



# Deafness

## Objectives:

- Definition of the HL (hearing loss).
- Etiology of:
  - Conductive deafness .
  - Congenital Sensorineural deafness.
  - Acquired Sensorineural deafness.
- Who's at risk to develop HL .
- Impact of HL.
- Investigation of HL.
- Management of HL.

[ Color index : **Important** | **Notes** | Extra ]

**Resources:** Slides+433team+Notes+Lecture Notes ENT.

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## Introduction :

- ❖ Deafness is the Impairment of sound perception more than 20 (15) decibel<sup>1</sup> on pure tone audiogram.
- ❖ Deafness is partial or complete inability to hear from one side or both sides of the ear.
- ❖ 50% of deafness and hearing loss is avoidable through prevention, early diagnosis, and proper management.
- ❖ How common is hearing loss?
  - Overall about 1 in 10.
  - 1 in 3 adults 65 - 75.
  - 1 in 2 older than 75 .
  - 1-2% school age children.
  - 4% children under 5.

it's advised to do newborn screening because it's the commonest congenital anomaly.

### ❖ Signs of Hearing Loss:

- Talking louder than necessary.
  - Turning up volume on the TV or radio.
  - Complaints that other people “mumble”.
  - Confusion of similar sounding words نحلة / نخلة.
  - inappropriate responses in conversation.
  - Ringing or buzzing in the ears.
  - Lip Reading.<sup>2</sup>
  - Watching a speaker's face intently.
  - Difficulty “hearing” someone behind.
  - Having difficulty speaking on the telephone.
- 
- ❖ The impact of hearing impairment:
    - Affects Speech - Language - Education - Social(depression).

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<sup>1</sup> decibel is a unit for expressing the relative intensity of sound on a logarithmic scale.

<sup>2</sup> تشوف بعض الناس يقولون للأطفال يسمع اللي بيبي ويترك اللي بيبي . وهو مسكين مايسمع لأنه ماشاف.

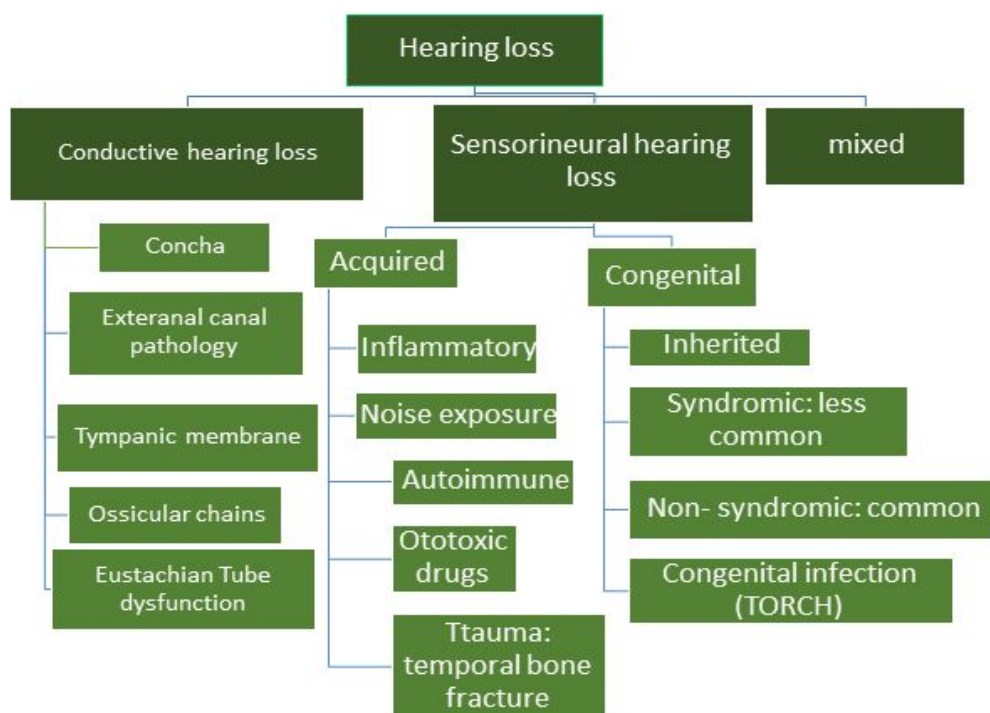
## High Risk Criteria For Hearing Loss in Infants:

- Family history of hereditary childhood sensorineural hearing loss.
- Hyperbilirubinemia and jaundice (kernicterus).
- Ototoxic medications.
- Bacterial meningitis.
- Birth weight less than 1500 grams & Premature babies.
- In utero infections “TORCH” (toxoplasmosis, syphilis, rubella, cytomegalovirus and herpes).
- Craniofacial anomalies (including pinna and ear canal).
- Birth asphyxia.
- Mechanical ventilation lasting 5 days or longer.
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.

## Types of hearing loss:

When the external and the middle ear are affected= conductive hearing loss. Inner ear (cochlea) and the nerve= sensory hearing loss.

Cochlea's job is tuning of the sound.(433)

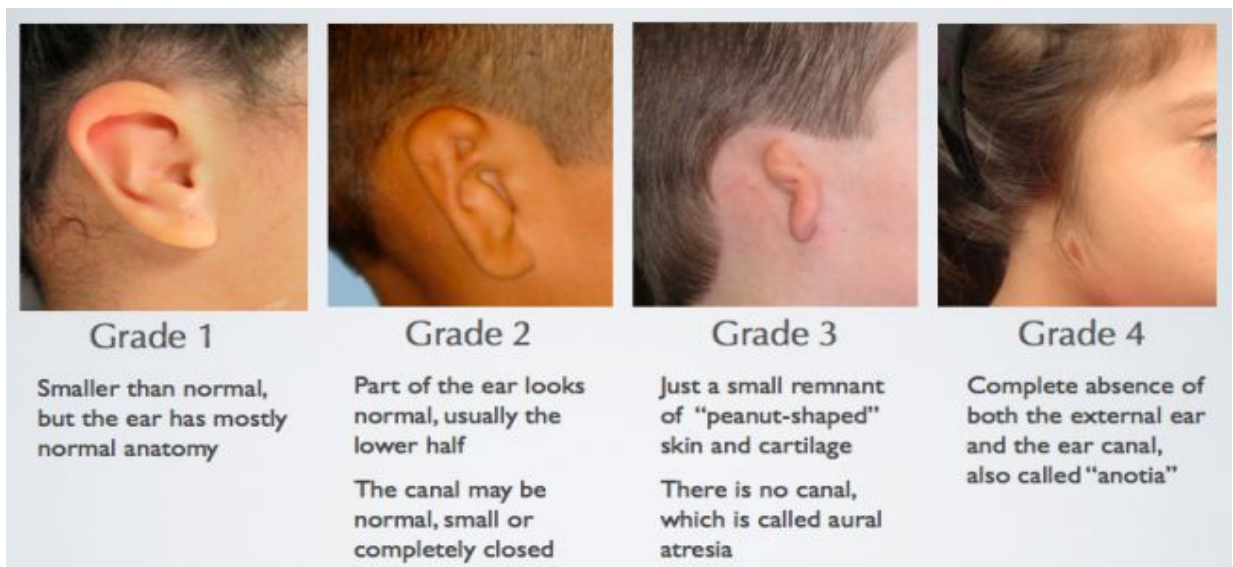


## Conductive hearing loss (CHL):

- ❖ Conduction of sound to the cochlea is impaired.
- ❖ Can be caused by **external** and **middle** ear disease
- ❖ .The lesion may lie in the external ear and tympanic membrane, middle ear or ossicles up to stapediovestibular joint.(433)

### ➤ External canal pathology:

- **Artesia (No ear canal) & Microtia:** Deformity of the ear auricle.
- **Inflammatory.**
- **Otitis media:** Acute suppurative (ASOM) – Otitis media with effusion (OME) - Chronic otitis media (CSOM).
- **Acute otitis externa:** It's a common condition involving inflammation of the ear canal. The acute form is caused primarily by bacterial infection, with *Pseudomonas aeruginosa* and *Staphylococcus aureus* the most common pathogens. **very painful.**
- **Wax:** the **commonest cause** of conductive hearing loss (CHL).
- **Foreign body** (Any form of obstruction can cause CHL). **furuncle(433)**
- **Trauma:** Skull base fracture blood goes to the external auditory canal> tympanic membrane perforation> blood in the middle ear gives **Raccoon eyes sign<sup>3</sup> and battle's sign.**
- **Tumors:** Benign or malignant.
  - **Osteoma:** Benign bone tumor, single, unilateral, broad-base.
  - **Exostosis:** Benign bone overgrowth, multiple, bilateral, pedunculated, in cold regions (Austria and Scandinavia).

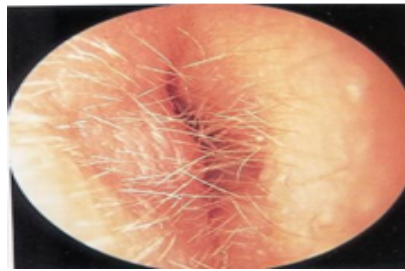


### Atresia

<sup>3</sup> periorbital ecchymosis



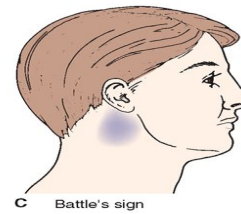
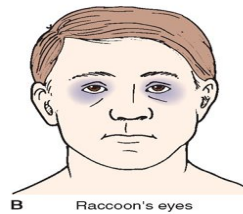
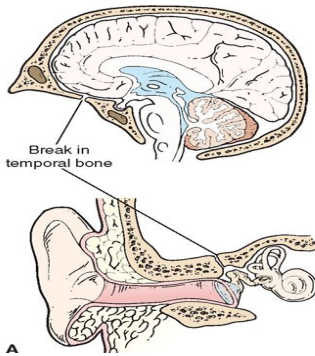
wax



Acute otitis externa



Foreign body



## ➤ Middle Ear:

### ● Tympanic membrane pathology:

- Absent TM caused by perforation: Fresh blood indicates a recent injury (acute injury).
- Too thick TM caused by tympanosclerosis.
- Multiple surgeries or infections (myringitis) causes TM scarring and thickening.
- Too thin TM caused by Secretory otitis media (SOM), Retraction.
- Tympanosclerosis: Calcification of an old inflamed tissue. (usually it's **asymptomatic -most of the time-**, but when it's symptomatic it causes CHL) (ask about previous infection in the ear while taking the history because it comes from recurrent infections) ,It's a condition characterized by the presence of masses of hard, dense connective tissue around the auditory ossicles in the middle ear, also know as myringosclerosis.



### ● Drum Retraction (Adhesive OM):

- It's also called Atresia, Atelectasis ear.
- The tympanic membrane gets sucked in because of eustachian tube dysfunction and negative pressure, which will suck the ear-drum inside. We treat it by ventilation tube, which prevents the ear from getting sucked inside by preventing the negative pressure. So perforation and retraction both of them are causes to conductive hearing loss.
- Treatment of adhesive OM is attachment of tube.

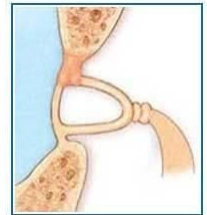


- **Ossicular chains:**

- Absent & erosion.
- Fixation: congenital or acquired otosclerosis.
- Disrupted trauma or dislocation.

- **Otosclerosis:**

- Congenital inherited autosomal recessive disease, causes fixation of the footplate (stapes) by new bone formation.
- A disease of the bony otic capsule characterized by abnormal replacement of mature bone of the otic capsule by woven bone of greater thickness.
- 10% otosclerosis lesions (10% symptomatic).
- Middle-age.
- Females: Male, 2: 1.
- Occur in Caucasian mostly.
- Worse during pregnancy and improve after delivery (due to hormonal changes).
- **Treatment: Stapedectomy** is a surgical procedure in which the innermost bone (stapes) of the middle ear is replaced with a small plastic tube of stainless-steel wire to improve the movement of sound to the inner ear.



- **Eustachian Tube dysfunction:**

- Retraction.
- Effusion.
- Otitis media: Acute suppurative (ASOM) Otitis media with effusion (OME)  
Chronic otitis media (CSOM).
- Congenital cholesteatoma.
- Squamous cell carcinoma

## Sensorineural hearing loss (SNHL):

- ❖ SNHL is a defect in the conversion of sound into neural signals or in the transmission of those signals to the cortex
- ❖ It can be caused by disease of the inner ear (cochlea), acoustic nerve (CNVIII), brainstem, or cortex.
- ❖ IT has **Two types:**
  - **Sensory** (the pathology is within hair cells in cochlea).



➤ **Neural** (the pathology is within the auditory nerve and its connection).

❖ **Etiologies:**

it may be congenital or acquired

● **Congenital hearing loss:**

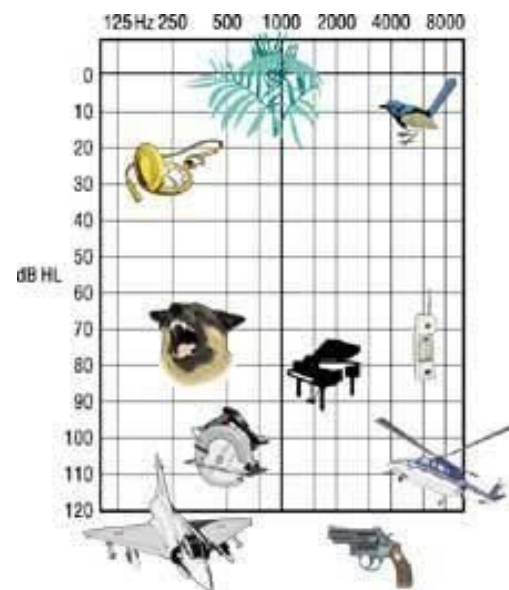
- Deafness affects 0.2%. (The most common congenital anomaly)
- SNHL attributed to:
  - ✓ 50% genetic factors.
  - ✓ 20-25% environmental.
  - ✓ 25-30% sporadic.
- Genetic (due to consanguinity marriages):
  - ✓ 75% AR (autosomal recessive).
  - ✓ 20% to AD (autosomal dominant).
  - ✓ 5% X-linked.
- Over 400 syndromes
- can lead to delay speech and language development.

● **Trauma:**

- Temporal bone fracture : affect hearing, balance and facial nerve.
- Longitudinal fracture:
  - ✓ Bleeding from ear.
  - ✓ Conductive hearing loss.
  - ✓ Uncommon facial nerve paralysis.
  - ✓ CSF.
- Transverse fracture:
  - ✓ SNHL.
  - ✓ Facial nerve paralysis common.
  - ✓ CSF.
- Mixed.

● **Noise induced SNHL:**

- Boilermaker's deafness.
- One of the most common occupationally induced disabilities.
- Follows chronic exposure to less intense sounds than seen in acoustic trauma and is mainly a hazard of noisy occupations. (That's why they have



devices that measure the noise in factories and airports) Load speakers in weddings have the same effects.

- Tinnitus (**only sign**).earliest sign
  - Commonly accompanied NISNHL.
  - Warning sign (**one gunshot could cause SNHL, and in KSA fireworks**).
    - 90 db for 8 hours
    - 95 db for 4 hours
    - 100 db for 2 hours
    - 105 db for 1 hours
- Causes damage to hair cells, starting in the basal turn of cochlea. Outer hair cells are affected before the inner hair cells.(433)

- **Ototoxic:**

- **Antibiotics (aminoglycosides). like Gentamicin.**

Patients particularly at risk are those: Concomitantly receiving other ototoxic drugs, Who have already received aminoglycoside antibiotics, Who are receiving high doses of ototoxic drugs with high serum level of drug, Who have genetic susceptibility to aminoglycosides. (433team).

- **Diuretics.(Furosemide)**They are known to cause oedema and cystic changes in the stria vascularis of the cochlear duct.
- Antineoplastics.
- Anti Inflammatories.
- Antimalarial agents.(chloroquine, quinine)
- Ototoxic agents.
- Others.
- **People at Higher risk:**
  - ✓ Renal failure (Elevated peak and trough levels).
  - ✓ Liver failure.
  - ✓ Immunocompromised.
  - ✓ Collagen-vascular disorders.
  - ✓ Advanced age (> 65 years).
  - ✓ Prior ototoxicity.
  - ✓ Concurrent use of known ototoxic agents.
  - ✓ Preexisting HL or Vestibular.
  - ✓ Bacteremia (fever).
  - ✓ Treatment course longer than 14 days.
  - ✓ + ve FHx of AG ototoxicity.

- **Acoustic neuroma (Vestibular Schwannoma):**

- Vestibular Schwannoma is a benign nerve tumor in the internal auditory meatus or cerebello-pontine (CP) angle at the base of the skull. It is usually unilateral, except in the very rare familial neurofibromatosis type 2 (NF2), when it may be bilateral. In its early stages, it causes progressive hearing loss and imbalance. As it enlarges, it may encroach on the trigeminal nerve in the CP angle, causing loss



of corneal sensation. In its advanced stage, there is raised intracranial pressure and brain stem displacement. Early diagnosis reduces the morbidity and mortality. Unilateral sensorineural deafness should always be investigated to exclude a neuroma. Audiometry will confirm the hearing loss. MR scanning will identify even small tumours.

- **Presbycusis:**

- Aging process of human beings, it's associated with grey hair, cataract and SNHL. most common type.
- Presbycusis = Deafness + Tinnitus + Recruitment (Out of proportion of loudness. (Meaning the patient can't hear, but when he hear, he hear everything louder than it's normal range) The cochlea normally acts as a filter; it decreases loud voices and amplifies the low sounds, here the cochlea is not functioning well.)
- **Overview of Hearing Loss:**
  - ✓ #1 Handicapping disorder.
  - ✓ 60% of Americans > 65 HL.
  - ✓ 90% of > 75 Y have HL.
  - ✓ HL + degenerative processes of aging.
  - ✓ ½ Vestibular symptoms.
- **Problems With Diagnosis includes:** Shame or embarrassment, HA social stigma, Embarrassment prevents 15 million elderly people from getting help.

- **Infection.**

## Conductive Hearing loss

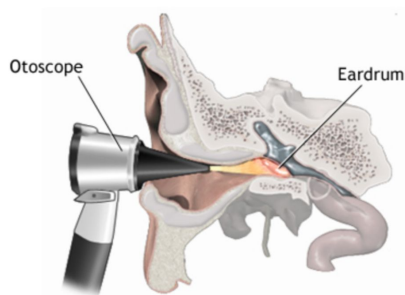
- Negative Rinne test ( $BC > AC$ )
- Weber lateralized to the poorer ear
- Normal absolute bone conduction
- Low frequencies affected more
- Audiometry shows bone conduction better than air conduction with air-bone gap. Greater the air-bone gap, more is the conductive loss
- Loss is not more than 60 dB.
- Speech discrimination is good

## Sensorineural Hearing loss

- A positive Rinne test ( $AC > BC$ )
- Weber lateralized to better ear
- Bone conduction reduced
- More often involving high frequencies
- No gap between air and bone conduction curve on audiometry
- Loss may exceed 60 dB.
- Speech discrimination is poor.
- There is difficulty in hearing in the presence of noise.

### Examination:

- Otoscopic



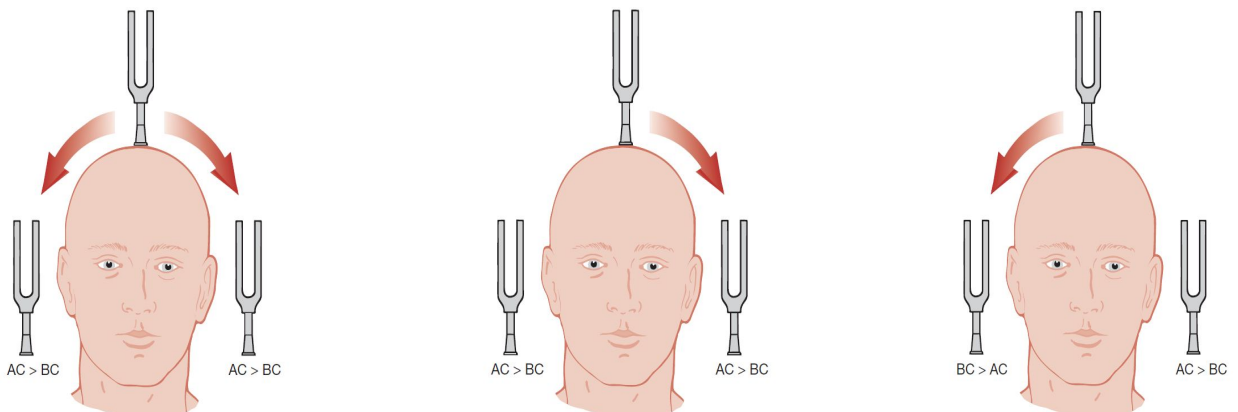
- Microscopic



### ◆ Clinical testing of hearing :

- **Tuning forks:** Tuning fork tests rely on the basic concept of classification of hearing loss. Deafness may be classified under one of these headings:
  - **Conductive** deafness.
  - **Sensorineural** deafness.

- **Mixed** conductive and sensorineural deafness.
- There are 2 tests:
  - **Weber test:** is a quick screening test for hearing.
    - It can detect:
      - Unilateral conductive hearing loss (middle ear hearing loss) deviated to affected ear.
      - Unilateral sensorineural hearing loss (inner ear hearing loss) deviated to better ear.
    - The test is useful in determining the type of deafness and in deciding which ear has the better-functioning cochlea. The base of a vibrating tuning fork is held on the middle of the skull and the patient is asked whether the sound is heard centrally or is referred to one or other ear.
      - In conductive deafness the sound is heard in the deafer ear.
      - In sensorineural deafness the sound is heard in the better-hearing ear.



**Figure 3.3** Tuning fork tests showing a positive Rinne in each ear and the Weber test referred equally to each ear, indicating symmetrical hearing in both ears with normal middle-ear function. **Figure 3.4** Sensorineural deafness in the right ear. The Rinne test is positive on both sides and the Weber test is referred to the left ear. **Figure 3.5** Conductive deafness in the right ear. The Rinne test is negative on the right, positive on the left, and the Weber test is referred to the right ear.

➤ **Rinne test:**

- It compares perception of sounds transmitted by air conduction to those transmitted by bone conduction through mastoid. evaluates hearing loss in one ear.
- This test compares hearing in one ear by air conduction (**AC**), and bone conduction (**BC**). It is usually performed as follows:
  - a tuning fork of 512Hz (cycles per second) is struck and held close to the patient's ear (AC); the base is then placed firmly on the mastoid process behind the ear (BC) and the patient is asked to state whether it is heard better by BC or AC (Fig. 3.3-check it above).

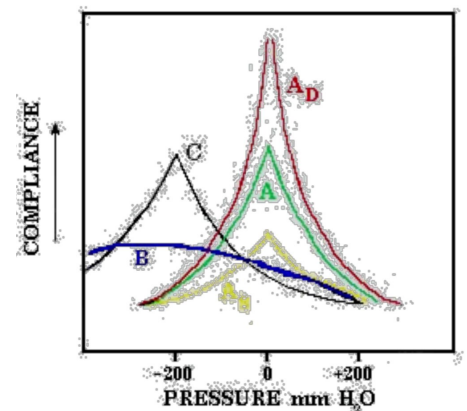
- Interpretation of Rinne's test:
  - If **AC > BC** (called Rinne **positive**) the middle and outer ears are functioning normally.
  - If **BC > AC** (called Rinne **negative**) there is defective function of the outer or middle ear (conductive deafness).
- Try this on yourself. Then gently occlude your outer ear by pressing the tragus, giving yourself a mild temporary conductive deafness. Now repeat the test and you should find that Rinne becomes negative, demonstrating the conductive loss.
- Rinne's test tells you little or nothing about the cochlea. It is a test of middle-ear function.

- **Impedance:**

- Acoustic reflex.

- **Tympanogram:**

- Is graphic representation of the relationship between the air pressure in the ear canal and the movement of the tympanic membrane.
- Type A: normal.
- Type B : fluid or perforated tympanic membrane.
- Type C : negative pressure in the middle ear.
- Type Ad : ossicular disruption with normal TM.
- Type As : ossicular fixation

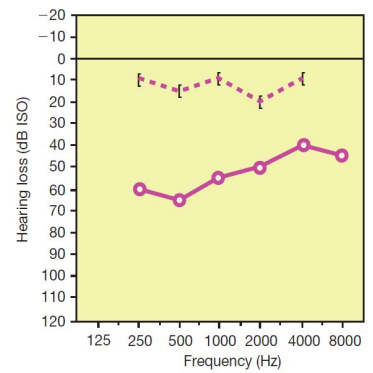
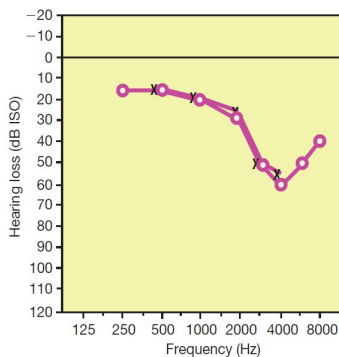
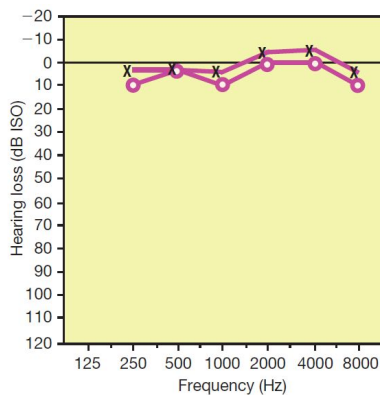


- **Audiogram:**

- Pure tone audiogram:
  - Pure tone audiometry provides a measurement of hearing levels by AC and BC and depends on the co-operation of the subject.
  - The test should be carried out in a soundproofed room.<sup>4</sup> The signal is presented to the patient through earphones (for AC) or a small vibrator applied to the mastoid process (for BC). Signals of increasing intensity at each frequency are presented to the patient, who indicates when the test tone can be heard.
  - The threshold of hearing at each frequency is charted in the form of an audiogram (Figs 3.6–3.8), with hearing loss expressed in decibels (dB)<sup>5</sup>.
  - When testing hearing by BC, it is essential to mask the opposite ear with narrow-band noise to avoid cross-transmission of the signal to that ear.

<sup>4</sup> The audiometer is an instrument that generates pure tone signals ranging from 125 to 12 000 Hz (12 kHz) at variable intensities.

<sup>5</sup> Decibels are logarithmic units of relative intensity of sound energy.



**Figure 3.6** A normal pure tone audiogram. o-o-o, right ear; x-x-x, left ear. **Figure 3.7** A pure tone audiogram showing sensorineural deafness maximal at 4 kHz typical of noise-induced deafness. **Figure 3.8** A pure tone audiogram showing conductive deafness. The BC (dashed line) is normal but the AC (solid line) is impaired. A case of otosclerosis.

○ **Speech audiogram:**

- Speech audiometry measures the ability of each ear to discriminate the spoken word at different intensities.
- A recorded word list is supplied to the patient through the audiometer at increasing loudness levels, and the score is plotted on a graph.
- In some disorders, the intelligibility of speech may fail above a certain intensity level.
- Above a critical threshold, sounds are suddenly perceived as having become excessively loud – loudness recruitment. This suggests a cochlear disorder and is common in elderly patients with presbycusis.

**Management of hearing impairment:**

● **Tow Types :**

➤ Medical

➤ **Hearing Aid :**

❖ **History:** 1550 by Girolamo Cardano when he saw that sound could be transmitted through the teeth.



## ❖ Cochlear implant:

- Putting tiny electrode in the cochlea.
- In congenital HL the cochlear implant is ineffective after 5 years (The child will be postlingual and the will have child already learned sign language so it will be difficult to adapt) , due to the disappearance of auditory segment from the brain (it gets used up by other centers of other senses that's why their other senses become better like vision for example) . But in people who used to hear and then lost their hearing there is no time limit for the usage of cochlear implant but we prefer to implant within 10 years.
- It's a device consisting of a microphone, signal processor, external transmitter, and implanted receiver; the receiver is surgically implanted under the skin near the mastoid process above and behind the ear.(source: medical dictionary)
- - So in the exam if they gave you a cochlear implant picture you should know it, it looks like a regular hearing aid but with a magnet from outside.

### Classical indication of cochlear implant:

Bilateral sensory-neural hearing loss not benefiting from hearing aids and less than 5 years of age if congenital hearing loss.

It is standard practice everywhere to implant for children under 5 because of the improvement in the quality of life.

We have to make sure that the patients has a cochlea and a nerve prior to implant.

## ❖ Bone Anchored Hearing Aids (B.A.H.A):

- Titanium implants, used in CHL (they use titanium because it doesn't react with the body)
- BAHA stimulates the cochlea by transmitting the sound waves through the bones in our skull, or bone conduction, thereby bypassing the outer and the middle ear.
- Atresia of external ear canal. and also microtia



We can also reach the brainstem and due to the presence of the cardiac center you can stimulate one and the patient may die but they still want the procedure to be done so it says a lot about the importance of hearing.

◆ **Auditory brainstem implant (A.B.I):**

Implant in the brains



◆ **Lip Reading**

◆ **Sign Language**

◆ **Myringoplasty & tympanoplasty:** in case of CSOM.

◆ **Ossiculoplasty:**

- in case of ossicular discontinuity.
- Either **partial** or **complete** (in case all 3 ossicles are involved) ossiculoplasty.