hearing disorders (loss, weakness,deafness,..) and tinnituss

Professor Khader J. Abdulbaqi
MD. PhD

The Auditory system

Gross Brief Anatomy & Physiology

- The auditory system comprises the ears and their connections to and within the central nervous system.
- The auditory system may be divided into

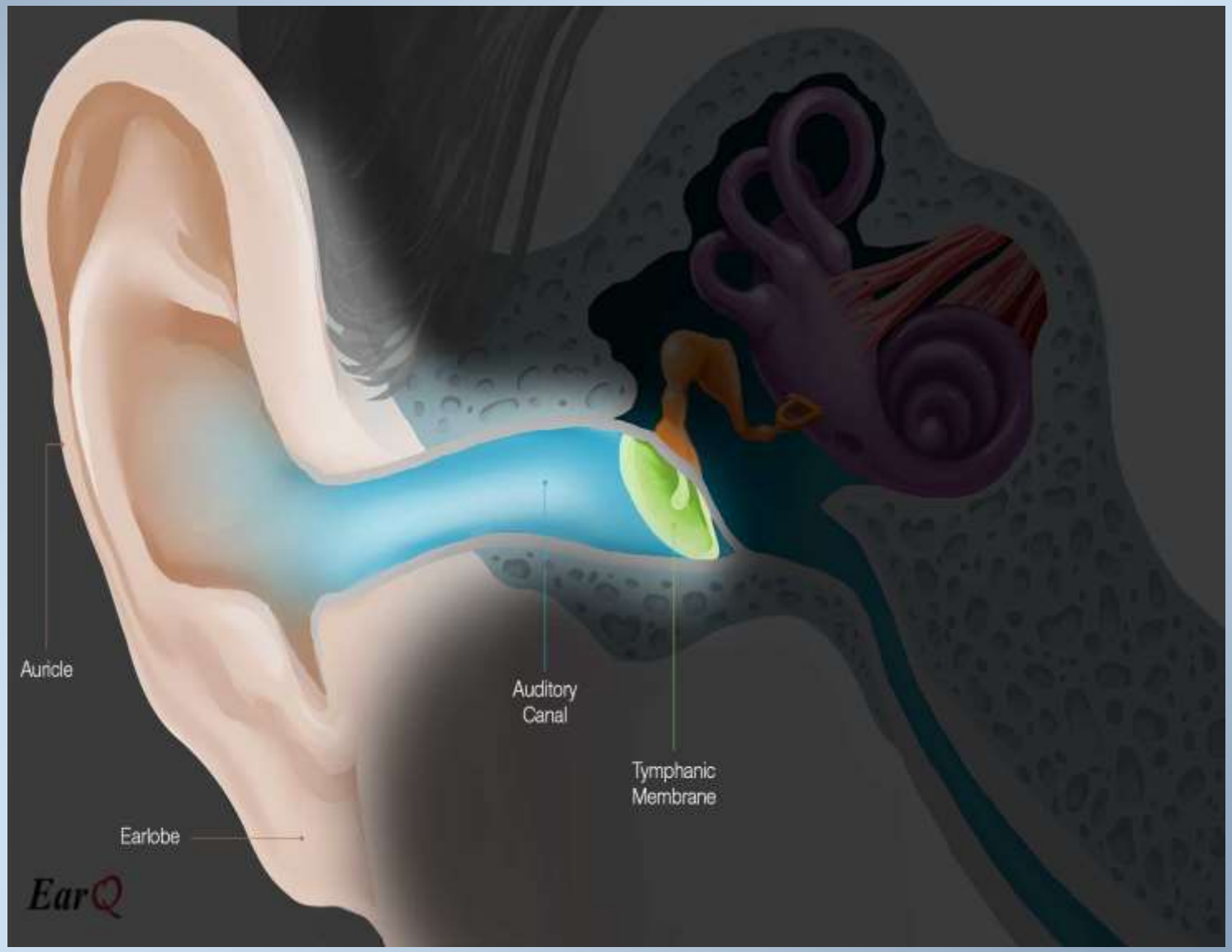
1-Peripheral system

- **Outer ear** (auricle & external auditory canal)
- **Middle ear**(tympanic membrane,cavity,ossicles & mussles)
- **Inner ear**(cochlear &vestibular systems)
- **Vestibulocochlear nerve** (CN VIII)

2-central system

Peripheral system

outer ear



Peripheral system

Middle ear consist of :

1 An air-filled space with a chain of **tiny bones (Ossicles)** :

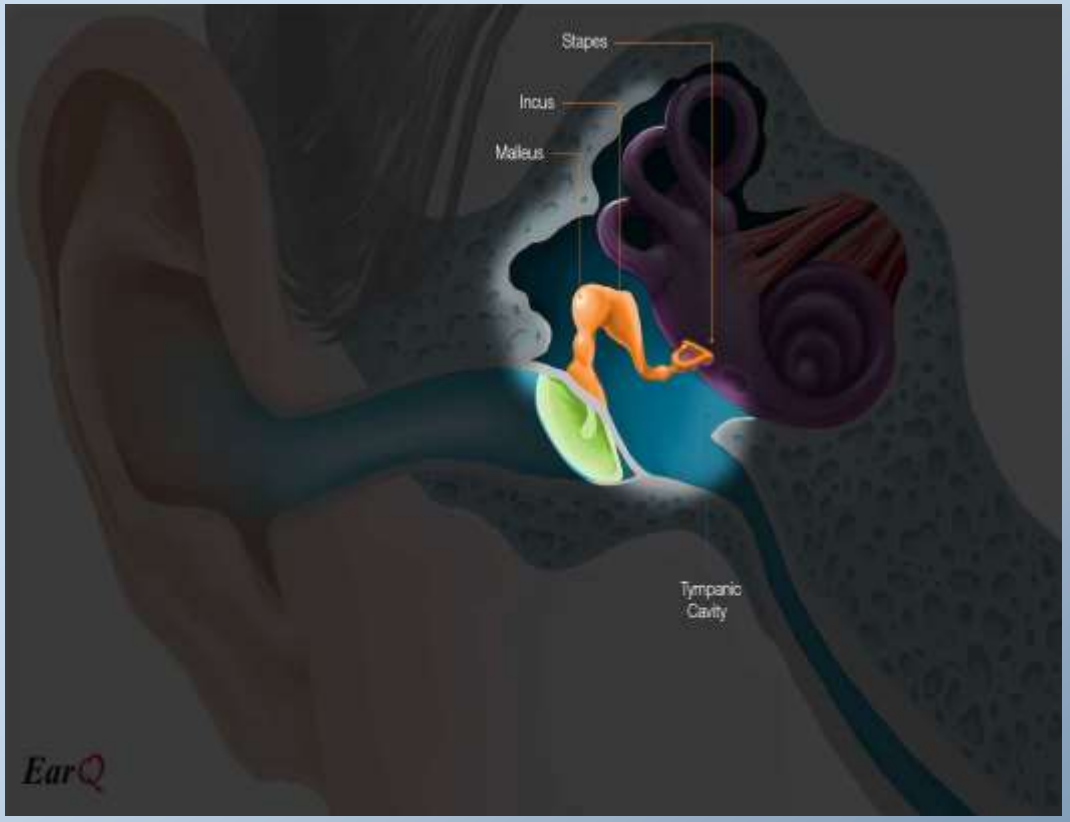
- malleus
- incus
- stapes

2 muscles:

-Tensor tympani

-Stapedius muscle

Eustachian tube



Peripheral system

Inner ear consist of :

.Cochlea : it is filled with fluids : perilymph and endolymph and many microscopic components .

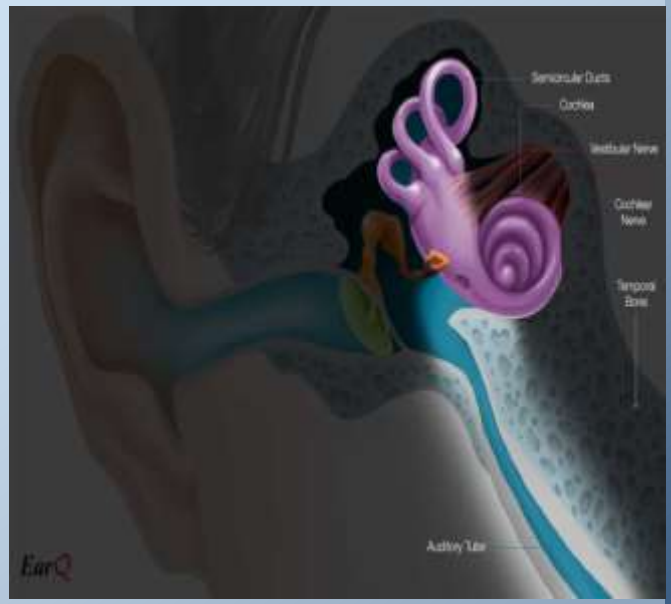
.Vestibular system:

-3 semi-circular canals : lateral or horizontal, superior or anterior, inferior or posterior

-2 otolithic organs(**the utricle and saccule**)

.Auditory and vestibular nerve (VIII CN)

.



Central Auditory system (CAS)

CAS consist of :

- Brainstem
- Cortex (temporal lobe)

BASIC PHYSIOLOGY

The process of hearing needs 3 Requirements :

1-Source of sound(acoustic energy)

2-Medium for transmission.

3-Sensory Receptor Organ { ear }

Acoustic energy transformation

- Acoustic **vibration** (external ear(
- **mechanical** energy (middle ear)
- **hydraulic** energy (perilymph and endolymph of cochlea)
- **electromechanical** energy (hair cells)
- **electrical** energy (VIII nerve)

Etiology(causes)of deafness(hearing loss

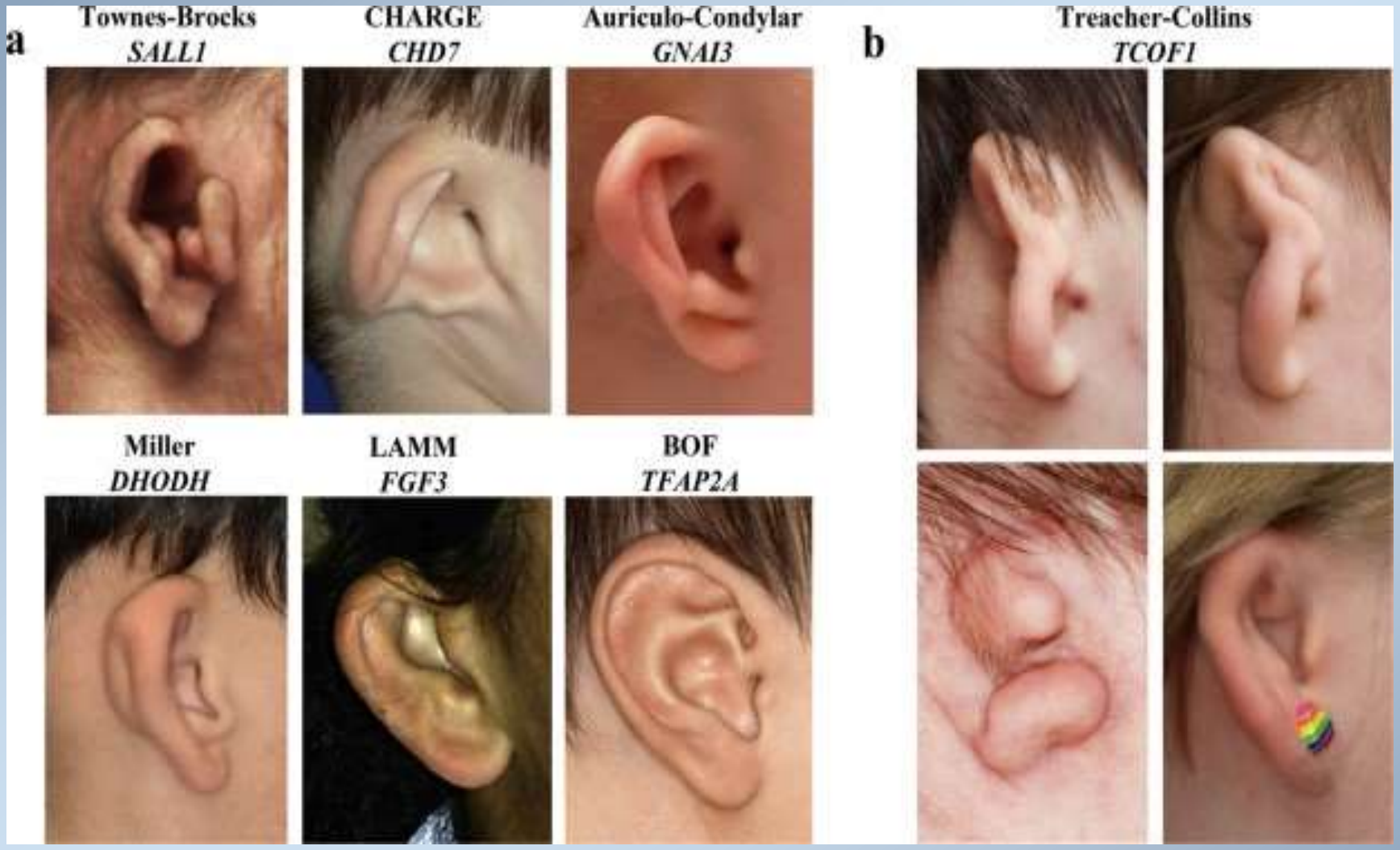
OUTER EAR CAUSES

1. Congenital :

- **Microtia** (the absence or malformation of the auricle) , may cause mild to moderate conductive hearing loss.

- **Atresia** or significant **stenosis** of the EAC causes moderate to maximal conductive hearing loss.

  maximum ABG 55-65 dBs.


a
Townes-Brocks
SALL1
CHARGE
CHD7
Auriculo-Condylar
GNAI3
Miller
DHODH
LAMM
FGF3
BOF
TFAP2A
b
Treacher-Collins
TCOF1

2. **Infection**:Infections may lead to blockage of the EAC due to the accumulation of debris, edema, or inflammation.
3. **Tumor**: the most common malignant tumor of the EAC is SCC, others basal cell carcinoma and melanoma, typically cause conductive hearing loss due to occlusion of the canal.
4. **Benign bony growths** may also occlude the EAC with a resulting conductive hearing loss. The two most common benign growths are **exostosis or osteoma**.

5 .Wax impaction : Some patients are not able to clear wax on their own or use Q-tips that push the wax mediallyto the to the TM. These individuals may need periodic cleaning of the wax to enhance their auditory capabilities.

6.Tympanosclerosis:

caused by previous infection or insertion of ventilation tubes result in white or yellow scarring within the tympanic membrane.

usually It does not cause a significant hearing loss.

Could affect ossicles & tendons

Rarely causes scarring and fixation of the ossicles, with a conductive hearing loss.

MIDDLE EAR CAUSES

1-Congenital :

- **atresia or malformation** of the ossicular chain can cause conductive hearing loss.

- the most common ossicular abnormality observed is a missing or malalignment of the crura of the stapes. others include abnormal incus or malleoincudal joint.

2.Infections:

Acute otitis media(AOM) :acute or chronic

- otitis media is a common childhood disorder that also frequently occurs in adults.
- 1-non-infective:OME (with effusion)
- 2-infective:APOM (Purulent)

It is normally associated with pain, fever, and ear fullness as well as decreased hearing.

- Conductive hearing loss is usually slight to mild
- The chronic OM causes more HL according to severity

3.Tympanic membrane perforation:

- The degree of conductive hearing loss depends upon the size and location of the perforation.

- can be caused by :
  - blast injury
  - barotrauma.during rapid decent or diving
  - foreign body trauma
  - temporal bone fractures
  - ear infections
  - self-inflicted trauma from a Q-tip or other object

Other causes in ME:

- **Tumors :** cholesteatoma :desquamated stratified squamous epithelium in M.E. space . usually found in CSOM . Erodes surrounding bones.

-**Otosclerosis :** bony growthon foot plate of the stapes. AD inheritance. Usually unilateral in young females (M:F=1-2) .ttt either HAs or Stapedectomy.

-Austacian tube dysfunction (C-type tymp) or obstruction (B-type tympanogram).

INNER EAR CAUSES

1.congenital :

-hearing loss that occurs at or shortly after birth.

-Hereditary :autosomal dominant or recessive;

usually autosomal recessive(90%)

-Acquired (Non-hereditary): involve an insult to the developing cochlea, including viral infections such as maternal rubella, cytomegalovirus (CMV), hepatitis, toxoplasmosis, HIV, and syphilis.

- **2-Presbycusis** :
- **age-related hearing loss**, is the most common cause of hearing loss in elrely.
- Is progressive, symmetric loss of high-frequencies
- Tinnitus usually present.
- Using Hearing Aids are usually successful in most patients with presbycusis,

3.Infection :

-The most common infection of the inner ear in **adults**

is viral cochleitis with sudden SNHL**,** vertigo, facial p paralysis, & pain occur rarely.

It is **meningitis** in young children .

- cause a profound sensorineural hearing loss affecting the ear hair cells

4- **Meniere disease**

characterized by triad episodes of fluctuating deafness, tinnitus, and vertigo. Typically the attacks are preceded by a sensation of fullness in the ear.

5.Noise exposure: either **:**

-NIHL. 8 hours daily exposure to noise more than 85 dBs in industrial works.

-Acoustic trauma:Sudden exposure to high intensity sound briefly

6-Tumors :

- The most common benign tumor that causes sensorineural hearing loss is an **acoustic neuroma (vestibular schwanoma) .**

- Originates from the vestibular portion of the eighth cranial nerve.

- The most common complaint is an asymmetric or unilateral sensorineural hearing loss and **tinnitus**.

7-Ototoxicity

- **Antibiotics** : (oral aminoglycosides , erythromycin ,gentamycin and tetracycline …)

- **chemotherapeutic agents** (cisplatin) are among the most commonly used drugs that cause hearing loss.
- Duretics
- Asperin ----reversible SNHL

- Any sensorineural hearing loss associated with antibiotic or chemotherapeutic drugs is permanent.

8-Neurogenic

- Cerebrovascular accident (CVA) or transient ischemic attack (TIA)
- Arnold-Chiari malformations may stretch the auditory vestibular nerve,
- Multiple sclerosis

9-Iatrogenic

- Iatrogenic inner ear injuries may occur during surgical procedures such as : **tympanomastoidectomy or stapedectomy**; following
- **radiotherapy**, either for intracranial or nasopharyngeal tumors; or they may be medication-related.

DEGREE OF HEARING LOSS

- Normally up to 25 db in adults and 15 in a child.
- Mild 25-40 db
- moderate 41-55 db
- Moderately severe 56-70 db
- Severe 71-85 db
- Profound >85 db

DIAGNOSIS

1-Thorough Proper detailed & complete History gives 90% of correct Diagnosis

2-examination: observation, ENT,General,TFTs

- Otoscopy,

3- Investigations :

- Audiogram
- tympanogram
- **if needed:speech :speech** audiometry **vestibular** tests

TREATMENT

-**Congenital : counseling** ,

-Underlying causes **: Medical &/or surgical**

- **Hearing aids** ,

-Speech therapy & special education programs ,

- **Suddenloss: corticosteroids**

Sound pathways or mechanisims of hearing

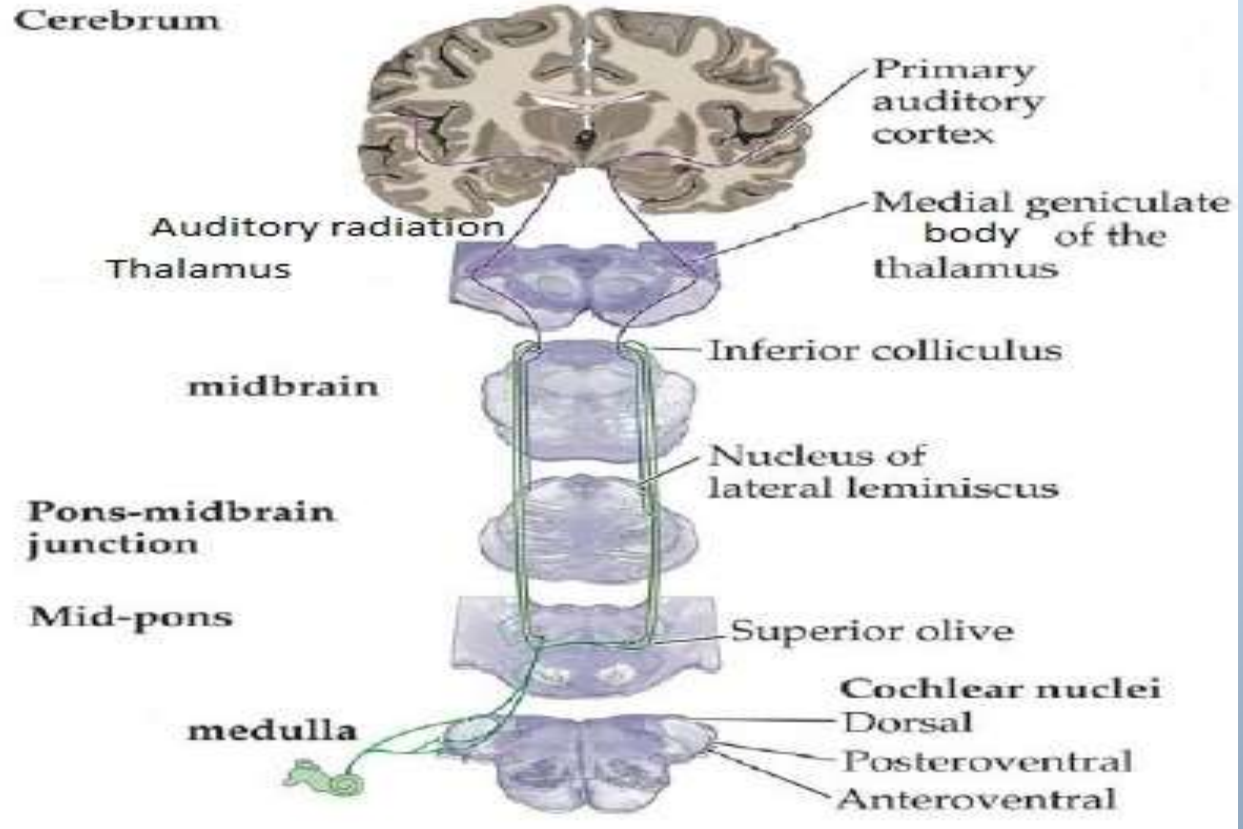
The sound is transmitted through two ways :

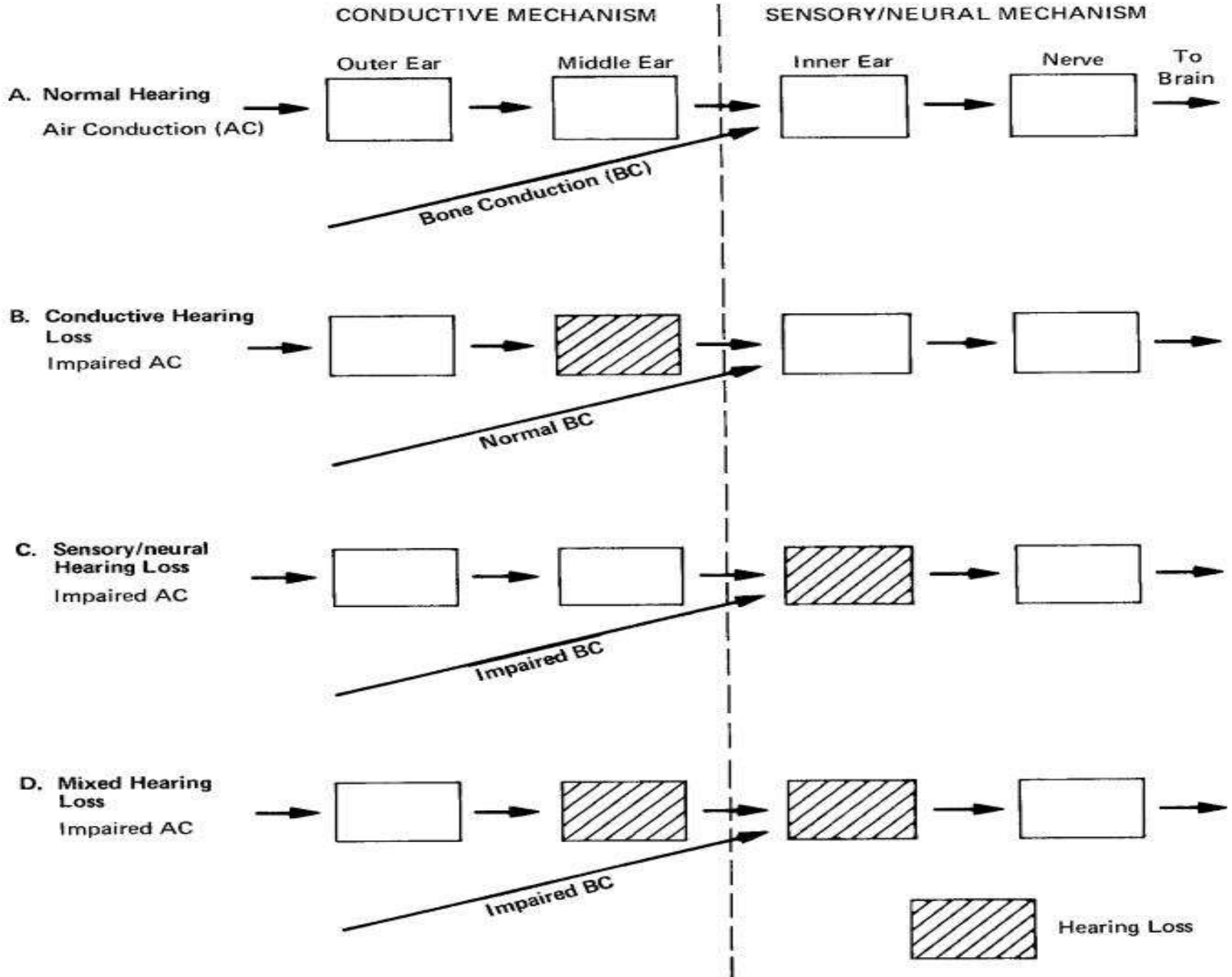
- **Air conduction** : In this pathway the sound passes through outer, middle and inner ear up to brainstem and auditory cortex in temporal lobe.
- **Bone conduction** : in this pathway the sound passes directly to the inner ear and up….
- Most sounds are heard via air conduction.

Cont.

- The sound passes through **outer, middle**, **inner** ears & through the **auditory nerve** to **brainstem stations** :upper medulla, pons and midbrains, (cochlear necleus **CN** , superior olivary comlex **SOC**, lateral leminiscus **LL** , inferior colliculus **IC**) to **medial geniculate body** in the thalamus and proceed as auditory radiations to the primary **auditory cortex** in temporal lobe .

The Primary Central Auditory Pathway:




CONDUCTIVE MECHANISM
SENSORY/NEURAL MECHANISM
Outer Ear
Middle Ear
Inner Ear
Nerve
To Brain
A. Normal Hearing
Air Conduction (AC)
Bone Conduction (BC)
B. Conductive Hearing Loss
Impaired AC
Normal BC
C. Sensory/neural Hearing Loss
Impaired AC
Impaired BC
D. Mixed Hearing Loss
Impaired AC
Impaired BC
Hearing Loss

Types of hearing loss

- **Conductive** Hearing Loss
- **Sensory/Neural** Hearing Loss
- **Mixed** Hearing Loss
- **Functional** or **nonorganic** Hearing Loss(**fake**)
- **Central and psychogenic** Hearing Loss : occurs due to insults or damage in brainstem or cortex, or both.

DEGREE OF HEARING LOSS

- Normally up to 25 db in adults and 15 in a child.
- Mild 25-40 db
- moderate 41-55 db
- Moderately severe 56-70 db
- Severe 71-85 db
- Profound >85 db

simple clinical tests
(Tuning Fork Tests)

- The tuning fork is a tool, usually made of steel, magnesium, or aluminum, that is used to test hearing in the clinic
- When the tuning fork is set in vibration properly, the tines move alternately away from and toward one another, and the stem moves with a piston action
- The tuning fork is set into vibration by holding the stem in the hand and striking one of the tines against a firm but resilient surface, many physicians prefer the **knuckle, knee, or elbow**… **application in clinic**
- By using tuning forks with various known properties, any diagnostic statement made on the basis of a tuning-fork test is absolutely limited to the pitch of the fork used because hearing sensitivity is often different for different pitches. **application in clinic with dB SPL, HL, SL**

Tuning Fork Tests

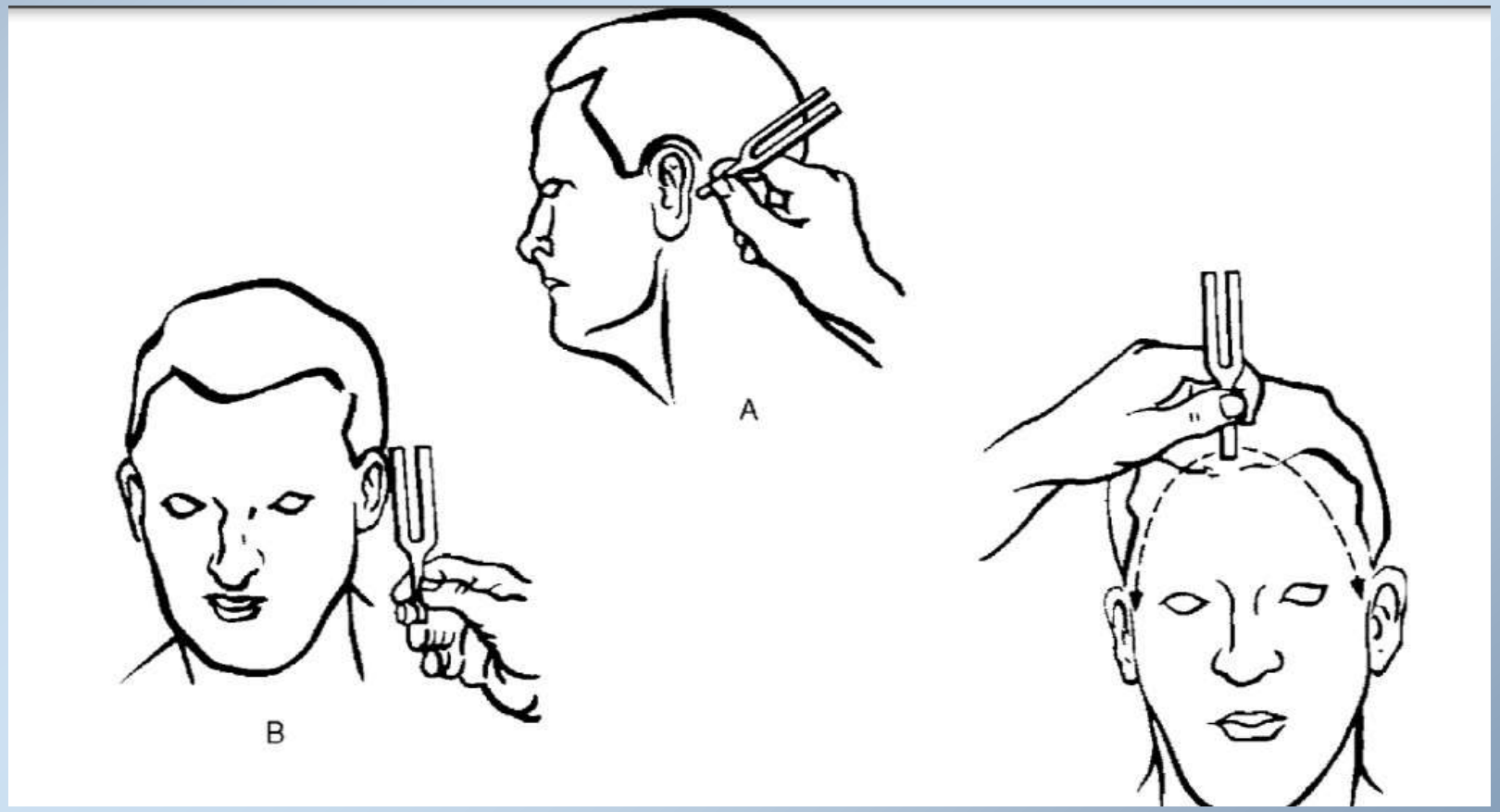
FIGURE 2.3 Several tuning forks. The larger forks vibrate at lower frequencies (produce lower-pitched tones) than the smaller forks.

FIGURE 2.4 Vibration pattern of tuning forks.

Tunning Fork Tests.

Three things should be done :

1-how we perform the test
2-how we report results and
3- how we interpret resuls



Tuning Fork Tests

see in clinic

Tuning fork tests

1. The Schwabach Test
2. The Rinne Test
3. The Bing Test
4. The Weber Test

The Schwabach Test

- The Schwabach test, introduced in 1890, is a **bone-conduction test.** It compares the hearing sensitivity of a patient and examiner.
- The tuning fork is set into vibration, and the stem is placed alternately against the **mastoid process** of the patient and of the examiner.
- This test assumes that examiners have normal hearing
- Each time the fork is placed on the patient's mastoid process, the patient indicates whether the tone is heard
- The vibratory energy of the tines of the fork decreases over time.

The Schwabach Test

- When the patient no longer hears the tone, the examiner immediately places the stem of the tuning fork behind his mastoid process and, using a watch, notes the number of seconds that the tone is audible after the patient stops hearing it.
- If both examiners and patients have normal hearing, both will stop hearing the tone emitted by the fork at approximately the same time (**normal Schwabach**).
- patients with **sensory/neural hearing loss**, they will stop hearing the sound much sooner than the examiner ***(diminished Schwabach).***
- The test can be quantified to some degree by recording the number of seconds an examiner continues to hear the tone after a patient has stopped hearing it. If **an examiner** hears the tone for 10 seconds longer than a patient, the patient's hearing is **"diminished 10 seconds."** In some **conductive hearing losses**, the patient's hearing in the low-pitch range may appear to be better than normal (**prolonged Schwabach).**
- If patients have a **conductive hearing loss**, bone conduction is normal, and they will hear the tone for at least as **long as the examiner, and sometimes longer**.

The Rinne Test

- The **Rinne test** compares patients' hearing sensitivity by bone conduction to their sensitivity by air conduction.
- This is done by asking the patient to state whether the tone is louder when the tuning-fork stem is held against the bone **behind the ear**, as in the Schwabach test, or when the tines of the fork that are generating an air-conducted sound are held **next to the opening of the ear**

The Rinne Test

- Because air conduction is a **more efficient means** of sound transmission to the inner ear than bone conduction, people with **normal hearing** will hear a louder tone when the fork is next to the ear than when it is behind the ear. This is called a **positive Rinne**. *A positive Rinne also occurs in patients with* ***sensory/neural hearing*** **loss**.
- The attenuation produced by a problem in the sensory/neural mechanism **produces the same degree of loss** by air conduction as by bone conduction.

The Rinne Test

- If patients have **more than a mild conductive hearing loss**, their bone-conduction hearing is normal, and they will hear **a louder** tone with the stem of the fork behind the ear (bone conduction) than with the tines at the ear (air conduction). This is called a ***negative Rinne.***
- Sometimes patients manifest what has been called the **false negative Rinne**, *which occurs* when the inner ear not deliberately being tested responds to the tone, this may happen readily during bone-conduction tests.

The Bing test

- when persons with normal hearing close off the opening of the ear canal, the loudness of a tone presented by bone conduction increases **(occlusion effect)**, and it is observed primarily for low-pitched sounds.
- The test : is used to see if there is conductive hearing loss or not by using bone conduction testing.

The Bing Test

- When performing the Bing test, the tuning fork handle is held to the mastoid process behind the ear while the examiner alternately closes and opens the ear canal with a finger.
- People with normal hearing and those with sensory/neural hearing loss, the result is a pulsating sound, or a sound that seems to get louder and softer **(a positive Bing)**
- patients with conductive hearing losses, no change in the loudness of the sound is noticed **(negative Bing).**
- Absence of conductive hearing loss **(positive Bing)**
- Presence of conductive hearing loss **(negative Bing).**
- The examiner must not suggest to patients what their responses should be.

The Weber Test

- It is a test of **lateralization;** that is, patients must state where they hear the tone (left ear, right ear, both ears, or midline).
- When performing the Weber test, the tuning fork is set into vibration, and the stem is placed on the midline of the patient's skull, Other sites are also used, such as the top or the back of the head, the chin, or the upper teeth(the loudest bone conducted sound)
- People with **normal hearing** or with **equal amounts of the same type of hearing loss** in both ears will report a **midline sensation**

The Weber Test

- Patients with sensory/neural hearing loss in one ear will hear the tone in their better ear , and the tone will be softer or will not be perceived at all in the poorer ear

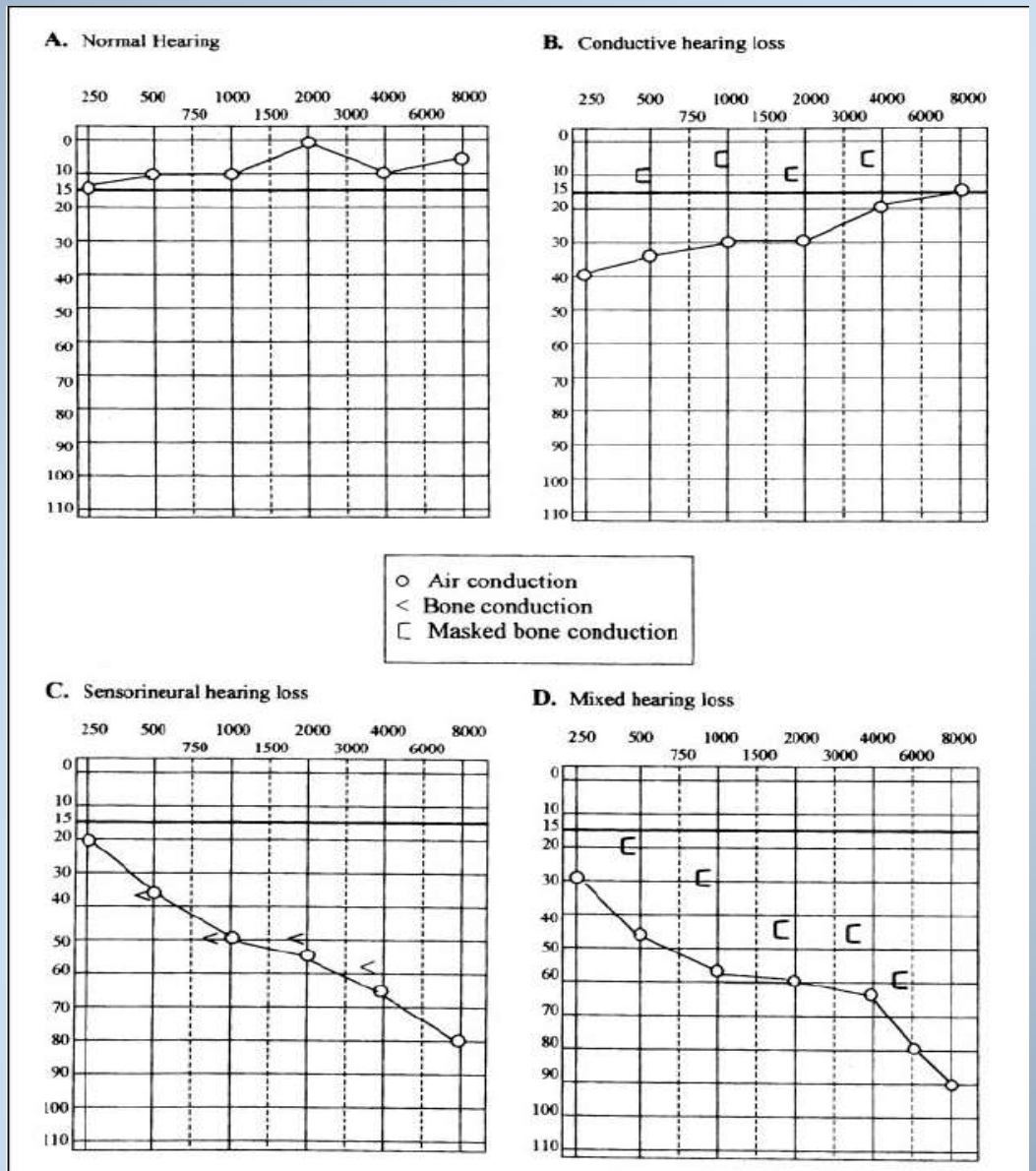
- Patients with conductive hearing loss in one ear will hear the tone in their poorer ear.
- One explanation of the Weber effect in sensory/neural cases is based on the Stenger effect.
- The **Stenger principle** states that if two tones that are identical in all ways except loudness are introduced simultaneously into both ears, only the louder tone will be perceived.

The Weber Test

- The Weber test has been known to avert misdiagnosis of unilateral sensory/neural hearing loss as conductive when false normal Schwabach or false negative Rinne results are seen, but the tone is heard in the poorer-hearing ear rather than the expected better-hearing ear
- Interpretation of the Weber test is also difficult in mixed hearing losses.

Test	*Purpose*	*Fork Placement*	*Normal Hearing*	*Conductive Loss*	*Sensory/neural Loss*
Schwabach	Compare patients BC to normal	Mastoid process	*Normal Schwabach* - Patient hears tone as long as examiner	*Normal or Prolonged Schwabach* - Patient hears tone as long as or longer than examiner	*Diminished Schwabach* - Patient hears tone for shorter time than examiner
Rinne	Compare patient's AC to BC	Alternately mastoid process and opening to ear canal	*Positive Rinne* - Louder at the ear	*Negative Rinne* - Louder behind the ear	*Positive Rinne* - Louder at the ear
Bing	Determine presence or absence of occlusion effect	Mastoid process	*Positive Bing* - Tone is louder with ear occluded	*Negative Bing* - Loudness does not change with ear occluded	*Positive Bing* - Tone is louder with ear occluded
Weber	Check lateralization of tone in unilateral losses	Midline of head	Tone equally loud in both ears	Tone louder in poorer ear	Tone louder in better ear

Pure tones Audiometry Audiogram



A-CLN 0285

Jordan University Hospital

Otolaryngology — Audiology clinic

Date : .....

Audiometric Evaluation

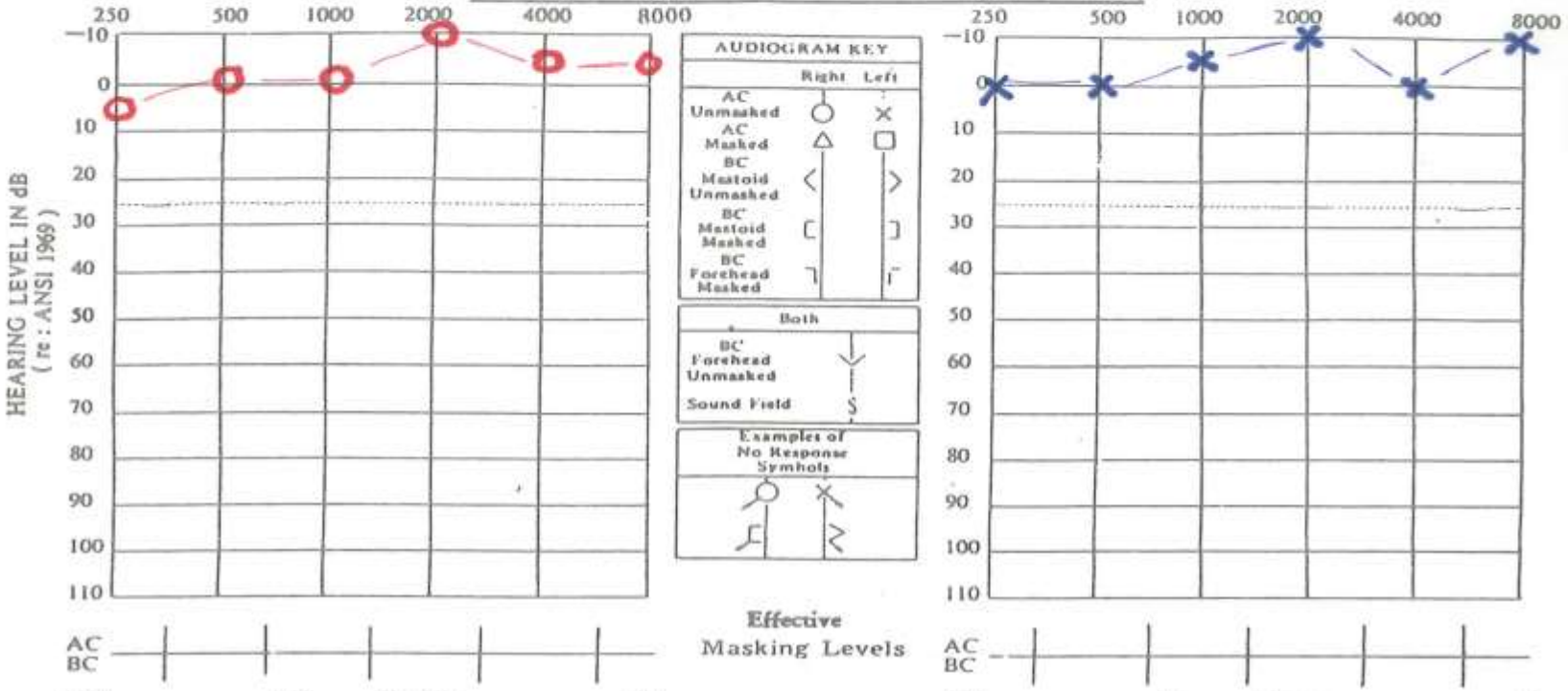
Name .. .. Age ..... Sex ..... Case No. .....

Referring Dr: Technician : Consultant :

RIGHT EAR
Frequency in Hz

Response consistency : good, moderate, poor.
Validity : acceptable, questionable.

LEFT EAR
Frequency in Hz



SRT _____ dB AC Ave. _____ dB

Speech Discrim. _____ % at _____ dB SL

_____ % at _____ dB _____ % at _____ dB SL

MCL : UCL :

	ACOUSTIC REFLEX 500	1000	2000	REFLEX DECAY 500	1000	2000
Contra.						
Ipsi						

SRT _____ dB AC Ave. _____ dB

Speech Discrim. _____ % at _____ dB SL

_____ % at _____ dB _____ % at _____ dB SL

MCL : UCL :

	ACOUSTIC REFLEX 500	1000	2000	REFLEX DECAY 500	1000	2000
Contra.						
Ipsi						

Special Populations

- **AGING**

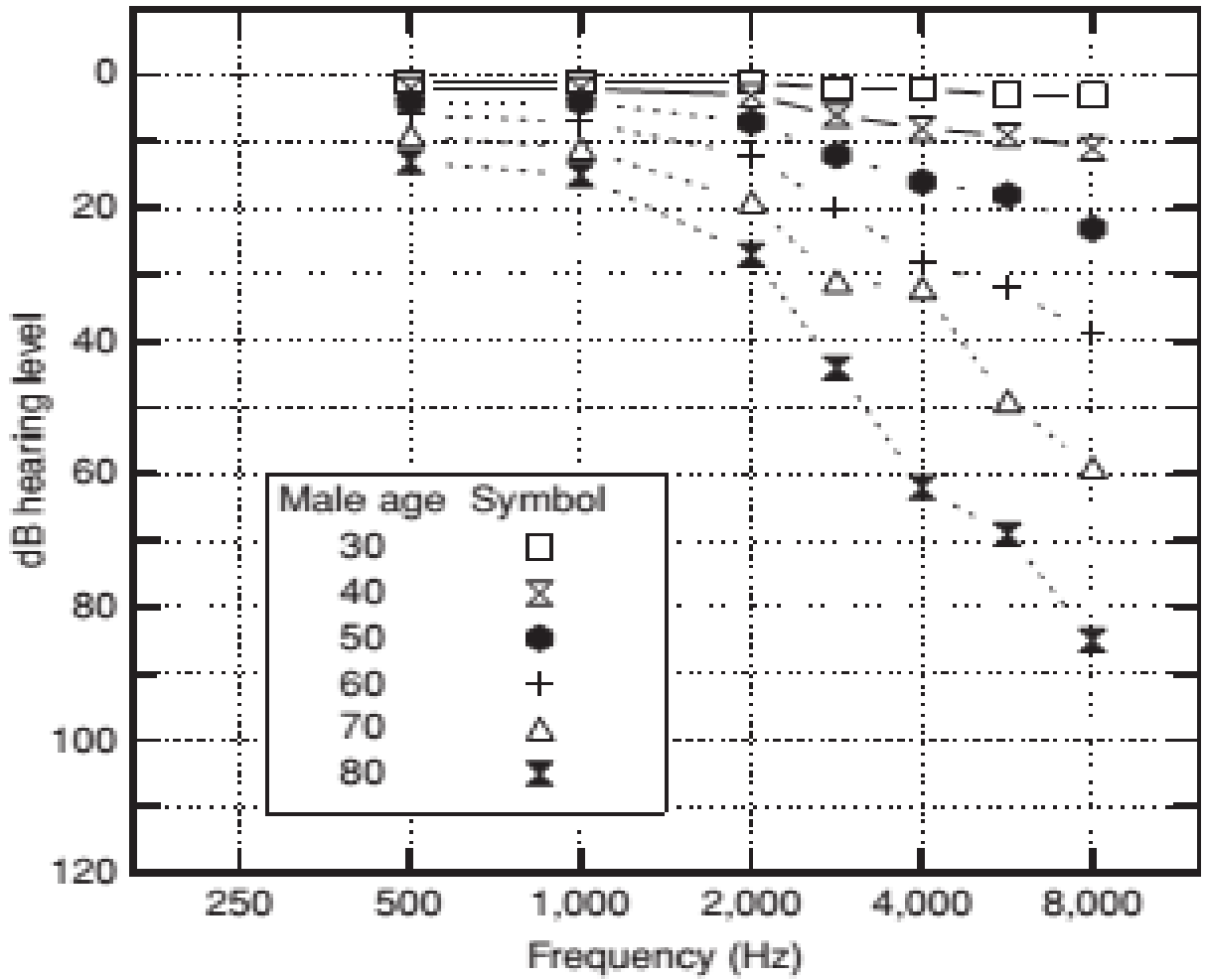
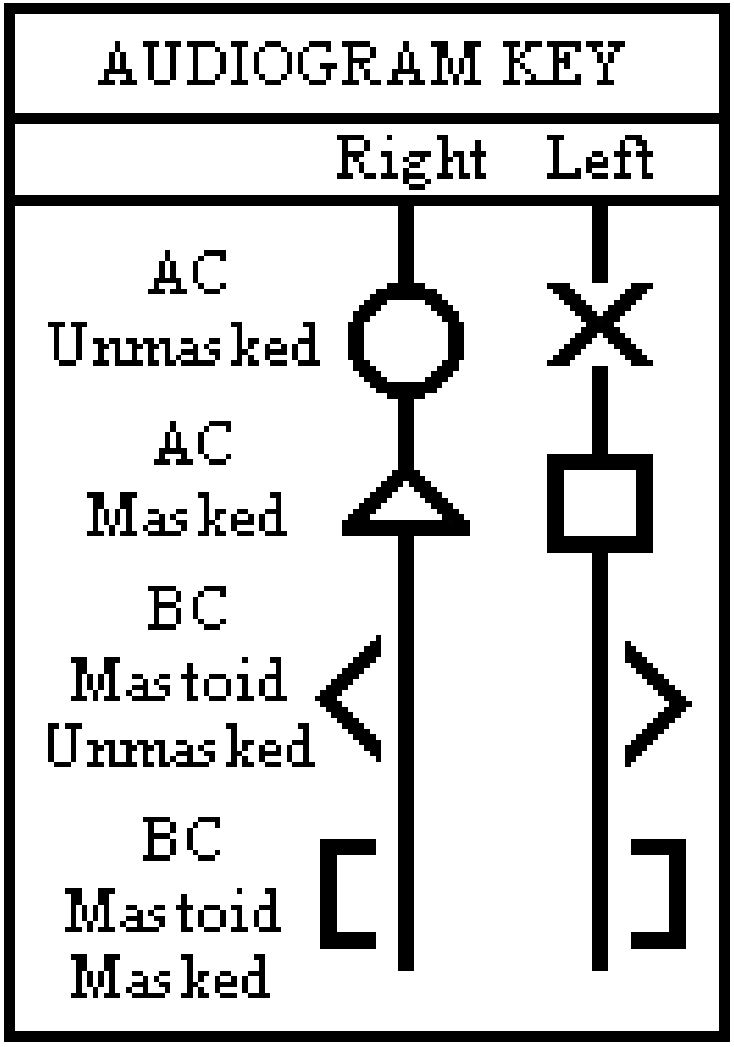
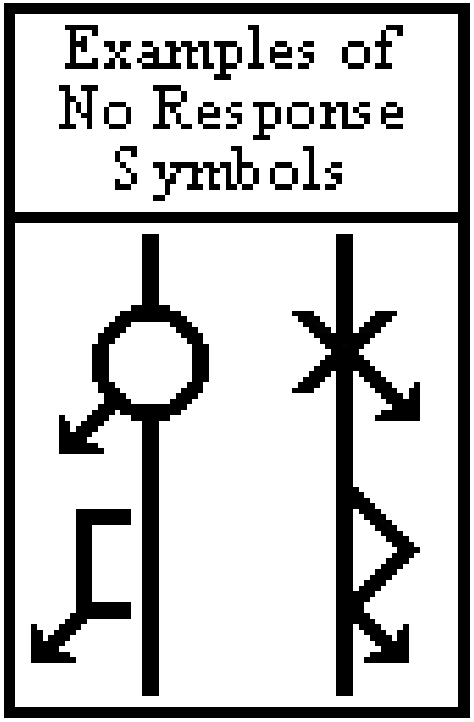
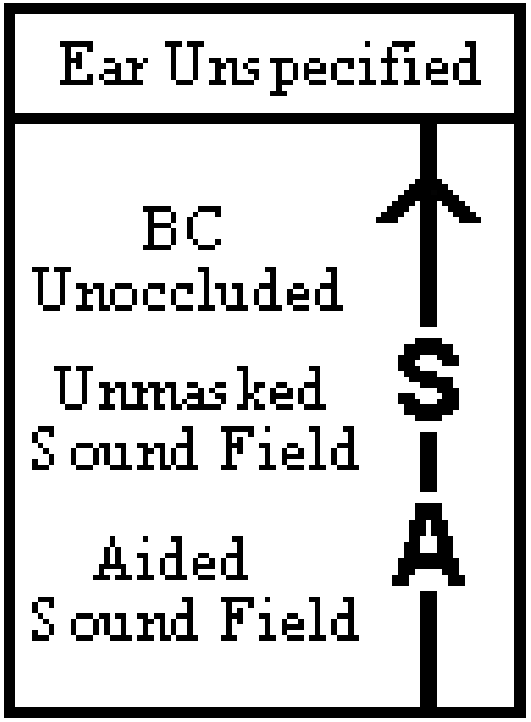


FIGURE 3.11 Average audiograms for adult males for different decades of life. Data from National Institute on Deafness and Other Communication Disorders [2005].







TEST TECHNIQUE: Conventional

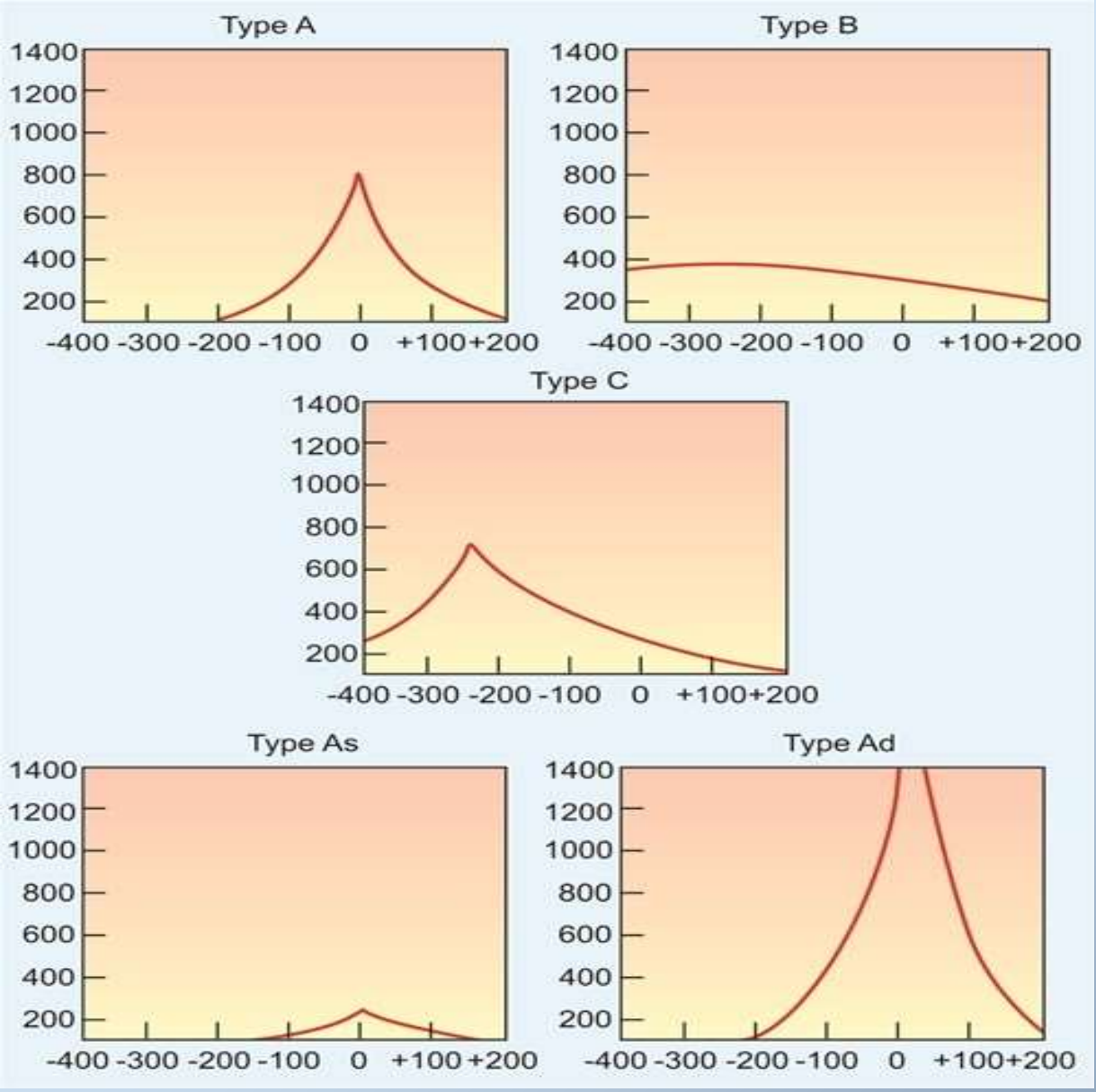
BOA VRA TROCA CPA

EARPHONE: TDH-49 TDH-50 ER3-A

RELIABILITY: good fair poor

VALIDITY: acceptable questionable

Tympanometry



TINNITUS

- defined as the aberrant perception of sound without any external stimulation, It is thought to be due to misinterpretation of signals in the central auditory pathways of the brain.

- it consists of an intermittent or continuous ringing, hissing or buzzing noise and it may be low, medium or high-pitched.

- Quality of tinnitus varies according to origin:
  high pitch (inner ear) / crackling (middle ear) /pulsating(vascular).

- it can be unilateral or bilateral.

- It is not a disease but a symptom.

- described as either subjective or objective:

¾ Subjective tinnitus: the most common type, occurs in the absence of any physical sound reaching the ear and is audible only to the patient.

¾ Objective tinnitus, which affects a minority of patients(1%),is generated in the body and reaches the ear through conduction in body tissue and is audible to the patient as well as the clinician.

- Objective: rare

causes:

¾ palatal myoclonus, middle ear myoclonus

¾ vascular (aneurysm, glomus tumor, AV shunt , atheroma of cranial vessels).

¾ acute middle ear infection

- Subjective: very common

Causes:

- acute trauma
- Meniere's disease
- ototoxicity , otosclerosis
- acoustic neuroma
- systemic disease: CVS disease,blood diseases (anemia), neurological (MS, neuropathy), drugs, fever, alcohol abuse.
- middle ear effusion and chronic otitis
- psychogenic(hallucinations)
- labyrinthitis, perilymph fistula
- idiopathic (majority)

DIAGNOSIS

- proper hx taking:
  localization , pitch , duration.
  audiological : deafness ,noise exposure
  Otological : discharge
  drug hx

- examination: ENT , neck ,TMJ, auscultation

- investigations:
  audiometry, tympanometry ,vestibulometry.
  Tinnitus test (pure tone matching)
  blood test (T3, T4 , lipid profile,…)
  radiograms(x-ray , CT, MRA)

TREATMENT

- counseling: informing the patients about the factors aggravating tinnitus which include : fatigue, anxiety, stress, depression.

- underlying cause: remove wax, tumors or aneurysms, control HTN,…

- Drugs : muscle relaxants, Antidepressants, tranquilizers, anticonvulsants.

- surgery: labyrinthectomy
- hearing aids.(if the patient is deaf)
- tinnitus maskers (white noise instrument), by producing quite noise .
- Tinnitus retraining therapy (TRT)

referance

- Introduction to audiology by Frederick.N. Martin &J. G. Clark . 11th edition.

- For more details look to folder :

Gross Anatomy and Physiology of Hearing

Muchas Gracias
Thank you