7- Hearing loss

Objectives:

- Definition of the HL (hearing loss).
- Prevalence of deafness.
- Etiology of:
  - Conductive deafness.
  - Sensorineural deafness.
- Who's at risk to develop HL.
- Impact of HL.
- Classification of hearing loss.
- Examination of the patient with HL.
- Investigation of patient with HL.
- Management of HL.

Resources: Slides+Lecture notes of ENT+Notes+435team+team 436 group A

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Introduction :

- Deafness is the Impairment of sound perception more than 20 (15) decibel[1] on pure tone audiogram. The normal is hearing at 20 if you need more than 20 its abnormal.
- Hearing impairment 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 نخلة/ نحلة.
    - inappropriate responses in conversation.
    - Ringing or buzzing in the ears.
    - Lip Reading.[2]
    - Watching a speaker's face intently.
    - Difficulty "hearing" someone behind.
    - Having difficulty speaking on the telephone.

- The impact of hearing impairment:
    - Affects Speech if the input is flawed then the output will be flawed too/ if you listen to something in a wrong way then you'll repeat it wrong too.
    - Language
    - Education
    - Social They'll feel left out/isolated and depressed. They'll have thoughts like "what if others are talking about me"

[1] decibel is a unit for expressing the relative intensity of sound on a logarithmic scale.

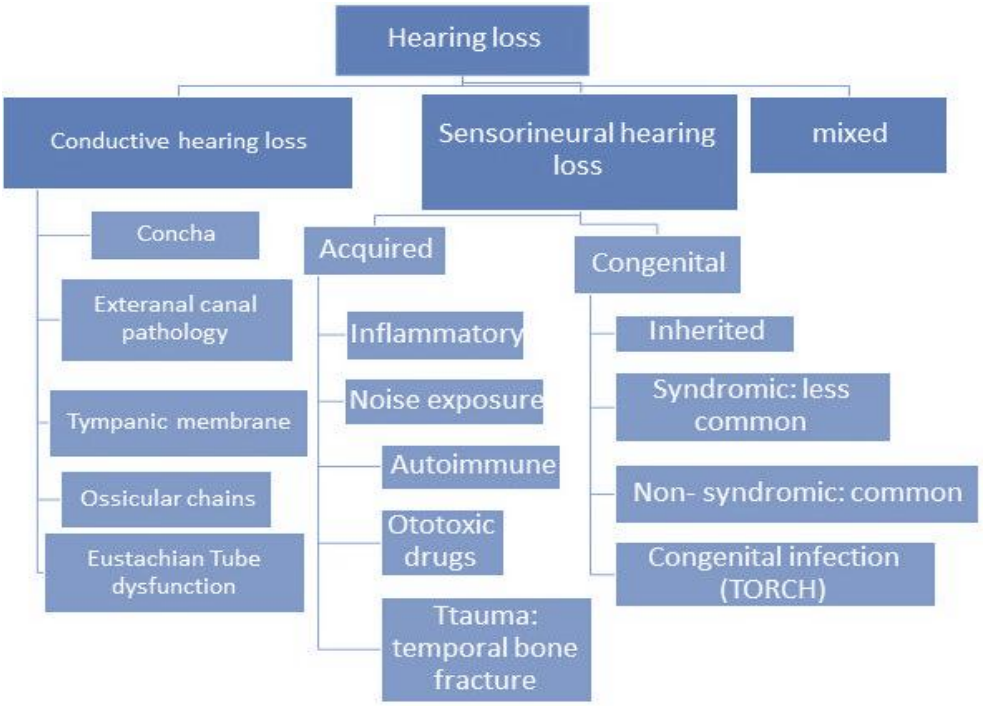
[2] وهو مسكين مايسمع لآنه ماشاف. تشوف بعض الناس يقولون للأطفال يسمع اللي يبي ويترك اللي يبي .

High Risk Criteria For Hearing Loss in Infants:

- Family history of hereditary childhood sensorineural hearing loss.
- Hyperbilirubinemia and jaundice (kernicterus) effecting the area if the brain responsible for the reception of language.
- Ototoxic medications. Such as Aminoglycosides or chemotherapy like Methotrexate
- Bacterial meningitis.
- Birth weight less than 1500 grams & Premature babies causing LBW and hypoxia affecting the brain which affects the area for language making them susceptible to deafness.
- In utero infections "TORCH" where the mother transmits it to the baby (toxoplasmosis, syphilis, rubella, cytomegalovirus and herpes).
- Craniofacial anomalies (including pinna and ear canal). Whether syndromic or non they may cause deafness
- 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. RF for adults such as Trauma or noise exposure in airplanes (not wearing ear plugs)

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:

1- Congenital

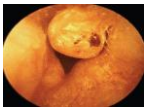
- **Atresia** (No ear canal) **&** **Microtia**: Deformity of the ear auricle

2- Inflammatory:

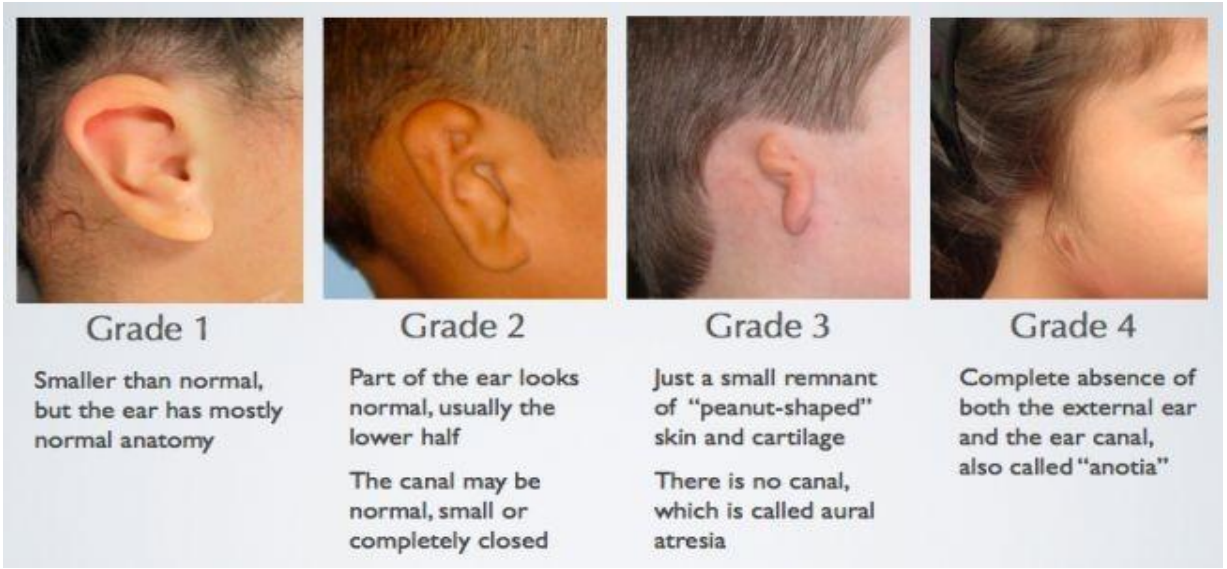
- **Otitis media**: Acute suppurative (ASOM) – Otitis media with effusion (OME) - Chronic otitis media (CSOM).
- **Acute otitis externa**: It's a common condition involving inflammation of the ear canal. The acute form is caused primarily by bacterial infection, with Pseudomonas aeruginosa and Staphylococcus aureus the most common pathogens. Very painful.

3-Obstruction:

- **Wax**: the **commonest cause** of conductive hearing loss (CHL).
- **Foreign body** (Any form of obstruction can cause CHL like insekt or Q-tips).
- **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).
- **Trauma**: Skull base fracture blood goes to the external auditory canal> tympanic membrane perforation> blood in the middle ear gives Raccoon eyes sign[3] and battle's sign.

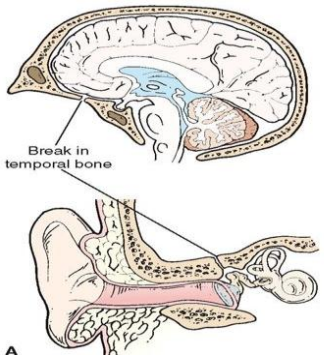


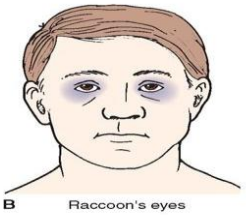
Atresia

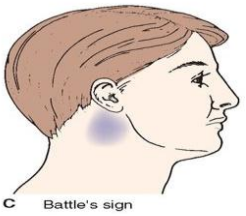


[3] periorbital ecchymosis

wax **Acute otitis externa** **Foreign body**







- **Tympanic membrane pathology:**

1- Absent TM caused by perforation: Fresh blood indicates a recent injury (acute injury).

2-Too thick TM caused by tympanosclerosis.

- Tympanosclerosis: Calcification of an old inflamed tissue. (usually it's **asymptomati**c -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 known as myringosclerosis.
- Multiple surgeries or infections (myringitis) causes TM scarring and thickening.

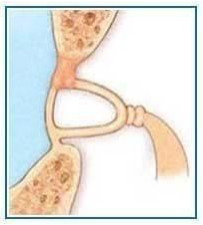
3-Too thin TM caused by Secretory otitis media (SOM), Retraction.

- **Drum Retraction (Adhesive OM):**

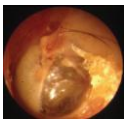
- It's also called Atresia, Atelectasis ear.
- The tympanic membrane gets sucked in because of eustachian tube dysfunction and negative pressure, which will suck the ear-drum inside. We treat it by ventilation tube, which prevents the ear from getting sucked inside by preventing the negative pressure.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

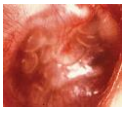
- **Ossicular chains:**
  - Absent & erosion.
  - Fixation: congenital or acquired otosclerosis.
  - Disrupted trauma or dislocation.

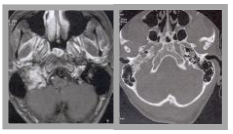
  - **Otosclerosis:**
    - Congenital inherited autosomal recessive disease causes fixation of the footplate (stapes) by new bone formation.
    - A disease of the bony otic capsule characterized by abnormal replacement of mature bone of the otic capsule by woven bone of greater thickness.
    - 10% otosclerosis lesions (10% symptomatic).
    - Middle-age.
    - Females: Male, 2: 1.
    - Occur in Caucasian and Europeans 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
  - Paraganglioma
  - schwannoma







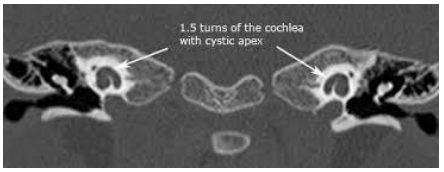
Sensorineural hearing loss (SNHL):

- 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:**



Mondini malformation

1-inherited

2- Syndromic (less common) such as waardenberg syndrome they have deafness and defects in their pigmentation.

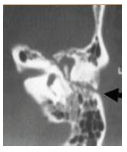
3- Non-syndromic (more common).

4- Congenital infection (TORCH)

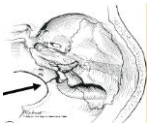
  - can lead to delay speech and language development.

- **acquired:**
  - **Trauma:**
    - Temporal bone fracture: affect hearing, balance and facial nerve.

      1. Longitudinal fracture:
         - Bleeding from ear.
         - Conductive hearing loss.
         - Uncommon facial nerve paralysis.
         - CSF.

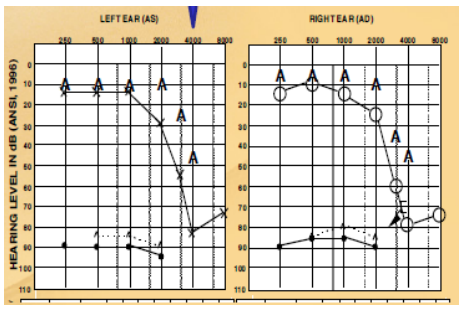



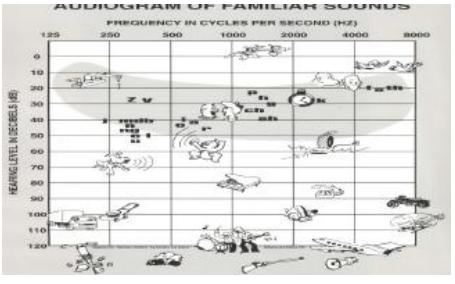
      2. Transverse fracture:
         - SNHL.
         - Facial nerve paralysis common.
         - CSF.

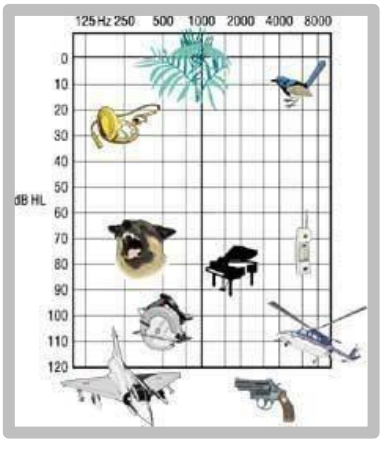


    - Mixed.
  - **Noise exposure induced SNHL:**
    - Boilermaker's deafness.
    - One of the most common occupationally induced disabilities.

      Follows chronic exposure to less intense sounds than seen in acoustic trauma and is mainly a hazard of noisy occupations. Here its cumulative like blow drying it is NOT REVERSIBLE (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) Hunting or explosions are one time highly intense shots causing hearing loss straight away.. Causes damage to hair cells, starting in the basal turn of cochlea. Outer hair cells are affected before the inner hair cells. (433)

- 90 db for 8 hours
- 95 db for 4 hours
- 100 db for 2 hours
- 105 db for 1 hours

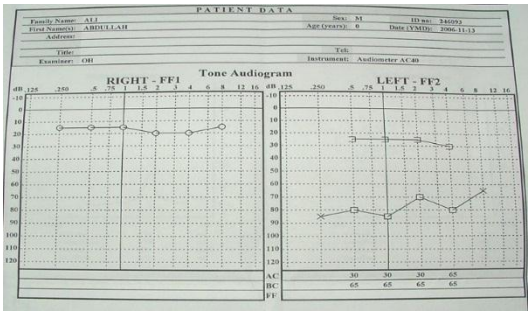
- **Ototoxic:**
  - **Antibiotics (aminoglycosides).** like Gentamicin.
  - Diuretics. (Furosemide)They are known to cause oedema and cystic changes in the stria vascularis of the cochlear duct.
  - Antineoplastics.
  - Anti Inflammatories.
  - Antimalarial agents. (chloroquine, quinine)
  - Ototopic agents.
  - Others.

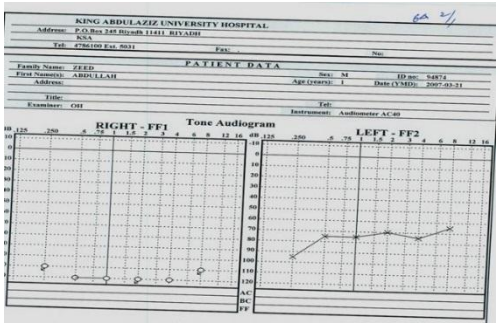
- Patients particularly at risk are those: Concomitantly receiving other ototoxic drugs, who have already received aminoglycoside antibiotics, who are receiving high doses of ototoxic drugs with high serum level of drug, who have genetic susceptibility to aminoglycosides. (433team)
  - **People at Higher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

- **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
    - ✓Half vestibular symptoms
  - **Problems With Diagnosis includes:** Shame or embarrassment, HA social stigma, Embarrassment prevents 15 million elderly people from getting help.

- **Inflammatory:** (labyrinthitis, meningitis): in meningitis it goes to cochlear through natural connection of the brain and ear causing ossification and the new bone being made causes death of OHC, some say the connection is through a blood supply, also it may affect Wernicke's and the temporal lobe causing neural hearing loss.
- **Autoimmune** (Cogan syndrome) with SLE or PRH

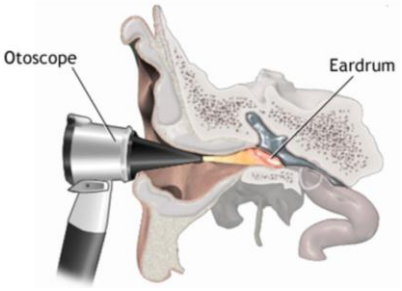




Conductive Hearing loss	Sensorineural Hearing loss
-Negative Rinne test (BC>AC)	-A positive Rinne test (AC > BC)
-Weber lateralized to the poorer ear	-Weber lateralized to better ear
-Normal absolute bone conduction	-Bone conduction reduced
-Low frequencies affected more	-More often involving high frequencies
-Audiometry shows bone conduction better than air conduction with air-bone gap. Greater the air-bone gap, more is the conductive loss	-No gap between air and bone conduction curve on audiometry
-Loss is not more than 60 dB.	-Loss may exceed 60 dB.
-Speech discrimination is good	-Speech discrimination is poor.
	-There is difficulty in hearing in the presence of noise.

Examination:

- General look (syndromic)
- Complete head and neck exam.
- Otoscopic / microscopic ear exam for both ears.
- Tuning fork test.



❖ 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. You need to know how to interpret the result

- 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 another ear.
- **Interpretation:**
    - In conductive deafness the sound is heard in the deafer ear.
    - In sensorineural deafness the sound is heard in the better-hearing ear.

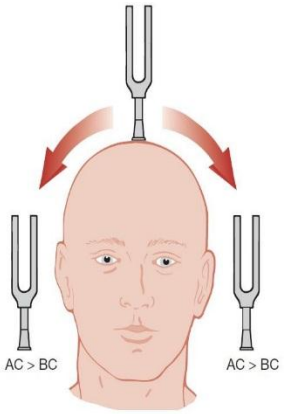


Figure 3.3 Tuning fork tests showing a positive Rinne in each ear and the Weber test referred equally to each ear, indicating symmetrical hearing in both ears with normal middle-ear function.

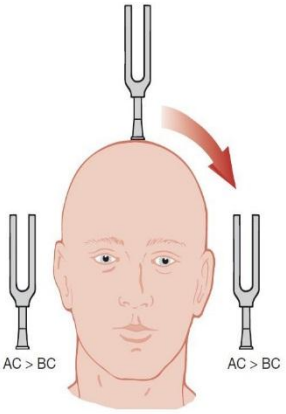
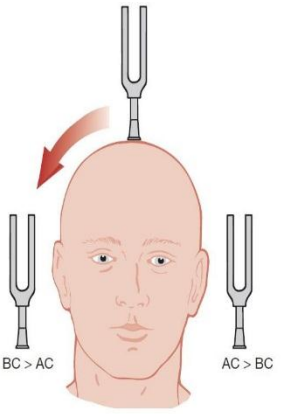


Figure 3.4 Sensorineural deafness in the right ear. The Rinne test is positive on both sides and the Weber test is referred to the left ear.



gure 3.5 Conductive deafness in the right ear. The Rinne test is negative on the right, sitive 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).

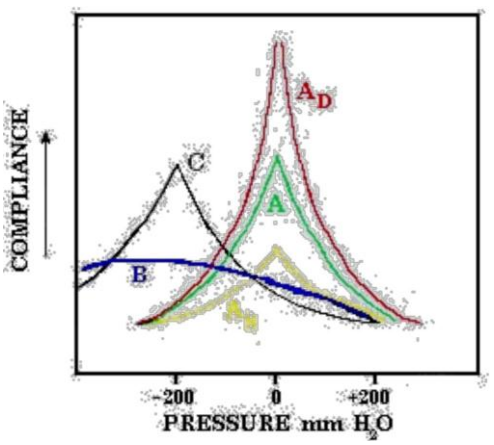
Interpretation of Rinne's test:

- 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[1]. 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)[2].
    - 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.
    - Tests the intensity of sound and its frequency, the patient decides the threshold by saying if he can or can't hear it.

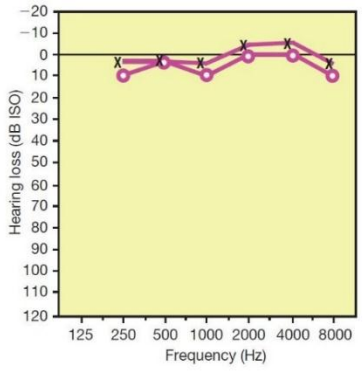


Figure 3.6 A normal pure tone audiogram. o–o–o, right ear; x–x–x, left ear.

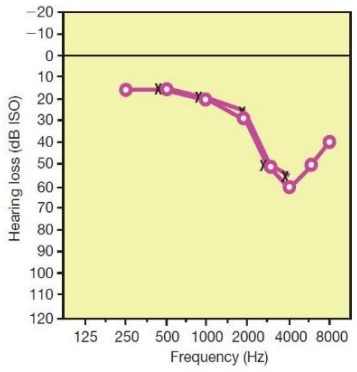
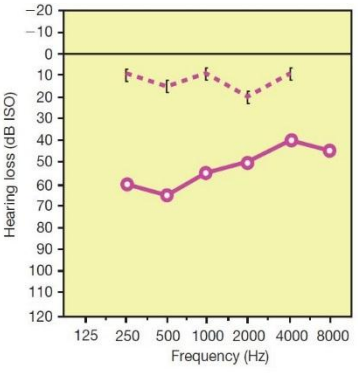
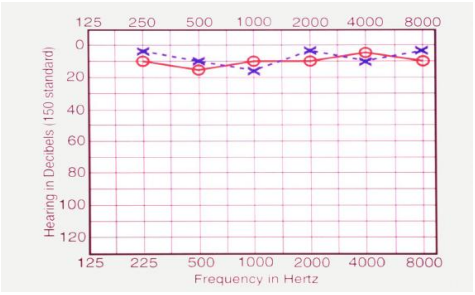


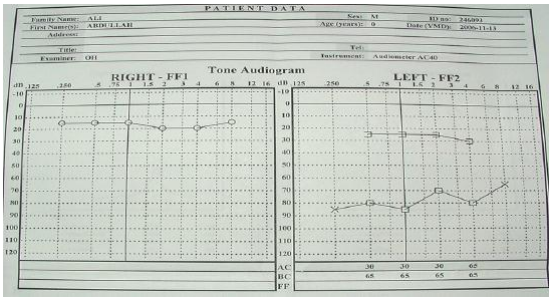
Figure 3.7 A pure tone audiogram showing sensorineural deafness maximal at 4 kHz typical of noise-induced deafness.



igure 3.8 A pure tone audiogram showing conductive deafness. The BC (dashed line) normal but the AC (solid line) is impaired. A case of otosclerosis.

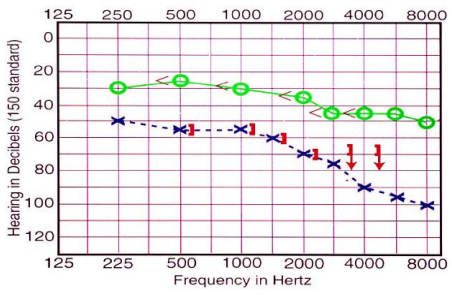
Normal hearing

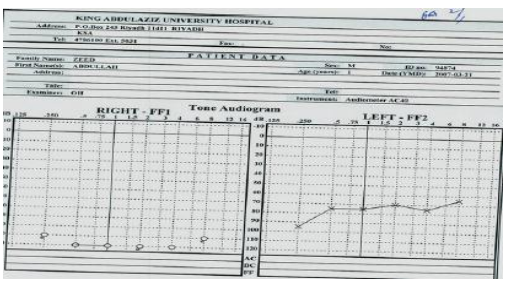




Conductive hearing loss

Mixed hearing loss



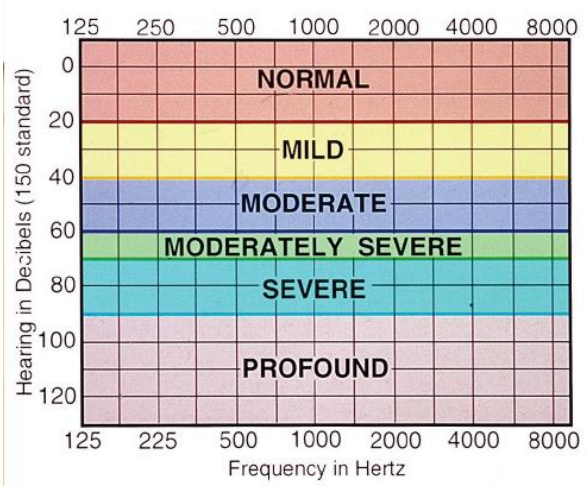


sensorineural hearing

[1] The audiometer is an instrument that generates pure tone signals ranging from 125 to 12 000 Hz (12 kHz) at variable intensities.
[2] Decibels are logarithmic units of relative intensity of sound energy.

- 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.

Degree of hearing impairment:



Management of hearing impairment:

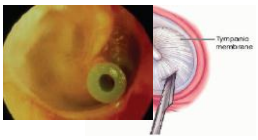
Treat the underlying cause

- Two Types :
  - ➢ Medical
  - ➢ Hearing Aid: they magnify the sound so it reaches the cochlear amplified. Hearing aids are better in sensory rather than neural loss.

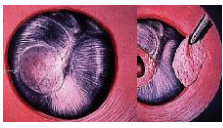
❖ **History:** 1550 by Girolamo Cardano when he saw that sound could be transmitted through the teeth.

○ Surgery:

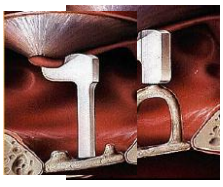
1- Myringoplasty & ventilation tube: in case of otitis media with effusion most common procedure performed (done in a eustachian tube infection) ventilation tube indication: 1-Persistent effusion 2-Eustachian tube dysfunction 3-Recurrent acute otitis media



2- Myringoplasty & tympanoplasty: in case of CSOM.



3- Ossiculoplasty:



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

4-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.

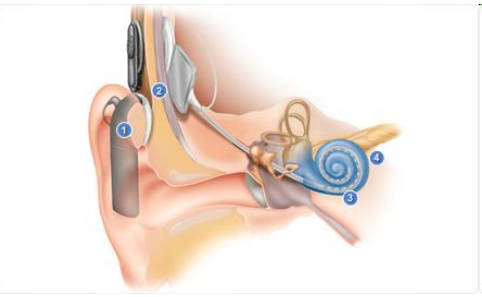
- Atresia of external ear canal. and microtia

- Chronic drainage ear not responding to surgery
- Done for all types of HL (mainly conductive) it skips the middle and internal ear and goes straight to the cochlear and stimulates the bone directly.
- 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.

5- Cochlear implant:

"audiogram shows bilateral profound sensorineural hearing loss can be an indication"

- Putting tiny electrode in the cochlea.
- Prelingual children and postlingual adult
- It by pass the external, middle and inner ear to stimulate the auditory nerve directly.
- In congenital HL the cochlear implant is ineffective after 5 years (The child will be prelingual and the child would've 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.
- 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

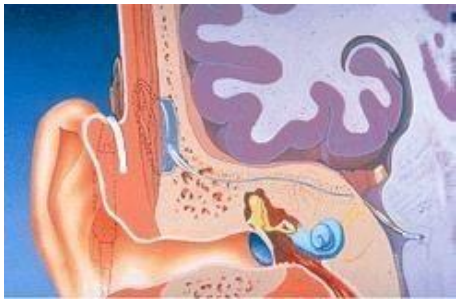
if congenital hearing loss.

- 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 patient has a cochlea and a nerve

prior to implant.

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

  Implant in the brains



- **Lip Reading**
- **Sign Language**