorders

AN40.2: Describe and demonstrate the boundaries, contents, relations and functional anatomy of middle ear and auditory tube.

Eustachian tube or the *pharyngotympanic tube or auditory tube* is a bony cartilaginous tube which connects the tympanic cavity (middle ear) with the nasopharynx. The tube shape is like two cones joined with each other at their apices. It can also be likened to the shape of a trumpet (Figs 7.1 and 7.2).

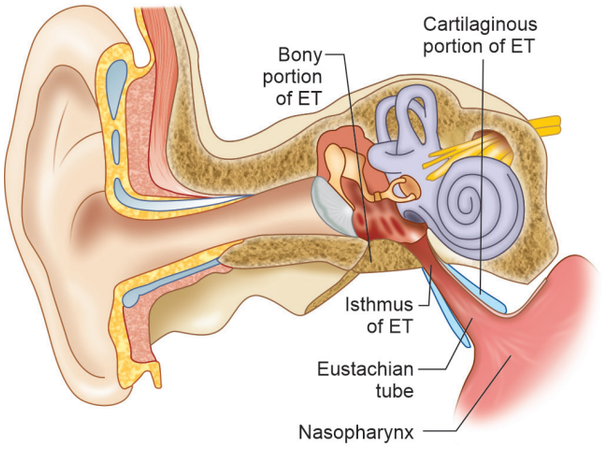


Fig. 7.1: Eustachian tube (ET) connecting the middle ear cavity with the nasopharynx

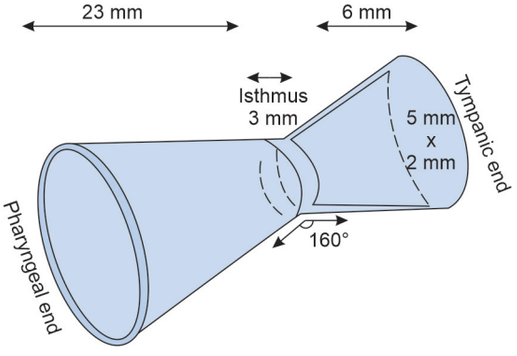


Fig. 7.2: Structure and shape of Eustachian tube

The length of the adult Eustachian tube is around 36 mm (33–38 mm) and it runs forwards, medially and downwards from the anterior wall of middle ear, making an angle of 45 degrees with the horizontal. The lateral one third (12 mm) is bony and it opens into the anterior mesotympanum (protympanum) above the level of the floor of the middle ear. The opening in the middle ear is oval or circular in shape, and its size is 5 × 2 mm (Fig. 7.3A).

The medial part of the Eustachian tube is fibrocartilaginous, and it is 24 mm in length. Junction of the two parts (bony and cartilaginous) is the narrowest portion of the tube and is called *isthmus* (2–3 mm).

The medial end (pharyngeal end) is like a slit and it produces an elevation in the lateral pharyngeal wall, which is called *torus tubarius*. Above the torus is the tubal tonsil (*of Geralch.*). Behind the torus tubarius is the pharyngeal recess or the *fossa of Rosenmuller*. The pharyngeal end is 1.25 cm behind and little below the posterior end of the inferior turbinate (Fig. 7.3B).

The pharyngeal end of the tube has respiratory mucosa but as we go towards the middle ear the number of goblet cells decrease and cilia number also go down.

Ostmann's pad of fat, also known as **Ostmann's fatty bodies**, is located inferomedial to the pharyngeal opening of Eustachian tubes. This has an important role in normal closure of the tubal opening. Absence of this triangular fat pad is an important feature of the patulous Eustachian tube.

Elastin hinge is an elastin rich zone present at the junction of medial and lateral lamina of the cartilage portion of the tube. It keeps the tube closed until acted upon by the tensor veli palatini muscle.

Muscles Related to Eustachian Tube

(Figs 7.3 and 7.4)

1. Tensor veli palatini arises from scaphoid fossa of the pterygoid bone and cartilaginous part of the Eustachian tube and descend to converge with the soft palate.
2. Levator veli palatini arises from the lower surface of the cartilaginous part of the tube and spreads out into the soft palate.

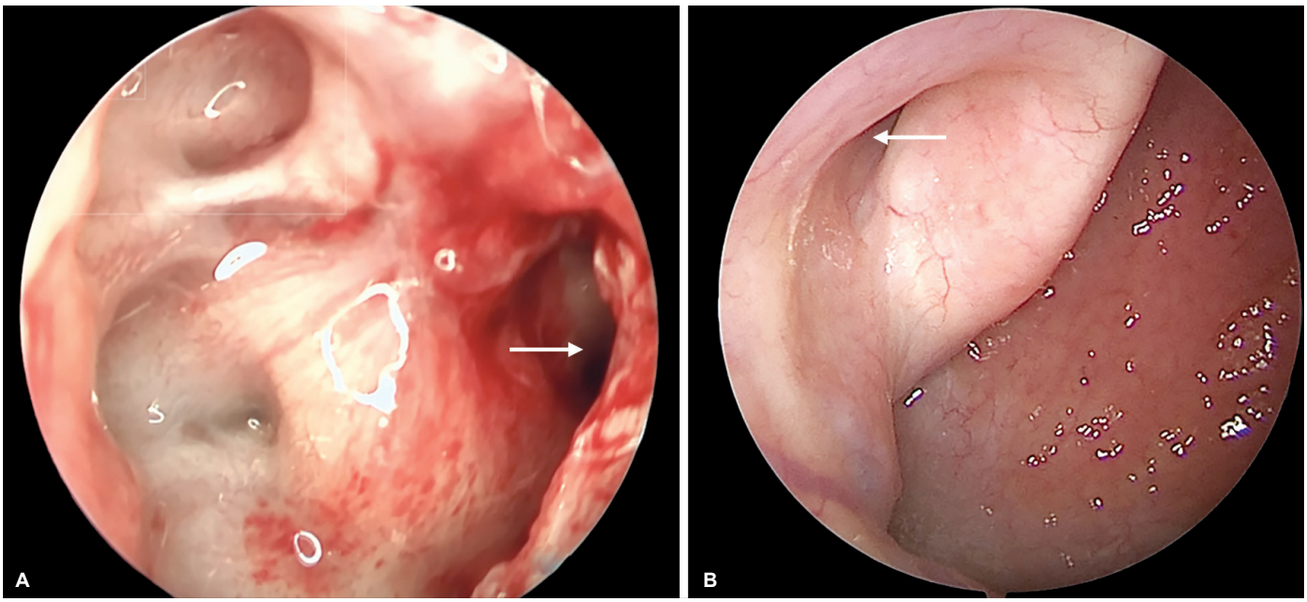


Fig. 7.3: (A) Lateral end of the Eustachian tube opening in the protympanum; (B) Medial end of Eustachian tube as seen by nasal endoscope (white arrows)

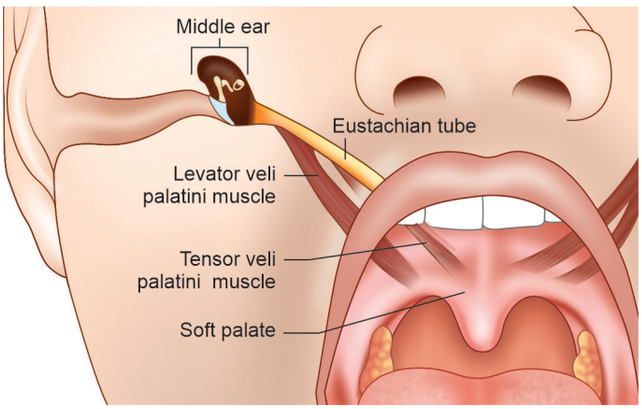


Fig. 7.4: Muscles of Eustachian tube

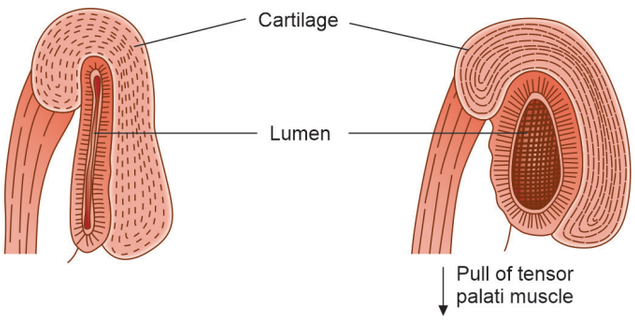


Fig. 7.5: Mechanism of Eustachian tube opening

The contraction of the tensor veli palatini causes the dilation and opening of the distal Eustachian tube. The contraction of the levator veli palatini results in elevation of the soft palate and medial rotation of the cartilaginous portion of the tube. In simultaneous contraction of these muscles, as in swallowing or yawning; leads to air passing through the tube to equilibrate pressure in the middle ear with atmospheric pressure (Fig. 7.5).

The closure of the tubal opening is by the elastic recoil of the elastin hinge cartilage and deformation of the Ostamann' fat.

3. Salpingopharyngeus: Its role in Eustachian tube function is doubtful.

Nerve Supply

Tensor veli palatini is supplied by the mandibular branch of the trigeminal nerve. Salpingopharyngeus and levator veli palatini are supplied by motor fibers from the pharyngeal plexus (fibres of XI nerve through X nerve). The parasympathetic secretomotor supply to the mucosa of the eustachian tube is received from the tympanic branch of IX nerve.

Eustachian Tube in Children

The Eustachian tube is not fully developed by birth. It is wider, shorter and more horizontal compared to the adult auditory tube. Due to these factors, the infant is more prone to have middle ear infections. Moreover, the infant tube is flaccid and ineffective in preventing ingress of secretions of gastroesophageal reflux (GER) to enter the middle ear (Table 7.1).

Milk can regurgitate into the middle ear of infants, if they are not fed in head up position. This happens more with bottle feeding with a large hole in the nipple.

Functions of the Eustachian Tube

1. Ventilation and regulation of the middle ear pressure
2. Clearance of the middle ear secretions
3. Protection against reflux of nasopharyngeal secretions.

Note: For any successful middle ear surgical procedures, a normal and functioning Eustachian tube is essential.

Table 7.1: Differences between infant and adult Eustachian tubes		
Eustachian tube	***Infant***	***Adults***
Length	It is comparatively shorter	Adult tube is 33–38 mm
Direction	It is more horizontal. At birth angle with horizontal is 10 degrees	At the age of 7 years it becomes angulated by 45 degrees
Angulations at isthmus	Present	Absent
Lumen of tube	Wider	Narrower
Cartilaginous part	Flaccid	Rigid
Elastic recoil of cartilage	Ineffective	Effective
Volume of Ostmann's fat	Less	More

Tests for Eustachian Tube Function and Patency

The patency of the Eustachian tube is not the functional parameter for the tube. Various tests have been devised for assessment of the function and patency of the Eustachian tube. These include Valsalva manoeuvre, Toynbee's manoeuvre, Frenzel manoeuvre, nine-step inflation–deflation test, saccharine or methylene blue test, sonotubometry, scintigraphy, ultrasound and lately the tubo-manometry (TMM).

1. **Valsalva test:** This simple test is done by generating positive air pressure in the nasopharynx. The patient is asked to pinch his nose and mouth and blow air to generate a positive pressure in nasopharynx, which he is told to direct it towards the bilateral ears. With patients of normal patency, the Eustachian tube opens and the air is allowed into the middle ear which leads to outward bulging of the tympanic membrane. This movement is visualized by otoscopy. The test is contraindicated in case of active URI, middle ear infection and patients with scarred and thinned tympanic membrane. Patients may not always be able to perform the test.
2. **Tonybee's maneuver:** It is the reverse of the Valsalva manoeuvre. A negative pressure is generated in the nasopharynx by asking the patient to swallow with nose pinched. The effect is seen in the inward movement of the tympanic membrane.
3. **Politerization:** It is an obsolete test, which was performed to check for the patency in noncompliant patients. It uses a Politzer bag with a tube having an olive shaped nasal end. The nasal end is introduced in one of the nostrils and the nose closed. The patient is asked to continuously say "k k k" (क क क) as the air is pumped by compressing the Politzer bag (Fig. 7.6). The amount of air pumped is regulated by the number of fingers used to compress the bag. As with valsalva manoeuvre, otoscopy is done to check for the bulging of the tympanic membrane.
4. **Frenzel manoeuvre (nasopharyngeal pressure test):** This is a more effective manoeuvre than Valsalva and Tonybee's manoeuvre. Here the nasopharyngeal air is compressed by the muscles of mouth and tongue, resulting in opening of the Eustachian tube. It is of obvious advantage in jet-pilots wearing helmets and masks.

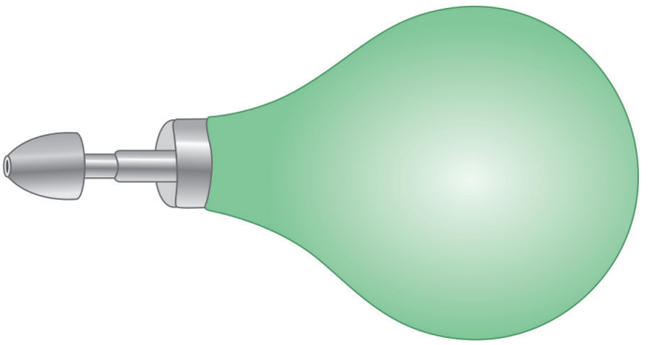


Fig. 7.6: Politzer bag

5. **Saccharine or methylene blue test:** It involves the placement of saccharine solution or methylene blue in the middle ear and time taken for sweet taste to appear or the dye to be seen in nasopharynx is noted. This test therefore provides evidence of clearance function of the Eustachian tube.
6. **Nine-step inflation and deflation tests:** In this test the middle ear pressure is recorded by tympanometry before and after application of a positive 300 daPa pressure to the ear canal and then before and after application of a 300 daPa negative pressure to the ear canal. Between steps, the subject is asked to swallow in an attempt to open the Eustachian tube and change middle ear pressure. A positive test is indicated by a minimum 10 daPa change in middle ear pressure after a swallow and excellent muscle-assisted Eustachian tube opening is indicated by an initial change to a more negative middle ear pressure after a swallow, followed by a return to 0 daPa after sequential swallows. The test is designed to measure the presence or absence of active Eustachian tube openings during swallowing at ambient pressures.
7. **Sonotubometry:** It is also known as *acoustic tube endoscopy*. A speaker is used to give white noise sounds to the nasal cavity (100 dB). The sound is conducted via the Eustachian tube if patent to the middle ear cavity. A microphone placed at the level of the external canal picks up the sound. The patient is then asked to perform the Valsalva or Tonybee's manoeuvre and the increased sound signals are

picked in case of patent Eustachian tube. The test is designed to measure the presence or absence of muscle-assisted Eustachian tube openings during manoeuvres at local ambient pressure.

8. **Tubomanometry (TMM):** It was described by Esteve in 2001. A controlled bolus of gas is applied to the nasopharynx during swallowing and the pressure sensor in the occluded ear canal records changes. The opening time of the Eustachian tube is recorded (R index). A 'R index' value of <1 indicates early opening of the tube, which is optimal (Fig. 7.7).
9. **Pressure chamber Eustachian tube test:** The set-up for the pressure chamber is expensive hence not popularly used worldwide. The patients is placed in a pressure chamber and exposed pressures from +2500 daPa to –1500 daPa. The pressures is changed up or down by 250 daPa and the subject is asked to swallow in an attempt to open the Eustachian tube as detected by sonotubometry and or a change in ear canal pressure.

   For perforated tympanic membrane, a small volume probe is used, which is connected to a pressure transducer, placed in the external auditory canal and the procedure repeated with continuous recording of middle ear pressure. Eustachian tube opening is recorded as a change in middle ear pressure.

DISORDERS OF EUSTACHIAN TUBE

1. Eustachian Tube Obstruction/Dysfunction (ETD)

Recent evidence shows that the gas diffusion between the middle ear and the blood mainly determines the middle ear pressure and the levels of all gases in the middle ear. The gases mainly involved are N_2, CO_2, O_2 and H_2O.

The pressure difference is created between the middle ear and the atmosphere due to the diffusion of gases into venous cell membranes of the middle ear. The carbon dioxide and oxygen have a tendency to quickly enter into the veins, hence producing a pressure vacuum in the middle ear compared to the atmospheric pressure. On the opening of the Eustachian tube the atmospheric pressure and middle ear pressure are again equated

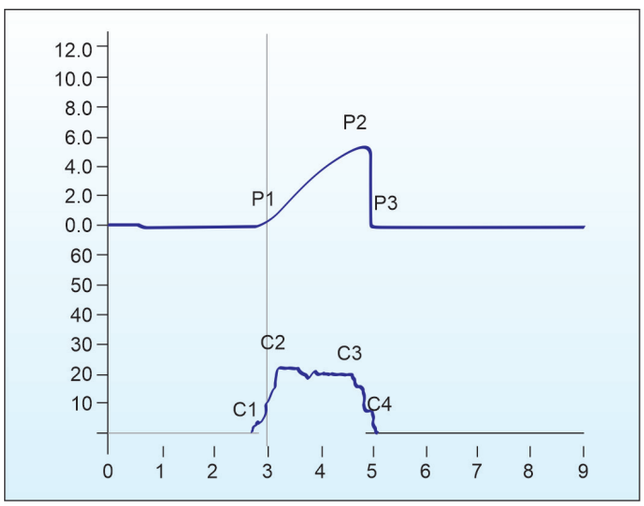


Fig. 7.7: TMM graph (Tubomanometry graph) upper tracing is the changes in the external ear, lower tracing is the changes in nasopharynx.

The Eustachian tube dysfunction leads to inability to equate pressures, The causes are either inflammatory or mechanical. Inflammatory causes within the lumen are either irritant or infective in nature. Irritant causes include allergies or chemical exposure. The infectious causes are viral or bacterial in nature. These causes lead to release of inflammatory mediators leading to oedema of the canal mucosa. The mechanical causes like a nasopharyngeal mass, lead to inability of Eustachian tube opening and thereby producing atmospheric and middle ear pressure difference.

Clinical Features

Hearing impairment or a feeling of fullness in the ear is the most common symptom of Eustachian tube dysfunction. The other symptoms are otalgia, tinnitus, balance disorders or vertigo.

On examination, the tympanic membrane is retracted with increased congestion of the handle of malleus and peripheral pars tensa. With continued retraction of the tympanic membrane, there is transudate collection in the middle ear leading to conductive hearing loss.

Causes

1. Hypertrophy of the nasopharyngeal adenoids. They cause mechanical obstruction to tubal opening and act as a source of pathogen to infect the middle ear and Eustachian tube mucosa.
2. Allergic tubotympanitis is a common cause of obstruction to tubal opening.
3. Acute viral and bacterial nasopharyngitis leads to the oedema of the tubal opening.
4. Sudden descent during flight, mines or deep-sea diving may lead to aero-otitis media or barotrauma.
5. Cleft palate patients, even after surgery, due to deficient structure and function of the tensor and levator muscles, freqeuently can cause tubotympanitis.
6. Extrinsic mechanical obstruction of the Eustachian tube may also be the result from a nasopharyngeal tumor. The invasion of the palate and pterygoid bone can interfere with tensor veli palatini muscle function and result in functional obstruction of the tube.
7. Nasal obstruction may also be involved in the pathogenesis of otitis media with effusion. Swallowing, when the nose is obstructed (due to inflammation or polyps) results in an initial positive nasopharyngeal air pressure, followed by a negative pressure phase, resulting in negative Eustachian tube pressure. Such a tube may get locked if the middle ear pressure is high.
8. Post-adenoidectomy scarring of tubal orifice or post syphilis or tuberculosis can lead to tubal dysfunction.

9. Malocclusion of the mandibular joint (*Costen syndrome*) can result in Eustachian tube dysfunction.
10. Injury to the mandibular branch of trigeminal nerve can result in functional obstruction of the Eustachian tube because of de-innervation of the tensor veli palatini.
11. Degenerative and metabolic diseases such as myasthenia gravis can alter the Eustachian tube function by affecting the muscles of the Eustachian tube.
12. If Eustachian tube dysfunction is associated with otitis media and the aetiology is obscure, it is considered to be idiopathic.

Treatment

1. Medical management includes use of nasal steroids, antihistaminics and nasal decongestants.
2. Use of Valsalva or Toynbee manoeuvres are suggested and have been found to be effective in mild cases.
3. **Ear popper** is an automated device used to deliver regulated air flow into the nasal cavity. It is easily administered at home by parents to children with otitis media with effusion.
4. **Balloon dilation Eustachian tuboplasty (BET):** It is a minimally invasive method to treat chronic Eustachian tube obstruction, which is done under general anaesthesia. The Eustachian tube opening is visualized by nasal endoscopy and a balloon catheter is inserted into it. The balloon is inflated with saline at a pressure of 3–5 bar for 2 minutes. This is repeated when needed (Fig. 7.8).
   Indications:
   - Inability to perform Valsalva
   - Having middle ear atelectasis
   - Chronic otitis media with effusion
   - Failed tympanoplasty
   - Uncomfortable sensation of pressure in the ears especially on having atmospheric pressure changes.

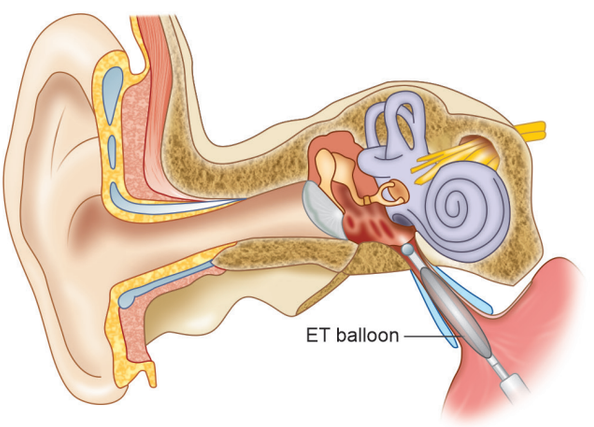


Fig. 7.8: Balloon dilatation Eustachian tuboplasty (BET)

5. **Laser tuboplasty:** A KTP-laser can be used to remove inflamed or hyperplastic tissue from the pharyngeal orifice. It is a safe and effective technique with a success rate of almost 70%.
6. **Myringotomy with tympanostomy tube insertion** is done for non-responsive patients to above techniques.

2. Patulous Eustachian Tube

A patulous Eustachian tube is also called *tuba aperta* or the *hyperpatent Eustachian tube*. It is of mainly two types: In the first type, the lumen of the tube remains anatomically patent even on rest, and in the other type the tube is semi-patulous which is closed at rest but having lesser airway tubal resistance. The second variant can develop with sudden weight loss, or any medical treatment.

Symptoms of the disease include autophony, breath synchronous tinnitus, pressure sensation in the ear and hearing loss. The examination mostly reveals the movement of the tympanic membrane with breath, which is easily seen on otoscopy. Though this is diagnostic of the disease but is not always present.

Tympanometry with reflex decay is useful to diagnose the disease, but this requires an intact tympanic membrane. Tubomanometry (TMM) does not require the intact tympanic membrane therefore is valuable in COM cases with patulous ET.

Treatment

1. Oestrogen nasal drops have been found to induce swelling in tubal opening. Similarly, potassium iodide (KI) solution has also been found effective for swelling induction.
2. Myringotomy and grommet insertion have the ability to reduce symptoms but are not definitive treatment.
3. Patulous tube reconstruction can be done by cartilage **(Kobayashi plug)** or **Vox implants** into the torus tubarius. This is done via endoscopic nasal approach to the nasopharynx.

Otitic Barotrauma

Otitic barotrauma is a pathological condition of the ear caused by sudden large air pressure changes in the environment. The common situations where barotrauma is seen are scuba diving and during air flights. There are three types of otitic barotrauma manifestations:

1. Compression injuries (Descent)
2. Injuries at high pressure
3. Decompression injuries.

1. *Compression Injuries*

A. External ear barotrauma/external ear squeeze

This is seen when a pocket of air is trapped in the external auditory canal (EAC) behind the ear wax, foreign bodies, ear plugs or exostosis. As we know, with descent of the aircraft the ambient air pressure increases. With a normal

functioning Eustachian tube, the middle ear pressure will increase and get equalised with outside pressure. But the air trapped in EAC, will be at negative pressure in EAC compared to middle ear while rapid descent. The tympanic membrane will therefore bulge laterally into the EAC. With sufficiently increased bulge, it can result in a tympanic membrane perforation. In severe cases there might be ossicular discontinuity. It can be prevented by not using earplugs and head gear during descent of the flight.

B. Middle ear barotrauma/middle ear squeeze

This is most commonly seen with Eustachian tube dysfunction. This occurs during descent of flight or ascend in scuba diving. In this situation, the EAC pressure will increase but due to the dysfunction of the Eustachian tube the middle ear pressure will not get equated with EAC pressure.

Initial symptoms are sensation of blocked ear and progressive otalgia. There is conductive hearing loss due to decreased tympanic membrane compliance. The tympanic membrane appears red and retracted. The intratympanic haemorrhage may occur.

Further there is development of middle ear effusion and sometimes hemotympanum. The perforation of the tympanic membrane may occur and this is followed by sudden severe ear pain.

It can be prevented by:

1. Not sleeping at the time of descent. While sleeping the number of swallowing movements are decreased and hence the opening of the Eustachian tube is minimised.
2. Avoid taking flights if suffering from upper respiratory tract infection.
3. Swallow repeatedly during flight descent. Use of chewing gums or candies is helpful.
4. Use of Valsalva manoeuvre can benefit a lot of patients.
5. Otovent device can be used if a patient finds it difficult to perform effective Valsalva manoeuvre.
6. Slowing the ascend rate in scuba diving.
7. Eustachian tube balloon dilatation is increasingly becoming the treatment of choice for those with recurrent flying equalization problems.

Treatment

No treatment is indicated with minimal signs of barotrauma. If significant signs of the barotrauma are seen, then oral and nasal decongestants are used. In case of perforation or ossicular trauma, tympanoplasty is indicated.

C. Inner ear barotrauma

Increase in the ambient pressure is reflected in the intracranial pressure and CSF pressure. The inner ear fluid pressure gets equalized with the CSF pressure via the cochlear and vestibular aqueduct. In the situation of Eustachian tube dysfunction, the middle ear pressure is not equated and it becomes negative compared to inner ear pressure. If this pressure difference is large, then it results in inner ear damage.

Two types of injuries are seen in this condition. Implosive injury when the middle ear pressure is very high and results in inward movement of the round window membrane causing rupture or disruption of the stapes footplate. In explosive type there is negative middle ear pressure and high inner ear pressure due to raised ICT, there is outward movement of the round window or the stapes footplate resulting in their rupture. Initial cases have haemorrhage in the inner ear. Further there are membranous labyrinth tears which finally may result in **perilymphatic fistula**.

Clinical features

The symptoms are sudden onset of vertigo, hearing loss and tinnitus. Tulio phenomenon and positive fistula are seen in perilymphatic fistula.

On examination, the tympanic membrane might be perforated. Nystagmus is present usually towards the opposite side. The hearing loss is commonly SNHL on audiometry.

Treatment is initially decided by the degree of the hearing loss.

1. Bed rest with head elevated at 30 degrees.
2. Daily audiometry is done for any deterioration of hearing loss.
3. Oral steroids are often recommended, though intratympanic steroids are favoured now.
4. If patients do not improve after 5 days of treatment or there is progressive increase in hearing loss, exploratory tympanotomy is done to find the defect and its subsequent closure is done.

2. *Injuries at High Pressure*

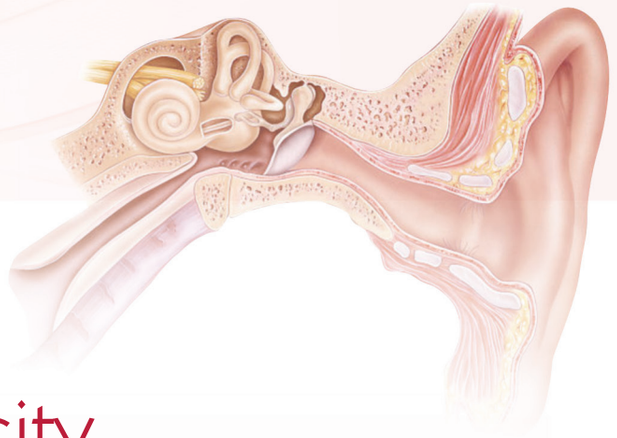
It is described as *high-pressure nervous syndrome*, which is caused by exposure to extremes of high pressure. Patients have dizziness, tiredness and tremors. Further increased pressure may result in ataxia and myoclonus. There is permanent vestibular damage but there is no hearing loss.

3. *Decompression Injuries*

Decompression sickness in divers occurs when dissolved gas, usually nitrogen, comes out of solution on ascent and forms bubbles in soft tissues and blood vessels, causing various symptoms including sensorineural deafness. This is a condition caused due to asymmetrical increase in middle ear pressure seen in divers and air crew. It usually starts within 2 minutes of ascent in divers. Divers have vertigo (alternobaric) and feel a tumbling sensation, or a tilting of their surroundings, rather than simple rotation. It has a maximum duration of 10 minutes and there is no permanent damage with decompression injuries.

Chapter

8



Ototoxicity

Ototoxicity is defined as the tendency of few therapeutic agents and chemical substances to cause functional impairment and cellular degeneration of the tissues of the inner ear and vestibular division of the VIII cranial nerve. Synergistic side effects of the drug might cause ototoxic damage. Therefore, ototoxic drugs used together, might cause damage to inner ear at lower doses than given alone, e.g. aminoglycosides with loop diuretics.

The primary effects of the ototoxic drugs include tinnitus, hearing loss and disequilibrium.

1. Tinnitus is high frequency at onset, often continuous and usually of pure tone.
2. Disequilibrium is caused by vestibulotoxic drugs and is usually symmetrical. The patient is unsteady with gross vestibular reduction of function. Vertigo and nystagmus are not seen.
3. Hearing loss is initially high frequency but subsequently it involves speech frequencies. Early diagnosis of ototoxicity can be done using high frequency audiometry.

EFFECTS OF FEW OTOTOXIC DRUGS

1. Aminoglycosides

Aminoglycosides can cause toxicity of the vestibular or cochlear systems of the inner ear in up to 10% of patients receiving these drugs intravenously. Gentamicin, tobramycin and streptomycin are preferentially vestibulotoxic, whereas amikacin and kanamycin are primarily cochleotoxic. The aminoglycosides cause loss of outer hair cells especially in the basal turn. The risk of ototoxicity increases with total cumulative exposure. Patients requiring aminoglycoside therapy, the duration of therapy should be minimized and monitoring of hearing thresholds should be done with pure tone audiometry (Table 8.1). Kanamycin is used as a second-line drug for treatment of multidrug resistant tuberculosis and can result in bilateral profound hearing loss. In all such patients regular surveillance by pure tone audiometry must be done to diagnose hearing loss at the earliest and stop the offending drug.

2. Cisplatin

It is an anticancer drug which causes strial atrophy hence leading to hearing loss. The damage is dose dependent.

3. Salicylates

These produce ototoxicity which is thought to be reversible but few patients might experience permanent damage. The hearing loss is mild to moderate and is symmetrical. Tinnitus is also an associated feature.

4. Loop Diuretics

These drugs like furosemide, bumetanide and pireatnide are responsible for the ototoxic effect. The effects include deafness and sometimes vertigo.

Table 8.1: List of ototoxic drugs

Ototoxic drug class	*Examples*
Aminoglycosides	Streptomycin, amikacin, tobramycin, gentamicin, kanamycin, capreomycin
Platinum-based chemotherapy	Cisplatin, carboplatin, oxaliplatin
Loop diuretics	Furosemide, torasemide, bumetanide, piratenide
Other antibiotics	Erythromycin, vancomycin
Antimalarials	Quinine
Salicylates	Aspirin
Phosphodiesterase type 5 inhibitors	Sildenafil, tadalafil, vardenafil

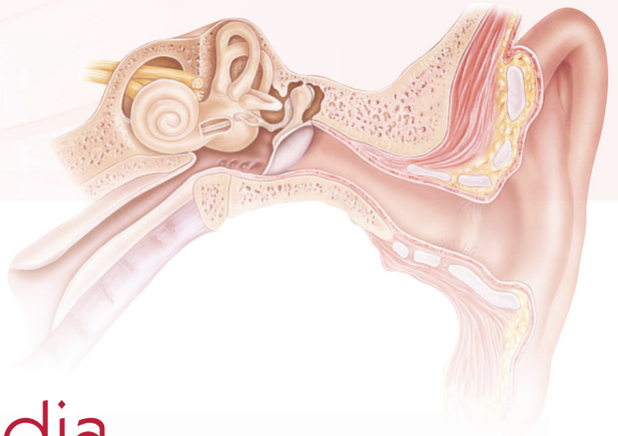
The environmental pollutants which can cause ototoxicity are mercury, carbon monoxide, manganese, styrene, xylene, toluene and arsenic.

MANAGEMENT

1. Prevention is better than cure. Avoid use of ototoxic drugs for prolonged periods. Environmental ototoxic agent contact should be avoided by use of masks and better ventilation conditions. Hazardous chemicals should be kept away from general contact.
2. Initiation of therapy with ototoxic drugs should be preceded by pure tone audiometry and subsequently hearing levels should be regularly monitored.
3. Early symptoms like tinnitus and mild hearing loss should be taken seriously and use of ototoxic drugs is immediately withheld.
4. Steroid and hyperbaric oxygen therapy have been found to be useful in early hearing loss cases.
5. Permanent hearing losses can be overcome by use of hearing aids.

Chapter

9



Otitis Media

PE28.4: Discuss the aetiopathogenesis, clinical features and management of acute otitis media (AOM).
EN4.3: Elicit document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of ASOM.

Otitis media includes a group of inflammatory and infectious conditions which affect the middle ear. Otitis media can be classified as:

A. **Acute otitis media (AOM):** Sign and symptoms of middle ear infection for <3 weeks.
B. **Sub-acute otitis media:** Sign and symptoms of middle ear infection for 3 weeks to 3 months.
C. **Chronic otitis media (COM):** Sign and symptoms of middle ear infection for >3 months. Usually there is a perforation of the tympanic membrane. It also includes tubercular otitis media and syphilitic otitis media.
D. **Otitis media with effusion (OME)**.
E. **Adhesive otitis media**.

ACUTE OTITIS MEDIA

Acute otitis media (AOM) is the inflammation of the middle ear cleft of rapid onset due to infective causes, associated with middle ear effusion. It is more commonly seen in children, especially less than 2 years of age, though it can occur in any age group.

It is broadly divided into four subgroups:

1. *Sporadic AOM*: In this episode of AOM are infrequent and are associated with upper respiratory tract infections.
2. *Resistant AOM*: It is the persistence of signs and symptoms of middle ear infection beyond 3–5 days.
3. *Persistent AOM*: It is the persistence or recurrence of symptoms and signs within 6 days after a course of antibiotics.
4. *Recurrent AOM*: It is the presence of three or more attacks of AOM within 6 months or 4–6 episodes in 12 months or continuous persistence of symptoms.

Aetiology

Acute otitis media is caused due to viral, bacterial or coexisting causes. The bacteria most commonly involved are *H. influenza*, *Streptococcus pneumonia*, *Morexella catarrhalis* and *Streptococcus aureus*. Viral infections also contribute to the AOM. The most common viral pathogens include the respiratory syncytial virus (RSV), corona viruses, influenza viruses, adenoviruses, and picorna viruses.

Risk Factors

1. Eustachian tube dysfunction due to adenoid hypertrophy, upper respiratory tract infection or sinusitis.
2. Immunity: Low levels of IgG_2 subclass have been reported with increased incidence of AOM because delayed maturation of anti-pneumococcal antibodies in such patients. Decreased expression of TNF, interleukins and cytokinins have been reported as risk factors.
3. Syndromes: Down syndrome and Turner syndrome children have increased incidence of AOM
4. Breast feeding for less than 3 months is a risk factor
5. Passive smoking
6. Iron deficiency anaemia.

Routes of Infection

1. The most common route of spread of infection is via the Eustachian tube. This is especially common in children, due to the Eustachian tube being more horizontal, shorter and patulous. This risk increases with careless bottle feeding or use of pacifiers.
2. The infection may directly travel to the middle ear via the tympanic membrane perforation.
3. Haematogenous route is also a possible route for spread of infection to the middle ear.

Pathophysiology, Clinical Features and Stages of AOM

1. **Stage of tubal occlusion:** There occurs an obstruction to the pharyngeal end of the Eustachian tube due

to oedema by infection or allergy. This leads to negative middle ear pressure formation, which causes tympanic membrane retraction. There can be an accompanying middle ear effusion.

Clinical features

Patient has complaints of ear block sensation with hearing loss.

On examination, the tympanic membrane is retracted (light reflex is lost and lateral process of malleus becomes prominent).

2. **Stage of pre-suppuration:** If the Eustachian tube obstruction persists, bacterial infection of the middle ear ensues. This leads to hyperaemia of middle ear mucosa with inflammatory exudate formation.

   Clinical features

   There is an increase in ear ache, which is of severe and throbbing nature. There is also hearing loss. Fever may develop and children may become irritable and stop feeding.

   On examination, the tympanic membrane is congested. It shows a typical ***cart-wheel appearance*** formed by engorged blood vessels supplying the membrane. Ultimately, the entire tympanic membrane gets congested (Fig. 9.1).

3. **Stage of suppuration:** Further, there is a formation of pus inside the middle ear. The collected pus may lead to increase in intratympanic pressure. This causes bulging of the tympanic membrane. The infection may spread to the mastoid air cell system.

   Clinical features

   The patient has a severe ear ache with fever and hearing loss.

   On examination, the tympanic membrane is congested and bulging (Fig. 9.2).

4. **Stage of resolution:** With time, necrosis of part of the tympanic membrane occurs which leads to perforation of the tympanic membrane (Fig. 9.3).

   Clinical features

   Symptoms of pain are relieved and the general condition of the child improves.

   On examination, a perforation is seen in the tympanic membrane. The perforation is commonly formed in the inferior three-third because it is the most dependent part of the middle ear and hence the area of maximum pus collection. There is a mucopurulent blood-tinged discharge seen in the middle ear and the external auditory canal. The middle ear mucosa is hypertrophied and congested.

5. **Stage of complication:** In cases where above stages are not treated timely or when the immunity of the patient is compromised along with high virulence of causative organisms, the infection might spread beyond bony confines of middle ear to cause complications like acute mastoiditis, petrositis and intracranial complications.

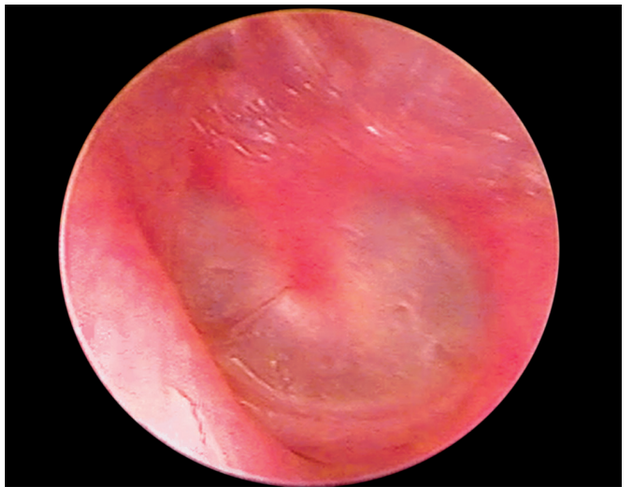


Fig. 9.1: Stage of pre-suppuration. The tympanic membrane appears congested

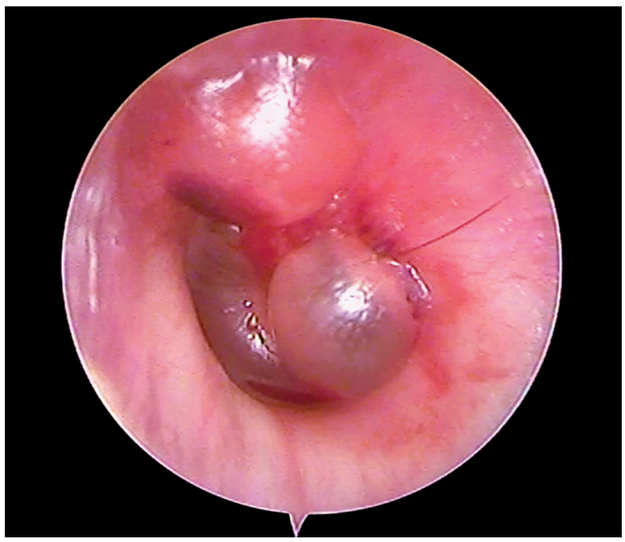


Fig. 9.2: Stage of suppuration: Congested tympanic membrane with a bulge with impending rupture



Fig. 9.3: Stage of resolution: Tympanic membrane perforation following AOM (Inferior division)

Clinical features

The complications may lead to rapid deterioration of a patient's general condition, along with worsening symptoms of ear pain, fever, ear discharge, and sometimes vomiting. Facial nerve palsy may occur, especially so in cases with dehiscent facial nerve in the tympanic segment.

Management

1. Analgesics and antipyretics are the first line of treatment, because most infections are viral to start with and are expected to recover fast.
2. Antibiotics are mainly indicated in children less than 6 months of age or who do not improve in 2–3 days of watchful waiting.

   Also the antibiotics are indicated on 1st day of infection, when the patient have:
   a. Pyrexia and vomiting
   b. High risk children like the children with craniofacial abnormalities or with immunodeficiency.

   The antibiotics of choice are amoxicillin, azithromycin and cefuroxime axetil.
3. Topical antibiotic drops may be started in conditions of tympanic membrane perforation with ear discharge.
4. Myringotomy is reserved for patients having impending perforation. This will avoid spontaneous rupture of the tympanic membrane and will relieve the pain with drainage of the pus.
5. Adenoidectomy is indicated in cases of recurrent AOM, who have failed medical management.

The use of antihistamines and decongestants are not recommended for AOM.

Outcomes

1. Complete rapid resolution with or without antibiotics
2. Persistent infection after short course of antibiotics
3. Recurrence of AOM
4. Progression to tympanic membrane perforation
5. Complications may be seen in a few cases especially in children with compromised immunity. The complications include: acute mastoiditis, facial nerve palsy, labyrinthitis, meningitis, extradural empyema, lateral sinus thrombophlebitis, brain abscess and otitic hydrocephalus.

EN2.14: Demonstrate the correct technique to instilling topical medications into the ear.

How to use eardrops

- Hands are sanitized or washed with soap and water.
- If eardrops are cool, then they should be warmed holding them your hand for one to two minutes. This will help decrease discomfort.
- After removal of the cap from the bottle, it should be placed on a clean and dry surface.
- The droppers should be checked for any cracks.

Putting in the drops

1. The head is positioned so that the concerned ear faces upward. If the patient instils the drops himself, then it is best by sitting or standing upright and head tilted to the side.
2. If the attendant gives the drops, then the patient tilts the head or lies down with concerned ear upwards (Figs 9.4 and 9.5).
3. The pinna is pulled upwards and backwards for adults and for children the pull is inferiorly and backwards.
4. The bottle is squeezed to instil the drops into the ear.
5. Gently pull the earlobe up and down to allow the drops to run into the ear.
6. Keep the head tilted for about two to five minutes so the drops can spread into the ear.
7. Put the cap back on the bottle.
8. Store the bottle as directed by the label or by your doctor or pharmacist.

Don't use a dropper that is cracked, chipped, or dirty. Don't share ear drops. Check expiry of the drops before use.

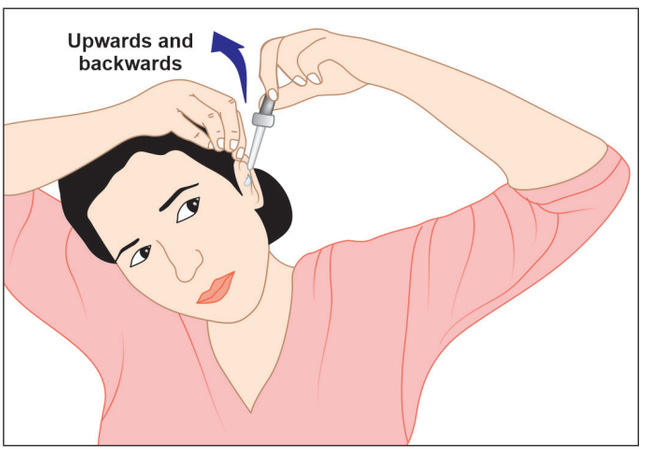


Fig. 9.4: Drop instillation in adults

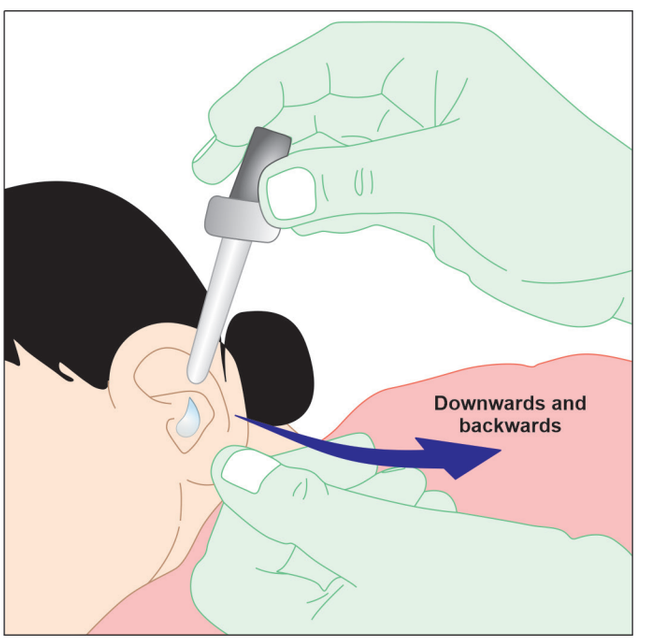


Fig. 9.5: Drop instillation in children



Chapter

10

Otitis Media with Effusion

EN4.5: Elicit document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of OME.

Otitis media with effusion (OME) is also known as Secretory otitis media (SOM) or "glue ear".

It is characterised by accumulation of mucoid secretions within the middle ear cavity or occasionally in the mastoid air cell system. This accumulation is chronic and usually has a history of more than three months.

PATHOGENESIS

The age group which is most commonly affected is 1–8 years, though it can be seen in adults. In children, the Eustachian tube is shorter, more horizontally oriented, and less functionally mature, hence predisposing children to OME.

The middle ear is normally lined by flat cuboidal respiratory mucosa. This is replaced by the thicker pseudostratified mucous secreting mucosa, which frequently contains goblet cells. This is less efficient to move the secretions from the middle ear to nasopharynx via the Eustachian tube. The underlying submucosa also becomes oedematous with dilated vessels.

Various factors which cause fluid accumulation in middle ear are:

1. **Ascending infection from Eustachian tube:** Earlier the serous fluid was thought to be sterile. Now respiratory bacteria like *S. pneumoniae*, *H. influenzae* and *Moraxella* have been isolated from the secretions by polymerase chain reaction (PCR). Biofilms have been recently diagnosed as probable organisms in OME formation. The recurrent infections of sinuses, oropharynx and nasopharynx may also lead to OME.
2. **Abnormal function of Eustachian tube:** This may occur due to adenoid hypertrophy, craniofacial abnormalities like Down syndrome or nasopharyngeal mass (benign or malignant).
3. **Allergy:** There is a strong correlation of OME with allergy. Serum IgE levels have been found to be elevated in many such patients.
4. **Gastro-oesophageal reflux:** It has been shown to be a risk factor for OME in children. This is due to silent (without any symptoms) reflux in children. In one study, pepsinogen has been isolated from 84% of OME effusions.
5. **Acute otitis media:** The recurrent attacks of AOM significantly increases the risk of OME.
6. **Immune deficiency:** It can lead to OME in children. There is strong association of OME with recurrent sinusitis, bronchitis or gastrointestinal (GI) abnormalities, in such patients.
7. **Secondary Eustachian tube block:** OME is also frequently seen in patients with prolonged nasal intubation or nasogastric tubes.

Inflammation of the mucosa of the Eustachian tube orifice due to various conditions such as upper respiratory infection (URI) or allergy and improper functioning of the Eustachian tube musculature lead to negative middle-ear pressure.* Thus, when the Eustachian tube opens, bacteria and viruses from the nasopharynx are drawn into the middle-ear space and generate an inflammatory response.

There is no difference among the sex groups or races. Parental smoking >20 cigarettes/day does contribute to some additional risk for OME.

HISTORY

The symptoms of patients include:

1. **Hearing loss:** It is the most common symptom but may not be noticed in patients with conductive hearing loss of less than 10 dB. The air-bone gap is usually between 10 and 40 dB. The parents or school teacher might complain about the child's lack of attention in home or class.

*Normally contractions of the tensor veli palatini and levator veli palatini dilate and open the auditory tube.

2. **Speech and language development delay:** The delay in speech and language might be the only complaint in very young children. There can be long-term outcomes of the OME leading to cognition defects.
3. **Balance problems:** There may be significant balance problems in few children which are cured on successful OME treatment.

EXAMINATION

The latest guidelines recommend otomicroscopy/oto-endoscopy along with pneumatic otoscopy as essential for OME examination. The otoscopic picture will show:

1. Dull tympanic membrane with absence of light reflex.
2. There may be a bulge in the tympanic membrane due to the fluid in the middle ear, which might show air bubbles (Fig. 10.1).
3. Retraction of the pars tensa or an attic retraction.
4. The pneumatic otoscopy **(siegelization)** will reveal decreased movement of the tympanic membrane.

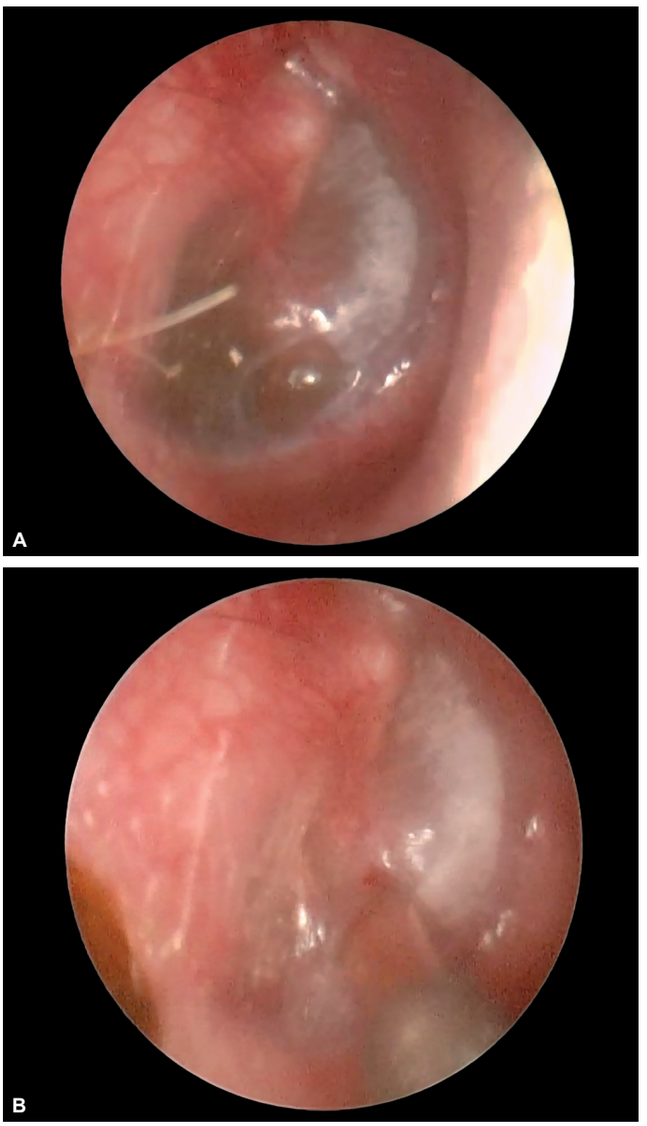


Fig. 10.1: Secretory otitis media

TESTS

1. **Tuning fork test:** As the OME occurs more in younger children, therefore they do not give reliable responses on tuning fork tests. In adults these tests will reveal conductive hearing loss.
2. **Tympanometry:** It shows a type B curve (due to fluid causing reduced tympanic membrane mobility) or sometimes type C curve (due to negative pressure in middle ear indicating Eustachian tube dysfunction).
3. **Acoustic reflex:** The reflex will be elicited at higher threshold due to conductive hearing loss or it might be absent.
4. **Audiometry:** There will be an air-bone gap indicating conductive deafness.
5. Radiological investigations are not desired.

MANAGEMENT

1. Medical therapy for OME including systemic antibiotics, systemic or intranasal steroids, antihistamines, mucolytics and decongestants have not been proven to be beneficial in recent studies, hence are now **not recommended**.
2. Manage the child with OME who is not at risk, with watchful waiting for 3 months from the date of effusion onset (if known) or 3 months from the date of diagnosis.
3. At present the best non-surgical approach to OME is **auto-inflation** (Valsalva manoeuvre).
4. Re-evaluate the child at 3–6 month intervals, until the effusion is no longer present.
5. **Tympanostomy tube insertion (Grommet insertion):** If OME persists with significant hearing loss, or structural abnormalities of the eardrum or middle ear are suspected, surgical intervention is done.
   a. In children less than 4 years of age only tympanostomy tube insertion is performed (Figs 10.2 and 10.3).
   b. Tympanostomy tube insertion with adenoidectomy for such children should only be performed if there is indication for adenoid removal, such as nasal obstruction or persistent nasal discharge due to chronic adenoiditis, is present. Currently there is **no role of myringotomy with aspiration**, i.e. without grommet insertion.
6. **Tympanostomy tube insertion with adenoidectomy:** It is done when surgery is performed in a child aged 4 years or older and there are two or more episodes of OME.
7. A child, who displays speech and language delay secondary to recurrent otitis media with effusion, should undergo **prompt myringotomy** with or without tympanostomy tubes (Fig. 10.3).
8. Additionally, certain subsets of children are more likely to need prompt myringotomy. These include:
   a. Children with permanent hearing loss independent of OME.

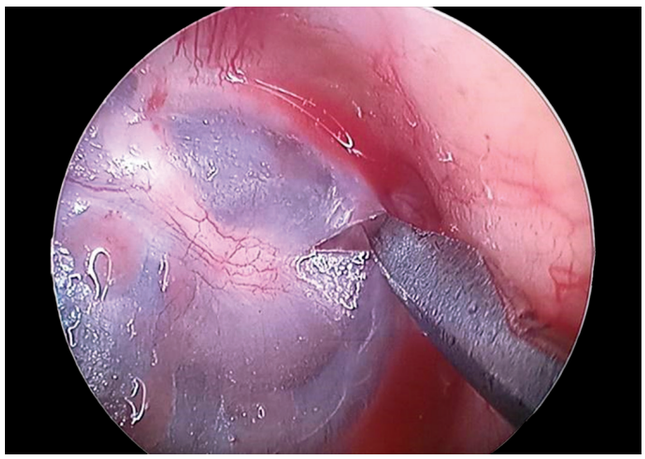


Fig. 10.2: Myringotomy done with sickle knife in right tympanic membrane (Anteroinferior quadrant)

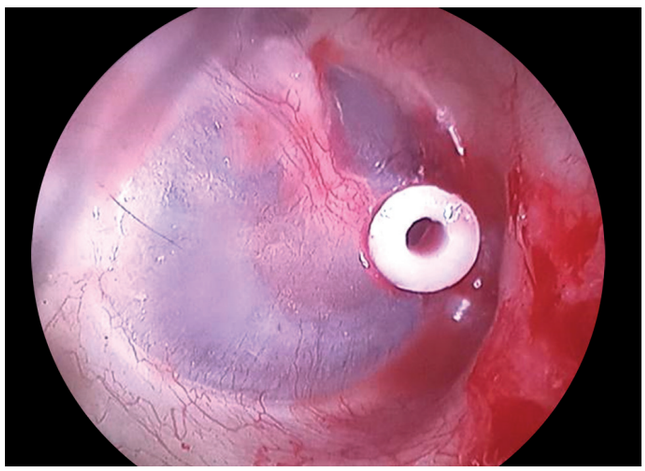


Fig. 10.3: Myringotomy with grommet insertion

   b. Children with autism-spectrum disorder and other developmental disorders.
   c. Children with syndromes (e.g. Down syndrome) or craniofacial disorders that result in Eustachian tube dysfunction.
   d. Children who are blind or have uncorrectable visual impairment.
9. In few cases of persistent OME, hearing aids may also be provided, so that the child does not develop cognition problems due to hearing loss.

SEQUELAE OF OME

1. Tympanosclerosis
2. Ossicular necrosis: Long process of incus and stapes suprastructure may get necrosed leading to conductive hearing loss.
3. Atrophic tympanic membrane: This occurs due to loss of fibrous layer of tympanic membrane.
4. Cholesterol granuloma.

There is no evidence of OME leading to cholesteatoma formation, as was earlier thought.

EN410: Observe and describe the indications for and steps involved in observe and describe the indications for and steps involved in myringotomy.
AN40.5: Explain anatomical basis of myringotomy.

MYRINGOTOMY PROCEDURE

Myringotomy is a procedure of making an incision through the layers of the tympanic membrane, so as to open up the middle ear space externally. This procedure may be combined with insertion of the tympanostomy tube (ventilation tubes) into the tympanic membrane so as to allow consistent drainage and ventilation of the middle ear space.

Indications

1. The most common indication is for children with OME of more than 3 months duration.
2. Children with recurrent acute episodes of otitis media (>4–5 infections in 6 months) also benefit from myringotomy.
3. AOM with bulging tympanic membrane when associated with severe ear ache.
4. AOM with facial nerve palsy: The paralysis of the nerve is due to either raised pressure, osteitis or acute inflammation of the nerve when the fallopian canal is dehiscent. The most commonly involved part of the facial nerve is the tympanic segment. The myringotomy is indicated as an early intervention in all such cases.
5. AOM with acute labyrinthitis.
6. Aero-otitis media.

Ventilation Tubes

Material: The ventilation tubes commonly made of teflon. titanium, gold or silver oxide coated and silicone (these inhibit the biofilm formation) are being used.

Types: Shepard tubes stay *in situ* for a shorter period than T-tube design but have lesser complications (Figs 10.4 and 10.5).

Technique

1. The procedure is performed by a permeatal approach in a sterile environment.
2. Ear speculum of the largest size is introduced into EAC while under an operating microscope. Otoendoscopes can be used, which does not require speculum insertion.
3. Local anaesthesia is infiltrated, if the procedure is done for adults under LA. In children the procedure is performed under general anaesthesia.
4. The EAC is cleaned of wax and debris.
5. Either the anteroinferior quadrant or the posteroinferior quadrant of the tympanic membrane can be used for myringotomy. A myringotomy knife (preferred in microscopic surgery due to its angulation; sickle knife can alternatively be used, especially in endoscopic ear surgery). The incision should be of approximately 3–5 mm in length. The incision made in the tympanic

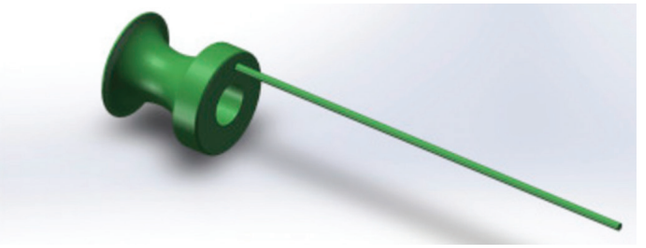


Fig. 10.4: Shepard type of grommet



Fig. 10.5: Goode T–tube (Ventilation tube)

membrane is either radial or circumferential. Both types of incision have similar extrusion rates of grommets (Figs 10.6 and 10.7).

6. Site of incision is decided on the basis of the time the ventilation tube is desired *in situ*.
   - The anteroinferior quadrant is the preferred site for grommet insertion because compared to posteroinferior quadrant, the extrusion rate is delayed. Also, the chances of residual perforation are lesser.
   - In cases of AOM, the posteroinferior quadrant is favoured because it is usually the site of maximum bulge and is the most dependent site for pus collection in tympanic cavity.
   - The posterosuperior quadrant is not preferred because of chances of damaging the incudostapedial joint.
   - For long-term ventilation anterosuperior quadrant is used because of the delayed extrusion of the grommet as the migration of epithelium is slowest in this region.
7. After incision, in OME patients, it might be difficult to suck out the thick fluid sometimes. In such cases an additional small incision may be given in the tympanic membrane which will allow the air to enter the middle ear and the glue can be then easily sucked out (beer can principle).
8. Topical ear drops may be used along with the insertion of ventilation tubes, to reduce the risk of blockage of the tubes due to blood and mucus, and to minimize risk of local infection.

Complications

A. Intraoperative

1. Displacement of ventilation tube into middle ear.
2. Damage to the ossicular chain.

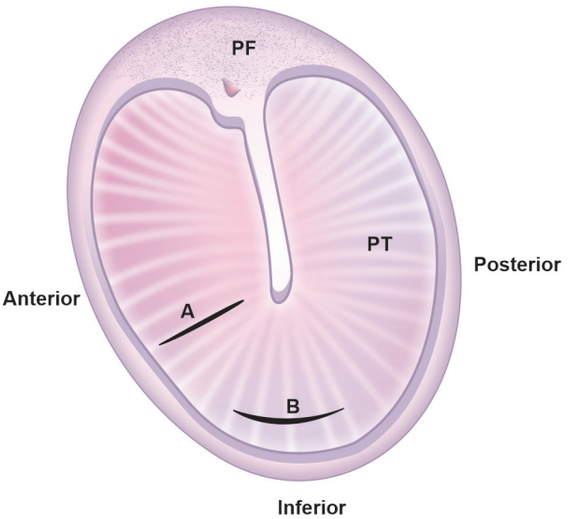


Fig. 10.6: Incisions for myringotomy: (A) Radial incision; (B) Circumferential incision

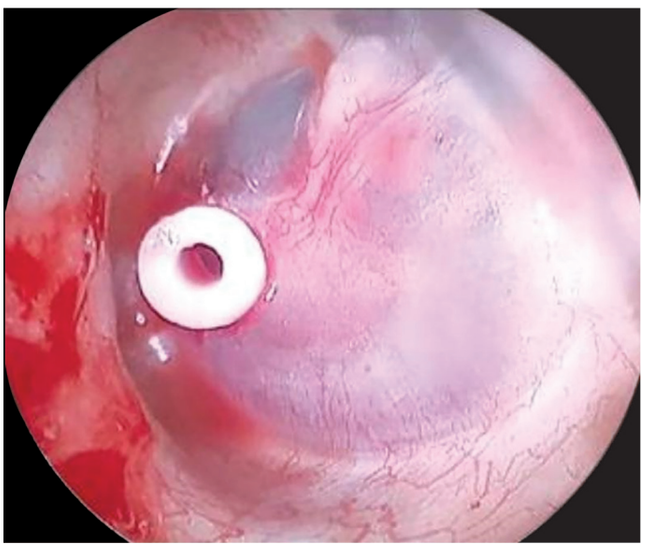


Fig. 10.7: Grommet *in situ*

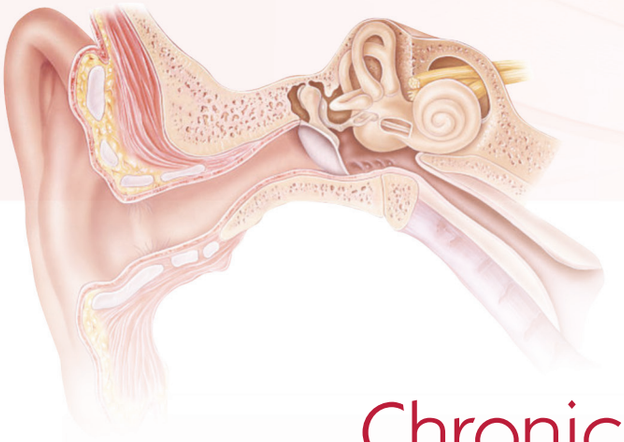
3. Rarely bleeding from a high dehiscent jugular bulb when injured.

B. Postoperative

1. Blockage of the lumen of grommet.
2. Local infection at insertion site.
3. Granulation formation at the insertion site.

C. Late

1. Hearing loss may occur due to tympanic membrane atrophy at the site of insertion.
2. Tympanosclerosis.
3. Persistent perforation may occur due to non-closure of defects after grommet extrusion.
4. Pars tensa atrophy.



Chapter

11

Chronic Otitis Media

EN4.6: Elicit, document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of a discharging ear.
EN4.7: Elicit document and present a correct history demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of mucosal type of CSOM.
AN40.4: Explain anatomical basis of otitis media.

The chronic otitis media (COM) is defined as inflammation of the middle ear cleft which results in permanent abnormality of the tympanic membrane involving pars tensa or flaccida. It was originally called chronic suppurative otitis media. This term is no more used because COM does not necessarily have suppurative findings. The earlier used terminology of tubotympanic/safe disease and atticoantral/unsafe disease are also no longer used, because these are incorrect and misleading.

Following is the latest classification of chronic otitis media (COM) with their otoscopic findings:

COM classification	*Otoscopic findings*
1. Active (mucosal) COM	Permanent defect of the pars tensa with ear discharge and congested middle ear mucosa.
2. Inactive (mucosal) COM	Permanent perforation of the pars tensa without discharge and normal middle ear mucosa.
3. Healed COM	Thinning of pars tensa without perforation. It can have associated tympanosclerosis.
4. Inactive (squamous) COM	Retraction pocket of pars flaccida or epidermisation.*
5. Active (squamous) COM	Retraction pocket with squamous debris or cholesteatoma.

*Epidermisation is an advanced type of retraction with replacement of middle ear mucosa by keratinizing squamous epithelium.

The recurrent episodes of acute otitis media (AOM) and otitis media with effusion(OME) in children are the leading causes of chronic otitis media. The COM is characterized by a permanent tympanic membrane perforation. The perforation becomes permanent due to epithelisation of its margins. Other features of COM are hearing loss, frequent episodes of ear discharge and middle ear pathologies like granulation tissue, cholesteatoma and cholesterol granuloma.

AETIOLOGY AND EPIDEMIOLOGY

The disease equally involves both the sexes. COM is a common disease in all developing countries with low socioeconomic status and poor hygiene. The higher incidence of recurrent upper respiratory tract infections, lead to increased episodes of acute otitis media (AOM). Inadequate treatment of these episodes of AOM, which along with otitis media with effusion (OME), significantly contribute towards increased incidence of COM. Moreover, recurrent episodes of AOM and OME lead to degeneration of the outer and inner fibrous layer of the tympanic membrane (TM), thereby making it less elastic and more susceptible to perforation and retraction.

The role of gastro-oesophageal reflux disease (GERD) in COM has been suggested. Other causes are craniofacial abnormalities, autoimmune disease and immune deficiency.

Organisms* involved in the middle ear infections are mainly gram-negative aerobes like *Pseudomonas aeruginosa*, *Proteus*, *E. coli*. Other being *S. aureus* and anaerobes.

TYPES OF COM

A. Inactive Mucosal Type of COM

Chronic mucosal otitis media usually has a milder course as compared with the squamous type of disease and has lesser frequency of complications. The inactive mucosal type has a dry perforation of

*_Biofilms_ are now being recognised as an important factor in aetiology chronic infections of the ear especially in relation to the squamous otitis media. 80% of cholesteatoma patients show the presence of biofilms.

the pars tensa.* The patient usually presents with the history of hearing loss or the small perforation may be detected on routine ear examination.

B. Active Mucosal Type of COM

This type of disease is characterised by a pars tensa perforation and episodes of ear discharge. The episodes of discharge are related with the upper respiratory tract infections reaching the middle ear cleft via the Eustachian tube or may be due to infection via the permanent perforation in the tympanic membrane.

Aetiology

1. **Environmental:** There is a close correlation between low socioeconomic status and overcrowding with the COM.
2. **Genetic:** The mastoid air cell system has been seen to be sclerosed in COM patients. This happens because of the disease process or it is the primary factor for disease formation is not established.
3. **Recurrent attacks of AOM:** It leads to thinning of the fibrous layer or a permanent defect in the tympanic membrane.
4. **Infections:** This has been established that bacteria in ears with COM are secondary invaders of the mucosa which has been inflamed by other factors like allergy, rather than the primary cause for disease.
5. **Upper respiratory tract infections:** These have a viral aetiology to start with, but later develop superadded bacterial infection which reach the middle ear cleft via Eustachian tube.
6. **Allergy**
7. **Nasal obstruction from adenoids**
8. **Autoimmune disorders:** Rheumatoid arthritis has shown a correlation with COM.
9. **Eustachian tube dysfunction**
10. **Ciliary dysfunction**
11. **Cigarette smoke and air pollution**.

Clinical Findings

Perforation of the pars tensa: The tympanic membrane may have perforation of variable size. Usual presentation is a central perforation because that is the region of least vascularity. The central perforation is defined as a perforation in pars tensa which is surrounded by a normal remnant of the tympanic membrane (Fig. 11.1).

Types of perforations associated with mucosal COM** (Figs 11.1 and 11.2).

1. *Small central perforation*: Perforations involving less than 25% of tympanic membrane.
2. *Large central perforation*: Perforations involving more than 25% of tympanic membrane.
3. Subtotal perforation (when only the annulus tympanicus is left).

The size of the perforation is perferably not termed as small or large; rather it should be mentioned in terms of percentage loss of tympanic membrane tissue.

Site of Pathology

The pars tensa can be divided into four quadrants but as the pathologies like tympanic membrane perforation are usually anterior, posterior or inferior therefore the division of the pars tensa into three divisions is preferred rather than four (Figs 11.3 and 11.4).

Clinical Features

1. **Ear discharge:** The discharge is the most common presentation of mucosal COM. The infection leads to chronic inflammation of middle ear mucosa with a varying degree of mucosal oedema, hypervascularity and presence of inflammatory infiltrates. There is

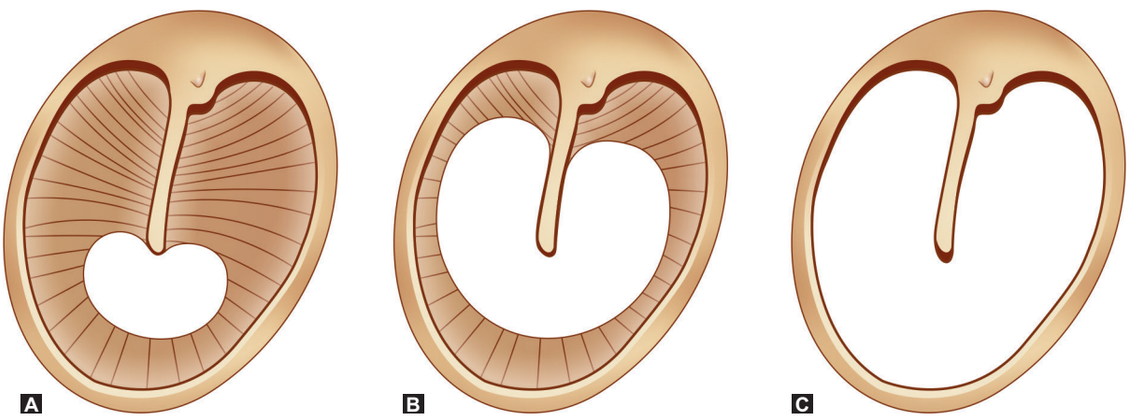


Fig. 11.1: (A) Small central perforation; (B) Large perforation; (C) Subtotal perforation

*The healed pars tensa perforation may mimic like a perforation sometimes. These can be correctly diagnosed by pneumatic otoscopy (siegelization), Valsalva manoeuvre or examination under microscope and otoendoscopy.

***Marginal perforation* is not a preferred terminology now and is only reserved for attic perforations because the annulus tympanicus is never absent until removed in surgery.

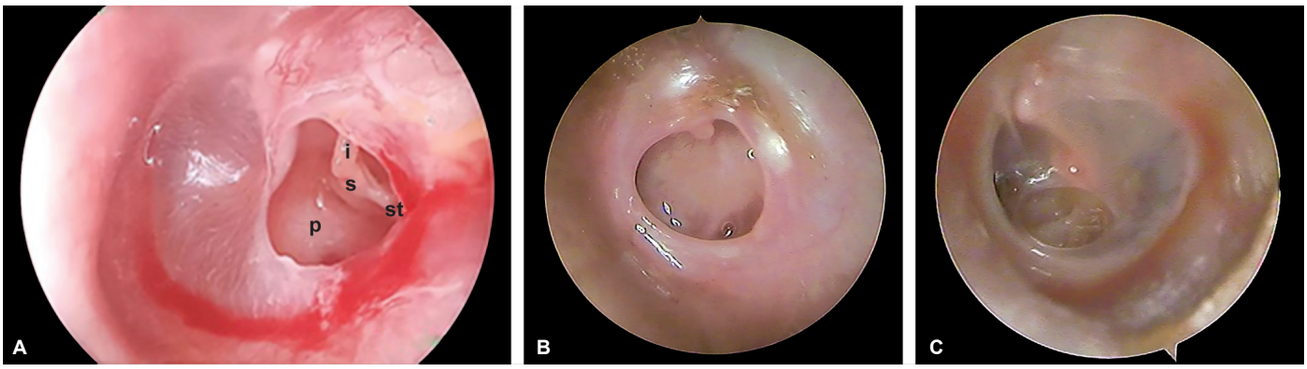


Fig. 11.2: Endoscopic pictures of central perforation of different sizes (ST: Stapedius tendon; S: Stapes, I: Incus; P: Promontory) are seen

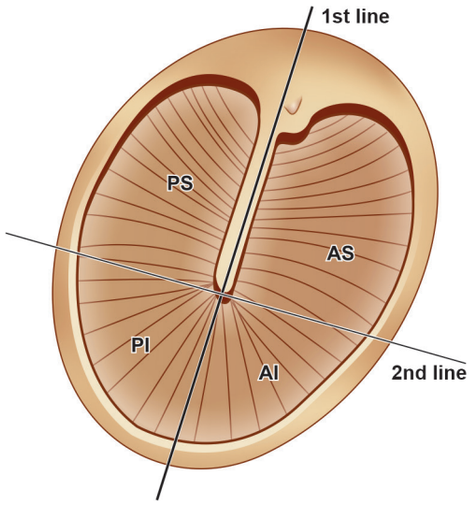


Fig. 11.3: Pars tensa division into four quadrants: Anterosuperior, anteroinferior, posterosuperior and posteroinferior (1st line passing through the length of the handle of malleus and the 2nd line perpendicular to the first line passing through the umbo)

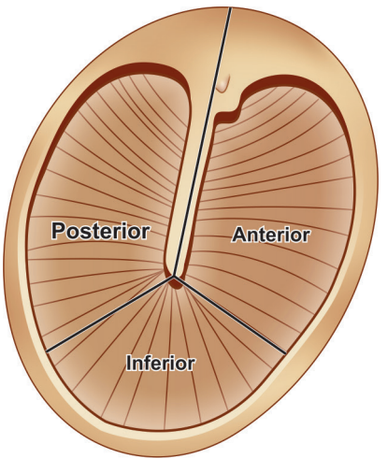


Fig. 11.4: Division of the pars tensa into three divisions/three-thirds: Anterior, posterior and inferior

basal cell hyperplasia and increase in number of goblet cells in middle ear mucosa.* The discharge is profuse and intermittent. The ear may start discharging during the episodes of upper respiratory infection and water entry into the perforated ear. It usually responds to antibiotic ear drops after which the ear becomes dry. The discharge may be mucoid, mucopurulent, or purulent depending on superadded bacterial infection. It is usually non-foul smelling (Fig. 11.5).



Fig. 11.5: Actively discharging ear

2. **Hearing loss:** Majority of patients will complain of hearing loss. It is usually conductive in nature, produced due to loss of pars tensa and but additional loss may be due to ossicular chain necrosis. In long standing cases some patients can develop mixed hearing loss. The main reason for cochlear damage is absorption of toxins via the round window or oval window.
3. **Polyps:** The mucosal oedema may lead to the formation of aural polyps, which may protrude from the perforation into the external auditory canal. In-fact these polyps are large granulations which bleed on touch or removal.
4. **Ossicular chain necrosis:** The ossicles are usually not involved in mucosal type of COM but may get

*The normal colour of middle ear mucosa is pale pink. On inflammation, it changes to pink or red.

necrosed in a few cases. The ossicular necrosis can occur due to the increased vascularity (hyperaemic decalcification)* and prominent granulations.** *The lenticular process of incus is most commonly involved because of its precarious blood supply followed by stapes crura, body of incus and manubrium of malleus*.

Note: If the perforation involves the posterior part of the pars tensa, then ossicular chain status (incudostapedial joint) can be seen on examination (Fig. 11.6).

5. **Tympanosclerosis:***** This occurs due to chronic inflammation of middle ear cleft mucosa, where upon healing the collagen in fibrous tissue hyalinises and become fused into a homogenous mass. There is subsequently calcification and rarely ossification. It is seen on tympanic membrane, ossicles, tendons, middle ear mucosa and oval and round window as a chalky deposit which leads to conductive hearing losses (Fig. 11.7).
6. **Fibrous sclerosis:** It is mainly a reparative phase of inflammation, where fibrous layer tissue is laid down in middle ear and mastoid air cells.
7. **Cholesterol granuloma:** There occurs a giant cell reaction around cholesterol crystals, which is probably derived from breakdown of extravasated blood cells. This indicates a middle ear inflammatory response. There may be a yellow fluid in the mastoid air cells. The eardrum might show a blue appearance. This is incorrectly called cholesterol granuloma, which is actually a histological pathology and better term for this is **cholesteatosis**.

Investigations

1. **Examination under microscope (EUM) or otoendoscopy** are done in all cases to confirm the otoscopic findings. Otoendoscope with video camera in OPD is an ideal equipment to demonstrate findings to the patient and record them for future reference. The status of ossicular chain, tympanosclerosis, and epithelial migration into the middle ear are all better visualised (Fig. 11.8).
2. **Pure tone audiometry** is done to assess the degree and type of hearing loss. Usually, it is of conductive type. Infrequently patients might develop mixed hearing loss. PTA is also a record for future comparison after surgical intervention and for medico legal purposes.
3. **Mastoid X-rays** do not have much role nowadays for for mucosal COM. In cases of non-drying ear, **HRCT temporal bone** is recommended to evaluate the status of mastoid pneumatization, any bony destruction and evidence of squamous disease.



Fig. 11.6: Necrosis of the lenticular process of incus along with pars tensa and flaccida retraction (Stapes suprastructure and malleoincudal joint are intact)



Fig. 11.7: Tympanosclerosis of the left tympanic membrane



Fig. 11.8: Otoendoscopy in OPD

*The role of bone remodelling molecules like RANK (which increase the osteoclastic activity) and expression of TNF-alpha and IL-1, 6, 10 have been established in bone destruction.

**The granulations are seen maximum around the ossicular chain in the middle ear cleft. This is probably due to the mucosal folds directing the inflammation on them.

***Though tympanosclerosis might seem like an end stage process, spontaneous recovery is seen in a few cases.

4. **Culture and sensitivity** is recommended in non-drying ears or in patients with infective complications of COM. It helps to decide the proper systemic and topical antibiotics. Gentamicin or ciprofloxacin are common topical antibiotics chosen without culture, as these are broad spectrum enough to cover most pathogens.

Treatment

Medical management: The aim of medical management is to make the ear dry and prepare it for surgery.

A. Cleaning of the ear properly and proper instillation of eardrops are the mainstay in medical management of chronic otitis media.

1. **Aural toilet:** The debris and discharge in the affected ear should be mopped with cotton swabs or wicks or it can be sucked using a suction tip. Mopping is recommended before instillation of drops. Continuous discharge patients may be subjected to ear toileting with suction under microscope. All patients are told to keep the ears dry especially during bathing and swimming by use of cotton plugs. Patients are also instructed not to blow their nose, so that ascending infections via Eustachian tube may be avoided.
2. **Eardrops:** The antibiotic ear drops are the mainstay of management of mucosal COM. The antibiotic ear drops used commonly are gentamicin, ciprofloxacin, chloramphenicol and polymyxin. These can be combined with steroids in case of oedema of middle ear mucosa or polyp formation. The antibiotics drops are preferred over systemic drugs because they achieve much higher concentration in ear tissue as compared to systemic drugs. The antibiotics used are known to be ototoxic when used systemically but these have shown to be non-ototoxic when given by means of topical eardrops.*
3. **Systemic antibiotics:** They are reserved for acute infections in COM and patients with associated URI or complications.
4. **Acetic acid irrigation (1%)** may be done in non-drying ears. It has antipseudomonal activity.

B. *Surgical Management*

1. **Aural polyp removal:** It should be done surgically by sharp cutting at its pedicle under vision. In no circumstance should a polyp be avulsed because it may arise from stapes supra-structure, facial canal or horizontal canal, therefore avulsion can lead to complications like facial nerve palsy or hearing loss. The removal of polyps facilitates the penetration of ear drops and to control ear discharge.
2. The aim of the surgery is **perforation closure** and to reconstruct the hearing mechanism. The decision how to manage the perforation depends upon the size of the defect.
   a. Small perforations can be managed by cautery patching or by **Pop-in technique**. The Pop-in technique is a type of inlay tympanoplasty using lobular fat as inlay graft (Fig. 11.9).

      Cautery patch or chemical cauterization is done for small central perforations which are dry. The chemical agents used are 50% trichloroacetic acid (TCA) or 25% silver nitrate. It can be repeated after 2 weeks if the entire perforation has not healed.
   b. Large perforations are managed by myringoplasty. The temporalis fascia is the most frequently used graft. If required the procedure can be combined with ossicular reconstruction, which is then termed as *tympanoplasty* (Fig. 11.10).

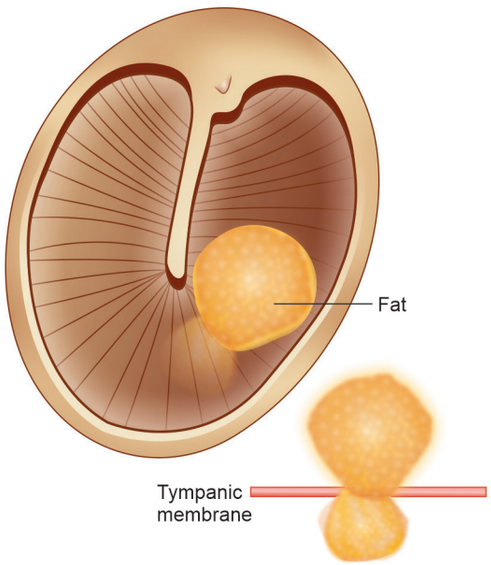


Fig. 11.9: Fat Inlay tympanoplasty (Pop-in technique)

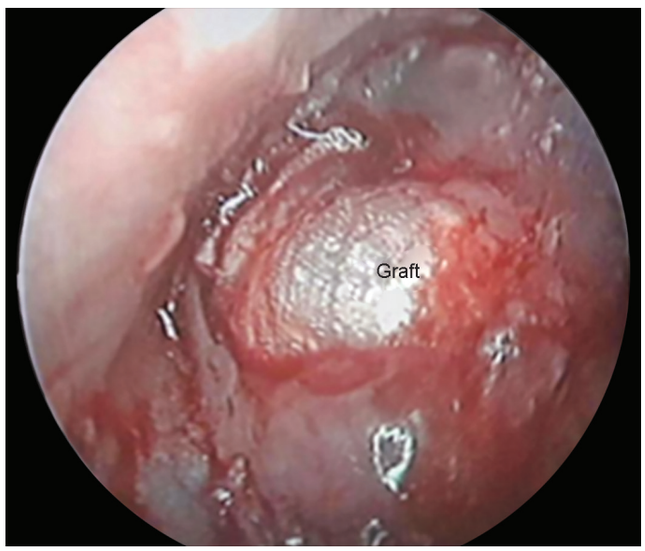


Fig. 11.10: Tympanoplasty with temporalis fascia

*The reason for this has been found to be due to the presence of a relatively deep round window niche and often this is protected by a pseudomembrane with selective permeability.

Tympanoplasty is not recommended in children less than 07 years especially if only one ear is involved. This is so because the number of attacks of upper respiratory tract infections are much more common in this age group, thereby compromising the success rate of the surgery.

EN4.8: Elicit document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of squamous type of CSOM.

C. *Inactive Squamous Chronic Otitis Media (Retraction, Atelectasis and Epidermization)*

The inactive squamous COM is defined as retractions in pars tensa or flaccida with the potential to be active with retained debris. They may be associated with damage to the ossicular chain and other middle ear structures. The pars tensa retraction grades are described in terms of structures on which the membrane is retracted onto and if it is adherent to them or not.

The pars tensa retraction pockets are classified into four grades according (*Sade*) (Fig. 11.11).

Grade 1: The pars tensa slightly retracted not touching the ossicles

Grade 2: The pars tensa retracted with adherence to ossicles.

Grade 3: The pars tensa is draping the promontory, without adhering to it.

Grade 4: The pars tensa is draped and adherent to the promontory.

The pars flaccida retractions are classified into four stages (*Tos*):* (Fig. 11.12).

Grade 1: There is a dimple in pars flaccid but not being adherent to malleus.

Grade 2: Pars flaccida is adherent to neck of malleus with no erosion of scutum.

Grade 3: Partial erosion of the bony outer attic wall (scutum).

Grade 4: Gross outer attic wall erosion.

It is important to note that if one encounters a pars flaccida retraction, it is mandatory to carefully examine for pars tensa retraction and vice-versa.

Management

1. **Observation:** The self-cleansing retraction pockets are usually stable** and therefore do not progress. Hence these do not require surgery and should be kept in regular follow up, especially children.
2. **Sinonasal treatments:** It is important to treat any associated allergy and sinonasal infections because they might be the only causes leading to Eustachian tube dysfunction and thereby producing retractions.
3. **Excision of retractions:** The retraction pockets which are deep accumulate squamous debris; therefore, have a tendency to progress. They are managed surgically. The area of retraction is excised and grafted preferably using a cartilage with perichondrium or temporalis fascia.***
4. **Marsupialisation:** Excision of the retraction pockets with repair can be combined with limited posterior-superior osteitic bone removal, which leads to marsupialisation of the retraction pocket.

Note: Exploration of the mastoid is not recommended in absence of active squamous changes like a cholesteatoma formation.

D. *Active Squamous Chronic Otitis Media*

In active squamous COM, there is an accumulation of keratinous debris in retraction pockets which actively expands to form cholesteatoma. The disease is characterised by the presence of decreased middle ear

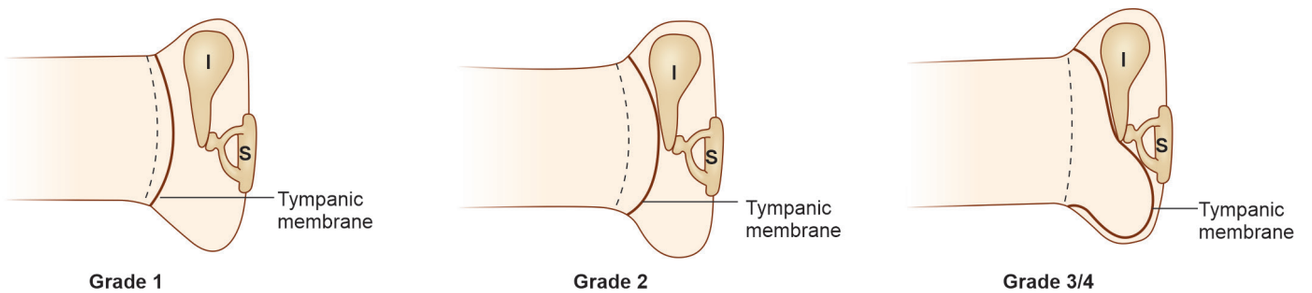


Fig. 11.11: Grades of pars tensa retraction pockets

*1. Though the classification is simple, it is sometimes difficult to differentiate grade 3 from 4, hence often may be grouped as grade 3/4.
2. Valsalva/autoinflation and tympanometry are helpful to determine the grading and outcomes.
3. Type A curve would indicate either an ossicular loss or tympanic membrane hypermobility due to atelectasis. Type B curve indicates presence of middle ear effusion but is sometimes associated with tympanic membrane retraction.
**The typical feature of a stable retraction pocket is that it still maintains the medial to lateral migration of the epithelium, thereby not allowing accumulation of epithelial debris.
***The recent data shows no role of ventilation tubes for prevention of progression of retractions, but may actually lead to retractions at application sites, in the long run.

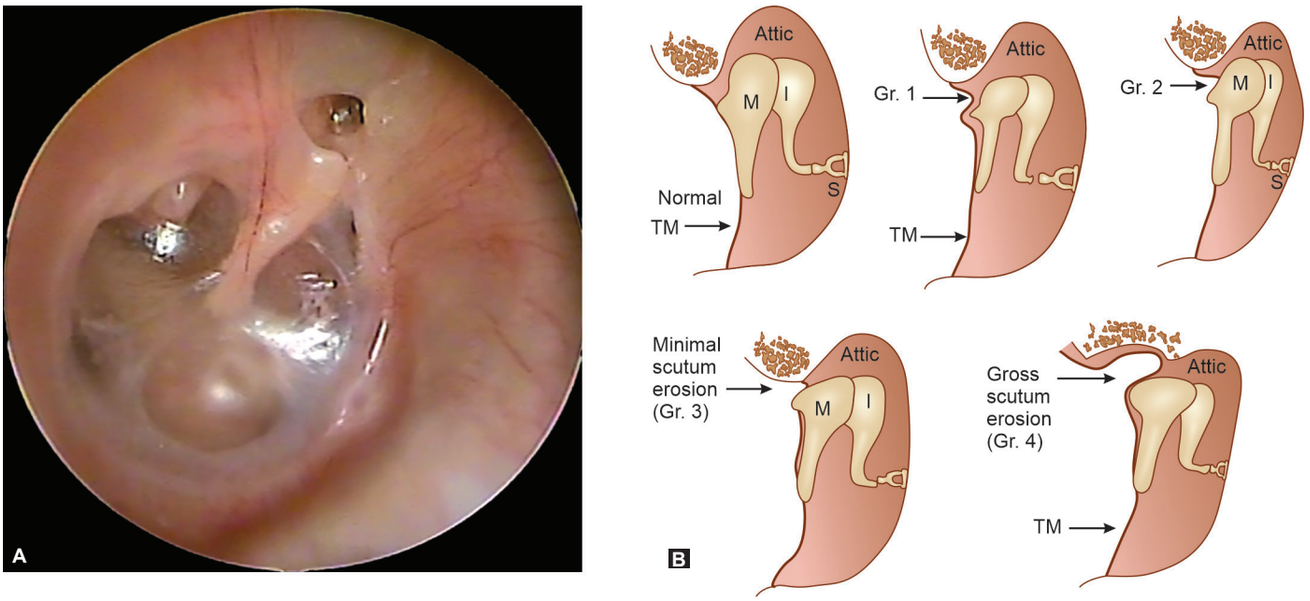


Fig. 11.12: (A) Grade 4 pars flaccida retraction; (B) Grade of pars flaccida retraction (*S: Scutum; M: Malleus*)

volume, poor Eustachian tube function and presence of **cholesteatoma**. The cholesteatoma has bone eroding property which involves the ossicular chain, mastoid air cell system and can lead to various intratemporal and intracranial complications, over a variable time scale. The other important feature is the presence of **granulations** which are formed due to osteitis of the adjacent bone (Figs 11.13 to 11.15).

CHOLESTEATOMA

The cholesteatoma is the presence of squamous keratinising epithelium in the middle ear cleft. The term cholesteatoma was first coined by a German physiologist Johannes Muller (1838). It is actually a misnomer because it is not a "cholesterol tumour", as the term denotes. It actually consists of inner keratin debris and outer matrix made of keratinising squamous epithelium resting on a fibrous stroma (Fig. 11.16).

Cholesteatoma is mainly of two types: **Congenital** and **acquired**.

Congenital cholesteatoma: *Levenson's criteria* are used for diagnosis of congenital cholesteatoma.

1. Presence of whitish mass behind intact tympanic membrane
2. No previous otological procedure
3. Normal pars tensa and flaccid
4. No prior history of tympanic membrane perforation or otorrhoea.

The congenital cholesteatoma is usually found in the middle ear or within the temporal bone, particularly the petrous apex. The source of congenital cholesteatoma

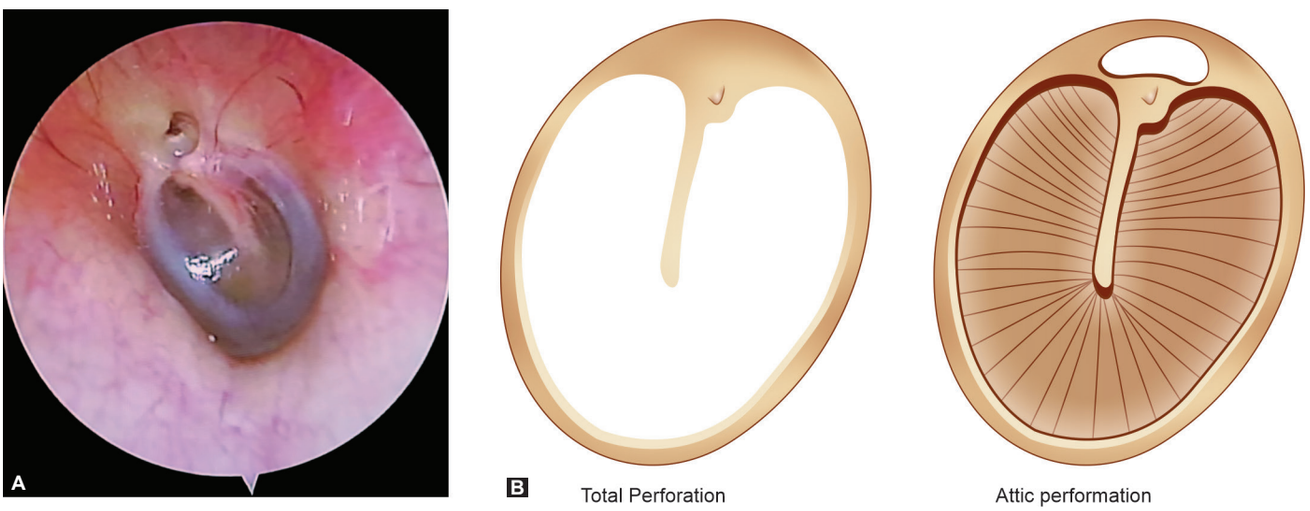


Fig. 11.13: (A) Attic perforation endoscopic view; (B) Perforation associated with squamous COM (schematics): (I) Total perforation (II) Attic perforation

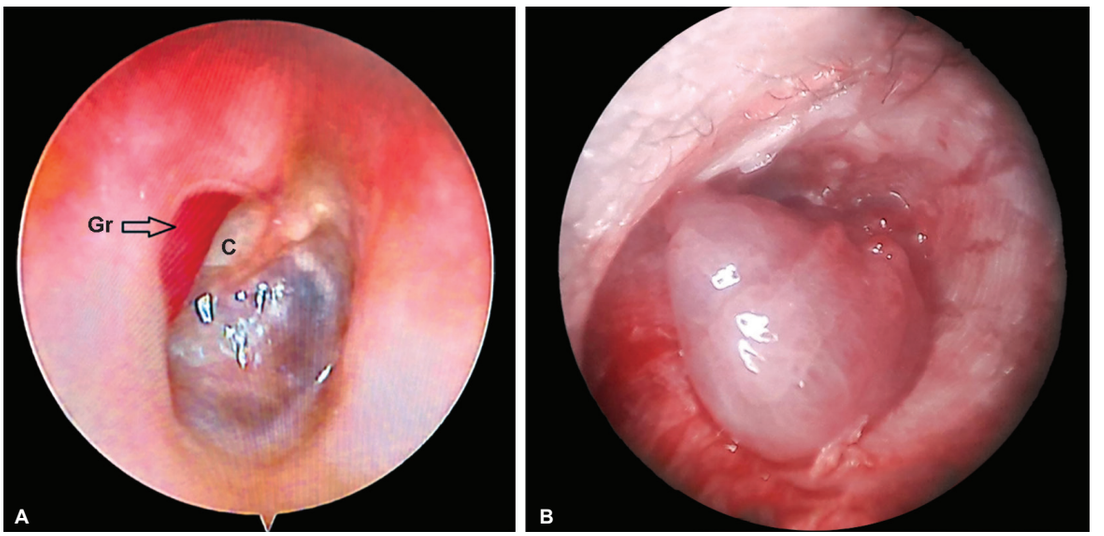


Fig. 11.14: (A) Otoendoscopic finding in a case of active squamous COM (Gr: *Granulation*; C: *Cholesteatoma*); (B) Aural polyp

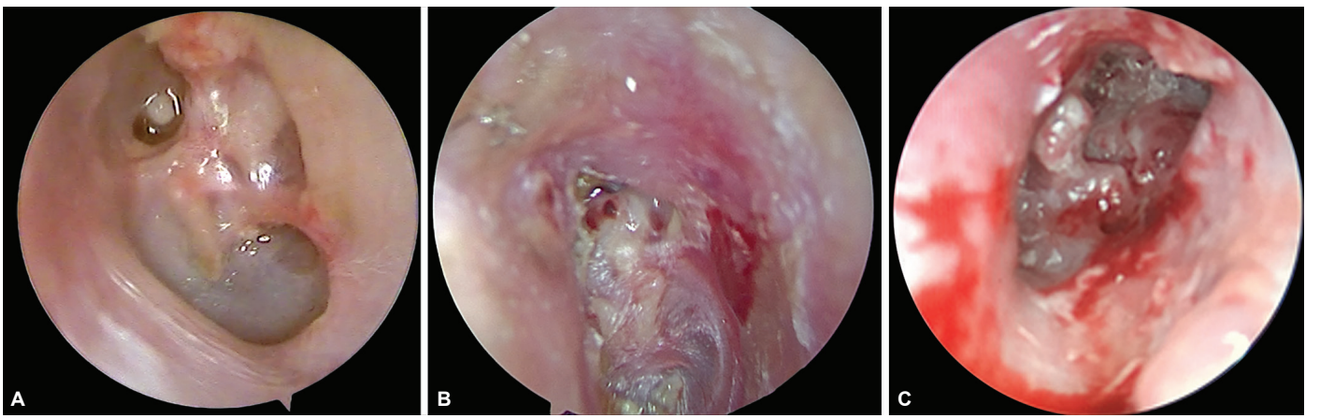


Fig. 11.15: Attic perforation with cholesteatoma

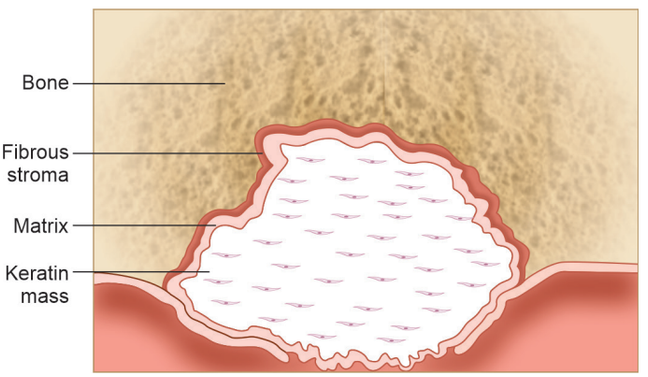


Fig. 11.16: Structure of cholesteatoma

is postulated to be due epidermoid formation which is a derivative of the 1st branchial groove. Normally it is found at the junction of the Eustachian tube and middle ear and it involutes by 33rd week of gestation.

Acquired cholesteatoma is of two types: Primary and secondary.

Primary acquired cholesteatoma results from enlargement of a retraction pocket.

Secondary acquired cholesteatoma is formed by infection, trauma, or surgical manipulation causing implantation of skin into the middle ear through a defect in the eardrum.

Over a period of time various theories of cholesteatoma formation have been described.

1. Congenital cell rest theory **(Teed, 1936)**: The epithelial rest theory involves the abnormal persistence of epithelial cell rests in middle ear cleft giving rise to cholesteatoma.
2. Invagination theory **(Wittmaack, 1939)**: It is also called the 'attic block theory'. The persistence of hyperplastic embryonic type of mucoperiosteum in epitympanum leads to the attic blocking of the ventilation, leading to negative pressure formation. This leads to the formation of a retraction pocket. Retraction pockets may become non-cleaning type with resultant precipitation of

keratin accumulation and formation of matrix from stratified squamous epithelium. This leads to the retraction pocket expansion.*

3. Epithelial invasion theory **(Habermann 1888)**: The cholesteatoma arises from the epithelial cells of the tympanic membrane and when it invades the middle ear around the margins of the perforation. It was postulated because the cytokeratins which form the main subunit in proteins of skin are also seen in middle ear cholesteatoma.
5. Basal cell hyperplasia or papillary in-growth of squamous epithelium **(Ruedi)**: Cone like extension from the basal layer of epidermis becomes invasive and then gets inflamed. This grows subepithelial through basal lamina to form cholesteatoma.
6. Mucosal metaplasia **(Sade 1971)**: There is metaplasia of middle ear mucosa to keratinizing squamous epithelium forming cholesteatoma.

Clinical Features

1. **Ear discharge:** The discharge is scanty and foul smelling due to putrefaction of desquamated epithelial cells in the cholesteatoma sac. Blood-tinged discharge can be seen after cleaning the ear due to the presence of granulations. The discharge is continuous, generally purulent in nature and does not respond to topical or systemic antibiotics.
2. **Hearing loss:** The patient generally has conductive hearing loss in the affected ear. It may sometimes be associated with sensorineural hearing loss because of inner ear damage due to toxins. In some patients even with ossicular necrosis, the patient hears well due the cholesteatoma bridging the defect (*cholesteatoma hearer*).
3. **Perforation and retraction pockets:** There may be attic perforation or a retraction pocket in posterior–superior quadrant. The deep retraction pocket with a narrow neck may collect debris and have cholesteatoma flakes.
4. **Complications:** In case of active squamous COM, there is a higher chance of complications; therefore, it is important to take relevant history and examinations to rule out these. The complications of COM have been discussed in the next chapter.

Investigations

The investigations are similar to that of the active mucosal type.

1. **EUM or otoendoscopy:** It is very important to perform examination under microscope or otoendoscopy because proper cleaning and suctioning under magnification will confirm the otoscopic findings and diagnosis. It might provide additional information about the ossicular status and extent of disease extension into attic and hypotympanum (Fig. 11.17).

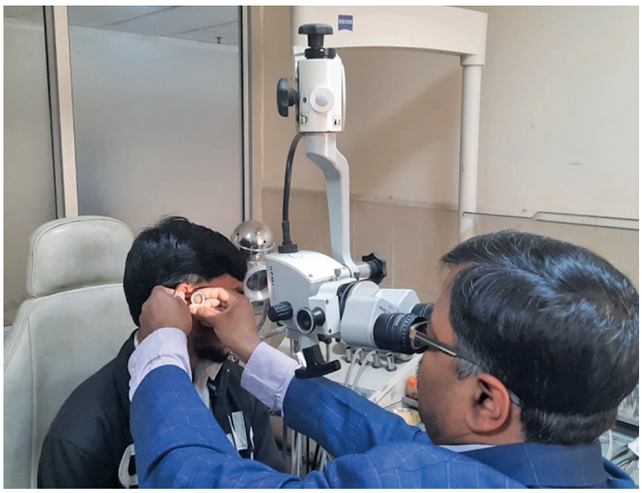


Fig. 11.17: Examination under microscope in OPD (EUM)

2. **X-ray mastoids:** Bilateral lateral oblique view (Law's view) and Towne's view are used. X-ray mastoids are of limited use for further management of the patient. X-rays can give some information regarding the degree of pneumatization of the mastoid air cell system and level of dural and sinus plate. The knowledge of the degree of pneumatization can be helpful in deciding the approach of mastoidectomy, i.e. *inside-out technique* (*anterior to posterior*) or out to in (*posterior to anterior*) technique. In sclerosed mastoid there might be difficulty in finding the antrum because it is contracted and there is low-lying dura and forward-lying sinus. Therefore, in such cases an inside-out technique is preferred (Figs 11.18 and 11.19).
3. **High-resolution computed tomography (HRCT) temporal bone** is a preferred radiological investigation nowadays because it gives much more information than an X-ray. All cases with suspected complications of COM should have a HRCT of the temporal bones (Figs 11.20 to 11.22).

Treatment

The treatment of active squamous COM is surgical.

Table 11.1 describes the differences between mucosal and squamous type of COM.

Surgery

AIIMS of mastoid surgeries: Various types of mastoid surgeries are performed for the management of squamous disease. The aims for the surgery are mentioned below according to their priority.

A. Eradication of disease from ear (especially cholesteatoma): The disease eradication is done by various mastoidectomy procedures which principally expose, excise and exteriorize (EEE) the diseased area.

*1. As the pars flaccida and posterosuperior quadrant of pars tensa has the maximum blood supply, therefore these areas have more inflammation during episodes of AOM and OME. This leads to thinning of the fibrous layer and subsequent retraction formation.
2. In case of pars flaccida, the presence of mucosal folds and bony plate above the supratubal recess, leads to differential negative pressures, which contributes towards retraction pocket propensity.

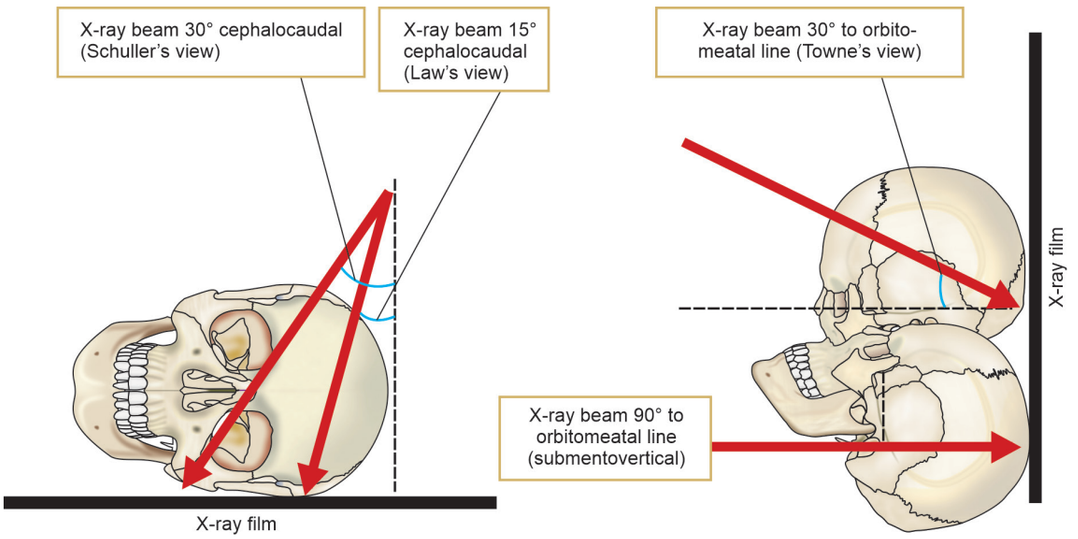


Fig. 11.18: How are the X-ray mastoids performed

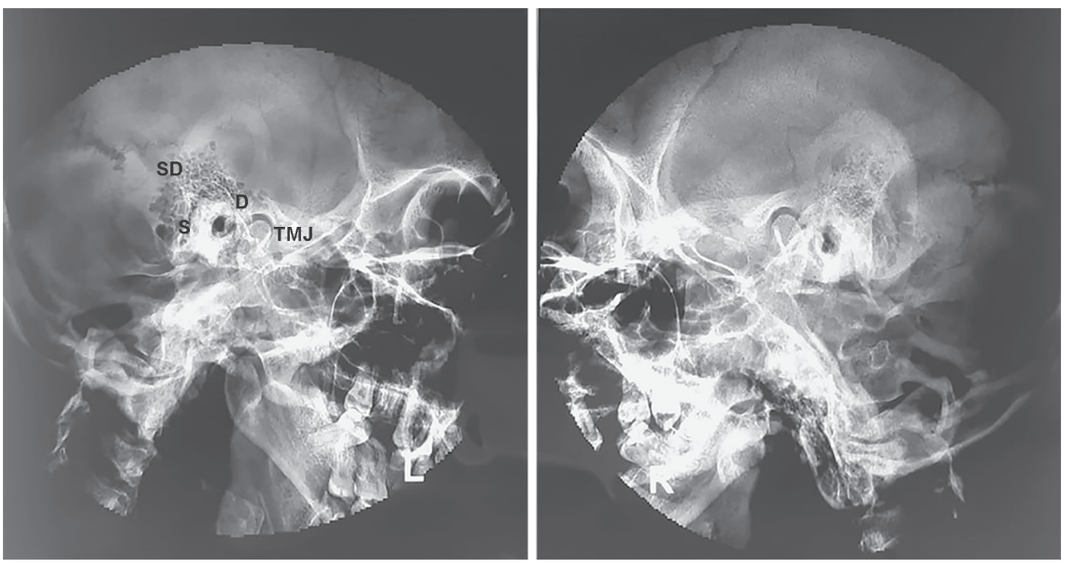


Fig. 11.19: X-ray mastoids: Lateral oblique view (both sides) (*D: Dural plate; S: Sinus plate; SD: Sinodural angle; TMJ: Temporomandibular joint*)

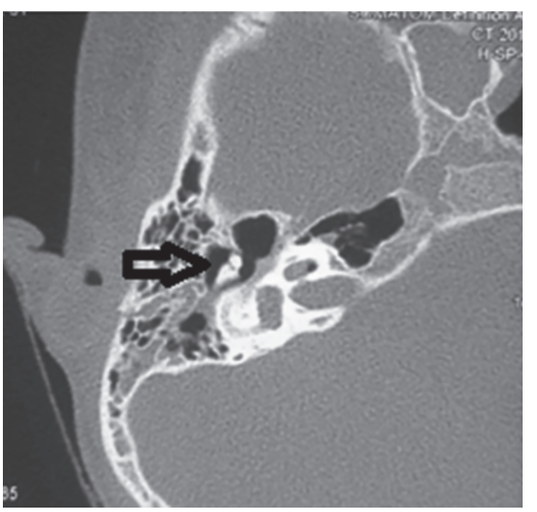


Fig. 11.20: HRCT temporal bone "Ice cream cone" appearance of malleus and incus (normal)

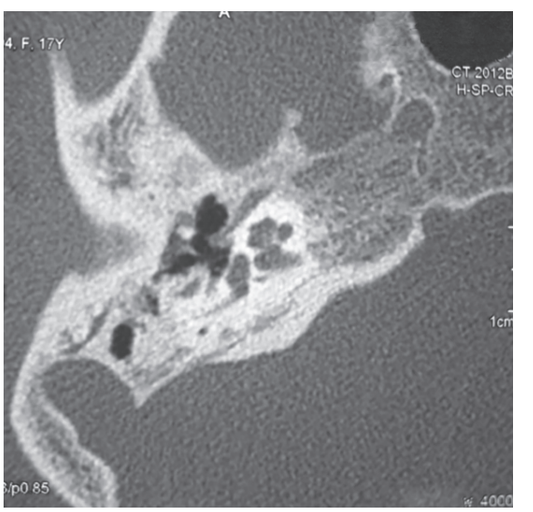


Fig. 11.21: Disease in right epitympanum

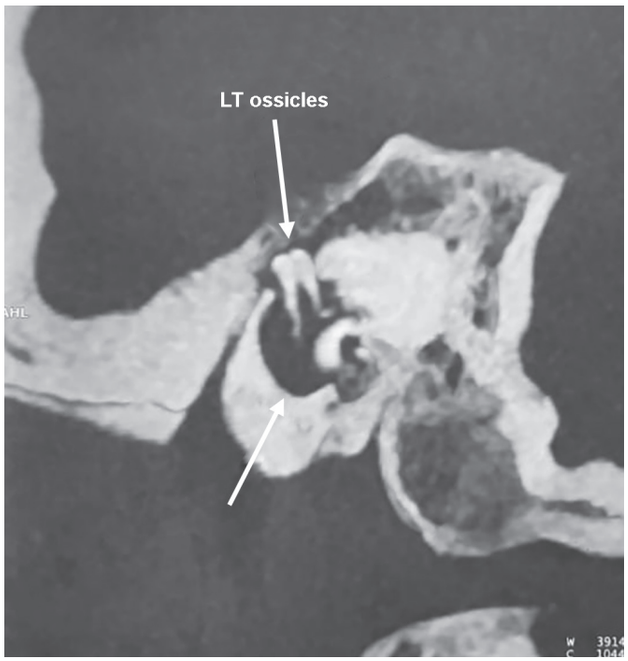


Fig. 11.22: Normal ossicles in left ear on HRCT temporal bone

Table 11.1: Differences between mucosal and squamous type of COM

		Mucosal COM	*Squamous COM*
1.	Ear discharge	Non-offensive profuse, mucoid or mucopurulent	Scanty, purulent, foul smelling
2.	Hearing loss	Usually conductive and mild to moderate	Moderate to severe, mixed also
3.	Perforation	Central or subtotal	Attic perforation or total perforation
4.	Retraction pocket or cholesteatoma	Not present	Retraction pocket or cholesteatoma is usually seen
5.	Bacteriology	Gram-positive cocci usually	*Pseudomonas*, *Proteus*, *E. coli* *Staph. aureus* and anaerobes
6.	Complications	Rare	Not uncommon

B. Achieving a dry, well epithelialized mastoid cavity: The healing of the mastoid cavity occurs by gradual epithelization of the mastoid cavity over a period of 6–10 weeks, depending upon the size of the cavity.

C. Hearing maintenance or improvement: The hearing restoration of the patient can be achieved partially or completely using autografts of incus or malleus. This can also be done using prosthesis (TORP and PORP) made of titanium or teflon. The ossiculoplasty may be done as a second stage procedure (after 9–12 months).

The postoperative mastoid cavities do affect the **acoustics** of the external auditory canal thereby producing additional hearing loss and difficulty in fitting the hearing aids, if so required.

APPROACHES FOR EAR SURGERY

Incisions and Approaches

1. **Permeatal/endomeatal approach:** In this approach *Rosen's incision* is used. It is an elliptical incision which starts from 12° o'clock position and ends at 6 o'clock position. The incision is approximately 5–7 mm from the annulus at the maximum width of the tympanomeatal flap (Fig. 11.23).
   The incision is mainly used for microscopic/endoscopic tympanoplasty and stapedotomy surgeries. Lately the endomeatal approach is being used for transcanal endoscopic mastoidectomy.
2. **Endaural approach:** This approach utilizes the Lempert's incision. The incision has two parts. The Lempert's I incision, which is a circumferential incision made from 6 o'clock to 12° o'clock in the external auditory canal. The second incision (Lempert's II) is continuation of the first incision at 12° o'clock position and ascends between the tragus and crus of helix in the preauricular crease. It is used for both tympanoplasty and mastoidectomy surgeries (Fig. 11.24).

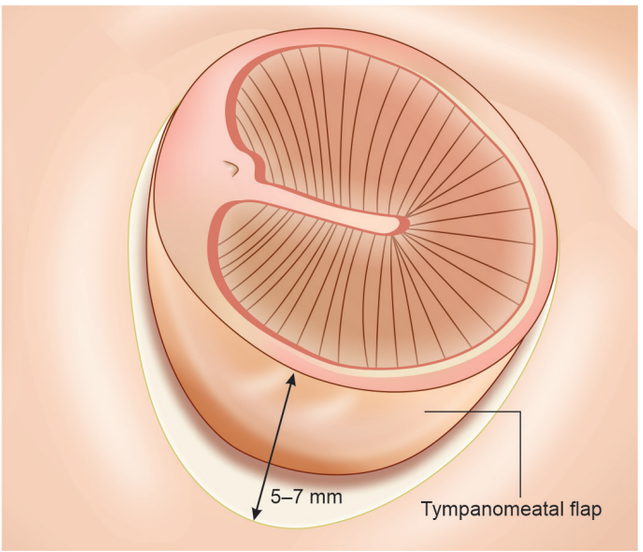


Fig. 11.23: Rosen's incision

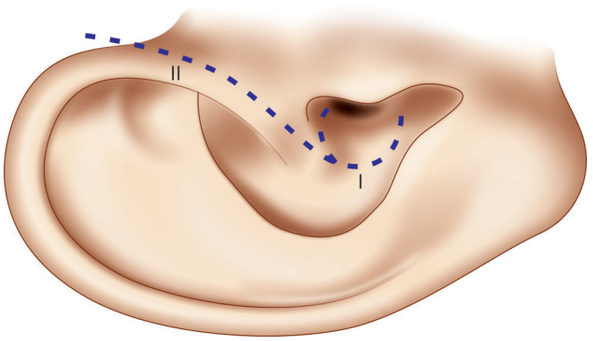


Fig. 11.24: Endaural incisions (Lempert's incision I and II)

3. **Postauricular approach:** This approach most commonly utilizes the *Wilde's postauricular incision*. The incision is made 8–9 mm behind the postauricular groove. It starts at the level of the superior most point of the pinna attachment and ends at the mastoid tip. For children below 2 years of age, the incision drifts away from the postauricular groove as it descends, because the mastoid tip is not developed till that age. This prevents any damage to the facial nerve, which will be lying very superficial and exposed. *Sulcus incision* is also used by few surgeons (Fig. 11.25).

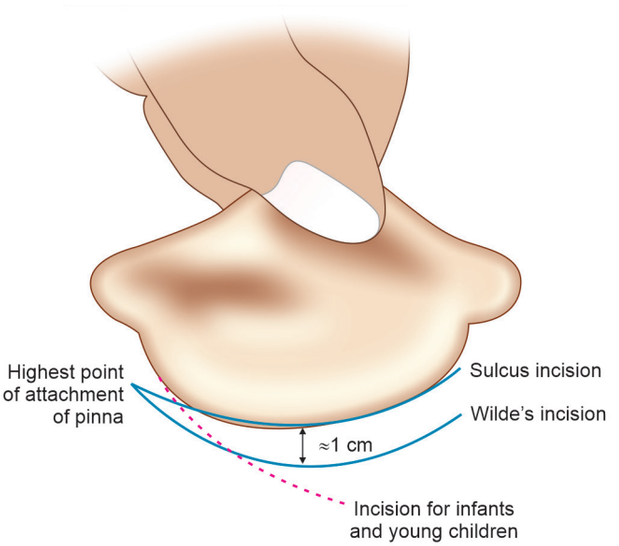


Fig. 11.25: Type of postauricular incisions

EN4.10: Observe and describe the indications for and steps involved in myringoplasty.

MYRINGOPLASTY AND TYMPANOPLASTY

Myringoplasty (*myringo*—eardrum and *plasty*—repair) is the surgical procedure of repair of pars tensa perforation.

Tympanoplasty involves reconstruction of the tympanic membrane and/or the ossicular chain after complete eradication of the disease from the middle ear cleft.

Tympanoplasty can be classified into five types according to the **Wullstein classification**.

Type I: There is only a perforation of the tympanic membrane and it is repaired. This is similar to myringoplasty except that the ossicular chain is intact on middle ear inspection (Fig. 11.26).

Type II: There is a perforation of the tympanic membrane along with erosion of the malleus. The graft is placed over the remnant of malleus or incus (Fig. 11.27).

Type III: Here both malleus and incus have been eroded or removed to clear the disease. The graft is placed over the head of the stapes. It is also called *myringostapediopexy* or *columella tympanoplasty* (Fig. 11.28).

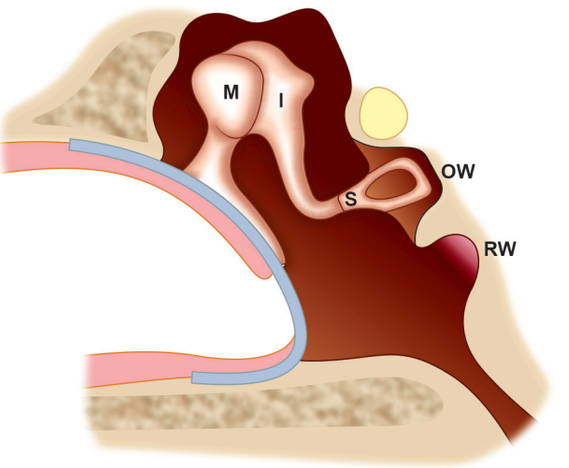


Fig. 11.26: Type I tympanoplasty

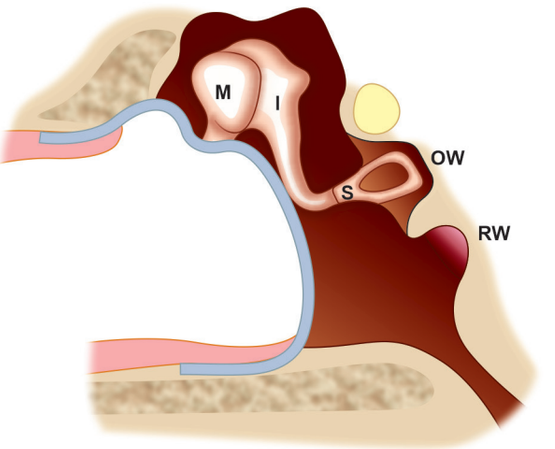


Fig. 11.27: Type II tympanoplasty

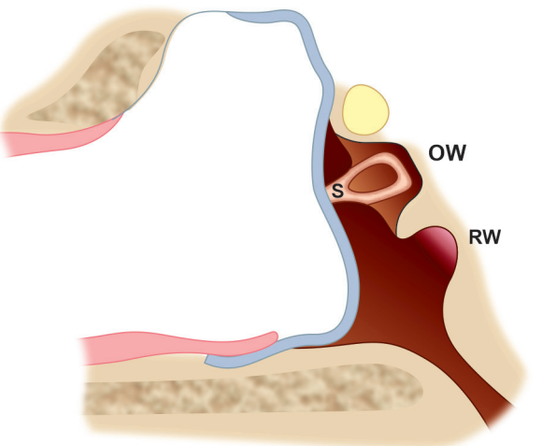


Fig. 11.28: Type III tympanoplasty

Type IV: Malleus, incus and suprastructure of stapes are completely eroded or have been removed to clear the disease and only footplate of stapes is present. The stapes footplate is left exposed and the graft is placed over the round window area in continuation with Eustachian

tube to create a small cavity called the **cavum minor** (*Round window baffle effect*). This leads to the sound waves directly striking the oval window (Fig. 11.29).

Type V: The stapes footplate is fixed. A second window is created by drilling the dome of horizontal semicircular canal bone and the procedure is called fenestration operation. Graft is also placed over this newly created second window. This is an obsolete procedure and no longer done nowadays (Fig. 11.30).

Indications

1. Mucosal type of pars tensa perforation
2. In case of residual perforation after a mastoidectomy. Tympanoplasty is essential to achieve dry postoperative mastoid cavity and decrease hearing loss.

Prerequisites

1. Dry/non-discharging ear for 4–6 weeks.
2. Good Eustachian tube function for ventilation of the middle ear cleft.
3. Age of the patient should be more than 12 years. Pediatric myringoplasty is not performed because in children there are frequent episodes of upper-respiratory tract infections and persistent Eustachian tube dysfunction. Thus there are high chances of repeated attacks of otitis media and reperforation of repaired tympanic membrane. Though now many centers are performing pediatric myringoplasty citing the lack of overall development because of hearing loss in young children with tympanic membrane perforation.
4. No active upper respiratory tract infection.
5. External auditory canal should be free of any infection.
6. Good cochlear reserve (it is not an essential prerequisite, because though following surgery, complete hearing recovery outcomes would not be achieved but it still would help achieve a dry ear).

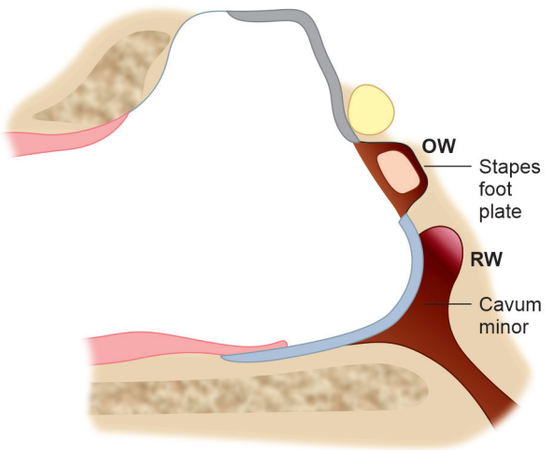


Fig. 11.29: Type IV tympanoplasty

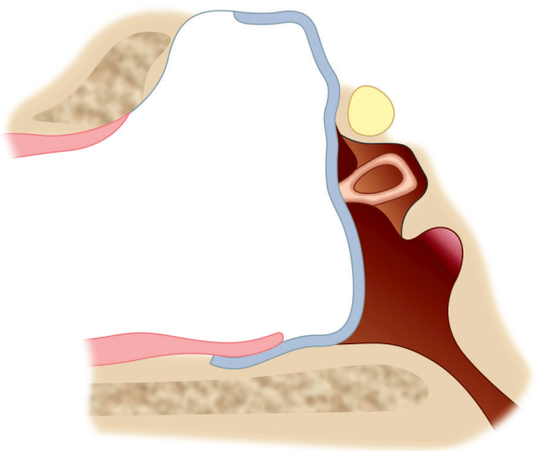


Fig. 11.30: Fenestration procedure—type V tympanoplasty

Note: In only hearing ears, myringoplasty should be undertaken by an experienced surgeon.

Techniques of Myringoplasty/Tympanoplasty

1. Underlay technique
2. Overlay technique
3. Inlay technique.

Instruments used in myringoplasty/tympanoplasty are shown in Fig. 11.31.

Steps

1. Underlay technique (graft placement is medial to mucosal layer of tympanic membrane)

1. Procedure can be done under general or local anaesthesia. For local anaesthesia 2% lignocaine with 1:30,000 adrenaline is used in each quadrant of EAC.
2. **Wilde's postauricular incision** is used for postauricular approach and *Lempert's incision* for endaural approach. For the permeatal approach *Rosen's incision* is used.
3. **Graft** is harvested. Temporalis fascia graft is obtained from the same incision, as for postauricular or endaural approach. In the permeatal approach, a separate 3–4 cm horizontal incision is made above the pinna for harvesting temporalis fascia. Tragal perichondrium is preferred by surgeons for permeatal approach (Figs 11.32 and 11.33).
   Grafts used in tympanoplasty: Temporalis fascia, tragal perichondrium, fascia lata, vein, periosteum, dura, cadaveric TM/dura and alloderm.
   Temporalis fascia is most preferred graft material because
   - It can be harvested from the same incision and no separate incision is required.
   - It is available in adequate size.
   - The thickness and consistency of temporalis fascia is similar to the tympanic membrane.
   - There is no donor site morbidity.
4. **Margins** of the perforation are freshened by removing the thin rim of the perforation edges using a micro-needle or a sickle knife.

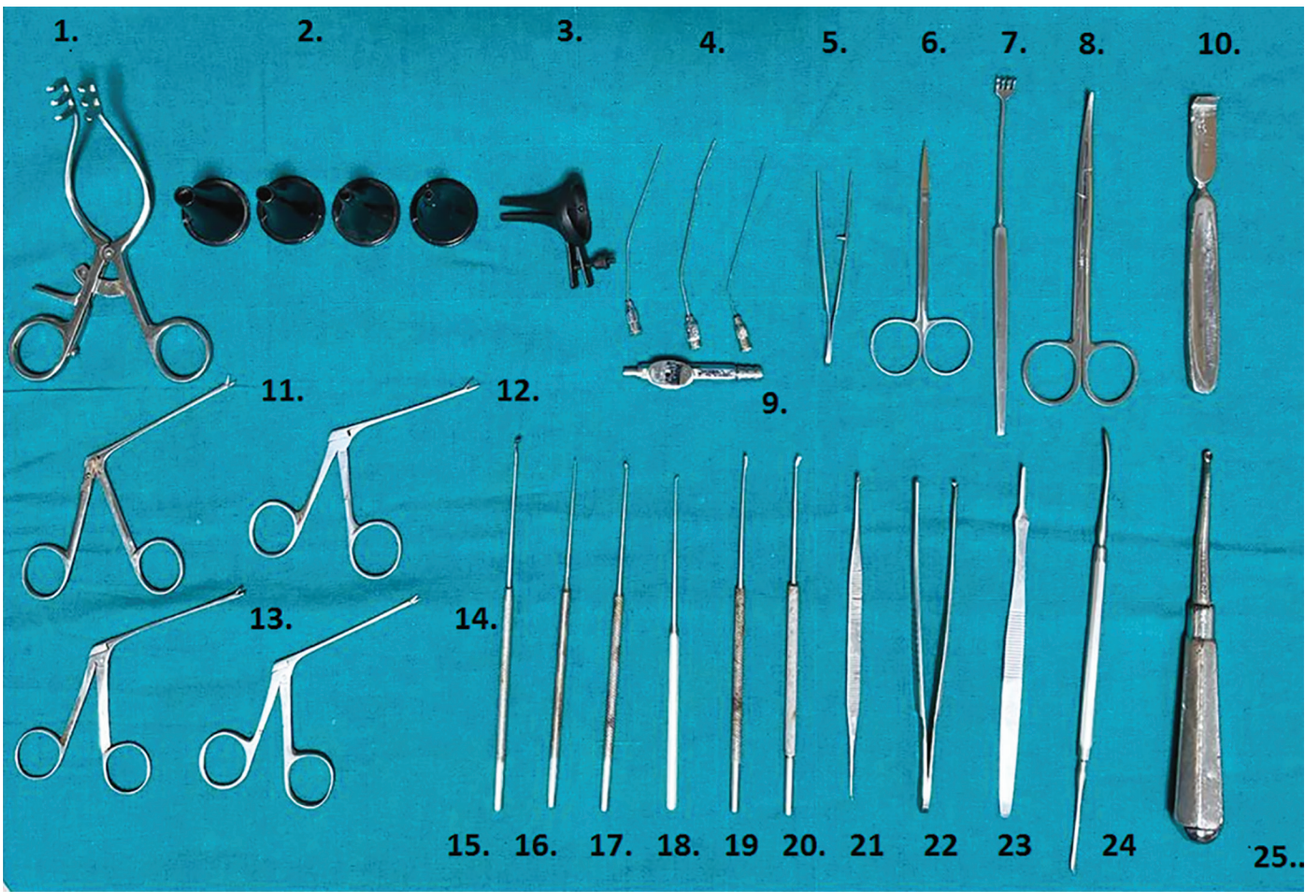


Fig. 11.31: Tympanoplasty set of instruments
1. Self-retaining mastoid retractor; 2. Set of ear speculum; 3. Self-retaining ear speculum; 4. Set of micro-suction tips; 5. Fine forceps; 6. Adson's scissors (straight); 7. Cat's paw retractor; 8. Tissue cutting scissors; 9. Suction tip connector; 10. Periosteal elevator; 11. Micro-cup forceps; 12. Crocodile forceps; 13. Micro-curved scissors; 14. Straight micro-scissors; 15–20. Micro-instruments for ear; 21. Micro curette; 22. Fine tooth forceps; 23. Bard Parker knife handle; 24. Freer's elevator; 25. Lempert's curette

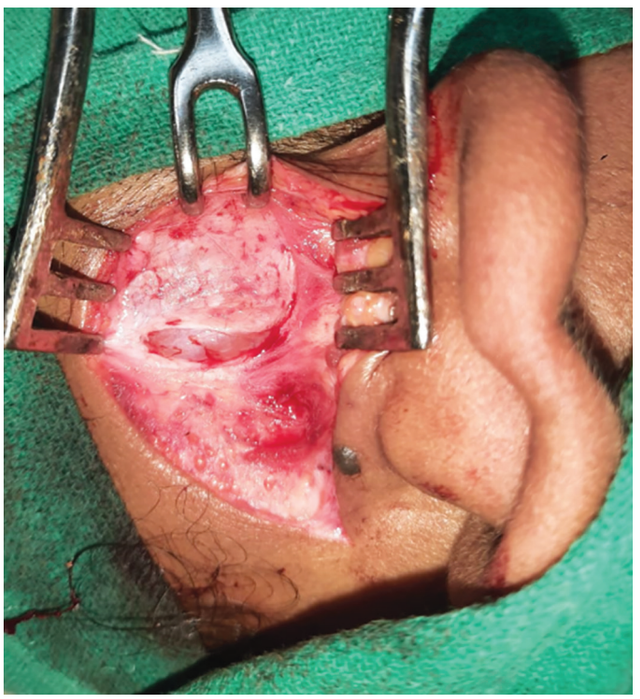


Fig. 11.32: Temporalis fascia graft



Fig. 11.33: Tragal perichondrium

5. Wide-based circumferential **canal incision** is made (5 o'clock to 12 o'clock for right ear and from 7 o'clock to 12 o'clock for left ear).
6. **Tympanomeatal flap** (meatal flap along with the annulus and remnant of the tympanic membrane) is

elevated by using a micro-circular knife and micro-flap elevator (Fig. 11.34).

7. The **middle ear is inspected** for tympanosclerosis, granulations or cholesteatoma. The ossicular chain integrity is checked. The ossicular chain reconstruction is done where required.
8. **Graft is then placed** by underlay technique (Fig. 11.35).

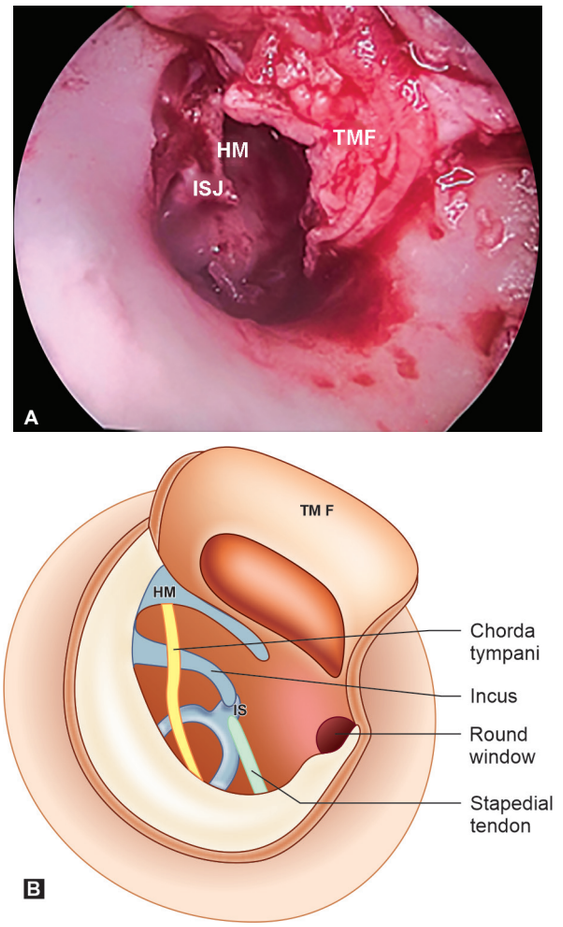


Fig. 11.34: (A) Endoscopic picture of the middle ear contents after elevation of tympano-meatal flap (TMF); (B) Diagram showing middle ear view on tympanomeatal flap elevation (*HM: Handle of malleus; IS: Incudostapedial joint*)

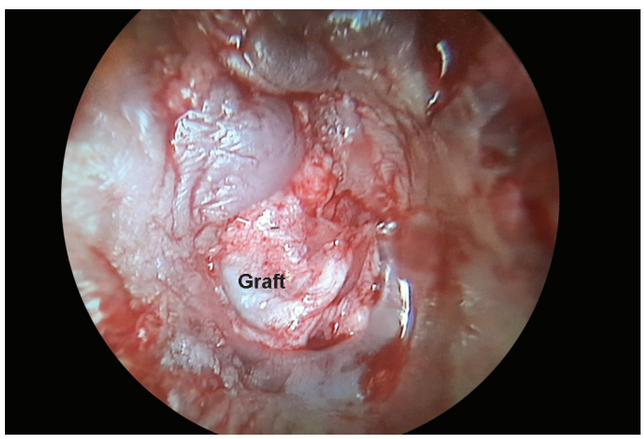


Fig. 11.35: Graft placed by underlay technique

9. The graft is stabilised by use of soft gelatine sponge pieces on both the sides.
10. The incision line is sutured in layers.

2. Overlay technique (graft placement between the epithelial and fibrous layer of tympanic membrane)

First 5 steps (above) remain the same. The tympanomeatal flap is elevated till the annulus. Now using sharp dissection, only the epithelial layer of the remnant of the tympanic membrane is carefully elevated. The graft is then placed between the epithelial layer and the fibrous layer of the tympanic membrane. The middle ear is not entered.

3. Inlay technique

First 4 steps are the same. The tympanomeatal flap is not elevated. Either a tragal cartilage with perichondrium on both sides or fat from lobule is used as graft. Cartilage is used for small to medium size of perforations but fat is only used for very small perforations (<2 mm) (Fig. 11.36).

Disadvantages of underlay technique

1. Medialization of graft
2. Postoperative anterior perforations
3. Reduction in middle ear space.

Disadvantages of overlay technique

1. Lateralization of graft
2. Cholesteatoma formation
3. Conductive deafness
4. Anterior blunting.

Complications

1. Postauricular hematoma
2. Facial nerve injury
3. Injury to chorda tympani
4. Ossicular dislocation.

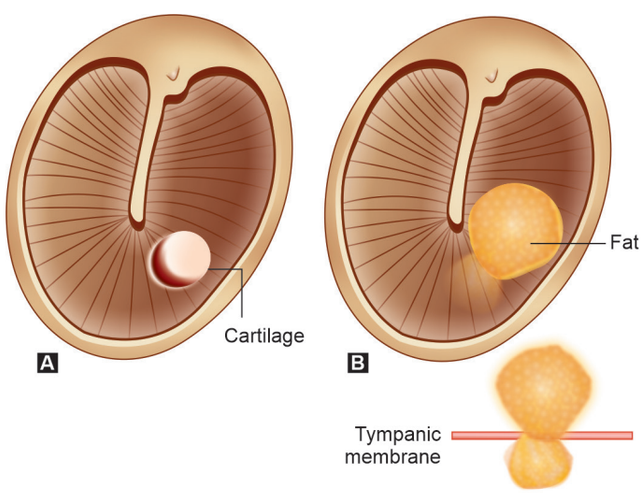


Fig. 11.36: (A) Cartilage inlay tympanoplasty; (B) Fat inlay tympanoplasty (Pop-in technique)

OSSICULAR RECONSTRUCTION AND PROSTHESIS

A tympanoplasty also involves reconstruction of the damaged ossicular chain, by using various autologous grafts and prosthesis. Grafts used commonly for reconstruction are autologous incus, cortical bone and tragal cartilage. Prosthesis which are used for construction of the hearing mechanism should be durable, biocompatible and easy to use during surgery. The various types of bio materials being used for making the prosthesis are: teflon, plastipore, ceramics, hydroxyapatite, metallic (gold and titanium) and hybrid.

Types

A. **PORP** or partial ossicular replacement prosthesis is used to bridge the gap between tympanic membrane or malleus handle and head of stapes. Commonly used materials are teflon and titanium. Titanium is preferred because it is lightweight and durable, though more expensive than teflon (Figs 11.37 and 11.38).

B. **TORP** or total ossicular replacement prosthesis is used to bridge gaps like a columella between tympanic membrane or malleus handle and footplate of stapes. Similar to PORP, commonly used materials are teflon and titanium (Fig. 11.39).

C. Autologous incus is commonly used as an autograft to reconstruct the ossicular chain. Most common defect of the ossicular chain is necrosis of the lenticular process, therefore incus is refashioned to be used as autograft.
   - It can either be placed between malleus handle and suprastructure of stapes and malleus-stapes assembly (MSA).
   - Between malleus and footplate of stapes and malleus to footplate assembly (MFA).
   - Incus can be also used to augment stapes remnants and directly connect to the tympanic membrane (Fig. 11.40).



Fig. 11.37: Titanium PORP



Fig. 11.39: Titanium TORP

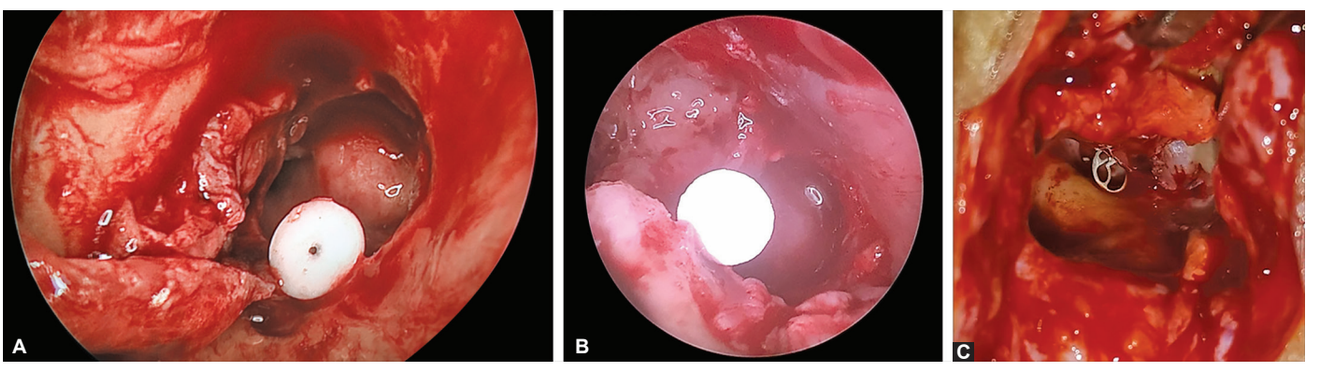


Fig. 11.38: (A) Teflon PORP; (B) Teflon TORP; (C) Titanium prosthesis

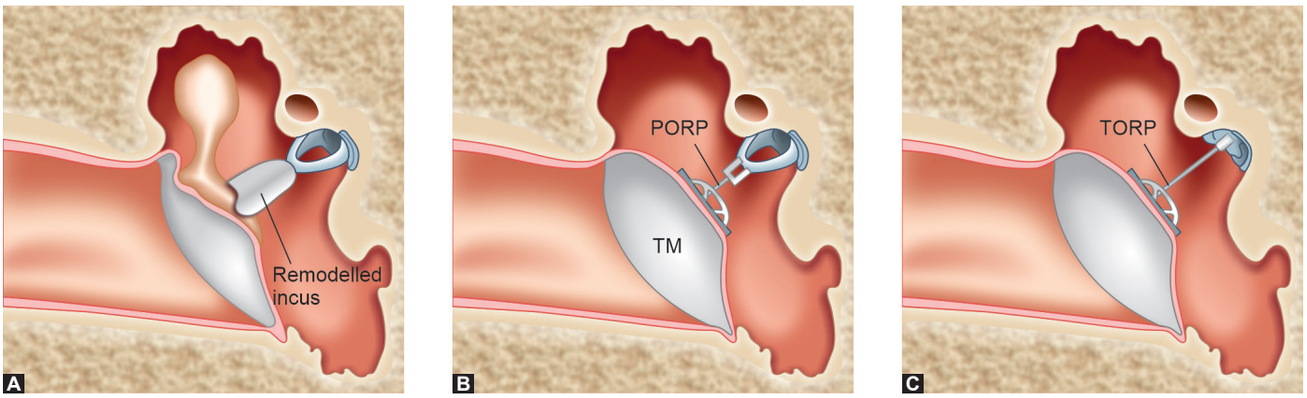


Fig. 11.40: Grafts and prosthesis for ossicular reconstruction

EN4.11: Enumerate the indications, describe the steps and observe a mastoidectomy.

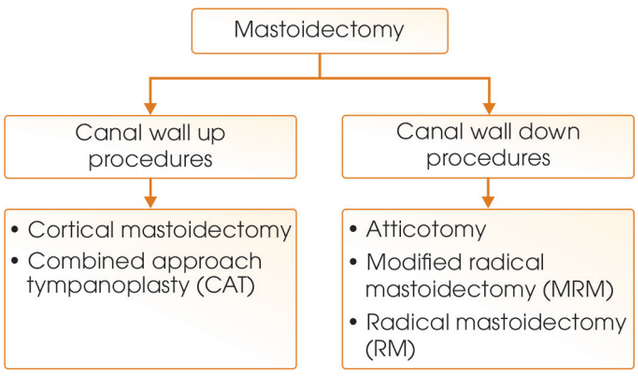
MASTOIDECTOMY

The eradication of disease from the middle ear cleft is done by various types of mastoid surgeries. Basically, two types of surgical techniques are used for disease eradication, namely the **canal wall up** (posterior canal wall is preserved) and **canal wall down surgery** (posterior canal wall is removed). The canal wall up surgeries are cortical mastoidectomy and *combined approach tympanoplasty* (CAT). The canal wall down surgeries include atticotomy, modified radical mastoidectomy (MRM) and radical mastoidectomy (RM) (Flowchart 11.1).

The **canal wall down procedures** include the traditional methods of disease removal. It can be done by posterior to anterior approach (*Outside-in technique*) or anterior to posterior (*Inside-out technique*) approach. These days anterior to posterior approach of mastoidectomy is preferred because it results in smaller cavity formation and subsequent less cavity problems.

The combined approach tympanoplasty **(canal wall up technique)** was started by Jansen in 1960. The technique involves performing a cortical mastoidectomy, tympanoplasty and posterior tympanotomy. Posterior tympanotomy is creation of a small window in the facial recess area to inspect and remove disease from the middle ear. Major advantage of the canal wall up procedures is there is no open cavity formation, thereby avoiding postoperative cavity problems. The disadvantage of the procedure is the high incidence of residual disease which necessitates a second look surgery after 12–18 months. It is certainly a technically difficult operation to be performed by an experienced surgeon. The facial recess approach to the middle ear is also used in insertion of cochlear implant electrodes.

Flowchart 11.1: Types of mastoid surgery



Facial recess is a triangular area bounded by facial nerve medially, chorda tympani laterally and fossa incudis superiorly.

All open cavities can be partially or completely obliterated or made into closed cavities by various procedures (discussed below).

Equipment: Apart from the tympanoplasty micro-instrument set, operating microscope or endoscopic camera monitor system, a high-speed drill is used. It comes with a set of diamonds and cutting burrs of various sizes. The otological drill has a speed between 10,000 and 40,000 rpm. The drill system has now completely replaced the mastoid gouge and hammer for bone removal during mastoidectomy.

1. Cortical Mastoidectomy (Simple Mastoidectomy or Schwartz Procedure)

Indications

1. Acute coalescent mastoiditis:
   i. Patient not responding to antibiotics.
   ii. Intracranial complications
   iii. Postauricular abscess, Bezold's abscess and zygomatic abscess
2. Masked mastoiditis
3. Tubercular mastoiditis
4. As an initial step in other surgeries
   i. Cochlear implantation
   ii. Endolymphatic sac surgery
   iii. Translabyrinthine approach for acoustic neuroma surgery, decompression of facial nerve.

Steps

1. The patient is ideally taken up under general anaesthesia and is put into supine position. The face is turned to the other side with the operated ear placed upwards. The head end is slightly raised to decrease bleeding.
2. Standard postauricular incision is given and the periosteum of the mastoid cut in T-shaped manner, elevated and retracted.
3. The **MacEven's triangle** then is identified and drilling is started in the area.
4. The antrum lies 12–15 mm deep to the outer mastoid cortex in adults. Antrum is the largest and the most consistent mastoid air cell. It might be contracted in cases of sclerosed mastoid. In such a case, the dural plate and sinus plate is first identified followed by the identification of the antrum. This helps to avoid any injury to the horizontal semicircular canal and facial nerve.

   Korner's septum if present may lead to a shallow cavity which is actually not the true antrum and the antrum actually lies below it. This can be confirmed by comparing the depth of the antrum formed and the middle ear cavity. Also false antrum will not have any communication with the aditus (Fig. 11.41).
5. Horizontal semicircular canal is identified.
6. All mastoid air cells from the posterior canal wall to the sinus plate, tegmen antri and mastoid tip are removed and sinodural angle **(Citelli's angle)** is cleared. This results in a common mastoid cavity.
7. It is essential to widen the aditus to achieve drainage and ventilation of the mastoid cavity thus produced. Surgeons should be careful here not to dislocate the incus while enlarging the aditus (Figs 11.42 and 11.43).
8. The incision line is closed in layers.
9. Mastoid dressing is applied.

Complications

1. Facial nerve injury
2. Horizontal semicircular canal injury
3. Dislocation of incus
4. Injury to dura
5. Injury to sigmoid sinus
6. Wound infection.

2. Modified Radical Mastoidectomy (MRM)

It is indicated in case of active squamous COM.

Steps

1. First 5 steps for the procedure are similar to cortical mastoidectomy. Apart from the postauricular approach, this procedure can also be done by endaural approach.
2. All the disease from the mastoid air cell system is removed. This requires following the disease and opening up the diseased air cells. Only diseased air cells are exenterated and following which a common mastoid cavity is formed.

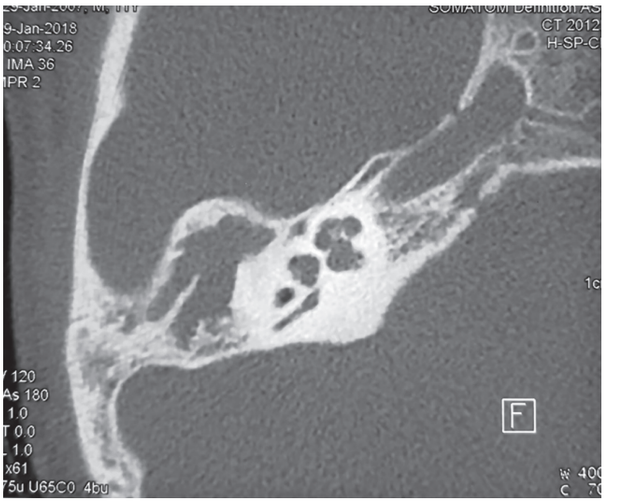


Fig. 11.41: Axial HRCT scan of temporal bone showing Korner's septum (incomplete)



Fig. 11.42: Opened mastoid antrum during cortical mastoidectomy. Note that the posterior canal wall is intact

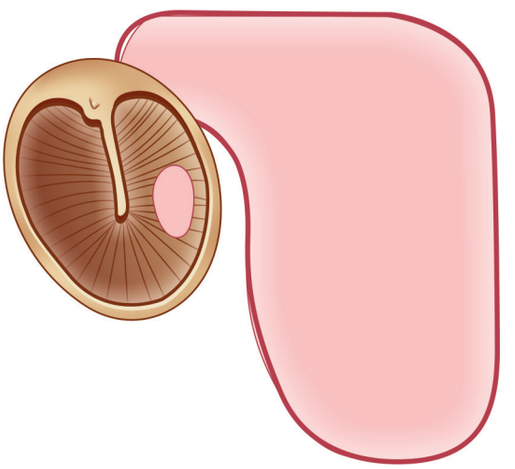


Fig. 11.43: Extent of surgery—cortical mastoidectomy

3. Facial bridge is now broken and the facial ridge lowered.
4. The anterior and posterior buttresses are removed.
5. All the disease from the epitympanum, mesotympanum, protympanum, retrotympanum and hypotympanum

is removed. This would require visualisation of all the hidden areas like sino-dural angle **(Citell's angle)**, sinus tympani and anterior epitympanum.

6. The procedure might require removal of ossicles if they are diseased or partially necrosed or the cholesteatoma is going medial to the ossicles. In such cases the disease clearance is not possible without removal of the ossicles.
7. Reconstruction of the hearing mechanism is done.
8. The mastoid cavity formed is then exteriorized by performing meatoplasty. The size of the meatoplasty should be sufficient enough to provide adequate drainage, ventilation and inspection of the mastoid cavity (Figs 11.44 and 11.45).
9. Closure of the wound in layers is done.

Complications

1. Facial nerve injury: 1–2% of the patients might have iatrogenic facial palsy. The most common site of injury is the 2nd genu of the facial nerve.
2. Horizontal semicircular canal injury.
3. Cavity problems: 20% of patients have problems of discharging mastoid cavity which is due to either infection, high facial ridge, tympanic membrane perforation, residual disease or recurrence.
4. Sensorineural hearing loss
5. Perichondritis of pinna
6. Injury to dura
7. Injury to sigmoid sinus
8. Wound infection.

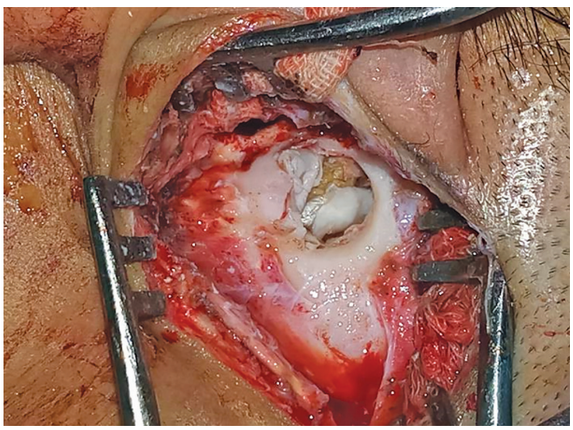


Fig. 11.44: Mastoid antrum filled with cholesteatoma

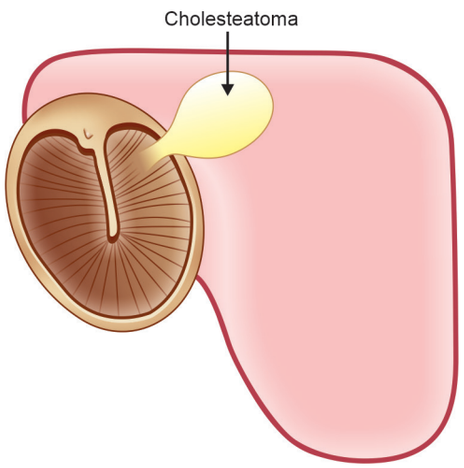


Fig. 11.45: Extent of modified radical mastoidectomy

3. Radical Mastoidectomy

Presently indications for the procedure are limited and include unresectable cholesteatoma involving the inner ear/Eustachian tube or when there is a cochlear promontory fistula.

Steps: The procedure is similar to MRM, but here all the mastoid air cells are exenterated and the Eustachian tube is blocked by muscle tissue and the middle ear is not grafted. All the ossicles are removed except for stapes remnant. Complications are similar to MRM.

4. Atticotomy

This procedure can be done with an endaural or postauricular approach using an operating microscope. Lately, transcanal endoscopic approach has been used to perform atticotomy.

It is done for limited disease in the attic and requires the removal of the outer attic wall (scutum) and part of the posterosuperior canal wall. After removal of the disease **atticoplasty** is done. This involves the repair of the scutum or the outer attic wall by using a conchal or tragal cartilage.

5. Attico-antrostomy

It is done for extension of the disease to the antrum. The aditus and antrum are opened and disease is cleared all areas.

Similarly, extensive disease spread to the other mastoid air cells will require **tympano-mastoidectomy**. It will require adequate lowering of the facial ridge, removal of anterior and posterior buttresses and inspection of the hidden areas of the middle ear cleft. A good meatoplasty is done similar to MRM.

Complications of the inside–out techniques are similar to those in MRM.

> ***Note:*** As the disease is followed from its place of origin in middle or epitympanum to the rest of the middle ear cleft, only minimal bone removal is required for complete clearance of the disease This leads to smaller cavity formation and obvious lesser cavity problems.

Transcanal endoscopic mastoidectomy is the recent advance in the management of active squamous COM. This makes use of rigid endoscopes with a diameter of 3 mm and 0-degree and 30-degree, along with HD camera and monitor. The procedure is similar to the microscopic inside out approach mastoidectomy (anterior to posterior technique). The advantages of the procedure are that the resultant cavity size is much smaller and all the hidden areas of the tympanic cavity, like sinus tympani, protympanum and hypotympanum can be easily seen with angled endoscopes and the disease

removal is complete. It is a relatively difficult technique to learn because one hand is busy holding the endoscope while the other hand performs the surgery, hence a single handed technique (Fig. 11.46).

Problems with the Postoperative Mastoid Cavity

Few mastoid cavities in the postoperative period keep on discharging even after 6–12 weeks. This is called discharging mastoid cavity. Factors which contribute to a discharging cavity are:

1. High facial ridge
2. Inadequate meatoplasty (meatal stenosis)
3. Perforation in the tympanic membrane
4. Residual disease
5. Large mastoid cavity (>3 cc).

Even after achieving the dry mastoid cavity there are some inherent problems of mastoid cavity which are lifelong. These can be termed as 5 D's.

5 D's of Mastoid Cavity

1. Debris: There is collection of squamous debris in the cavity due to altered migration of epithelium.
2. Discharge: The cavity starts discharging when there is infection of the collected debris due to entry of water during bathing or swimming or when there is extensive moisture as in monsoon season.
3. Dependence: The patient needs to regularly visit an ENT specialist for cleaning the cavity, especially wax.
4. Deafness: The middle ear volume is reduced because of the removal of the posterior canal wall resulting in conductive deafness.
5. Dizziness: The dome of the lateral semicircular canal is exposed after canal wall down mastoidectomy. The patient feels dizzy whenever there is sudden change in temperature or cold or warm air enters the cavity due to caloric stimulation of the lateral semicircular canal.

The problems of mastoid cavities can be reduced by:

i. Good surgical technique thereby complete disease removal, saucerization of mastoid cavity, adequate meatoplasty and adequate lowering of the facial ridge (Fig. 11.47).

ii. Inside out mastoidectomy technique and transcanal endoscopic mastoidectomy produce smaller mastoid cavities due to less bone removal for complete disease eradication. The smaller mastoid cavities have better healing rates and less cavity problems.

iii. Large mastoid cavities have higher chances of producing discharge, therefore it is recommended to perform mastoid cavity obliteration in such cases.

Partial or complete obliteration of the mastoid cavities can be done by using:

a. Pedicled flaps, e.g. Rambo's flap-temporalis muscle flap, Palva's flap–musculo-periosteal flap, Hong Kong flap-pedicle temporalis fascia flap, Singapore flap–pedicled temporoparietal fascial flap (TPFF) flap (Fig. 11.48).

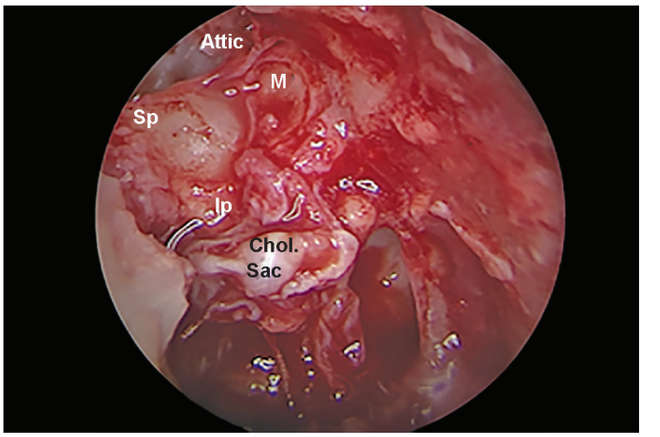


Fig. 11.46: Transcanal endoscopic atticotomy



Fig. 11.47: Healed mastoid cavity and tympanic membrane

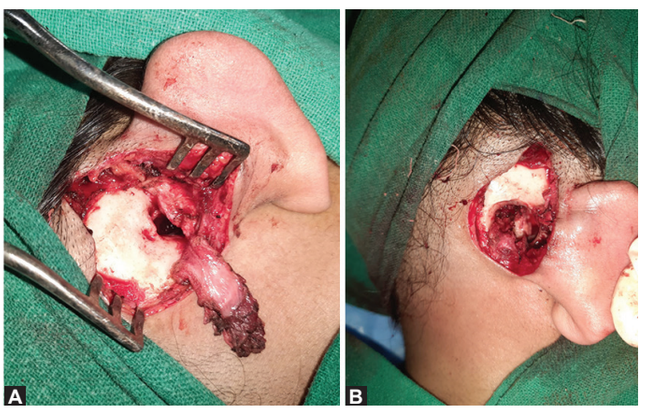


Fig. 11.48: (A) Palva's flap elevation (Musculo-periosteal flap); (B) Palva's flap rotated inside the cavity

b. Free grafts of skin, bone, cartilage and fascia.

c. Hydroxyapatite, bone pate and cartilage pieces.

iv. Reconstruction of posterior canal wall by cartilage or prosthetic material like porous hydroxyapatite, ceramic, bioglass and ceravital.



Chapter

12

Complications of Otitis Media

The complication of chronic otitis media (COM) is defined as the spread of infection beyond the middle ear cleft to surrounding structures separated by the bone. Though with the advent of good antibiotics and early management of ear infections, the rate of complications of COM have drastically decreased. In developing countries like India, ear related complications are still not rare. This is due to relative lack of medical facilities, awareness, and low socioeconomic status.

The main factors which contribute to complications are:

1. **Age:** Most of the patients with complications are young children because the disease process in chronic otitis media is more aggressive in younger age groups.
2. **Immunodeficiency:** Patients with lack of immunity, as in AIDS, diabetes, immunosuppressive drug intake or cancer, are more prone to develop complications.
3. **Inadequate management:** Lack of proper management of subacute and chronic infections of the ear.

PATHWAYS OF SPREAD OF INFECTION

1. **Direct extension:** The spread occurs through the bone either due to hyperaemic decalcification in acute infection, resorption of bone in cholesteatoma or osteitis in chronic infections.
2. **Venous thrombophlebitis:** The small veins (emissary) transmit infection through bones and dura to the venous sinuses. The venous sinuses mostly implicated are lateral (sigmoid) and superior petrosal sinus. From lateral petrosal sinus the infection may spread to cerebellum and from superior petrosal sinus to temporal lobe.
3. **Preformed pathways:**
   i. Congenital dehiscence like the dehiscent facial nerve and high jugular bulb which has a dehiscent floor of middle ear.
   ii. Normal anatomical pathways like oval and round windows.
   iii. Post-surgical pathways like after stapedectomy, fistula in lateral semicircular canal or dura exposure in mastoid surgery.
   iv. Previous fractures of the temporal bone or by neoplastic erosions.
   v. Patent sutures like the petrosquamous suture.
   vi. Periarteriolar spaces of Virchow–Robin.

Risk of Complications

The risks of complications increase with general debilitating illnesses and immunodeficiency. The timely efficacious treatment surely prevents many complications.

TYPES*

The complications of the chronic otitis media have been divided into two main types: **intratemporal/extracranial** and **intracranial complications**. The incidence of extracranial complications is around 32%, intracranial is around 56% and combined intracranial and extracranial is around 12%.

Intratemporal/ Extracranial complications	*Intracranial complications*
1. Acute mastoiditis	1. Extradural abscess
2. Masked mastoiditis	2. Subdural abscess (empyema)
3. Petrositis[a]	3. Meningitis
4. Facial nerve palsy	4. Brain abscess
5. Labyrinthitis	5. Lateral sinus thrombophlebitis
	6. Otitic hydrocephalus

[a]Petrositis may be taken as an extension of acute mastoiditis.

It was always thought that mucosal type of COM had lesser risk of complications therefore were referred as "safe COM" but recent studies have shown incidence of extracranial complications was 59% in mucosal COM,

*Rarer complications, e.g. subclavian thrombosis and internal carotid artery aneurysm have also been reported.

as against 41% in squamosal COM. The incidence for intracranial complications was 59% with squamosal type and 41% with mucosal type. Therefore, the terminology of safe and unsafe COM is not used now.

SYMPTOMS

1. **Ear discharge** is *not an obvious symptom for complications*; it may or may not be present. Profuse ear discharge with pain may occur in acute mastoiditis.
2. **Pain** in a discharging ear is a sinister sign and may indicate a COM complication. It indicates the formation of abscess under pressure in the middle ear or mastoid. The pain may also be due to dural irritation, as in extradural abscess.
3. **Fever** in COM indicates pus formation in case of abscess around the ear, mastoiditis or extra/subdural or intracranial abscess formation.
4. **Headache**
5. **Nausea and vomiting**
6. **Postaural swelling**
7. **Facial weakness** is usually not a common feature. It mainly occurs due to compression of the dehiscent facial nerve by the pus under pressure in the middle ear or due to granulations involving the facial sheath.
8. **Vertigo**
9. **Signs of meningitis**.

A. INTRATEMPORAL COMPLICATIONS

1. Acute Mastoiditis

It is the inflammation of the mucosal lining of the antrum and the rest of the mastoid air cell system and later involves the bony septa of the mastoid air cells. The inflammation of the mastoid air cells usually occurs following acute otitis media (AOM). Earlier, it was thought that mastoiditis occurs 10–14 days following AOM but it has been now observed that it may occur even after 2 days of AOM onset.

Age group: It is a disease of childhood, and most patients are less than 4 years of age. This is due to the presence of higher inflammatory markers in the children.

Organisms: *Streptococcus pneumonia*, *Streptococcus pyogenes*, *Pseudomonas* and *S. aureus* are the most common organisms.

Stages

1. **Acute otitis media infection:** It may cause the inflammation of mucosa of the mastoid air cell system. This stage is usually not associated with signs of mastoiditis.
2. **Acute mastoiditis with periostitis:** Ironing of the mastoid crease occurs. There may be mild swelling of the mastoid with pinna pushed forward. At this stage no abscess formation is seen.
3. **Acute mastoiditis with osteitis:** As the inflammatory process increases, there is mucosal oedema of middle ear and mastoid air cells. This blocks the natural removal of the discharge, leading to the accumulation of discharge under pressure. The **hyperaemic decalcification** of the bony septa occurs, resulting in formation of a common mastoid cavity filled with pus. Further the inner and outer mastoid cortex may get eroded. This leads to formation of abscess around the ear (Fig. 12.1).

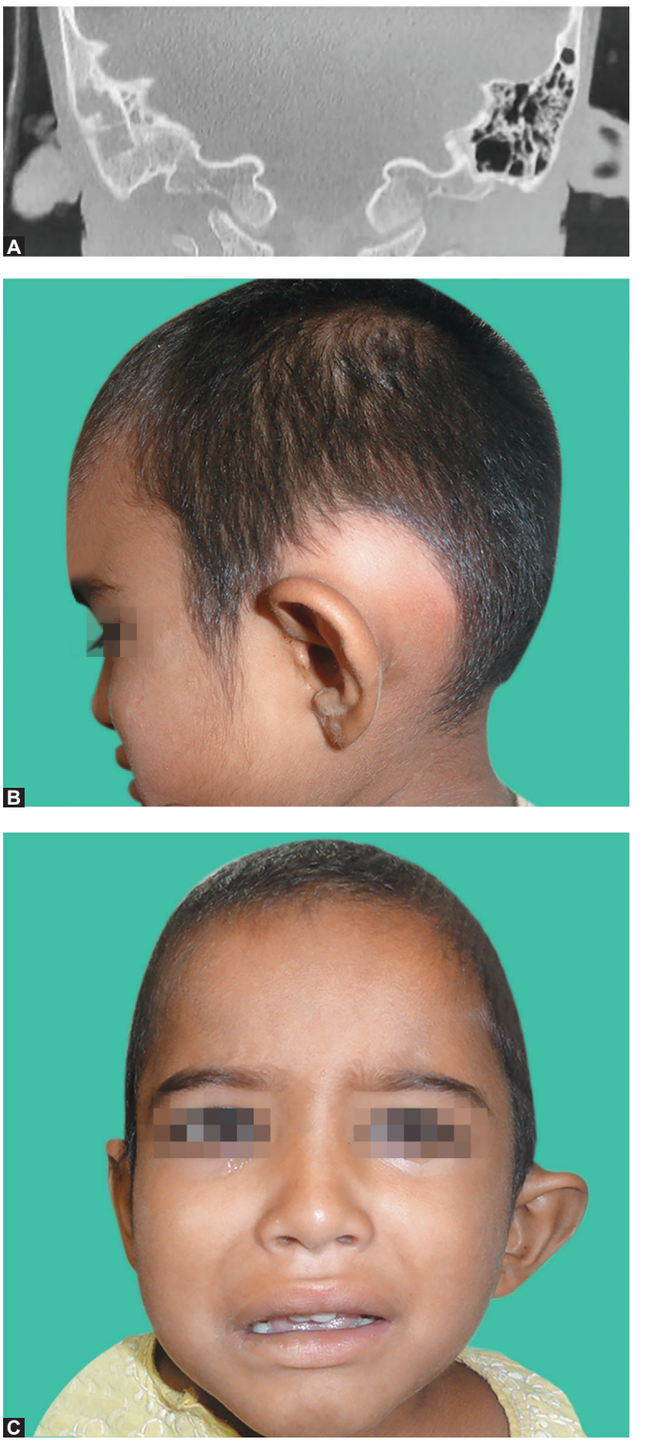


Fig. 12.1: (A) HRCT temporal bone–Right acute mastoiditis with common cavity formation; (B and C) Postauricular abscess following acute mastoiditis



Fig. 12.1D: Incision and drainage of a post-aural abscess

Symptoms of Mastoiditis

1. **Otalgia:** A persistent pain in or behind the ear is the most common symptom of acute mastoiditis. Pain is typically worse at night, probably due to venous congestion. It is typically worse for patients with intact but bulging tympanic membrane due to pus under tension in the middle ear. It usually resolves on antibiotics or after rupture of tympanic membrane. The bulging tympanic membrane may be surgically incised (myringotomy) to release the pus.
2. **Postauricular swelling:** 80–90% of these patients will have some postauricular swelling.
3. **Irritability**
4. **Ear discharge:** The discharge of AOM increases with association of infection of the mastoid air cell system. It becomes more purulent and copious. The discharge may decrease due to blockage in its outflow but then the symptoms of otalgia and fever usually worsen.
5. **Fever:** It is usually low grade and intermittent. It is caused by absorption of toxins from trapped pus under pressure.
6. **Hearing loss:** Patients will complain of mild-to-moderate hearing loss. It persists till there is resolution of pus from middle ear or oedema of mesotympanic and epitympanic mucosa.

Signs

1. **Discharge:** The discharge is initially mucopurulent and then becomes purulent. The discharge may appear pulsatile if seen via small perforation in the tympanic membrane. It is also called the *light house sign*. This discharge has the tendency to fill up again after dry mopping or suction (*Reservoir sign*).
2. **Mastoid tenderness:** It is an important sign of mastoiditis. It can be elicited by applying pressure over the mastoid process. It is elicited over the mastoid bone overlying the antrum (suprameatal triangle area/cymba concha), body and tip of mastoid (*three-finger test*).
3. **Sagging of posterosuperior meatus:** This occurs due to the inflammation of the periosteum (periostitis) of the bony canal adjacent to the antrum.
4. **Mastoid swelling:** It is initially caused by the oedema of the periosteum, leading to the loss of bony grooves (*Ironing of the mastoid*). Further, the pus might form below the periosteum, leading to increase in swelling and pain. This is a subperiosteal abscess also called postaural abscess.
5. **Hearing loss:** It is mainly conductive in type due to pus in the middle ear.
6. **Tympanic membrane perforation:** It occurs due to necrosis of the tympanic membrane tissue by pus under tension in the middle ear.
7. Generally, the child is sick and toxic and requires hospitalization.

Investigations

1. X-ray of the mastoids: It will reveal clouding of air cells due to pus formation. As the disease progresses, there is destruction of the septa of mastoid air cells, which leads to the formation of a common cavity.
2. Blood count: There is leukocytosis with increased ESR levels.
3. Pus culture is sent to know the organisms involved and their drug sensitivity.

Treatment

1. Intravenous antibiotics with close monitoring.
2. Myringotomy: This may be required in cases with pus under pressure (bulging membrane) in middle ear with tympanic membrane intact. It is done preferably in the posteroinferior quadrant/posterior trident because that is the post dependent part of the middle ear cavity.
3. Incision and drainage: All the abscesses in and around the ear require early incision and drainage.
4. Cortical mastoidectomy (simple mastoidectomy) may be required in few cases. The indication include:
   a. Non-drying ear not responding to antibiotics.
   b. Mastoiditis with complications like the facial nerve palsy and labyrinthitis.
   c. Masked mastoiditis.

Differential Diagnosis of Acute Mastoiditis

1. **Otitis externa:** This can be differentiated from mastoiditis by:
   - No history of AOM
   - Tragal sign is positive
   - The external ear does not have goblet cells hence there is absence of mucoid or mucopurulent discharge.
   - There may be acute lymphadenitis of preauricular and postauricular nodes.

- Tympanic membrane will be normal.
- X-rays of mastoids will reveal normal study with absence of haziness of the mastoid air cell system.

2. **Postauricular lymphadenitis**.
3. **Infected sebaceous cyst**.

Various abscesses around the ear

i. *Postauricular abscess*: It is most commonly seen abscess and is seen over the mastoid process. The pinna is pushed anteriorly and inferiorly due to the mass effect of the swelling. The abscess may erode the outer/lateral mastoid cortex and burst onto the skin, leading to a *post aural fistula* formation (Figs 12.2 and 12.3).

ii. *Bezold abscess*:* It is formed when there is erosion of the medial aspect of the mastoid cortex resulting in tracking down of pus along the sheath of sternomastoid muscle. This leads to the formation of the swelling in the upper part of the neck.

This abscess is more commonly seen in well pneumatized mastoids because of thin bone. It may rarely involve the trapezius and splenius capitis muscles.

Differential diagnosis

1. Infected upper jugular lymph nodes (level II neck nodes).
2. Infected branchial cyst
3. Parapharyngeal abscess
4. Infected lymphangioma.

Management: Contrast enhanced CT scan of the neck is essential to evaluate the extent of the abscess. This may be combined with the high-resolution computed tomography (HRCT) scan of the temporal bone, to evaluate the ear involvement before surgery.

Treatment

1. Broad spectrum intravenous antibiotics are started.
2. Cortical mastoidectomy is performed to drain the pus from the mastoid air cell system and middle ear. The osteitic bone and granulations are also removed.

   The pus removed should be sent for culture sensitivity for further antibiotic management. The tissues removed are to be sent for histopathology to especially rule out tubercular mastoiditis.
3. Neck abscess is incised and collected pus is drained. A suction drain is kept in the wound for a few days.

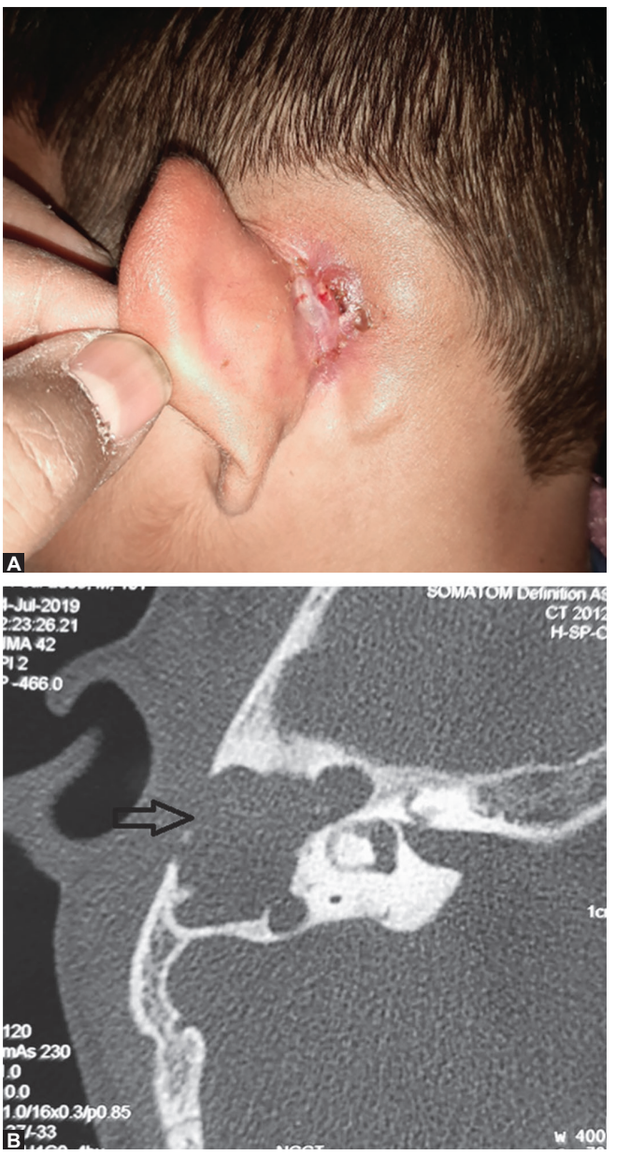


Fig. 12.2: (A) Left ear postaural fistula; (B) HRCT of temporal bone of the patient showing postaural fistula

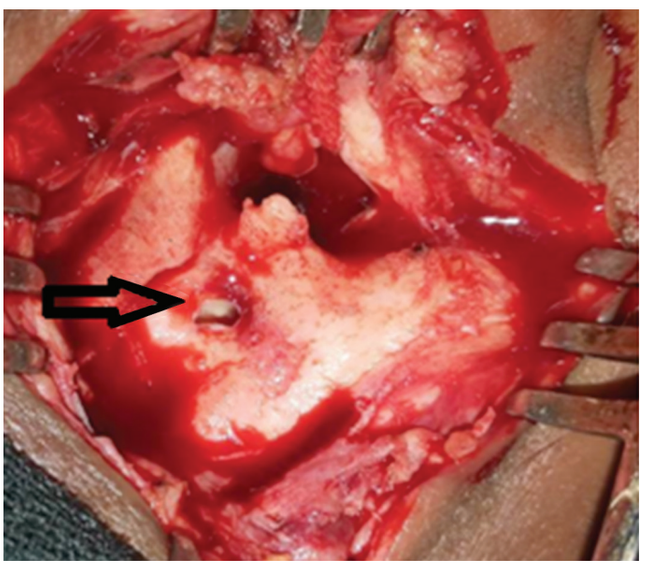


Fig. 12.3: Erosion of the lateral cortex of mastoid

*This was first described in 1882 by Dr Frierich Bezold. He was also first to describe the Citell's abscess.

iii. *Citelli's abscess*: This involves the direct extension of pus from the diseased mastoid process along the posterior belly of the digastric muscle.

In a few cases the pus may track into occipital and cervical regions. The pus extension from the mastoid tip to these regions is either through the facial and subcutaneous planes or via the mastoid emissary vein.

Treatment: Intravenous antibiotics are started, and the pus is drained.

iv. *Luc's abscess* (*temporal abscess*): It is a very rare abscess, where the pus collects deep to the temporalis muscle. It has nothing to do with the mastoid process. Actually, it is the spread of infection from the infected middle ear via the notch of Rivinus and the branch of the deep auricular artery. These patients do not require mastoidectomy for this complication. Incision, drainage or repeated aspiration and intravenous antibiotics are the treatment of choice. Pus under tension in the middle ear may require a temporary grommet insertion.

v. *Zygomatic abscess* (*temporoparietal abscess*): This is a rare complication which involves the zygomatic air cells near the root of zygoma. A fluctuant swelling appears anterior and superior to pinna. It usually occurs in association with postauricular abscess.

Treatment:

1. Antibiotics.
2. Cortical or modified radical mastoidectomy.
3. Incision and drainage of the abscess.

vi. *Retropharyngeal or parapharyngeal abscess*: This abscess is formed when the peritubal air cells are involved and the pus from here tracks down into the retropharyngeal space and parapharyngeal space.

2. Subacute (Masked) Mastoiditis

It is a subclinical infection and inflammation of the mucosal lining and bony structure of the mastoid air cells with an intact tympanic membrane.

Aetiopathogenesis: This is formed due to incomplete treatment of AOM and obstruction of the aditus by the mucosal oedema and granulation tissue. This leads to continuous isolated inflammation of the mastoid air cell system. This may go unnoticed for weeks, months or even years, thereby causing silent bony destruction till it resolves spontaneously or causes complications.

Symptoms of the disease are usually vague. Patients may present with dull pain behind the ear with mild hearing loss.

On examination the tympanic membrane is normal. The pure tone audiometry is usually normal or may reveal mild conductive hearing loss. The impedance audiometry is normal, as there is no fluid secretion in the middle ear.

X-ray mastoids: It reveals clouding of air cells with loss of bony septa. HRCT temporal bone is necessary to reveal the extent of the bony destruction and associated intratemporal complications.

Treatment

1. Intravenous antibiotics.
2. Cortical mastoidectomy is done to drain pus from mastoid air cells and to achieve the patency of the aditus.

3. Petrositis

This involves spread of infection from the mastoid to the petrous part of the temporal bone through anterior and posterior group of air cells which connect mastoid air cells to petrous air cells. Hence it is an extension of infection of acute mastoiditis to the petrous part of temporal bone. The petrous bone has numerous air cells and has highly vascular marrow. This makes it prone to spread of infections from the middle ear and mastoid. The extent of the pneumatization of the petrous bone is variable and only 30% of the cases have pneumatization till the apex. The extension of infection to the petrous apex mainly leads to complications associated with petrositis (*Petrous apicitis*).

Clinical Features

As petrous bone is directly related to various vascular and neurological structures, the infection of this area can have severe consequences. The petrous apex infection causes the local meningeal irritation leading to involvement of adjacent cranial nerves. The *Gasserian ganglion* (of Vth nerve) and abducens nerve (VI Cr N in *Dorello's canal*) are frequently involved in apical petrositis. Dorello's canal is a bow-shaped bony enclosure at the tip of the petrous part of the temporal bone formed by the petrosphenoidal ligament (*Gruber's ligament*, or petroclinoidal ligament). The canal allows passage of the abducens nerve (CN VI) and the inferior petrosal sinus as it travels to merge with the cavernous sinus.

Clinical Features

1. Persistent ear discharge with deep seated ear pain.
2. Retro-orbital pain due to the involvement of the trigeminal nerve.
3. Diplopia due to lateral rectus palsy (patient will not be able to move the involved eye laterally) due to the involvement of the abducens nerve (Fig. 12.4).

   The presence of above three symptoms of persistent ear discharge, retro-orbital pain and diplopia form the part of ***Gradinego's syndrome***.
3. Other symptoms include fever, headache and sometimes signs of meningeal irritation may be present.
4. The carotid artery and dural venous sinus may also be involved.



Fig. 12.4: Left rectus palsy in acute petrositis



Fig. 12.5: Left facial palsy (Grade VI)

Differential Diagnosis

1. Cholesteatoma in petrous apex
2. Meningioma of petrous apex
3. Extension of acoustic neuroma
4. Metastatic carcinoma affecting petrous bone
5. Histiocytosis X.

Diagnosis is made on HRCT temporal bone and MRI head. It reveals the involvement of the petrous apex and helps rule out other intracranial complications.

Treatment

1. Surgical intervention is rarely required nowadays and it is treated by intravenous antibiotics for 2–3 weeks.
2. In cases of associated mastoiditis, cortical mastoidectomy may be done and the petrous part of abscess drained via the same route.

4. Facial Nerve Palsy

The facial nerve paralysis is a complication seen with both acute otitis media and chronic otitis media.

In 30–40% of the patients have congenital dehiscence of fallopian canal* (absence of the bony covering over the facial nerve). This puts the nerve at risk during infections of the middle ear cleft. Though the risk of facial nerve palsy is very low (0.005%) in acute otitis media.

The possible causes for facial nerve paralysis in **acute otitis media** are:

1. *Raised middle ear pressure*: This occurs due to pus formation in the middle ear which is not able to escape due to blocked drainage pathways because of oedema. The increased pressure is more so in cases of intact tympanic membrane.
2. *Osteitis*
3. *Tschiassny's theory*: It suggests that there is an infectious involvement of facial nerve via the bony facial canal dehiscence or due to neurovascular communication between the nerve and inflamed middle ear mucosa.

These causes lead to the oedema of the nerve of the nerve, which is already in a tight bony canal, leading to nerve conduction defects (Fig. 12.5).

Management: In AOM the first line of management is intravenous antibiotics and steroids. Myringotomy with grommet insertion is done to drain the pus under tension from the middle ear and relieve the nerve of pressure. Early intervention leads to recovery in most cases and facial nerve decompression is usually not required.

In **chronic otitis media**, the involvement of the nerve is due to the erosion of the fallopian canal by the cholesteatoma or direct involvement of the exposed dehiscent nerve.

- The patient is treated surgically by mastoidectomy to remove the disease from the middle ear and mastoid. The cholesteatoma matrix covering the facial nerve should be removed very carefully under magnification of microscope or otoendoscope.
- In case of florid granulations covering the facial canal, an experienced surgeon can try to remove granulations under high magnification. Sometimes it may be difficult to remove all the granulations without damaging the nerve and hence, they can be left behind over the facial nerve.

Note: The granulations or polyps from the facial sheath are preferably cut under magnification. They should never be avulsed.

5. Labyrinthitis

Inflammation of the labyrinth is termed as labyrinthitis.

This condition can occur following acute otitis media also. The common route of spread is via the round window. The other route of spread to the labyrinth is

*Most common site of dehiscence of the facial nerve is the tympanic segment just above the oval window.

via the preformed fistulae, as in post-stapedectomy or erosion the bony labyrinth by cholesteatoma in chronic destructive ear disease.

The labyrinthitis can be of following types:

1. *Perilabyrinthitis (Labyrinthine Fistula)*

Vestibular irritation by the inflammatory disease close to the endosteum of the bony labyrinth is termed as paralabyrinthitis and is seen in COM. The chronic osteitis around the bony labyrinth sometimes causes destruction of the otic capsule with normal vestibular function, this is called as perilabyrinthitis. It is generally seen in COM with cholesteatoma (Fig. 12.6).

The causes are chronic osteitis, malignancies of the ear, and surgical or accidental trauma to labyrinth.

Symptoms: As the labyrinth gets exposed, the pressure changes of the middle ear frequently produce vertigo or unsteadiness. This is known as **Tulio's phenomenon**.

On examination

1. Fistula test is positive: Fistula test is performed by pressing the tragus intermittently. This causes a change in pressure in the middle ear thus stimulating exposed labyrinth. It produces sensation of vertigo and nystagmus which is seen towards the normal side.*
2. Siegle's speculum can also be used to perform fistula tests and elicit vertigo.

Treatment: The labyrinthine fistula is treated by performing mastoid exploration and removing the disease process like granulations or cholesteatoma from the middle ear cleft. The fistula is tackled at the end of the surgery. The cholesteatoma matrix is gently elevated from the fistula site and direct suction is avoided over the fistula. It is immediately covered with temporalis fascia graft, perichondrium or thin sliced conchal or tragal cartilage. Tissue glue can be used to further augment the cover over the fistula.

2. *Serous Labyrinthitis*

It is the inflammation of the labyrinth without the pus formation and is a reversible process if the treatment is started early.

Clinical features include vertigo and dizziness. On fistula test there is The nystagmus can be seen on fistula test beating towards the affected ear. The hearing assessment will reveal mild to moderate sensorineural hearing loss.

Treatment

1. The patient is advised bed rest and asked to avoid head movements. (All tests of audiology should be performed preferably in bed lying position, not sitting).
2. Parental prochlorperazine or cinnarizine is given to control vertigo and vomiting. Intravenous fluids may be required for hydration.
3. Steroids are given to prevent sensorineural hearing loss.**
4. Myringotomy is required in cases with pus in the middle ear with bulging tympanic membrane.
5. Mastoidectomy may be required to make the ear safe but this is deferred till acute symptoms subside.
6. *Cawthorne–Cooksey exercises* (vestibular head exercises) are added after full infective recovery. They accelerate central compensation of vestibular function.

3. *Suppurative Labyrinthitis*

It is the formation of frank pus or pyogenic infection of the labyrinth. It usually follows uncontrolled or untreated serous labyrinthitis. The patient complains of violent vertigo along with nausea and vomiting. There is severe sensorineural hearing loss. The patient usually lies in bed with the infected ear upwards.*** The patients avoid any head movements.

Treatment: The management is similar to that of serous labyrinthitis.

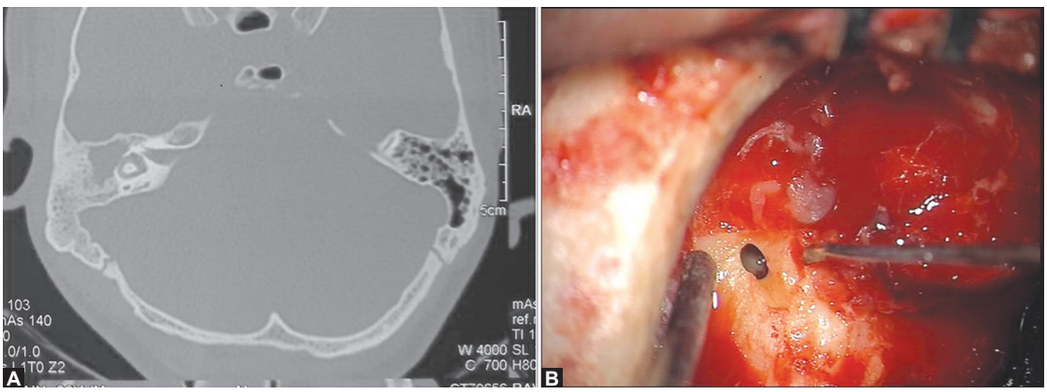


Fig. 12.6: Lateral semicircular fistula as seen on (A) HRCT temporal bone; (B) Peroperative

*The deviation towards the normal side is the most common finding as the dome of the lateral semicircular canal is involved. A lateral canal fistula anterior to the ampulla, however, will cause deviation towards the side of fistula. In erosion of the vestibule, rotatory horizontal deviation towards the diseased ear is seen.
**Suppression of the expression of NF-kB (expressed by fibrocytes in spiral ligament) by use of steroid therapy is suggested as the mechanism of hearing preservative role of steroids in early cases.
***As the infected ear has become hypofunctional; therefore, the vestibular signals from the labyrinth decreases. Hence, the nystagmus is towards the normal side and patients will find having less nystagmus and vertigo when looking towards the affected ear.

4. Healed Labyrinthitis

This follows after successful control of inner ear suppurative infection. Dense fibrous tissue response occurs, which obliterates the labyrinthine spaces with resulting complete loss of vestibular and auditory function. Calcification and osteogenesis of some or all labyrinthine spaces results **(*labyrinthitis ossificans*)**.

Viral Labyrinthitis

Viruses commonly involved are mumps, influenza, adenovirus and cytomegalovirus. The viruses reach the inner ear either directly from the bloodstream or by way of subarachnoid space. They affect the structures within the scala media. The hair cells of the organ of Corti, stria vascularis and spiral ganglion may be affected. Symptoms include sustained vertigo and nystagmus lasting 3–5 days with gradual decrease of symptoms. The viral labyrinthitis is usually unilateral and may result in permanent hearing loss.

B. INTRACRANIAL COMPLICATION

1. Extradural Abscess

The bone erosion can occur in COM due to cholesteatoma and in AOM by hyperemic demineralization. It leads to the spread of infection from the middle ear cleft to over the dural surface. The infections may lead to the formation of pus collection between the dura and the bone (Fig. 12.7). This is the most common intracranial complication. The extradural abscess usually precedes the lateral sinus thrombophlebitis and brain abscess.

As the size of the abscess increases, it strips the dura from the overlying bone, eventually producing sizable mass which produces pressure effects over the brain.

Clinical Features

Most of the patients of extradural abscess are asymptomatic and it may be discovered as a chance finding during mastoid surgery. Patients may have vague and non-specific symptoms, though specific neurological signs and papilloedema may be seen in few cases.

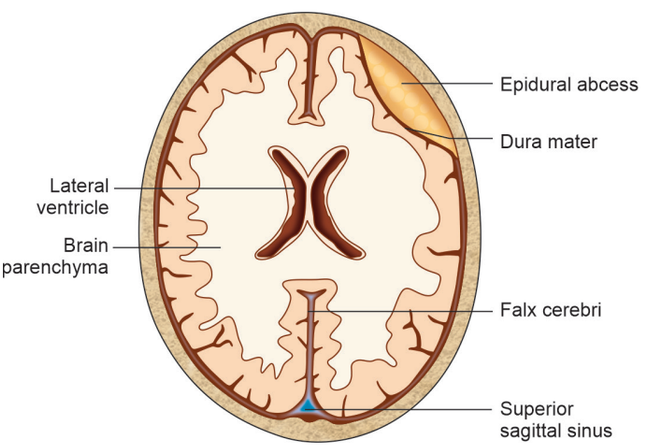


Fig. 12.7: Location of the extradural/epidural abscess

Any patient with a persistent headache over the side of the otitis media should draw suspicion and should be investigated.

Irritation of the Gasserian ganglion (of the V nerve) and VI nerve may produce the *Gradenigo's syndrome* of facial pain, diplopia, and aural discharge. Even the outer table of the bone may get eroded producing a subperiosteal abscess or *Pott's puffy tumour*.

Diagnosis: In cases of suspicion of intracranial complications CECT of the brain is essential (Fig. 12.8).

Management

a. Extradural abscess can be drained through the mastoid route. Surgical exploration of the extradural abscess is done by performing mastoidectomy and removal of the dural plate and granulations around the pus, to allow its drainage. The dura is not touched.
b. Broad spectrum antibiotics are given for 4–6 weeks.
c. There is always a need to be vigilant and look for co-existing intracranial complications associated with extradural abscess.

2. Subdural Abscess (Empyema)

This is the presence of a collection of pus in the subdural space. It is usually caused by non-hemolytic streptococci, especially microaerophilic bacteria. Usually the infection enters the subdural space via the dura. The dura is a tough structure which prevents any infection transfer inside the brain. With prolonged exposure to infection, granulations are formed which ultimately produce dural fibrosis. Fibrosed sections of dura may get necrosed and allow transmission of infection.



Fig. 12.8: Left extradural abscess

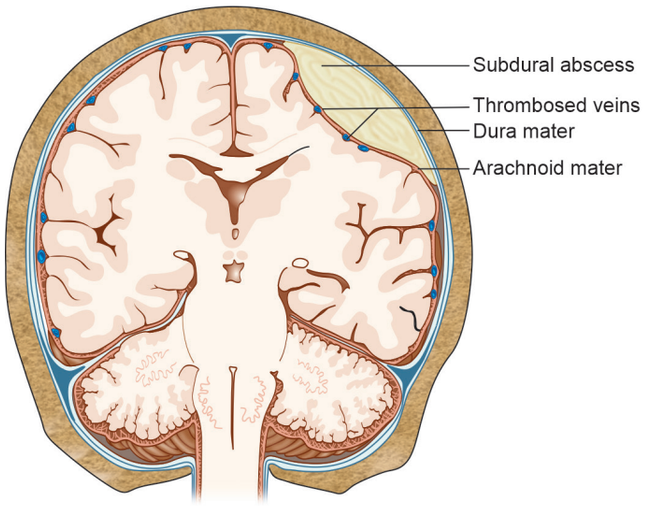


Fig. 12.9: Subdural empyema between the dura and arachnoid mater

Clinical Features

Initially, the abscess is small and confined to the site of dural penetration, but it may soon enlarge to spread to falx cerebri, tentorium cerebelli and intrahemispheric spaces. This leads to rapid deterioration of the patient (Fig. 12.9). The patient has a severe headache, fever and drowsiness. These may be followed by development of focal neurological symptoms which can be irritative (like Jacksonian epileptic fits) or paralytic. The paralysis of a limb may rapidly progress to become hemiplegia.

Diagnosis

It is confirmed by contrast enhanced CT scan of the brain. In the case of equivocal or normal pictures in CT scans, MRI is more reliable. Lumbar puncture is contraindicated because raised ICT may lead to coning of brainstem.

Treatment

1. Intravenous antibiotics are administered.
2. Neurosurgical intervention (burr holes or craniotomy) is needed to drain the empyema.
3. Once the patient has been stabilized, mastoidectomy is done to remove the disease from the middle ear cleft.

3. Otogenic Brain Abscess

The most common site of otogenic brain abscess is the temporal lobe followed by the cerebellum. The otological causes still remain the most common cause for brain abscess.

- For temporal lobe abscess, the common route of spread is by direct extension of the infection through the osteitic tegmen antri and tympani. This leads to the middle cranial fossa extradural abscess formation which subsequently leads to temporal lobe abscess formation.
- The cerebellar abscess usually follows lateral sinus thrombophlebitis. The infection to the cerebellum spreads via the retrograde thrombophlebitis or via dura underlying the *Trautmann's triangle* (Fig. 12.10).

Streptococci and *Staphylococcus* are the most common organisms involved. Gram-negative organisms like *Proteus mirabilis*, *E. coli*, and *Pseudomonas* are also frequently involved.

Clinical Features

1. **Stage of encephalitis:** In the initial phase of cerebral oedema and encephalitis, there is headache, fever, malaise and vomiting.
2. **Stage of localisation:** The collection may get confined by encapsulation (abscess localization) and there might be a decrease in symptoms. The patient might continue to have raised temperature and headache.
3. **Stage of enlargement:** In the next stage of enlarging abscess, the intracranial pressure increases, leading to severe headache, projectile vomiting, papilledema, drowsiness, progressing to stupor and coma. The pulse is slow and temperature subnormal. Eventually death may result from tentorial herniation.
4. **Stage of rupture:** Then finally the abscess ruptures into ventricles or subarachnoid space with resulting overwhelming meningitis and death (Fig. 12.11).

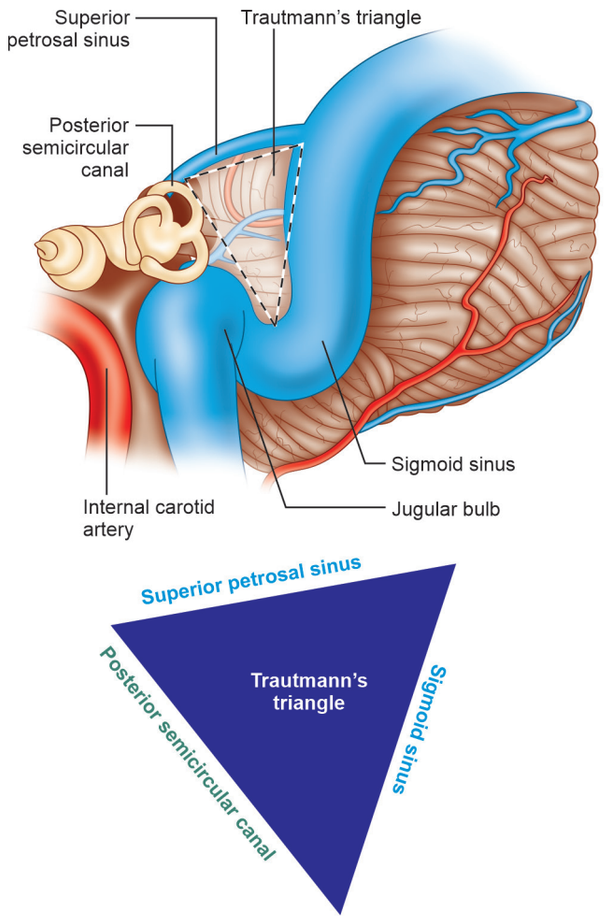


Fig. 12.10: Boundaries of the Trautmann's triangle

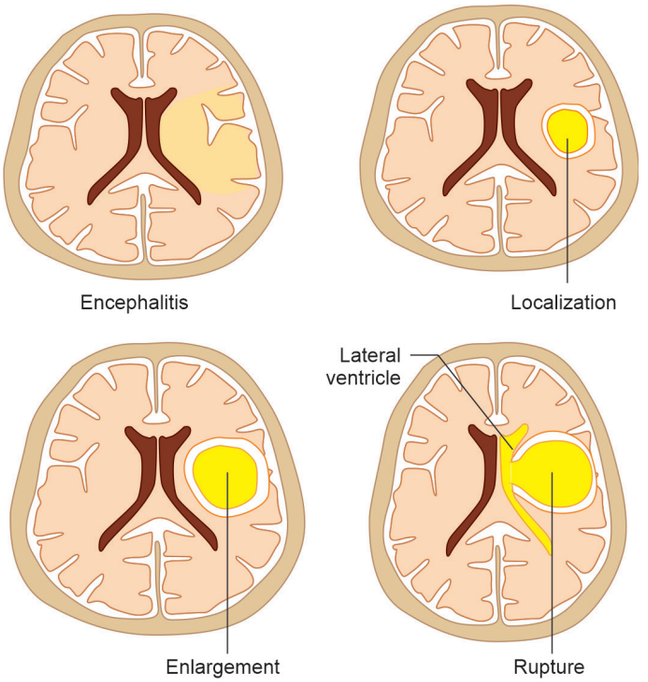


Fig. 12.11: Progression of the brain abscess



Fig. 12.12: Axial cut of NCCT brain showing left cerebral abscess

Temporal lobe abscess are generally multiple while cerebellar abscess are usually single large. *The most common site of otogenic abscess is the temporal lobe and the most common cause of cerebellar abscess is otogenic.*

Patients with cerebral abscess (temporal lobe) may have nominal aphasia, visual field defects (commonly have quadrantic homonymous hemianopia), and contralateral motor paralysis.

The patients with **cerebellar abscess** show weakness, difficulty in walking and incoordination on the side of lesion. The cerebellar function tests are positive for intention tremor, past pointing, irregular dysdiadochokinesia and ipsilateral ataxia. Spontaneous nystagmus may be seen which beats to the side of the lesion. Abscesses of the posterior cranial fossa cause early raised ICT (compared to cerebral abscess), leading to *coning* (impaction of the flocculus and brainstem into the foramen magnum) and death.

Diagnosis

1. **CT scan** head with contrast is the investigation of choice. It shows an abscess cavity with peripheral enhancement and is classically known as "*ring sign*" (Figs 12.12 and 12.13).
2. **MRI scans** are sometimes required for differentiation of brain oedema and abscess or normal brain tissue.
3. **Lumbar puncture** must be done only after decreasing the raised ICT. The CSF shows raised protein levels, raised WBC count in CSF but the count is less raised compared to that seen in meningitis. The glucose levels of CSF are normal.

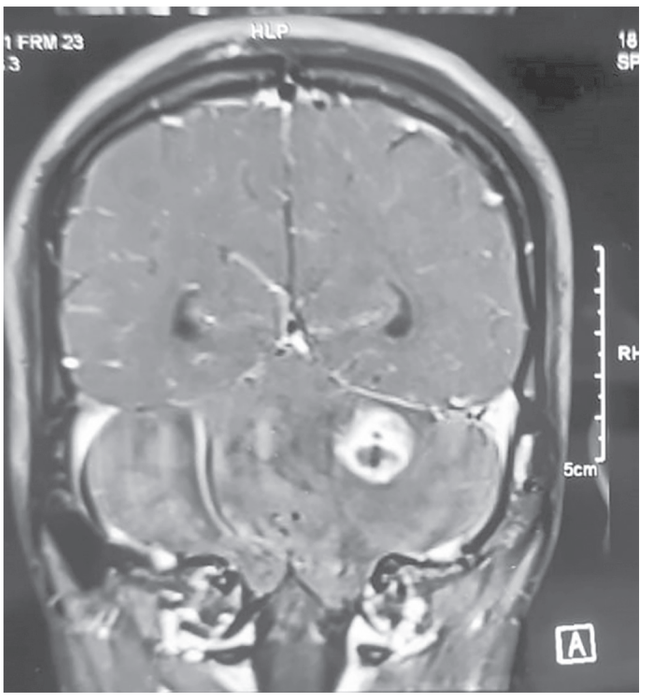


Fig. 12.13: Coronal cut of MRI Brain showing left cerebellar abscess

Treatment

1. High dose of systemic antibiotics.
2. If the intracranial pressure is raised, dexamethasone in dose of 4 mg i/v 6 hourly or 20% mannitol in dose of 0.5 g/kg are used especially in rapidly deteriorating patients while neurosurgical intervention is awaited.
3. In early stages of encephalitis surgical drainage is avoided, but it will be required if the abscess is expanding and the patient is not responding to antibiotics.

4. Surgical treatment: The abscess may be repeatedly aspirated or excised by neurosurgical procedures. If repeated drainage fails then open neurosurgical procedure for excision may be required. The cerebral abscesses may be drained through trautman's triangle.
5. Once the intracranial complications are controlled, the mastoidectomy is performed to remove the primary focus of infection from the ear. This would prevent any recurrent intracranial complication.
6. The temporal lobe abscess has a high risk of epileptic seizures. Almost 70% of patients will have fits therefore long-term anticonvulsant medication is needed.

4. Lateral Sinus Thrombophlebitis

The thrombophlebitis of the lateral sinus (sigmoid and transverse sinus together) can occur as a serious complication of middle ear disease. It is usually because of infection spreading from middle ear to sigmoid sinus via retrograde thrombophlebitis of mastoid emissary veins.

Stages of pathological process and associated clinical features (Fig. 12.14):

1. The perisinus abscess is formed around the vein due to osteitis, further leading to the formation of the **mural thrombus formation** (thrombus over a wall of the vessel). The patient develops a headache along with symptoms of underlying ear disease.
2. The thrombus subsequently enlarges to become an **occluding thrombus**. It further gets organised and partly breaks due to the suppuration by the involved organisms. This leads to the dissemination of the infected emboli into the systemic circulation.
3. Further **propagation of the thrombus** occurs proximally and distally. It spreads proximally to superior sagittal sinus and *torcula Herophili* (confluence of sinuses) and distally to internal jugular vein in the neck.

Clinical Features

- High grade fever which has a *picket-fence pattern*. Rapid rise of fever and subsequent fall, is accompanied with rigors and profuse sweating. This corresponds to release of infected thrombus in blood circulation
- Patients have increased headache, emaciation and progressive anaemia.
- There may be infiltrates in the lung fields and septic embolic spread to the large joints and subcutaneous tissue.
- **Griesinger's sign** is sometimes seen due to thrombosis of the mastoid emissary vein. There is bluish discoloration and pitting oedema over the mastoid region. It is observed better in fair skinned patients.
- Tenderness along the jugular vein with associated acute lymphadenitis (upper jugular lymph nodes).
- Perivenous inflammation at the jugular foramen may lead to the paralysis of the X, XI and XII cranial nerves.
- Papilloedema develops in half the number of patients.
- **Tobey–Ayer test or Queckenstedt test:** This test involves the measurement of the cerebrospinal pressure (by doing lumbar puncture) and the changes on compression of each side's internal jugular vein in neck by fingers on the neck. In healthy individuals, compression of each internal jugular vein is followed by rapid rise of cerebrospinal fluid pressure by 50–100 mm Hg and fall after release. There is no such effect produced on the unhealthy thrombosed side. The test has low sensitivity and specificity and is not done now.
- **Crowe–Beck test** involves observation of retinal veins engorgement as seen on ophthalmoscopy) on pressure of the jugular vein on the healthy side. The engorgement subsides on release of pressure. Pressure on the affected side does not produce such change.

There is no single pathognomonic sign of lateral sinus thrombophlebitis.

Investigations

1. The blood count shows anaemia with raised erythrocyte sedimentation rate.
2. Blood samples for culture should be taken at the time of chills. This reveals organisms released into circulation with septic emboli.
3. Blood smear to rule out malaria.
4. CSF examination to rule out meningitis.
5. Contrast enhanced CT scan of brain will reveal an *empty triangle* or *delta sign* formed due to enhancement of the inflamed sinus wall without the contents. It also

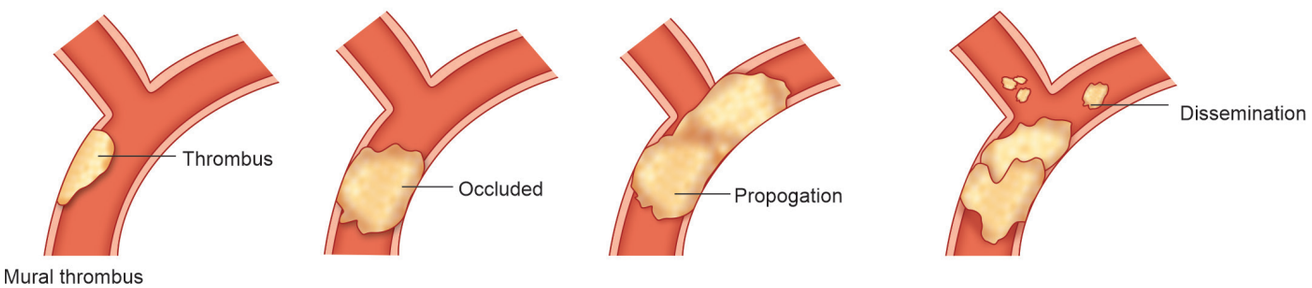


Fig. 12.14: Pathological stages of lateral sinus thrombophlebitis

helps to identify accompanying other complications like the brain abscess, extradural and subdural abscess. *The MRI with contrast will also reveal the same *delta sign*. MR venography is the gold standard as it can confirm the presence of thrombus by absence of blood flow on the involved side (Figs 12.15 and 12.16).

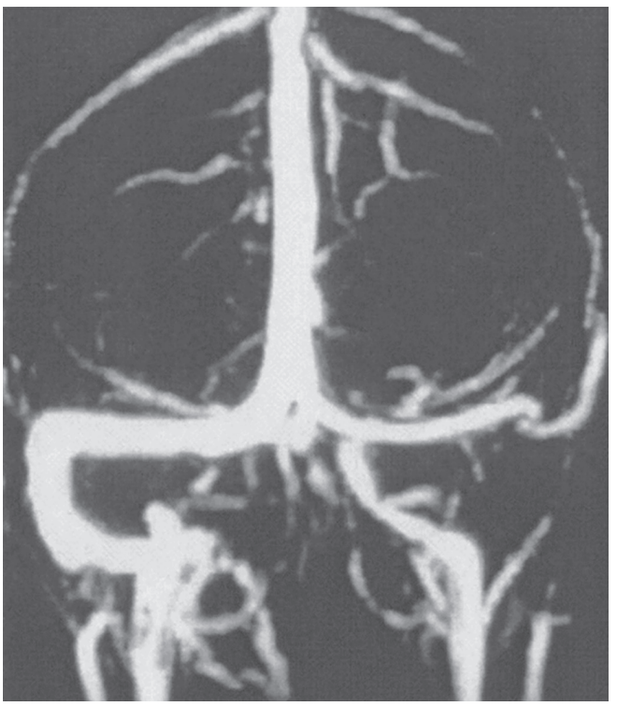


Fig. 12.15: Absence of flow in lateral sinus on the involved side as seen on MR venography



Fig. 12.16: Empty triangle sign on CECT of brain (Left side)

*20% of patients with LST have associated intracranial complications.

Treatment

1. **Intravenous antibiotics:** The antibiotics need to be changed once the culture sensitivity reports are available. Non-pneumococcal streptococcal, anaerobic and staphylococcal species are commonly implicated organisms. Combination of ceftriaxone, metronidazole or clindamycin is the usual first line of antibiotic treatment.
2. **Anticoagulants:** They are not routinely recommended now as they unnecessarily increase the risk of cerebral haemorrhage. They may be rarely indicated when there is extension of the thrombus to the cavernous sinus or jugular vein.
3. **Surgical management:** The patient is taken up for mastoidectomy as soon as the general condition of the patient allows. The sinus plate is removed to expose the sinus adequately. The perisinus necrotic debris and pus around the area are removed. Presently, it is not recommended to open the sinus to remove the thrombus from inside. Mastoidectomy is completed and all the disease from the middle ear cleft is removed.

The ligation of the internal jugular vein is required only in cases where there is repeated septic embolization in systemic circulation and which does not respond to medical and surgical management.

5. Meningitis

It is the inflammation of the pia-arachnoid (leptomeninges) caused commonly by bacterial or viral infection of CSF. It can also be due to fungal or tubercular infection.

The infection for otological meningitis spreads through following routes:

1. Direct bone erosion of the middle ear cleft.
2. Spread of infection from suppurative labyrinthitis.
3. Rarely, it may be caused from rupture of the brain abscess.

The organisms responsible are *Haemophilus influenzae* type B and *Streptococcus pneumoniae*. The initial inflammation of the pia-arachnoid is soon followed by CSF getting permeated by WBC's and rapid bacterial multiplication. The exudate thus formed is sticky, which leads to the obstruction in CSF flow. Spread of infection may occur to the Virchow-Robin spaces, which can further lead to brain abscess formation.

Clinical Features

1. Initially there is headache and neck stiffness. Irritation of the upper cervical nerve roots by inflammatory exudates is responsible for the neck pain (initially only flexion is affected but finally rigidity develops). Kernig's sign and Brudzinki's sign are positive (Fig. 12.17).

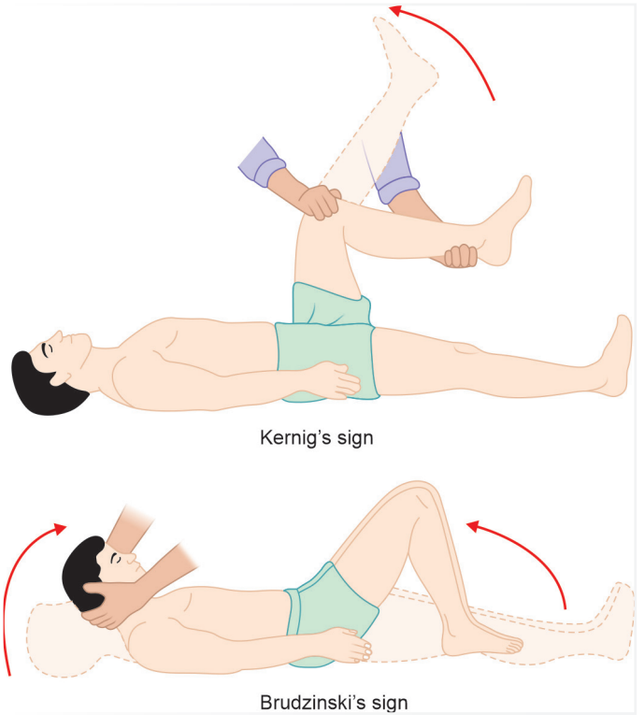


Fig. 12.17: Clinical signs for meningitis

2. High grade fever.
3. Mental hyperactivity with restlessness occurs in early cases.
4. Photophobia.
5. Nausea and projectile vomiting due to raised ICT.
6. Tendon reflexes become less marked in later stages.
7. Once the patient deteriorates, delirium and stupor results.
8. Epileptic fits and focal neurological signs are usually not seen. If observed, then other intracranial complications should be investigated.

Diagnosis

1. All patients with otitis media who develop headache and neck stiffness, should undergo a lumbar puncture. In early stages there is increased fluid pressure on lumbar puncture. (>100–150 mm Hg). As disease evolves, CSF turns cloudy and then turbid. There is an increase of leucocyte count, protein content is raised from normal 150–400 mg/L to 2–3 g/L, chloride content falls from 120 to 80 mmol/L and glucose levels fall from 1.7–3.0 mmol/l to zero. Culture is done for finding the causative organism. Lately polymerase chain reaction (PCR) has been used to detect bacterial DNA in apparently sterile cultures.
2. MRI is the most sensitive imaging technique to show early exudates and adhesions in the basal cisterns. It will also help to show other associated intracranial complications.

Treatment

A. Medical management

1. High doses of systemic antibiotics are the mainstay for treatment. The antibiotics should be active against both aerobic and anaerobic organisms and should cross the blood brain barrier. Third generation cephalosporin is a good choice with addition of vancomycin to cover for resistant strains.
2. Dexamethasone is used along with antibiotics because it reduces chances of neurological sequelae and hearing loss.

B. Surgical management

1. Usually, patients respond well with medical management.
2. Otogenic meningitis will require mastoidectomy once the meningitis is under control.
3. Acute otitis media with bulging tympanic membrane will require myringotomy.

6. Otitic Hydrocephalus

There is raised intracranial pressure due to middle ear infection. It is more common in children and adolescents.

Pathogenesis

The exact aetiology is unknown. It is said to be due to lateral sinus thrombosis. The thrombus extends to superior sagittal sinus thereby impeding CSF resorption (by *Pacchionian bodies*) or causes obstruction of the lateral sinus and affects cerebral venous outflow.

Clinical Features

Main symptoms are headache, nausea, vomiting and sometimes diplopia (due to stretching of VI nerve causing lateral rectus palsy) which may be unilateral or bilateral. The onset of symptoms can be many months or years after the chronic otitis media.

Clinical examination shows papilloedema and raised lumbar puncture pressure. The CSF examination is normal.

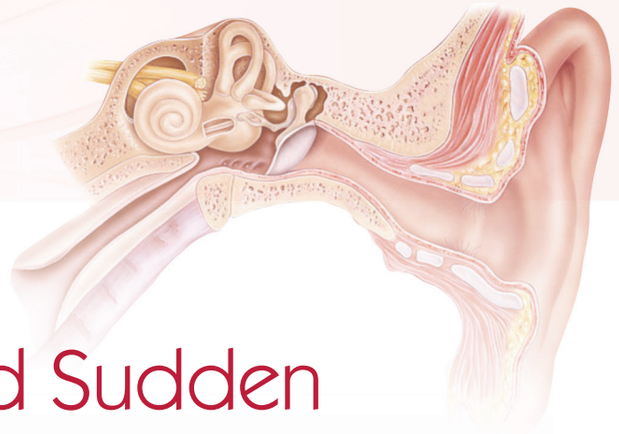
Radiological investigations like MRI and MR venography (MRV) help in diagnosis and rule out other intracranial complications, specifically any space-occupying lesion which is necessary, before carrying out lumbar puncture. MRV facilitates detection of sigmoid sinus thrombosis. There is absence of ventricular dilatation on MRI.

Treatment

1. Steroids, diuretics, acetazolamide and hyperosmolar dehydrating agents (mannitol) are used to decrease intracranial pressure.
2. Long-term ventriculoperitoneal shunting may be required in few cases.
3. Repeated lumbar puncture has been suggested as a method to decrease, but is not without complications.

Chapter

13



Presbycusis and Sudden Sensorineural Hearing Loss

EN4.12: Elicit document and present a correct history demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of hearing loss.
IM24.17: Describe and discuss the aetiopathogenesis, clinical presentation, identification, functional changes, acute care, stabilization, management and rehabilitation of hearing loss in the elderly.
EN4.14: Describe the clinical features, investigations and principles of management of sudden sensorineural hearing loss.

PRESBYCUSIS

The sensorineural type of hearing loss associated with aging is called presbycusis. (*presby* means 'elder' and *akusis* means 'hearing').

There are four pathological types of presbycusis:

1. *Sensory presbycusis*: There is a loss of hair cells and sustentacular cells at the basal turn of the organ of Corti, which can progress toward the apex. Therefore, higher frequencies are affected earlier. The speech discrimination remains relatively good in most cases.
2. *Neural presbycusis*: There is degeneration of cochlear nerve neurons and cochlear spiral ganglion neurons. The loss of 50% (of the total 35,500 neurons) or more of the cochlear neurons is used as the criteria for neural presbycusis. The speech discrimination is poor.
3. *Strial or metabolic presbycusis*: It occurs due to atrophy of stria vascularis of the cochlea. The hearing loss is directly related to the degree of degeneration of stria. The speech frequencies are largely unaffected. It is slowly progressive and may be hereditary in nature.
4. *Mechanical or cochlear conductive*: It is due to stiffening of the cochlear basilar membrane. Mechanical presbycusis is slowly progressive and audiogram shows a sloping high-frequency sensorineural hearing loss.

There are two other subcategories of presbycusis.

- Mixed presbycusis term is used when there are a combination of the two more types of presbycusis.
- Indeterminate presbycusis term is used in patients who do not have any of the above causes for presbycusis. It may be due to the loss of the peripheral dendrites of the spiral ganglion or alteration in endolymph composition.

Management

1. *Non-specific Management*

It is always beneficial to have a good acoustic environment, i.e. with less background sounds. Patients with presbycusis have difficulty in speech perception in noisy surroundings. Face to face communication along with non-verbal cues help speech understanding. For individuals with severe hearing loss, wireless headphones for use with television and louder doorbells are useful for these patients. Lip reading classes certainly help in profound uncorrected hearing loss patients. Cooperation of the family members goes a long way in boosting confidence and hearing aid hygiene.

2. *Specific Management*

- Binaural digital hearing aids can substantially improve the quality of life of patients.
- Patients with profound hearing loss or having no improvement with hearing aids can be candidates for cochlear implant.

SUDDEN SENSORINEURAL HEARING LOSS

Sudden sensorineural hearing loss (SSNHL) is most commonly defined as sensorineural hearing loss of 30 dB or more, occurring in three or more contiguous audiometric frequencies, precipitating within a span of 3 days. The loss is usually unilateral and patients usually complain of sudden blockage of an ear and hearing loss especially described on getting up in the morning (Table 13.1).

Mainly four factors affect the recovery from SSNHL:

1. Time of presentation of the patient: Earlier the presentation, better is the prognosis.
2. Age of the patient: The patients over 60 years have poorer prognosis.

Table 13.1: Causes of sudden sensorineural hearing loss	
Infections	1. Meningococcal meningitis
	2. Cryptococcal meningitis
	3. HIV
	4. Mumps
	5. Measles
	6. Lyme disease
	7. Syphilis
Trauma	1. Barotrauma
	2. Perilymph fistula
	3. Temporal bone fracture
	4. Stapedotomy
	5. Intense noise exposure
Toxic	1. Aminoglycosides
	2. Loop diuretics
	3. Salicylates
	4. Chemotherapeutics
Functional	1. Conversion disorder
	2. Malingering
Neoplastic	1. CP angle tumour
	2. Leukemia
	3. Myeloma
Autoimmune	1. Wegner's granulomatosis
	2. Relapsing polychondritis
	3. Lupus erythematosus
	4. Polyarteritis nodosa
	5. Cogan's syndrome
	6. Sjögren's syndrome
Metabolic	1. Diabetes
	2. Hypothyroidism
Circulatory	1. Cardiopulmonary bypass
	2. Sickle cell anaemia
	3. Vertebrobasilar insufficiency

3. All patients with associated vertigo have poorer prognosis.
4. Patients with profound hearing loss and with downward sloping audiograms have a poorer prognosis.

The recovery which occurs within the first 2 weeks, usually decides the final outcomes. About 50% of patients have spontaneous recovery with no treatment. The chances of recovery are poor if no improvement is seen within the first 1 month.

Therapies for Treatment

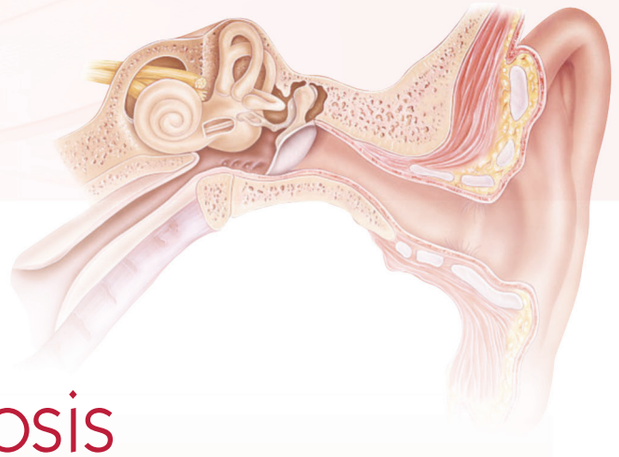
1. Systemic corticosteroids: Oral prednisolone 1 mg/kg/day up to a maximum of 60 mg daily for 10–14 days is recommended. Steroids are the only drug which has proven efficacy in treatment of SSNHL.
2. Intratympanic steroids: This has gained popularity because of reduced systemic side effects and higher levels of inner ear steroid levels. The steroids like dexamethasone or methylprednisolone are administered via grommet or myringotomy via *microwick* or *transtympanic needles*.
3. Hyperbaric oxygen delivers 100% oxygen to a patient at a pressure >1 atm. It helps in increased oxygenation of ischemic tissues of cochlea. It is an expensive and time-consuming modality but has shown promising results in some of the studies. Patient has to attend multiple sessions of 1–2 hours daily for at least a week.
4. Antiviral therapy: Acyclovir has been given in such patients, either alone or in conjunction with steroids.
5. Low molecular weight dextran, papaverine and pentoxifylline have been used to decrease blood viscosity and improve inner ear microcirculation.

Fluctuating Hearing Loss

The most common cause of fluctuating hearing loss is a change in middle ear pressure and resulting in mild conductive hearing loss. Next important cause is serous otitis media. The most common inner pathology causing fluctuating sensorineural hearing loss is endolymphatic hydrops due to Meniere's disease and syphilitic labyrinthitis.

Chapter

14



Otosclerosis

EN4.13: Describe the clinical features, investigations and principles of management of otosclerosis.

DEFINITION

Otosclerosis is a disorder of bone metabolism affecting the endochondral bone of the otic capsule resulting in progressive hearing loss sometimes (Table 14.1).

Table 14.1: History of otosclerosis and stapes surgery

Year	*Surgeon*	*Surgery*
1800	Kessel	1st stapes surgery by doing mobilization
1869	Von Troeltsch	Coined the term otosclerosis
1923	Holmgren	Father of fenestration surgery
1953	Samuel Rosen	Popularized mobilization surgery
1953	Julius Lempert	Single stage fenestration surgery
1956	John Shea	Father of modern stapes surgery
1980	Perkins	First use of LASER in stapes surgery
2006	S Jovanovic	One shot LASER stapedotomy
2006	Robert Vincent	Largest series of stapedotomy

Histologic otosclerosis: The term is used when the histological changes of otosclerosis are present in the otic capsule without any clinical symptoms. It can be confirmed only by temporal bone study during autopsy. Various studies have reported the incidence of histological otosclerosis to about 3% to 10% in white population.

Clinical otosclerosis: It occurs when the disease process affects the stapedio-vestibular joint and interferes with the motion of the stapes, and the patient presents with conductive hearing loss.

Cochlear otosclerosis: The patients present purely with sensorineural hearing loss. It is diagnosed on CT scans of temporal bone where the otosclerotic focus surrounds the cochlea without involvement of the oval window or stapediovestibular joint.

COMMON SITES

The common sites for the otosclerosis foci are (Fig. 14.1):

1. The cochlear wall anterior to the oval window (*Fistula ante fenestrum*) is the most common site.
2. The round window niche is the next most common site.
3. Focus posterior to the oval window.
4. The entire footplate is involved with delineated margins (*Biscuit or rice grain footplate*) or rarely, the entire foot plate may be obscured by a large mass of otosclerotic foci (*obliterative otosclerosis*).

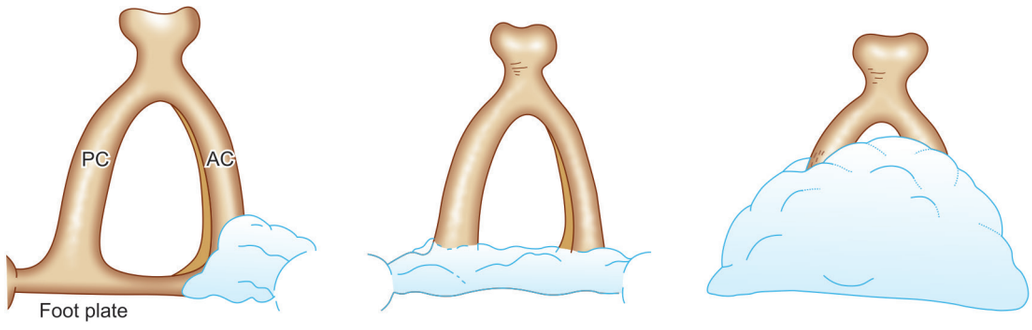


Fig. 14.1: Types of stapes involvement (*PC: Posterior crura; AC: Anterior crura*)

5. Rarely, very large cavities form within the otosclerotic foci, leading to the formation of the "third window".* (*Cavitating otosclerosis*).

AETIOLOGY

Various hypotheses have been advocated but none of these can conclusively explain the aetiopathogenesis of otosclerosis.

1. Genetic predisposition: Though many genes have been identified on chromosome 3, 6 and 7 to be associated with otosclerosis but none of them play a key role and the exact genetic defect is still not known. Two third of the patients with otosclerosis have a family history of hearing loss and have autosomal dominant inheritance.
2. Viral infections, especially measles have been implicated as a cause.
3. Autoimmune factors may be a cause. Autoimmunity to collagen II is abundantly present in regions of the world with increased predilection.
4. Cytokines like TGF-beta 1 and bone morphogenic protein (BMP) are associated with otosclerosis.

PATHOLOGY

The basic difference between the otic capsule and the rest of the bones in the body is the *gradient remodeling*, i.e. the rate of remodeling decreases from periphery to the membranous labyrinth. Osteoprotegerin (OPG) and **receptor activator of nuclear factor kappa-β ligand** (RANKL) are the two cytokines which respectively produce inhibition and activation of osteoclast maturation. The OPG is expressed more inside and RANKL outside.

Three phases of disease propagation: Active stage, Intermediate stage and Inactive stage.

a. **Active stage** is called *otospongiosis* in which the normal mature lamellar bone of an otic capsule is replaced by irregularly laid new spongy, more vascular and thick bone. On H and E staining it stains blue and hence is also known as *blue mantles*.
b. **Intermediate stage** an intermediate stage which has features of both active and inactive disease with some portions having otospongiotic foci and some showing otosclerotic foci.
c. **Inactive stage** (final stage) or "otosclerosis" there is deposition of new bone (sclerotic bone which stains red-acidophilic). Osteoclasts disappear in this stage and osteoblasts are seen. The vascular spaces are narrowed or obliterated by new bone formation and lamellar bone which is thicker and more cellular.

INCIDENCE

Clinical otosclerosis is more common in females than males in ratio of 2 : 1.** It has been confirmed now that the incidence of histological otosclerosis is same in both sexes but due to hormonal influences the clinical otosclerosis is twice more common in females than males. At times of increased endocrinal activity such as pregnancy and menopause, the disease rapidly progresses. *There is overall 10% prevalence of histologic otosclerosis while the prevalence of clinically significant otosclerosis is only 1%*.

The disease is uncommon before the age of 5 years. The symptoms of hearing loss usually appear between 15 and 45 years.

CLINICAL FEATURES

1. Deafness: Patients present with slow progressive hearing loss. In 70–80%, it is bilateral and asymmetrical. The patients hear better in noisy surroundings (*paracusis willisii*). It is said to be due to the tendency of people to speak louder in noisy environment thereby overcoming the hearing threshold of the person and is seen in any conductive hearing loss.
   Patients with otosclerosis usually have soft voice and this is usually detected by the close relatives.
2. Tinnitus is seen in 65–85% of patients. It may not be presenting complaints in many patients. It is a result of increased vascularity of the otospongiotic bone. Tinnitus is an indication of sensorineural degeneration.
3. Vertigo which is present in around 24% patients. It may be due to the release of toxins or due to vestibular hydrops secondary to capsular otosclerosis.

CLINICAL EXAMINATION AND TESTS

1. On otoscopy the tympanic membrane usually appears normal. In some cases, there can be red hue due to active foci giving rise to *Flamingo pink colour* of the tympanic membrane (also called as **Schwartze's sign**). It is a sign of active otospongiosis which has an increased vascularity.
2. Tuning fork tests shows negative Rinne's test (BC>AC). Weber's is lateralized to the worse ear (more conductive hearing loss) and absolute bone conduction is normal.
3. Tympanometry shows reduced compliance of the middle ear because of the stapes footplate fixation. This results in reduction in the height of the peak in impedance audiometry with a normal middle ear pressure. The type of tympanogram curve formed is **As (s-stiffness curve) type**. As the disease progresses both the ipsilateral and contralateral acoustic reflex become absent.

*It is one of the reasons for persistence of air bone gap after successful stapedotomy. Another common reason is the complete obliteration of the round window niche by the otosclerotic foci.
**Otosclerosis affects both sexes equally especially when hearing loss is mild (not always diagnosed). For moderate-to-severe losses the females have a higher propensity.

4. On pure tone audiometry there is conductive deafness and typically shows a **Carhart's notch**, which is a hallmark sign of otosclerosis.* Carhart's notch is a dip in the bone conduction at 2000 Hz in pure tone audiometry (Fig. 14.2). Mixed hearing loss may be seen in few cases suggestive of cochlear involvement.
5. Speech audiometry shows normal discrimination score.
6. HRCT temporal bone shows an irregular margin, increase in thickness and scalloped edges of otic capsule producing a classical '***double ring***' or '***halo***' sign (Fig. 14.3).

> ***Note:*** Carhart's notch is seen with any case of ossicular chain fixation and is not specific to otosclerosis.

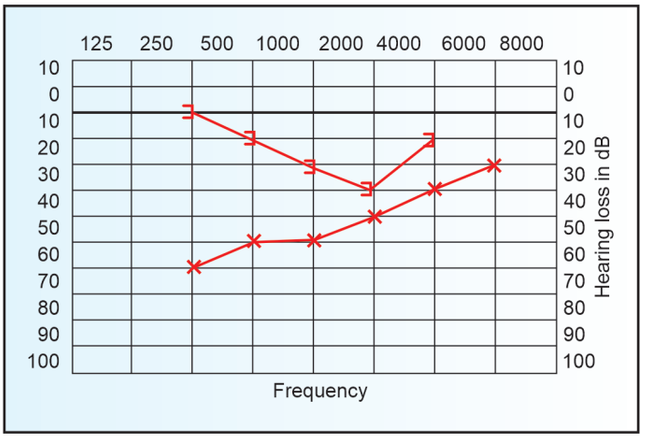


Fig. 14.2: Carhart's notch is dip in the bone conduction at 2000 Hz

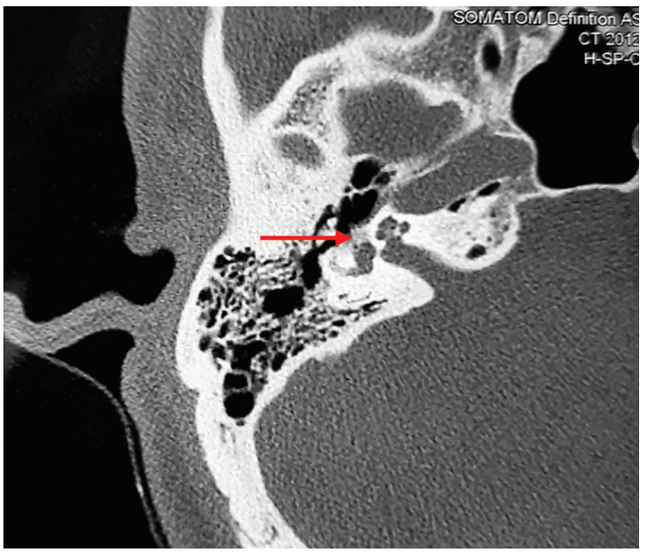


Fig. 14.3: Otospongiotic changes are seen in the anterior part of oval window (marked by red arrow) in otosclerosis

Differential Diagnosis

a. Hypermobile tympanic membrane
b. Otitis media with effusion
c. Tympanosclerosis
d. Ossicular discontinuity
e. Congenital stapes fixation
f. Malleus head fixation
g. Congenital cholesteatoma
h. Paget's disease
i. Osteogenesis imperfecta**
j. Superior semicircular canal dehiscence.

MANAGEMENT

A. *Medical management*

1. **Sodium fluoride** is used to treat the active stage of disease (otospongiosis). The fluoride ion replaces hydroxyl group in bone forming fluorapatite which is resistant to resorption and increases calcification of new bone. Ultimately it causes maturation of active foci of otosclerosis. It reduces tinnitus, reverses Schwartze's sign, stabilizes SNHL and vertigo. On CT there is resolution of otospongiosis. It is given in a daily dose of 50 mg for 2 years.

   Indications of fluoride

   a. Non-surgical candidates/patients who do not want surgery
   b. Patients who have a positive *Schwartze's sign*
   c. Progressive sensorineural hearing loss.

   The most frequent side effects of fluoride therapy are rash, arthritis, bone pain, ligamentous calcification, dental caries, and gastritis. The gastric side effects can be decreased by using enteric coated formulations or by buffering with calcium carbonate.

   Contraindications of fluoride

   a. Chronic renal failure
   b. Pregnancy and lactation
   c. Children
   d. Allergy
   e. Fluorosis.

2. **Bisphosphonates** can be used as an alternative to sodium fluoride. It is an analog of organic pyrophosphate that binds with calcium ions. It blocks osteoclastic activity. The common side effects include osteonecrosis of the jaw, renal toxicity and erosive oesophagitis.

B. *Amplification*

Patients who do not desire surgery can be offered hearing aid. Hearing aids work by amplifying the sound beyond the hearing threshold of the patient. Patient satisfaction rates are lower than that of a successful surgery.

*The proposed explanation for the dip in Carhart notch at 2,000 Hz is that the stapes fixation disrupts the normal ossicular resonance which is maximum at 2000 Hz. This is reversed with stapes surgery.
**In osteogenesis imperfecta, there occurs blue sclera, multiple fractures, progressive hearing loss.

C. *Surgery*

Surgical candidates for the surgery are those with air bone gap (AB gap) >15 dB and speech discrimination of more than 60%.

Contraindications of surgery include active ear disease, only hearing ear, vestibular symptoms (hydrops), SNHL (cochlear otosclerosis), professional athletes, pilots and during pregnancy.

Preoperative counselling including informed consent about the risk of total sensorineural hearing loss must be taken though the incidence is less than 0.1–0.2% in good surgical hands. Mild dizziness may occur postoperatively which is usually transient. There can be tinnitus, facial paralysis, recurrent conductive hearing loss and alteration of taste. Patients are advised to avoid flying, heavy work in the immediate postoperative period.

Surgical Steps

A. *Stapedotomy**

1. 1 ml of 2% lidocaine with 1:30,000 is injected in all the quadrants at the bony cartilaginous junction. Preferred approach for stapes surgery is permeatal. Only in cases with tortuous/stenotic external ear canal, endaural incision may be given.
2. Rosen's incision (permeatal) is then given from 6–12 o'clock position, having 6–8 mm maximum width from the annulus.
3. Tympanomeatal flap is elevated, and adequate middle ear exposure achieved.
4. Next chorda tympani nerve is seen and preserved without causing any undue stretching.
5. Posterosuperior canal wall is curetted or drilled using a micromotor drill, till the base of the pyramid is visualized.
6. The facial nerve is identified.
7. The mobility of ossicles is checked, and malleus or incus fixation are ruled out. Fixation of stapes is confirmed.
8. Incudostapedial joint is separated with a curved sickle knife or a pick (Fig. 14.4).
9. Stapedial tendon is then cut.
10. Stapes superstructure is removed using a right-angle pick or crurotomy scissors (Fig. 14.5).
11. Size of the prosthesis to be inserted is determined by using a measuring rod. The length is measured from the medial aspect of the long process of the incus to the footplate. 0.25 mm is added to it for the teflon piston.
12. By using 0.7 mm diamond burr at 1000 rpm a fenestra is made in the posterior part of the footplate. It can be also made using a hand burr (Larkin's burr) or by a LASER.
13. Oval window seal can be made by using a vein graft (hand or wrist vein is used), tragal perichondrium, temporalis fascia, blood or fat.
14. Finally, an appropriate sized piston is placed in the fenestra and crimped to long process of incus. A newer prosthesis (nitinol piston) needs only heat of cautery for crimping (Figs 14.6 and 14.7).

The various types of pistons used are wire, teflon, stainless steel, titanium, nitinol, soft clip titanium (Fig. 14.8).

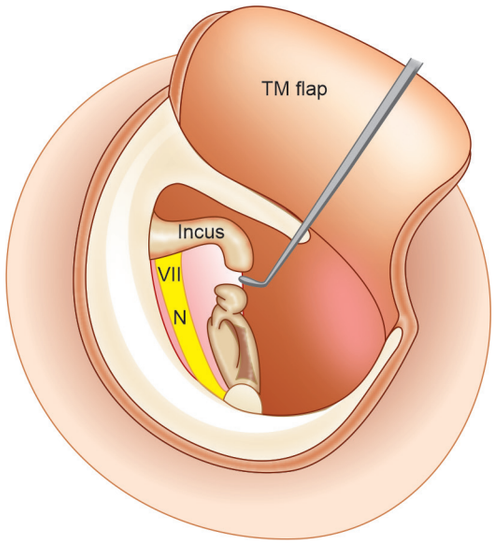


Fig. 14.4: Incus separated from stapes head

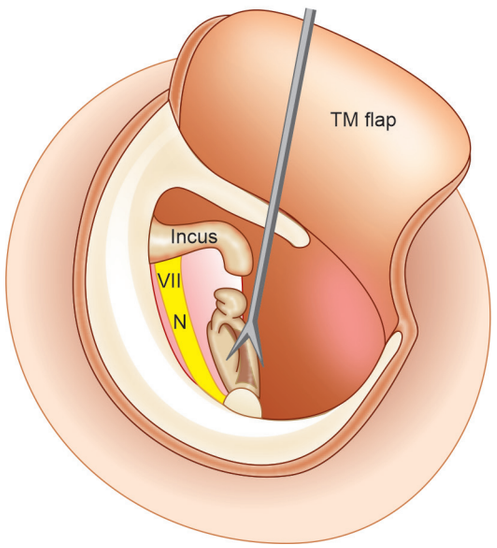


Fig. 14.5: Crurotomy

*Diffuse obliterative otosclerosis is a condition in which the whole footplate including the annular ligament, and oval window niche are involved with the otosclerotic focus. Because of the thickness of otosclerotic focus it is not possible to make the fenestra. In such a situation the obliterative focus may be drilled using a diamond burr till the blue line of the footplate is visualised. Stapedotomy is then done, and a piston inserted. There is a high risk of SNHL in such procedures.

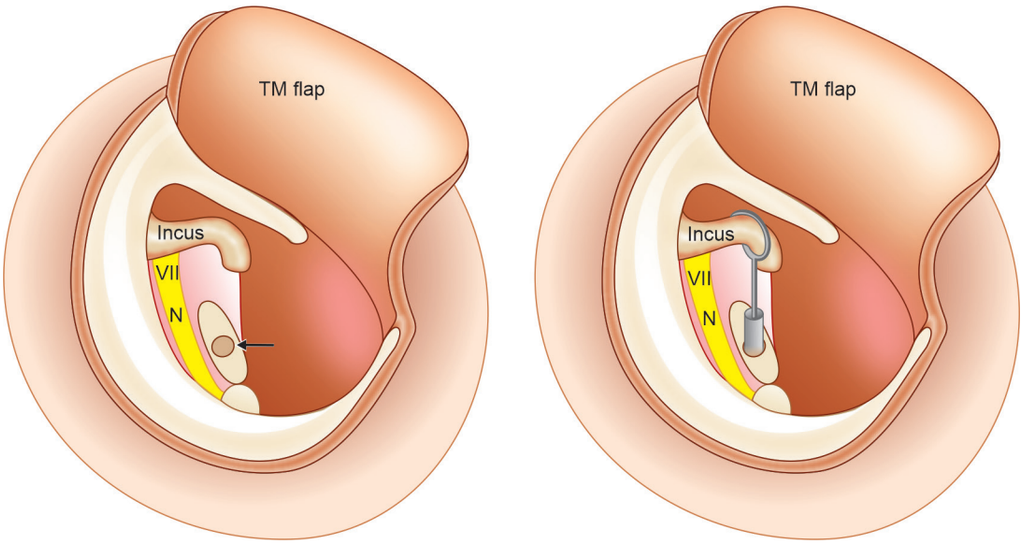


Fig. 14.6: Fenestra formation and insertion of piston

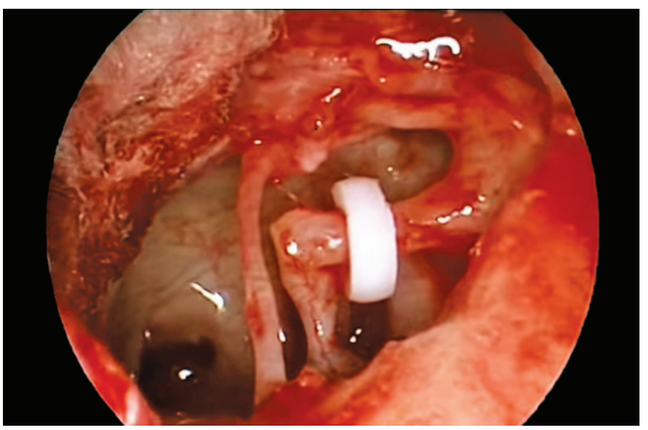


Fig. 14.7: Teflon piston crimped to long process of incus

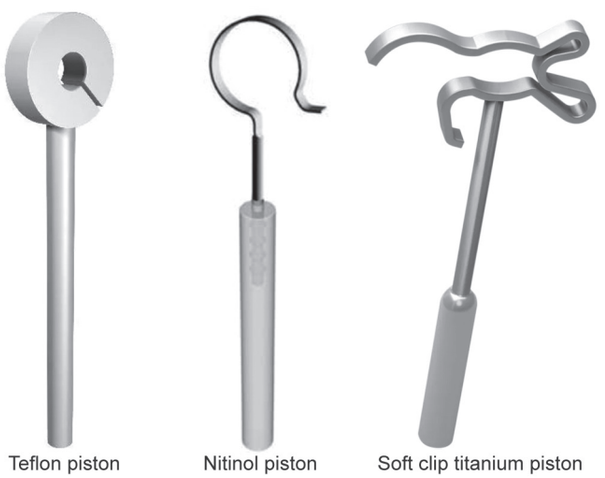


Fig. 14.8: Pistons for stapes surgery

B. *Stapedectomy*

Total stapedectomy is usually not preferred but total removal of stapes may happen inadvertently. It may be required in a situation of floating footplate.* It involves complete removal of the stapes suprastructure and footplate. Rest of the steps are the same.

Disadvantages of stapedectomy over stapedotomy

1. Increased postoperative vestibular symptoms
2. More technically difficult
3. Increased chances of prosthesis migration
4. Increased chances of SNHL due to less cochlear trauma and more release of proteolytic enzymes from foci.
5. Increased chances of perilymph leak (fistula)
6. Increased chances of blood entry in vestibule
7. More chances of fixation of prosthesis by scar tissue.

C. *Laser Stapedotomy*

It was first performed by Perkins in 1980. This stapedotomy uses a CO_2 laser for cutting the stapedial tendon and anterior and posterior crura. Laser is further used to make a fenestra of 0.6 mm. 10–15 seconds of cooling time between the pulses is given to avoid inner ear thermal damage. It has an advantage of giving a blood less field, minimal chances of the footplate subluxation and is atraumatic way to perform stapedotomy leading to better hearing outcomes.

D. *STAMP*

It is a technically challenging procedure where a LASER removal of limited anterior otosclerotic foci is done.

*Complication of floating footplate can occur in case of inadvertent excessive movement of the stapes footplate while making a fenestra. Footplate dislodges from the surrounding oval window niche. It is a challenging complication, where experienced otologists may be able to extract the footplate with the help of microneedles/hooks. Alternatively, the footplate can be covered with a vein or fascia graft and then the piston is placed. Chances of this complication can be avoided by using LASER for making the fenestra or doing reverse stapedotomy, i.e. making fenestra first before removing the suprastructure.

It involves vaporizing the anterior crus along with a linear stapedotomy across the anterior one third of the stapes footplate. The ossicular chain is not disturbed and therefore no prosthesis is required.

The advantages of the procedure are that the stapedius tendon and most of the normal stapes remains intact, eliminating chances of hyperacusis after surgery. There is also a reduction in risk for preoperative inner ear injuries and postoperative prosthesis complications. It has a slight risk of thermal damage to the vestibule.

COMPLICATIONS OF STAPES SURGERY

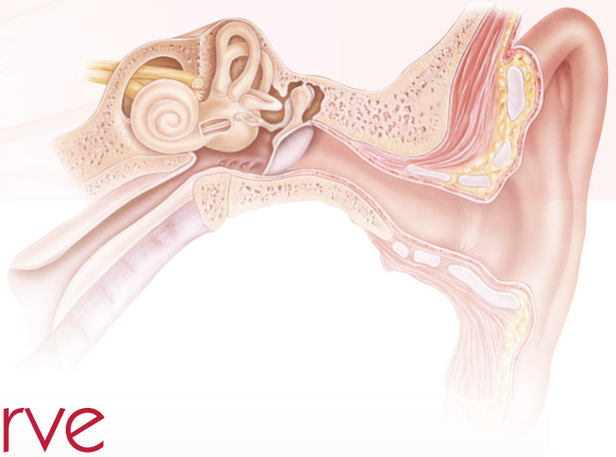
1. **Facial nerve injury** may occur especially in the situation of an overhanging facial nerve: In 0.5% of cases there can be significant dehiscence of the facial nerve. The procedure can be aborted if the nerve cannot be displaced.
2. **Perilymph fistula** is a common complication. It can be primary at the time of surgery or secondary due to barotrauma or sudden increase in middle ear pressure due to blowing of nose or lifting weight in post operative period.
3. **Excessive bleeding:** Persistent stapedial artery* is present in 0.2% cases. It is a vestigial artery, which can be a source of troublesome bleeding during surgery. If possible, it may be inferiorly displaced, and surgery completed.
4. **Disarticulation of the prosthesis** due to improper crimping during surgery.
5. **Floating footplate**.
6. **Dislocation of incus** may occur during removal of the posterosuperior bony overhang. If incus is out of position, either surgery is abandoned or IRP(Incus replacement prosthesis) or Total ossicular replacement prosthesis is used.
7. **Perilymphatic gusher****
8. **Sensorineural hearing loss**
9. **Damage to chorda tympani nerve**
10. **Perforation of the tympanic membrane**
11. **Persistent vertigo**
12. **Refixation of the stapes footplate**
13. **Labyrinthitis**.

*Stapedial artery is transiently present in normal foetal development, connecting the branches of the future ECA to the ICA.

****Perilymphatic gusher** is associated with patent cochlear aqueduct or abnormal vestibule. It has an increased risk of SNHL. The chances of gusher are less with stapedotomy than with stapedectomy. The head end is elevated during surgery and injection mannitol given to lower the CSF pressure. The gusher usually decreases after some time. Rapid placement of the oval window seal (fascia or vein) is advocated. The prosthesis is then placed. Postoperative bed rest and straining and heavy exercise are avoided.

Chapter

15



Facial Nerve

EN4.18: Describe the clinical features, investigations and principles of management of Facial Nerve palsy.

ANATOMY OF THE FACIAL NERVE

The facial nerve is the seventh cranial nerve. There are approximately 7000–9000 nerve fibres of motor type and 3000–5000 secretory in main trunk and somatosensory type present in nervus intermedius (*Nerve of Wrisberg*). The motor nerve fibres originate from the precentral gyrus area of the brain. These travel in corticobulbar tract, internal capsule, upper midbrain, lower brainstem, and then finally to the facial nerve nuclei in the pons (Fig. 15.1).

The facial nerve nucleus lies in the pons, ventrolateral to VI nerve nucleus and medial to spinal nucleus of V nerve. The upper part of the nucleus receives bilateral supranuclear innervations from both cerebral hemispheres, but the lower part of the nucleus receives only contralateral hemisphere motor fibres. The upper part of the nucleus supplies the forehead and the lower part supplies the middle and lower part of the face. Due to bilateral innervations of the upper nucleus, the forehead muscles are spared off in supranuclear pathologies of the facial nerve (Fig. 15.2).

Facial nerve is a mixed cranial nerve. There are four major functions of the facial nerve:

1. **General somatic efferent:** This the motor root of the facial nerve is composed of branchial efferent fibres that supply the 2nd branchial arch muscles, which include facial muscles, buccinator, stapedius, platysma and posterior belly of digastric.

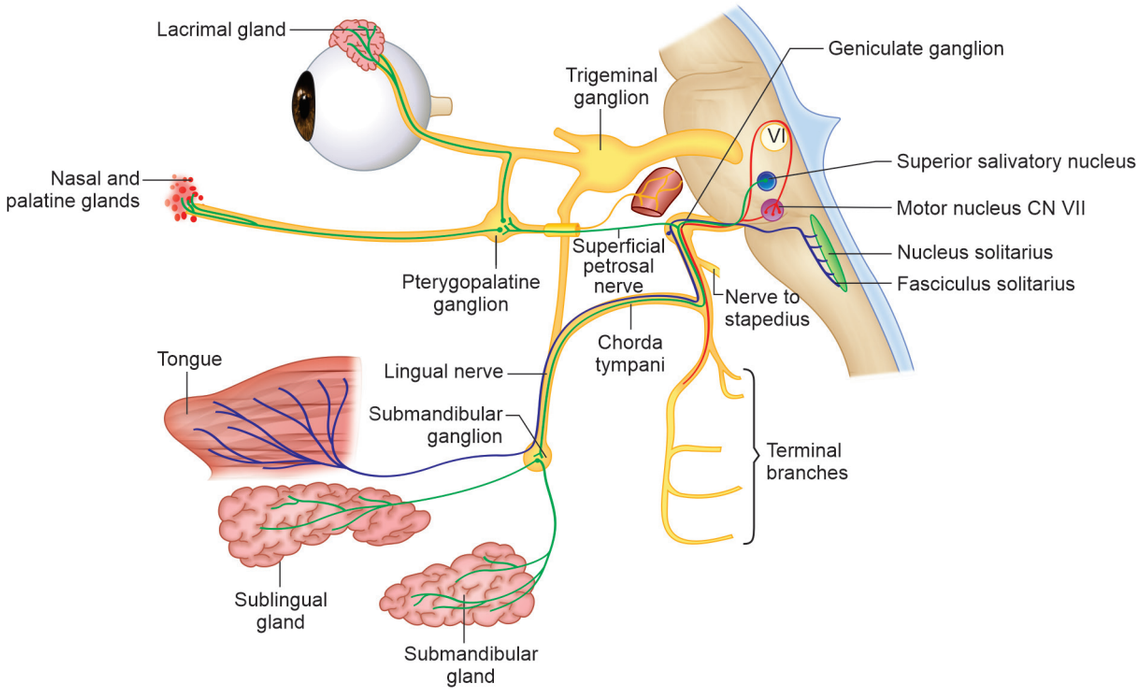


Fig. 15.1: Facial nerve and its branches

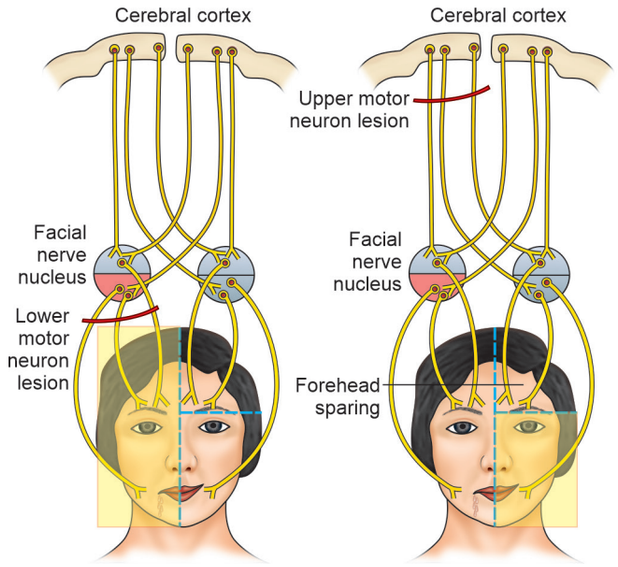


Fig. 15.2: Upper and lower motor neuron lesions of facial nerve

2. **General visceral efferent (parasympathetic secretomotor supply):** These fibres mainly arise from superior salivatory nucleus. They supply lacrimal and palatine glands (via greater petrosal nerve through sphenopalatine ganglion), parotid gland (via lesser petrosal nerve through otic ganglion) and submandibular and lingual gland (via chorda tympani).
3. **Special visceral afferent:** These travel through the chorda tympani via the geniculate ganglion, nervous intermedius and tractus solitarius, reach the rostral part of the VII nerve nucleus. It carries the taste sensation from the anterior two-thirds of the tongue.
4. **General somatic afferent:** These carry cutaneous sensations like pain and touch from the posterior aspect of the external auditory canal. **Hitzelberger's sign** is seen in acoustic neuroma, where there is anaesthesia over the floor of EAC and is the cause for severe pain due to vesicular eruptions in **Ramsay–Hunt syndrome**.

Note: The EAC area also receives supply from the vagus and this is the reason for cough reflex during ear cleaning.

Course of Facial Nerve

The further course of the facial nerve can be divided into three parts, namely **cisternal/intracranial, intratemporal and extratemporal** (Fig. 15.3).

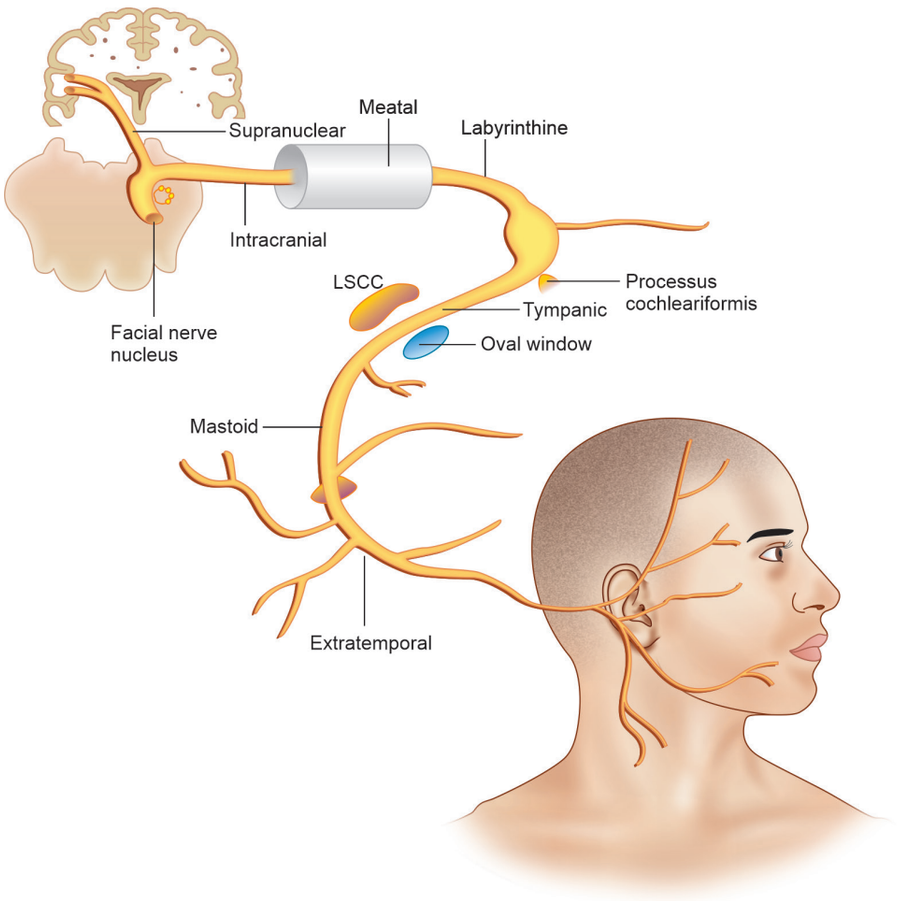


Fig. 15.3: Course of facial nerve (LSCC: Lateral semicircular canal)
Note the important landmarks for facial nerve [oval window, processus cochleariformis and lateral semicircular canal (LSCC)]

LSCC

A. **Intracranial part/cisternal:** This part is of 23–24 mm long and streches from pontocerebellar cistern (CPA) to the *porus* of Internal acoustic meatus (IAM). The facial nerve emerges from the pontomedullary junction and then enters into the internal acoustic meatus. The orifice of the meatus is only 0.68 mm in diameter.

B. **Intratemporal part:** This part extends from the IAM to the stylomastoid foramen and is about 28–30 mm in length. The intratemporal part of the facial nerve has a Z-shaped course with two bends or genu.

It is divided into four parts: Meatal part, labyrinthine part, tympanic part and the mastoid part.

1. *Meatal part* is only 5–12 mm in length. The fibres of nervous intermedius join the motor fibres in this segment. The IAM is divided into four quadrants internally. The **crista falciformis** divides the meatus horizontally. The superior part of the IAM is further divided by **Bill's bar** vertically. The facial nerve lies in the anterosuperiorly quadrant of the meatus. The superior vestibular nerve lies in the posterosuperior quadrant, cochlear nerve in anteroinferior and inferior vestibular nerve in posteroinferior (Fig. 15.4).
2. *Labyrinthine part* extends from the IAM to the geniculate ganglion and is the shortest of all the parts and is only 3–5 mm in length.
3. *Tympanic part*: The segment is about 8–11 mm in length. As it emerges from the geniculate ganglion it takes its first bend (1st genu) with an angle of around 75 degrees posteriorly and runs backwards as the horizontal/tympanic portion of the facial nerve. The nerve courses along the medial wall of the middle ear below the *cog* anteriorly and above the oval window and below the lateral semicircular canal bulge (very constant landmark) posteriorly. Thus the tympanic part lies between the first and second genu of the facial nerve. This part of the facial nerve shows maximum incidence of dehiscence of the bony facial canal (fallopian canal) especially just above the oval window (10–40%).
4. *Mastoid part*: The facial nerve just at the level of the pyramid behind the oval window and 3–4 mm medial to the tympanic sulcus it takes a gradual. This is the 2nd genu (bend of 95–125°) and it further courses as a vertical/mastoid segment of the facial nerve. Thus the mastoid portion of the facial nerve lies between the second genu to the stylomastoid foramen. The length of this segment is 10–14 mm. The nerve can be damaged while elevating the tympanic annulus and lowering the facial ridge in the mastoid surgeries. The branches which arise from the vertical part are the nerve to stapedius, chorda tympani and the sensory auricular nerve (Fig. 15.5).

C. **Extratemporal part:** This is the part of the facial nerve from the stylomastoid foramen to the terminal branches of the facial nerve in the parotid gland.

The stylomastoid foramen is at the base of the temporal bone between the mastoid process and the styloid process. It can be located during parotid surgery behind the middle of the **digastric groove**.* After emerging

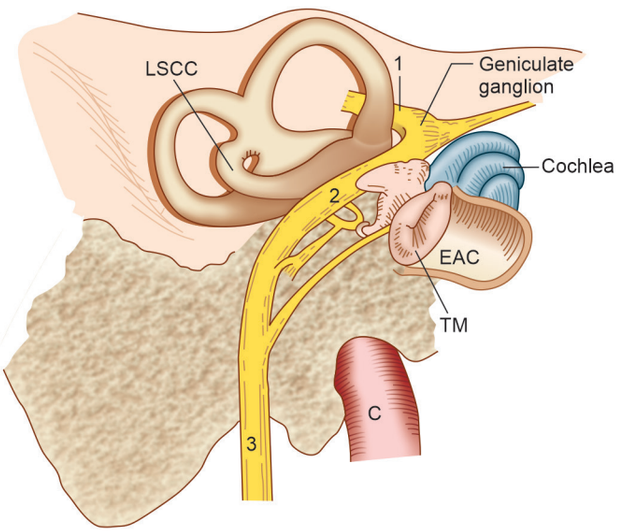


Fig. 15.5: Intratemporal course of the facial nerve—meatal, tympanic and mastoid segment of facial nerve (1 meatal part, 2. tympanic part, 3. vertical or mastoid part)

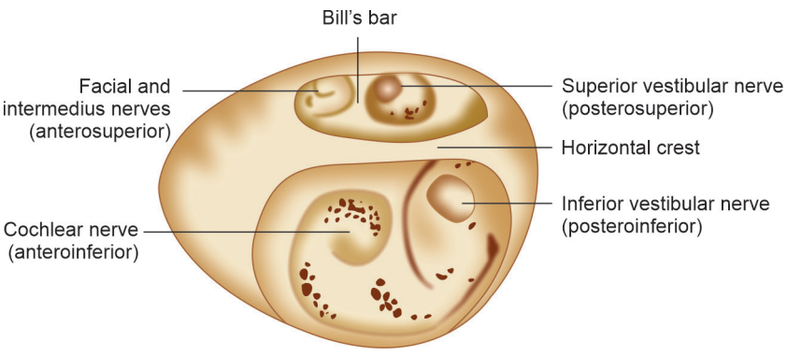


Fig. 15.4: Internal acoustic meatus (IAM)

*Importantly, this foramen is located higher up in newborn almost at the level of the antrum. With the growth of the squamous portion of the mastoid, the facial nerve canal is pushed down.

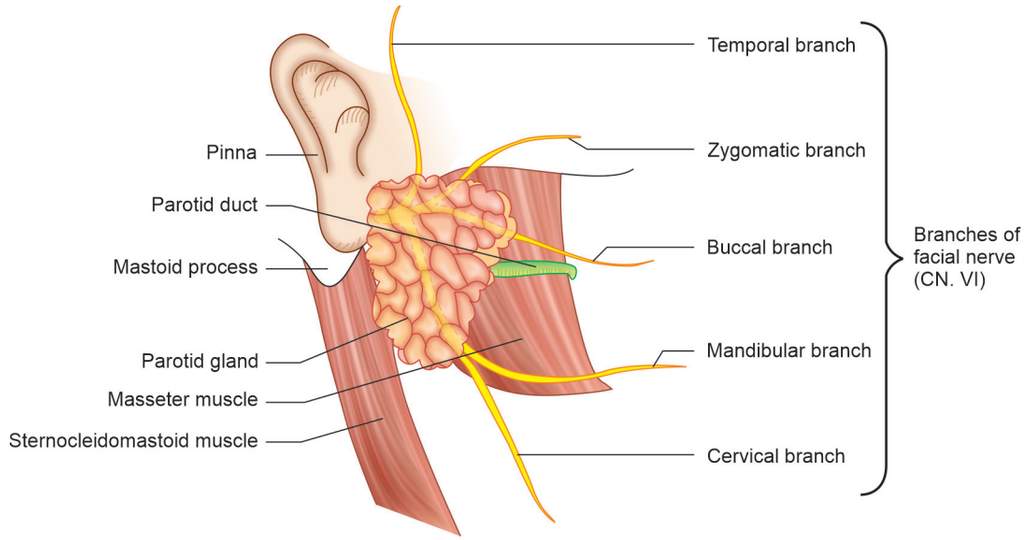
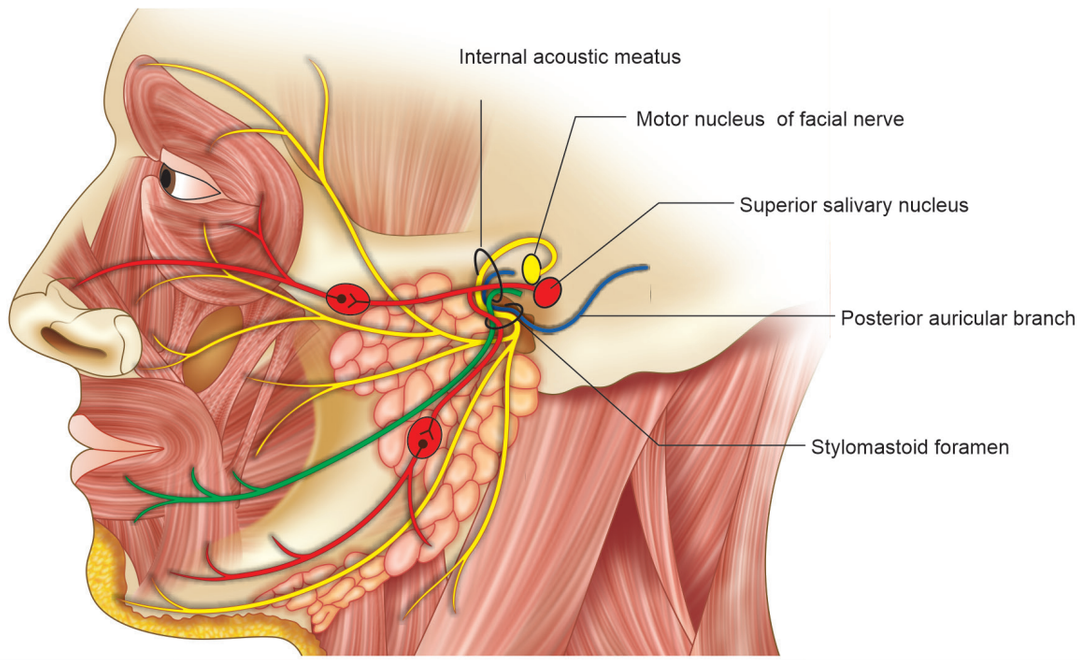


Fig. 15.6: Extratemporal course of facial nerve

from the stylomastoid foramen, the nerve enters the posteromedial surface of the parotid gland. Within the substance of the parotid gland, the nerve divides into two main trunks, namely the upper *temporofacial* and lower *cervicofacial*. This branching of the nerve is called the *pes anserinus* **('goose foot')**. The trunks then further divide into temporal, zygomatic, buccal, marginal mandibular and cervical branches (Fig. 15.6).

BRANCHES OF THE FACIAL NERVE (Fig. 15.7)

1. **Greater superficial petrosal nerve (GSPN)** arises from the geniculate ganglion and it supplies the secretomotor fibres to the lacrimal and palatine glands via the sphenopalatine ganglion.
2. **Nerve to stapedius** arises below the level of the second genu and supplies the stapedius muscles.



Branchial motor (special visceral efferent)	Visceral motor (general visceral efferent)	Special sensory (special afferent)	General sensation somatic afferent (posterior auricular)
Muscles of facial expression, posterior belly of digastric, stapedius and stylohyoid by branches of facial nerve	Submandibular, sublingual glands, mucous glands of nasopharynx and palate by lingual nerve	Taste sensation from anterior 2/3rd of tongue by chorda tympani	General sensation from skin of pinna and posterosuperior part of canal by posterior auricular nerve

Fig. 15.7: Structures supplied by facial nerve

3. **Chorda tympani nerve** arises from the mastoid part of the facial nerve, about 5 mm above the stylomastoid foramen. It passes between the mucosal and fibrous layer of tympanic membrane, behind the neck of the malleus to finally exit the tympanic cavity via the ***canal of Huguier/anterior canaliculus*** within the medial part of petrotympanic fissure and goes on to join the lingual nerve to supply the anterior 2/3rd of the tongue. It is an important surgical landmark for the **posterior tympanotomy*** approach. It is also a useful landmark for identification of the facial nerve during canal wall down mastoidectomy.
4. Ansa of Haller: Anastomosis with the IX nerve
5. Postauricular branch supplies the muscles of the pinna and occipital belly of occipitofrontalis muscles.
6. Stylohyoid branch to the stylohyoid muscle
7. Digastric branch supplies the posterior belly of digastric.
8. Pes anserinus and its terminal branches: They supply the muscles for facial expression and eye closure.

BLOOD SUPPLY

The blood supply to the facial nerve is derived from the branches of the external carotid artery and the vertebrobasilar artery. The labyrinthine artery, which is a branch of anterior-inferior cerebellar artery (AICA), supplies the cisternal, meatal and labyrinthine parts of the nerve. The tympanic and mastoid segments are supplied by an anastomotic network between the superficial petrosal branch of middle meningeal artery and the stylomastoid branch of occipital and posterior auricular arteries.

PATHOGENESIS OF NERVE DAMAGE

Sir Herbert Seddom (1943) classified the localized nerve injuries into three categories, namely neuropraxia, axonotmesis and neurotmesis.

a. **Neuropraxia** is the result of compression or stretch on the nerve fibres but for a short duration. This leads to blockage of axoplasmic transport and ion channel function in the affected neurons. The loss of function will be temporary and complete recovery is anticipated on removal of the causative factor.
b. **Axonotmesis** will occur when there is partial sectioning of the nerve or a severe compression. With the surrounding connective tissue intact, the axons tend to re-grow (at the rate 1 mm/day) towards the effector site and then try to form re-innervation of the target. Often this is not completed thereby the functional outcomes are not complete.

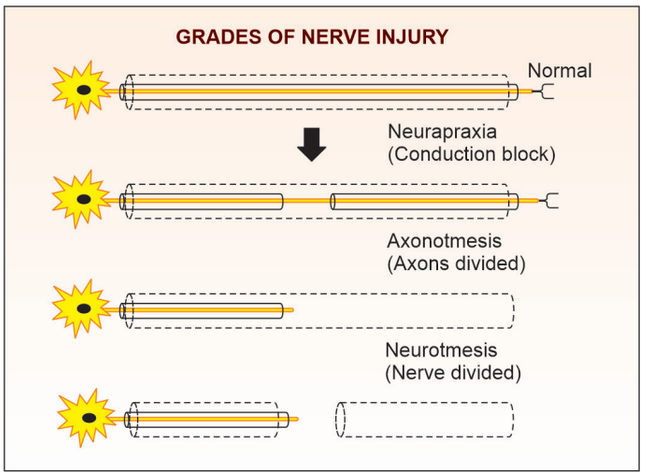


Fig. 15.8: Grades of nerve injury

c. **Neurotmesis** will occur when the nerve is divided completely or damaged. Recovery is only possible if the surgical intervention is done (Fig. 15.8).

FACIAL NERVE PALSY

Clinical Features

The facial nerve palsy (lower motor neuron type) patient presents with following clinical features on the involved side

- Facial weakness on one side
- Inability to close eye
- Loss of nasolabial fold
- Bell's phenomenon (up rolling of eyeball when patient tries to close eye)
- Posterior auricular pain/otalgia
- Decreased tearing
- Hyperacusis
- Taste disturbances
- Drooling.

EVALUATION OF FACIAL PALSY

The history of the patient is taken to mainly to find out the duration of the facial palsy, its progressive nature, ipsilateral or bilateral, recurrent attacks, associated complaints (like loss of taste sensation, facial oedema, fissured tongue, vesicles, otalgia), alternating side of palsy and associated systemic illnesses (Fig. 15.9).

The clinical examination should include a complete head and neck, ear and cranial nerves examination. The examination of the face and motor function of the facial muscles is done. The parotid area is examined for any swelling because malignancy of the parotid can involve the facial trunk or its branches. The facial function has

*Posterior tympanotomy is done to visualise the hidden areas of the middle ear, especially the sinus tympani. The boundaries of posterior tympanotomy (facial recess) are, the facial nerve medially, chorda tympani laterally and fossa incudis superiorly.



Fig. 15.9: Right lower motor neuron facial palsy

now been graded by an internationally accepted system by **House–Brackmann**. The grade I is normal, grade VI is the complete absence of facial motor function and the rest are in between (Table 15.1).

TOPODIAGNOSTIC TESTS

These are done to find out the anatomical level of facial nerve injury.

1. **Schirmer's test:** This is done to assess the function of the greater superficial petrosal nerve. The *Whatson paper strip* no. 21 is placed in the inferior fornix at the junction of medial 2/3rd and lateral 1/3rd for five minutes. The length of paper moistened is checked and compared with both eyes. A moistened length less than 10 mm or unilateral 75% less compared to the other eye, is taken as significant.
2. **Stapedial reflex:** As described earlier, the loss of reflex points towards the facial injury proximal to the second genu of the facial nerve.
3. **Taste test/Electrogustometry:** Clinical bedside the taste can be tested by asking the patient to protrude the tongue and placing over it the sugar solution or salt. Loss of function would indicate injury proximal to the origin of chorda tympani. Alternatively electric stimulation would produce metallic taste and this compared on both sides.
4. **Salivary flow test:** In this test the submandibular duct (Wharton's duct) is cannulated and salivary flow is measured over a gustatory stimulation (6% citric acid over the anterior 2/3rd of tongue).

ELECTROPHYSIOLOGICAL TEST

The nerve excitability test (NET) and maximal stimulation test (MST) are not frequently used these days because of limitations in accuracy, reproducibility, inter-observer variation and low prognostic value.

1. Electroneuronography (ENoG)

The test is performed by giving supramaximal stimulus to the facial nerve at the stylomastoid foramen and evoked compound muscle action potentials are recorded (CAP). The recording electrode is placed at the nasolabial fold. The paralysed side responses are compared with the normal side. This is done after fourth day of injury because most of the axons start to degenerate after 3–4 days of injury.* The diminution of CAP is of significance. Values of more than 90% loss within 14 days of paralysis indicates a poor prognosis. At present it is the most reliable electrophysiological test.

Table 15.1: House–Brackmann grading

Degree of injury	*Grade*	*Definition*
Normal	I	Normal function in all areas of face
Mild dysfunction(barely seen)	II	Slight weakness which is seen on close inspection
		Complete eye closure with slight effort but slight asymmetry on smile with maximum effort. No spasm
Moderate dysfunction	III	Gross obvious weakness but not disfiguring
		Complete eye closure with effort but asymmetrical smile
		Obvious but no disfiguring synkinesis or spasm
Moderately severe	IV	Gross obvious weakness with disfiguring
		Incomplete eye closure, mouth asymmetric
Severe dysfunction	V	Barely perceptible motion
		Incomplete eye closure, mouth has slight movement
		Synkinesis, spasms and contracture
Total	VI	No movement or tone

*It is best done between 3 days and 3 weeks of paralysis.

2. Electromyography (EMG)*

It is performed by recording the active motor unit potentials of the orbicularis oculi and orbicularis oris muscles on rest and on voluntary contraction. Following responses may be seen:

a. Polyphasic motor unit potentials indicate regenerative processes in paralysed nerve. In these cases, close follow-up is required with nerve studies, though no surgical intervention is indicated.
b. Fibrillation potentials indicate Wallerian degeneration,** leading to loss of motor neurons but the motor end plates are viable.
c. No activity on EMG would indicate long-standing denervation and there is a suggestion of replacement of muscle with fibrous tissue.

There are few **limitations** to the nerve conduction studies.

1. No clinical information is available in first 3 days.
2. It provides no information in cases of incomplete nerve paralysis.
3. Neurotmesis and axonotmesis cannot be distinguished.

COMMON CAUSES OF FACIAL NERVE PALSY (Table 15.2)

A. Bell's Palsy

It is an acute (onset in <72 hours), idiopathic, unilateral, lower motor paralysis of the facial nerve. It was first described by Sir Charles Bell and is the most common acute mononeuropathy (involvement of single nerve) in the body.

The minimum diagnostic criteria for the Bell's palsy are:

1. Paralysis or paresis of one side of the facial muscles.
2. Absence of any central nervous system disease.
3. Absence of signs of any ear disease.
4. Absence of cerebellopontine angle disease.

Bell's palsy is diagnosed when there is no other medical cause identified for the facial weakness. It is a diagnosis of exclusion, which requires elimination of other conditions which may cause facial paralysis like stroke, brain tumours, infections, neoplasm or systemic illnesses. Though it is unilateral in presentation, rarely it may occur bilaterally. Both sexes are equally affected and the age group commonly affected is 15–45 years.

Table 15.2: Causes of facial palsy

At birth	*Trauma*	*Infection*	*Metabolic*	*Neoplastic*	*Toxic*	*Iatrogenic*	*Idiopathic*	*Neurological*
Forceps delivery	Base skull fracture	AOM	Diabetes mellitus	VII schwannoma	Tetanus	Mastoid Sx	Bell's palsy	Opercular syndrome
Moebius syndrome	Facial injuries	Otitis externa	Hyperthyroidism	Jugular paraganglioma	Diphtheria	Parotid Sx	Melkerson–Rosenthal syndrome	Millard–Gubler syndrome
	Barotrauma	COM	Pregnancy	Tympanic paraganglioma	Carbon monoxide	Anti-tetanus serum	Temporal arteritis	
	Penetrating	Mastoiditis	Hypertension			Rabies vaccination	Polyarteritis nodosa	
	Middle ear injury	Ramsay Hunt syndrome	Acute porphyria			Mandibular block anaesthesia	Hereditary hypertrophic neuropathy	
		Otitis externa					Multiple sclerosis	
		Mumps					Myasthenia gravis	
		Poliomyelitis					Sarcoidosis	
		Syphilis						
		Malaria						
		Mucormycosis						
		Tuberculosis						
		Lyme disease						
		Encephalitis						
		Chickenpox						
		Botulism						
		Infectious mononucleosis						

*EMG is complementary to ENoG after 2 weeks of paralysis.
**Wallerian degeneration is the process of degeneration of neurons which occurs distal to the site of nerve injury. This happens due to the loss of the neurons where the retrograde flowing neurotrophins pathway is obstructed, especially when the injury is close to the cell body.

Aetiology

The causes for disease are unclear but few factors which are described include infections, genetic, immunological and ischemic neuropathy. Out of these, infectious pathology is considered as the most likely cause. Pathogens which have been implicated include HSV-1, VZV, influenza B virus, adenovirus, coxsackie virus and Epstein–Barr virus. All the probable etiologies ultimately cause edema and inflammation of the nerve in the compromised space of the fallopian canal, leading to microcirculatory failure of the nerve and ultimately nerve dysfunction.

Symptoms include unilateral facial weakness or paralysis, loss of lacrimation, dryness of mouth, hyperacusis and taste alterations.

Various **tests** have been recommended to evaluate patients with acute facial paresis/paralysis for identifiable causes. It may include

A. CT or MRI to identify potential infection, tumours, fractures or other causes
B. Electrodiagnostic testing
C. Serological tests for Syphilis
D. Audiometry and stapedial reflex
E. Schirmer's test.

Most patients with Bell's palsy show some recovery in 2–3 weeks and complete recovery is seen in 3–4 months of onset of symptoms. Almost 80% of the patients recover fully within 3 months. Rest of the patients who have incomplete recovery, will improve slowly till a maximum of 6 months.

Treatment

Even though 80% of patients recover fully without any medications but considering the negative outcomes of facial asymmetry in the rest of the 20%, empirical steroid therapy is started in all the patients.

1. Oral corticosteroids: The recommended dose of prednisolone is 1 mg/kg for 7 days. The patient is reviewed after 1 week to stop steroids if there is good improvement in facial nerve function.
2. Antiviral treatment: Addition of antiviral medication improves the rates of incomplete recovery and reduces the long-term after-effects of idiopathic palsy. Hence it is recommended to start antiviral treatment along with the steroids. Acyclovir in dose of 400 mg five times a day for 10 days and valacyclovir in dose of 500 mg twice a day for 5 days. It is effective against the varicella-zoster virus (VZV) which is the most probable infective cause for Bell's palsy.
3. Facial physiotherapy
4. Eye care is important to prevent exposure keratitis. Use of dark goggles can help to protect the eyes from dust and debris. Lubricating eyedrops should be used frequently during the daytime to prevent dryness of cornea and ointment is preferred during the night. Following the application of the ointment the eye should be taped and closed. Temporary tarsorrhaphy can also be done till the facial nerve function improves.
5. Surgical decompression: There is a role of facial nerve decompression by middle cranial fossa approach, which was done to provide access to the primary site of lesion (meatal segment) in patients of Bell's palsy. It is not preferred now, due to the morbidity associated with such a procedure.

Poor prognosis is anticipated in patients who have any of the following:

1. Complete paralysis at onset or incomplete paralysis with late onset of recovery
2. Old age,
3. Dry eye, or loss of taste sensation
4. Absent stapedius reflex postauricular pain.

These patients may have residual facial asymmetry and synkinesis.

B. Ramsay Hunt Syndrome

This is a viral infection caused by reactivation of varicella zoster virus (VZV) in the geniculate ganglion. Ramsay Hunt syndrome was first described in 1907 by James Ramsay Hunt. The presence of the VZV infection was confirmed by using PCR (polymerase chain reaction), which demonstrated the viral DNA in the geniculate ganglion. The cause for reactivation of the virus still remains unclear.

Ramsay Hunt syndrome is a herpes zoster infection of the facial nerve. It is frequently associated with VIII cranial nerve involvement. It can rarely involve V, VI, IX, and X cranial nerves. XII nerve involvement has also been reported in few cases. The hypothesis for involvement of multiple cranial nerves includes factors like anatomical proximity, same embryological origin of the nerves, haematogenous spread, and transaxonal spread of infection.

Symptoms

1. Often there is a prodrome of ear pain, fever, and fatigue for 1–3 days.
2. The classic triad of Ramsay Hunt syndrome includes ipsilateral facial paralysis, otalgia, and vesicles on the auricle. The erythematous vesicular rash is painful and it involves the auricle and external auditory canal. Rarely the rash may appear over the soft palate, mouth and throat Fig. 15.10).
3. Hearing loss with tinnitus may occur due to the involvement of vestibulocochlear nerve.
4. Few patients also complain of vertigo, nausea, vomiting, dysphagia, hoarseness of voice and headache.

Diagnosis

1. Diagnosis of Ramsay Hunt syndrome is mainly based on history and clinical examination which classically shows facial palsy and vesicular rash.



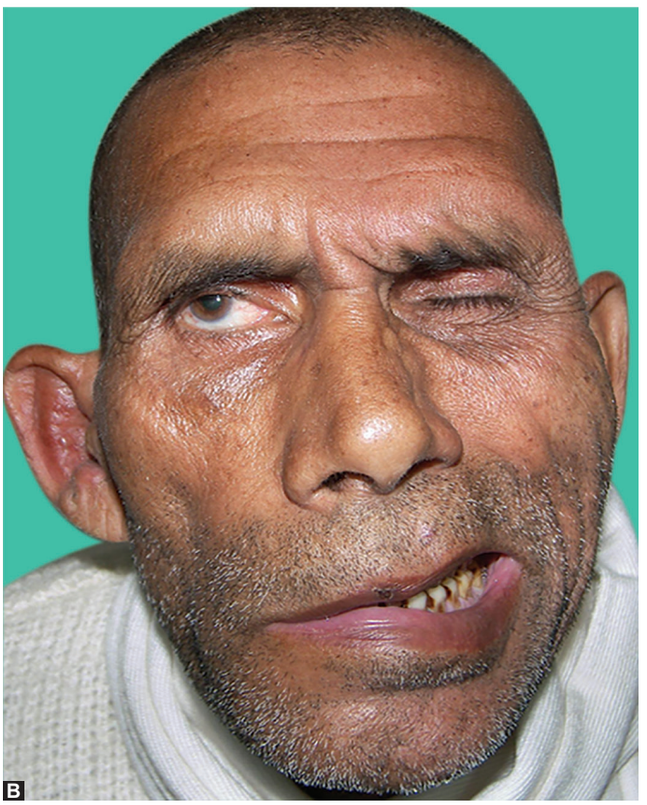


Fig. 15.10: Ramsay Hunt syndrome patient (vesicles in conchal region) with right sided facial nerve palsy.

2. **Tzanck smear** can be performed on fluid from vesicles showing multinucleated giant cells.
3. PCR analysis of tears, saliva, or fluid from the vesicles can be done.
4. Magnetic resonance imaging (MRI) may be used to find the inflammation near the geniculate ganglion of the affected facial nerve. Though imaging modalities are not necessary for management.
5. Pure tone audiometry to look for VIII nerve involvement.
6. Laryngoscopy to look for X nerve involvement.

Treatment

1. Antiviral drugs like acyclovir (800 mg × 5 times/day for 10 days) or valacyclovir (1000 mg given three times a day) or famciclovir (500 mg given three times a day) is started with onset of symptoms.
2. Antivirals are given in combination with oral steroids for best results. Tab. prednisolone 1 mg/kg body weight should be given for 21 days followed by tapering of dose for 7 days.
3. Eye care is essential to prevent exposure to keratitis. Artificial tear drops and lubricating ointments are recommended.
4. Analgesics are given for pain relief.

C. Iatrogenic Facial Nerve Injury

1. In **middle ear surgery** and **mastoid surgery**, the incidence of facial nerve injury is around 1–2%. The most common sites for injury are the second genu and distal end of tympanic segment.
2. In parotid surgery might lead to paresis or paralysis of the facial nerve or its branches. This is especially more commonly seen in cases of malignant parotid tumours, revision parotid surgery and parotid sialadenitis.
3. Other causes of iatrogenic facial nerve injury include, cerebellopontine angle tumour surgery, submandibular gland surgery (marginal mandibular nerve), neck dissection, submandibular abscess drainage and forceps delivery.

Management

The knowledge of surgical anatomy of facial nerve and its landmarks, as it courses through the mastoid and parotid substance is extremely important. A facial nerve monitor may be used during the surgery to identify and preserve the facial nerve and its branches during surgery (Fig. 15.11).

i. If there is superficial injury to the sheath and is recognized at the time of surgery, segments distal and proximal to the nerve injury site are decompressed. Decompression of nerve is done by removing the bone of the fallopian canal and allowing the nerve to bulge out or expand.

ii. If the nerve fibres are seen to be damaged, then epineural sheath is opened to release oedema. In the situation of more than 50% of the nerve fibres being damaged, end to end anastomosis is done. In case a segment of nerve is missing, cable grafting with a greater auricular nerve is done.

iii. In the situation of facial paresis or paralysis being recognized postoperatively and the surgeon has identified the nerve intraoperatively, then a few hours of observation might be sufficient to clear off the effect of local anaesthesia. The mastoid pack should be loosened or removed. In case of incomplete recovery, oral steroids are started.

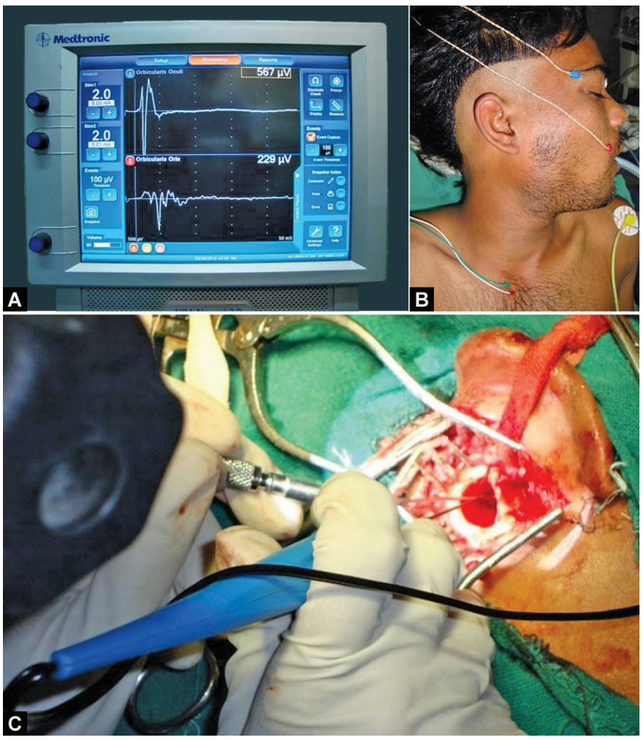


Fig. 15.11: (A) Facial nerve monitor; (B) Patient with electrodes placed in facial muscles; (C) Intraoperative monitoring during mastoidectomy using a facial nerve probe.

If there is complete paralysis and the surgeon is not sure of identifying the facial nerve in its entire course during the surgery, the patient must be taken up for a revision surgery for decompression of the nerve. Also, nerve conduction study when shows more than 90% degeneration on the involved side, then exploration of the nerve should be done. It is not very essential to do urgent decompression, as was advised earlier. Planned re-exploration surgery, by an expert otologist within the next 3–7 days is the current norm.

D. Otitis Media

Acute otitis media and chronic otitis media both can lead to facial nerve paralysis. The presumed cause is the direct involvement of the nerve through fallopian canal dehiscence or due to fallopian canal osteitis with bone erosion resulting in compression and secondary thrombosis of vasa vasorum. This results in subsequent ischemia and paralysis of the nerve.

The mainstay of the treatment of AOM with facial palsy is drainage of pus from the middle ear by myringotomy. This followed by use of antibiotics and steroids. In cases of COM, exploration of the mastoid with facial decompression is done.

E. Temporal Bone Fractures

The temporal bone fractures are caused due to blunt trauma to the head which sometimes involves ear structures leading to hearing loss, vertigo, balance disturbances and facial nerve paralysis. The ear involvement should be suspected when there is postauricular ecchymosis **(*Battle sign*)**, bleeding from ear or facial nerve paralysis.

Clinical examination may reveal blood clots in EAC, step deformity of EAC with ruptured tympanic membrane or hemotympanum with intact tympanic membrane, facial weakness, CSF otorrhea and hearing loss which may be conductive or sensorineural depending upon the structures involved.

The temporal bone fractures have classically described on the basis of the relation of fracture line with the long axis of petrous bone (Fig. 15.12).

1. Longitudinal
2. Transverse
3. Combination (mixed).

Longitudinal fractures (80–90%) are those where the fracture line passes parallel to the petrous bone long axis. They usually occur with head injury due to blow at the temporal or parietal areas. The fracture line passes along the roof of the external auditory canal, tympanic membrane, roof of middle ear travels anterior to labyrinthine capsule to end finally at foramen spinosum or foramen lacerum. There can be tympanic membrane perforation and step deformity in the external auditory canal. Risk of facial nerve injury is 10–25%. The middle ear involvement is usually not severe and there is usually no residual hearing loss in most cases (Fig. 15.13).

Transverse fractures (10–20%) are caused due to frontal or occipital blow to head. The fracture line passes transversely to the long axis of petrous bone. The risk

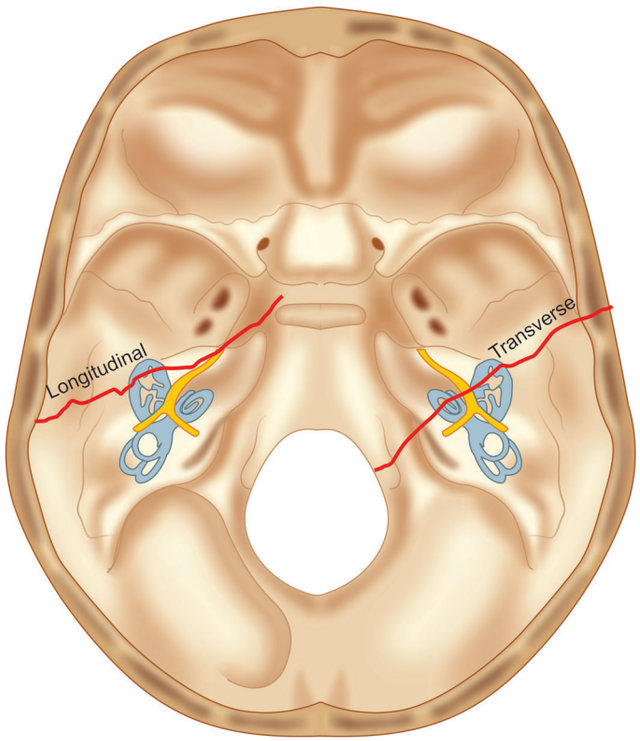


Fig. 15.12: Types of temporal bone fractures



Fig. 15.13: Longitudinal fracture of the right temporal bone



Fig. 15.14: Transverse fracture of the right temporal bone

of involvement of facial nerve is 40–50% and there is a much higher chance of other inner ear injury. The formation of hemotympanum, dislocation of ossicles and incidence of sensorineural hearing loss is much higher. There are much higher chances of patients having vertigo and balance problems.

Due to fractures being randomly placed or being comminuted, therefore the above classification does not always hold adequate. Recently, the emphasis is on describing the fractures as **otic capsule violating** and **otic capsule non-violating type**. The otic capsule violating fracture has higher chances of serious sequelae like facial nerve injury, CSF leak and profound hearing loss (Fig. 15.14).

Management of Facial Nerve Palsy in Temporal Bone Fractures

Around 7% of temporal bone fractures are associated with facial nerve palsy

1. HRCT scan is the investigation of choice for diagnosing temporal bone fractures.
2. The incomplete and delayed onset facial palsy will improve in almost all cases and such cases should be managed conservatively.
3. Patients who have complete and immediate onset facial nerve palsy may require surgical decompression of facial nerve.
4. Recent study reveals, that an undisplaced longitudinal temporal bone fracture, with facial palsy does not required a facial nerve decompression. All such patients improve with conservative management in 6–9 months of injury.

F. Melkersson–Rosenthal Syndrome

Melkersson–Rosenthal syndrome is characterized by a triad of recurrent facial nerve palsy, oro-facial edema, and fissuring of the tongue (*lingua plicata*). The most commonly affected age group is between 25 and 40 years. It is more common in females. The aetiology of the syndrome is unclear. Corticosteroid is the mainstay of treatment. Refractory and recurrent cases may require immunosuppressant drugs like methotrexate. The facial nerve decompression is indicated in recurrent facial nerve palsies or cases refractory to medical management (Fig. 15.15).

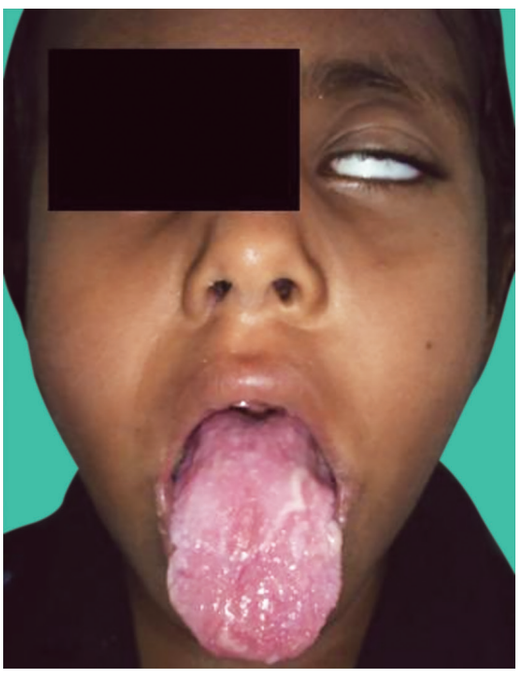


Fig. 15.15: Melkerson–Rosenthal syndrome

SEQUELAE OF FACIAL NERVE PALSY

1. **Exposure keratitis** can occur due to the ability to close eyes leading to dryness of eyes and corneal damage. Artificial tear drops and lubricating eye ointments are advised. Temporary tarsorrhaphy can be done if a longer time of facial paralysis is anticipated. Gold implants in the upper eyelid are used for permanent facial nerve paralysis.
2. **Crocodile tears (Gustatory lacrimation)** occur due to erroneous innervations of the parasympathetic fibres of the facial nerve of salivary glands to the lacrimal gland. This leads to production of tears while eating. It can be treated by tympanic neurectomy.
3. **Frey's syndrome (Gustatory sweating/auriculotemporal syndrome)** occurs after a parotid surgery. This occurs due to erroneous innervations of the postganglionic parasympathetic fibres of the auriculotemporal nerve, which supplies the parotid gland to the sympathetic fibres supplying the local sweat glands. This leads to sweating and flushing over the parotid area, while masticating. The diagnosis can be confirmed by an iodine-starch test. It can be prevented by performing the skin flap raise below the **SMAS plane** (superficial musculoaponeurotic system) during parotidectomy. The treatment is mainly surgical, in terms of placing a facial graft or fascial flap (temporoparietal flap) between the skin and the parotid tissue. Local anti-perspirant can be used or subdermal injections of botulinum toxin are helpful in decreasing the symptoms.
4. **Tics and spasms** occur due to faulty innervations of damaged facial nerve branches leading to involuntary movements of the face.
5. **Synkinesis** is the mass movement of facial muscles usually after cross-innervation of facial nerve fibers after trauma.

Chapter

16



Ménière's Disease

EN4.20: Describe the clinical features, investigations and principles of management of Ménière's disease.

Ménière's disease is a disease of the membranous labyrinth, characterised by deafness, vertigo and tinnitus. Aural fullness is now added as another important feature of the disease. The probable cause for this disease is a sudden increase in endolymphatic pressure leading to the stretching of the round window.

The disease was first described by Dr Prosper Meniere in 1861. Though the disease circumscribes diverse clinical features favouring the use of term Ménière's syndrome, but to keep uniformity and simplicity the term Ménière's disease is still favoured. It is sometimes referred to as *endolymphatic hydrops*. The hydrops in Ménière's disease is idiopathic in origin. There are many other conditions also where hydrops can be seen, hence the term *secondary endolymphatic hydrops* is used for condition producing endolymphatic hydrops other than Ménière's disease.

PATHOPHYSIOLOGY

The main pathology of the disease is the distension of the endolymphatic system due to excessive endolymphatic fluid accumulation. The distension is always seen in the cochlear duct and in most cases, also the saccule. The utricle is not commonly involved (Fig. 16.1).

The reasons for the distension, is not clear and possible cause is either overproduction of endolymph or reduced reabsorption of endolymph at the endolymphatic sac. The acute attacks are explained by the rupture of the membranous labyrinth. Schuknecht theory suggests that the ruptures of the membranous labyrinth leads to mixing of potassium rich endolymph into the perilymph, causing depolarisation of nerve cells resulting in their inactivation. This results in decreased cochlear and vestibular function and symptoms of *Ménière attack*. When the rupture membrane heals the attacks subside. Other possible origins of the disease are perisaccular fibrosis, atrophy of the endolymphatic sac and loss of epithelial integrity, hypoplasia of the vestibular aqueduct, and narrowing of the lumen of the endolymphatic duct (Sajjadi and Paparella, 2008).

AETIOLOGY

The exact cause of Ménière's disease is not known but few causes have been closely associated with the disease. Both the sexes are equally affected. The common age is between the second to sixth decade.

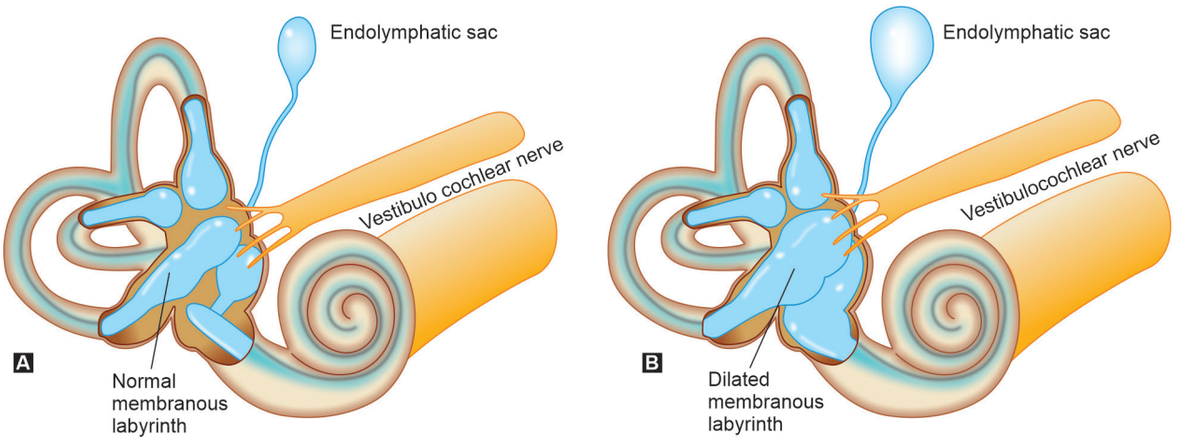


Fig. 16.1: Ménière's disease

1. Genetic

The familial tendency of the disease is described and has been found in 14–20% of the patients. The possible locus lying between the HLA-C and HLA-A on the short arm of chromosome 6 has been described.

2. Viral infections

The damage to the endolymphatic sac and duct by viral infections has been suggested as the cause. The commonly implicated viruses are herpes simplex type 1 and enterovirus. Antiviral treatment has been shown to produce vertigo control in 90% of patients with Ménière disease.

3. Anatomical variations

A small vestibular aqueduct has been proposed as the cause for the disease. The smaller size of the endolymphatic sac and duct have also been suggested as the cause.

4. Traumatic

Trauma may lead to biochemical dysfunction in the cells of the membranous labyrinth. Trauma may also lead to release of debris which can block the endolymphatic sac and duct.

5. Allergy

80% of the patients with Meniere' disease have a history of childhood allergy. The circulating immune complexes can produce damage to blood vessels of the endolymphatic sac and stria vascularis. Allergy treatment also controls vertigo in 47% of the patients of Ménière's disease.

6. Vascular Factors

Endolymphatic hydrops impairs inner ear blood flow autoregulation leading to venous congestion. The combination of hydrops and venous congestion cause vertigo symptoms of the disease.

7. Autoimmunity

Antibodies against mesenchymal elements of the normal inner ear have been seen in Ménière's patients. Severe immunological abnormalities as measured by ESR, serum immunoglobulin levels, complement assays and autoantibodies levels have also been described.

Causes of secondary endolymphatic hydrops

1. Abnormal metabolic and endocrine states
2. Developmental insult
3. Syphilis
4. COM
5. Viral infection
6. Autoimmunity
7. Otosclerosis
8. Abnormal fluid balance
9. Leukaemia.

CLINICAL FEATURES

1. Episodic attack of **vertigo** is the most common symptom, which is seen in 97% of the patients. Usually, the attack lasts for 2–3 hours. The thumb rule is that Ménière's vertigo lasts between 24 minutes and 24 hours in the majority of the cases. There is no loss of consciousness or any focal neurological deficit during the attack of vertigo. Head movement exacerbates the symptom.

   Nystagmus is seen in all patients. Initially as symptoms start, irritative nystagmus is seen which beats towards the affected ear. After a short interval the paralytic nystagmus is seen which beats to the healthy side. Hours into the attack the recovery nystagmus is seen which again changes to the healthy ear.

   Hennebert's sign performed by application of the negative pressure to the external auditory canal leads to horizontal nystagmus in 90% of the cases. This is due to the close proximity of the stapedial footplate with dilated saccule.
2. **Sensorineural hearing loss:** It is another important feature of the disease which is typically fluctuant and progressive. Early in the disease, the characteristic pattern is of the involvement of the low frequencies leading to a rising curve. With progression of the disease the curve tends to flatten due to the higher frequencies getting involved. Profound deafness is rarely seen in the end stage of Ménière's disease.

   Dysacusis is hearing impairment characterized by difficulty in processing the details of sound due to distortion in frequency or intensity rather than primarily a loss of the ability to perceive sound. The patient feels pain or discomfort due to sound.

   Diplacusis binauralis dysharmonica is the perception of the same frequency sound as of different pitch in the two ears. Usually, the affected ear perceives higher pitch to the sound.
3. **Recruitment** is another abnormal feature seen with the disease. It is seen in 56% of the patients, leading to loudness intolerance. This phenomenon of abnormal loudness perception to suprathreshold stimuli is a cochlear phenomenon and is not seen with retrocochlear lesions.
4. **Tinnitus:** It may be the presenting symptom of the disease. It may be continuous or intermittent and is non-pulsatile. Patients usually describe it as roaring in nature and it mainly precedes onset of vertigo.
5. **Aural fullness:** The feeling of aural fullness is commonly felt by patients. Interestingly the fullness may be perceived in the head and neck too.
6. **Somatopsychic effects:** Commonly seen in elderly patients, who may complaint of secondary *agoraphobia* (anxiety disorder of places which make you feel trapped).

Atypical Ménière's disease is a term used for a condition where a complete set of conditions associated with Ménière's disease are not present.

a. **Lermoyez syndrome** is a variant of the disease which shows vertigo being preceded by tinnitus and hearing loss. Hearing loss and tinnitus dramatically improves during or shortly after vertigo initiation.
b. **Tumarkin's crisis (drop attacks)** is characterised by sudden falling attacks without loss of consciousness or vertigo. This is probably caused by the acute otolithic organs dysfunction. These attacks often remit, unlike Ménière's disease.
c. **Abnormal oculovestibular response** are seen in a set of patients who show all symptoms like Ménière's disease but have unusual symptoms of nausea and vomiting and diaphoresis when exposed to optokinetic stimuli like riding in a moving bus or car.

DIFFERENTIAL DIAGNOSIS (Table 16.1)

Table 16.1: Differential diagnosis for Ménière's disease

Vertigo with normal hearing	*Auditory symptoms without vertigo*	*Vertigo with auditory disturbances*
1. Vestibular neuronitis 2. Benign paroxysmal positional vertigo	1. Sudden deafness 2. Vestibular schwannoma	1. Cogan's syndrome 2. Craniovertebral junction anomalies 3. Vertebrobasilar insufficiency 4. Migraine 5. Non-specific cochlea-vestibulopathy 6. Vestibular migraine 7. Vestibular paroxysmia 8. Chronic subjective dizziness

INVESTIGATIONS

I. Metabolic and screening test
II. Assessment of the cochlear function
III. Assessment of vestibular function.
IV. Radiology.

I. Metabolic and Screening Test

These are performed to exclude causes of secondary endolymphatic hydrops. As the Ménière's disease is the idiopathic endolymphatic hydrops, therefore a range of diagnostic tests are required.

1. Complete blood count
2. Random blood sugar
3. ESR
4. Serum urea
5. Serum electrolytes
6. VDRL and *Treponema pallidum* haemagglutination antibody (TPHA)
7. Thyroid function test
8. Lipid profile.

II. Assessment of Cochlear Function

1. **Audiogram:** Most of the patients reveal moderate to severe SNHL There is involvement of low frequency initially.
   Glycerol dehydration test: It involves performing a PTA during the course of Ménière's attack and then repeating the test after giving parental glycerol (1.5 mg/kg). Marked improvement is noticed in hearing threshold results. In clinical practice a positive glycerol test indicates reversibility and that treatment with diuretic drugs may be of value.
2. Alternate binaural loudness balance test (ABLB) can be done to elicit recruitment phenomenon.
3. Speech discrimination scores are abnormal. This helps to rule out retrocochlear lesions. Also, there is no ***roll over phenomenon***.

> ***Note:*** Marked decrease in speech discrimination is not seen with retrocochlear lesions.

4. Auditory brainstem evoked potentials can be recorded. The increase of pressure in scala media leads to stiffening of the basilar membrane. This further will lead to increased speed of sound travelling waves. Hence the latency of wave V is decreased.
5. **Electrocochleography:** ECochG measures the ratio of the summating potential (SP) and the nerve action potential (AP) in response to auditory stimuli. Hydrops (elevated pressure) is suggested when the ratio of SP/AP is greater than 0.35. The test is most accurate when there is an acute episode of Ménière's disease.

III. Assessment of Vestibular Function

1. **Vestibulospinal reflex arch examination** would include Romberg's test and *Unterberger stepping test*. Approximately 30% of patients show positive findings.
2. **Caloric test and Furosemide test:** Large number of patients will show hypoactivity of the ***vestibulo-ocular reflex***. The furosemide test is also called vestibular glycerol test. Administration of the diuretic leads to improvement in vestibular symptoms.
3. **Vestibular evoked myogenic potential (VEMP)** test could be useful in the analysis of saccular function, and diagnosis of Ménière's disease. The low amplitude of cVEMPs may be found in the affected ear with Ménière's disease.

IV. Radiology

Delayed MRI after intravenous or intratympanic injection of gadolinium-based contrast agent can visualize endolymphatic hydrops and can be helpful in confirming the diagnosis. It also helps to rule out inner ear malformations, vestibulocochlear nerve abnormality and cerebellopontine angle lesions like vestibular schwannoma.

Diagnostic Scale

This is the internationally accepted scale (American Academy of Otolaryngology-Head and Neck Surgery) for the diagnosis of Ménière's disease.

a. **Certain Ménière's disease:** The certain diagnosis of disease can only be made on histopathology (autopsy).

b. **Definite Ménière's disease**
   1. Two or more episodes of vertigo which are 20 minute or longer
   2. Audiometric documented hearing loss
   3. Tinnitus and aural fullness.

c. **Probable Ménière's disease**
   1. One episode vertigo
   2. Audiometric documented hearing loss
   3. Tinnitus and aural fullness.

d. **Possible Ménière's disease**
   1. Episodic vertigo with no hearing documentation
   2. Fluctuating sensorineural hearing loss.

STAGING OF MÉNIÈRE'S DISEASE

The staging of Ménière's disease is solely made on the basis of hearing loss over the last six months for frequencies between 500 Hz and 3 KHz frequencies (Table 16.2).

Table 16.2: Staging of Ménière's disease

Stage	*Four tone average (dB)*
1.	≤25
2.	26-40
3.	41–70
4.	>70

TREATMENT

A. Medical Treatment

1. Prophylaxis in between acute attacks

i. Dietary restrictions include salt and caffeine restriction.
ii. Cessation of smoking is advisable because of nicotine having vasoconstrictive properties.
iii. Avoiding over hydration
iv. Diuretic therapy: Acetazolamide (carbonic anhydrase inhibitor) and hydrochlorothiazide.
v. Steroids
vi. Vasodilators: Betahistine dihydrochloride is the effective drug in Ménière's disease especially for maintenance therapy. This has been known to improve microcirculation of the inner ear, thereby decreasing hydrops. Normal recommended dose is 16 mg thrice a day. The drug has limited side effects and must be avoided in asthmatics.
vii. Hyperbaric pressure chamber therapy
viii. Avoidance of allergens does produce reduction of attacks of acute symptoms.
ix. **Meniett device therapy** is a minimally invasive form of therapy in which pressure pulses are delivered to the ear through a small device following placement of a tympanostomy tube in the tympanic membrane. These pressure changes are thought to help stimulate the flow of endolymph, which can result in a reduction of vertigo symptoms.
x. Immunotherapy.

2. Acute phase management

a. Vestibular suppressants: These are mainly required in cases of acute episodes of symptoms of the disease. These include the drugs with anticholinergic, antiemetic and sedative properties. These include

- Phenothiazines: Prochlorperazine, perphenazine
- Anticholinergics: Cinnarizine, dimenhydrinate, promethazine hydrochloride. Also, transdermal scopolamine hydrobromide can be used.
- Benzodiazepines: Lorazepam, diazepam.

b. Vasodilators: Betahistine is recommended in high doses. The usual dose is 16 mg given TDS. Few studies recommend doses as high as 48 mg given TDS.

B. Surgical Treatment

This is reserved for patients with severe and repeated episodes of vertigo and who show no improvement on medical management (intractable vertigo). These include only 10–20% of the patients.

The procedures have been divided into:

I. Procedures with hearing and vestibular preservation.
II. Procedures with hearing preservation and vestibular ablation.
III. Procedures with hearing and vestibular ablation.

I. Procedures with hearing and vestibular preservation

1. *Intratympanic injection* of steroids after grommet insertion.
2. *Endolymphatic sac decompression* is a relatively safe procedure where the sac is opened medially into the subarachnoid space or the mastoid cavity. This surgical fistula is kept patent by inserting a silastic sheet. The preservation of hearing and vestibule is achieved.
3. *Sacculotomy* where a fenestra is made in the stapes footplate or round window membrane and a permanent shunt for draining of the saccule is created. It is associated with hearing loss in quite a number of patients.

Tenotomy of tensor tympani and stapedial tendon has no beneficial role in management of Ménière's disease and is now obsolete.

II. Procedures with hearing preservation and vestibular ablation

a. Intratympanic injection of gentamicin can be given after insertion of grommet in such patients. It is vestibulotoxic therefore causes *medical labyrinthectomy* and is effective for controlling vertigo in 80% of the patients. A solution of 26.4 mg/ml of gentamicin is instilled at a volume of 0.8 ml into the middle ear. Though the side effect of hearing loss is seen in 4–10% of the patients.

b. *Ablation of vestibular nerve*: This involves the selective sectioning of the vestibular part of the VIII nerve by the transtemporal supralabyrinthine approach (modified middle cranial fossa) or retrolabyrinthine approach.

III. Procedures with hearing and vestibular ablation: Transmastoid labyrinthectomy: In this procedure the inner ear is completely destroyed. It is only reserved for patients with intractable episodes of vertigo with severe hearing loss due to Ménière's disease.



Chapter

17

Hearing Impairment in Children

EN4.12: Elicit document and present a correct history demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of hearing loss.

"Earlier you catch the better it is." This essentially holds true for many causes of hearing impairment. It has been seen that after 5 years of age the results of hearing correction are not good. This is due to the phenomenon of neural plasticity of the brain which enables the brain for sound perception and its understanding, which is lost by the age of 5 years. Therefore, early detection of hearing impairment and its treatment is essential. A good history, examination and full audiological investigations are required for classification of deafness in children.

The hearing impairment is broadly classified as Flowchart 17.1

- Congenital—prenatal and perinatal causes
- Acquired.

CONGENITAL DEAFNESS

The term congenital deafness means the hearing loss which is present at the time of birth. These can be due to genetic or non-genetic factors. The genetic hearing loss can be classified into: syndromic and non-syndromic. 70% of the deafness is non-syndromic and the rest 30% is syndromic. The non-syndromic can be further divided into autosomal recessive, autosomal dominant and mitochondrial X-linked.

The congenital deafness can also be divided on the basis of type of hearing loss: **Conductive hearing loss** and **sensorineural hearing loss**.

A. Conductive Congenital Hearing Loss
(Table 17.1)

Down syndrome (Trisomy 21): The defect is seen in 1:600 births. The child has a narrow external auditory canal, otitis media with effusion and ossicular chain defect. The most common finding is small cochlea leading to sensorineural hearing loss (Fig. 17.1).

Flowchart 17.1: Type and prevalene of hearing impairment

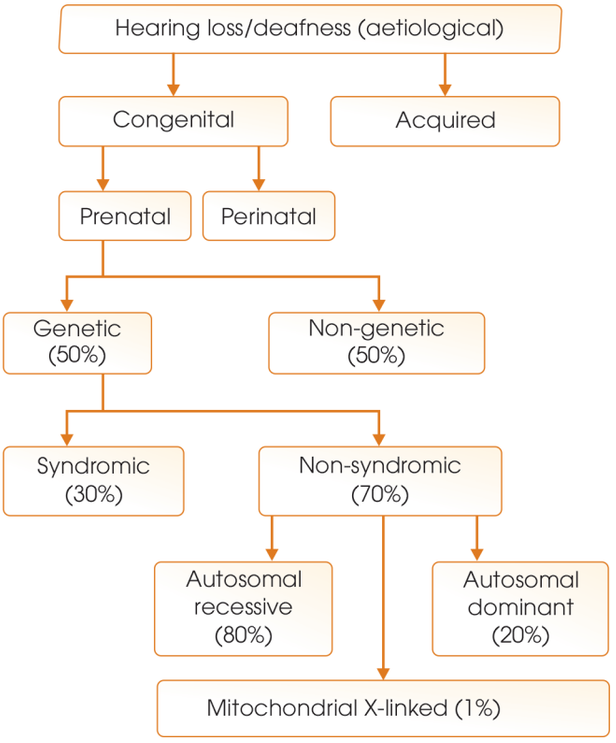


Table 17.1: Conductive congenital hearing loss causes

Genetic conductive causes	*Non-genetic conductive causes*
a. Down syndrome	a. Cystic fibrosis
b. Crouzon's disease	b. Immotile cilia syndrome
c. Marfan's syndrome	c. Cleft palate
d. Treacher–Collins syndrome	d. Immune deficiency disease
e. Pierre–Robin syndrome	e. Congenital cholesteatoma
f. Branchio-oto-renal syndrome	
g. Apert's disease	
h. Achondroplasia	
i. Osteogenesis imperfecta	
j. Otosclerosis	



Fig. 17.1: Down syndrome



Fig. 17.3: Treacher Collins syndrome

Crouzon' disease (craniofacial dysostosis): There is atresia of the external auditory canal with absent or fused malleus head to epitympanic wall. There may be deformed stapes with narrow round window niche (Fig. 17.2).

Treacher Collins syndrome (mandibulofacial dysostosis): The prominent feature is hypoplasia of the malar bones and mandible. There is antimongoloid slant to the palpebral fissures. There is deformity of pinna, atresia or stenosis of external auditory canal and tympanic membrane defects. Middle ear muscles may be absent. The ossicular chain can have a variety of malformations (Fig. 17.3).

Branchio-oto-renal syndrome: This syndrome is characterised by auricular or preauricular pits in association with ear malformations, branchial fistula and renal malformations.



Fig. 17.2: Crouzon's syndrome

B. Sensorineural Congenital Hearing Loss (Table 17.2)

Klippel–Feil syndrome (Brevicollis): The external ear has microtia with preauricular tags with atresia of the external auditory canal. There may be deformity of the incudostapedial joint and short process of incus fixation to the floor of the attic. Most patients have sensorineural hearing loss.

Goldenhar's syndrome (oculoauriculovertebral dysplasia): They have eye lesions which may be a cleft upper lid, dermoids and extraocular muscle defects. There may be auricular appendages, microtia and atresia of external auditory canal. Also there is club foot and hemivertebrae.

Waardenburg's syndrome: It is an autosomal dominant trait and accounts for 2% of all congenital deafness. 20% of these patients have characteristic white forelock. Another important feature is partial heterochromia iridis. The hearing loss is sensorineural and is present in 20–50% of patients. Vestibular hypofunction is a commonly overlooked feature (Fig. 17.4).

Pendred's syndrome: The patient has a defect in thyroxine synthesis with severe to profound hearing loss.

Table 17.2: Sensorineural congenital hearing loss causes

Genetic SNHL	*Non genetic SNHL*
a. Klippel–Feil syndrome	a. Torch infections
b. Turner's syndrome	b. Congenital syphilis
c. Fanconi's syndrome	c. Herpes simplex
d. Pendred's syndrome	d. Ototoxic drugs
e. Waardenburg's syndrome	e. Irradiation
f. Goldenhar's syndrome	f. Maternal diabetes
g. Jervel and Lange–Nielson syndrome (deafness with sudden death syndrome due to QT interval prolongation)	
h. Congenital hypothyrodism	



Fig. 17.4: Waardenburg syndrome

TORCH infections

- **Toxoplasmosis:** Adequate treatment initiated early in neonatal life can prevent SNHL due to it.
- **Rubella:** Rubella infection in intrauterine period causes deafness in one third of patients. First trimester infections are more likely to cause hearing loss. The hearing loss is usually bilateral mild to severe hearing loss.
- **Cytomegalovirus:** It produces bilateral sensorineural hearing loss. It may be mild at birth and it may increase progressively to a severe or profound category.
- **Herpes simplex virus:** Intrauterine HSV infection is rare and most neonatal infections are perinatally acquired via contact with infected lesions in the maternal genital tract. It causes SNHL which may be unilateral or bilateral.

Congenital syphilis is an infection vertically transmitted especially the maternal syphilis is untreated. It is presented as the "*Huctchinson triad*". This includes, the inflammation of the cornea, peg shaped upper teeth (Hutchinson teeth) and VIII cranial nerve palsy causing hearing loss. The hearing loss appears at 8–10 years of age. It is treated by high doses of penicillin.

Congenital hearing loss which are normal at birth but show hearing loss after birth.

a. Renal tubular acidosis
b. Cogan's syndrome
c. Norrie's syndrome
d. Alport's syndrome.

Cochlear deformity with sensorineural hearing loss

1. **Michael dysplasia:** This is the most severe with total absence of labyrinth, may be caused by failure of the otic vesicle to separate from the neural ridge.
2. **Mondini's dysplasia:** It affects the cochlea and semicircular canals. The cochlear duct is reduced to basal turn only. The organ corti may be absent or replaced by deformed cells. This is seen in Klippel–Feil and Penderd's syndromes.
3. **Bing–Siebenmann dysplasia:** In this the bony labyrinth is normal but the membranous labyrinth is underdeveloped.
4. **Scheibe (cochleosaccular) dysplasia:** This is the least severe deformity and luckily the most common, seen in 70% of the congenital defects. The utricle and semicircular canals are normal. The saccule is collapsed. The cochlea shows stria vascularis with alternate areas of aplasia and hyperplasia. This is seen in Waadenburg's syndrome, Usher's and Refsum's syndromes. It is also seen in rubella.
5. **Alexander's dysplasia:** Only the basal turn of cochlea is involved, leading to difficulty in perception of high frequency sounds.
6. **Enlarged vestibular aqueduct:** It causes fluctuant and progressive sensorineural hearing loss and is usually bilateral. It is said to be enlarged when the aqueduct is wider than 1.5 mm at midpoint or >2 mm at its operculum. It is often seen in Waardenburg syndrome and sometimes with Pendred syndrome. It is often associated with Mondini's deformity.

ACQUIRED DEAFNESS (Table 17.3)

Meningitis in childhood leads to SNHL in 10% of cases. All patients of meningitis should have screening for hearing loss. Measles infections can lead to hearing loss which is usually mild to moderate in most cases. It is usually conductive hearing loss but can be sensorineural. The perinatal causes which lead to deafness, are hypoxia, hyperbilirubinemia, low birth weight and prematurity (preterm birth).

HEARING LOSS MANAGEMENT

The early screening and timely management of such children will maximise hearing results. As, speech is dependent on hearing, therefore the early hearing rehabilitation is essential for proper speech and language development.

With children due to their innocent non-cooperation, requires special tests and methods to evaluate the hearing loss. If the hearing problem is encountered further investigations are done to find the cause and then finally appropriate management and rehabilitation is done.

Table 17.3: Acquired deafness causes

Conductive acquired	*SNHL acquired*
a. Otitis externa	a. Mumps
b. Acute otitis media	b. Measles
c. Chronic otitis media	c. Immunization
d. Otitis media with effusion	d. Meningitis (SNHL due to labyrinthitis ossificans)
e. Trauma	e. Ototoxicity
f. Foreign body	f. Trauma
g. Wax	

WHOM TO TEST

According to the new guidelines of National Programme for Prevention and Control of Deafness (NPPCD) it is mandatory to have a hearing screening for all the newborn within the first month of the birth. High risk screening involves screening of only high-risk children and it will fail to identify 50% of neonatal deaf cases, hence universal screening of all the new-borns is indicated. In cases of high degree of suspicion, it is mandatory to take detailed prenatal, perinatal and postnatal history.

The high-risk children include:

1. Family history of hearing loss
2. Syndromic children with craniofacial anomalies, including ear tags, ear pits and anomalies of outer ear, external auditory canal and temporal bone, telecanthus, heterochromia iridis, palatal abnormalities, white forelock, etc.
3. Prenatal infections (TORCH)
4. Toxaemia during pregnancy
5. Maternal illness especially diabetes
6. APGAR score less than 7 at 5 minutes
7. Low birth babies, less than 1500 g
8. Hyperbilirubinemia requiring exchange transfusion.
9. Ototoxic drug intake by mother during pregnancy.
10. Stay in neonatal ICU (NICU) for more than 5 days or receive assisted ventilation or exchange transfusion.

METHODS FOR HEARING LOSS DETECTION

I. Screening Methods

In a well-established screening programme, 50% of permanent hearing loss in children can be detected at birth. However, passing of screening does not guarantee normal hearing. Progressive or late onset hearing loss are seen with CMV infection or in some genetic conditions. Skilled observation and recording of child responses to speech and non-speech stimuli is referred to as behavioural hearing assessment.

Skilled testing by trained personnel in a suitable environment is essential. Even the best established neonatal hearing assessment programmes in developed countries have sensitivity of around 94% and specificity of 99.9%.

The documented limitations of screening methods are:

1. Newborn who show progressive hearing loss may be missed.
2. Mild loss in hearing at birth may be missed in screening test.

Also, false-positive test results might cause unnecessary anxiety in parents during the vulnerable first weeks of newborn.

The hearing test in newborn may be classified as behavioural tests and objective tests.

II. Following are the Behavioural Tests

A. *Auditory Response Cradle (ARC)*

It is a device where a microprocessor designed to record behavioural responses to noise of 85 dB and frequency of 2–4 kHz. The responses include head turn, body activity, startle response and respiration changes. The ARC is quite a reliable technique (failure rate of <2%) but may not be so consistent for special care babies.

B. *Behavioural Observation Audiometry (BOA)*

Responses to sound mature with age, as do other skills. A good knowledge of the reflexes and responses of the neonatal period is essential to estimate the hearing status of the child concerned.

Traditional methods of sound production are still used in some centres:

1. The cup and spoon methods for sound production.
2. Human voice is also used in some centres. The sound of 'oo' is usually around 500 kHz, hummed 'mm' is around 1 kHz and 's' is around 3 kHz.

Warble tones are now being used. These devices can produce sound up to 120 dB for a frequency range of 125 Hz to 8 kHz.

1. **Neonatal responses (startle reflexes):** In the early weeks after birth, following reflexes can be seen.
   - Aural–palpebral reflex
   - Change in heart rate and respiratory rate
   - *Moro's reflex*: Backward jerk of the head and body movements on loud noise.

   The above sounds require a sound of 80–90 db for the response to be elicited.
2. **1 month to 4 months:** The child by this age responds by Stilling and Listening to sounds which are familiar like of parents and relatives.
3. **4–6 months:** Child by this age learns to turn his head towards the sound and tries to localise the sound with increasing frequency. Infant will not only be more consistent now but also will respond to low intensity sounds. In children with hearing loss the response is delayed and a longer presentation of sound is required.
4. **7–9 months:** Child is more accurate and quicker with his responses. Also, may start to babble tunefully.
5. **10–12 months:** A child by this age starts to identify quiet sounds. By their birthday, vocalisation increases and there is an attempt to repeat a few words.
6. **1–2 years:** By this age the child has fast responses to sounds and also starts to anticipate and search sounds. There may be a requirement for distraction for performing tests. By 18–21 months may start to join two words.

C. *Distraction Testing for Hearing*

This is done for children who are above the mental age of 2 years. Here a **distracter** will keep the child involved in an activity and also would check the response of the child. The **tester** will produce the sound but staying away from his visual field. The sound production is done 1 m from the child's ear and on a horizontal plane. At no point of time does the distractor and tester communicate, even with eyes.

D. *Visual Reinforcement Audiometry (VRA)*

In this test the supra threshold sound is given along with visual stimulus. This test helps to perform the hearing test by using standard hearing tests. The sound is delivered via headphones or earphones and a flashing light or toy is used for visual reinforcement.

E. *Conditioning Audiometry*

1. **Play audiometry:** In this method the child is taught to join a game with the tester. A child above 2 years can participate in the test. Child is trained to carry out very simple tasks in response to the sound. Initially the task is taught with visual reinforcement and lip movements. Later the visual input is withdrawn and only auditory stimulus is given and response of the child seen. Important point to note here is not that the child performs action on sound stimulus but actually waits for the sound stimulus to be given.
2. **Speech audiometry:** The child is presented with speech above the threshold and the child is asked to repeat words. The intensity of sound is then lowered. This test also tests the speech discrimination.

F. *Speech Perception*

The recognition of sound is not the same as speech perception. Most of the current emphasis is on the detection of auditory disability and very less emphasis is on speech recognition skills. But with the advent of cochlear implants the focus has been widened to practically encompass speech perception as a very important area for child rehabilitation.

Geers and Moog in 1987 made five speech perception categories on the basis of variety of cognitively and linguistically appropriate test:

Level 1: Detection of sound but inconsistent pattern perception

Level 2: Consistent pattern perception, e.g. discrimination of monosyllable and multisyllable words.

Level 3: Inconsistent closed set word identification

Level 4: Consistent closed set word identification

Level 5: Open set word recognition.

Difficult child for testing

1. Behavioral testing is not suitable for children who remain below 4 months of mental age.
2. A child with poor vision may respond to sounds only by stilling and listening. Also, it has been seen that such children respond to voice not to warble notes. It may be difficult to be sure with their responses.
3. Patients of cerebral palsy are also difficult to test because of their voluntary movement limitations.
4. Autism
5. Hyperactive child is also difficult to test but patiently performing the test is vital.

III. Following are the Objective Tests

(Describe in Chapter 3: Audiology)

1. **OAE**
2. **Auditory brainstem response**
3. **Impedance audiometry**
4. **Acoustic stapedial reflex and acoustic reflex threshold decay**.

Electrocochleography **(ECoG)** was earlier being used for testing but has fallen out of use because it is an invasive test.

MANAGEMENT OF HEARING LOSS IN A CHILD

A holistic approach to such patients is required with the involvement of the child, parents and family. It leads to better hearing outcomes. The management of such a child has to start from finding the cause and the age at which it started. Most of such children are pre-lingual deaf but few may be post-lingual deaf. The next is to find out the degree of hearing loss.

Major aims of the treatment are early hearing perception (sound and speech) and development of speech and language.

Investigations and Referrals

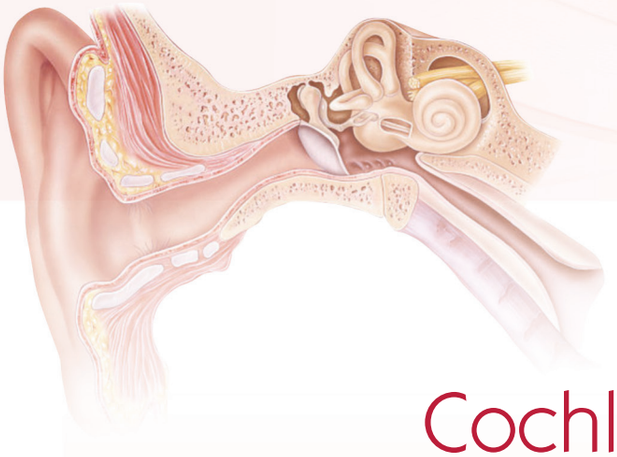
The hearing impairment is a multifactorial problem. This involves the role of multiple specialities.

1. *Radiology*: CT including 3D reconstruction is useful to define the inner ear bony dysplasia, integrity of the ossicular chain and enlarged vestibular aqueduct. MRI could help to delineate the membranous labyrinth, presence of auditory nerve and central auditory pathway.
2. *Blood tests*: Full blood counts, thyroid function test, ESR, VDRL, serum electrolytes, and kidney function test.
3. *Urine test*: For proteinuria and haematuria
4. *Serology*: For CMV infection and rubella
5. *Ophthalmological assessment*: These have to be picked up early to optimize language development.
6. *ECG*: Particularly required for Jervell and Lange–Nielson syndrome patients.
7. *Genetic screening*: GJB2 gene mutation caused by recessive deafness is particularly screened. Another important mutation is the GJB6 gene.

Treatment and Care

1. **Counselling of the parents** is the most important step in the hearing rehabilitation process of a deaf child. A realistic and holistic understanding of the problem by the parents should be done. Various modalities available and usable for the specific child should be discussed. They can also counsel them regarding the NGO groups, who provide financial support for treatment of such children.
2. **Hearing aids and devices** should be provided to the child as early as possible because the earlier perception of sound leads to better hearing outcomes.

   Accessories such as radio hearing aids systems (FM) or wireless streaming systems or sound field systems should be used in classrooms for the benefit of such children.
3. **Cochlear implantation** should be performed at the earliest in indicated children with profound hearing loss.
4. **Speech therapy** is very essential in children who have been provided with hearing aids. For postcochlear implantation a dedicated speech therapy is vital for gaining speech in such children.
5. **Sign language** is offered for children who have not gained hearing on treatment.
6. **Social guidance and confidence building** is essential to rehabilitate children so that they earn a living by taking vocational training courses and gain confidence to face various challenges in life.



Chapter

18

Cochlear Implant

EN4.12: Elicit document and present a correct history demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of hearing loss.

The cochlear implant (CI) is an electronic device which is implanted in children and adults having severe to profound sensorineural hearing loss. It can be implanted in a pre-lingual child who has not developed speech and language or in a post-lingual child or an adult who has already developed speech and language. Unlike hearing aids, which amplify sound, a cochlear implant bypasses the damaged portions of the inner ear to deliver electric signals to the auditory nerve endings directly. From here the VIII nerve carries the signals to the auditory cortex in the brain.

The implant consists of two main parts, namely the external part and the internal part.

1. The external part consists of a *Microphone*, *Speech processor* and a *Transmitter coil*.
2. The internal part consists of an *Internal receiver-stimulator package* and *Intracochlear electrode array*.

EXTERNAL PART

The external part detects the acoustic signals and converts it into an electric signal. The encoded signal is transmitted to the internal device using radiofrequency via the external coil. This part is worn behind the ear. Newer implants have the facility to provide communication with the hearing aid on the contralateral ear for bimodal hearing.

INTERNAL PART

This part of CI is surgically implanted behind the ear. It consists of a receiver stimulator package which has a magnet, electronic system and a stimulating electrode array. The electronic signals are decoded by the receiver stimulator package, which transmits them to the electrodes by using a speech strategy, which is specific for the manufacturer of the implant. The examples are the CIS strategy for Clarion® and SPEAK strategy for Nucleus® implant. The electrode array is inserted into the scala tympani of the basal turn of cochlea, from where the bipolar spiral ganglion cells are stimulated and further the signals are sent to the auditory cortex via the auditory nerve.

The normal auditory nerve has approximately 35,000 nerve fibres and it is essential that at least 10,000 of them should get stimulated for good speech perception using the cochlear implant. The electrode array ideally should be completely inserted along the entire length of the cochlear duct, but it may be incomplete depending upon the congenital abnormalities of the inner ear.

COCHLEAR IMPLANT TEAM

The cochlear implantation team is multidisciplinary. The team includes:

1. Teacher for deaf
2. Speech and language therapist
3. Psychologist
4. Audiologist
5. Radiologists
6. Surgeon
7. Parents.

CANDIDACY FOR COCHLEAR IMPLANTATION

1. A cochlear implant should be considered for any person having severe to profound hearing loss and does not gain adequate benefit from acoustic hearing aids. Severe to profound deafness is defined as the inability to hear sounds of intensity less than 90 dB HL at 2 kHz and 4 kHz without hearing aids. Hearing aids should be used for at least 3 months unless inappropriate or contraindicated.
2. Elderly with SNHL also do well with CI. Recent studies show better cognitive skills in patients with hearing loss who underwent CI.
3. Cochlear fractures following temporal bone fracture are good patients for CI, if implanted early before cochlear fibrosis sets in.

Special situations

4. Bilateral CI implantation can be done in all who are blind and auditory stimulus is of primary importance for spatial awareness.
5. Single ear deafness (SSD) is no more a contraindication for CI and dual hearing improves speech recognition in noise.
6. Auditory neuropathy/auditory dyssynchrony (ANDS) patients also are benefitted by CI.
7. Neurofibromatosis type 2 was earlier a contraindication, and such patients were referred for auditory brainstem implants (ABI). Recent advances in tumour resection keeping the auditory nerve intact, makes the CI possible in such cases.

WORKUP FOR COCHLEAR IMPLANTATION

The workup for the patient is of critical importance. The patient should be medically fit and also free of ear infections. Mucosal chronic otitis media should be treated by myringoplasty three months before CI surgery. A squamosal otitis media patient should undergo radical mastoidectomy with cavity obliteration and cul-de-sac closure of external auditory canal before cochlear implantation.

The child should be fully vaccinated against meningitis, i.e. *H. influenzae* and *Meningococcus*.

Radiology

HRCT (high-resolution CT scan) of temporal bone helps to identify inner ear abnormalities, cochlear lumen obliteration, middle ear pathology and other anatomical variations like low-lying dura or anterior position of sigmoid sinus. CT can pick up a cochlear hypoplasia and enlarged vestibular aqueduct where an abnormal communication between CSF and cochlea exist. This situation may lead to *perilymph gusher* during surgery. It also gives better information in cases of labyrinthitis ossificans, which occurs after meningitis and is a contraindication for CI.

MRI helps in diagnosing cochlear nerve aplasia, wherein the CI is contraindicated and Auditory brainstem implant is required (Fig. 18.1).

Parental expectations should be realistic and they should be motivated for rehabilitation. Age less than 5 years is desirable for prelingually deaf children due to the reasons of neural plasticity. Neural plasticity is the ability of the brain to learn new skills by changing the growth and reorganisation of neural network. For auditory function this can occur till the age of around 5 years. The neural area of the brain is predetermined for a particular skill like hearing, speech and language development. The areas in the brain cortex responsible for auditory function and speech are lost and encroached by other areas, if not stimulated during early years of life. Hence it is important to diagnose congenital hearing loss at the earliest and perform early rehabilitation.

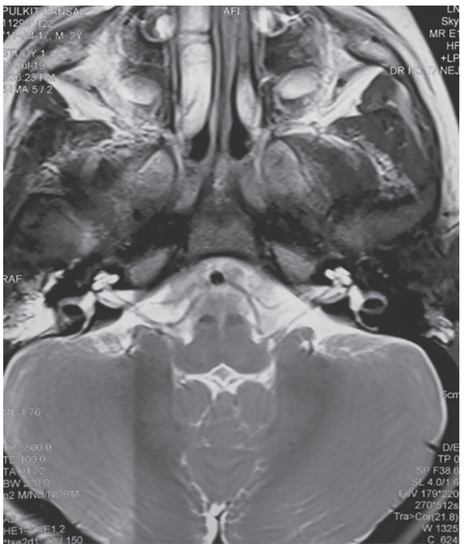


Fig. 18.1: MRI brain showing bilateral VII–VIII nerve complexes

Audiology

Audiological investigations which are mandatory before taking up for CI are:

1. Pure tone audiogram (PTA)
2. Tympanometry
3. Otoacoustic emissions (OAE).
4. Auditory brainstem responses (ABR).

The psychological evaluation should also be done in all cases.

Devices

Various devices available in the marker are shown in Fig. 18.2.

Cochlear Implantation Surgical Steps

1. The surgery is performed under general anaesthesia with the patient in supine position and the head turned 45–60 degrees away from the surgeon.
2. Post auricular incision is given, usually of C shape.
3. The skin flap is elevated along with subcutaneous tissue and part of the temporalis muscle.
4. A cortical mastoidectomy is performed with keeping overhanging edges.
5. Middle ear is then opened via posterior tympanotomy through the facial recess. The round window niche then is visualised (Fig. 18.3).
6. Then a bony cochleostomy is done anteroinferior or inferior to the round window by creating an opening in the basal turn of the cochlea, with an axis of drilling towards the cochlear apex (Fig. 18.4A). This will ensure that the implant electrode goes into the scala tympani.
7. Through this opening the electrode array is passed into cochlea. Electrodes may also be inserted via the round window. The latter is preferred these days, because it causes less damage to intracochlear



Fig. 18.2: Various devices available in the market

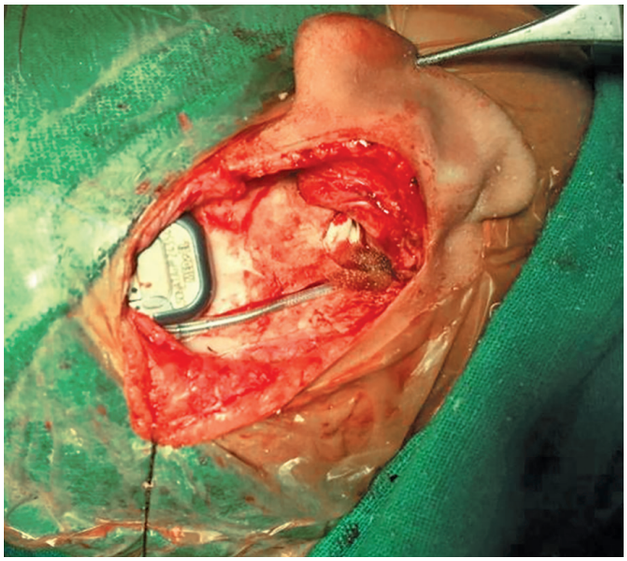


Fig. 18.5: Internal receiver stimulator unit

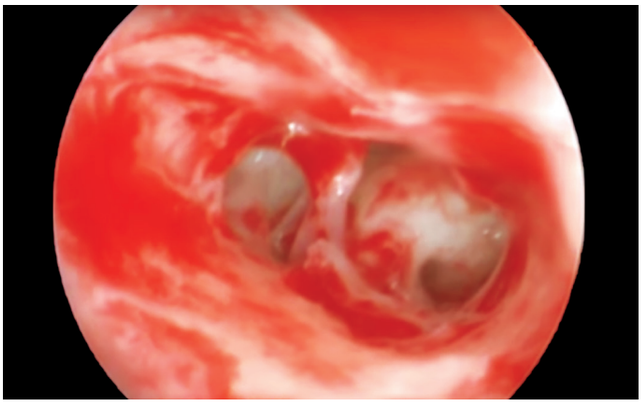


Fig. 18.3: Round window identification via posterior tympanotomy

structures, less incidence of postoperative vertigo and is more physiological.

8. A bony well is created for the receiver-stimulator, which is fixed with non-absorbable suture (Fig. 18.5).
9. Finally, the electrodes are inserted by using fine forceps (Fig. 18.4B). Hyaluronic acid or 50% glycerine may be used to keep blood out of the scala during electrode insertion and to lubricate the electrode.
10. Muscle pieces from temporalis muscle are used to plug 360 degrees around the rest of the cochleostomy site.
11. Neural response telemetry is performed to check for the functional status of electrodes once the implant is in position.
12. The incision is then closed in layers.
13. Postoperative X-ray (Stenver's view) is done to confirm the electrode position (Fig. 18.6).

COMPLICATIONS

The complications due to cochlear implant surgery can be classified as major and minor, according to the severity (Table 18.1).

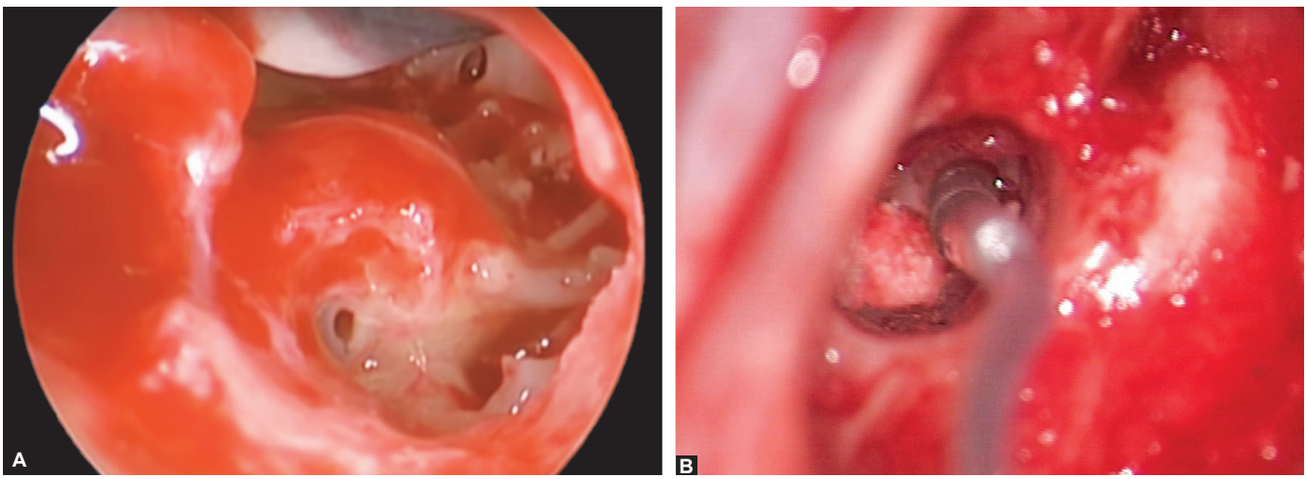


Fig. 18.4: (A) Round window opening made for electrode insertion; (B) Electrode inserted through the cochleostomy

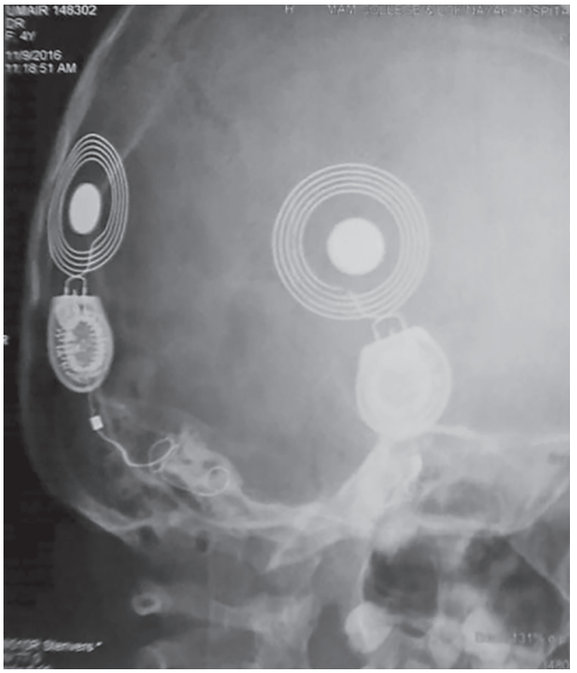


Fig. 18.6: Electrode array in cochlea (Stenver's view)

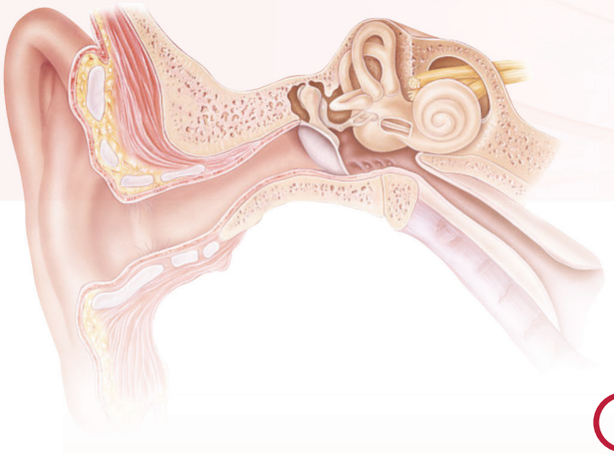
Table 18.1: Complications of cochlear implantation	
Major	***Minor***
1. Facial nerve injury	1. Wound infection
2. Displacement of device	2. Otitis media
3. Device failure	3. Vertigo
4. Meningitis	
5. CSF leak	
6. Flap necrosis	



Fig. 18.7: Postoperative picture of a child with external device (speech processor and transmitting coil)

OUTCOMES

- Most children who are implanted below 2 years of age show similar language and speech development as normal children. Those who are implanted after 3 years usually struggle to catch up with normal peer group. This is due to the sensitive period of neural plasticity, where 2–3 years is the most important time (Fig. 18.7).
- Cognitive impairment does influence the language and speech outcomes.
- For adults, the duration of hearing loss (profound or severe) before intervention, does influence the outcomes. This is due to the deterioration of unstimulated spiral ganglion cells.



Chapter

19

Otalgia

EN4.1: Elicit document and present a correct history, demonstrate and describe the clinical features, choose the correct investigations and describe the principles of management of otalgia.

Otalgia is pain in the ear caused by pathologies inside the ear (primary otalgia) or it may be due to pain referred from pathologies elsewhere (referred otalgia).

PRIMARY OTALGIA

The sensory supply to the ear is from greater auricular nerve (C2, 3) lesser occipital (C2, 3), auriculotemporal (V3), Jacobson nerve (branch of IX N), Arnold's nerve (auricular branch of X N) and the facial nerve. The causes arising within the ear produce pain due to direct stimulation of these nerves.

Causes

A. *Pinna*
   1. Trauma
   2. Cellulitis
   3. Herpes zoster oticus
   4. Malignancy
   5. Chronic relapsing perichondritis
   6. Chondrodermatitis nodularis helicis.

B. *External auditory canal*
   1. *Otitis externa*:
      - Acute or chronic
      - Malignant otitis externa
   2. Furuncle
   3. Keratosis obturans (*Canal cholesteatoma*)
   4. Herpes zoster oticus
   5. Malignancy
   6. Bullous myringitis.

C. *Middle ear*
   1. Otitis media
   2. Malignancy.

D. *Inner ear*
   Noise injury.

REFERRED OTALGIA

The nerves involved in referred pain to the ear are:

1. **Trigeminal nerve:** It is the most common route of referred pain. It supplies the anterior two-thirds of tongue, floor of mouth, palate, lower teeth, major salivary glands, temporomandibular joint and mandible.
   Site of perception: Pain arising from these structures is referred commonly to the pinna, tragus, anterior wall of EAC.
   Causes: Dental caries, impacted molars, malignancy of tongue, osteoarthritis of temporomandibular joint, ill-fitting dentures, parotid infection.
2. **Glossopharyngeal nerve:** It receives sensation from the posterior third of tongue, palatine tonsil, nasopharynx, hypopharynx, parapharyngeal space and retropharyngeal space.
   Site of perception: Produce deep ear pain-middle ear and Eustachian tube.
   Causes: Acute tonsillitis, peritonsillar abscess, malignancy of base tongue and tonsil, Eagle syndrome, parapharyngeal abscess.
3. **Vagus nerve:** This supplies the valleculae, supraglottis, thyroid gland, tracheobronchial tree and oesophagus.
   Site of perception: Inferior and posterior aspect of EAC.
   Causes: Malignancy of the supplied structures, thyroiditis.
4. **Upper cervical nerves (C2 and C3):** They supply the muscles and facet joints of the cervical spine.
   Site of perception: Posterior part of pinna and skin over mastoid.
   Causes: Caries spine, cervical spondylitis, spinal stenosis, osteoarthritis.

TEMPOROMANDIBULAR JOINT DYSFUNCTION SYNDROME

The syndrome is characterised by diffuse pain in and around the joint with crepitus and trismus. Females are more predisposed, and bruxism is the most common aetiological factor, being present in 50% of the patients.

The most reliable clinical finding is muscle tenderness and 85% of the patients have tenderness of the lateral pterygoids. This can be confirmed via intraoral palpation. The medial pterygoids may also be tender. Patients with bruxism have flattening of the occlusal surfaces of the molar teeth, worn incisors and linear scarring of the buccal mucosa and lateral border of tongue. Treatment is a soft diet, hot fomentation and muscle relaxants.

COSTEN SYNDROME

It is also called as temporomandibular joint syndrome or mandibular joint neuralgia.

The patient has hypertonia of masticatory muscles leading to radiation of pain to various head and neck regions. Symptoms include ear pain, tinnitus, dizziness with impaired hearing and aural congestion. The treatment of such patients includes rest and a soft diet. Anti-inflammatory drugs like ibuprofen or diclofenac can be used to control pain and inflammation. Benzodiazepines or low dose tricyclic antidepressants may be used for refractory cases. Muscle relaxants are helpful to reduce stiffness.

TMJ surgery (condylotomy) with occlusal splints may be offered as the last resort.

GLOSSOPHARYNGEAL NEURALGIA

It is characterized by severe transient pain in ear, base of tongue, tonsillar fossa or below the angle of jaw. It is mainly of two types: (a) *tympanic type*: this mainly affects the ear, (b) *oropharyngeal type*: which mainly affects the oropharynx.

The onset of symptoms is usually by swallowing or sometimes by coughing, yawning or talking.

Usually there is no evident lesion affecting the nerve. The most common cause is vascular compression of the nerve by posterior inferior cerebellar artery.

The first line of treatment for these patients is carbamazepine. The surgery for the neuralgia will involve microvascular decompression. The nerve section may be performed as the last resort via the posterior fossa approach, via neck or the transtonsillar approach.

EAGLE'S SYNDROME (STYALGIA)

This syndrome involves neuralgia secondary to an elongated styloid process. The elongation results from calcification of the stylohyoid ligament.

The normal length of the styloid process is 2.5 cm, but is considered elongated when it is more than 4 cm long. The symptoms are postulated to be due to compression/irritation of the glossopharyngeal nerve, impingement of the carotid vessels or due to inflammation at the insertion of the stylohyoid ligament.

Clinical Features

The main symptoms consist of dull and persistent throat pain that may radiate to the ear via glossopharyngeal nerve. Also, there is odynophagia and foreign body sensation in the throat.

The styloid process may be palpable in the tonsillar fossa and local injection of local anaesthetic can temporarily relieve symptoms.

X- ray skull AP view, orthopantomogram (OPG) or a CT scan will confirm the elongation of the styloid process (Fig. 19.1).

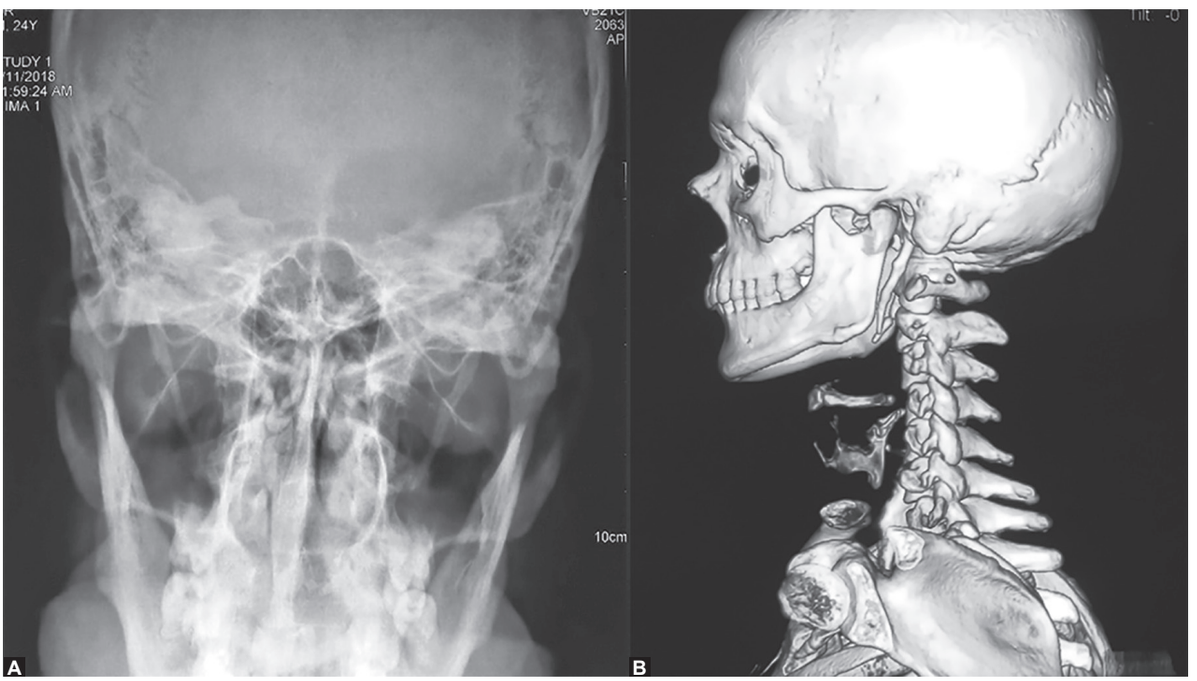


Fig. 19.1: Bilateral enlarged styloid process as seen on X-ray and 3D CT reconstruction

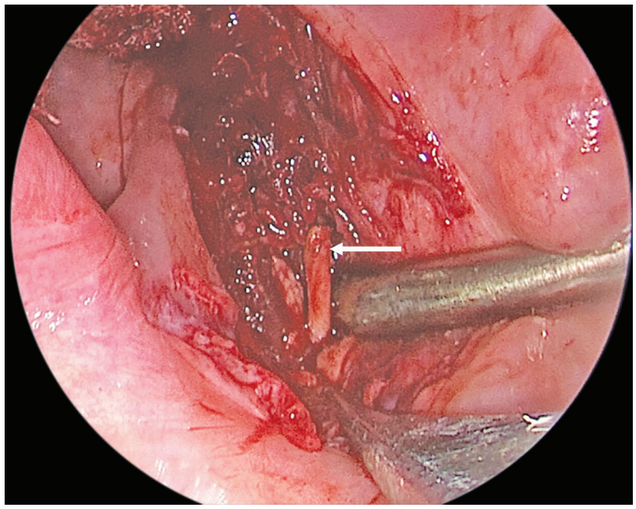


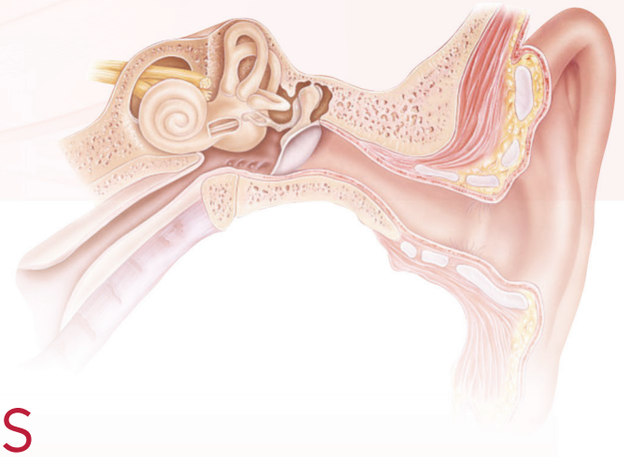
Fig. 19.2: Intraoperative picture of enlarged styloid process (marked as white arrow) after performing tonsillectomy

Management

- The conservative management includes steroids, local anaesthetic and carbamazepine.
- The surgical management requires the removal of the elongated process. This procedure can be done via the transoral approach (approached via the tonsillar fossa following tonsillectomy) or via a transcervical (external approach). Recently there has been increased use of endoscopes to better visualise and remove the enlarged styloid process by transoral approach (Fig. 19.2).

Chapter

20



Tinnitus

EN4.21: Describe the clinical features, investigations and principles of management of Tinnitus.

The word tinnitus is derived from the Latin word ***tinnire*** ('to ring'). Dennis McFadden first described this condition and defined it as the conscious expression of sound that originates in an involuntary manner in the head of its owner or may appear to do so, in absence of an external sound or electrical stimulus.

Tinnitus can also be classified into subjective or objective types.

In the **subjective type**, only the patient is aware of the sound sensation.

In the **objective type**, the sound can also be perceived by the examiner, either unaided or with a stethoscope or microphone with amplifier. It can also be divided into pulsatile and nonpulsatile objective type tinnitus.

Meniere's disease, otosclerosis and vestibular schwannoma are associated with an increased risk of tinnitus and such cases are sometimes referred to as cases of '*syndromic tinnitus*'.

The prevalence of tinnitus is about 10–15% and is equally found in both sexes. The sound is usually heard in the middle of the head or ears in half of the sufferers. The majority of the patients report tinnitus, which is subjective non-pulsatile without any pathological association. Mostly the tinnitus sounds can be described as whistling, humming, or ringing. These sounds can be complex like musical tunes. Careful history should rule out meaningful sounds which might be due to *auditory hallucinations* (Flowchart 20.1).

The **risk factors** for tinnitus are smoking, alcohol, head injury, and cardiovascular diseases.

Various causes of tinnitus are mentioned in Table 20.1.

SUBJECTIVE TINNITUS

Investigations

1. Pure tone audiometry
2. Impedance audiometry
3. Speech audiometry
4. Specific tinnitus tests assess loudness discomfort levels, tinnitus pitch matching, tinnitus loudness matching and minimal masking levels.
5. MRI brain: In unilateral tinnitus, MRI is required to rule out retrocochlear pathologies like acoustic neuroma.

The patients can be evaluated by Tinnitus Handicap questionnaire or visual analogue score (0–10) to know the degree of tinnitus.

Treatment

1. **Counselling and reassurance:** It is the most important step in the management of these patients. The clinicians must avoid any negative counselling, which can be very damaging to the patient.
2. **Hearing aids:** Tinnitus has a strong association with hearing loss, therefore providing hearing amplification can reduce symptoms. Hearing aids amplify external sounds and mask tinnitus.

Flowchart 20.1: Pathophysiology of tinnitus

Pathophysiology

Peripheral causes

1. Discordant damage to cochlear hair cells (noise and aminoglycoside drugs)
2. Calcium channel dysfunction (salicylates and quinine)
3. Glutamate receptors excitation (loss of protection from noise and salicylates)

Central causes

1. Increased spontaneous firing by the auditory cortex
2. Increased central neural synchrony
3. Reorganisation of cortical auditory map

Table 20.1: Causes for tinnitus		
Subjective		*Objective*
Ear causes	**Non-ear causes**	**A. Pulsatile**
• Wax	• Anaemia	• Atherosclerotic carotid artery disease
• Impacted ear foreign body	• Hypertension	• Arteriovenous fistula
• Acute otitis media	• CAD	• Arteriovenous malformation
• Chronic otitis media	• Migraine	• Carotid artery aneurysm
• Secretory otitis media	• Drugs-like salicylates, quinine, aminoglycoside antibiotics, cisplatin	• Dissection of the carotid artery
• Meniere's disease		• Vascular compression of the VIIIth nerve
• Otosclerosis		• Glomus tumour
• Noise trauma		**B. Nonpulsatile**
• Ototoxic drugs		• Patulous Eustachian tube[1]
• Presbycusis		• Palatal myoclonus[2]
• Tumours of VIII nerve		

[1]Patients of the patulous Eustachian tube have tinnitus which is synchronous with respiration.
[2]In palatal myoclonus clicking sound is produced due to clonic contraction of the muscles of the soft palate. Rarely contraction of muscles of middle ear can also cause tinnitus.

3. **Sound therapies:** Use of tinnitus maskers in the ear, which generate wide-band sound. Such devices deliver masking or sound therapy so that the tinnitus is masked. An alternative is environmental sound enrichment, meaning sound from the environment like electric fans, air-conditioners, radio or soft music, etc. may be used to mask the tinnitus.
4. **Novel sound therapy:** Several devices claim to target tinnitus by sound therapies. The devices include Neuromonics, Serenade, Noise cancellation and Acoustic CR neuromodulation.
5. **Combination treatment modalities:** It combines the directive counselling and sound therapy and promotion of habituation to the sound **(TRT: Tinnitus-retraining therapy)**.
6. **Cognitive behavioural therapy:** It helps to improve the quality of life of such patients.
7. **Psychological treatments:** These have been found to be effective in many patients. These include *Mindful meditation, Acceptance and commitment therapy*.
8. **Surgery:** It has a definitive role in management of tinnitus associated with otosclerosis, Meniere's disease and glomus tumours.

The use of systemic drugs, intratympanic drugs and dietary supplements is still not proven efficacious.

OBJECTIVE TINNITUS

These can either be of pulsatile/synchronous or non-synchronous/nonpulsatile type.

Synchronous Objective Tinnitus

The pulsatile tinnitus is in synchrony with heartbeat, and it is because of abnormal vascular anatomy in relation to the ear. The list of causes for pulsatile tinnitus are mentioned in Table 20.1.

Investigations

1. Contrast enhanced computed tomography (CT) of the temporal bone.
2. Doppler USG in case of suspected atherosclerotic carotid artery disease.
3. Magnetic resonance angiography (MRA).

Management

Management of pulsatile tinnitus requires surgery depending on the aetiology. Treatments may include decompressive and destructive procedures. Surgery for carotid aneurysm and AV malformation may be attempted by experts. Microvascular decompression of vascular loops around the VIII nerve may be performed in severe intractable tinnitus and vertigo.

Nonsynchronous Objective Tinnitus

Tinnitus that is not synchronous with the pulse is termed as non-synchronous pulsatile tinnitus. These sounds are often related to myoclonic activity of the middle ear muscles (tensor tympani and/or stapedius). It can also develop due to the palatal muscle's myoclonic contraction.

Investigation

For palatal myoclonus, the involuntary palatal movements may be observed during oropharyngeal examination or it can be confirmed by fiberoptic endoscopy.

On impedance audiometry, there is change in middle ear compliance with contraction of the middle ear muscle.

Treatment

Trial of medical treatment with benzodiazepines, carbamazepine, and piracetam can be done. If not responding, then surgical division of the tendons of middle ear muscles is recommended and it has good results.

Chapter

21



Paragangliomas of Temporal Bone

The paraganglia are cells derived from the neural crest and are found in sites of autonomic nervous system usually at carotid, ciliary and vagal bodies, aorta and main branches of glomus jugulare complex, bladder and adrenal medulla. The paraganglia of the adrenal gland secrete catecholamines, adrenaline and nor-adrenaline. These stains with chromaffin and hence are called **chromaffin positive**. Rest of the paraganglia do not stain with chromaffin therefore are **chromaffin negative**.

The paragangliomas of the temporal bone are tumours which are derived from paraganglia present in adventitia of the jugular bulb and along the IX and X cranial nerve and with Jacobsen's plexus on promontory.

Temporal bone paragangliomas can be divided into three types:

1. Jugular
2. Jugulotympanicus
3. Tympanic (glomus tympanicus).

ETIOLOGY

There is a female preponderance of 6 : 1. It is more common in the middle age group. About 40% of the patients with head and neck paragangliomas have a mutation of the succinate dehydrogenase gene. A portion of patients have multiple paragangliomas. Their spread is mainly local, hence these are considered as low grade malignant tumours.

CLASSIFICATION

Fisch classification widely followed for paragangliomas is based on MRI and CT imaging (Table 21.1).

Table 21.1: Classification of paragangliomas

Classification	*Spread*
Class A	Limited to mesotympanum
Class B	In the middle ear-mesotympanum and hypotympanum
Class C	Tumour originates in the dome of the jugular bulb, destroying the overlying bone, and may spread inferiorly, posteriorly, superiorly, laterally and or medially.
Class D	Intracranial extension

CLINICAL FEATURES

These are very slow growing tumours but without any substantial increase in size, these can produce symptoms due to local erosion and compression.

1. Pulsatile tinnitus and conductive deafness (69%) are the most common symptoms of the patients.
2. Hoarseness and dysphagia
3. Otorrhea
4. Endocrine disturbances
5. Headache and visual disturbances.

On Examination

1. On otoscopy: A red coloured middle ear mass **(rising sun sign)** can be seen in 75% of the patients (Fig. 21.1).
2. Cranial nerve palsies are reported in about 30% of the patients. These nerves are mainly involved in the jugular fossa.
3. Facial nerve palsy.

DIFFERENTIAL DIAGNOSIS

1. High jugular bulb
2. Aberrant carotid artery

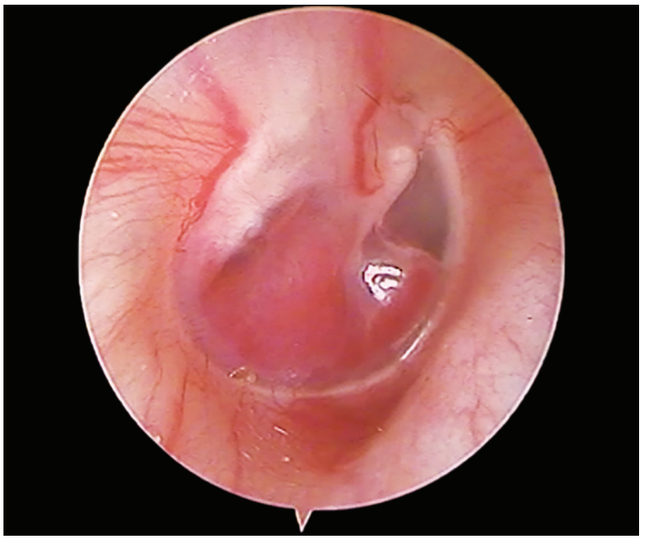


Fig. 21.1: Rising-sun sign seen in case of glomus tympanicus

3. Squamous cell carcinoma
4. Neuromas of the last four cranial nerves.

INVESTIGATIONS

1. Pure tone audiometry will reveal conductive hearing loss in most cases.
2. Contrast-enhanced CT scan of temporal bone will delineate the bony destruction and extent of the tumour and help in surgical planning (Fig. 21.2).
   Class C tumours cause erosion of the bone around the jugular bulb. The absence of bony crest between carotid canal and jugular fossa is virtually diagnostic of jugulare type of tumour **(Phelp's sign)**.



Fig. 21.2: HRCT scan showing glomus tympanicus tumour (Black arrow)

3. MR angiography can help in identifying the feeder vessels to the tumour and decision regarding preoperative embolization.

Note: The biopsy of the tumour is unnecessary especially when radiology is strongly suggestive of the disease. It may rarely be done to rule out squamous cell carcinoma but is not without excessive bleeding requiring ear packing.

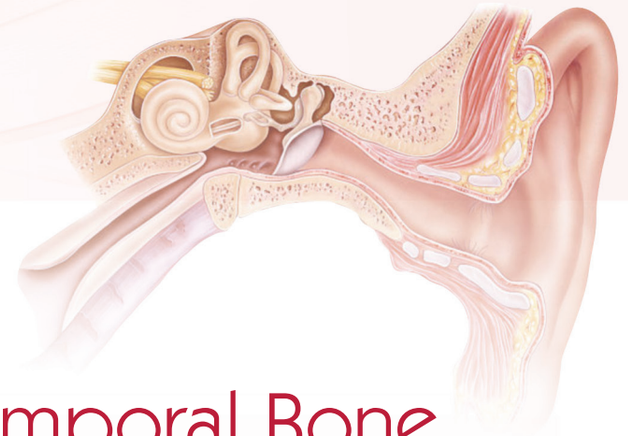
TREATMENT

The current treatment guidelines suggest following options for management:

1. **Observation:** This is suggested for patients who present with small tumours in the sixth or seventh decade. These patients can be followed up on serial CT scans. No treatment is offered apart from counselling and reassurance.
2. **Primary radiotherapy:** This is indicated for small tumours especially in elderly. The symptoms of tinnitus and vertigo may improve with radiotherapy but deafness and cranial nerve palsies persist. It acts by inducing fibrosis which sets in about 6–12 months post radiotherapy.
3. **Surgery:**
   - Preoperative selective embolization of the feeder vessels supplying the tumour is helpful to reduce per-operative bleeding.
   - A small paragangliomas **(glomus tympanicus)** are surgically managed by permeatal/endaural approach, by using an operating microscope or otoendoscopes.
   - Extensive tumours will require extended facial recess approach or the lateral approach (Fish infratemporal fossa approach) for complete excision.

Chapter

22



Tuberculosis of Temporal Bone

TUBERCULOSIS

Tuberculosis of the temporal bone is a chronic infection which is commonly caused by *Mycobacterium tuberculosis* but rarely by *Mycobacterium avium* and *Mycobacterium bovis*. The incidence of tubercular infection of temporal bone is very rare in western world, but it is not uncommon to be seen in developing countries.

Pathogenesis

The usual route of involvement is through direct spread via the perforation of the tympanic membrane. It can though be due to ascending infection into the middle ear via the Eustachian tube or rarely by haematogenous spread.

There are three stages of infection:

1. Acute phase: There is congestion of the tympanic membrane which is followed by thickening.
2. Chronic phase is very slow and is characterised by multiple perforations and pale pink granulations.
3. Caseous phase or necrotic phase has destruction of bone with sequestrum formation.

Clinical Features

1. Chronic ear discharge not responding to usual antibiotic therapy.
2. The characteristic features are multiple perforations (which may join later to form a larger perforation) is classically described but rarely seen (Fig. 22.1).
3. Pale granulations.
4. Early severe hearing loss.
5. Painless otorrhoea (absence of otalgia).
6. Facial weakness (10–20%).
7. Bone necrosis often presents as mastoid fistula (Fig. 22.2).
8. Pale middle ear mucosa.

Any patient with facial palsy without any squamosal COM disease, hearing loss out of proportion of otoscopic findings and absence of otalgia, should raise a suspicion of tubercular otitis media.

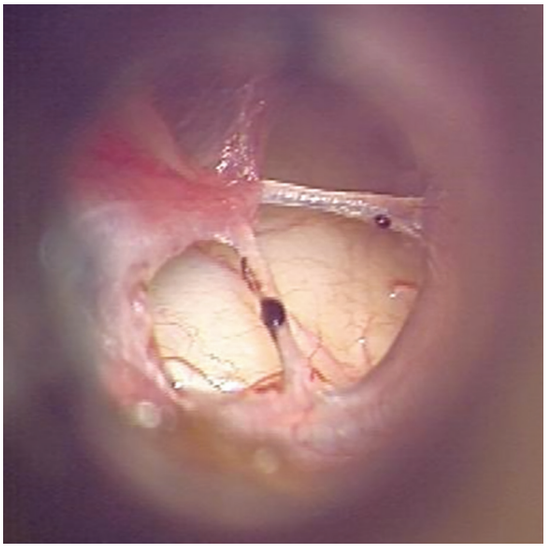


Fig. 22.1: Multiple perforation in a case of tubercular otitis media



Fig. 22.2: Postaural mastoid fistula with granulations

Diagnosis

1. The diagnosis is confirmed by a biopsy of granulations, polyps from middle ear or tissue removed from mastoids or external auditory canal. Many times, it is diagnosed after mastoidectomy when the suspected granulations(pale and fleshy which look like chicken fat) from the middle ear are positive for tuberculosis on histopathology.
2. In case of high suspicion the ear discharge may be sent for CBNAAT(Cartridge-based nucleic acid amplification test). It is a rapid test based on polymerase chain reaction and can detect DNA of *Mycobacterium tuberculosis*.
3. An HRCT scan of temporal bone is the radiological investigation of choice. It usually shows soft tissue contents in the middle ear cleft with destruction of the mastoid air cell system. In initial cases scutum is not eroded. In advanced cases there is extensive destruction of temporal bone. The CT scan also helps to delineate complications of facial nerve involvement and labyrinthine fistula (Fig. 22.3).



Fig. 22.3: HRCT temporal bone showing postaural fistula

CT scan of the same patient showing a large mastoid cavity with extensive destruction of the mastoid cortex.

Complications

1. Facial nerve palsy
2. Profound hearing loss (cochlear involvement)
3. Postaural fistula
4. Meningitis or tuberculoma
5. Otitic hydrocephalus
6. Multiple cranial nerve palsies.

Management

- The main line of treatment is the anti-tuberculous therapy for 6–9 months. In case of complications like meningitis, an extended course of ATT is required.
- Mastoid exploration is done in case of extensive bone destruction or sequestrum formation. Facial nerve decompression is indicated in patients who do not respond to ATT.

OTOSYPHILIS

Otosyphilis is usually diagnosed by various cochleovestibular symptoms and serological tests. The syphilis is of two types, congenital and acquired syphilis The congenital syphilis usually presents at around 2 years. The acquired form manifests at the age of 8–20 years. The features of the disease include:

1. Early neurosyphilis mainly presents as meningitis with or without cranial nerve involvement. Hearing symptoms are of a sudden hearing loss.
2. Late neurosyphilis presents with hearing loss (fluctuating hearing loss) and/or vertigo.
3. False-positive fistula test (*Hennebert's sign*) seen due to formation of fibrous adhesions between the stapes footplate and membranous labyrinth.
4. Tullio phenomenon is sometimes seen, which is the presence of vertigo on exposure to loud sounds.

Treatment recommended is aqueous penicillin G, 18–24 million units/day for 10–14 days. Alternatively, procaine penicillin 2.4 million units may be given IM daily along with probenecid 500 mg by mouth 4 times/day for 10–14 days.

Chapter

23

Vestibular Schwannoma

Vestibular schwannoma is also called acoustic neuroma or acoustic neurilemmoma. These are the most common tumours of the cerebellopontine angle* (Fig. 23.1).

The vestibular schwannoma comprises only 10% of the all tumours of the brain but are by far the most common tumour of the cerebellopontine angle (80–95%) (Table 23.1).

The vestibular schwannoma is a benign neuroectodermal tumour of the Schwann cells. It arises from vestibular division of the vestibulocochlear nerve (VIII N) at the transition point between the glial cells and Schwann cells **(Obersteiner–Redlich zone)**. This zone lies with in the internal acoustic meatus (IAM). The structures in the IAM are described in Fig. 23.2.

They are very slow growing tumours and usual growth is less than 2 mm/year. The growth of the tumour though may be more in few cases where the symptoms appear earlier than usual. The tumour of the nerve initially causes the expansion of the IAM by canal erosion, and then further expands into the

Table 23.1: Lesions of the cerebellopontine angle

Benign	*Malignant*
Vestibular schwannoma	Metastases from breast, NP, parotid
Meningioma	Malignant melanoma
Epidermoid cyst	Lymphoma
Arachnoid cyst	Chondrosarcoma
Facial nerve schwannoma	SCC
Cochlear schwannoma	Hemangiopericytoma
Trigeminal schwannoma	
Cholesterol granuloma	
Tuberculoma	
Dermoid cyst	
Lower cranial nerve schwannoma	
Cysticercosis	
Haemangioma	

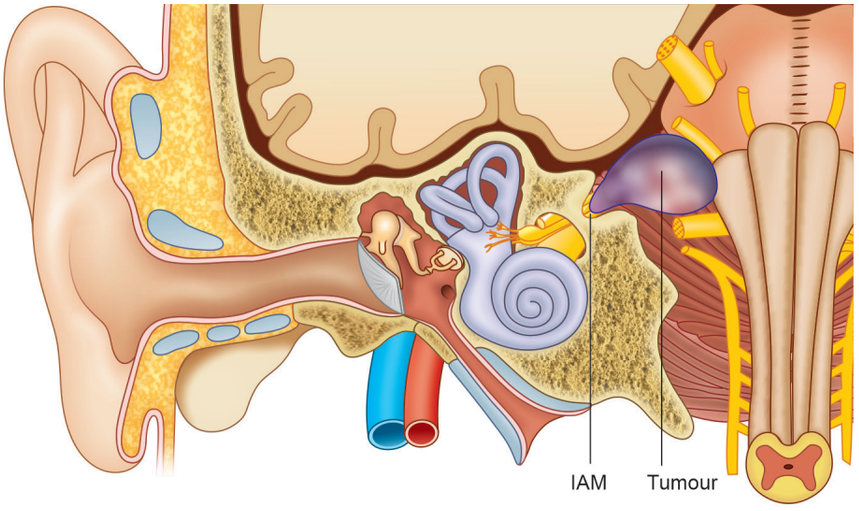


Fig. 23.1: Vestibular schwannoma tumour

*Cerebellopontine angle or cerebellopontine cistern is a three-dimensional triangular space bounded by brainstem medially, petrous part of temporal laterally, cerebellum superiorly and posteriorly and the arachnoid space containing lower cranial nerves IX to XII inferiorly.

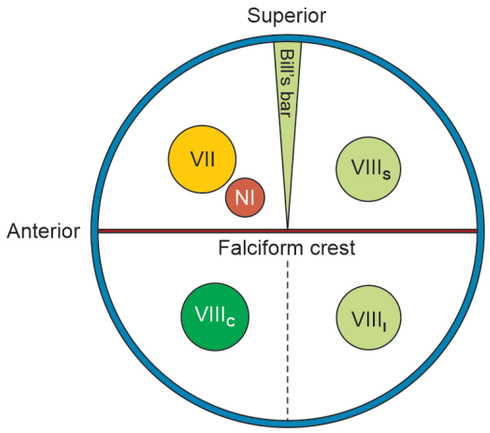


Fig. 23.2: Diagram depicts the cross section at the fundus (lateral end) of the Internal auditory canal. The transverse crest/falciform crest divides the IAM into superior and inferior parts and Bill's bar divides the superior part into anterior and posterior divisions. (VII$_S$: Facial nerve; NI: Nervus intermedius; VIII$_S$: Superior vestibular nerve; VIII$_I$: Inferior vestibular nerve; VIII$_C$: Cochlear division of the 8th nerve)

cerebellopontine angle (CPA). Here it may involve the Vth cranial nerve in the *Meckel's cave* and the lower cranial nerves. Further growth of the tumour results in displacement of the cerebellum and brainstem.

The aetiology of the disease is unknown. Most of the cases are unilateral but bilateral presentation vestibular schwannoma is associated with neurofibromatosis type 2 (NF-2). Age of presentation for the unilateral sporadic cases is between 4th and 6th decade but for vestibular schwannoma associated with NF-2, it is 3rd decade of life.

The classification of the tumours according to the size only (Table 23.2).

Table 23.2: Grading of vestibular schwannoma according to size

Classification	*Grade*	*Size (mm)*
Grade 0	Intrameatal	0
Grade 1	Small	1–10
Grade 2	Medium	11–20
Grade 3	Moderately large	21–30
Grade 4	Large	31–40
Grade 5	Giant	>40

Tumours can be classified according to the involvement of adjacent structures:

Koo's Classification

(Based on site and size)

Grade 1: Small intracanalicular tumour.

Grade 2: Small tumour with protrusion into the cerebellopontine cistern (CPA) but no contact with the brainstem.

Grade 3: Tumour occupying the cerebellopontine cistern with no brainstem displacement.

Grade 4: Large tumour with brainstem and cranial nerve displacement.

Signs and Symptoms

Signs and symptoms appear due to the involvement of cranial nerve VIII, compression of surrounding cranial nerves, cerebellum, brainstem and raised intracranial pressure.

I. VIII nerve involvement

a. High frequency retrocochlear progressive hearing loss is seen as the most common symptom, which occurs due to the involvement of the cochlear nerve. There is also associated decreased speech discrimination. Few patients may have sudden SNHL (12–22%) and about 5% of patients may have normal hearing initially.
b. Tinnitus: This is a common symptom but rarely is the only symptom.
c. Vestibular symptoms are found in 40–60%. Vertigo and disequilibrium are the main vestibular symptoms.

II. Compression of other cranial nerves

a. Facial nerve paresis is a late feature of the disease but due to slow growth of the tumour. The sensory fibers of the nerve may be affected earlier leading to hypoaesthesia of posterior meatal wall **(Hitselberger's sign)**.
b. Trigeminal nerve involvement occurs as the tumour becomes extracanalicular (Stage 2). This produces loss of corneal reflex (*commonly the first sign of extrameatal tumour growth*) and numbness or tingling over the area of distribution of three divisions of the trigeminal nerve.
c. Glossopharyngeal and vagus nerve compression leads to palatal palsy, hoarseness of voice and dysphagia.

III. Cerebellar compression leads to cerebellar signs like ataxic gait, in-coordination of upper limb and dysarthria.

IV. Brainstem compression leads to contralateral lower nerve involvement, pyramidal weakness and nystagmus.

V. *Raised intracranial* pressure may occur at late stages of the disease and this leads to severe decrease in vision, headache and vomiting.

Investigations

1. Schirmer's test can be done to evaluate the compression of nervus intermedius (part of the facial nerve which joins the main trunk of the facial nerve in IAC.
2. Pure tone audiometry shows sensorineural hearing loss especially at high frequencies.
3. Speech discrimination score is decreased.*
   Grading of tumour can be done on the basis of result of PTA and SDS score (Table 23.3).
4. Stapedial reflex will show delayed response.

*AAO-HNS/Gardner Robertson grading
(Based on average hearing loss at 0.5 kHz to 4 kHz and speech discrimination test)

Table 23.3: Grading of tumours on the basis of hearing loss and SDT where A, B, C, D are grades of tumours of increasing size
A: ≤30 dB of hearing loss and ≥70% SDT
B: ≥30 ≤50 dB of hearing loss and ≥50% SDT
C: >50 dB of hearing loss and ≥50% SDT
D: <50% SDT

(SDT: Speech discrimination test—sound given at 40 dB above the speech reception threshold in form of spondee words and patient is asked to repeat the words)

5. Brainstem evoked response audiometry (BERA): Interaural difference in wave I–V delay is greater than 0.2 ms or if the interaural difference in absolute wave V latency is greater than 0.2 ms. The BERA for diagnosis of vestibular schwannoma is limited in small lesions, where it can be normal.
6. Vestibular test: Caloric testing reveals canal paresis in 70% of the cases. Actually, the test is most sensitive for stimulation of the horizontal semicircular canal which is supplied with superior vestibular nerve. Any tumour arising from an inferior vestibular nerve will not show good caloric response.
7. *MRI with gadolinium contrast is now the gold standard for diagnosis of vestibular schwannoma*. All patients with unilateral audiovestibular symptoms are advised MR imaging. The tumour is isointense or hypointense on T1 images and hyper intense on T2 images (Figs 23.3 and 23.4).

Earlier X-ray (perorbital view) was used to compare the size of both the internal acoustic meatus for diagnosis but it is obsolete now. CECT scan can be used with some limitations, if the MRI is not available.

Management

The management strategies for vestibular schwannoma are:

A. Observation and scan
B. Radiosurgery
C. Surgery.

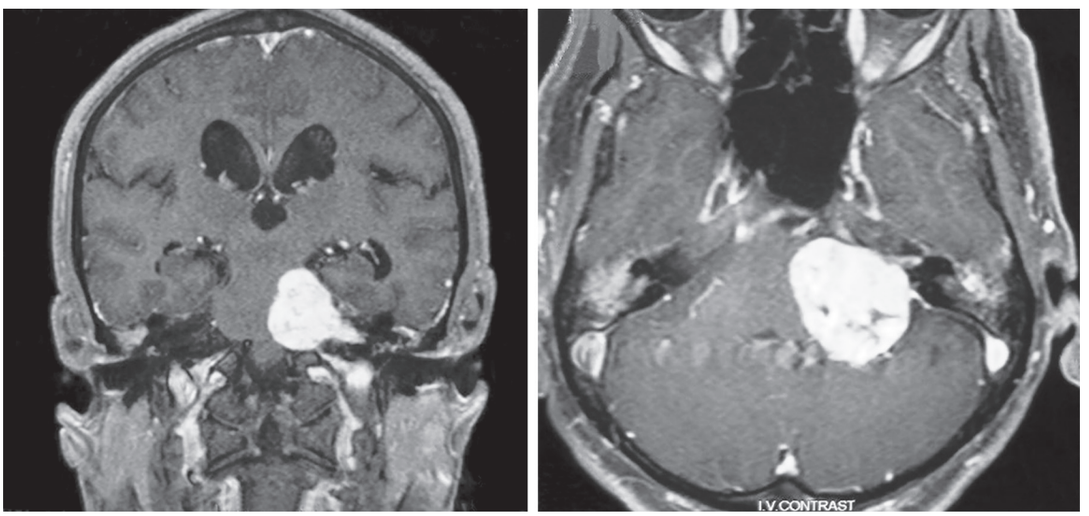


Fig. 23.3: MRI brain showing a vestibular schwannoma

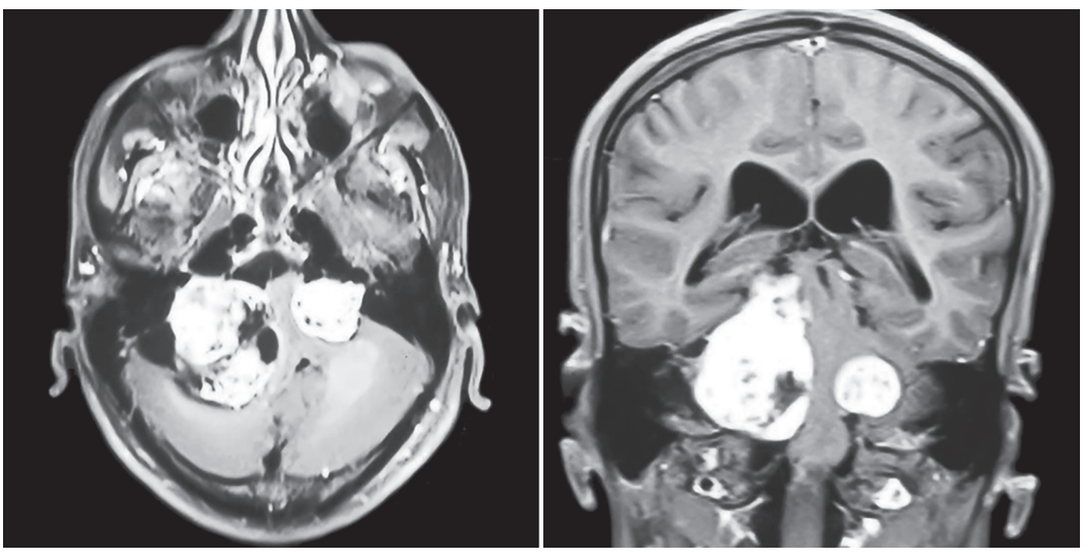


Fig. 23.4: Bilateral vestibular schwannoma associated with neurofibromatosis type 2

A. Observation and scan: It is one of the options for management which requires due consideration because these tumours are very slow growing and a lot of them present at late age of life, therefore their chances of producing severe symptoms are remote. It is the best approach for small and slow growing tumours whose growth rate <2.5–3.0 mm/annum and size is <3 cm. Any sudden increase in size will demand step-up management.

B. Radiosurgery: Suitable candidates for treatment by this modality are those with tumors size less than 2.5 cm in the greatest dimension and are of non-cystic type of vestibular schwannoma. It is recommended for old patients and those with comorbidities.

The radiosurgery modalities which are being used are:

1. Stereotactic radiosurgery (SRS)—single dose radiation.
2. Stereotactic radiotherapy (SRT)—fractionated radiation.

The technique utilizes high precision localization of the tumour and high-resolution collimation beam radiation for a small, targeted lesion.

Two types of machines which are in use:

1. **Gamma knife** (*Elekta*): Head is fixed with four screws through the skull.
2. **Cyber knife** (*Accuray*): The head is stabilized by a facial mask similar to a hockey mask and it uses a linear accelerator to target the radiotherapy beam.

The dose given is 12–13 Gy. Caution: Higher dosages have been associated with SNHL.

C. Surgery: There are four approaches which are mainly used now.

1. *Translabyrinthine Approach*: This approach can tackle all tumour sizes but labyrinthine destruction leads to permanent hearing loss. It is therefore used for patients with preoperative profound hearing loss.
2. *Retrosigmoid Approach*: It can tackle all tumour sizes and preserves hearing. It has drawbacks of postoperative headaches and chances of imbalance due to cerebellar retraction at the time of surgery. This approach can be done using endoscopic assistance and is called Minimally invasive retrosigmoid approach (MIRA).
3. *Middle Cranial Fossa Approach*: It is used for small intracanalicular tumours only when the hearing of the patient is preserved.
4. *Endoscopic Transcanal Transpromontorial Approach (ETTA)*: This is an endoscopic transcanal approach through the cochlea and vestibule to internal acoustic meatus. It is used for grade I and II tumours (Koo's grading system). It is ideal for small tumours with preoperative group D hearing loss.

Chapter

24

Noise-induced Hearing Loss (NIHL) and Noise Pollution

CM3.1: Describe the health hazards of air, water, noise, radiation pollution.
EN2.15: Describe the national programs for prevention of deafness, cancer, noise and environmental pollution.
EN4.15: Describe the clinical features, investigations and principles of management of Noise Induced Hearing Loss.

Noise is any unwanted sudden or chronic excessive sound, which is capable of producing damage to the ear (Fig. 24.1).

EFFECTS OF NOISE

- Auditory (NIHL) effects: It mainly refers to decrease in hearing acuity due to long standing noise exposure leading to gradual progressive sensorineural hearing loss.
- Non auditory effects.

AUDITORY EFFECTS (NIHL)

It is almost always bilateral and symmetrical and does not progress once noise exposure is stopped. The earliest finding on pure tone audiometry is dip in bone conduction at 4 kHz. This typically occurs after 10–15 years of chronic exposure to noise. Asymmetrical hearing loss can occur in military personnel due to head shadow effect from head and shoulder borne weapons.

NIHL may be of two types depending on reversibility of the hearing loss.

Temporary threshold shift: It is the temporary reduction in hearing acuity caused by exposure to loud sound and it is reversible (reverts to normal within 48 hours). The probable hypothesis is that the hair cells' kinocilium get bent due to sudden noise.

Permanent threshold shift: It is the permanent reduction in hearing acuity caused by exposure to loud sound and it is irreversible. It is usually symmetrical loss and associated with tinnitus. Extent of hearing loss in permanent threshold shift is directly proportional to intensity of noise and duration of exposure. It occurs

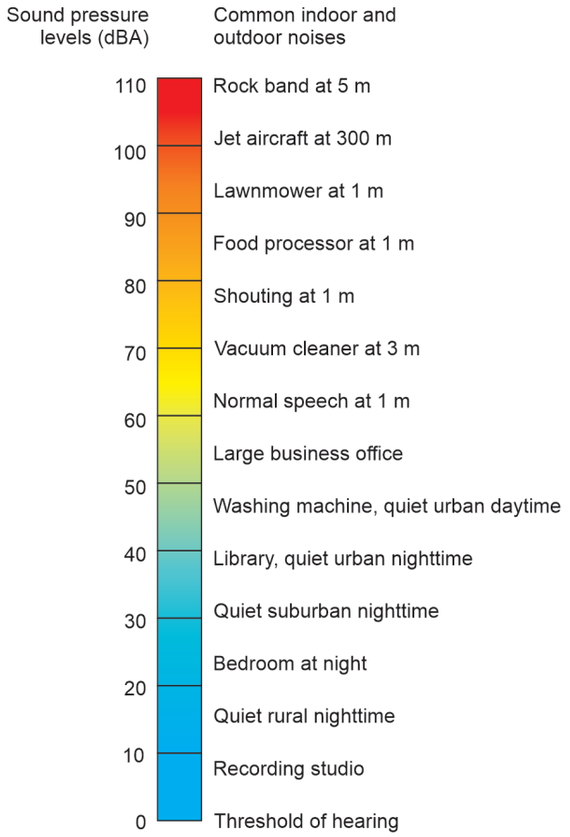


Fig. 24.1: Intensity of sounds in various settings

when the intensity of noise is more than 90 dB and the exposure is more than 8 hours per day for 5 days a week for years. There occurs a permanent micromechanical damage to cochlea resulting in depolarisation of actin filaments. There is also apoptosis of outer hair cells (OHC). The inner hair cells (IHC) and spiral ganglion neurons might show synaptic disconnection (hidden hearing loss).

Acoustic trauma leading to immediate hearing loss occurs when there is a single exposure to an intense sound, for example crackers, bomb blast, explosions, slap injury. It can improve if the insult is minimal, but there can be permanent varying degrees of sensorineural

hearing loss if there is trauma more severe. It is often associated with tinnitus. Aetiology in such a case is mechanical injury to the sensory cells of the cochlea. Conductive hearing loss can also occur following acoustic trauma in cases of tympanic membrane disruption or ossicular dislocation.

Early onset presbycusis has been seen to be due to noise exposure from early childhood. This might have a cumulative effect on hearing impairment in adulthood and can lead to early onset presbycusis. This can be one of the reasons for early onset presbycusis in people living in cities.

Socioacusis or non-occupational NIHL occurs because of loud music like concerts and music shows or headphones, personal entertainment devices, open vehicles such as motorcycles, snowmobiles or ATV and formula one racing cars. On examination there is high frequency SNHL.

Certain **predisposing factors** lead to increased susceptibility to NIHL, as is seen in patients with smoking, diabetes and cardiovascular disease, ototoxic drugs and recreational drug users.

Diagnosis of NIHL is clinical and it requires a history of noticeable noise exposure and presence of hearing loss greater than expected from the age. It is frequently associated with tinnitus. Pure tone audiometry shows significant high tone hearing loss with classical dip at 4000 Hz also known as ***boiler's notch***, seen in boiler makers (Fig. 24.2). In late cases PTA will show involvement of all the frequencies.

Management

A. Prevention is better than cure

For industrial workers

- Medical surveillance of all industrial workers. They must have baseline audiometric data (PTA) before they are posted in a noisy environment.
- The workers should have annual audiological testing by pure tone audiometry.
- The exposure to noise should be regulated, as recommended (Table 24.1).

Table 24.1: Permissible exposure to noise for industrial workers

Exposure time (hours)	*Noise exposure (dB)*
8	90
4	93
2	96
1	99
½	102
1/8	108
1/16	111
1/32 (2 minutes) or less	114

- Training and education programmes for the employees to educate them about the adverse effects of noise and how to avoid NIHL.

  Continuous or intermittent noise exposure above 115 dB is not permitted. Pulse noise of >140 dB should be avoided.
- Use of personal hearing protective devices is recommended for workers. These include:
  i. **Earplugs:** It is available as foam type and premoulded canal caps. These produce 10–15 dB sound attenuation. It is effective only if fitted properly in the canal having complete occlusion of the canal.
  ii. **Ear Muffs:** These reduce the sound by 15 dB and are better than ear muffs in terms of sound attenuation. In industrial areas helmets with protective ear cups can be used for both head protection and noise attenuation (Fig. 24.3).
  iii. **Active noise reduction:** It is an electronic method of sound reduction. It uses electronic devices which provide sound to the ear muff. The provided sound is 180 degrees out of phase to the ambient noise. It is a very effective way to reduce noise particularly in low frequency. This is useful especially in aircrafts where multiple communication devices are used.
  iv. **Rotation** to non-noisy job areas.

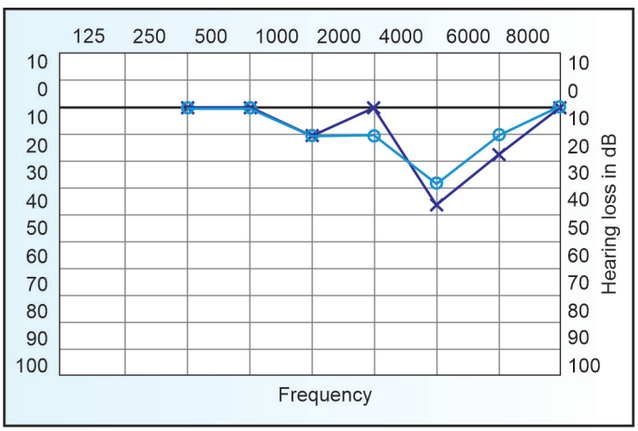


Fig. 24.2: Dip in hearing threshold seen at 4000 Hz in NIHL



Fig. 24.3: Ear plugs and muffs

For personal noise trauma prevention

i. The volume of the music player should not be loud. If you are not able to hear external sounds from surrounding environment, then the volume of the music player is too loud. Ideally the person sitting next to you should not be able to hear your played music.
ii. Good quality noise cancelling muff-type headphones are advisable.
iii. The muff type of headphones is better than ear bud style earphones. This is due to the fact that they have are better background noise cancellation and hence require lower volume to listen.
iv. It is advisable to take regular breaks from using the headphones. The breaks give time for the ear to recover from any insult and prevent the permanent hearing damage.
v. Immediate ENT consultation is recommended If one develops tinnitus or have hearing lose.
vi. *60% rule*: Always ensure that earphone volume is no more than 60% of its maximum and it is used for less than 60 minutes a day with volume not more than 60 dB.

B. Combined treatment with a steroid (prednisolone) and the nootropic drug piracetam have shown some benefit in recovering losses after sudden noise exposure.

C. If the hearing loss is severe and permanent, it can be corrected by use of hearing aids.

D. Tinnitus will require neurophysiological methods like tinnitus retraining therapy, in combination with counselling and sound therapy with hearing aids.

E. In a case of profound hearing cochlear implantation is indicated.

The Noise Pollution (Regulation and Control) Rules, 2000

Legislation to control noise levels in industry, silent zones, use of loudspeakers, noise levels of household equipment and use of horns.

- *Silence zone*-is defined as an area with 100 m radius of the hospitals, educational institutions and courts.
- A *loud speaker or a public address system* shall not be used at night (between 10.00 pm to 6.00 am).
- Manufacture and sale of *crackers* having an impulsive noise of more than 90 dB at 5 meters distance from the site of bursting is not allowed.

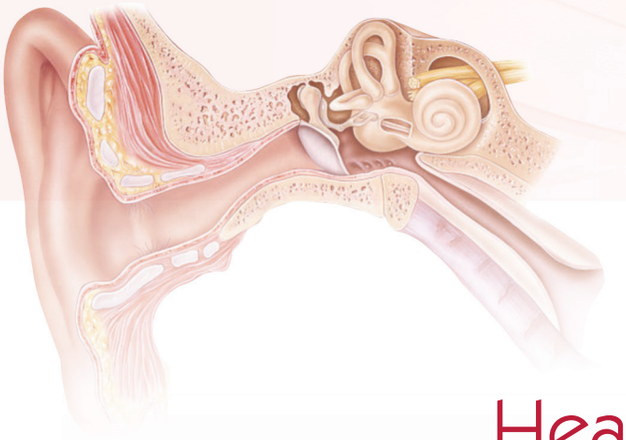
Permissible Level of Noise by Central Pollution Control Board in India

The permissible noise limits for industrial areas, is 75 dB for daytime and 70 dB at night. In residential areas it is 55 dB and 45 dB during daytime and night respectively. For commercial areas limits are 65 dB and 55 dB respectively.

NON-AUDITORY EFFECTS

Due to prolonged noise exposure, there occurs an increase in levels of catecholamines.

1. Noise exposure may increase blood pressure and heart rate
2. Mood disturbances and behavioural changes
3. Poor reaction time
4. Annoyance factor
5. Sleep disturbance
6. Decreased performance at work and memory loss.



Chapter

25

Hearing Aids

Hearing aids are devices which amplify the sounds, so as to assist a hearing impaired individual.

TYPES

A. Air conduction hearing aids
B. Bone conduction hearing aids
C. Implantable hearing aids.

Components: Hearing aids are made of following interconnected units.

1. *Microphone*: It receives acoustic signals and converts them to electricity. Directional microphones are now available, which have two acoustic entry ports. This makes them more sensitive to different directions of sounds.
2. *Amplifier*: The sound signals received by the microphone are made more powerful by the amplifier. It can represent sound in an analogue or digital manner. Filters can be used to change the relative amplitude of the low, mid and high frequency components in a signal.
3. *Receivers*: These are small headphones that use electromagnetism to convert modified electrical signals back into sound. A damper is used in a few hearing aids to absorb energy at the frequencies corresponding to the peaks and improve sound quality and listening comfort.
4. *Battery*: The hearing aid takes electrical power from a battery. These batteries have various physical sizes and capacities, depending on the power needed and space availability in the hearing aid. Latest generation hearing aids have rechargeable batteries.

Hearing aid amplification systems are programmed by a computer using a suitable wired or wireless interface. Now hearing aids are available with bluetooth connectivity which helps to attend mobile phone calls and listen to music, watch TV via a bluetooth device.

AIR CONDUCTION HEARING AIDS

They can be of analog type and digital type. The analog type amplifies all sounds reaching the aid, but the digital type is programmable according to the frequency specific hearing losses of the patient.

These hearing aids are mainly categorised according to the site where they are worn. The site therefore decides the size of the hearing aid. These can be of following types (Fig. 25.1):

- *Body-worn type*: These have a comparatively large microphone, amplifier and battery unit, which can be clipped to a shirt or kept in the pocket. The receiver is placed in the outer external auditory canal.
- *Behind the ear type (BTE)*: These have the microphone, amplifier, battery and receiver unit, placed behind the ear. These are connected to the ear canal mould by a tubing.
- *Receiver-in-canal (RIC)*: These are similar to BTE but the speaker is placed in the canal. This leads to better sound quality and noise reduction.
- *In-the-ear type (ITE)*: All components of the hearing aid are in a single unit, which is placed in the ear. It is cosmetically better than BTE.
- *In-the-canal (ITC)*: Complete unit is placed within the ear canal but can be seen.
- *Completely-in-canal (CIC)*: The complete unit of the hearing aid is within the ear canal and is not visible without close inspection. These are very small in size and are difficult to use in very old patients because of poor dexterity and eye sight.
- *Spectacle type*: The unit of microphone, amplifier, battery and receiver unit, are placed on the spectacle frame behind the pinna.
- *Contralateral routing of signal (CROS)*: In this type, a microphone is placed on the side of the deaf ear to pick sound signals. The sound signals are then sent to the better ear for perception. It is useful only when there is single sided deafness and helps by providing the direction of the sound and also hearing any person or thing which is on the side of deaf ear.

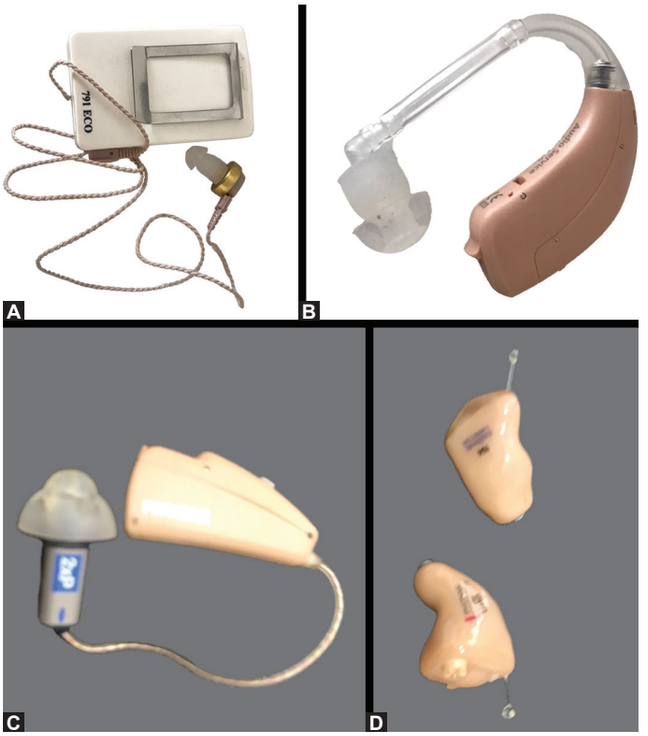


Fig. 25.1: (A) Body worn type; (B) Behind-the-ear (BTE); (C) Receiver-in-canal (RIC); (D) In-the-canal (ITC) and completely in-the canal (CIC)

Use of Bilateral Hearing Aids

Advantages: Wearing bilateral hearing aids increases the range of sound level perception at a much lower intensity. It is recommended for severe hearing losses. They also increase the localisation of sound.

Disadvantages: 1. The hearing aids are costly. 2. These are difficult to manage by elderly. 3. They are more susceptible to wind noise.

BONE-ANCHORED HEARING AIDS

Bone-anchored hearing aids (BAHA) can be used in patients with purely conductive hearing loss, mixed or mild to moderate sensorineural hearing loss (SNHL), and for single-sided deafness (SSD).

Candidacy Criteria for BAHA

1. External and middle ear malformations
2. Chronic otitis media with ossicular chain defects
3. Craniofacial abnormalities
4. Inability to wear hearing aid due to mastoid cavity produced after radical mastoidectomy, extremely narrow ear canal, cul-de-sac ear canal closure after lateral temporal bone resection or extensive skull base surgery.
5. Single-sided deafness (SNHL or mixed hearing loss).

Candidacy Criteria for Bilateral BAHA

A. Congenital deformities in both ears
B. Active discharge in bilateral ears
C. Conductive/mixed hearing loss in both ears.

All patients must receive and show benefit by a **softband** or **headband bone conduction device** for a minimum of 14 days before taking up for surgical implantation of the device.

Bone Conduction Hearing Devices can be of Two Types

A. **Percutaneous BAHA:** This device is in direct contact with the skull bone and connected to the external device going through the skin. Examples of percutaneous BCHDs are BAHA by Cochlear™ and the Ponto bone-anchored hearing system by Oticon®.
B. **Transcutaneous BAHA:** Where the device has the normal skin and soft tissue are interposed between the internal and external component. Examples of transcutaneous devices are Sophono™ and the Cochlear™ Baha® Attract and Vibrant Sound bridge middle ear implant by Medel®.

A. *Percutaneous BAHA*

This has three components:
i. Titanium implant or screw
ii. Percutaneous abutment
iii. Sound processor.

The titanium implant or screw is surgically implanted in the skull bone with percutaneous abutment going through the soft tissue and skin. The titanium implant attaches with the bone by the process called osseointegration and it takes around 3–6 months. This process is similar to osseointegration of orthopaedic implants. The sound processor transmits vibrations to the abutment, which then transmits the vibrations to both the cochleae directly via skull. The main complication with the surgical procedure is failure of osseointegration of abutment (Fig. 25.2).

B. *Transcutaneous Devices*

In a transcutaneous device an internal magnet is fixed to the skull and an external hearing device with an oscillator is used which is held in place by the magnet of the internal device. The external sound processor magnetically delivers auditory stimulation through the closed skin to internal component (Fig. 25.3). The major advantage of this system is that there is no local skin infection, inflammation and failure of osseointegration of the implant.

ASSISTIVE LISTENING DEVICES

1. For patients with profound hearing disability, the assistive listening devices enable the wearer to detect alerting sounds like doorbell, alarm and telephone ring. The sounds are converted into other sensory modalities like flashing lights or vibrating shakers, to alert the users.
2. These can also provide hearing aid users with receiving sounds other than amplified by hearing aid, like distant music or speech.

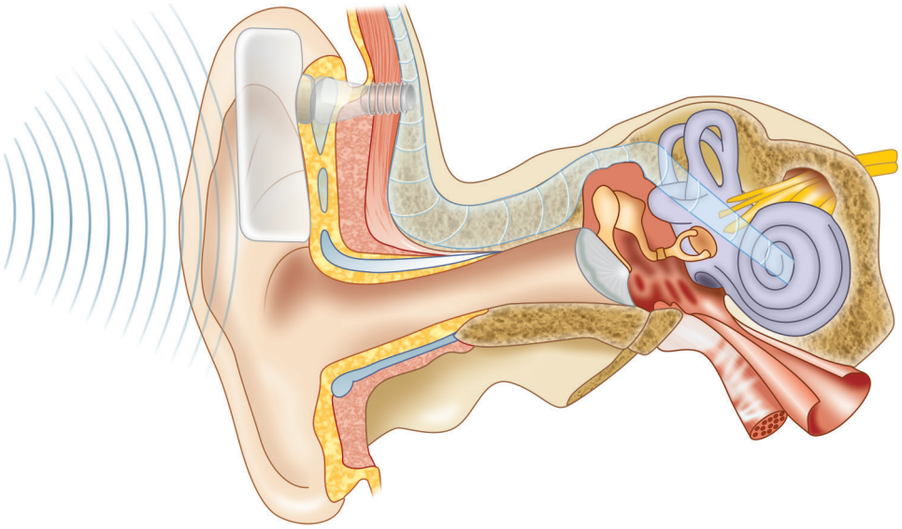


Fig. 25.2A: Functioning of percutaneous BAHA



Fig. 25.2B: Parts of BAHA: Titanium screw, percutaneous abutment, sound processor

Vibrotactile or electrotactile aids: It is used as an alternative for patients with profound hearing loss who are non-responsive to auditory stimulation from hearing aids but who do not want cochlear implant or medical or psychologically unfit. Training is required in integrating tactile information with visual information. These are also beneficial in patients who are deaf and blind.

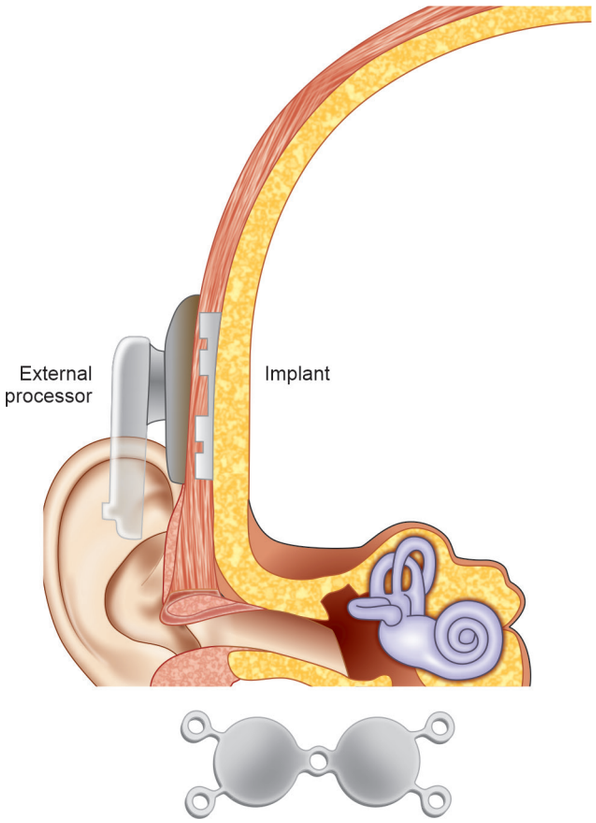


Fig. 25.3: Transcutaneous BAHA

Chapter

26



National Programme for Prevention and Control of Deafness (NPPCD)

EN2.15: Describe the national program for prevention of deafness and noise.

Presently, the hearing loss is the most common type of sensory deficit in man. As per WHO estimates in India, there are approximately 63 million people, who are suffering from significant auditory impairment. As per National Sample Survey Office (NSSO) 2001, currently there are 291 persons per one lakh population who are suffering from severe to profound hearing loss.

According to the latest survey by ICMR the prevalence of hearing loss varies from 9 to 11% with a higher percentage in the rural population. Importantly, large percentage of these are children who are between age of 0 to 14 years. Due to the large number of hearing-impaired young Indians being affected, it certainly leads to severe losses in productivity. The milder degree hearing loss percentages are even higher in our country.

OBJECTIVES OF THE NPPCD[1,2]

1. To prevent avoidable hearing losses due to ear disease or injury.
2. Early identification, diagnosis and treatment of ear problems responsible for hearing loss.
3. To rehabilitate all who are suffering from deafness.
4. To strengthen the existing inter-sectoral linkages for continuity of the rehabilitation programme, for persons with deafness.
5. To develop institutional capacity for ear care services by providing support for equipment, material and training personnel.

Long-term objective: To prevent and control major causes of hearing impairment and deafness, to reduce the total disease burden by 25% of the existing burden by the end of 12th Five Year Plan (2012–2017).

COMPONENTS OF THE PROGRAMME

1. **Manpower training and development:** For prevention, early identification and management of hearing impaired and deafness cases, training would be provided from medical college level specialists (ENT and Audiology) to grass root level workers.
2. **Capacity building:** For the district hospital, community health centres and primary health centre in respect of ENT/Audiology infrastructure.
3. **Service provision:** Early detection and management of hearing and speech impaired cases and rehabilitation, at different levels of the health care delivery system.
4. **Awareness generation through IEC/BCC activities:** For early identification of hearing impaired, especially children so that timely management of such cases is possible and to remove the stigma attached to deafness.

STRATEGIES

- To strengthen the service delivery for ear care.
- To develop human resources for ear care services.
- To promote public awareness through appropriate and effective IEC strategies with special emphasis on prevention of deafness.
- To develop institutional capacity of the district hospitals, community health centres and primary health centres selected under the programme.

PROGRAMME EXECUTION AND EXPANSION

The Programme was initiated in 2007 on pilot mode in 25 districts of 11 State/UTs. At present the program is running in more than 590 districts across the country.

Expected Benefits of the Programme

i. Availability of various services like prevention, early identification, treatment, referral, rehabilitation, etc. for hearing impairment and deafness as the primary health center/community health centres/district hospitals largely cater to their need.
ii. Decrease in the magnitude of hearing-impaired persons.
iii. Decrease in the severity/extent of ear morbidity or hearing impairment.

iv. Improved service network/referral system for the persons with ear morbidity/hearing impairment.
v. Awareness creation among the health workers/ grassroot level workers through the primary health centre medical officers and district health officers, which will percolate to the lower-level health workers functioning within the community.
vi. Capacity building at the district hospitals to ensure better care.

Central Coordination Committee (CCC)

A central coordination committee will be constituted at the central level with the following members to provide technical inputs and facilitate implementation of the programme.

- Representatives of Directorate General of Health Services/Ministry of Health and Family Welfare
- Representative of WHO
- ENT specialists/experts
- Audiologists and Speech therapists
- Public Health expert
- Representative of Rehabilitation Council of India (RCI).

I. Central Level: Central Cell-A support to the Programme Division in the Ministry of Health and Family Welfare will be provided by a team with necessary technical support to the States/District level functionaries in successfully implementing the programme. The team will put in place an effective Management Information System (MIS) for collection of data for effective supervision and monitoring of the programme.

II. State level: State Cell-To strengthen Monitoring of the programme at State level. Already constituted state mission under National Rural Health Mission (NRHM) will look into the activities at state level. Under the programme, funds will be transferred to the State Health Society, for carrying out various activities through the District Health Society.

Functions

- Preparation of District plans for implementation of National Programme for prevention and control of Deafness.
- Monitor and supervise Implementation of National Programme for Prevention and Control of Deafness in the state in reference to the programme activities related to Training of Medical and non-medical professionals, IEC and Social mobilization and Manpower development.
- Release and monitor flow of funds to District Health Societies.
- Review and take appropriate measures in the expenditure of funds by District Health Society.

State Nodal Officer-State Nodal Officer preferably an ENT surgeon at the directorate/Secretariat level will provide technical guidance and expertise to the State Health Society for the purpose of implementation of the programme in the various districts of the state.

III. District level: District Health Society and Programme Committee (under NRHM) Functions

- Planning: Preparation of District Micro-plan based on magnitude and distribution of deaf/hearing impaired persons and resources available for ear care.
- Implementation of the programme through utilization of government facilities, involvement of NGOs and community participation.
- Monitoring of programme: Activities and quality control.
- Financial and Material Management
- Social mobilization and public awareness
- Orientation of various functionaries of health and other related sectors.
- Procurement of equipment and other materials.
- Arrangements for screening camps-through identified NGOs, having adequate infrastructure for carrying out activities under the programme.
- Monitoring and Financial Assistance for organizing camps.

District hospital: It is proposed to strengthen the ear care services at district level by providing manpower support such as one ENT Surgeon, one Audiologist, one Audiometric Assistant and one Instructor for the hearing impaired at each district on contractual basis.

SERVICE DELIVERY AND REFERRAL SYSTEM

I. Primary level: CHCs/PHCs/SCs/Primary School teachers/Health Workers/Panchayat Functions:

- Early identification of cases of hearing impairment and their management in collaboration with NRHM (RBSK-Rashtriya Bal Swasthya Karyakram).
- Primary ear care.
- Promoting public awareness in respect of prevention of deafness.
- Sensitization training of health workers.
- Support for the School Ear Care Programme.

II. Secondary level: District Hospital Functions:

- Management of cases referred from PHCs/CHCs.
- Organization of Ear care screening camps in collaboration with NRHM and Ministry of Social Justice and Empowerment.
- Organization of School Ear Care Programme.
- Training of manpower—PHC doctors, nurses, audiometric assistants, health workers, and school teachers.

III. Tertiary Level: State Medical College In each state, one medical college has been identified to act as the Centre of Excellence and referral centre for the districts covered under the programme in that state. Two ENT surgeons and one Audiologist from the medical college will be trained under the programme.

Functions

- Training of manpower, i.e. sensitization programmes and surgical training workshops.

- Management of referral cases, especially the difficult cases for diagnosis and management.

PROGRAMME COMPONENTS

Manpower Training and Development

Objectives

1. To orient all the healthcare personnel from the district to grassroot level about prevention, promotion, early identification and rehabilitation of all types of ear diseases leading to deafness.
2. To make these personnel aware of the existing facilities available for deafness management in order to facilitate appropriate referral.
3. To sensitize the health care personnel regarding their specific roles in the programme.
4. To enable the health providers to provide a leadership role in creating awareness about hearing impairment.
5. To facilitate development of suitable manpower, in order to be able to implement this programme in the entire nation, in a phased manner.

Training would be imparted to the following functionaries at all levels:

A. Sensitization of the State medical college experts
B. District level ENT officers
C. Sensitization of Audiologists
D. Medical Officers under CHC/PHC
E. Obstetricians and paediatricians at the district and CHC level
F. Medical Officer under School Health Scheme
G. Medical Officers involved in industrial health available in the district
H. Multi-purpose Workers (MPW) male and female
I. Primary School teachers at village level functionaries at grass root level such as Anganwadi workers (AWW), accredited social health activists (ASHA) and trained birth attendants.

Service Provision Including Rehabilitation in collaboration with NRHM and Ministry of Social Justice and Empowerment.

1. *Early detection*

- The detection would be by sensitized personnel at grass root level including family members/parents, selected school teachers, MPWs at sub centre level, Public Health Nurses and medical officers in PHCs and CHCs and district level.
- House to house surveys will be conducted by the AWWs and ASHAs, under the supervision of the male and female MPWs for detection of cases of hearing impairment and deafness. The deafness cases will be noted in the disability column of ANM's village register.
- The MPWs will maintain records of each family based on a Family Performa provided to them.
- The district level Paediatricians and Gynaecologists will be responsible for referring any child born of a high-risk pregnancy or delivery. These children will be screened by the district level ENT doctor/audiologist with OAE and then subjected to diagnostic tests.
- School teachers will undertake to screen the children in the school with the help of pre-prepared proformas. These will help to identify children with any ear or hearing problem. They will then be referred to the School Health doctor.

2. *Ear screening camps*

Functions

- Screening camps will be organized in collaboration with NRHM (RBSK)/Ministry of Social Justice and Empowerment at the PHC/CHC and District level for screening the general population in respect of ear problems, hearing impairment and deafness.
- Detection and treatment of common ear problems.
- Spreading awareness regarding ear problems and services provided by health facilities.
- Education of the community, especially the parents of young children regarding the importance of right feeding practices and various common ear problems.
- Education of Panchayat members, members of Mahila Mandals and Youth leaders.

Conduct of screening camps

Ear screening camps will be conducted by the PHC/CHC doctors and district level ENT specialists, trained under the programme.

- The screening camps will be facilitated in collaboration with NRHM (RBSK) or by the NGOs, identified by the Ministry of Social Justice and Empowerment/ District Health Society. These NGOs will require adequate infrastructure to carry out screening camps and experience of work at the community level.
- One screening camp will be organized per month at any PHC or CHC or District hospital by rotation.

3. *Treatment:* Treatment of all affected persons would be undertaken at the following levels:

- Public Health Nurses and MPWs: Would provide treatment of common ear ailments such as Wax, Acute Suppurative Otitis Media etc. under the guidance of the PHC doctor. The Public Health Nurses and MPWs will have the capacity to distribute relevant ear drops and medicines under the guidance of the PHC doctor.
- Trained PHC/CHC doctors will provide early diagnosis of ear diseases and treatment of all common ear ailments. All persons requiring special diagnostic facilities, complicated cases and those needing surgical intervention will be referred to the District hospital.
- District hospital: The District level ENT doctors and Audiologists will provide comprehensive preventive, promotive and curative and medical rehabilitative services.
- The District level Paediatricians will also be responsible for treating ear diseases such as Acute Otitis media, so that progress to Deafness can be prevented.

4. *Referral services*: Effective linkages would be developed from peripheral level to district level with the help of functionaries and personnel from grass root level (AWW, ASHA and sensitized parents and PRIs), sub centre level (Male and female MPWs), PHC level medical officers, Public health nurses, School teachers and School health doctors, ENT private practitioners and District level officers.

5. *Rehabilitation and Hearing Aid provision*

- All patients who are identified as having an ear problem that either requires surgery, hearing aid fitting or rehabilitative therapy will be referred to the ENT doctor and Audiologist at the district level.
- Those who need surgery will be given the appropriate treatment at the district hospital.
- Complicated cases that cannot be adequately handled at the district hospital will be further referred to the State Medical College for expert treatment.
- Patients who suffer with sensorineural hearing loss that is not amenable to medical or surgical correction and which requires hearing aid, will be fitted with the same at the district level which will be provided by the Ministry of Social Justice and Empowerment. The hearing aids will be issued as per existing rules.
- The requirement for Speech therapy and Hearing therapy will be met by the Audiologist at the District level.
- Budget-Under NPPCD, no budgetary allocation has been kept separately for provision of hearing aids.

IEC ACTIVITY

- Community level health workers and doctors will undertake this activity on a continuous basis. This will also form a part of the IEC activities at various levels.
- Sensitization will be done regarding various aspects relating to early detection of hearing loss and if untreated, its ill-effects on the speech, mental and social heath.
- Information regarding various treatment modalities as well as techniques of rehabilitation.
- Sensitization to ill effects of hearing loss in the elderly so that they may be referred for suitable management/ rehabilitation.
- Budget: Provision of ₹ 20.00 lakhs at State level and in addition ₹ 2.00 lakhs at each district level has been kept for IEC activities under NPPCD.

CAPACITY BUILDING

1. At the PHC and CHC level: For screening of ear morbidity and detection of hearing loss, following equipment will be provided
   a. PHC Kit which will consist: (a) Head Lights (b) Ear Speculum (c) Ear Syringes (d) Otoscope (e) Jobson Horne Probes (f) Tuning forks of 256, 512, 1024 Hz (g) Noise Maker (h) Pure tone audiometer (only for CHC).
   b. Budget: Provision of ₹ 20,000/- per kit for PHC and ₹ 50,000 per kit for CHC has been kept.
   a. District hospital: The District hospital is to be an important centre for the management of ear problems and deafness cases, which are referred from the health care facilities at various levels. They should be provided with operating microscope, micro drill system, micro-surgery instruments, otoscope, indigenous digital audiometer, impedance audiometer and OAE analyser.

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