



Deafness



It can be HEARD without having to SPEAK

Objectives:

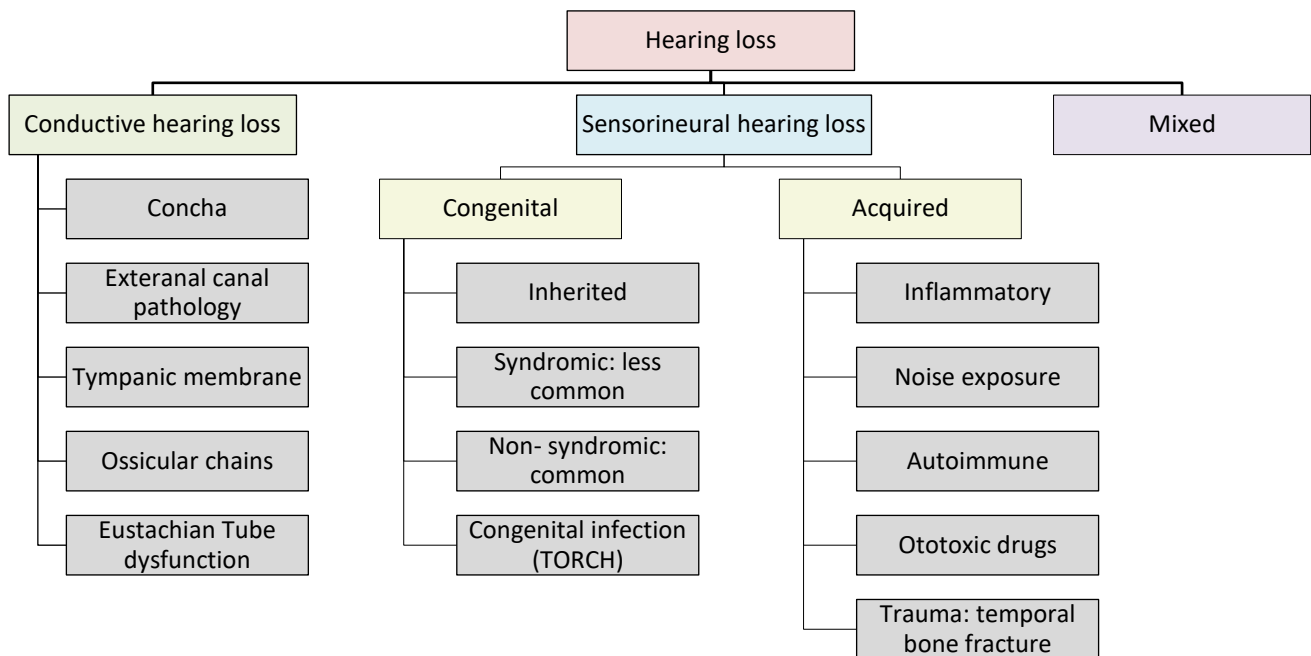
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| <ul style="list-style-type: none">• Definition• Prevalence of deafness• Impact of deafness• Who is at risk of deafness• Classification of deafness | <ul style="list-style-type: none">• Etiologies of conductive deafness• Etiologies of sensorineural deafness• Examination of patient with deafness• Investigation of patient with deafness• Management of deafness |
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Correction File

Color Index : Slides - **Team 433** - **Important Notes** - **Doctors' Notes** - **toronto notes**



Definitions:

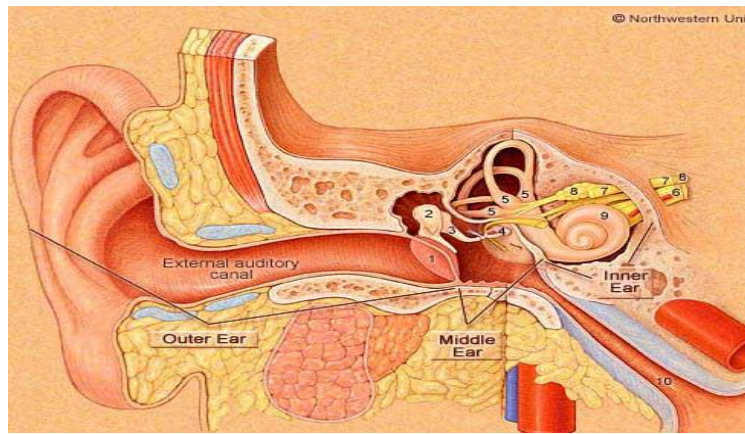
- Impairment of sound perception more than 20 (15) decibel on pure tone audiogram.
- Deafness is partial or complete inability to hear from one side or both sides of the ear.
- 50% of deafness and hearing loss is avoidable through prevention, early diagnosis, and management

The impact of hearing impairment:

- Speech
- Language
- Education
- Social

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 asphexia
- 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



- From auricle to the oval window → conductive hearing
- Inner ear (cochlea) + Nerve → sensorineural hearing
- Drum → magnify sounds x18 to deliver enough sound energy to the inner ear

Hearing loss Types:

1) Conductive hearing loss:

1. Concha
2. Ear Canal
3. Drum
4. Ossicular Chain
5. Eustachian Tube

2) Sensorineural hearing loss

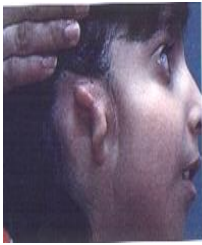
3) Mixed hearing loss: combination of conductive and sensorineural hearing loss

Conductive hearing loss (CHL):

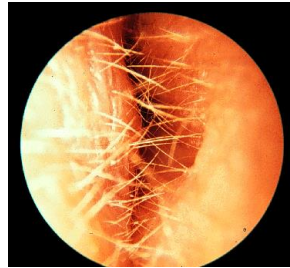
- Conduction of sound to the cochlea is impaired
- Can be caused by external and middle ear disease

❖ **External canal pathology:**

1. **Artesia (No ear canal) & Microtia:** Deformity of the ear auricle.
2. Inflammatory
3. **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.
4. **Wax:** the commonest cause of conductive hearing loss (CHL).
5. **Foreign body (Any form of obstruction can cause CHL)**
6. **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



Artesia



Acute otitis externa



Wax



Foreign body



Bone growth from external canal (Osteoma)



Exostosis

❖ **Tympanic membrane:**

- Absent → Perforated: Fresh blood indicates a recent injury (acute injury).
- Too thick → tympanosclerosis
- Multiple surgeries or infections (myringitis) → TM scarring → thinking
- Too thin → SOM, Retraction



TM Perforated

Tympanosclerosis:

- Calcification of an old inflamed tissue. (usually it's asymptomatic, 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.



Tympanosclerosis

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 conductive hearing loss.
- Treatment of adhesive OM is attachment of tube.



❖ Ossicular chains

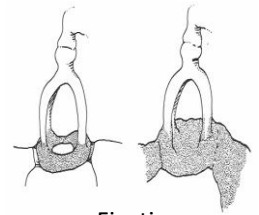
- Absent & erosion
- Fixation: congenital or acquired otosclerosis
- Disrupted trauma or dislocation

Otosclerosis:

- Congenital inherited autosomal recessive disease, cause 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 inner most 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.



Absent & erosion



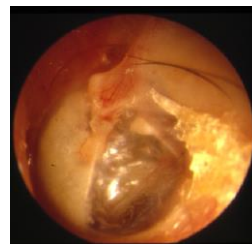
Fixation



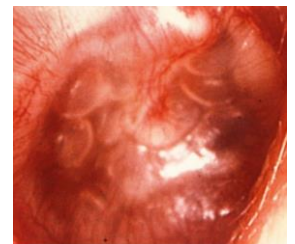
Disrupted traum

• Eustachian Tube dysfunction:

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



Retraction



Effusion

Sensorineural hearing loss:

- Defect in the conversion of sound into neural signals or in the transmission of those signals to the cortex
- Can be caused by disease of the inner ear (cochlea), acoustic nerve (CNVIII), brainstem, or cortex

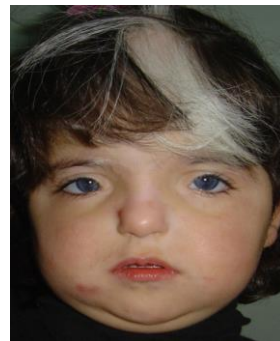
Two types:

1. Sensory (the pathology is within hair cells in cochlea)
2. Neural (the pathology is with in the auditory nerve and it's connection)

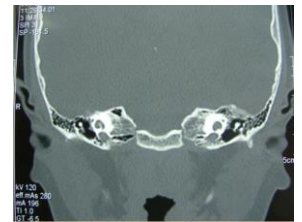
Etiologies:

1/ Congenital: Mondeni deformity

- Inherited
- Syndromic: less common
 - ♦ Wartenberg syndrome: white hair, blue eyes, SNHL
 - ♦ Teacher Collins syndrome: CHL, difficult intubation
- Non- syndromic: common
- Congenital infection (TORCH)



wartenberg syndrome



Mondeni deformity



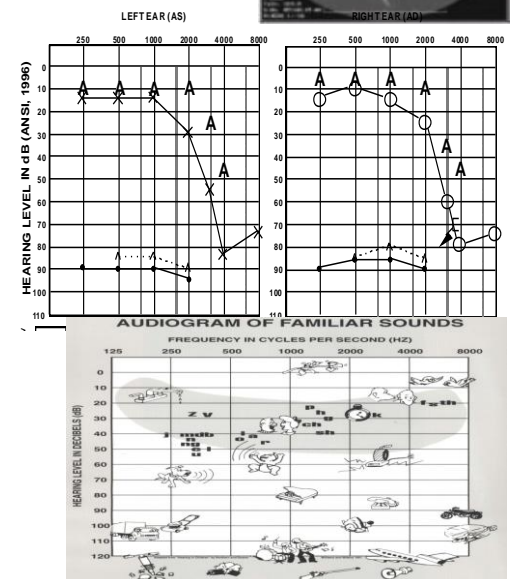
Teacher Collins syndrome

2/ Acquired:

1. **Inflammatory** -labyrinthitis ,meningitis → infection can be transmitted from the brain to the inner ear → cause ossification and fibrosis , hearing loss is immediate, reversible if treated early
2. **Noise exposure:** can be immediate or progressive hearing loss

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.
- Causes damage to hair cells, starting in the basal turn of cochlea. Outer hair cells are affected before the inner hair cells.
- Tinnitus (only sign)
 - Commonly accompanied NISNHL
 - Warning sign (One gunshot could cause SNHL, and in KSA fireworks)



3. **Autoimmune:** -Cogan syndrome, SLE, rheumatoid arthritis. → usually bilateral HL with dizziness.

4. **Ototoxic drugs:** -Aminoglycosides groups , diuretics, chloroquine, chemotherapy

Ototoxicity:

• Antibiotics (aminoglycosides) 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.

• Diuretics (Furosemide): They are known to cause oedema and cystic changes in the stria vascularis of the cochlear duct.

• Antineoplastic

• Antinflammatories

• Antimalarial agents (chloroquine, quinine)

• Ototoxic agents

• Others

Higher risk:

1. Renal failure (Elevated peak and trough levels)

2. Liver failure

3. Immunocompromised patients

4. Collagen-vascular disorders

5. Advanced age (> 65 years)

6. Prior ototoxicity

7. Concurrent use of known ototoxic agents

8. Preexisting HL or Vestibular

9. Bacteremia (fever)

10. Treatment course longer than 14 days

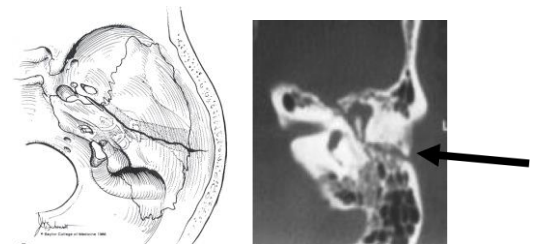
11. Positive family history of AG ototoxicity

5. Trauma

☑ Temporal bone fracture : affect hearing, balance and facial nerve

1. Longitudinal fracture:

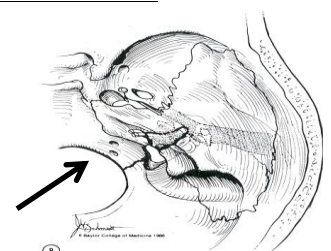
- Bleeding from ear
- Conductive hearing loss
- Uncommon facial nerve paralysis
- CSF



Longitudinal fracture

2. Transverse fracture

- SNHL
- Facial nerve paralysis common
- CSF



Transverse fracture

3. Mixed

6. Vestibular schwannoma

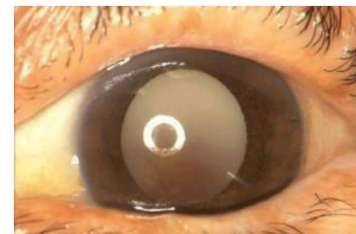
7. Meniere's disease

8. Presbycusis:

- It's associated with grey hair, cataract and SNHL.
- Sensorineural hearing loss associated with physiological aging process in the ear is called presbycusis. It usually manifests at the age of 65 years but may do so early if there is hereditary predisposition, chronic noise exposure or generalized vascular disease.
- Treatment: Hearing aid
- Presbycusis = Deafness + Tinnitus + Recruitment

9. Acoustic neuroma

10. Systemic disorders (DM, hypo- thyroidism, kidney disease and multiple sclerosis)



Diagnosis:

1. History

2. Severity of deafness (mild, moderate, moderately severe, severe, profound or total). This can be found out on audiometry.

3. Site of lesion

4. Laboratory tests (X-rays or CT scan of temporal bone for evidence of bone destruction (congenital cholesteatoma, glomus tumor, middle ear malignancy or acoustic neuroma), blood counts (leukemia), blood sugar (diabetes), serology for syphilis, thyroid functions (hypothyroidism), kidney function tests).

Examination:

- General look (syndromic)
- Complete head and neck exam
- Otoloscopic / microscopic ear exam of **both ears**

Clinical testing of hearing:

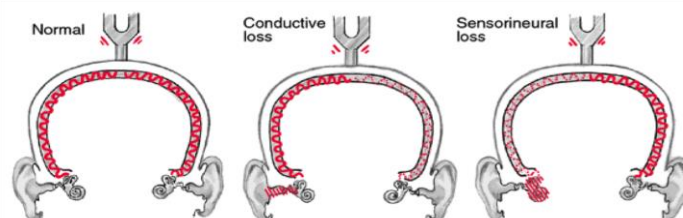
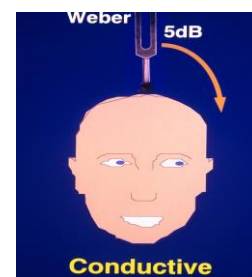
• Tuning forks:

○ Weber: hit the forks then place them on the middle of the forehead (on vertex of head) and patient states whether it is heard centrally (Weber negative) or is lateralized to one side (Weber right, Weber left)

- Normally: central
- In unilateral conductive hearing loss patient will hear better in the affected side
- In sensorineural hearing loss patient will hear better in the normal side
- In bilateral hearing loss weber is not used

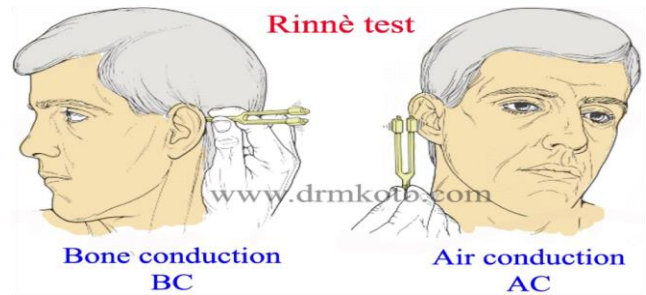
○ Weber Test lateralization = ipsilateral conductive hearing loss or contralateral sensorineural hearing loss

○ The Weber test is more sensitive in detecting conductive hearing loss than the Rinne test



Rinne: hit the two forks and held firmly on mastoid process to test bone conduction then place the tuning fork in front of the ear (beside the pinna) to test air conduction.

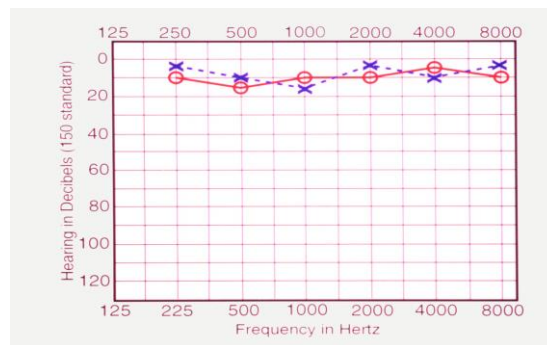
- +ve test: air conduction > bone conduction → in normal patients or SNHL
- -ve test: air conduction < bone conduction → unilateral CHL



- Audiogram battery:
 - Pure tone audiogram
 - Speech audiogram
- Impedance
 1. Acoustic reflex
 2. Tympanogram
 3. Volume
 4. Acoustic reflex decay

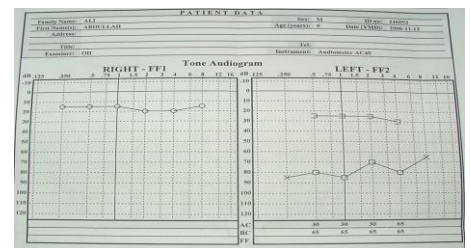
pure tone audiogram

- ◆ Normal hearing:



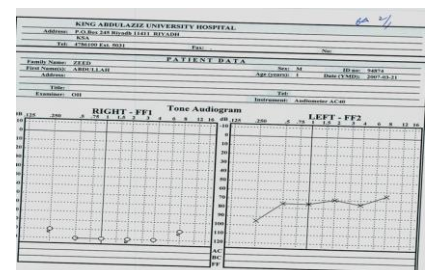
- ◆ Conductive hearing loss:

- BC → normal
- AC → less than normal range
- gap between AC and BC thresholds > 10 dB (an air-bone gap)



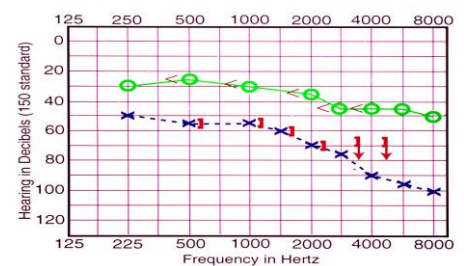
- ◆ Sensorineural hearing loss:

- Air and bone conduction → both are decreased
- No gap between AC and BC < 10 dB (no air-bone gap)



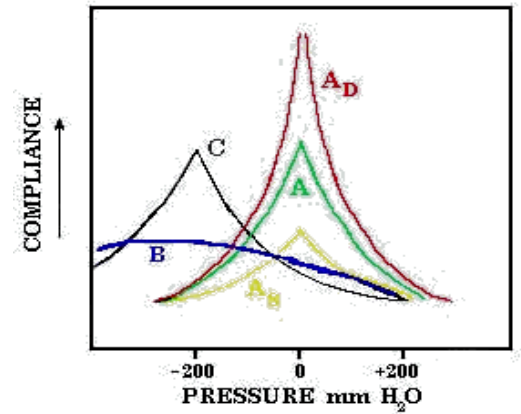
- ◆ Mixed hearing loss:

- Air and bone conduction → both are decreased
- Gap between AC and BC thresholds > 10 dB (an air-bone gap)

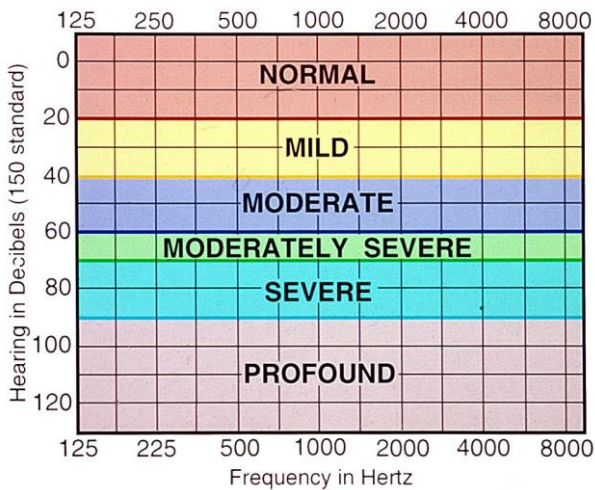


❖ Tympanogram

- Type A → Peak at zero pressure → normal
- Type B → flat curve → effusion (fluid) or perforation
 - how to differentiate between effusion (fluid) or perforation? by Ear Canal Volume (ECV)
 - if increased → perforation
 - if nearly normal → effusion (fluid)
- Type C → negative pressure: eustachian tube dysfunction

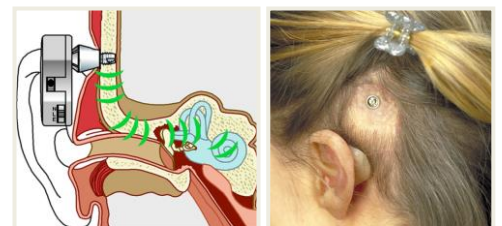


❖ Degree of hearing impairment



❖ Managemnt of deafness:

- **Medical management**
- **Hearing aids** → cause magnification
- **BAHA (Bone anchored hearing aid):**
 - Atresia of external canal
 - Chronic drainage ear not responding to surgery
 - **indication: congenital or acquired conductive hearing loss**
 - **mechanism: cause vibration of bone so the voice goes from the bone to the inner ear**
 - Titanium implants, used in conductive hearing loss (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 by- passing the outer and the middle ear.

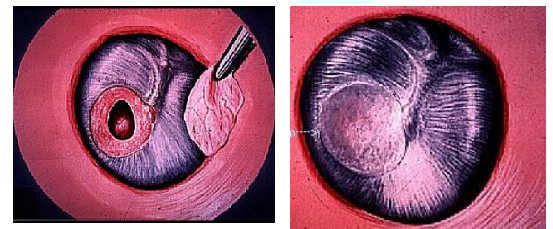
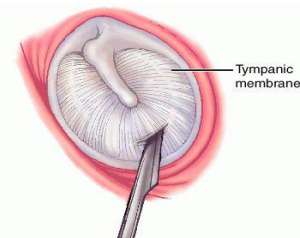


Management of deafness:

• Surgery

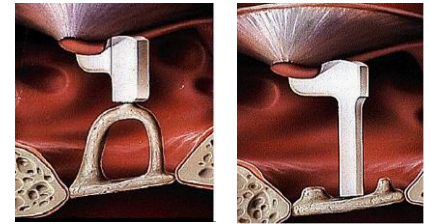
A. Myringotomy and ventilation tube Otitis media with effusion

- indication: resistant effusion, recurrent acute otitis media, eustachian tube dysfunction
- complication: bleeding, infection, obstruction, otorrhea, multiple surgeries → myringosclerosis



B. Myringoplasty & tympanoplasty in case of CSOM

C. Ossiculoplasty



D. Cochlear implant:

- prelingual children and post-lingual adult
- it pass by the external, middle and inner ear to stimulate auditory nerve directly
- Indication: congenital or acquired sensory neural hearing loss
- mechanism: by putting tiny electrode → cause digitalization of voice (convert voice to electrical current) that goes to the nerve directly and stimulate it
- In congenital HL the cochlear implant is ineffective after 5 years, due to the disappearance of auditory segment from the brain. But in people who used to hear and then lost their hearing there is no time limit for the usage of cochlear implant.
- 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.



Type of Hearing loss	Conductive Hearing loss	Sensorineural Hearing loss
Rinne test	Negative Rinne test (BC>AC)	A positive Rinne test (AC > BC)
Weber test	weber lateralized to the poorer ear	Weber lateralized to better ear
Bone conduction	Normal absolute bone conduction	Bone conduction reduced
Frequencies	Low frequencies affected more	More often involving high frequencies
Audiometry	- 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.	- No gap between air and bone conduction curve on audiometry - Loss may exceed 60 dB.
Speech	Speech discrimination is good	Speech discrimination is poor.
Hearing in the presence of noise	—	There is difficulty in hearing in the presence of noise.

