Dr. Rahul Bagla ENT Textbook
Ultimate Guide – Makes You Best

Types of Hearing Loss

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Hearing is one of our most important senses, which connects us to the world and facilitates communication. **Hearing loss** is defined as an impairment of auditory function, which can significantly impact an individual's quality of life. This impairment can range from mild to profound, or even result in total loss of hearing, often referred to as deafness. The term **"deafness"** refers to a condition characterised by little to no hearing ability.

Understanding the various types of hearing loss, their underlying mechanisms, and appropriate management strategies is fundamental for every **medical student** and future **ENT specialist**. Therefore, this chapter provides a clear, comprehensive, and exam-oriented overview, ensuring you master this critical topic.

Classification of Hearing Loss

The **World Health Organisation** (WHO) established a grading system for hearing loss in 1980. This system is based on pure tone audiometry, which measures the average hearing threshold at frequencies of 500, 1000, and 2000 Hertz in the better ear. The following table summarises the WHO grading of hearing loss:

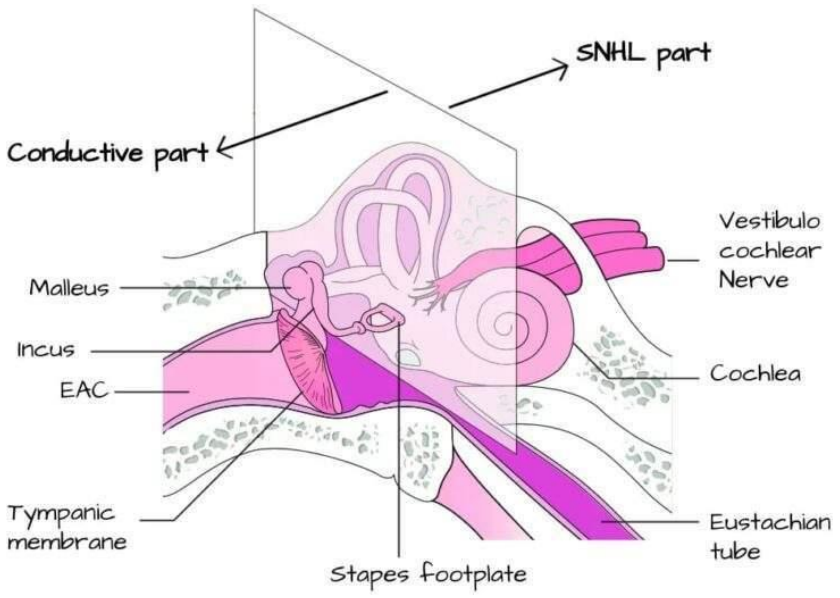
Table: WHO grading of degree of hearing loss

Degree of hearing loss	Hearing threshold in better ear (average of 500, 1000 and 2000 Hz)
Not significant	0-25 dB
Mild	26-30 dB
Moderate	41-55 dB
Moderately severe	56-70 dB
Severe	71-91 dB
Profound	Above 91 dB
Total	

Assessment of Hearing Loss

In evaluating auditory function, it is essential to determine several factors:

1. **Type of Hearing Loss**: Depending upon the anatomical location affected, hearing loss can be classified as conductive, sensorineural, or mixed.
2. **Degree of Hearing Loss**: This can range from mild to total loss, categorised as mild, moderate, moderately severe, severe, profound, or total.
3. **Site of the Lesion**: In conductive hearing loss, the lesion may occur in the external ear, tympanic membrane, middle ear, ossicles, or eustachian tube. Clinical examination and **tympanometry** are useful in identifying these lesions. In sensorineural hearing loss, it is important to ascertain whether the lesion is **cochlear** (sensory), **retrocochlear** (neural, affecting the 8th nerve), or **central** (affecting higher auditory pathways). Special hearing tests, such as auditory brainstem response (ABR) or otoacoustic emissions (OAE), may be necessary for accurate differentiation.
4. **Cause of Hearing Loss**: Potential causes include congenital factors, trauma, infections, neoplasms, degenerative conditions, metabolic disorders, ototoxicity, vascular issues, or autoimmune processes. A comprehensive patient history and laboratory investigations are crucial for identifying the underlying cause.



Types of Hearing Loss

Hearing loss can be categorised into several distinct types, each characterised by different underlying mechanisms and causes:

1. **Conductive hearing loss**

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2. **Sensorineural hearing loss**
3. **Mixed hearing loss**
4. **Non-organic/ Functional hearing loss**
5. **Central hearing loss**

1. Conductive Hearing Loss (CHL)

Conductive hearing loss arises from obstructions in the transmission of sound waves along the auditory pathway, which extends from the external ear to the cochlea, specifically at the stapediovestibular joint.

Common causes of conductive hearing loss include: **(Mnemonic: O-T-M-E-O)**

- **O**bstruction in **External Ear Canal**:
  - Impacted **cerumen (earwax)** is the most common cause.
  - Foreign bodies.
  - Osteomas or exostoses (bony growths in the canal).
  - Keratotic masses (e.g., cholesteatoma of EAC).
  - Benign or malignant tumours of the EAC.
  - Meatal atresia (congenital narrowing or absence of the canal).
- **T**ympanic Membrane (TM) Pathology:
  - **Perforations** (due to trauma, infection).
  - Tympanosclerosis (scarring/calcification of TM).
  - Retraction pockets.
- **M**iddle Ear Disorders:
  - Presence of fluid: **Otitis Media with Effusion (OME)**, also known as "glue ear."
  - Acute otitis media (AOM) or Chronic Suppurative Otitis Media (CSOM).
  - Hemotympanum (blood in the middle ear, often post-traumatic).
- **O**ssicular Chain Pathology:
  - **Ossicular fixation:** Most commonly **otosclerosis** (fixation of stapes footplate). Also, malleus head fixation, fibrous ankylosis.
  - **Ossicular disruption/discontinuity:** Due to trauma (e.g., temporal bone fracture), cholesteatoma erosion, or chronic infection.
- **E**ustachian Tube Dysfunction:
  - **Obstructions** affecting pressure equalisation in the middle ear, leading to fluid accumulation (OME).
  - Barotrauma.

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Table: Congenital and Acquired causes of conductive hearing loss

Table: Causes of conductive hearing loss	
Congenital causes	**Acquired causes**
• Meatal atresia • Fixation of stapes footplate • Fixation of malleus head • Ossicular discontinuity • Congenital cholesteatoma • Aplasia of the oval or round window	• **External ear:** Any obstruction in the ear canal, e.g. wax, stenosis, benign or malignant tumour, foreign body, furuncle. • **Middle ear:** - Tympanic membrane perforation. - Fluid in the middle ear (acute otitis media, serous otitis media or hemotympanum). - Mass in middle ear, e.g. benign or malignant tumour - Ear ossicles, e.g. disruption, erosion and fixation of ear ossicles (otosclerosis, trauma). - Eustachian tube obstruction as in retracted tympanic membrane.

Table: Characteristics of Conductive Hearing Loss

Feature	Conductive Hearing Loss Characteristics
Rinne Test	Negative (BC > AC in affected ear). This is a hallmark.
Weber Test	Lateralizes to the poorer (affected) ear. The sound is perceived louder in the ear with the conductive loss.
Absolute Bone Conduction	Normal (or near normal) levels. The cochlea and nerve are intact.
Frequency Impact	Greater impact on low frequencies. (However, large perforations can affect all frequencies).
Degree of Loss	Typically does not exceed 60 dB HL. This is because bone conduction is intact.
Speech Discrimination	Remains good. Patients hear speech but find it difficult to hear quiet sounds. They often speak softly themselves.
Hearing Aid Benefit	Excellent candidates for hearing aid amplification as the inner ear can effectively process amplified sound.

Characteristics of Conductive Hearing Loss on Pure Tone Audiometry (PTA)

- A difference of more than 10 dB between AC and BC thresholds at any frequency.
- BC thresholds are generally at normal hearing levels (equal to or better than 20 dB HL) in cases of pure conductive hearing loss.
- If BC thresholds exceed 20 dB HL with a significant air-bone gap (ABG), this condition is classified as mixed hearing loss.

Management of Conductive Hearing Loss

Management strategies for conductive hearing loss include:

1. **Removal of Canal Obstructions:**
   - Impacting cerumen or foreign bodies: Removal through irrigation or suction clearance.
   - Osteomas/Exostoses: Surgical excision.
   - Meatal atresia: Canaloplasty (surgical reconstruction of the EAC).
   - Benign or malignant tumours: Surgery
2. **Medical Management:**
   - Otitis Media with Effusion (OME): Decongestants, antihistamines, and watchful waiting.
   - Acute Otitis Media: Antibiotics.
3. **Surgical Interventions:**
   - **Myringotomy with or without grommet insertion:** To drain fluid from the middle ear (e.g., in OME).
   - **Myringoplasty:** Surgical repair of the tympanic membrane perforation.
   - **Tympanoplasty:** A broader operation to (i) eradicate disease from the middle ear and (ii) reconstruct the hearing mechanism, potentially with or without mastoid surgery and with or without tympanic membrane grafting.
   - **Ossiculoplasty:** Surgical reconstruction of the ossicular chain (e.g., using prostheses or autologous ossicles).
   - **Stapedectomy/Stapedotomy:** For otosclerosis, replacing the fixed stapes with a prosthesis.
4. **Hearing Aids:**
   - These devices are utilised when surgical options are not feasible, declined by the patient, or have previously failed.

2. Sensorineural Hearing Loss (SNHL)

Sensorineural hearing loss is the result of damage to the cochlea **(sensory type)** or the eighth cranial nerve and its central auditory pathways **(neural type)**. This damage disrupts the process of **cochlear transduction**, where mechanical energy is converted into electrical impulses for transmission to the brain.

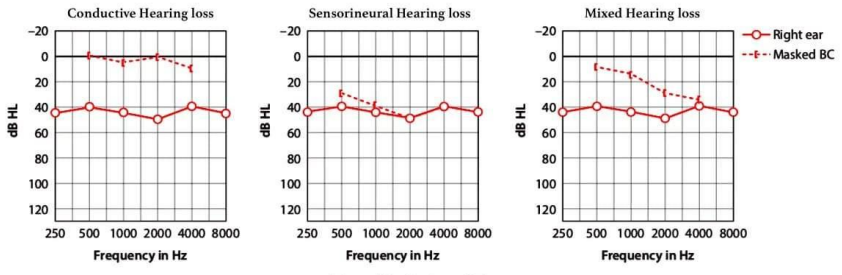
- **Sensory SNHL (Cochlear):** It results from damage to the hair cells or other structures within the cochlea. This is the most common type of SNHL.
- **Neural SNHL (Retrocochlear):** It is caused by lesions affecting the eighth cranial nerve (vestibulocochlear nerve).
- **Central Deafness:** It results from lesions in the central auditory pathways. (Sometimes classified separately or as a subtype of SNHL).

Characteristics of Sensorineural Hearing Loss:

Feature	Sensorineural Hearing Loss Characteristics
Rinne Test	Positive (AC > BC), but both are reduced. The sound is heard better by air, but softly, in the affected ear.
Weber Test	Lateralizes to the better (unaffected) ear. Sound is perceived louder in the ear with better cochlear function.
Bone Conduction	Reduced on Schwabach and absolute bone conduction tests. This reflects inner ear or neural damage.
Speech Discrimination	Poor speech discrimination. Patients often report, "I can hear people talking, but I can't understand what they're saying."
Hearing in Noise	Significant difficulty in hearing in the presence of background noise.
Recruitment Phenomenon	May be present (abnormal loudness growth). Loud sounds become disproportionately louder, often painful. Typical of cochlear SNHL.
Tinnitus	Frequently accompanied by tinnitus (ringing or buzzing in the ears).
Degree of Loss	Can range from minimal to profound or total loss, and is usually permanent.

The characteristics of sensorineural hearing loss on PTA:

1. AC and BC thresholds being within 10dB of each other.
2. High frequencies are mostly involved.
3. Loss may exceed 60 dB, generally complete hearing loss.



Types of Hearing loss on PTA.
CHL - BC thresholds are normal, increased AC thresholds with significant AB gap.
SNHL - Both AC, BC thresholds increased with no significant AB gap.
Mixed HL - Both AC, BC thresholds increased with significant AB gap.

Aetiology of Sensorineural Hearing Loss:

SNHL can be broadly categorised into congenital (present at birth) and acquired (developing later in life) causes.

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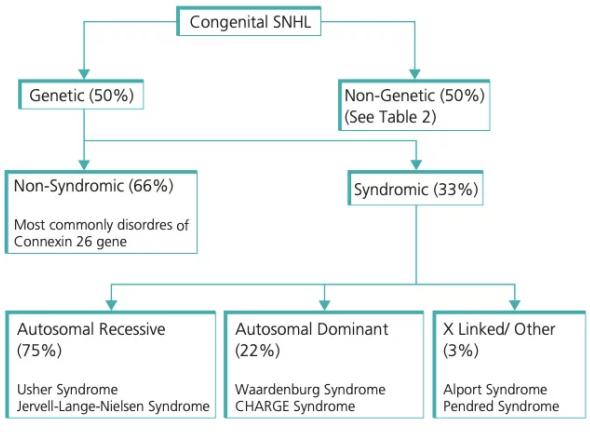
A. Congenital SNHL:

Congenital SNHL can result from inner ear anomalies (e.g., Mondini dysplasia) or prenatal/perinatal damage to the hearing apparatus.



INNER EAR ANOMALIES GENETICS

Syndromes Associated with Congenital SNHL

Syndrome	Associated Features	Hearing Loss Characteristics
Waardenburg Syndrome	Pigmentation abnormalities (white forelock, heterochromia iridis), wide-set eyes, synophrys.	Unilateral or bilateral, progressive SNHL.
Usher Syndrome	Progressive retinitis pigmentosa (leading to blindness) and vestibular dysfunction.	Bilateral SNHL (often severe to profound).
Pendred Syndrome	Goiter (thyroid dysfunction), inner ear malformations (Mondini dysplasia).	Bilateral, profound SNHL.
Alport Syndrome	Nephritis (kidney disease), ocular abnormalities (anterior lenticonus).	Progressive SNHL.
Jervell and Lange-Nielsen Syndrome	Prolonged QT interval on ECG (risk of sudden cardiac death due to arrhythmias).	Bilateral, profound SNHL.
Branchio-Oto-Renal (BOR) Syndrome	Branchial cleft cysts/fistulae, ear anomalies (preauricular pits, malformed pinna), renal anomalies.	Variable SNHL (conductive, SNHL, or mixed).

B. Acquired SNHL:

Acquired SNHL develops after birth. It can have genetic (non-syndromic or syndromic) or non-genetic causes.

Common Causes of Acquired SNHL (CBME Focus):

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1. **Infections of Labyrinth:**
   - **Viral:** Mumps, Measles, Rubella (congenital), Cytomegalovirus (CMV, congenital or acquired), Herpes Zoster Oticus (Ramsay Hunt syndrome – often unilateral, with facial paralysis and vesicles).
   - **Bacterial:** Labyrinthitis as a complication of otitis media or meningitis.
   - **Spirochaetal:** Syphilis, Lyme disease.
2. **Noise-Induced Hearing Loss (NIHL):**
   - Chronic exposure to loud noise (e.g., occupational, recreational). Typically presents as a **notch at 4000 Hz** on audiometry.
   - Acoustic trauma (single loud blast).
3. **Ototoxic Drugs:**
   - **Aminoglycosides** (e.g., gentamicin, amikacin) – affect high frequencies initially.
   - **Loop diuretics** (e.g., Furosemide) – usually reversible.
   - **Salicylates** (e.g., aspirin in high doses) – usually reversible.
   - **Chemotherapeutic agents** (e.g., cisplatin, carboplatin).
   - **Antimalarials** (e.g., quinine, chloroquine).
4. **Presbycusis:** Age-related hearing loss. The most common cause of SNHL in adults. Typically bilateral, symmetrical, and affecting high frequencies first.
5. **Sudden Sensorineural Hearing Loss (SSNHL):** Acute onset SNHL (≥30 dB loss over ≥3 contiguous frequencies within 72 hours). Often idiopathic. Requires urgent evaluation.
6. **Meniere's Disease:** Characterised by episodic vertigo, fluctuating SNHL (typically low-frequency), tinnitus, and aural fullness. Due to endolymphatic hydrops.
7. **Acoustic Neuroma (Vestibular Schwannoma):** Benign tumour of the vestibular division of the 8th cranial nerve. Typically causes **unilateral, progressive SNHL**, often with tinnitus and disequilibrium. **Asymmetrical SNHL mandates an MRI brain with IAC protocol.**
8. **Trauma to Labyrinth or VIIIth Nerve:**
   - Temporal bone fractures (especially trans-labyrinthine fractures).
   - Barotrauma (e.g., diving injuries leading to perilymph fistula).
9. **Familial Progressive SNHL:** Genetically inherited SNHL that develops and progresses over time.
10. **Systemic Disorders:**

- **Metabolic:** Diabetes Mellitus, Hypothyroidism, Kidney disease.
- **Autoimmune:** Systemic lupus erythematosus (SLE), Rheumatoid arthritis, Cogan's syndrome.
- **Vascular:** Vasculitis, stroke.
- **Neurological:** Multiple Sclerosis.
- **Haematological:** Sickle cell anaemia, polycythemia.

Table: Causes of Bilateral vs. Unilateral SNHL

Causes of Bilateral SNHL	Causes of Unilateral SNHL
Presbycusis	Chronic Suppurative Otitis Media (CSOM) – if labyrinth involved
Noise-induced hearing loss	Mumps (can be unilateral SNHL)
Ototoxic drugs	Herpes Zoster Oticus
Systemic disorders (e.g., diabetes, autoimmune)	Acoustic Neuroma (Vestibular Schwannoma)
Familial progressive SNHL	Sudden Sensorineural Hearing Loss (often unilateral)
Congenital SNHL (syndromic/non-syndromic)	Meniere's Disease (initially unilateral, can become bilateral)
Bilateral temporal bone fractures	Unilateral temporal bone fracture

Management of Sensorineural Hearing Loss:

For most SNHL, medical or surgical interventions are very limited, as the damage to the cochlea or nerve is often irreversible.

1. **Hearing Aids:** The primary mode of rehabilitation for most SNHL. These amplify sound to compensate for the reduced sensitivity.
2. **Cochlear Implants:** For severe-to-profound SNHL where hearing aids provide insufficient benefit. These surgically implanted devices bypass the damaged cochlea and directly stimulate the auditory nerve.
3. **Auditory Brainstem Implants (ABIs):** For SNHL caused by damage to the auditory nerve (e.g., bilateral acoustic neuromas due to Neurofibromatosis Type 2), bypassing the nerve and directly stimulating the brainstem.

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4. **Medical Management (for specific causes):**

- **Sudden SNHL:** Oral corticosteroids (often with intratympanic steroids).
- **Meniere's Disease:** Dietary modifications (low salt), diuretics, intratympanic steroids/gentamicin, sacculotomy, endolymphatic sac decompression.
- **Autoimmune inner ear disease:** Immunosuppressants (e.g., steroids).
- **Infections:** Appropriate antimicrobials.
- **Acoustic Neuroma:** Observation, Stereotactic Radiosurgery, or Surgical Excision.

5. **Aural Rehabilitation:** Speech therapy, auditory training, communication strategies.

3. Mixed Hearing Loss. **Mixed hearing loss** is diagnosed when both conductive and sensorineural components coexist in the same ear. This implies a problem in sound conduction to the inner ear *and* a problem within the inner ear or auditory nerve itself.

Identification on Audiometry: Mixed hearing loss is characterised by:

- The presence of an **air-bone gap (ABG > 15-20 dB)**, indicating a conductive component.
- **Impaired bone conduction thresholds (BC > 20 dB HL)**, indicating a sensorineural component.
- *Therefore, in mixed hearing loss, both AC and BC thresholds are elevated, but AC thresholds are significantly poorer than BC thresholds, creating the ABG.*

Common Causes of Mixed Hearing Loss: This type often arises from conditions that can affect both conductive and sensorineural mechanisms.

- **Otosclerosis with Cochlear Involvement:** While primarily conductive (stapes fixation), prolonged otosclerosis can sometimes lead to secondary cochlear damage (cochlear otosclerosis).
- **Chronic Suppurative Otitis Media (CSOM) with Labyrinthine Fistula/Involvement:** Chronic middle ear infection can erode ossicles (conductive component) and, if it extends to the labyrinth (inner ear), cause sensorineural damage.
- **Temporal Bone Fracture:** Can cause ossicular disruption (conductive) and inner ear/nerve damage (sensorineural).
- **Head Trauma:** Similar to temporal bone fractures.

- **Ageing with Earwax Impaction:** Presbycusis (SNHL) coexisting with cerumen impaction (CHL).

Management of Mixed Hearing Loss: Management involves addressing both components where possible.

- **Prioritise Conductive Component:** Often, addressing the conductive component first (e.g., surgery for CSOM, stapedectomy for otosclerosis) can significantly improve hearing.
- **Address Sensorineural Component:** Hearing aids or cochlear implants may be necessary if the SNHL component is significant.

4. Non-Organic / Functional Hearing Loss. Functional hearing loss, also known as **non-organic hearing loss**, describes a situation where an individual exaggerates or feigns hearing impairment without a corresponding organic pathology that fully explains the observed hearing thresholds. Various terms have describe this phenomenon, including **pseudo-hyperacusis, malingering** (conscious feigning for gain), and **factitious hearing loss** (unconscious feigning). However, it is crucial to recognise that in many instances, individuals may indeed possess an underlying organic hearing loss that is then intentionally or unintentionally exaggerated. In other cases, particularly following trauma or for compensation claims, the individual may entirely feign hearing loss. Given the potential presence of an organic component, functional hearing loss is best understood as an exaggerated hearing loss or a functional overlay to an existing organic loss.

Clinical Presentations

Patients may present in any of the following three scenarios:

1. **Total hearing loss in both ears**
2. **Total hearing loss in one ear**
3. **Exaggerated hearing loss in one or both ears**

1. High Index of Suspicion: Watch for **exaggerated behaviours**, such as frequently requesting repetition of questions, cupping the ear excessively and inconsistent complaints with clinical findings. These actions raise red flags for **functional overlay**.

2. Inconsistent Audiometry Results: In **pure tone and speech audiometry**, normal variation on repeat testing is **±5 dB**. A variation **greater than 15 dB** strongly suggests NOHL. **Audiometric Zig-zag" or "Saucer" Pattern** is sometimes seen on PTA.

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3. Absence of Shadow Curve in Bone Conduction. In unilateral deafness without masking, a **shadow curve** should appear on bone conduction due to transcranial sound transmission. If this shadow curve is absent, despite claims of total unilateral hearing loss, it is diagnostic of **functional hearing loss**.

4. Discrepancy Between PTA and SRT. Normally, the Pure Tone Average (PTA) at 500, 1000, and 2000 Hz closely matches the Speech Reception Threshold (SRT) (within 10 dB). If SRT is better than PTA by more than 10 dB, this suggests exaggeration or malingering.

5. Stenger Test (Classic Clinical Test)

Principle:
When the same tone is played in both ears with different intensities, the patient will only perceive the **louder tone**.

Procedure: Always blindfold the patient during the test to avoid bias.

- Take **two identical tuning forks** or use a **two-channel audiometer**.
- Strike the forks and hold both 25 cm away from each ear.
- The patient will report hearing in the **normal ear**.
- Now, bring the tuning fork **closer (8 cm)** to the suspected ear, keeping the normal side unchanged.
- A **malingering patient** will now claim to hear **nothing**, even though the louder stimulus should be heard in the "deaf" ear.
- A truly deaf patient will continue hearing in the normal ear.

Interpretation:

- **Positive Stenger** (patient says he hears nothing): Indicates **malingering**.
- **Negative Stenger** (hears in one ear): Likely genuine loss.

6. Acoustic Reflex Threshold. Normally, the stapedial (acoustic) reflex is present at 70–100 dB SL. If a patient claims total deafness but the reflex is still elicited, it clearly indicates functional hearing and NOHL.

7. Electric Response Audiometry (ERA). ERA evaluates auditory evoked potentials, bypassing the patient's conscious effort. It can accurately estimate the hearing threshold within 5–10 dB, even in cases of suspected malingering. Highly reliable in children, non-cooperative patients, and suspected NOHL cases.

Management:

- Requires a sensitive, non-confrontational approach.
- Counseling and psychological support are often beneficial.
- Reassurance that hearing is better than reported.

5. **Central Hearing Loss / Auditory Processing Disorder (APD).** Individuals with **central hearing loss**, more accurately termed **Auditory Processing Disorder (APD)**, typically have **normal pure-tone hearing thresholds** and pass other standard hearing tests. However, they exhibit difficulties in processing distorted or unclear speech, particularly in challenging listening environments such as the presence of background noise (e.g., in social settings or classrooms). Consequently, these individuals struggle to follow verbal instructions, understand rapid speech, or distinguish similar-sounding words. These challenges are attributed to deficits in the processing of auditory information within the higher auditory pathways of the brain, rather than problems with the peripheral auditory system (ear or auditory nerve).

Key Features:

- Normal pure tone audiogram.
- Difficulty understanding speech, especially in noise.
- Difficulty localising sound.
- Problems with auditory memory or sequencing.
- Can hear, but cannot understand.

Diagnosis:

- Specialised audiometric tests are designed to assess central auditory function (e.g., speech-in-noise tests, dichotic listening tests).

Management:

- Auditory training.
- Environmental modifications (e.g., reducing background noise).
- Compensatory strategies.
- Frequency modulation (FM) systems in educational settings.

--------End of the Chapter----------