



Clinical Audiology

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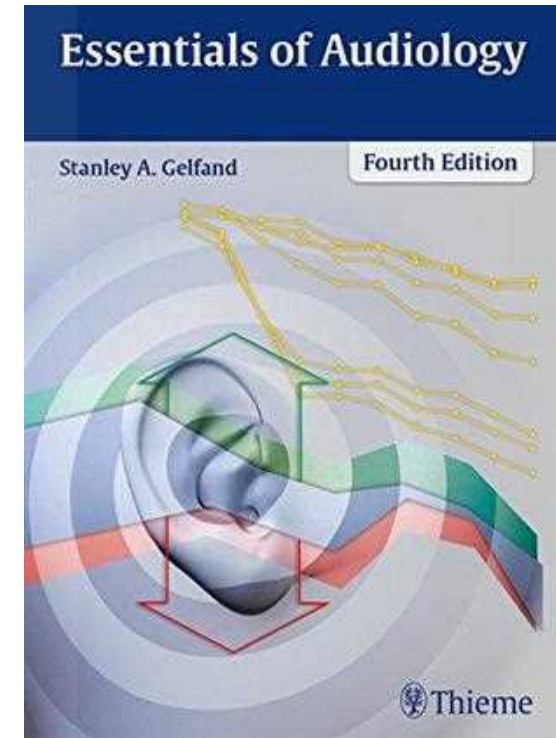
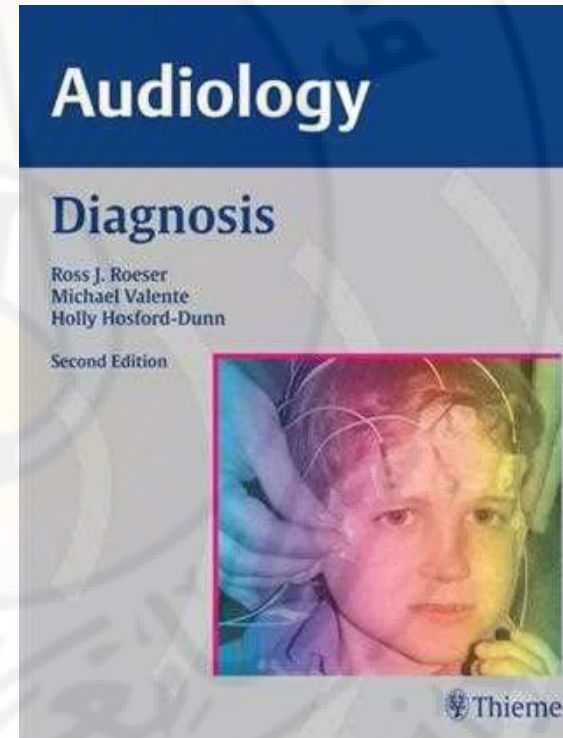
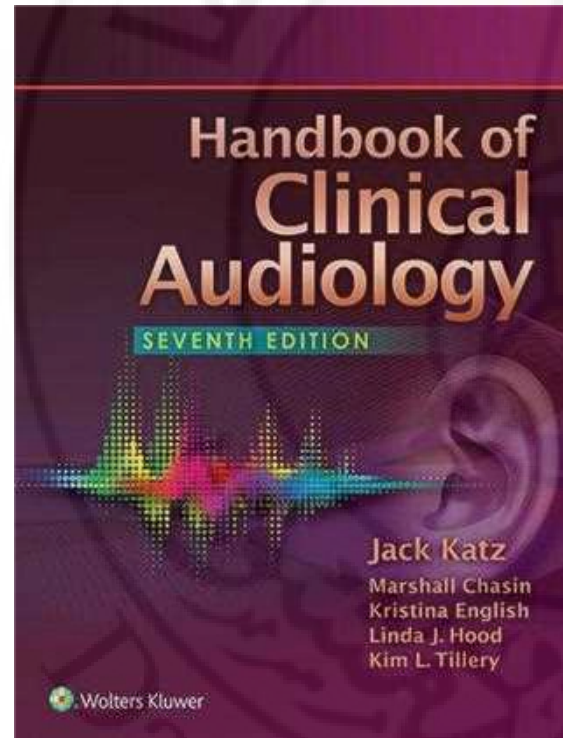
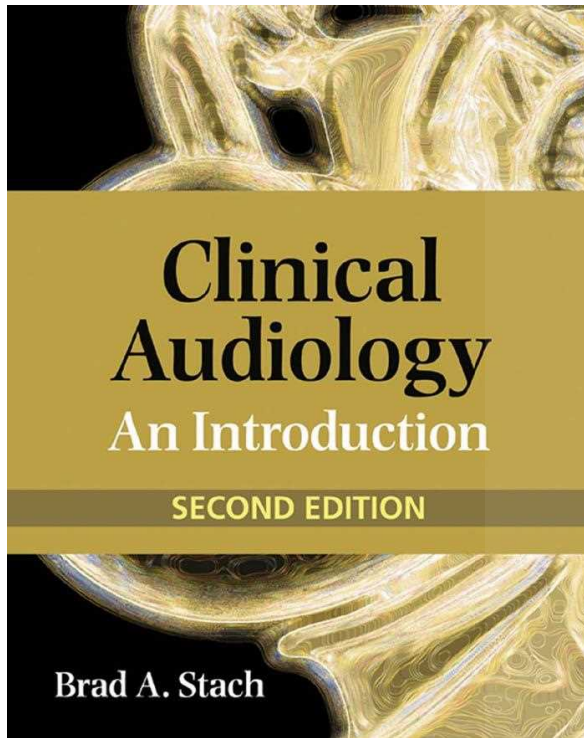
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Main Topics

- The Profession of Audiology
- Basic evaluation and test battery approach
 - Clinical Process
 - Case History
 - Physical examination
 - Tuning Forks examination
- Otologic considerations in clinical audiology
 - Outer ear pathologies
 - Middle ear pathologies
 - Inner ear pathologies
- Neurologic considerations in clinical audiology
- General Medical considerations in clinical audiology
- Audiologic test battery approach



References



E. references

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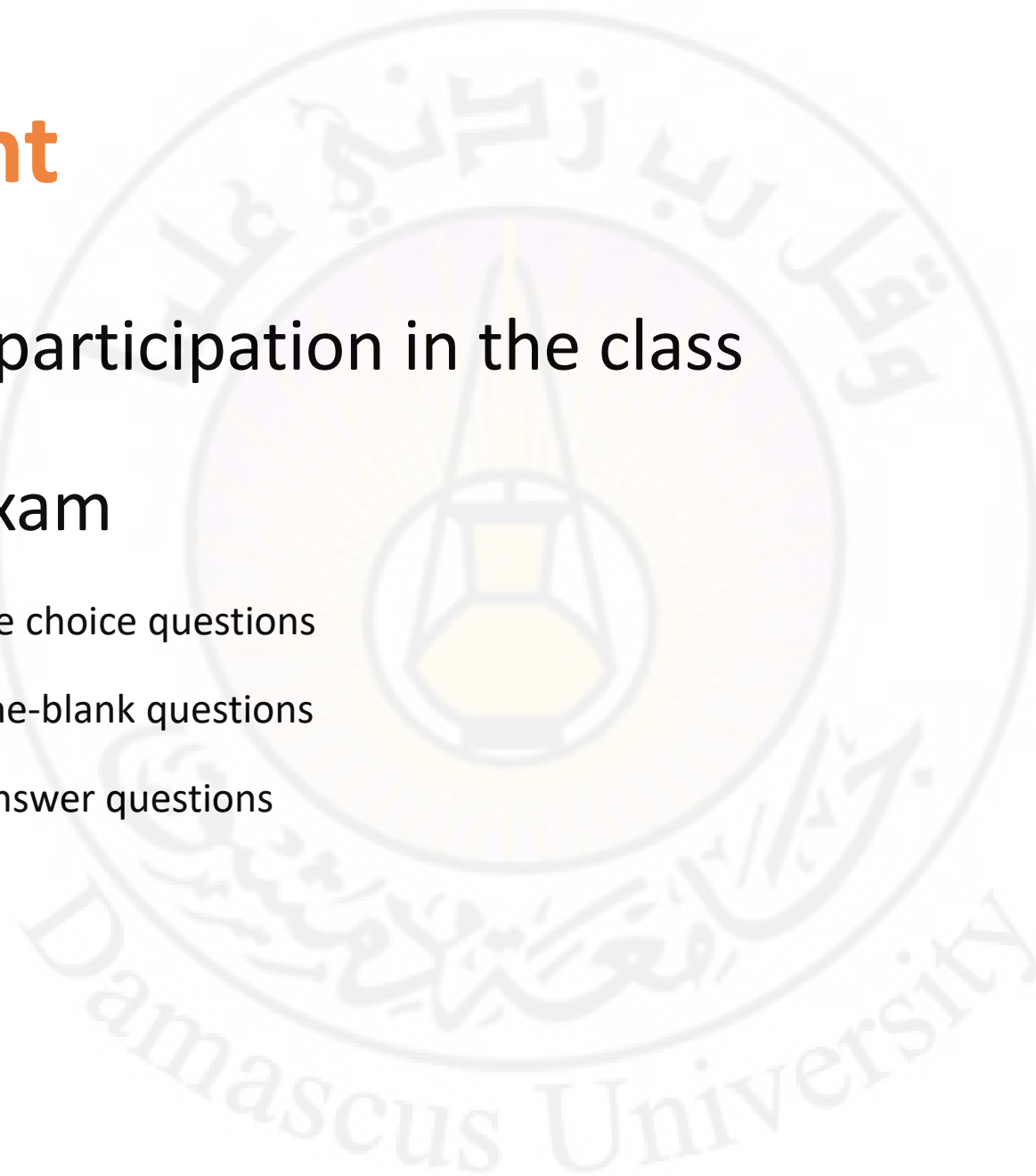
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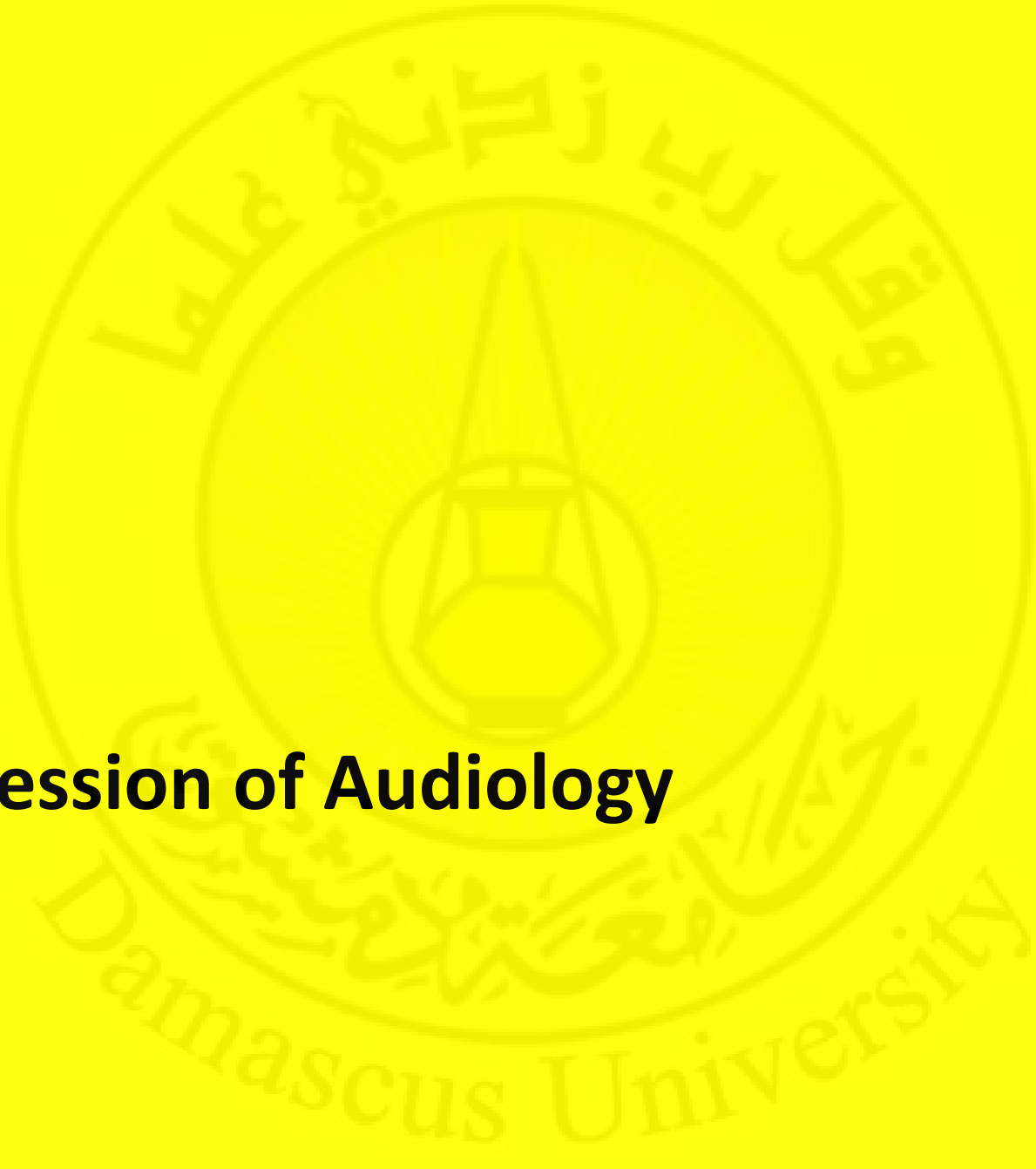
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Assessment

- Active participation in the class
- Final exam
 - Multiple choice questions
 - fill-in-the-blank questions
 - short answer questions





1. The Profession of Audiology

Audiology!

- **Audiology** is the study of hearing and hearing disorders, a field devoted to helping those with auditory and vestibular dysfunctions.
- This work may involve evaluation, re/habilitation, counseling, education, research, and/or screening/ prevention.



What is an Audiologist?

American Academy of Audiology (AAA)

An audiologist is a person who, by virtue of academic degree, clinical training, and license to practice and/or professional credential, is uniquely qualified to provide a comprehensive array of professional services related to the **prevention** of hearing loss and the audiologic **identification, assessment, diagnosis, and treatment** of persons with impairment of auditory and vestibular function, and to the prevention of impairments associated with them.”

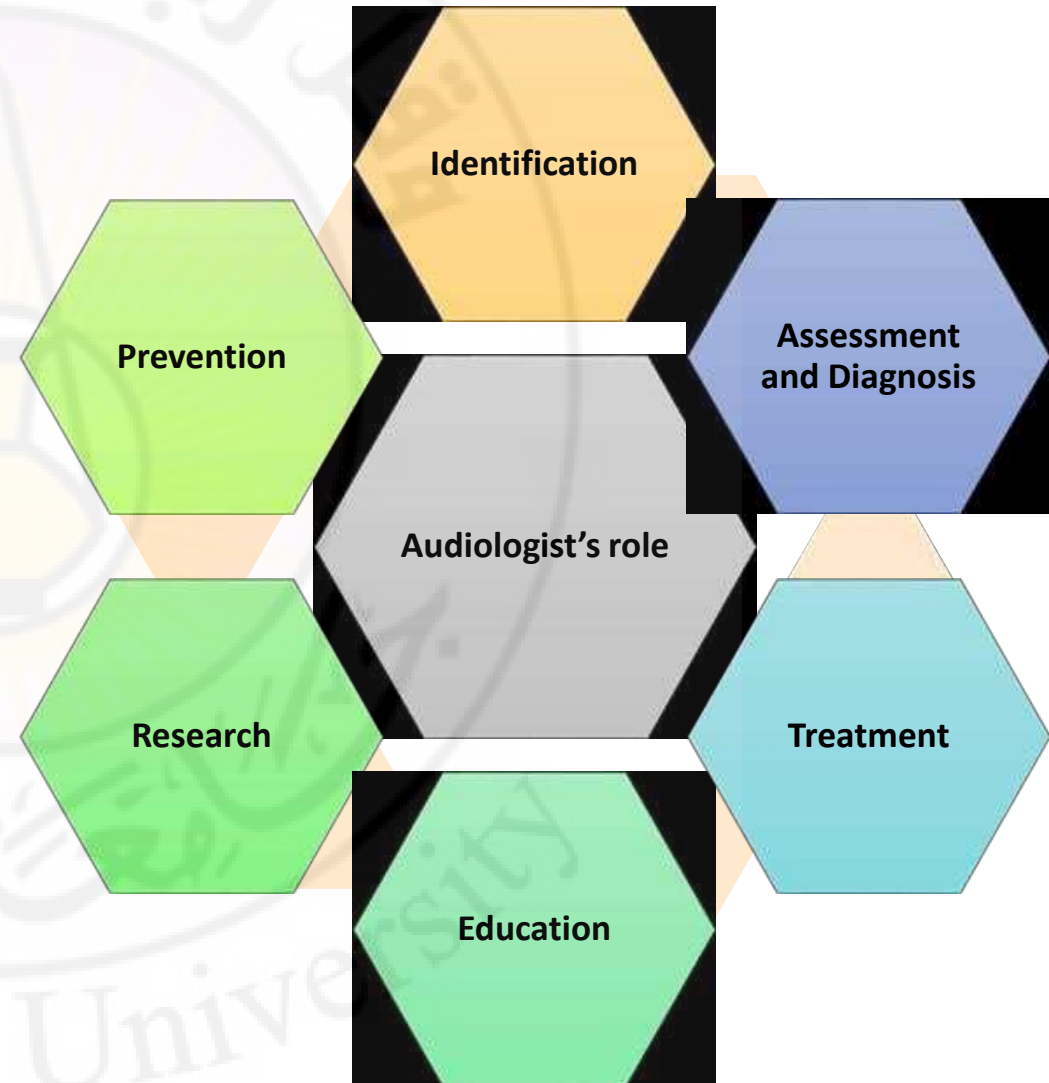
American Speech-Language-Hearing Association (ASHA)

Audiologists are professionals engaged in autonomous practice to promote healthy hearing, communication competency, and quality of life for persons of all ages through the **prevention, identification, assessment, and rehabilitation** of hearing, auditory function, balance, and other related systems.



What is an Audiologist's role?

- The audiologist identifies, assesses, diagnoses, and treats individuals with impairments of **hearing** and/or **vestibular** function.
- The audiologist also evaluates and fits hearing aids, and assists in the implementation of hearing loss treatment.

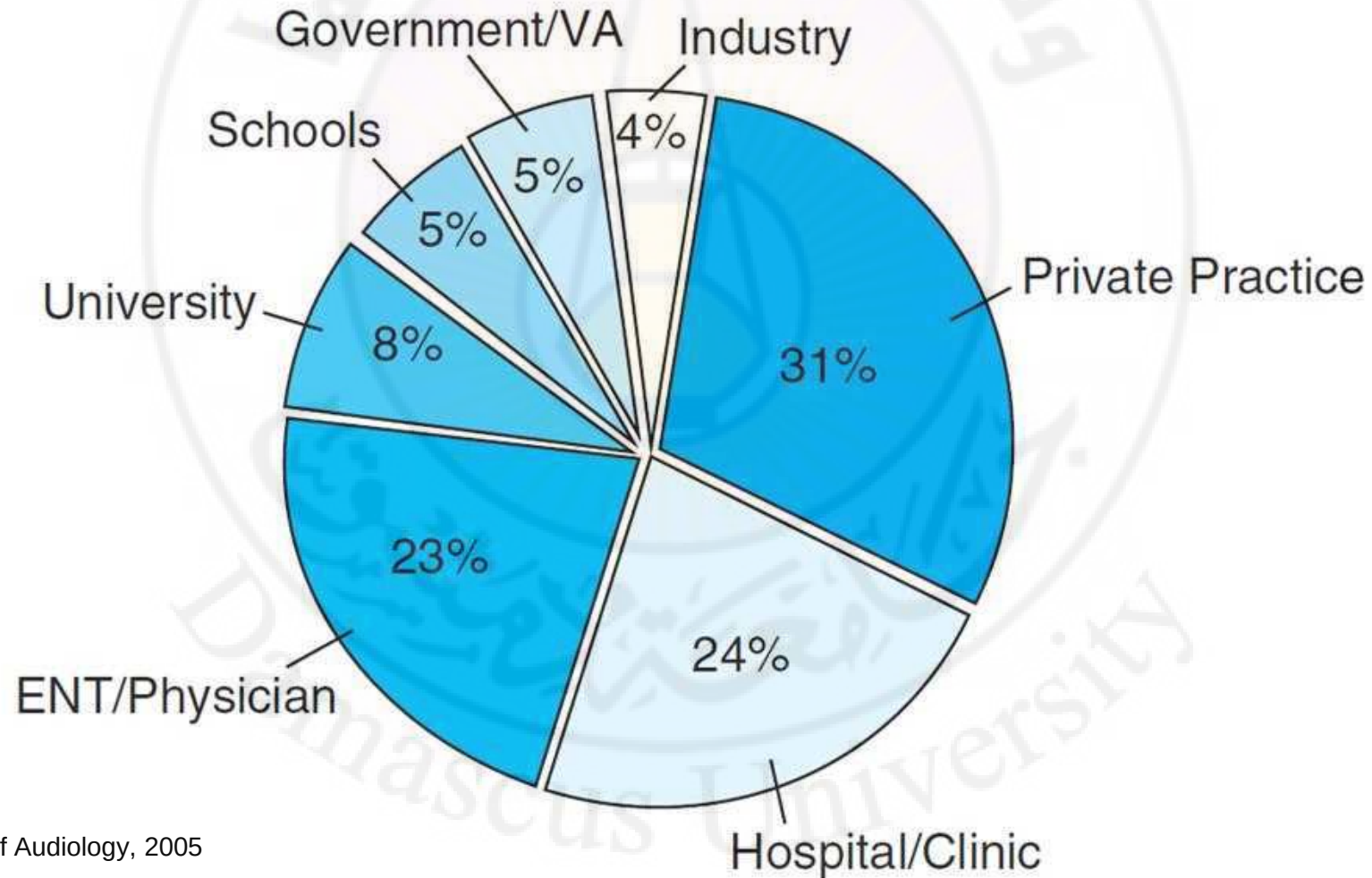


Scope of Practice

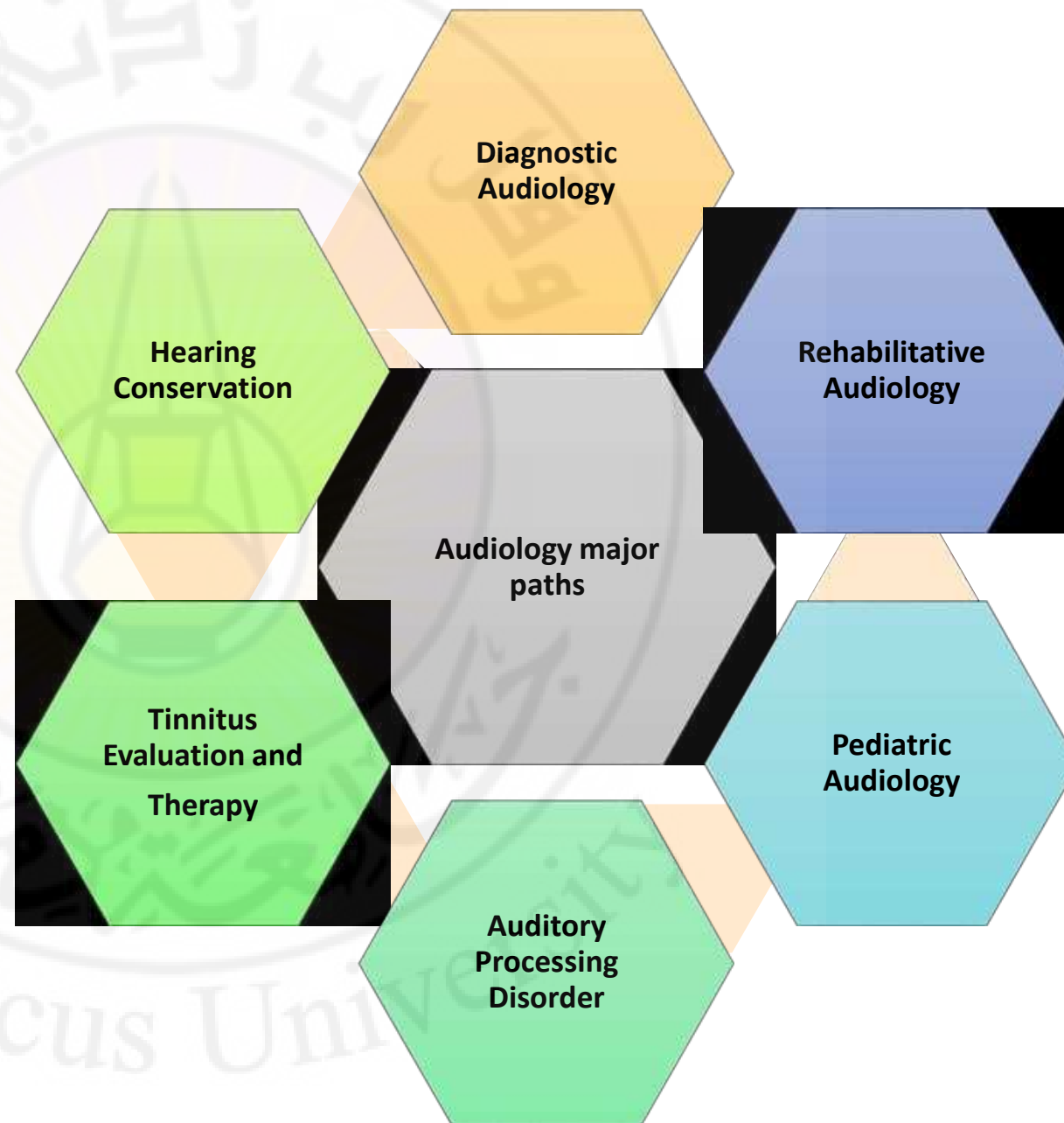
- It is incumbent on all professions to define their boundaries. They must delineate the professional activities that lie within their education and training and, by exclusion, the activities outside their territory.
- Audiology is an **autonomous** profession. As long as audiologists are practicing within their boundaries, they are acting as experts in their field.
- **Two examples:**
 - [American Academy of Audiology](#)
 - [American Speech-Language-Hearing Association](#)



Where do Audiologists practice?



Six Divergent Paths!



2. Basic evaluation and test battery approach

- **Clinical Process**
- **Case History**
- **Physical examination**
- **Tuning Forks examination**

Clinical Process

Domain of Information	Clinical Process 				
Patient	(1) Presenting Problem				(6) Outcome
Profession		Collecting Evidence		(4) Assessment: Problem List; Clinical impressions; “Diagnosis”	(5) Plan: Treatment or Management
		(2) Subjective: Chief Compliant; Symptoms; History	(3) Objective: Physical signs; Special Tests		
Discipline		(7) Learned theories of Cause and Effect			

Domain of Information	Clinical Process 				
Patient	(1) Presenting Problem: Difficulty understanding speech of family members and friends				(6) Outcome: Decreased noise exposure and no progression in hearing loss over years; uses hearing aids with benefit in speech understanding; family helps with communication misunderstandings; person participates in communication activities to a fuller extent
Profession		Collecting Evidence		(4) Assessment: Noise induced (firearms) high frequency hearing loss; communication deficits and activity restrictions compatible with hearing loss; continued risk of noise induced hearing loss	(5) Plan: Conservation counseling; audiologic rehabilitation (hearing aids, effective use of residual hearing)
Discipline		(2) Subjective: Slowly developing hearing problem primarily in background noise, limiting participation in family and outside activities. History: Right-handed hunter	(3) Objective: High frequency “noise notch” type sensorineural hearing loss, more severe on the left side	(7) Learned theories of Cause and Effect: Effects of firearm noise exposure on hearing; audibility theory and the effects of hearing loss on speech perception	

Clinical information



- **Subjective** (Chief complaint, sign & symptoms, history)
- **Objective** (Observation and examination- Audiologic testing)
- **Assessment** (Problem lists, clinical impressions, and diagnosis)
- **Plan** (Treatment or management)
- *Outcome*

Data >>> Fact >>> Clinical information

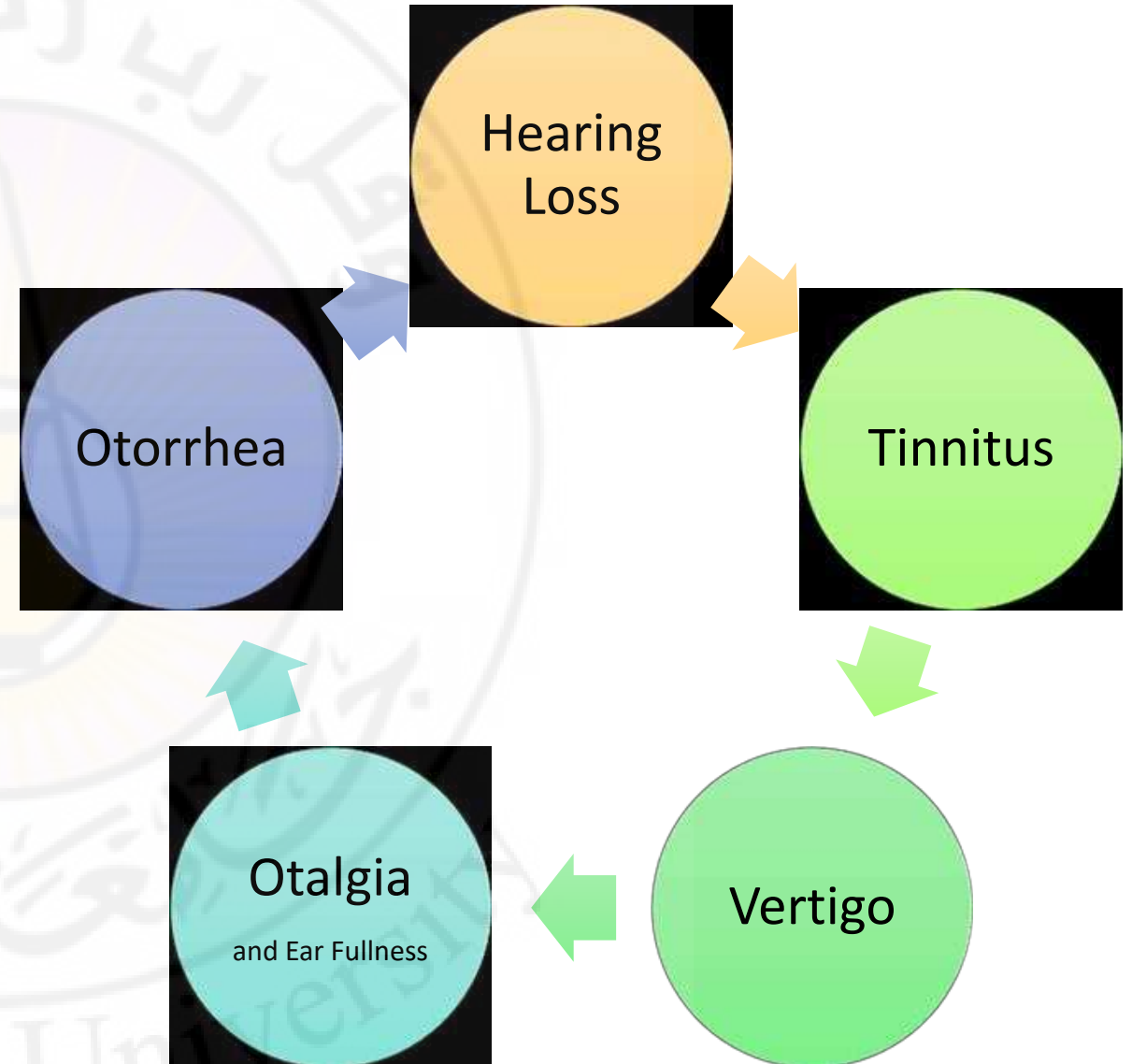
Before testing...

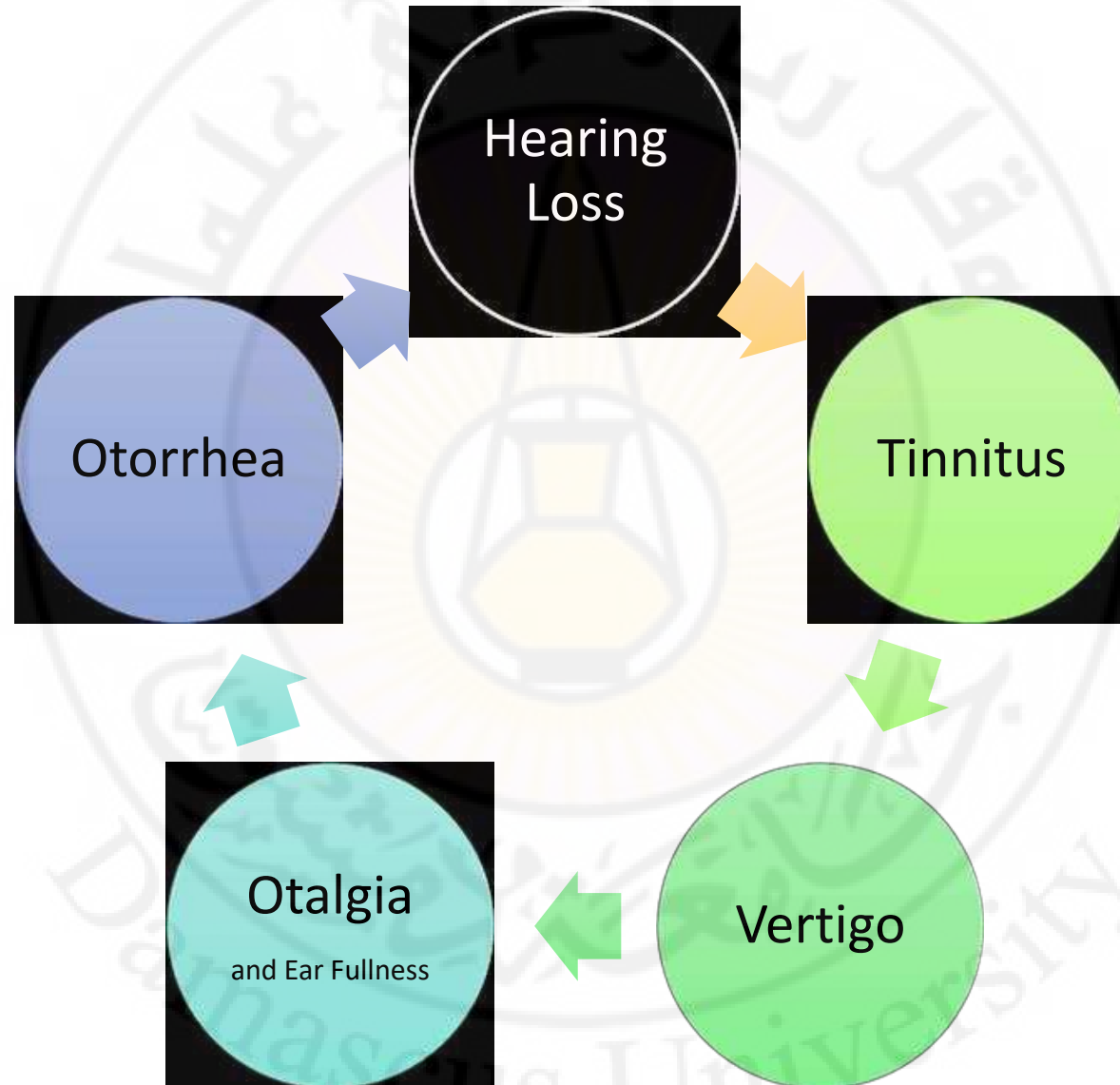
- Chief complaints
- Sign & Symptoms
- Case History
- Physical Examination
- Tuning Forks



Otologic Symptoms

- Hearing loss, tinnitus, otalgia, otorrhea, and vertigo are symptoms of otologic disease.
- Queries into each of these symptom categories will assist the clinician in formulating a **differential diagnosis**.
- Assessment of the patient with otologic complaints requires careful **history taking** in addition to a focused **clinical exam**.
- Many otologic complaints can be diagnosed by history alone, with the clinical exam frequently supporting the expected findings.





1. Hearing Loss

- HEARING disorder results from a number of causes and is usually characterized by the type and degree of hearing loss.
- **Type of hearing loss** is related to the site of the disorder within the auditory system, and **degree of loss** is related to the extent that the disorder is infringing on normal function.
- Defining both the type and degree of hearing loss is a cornerstone of audiology.
- Hearing disorders are of two major types:
 - Hearing sensitivity loss, and
 - Suprathreshold hearing disorders.
- *...and functional hearing loss*



In addition to type of loss, a hearing disorder can be described in terms of time of onset, time course, and whether one or both ears is involved.

Time of onset

- Congenital: present at birth
- Acquired: obtained after birth

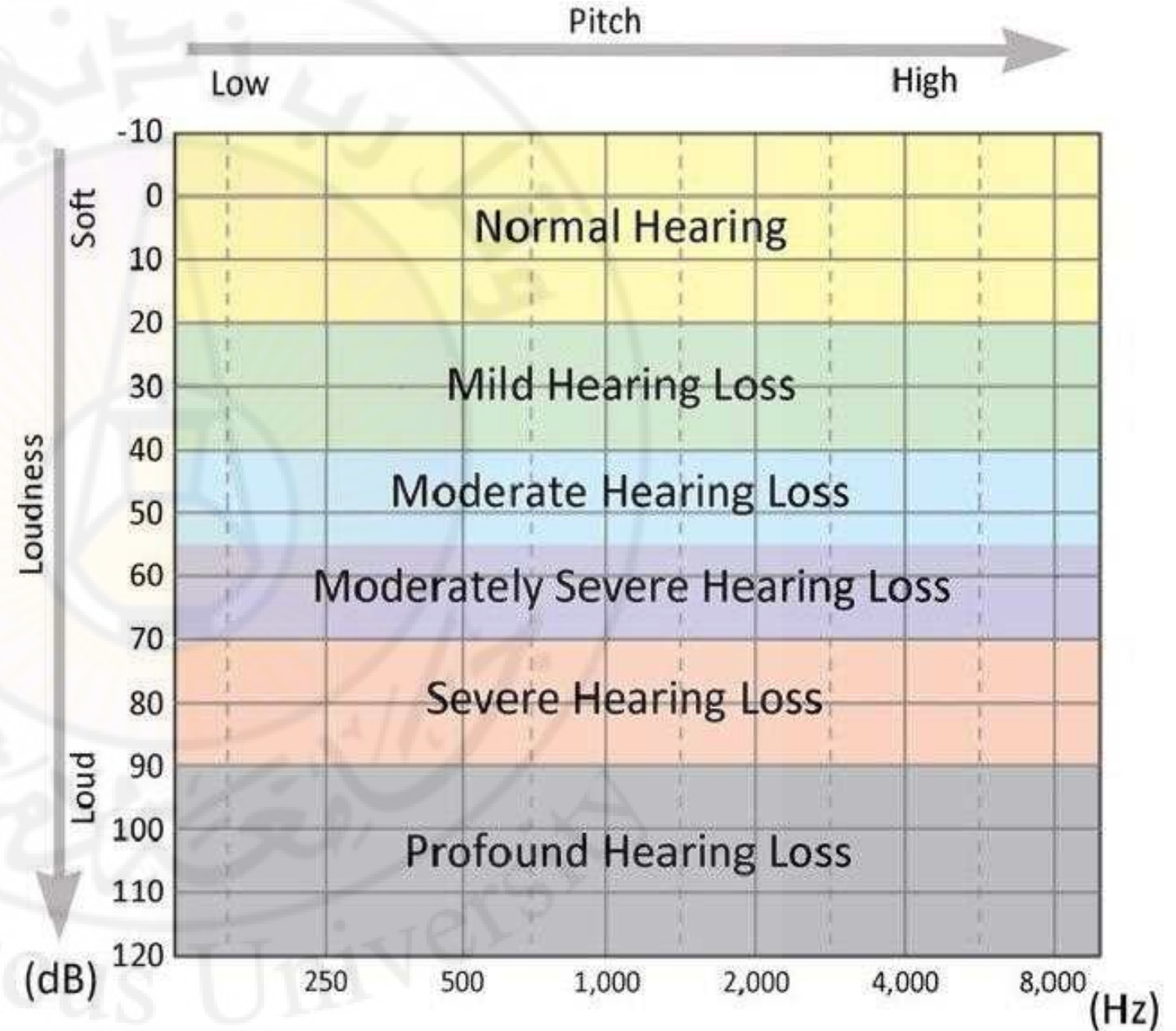
Number of ears involved

- Unilateral: pertaining to one ear only
- Bilateral: pertaining to both ears

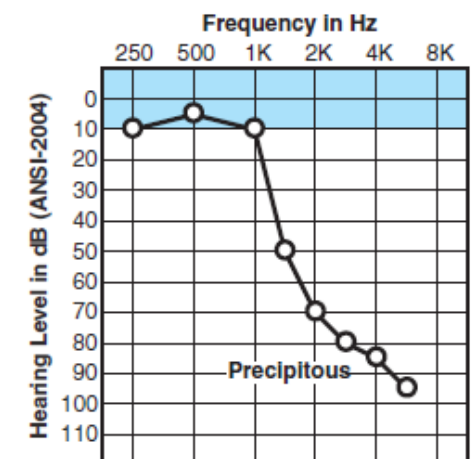
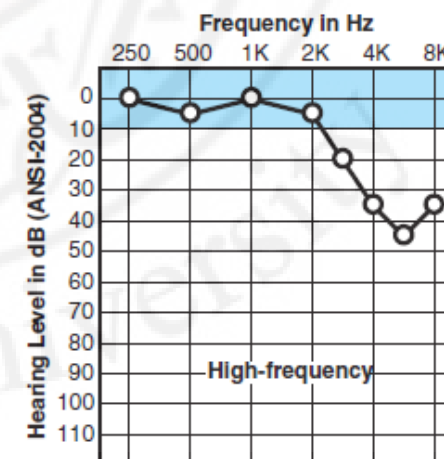
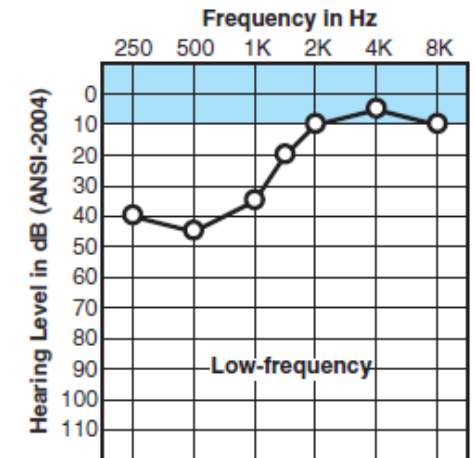
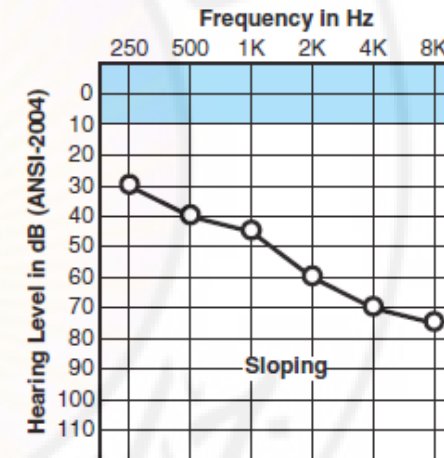
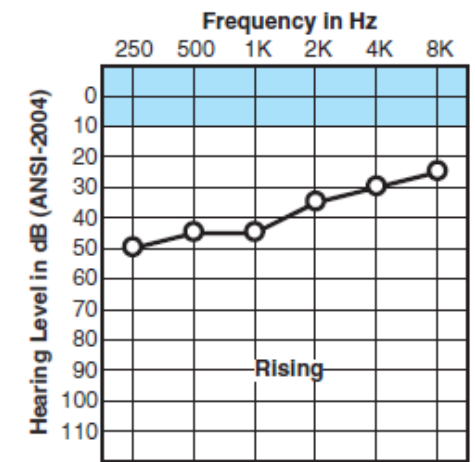
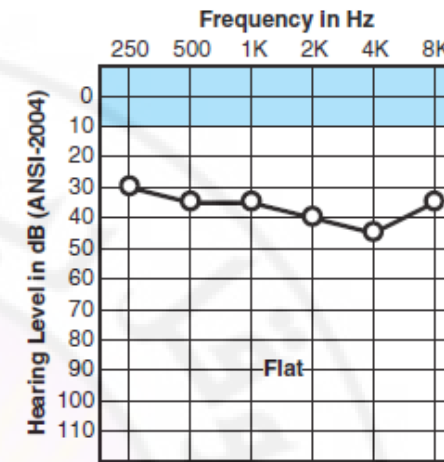
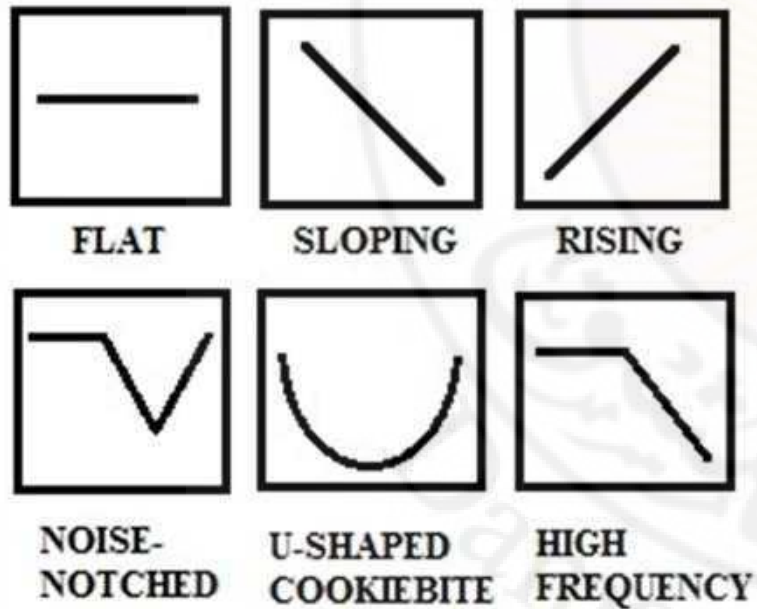
Time course

- Acute: of sudden onset and short duration
- Chronic: of long duration
- Sudden: having a rapid onset
- Gradual: occurring in small degrees
- Temporary: of limited duration
- Permanent: irreversible
- Progressive: advancing in degree
- Fluctuating: aperiodic change in degree

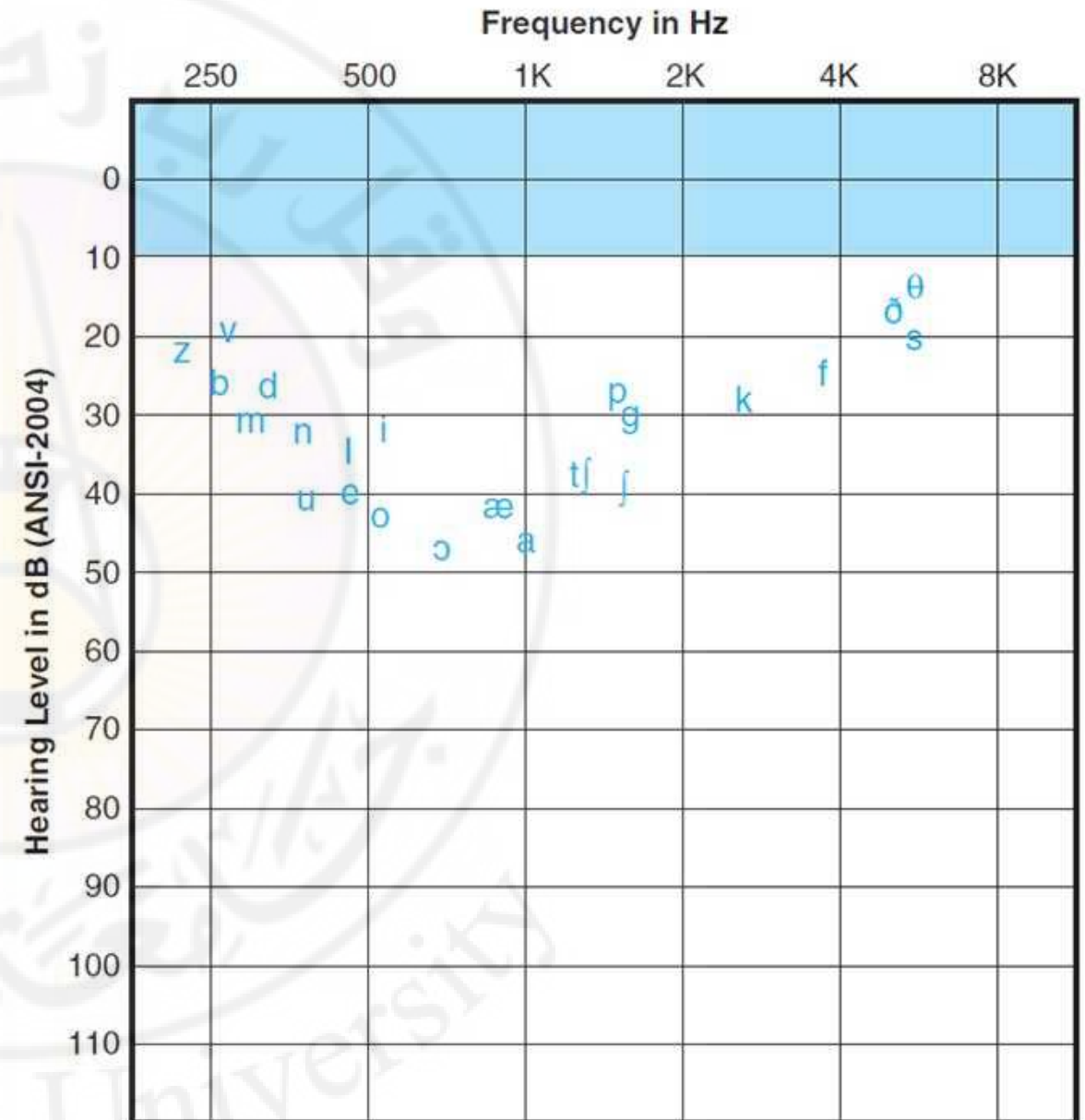
Degree of Hearing Loss



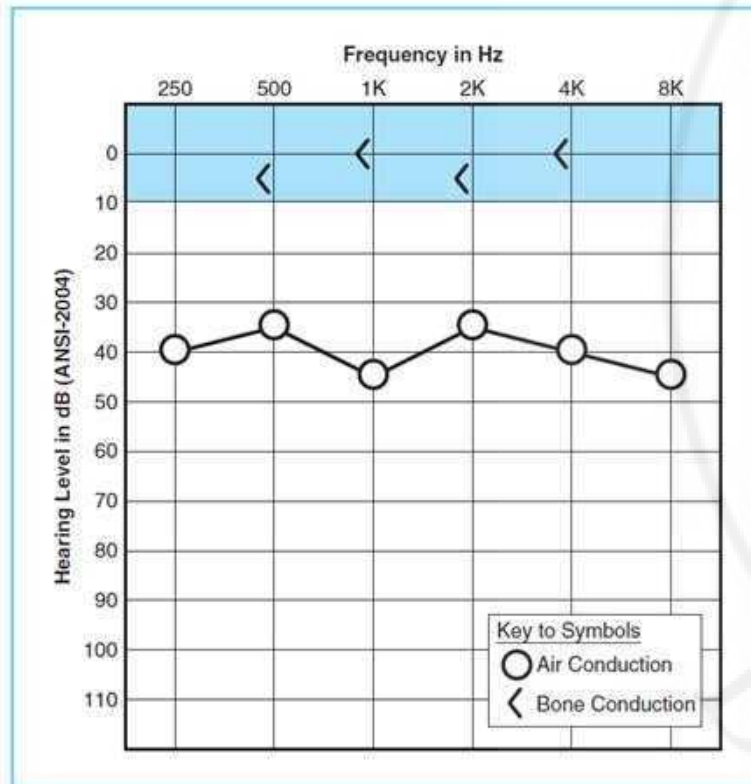
Configuration of Hearing Loss



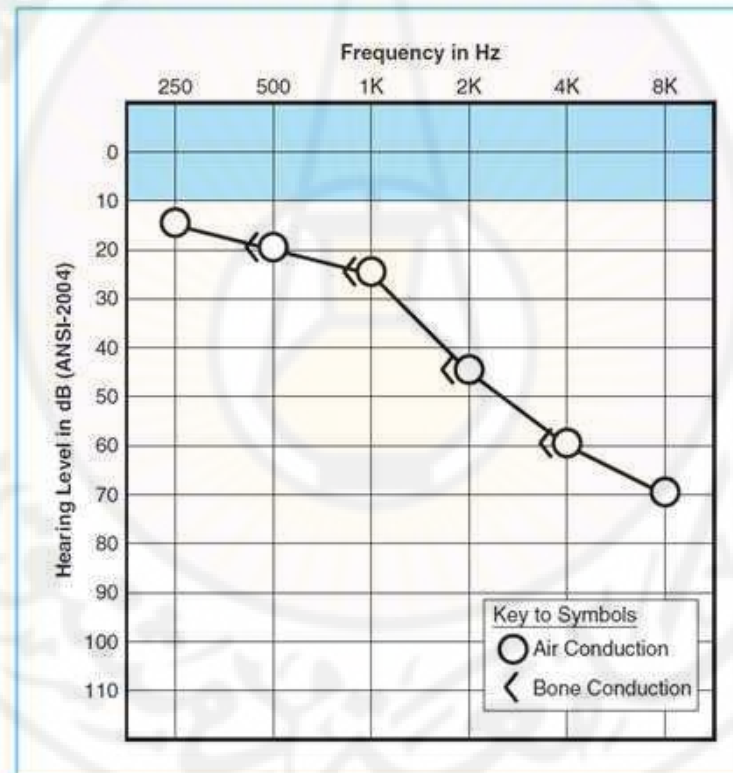
Generalized phonetic representations of speech sounds occurring at normal conversational levels plotted on an audiogram.



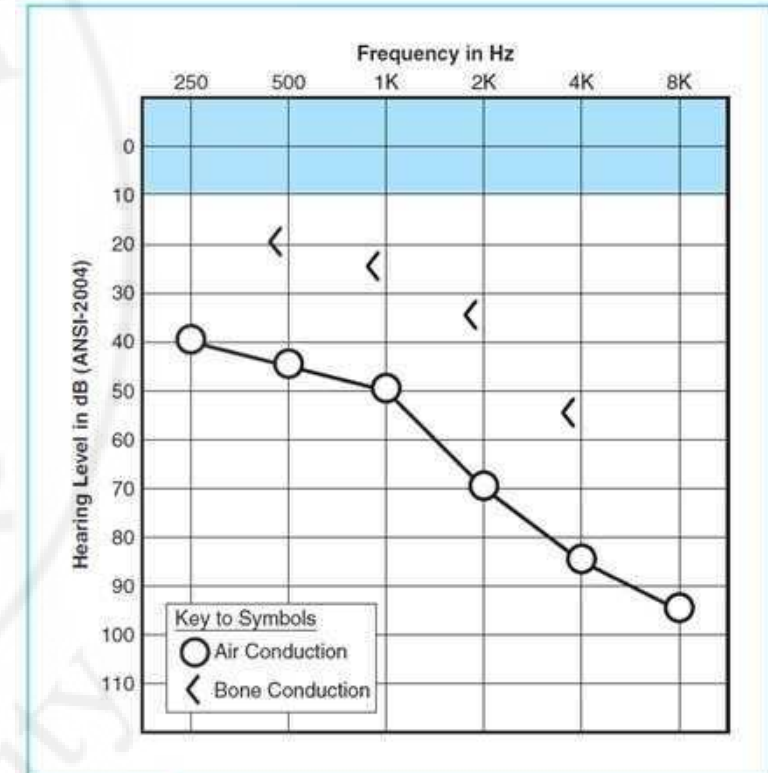
Type of Hearing Loss



CHL



SNHL



Mixed

Hearing Loss: CHL

- The configuration of a conductive hearing loss is generally either **flat or low-frequency** in nature.
- In addition, conductive hearing loss has a **maximum** limit of approximately **60 dB HL**. This represents the loss due to both the lack of impedance-matching function of the middle ear and additional mass and stiffness components related to the nature of the disorder.
- At **suprathreshold level**, the ear with a conductive hearing loss acts quite normally. As long as sound can be made sufficiently loud, hearing will be normal.
- Usually **transient** and needs medical treatment
- Sometimes **permanent** and needs to be treated with hearing aid amplification
- Some children have chronic middle-ear disorder that results in **fluctuating** conductive hearing loss throughout much of their early lives.

Hearing Loss: CHL

- Although examination and audiometric testing are confirmatory, there are several historic components which may suggest CHL:
- **Hyperacusis of Willis (Paracusis Willisii)** is a phenomenon of CHL in which the patient claims to hear better in background noise, such as a cocktail party.
- **Autophony** may also suggest CHL, although it does not define cause.
- A personal history of **ear infection/trauma/or surgery** could cause CHL through ossicular discontinuity or fixation.
- **Otosclerosis** is a condition which frequently presents with CHL secondary to stapedial fixation. Patients may present with unilateral or bilateral progressive hearing loss without a significant otologic history. In such a case, family history may reveal relatives who had surgery for loss of hearing.

Hearing Loss: SNHL

- A sensorineural hearing loss has at least three fundamentally important effects on hearing:
 - a reduction in the cochlear sensitivity,
 - a reduction in frequency resolution, and
 - a reduction in the dynamic range of the hearing mechanism.
- One of the consequences of sensorineural hearing loss is **recruitment**, or abnormal loudness growth. loudness grows more rapidly than normal at intensity levels just above threshold in an ear with sensorineural hearing loss.
- Reduction in frequency resolution and in dynamic range affect the **perception of speech**. In most sensorineural hearing loss, this effect on speech understanding is predictable from the audiogram and is poorer than would be expected from a conductive hearing loss of similar magnitude.

Hearing Loss: SNHL

- Sensorineural hearing loss (SNHL) may occur at any age. The **age of the patient** should define the line of questioning.
- For **children**, prenatal and birth history, as well as family history, may suggest ototoxicity, prenatal infection, auditory neuropathy, or genetic hearing loss of syndromic or nonsyndromic nature.
- **Post-lingual or adult-onset** SNHL has a wide range of etiologies that may be evaluated by a combination of careful history, exam, audiometric testing, and occasionally imaging.
- **Family history** is important in adults as well; many patients may have a genetic predisposition to hearing loss.
- A history of **noise exposure**, whether occupational or recreational, is also important to elicit. Many patients with noise-induced SNHL will complain of the significant effort needed to understand speech. This difficulty is a result of the loss of mid- and high-frequency sounds with relative preservation of the low frequencies; the patient will say that **they can hear, but cannot understand**, what people are saying.

Hearing Loss: SNHL of retrocochlear origin

- Hearing loss from retrocochlear disorder is distinguishable from cochlear or conductive hearing loss by the extent to which it can adversely affect **speech perception**.
- Loudness growth can be abnormal in patients with retrocochlear disorder. Instead of the abnormally rapid growth of loudness characteristic of cochlear hearing loss, retrocochlear disorder can show **no recruitment or decruitment**, an abnormally slow growth in loudness with increasing intensity.
- Another abnormality that can occur as a result of retrocochlear disorder is **abnormal auditory adaptation**. The normal auditory system tends to adapt to ongoing sound, especially at near-threshold levels, so that, as adaptation occurs, an audible signal becomes inaudible. At higher intensity levels, ongoing sound remains audible without adaptation. However, in an ear with retrocochlear disorder, the audibility may diminish rapidly due to excessive auditory adaptation, even at higher intensity levels.



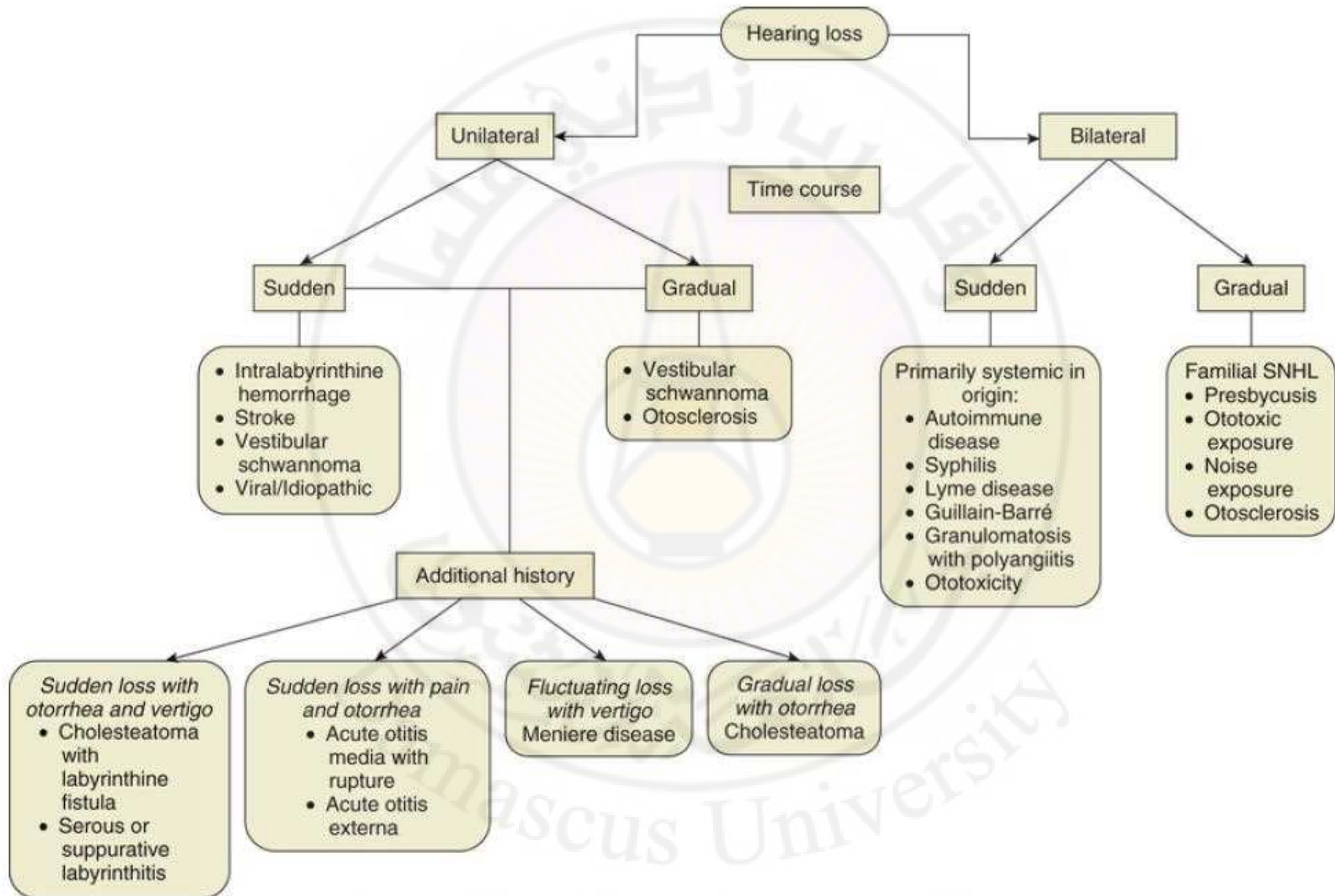
History Taking for a Chief Complaint of Hearing Loss

Duration	What difficulty is this causing the individual?
Progression	Prior ear infection/trauma/surgery
Slow versus rapid	Noise exposure
Sudden	Family history of hearing loss
Fluctuating	Ototoxic medication exposure
Symmetric or asymmetric?	Prior amplification experience
Associated ear symptoms	
Tinnitus	
Vertigo	
Otalgia	
Otorrhea	

Red Flags for Hearing Loss History

- Unilateral
- Sudden onset
- Fluctuation
- Rapidly progressive
- Vertigo or ataxia
- Otorrhea
- Otalgia
- Accompanying cranial nerve signs
- Numbness
- Diplopia
- Facial paresis
- Voice/swallowing changes



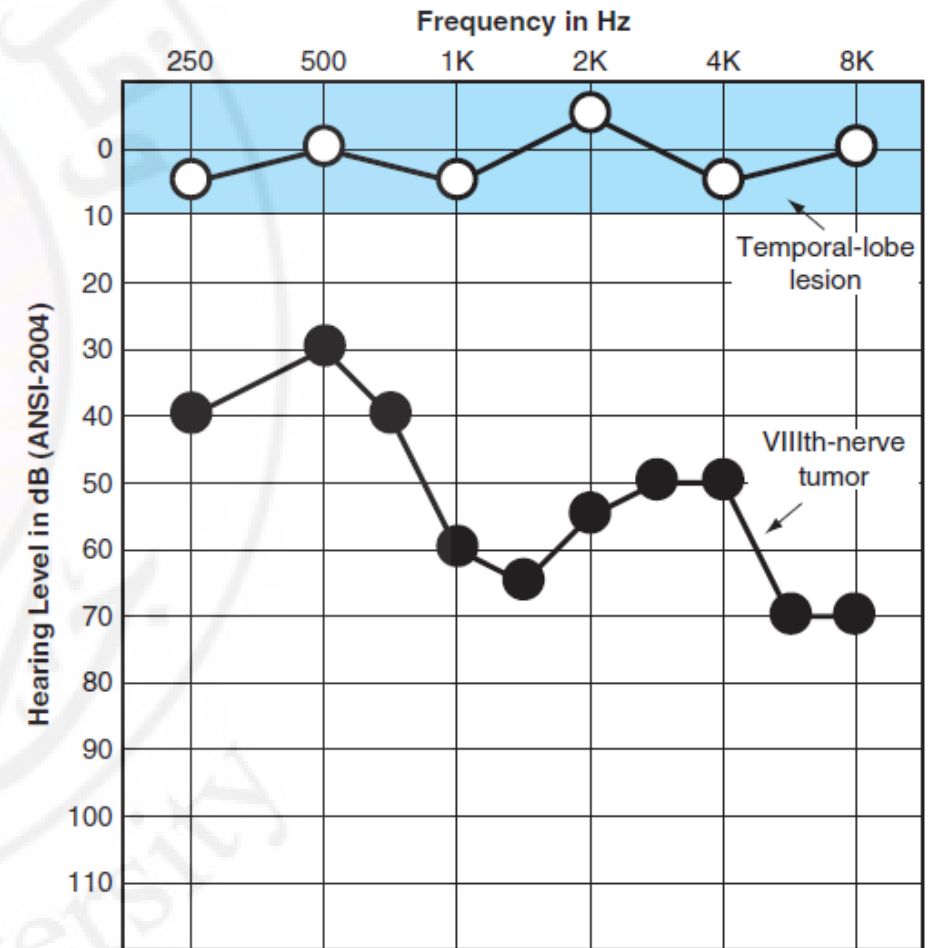


Suprathreshold Hearing Disorder

- Although there is a tendency to think of hearing disorder as the sensitivity loss that can be measured on an audiogram, there are other types of hearing disorders that may or may not be accompanied by sensitivity loss.
- These other disorders result from disease or damage to the central auditory nervous system in adults, or delayed or disordered auditory nervous system development in children.
 - When a disorder is caused by an active, measurable disease process, such as a tumor or other space-occupying lesion, or from damage due to trauma or stroke, it is often referred to as a **retrocochlear disorder**. That is, retrocochlear disorders result from structural lesions of the nervous system.
 - When a disorder is due to developmental dysfunction or delay or from diffuse changes such as the aging process, it is often referred to as a **auditory processing disorder**. That is, auditory processing disorders results from “functional lesions” of the nervous system.

Retrocochlear Hearing Disorder

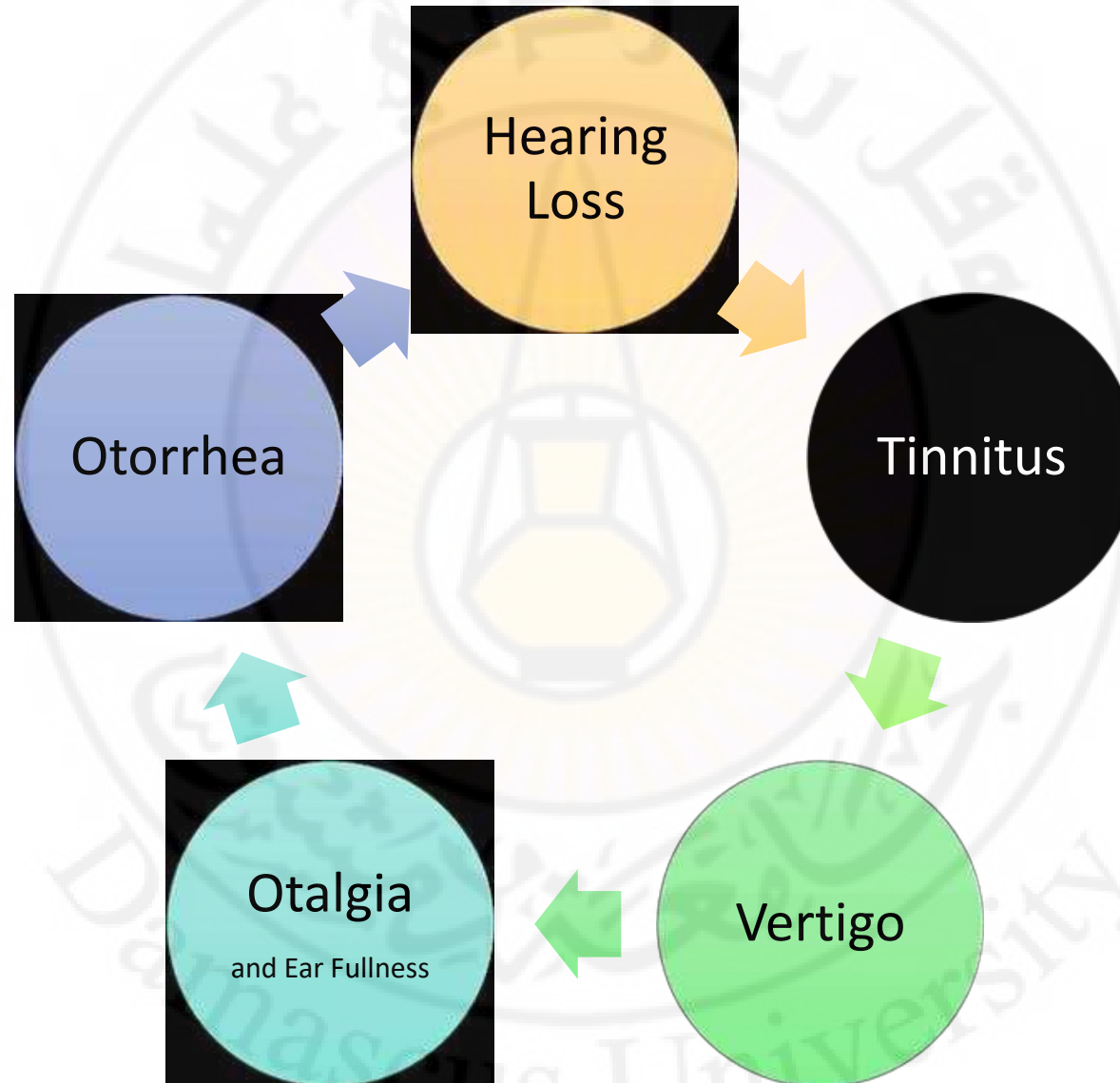
- A retrocochlear disorder is caused by a change in neural structure and function of some component of the peripheral or central auditory nervous system. As a general rule, the **more peripheral** a lesion, the greater its impact will be on auditory function. Conversely, the **more central** the lesion, the more subtle its impact will be.
- A retrocochlear lesion may or may not affect auditory sensitivity. This will depend on many factors, including lesion **size**, **location**, and **impact**.
- More subtle hearing disorders from retrocochlear disease are often noted in measures of **suprathreshold function such as speech recognition** ability. Using various measures that stress the auditory system, the audiologist can detect some of the more subtle changes resulting from retrocochlear lesions.



Auditory Processing Disorder

Classification system for describing types of disorders that can affect the ability to turn sound into meaning

Disorder Type	Nature of Deficit
Auditory processing disorders — speech-in-noise problems — dichotic deficits — temporal processing disorders	Auditory
Receptive language processing disorders — linguistically dependent problems — deficits in analysis, synthesis, closure	Linguistic
Supramodal disorders — attention — memory	Cognitive



2. Tinnitus

- Tinnitus is often described as “ringing in the ears” and is the medical term to explain the perception of sound in one or two ears when no sound is actually present.
- Tinnitus may be the chief complaint of the patient or an incidental symptom uncovered during questioning.
- Tinnitus is a **common** condition that affects 30% of people older than age 55.



Term	Definition
Tinnitus	The perception of sound when there is no external source of the sound
Primary tinnitus	Tinnitus that is idiopathic ^a and may or may not be associated with sensorineural hearing loss
Secondary tinnitus	Tinnitus that is associated with a specific underlying cause (other than sensorineural hearing loss) or an identifiable organic condition
Recent onset tinnitus	Less than 6 months in duration (as reported by the patient)
Persistent tinnitus	6 months or longer in duration
Bothersome tinnitus	Distressed patient, affected quality of life ^b and/or functional health status; patient is seeking active therapy and management strategies to alleviate tinnitus
Nonbothersome tinnitus	Tinnitus that does not have a significant effect on a patient's quality of life but may result in curiosity of the cause or concern about the natural history and how it might progress or change

Sound features of tinnitus

TINNITUS CRITERIA	POSSIBLE FEATURES
Onset	Sudden, gradual
Pattern	Pulsatile, intermittent, constant, fluctuating
Site	Right or left ear, both ears, within head
Loudness	Wide range, varying over time
Quality	Pure tone, noise, polyphonic
Pitch	Very high, high, medium, low



History Taking for a Chief Complaint of Tinnitus

Duration	Associated ear symptoms
Progression	Vertigo
Slow versus rapid	Hearing loss
Sudden	Otalgia
Fluctuating	Otorrhea
Symmetric or asymmetric?	Prior ear infection/trauma/surgery
Pulsatile or non-pulsatile?	Noise exposure
How does it sound?	Family history of hearing loss
Mitigating factors	Caffeine or other stimulant consumption
Prior therapies	

Subjective vs. Objective Tinnitus

- **Objective tinnitus**
 - can be detected by an observer using a stethoscope or ear canal microphone.
 - **Somatosounds** that reflect the perception of internally generated sounds from joints, muscles, turbulent blood flow, or, rarely, otoacoustic emissions.
 - Objective tinnitus usually has a pulsatile or rhythmic quality.
 - Is less common
- **Subjective tinnitus** is not audible to an observer



Subjective Tinnitus

- **Most commonly related to**
 - Sensorineural hearing loss (SNHL)
 - Acoustic trauma
 - Presbycusis
- **Less commonly the result of**
 - Conductive hearing loss,
 - Endolymphatic hydrops,
 - Cerebellopontine angle tumors

Box 151-2. SUBJECTIVE TINNITUS SUBTYPES

Pattern of hearing loss

Noise-induced hearing loss

Presbycusis

Unilateral

High-frequency hearing loss

Outer hair cell dysfunction

Somatic tinnitus

Temporomandibular joint dysfunction

Cervical dysfunction

Gaze evoked

Cutaneous evoked

General somatosensory modulated

Typewriter tinnitus

Exacerbated by sleep or rest

Musical/complex

Associated affective disorder

Intrusive (versus habituated)

Box 151-1. OBJECTIVE TINNITUS SUBTYPES

Pulsatile

Synchronous with Pulse

Arterial etiologies

Arteriovenous fistula or malformation

Paraganglioma (glomus tympanicum or jugulare)

Carotid artery stenosis

Other atherosclerotic disease (subclavian, external carotid)

Arterial dissection (carotid, vertebral)

Persistent stapedial artery

Intratympanic carotid artery

Vascular compression of cranial nerve VIII

Increased cardiac output (pregnancy, thyrotoxicosis)

Intraosseous (Paget disease, otosclerosis)

Venous etiologies

Pseudotumor cerebri

Venous hum

Sigmoid sinus and jugular bulb anomalies

Asynchronous with Pulse

Palatal myoclonus

Tensor tympani or stapedius muscle myoclonus

Nonpulsatile

Spontaneous otoacoustic emission

Patulous eustachian tube

Tinnitus

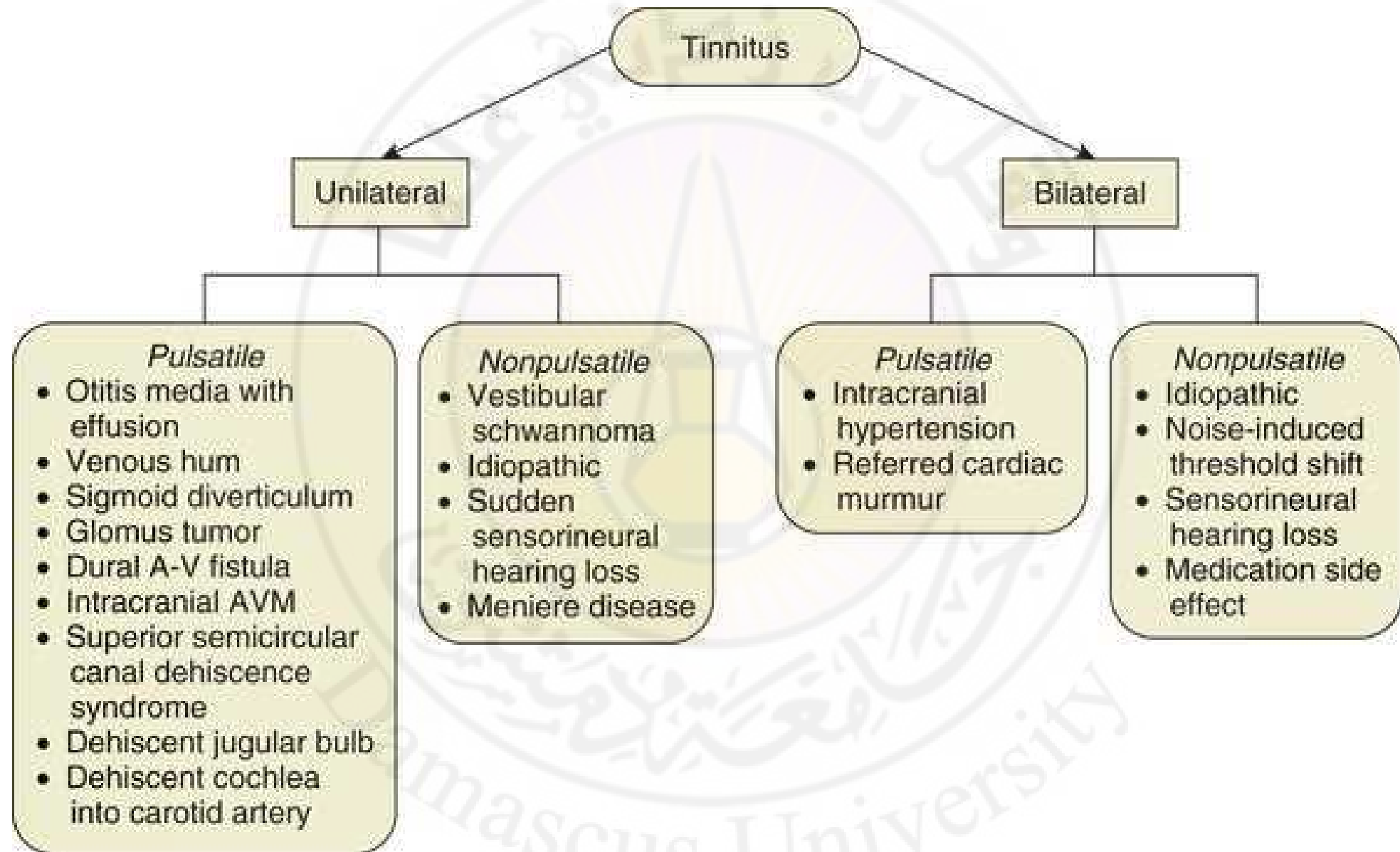
- It is specifically important to the tinnitus sufferer to evaluate their **state of mind**. Patients may come in very emotionally distressed by their tinnitus, occasionally with suicidal intent.
- It is a moral imperative for the clinician to assess the patient for the risk of self-harm during the clinical visit.
- Additional **exacerbating conditions** such as poor sleep quality and significant situational stress (such as work environment or familial strife) should be asked.

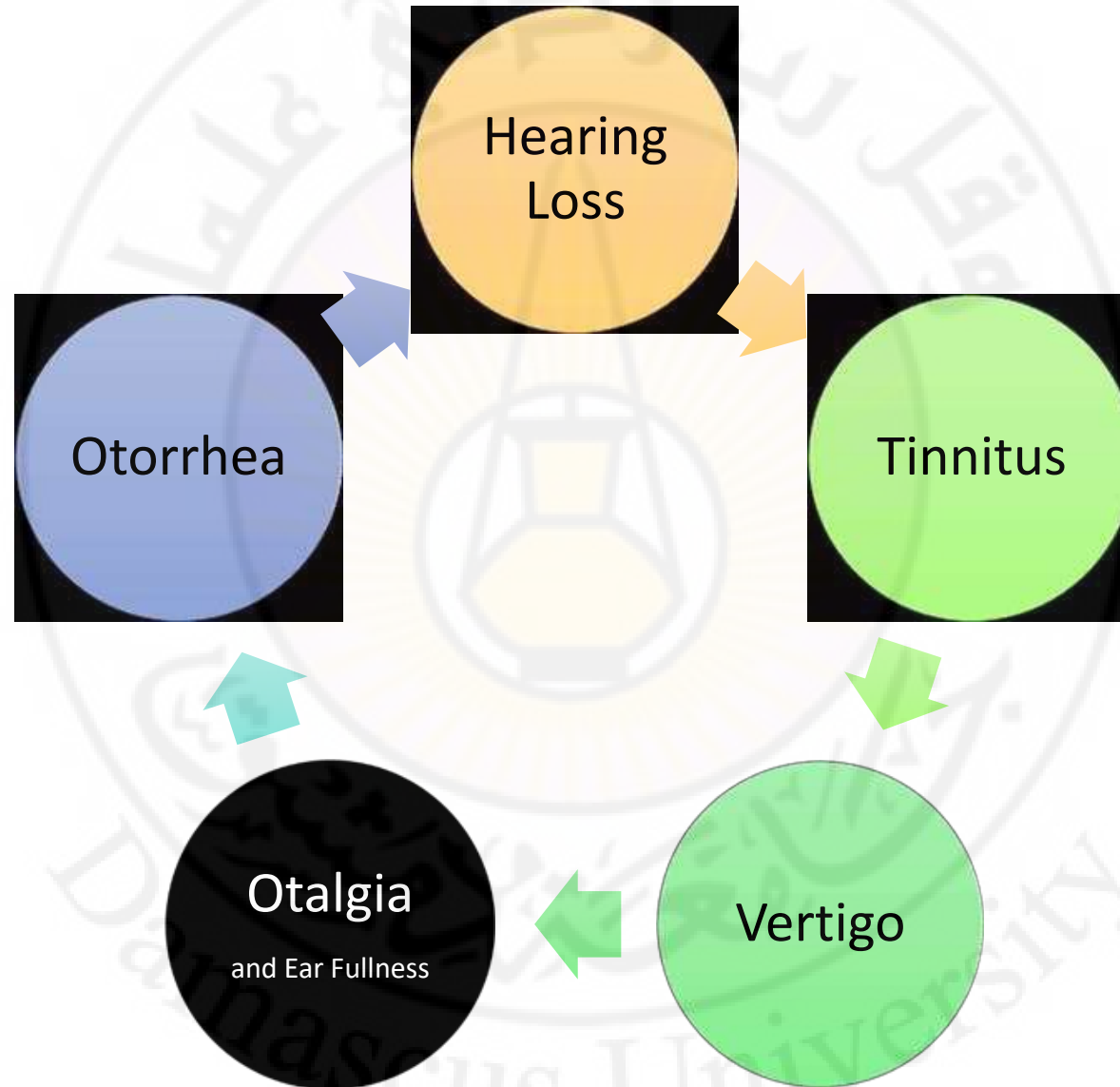


Tinnitus

- **Pulsatile tinnitus** requires a very different line of inquiry than nonpulsatile tinnitus. The origin may be completely benign but could also be the sign of a serious cerebrovascular problem.
- **Neurologic symptoms** should be obtained, such as headaches, vision changes, and cranial nerve (CN) changes, as well as symptoms of **stroke** such as aphasia and face/body weakness now or in the past.
- **Cardiac and vascular history** should be obtained.
- Vertigo, hyperacusis, and other autophonies should be asked.
- Prior head or neck trauma should also be reviewed.







3. Otalgia and Ear Fullness

- A proper assessment of **ear pain** must include a thorough inquiry of the pain characteristics and associated symptoms.
- Understanding the relationship of otalgia and ear fullness to other otologic symptoms can differentiate between **primary otologic** disease and **referred otalgia**.
- Pain may be characterized as **acute and chronic**, as well as **nociceptive** (due to damaged tissue and responding nociceptors) or **neuropathic** (originating from nerve damage and/or abnormal nerve signals).



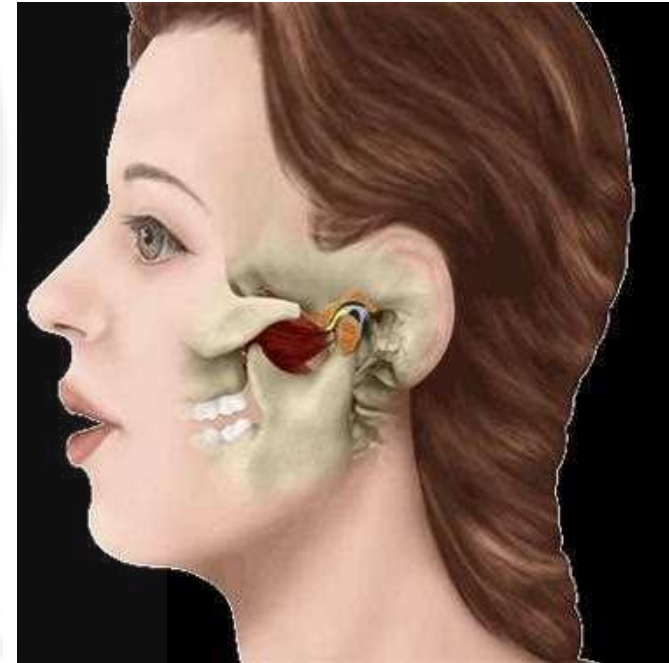


History Taking for a Chief Complaint of Ear Pain

Onset: Timing; context	Associated ear symptoms
Pain scale 1–10	Vertigo
Presence: Continuous vs intermittent	Hearing loss
Symmetric or asymmetric	Otalgia
Qualities: Stabbing; dull; burning	Otorrhea
↑ Exacerbating factors	Prior ear infection/trauma/surgery
↓ Mitigating factors	Prior head and neck infection/surgery/radiation
Masses in mouth/throat/neck	Tobacco use
Voice/swallowing changes	Alcohol use
Weight loss	

Otalgia

- **Primary otalgia** is frequently accompanied by hearing loss and otorrhea. The combination of these symptoms may indicate common self-limited disease such as otitis media or externa. These disorders, as well as more serious causes such as primary otologic malignancy, mastoiditis, cholesteatoma, or skull base osteomyelitis (malignant otitis externa), can be further evaluated by physical exam.
- Otalgia may be referred from numerous sources, because of the anatomic complexity of the head and neck. A most common source of **referred otalgia** is dental. This may include primary **dental disease** (such as periodontal disease, caries, and dental abscesses) or conditions relating to the **temporomandibular joint**, such as bruxism, clenching, primary temporomandibular joint osteoarthritis, or musculoskeletal pain from the muscles of mastication.



Otalgia

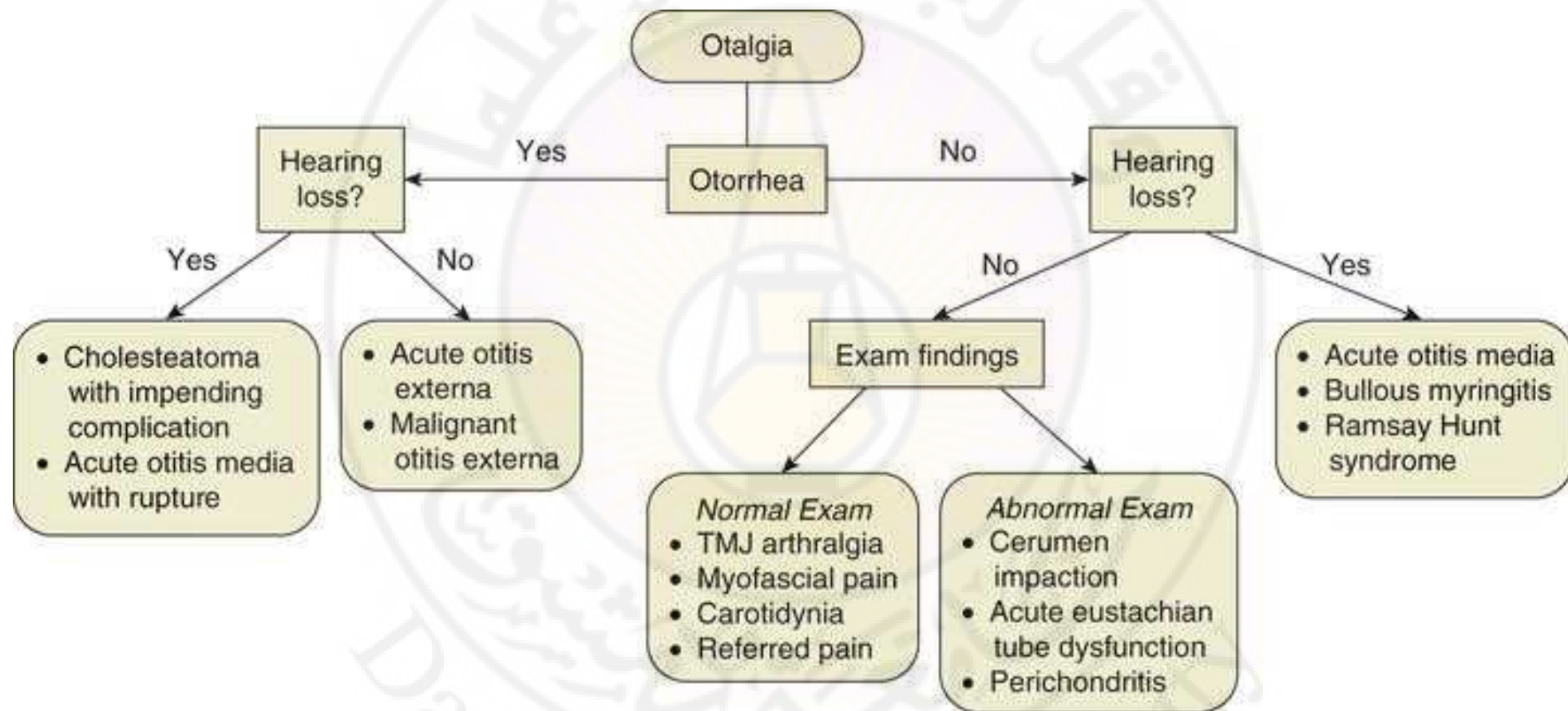
- **Chronic ear pain** may be more difficult for the patient to describe, and as such, the patient may have a complex history of this pain over time.
- Chronic pain may be classified as neuropathic, nociceptive or both.
- Chronic pain conditions such as post-herpetic neuralgia, fibromyalgia, and reflex sympathetic dystrophy could be suggested by the patient's past and current medical history.
- **Psychological stressors** should be explored.
- Prior therapeutic interventions should be comprehensively reviewed, especially opioid use.

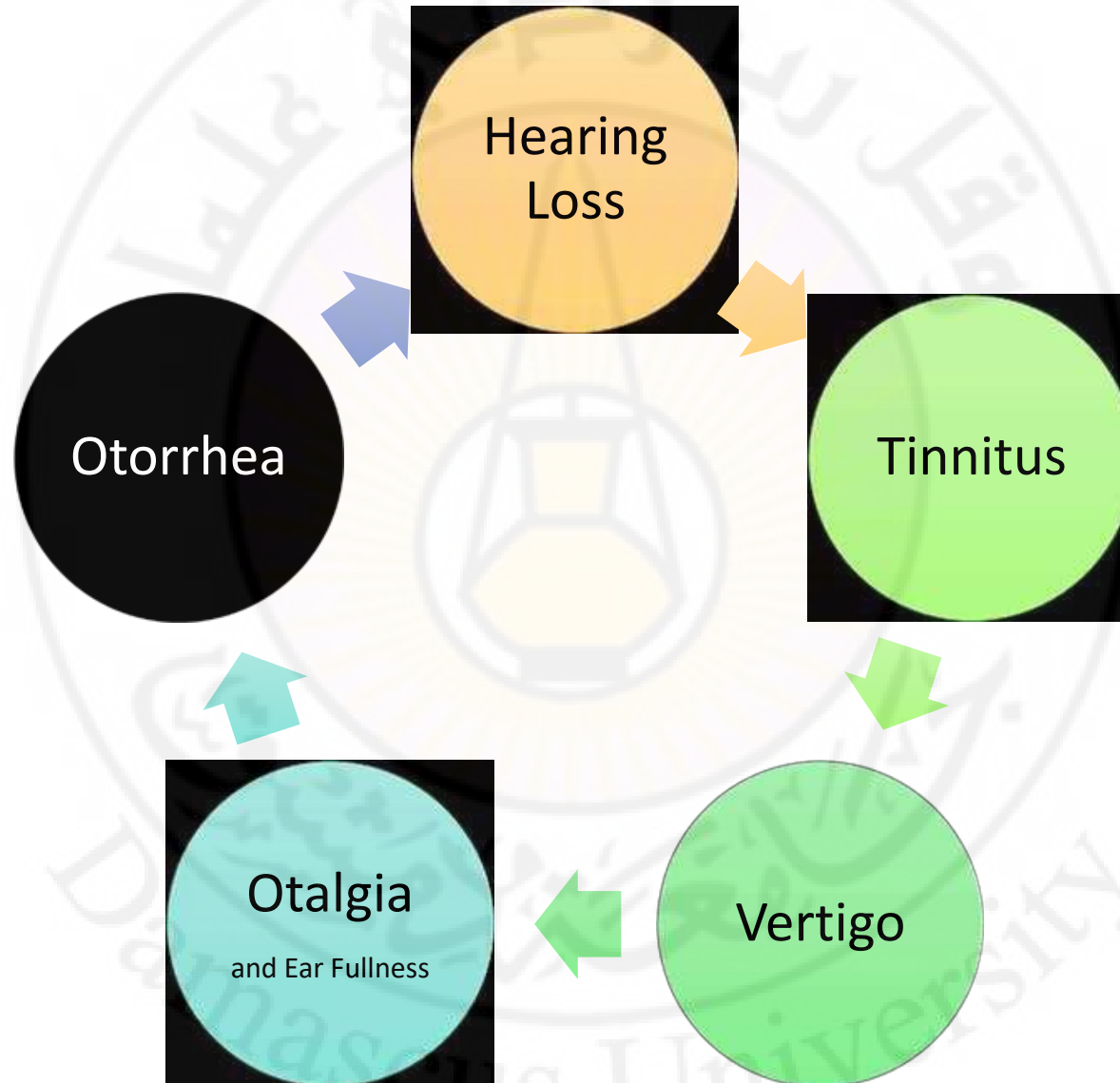


Ear Fullness

- **Ear fullness** is a vague descriptor for a sensation that could be originating in or outside of the ear.
- Patients may describe a sensation of pressure and may say “if I could just pop my ear it would be better.” Other patients may describe a sensation of being underwater or a “clogged” feeling.
- The clinician should evaluate for hidden hearing loss, eustachian tube dysfunction, chronic otitis media, and temporomandibular joint dysfunction







4. Otorrhea

- **Otorrhea**, or drainage from the ear, suggests a primary otologic pathology.
- Otorrhea can result from external ear canal pathology or middle ear disease with tympanic membrane perforation.
- It can also occur after a significant skull injury.
- Otorrhea in the setting of a traumatized or immunocompromised patient may indicate a serious life-threatening condition, and so prompt and thorough evaluation is necessary if trauma has not been ruled out.
- The physical examination and the history will differentiate among most causes of otorrhea in children.



Otorrhea

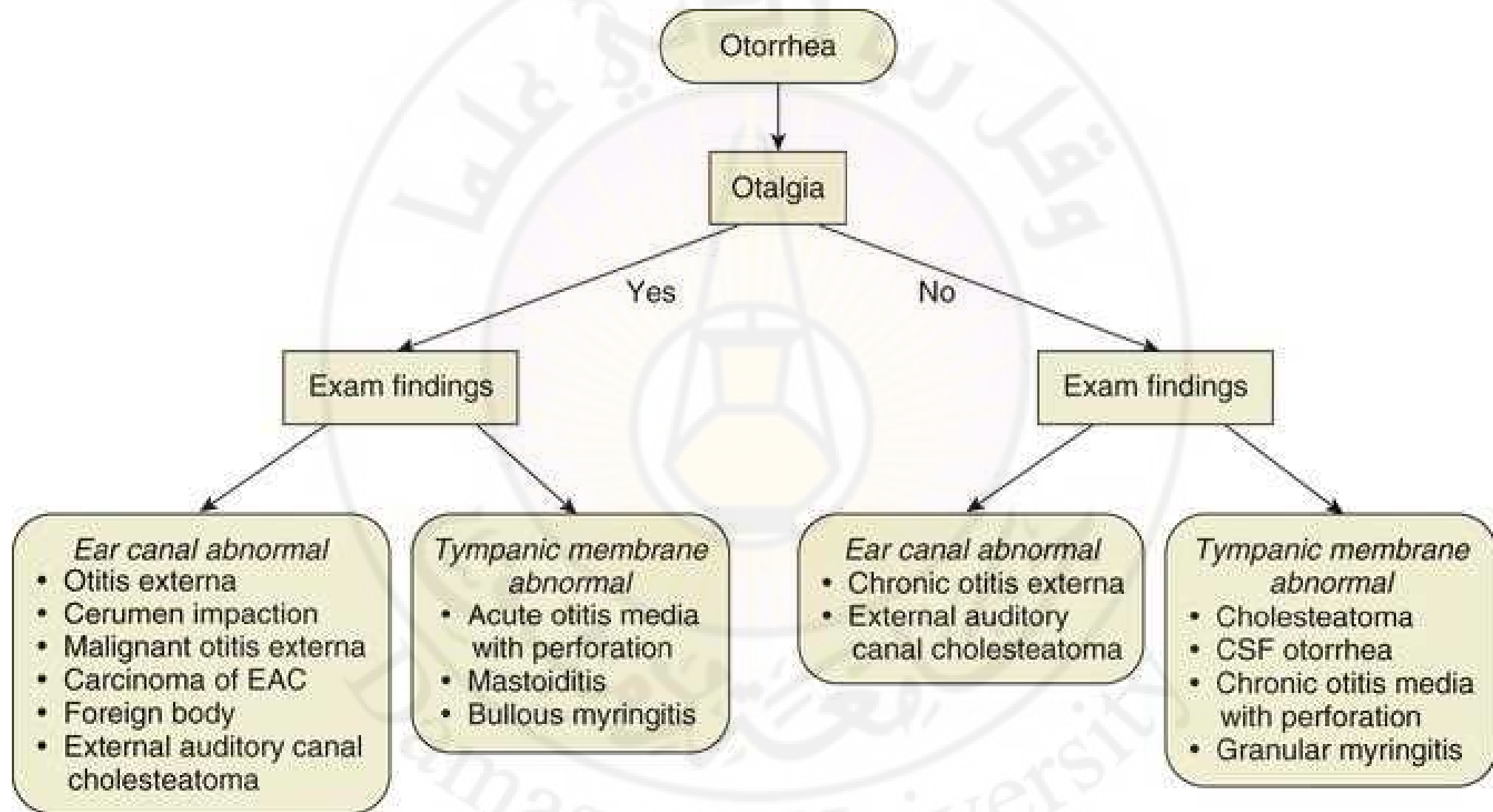
- **Acute otitis externa** is the most common cause of otorrhea and should be easily confirmed by physical exam.
- Physical exam may instead reveal otitis media, which may be acute or chronic drainage via ventilation tube or perforated tympanic membrane (TM).
- **Chronic painless otorrhea** should raise one's clinical suspicion for cholesteatoma.
- Sometimes, chronic otorrhea may be secondary to chronic otitis externa.
- Moisture from de-epithelialized regions of the ear canal or TM may also accumulate and occasionally drain. The appearance of mucosa-like areas or myringitis should be sought in the setting of an intact TM or dry perforation.

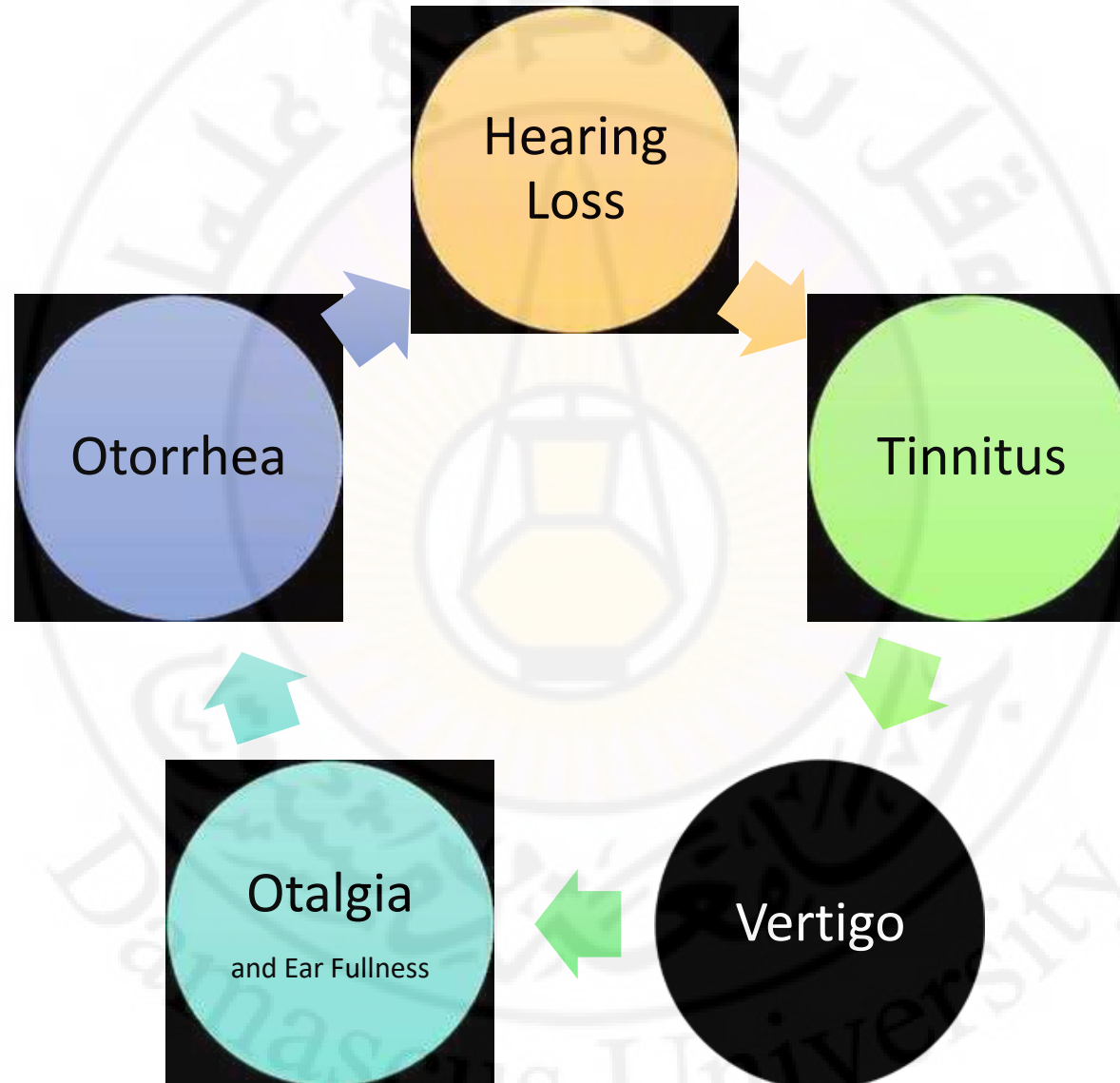


Otorrhea

- **Cerebrospinal fluid (CSF) otorrhea** has a distinct presentation. It is clear and copious. The patient may report prior head trauma or temporal craniotomy, but it may also occur spontaneously.
- CSF otorrhea may occur during activities or positions which raise intracranial pressure such as the Valsalva maneuver.
- CSF otorrhea should be ruled out in any patient having copious watery otorrhea after ventilation tube placement or any patient with a pulsatile clear copious otorrhea at the time of tube placement.
- If it is feasible to collect the otorrhea, it should be tested for beta-transferrin.
- The middle fossa floor should be examined for the location of the leak via high-resolution CT of the temporal bone.







5. Vertigo

- **Vertigo** is a symptom, rather than a condition itself. It's the sensation that you, or the environment around you, is moving or spinning. This feeling may be barely noticeable, or it may be so severe that you find it difficult to keep your balance and do everyday tasks
- Understanding the nature of a patient's vertigo is critical to developing a differential diagnosis, evaluation scheme, and treatment plan.
- Patient history accounts for 85% of the information considered in the diagnosis of the dizzy patient





History Taking for a Chief Complaint of Dizziness

Vivid description of the dizziness	Neurologic symptoms
Onset: Timing; situation?	Extremity weakness, numbness Cranial nerve symptoms Headaches Memory loss Personality change Visual symptoms Auras Diplopia Light sensitivity
Presence: Continuous vs intermittent; duration	
Mitigating factors	
Exacerbating factors	
Associated ear symptoms	
Fullness	
Hearing loss	
Tinnitus	
Autophony	Medication use

Vertigo

- Essential in the query of the dizzy patient is determining if they are, in fact, **vertiginous** or if their symptoms are pre-syncope, imbalance, lightheadedness, or even just a vague sense of disequilibrium. Ask the patient to describe what they are actually feeling. Disallow the use of the nondescriptive term “dizzy.”
- Second, clarify the **temporal cues** to the patient's symptoms: When did it start? Is it continuous or episodic? How long do the episodes last? How often do they occur? When was the last episode? Do the symptoms completely resolve between episodes?
- Next, ask patients about **triggers** (e.g., rolling over in bed as a classic trigger of vertigo with benign paroxysmal positional vertigo). What makes it better? What makes the vertigo worse?
- Lastly, explore **associated symptoms**. If the patient is experiencing labyrinthine vertigo, aural symptoms may accompany the vertigo and can assist the clinician in formulating a differential diagnosis. Aural fullness, tinnitus (pulsatile or ringing), autophony, and perceived hearing loss should be correlated with the spells if present. True labyrinthine vertigo may have accompanying autonomic symptoms, classically nausea/vomiting, but occasionally diarrhea, sweating, or flushing are also present.

Vertigo

- **Neurologic symptoms** such as headache, visual aura, photophobia/phonophobia, extremity weakness or numbness, or other CN deficits should NOT be present with true labyrinthine vertigo and are suggestive of neurologic etiologies such as a posterior circulation cerebrovascular issue, neoplasm, or migraine headache.
- **Loss of consciousness** is not associated with labyrinthine vertigo. Patients who endorse syncope or pre-syncope should be evaluated for other causes of their symptoms. Cardiac causes are common in this population and should be evaluated by the appropriate specialist.



Vertigo

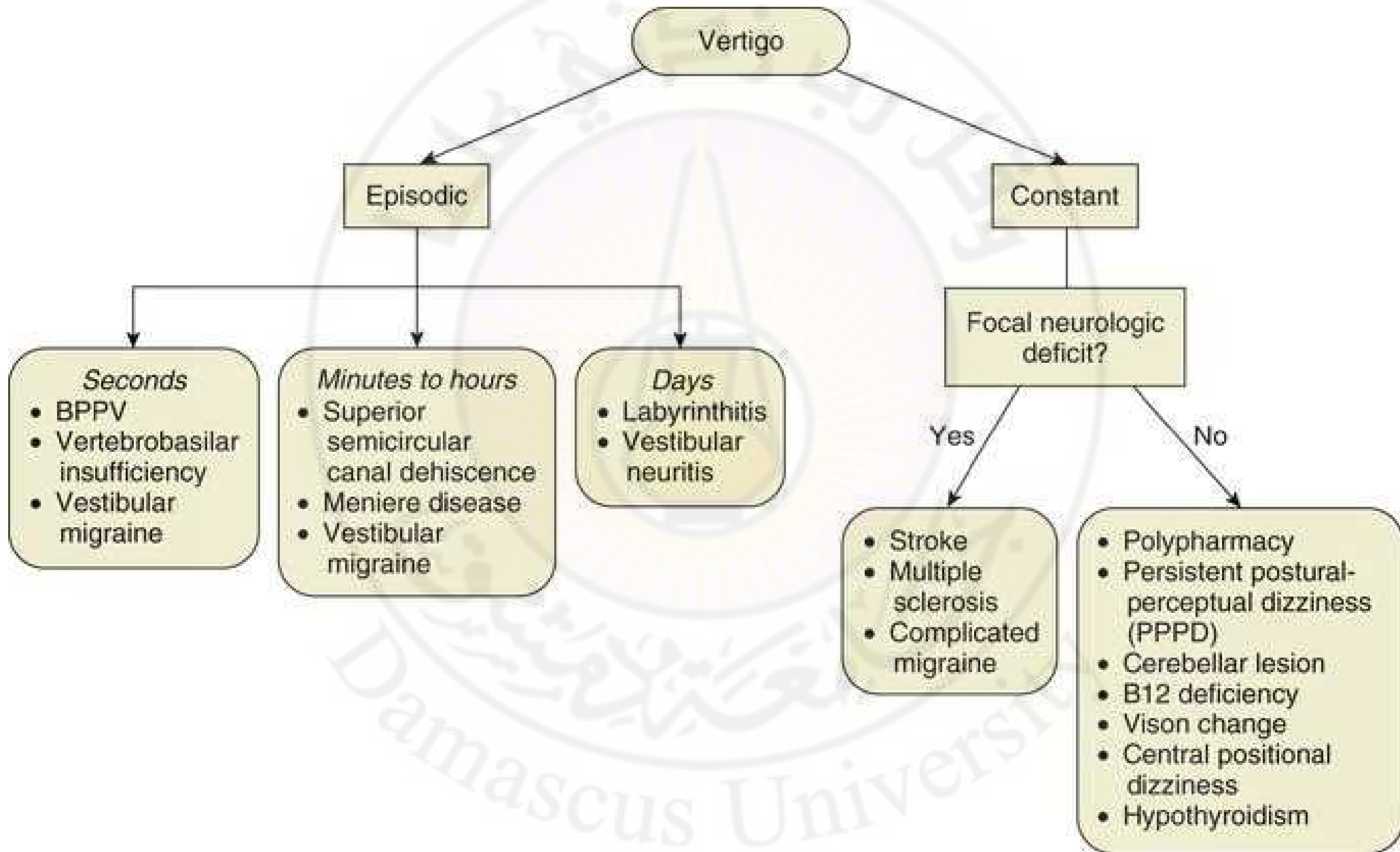
- **Imbalance** as a symptom should also be explored, especially those factors that influence stability or ability to compensate after vestibular insult.
- A patient's sense of balance and equilibrium is affected by all peripheral inputs from their environment. Therefore visual acuity, cervical immobility/pain, strength of core (abdominal) and leg muscles, ankle mobility/flexibility, and peripheral sensation (feet, toes) may all impact a patient's stability.
- The **brain** is responsible for receiving and integrating all this information to keep the individual upright and moving. While many patients can adapt to loss of or alterations in input from one of these systems (visual, proprioceptive, or vestibular), alterations to two or more systems significantly impair the ability of the CNS to accurately interpret the signals and maintain a sensation of normal equilibrium.



Vertigo

- **Polypharmacy** is a significant cause of balance dysfunction and is exceptionally common, particularly in older patients or those with multiple medical problems.
- Specifically, the presence of psychotropic medications or vestibular suppressants should be asked.
- Patients may have numerous medications listed as “as needed,” but some patients will take those medications without interruption, several times a day. This not only affects the patient's sense of equilibrium on an ongoing basis but also impacts the in-office exam and subsequent objective testing such as nystagmography or rotary chair examinations.







Clinical Audiology

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Physical examination



Physical Exam

- The physical exam is a key component in the diagnosis of ear related symptoms.
- The exam can be divided into these different areas: Pinna, external auditory canal (EAC), TM, middle ear, auditory system, and vestibular system.
- Examination of most of these anatomic areas involves both visual inspection and functional assessments.



Direct Examination of the Ear

- Wash your hands or wear gloves
- Preferably, sit at the level of the patient and in such a way that you have access to both ears
- First, check the earlobe (abnormalities and skin changes).
- Check for scars or pits in front or behind the earlobe
- If the patient wears a hearing aid, inform him and remove the hearing aid with his permission.
- Touch the earlobe slowly and check the possibility of inflammation.



FIG. 137.7 Normal pinna. (1) tragus, (2) antitragus, (3) root of the helix, (4) helix, (5) concha cavum, (6) concha cymba, (7) triangular fossa, (8) lobule, (9) antihelical fold, (10) external auditory canal meatus.

Pre-auricular pits or tags should be noted, and the patient should be asked if they ever have drainage or swelling around the pit.



Skin tag /
accessory
auricle

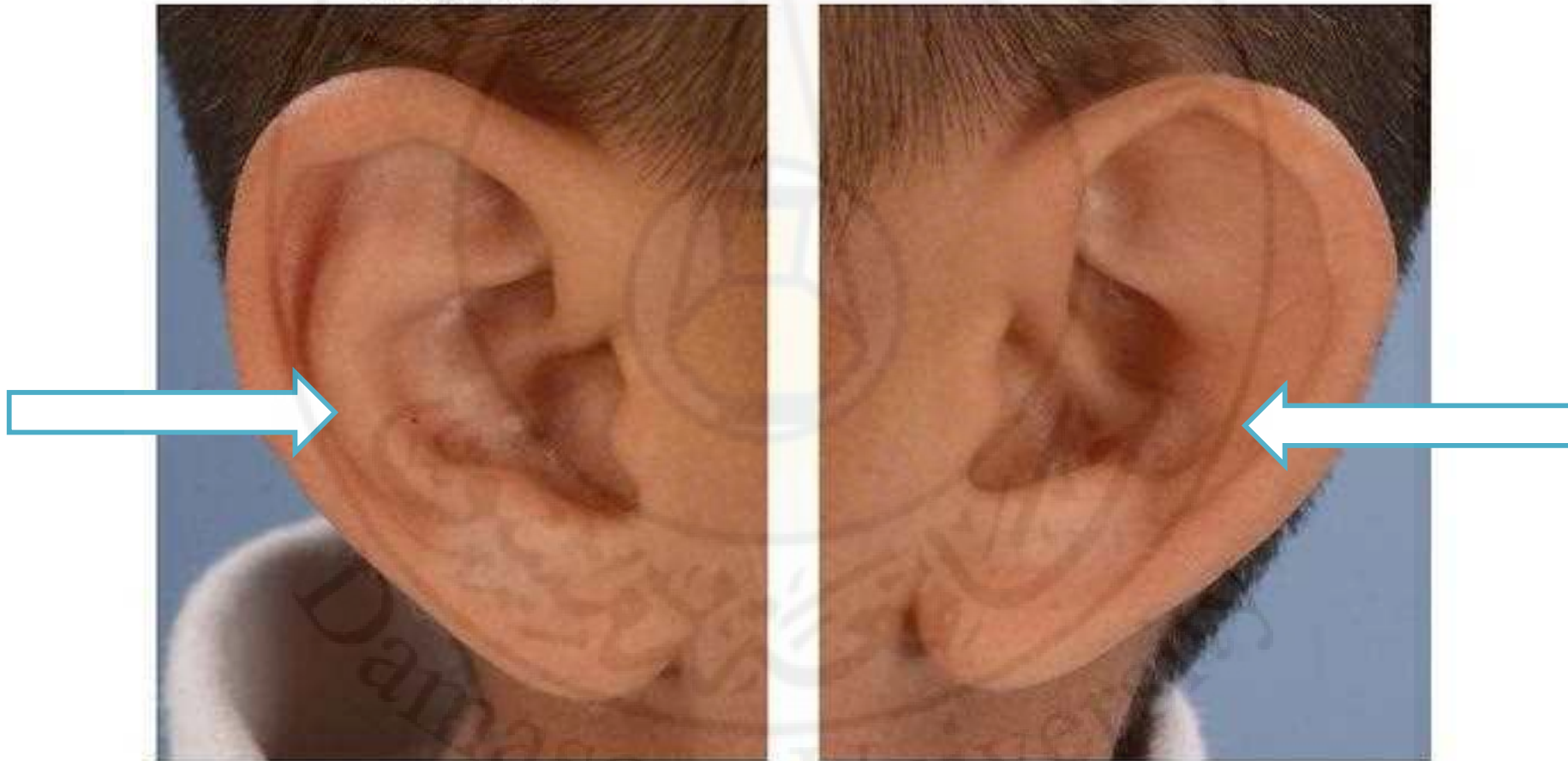


Preauricular pit

Loss of antihelical fold

RIGHT EAR

LEFT EAR



Microtia & Anotia



Grade 1

Smaller than normal, but the ear has mostly normal anatomy



Grade 2

Part of the ear looks normal, usually the lower half

The canal may be normal, small or completely closed



Grade 3

Just a small remnant of "peanut-shaped" skin and cartilage

There is no canal, which is called aural atresia



Grade 4

Complete absence of both the external ear and the ear canal, also called "anotia"

Microtia is commonly associated with atresia of the EAC

Cauliflower ear



Injury and devascularization of the helical cartilage due to trauma (i.e., cauliflower ear) results in loss of the cartilage structure and excessive thickening of the perichondrium and a cosmetic deformity.

Perichondritis

Infection or inflammation of the perichondrium is manifest by exquisitely tender erythema and edema of the pinna.

In **infectious** cases of perichondritis, the lobule may also be involved, and the infection can spread into the surrounding periauricular tissues in severe cases.

Non-infectious inflammation, as in autoimmune relapsing polychondritis, may involve both the helical and tragal cartilages and may spare the lobule.

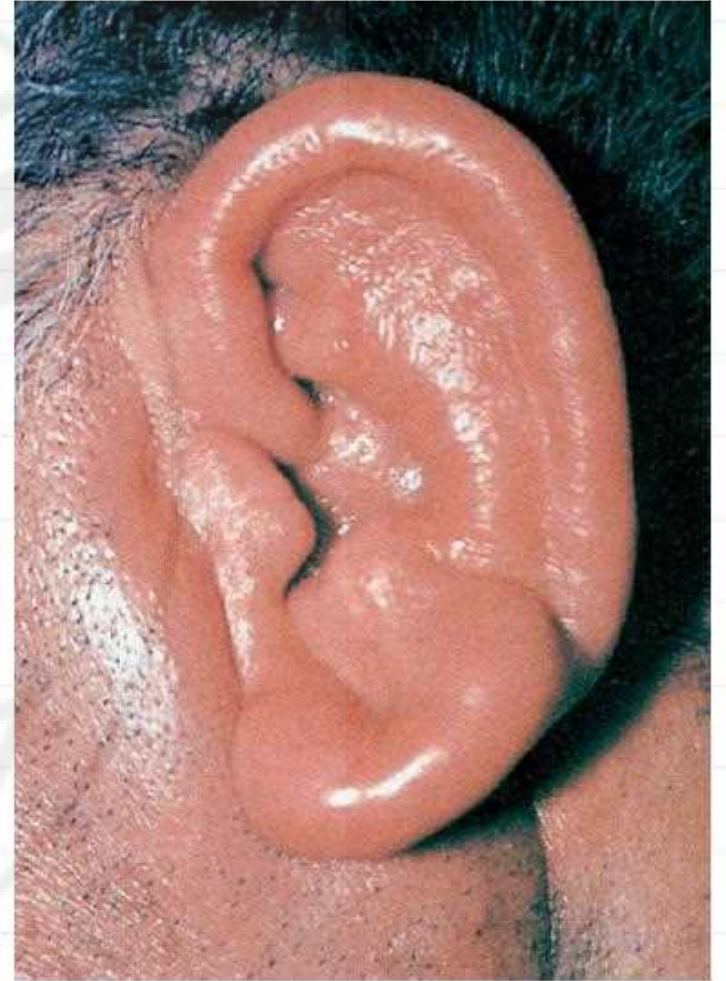




Mastoiditis



Perichondral
haematoma



Erysipelas

Seborrheic dermatitis (ear Eczema)



Examination of the Ear canal & TM

- Inspect the **external auditory canal**.
- Evaluate **tympanic membrane**
 - Note the **color** (red, white, yellow) and **translucency** (transparent, opaque) and **position** (retracted, neutral or bulging) of the drum
 - Identify the **pars tensa** with its **cone of light**, the handle and short process of malleus, and the anterior and posterior folds of the pars flaccida and position of the malleus handle..
- Air inflation otoscopy (**pneumatic-otoscope**) is very useful to evaluate middle ear disease. Assess the mobility of tympanic membrane by applying positive and negative pressures with the rubber squeeze bulb.

Eardrum seen through otoscope



Tools for Examination



Head mirror or Headlight



Otoscope

Otoscope Speculum



Tools for Examination



Video Otoscope



iPhone Otoscope

Tools for Examination



ENT Binocular microscope

Siegel Pneumatic Otoscopy

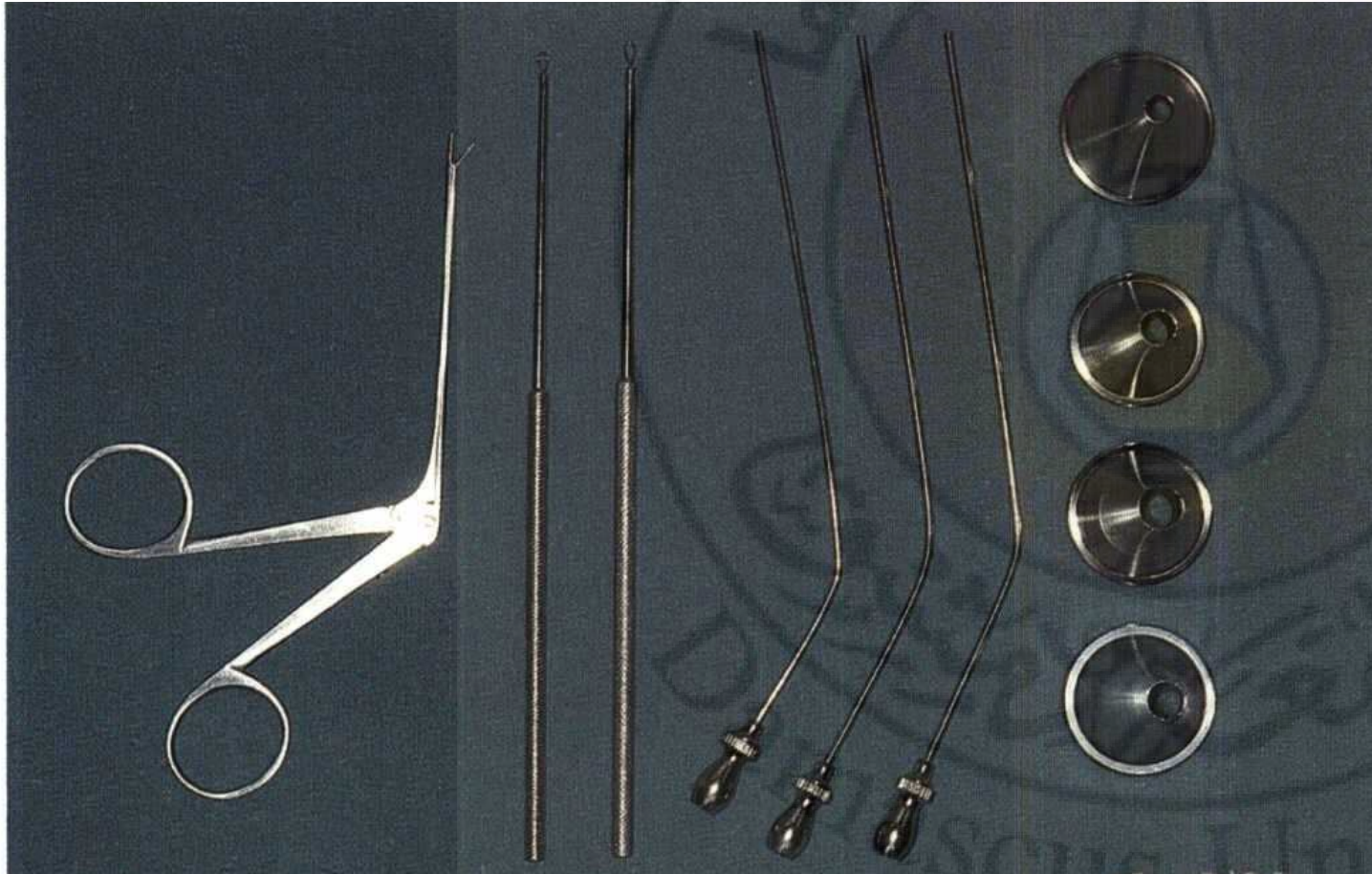


Pneumatic Otoscopy

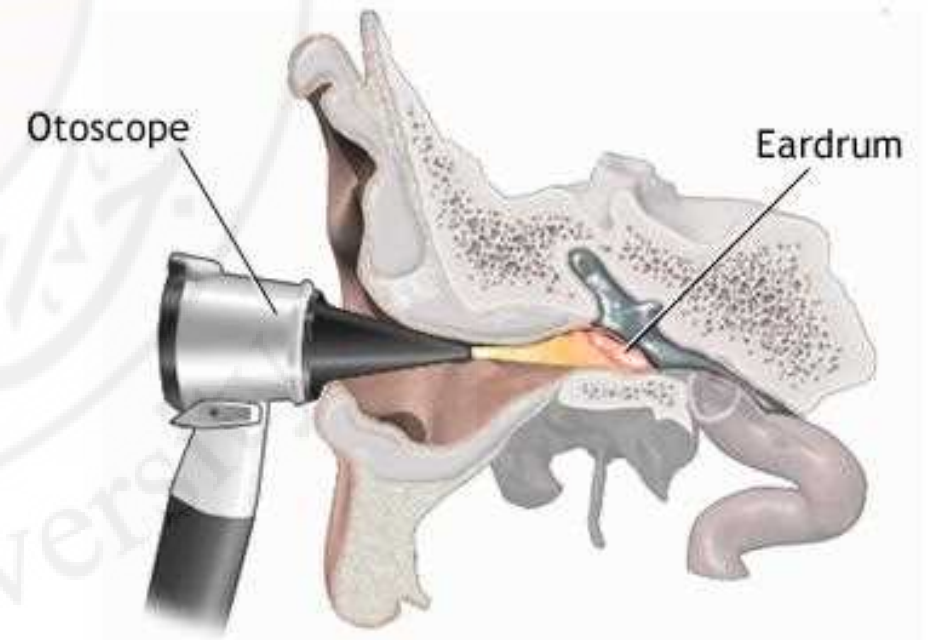
- **Pneumatic otoscopy** allows for visualization of the movement of the ear drum in response to a change in pressure within the ear canal.
- Similar to tympanometry, the movement of the TM depends on the pressure differential of the middle ear space. Unlike tympanometry, the examiner can visualize that movement.
- Relative movement of the TM during insufflation can predict negative middle ear pressure such as seen in eustachian tube dysfunction, or otitis media with effusion.
- This test can also be used to apply enough differential pressure for a bedside **Hennebert's (fistula) test**.



Make sure that ear canal is clean and open before otoscopy



- Aural speculi
- Billeau ear loop
- Hartman auricular forceps
- Suction tips



Important points during otoscopy

- Make sure that the speculum is clean and disinfected
- Choose the largest possible speculum that fits comfortably in the ear.
- Preferably hold the otoscope like a pencil
- To examine the right ear, hold the otoscope in the right hand and vice versa.
- During an adult examination, pull the earlobe up and back to straighten the ear canal.
- First, examine the better ear. In this way, you reduce the possibility of spreading a possible infection from the worse ear to the better one.



Maneuvering the ear in otoscopy



Correct



Incorrect

The otoscopic exam is performed by gently pulling the auricle upward and backward.

Maneuvering the ear in otoscopy



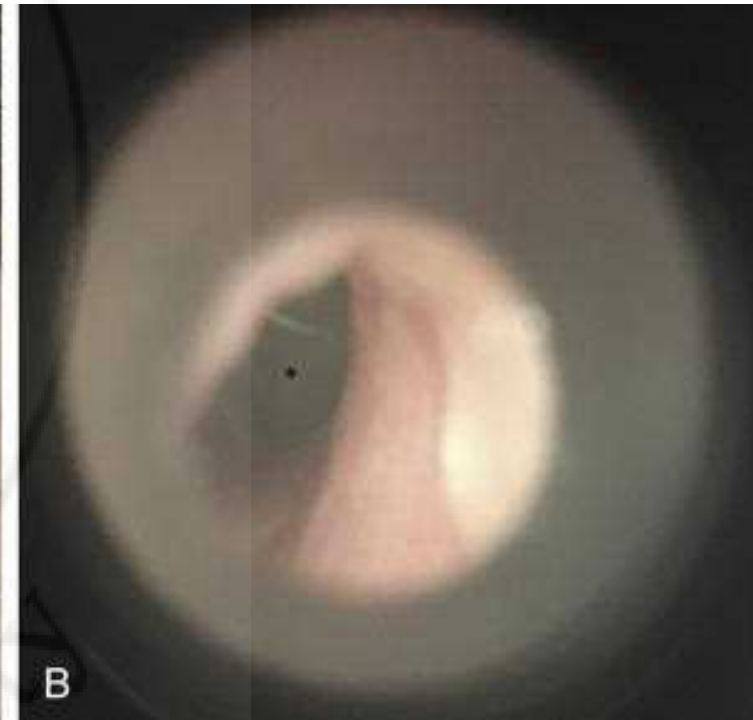
Maneuvering the ear in children's otoscopy



In children, the auricle should be pulled downward and backward. This process will move the acoustic meatus in line with the canal.

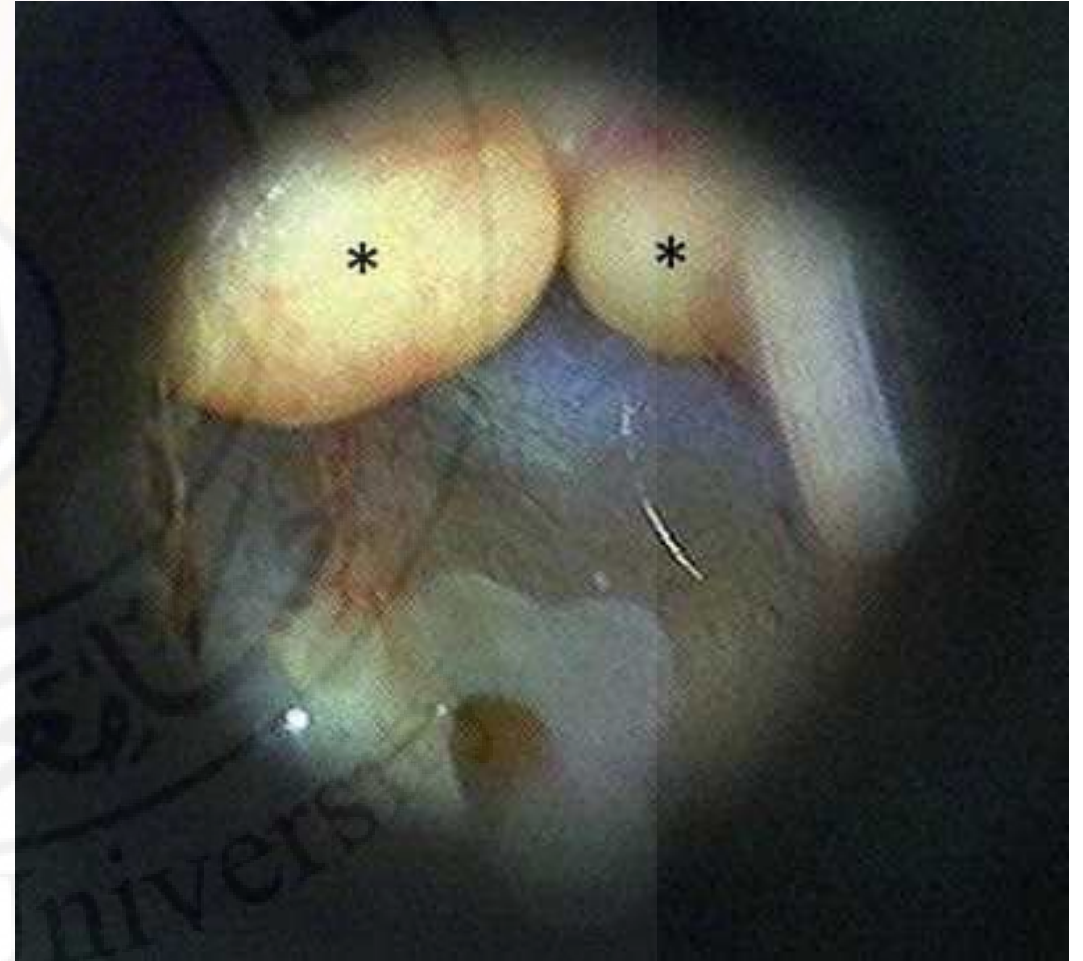
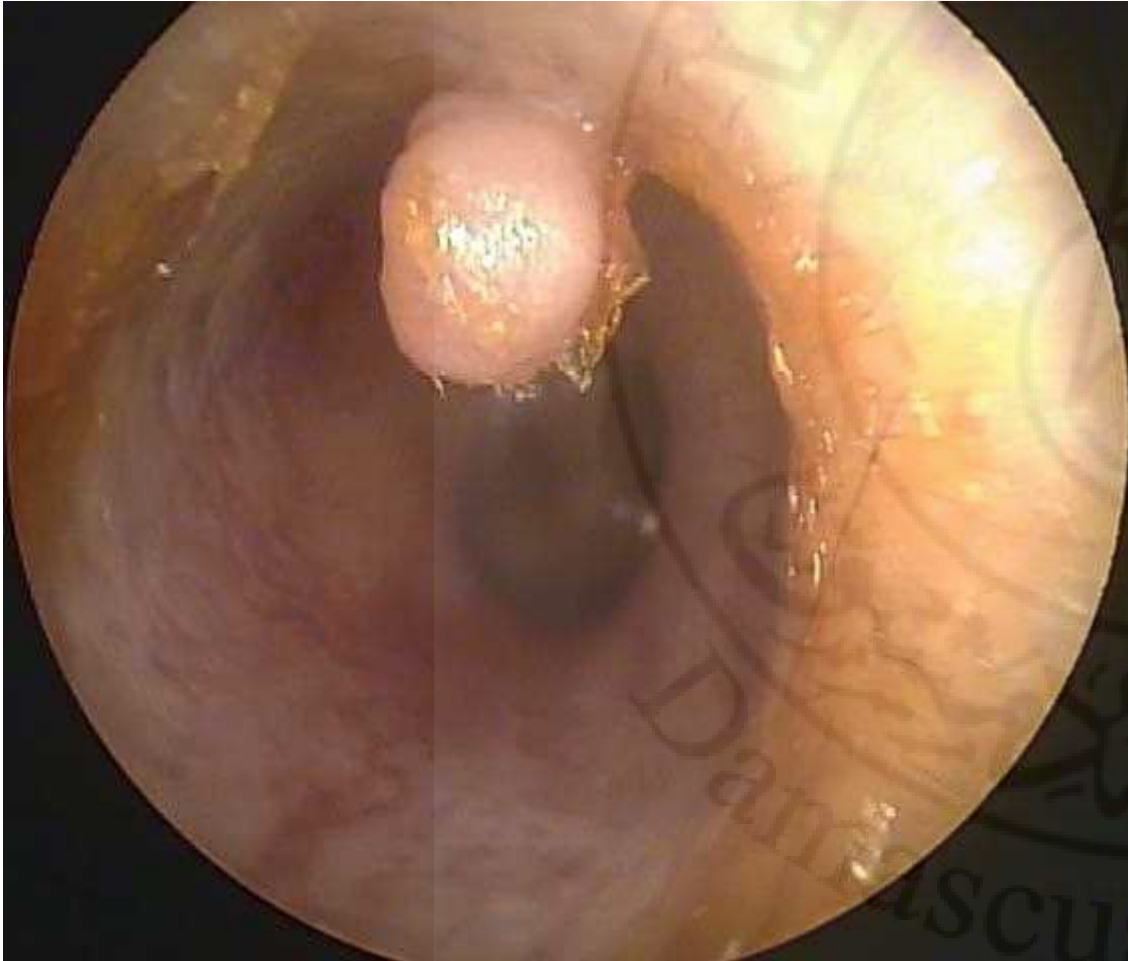
The External Auditory Canal

- Infections (Bacterial, Fungal)
 - Edema
 - Erythema
 - Discharge
- Neoplastic processes
- Mass lesions of the EAC
 - Osteomas
 - Cholesteatomas eroding into the EAC
- Canal stenosis & Exostosis
- Congenital obliteration of EAC

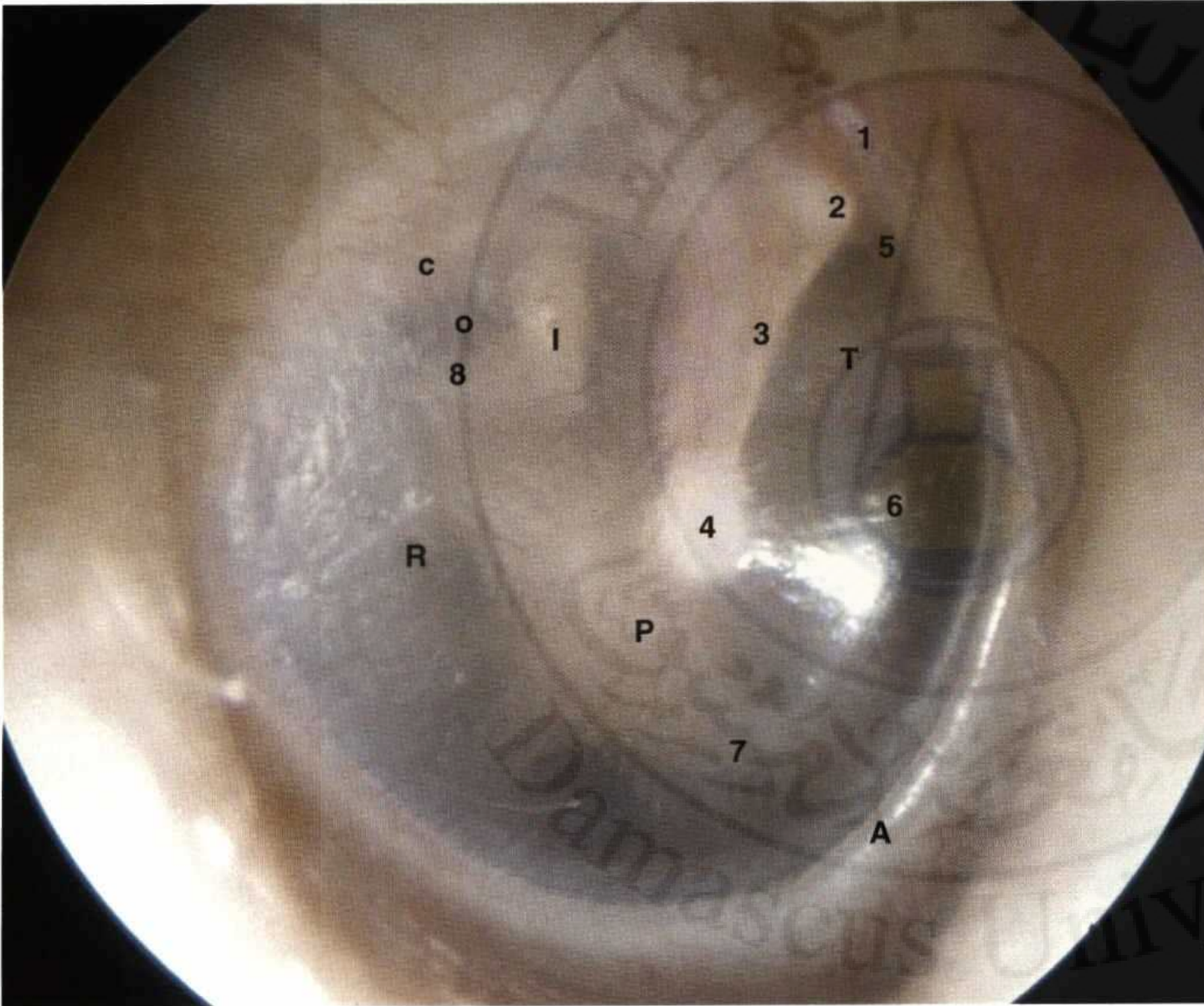


Severe otitis externa

Osteoma of external auditory canal

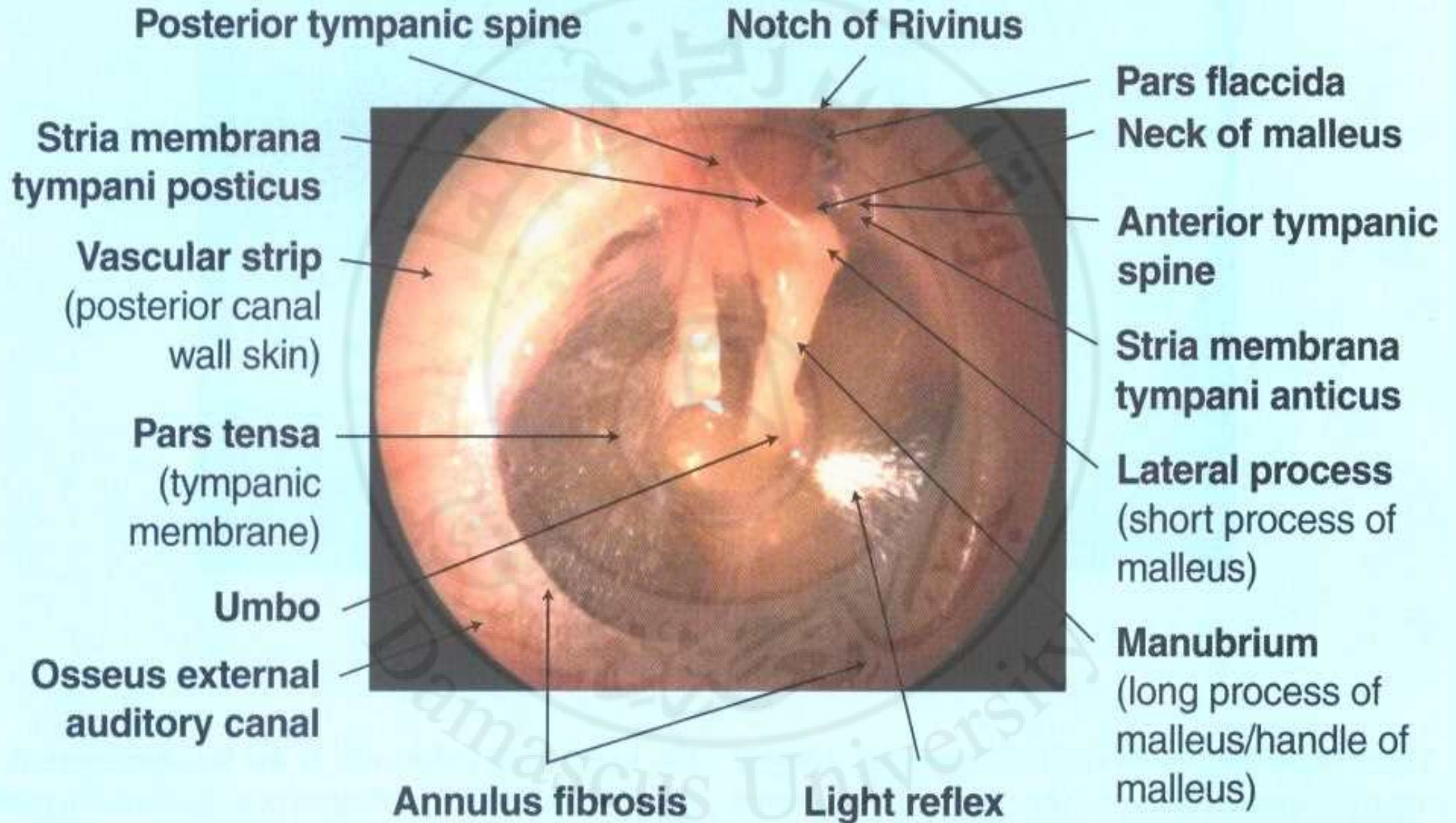


Normal tympanic membrane



- Pinkish gray in color , translucent and in neutral position.
- Malleus lies in oblique position behind the upper part of drum.
- Mobile with air inflation.

Figure 2.1 Right ear. Normal tympanic membrane. 1 = pars flaccida; 2 = short process of the malleus; 3 = handle of the malleus; 4 = umbo; 5 = supratubal recess; 6 = tubal orifice; 7 = hypotympanic air cells; 8 = stapedius tendon; c = chorda tympani; l = incus; P = promontory; o = oval window; R = round window; T = tensor tympani; A = annulus.



Chorda tympani nerve

Long crus (process) of incus

**Lenticular
process of incus**

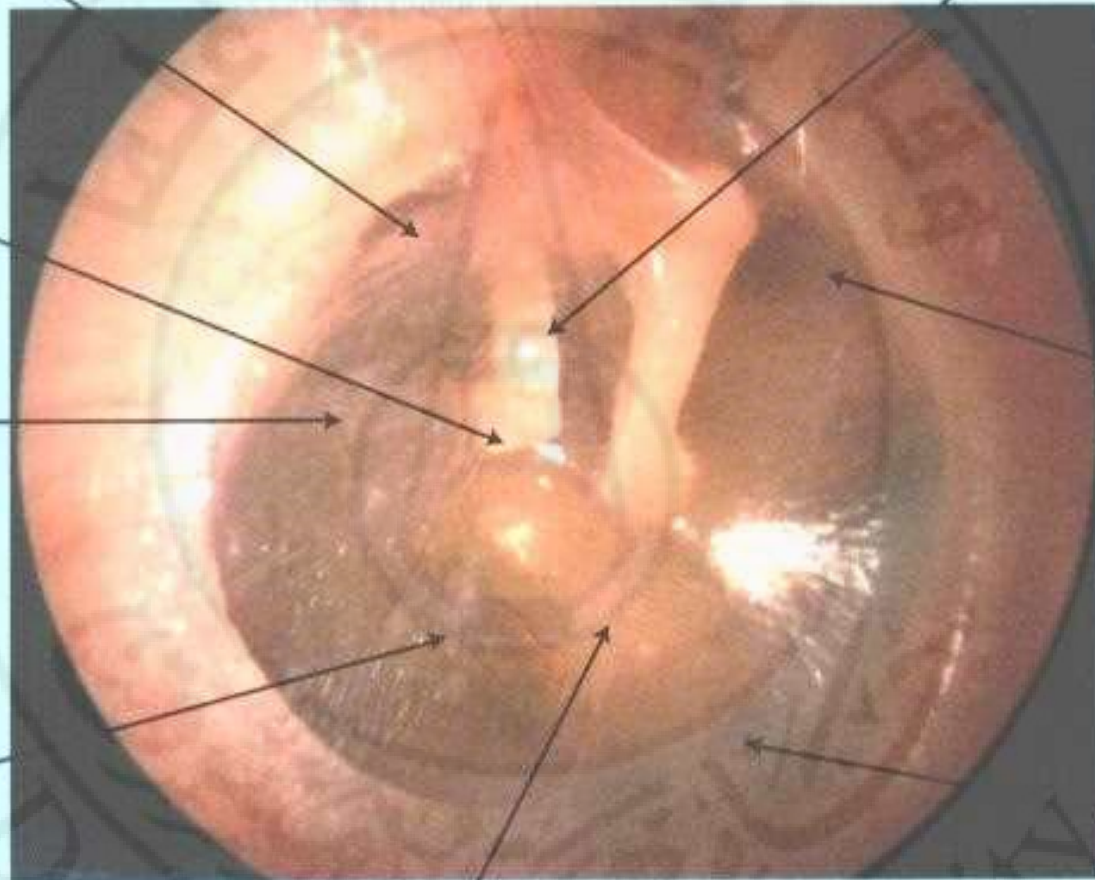
Stapes

**Protympanum
(tubotympanum)**

**Fossula fenestrae
cochleae (round
window niche)**

Hypotympanum

Promontory



Normal tympanic membrane

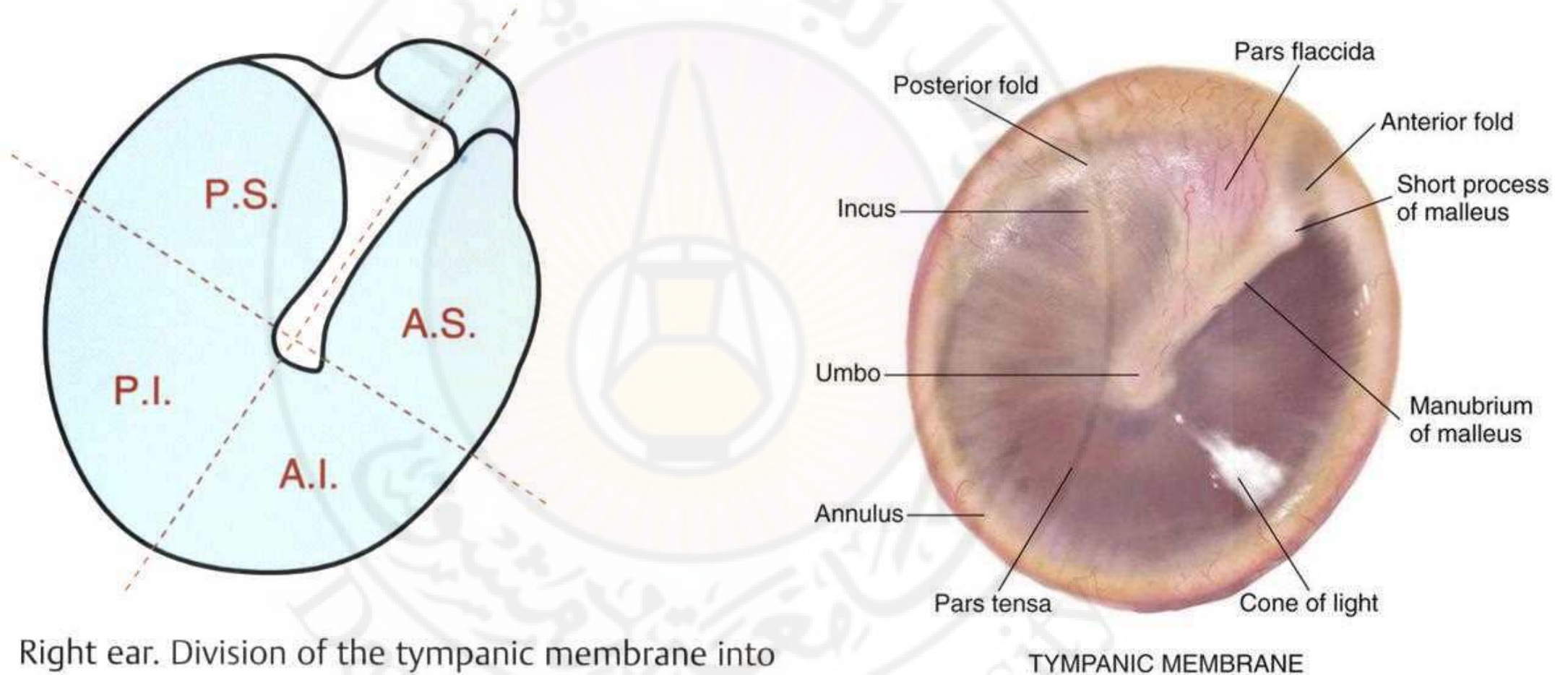


Figure 2.3 Right ear. Division of the tympanic membrane into four quadrants: A.S. = anterosuperior; A.I. = anteroinferior; P.S. = posterosuperior; P.I. = posteroinferior. This division facilitates the description of different pathologic affections of the tympanic membrane.

Normal tympanic membrane

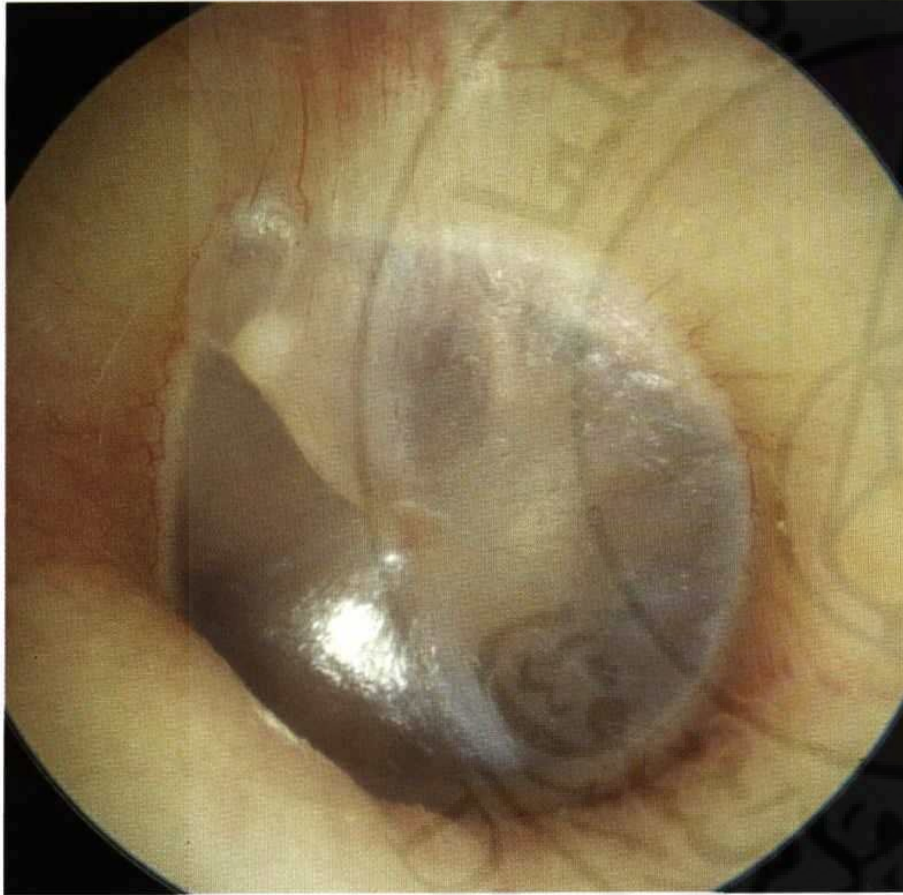


Figure 2.6 Left ear. Normal tympanic membrane. The handle of the malleus and cone of light are well visualized through the tympanic membrane; the promontory, the area of the round window, and the air cells in the hypotympanum can be appreciated. The pars flaccida is visualized superior to the short process of the malleus.

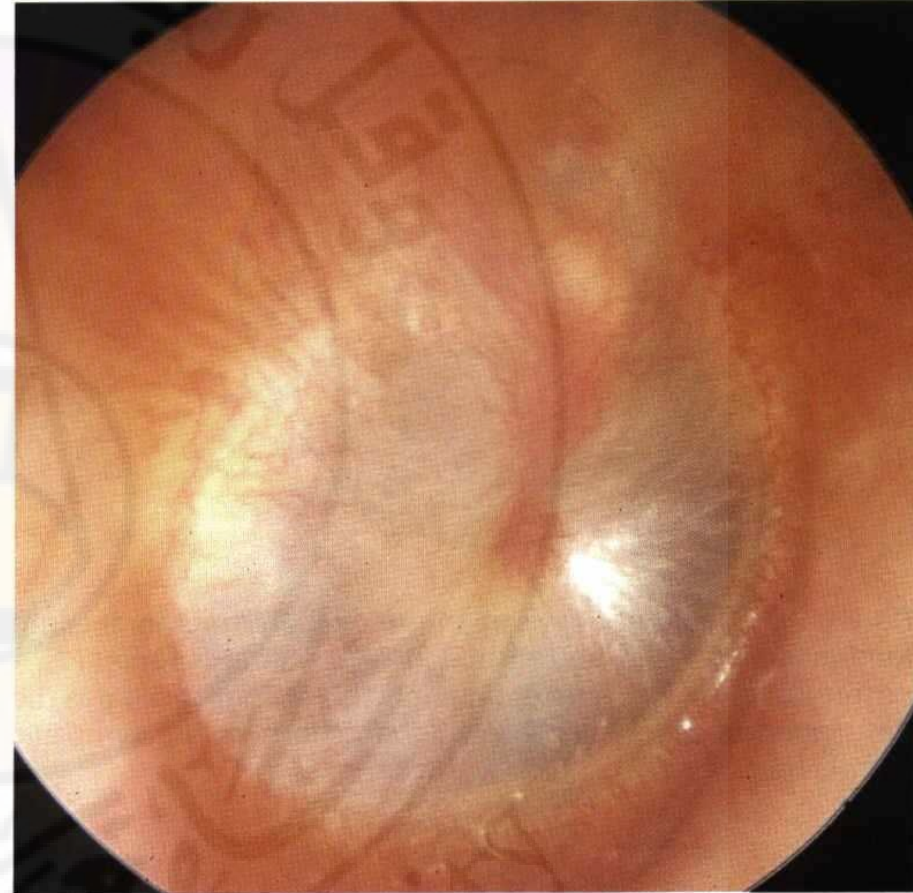
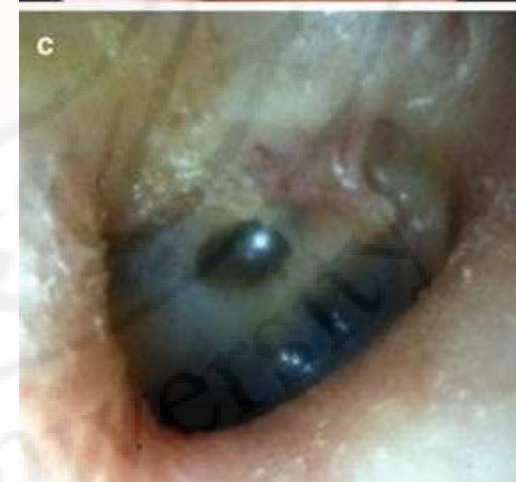
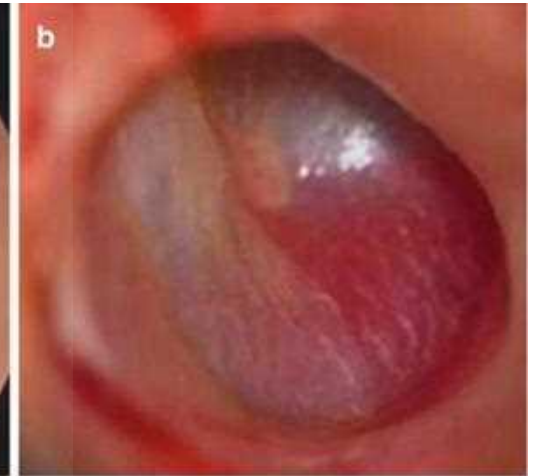


Figure 2.7 Right ear. Normal tympanic membrane. The drum, however, is slightly thickened with an accentuated capillary network along the handle of the malleus. The increased thickness of the tympanic membrane obscures all the structures in the middle ear.

Abnormal tympanic membrane

- The TM should be examined for retraction pockets, perforations, and mobility.
- Color:
 - Red and loss of landmarks (Acute purulent otitis media)
 - Amber: Serous effusion (Barotrauma)
- Bulging: (Acute otitis media)
- Retraction: (Tympanosclerosis)
- Translucent: Air fluid level (Otitis media)
- Hemorrhagic vesicles: (Bullous myringitis)
- Hole: (Perforation)



Abnormal otoscopy: foreign body in the ear canal



Abnormal otoscopy: Canal bony mass (Osteoma)



Abnormal otoscopy: Acute otitis externa



Abnormal otoscopy: Perforation (rupture) of the tympanic membrane



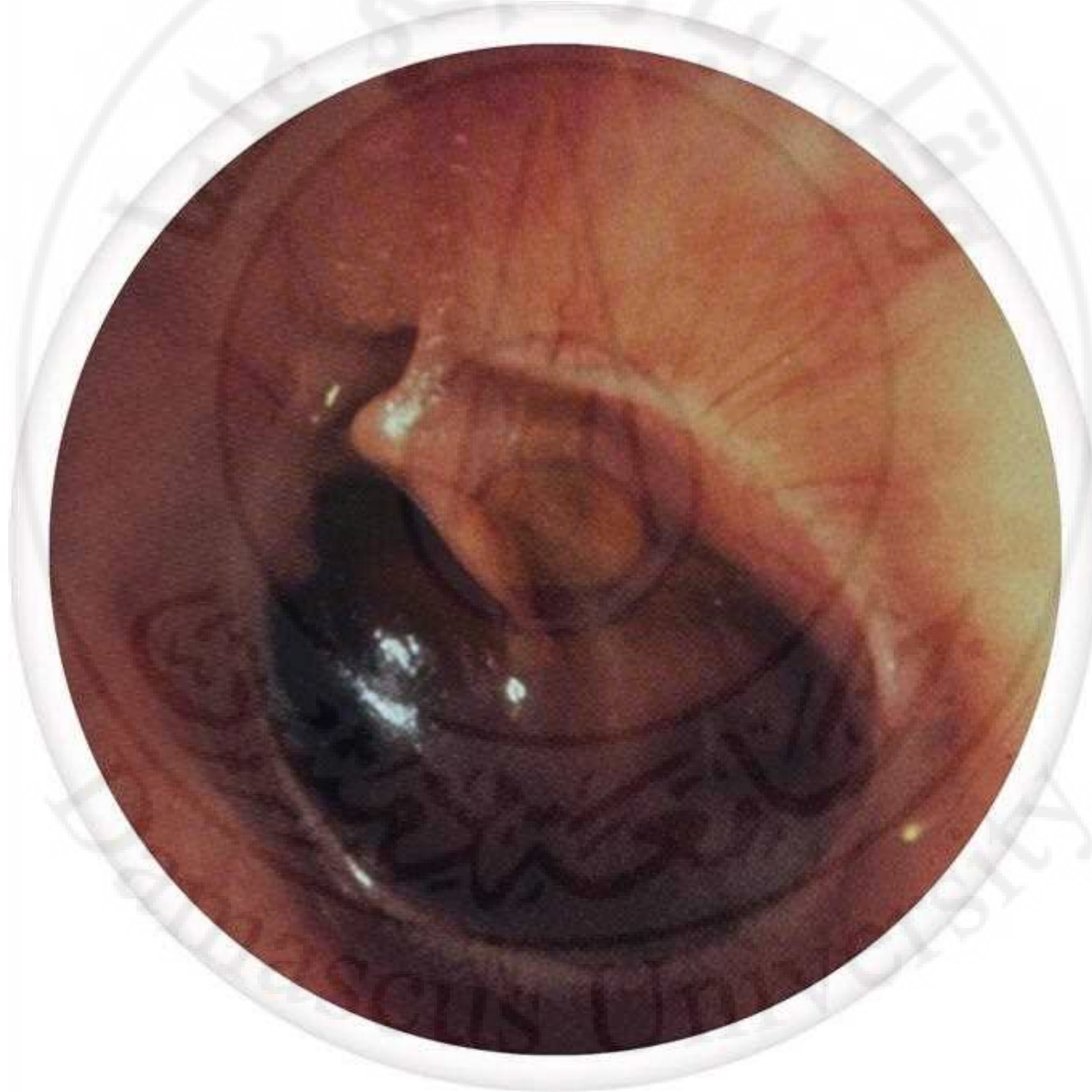
Abnormal otoscopy: Tympanosclerosis



Abnormal otoscopy: Ventilation tube (VT) in the tympanic membrane

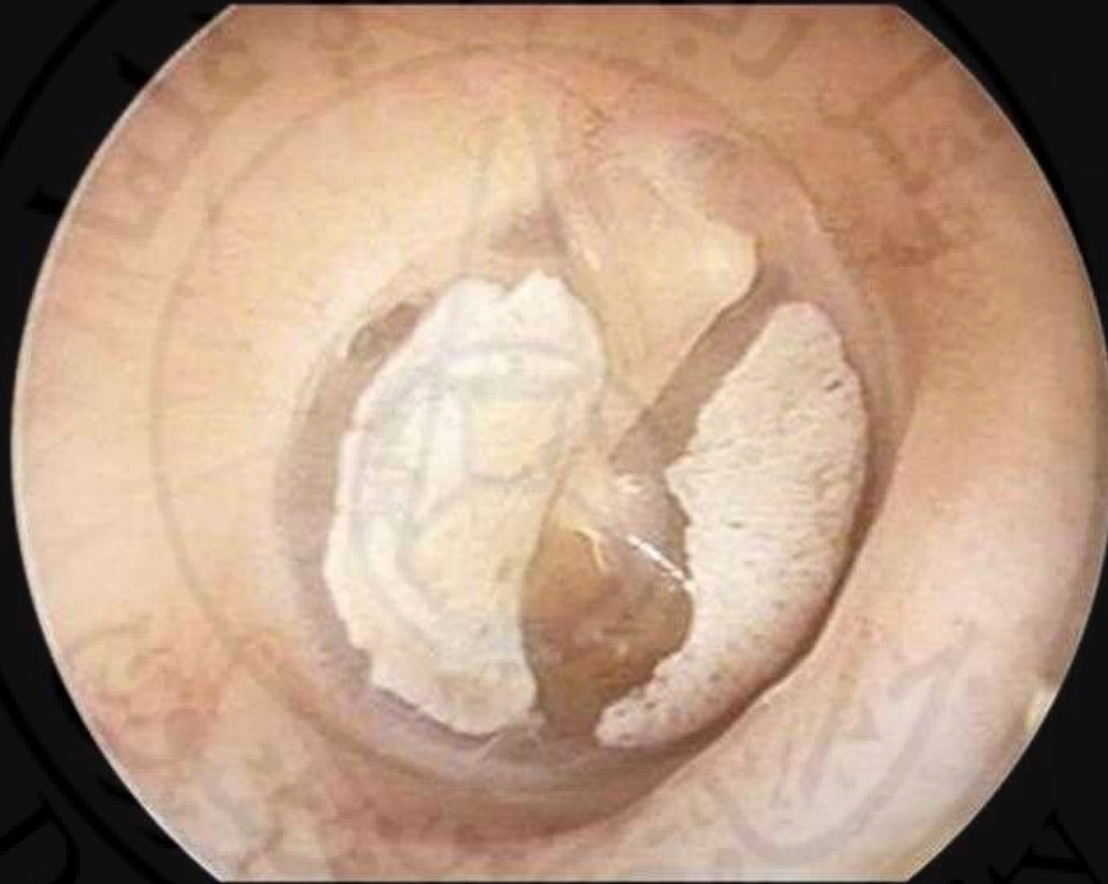


Abnormal otoscopy: Serous otitis media



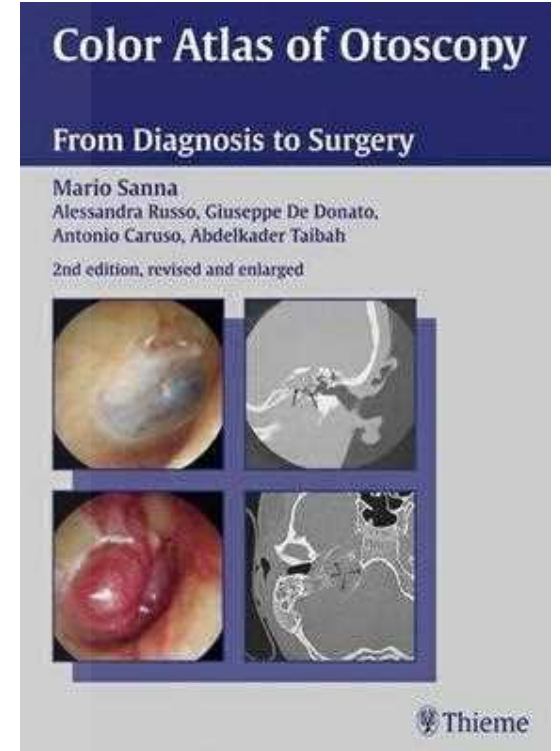
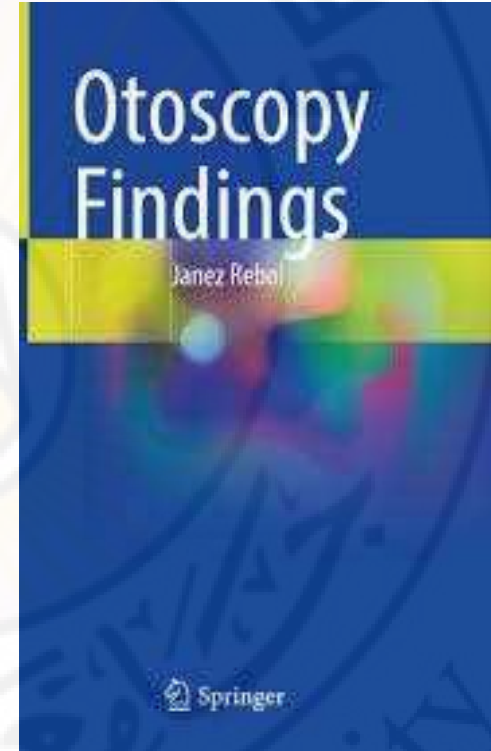
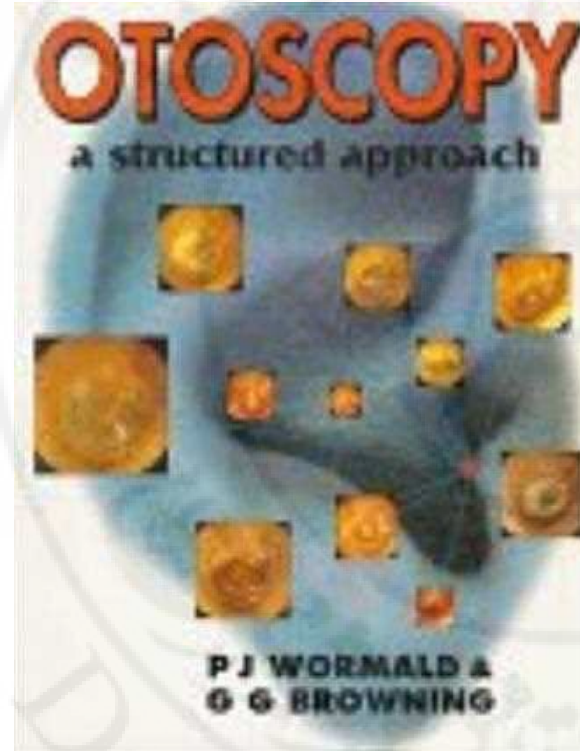
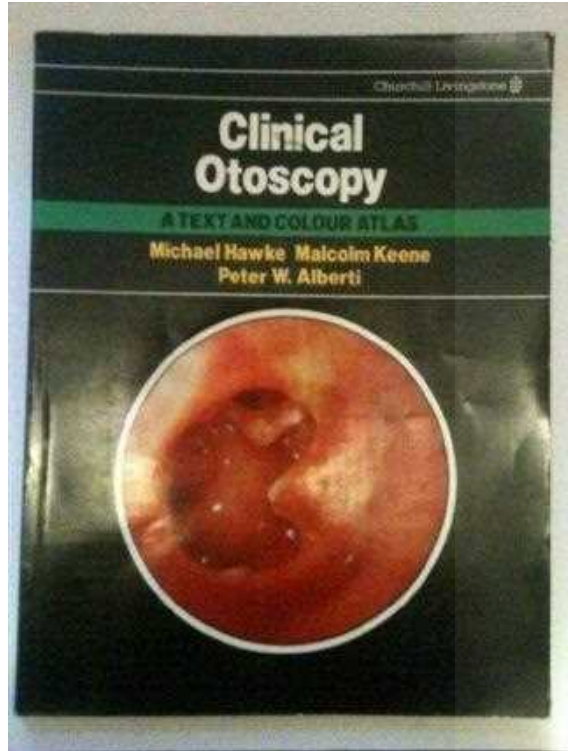


Air Bubbles or Air Fluid Level



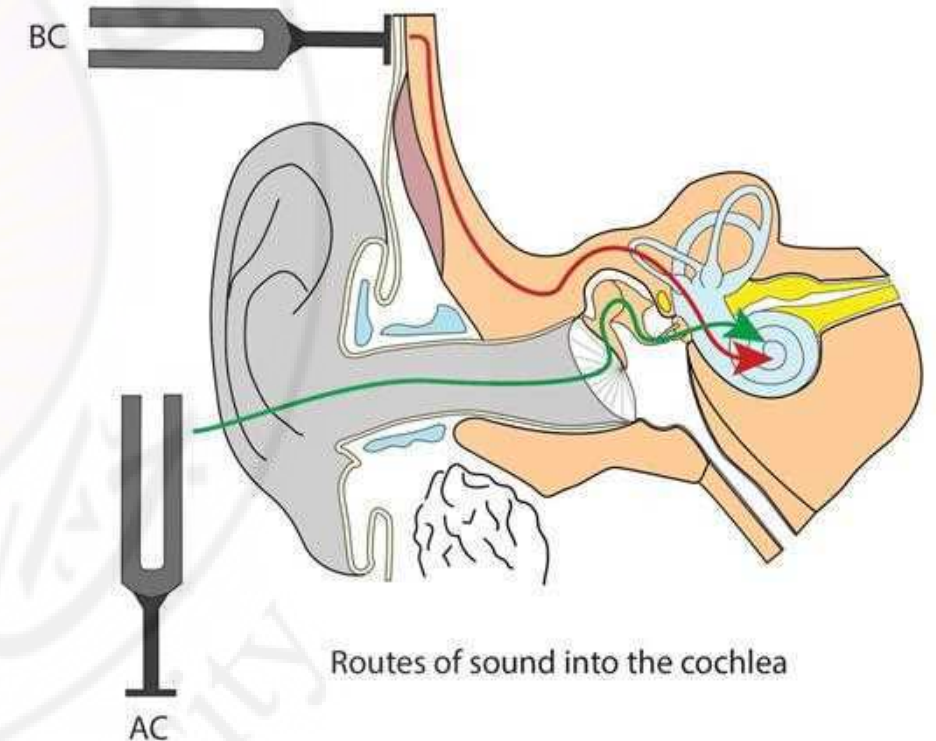
Tympanosclerosis

Suggested reading



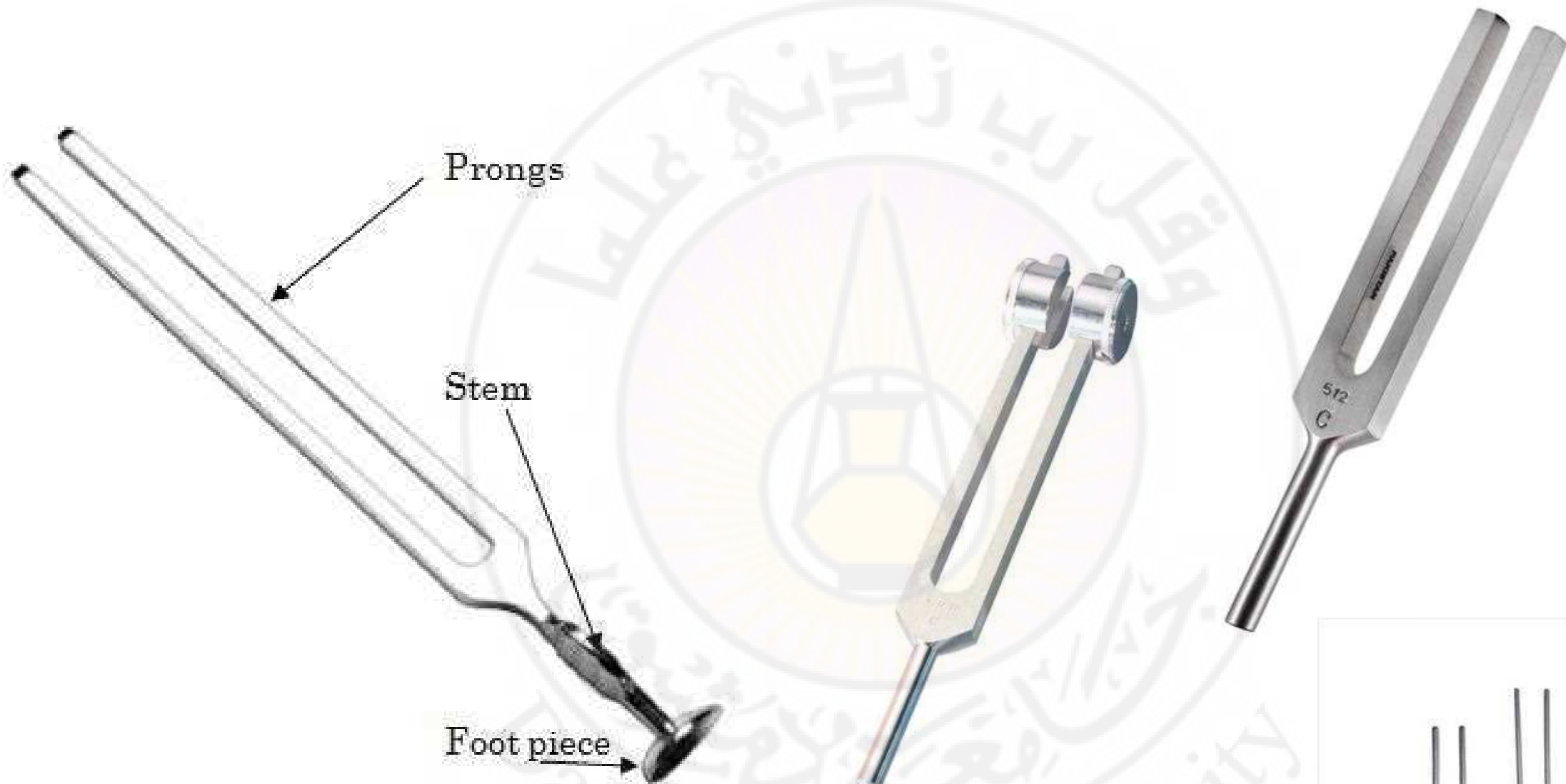
Tuning Forks examination

- A tuning fork examination allows the practitioner to quickly assess the hearing of the patient and provides information about the **type** of hearing loss.
- Additionally, tuning forks are particularly valuable in the situation of a masking dilemma or when there is uncertainty about the viability of the cochlea.
- A complement of frequencies offers the most complete approach to the tuning fork exam and should include 256, 512, and 1024 Hz.
- The two most commonly used tuning fork assessments are the **Weber** and the **Rinne** tests.



Tuning Forks





Weber Test

- It is a type of lateralization test that identifies the sensorineural or conductive origin of **unilateral hearing losses**.
- The tuning fork is vibrates and its base is placed on the person's forehead. Then the person is asked to say on which side they hear the sound (better ear, worse ear, or middle/inside of the head).



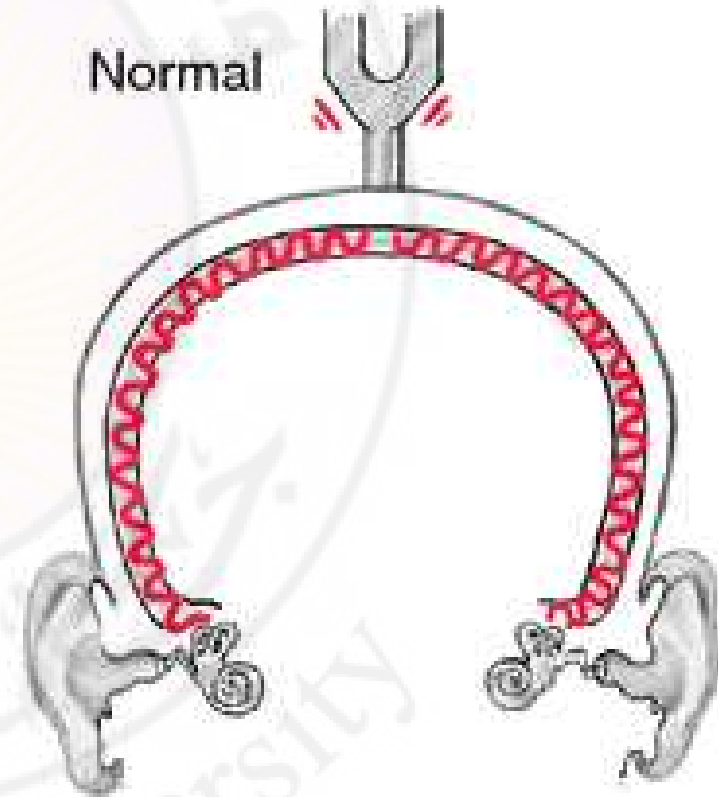
Ernst Heinrich Weber



Interpretation of Weber's test

Normal Hearing:

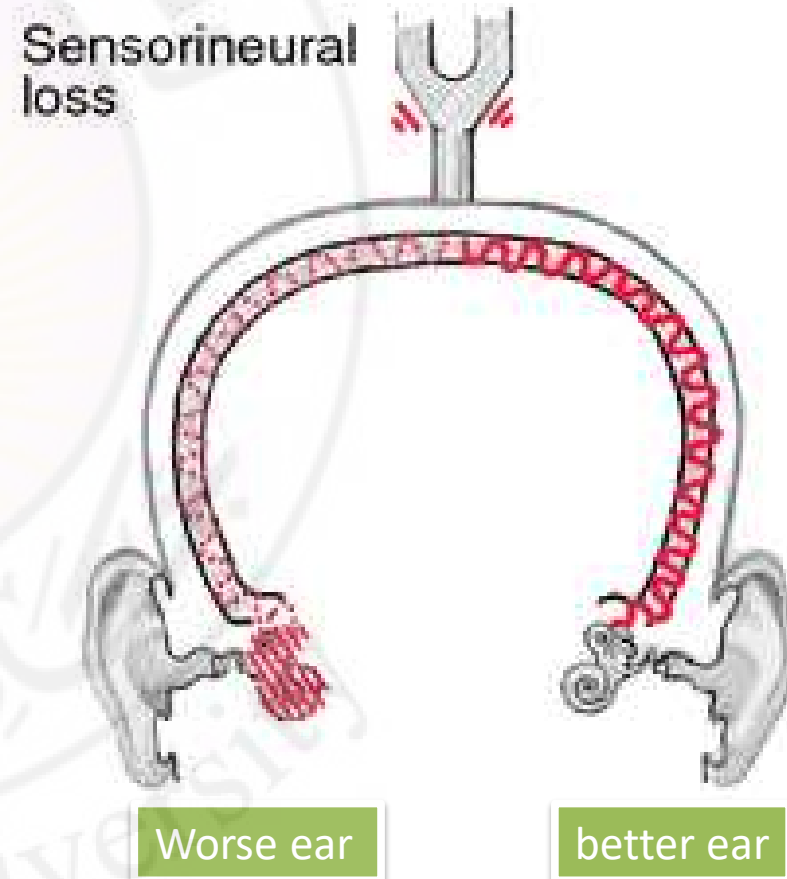
Weber test does not demonstrate lateralization: In a normal subject, the sound should be heard in the middle and equally on both sides.



Interpretation of Weber's test

Unilateral SNHL:

Weber test lateralizes to the unaffected ear. In other words, it is heard louder in the better ear.



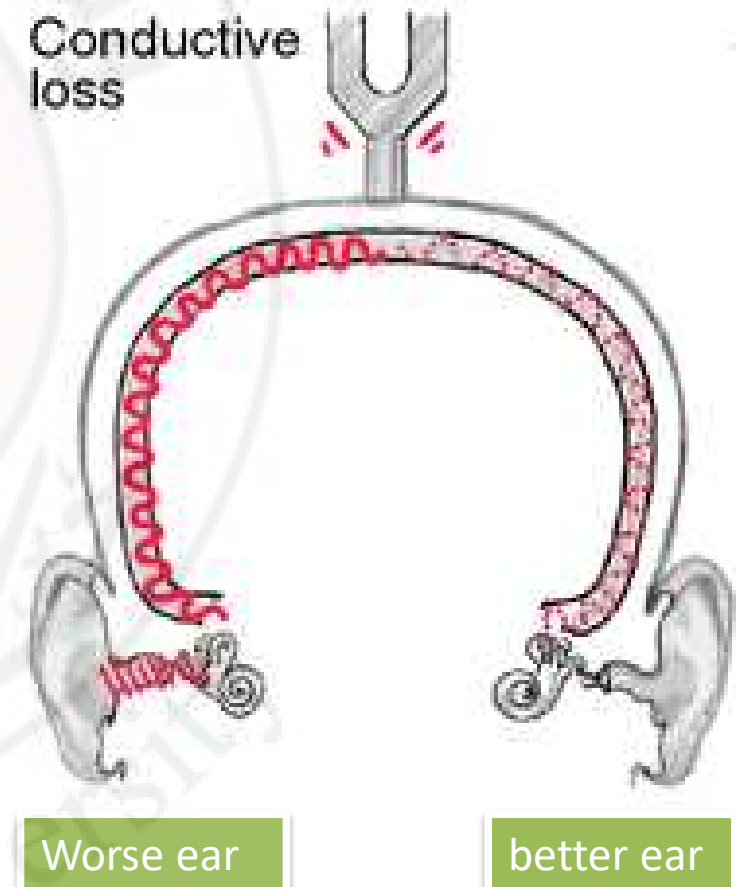
Interpretation of Weber's test

Unilateral CHL:

Weber test lateralizes to the affected ear. In other words, it is heard louder in the poorer ear.

Symmetrical CHL:

Weber test does not demonstrate lateralization.

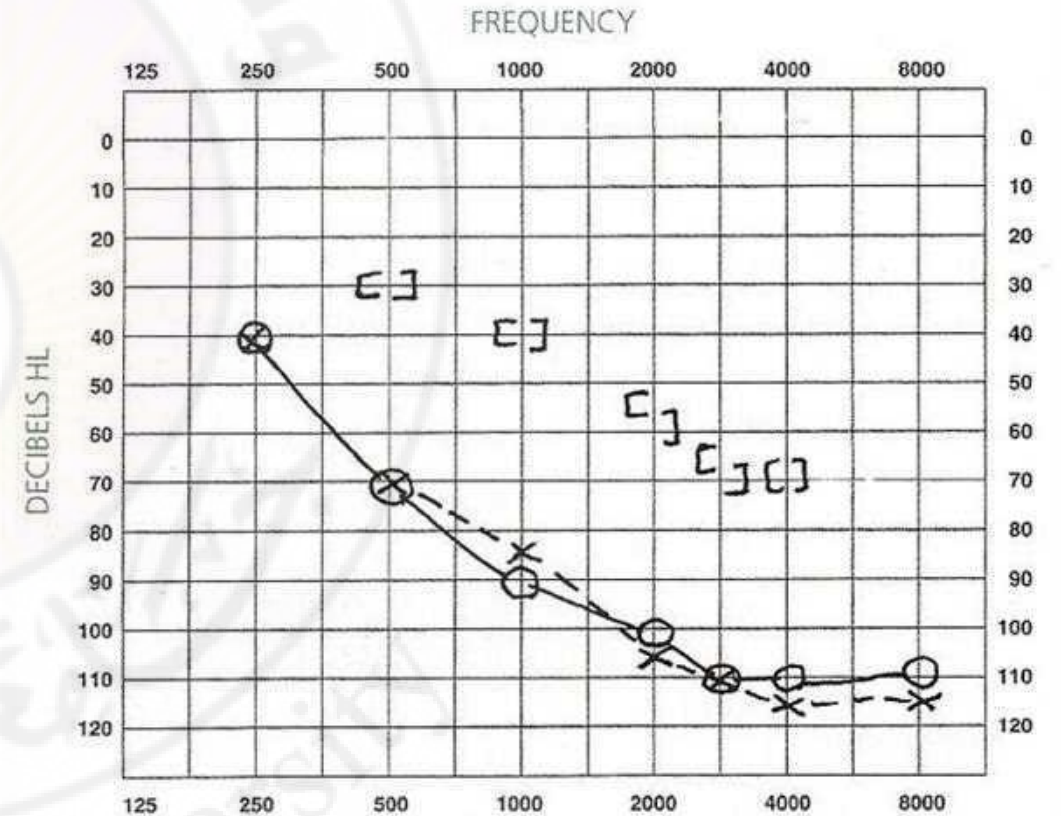


Interpretation of Weber's test

Mixed loss:

If there is a mixed loss, the tone will be lateralized to the better ear if its level is below the poorer ear's bone-conduction threshold.

The Weber test will fail to detect the conductive component of a mixed loss in such cases.



Rinne Test

- When performing the Rinne test, the tuning fork is struck and then placed on the mastoid tip, when the patient can no longer hear the tone the fork is moved in front of the meatus and patient reports if they can hear the tone.
- When conducting the Rinne, where there is a 30 dB or more difference between ears, it is advised to **mask the better ear** when conducting the test.
- For the Rinne, if the fork is louder at the mastoid, it is reported: **Rinne BC > AC (⊖ Negative / Abnormal response)**



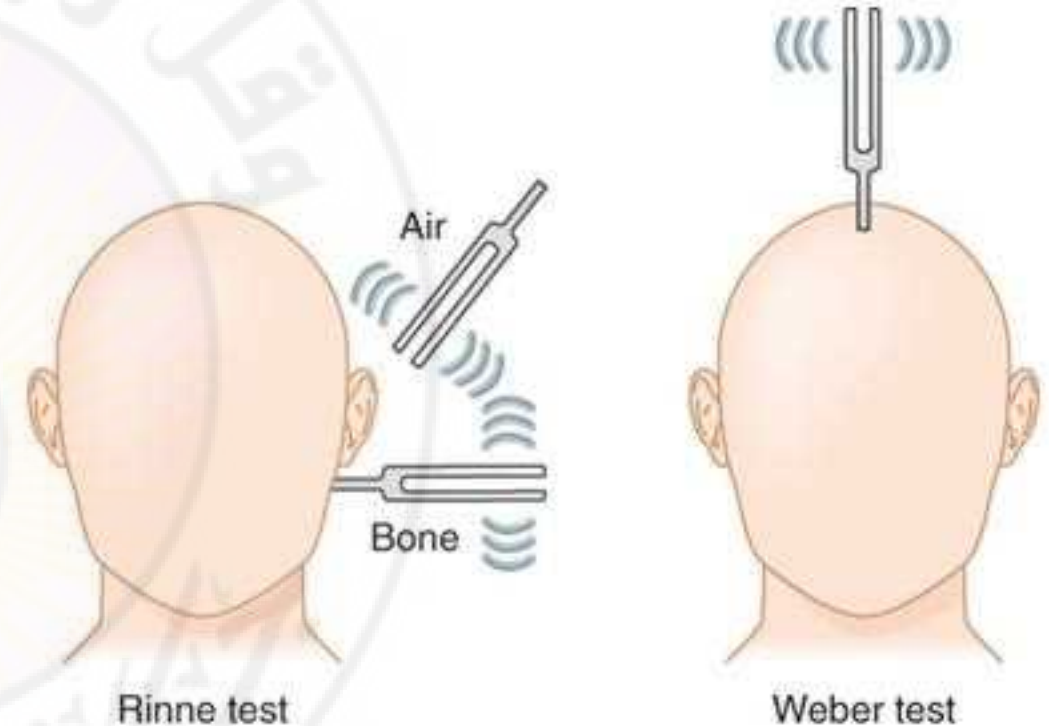
Interpretation of Rinne test

Normal Hearing: Normal/positive $\oplus$ in both ears (AC greater than BC)

Unilateral SNHL: Normal/positive on the affected ear (AC greater than BC); normal/positive on the unaffected ear (AC greater than BC).

Point: an abnormal/negative $\ominus$ response on the affected ear (BC greater than AC) can also occur in a severe sensorineural hearing loss, also called a dead ear. This is termed a "false negative."

Unilateral CHL: Abnormal/negative $\ominus$ on the affected ear (BC greater than AC); normal/positive $\oplus$ on the unaffected ear (AC greater than BC)



Hearing loss	Rinne test (Conduction)	Weber test (Localization)
None	Air > bone	Midline
Sensorineural	Air > bone	Normal ear
Conductive	Bone > air	Affected ear

Minimum air-bone-gaps needed for Rinne test to identify conductive losses with 75% accuracy (Gelfand 1977)

Frequency (Hz)	Air-bone-gaps (dB)
128	25–30
256	5–40
512	55–60
1024	45–50
2048	None ^a

^aCorrect identification of conductive losses failed to reach chance at 2048 Hz, no matter how large the size of the air-bone-gap.

Degree of Hearing Loss Estimated by Rinne Testing

Rinne Test Result	Estimated Conductive Loss
Negative: 256 Hz	Mild conductive loss of 20–30 dB
Positive: 512 and 1024 Hz	
Negative: 256 and 512 Hz	Moderate conductive loss of 30–45 dB
Positive: 1024 Hz	
Positive: 1024 Hz	Severe conductive loss of 45–60 dB
Negative: 256, 512, and 1024 Hz	

Negative Rinne test: bone conduction greater than air conduction (abnormal).

Positive Rinne test: air conduction greater than bone conduction (normal).

Interpreting the Rinne & Weber tests

Weber test \ Rinne test		lateralizes to left		no lateralization	lateralizes to right	
		left ear	right ear	both ears	left ear	right ear
⊕	⊕	Normal	SN loss	Normal SN loss	SN loss	Normal
⊖	⊕	Conductive loss	Normal	(no such condition)	Combined loss	Normal
⊕	⊖	Normal	Combined loss		Normal	Conductive loss
⊖	⊖	Conductive loss	Combined loss	Conductive loss	Combined loss	Conductive loss
SN loss = S ensorineural loss, Combined loss = C onductive & S ensorineural loss						

Bing Test

- If the **occlusion effect** is present, covering the ear canal should cause the tuning fork to sound louder.
- This is called a **positive** $\oplus$ result and implies that the ear is either **normal** or has a **sensorineural** hearing loss.
- A **negative** $\ominus$ result occurs if closing the ear canal fails to make the tuning fork sound louder, and implies that there is either a **conductive** or mixed hearing loss.
- In patients with pronounced deafness if the vibrating fork is heard after occlusion of external canal then deafness is interpreted to be due to labyrinthine causes.



How to Conduct Tuning Fork Tests



AMERICAN ACADEMY OF
OTOLARYNGOLOGY-
HEAD AND NECK SURGERY

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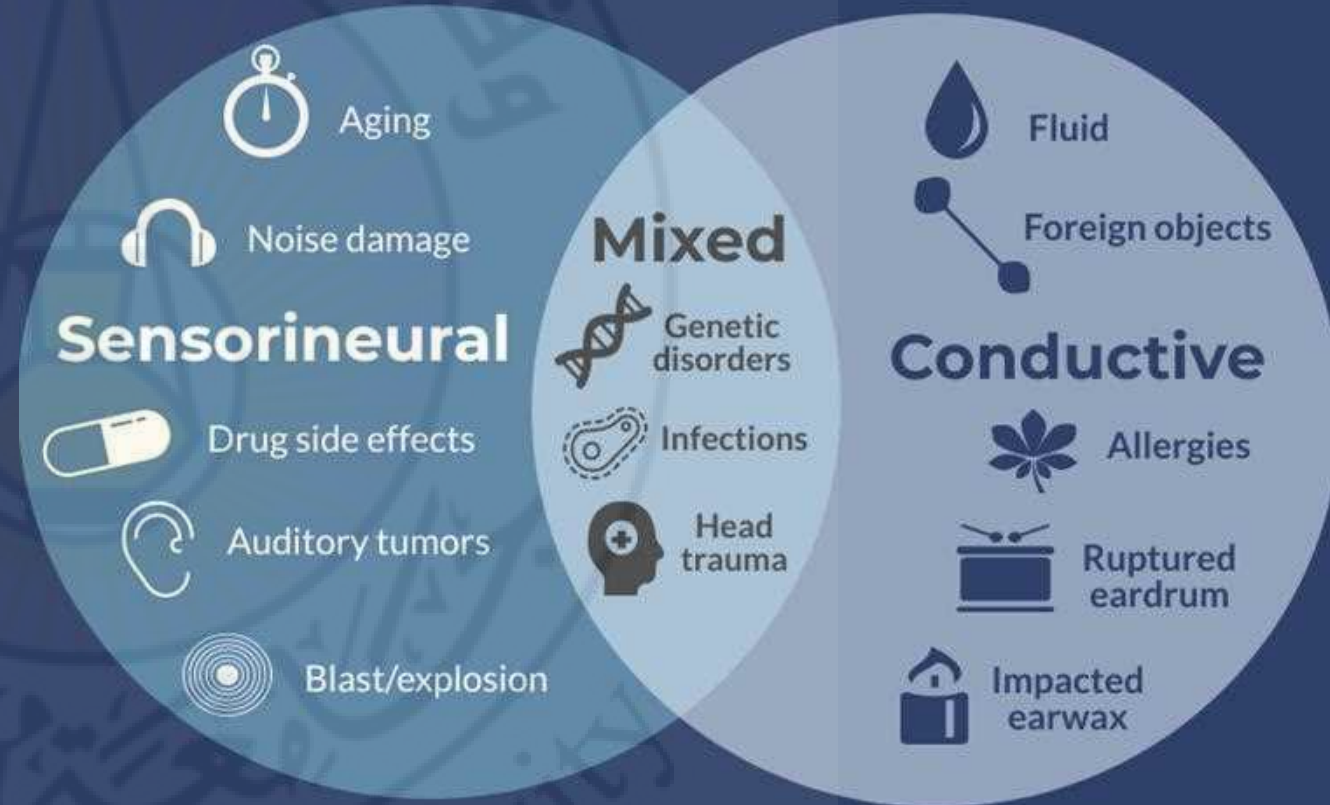
The background of the slide features a large, faint watermark of the Damascus University logo. The logo is circular, with the university's name in Arabic 'جامعة دمشق' at the top and 'Damascus University' at the bottom. In the center is a stylized emblem consisting of a triangle above a circle, which contains a smaller circle and a square.

3. Otologic considerations in clinical audiology

- b. Outer ear pathologies**
- c. Middle ear pathologies**
- d. Inner ear pathologies**

Causes of Hearing Disorder

- There are a number of causes of hearing disorder, some affecting the growing embryo, some the newborn and young child, others adults and the elderly.
- Some of the causes affect the outer- or middle-ear conductive mechanisms. Others affect only the sensory system of the cochlea or the auditory nervous system. Still others can affect the entire auditory mechanism.



Audiological evaluation

- Although the overall goal of an audiological evaluation is to characterize hearing ability, the approach used to reach that goal can vary considerably across patients:
 - **Age**
 - **Reason for referral**
 - **Needs and expectations of individual patients**
- There are several broad categories of patients that present common challenges and can be approached in a similar clinical manner, including **pediatric patients**, those being **assessed for otologic reasons**, those being **assessed for audiologic reasons**, and patients with **functional hearing loss**.



Otologic Referrals

- An otologist's evaluation of a patient, aimed at diagnosing and treating ear disease, often includes ancillary testing, such as imaging studies, laboratory tests, and audiologic evaluations.
- **Audiologic assessment** in this context serves to provide additional information to the physician to aid in diagnosis.
- In addition, because hearing loss is often a quantifiable consequence of auditory disorder, results can serve as a metric for the success or failure of treatment approaches.
- In general, otologists are faced with three main categories of patients: those with **outer- or middle-ear disorders**, those with **cochlear disorders**, and those with **neurologic disorders** affecting the peripheral or central auditory nervous system.



Outer- or Middle-Ear Disorders: Evaluative Goals

- Physicians who note the presence of an outer- or middle-ear disorder are likely to refer the patient for an audiologic consultation.
- For the audiologist, there are two primary goals in the assessment of outer- and middle-ear disorders:
 1. Investigation of the nature of the disorder and;
 2. Assessment of the impact of that disorder on hearing sensitivity
- ***Test Strategies***
 - Immittance Audiometry
 - Pure-Tone Audiometry (AC & BC)
 - Speech Audiometry

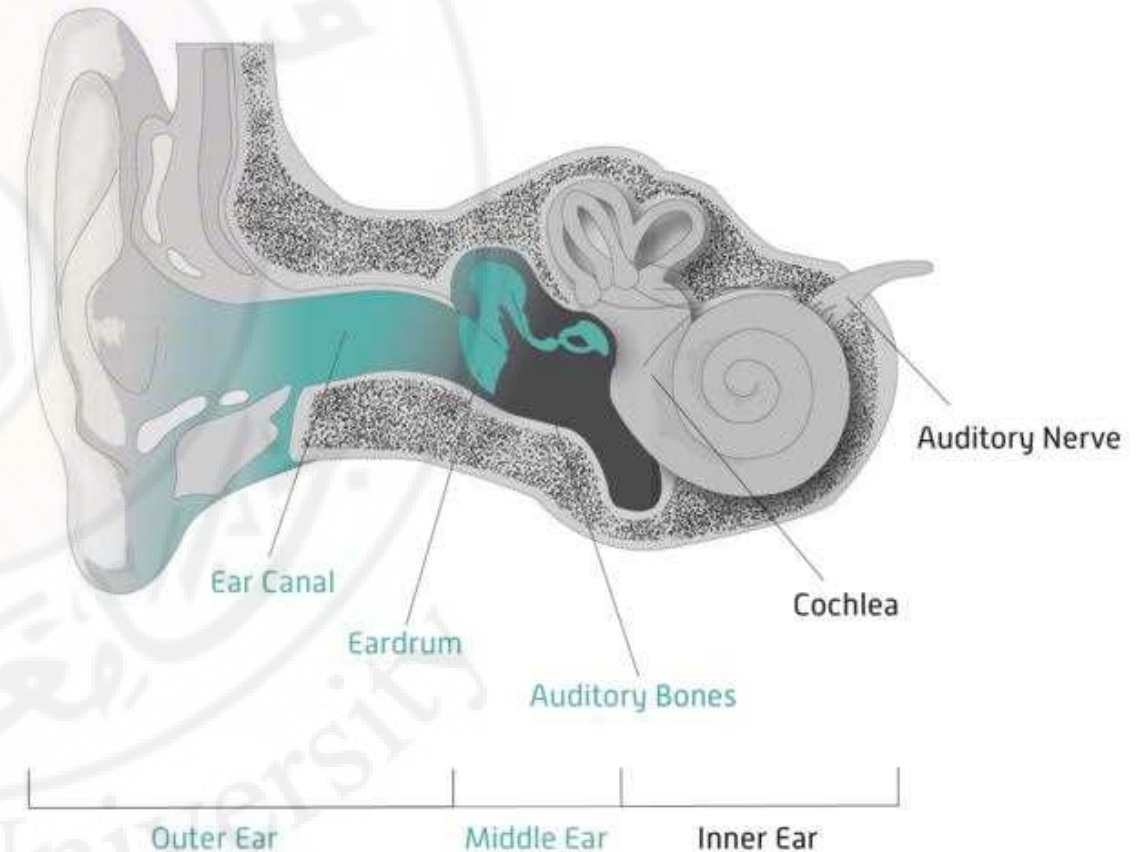
The background of the slide features a large, faint watermark of the Damascus University logo. The logo is circular, with Arabic calligraphy at the top and "Damascus University" at the bottom. In the center is a stylized yellow and orange emblem resembling a minaret or a sunburst.

Conductive Hearing Disorders

- External ear disorders (Pinna & Ear canal)
- Tympanic membrane & Middle ear disorders

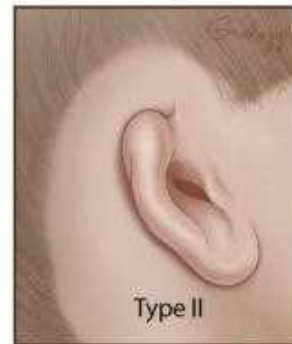
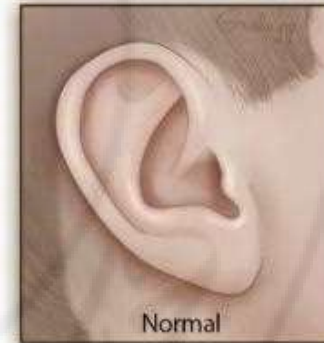
Conductive Hearing Disorders

- Disorders of the outer and middle ear are commonly of two types, either structural defects due to embryologic malformations (**congenital**) or structural changes secondary to infection or trauma (**acquired**). Another common abnormality, otosclerosis, is a bone disorder.
- Sound waves are not able to efficiently go through the outer ear canal to the eardrum and the small bones of the middle ear. There is typically a reduction in sound levels or the ability to hear faint sounds. This type of hearing loss can often be corrected medically or surgically, and bone conduction hearing aids are available when medical or surgical correction are insufficient.



Congenital Outer- and Middle-Ear Anomalies

- **Microtia** and **atresia** are congenital malformations of the auricle and external auditory canal.
- **Microtia** is an abnormal smallness of the auricle. It is one of a variety of auricular malformations. **Microtia in isolation may not affect hearing in any substantial way.** The auricle serves an important purpose in horizontal sound localization and in providing certain resonance to incoming signals. But the absence of auricles, or the presence of auricles that are deformed, does not in itself create a significant communication disorder for patients.
- Ears with microtia or other deformities will often be surgically corrected at an early age.



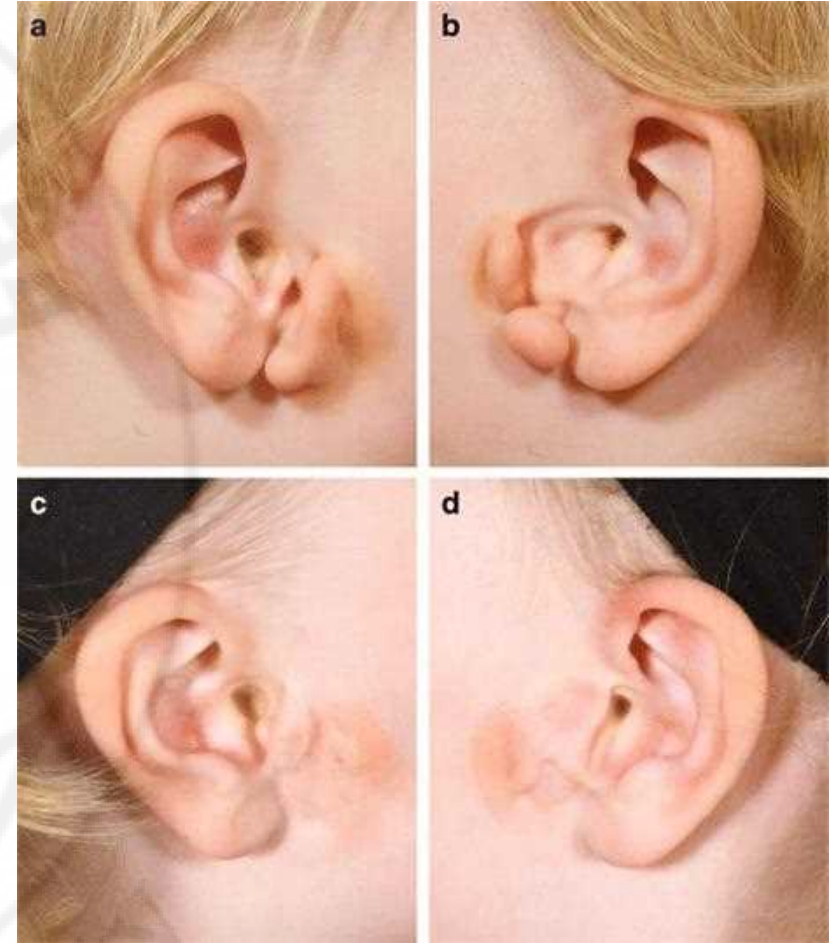
congenital auricular malformations	Definition
Accessory auricle	an additional auricle or additional auricular tissue
Anotia	congenital absence of an auricle
Auricular aplasia	anotia
Cleft pinna	congenital fissure of the auricle
Coloboma lobuli	congenital fissure of the earlobe
Macrotia	congenital excessive enlargement of the auricle
Melotia	congenital displacement of the auricle
Low-set ears	congenitally displaced auricles
Polyotia	presence of an additional auricle on one or both sides
Preauricular pits (Fistula auris)	small hole of variable depth lying anterior to the auricle
Preauricular tags	small appendage lying anterior to the auricle
Scroll ear	auricular deformity in which the rim is rolled forward and inward



Accessory auricle



Cleft pinna



Polyotia



Preauricular pits



Preauricular tags



Scroll ear

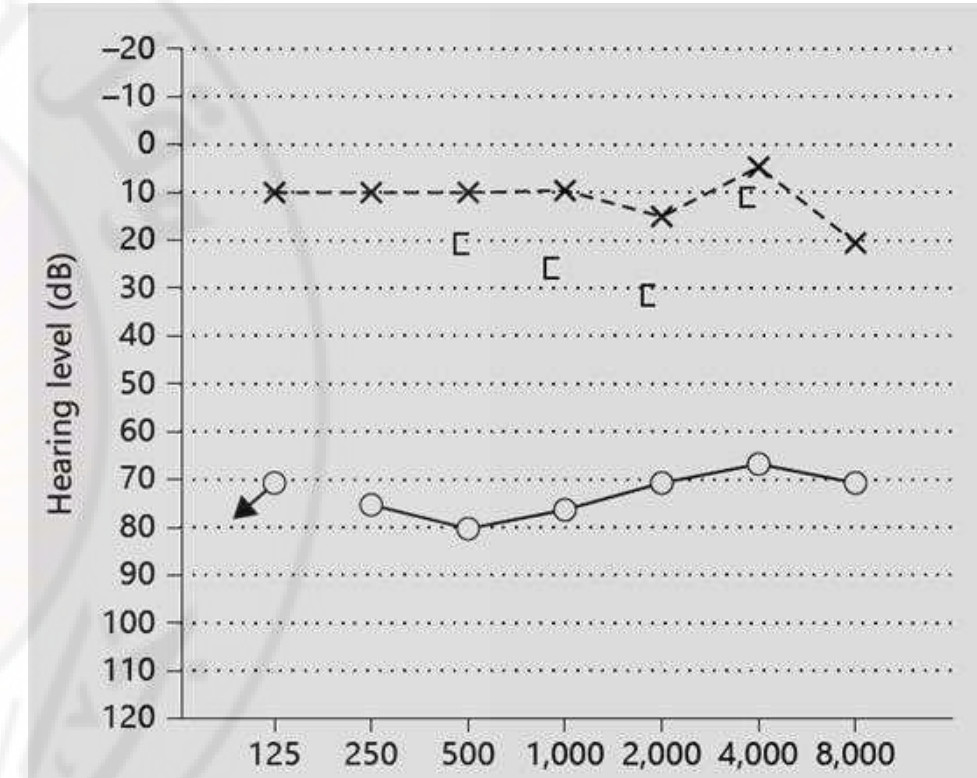
Congenital Outer- and Middle-Ear Anomalies

- **Atresia** is the absence of an opening of the external auditory meatus. An ear is said to be **atretic** if the ear canal is closed at any point.
- Atresia is a congenital disorder and may involve one or both ears.
- **Bony atresia** is the congenital absence of the ear canal due to a wall of bone separating the external auditory meatus from the middle ear.
- **Membranous atresia** is the condition in which a dense soft tissue plug is obstructing the ear canal.



Atresia: Audiometry

- Pure tone audiometry is performed by **air** conduction and **bone** conduction hearing tests.
- Most patients with microtia/atresia of the external auditory canal suffer from conductive hearing loss, and mixed hearing loss is observed in some patients, but sensorineural hearing loss is very rare.
- On pure tone audiogram, the air conduction hearing level is classified as the **flat type** and the type in which the threshold of the **high pitch area is reduced** when frequency becomes higher.
- Bone conduction threshold is close to 0 dB in most patients, like the threshold in healthy subjects. Retesting is recommended when the threshold for bone conduction is high.



Fused Malleus and Incus complex

Atresia: Audiometry

- Atresia causes a significant conductive hearing loss, which can be as great as **60 dB**. The additional bone or membranous plug adds mass and stiffness to the outer ear system, resulting in a relatively **flat** conductive hearing loss.
- In cases of partial atresia where a TM is present, **impedance studies** may be of great help. The audiologist can help determine whether any surgical attempts should be made.

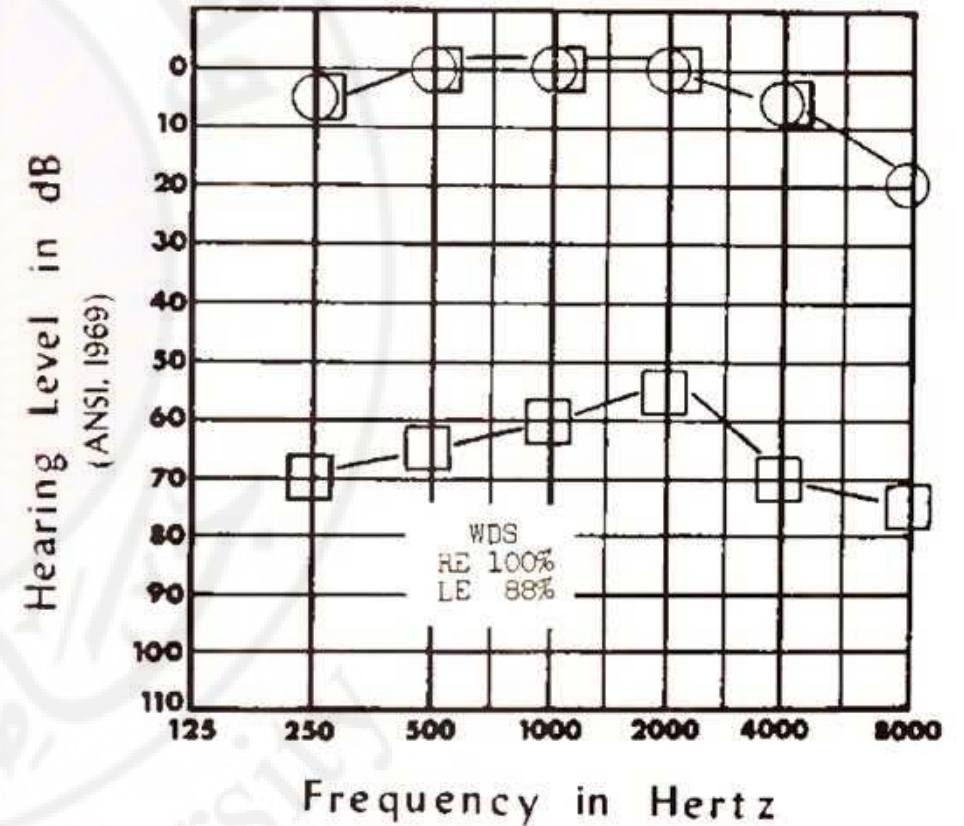
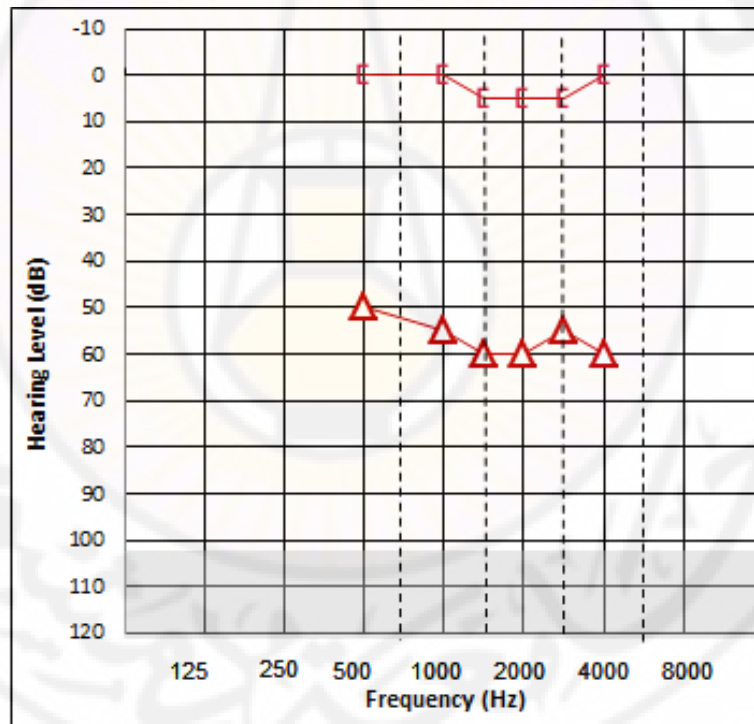


Fig. 2.1. Audiometric results of 14-year-old boy with congenital atresia, left ear.

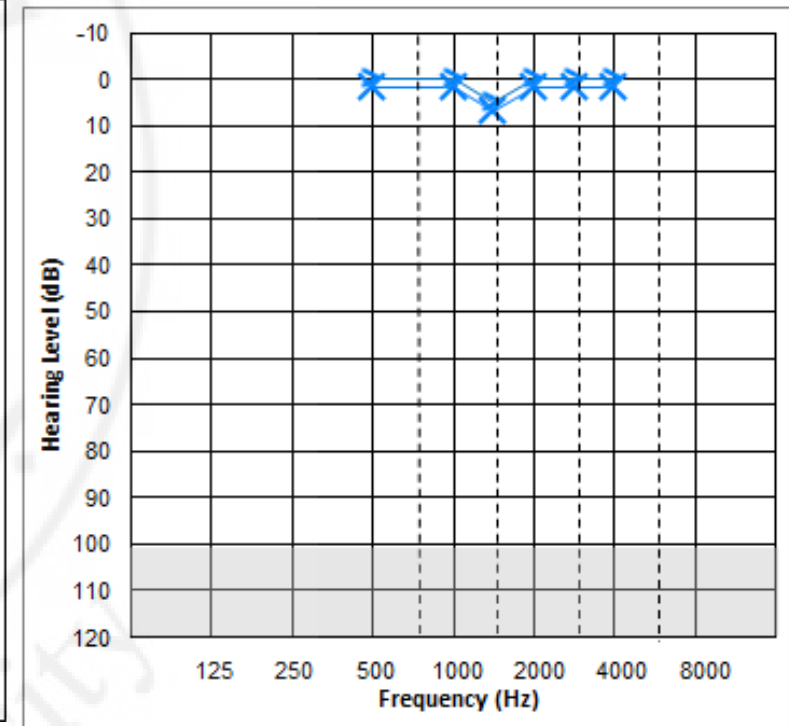
Case Example

- Aural Atresia with malformations
- of the Ossicles
- Tympanometry
 - Left: Type A
 - Right not measureable

Right

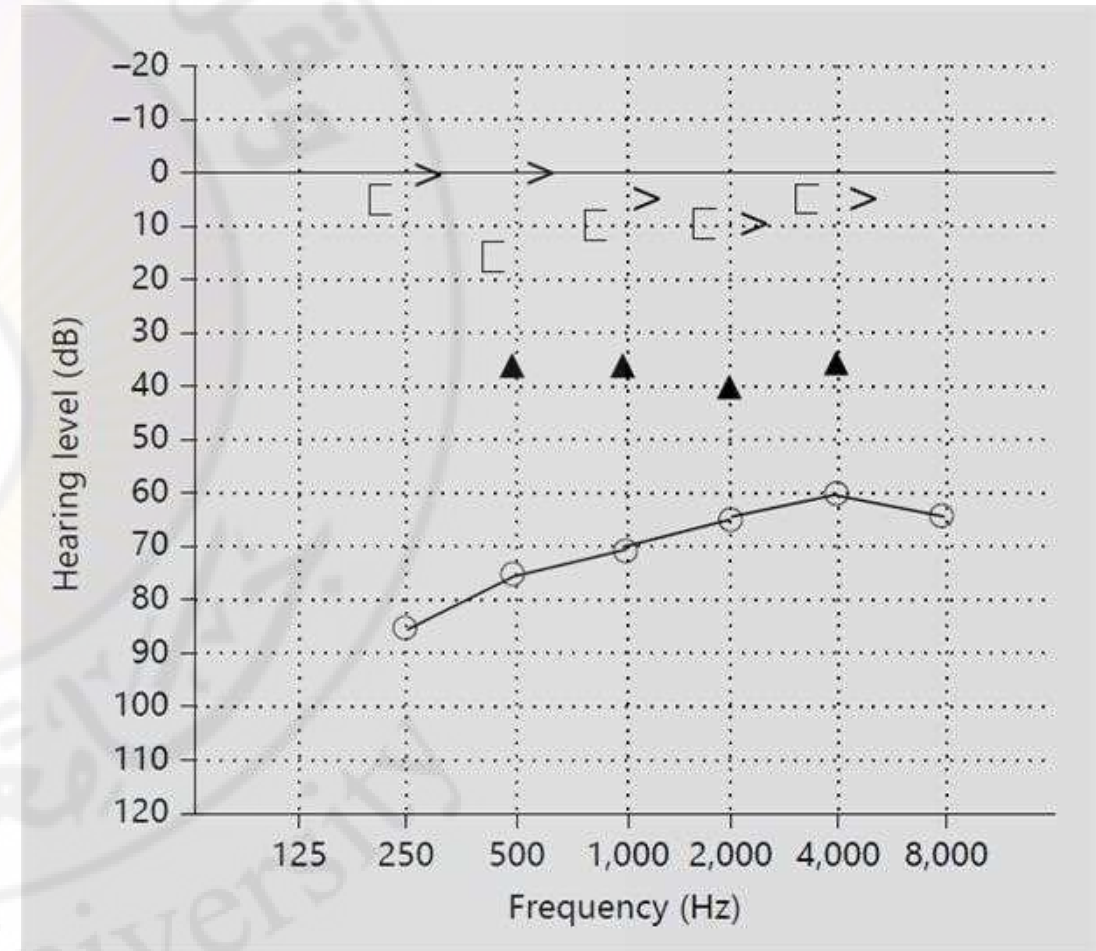


Left



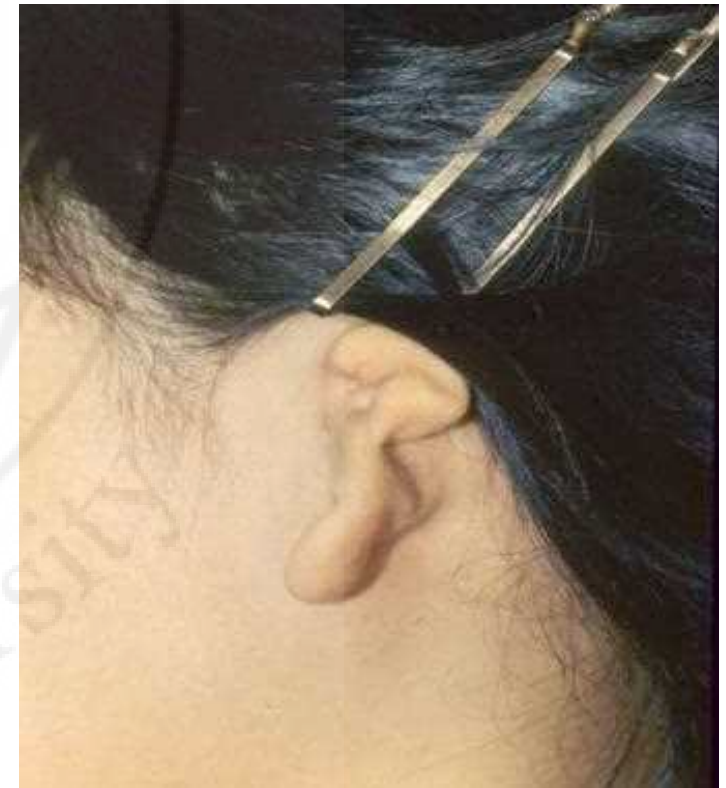
Atresia: Sound field Audiometry

- Once bone conduction hearing aid is used with unilateral or bilateral atresia, threshold test can be performed in a free sound field to test the effect of the hearing aid(s).
- **The effect is between 25 and 35 dB.**



Atresia: BC-ABR

- The goal of BC-ABR is to estimate **cochlear function** and to help identify the **type** of hearing loss present.
- It enables differential diagnosis of conductive hearing loss and sensorineural hearing loss in Microtia/Atresia.
- Problems with the current bone-conduction ABR are poor reproducibility, difficulty estimating bone conduction threshold at specified frequencies (as with air conduction ABR), and masking.



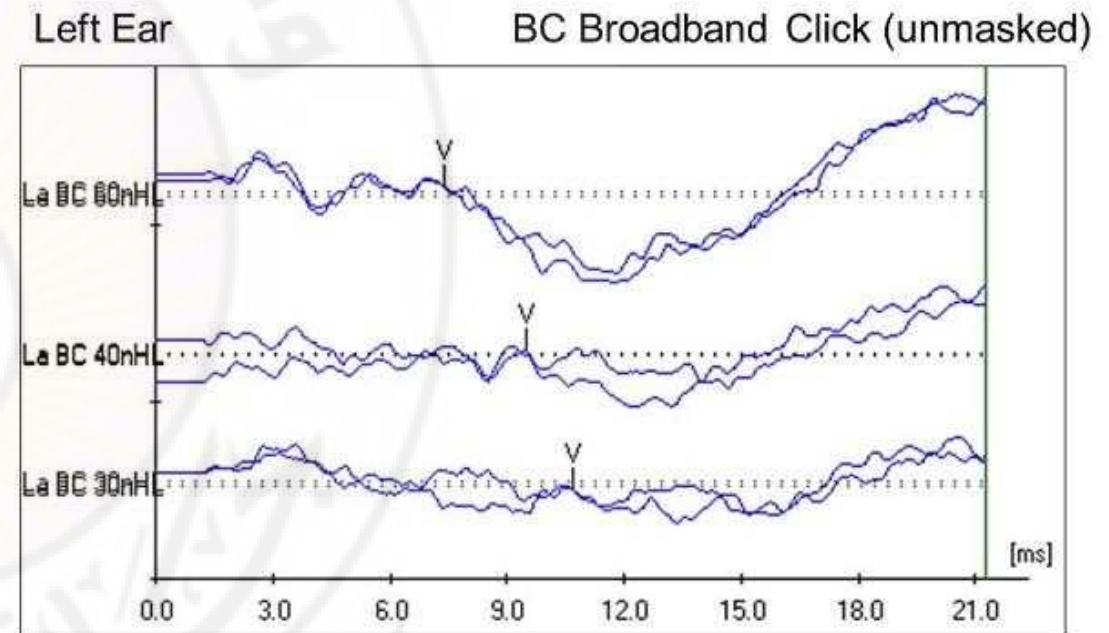
Placements for Bone Oscillator



The tension of the headband can be adjusted to a recommended coupling force of 400 to 450 g using a spring scale.

BC-ABR threshold investigation method

- Start recording at a moderate level so as not to wake up the baby with the vibrations from the oscillator; around 30 dBnHL.
- Do not exceed 50 dBnHL because you will overdrive the oscillator and will end up collecting artifact.
- Decrease in 10 dB steps to find threshold; 20 dBnHL or below is within normal limits.



Problems with BC-ABR

Artifacts

- It is very common to observe a large amount of artifact while using bone-conduction ABR. This is especially true at high intensities (~50 dB nHL) and at earlier waves (i.e. Wave I).
- To avoid stimulus artifact, it is recommended that the bone oscillator be placed high on the temporal bone and that the inverting electrode is placed on the earlobe, mastoid, or nape of the neck.
- Using an **alternating phase** stimuli should be used to reduce artifact.

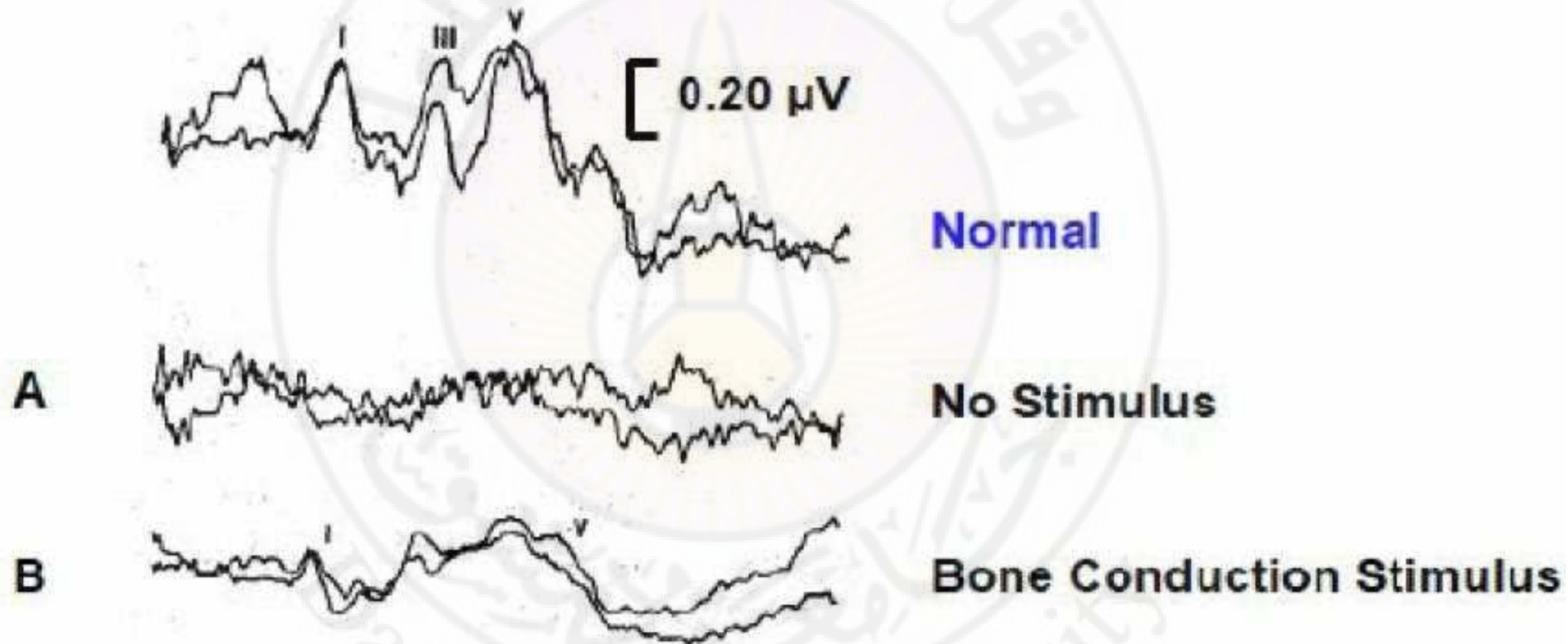
Output Limitation

- Since the output of most bone oscillators is around 45 to 55 dB nHL, it becomes difficult to distinguish between sensorineural or mixed hearing losses when the losses by bone exceed this number.

Threshold of Bone-Conduction ABR

- With BC-ABR, the waves are typically more rounded compared with AC-ABR.
- The wave V latency of BC-ABR is approximately **0.5 ms longer** than AC ABR. Such a latency discrepancy has been thought to be due to the low pass filter characteristics of the bone vibrator. In other words, BC stimuli would excite more apical cochlear regions with a longer traveling wave delay than the corresponding AC stimuli
- For **infants**, Wave V latencies for bone-conduction clicks are shorter than the air conduction clicks. These differences can be attributed to changes to the skull due to aging.



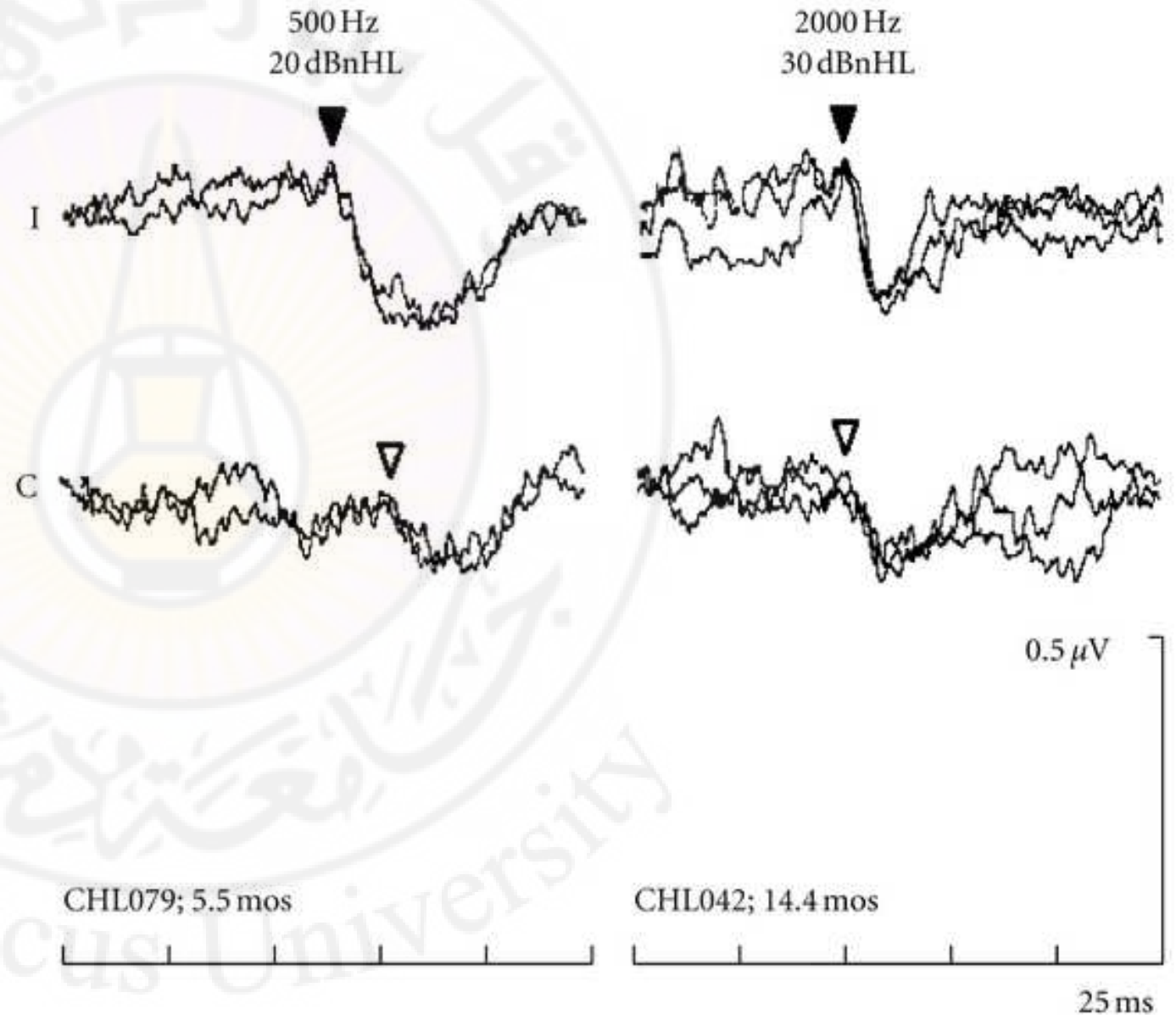


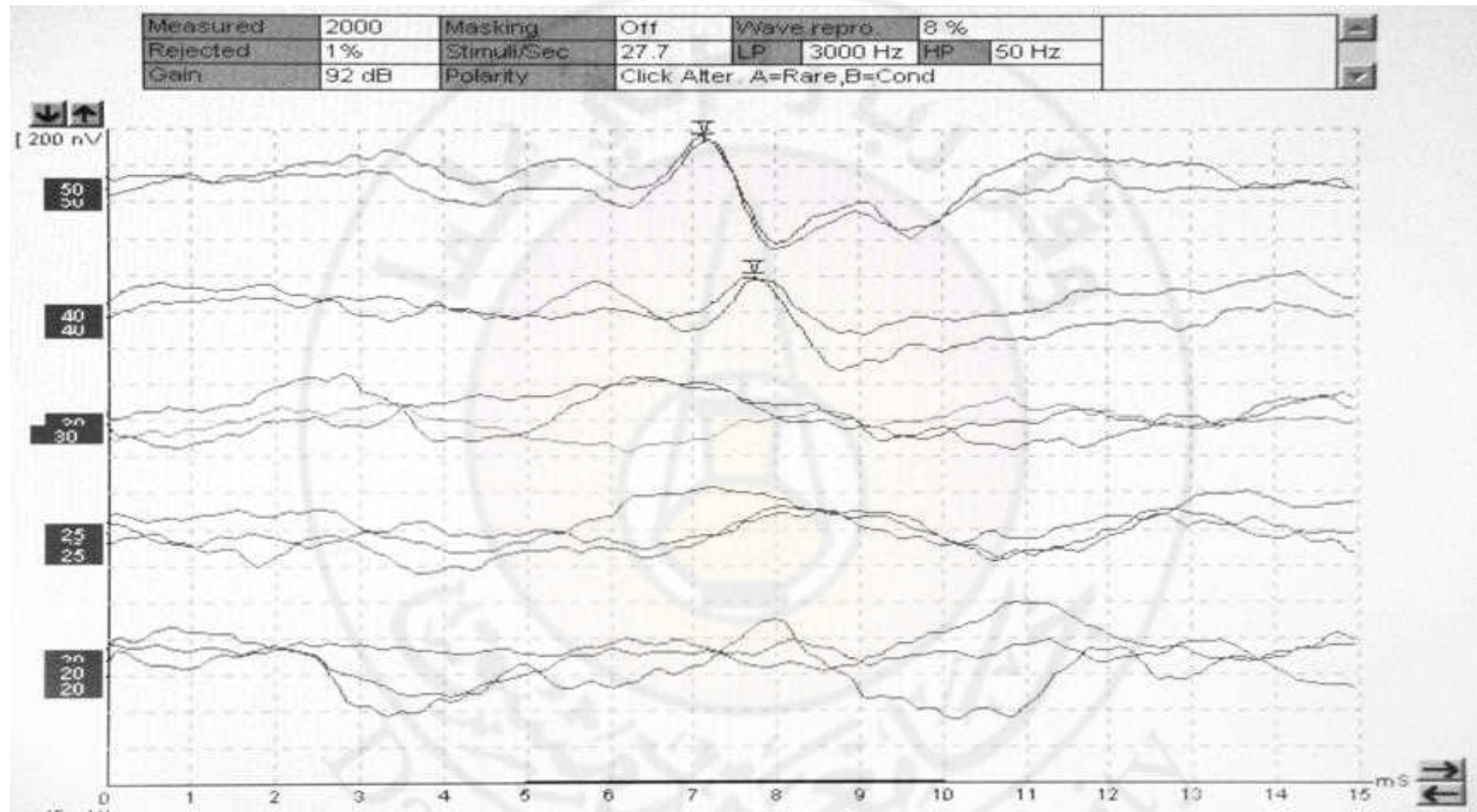
Masking in BC-ABR

- Masking of the contralateral ear, typically required with bone-conduction testing in **adults**, but not necessary in young **infants** (under 6 months); because the Inter-aural attenuation is high (as much as 25 dB) due to inadequate fusion of the cranial bones.
- Clear **wave I** in an ABR elicited with bone conduction stimulation confirms that the response is ear-specific and masking is not needed.
- If a reliable wave I is not identified then masking of the non-test ear is indicated. A general guideline is masking noise of 50 dB presented via insert earphones (in unilateral atresia).
- **Bilateral Atresia present a masking dilemma!**

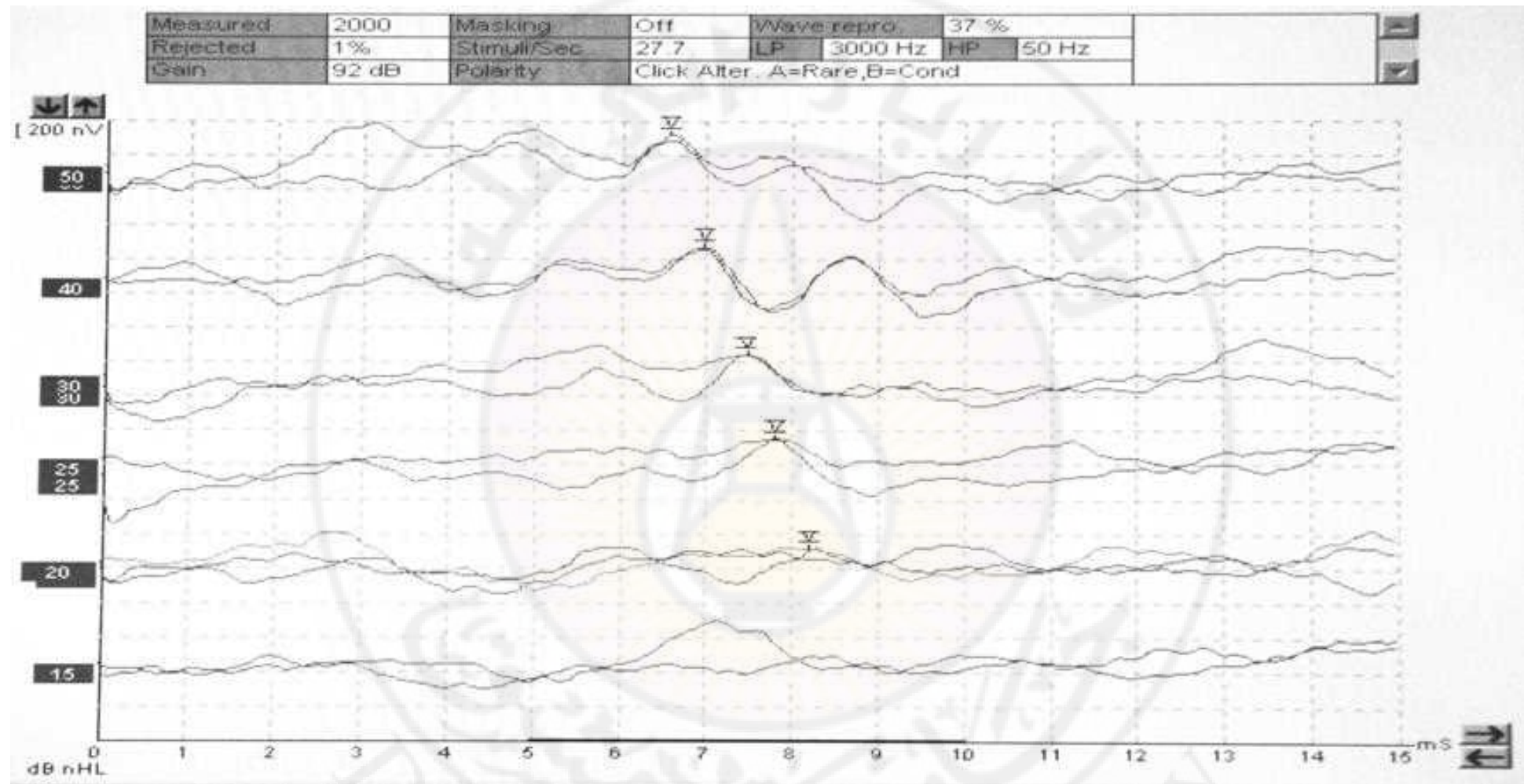


ABRs to 500 and 2000 Hz BC tones, presented at 20 and 30 dBnHL respectively, recorded from two infants with conductive hearing loss at the time of ABR testing.





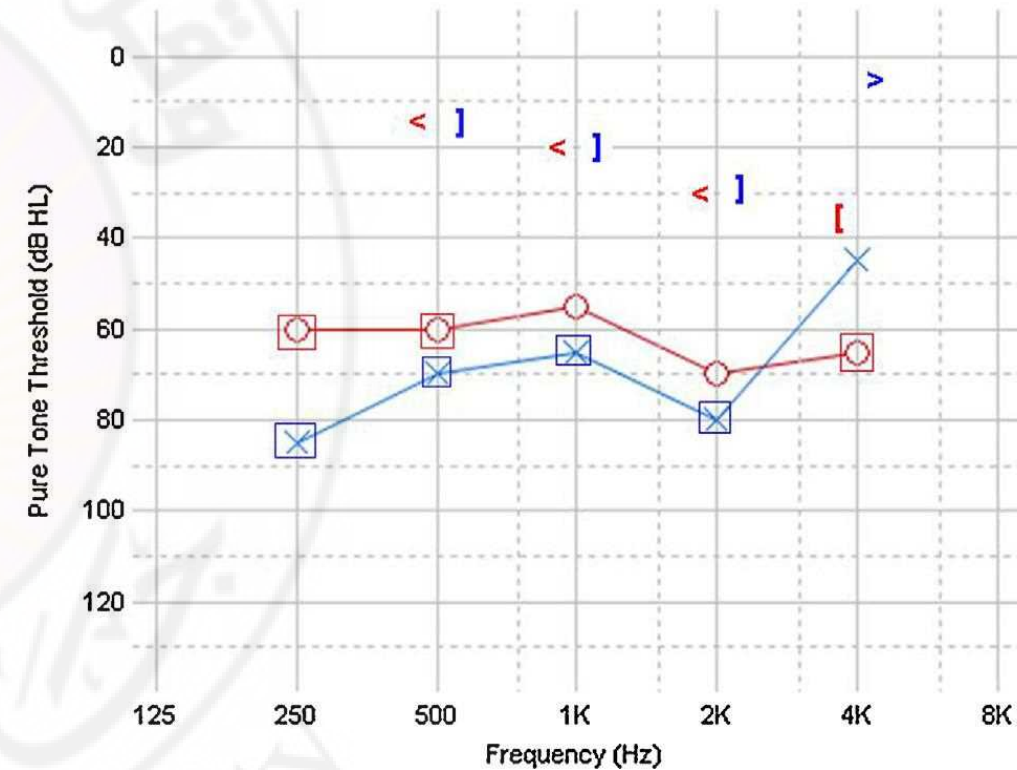
BC-ABR thresholds in a female subject, with the vibrator positioned on the **forehead**.



BC-ABR thresholds obtained in a female subject, with the vibrator positioned on the **mastoid**

Congenital Outer- and Middle-Ear Anomalies

- **Middle-ear anomalies** include malformed ossicles (ossicular dysplasia), malformed openings into the cochlea (fenestral malformations), and congenital cholesteatoma.
- **Ossicular dysplasia** can result in fixation of the bones, deformity of the bones, and disarticulation of the bones, especially the incus and stapes.
- In cases of **congenital stapes fixation**, the stapes footplate is fixed into the bony wall of the cochlea at the oval window.
- **Lack of oval window** development is an example of **fenestral malformation**.
- **Congenital cholesteatoma** is a cyst that is present in the middle-ear space at birth.



Oval window atresia in a 14 year old male patient

Congenital Outer- and Middle-Ear Anomalies

- **Congenital middle ear anomalies** usually accompany the atresia or stenosis of external ear canal; however, in the presence of a normal ear canal, diagnosis is more difficult.
- Some of these children may have a ventilation tube in place.
- It is also possible to **misdiagnose** this condition as sensorineural hearing loss (SNHL) unless bone conduction auditory brainstem response testing (ABR) is done.
- Bone conduction ABR is very important in the preoperative evaluation for differentiation between conductive and sensorineural hearing loss.

Clinical characteristics:

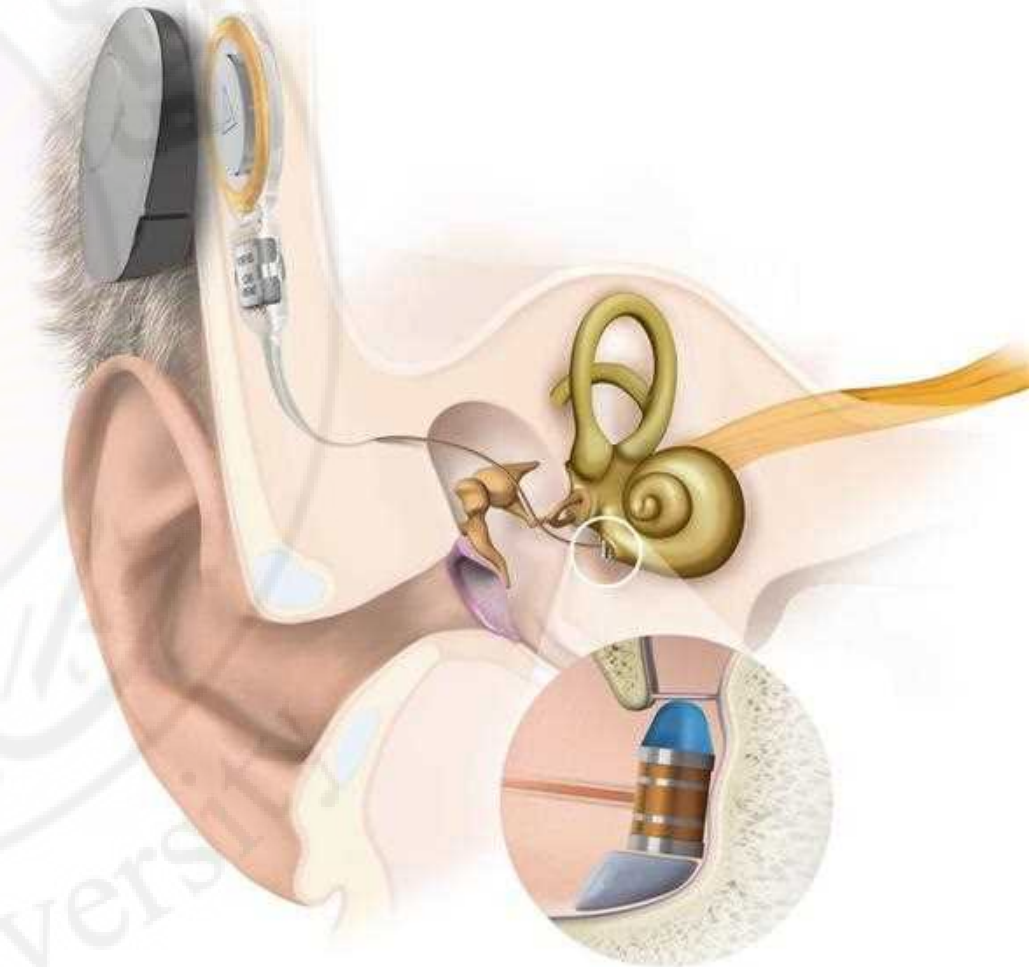
- Non-progressive hearing loss since birth
- Conductive hearing loss with air-bone gap of over 40 dB
- Excellent language development with hearing aids

Cremers classification of middle ear anomalies

Class	Anomaly	Subclassification
I	Isolated stapes footplate fixation	
II	Stapes fixation with another congenital ossicular chain anomaly	a. Ossicular discontinuity b. Pitympanic fixation c. Tympanic fixation
III	Anomaly of ossicular chain but mobile stapes footplate	a. Ossicular discontinuity b. Epitympanic fixation c. Tympanic fixation
IV	Congenital aplasia/dysplasia of the oval window or round window	a. Aplasia b. Dysplasia b1. Abnormal facial nerve b2. Persistent stapedial artery

Congenital Outer- and Middle-Ear Anomalies: Management

- Hearing aids
- Bone conduction hearing aids
- Middle ear implants
- Ossiculoplasty & Canaloplasty



Bone Conduction Devices

Invasive

Vibration directly to bone

Percutaneous

Bone Anchored
Hearing Aids

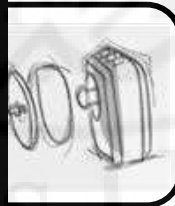
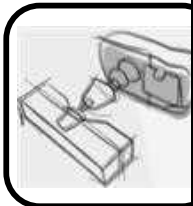
Transcutaneous
(intact skin)

Active Bone
Conduction Implant

Vibration through skin

Fixed by implanted
magnet

Passive
transcutaneous BC-
system



Non-Invasive

Skin Pressure

Undefined
Transducer Pos.

Flashy

Contact Pressure

No Pressure

Defined
Transducer Pos.

Discreet

Adhesive



- **Bone Conduction Devices**



- **Active Middle Ear Implants**







Impacted Cerumen

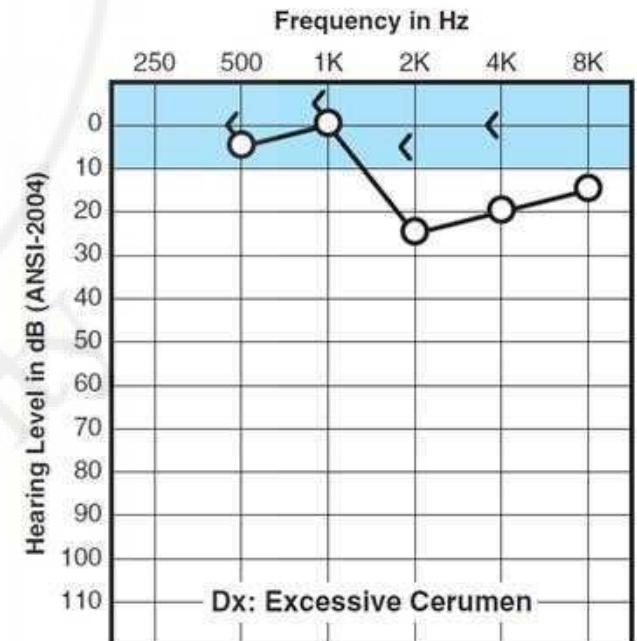
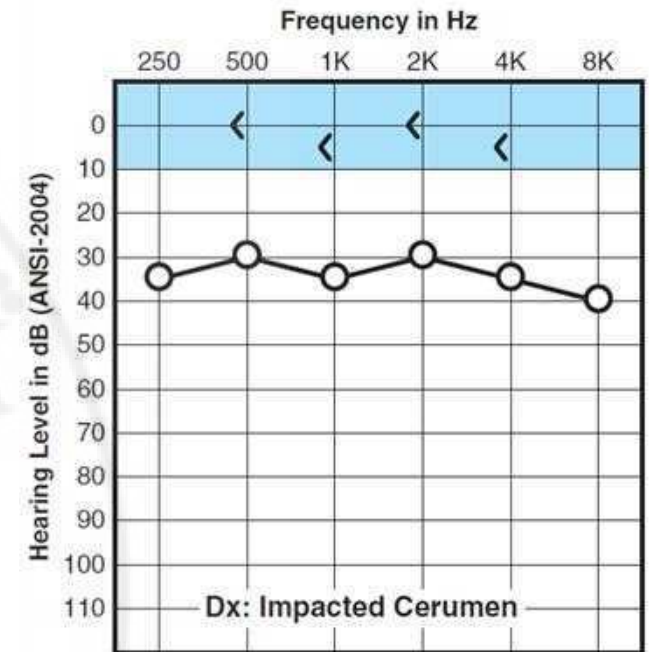
- One common cause of transient hearing disorder is the accumulation and impaction of cerumen in the external auditory canal. Cerumen has a natural tendency to migrate out of the ear canal and, with proper aural hygiene, seldom interferes with hearing.
- In some individuals, however, cerumen is secreted excessively, a condition known as **ceruminosis**. In these patients, routine ear canal cleaning may be required to forestall the effects of excessive cerumen accumulation.
- Impacted cerumen is often managed first by a course of eardrops to soften the impaction, followed by irrigation of the ear canal to remove it.



Worm in the otologic apple!

Impacted Cerumen

- **Losses of as much as 40 dB are possible!**
- A moderate level of impacted cerumen affect one of both mass and stiffness, resulting in attenuation across the frequency range, giving a **flat audiogram**.
- Sometimes cerumen is pushed down into the ear canal and onto the tympanic membrane without occluding the ear canal. This results in an increase in the mass of the tympanic membrane, resulting in a **high-frequency conductive hearing loss**.



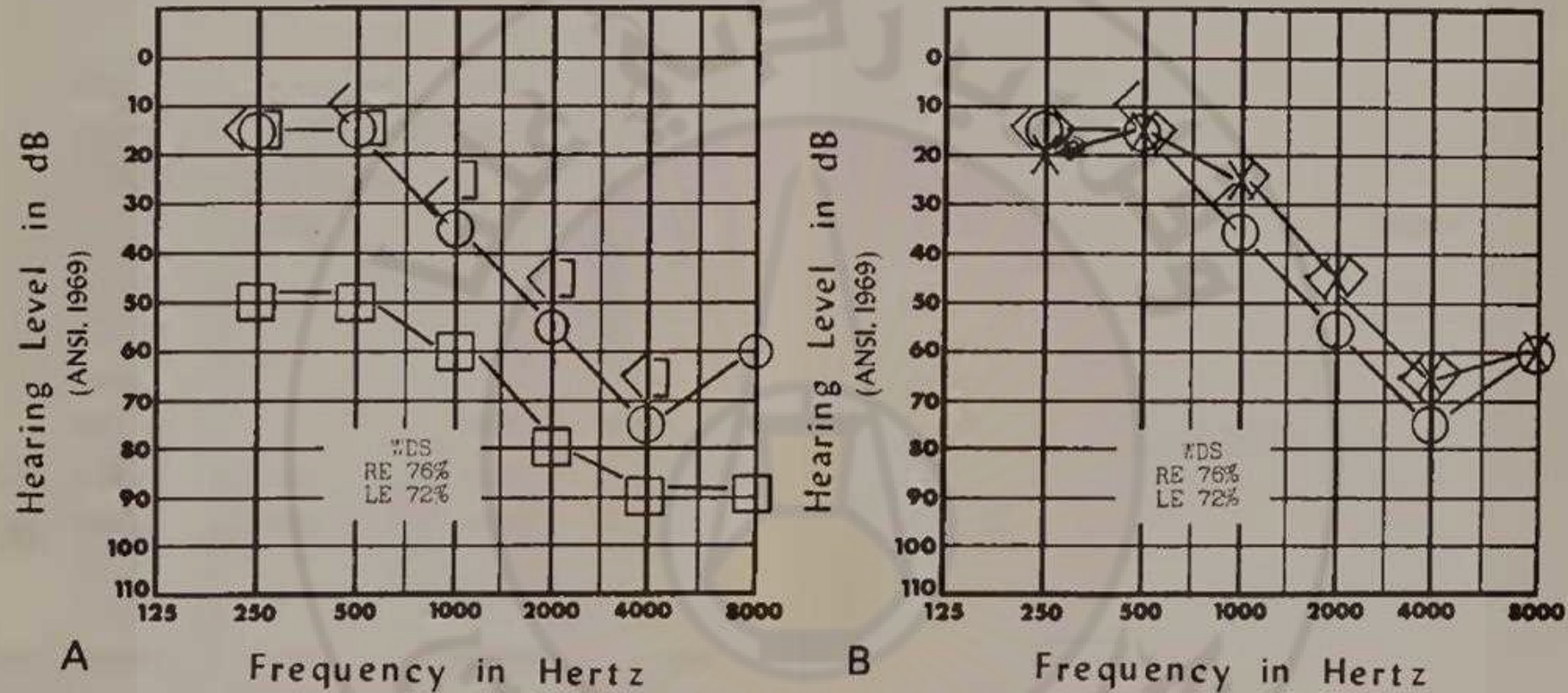
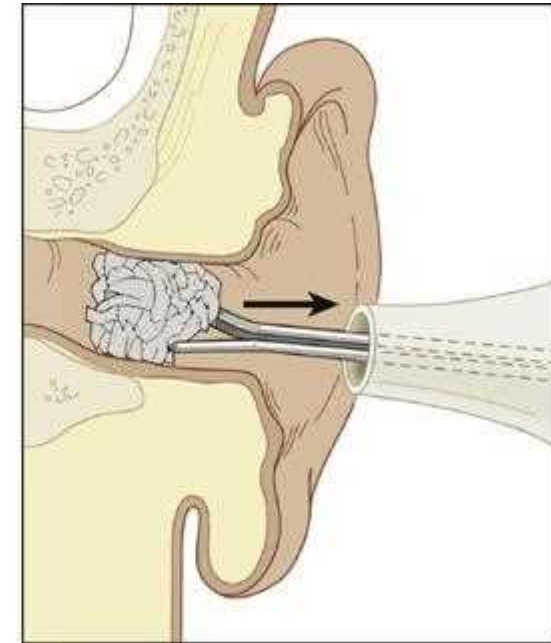
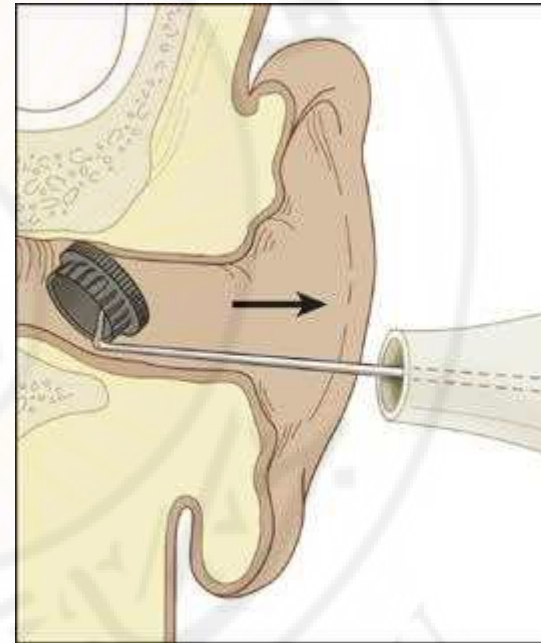


Fig. 2.2. (A) Hearing test results of patient showing effects of impacted cerumen, left ear. (B) Hearing test results of same patient as A, after removal of impacted cerumen.

If cerumen is present, Audiologist should refer the patient for its removal. A clean external canal is even of greater importance in impedance measurement since the presence of cerumen will give misleading information regarding the status of the middle ear.

Foreign Body Obstruction

- Obstruction of the external canal by foreign bodies can produce a conductive impairment especially in children.
- Small foreign bodies leave no audiologic consequences, thus a hearing loss discovered on routine school audiometry rarely is caused by this problem.
- The foreign object would have to be totally obstructive such as a bead or a pencil eraser.
- Foreign bodies should always be thought of in cases of **unilateral conductive loss** but their presence is soon discovered on **otoscopy**.



Collapsed Canal

- Audiologists are sometimes baffled by an air-bone gap with a flat air- conduction curve for which there is no apparent cause.
- The TM appears quite normal as does the canal, and tuning fork tests fail to confirm the presence of an air-bone gap.
- This proves to be due to an anatomical variation of the outer-most part of the cartilaginous canal and tragus.
- The pressure of the ear phone produces a valve like closing of the meatus, so complete as to produce a very significant conductive loss.
- This must be looked for and, if suspected, a device should be placed in the canal to prevent collapse when the phones are applied.

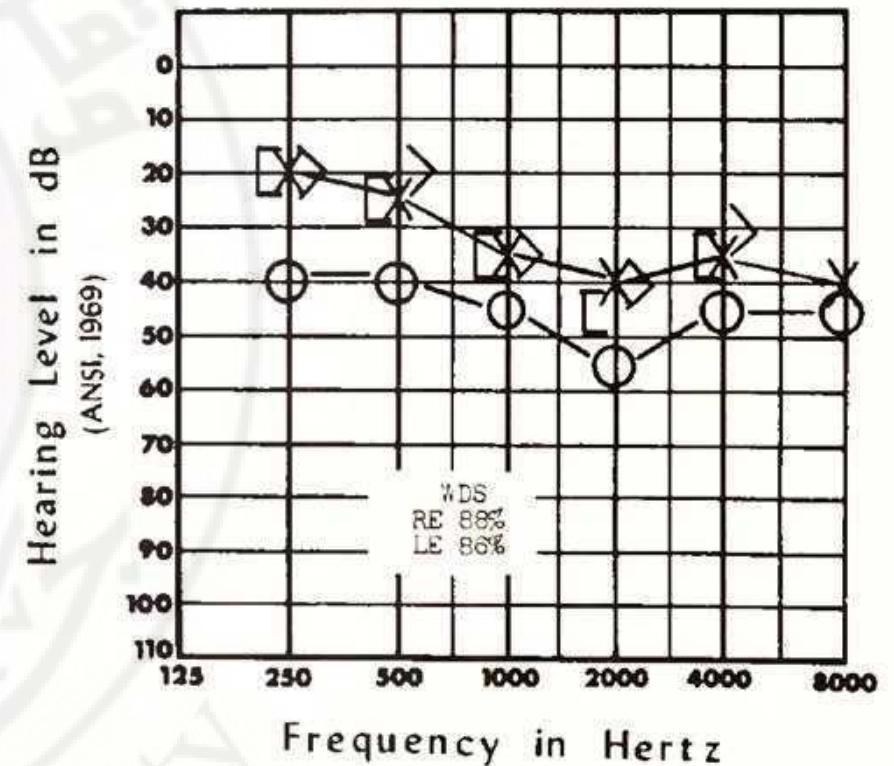


Fig. 2.3. Audiometric findings of individual showing influence of collapsed canal while under test (right ear).

Case Example

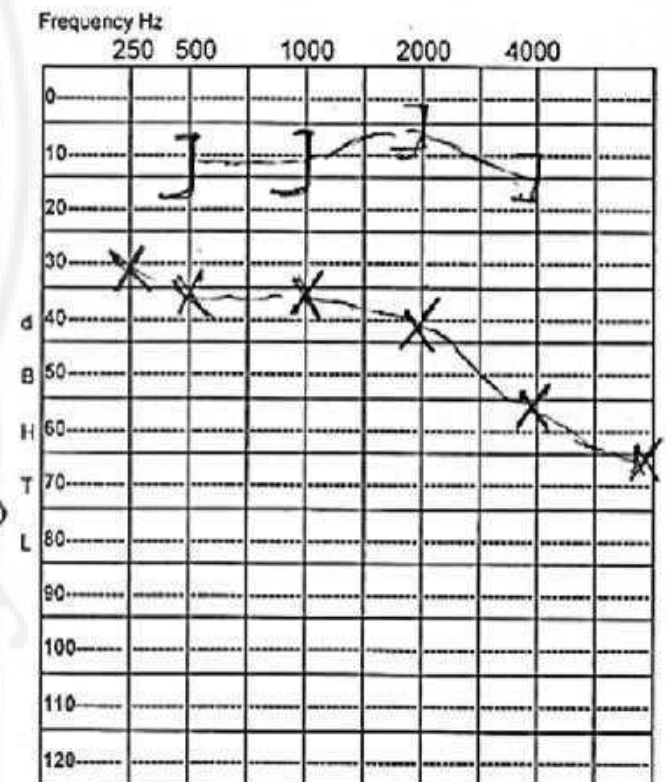
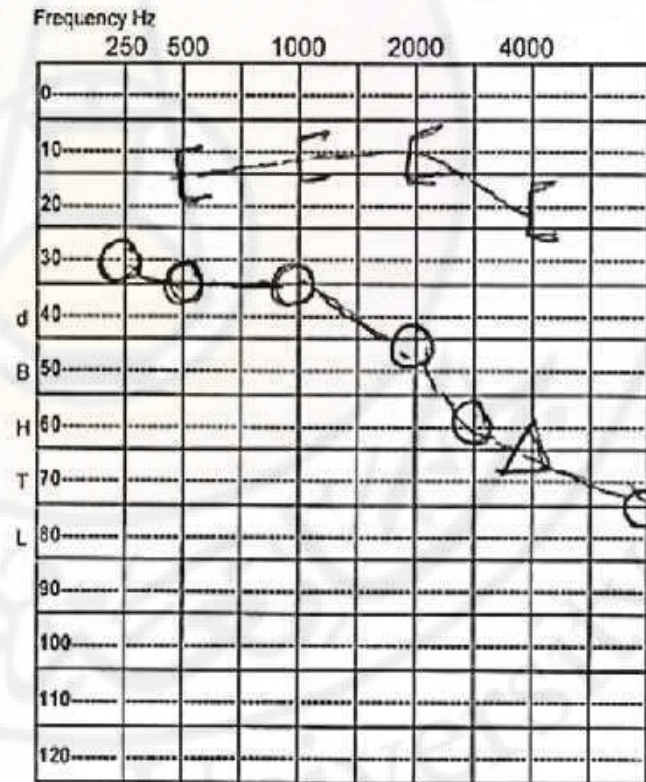
Bilateral Conductive Hearing Loss due to Collapsed Ear Canals in a 35 Years Old Female.



Acoustic Reflex pattern in Bilateral Collapsed Ear Canals:

- All contralateral ARTs are absent.
- All ipsilateral ARTs are present.

	Ipsilateral	Contralateral
Right	Normal	Absent
Left	Normal	Absent



Osteoma, Hyperostosis, Exostosis

- Growths of the bony external canal are not uncommon but rarely produce a hearing loss, unless they result in ear canal blockage.
- **Osteoma:** Benign tumors leading to obstruction of the external auditory canal. Usually occur medially in the bony canal rather close to the tympanic membrane. Typically arise *unilaterally* and are true bony neoplasms.
- **Exostosis:** The overgrowth of the external auditory canal's compact bone in adults that occur *bilaterally* and may be secondary to a periosteal reaction triggered by repeated cold water exposure (**Surfer's ear**).
- **Hyperostosis:** The diffuse narrowing of the bony canal wall, also must be completely obstructing to produce a change in the pure tone results.



Osteoma



Exostosis

Otitis Externa

- Infections of the ear canal or auricle are referred to collectively as otitis externa, or inflammation of the external ear.
- They are often caused by **bacterium, virus, or fungus** cultivated in the external ear canal.
- One common type is known as **swimmer's ear**, or *acute external otitis*, and is characterized by diffuse reddened pustular lesions surrounding hair follicles in the ear canal due to a gram-negative bacterial infection during hot, humid weather and often initiated by swimming.
- Otitis externa can cause a conductive hearing loss. If the swelling of the canal is so marked that it obstructs the canal or if the discharge is so copious, a conductive element will result.
- **Chronic Otitis Externa: COE**



Otitis Externa

Otomycosis is a fungal infection in the ear canal. It causes pain and itching. Sometimes fluid leaks from the ear. The infection can also cause a feeling of fullness in the ear and sometimes hearing loss.

Furunculosis is the infection of the hair follicles with staphylococcus and is limited to the outer part of ear canal.



Complications of Acute and Chronic Otitis Externa

- **Cellulitis/perichondritis/chondritis:**
 - Extension of infection into soft tissues and cartilage of the auricle
- **Malignant otitis externa:**
 - Extension of infection beyond the EAC into soft tissue, mastoid and skull base; can evolve into temporal bone
- **Medial canal fibrosis:**
 - Fibrous scar of the medial EAC; a sequela of COE
- **Perforation of the tympanic membrane:**
 - Often seen in the setting of fungal OE



Malignant otitis externa

Other Outer-Ear Disorders

- **Carcinoma** of the auricle is not uncommon. Basal-cell, epidermoid, and squamous-cell carcinoma can all occur around the auricle and external auditory meatus. In addition, tumors of various types can proliferate around the external auditory meatus and auricle.
- **Hematoma** Is a collection of blood underneath the perichondrium of the ear and typically occurs secondary to trauma (Boxers, wrestlers, rugby players). Auricular deformity, commonly known as "cauliflower ear" is the result of untreated or inadequately treated auricular hematoma.
- Acute dermatitis
- Chondrodermatitis chronica helices
- Keratosis Obturans
- Cholesteatoma of the External Auditory Canal





Clinical Audiology

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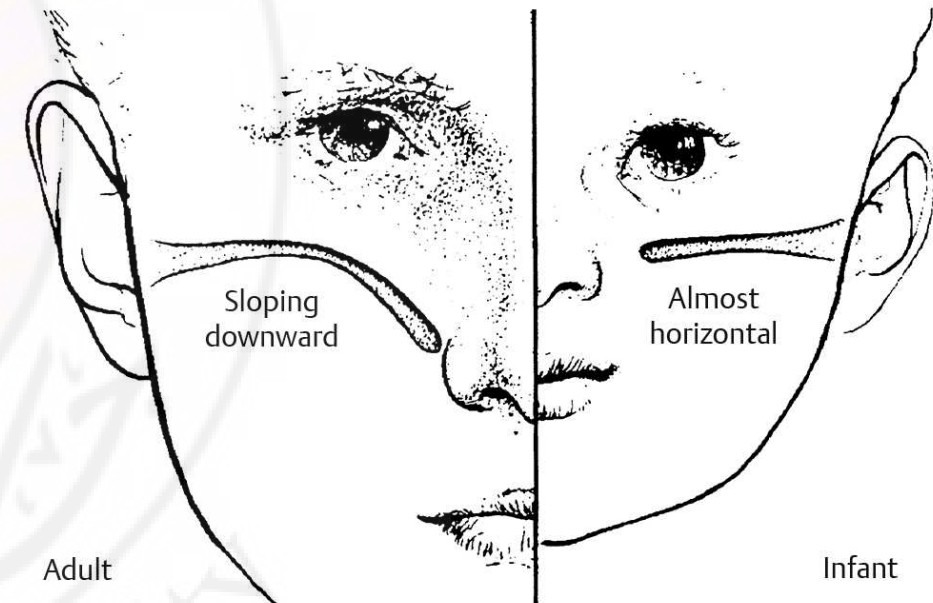


Middle Ear Disorders



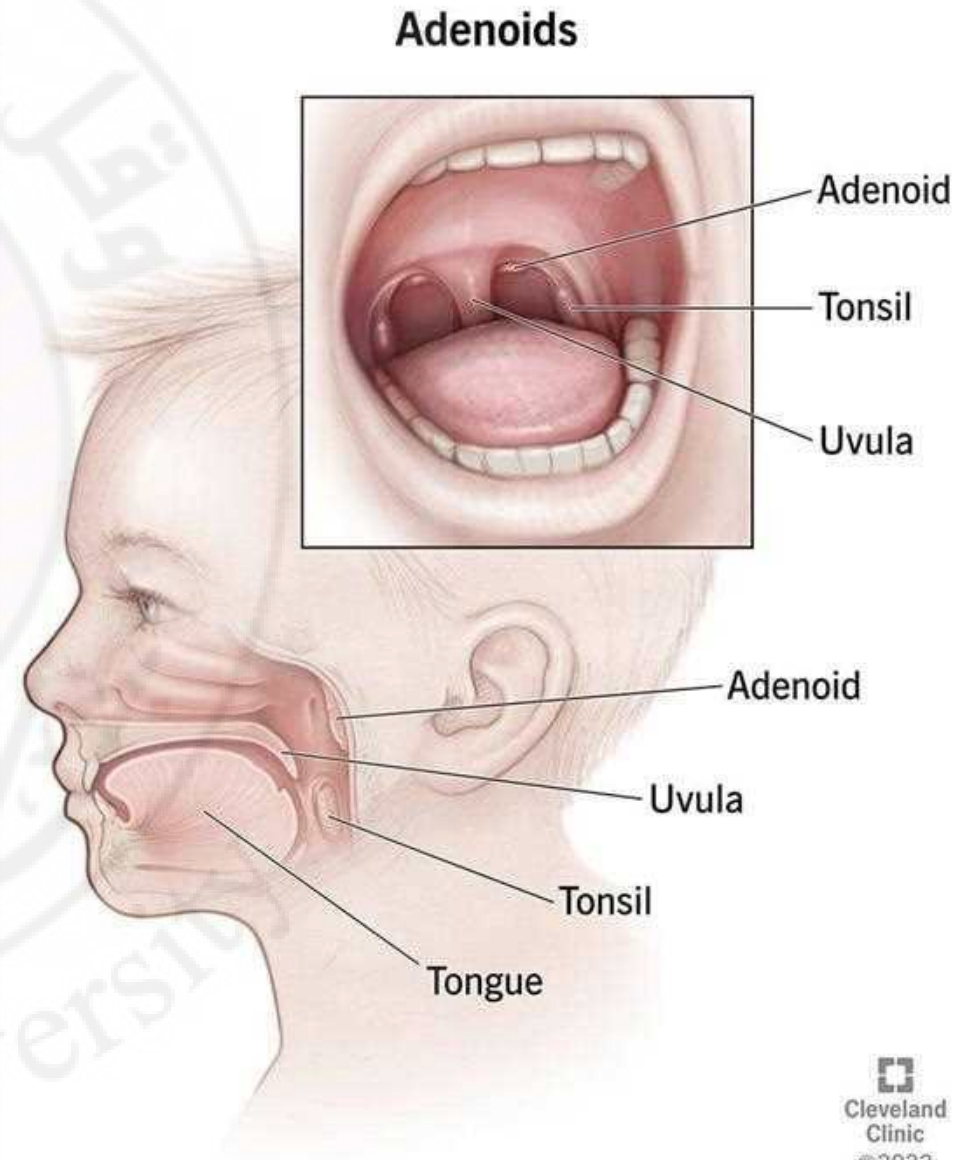
Eustachian Tube Dysfunction

- A properly functioning Eustachian tube provides the middle ear with **ventilation** and **drainage**, and maintains the same amount of **pressure** on both sides of the tympanic membrane.
- The Eustachian tube is normally closed, and that it is opened by the **tensor palatini muscle** during activities such as chewing and yawning.
- In fact, middle ear fluid appears to be significantly less common in 2- to 6-year olds who regularly chew gum compared with those who do not.
- Problems with this process can lead to an accumulation of fluid in the middle ear and the possible development of more serious disease.



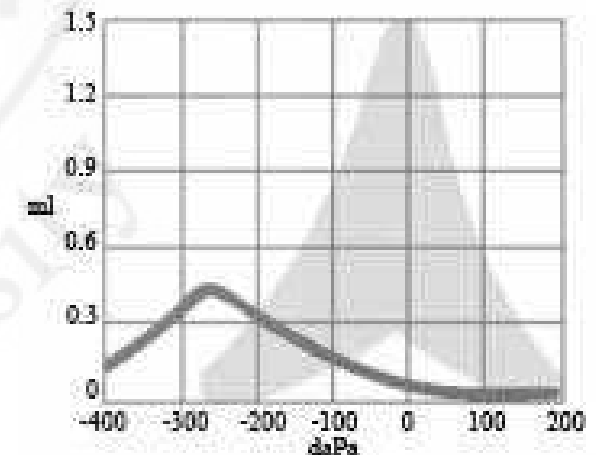
Eustachian Tube Dysfunction

- Eustachian tube dysfunction exists when the tube does not open properly, is not patent, or is **blocked** in one way or another.
- The Eustachian tube might be blocked by edema (swelling) and/or fluid caused by an upper respiratory infection, sinusitis, inflamed adenoids, or allergies obstruction by hypertrophic (enlarged) adenoids or obstruction or encroachment by a nasopharyngeal tumor or other growth.
- The Eustachian tube may also fail to open appropriately (e.g., upon swallowing or yawning) due to structural or functional abnormalities affecting the tensor palatini muscle. This problem is common among patients with cleft palate.



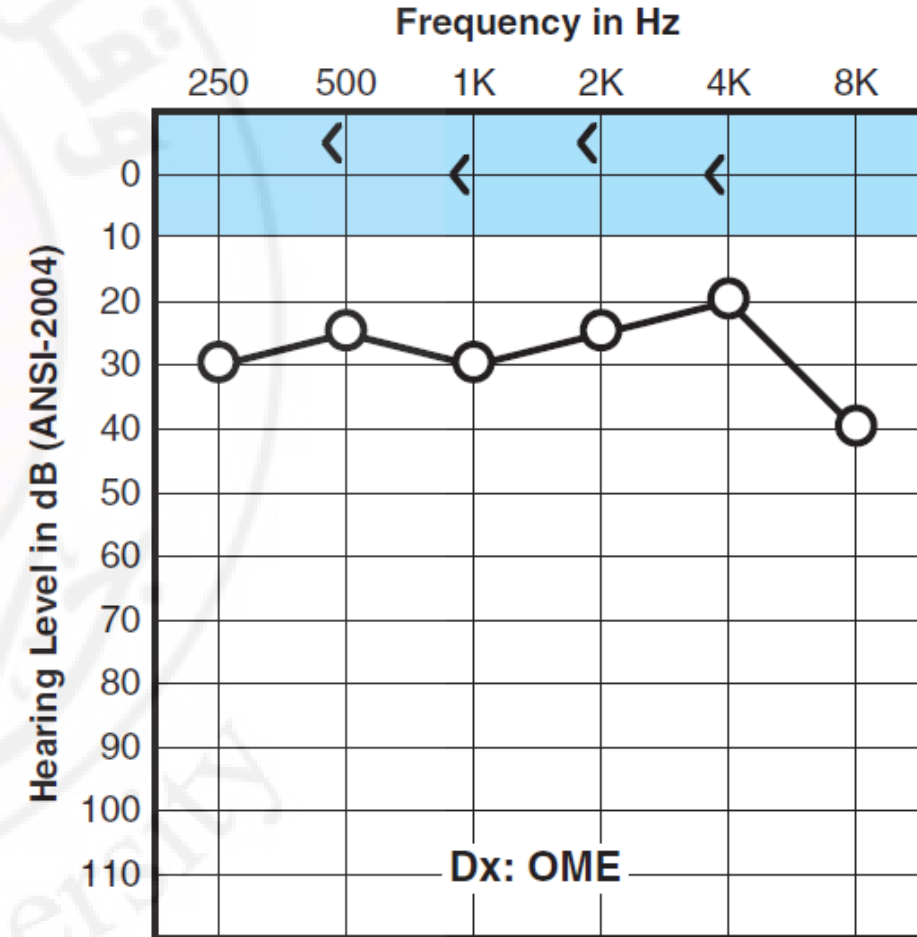
Eustachian Tube Dysfunction

- Eustachian tube dysfunction prevents the middle ear from being ventilated, causing the air within the middle ear cavity to become stagnant. Part of the stagnant air is then absorbed by the tissues of the middle ear.
- As a result, the air pressure will be lower inside the middle ear than it is outside in the surrounding atmosphere. This situation is called **negative pressure** in the middle ear.
- This pressure difference on the two sides of the tympanic membrane (higher outside, lower inside) causes it to be drawn inward, or **retracted**.



Otitis Media

- The most common cause of transient conductive hearing loss in children is **otitis media with effusion**.
- Otitis media is inflammation of the middle ear. It is caused primarily by **Eustachian tube dysfunction**. When it is accompanied by middle-ear effusion, otitis media often causes conductive hearing loss.
- There are a number of ways to classify otitis media, including by type, effusion type, and duration.
- Estimates are that from 76 to 95% of all children have one episode of otitis media by 6 years of age. The prevalence of otitis media is highest during the first 2 years and declines with age.



Various descriptions of otitis media, based on type, effusion type, and duration (1)

Description	Definition
otitis media	inflammation of the middle ear, resulting predominantly from Eustachian tube dysfunction
otitis media, acute	inflammation of the middle ear having a duration of fewer than 21 days
otitis media, acute serous	acute inflammation of middle-ear mucosa with serous effusion
otitis media, acute suppurative	acute inflammation of the middle ear with infected effusion containing pus
otitis media, adhesive	inflammation of the middle ear caused by prolonged Eustachian tube dysfunction resulting in severe retraction of the tympanic membrane and obliteration of the middle-ear space
otitis media, chronic	persistent inflammation of the middle ear having a duration of greater than 8 weeks
otitis media, chronic adhesive	longstanding inflammation of the middle ear caused by prolonged Eustachian tube dysfunction resulting in severe retraction of the tympanic membrane and obliteration of the middle-ear space

Various descriptions of otitis media, based on type, effusion type, and duration (2)

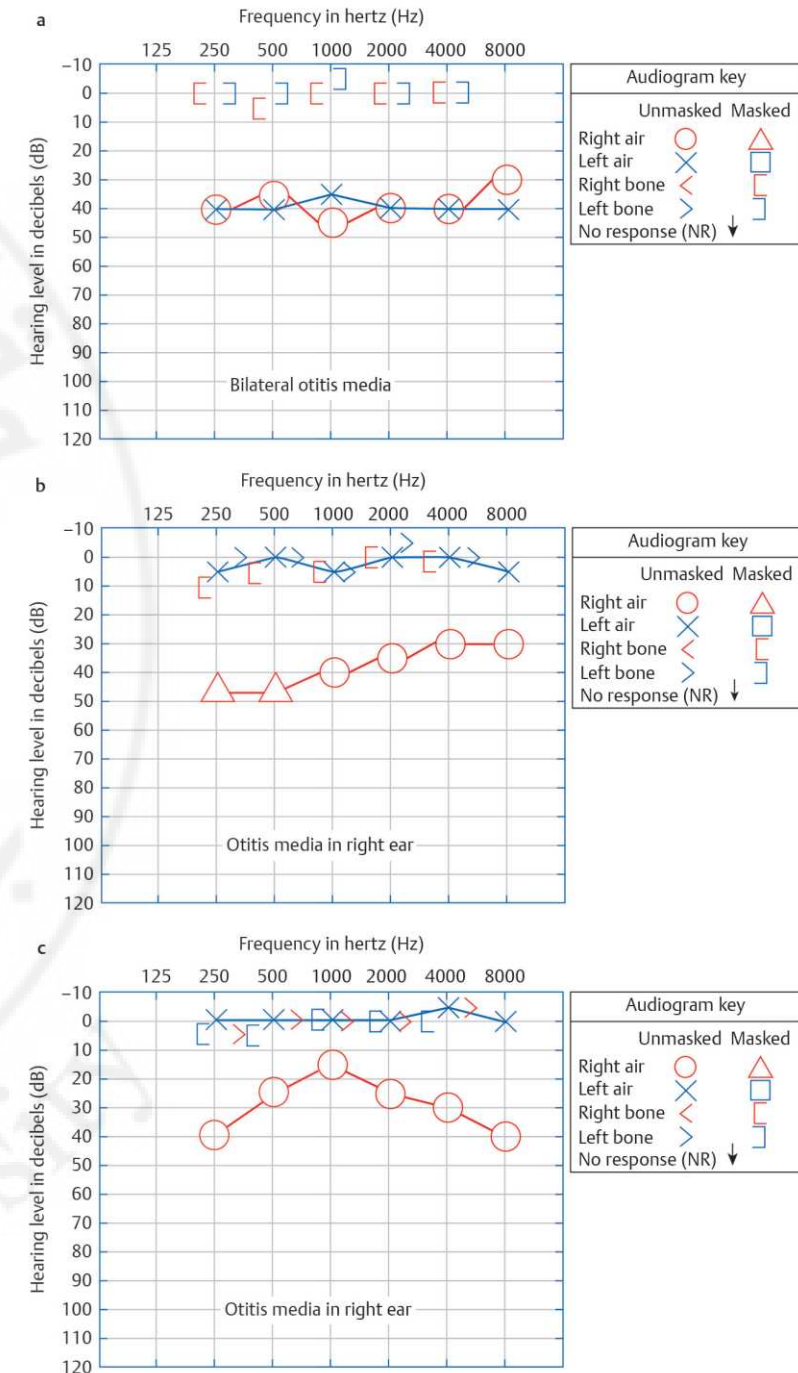
Description	Definition
otitis media, chronic suppurative	persistent inflammation of the middle ear with infected effusion containing pus
otitis media, mucoid	inflammation of the middle ear, with mucoid effusion
otitis media, mucosanguinous	inflammation of the middle ear with effusion consisting of blood and mucus
otitis media, necrotizing	persistent inflammation of the middle ear that results in tissue necrosis
otitis media, nonsuppurative	inflammation of the middle ear with effusion that is not infected, including serous and mucoid otitis media
otitis media, persistent	middle-ear inflammation with effusion for 6 weeks or longer following initiation of antibiotic therapy
otitis media, purulent	inflammation of the middle ear with infected effusion containing pus; synonym: suppurative otitis media
otitis media, recurrent	middle-ear inflammation that occurs 3 or more times in a 6-month period

Various descriptions of otitis media, based on type, effusion type, and duration (3)

Description	Definition
otitis media, secretory	otitis media with effusion, usually referring to serous or mucoid effusion
otitis media, serous	Inflammation of middle-ear mucosa, with serous effusion
otitis media, subacute	inflammation of the middle ear ranging in duration from 22 days to 8 weeks
otitis media, suppurative	inflammation of the middle ear with infected effusion containing pus
otitis media, unresponsive	middle-ear inflammation that persists after 48 hours of initial antibiotic therapy, occurring more frequently in children with recurrent otitis media
otitis media with effusion	inflammation of the middle ear with an accumulation of fluid of varying viscosity in the middle-ear cavity and other pneumatized spaces of the temporal bone; synonym: seromucinous otitis media
otitis media without effusion	inflammation of the middle ear
otitis prone child	if otitis media occurs before the age of 1 year or if six bouts occur before the age of 6 years ***

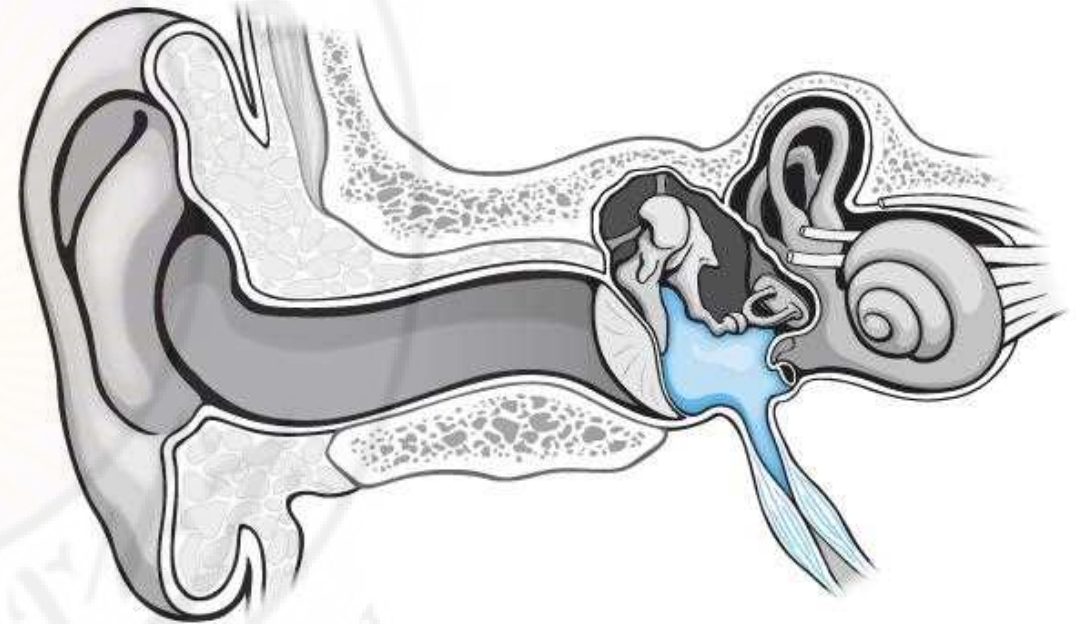
Otitis Media & CHL

- Conductive hearing loss is certainly the effect with which we are most directly concerned. However, any amount of conductive loss can occur with otitis media, and fluctuation is common.
- In addition to flat conductive losses, patients with otitis media may also have audiograms with poorer thresholds in the low frequencies, and others that are somewhat tent-shaped.
- Conductive losses associated with middle ear effusion in children tend to come with relatively flat audiograms with thresholds (at 500, 1000, & 2000Hz) averaging in the 18–35 dB HL range across studies.
- Moreover, otitis media can also have a sensorineural component, particularly in the high frequencies. One of the possible mechanisms for a sensorineural component may be the transmission of toxins to the perilymph via the round window.



Otitis Media in Children

- developmental deficits in communication and behavioral/attention problems experienced by some children with recurrent otitis media are, for the most part, auditory-based.
- There is increasing evidence that the age of onset, as well as the nature, degree, and configuration of the peripheral conductive hearing loss which occurs secondary to otitis media, are critical components that place children at risk for developing [communication and learning disorders](#).

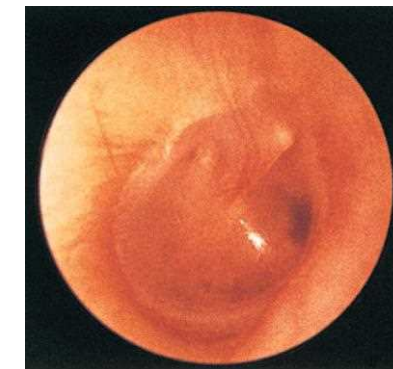


Otitis Media in Children

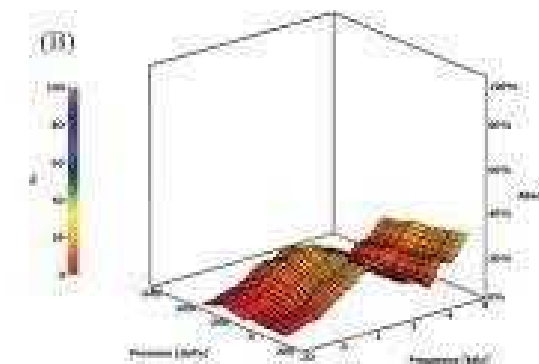
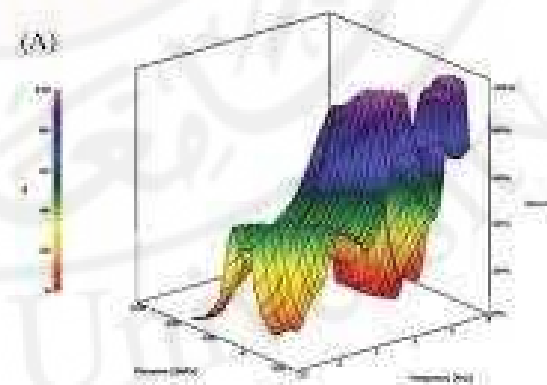
- It is not the degree of hearing loss alone that is an issue, but the intrinsic nature of the conductive hearing loss associated with otitis media and middle ear effusion particularly when it occurs in early life.
- The hearing deficit is characterized as **fluctuant**; that is, existing only during the duration of the otitis media episode. At resolution, or between otitis media episodes, hearing presumably returns to the "normal" range. Therefore, hearing sensitivity may vary within the same episode of otitis media, as well as between episodes within the same child; the actual number of episodes the child experiences within a particular time period is an additional consideration.
- Finally, **asymmetries in hearing sensitivity** may exist between the child's two ears, thereby potentially disrupting critical binaural auditory processing skills.



Audiologic findings in Otitis Media



- **Otoscopy** (Middle ear fluid sometimes can be detected otoscopically by seeing a fluid line or **meniscus** or the presence of bubbles)
- **Audiometry (AC & BC):**
 - One of the consequences of otitis media with effusion is conductive hearing loss. The degree and configuration of the loss depends on the amount and type of fluid and its influence on functioning of the tympanic membrane and ossicles. **Air-bone gap of less than 60 dB, with excellent speech recognition scores at MCL.**
- **Tympanometry:**
 - Flattened tympanogram (Type B) or negative MEP (Type C). Tympanogram could be abnormal even with normal pure tone audiogram.
- **Acoustic reflexes:**
 - Absent
- **Wideband tympanometry**
- **Tuning Fork tests**
- **OAEs**
 - Absent



Diagnostic criteria

Middle ear effusion demonstrated by pneumatic otoscopy and/or tympanogram

and

Moderate-to-severe eardrum bulging

or

Recent onset of otorrhea (not caused by external otitis)

or

Mild eardrum bulging

and

Recent onset of pain

or

Intense reddening of the eardrum

Severe symptoms

Temperature $\geq 102.2^{\circ}\text{F}$ ($\geq 39^{\circ}\text{C}$)

or

Moderate or severe pain

or

Pain for ≥ 48 hours

Nonsevere symptoms

Temperature $\leq 102.2^{\circ}\text{F}$ ($\leq 39^{\circ}\text{C}$)

and

Mild pain for <48 hours

Diagnostic and severity criteria for acute otitis media in children

Case Example

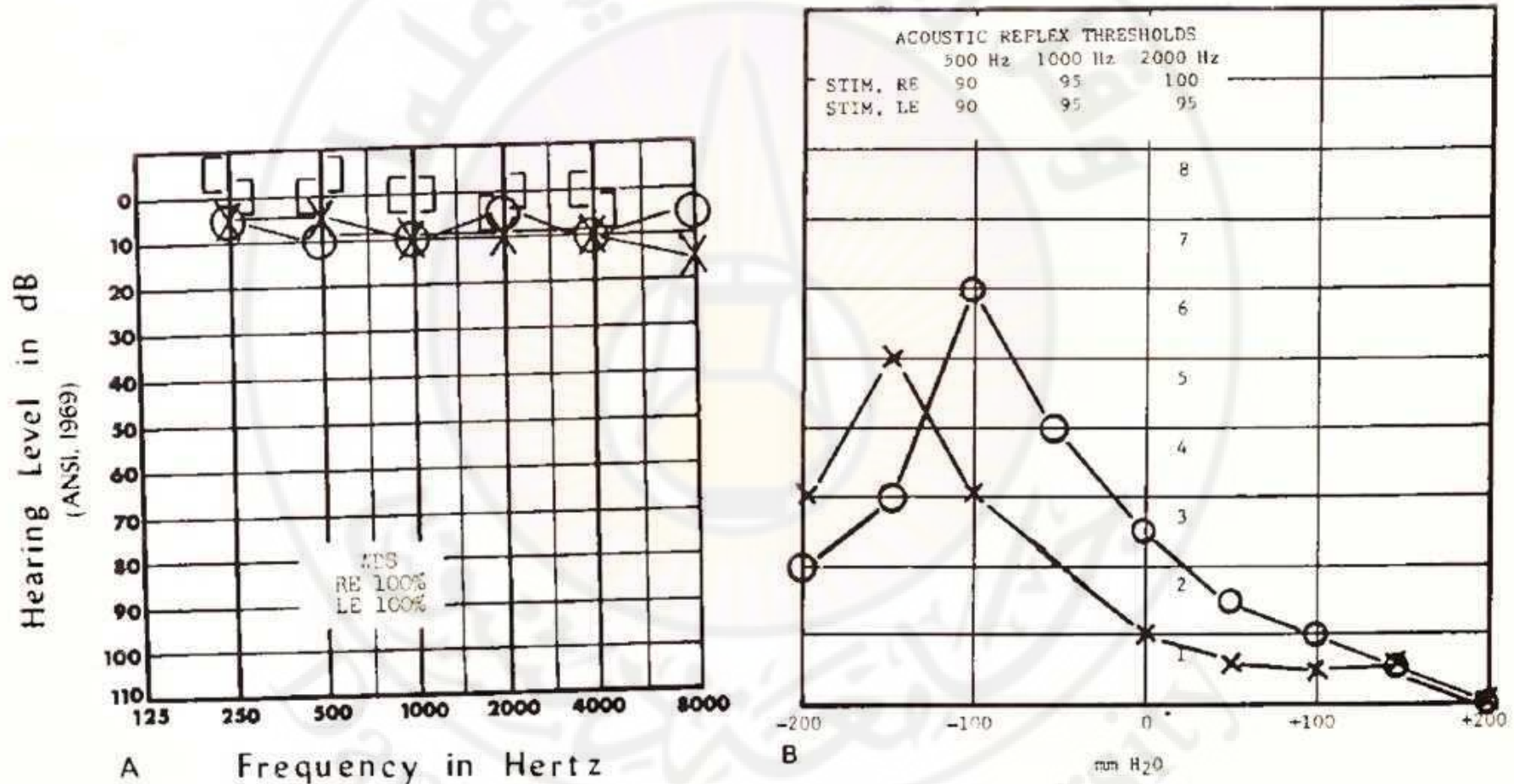


Fig. 2.5. (A) Audiometric results of girl, age 5, with bilateral acute otitis media. (B) Tympanometry curves of girl, age 5, with bilateral acute otitis media.

Case Example

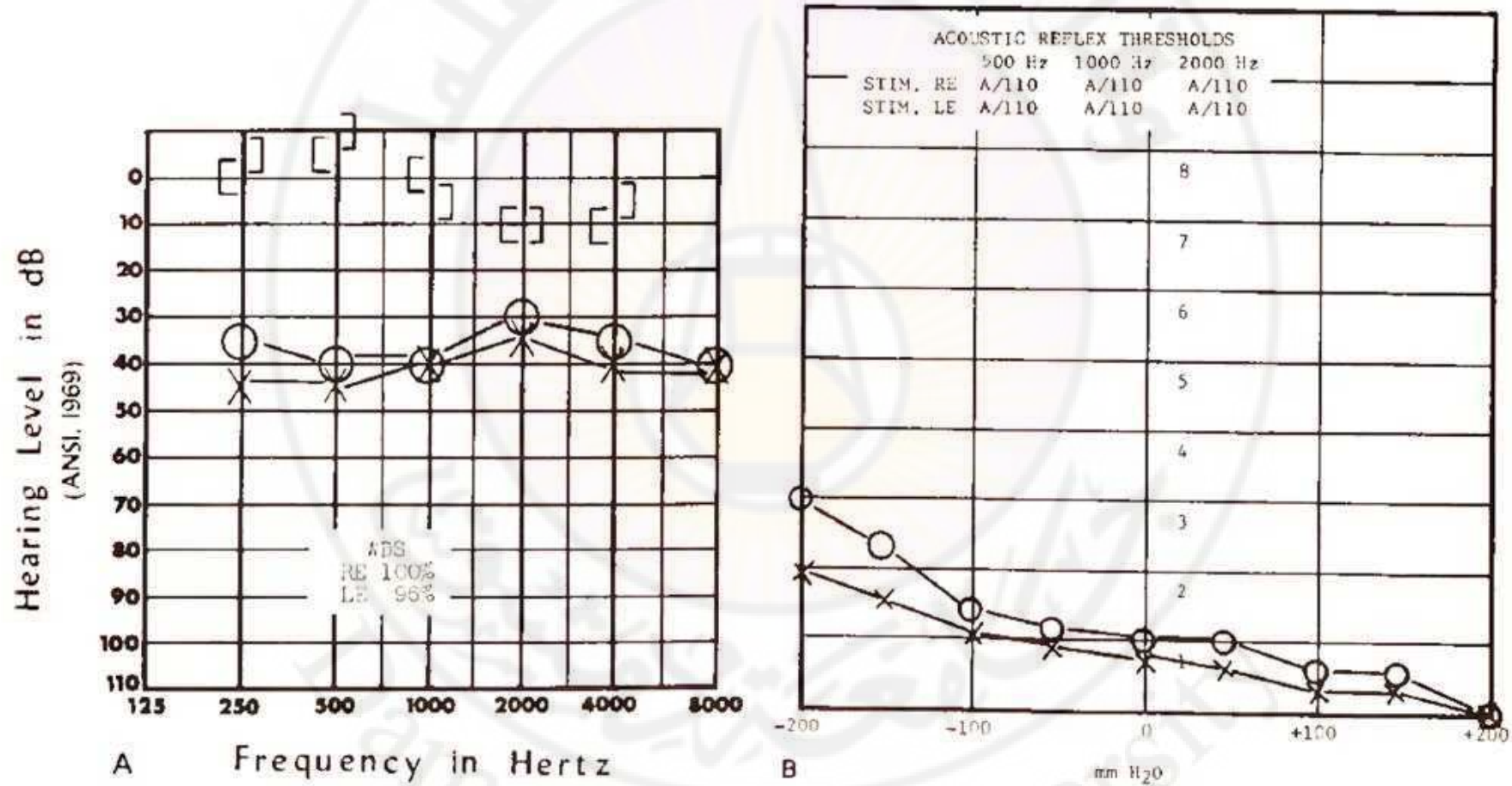
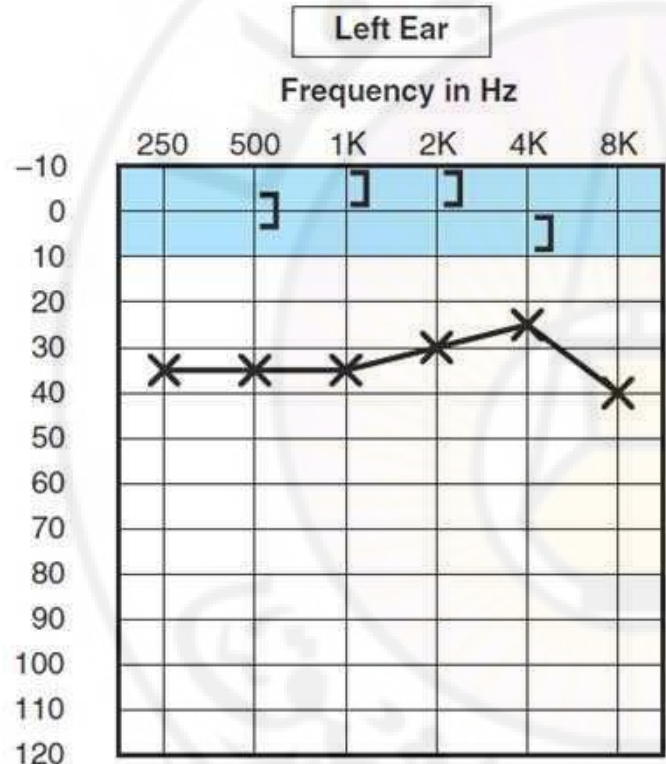
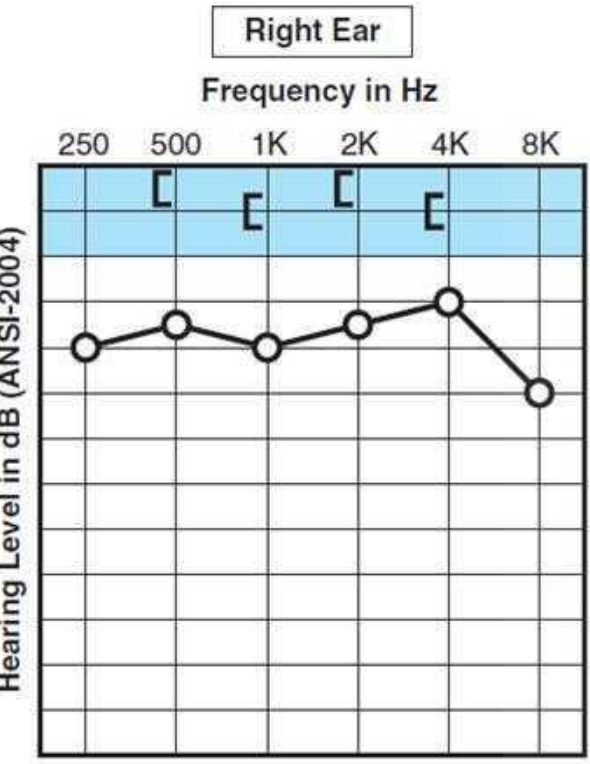


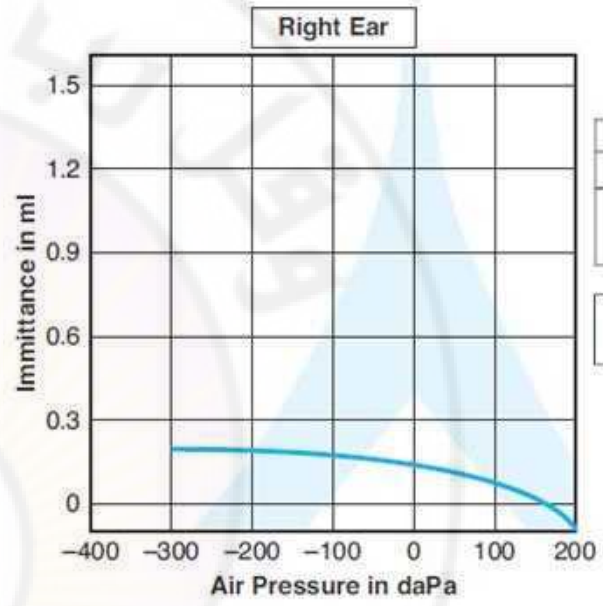
Fig. 2.7. (A) Audiometric findings of 7-year-old girl with bilateral serous otitis media. (B) Tympanograms of same patient as shown in A, with bilateral serous otitis media.

Case Example



Air Conduction	Right	Left
Unmasked	○	×
Masked	△	□
Bone Conduction	Right	Left
Unmasked	<	>
Masked	[	]

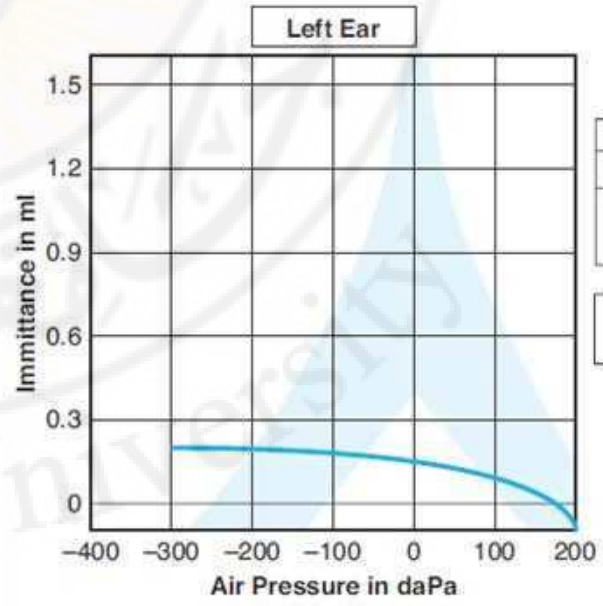
Speech Audiometry					
Right			Left		
25	dB	SRT	25	dB	
100	%	PSI-W _m	100	%	
100	%	PSI-S _m	100	%	
100	%	PSI-CCM	90	%	



Signal to RE	BBN	500	1K	2K	4K
Crossed	>110				
Uncrossed			>110		

PTA - BBN + 5 = CNE
Corr SPAR

Static Immittance = 0.2

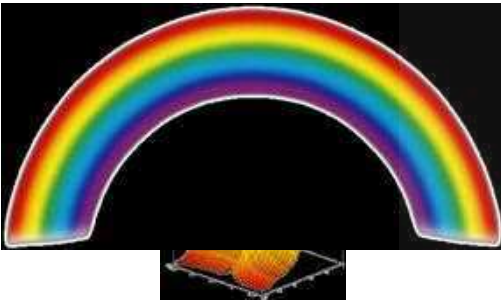


Signal to LE	BBN	500	1K	2K	4K
Crossed	>110				
Uncrossed			>110		

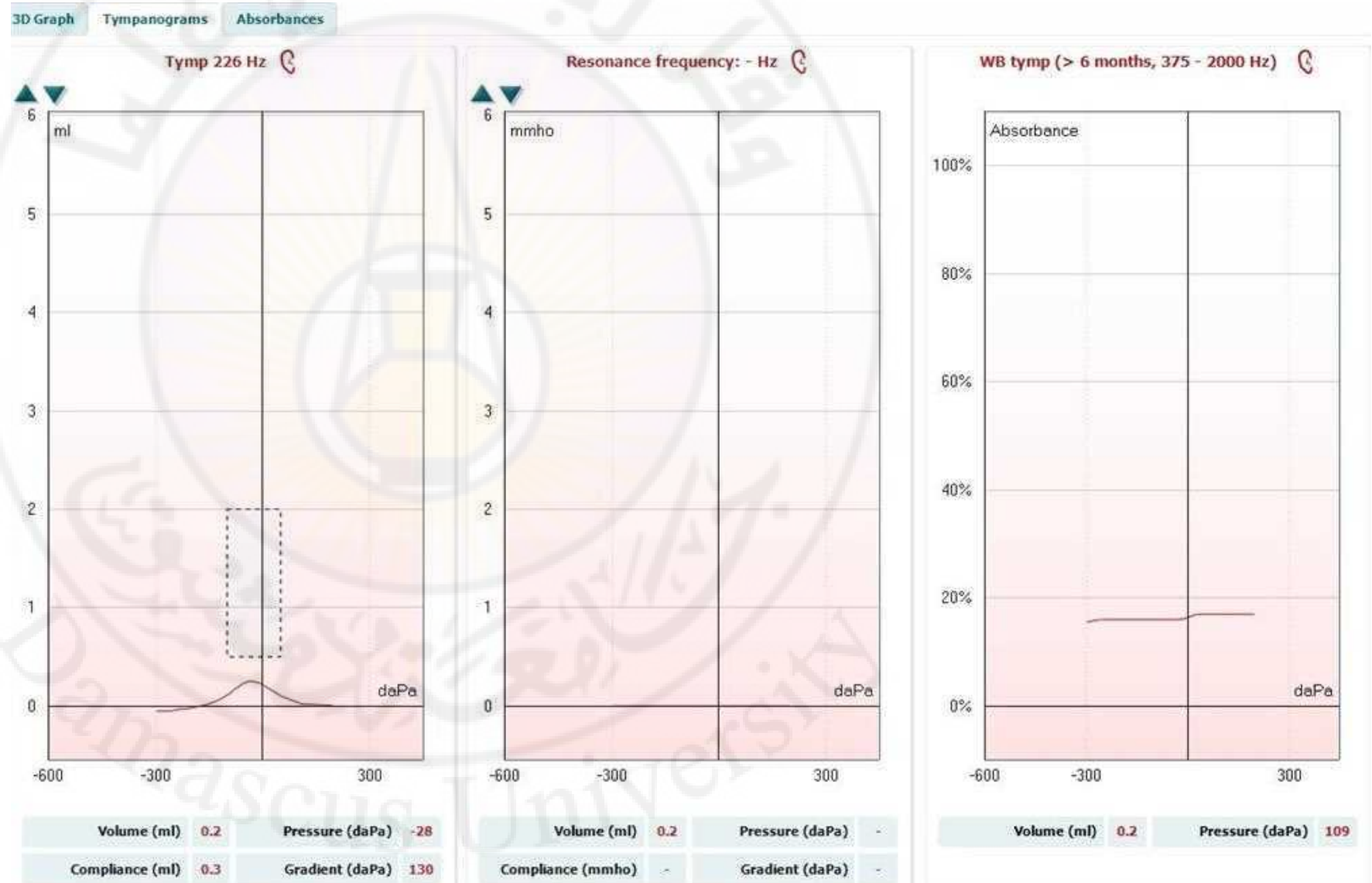
PTA - BBN + 5 = CNE
Corr SPAR

Static Immittance = 0.2

Case Example



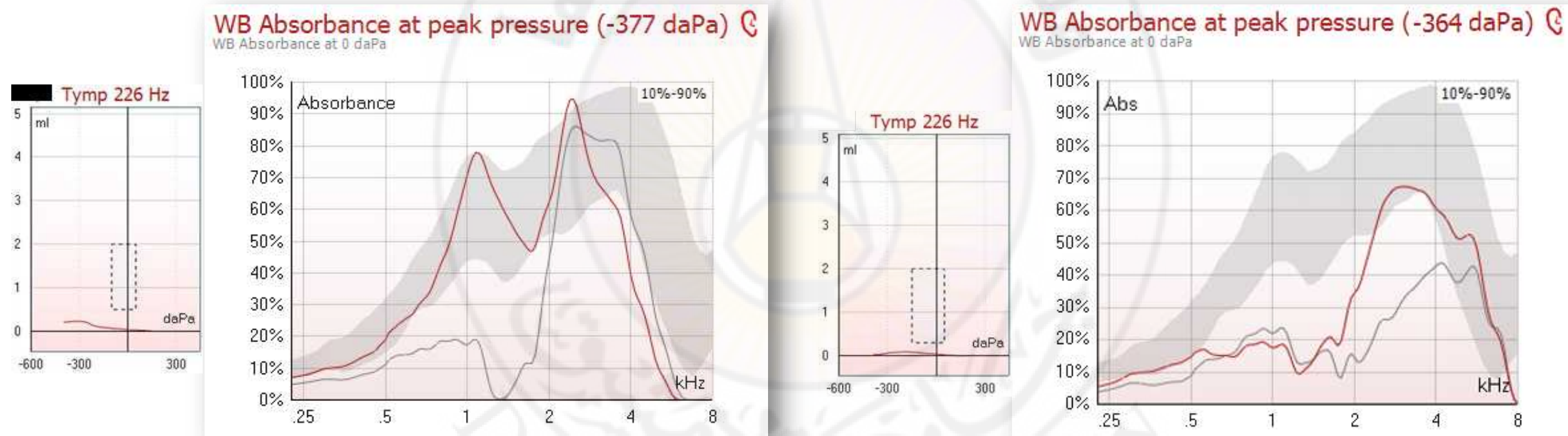
Improved Sensitivity
Improved accuracy over
standard tympanometry

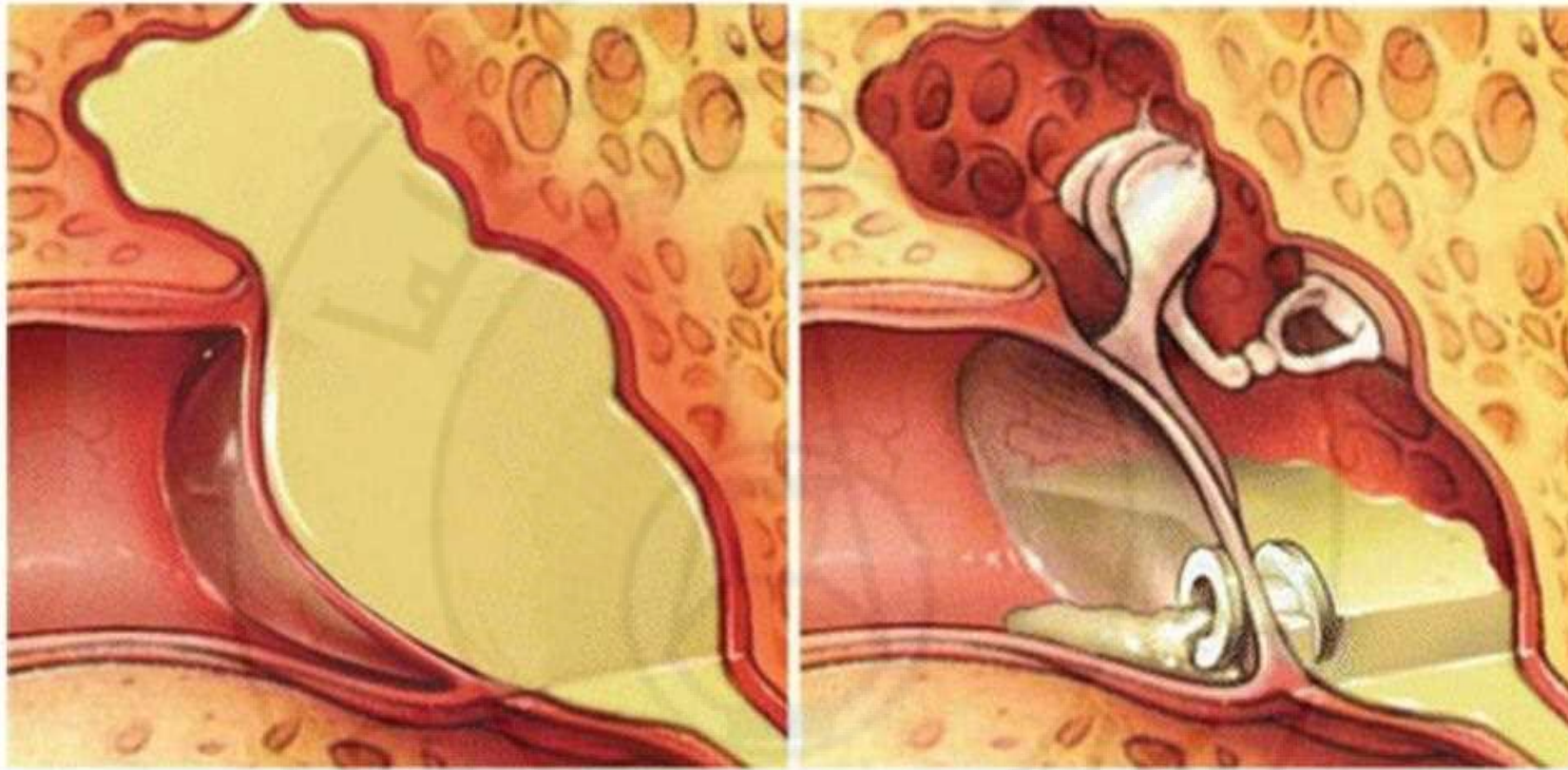


Know your Type C Tympanogram

Know when OME is
present in the middle ear

Case Example





Treatment of Otitis Media with Effusion

A **ventilation tube**, also known as a **tympanostomy tube** or **grommet**, is a small tube that is inserted into the eardrum to help drain fluid that has accumulated in the middle ear. This procedure is often used to treat middle ear effusion. The ventilation tube allows air to enter the middle ear space, equalizing pressure and preventing the buildup of fluid. It can also help prevent future infections by allowing drainage of any fluid that accumulates.

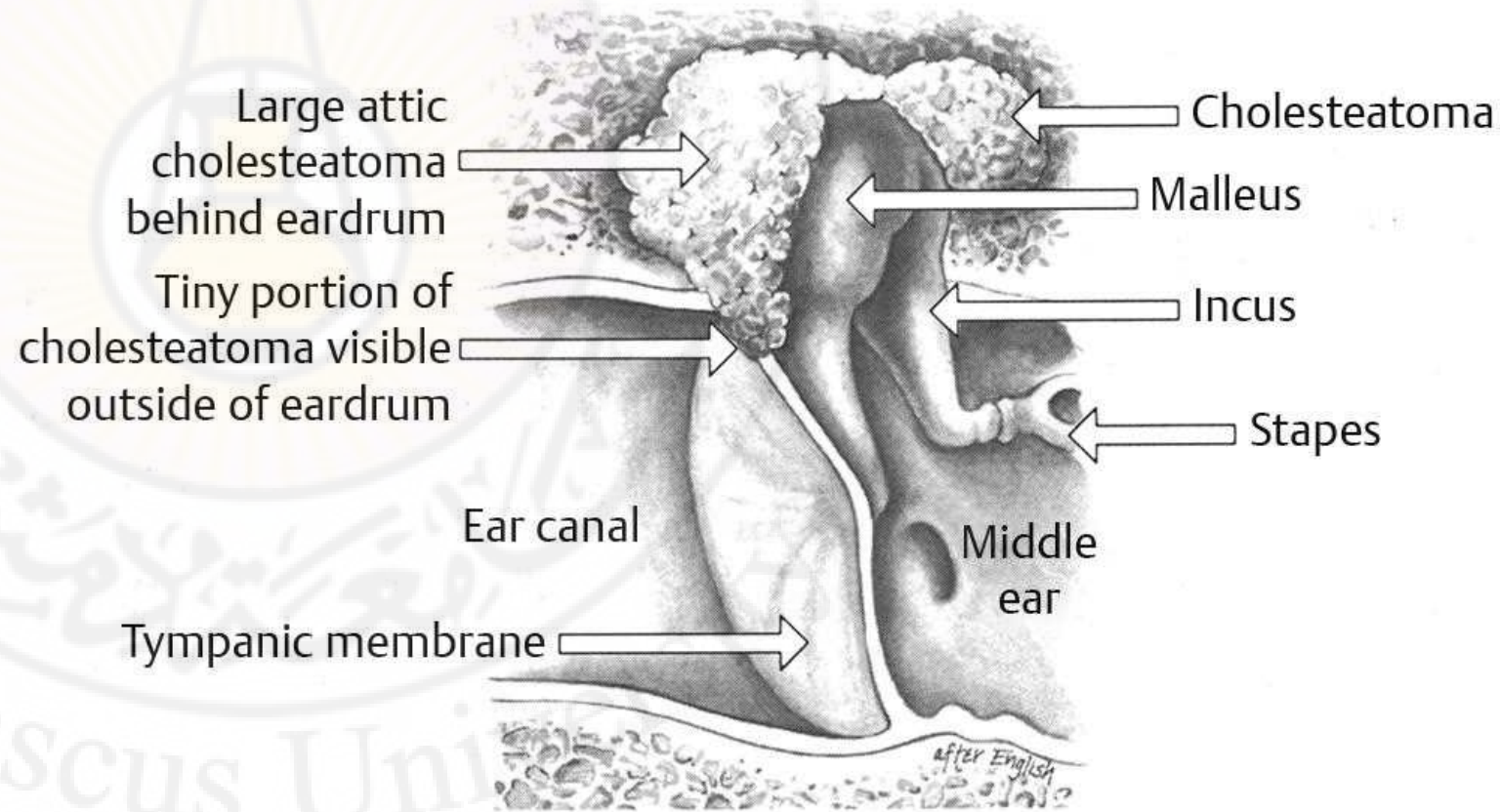
Audiological services for patients with ventilation tubes:

- (1) **Custom-made earplugs** often called “swimming earmolds” or “earplugs” are often used to prevent water from entering the middle ear via the tympanostomy tubes when the patient swims, showers, and engages in similar activities. However, the routine use of earplugs and other water precautions is not recommended for children except when the child has current or prior otorrhea or problems when exposed to water.
- (2) Periodic **monitoring with acoustic immittance** tests is needed to determine if the tympanostomy tubes is still in place and patent, or if it has become clogged or dislodged.
- (3) Audiological assessments are necessary so that **residual hearing** impairments or changes in hearing over time can be identified and addressed.



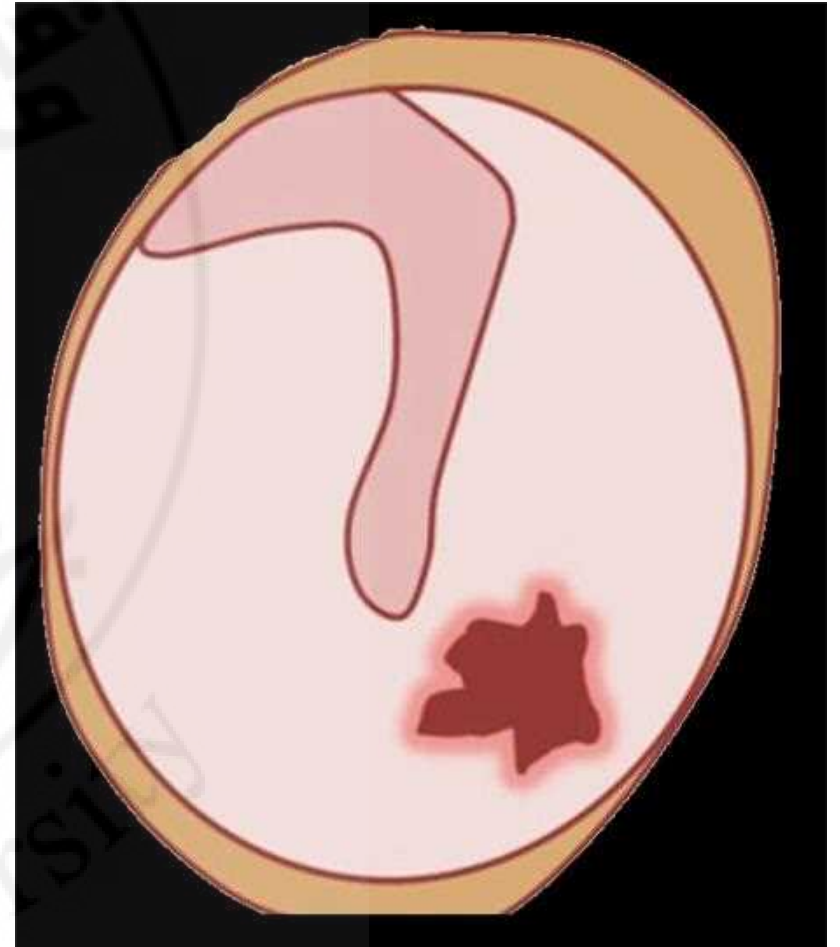
Complications and Sequelae of Otitis Media

- Cholesteatoma
- Facial paralysis
- Labyrinthitis
- Mastoiditis
- Ossicular discontinuities
- TM perforation



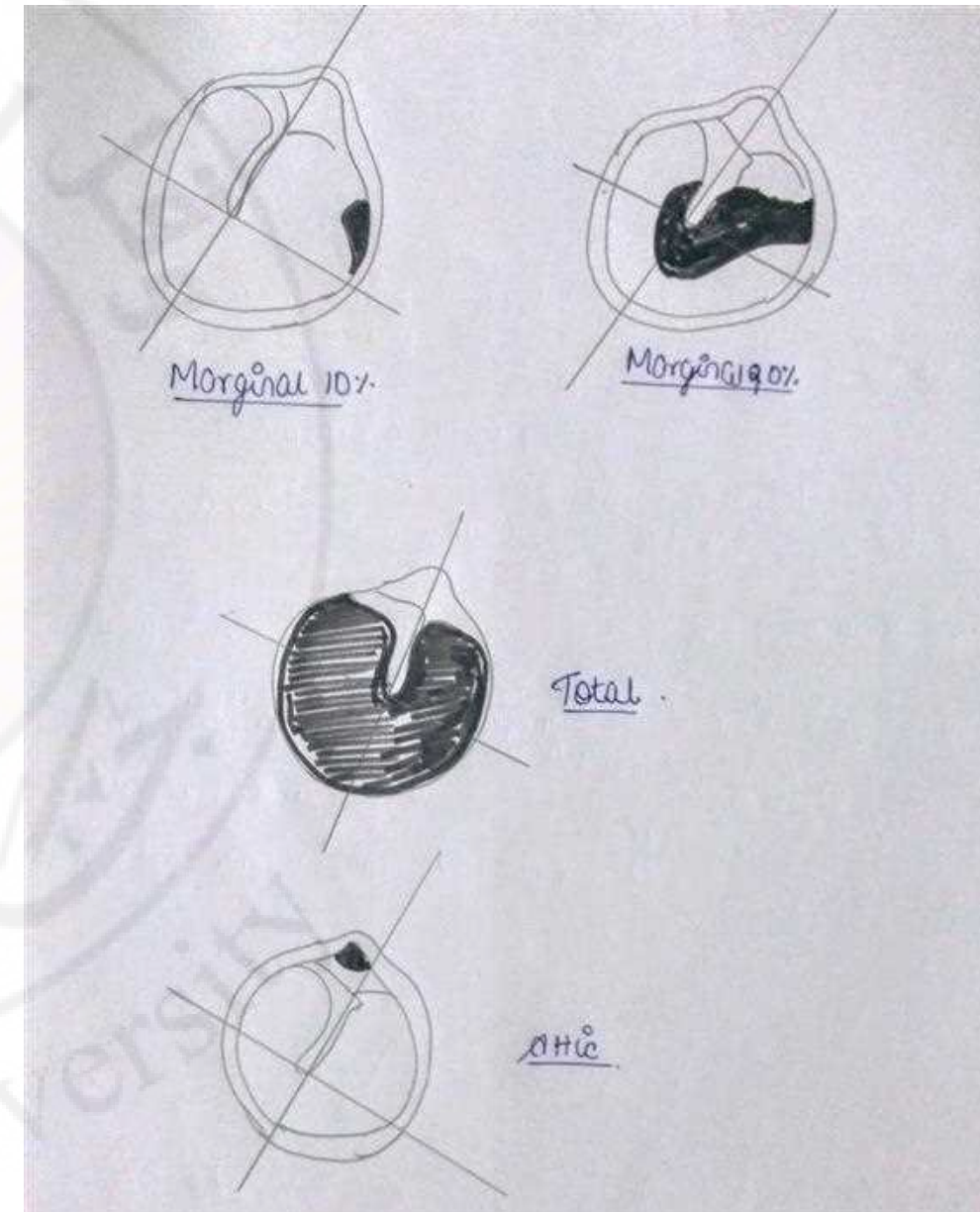
Tympanic-Membrane Perforation

- Perforation of the tympanic membrane is a common **complication of middle-ear infection**. Sometimes, in the later stages of an acute attack of otitis media, the fluid trapped in the middle-ear space is so excessive that the membrane ruptures to relieve the pressure.
- In other cases, chronic middle-ear infections erode portions of the tympanic membrane, weakening it to the point that a perforation forms.
- The tympanic membrane can also be perforated by **trauma**. Traumatic perforation can occur when a foreign object is placed into the ear canal in an unwise effort to remove cerumen. It can also occur with a substantial blow to the side of the head.

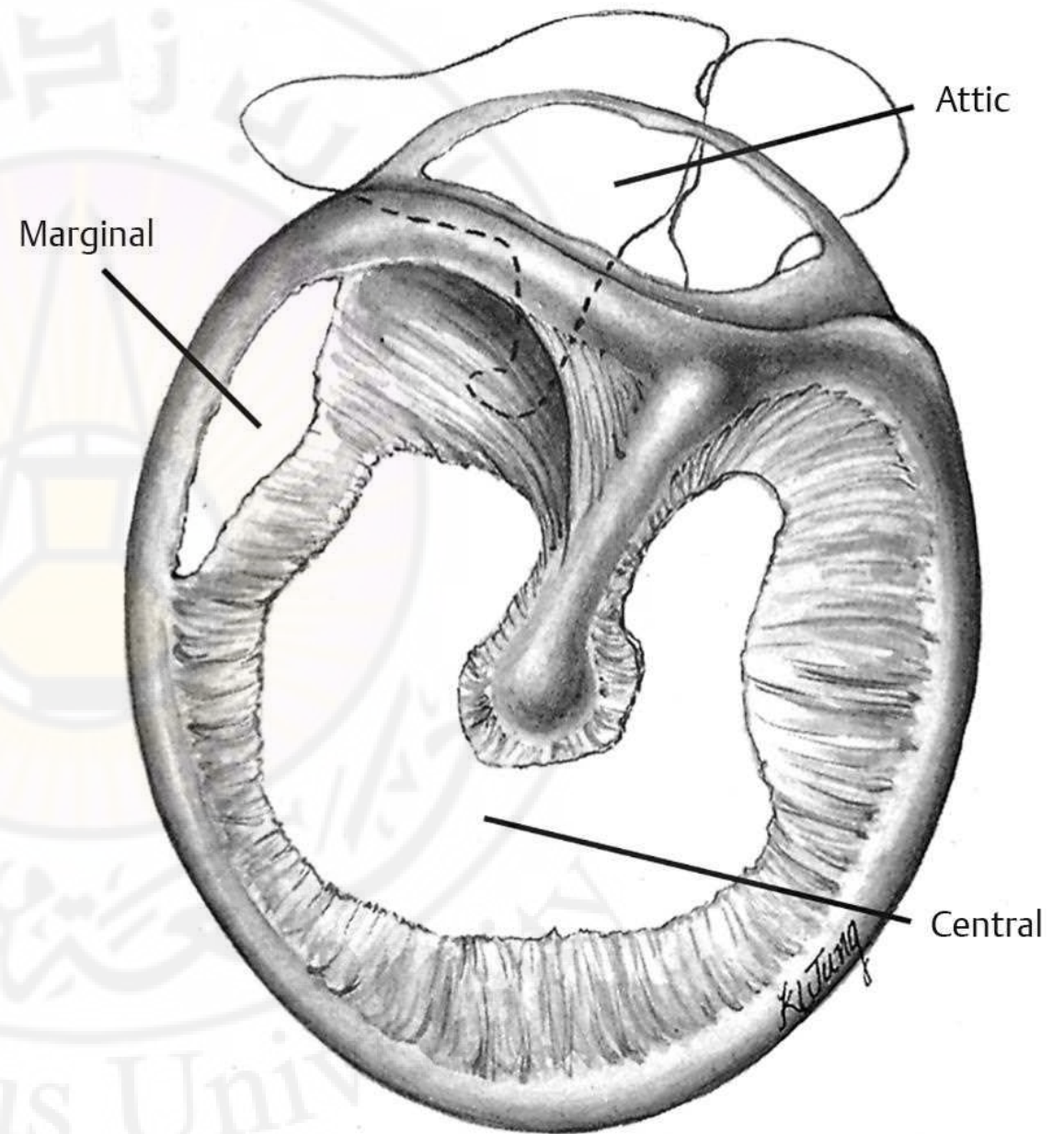


Tympanic-Membrane Perforation

- Perforations of the tympanic membrane are of three types, defined by **location**.
- A **central perforation** is one of the pars tensa portion of the tympanic membrane, with a rim of the membrane remaining at all borders.
- An **attic perforation** is one of the pars flaccida portion.
- A **marginal perforation** is one located at the edge or the margin of the membrane.
- Regardless of type, most perforations heal spontaneously. Those that do not are usually too large or are sustained by recurrent infection. In such cases, **surgery** may be necessary to repair or replace the membrane.

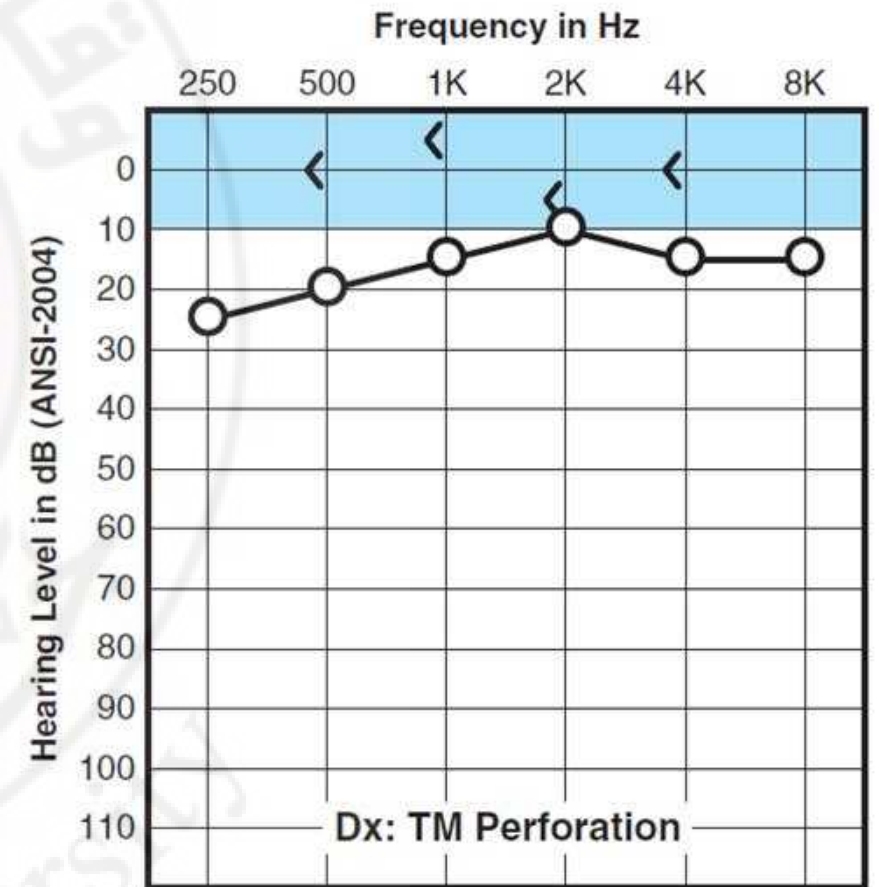


Examples of central, marginal, and attic perforations of the tympanic membrane.



Tympanic-Membrane Perforation

- A perforation in the tympanic membrane may or may not result in hearing loss, depending on its **location and size**. In general, if a perforation causes a hearing loss, it will be a mild, conductive hearing loss.
- The larger the perforation the more the loss of sound pressure which can be transmitted to the inner ear. However, if the perforation is small but located directly over the round window, the loss may be even greater than that due to a larger perforation located elsewhere. Size and location are therefore both important factors.



Tympanic-Membrane Perforation

- Eardrum perforations produce conductive hearing losses because they:
- (1) reduce the size of the drum's vibrating surface area, which reduces the eardrum-to-oval window area advantage,
- (2) impair the coupling of the eardrum to the ossicular chain so that signal transmission is reduced, and
- (3) interfere with the proper phase relationship at the oval and round windows by allowing sound to impact upon the round window.
- **In general, the magnitude of the conductive hearing loss gets worse as the size of the perforation increases.**

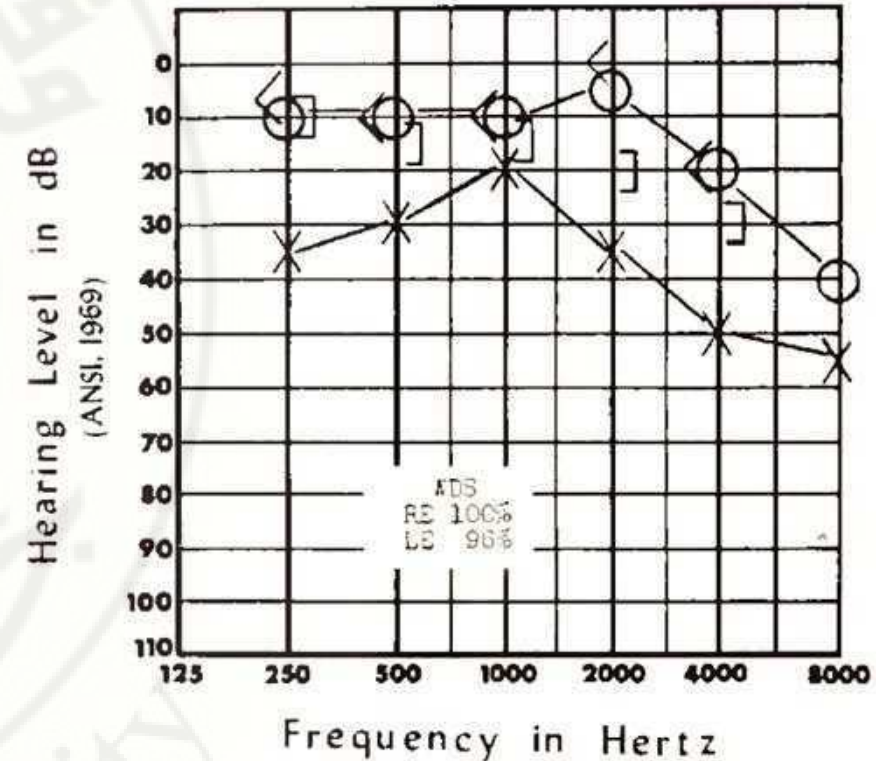
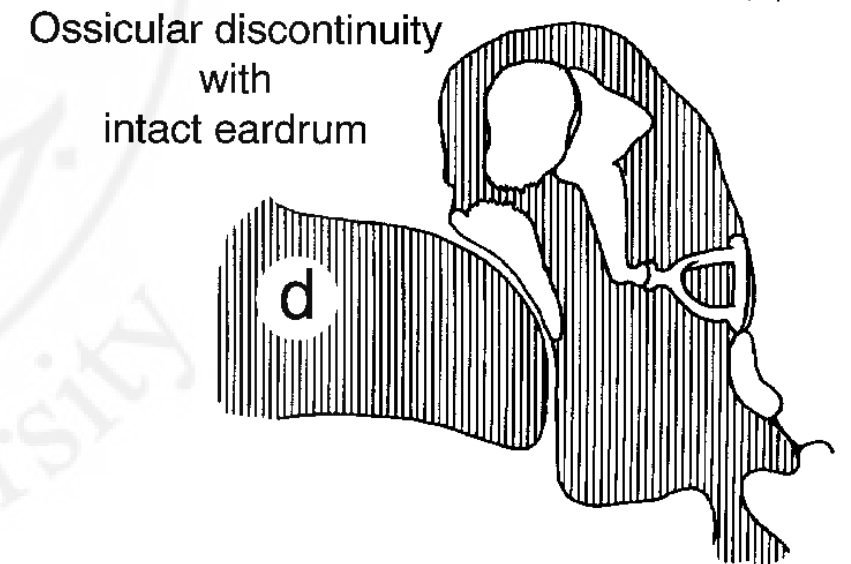
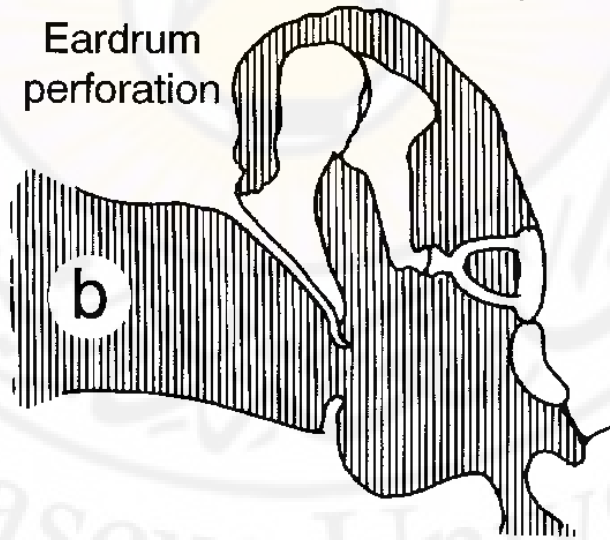
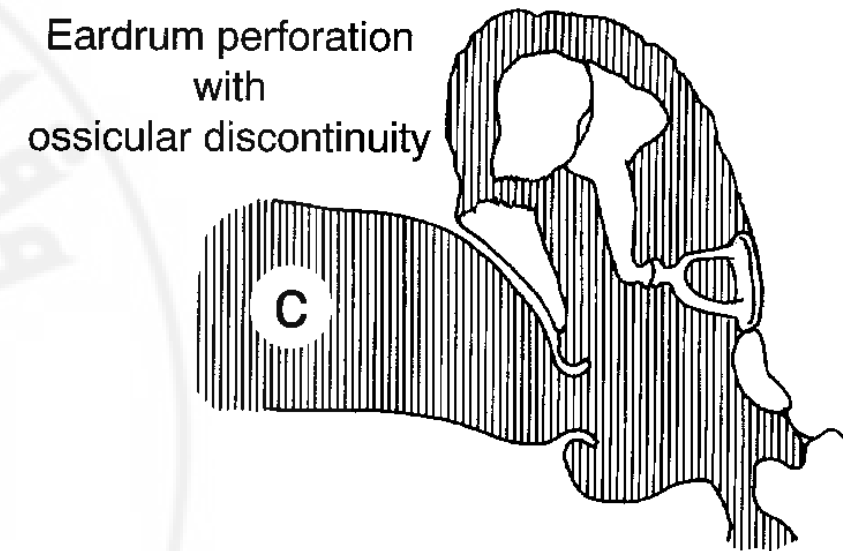
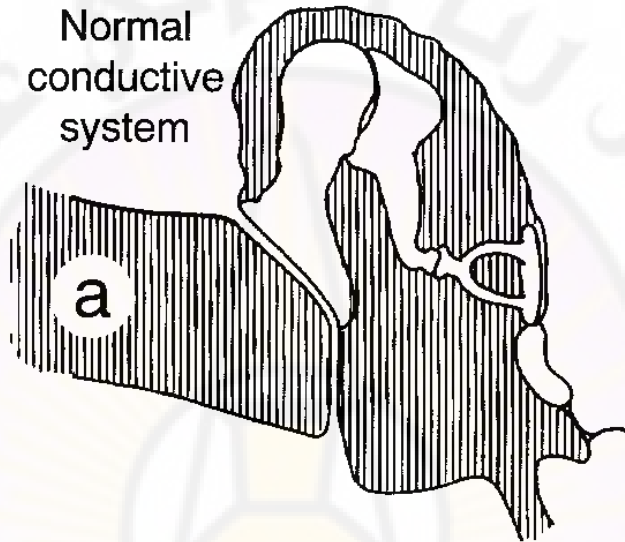
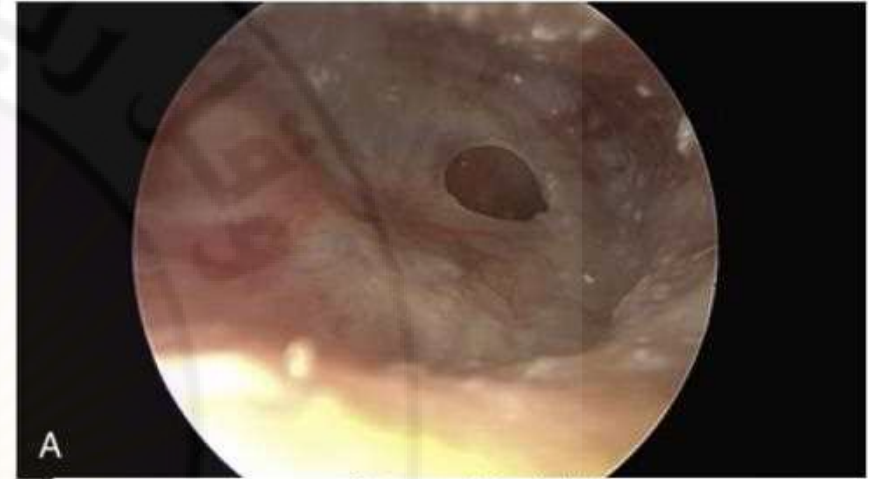


Fig. 2.6. Hearing test results of 50-year-old man with a large central tympanic membrane perforation, left ear.

(a) Normal tympanic membrane and ossicular chain; (b) perforated eardrum with intact ossicles; (c) perforated eardrum with interrupted ossicular chain due to eroded malleus; (d) intact eardrum with interrupted ossicular chain due to eroded malleus.

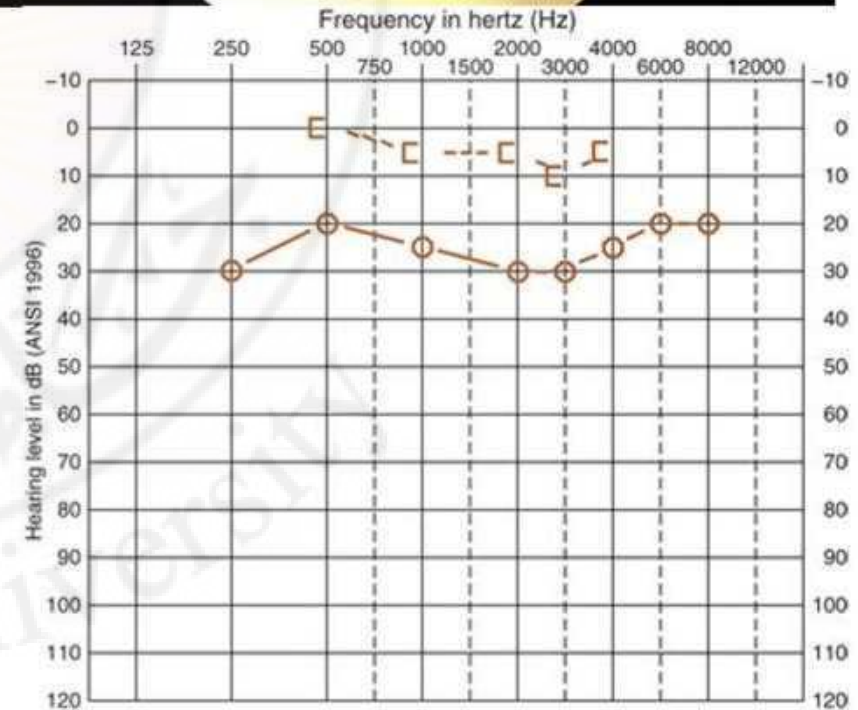


Case Example



TM perforation with undisturbed ossicular continuity

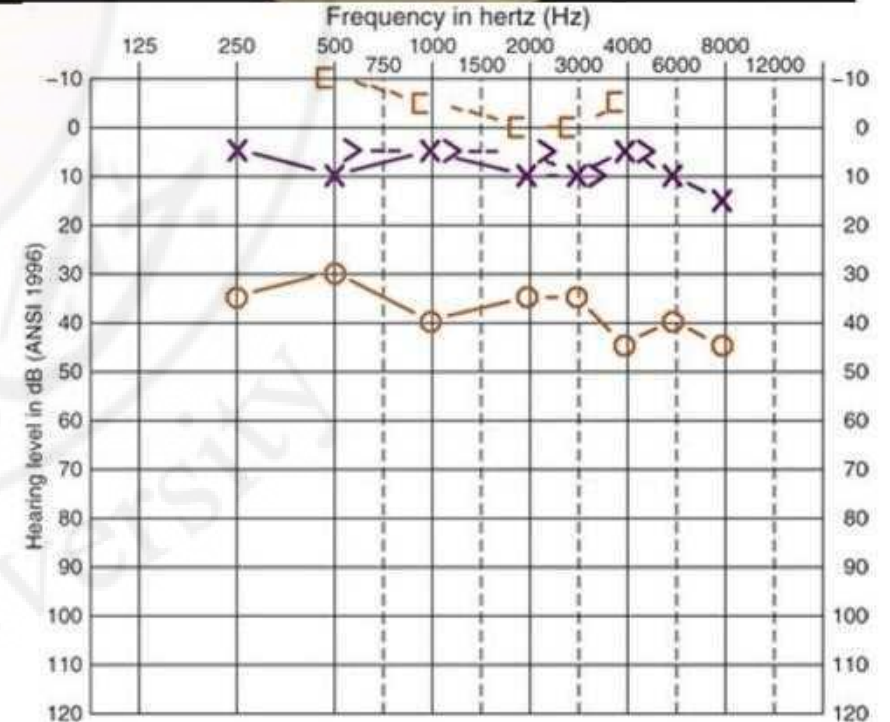
produced a hearing loss that was linearly proportional to the size of the perforation (loss of areal ratio plus loss of catenary lever). The degree of hearing loss, flat across speech frequencies, was not altered by the location of the perforation on the drumhead.



Case Example

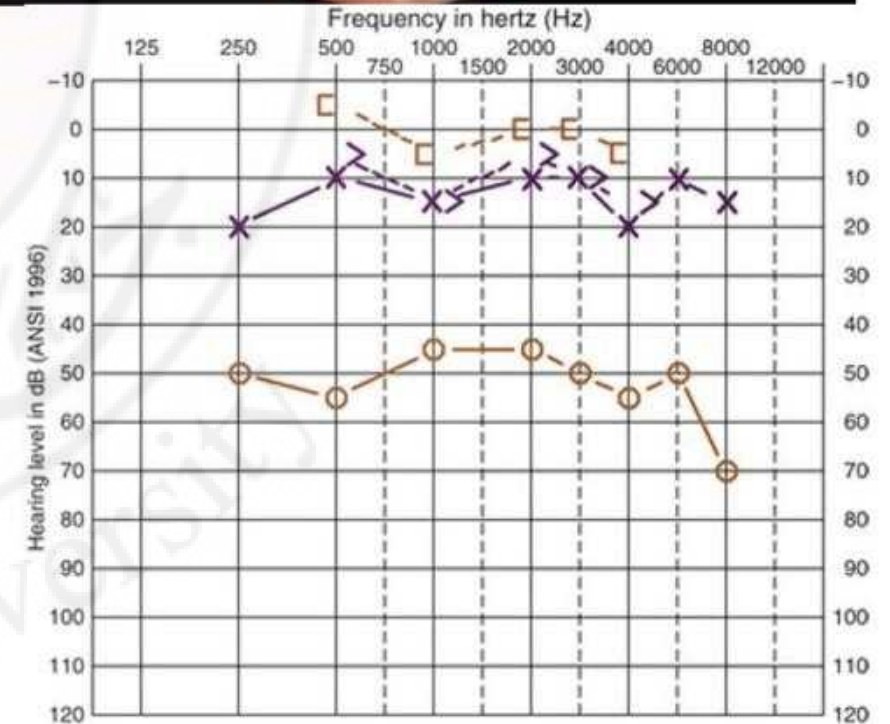
TM perforation combined with ossicular disruption

requires surgical therapy. An audiogram may falsely show depressed bone-conducted hearing levels of up to 10 dB in speech frequencies. Correction of the ossicular problem typically increased bone levels, reflecting the reestablishment of impedance matching at the oval window. Perforation size and the condition of the stapes footplate accounted for variations in this patient group.



Case Example

TM perforation combined with Incudostapedial joint erosion
requires surgical therapy.



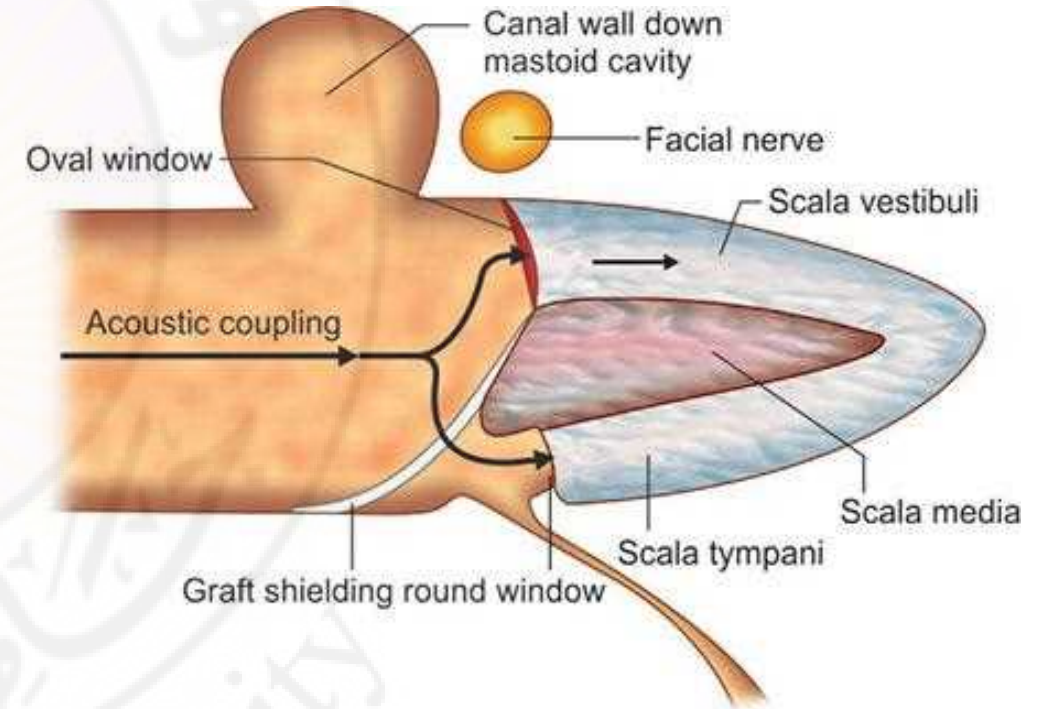
Case Example

Total loss of the TM and ossicles

Will create a condition wherein sound pressure contacted the oval and round windows simultaneously, resulting in partial phase cancelation of the sound wave in the cochlear perilymph.

Conductive hearing loss would be flat across speech frequencies and averaged **50 dB** when corrected for misleading bone levels.

More complete phase cancelation caused increased hearing loss as compared with patients who had only partial perforations.

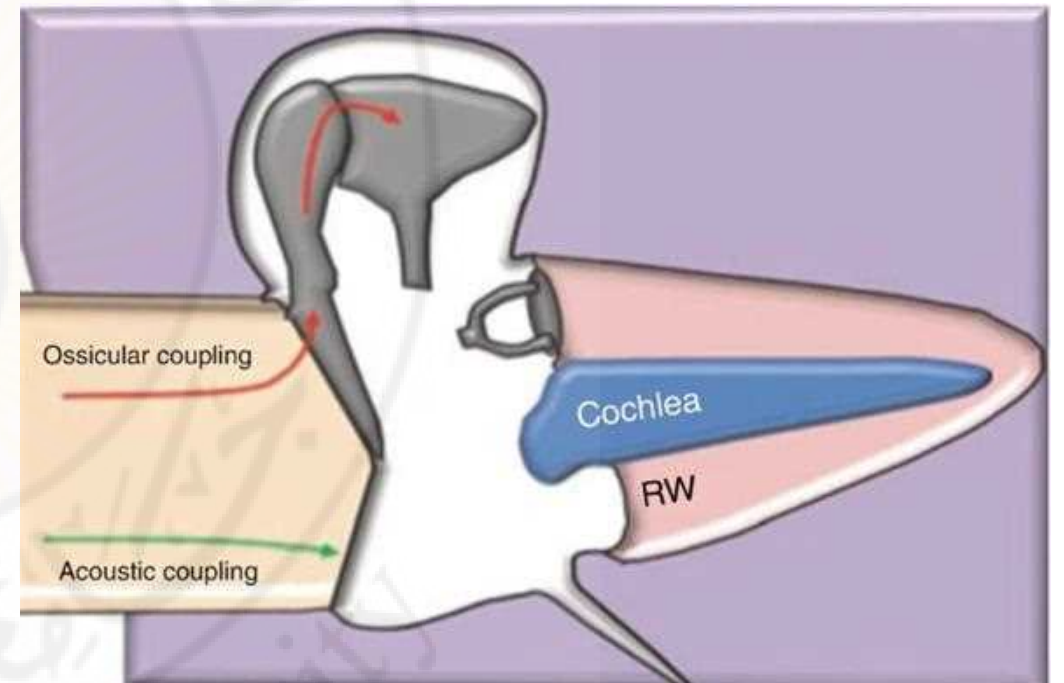


Case Example

Ossicular disruption behind an intact TM

resulted in a maximal conductive hearing loss of **55 to 60 dB**. In the absence of functional impedance matching, the intact eardrum reflected sound energy back into the EAC, which caused an additional 17-dB conductive loss above what was expected from removal of the hydraulic and catenary/ossicular lever action.

The decreased sound pressure also reached the round and oval windows almost simultaneously and induced phase cancelation in the labyrinthine fluids.

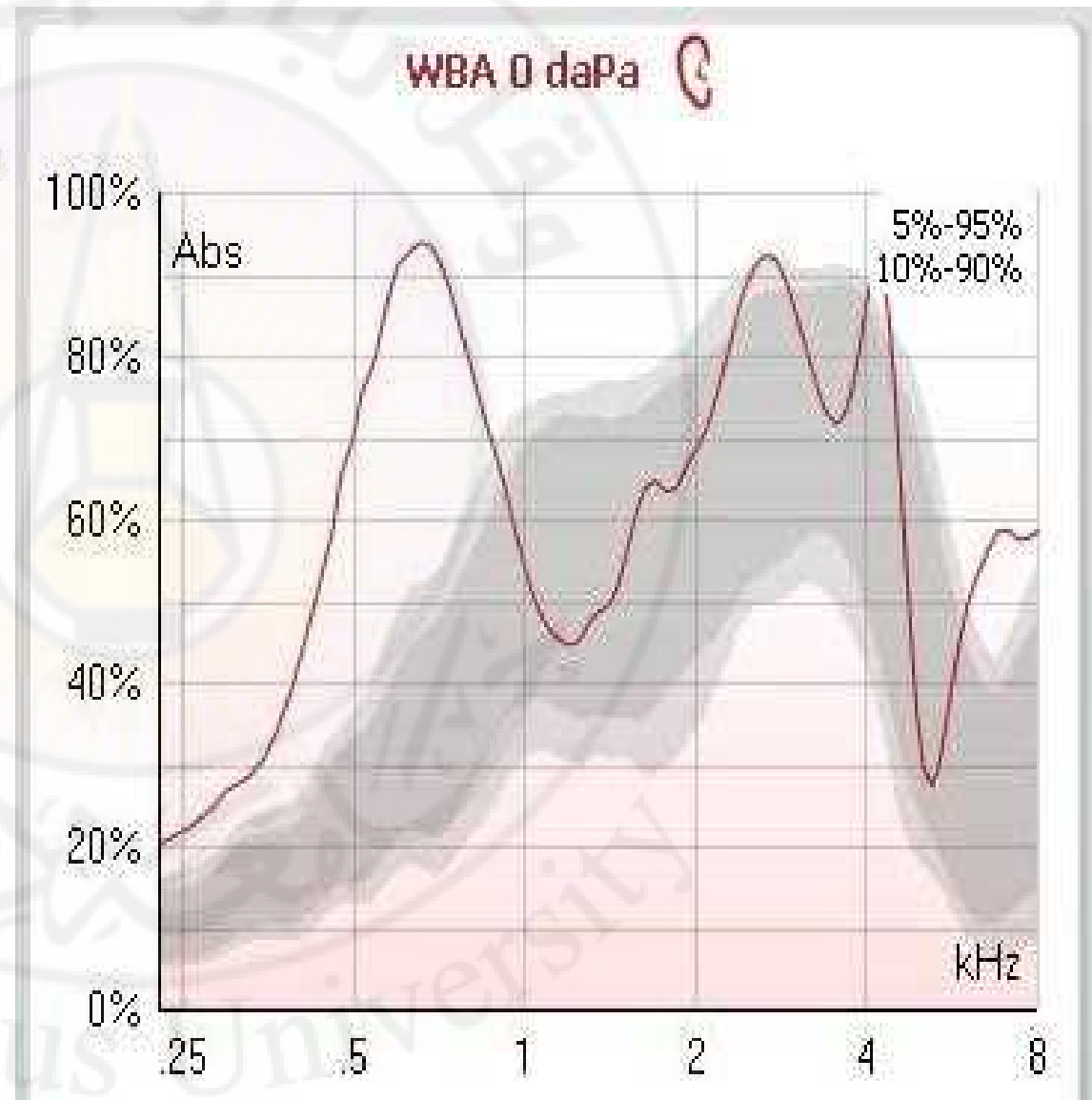
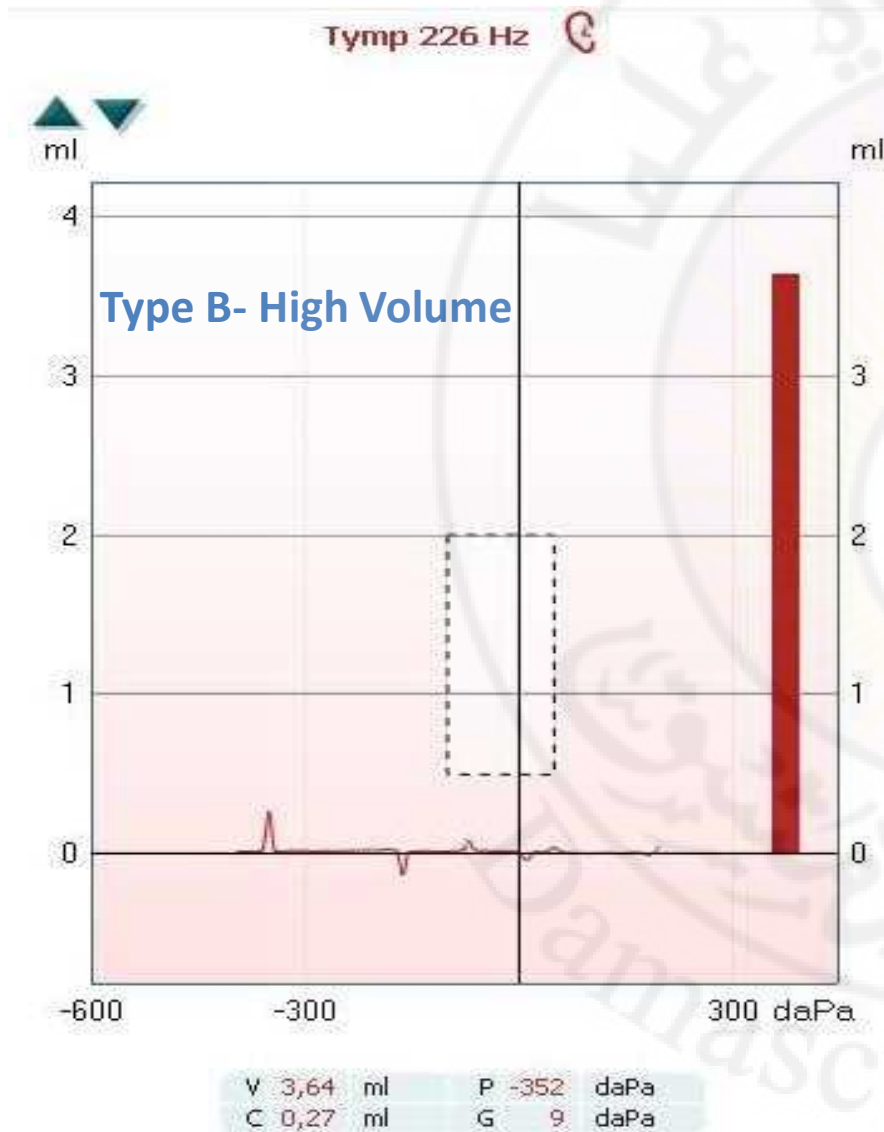


Specific Lesions of the Conductive Apparatus and the Associated Hearing Loss

Classification	Component Disrupted	Expected Loss
Perforation of tympanic membrane	Loss of areal ratio, catenary lever	Proportional to size of perforation
Perforation of tympanic membrane with ossicular interruption	Hydraulic lever, areal ratio, catenary lever	38.3 dB
Total loss of tympanic membrane and ossicular chain	Hydraulic lever, areal ratio, catenary lever, phase cancelation	50 dB
Ossicular interruption with an intact tympanic membrane	Hydraulic lever, areal ratio, catenary lever, phase cancelation, reflection of sound energy away from the middle ear at the tympanic membrane	55–60 dB
Ossicular interruption with an intact tympanic membrane and closure of the oval window (distinct congenital malformation)	Hydraulic lever, areal ratio, catenary lever, phase cancelation, reflection of sound energy away from the middle ear at the tympanic membrane	55–60 dB

From Austin DF: Sound conduction of the diseased ear. *J Laryngol Otol* 92:367, 1978; and Austin DF: Acoustic mechanisms of middle ear sound transfer. *Otolaryngol Clin North Am* 27:641, 1994.

Case Example



Monomeric membranes & Myringoplasty

- Many tympanic membrane perforations heal spontaneously. Healed perforations are associated with the development of scar tissue and thin areas that do not have a fibrous middle layer, which are sometimes called **monomeric membranes**.
- Larger perforations and those associated with chronic middle ear infections generally do not heal on their own, and require a surgical repair of the eardrum called a **myringoplasty**

Graft tissue is taken from temporalis fascia



Graft patch in place over defect





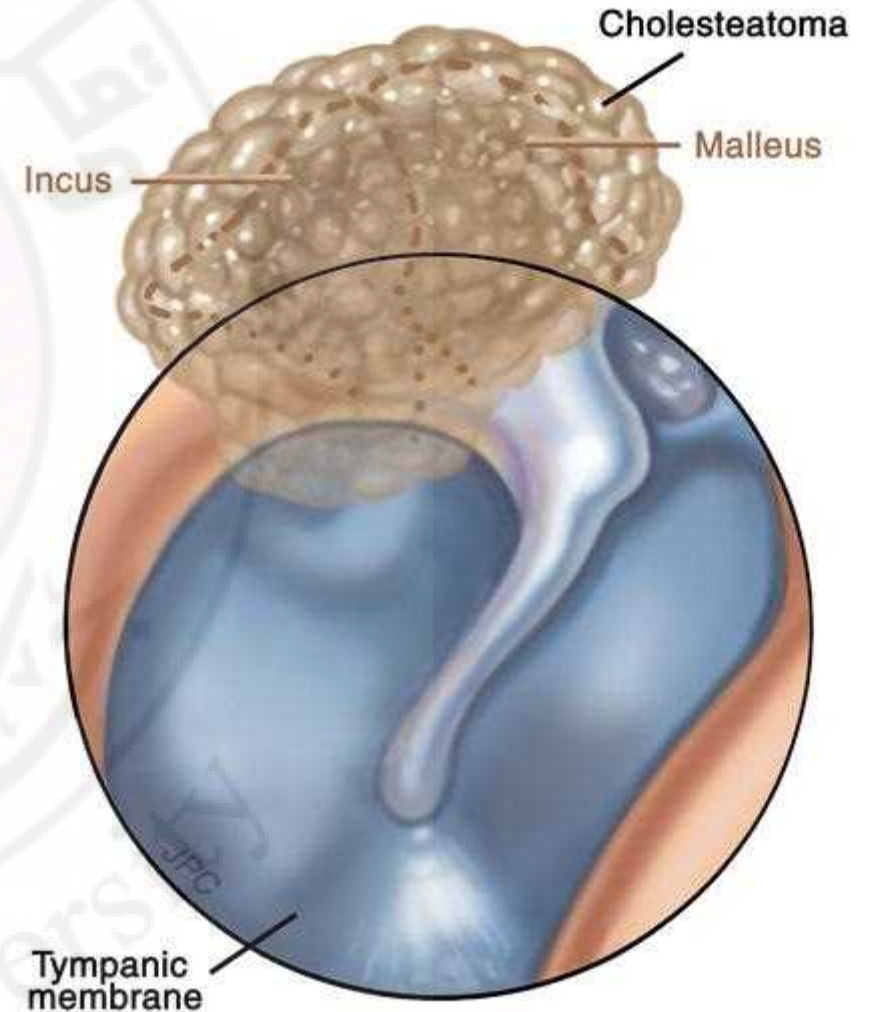
Clinical Audiology

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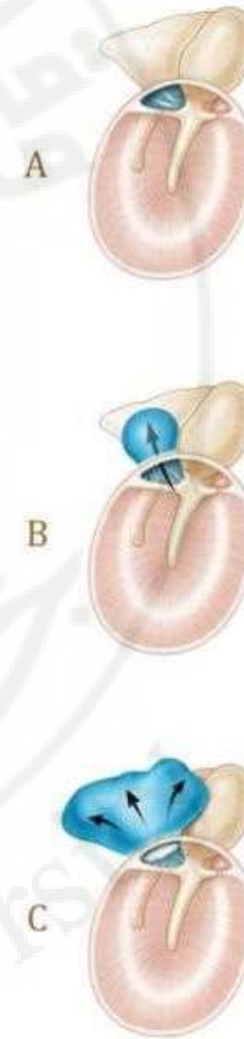
Cholesteatoma

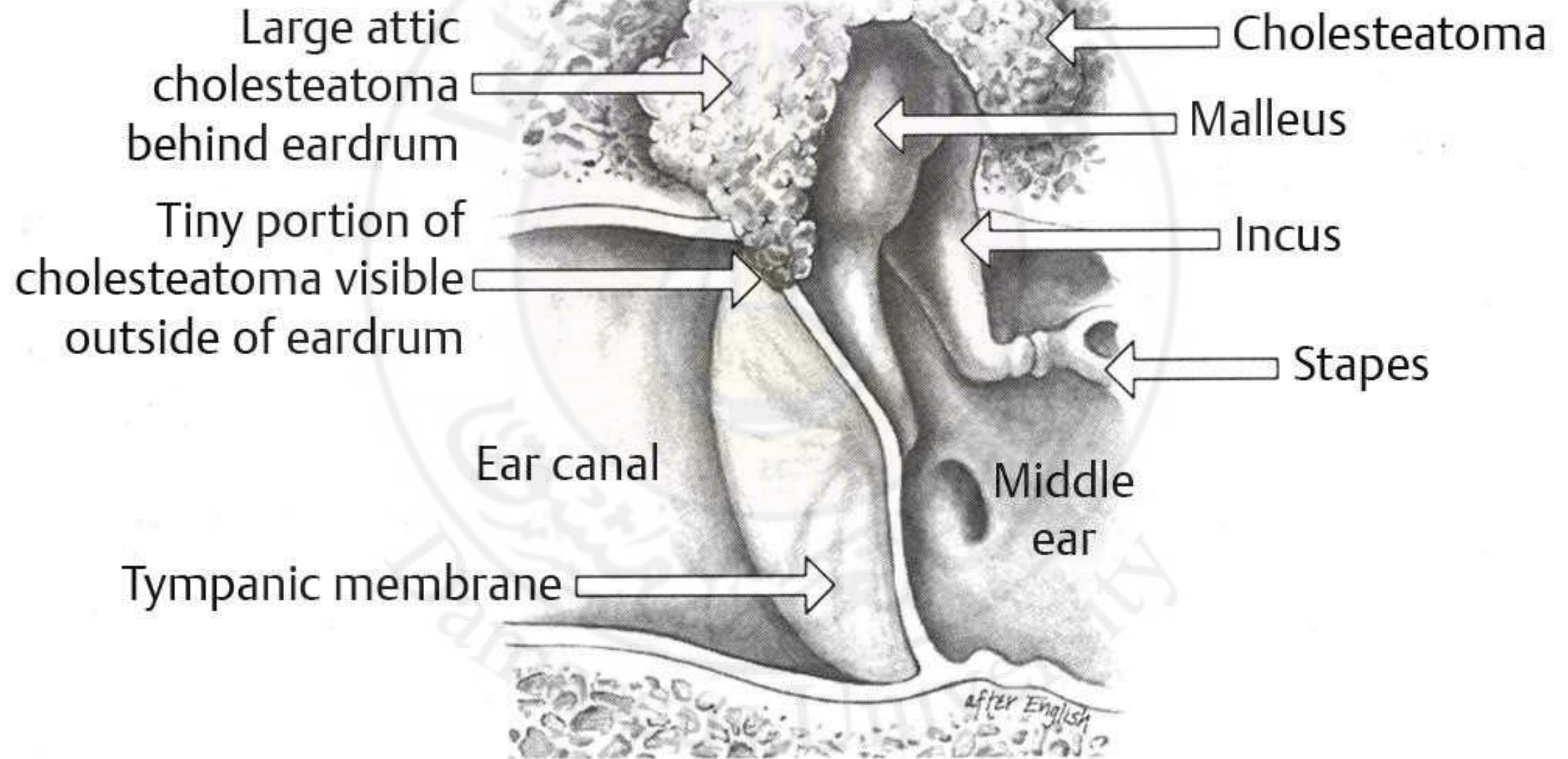
- A **cholesteatoma** is a cyst made of layers of keratin producing squamous epithelium filled with an accumulation of keratin and cellular debris.
- It is also- known as a **keratoma** or an **epidermoid inclusion cyst**.
- Even though keratoma is the less popular term, it is actually more descriptive because cholesteatomas contain considerable amounts of keratin but little if any cholesterol.
- Cholesteatomas of the temporal bone may be **congenital** or **acquired**.



Cholesteatoma

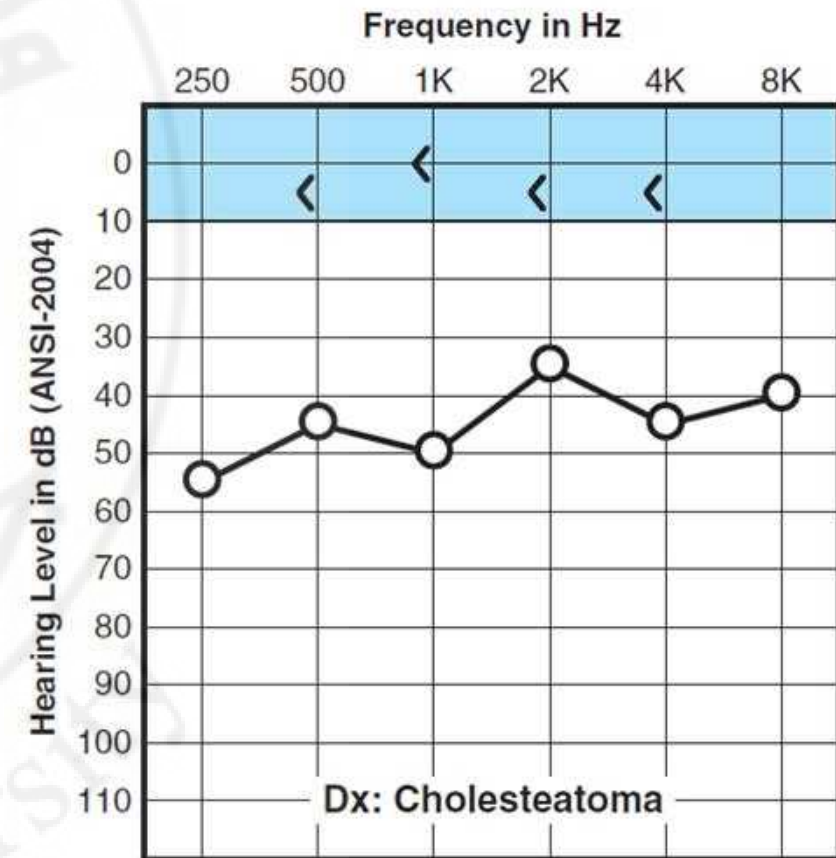
- Following chronic otitis media, a common pathologic occurrence is the formation of a cholesteatoma (“**pearly tumor**” of the temporal bone).
- A cholesteatoma is an epithelial pocket that forms, usually in the epitympanum (**attic cholesteatoma**). Typically, a weakened portion of the tympanic membrane becomes retracted into the middle-ear space. The outer layer of the membrane then sheds the cells naturally, and they become entrapped in the pocket formed by the retraction, resulting in growth of the cholesteatoma.
- As the cholesteatoma grows, it can begin to erode adjacent structures with which it has contact. The result can be substantial **erosion of the ossicles** and even **invasion of the bony labyrinth**.





Cholesteatoma

- Depending on the location of cholesteatoma growth, the magnitude of conductive hearing loss can vary from slight to total. Typically, the cholesteatoma will impede the ossicles, resulting in a significant **conductive hearing loss**. (Mixed or SNHL is also possible)
- Discharge** (purulent otorrhea) has usually been present for many years and usually has a very unpleasant smell. **Dizziness and true vertigo** may result if the lateral semicircular canal is involved.
- They erode the bone with which they come in contact and can produce serious **intracranial complications** if they erode through the dura of the middle or posterior fossa of the skull, through the lateral sinus, or into the lateral semicircular canal. They may also produce a **facial paralysis** if the facial nerve is eroded in the middle ear or mastoid.



Case Example

Sometimes members of the ossicular chain may be destroyed and yet the hearing remains fairly good with the cholesteatoma itself bridging the gap and helping in sound transmission.

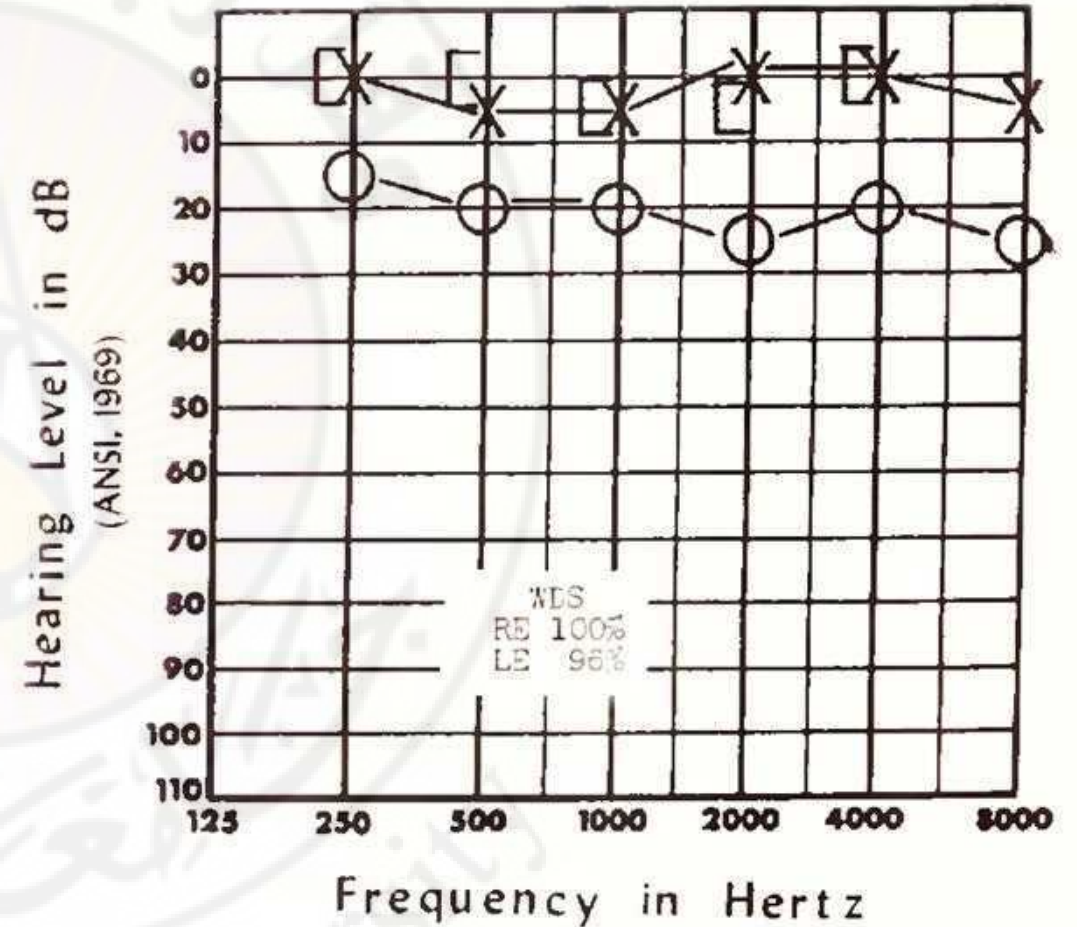


Fig. 2.8. Audiometric results of 34-year-old woman with large cholesteatoma, right ear.

Case Example

The otoscopic appearance of an aural cholesteatoma is variable. A typical **attic retraction cholesteatoma** appears as a defect of variable size adjacent to the posterosuperior portion of the tympanic membrane. The center of the defect contains keratin debris (*primary acquired cholesteatoma*).



- **Cholesteatoma of the attic**, left ear.
- Keratin debris fills a perforation of the pars flaccida superiorly, and a ventilation tube is in position on the mesotympanum inferiorly.



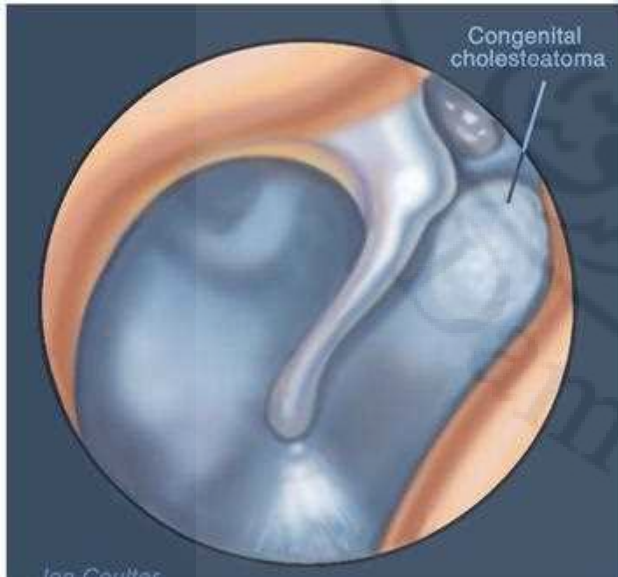
**Cholesteatoma developing at the
margin of perforation**

(secondary acquired cholesteatoma) with
secondary infection



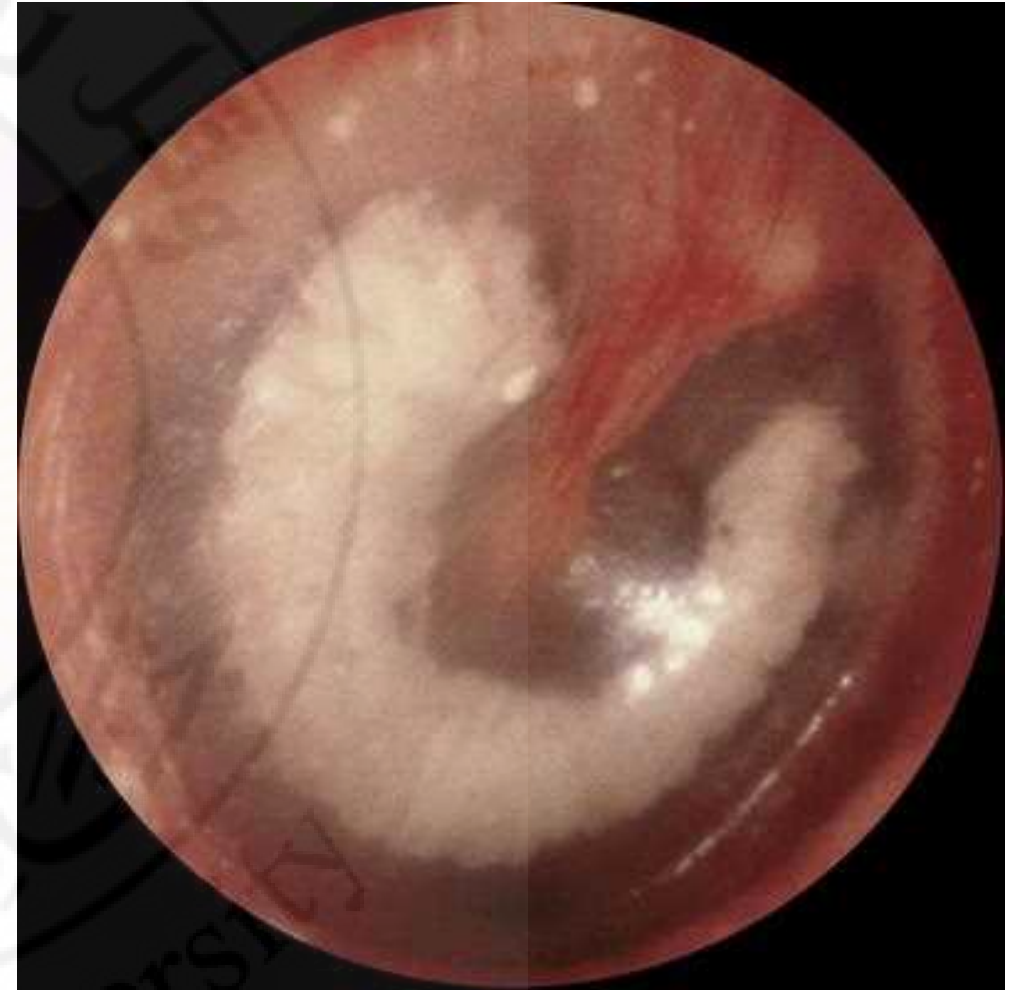
Case Example

Cholesteatomas sometimes appear behind or within an *intact* tympanic membrane, so-called **congenital cholesteatoma**.



Tympanosclerosis

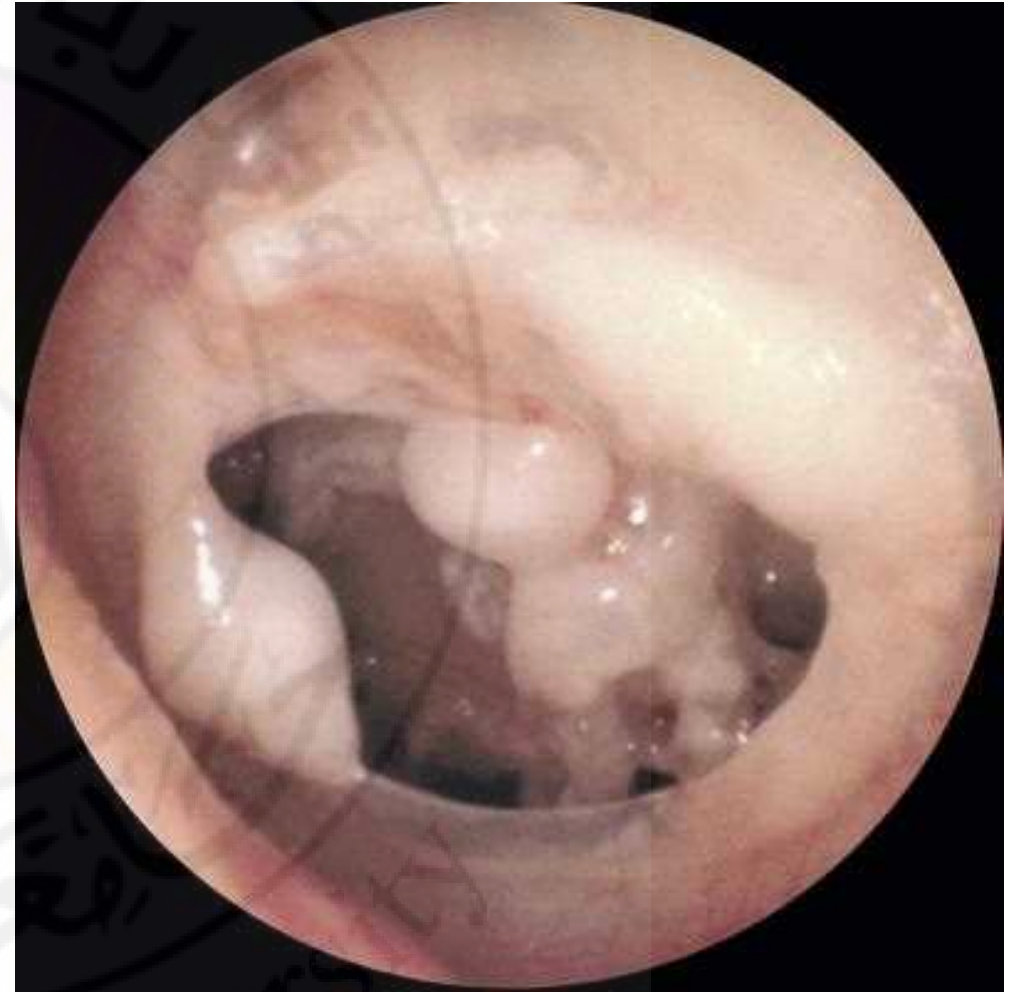
- Another consequence of chronic otitis media, is tympanosclerosis.
- Tympanosclerosis is the formation of whitish horseshoe-shaped plaques on the tympanic membrane, with nodular deposits in the mucosal lining of the middle ear. These plaques can cause an increased stiffening of the tympanic membrane and ossicles and even fixation of the ossicular chain in rare cases.
- Conductive hearing loss can occur, depending on the severity and location of the deposits.



Tympanic membrane tympanosclerosis
(myringosclerosis)

Case Example

Tympanosclerosis within the middle ear is histologically similar to that occurring within the tympanic membrane, but it often leads to conductive hearing loss caused by ossicular fixation.

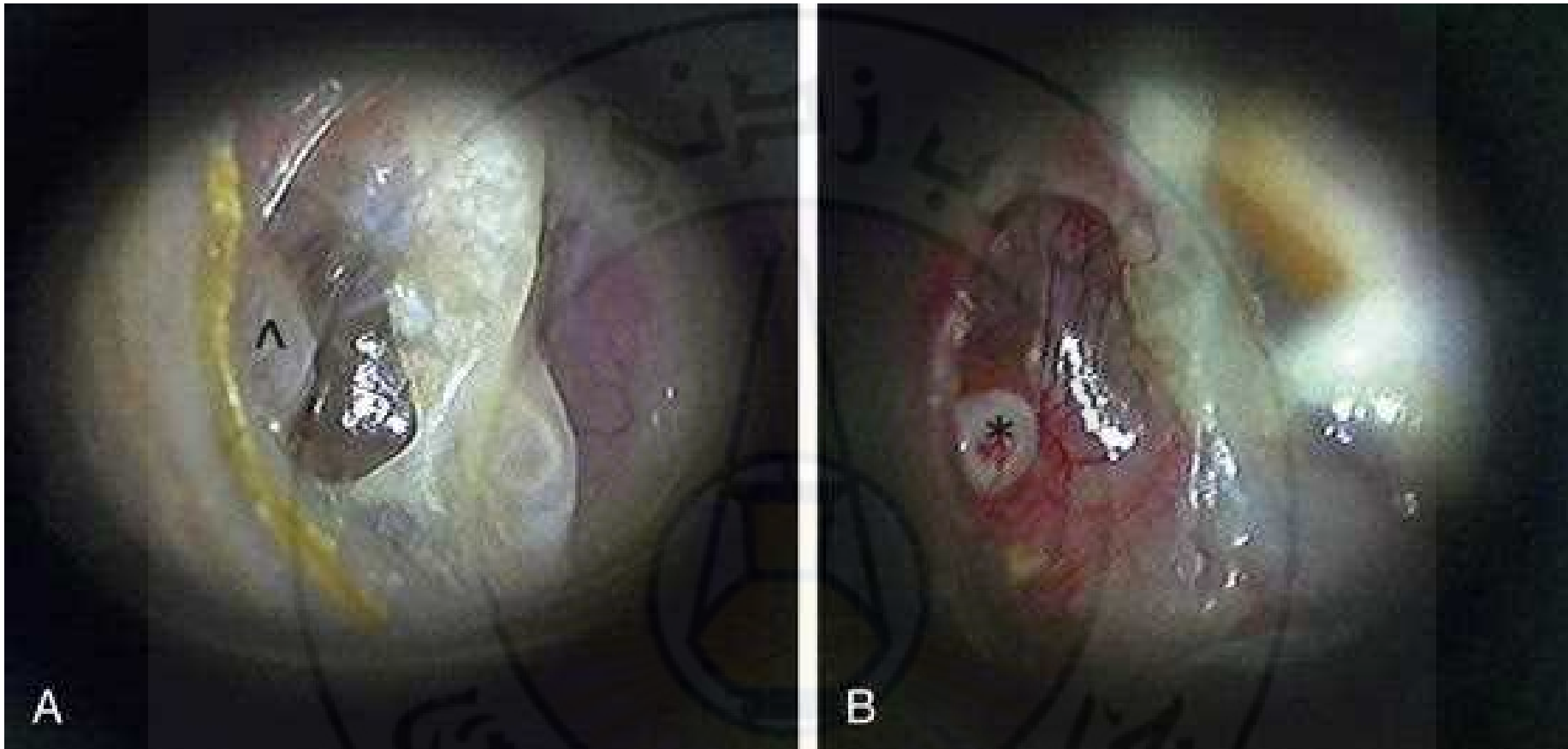


Advanced tympanosclerosis of the tympanic membrane and the middle ear. White avascular lesions attached to the promontory and the malleus.

Myringostapediopexy

- A perforation may be associated with an incus necrosis but the drum remnant may have attached itself to the stapes head producing a *spontaneous "tympanoplasty"* known as a myringostapediopexy.
- In this case, if the perforation is not very large and is not located directly over the round window, the hearing loss may be very slight or nonexistent.
- Myringostapediopexy, when it occurs **naturally**, can be stable and function well. When hearing is normal, no treatment is necessary. **Purposeful** creation of myringostapediopexy surgically has been very difficult to accomplish.



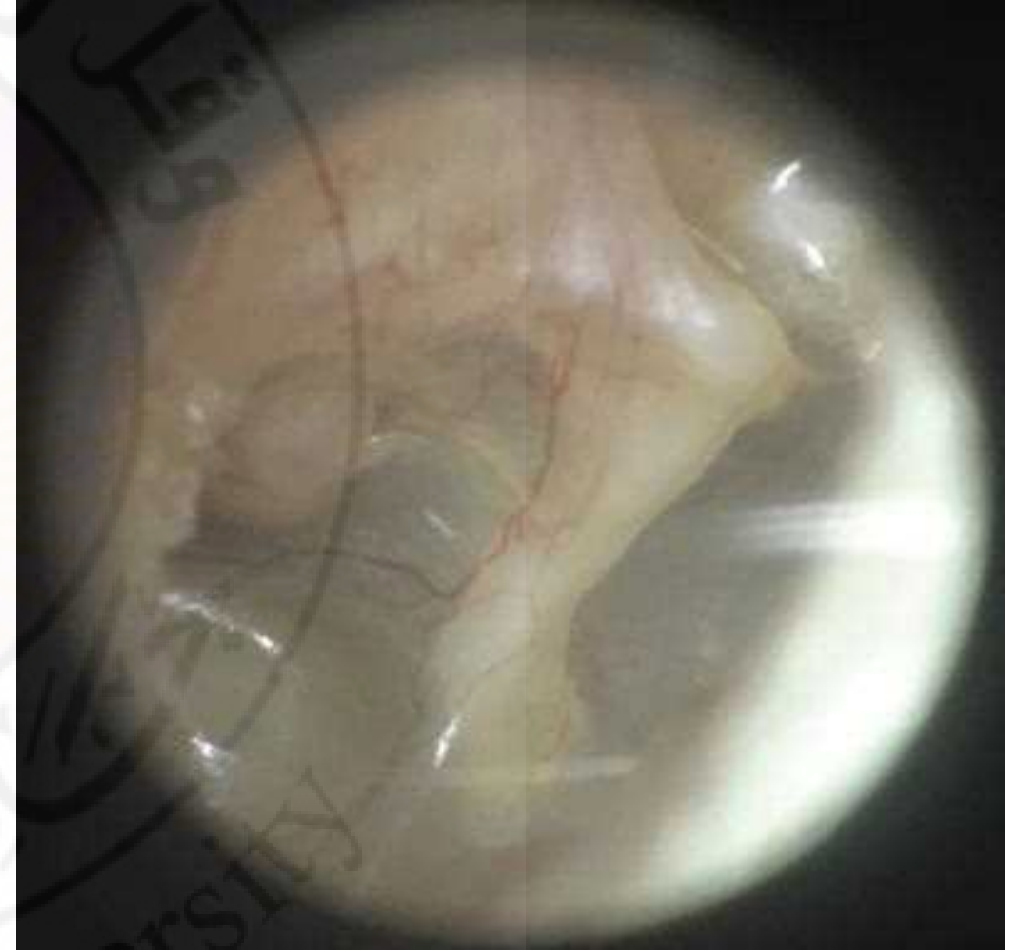


(A) Myringoincudopexy: The tympanic membrane is resting on the long process of the incus (^).

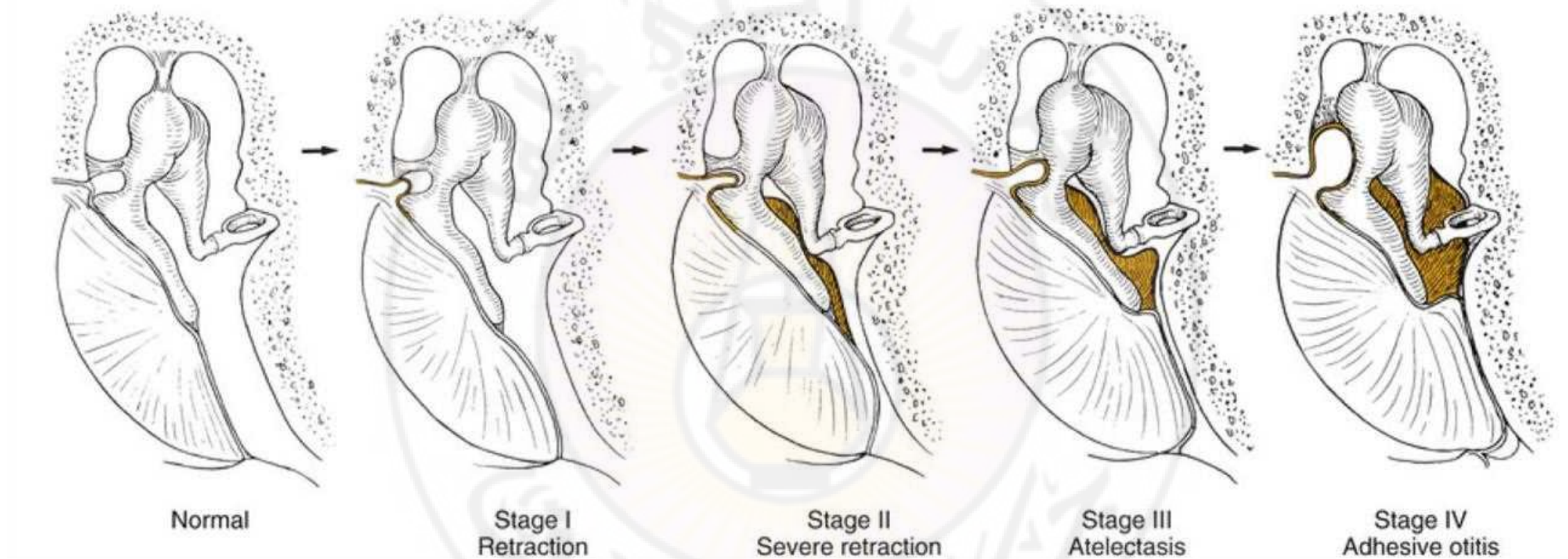
(B) Myringostapediopexy: The atelectatic tympanic membrane has eroded the long process of the incus, allowing the tympanic membrane to rest directly on the stapes superstructure

TM Atelectasis

- Severe retraction of the entire TM is referred to as **atelectasis**.
- The TM in this situation takes on a “cellophane” appearance and is not amenable to placement of a tympanostomy tube due to structural changes within the collagen layer of the TM.
- If atelectasis develops, the tympanic membrane becomes retracted onto the promontory and the ossicles of the middle ear.



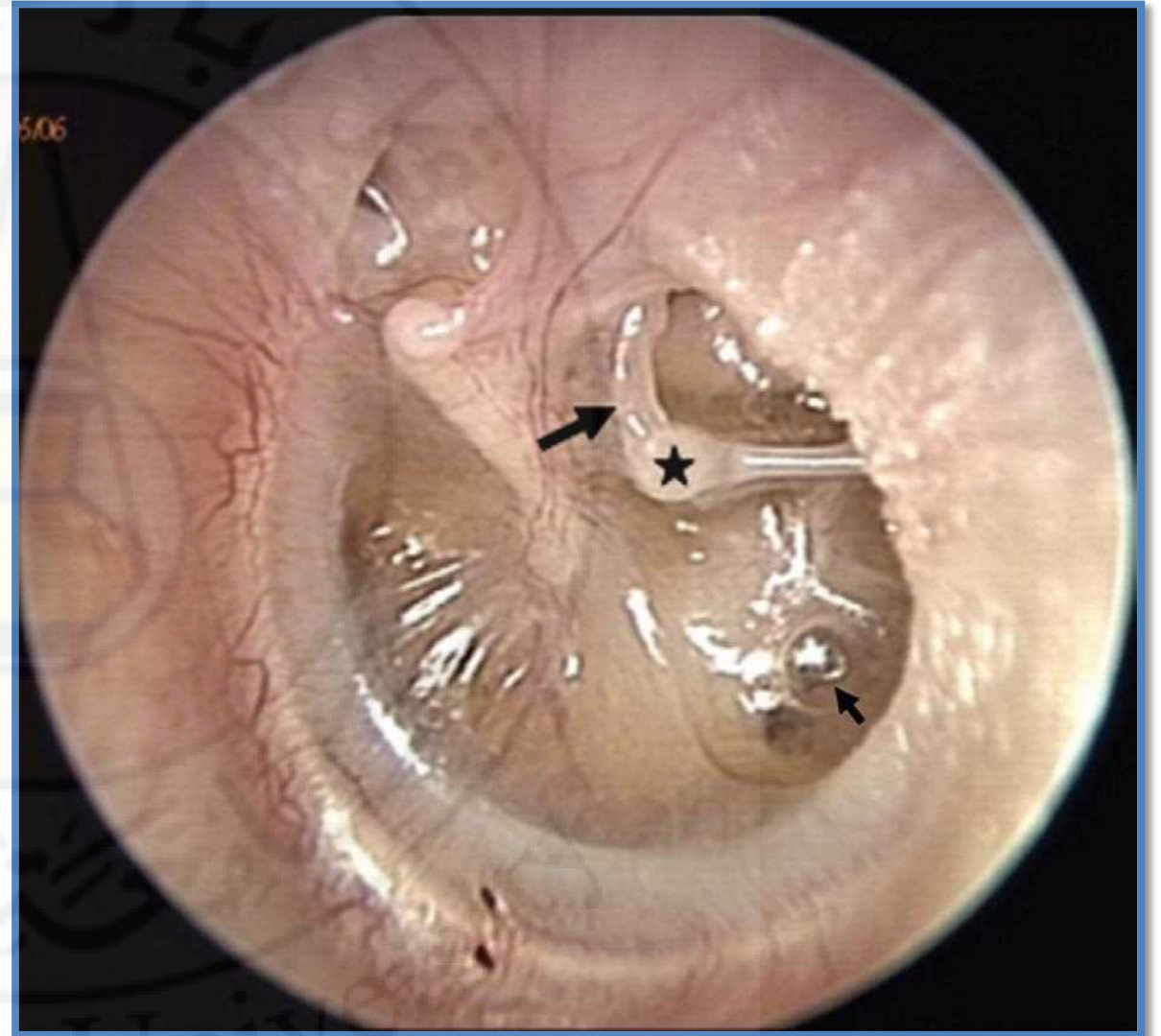
Atelectatic middle ear space. Note the tympanic membrane is retracted around the malleus handle and onto the promontory and the ossicular chain



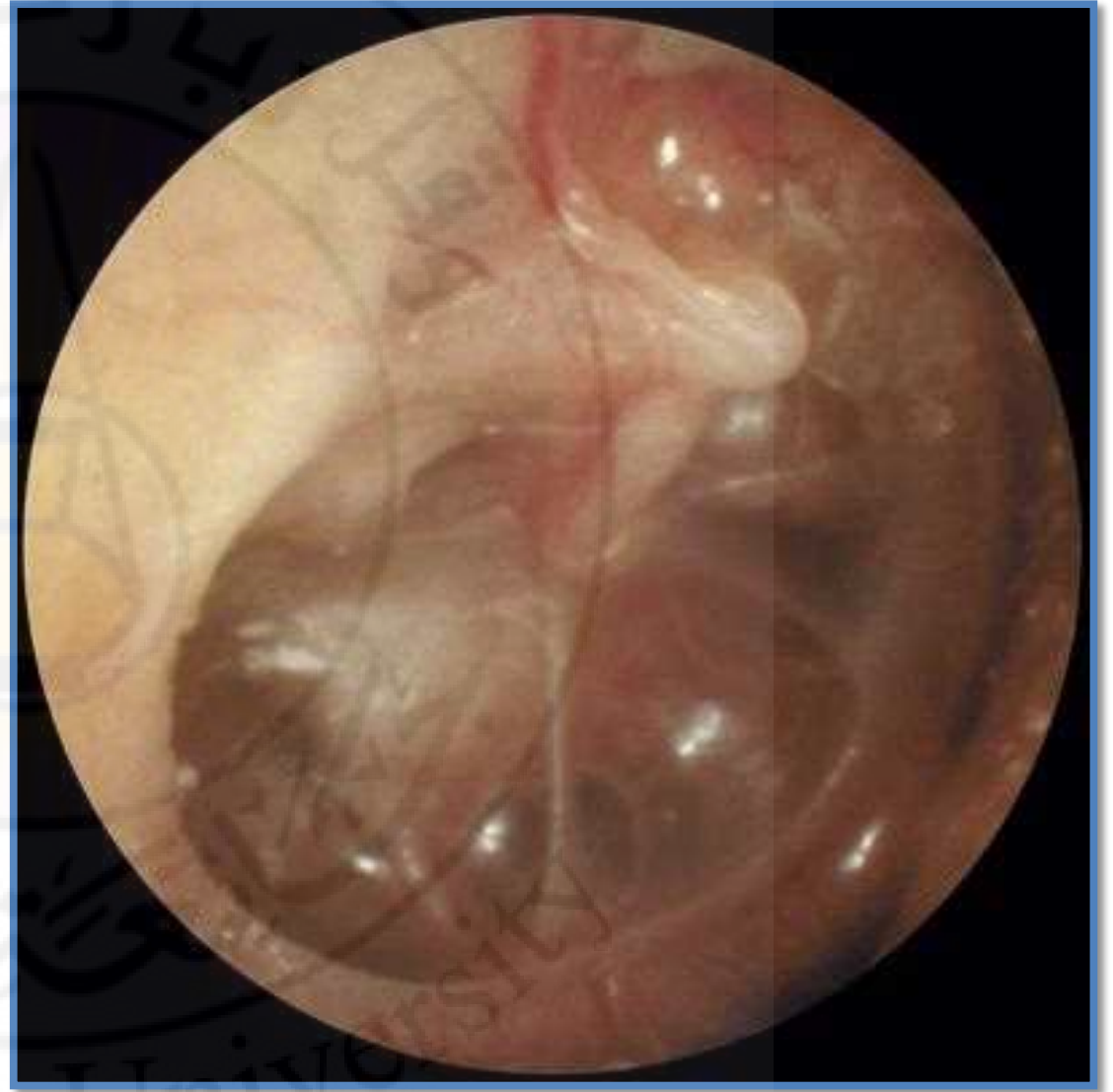
The four stages of middle ear atelectasis.

Middle ear atelectasis with effusion and ossicular chain disruption.

Otoscopic view shows the air bubble (small arrow) in the middle ear space. The eardrum is retracted onto the ossicles, resulting in erosion of the long process of the incus (large arrow). The stapes head (star) and the stapedial tendon are clearly visible.



Middle ear atelectasis with effusion

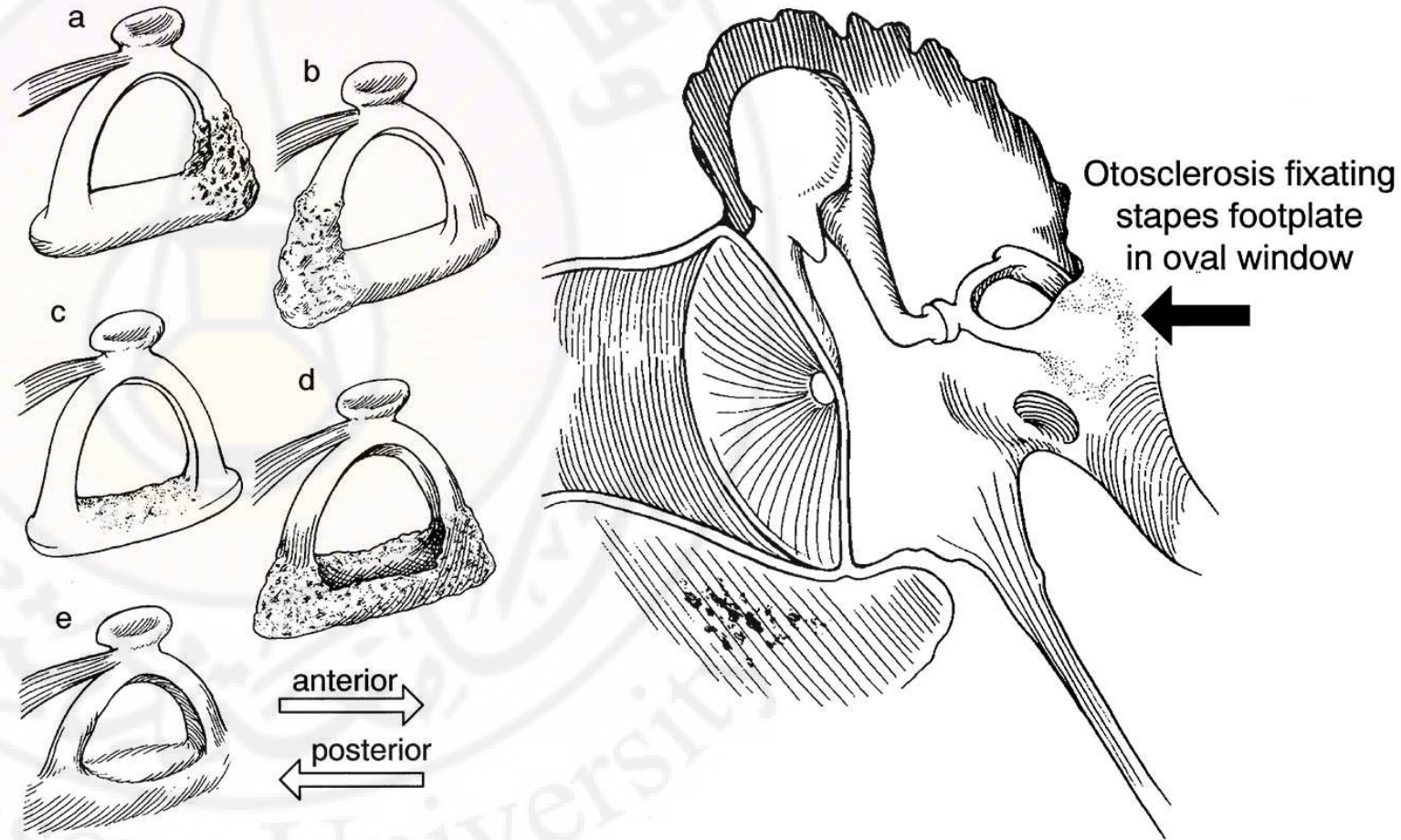


Otosclerosis

- Otosclerosis is a disorder of bone growth that affects the **stapes** and the **bony labyrinth** of the cochlea. The disease process is characterized by resorption of bone and new spongy formation around the stapes and oval window. Gradually the stapes becomes **fixed (ankylosis)** within the oval window, resulting in conductive hearing loss.
- Otosclerosis is a common cause of middle-ear disorder. A **family history** of the disease occurs in over half of the cases (transmitted in an autosomal dominant fashion with variable penetrance).
- **Women** are more likely to be diagnosed with otosclerosis than men (2:1), and its onset can be related to **pregnancy**.
- Otosclerosis usually occurs **bilaterally** (75% of cases), although the time course of its effect on hearing may differ between ears.
- Otosclerosis is the most common cause of **progressive** conductive hearing loss (CHL) in adults.



Otosclerotic involvement affecting various parts of the stapedial footplate (a–d) as well as complete obliteration of the oval window (e). Right panel: Otosclerosis fixating the stapes in the oval window.



Otosclerosis- History

- **Hearing loss** is typically of gradual onset and progresses slowly over several years
- 75% of otosclerosis cases are bilateral and present in individuals in their late teens or 20s.
- Hearing loss may not become apparent to the patient until age 30 or 40 years
- Most patients report difficulty hearing conversation while chewing and may hear better in noisy rooms due to a phenomenon known as **paracusis of Willis**.
- **Tinnitus** often accompanies otosclerosis and may be described as pulsatile (During the early or active phase) or continuous.
- Two-thirds of patients with otosclerosis have a **family history** of hearing loss
- Associated symptoms such as autophony and/or dizziness with loud sounds should also be elucidated in order to rule out other conditions such as a superior canal dehiscence.



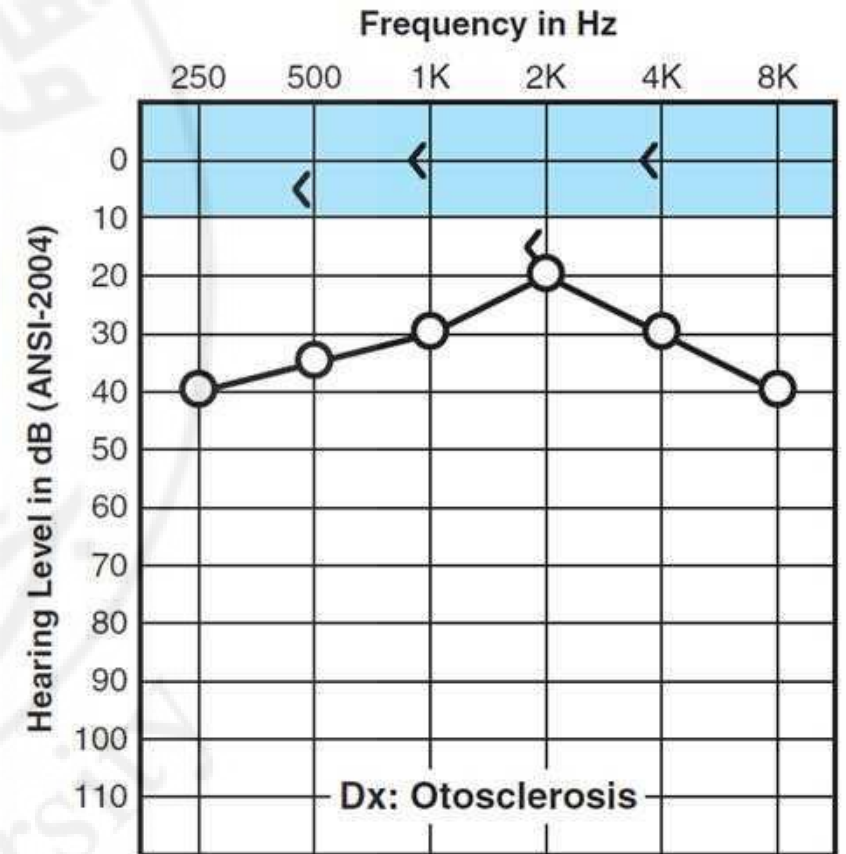
Otosclerosis- Physical exam

- In otosclerosis, the **Schwartz sign (Flemingo's flush sign)** refers to a characteristic reddish-blue discoloration of the promontory (a bony prominence) in the middle ear. This sign is seen in about 10% of cases during otoscopy (an examination of the ear using an otoscope), and indicates **otospongiosis** (active otosclerosis).
- The Schwartz sign is believed to result from increased vascularity (blood flow) in the area due to the formation of new blood vessels (neovascularization) in response to bone remodeling associated with otosclerosis.
- While the Schwartz sign can be helpful in diagnosing otosclerosis, it is not always present and can also be seen in other middle ear conditions. Therefore, additional diagnostic tests, such as audiometry and imaging studies, may be needed to confirm a diagnosis of otosclerosis.



Otosclerosis- Audiometric Patterns

- As with other middle ear disorders, otosclerosis reduces sound-related energy passing from the tympanic membrane to the inner ear. Fixation and resultant stiffening of the ossicular chain almost always produces a hearing loss, particularly for **lower frequency sounds**.
- The degree of the conductive hearing loss varies with the progression of fixation, but the configuration of the bone-conduction thresholds is almost a **signature** of the disease.
- Note that the **bone-conduction threshold dips slightly at 2000 Hz**, reflecting the elimination of factors that normally contribute to bone-conducted hearing due to the fixation of the stapes into the wall of the cochlea. This **2000 Hz notch** is often referred to as **“Carhart’s notch,”** named after Raymond Carhart who first described this characteristic pattern.

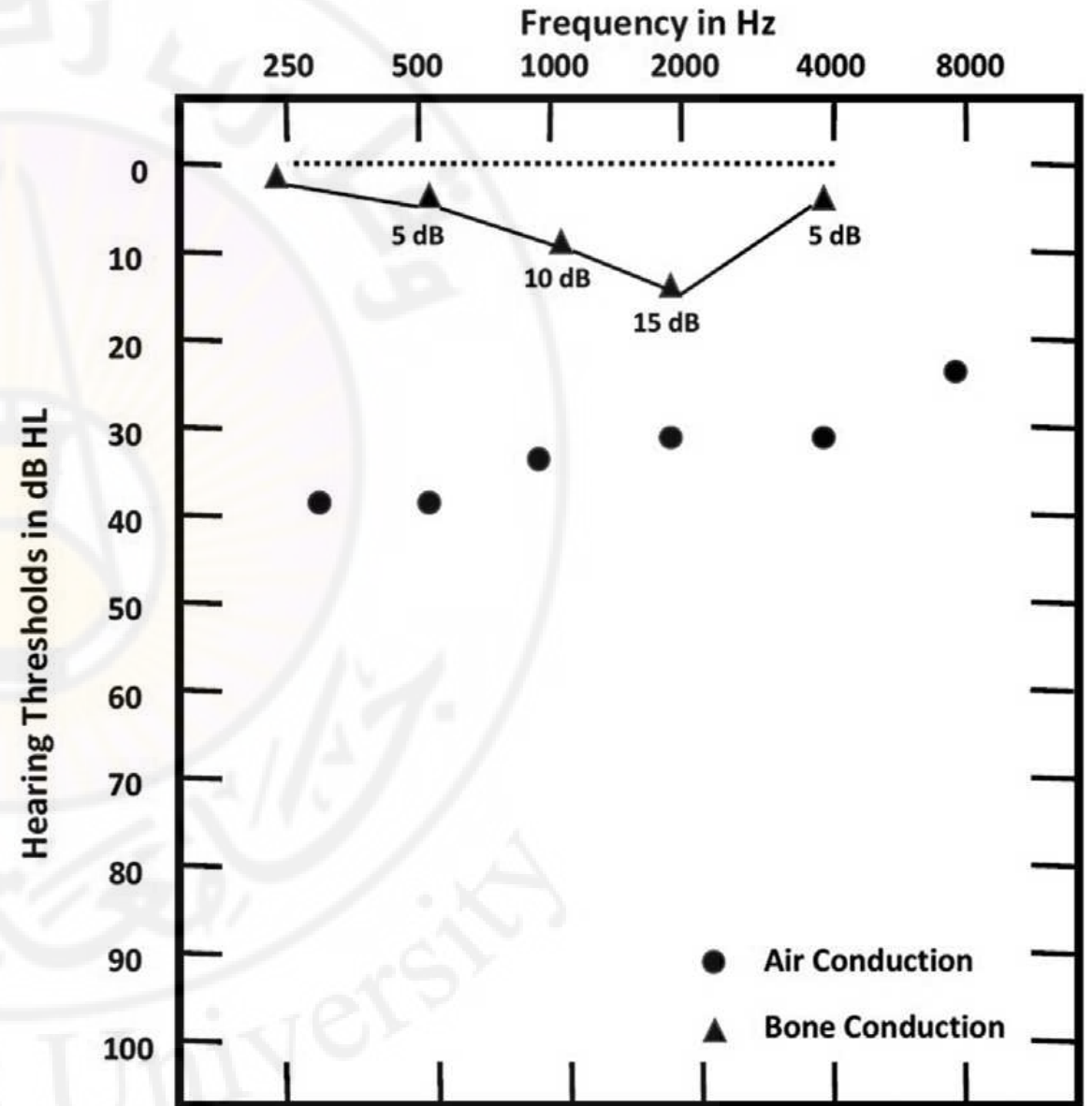


Patterns of basic auditory findings in patients with the diagnosis of otosclerosis

Procedure	Findings
Pure tone audiometry	
Air conduction	Hearing loss greater for low frequencies.
Bone conduction	Apparent decrease in bone conduction thresholds sometimes with a notching deficit at 2000 Hz (Carhart notch). Actual bone conduction hearing is typically normal.
Audiometric Weber test	Perception of low-frequency pure tone stimuli in the ear with conductive hearing loss.
Sensorineural acuity level test	Presence of an air-bone gap and confirmation of normal bone conduction hearing.
Acoustic immittance measures	
Tympanometry	Shallow type A tympanogram reflecting increased stiffness of the ossicular chain (sometimes Type As or type An tympanograms)
Acoustic reflexes	Absence of stapedial acoustic reflex activity even in patients with minimal air-bone gap and conductive hearing loss. Atypical acoustic reflex pattern in patients with very early subclinical otosclerosis.
Otoacoustic emissions	Otoacoustic emissions cannot be detected in patients with otosclerosis and conductive hearing loss. Recovery of detectable otoacoustic emissions is possible in patients following microtraumatic stapedotomy.

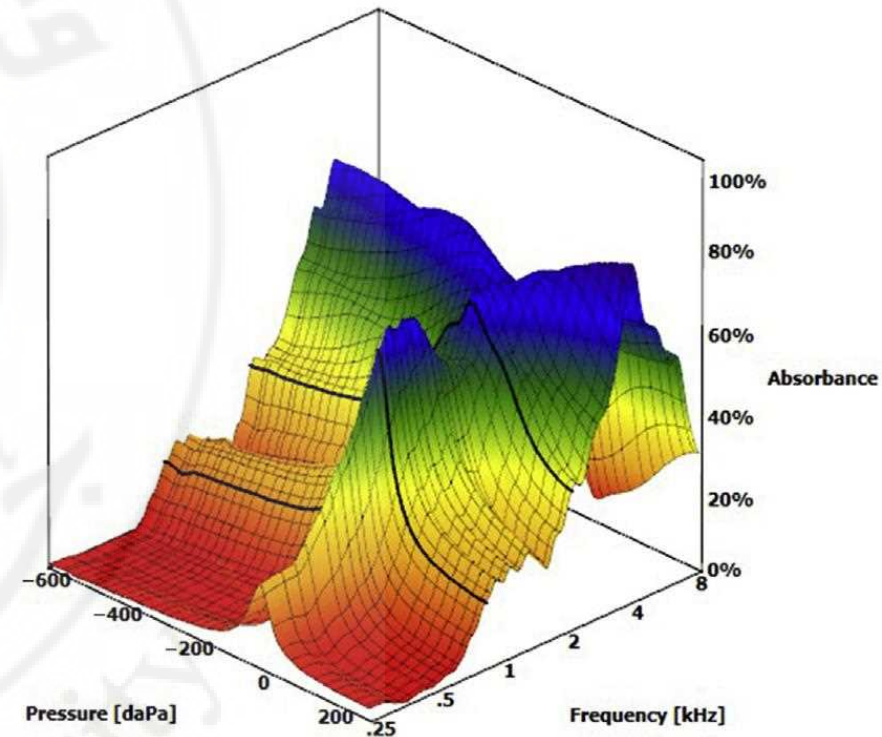
Otosclerosis- Audiometric Patterns

- Typical air and bone conduction hearing threshold patterns for a patient with otosclerosis.
- Notice the appearance of **Carhart notch** in bone conduction hearing at 2000 Hz.
- The dotted line indicates true bone conduction hearing or "**cochlear reserve.**"
- Some patients show Mixed HL.



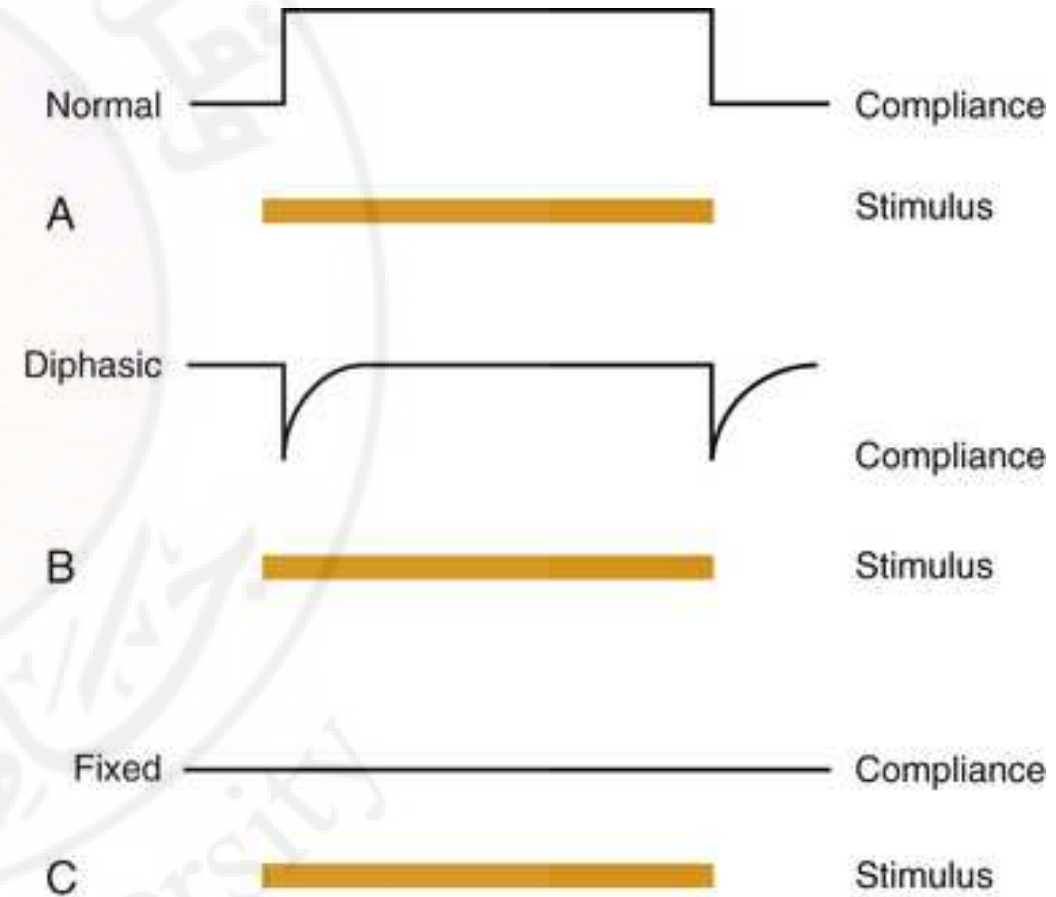
Otosclerosis- Wideband Acoustic Immittance (WAI)

- In contrast to conventional 226-Hz tympanometry, WAI is significantly more sensitive to ossicular pathologies.
- Generally, a stiffening pathology results in decreased absorbance over a specific frequency range. For example, otosclerotic ears demonstrate significantly increased reflectance between 400 Hz and 1000 Hz.
- Researchers found that **Power absorbance (PA)** was the most effective way of identifying ears with otosclerosis compared with 226-Hz tympanometry and multifrequency tympanometry. PA was able to identify otosclerosis in 82% of their sample and had a false-positive rate of 17.2%.



Otosclerosis- Acoustic reflexes

- Acoustic reflexes are a sensitive measure of the movement of the stapes. In the presence of otosclerosis, the reflex is absent.
- With **early stapes fixation**, the reflex may be abnormal, demonstrating a negative on-off effect, or diphasic reflex.
- With **more advanced disease**, the reflex is absent when the probe is in the involved ear. As the disease and hearing loss worsen, the contralateral reflex is affected as a result of the degree of CHL in the otosclerotic ear.



Case Example

- The classical example shows a **conductive impairment** with a greater loss in the low than in the high frequencies and follows the impedance formula for stiffness.
- The less typical case is a patient with a **mixed loss** and a greater or lesser degree of sensory-neural involvement.
- Otosclerosis also may cause a **purely sensory-neural loss (Cochlear otosclerosis)**. In these cases the conduction mechanism is not involved but the sensory-neural elements are directly or indirectly affected. The diagnosis of sensory-neural loss due to otosclerosis can only be made after special x-ray studies of the temporal bone are done.

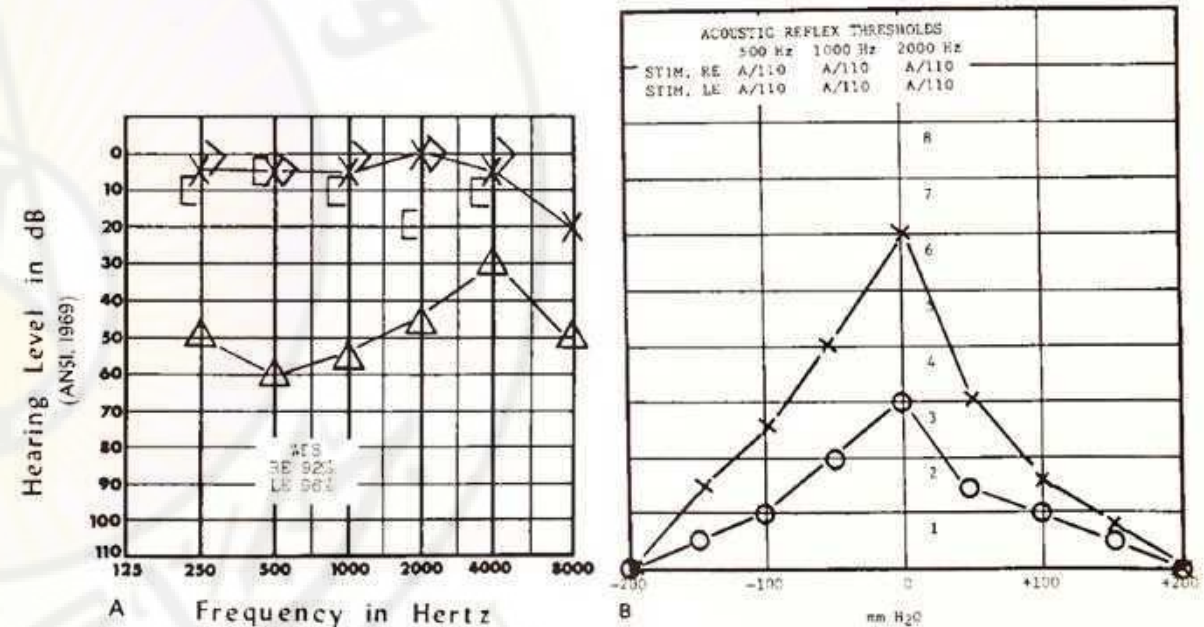
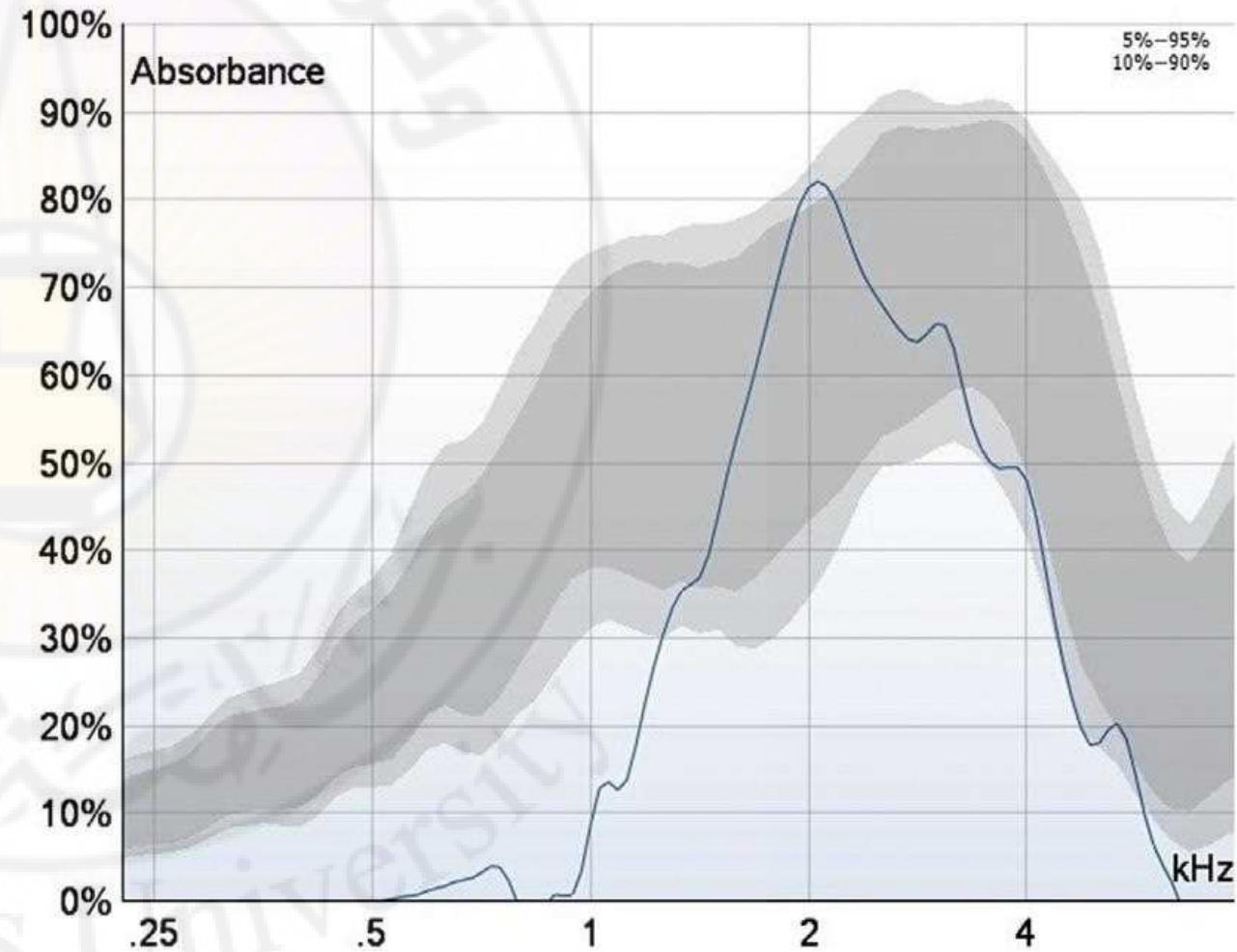


Fig. 2.10. (A) Results of audiometric studies performed on 46-year-old woman with surgically confirmed otosclerosis, right ear. (B) Tympanometric results of patient described in A.

Case Example

Power absorbance in a surgically confirmed otosclerotic ear. Note that the absorption less than 1500 Hz is significantly reduced compared with the 90% range for normal individual (shaded area). The y-axis is absorbance in% and the x-axis is the frequency in Hz. (Courtesy of Navid Shahnaz, PhD, University of British Columbia, Vancouver, BC, Canada.)



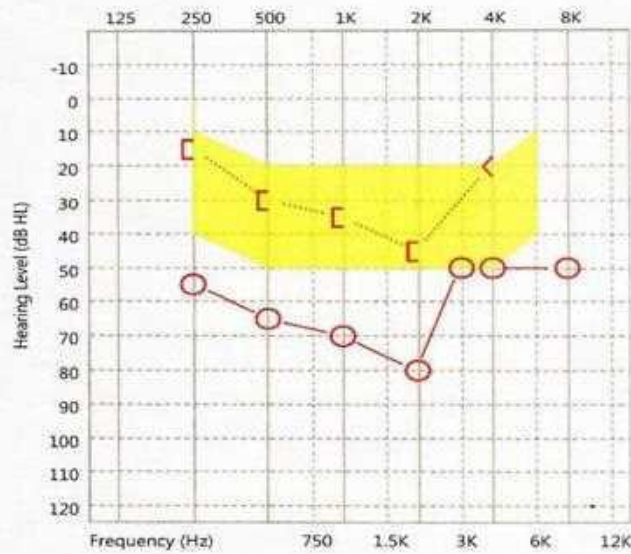
Case Example

Person ID
First name
Last name
Birth date
Gender Female



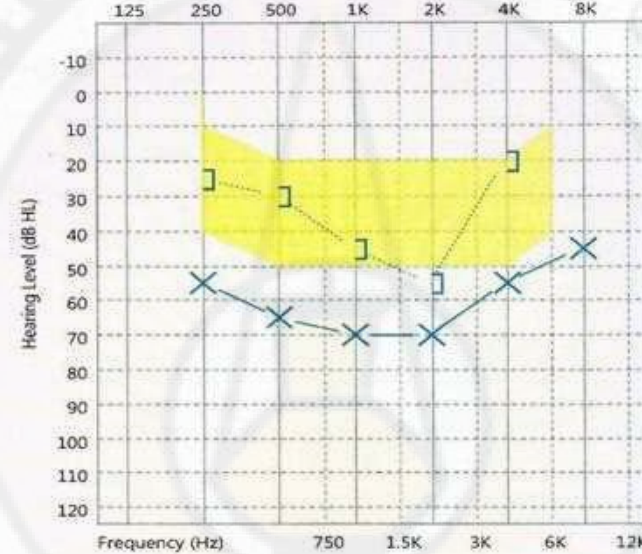
Tehran Ear Clinic (Audiology Unit)
Day Medical Tower
Unit C, 9th Floor
Tehran
021-88205806

Right Ear Audiogram

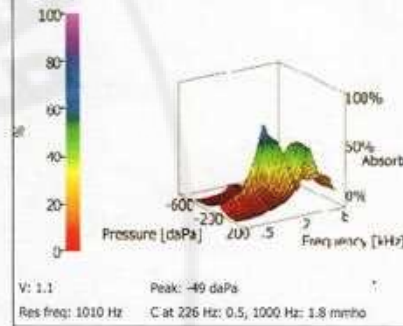


Symbol	Frequency	Intensity
○	250	50
○	500	60
○	1000	70
○	2000	80
○	4000	50
○	8000	50

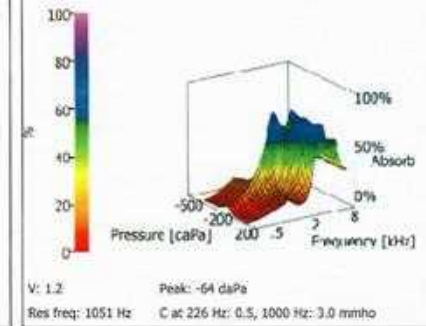
Left Ear Audiogram



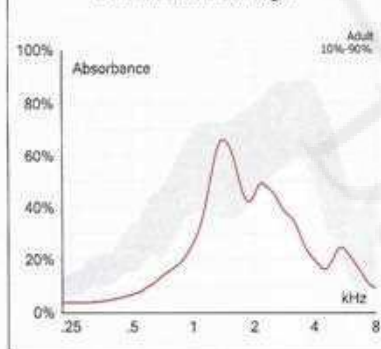
3DT - Right



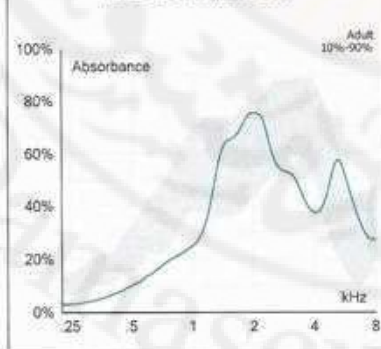
3DT - Left



WB absorbance Right



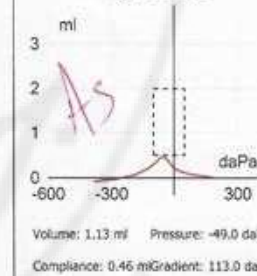
WB absorbance Left



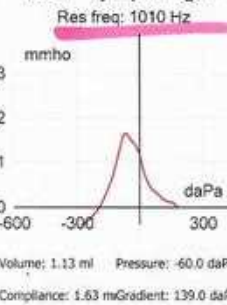
Reflex Right	F:226 Hz	P:-47 daPa
500 Hz I	0.00	0.00
1 kHz I	0.00	0.00
2 kHz I	0.00	0.00
4 kHz I	0.00	0.00

Reflex Left	F:226 Hz	P:-67 daPa
500 Hz I	0.00	0.00
1 kHz I	0.00	0.00
2 kHz I	0.00	0.00
4 kHz I	0.00	0.00

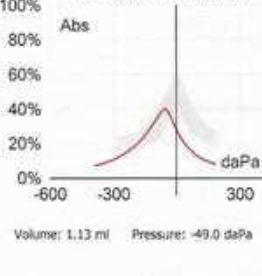
Tymp 226 Hz



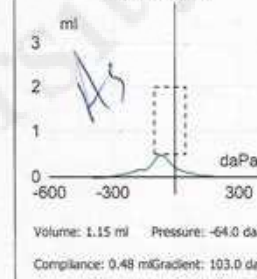
3DT - tymps Right



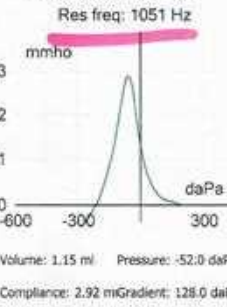
WB tymp (>6 months)



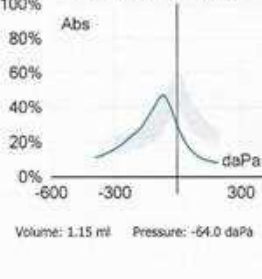
Tymp 226 Hz



3DT - tymps Left

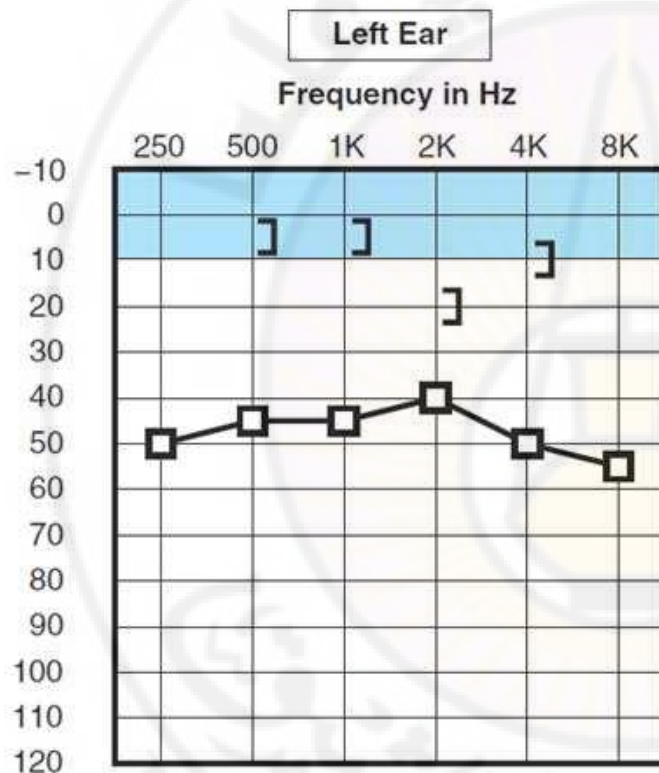
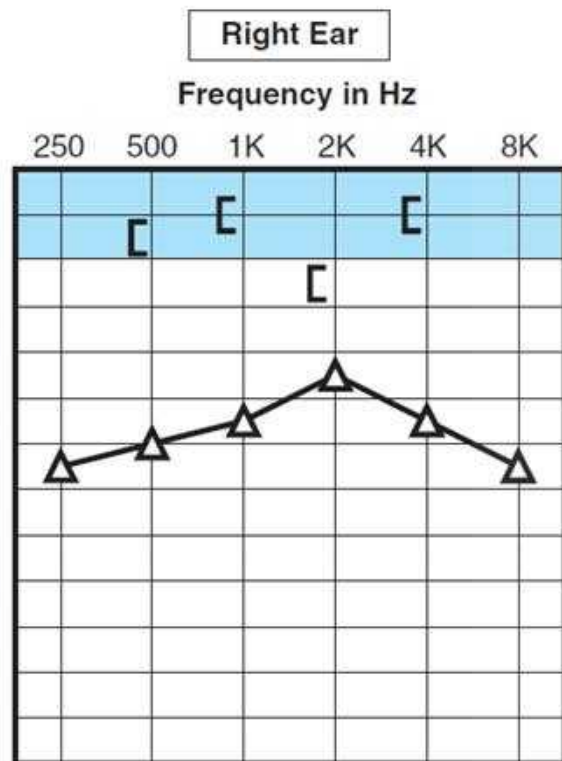


WB tymp (>6 months)



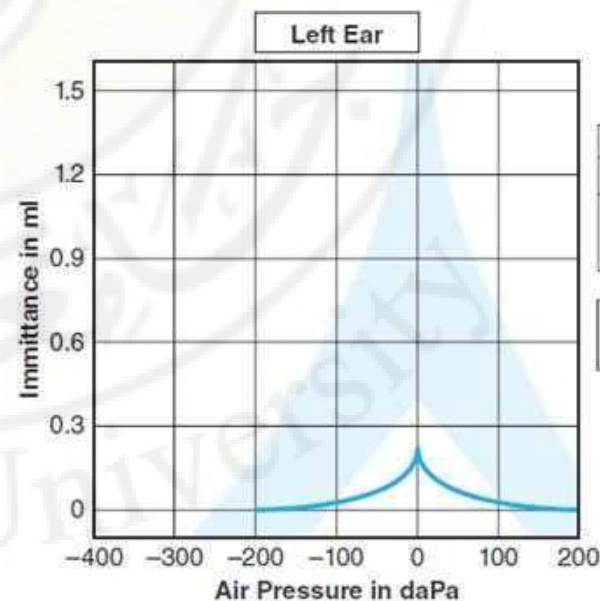
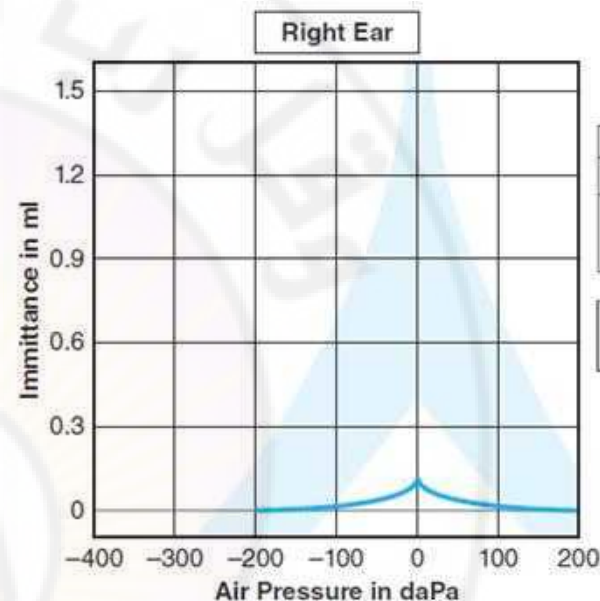
Case Example

Hearing Level in dB (ANSI-2004)



Air Conduction	Right	Left
Unmasked	○	×
Masked	△	□
Bone Conduction	Right	Left
Unmasked	<	>
Masked	[	]

Speech Audiometry					
Right			Left		
40	dB	SRT	45	dB	
96	%	WRS _m	100	%	
100	%	SSI _m	100	%	
100	%	DSI	100	%	



Signal to RE	BBN	500	1K	2K	4K
Crossed	>110				
Uncrossed			>110		
$\frac{\text{PTA}}{\text{BBN}} - \frac{5}{\text{Corr}} = \frac{\text{CNE}}{\text{SPAR}}$					

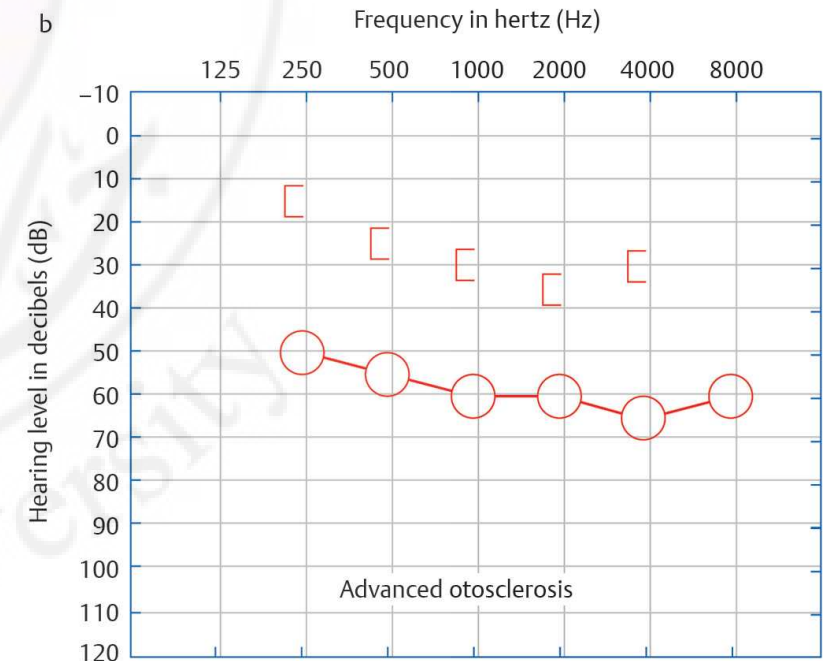
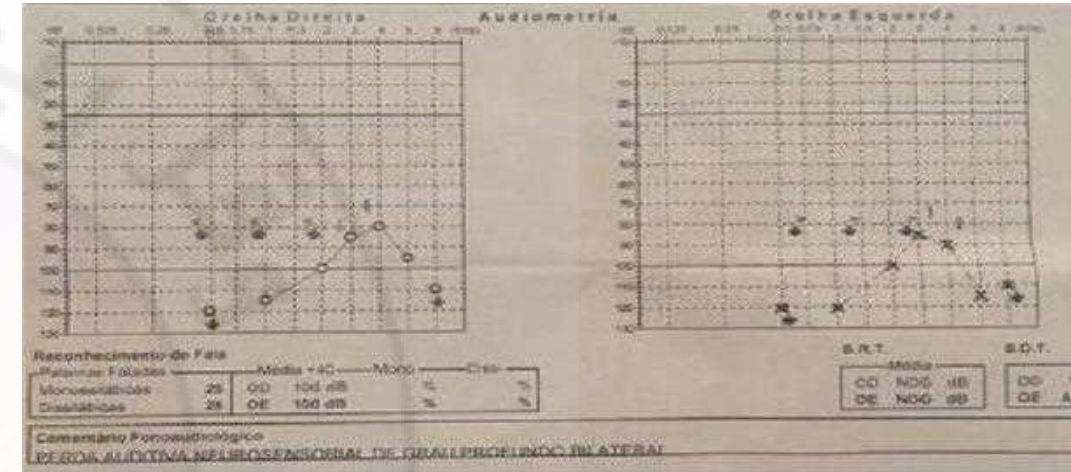
Static Immittance	=	0.1
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Signal to LE	BBN	500	1K	2K	4K
Crossed	>110				
Uncrossed			>110		
$\frac{\text{PTA}}{\text{BBN}} - \frac{5}{\text{Corr}} = \frac{\text{CNE}}{\text{SPAR}}$					

Static Immittance	=	0.2
-------------------	---	-----

Far Advanced Otosclerosis (FAO)

- Far-advanced otosclerosis (FAO) is an uncommon diagnosis. Patients with far advanced otosclerosis often present with what seems to be a **profound SNHL**, yet they may benefit from a stapedectomy.
- The hearing loss appears profound because the limit of the audiometer for bone conduction is 70 dB.
- Despite the severity of hearing loss, these patients often have **relatively normal speech and good speech comprehension**.
- There is often a family history of otosclerosis and a history of a progressive hearing loss since the second or third decade of life.
- At surgery, a fixed stapes is found, and hearing is improved postoperatively.
- Such patients subsequently obtain better use of a hearing aid with improved speech discrimination scores



Otosclerosis & van der Hoeve's syndrome

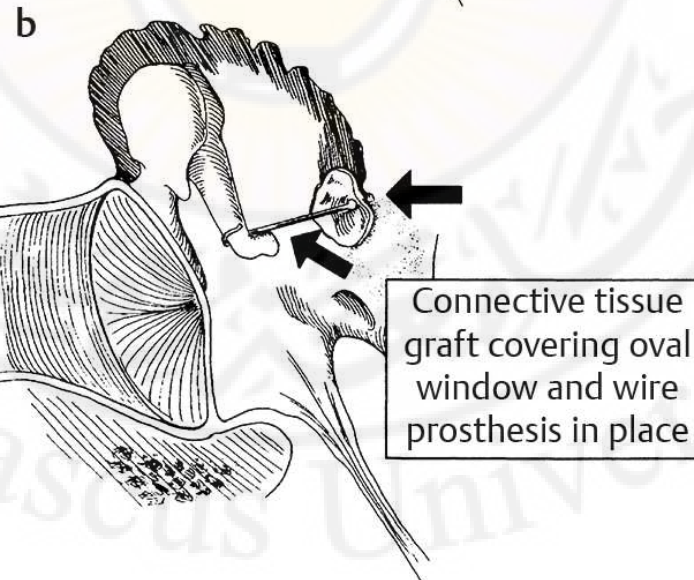
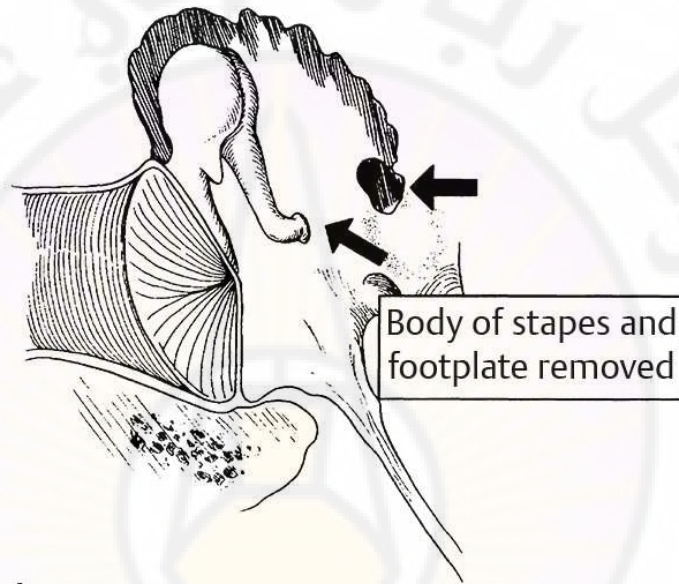
- Stapedial fixation that occurs in association with fragile, easily fractured bones (*osteogenesis imperfecta tarda*) and a bluish tint to the whites of the eyes (blue sclerae) is known as **van der Hoeve's syndrome**, and may be related to otosclerosis.



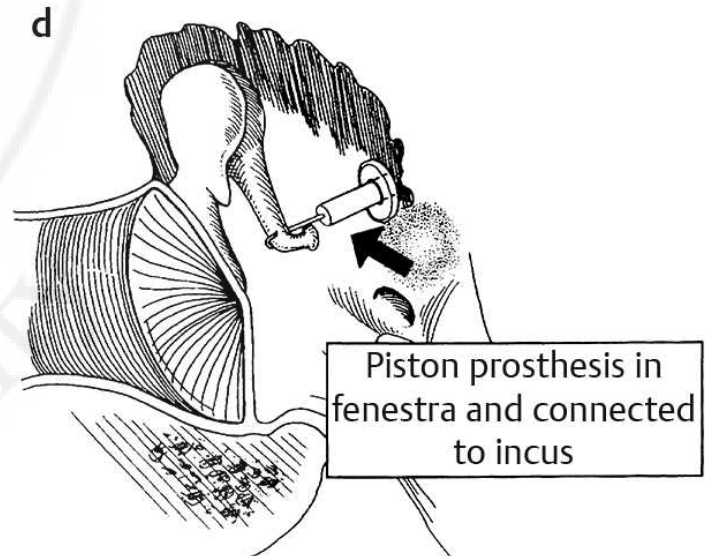
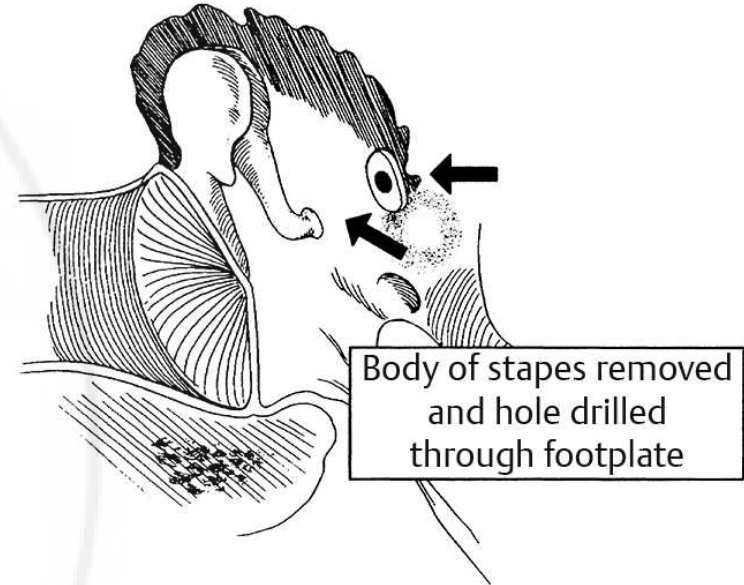
Otosclerosis Surgery

- Fenestration
- Stapes mobilization
- Stapedectomy
 - Total stapedectomy
 - Partial stapedectomy
- Stapedotomy

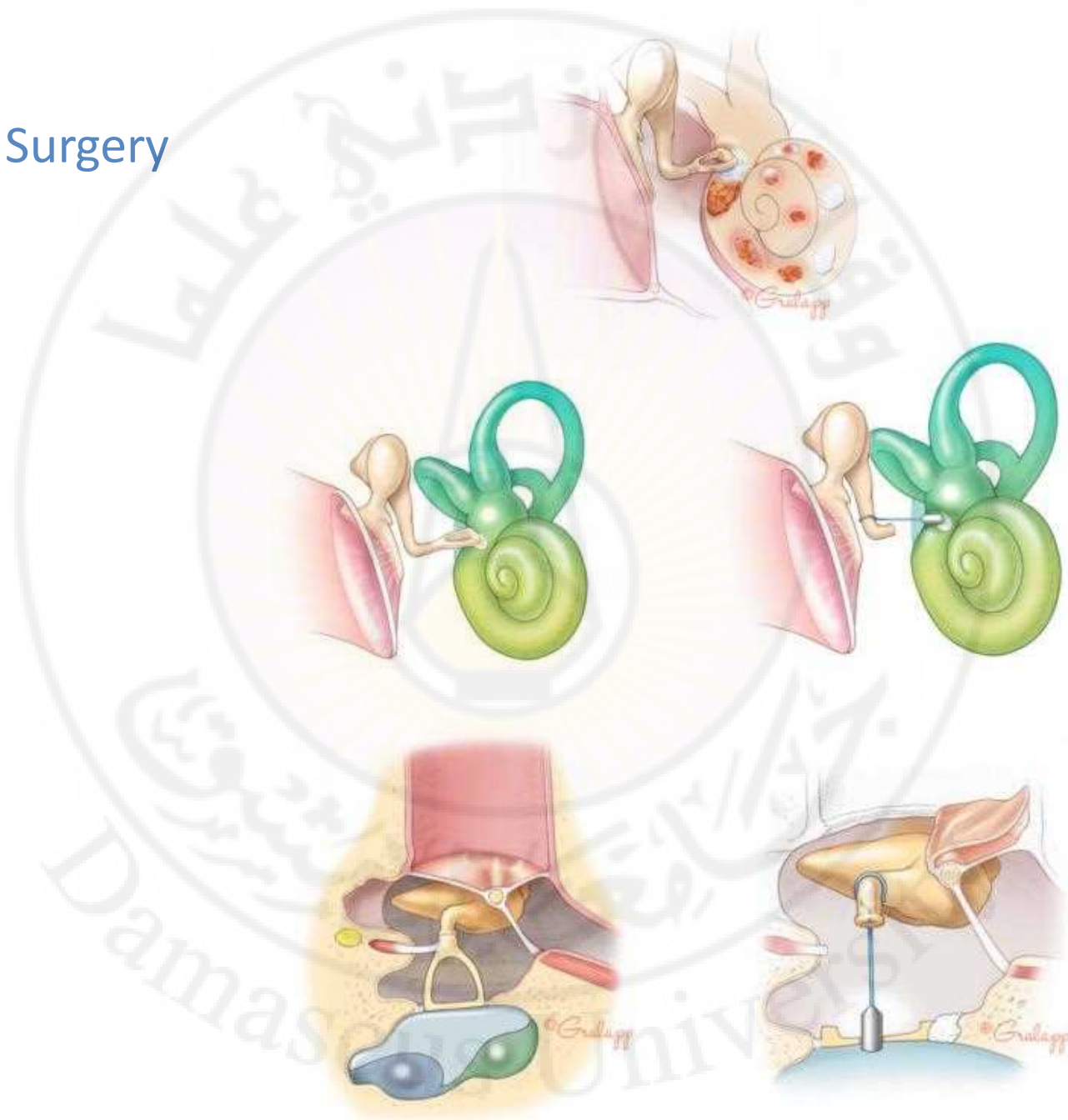
a Stapedectomy



c Stapedotomy

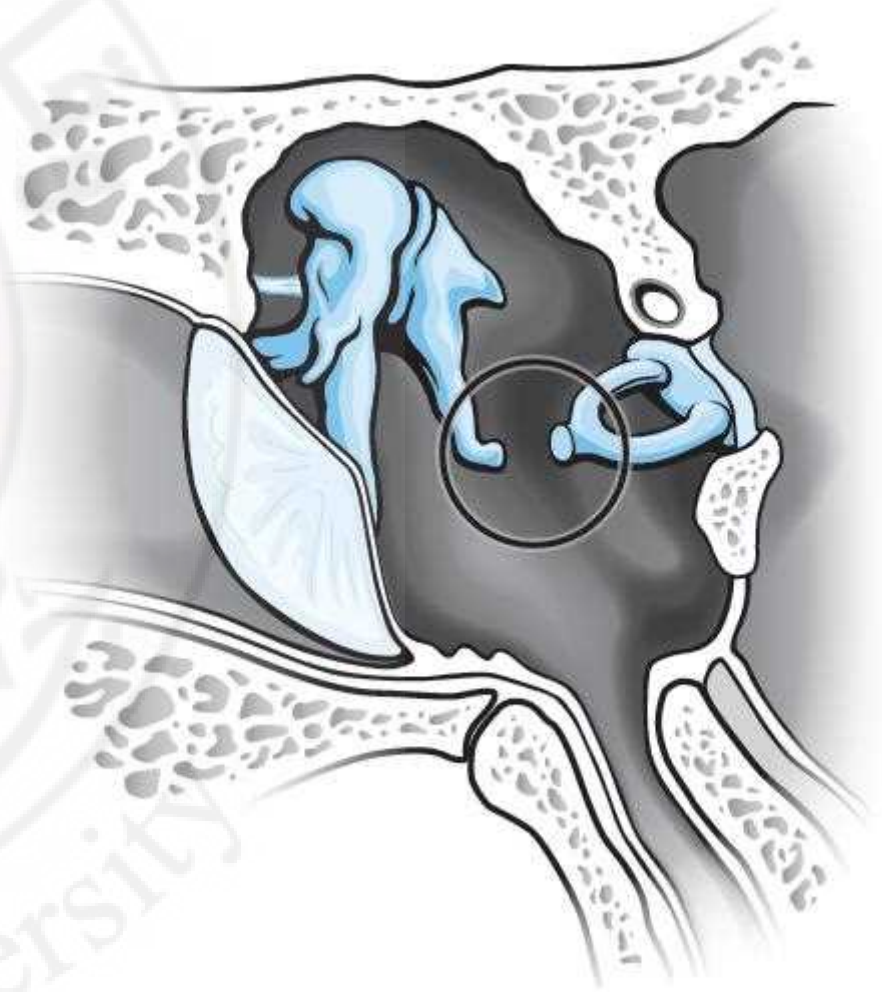


Otosclerosis Stapes Surgery



Ossicular discontinuities

- Physical **trauma** (as well as **infections**) can cause various kinds of middle-ear disorder. One consequence of trauma is a partial or total disarticulation of the ossicular chain.
- **Ossicular discontinuities** include partial fracture of the incus with separation of the incudostapedial joint, complete fracture of the incus, fracture of the crura of the stapes, and fracture of the malleus.
- Any of these types of ossicular disruptions can result in substantial conductive hearing loss.
- A complete disarticulation terminates the impedance matching capabilities of the middle ear by eliminating the stiffness component.
- In addition, the remaining unattached ossicles add mass to the system, resulting in a **maximum, flat conductive hearing loss**.

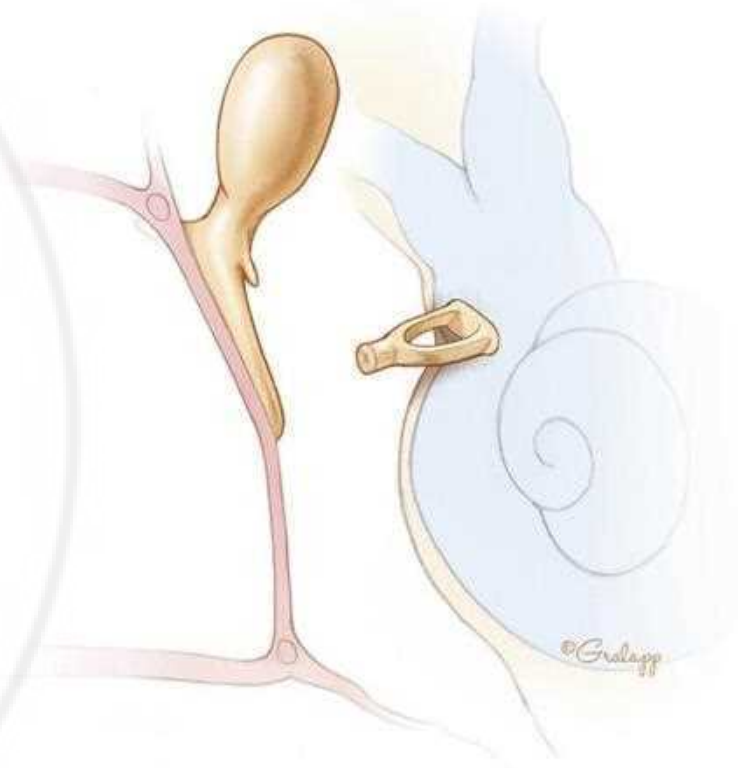




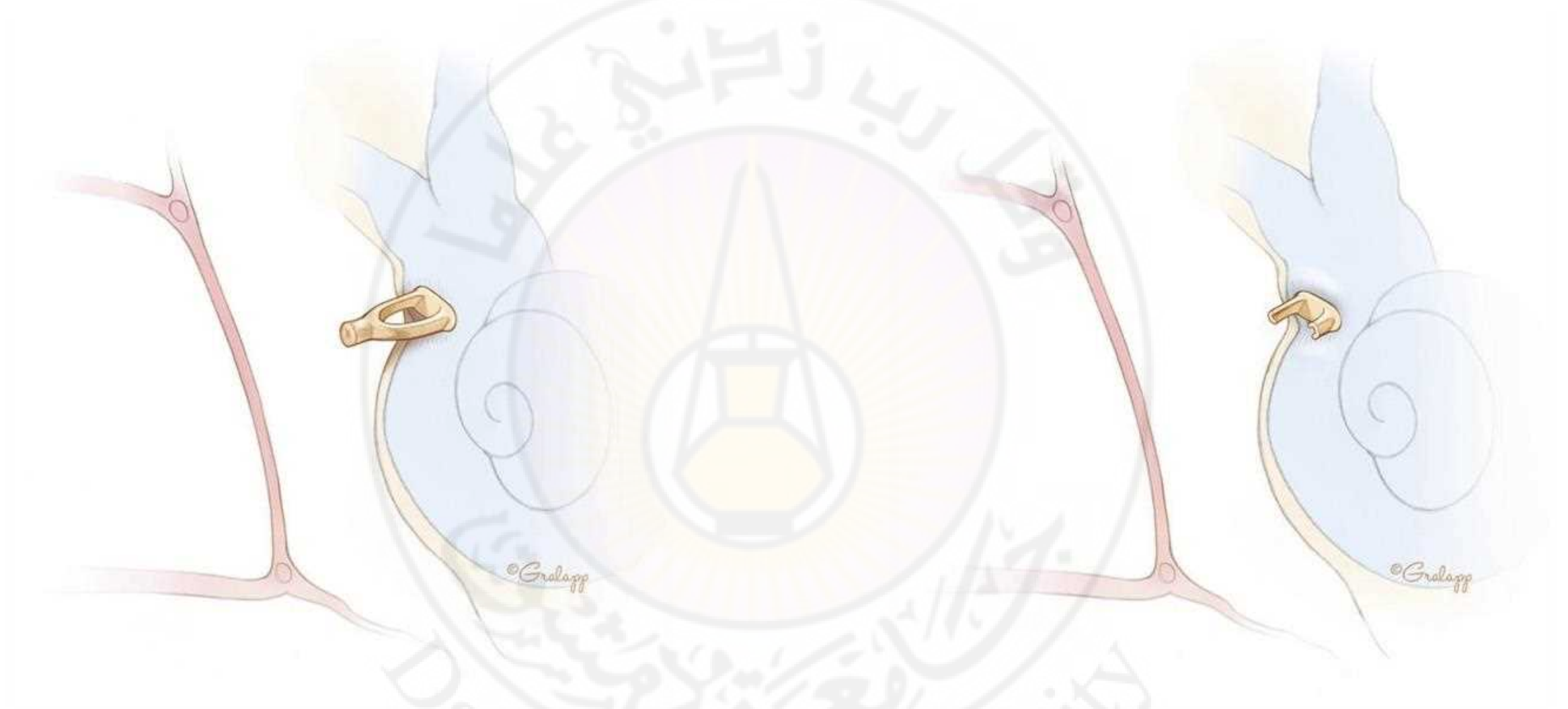
Incudostapedial joint separation most commonly occurs after temporal bone fracture.



Erosion of the long process of the incus most often occurs as a consequence of chronic otitis media or cholesteatoma. The long process sometimes fractures, especially following stapes surgery or in osteogenesis imperfecta.



Absence of a functional incus is common in chronic otitis media and cholesteatoma.

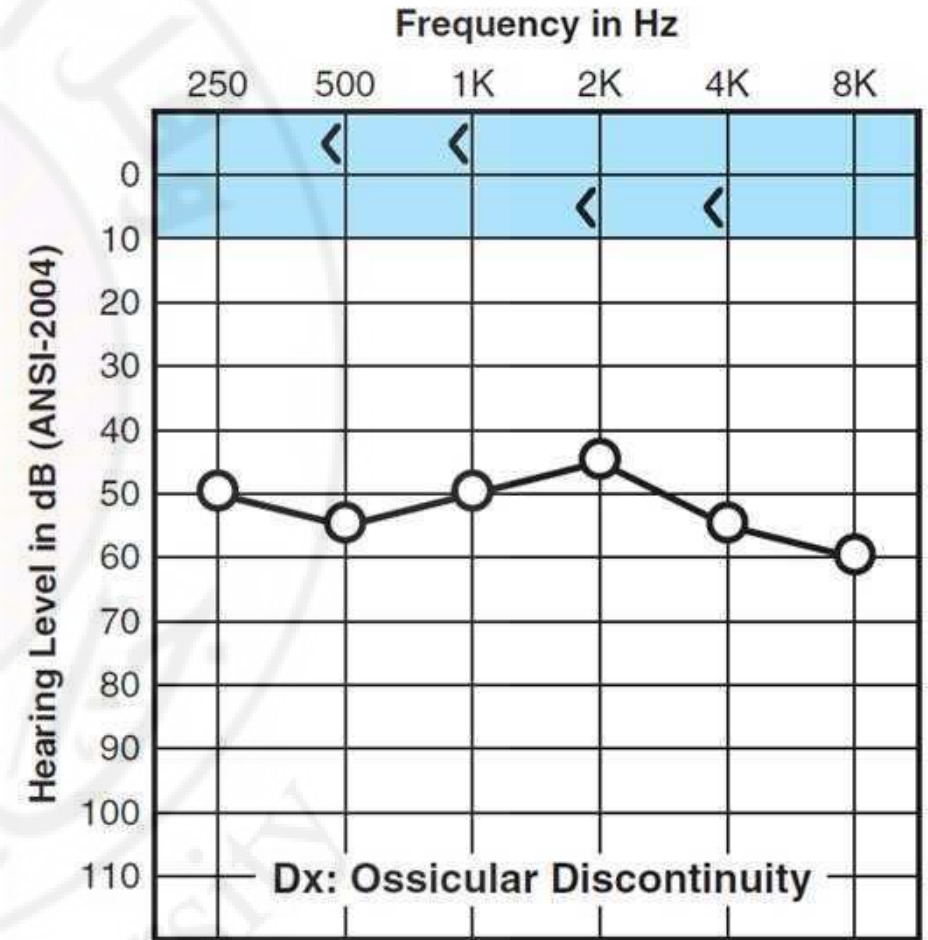


Absence of malleus and incus.

Cholesteatoma often involves the malleus, incus, and stapes arch leaving a crural remnant

Ossicular discontinuities

- Because of the frequent **medico-legal implications** of ear trauma, especially in cases of automobile accidents or industrial accidents, a very complete battery of audiometric tests is needed.
- Many times in these cases, the audiologist must be particularly astute. When litigation and possible enrichment exist together it is not uncommon for the patient to exaggerate the loss. In some cases there may be actual **malinger**ing.



Case Example

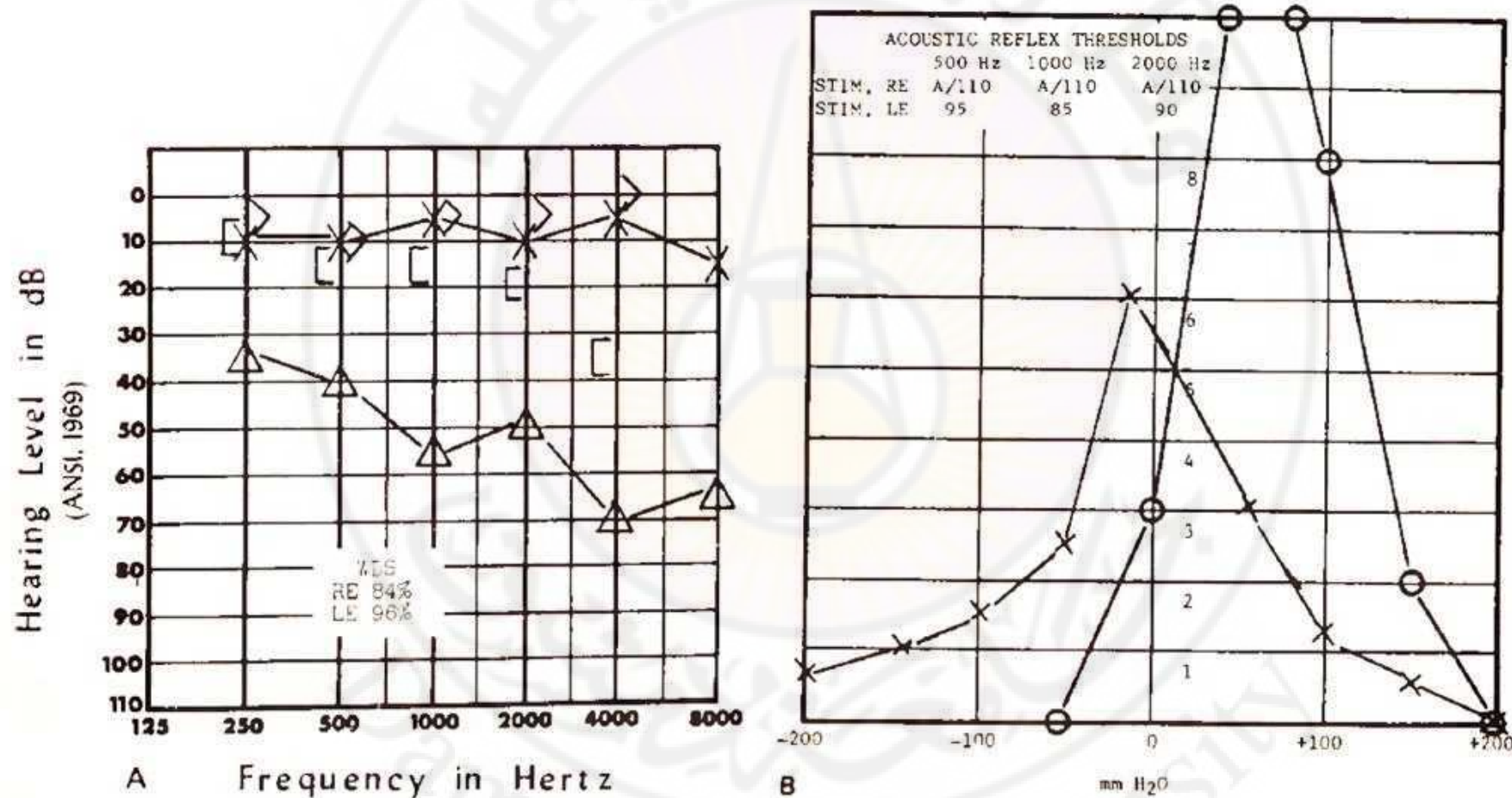


Fig. 2.9. (A) Hearing test results of 14-year-old girl with reported history of head trauma and subsequent surgically confirmed ossicular discontinuity (right ear). (B) Tympanograms of patient shown in A.

Case Example

	Left	Right
MEV:	1.4 ml	2.5 ml
COMP:	.6 ml	1.3 ml
MEP:	+18 daPa	-20 daPa
ACOUSTIC REFLEX:	Present	Absent

Pt. Information:

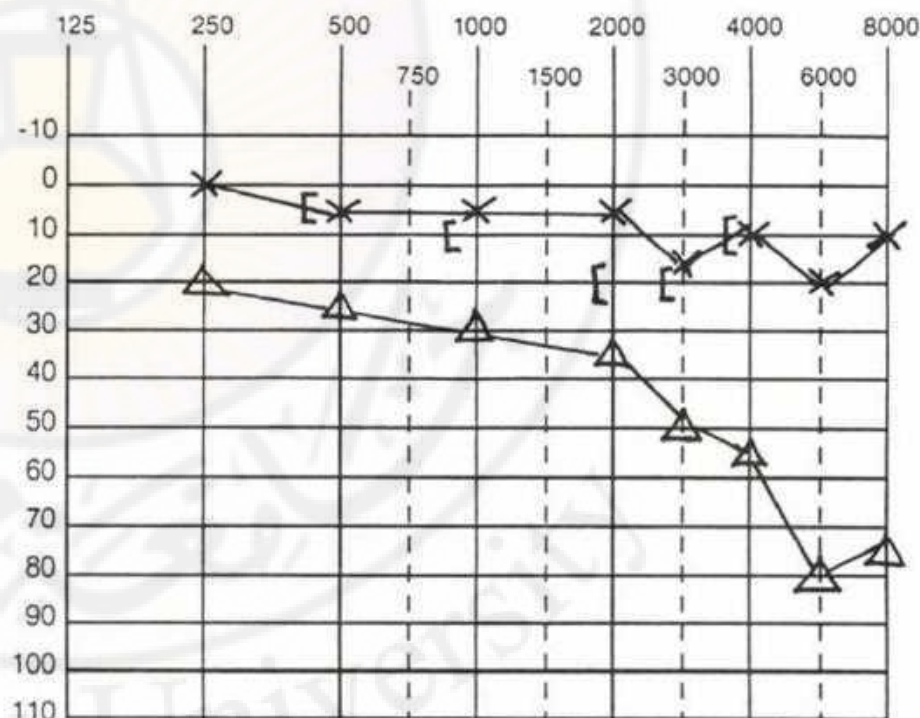
M X F Age: 24

SPEECH AUDIOMETRY					
	SRT		WORD RECOGNITION		
	dB	Mask	%	Mask	SL
RIGHT	40	45	100	55	30
LEFT	5		100		30
RIGHT					
LEFT					
BIN					

MASKING LEVELS USED

In Right (testing left)	AIR										
	BONE										
In Left (testing right)	AIR	← 45	50	55	65	70					
	BONE	45	50 →	55							
WEBER Lateralizes To	R	✓	✓								
		250	500	750	1000	1500	2000	3000	4000	6000	8000

PURE-TONE AUDIOGRAM
FREQUENCY IN HERTZ (Hz)



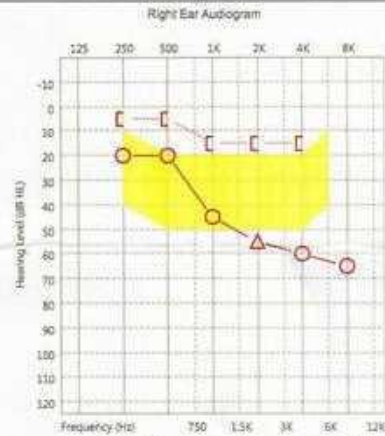
Case Example

First Name: [Redacted]
 Name: [Redacted]
 Age: 25 years
 Gender: Male



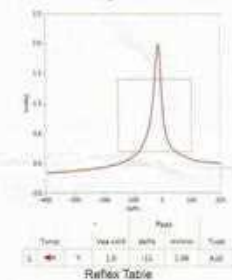
Facility Name
 Address (line 1)
 Address (line 2)
 Phone

4/26/2021 5:53 PM



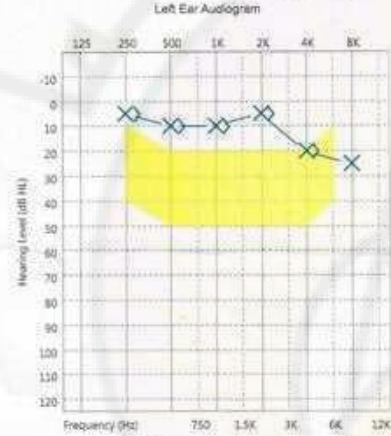
Ear	Test Type	% dB HL
R	SRT	35
R	MCL	60
R	WRS	96

Right Tym 226 Hz



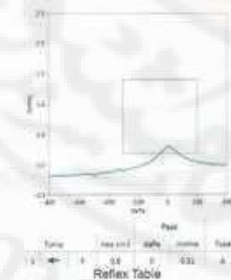
Reflex Table

Threshold	Ipsilateral					R
	500	1000	2000	250	4000	
dB HL	90#	90#	90#			
	Contralateral					L
	250	500	1000	2000	4000	
dB HL						



Ear	Test Type	% dB HL
L	SRT	15
L	MCL	40
L	WRS	95

Left Tym 226 Hz



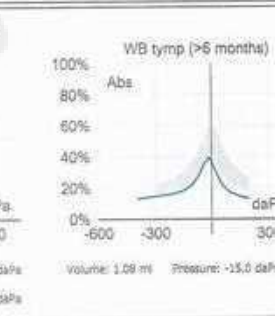
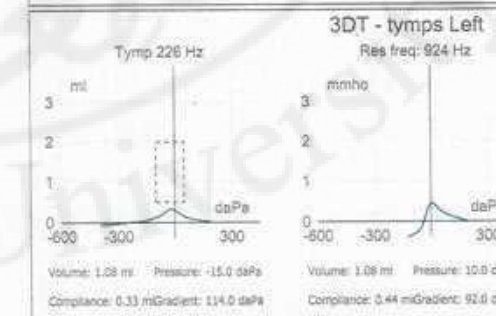
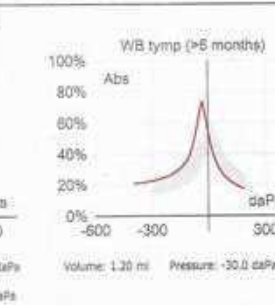
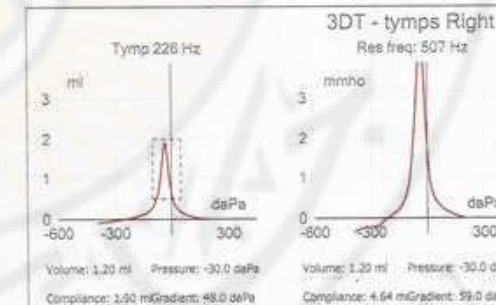
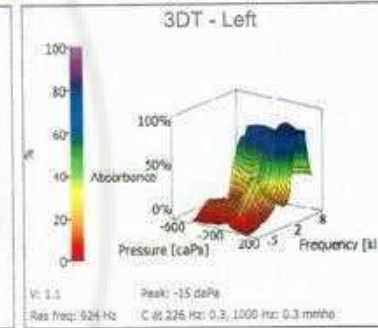
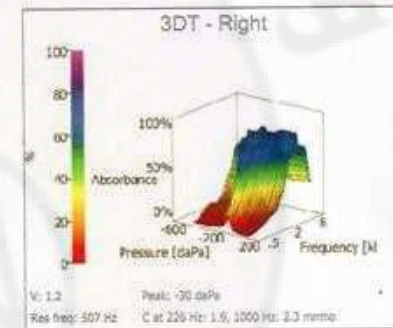
Reflex Table

Threshold	Ipsilateral				
	500	1000	250	2000	4000
dB HL	95#	100#			
	Contralateral				
	250	500	1000	2000	4000
dB HL					

Person ID: [Redacted]
 First name: [Redacted]
 Last name: [Redacted]
 Birth date: [Redacted]
 Gender: Male



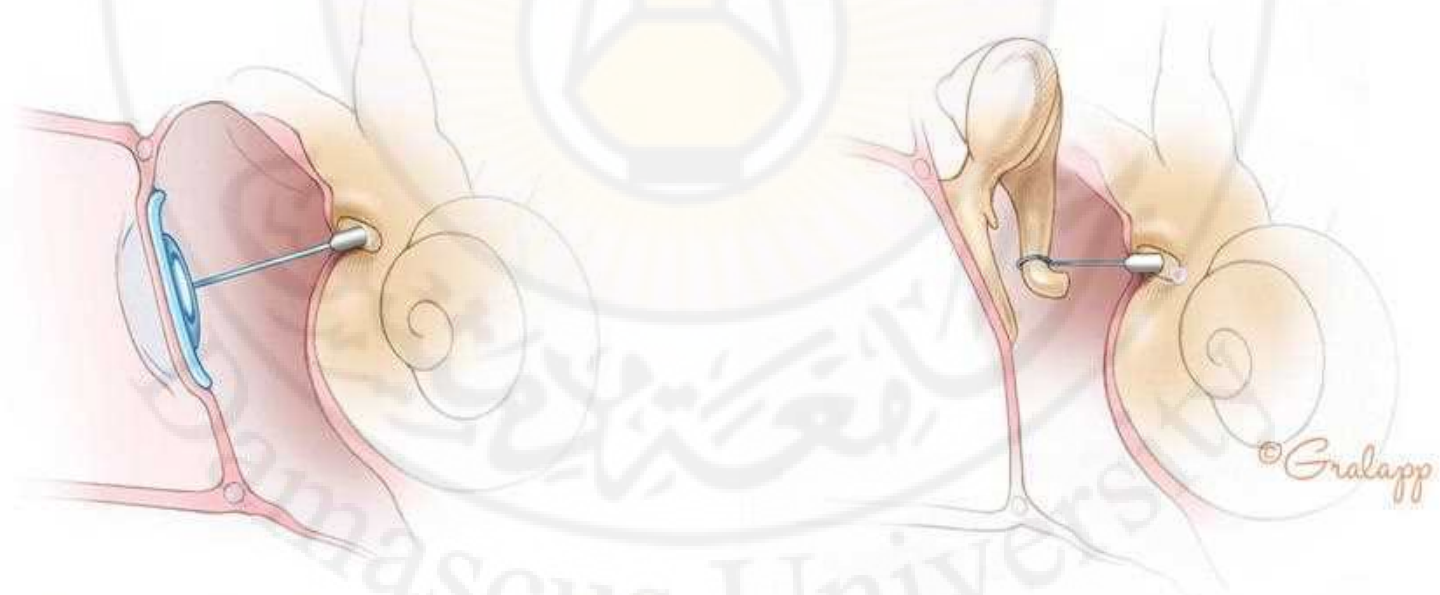
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 Day Medical Tower
 Unit C, 9th Floor
 Tehran
 021-88205808





Partial Ossicular Chain Reconstruction (PORP)

Crutch and Cup



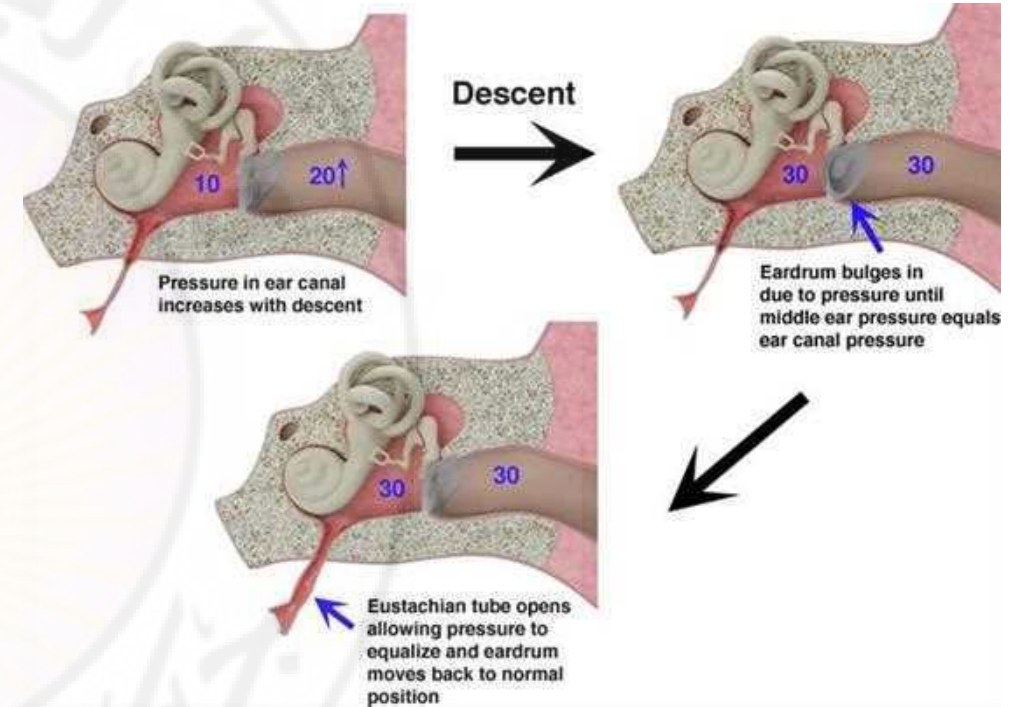
Total Ossicular Chain Reconstruction (TORP)

Stapes Replacement

© Ghalapp

Barotrauma

- Another form of insult to the middle ear results from **barotrauma (aerotitis)**, or trauma related to a sudden, marked change in atmospheric pressure.
- This often occurs when an airplane descends from altitude too fast without proper airplane cabin pressure equalization or when a diver ascends too rapidly in the water.
- It can also occur under more normal ascending and descending conditions if a person's Eustachian tube is not providing adequate pressure equalization.
- Under both circumstances, a severe negative air pressure is created in the middle-ear space due to failure of the Eustachian tube to open adequately.



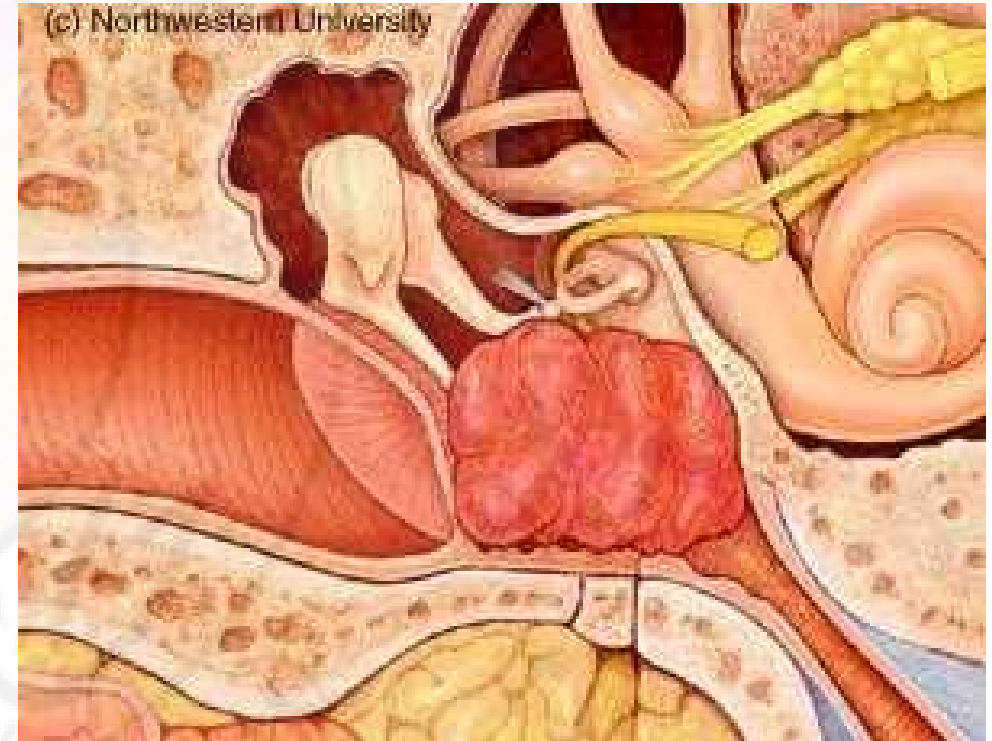
Barotrauma

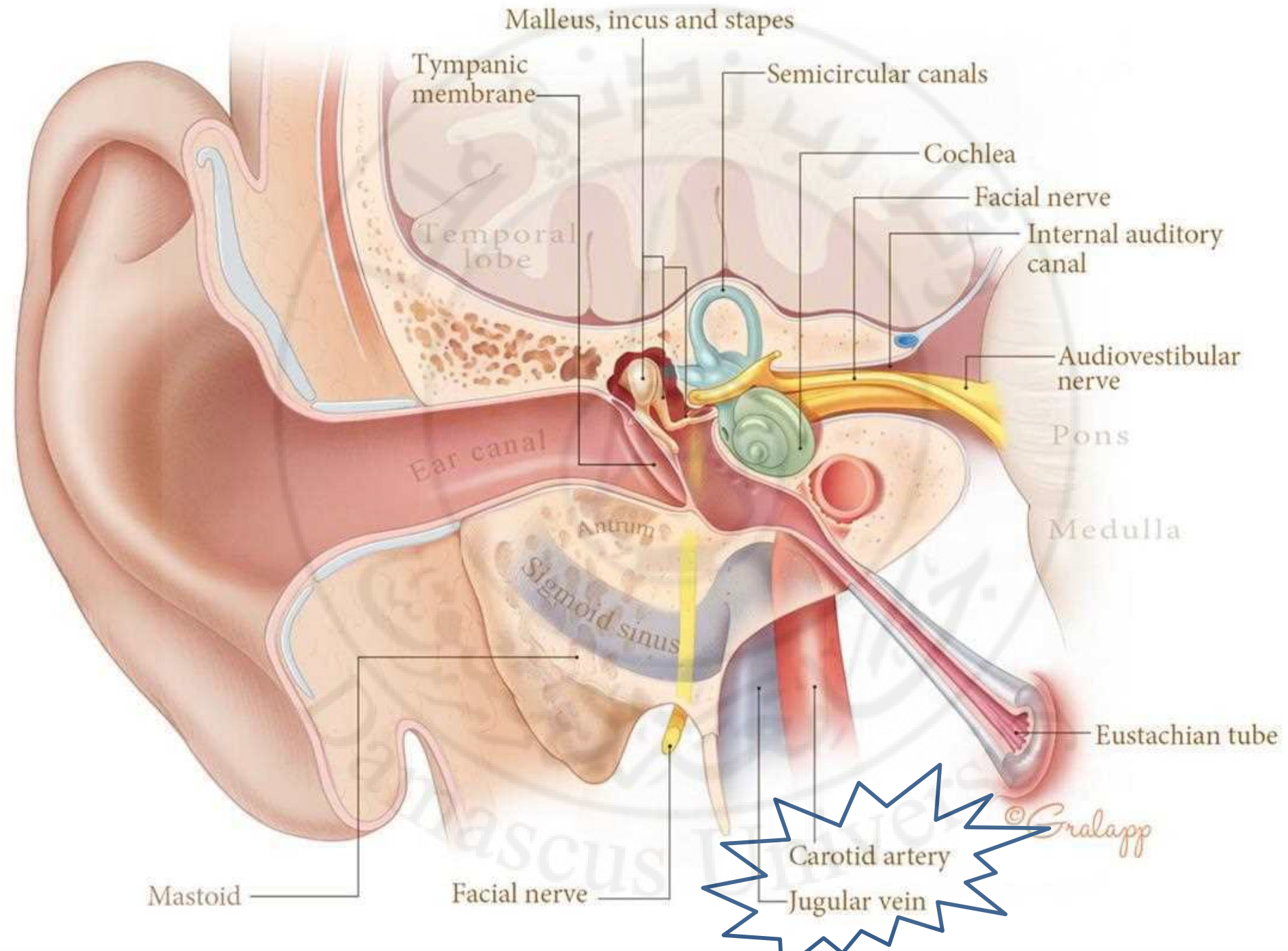
- If air pressure changes are sudden and intense, the tympanic membrane may rupture. If not, it is stretched medially, followed by increased blood flow, swelling, and bruising of the mucosal lining of the middle ear.
- Effusion then forms within the middle ear from the traumatized tissue.
- Barotrauma can cause TM perforation, CHL, and, in rare cases, a fistula of the oval window.



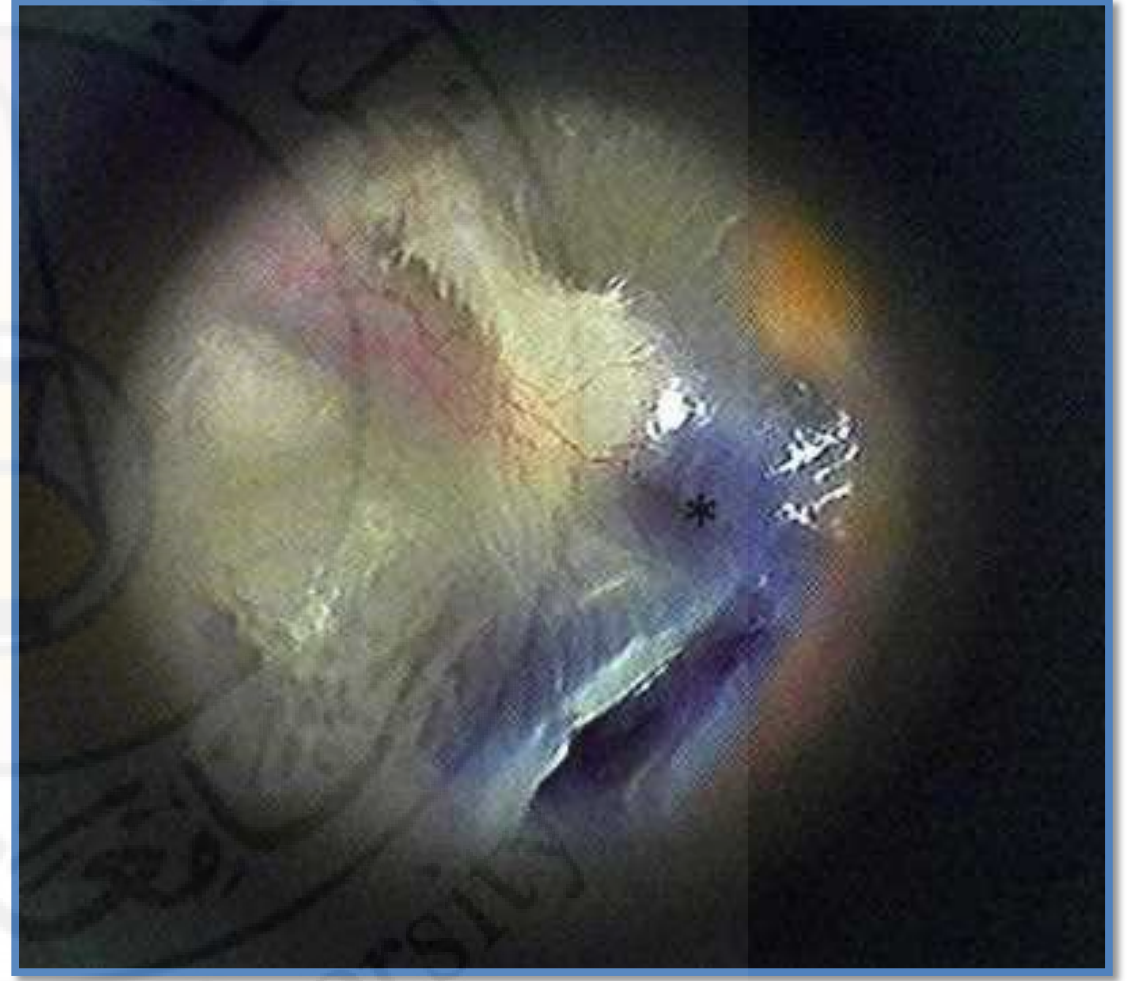
Glomus tumor

- Tumors may also occur within the middle ear. One neoplasm commonly found in the middle ear is a **glomus tumor, or glomus tympanicum**.
- Glomus tumors, are slow-growing, usually benign tumors in the carotid arteries (major blood vessels in neck), the middle ear or the area below the middle ear (jugular bulb), and can result in significant conductive hearing loss.
- They are called **glomus tympanicum tumors** when they develop within the middle ear cavity and **glomus jugulare tumors** when they develop from the crest of the jugular bulb under the floor of the middle ear.





Jugular bulb in the middle ear space



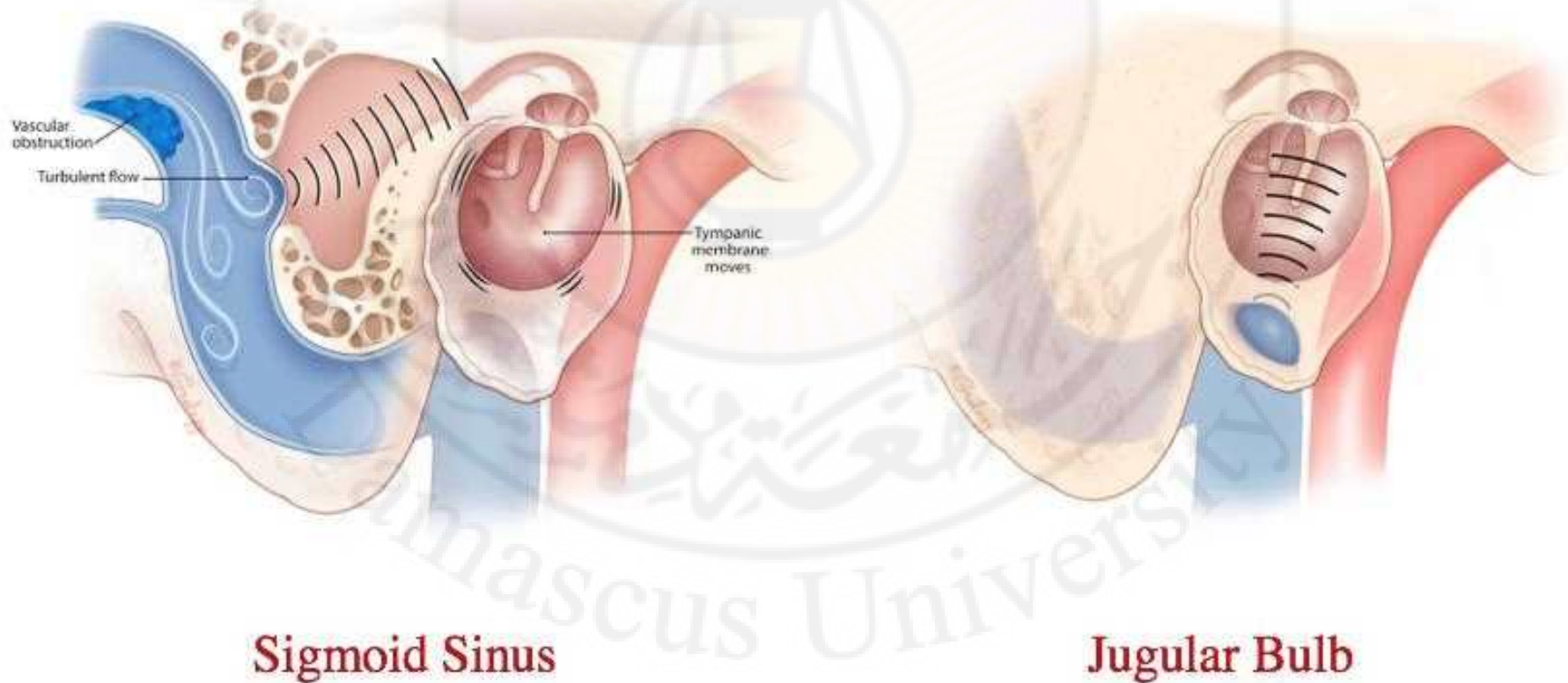
Glomus tumor

- Glomus tumors invade the middle ear and other parts of the temporal bone, and can also extend intracranially.
- They are usually **unilateral** and are more common in **women** than in men by a ratio of roughly 3 to 1.
- The most common characteristics of patients with glomus tumors are **pulsing tinnitus** that is synchronized to the patient's vascular pulse and a **conductive or mixed hearing loss**, which is unilateral in a wide majority of cases.
- **Bleeding** from the ear occurs in some cases, and patients may also experience a feeling of **pressure** in the ear. Other complaints may also occur but are highly variable.
- Glomus tumors are usually seen as a **reddish mass behind the tympanic membrane**, often with a "**rising sun**" appearance one might also see it pulsating, and possibly causing the eardrum to bulge.
- Glomus tumors are treated surgically.



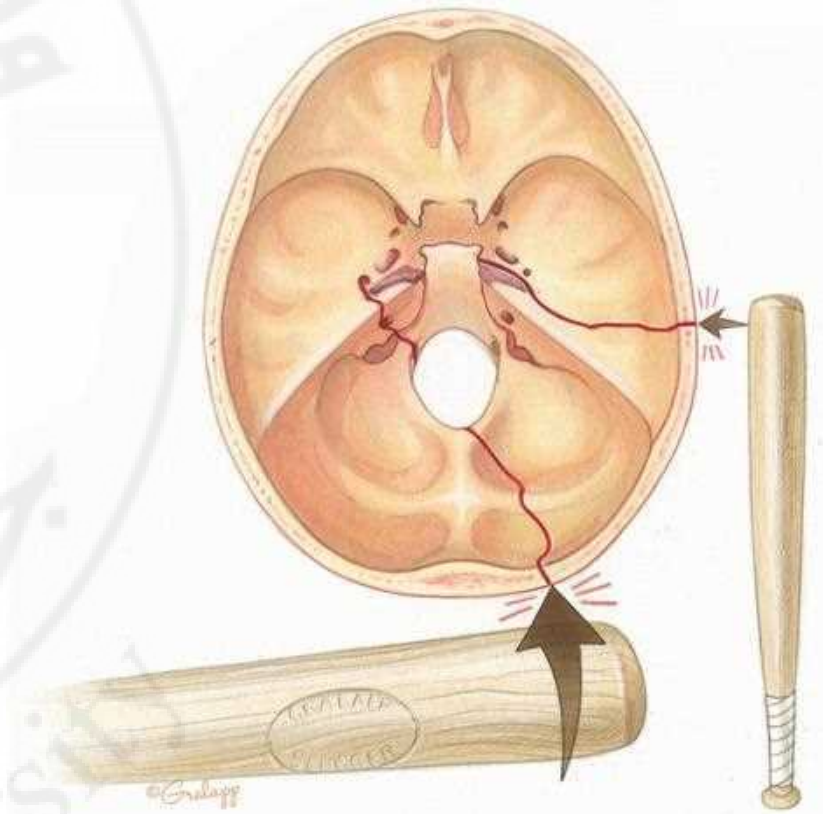
Glomus tumor & Audiology

Type B tympanogram + ABG + bulged TM + **Pulsatile tinnitus** + Bleeding

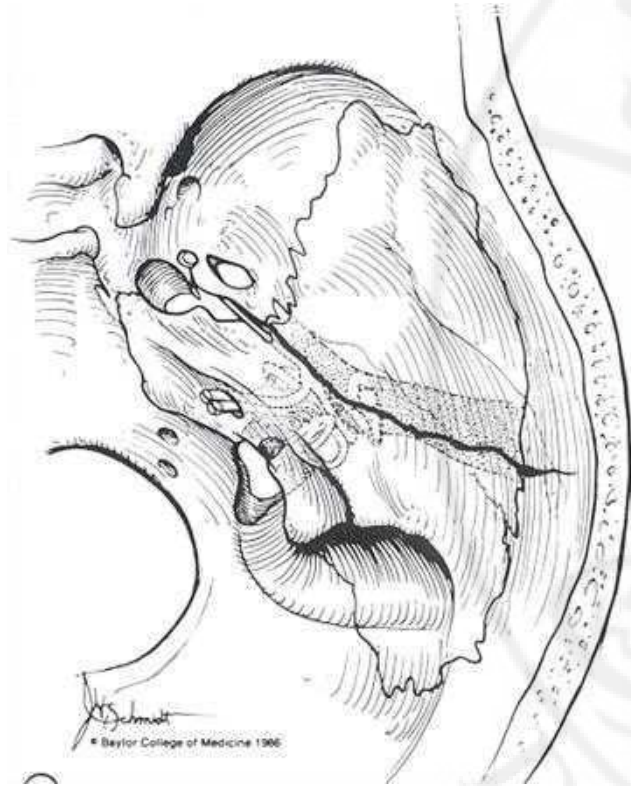


Fracture of temporal bone

- The temporal bone is the thickest bone in the body, requiring significant force to fracture (**850 kg of lateral force** in one study).
- The temporal bone houses portions of the internal carotid artery and internal jugular vein, cranial nerves (CN) VII-XI, the middle and inner ear structures, and forms a portion of the middle cranial fossa. Such high-velocity injury to these crucial structures carries the risk of significant morbidity and mortality.
- Approximately one-third of temporal bone fractures in the general population result from motor vehicle accidents, assaults, and falls, and an additional ~25% are attributed to sports injuries, gunshot wounds, bicycle accidents, and other miscellaneous causes.
- **Males** are at least 3-times more likely than females to suffer temporal bone fractures. Temporal bone fractures are typically **unilateral**.



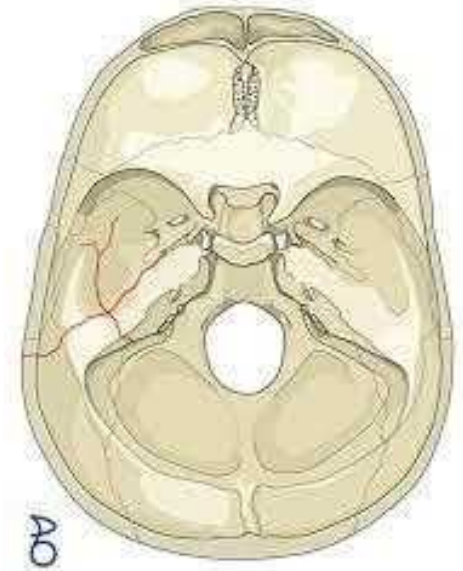
Fracture of temporal bone - Classifications



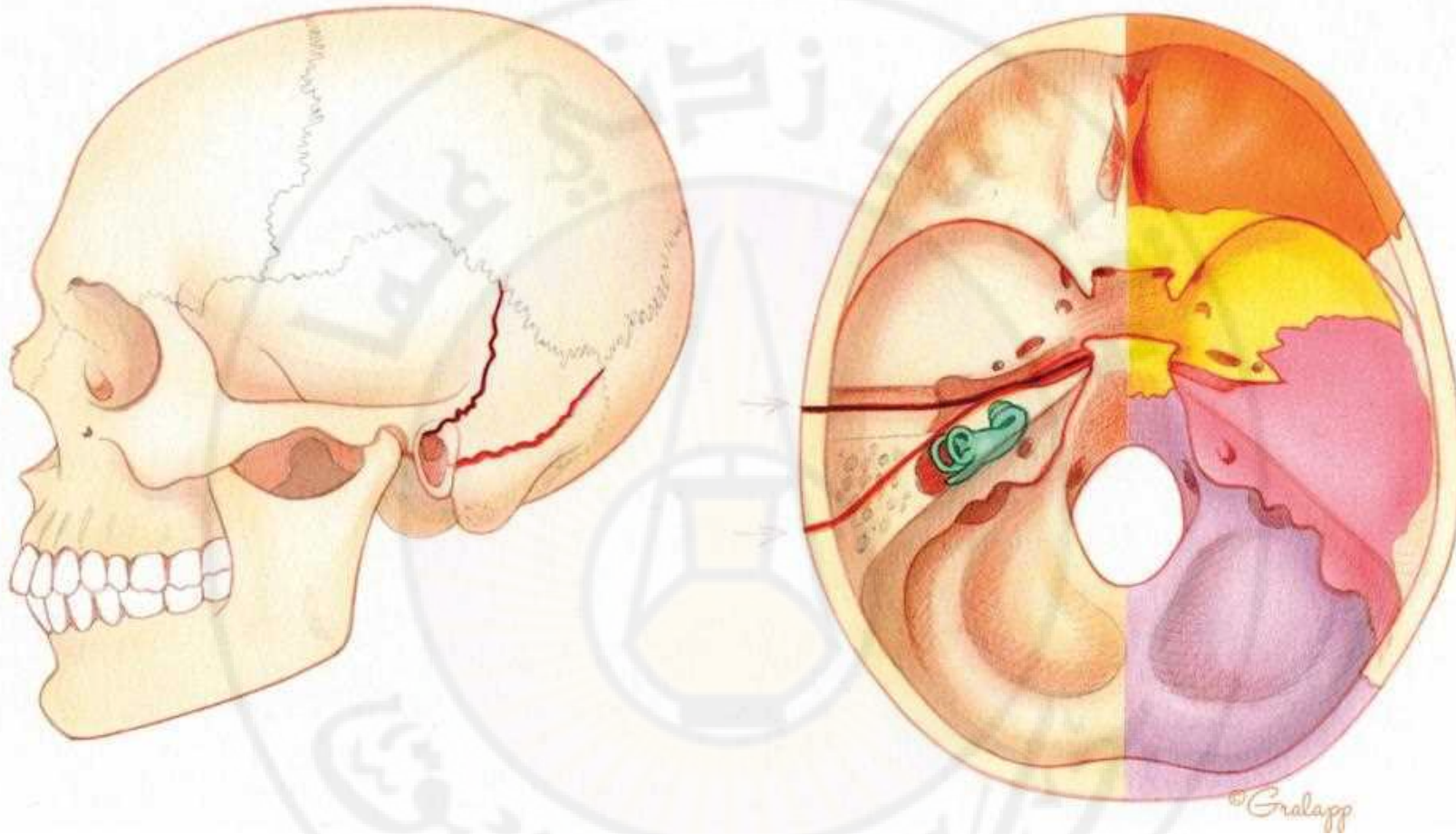
Longitudinal fractures



Transverse fractures

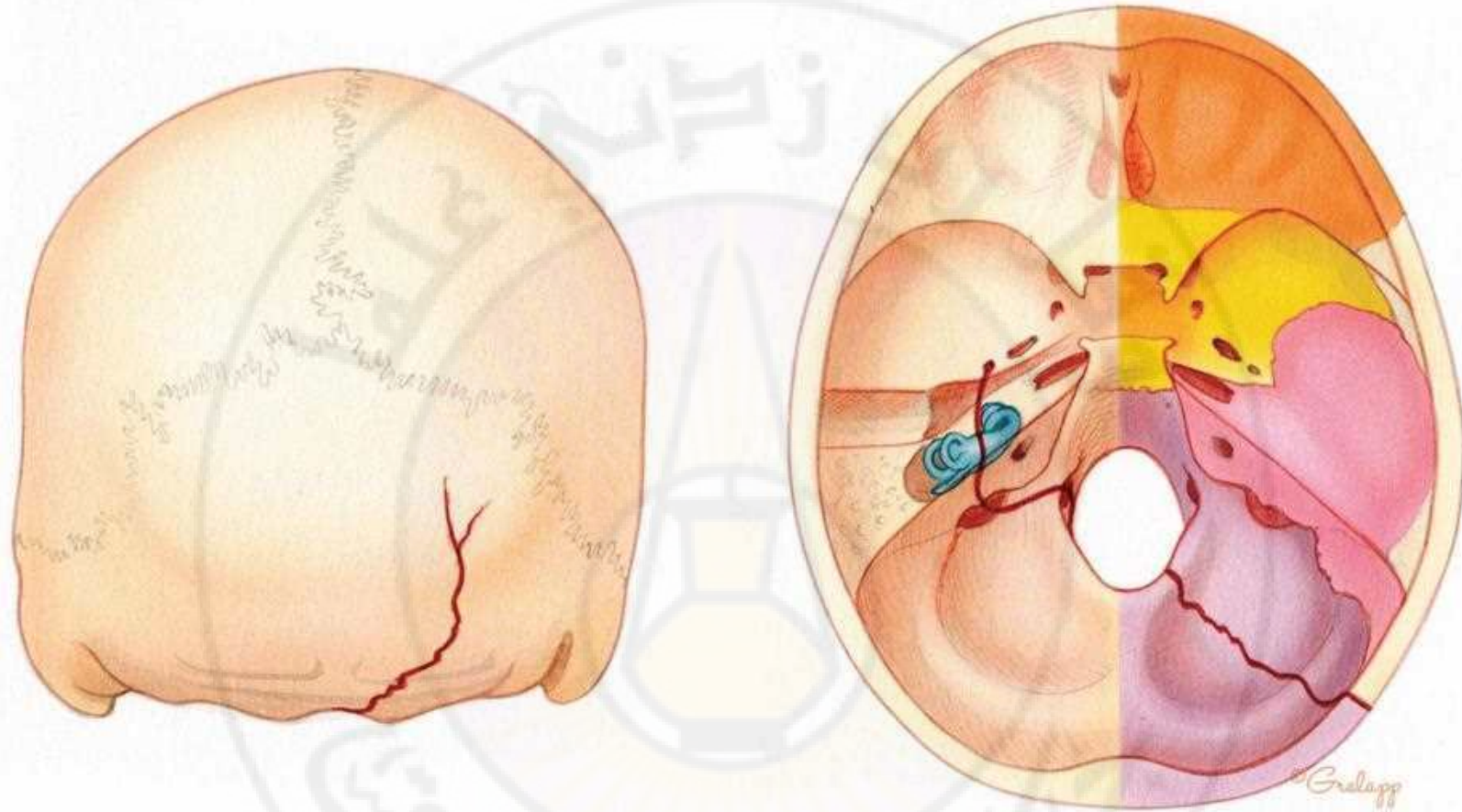


Mixed fractures



fractures that course parallel to the long axis of the petrous ridge are considered **"longitudinal"**

"Longitudinal fractures made up approximately 70-90% of all temporal bone fractures"



fractures that course perpendicular to the long axis of the petrous ridge are considered **"transverse"**

"transverse fractures made up approximately 10-30% of all temporal bone fractures"

Fracture of temporal bone - symptoms

- **Hearing Loss & tinnitus**

- 71% of patients with Temporal Bone Trauma have hearing loss
- TM Perforations
- CHL > 40db suspicion for ossicular discontinuity

- **Longitudinal Fractures**

- Conductive or mixed hearing loss
- 80% of CHL resolve spontaneously

- **Transverse Fractures**

- Sensorineural hearing loss
- Less likely to improve

- **Dizziness & Vertigo**

- **CSF Otorrhea and Rhinorrhea**

- **Facial Nerve Injuries**





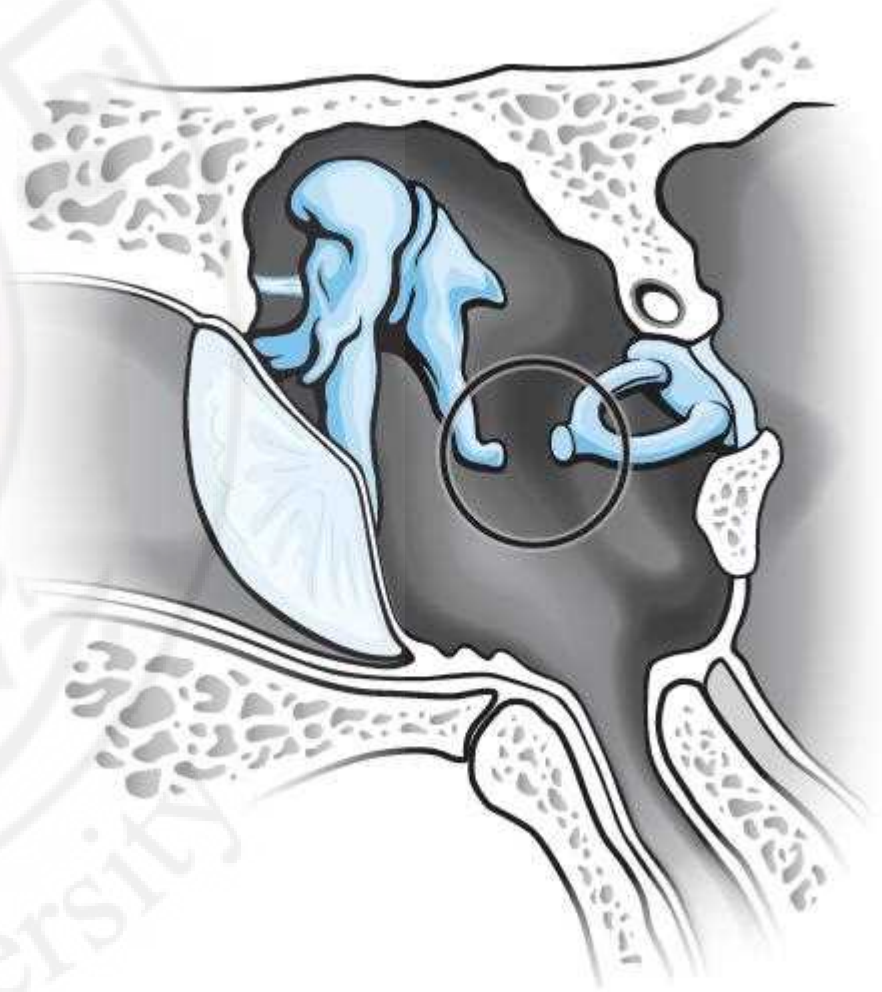
Clinical Audiology

Mohsen Ahadi, Ph.D.

Associate Professor of Audiology
Iran University of Medical Sciences
Rehabilitation Research Center
ahadi.m@iums.ac.ir

Ossicular discontinuities

- Physical **trauma** (as well as **infections**) can cause various kinds of middle-ear disorder. One consequence of trauma and ME infections is a partial or total disarticulation of the ossicular chain.
- **Ossicular discontinuities** include partial fracture of the incus with separation of the incudostapedial joint, complete fracture of the incus, fracture of the crura of the stapes, and fracture of the malleus.
- Any of these types of ossicular disruptions can result in substantial conductive hearing loss.
- A complete disarticulation terminates the impedance matching capabilities of the middle ear by eliminating the stiffness component.
- In addition, the remaining unattached ossicles add mass to the system, resulting in a **maximum, flat conductive hearing loss**.

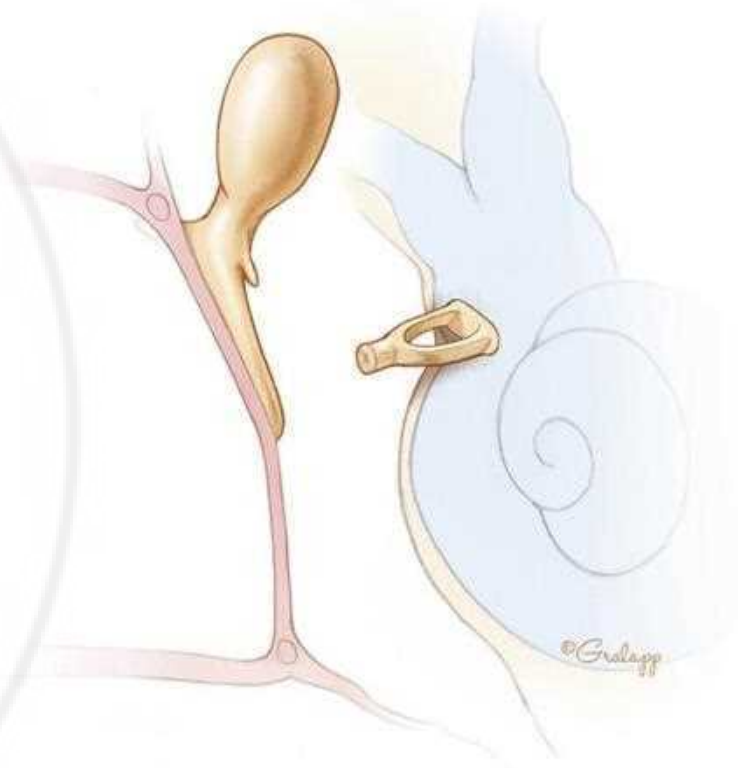




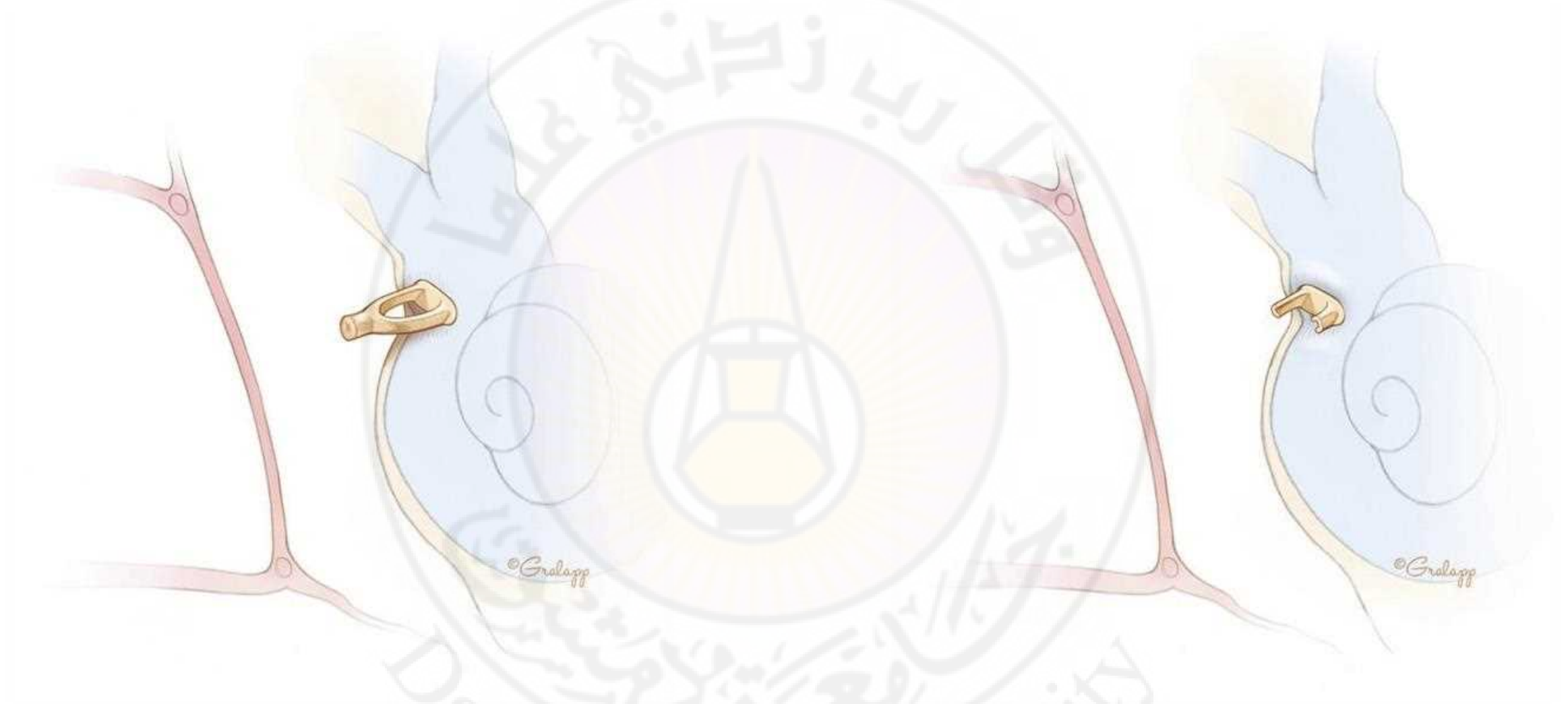
Incudostapedial joint separation most commonly occurs after temporal bone fracture.



Erosion of the long process of the incus most often occurs as a consequence of chronic otitis media or cholesteatoma. The long process sometimes fractures, especially following stapes surgery or in osteogenesis imperfecta.



Absence of a functional incus is common in chronic otitis media and cholesteatoma.

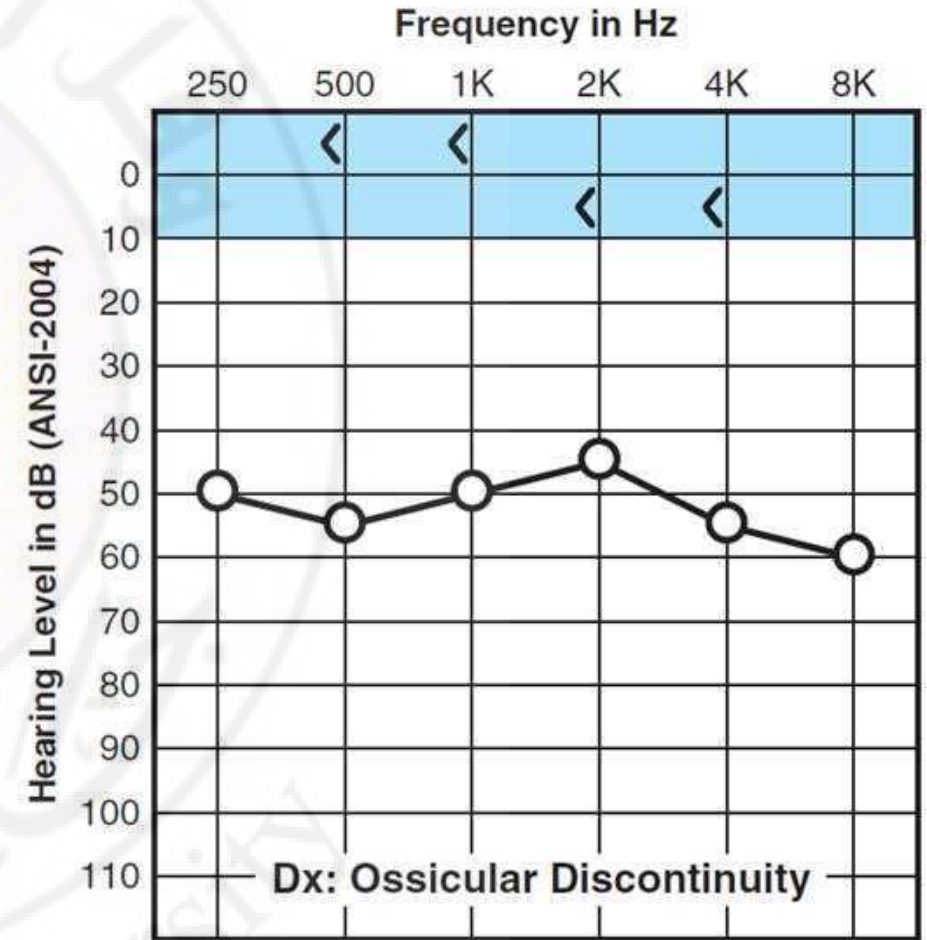


Absence of malleus and incus.

Cholesteatoma often involves the malleus, incus, and stapes arch leaving a crural remnant

Ossicular discontinuities

- Because of the frequent **medico-legal implications** of ear trauma, especially in cases of automobile accidents or industrial accidents, a very complete battery of audiometric tests is needed.
- Many times in these cases, the audiologist must be particularly sharp. When litigation and possible enrichment exist together it is not uncommon for the patient to exaggerate the loss. In some cases there may be actual **malinger**ing.



Case Example

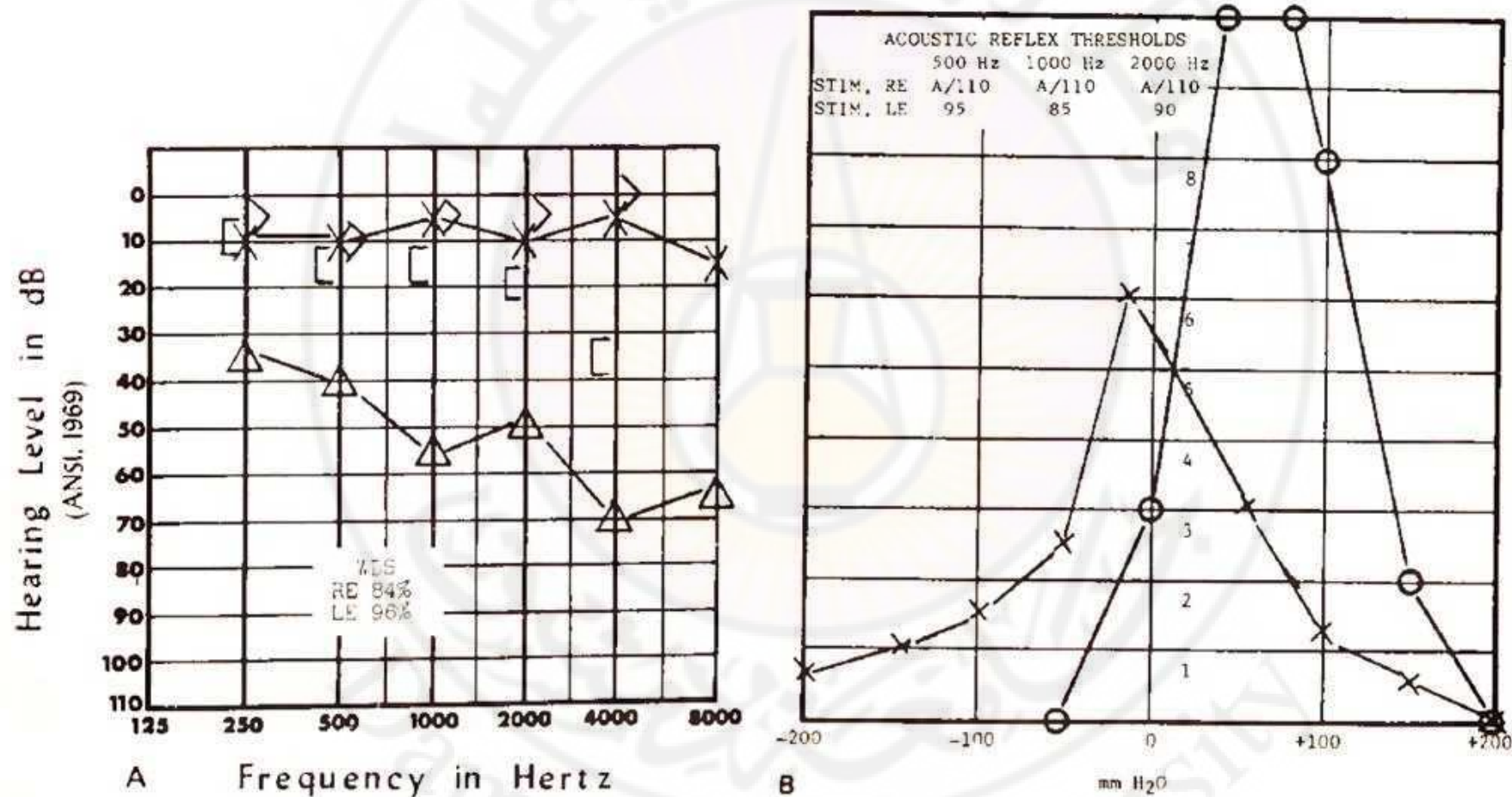


Fig. 2.9. (A) Hearing test results of 14-year-old girl with reported history of head trauma and subsequent surgically confirmed ossicular discontinuity (right ear). (B) Tympanograms of patient shown in A.

Case Example

	Left	Right
MEV:	1.4 ml	2.5 ml
COMP:	.6 ml	1.3 ml
MEP:	+18 daPa	-20 daPa
ACOUSTIC REFLEX:	Present	Absent

Pt. Information:

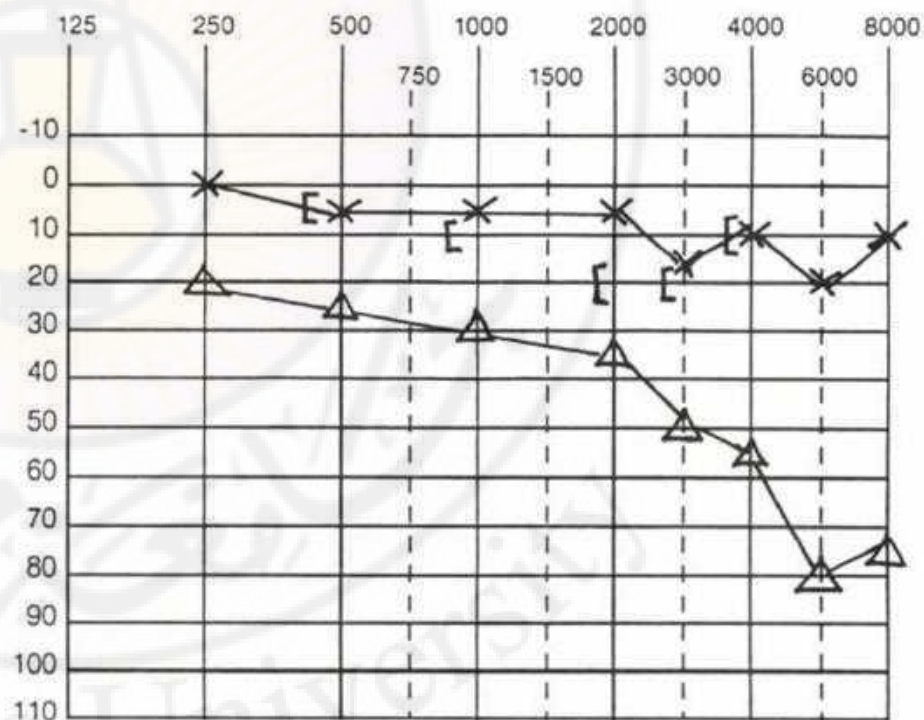
M X F Age: 24

SPEECH AUDIOMETRY					
	SRT		WORD RECOGNITION		
	dB	Mask	%	Mask	SL
RIGHT	40	45	100	55	30
LEFT	5		100		30
RIGHT					
LEFT					
BIN					

MASKING LEVELS USED

In Right (testing left)	AIR										
	BONE										
In Left (testing right)	AIR	← 45	50	55	65	70					
	BONE	45	50 →	55							
WEBER Lateralizes To	R	✓	✓								

PURE-TONE AUDIOGRAM
FREQUENCY IN HERTZ (Hz)



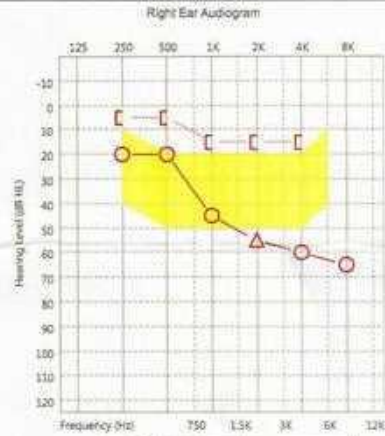
Case Example

First Name: [Redacted]
 Name: [Redacted] Andoust
 Age: 25 years
 Gender: Male



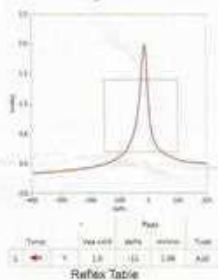
Facility Name
 Address (line 1)
 Address (line 2)
 Phone

4/26/2021 5:53 PM



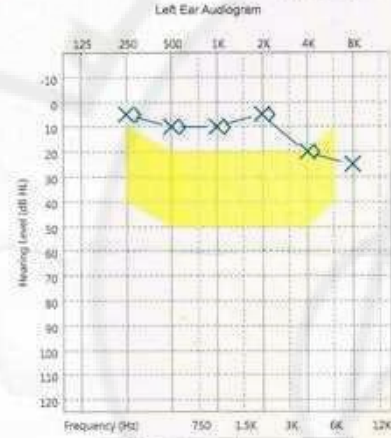
Ear	Test Type	% dB HL
R	SRT	35
R	MCL	60
R	WRS	96

Right Tym 226 Hz



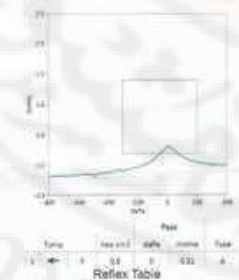
Reflex Table

Threshold	Ipsilateral					R
	500	1000	2000	250	4000	
dB HL	90#	90#	90#			
	Contralateral					R
	250	500	1000	2000	4000	
dB HL						



Ear	Test Type	% dB HL
L	SRT	15
L	MCL	40
L	WRS	95

Left Tym 226 Hz



Reflex Table

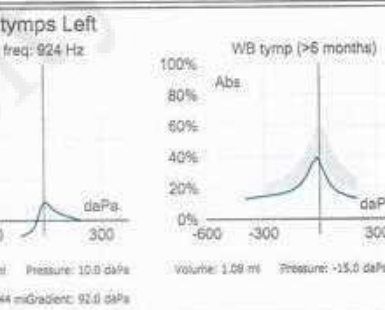
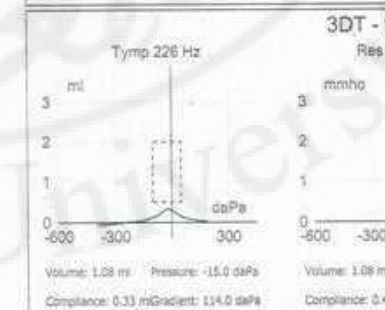
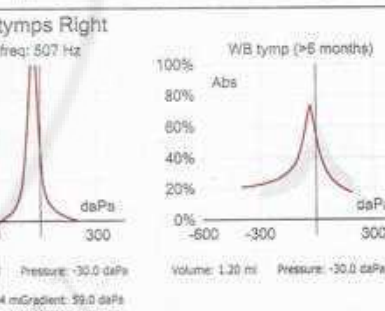
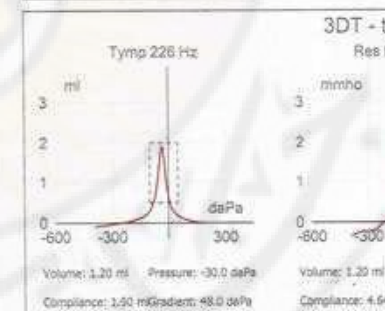
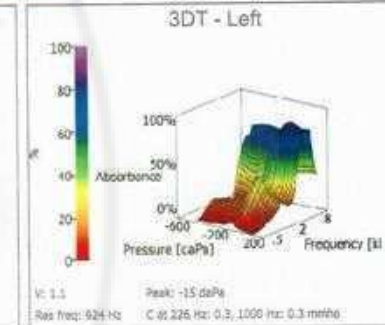
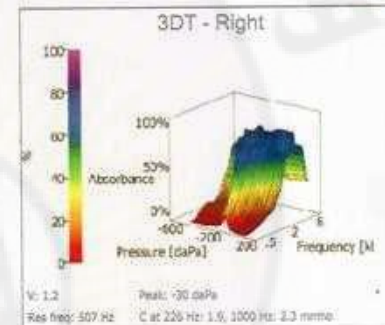
Threshold	Ipsilateral				
	500	1000	250	2000	4000
dB HL	95#	100#			
	Contralateral				
	250	500	1000	2000	4000
dB HL					



کلینیک گوش تهران

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 Tehran
 021-88205808

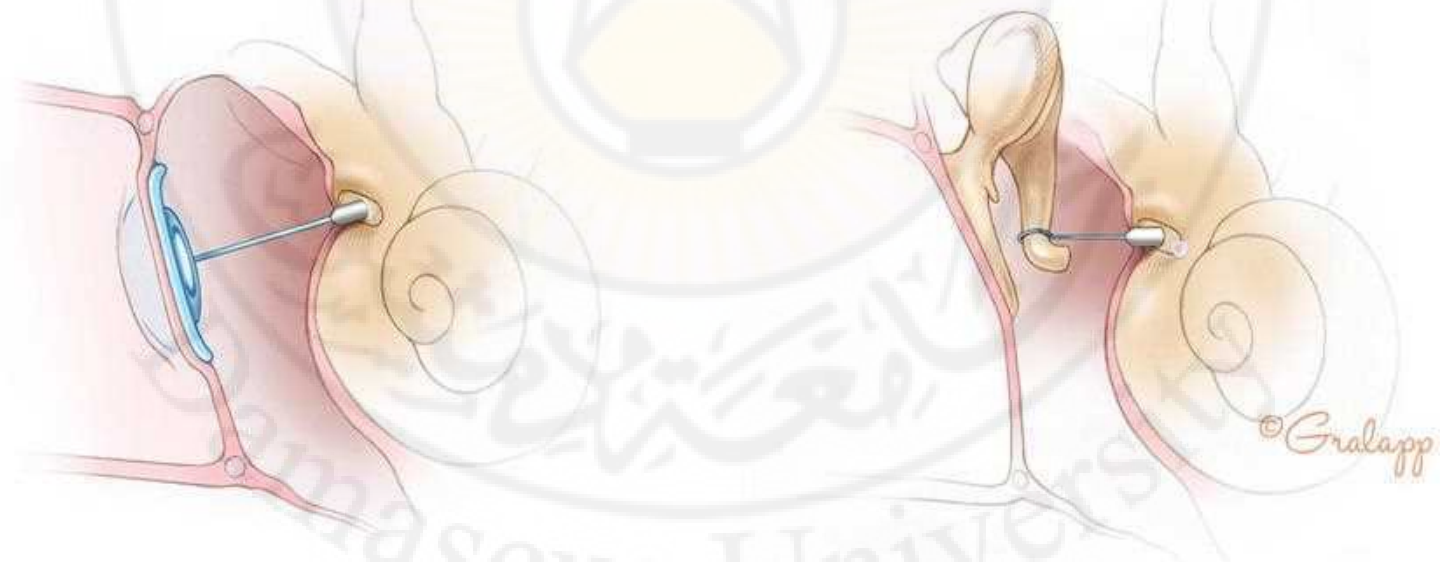
Person ID: [Redacted]
 First name: [Redacted]
 Last name: [Redacted]
 Birth date: [Redacted]
 Gender: Male





Partial Ossicular Chain Reconstruction (PORP)

Crutch and Cup



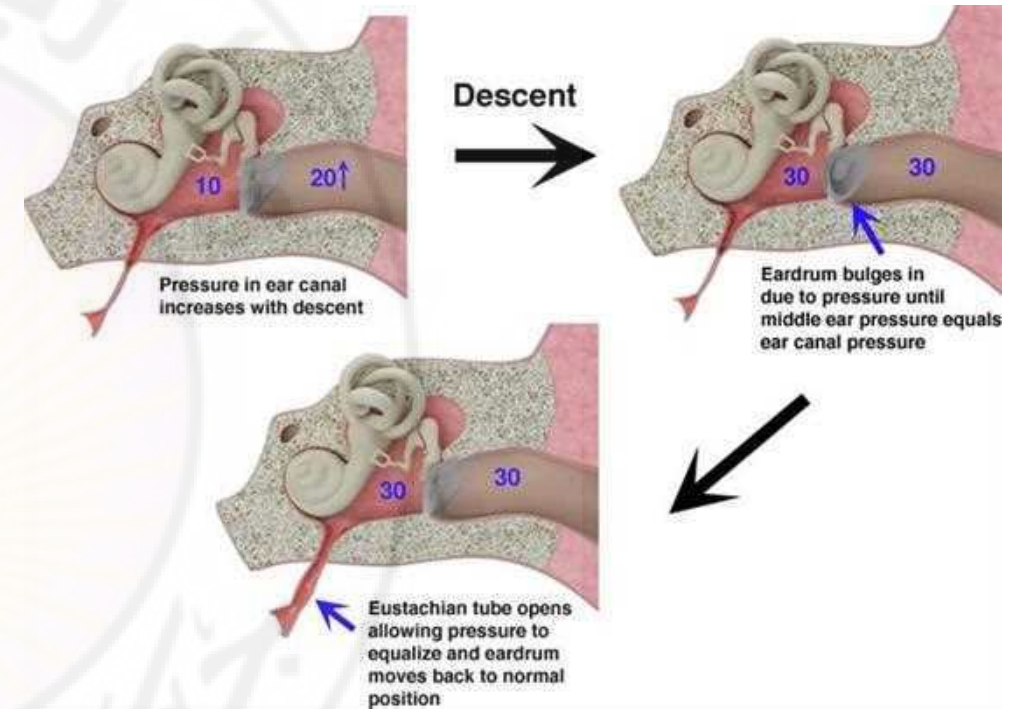
Total Ossicular Chain Reconstruction (TORP)

Stapes Replacement

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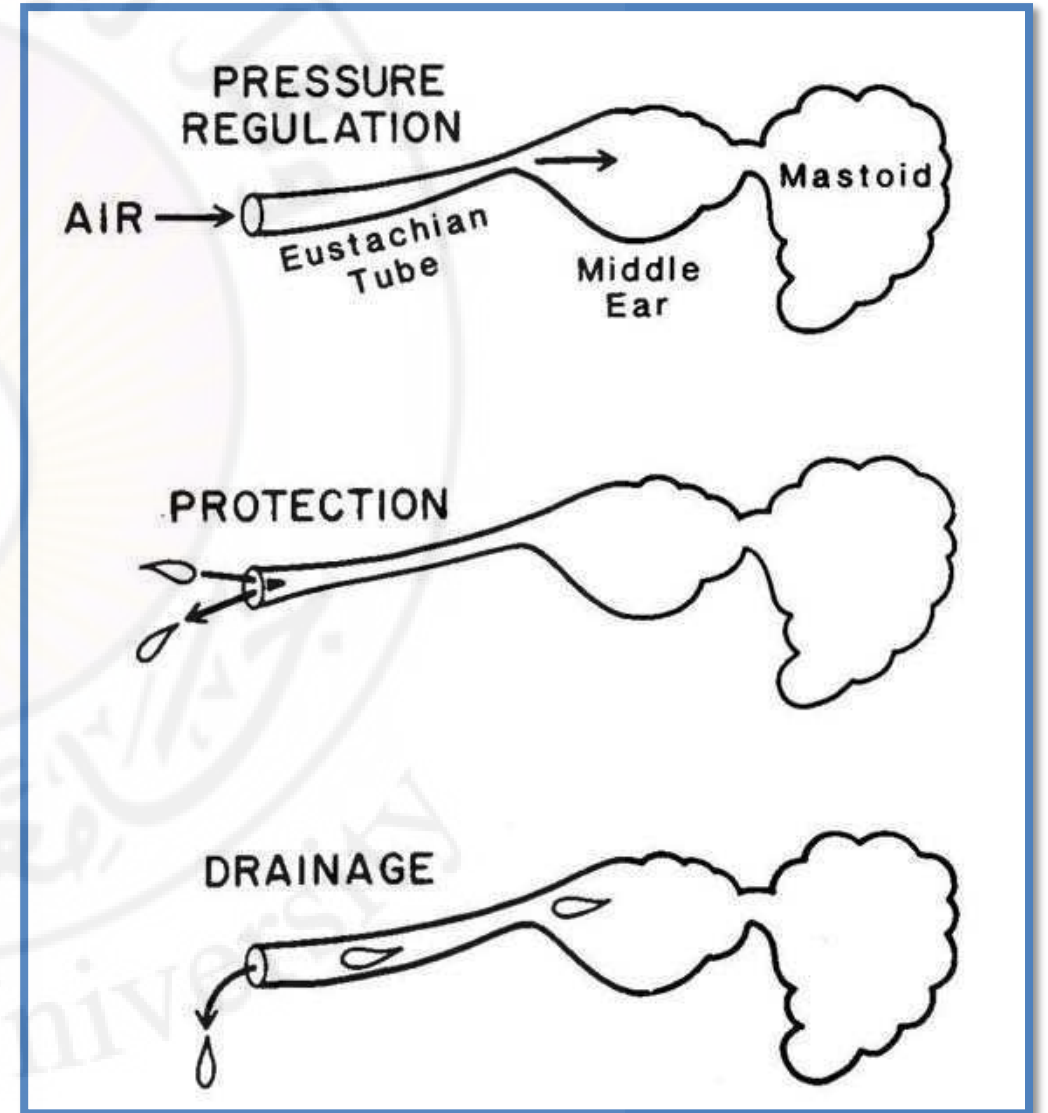
Barotrauma

- Another form of insult to the middle ear results from **barotrauma (aerotitis)**, or trauma related to a sudden, marked change in atmospheric pressure.
- This often occurs when an airplane descends from altitude too fast without proper airplane cabin pressure equalization or when a diver ascends too rapidly in the water (scuba diving).
- It can also occur under more normal ascending and descending conditions if a person's **Eustachian tube** is not providing adequate pressure equalization.
- Under both circumstances, a severe negative air pressure is created in the middle-ear space due to failure of the Eustachian tube to open adequately.



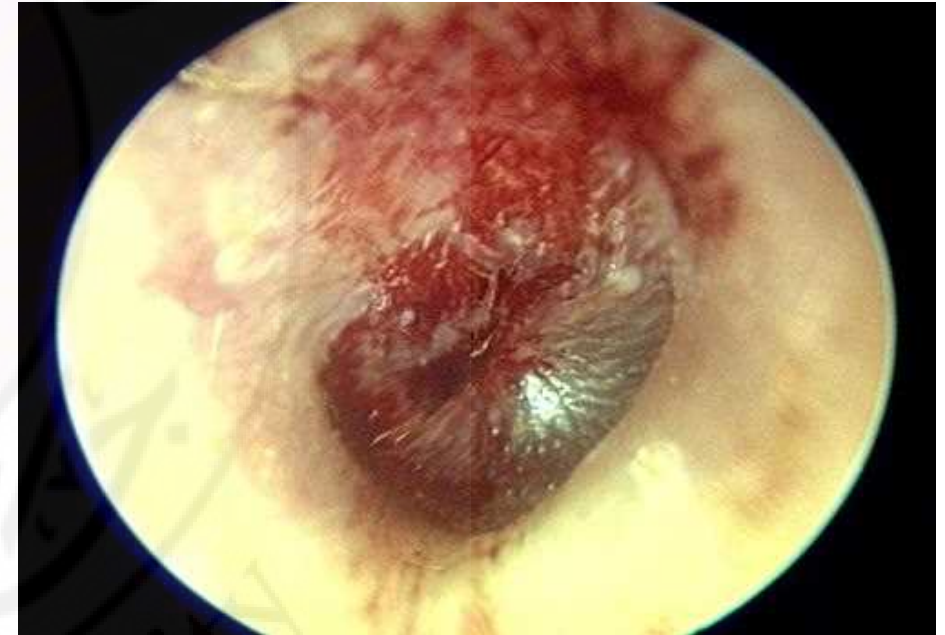
ET Function

1. When there are changes in atmospheric pressure, the Eustachian tube is required to equalize pressure in the middle ear.
2. Normally, ET is closed to protect the middle ear
3. The tube also drains mucus from the middle ear because of its ciliated epithelium and aerates the middle ear.



Barotrauma

- If air pressure changes are sudden and intense, the tympanic membrane may rupture. If not, it is stretched medially, followed by increased blood flow, swelling, and bruising of the mucosal lining of the middle ear.
- Effusion then forms within the middle ear from the traumatized tissue.
- Barotrauma can cause TM perforation, CHL, and, in rare cases, a fistula of the oval window.



Eustachian Tube Function test

- **Non-Perforated TM**

- Valsalva Test
- Toynbee Test
- Inflation / Deflation test

- **Perforated TM**

- Inflation Test



Valsalva Test

- The Valsalva test (Bluestone, 1975) introduces positive pressure into the middle ear via the ET using the classic Valsalva maneuver.

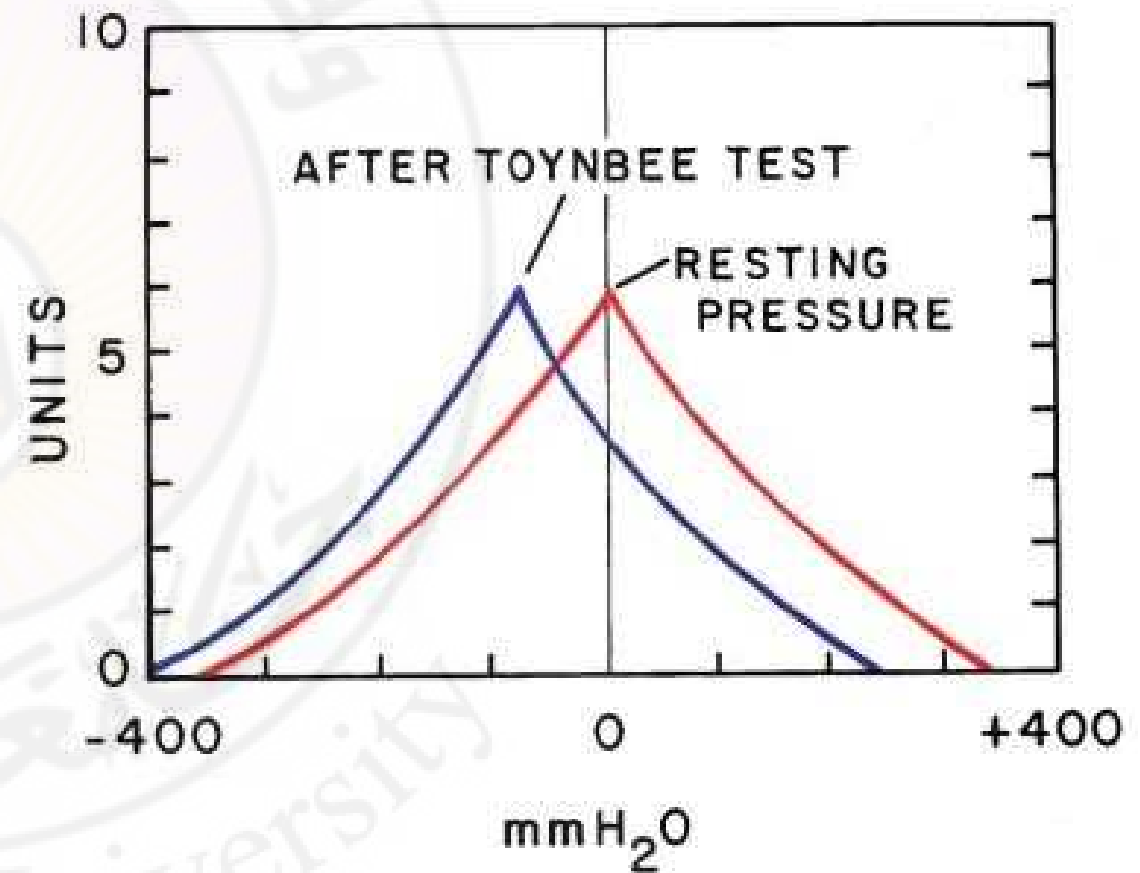
1. *A pretest tympanogram is recorded,*
2. *the patient is instructed to perform the Valsalva maneuver by holding the nose and gently blowing air into the posterior nasopharynx.*
3. *Then, a posttest tympanogram is recorded.*

- Tubal opening is indicated by a [positive shift in TPP.](#)

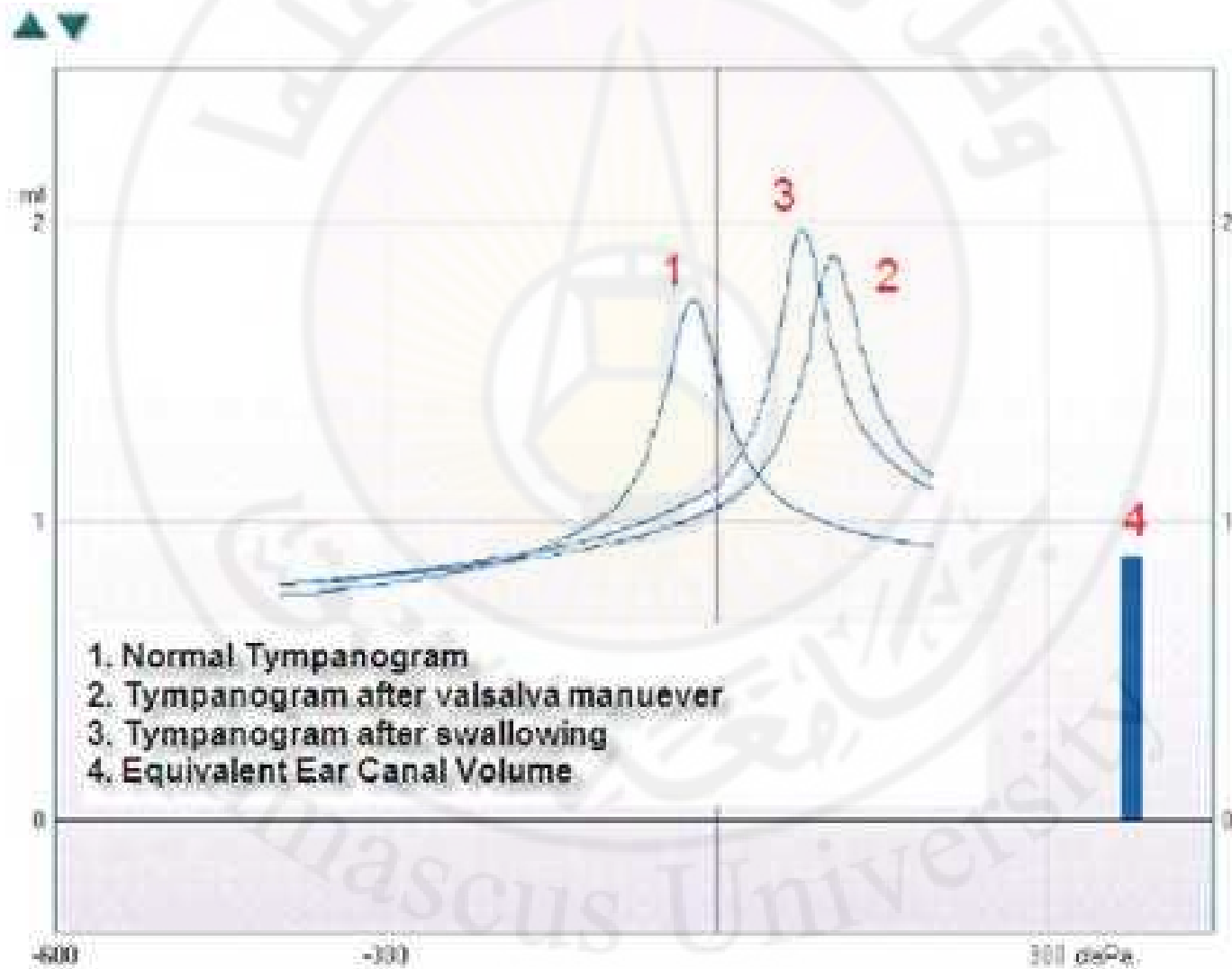


Toynbee Test

- In the Toynbee maneuver the patient is instructed to swallow while holding his nose closed (by pinching his nostrils).
- The Toynbee maneuver is typically expected to make the middle ear pressure more **negative**, but it actually causes *either* a positive *or* negative shift in middle ear pressure.
- The Toynbee test follows the same procedures used for the Valsalva test, except that the Toynbee maneuver is used.

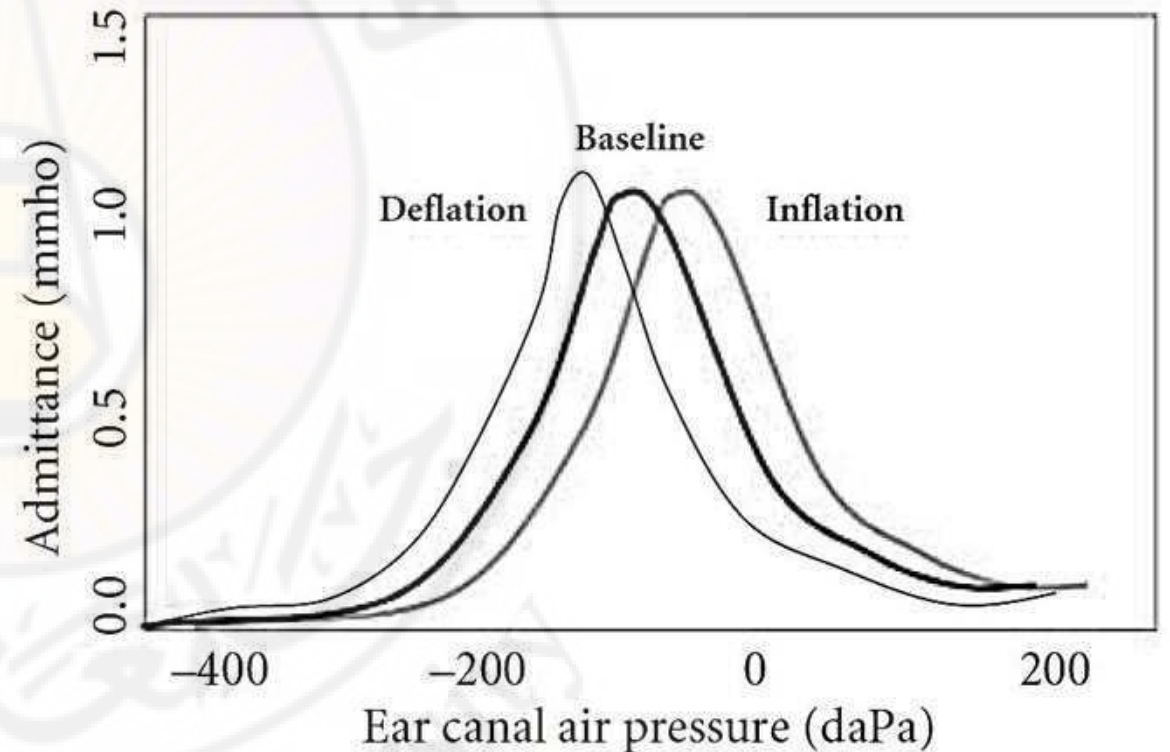


Suggested 3-stage ETF testing



Inflation / Deflation test

- The inflation–deflation test uses high positive pressure (inflation) or negative pressure (deflation) introduced into the ear canal using the tympanometers (± 400 daPa) while the patient is asked to swallow several times.
- Pre- and posttest tympanograms are recorded.
- Tubal opening is indicated by a shift in the pressure peak in the opposite direction of applied pressure.



Inflation test for Perforated TM

- Put the positive pressure in the ear canal, with sealing of the probe tip.
- Patient swallow several times and decreasing the pressure is an indicator of normal ET function.



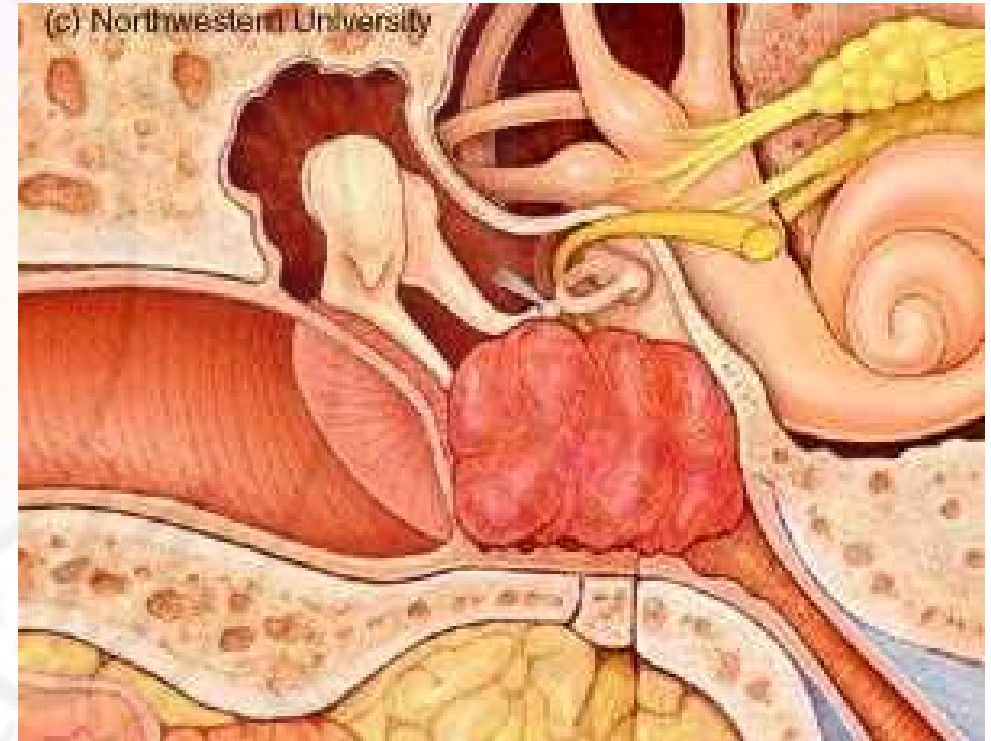
# 1	255 daPa	222 daPa
# 2	216 daPa	95 daPa
# 3	83 daPa	68 daPa

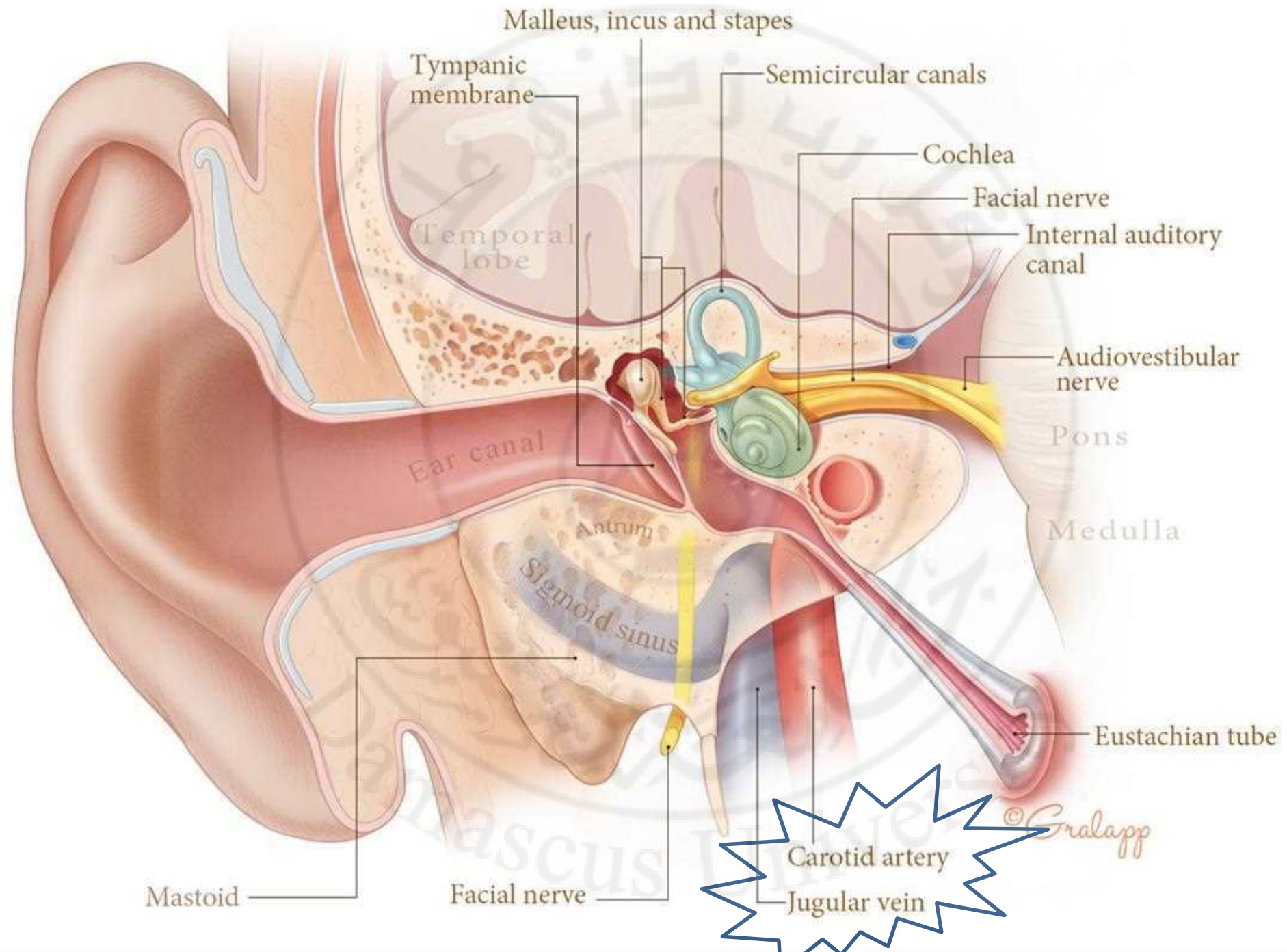
Table 4-5. Normative Values (in daPa) for Eustachian Tube Function Tests for 24 Adults with Normal Middle Ear Function

<i>Test</i>	<i>Mean</i>	<i>Standard Error</i>	<i>95% Confidence Interval</i>
Valsalva			
Baseline	9	1	7-11
Posttest	72	8	55-87
Shift	63	8	47-79
Toynbee			
Baseline	9	1	7-11
Posttest	-6	5	-17-5
Shift	-15	5	-26 to -4
Inflation			
Baseline	8	1	6-10
Posttest	-3	1	-6-0
Shift	-11	1	-13 to -8
Deflation			
Baseline	8	1	6-10
Posttest	23	2	20-26
Shift	15	2	12-18

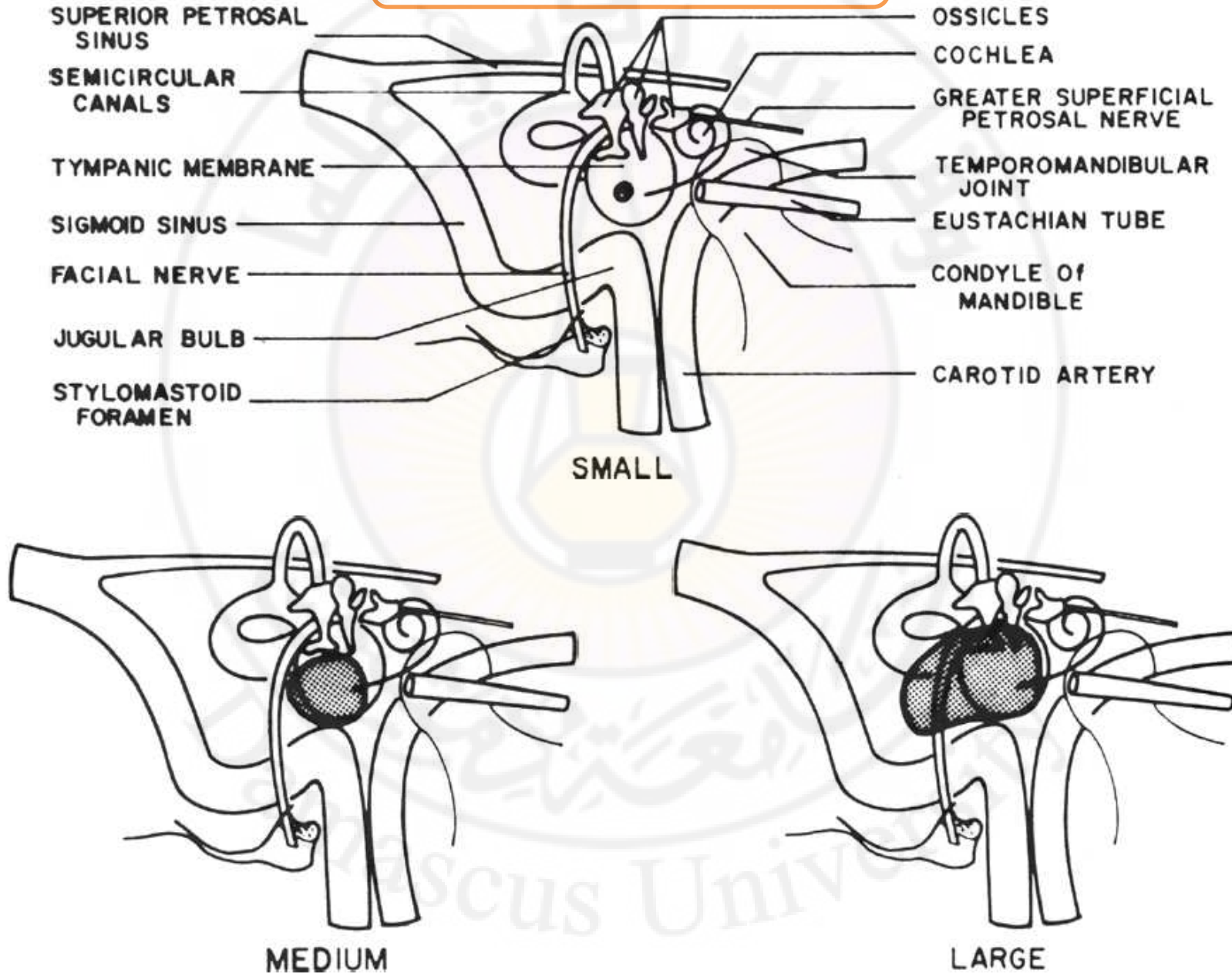
Glomus tumor (Tympanic Paraganglioma)

- Tumors may also occur within the middle ear. One neoplasm commonly found in the middle ear is a **glomus tumor, or glomus tympanicum**.
- Glomus tumors, are slow-growing, usually benign tumors in the carotid arteries (major blood vessels in neck), the middle ear or the area below the middle ear (jugular bulb), and can result in significant conductive hearing loss.
- They are called **glomus tympanicum tumors** when they develop within the middle ear cavity and **glomus jugulare tumors** when they develop from the crest of the jugular bulb under the floor of the middle ear.

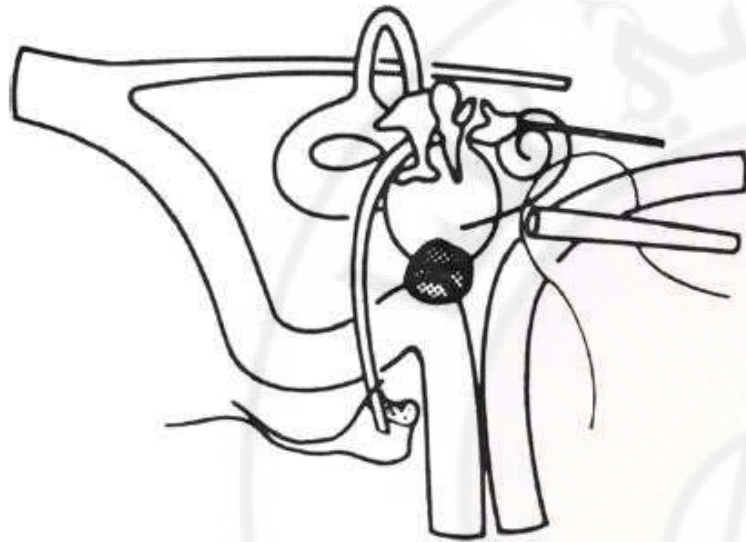




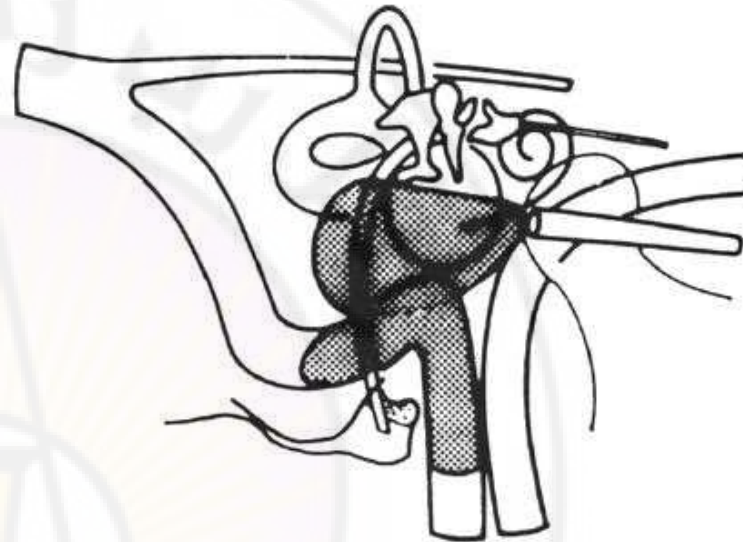
GLOMUS TYMPANICUM



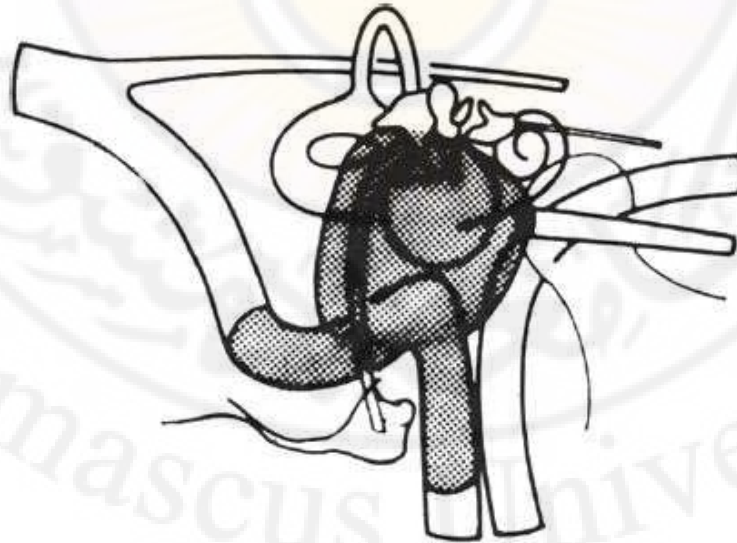
GLOMUS JUGULARE



SMALL

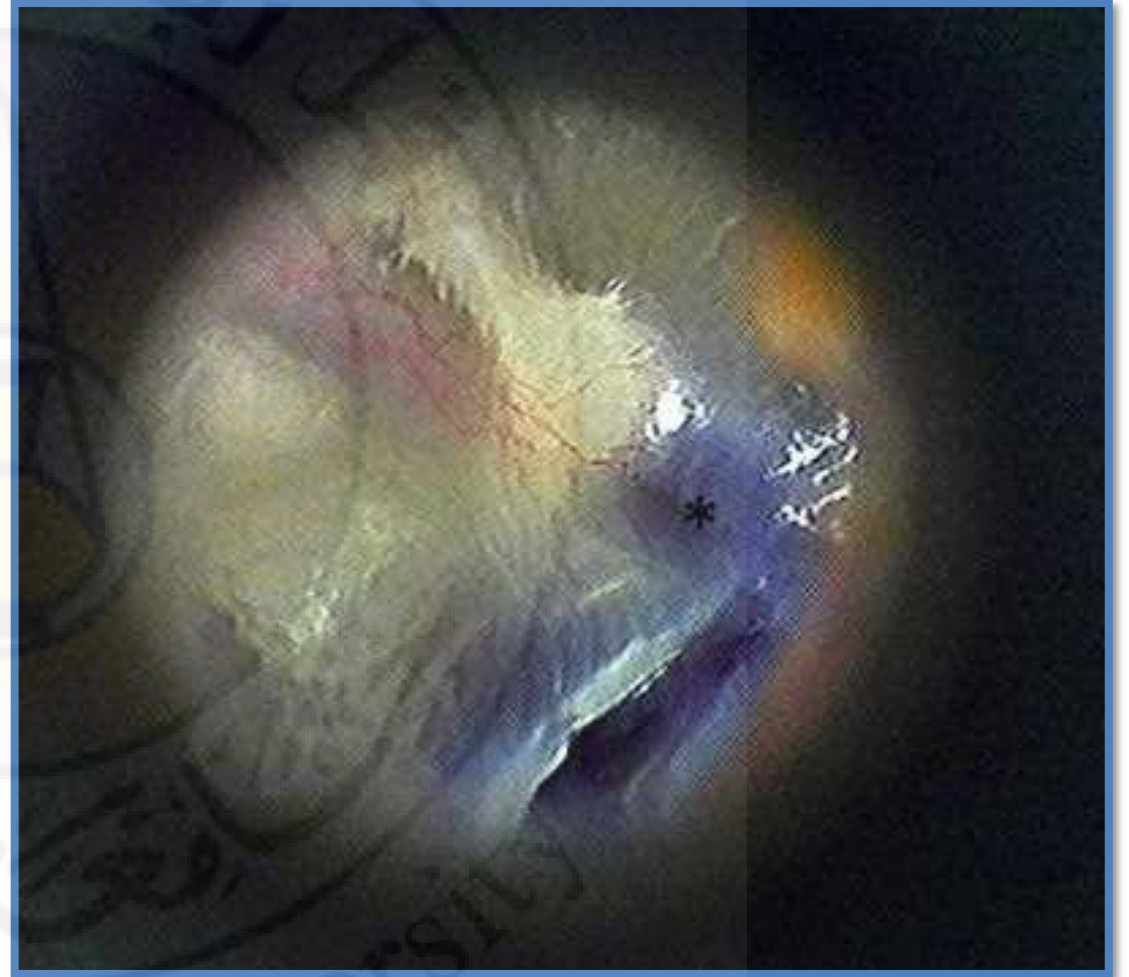


MEDIUM



LARGE

Jugular bulb in the middle ear space



Glomus tumor

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- They are usually **unilateral** and are more common in **women** than in men by a ratio of roughly 3 to 1.
- The most common characteristics of patients with glomus tumors are **pulsing tinnitus** that is synchronized to the patient's vascular pulse and a **conductive or mixed hearing loss**, which is unilateral in a wide majority of cases.
- **Bleeding** from the ear occurs in some cases, and patients may also experience a feeling of **pressure** in the ear. Other complaints may also occur but are highly variable.
- Glomus tumors are usually seen as a **reddish mass behind the tympanic membrane**, often with a "**rising sun**" appearance one might also see it pulsating, and possibly causing the eardrum to bulge.
- Glomus tumors are treated surgically.



Glomus tumor & Audiology

Oscillations in tympanogram + ABG + bulged TM + **Pulsatile tinnitus** + Bleeding

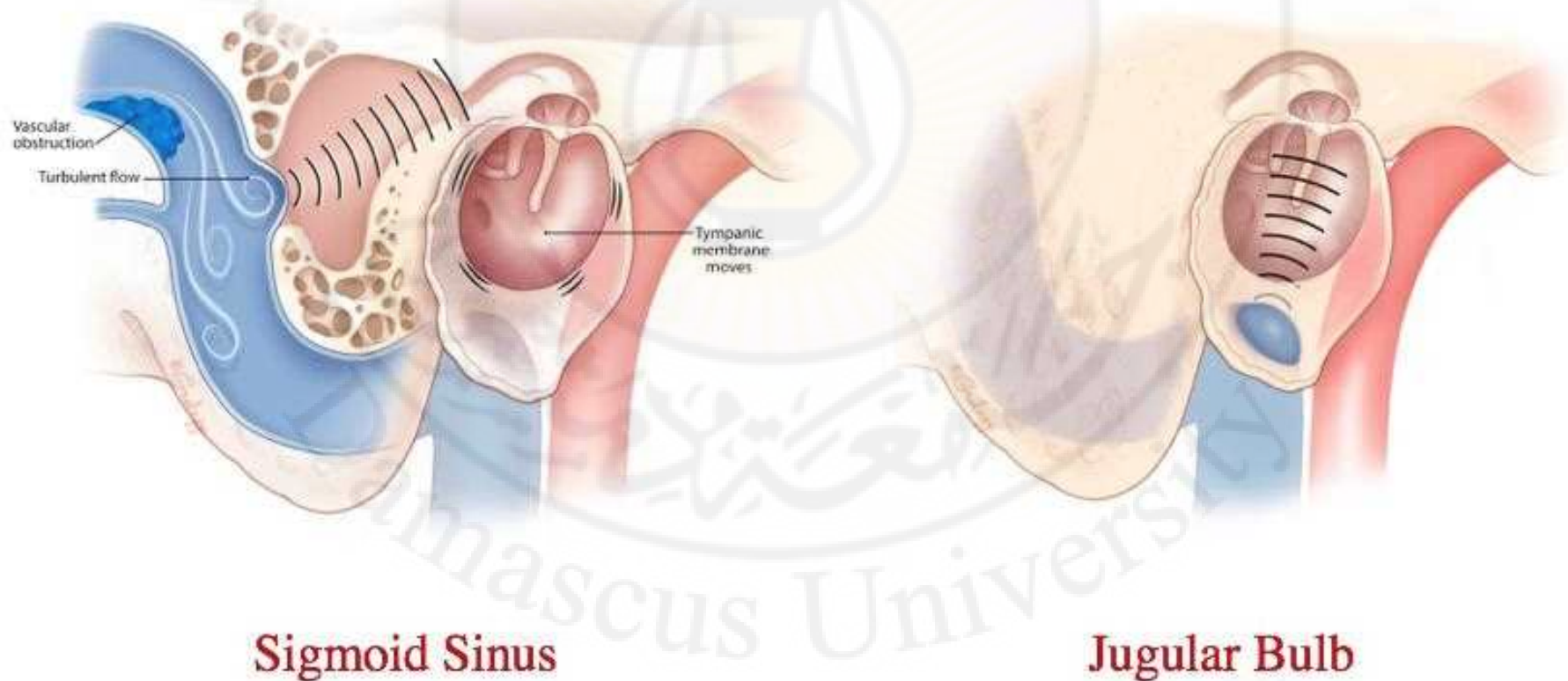


TABLE 1. Symptoms presented by glomus tumor patients at first consultation.

<i>Symptom</i>	<i>Jugulare</i> (%)	<i>Tympanicum</i> (%)
Tinnitus (N = 40)	90	85
Pulsatile	70	55
Nonpulsatile	20	30
Hearing loss (N = 39)	79	75
Dizziness (N = 39)	32	55
Ear pain (N = 39)	32	30
Feeling of ear pressure (N = 39)	21	25
Bleeding from ear (N = 39)	11	10
Hoarseness (N = 39)	11	0

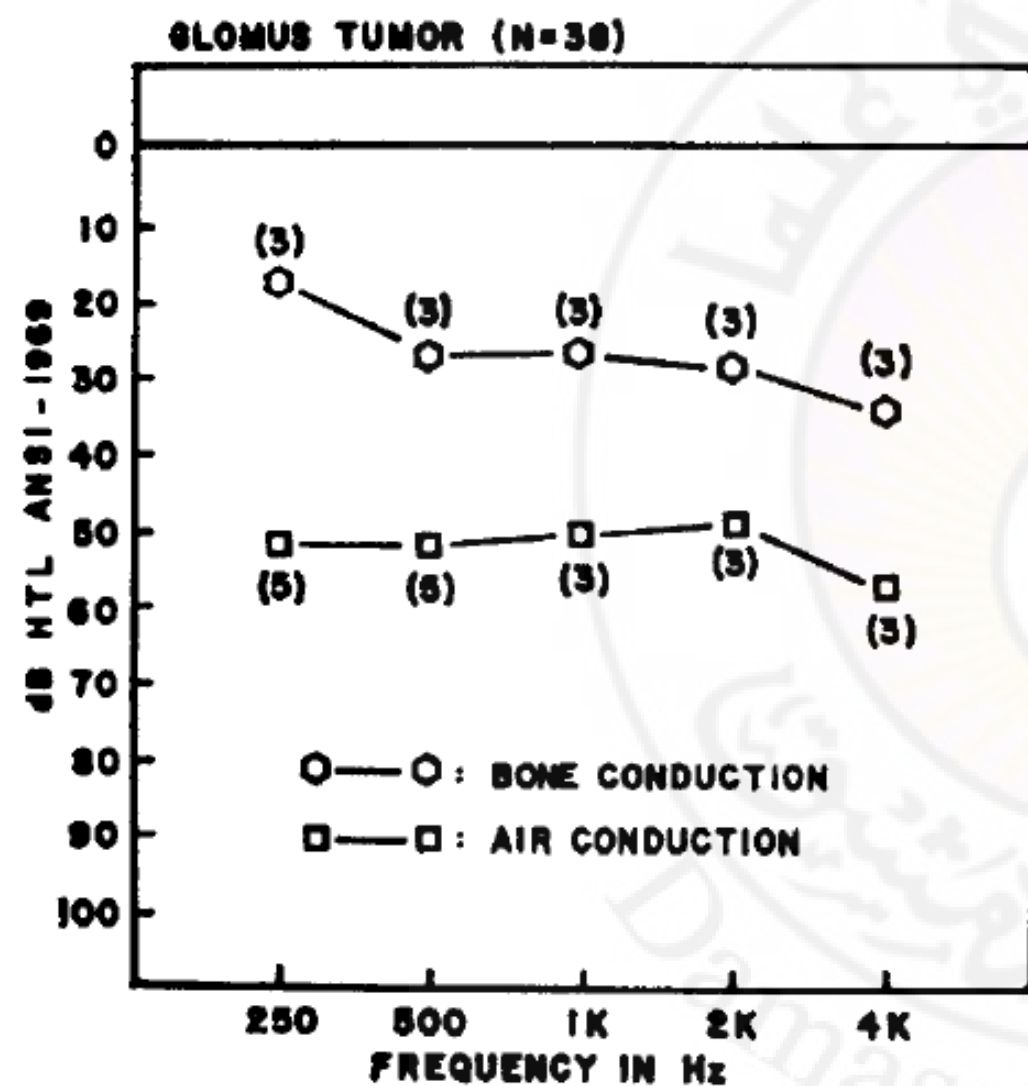


Figure 3. Mean air and bone conduction thresholds for the entire glomus tumor series (N = 38). The numbers in parentheses indicate the incidence of no response at the limits of the audiometer at each frequency. When necessary, audiometer intensity limits are included in determination of mean thresholds.

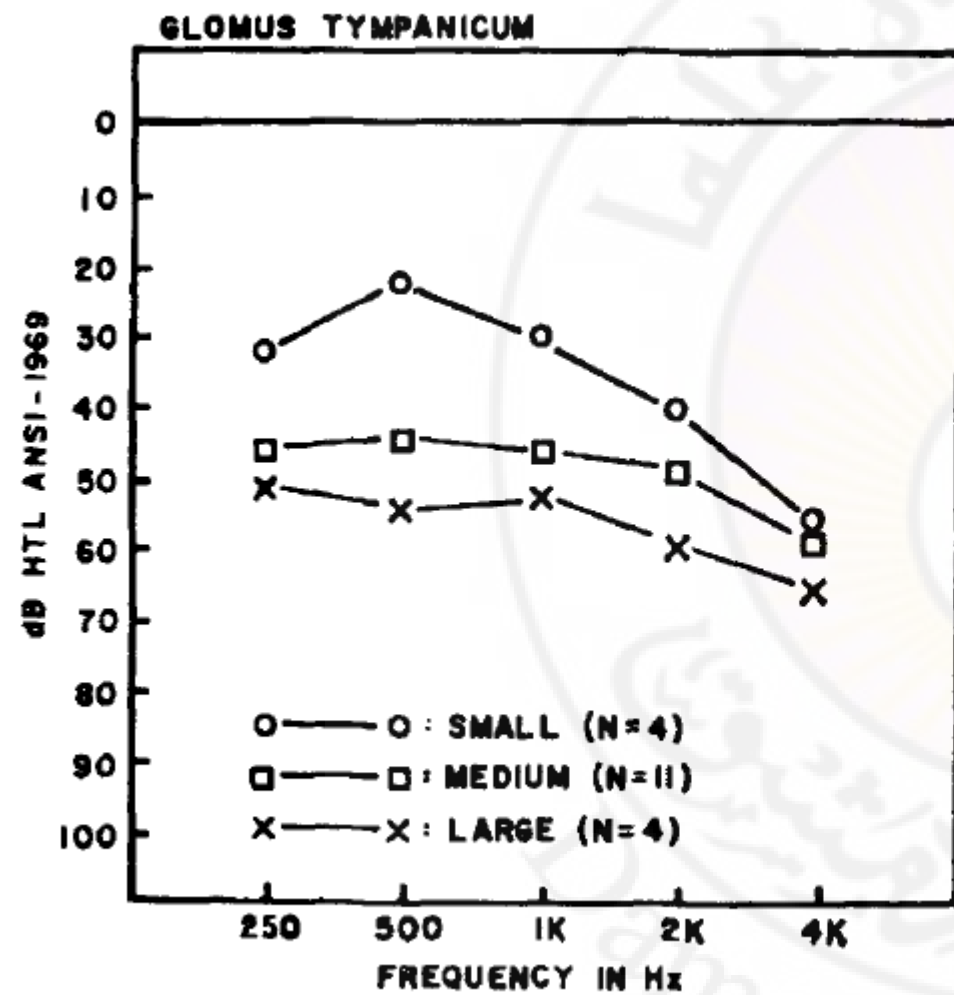
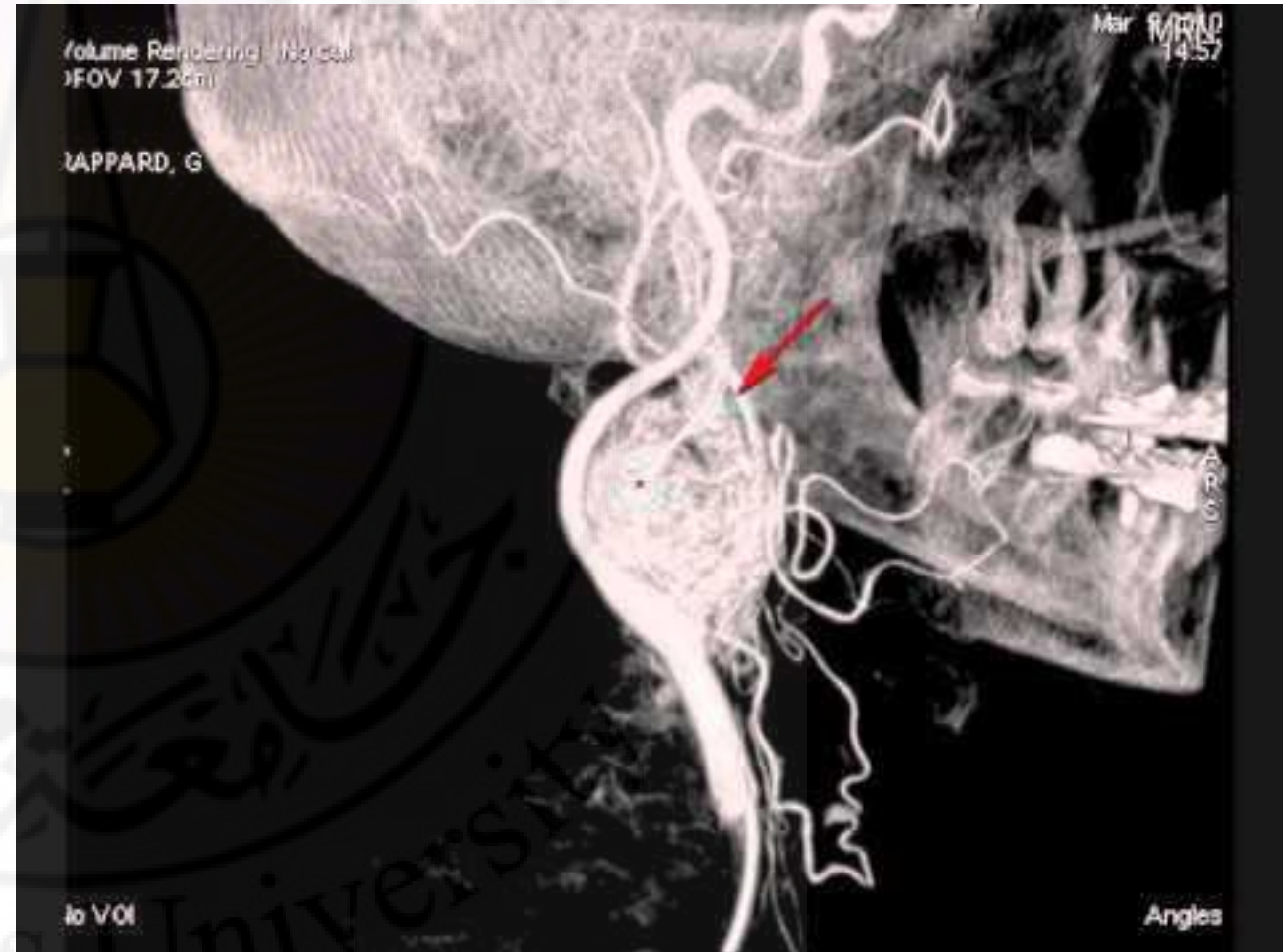


Figure 5. Mean air conduction thresholds for the glomus tympanicum tumor as a function of tumor size.

Glomus tumor & Vascular Pulsing

Although most **tympanograms** are smooth, they sometimes have regular ripples or undulations that are synchronized with the patient's pulse, which is their origin .

Medical referral is indicated when vascular pulsing is present on the tympanogram because it tends to occur in patients with glomus **jugular tumors**.



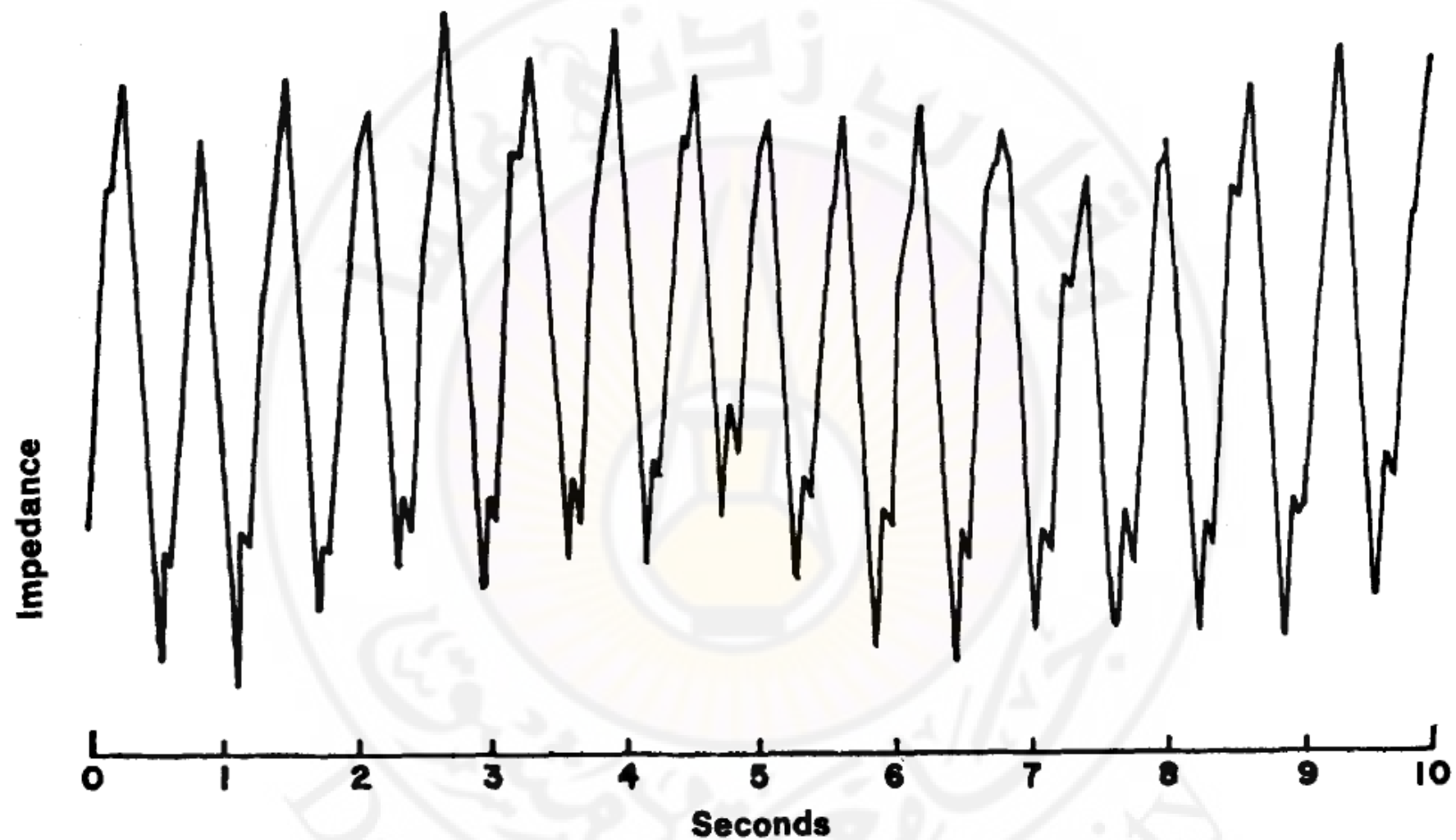
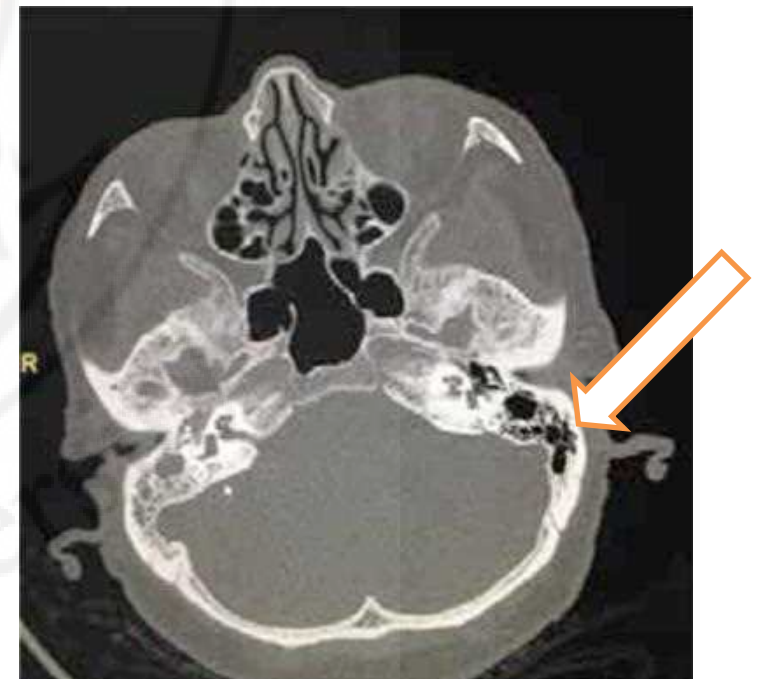
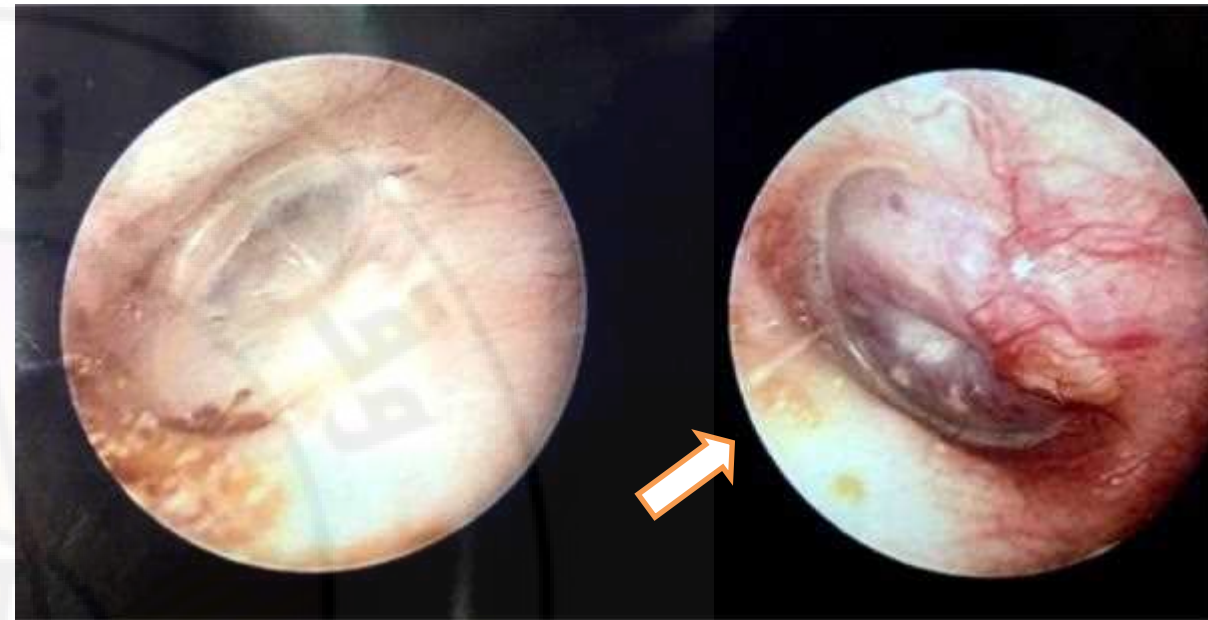
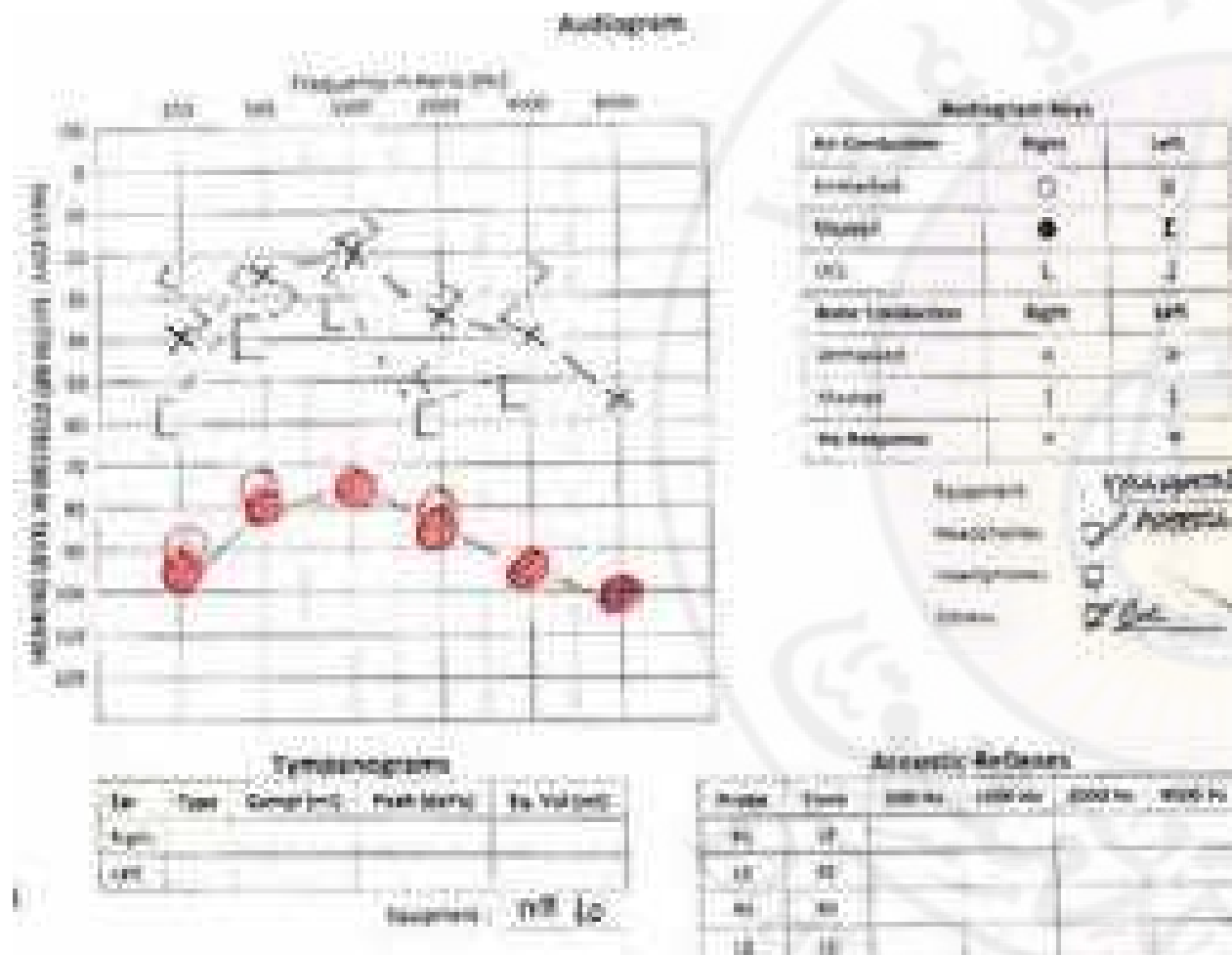


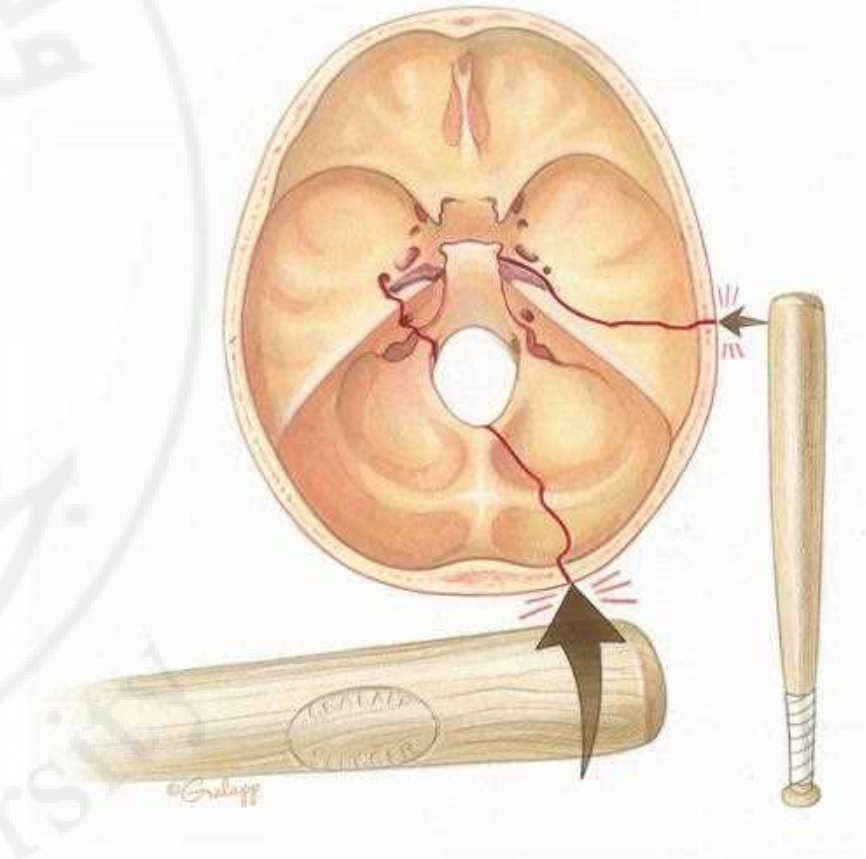
Figure 6. Electroacoustic impedance variation in a 31-year-old female with a diagnosed glomus jugulare tumor expanding into the middle ear space. Cyclical oscillations in impedance are seen to occur in synchrony with the pulse.

Case Example

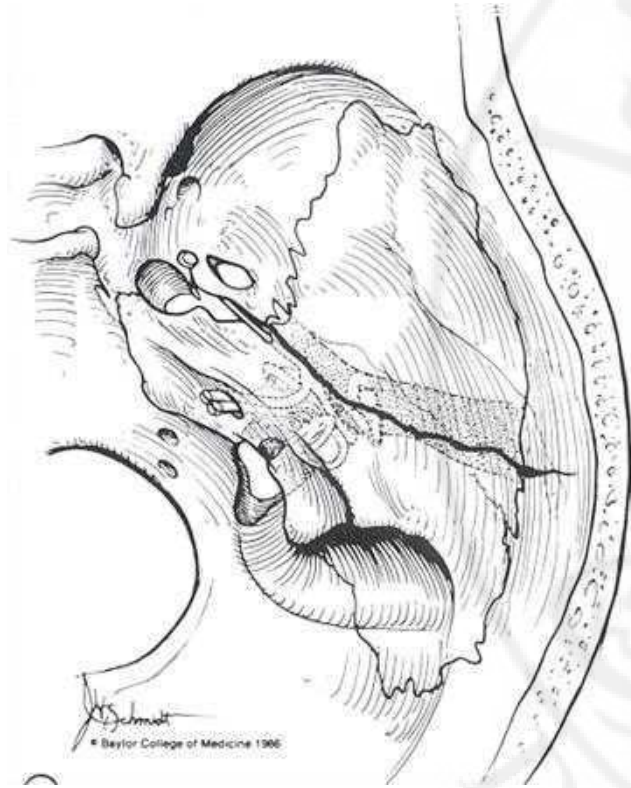


Fracture of temporal bone

- The temporal bone is the thickest bone in the body, requiring significant force to fracture (**850 kg of lateral force** in one study).
- The temporal bone houses portions of the internal carotid artery and internal jugular vein, cranial nerves (CN) VII-XI, the middle and inner ear structures, and forms a portion of the middle cranial fossa. Such high-velocity injury to these crucial structures carries the risk of significant morbidity and mortality.
- Approximately one-third of temporal bone fractures in the general population result from motor vehicle accidents, assaults, and falls, and an additional ~25% are attributed to sports injuries, gunshot wounds, bicycle accidents, and other miscellaneous causes.
- **Males** are at least 3-times more likely than females to suffer temporal bone fractures. Temporal bone fractures are typically **unilateral**.



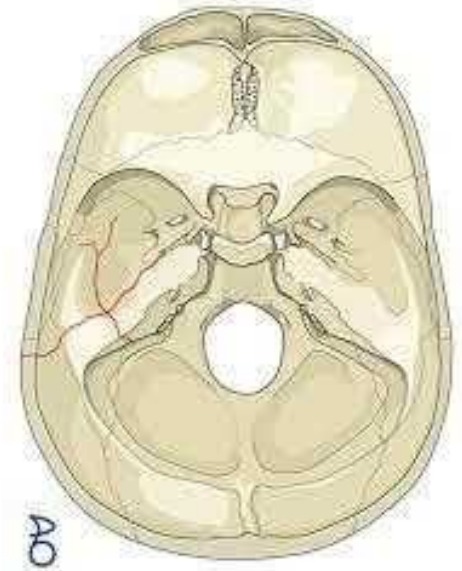
Fracture of temporal bone - Classifications



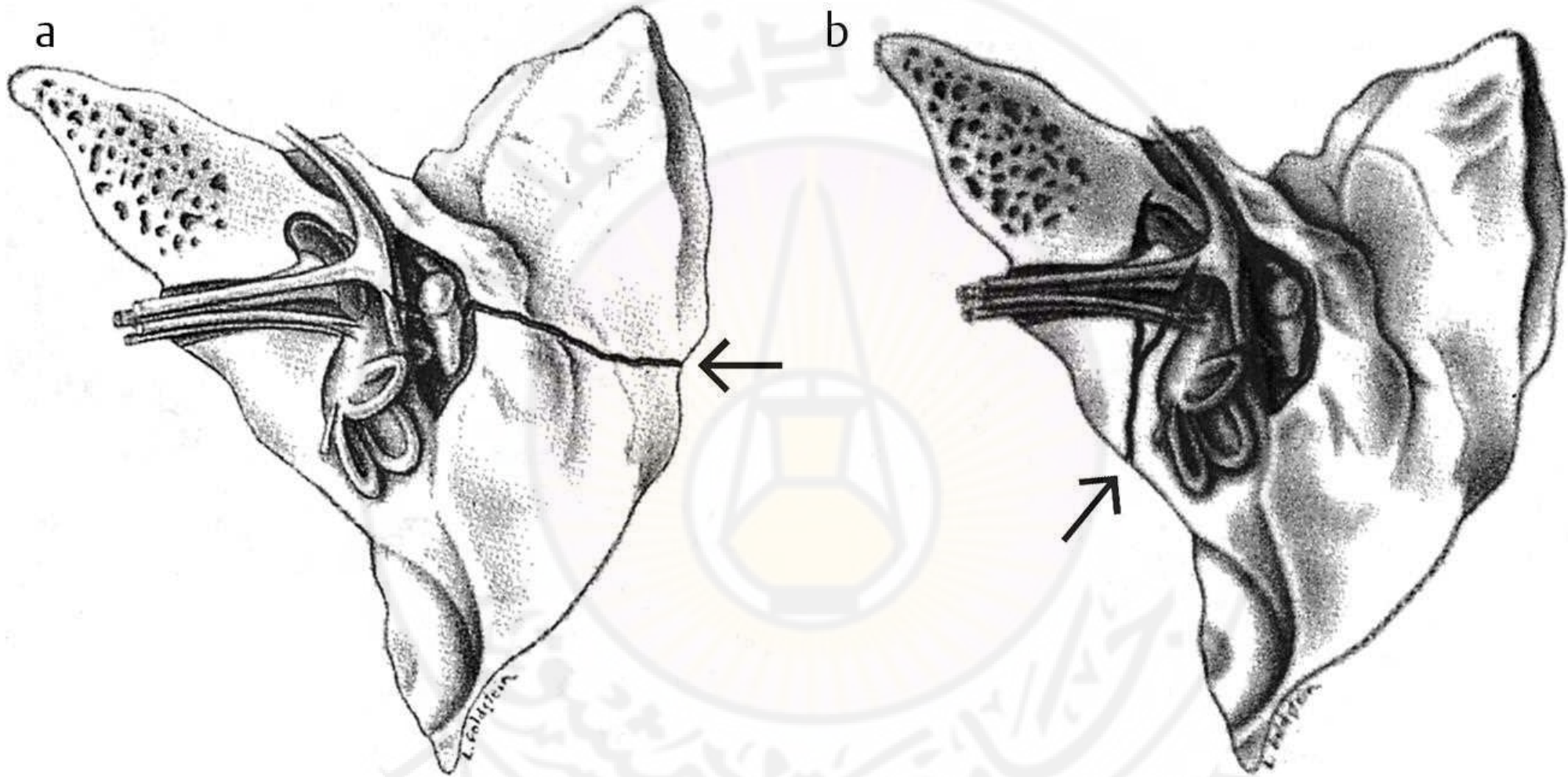
Longitudinal fractures



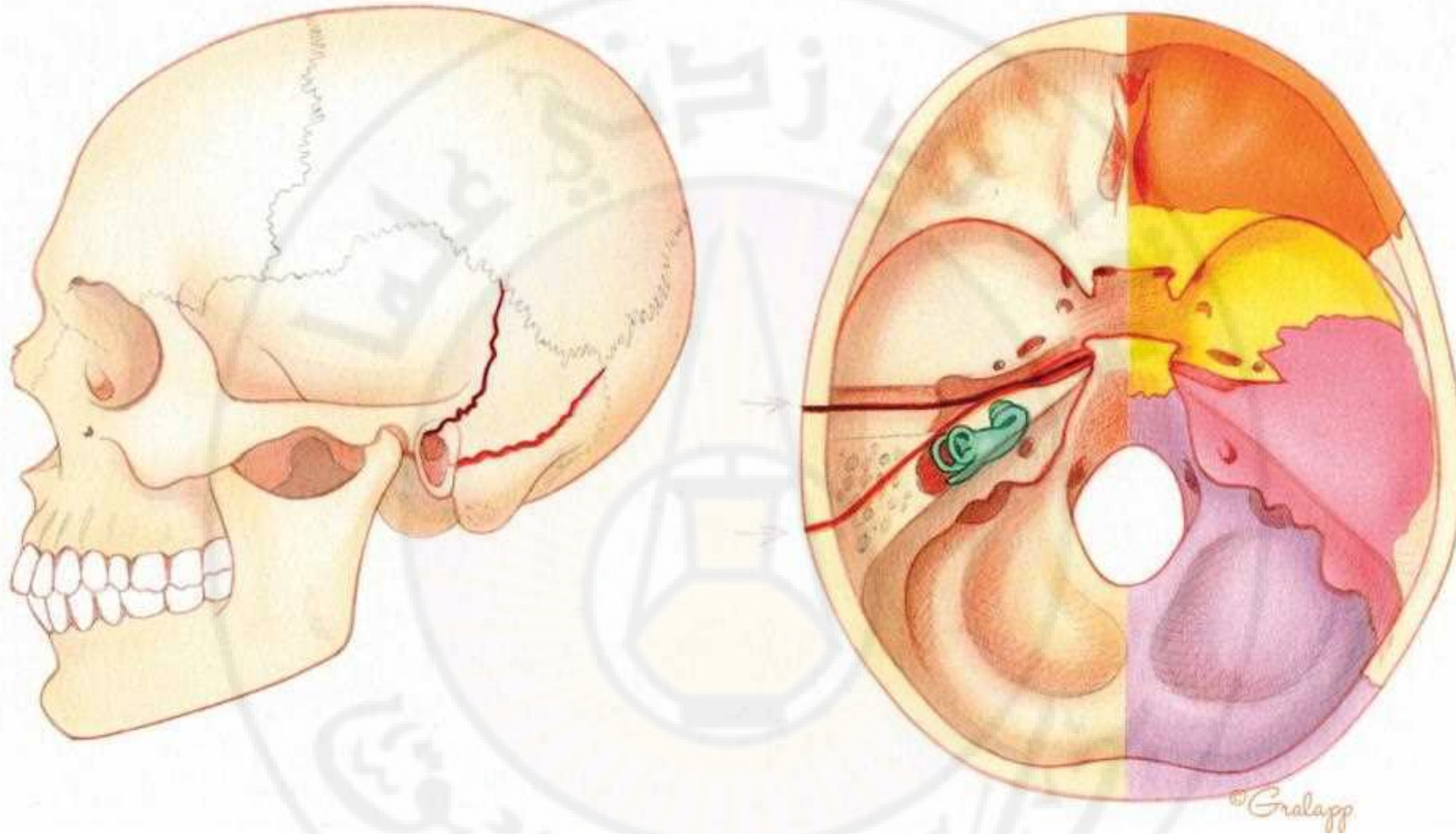
Transverse fractures



Mixed fractures

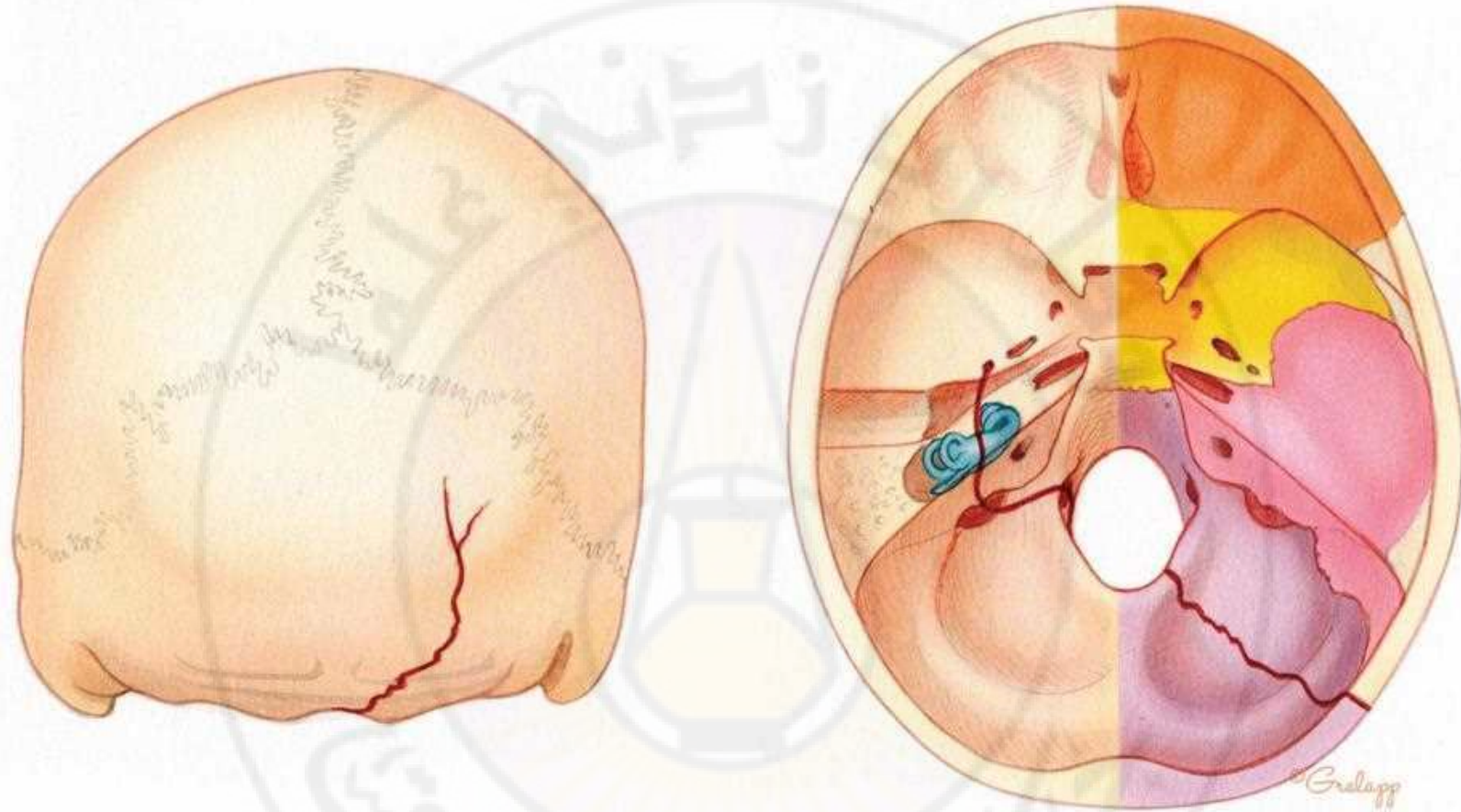


Schematic representations of (a) longitudinal and (b) transverse fractures of the temporal bone and how they relate to ear structures.



fractures that course parallel to the long axis of the petrous ridge are considered **"longitudinal"**

"Longitudinal fractures made up approximately 70-90% of all temporal bone fractures"



fractures that course perpendicular to the long axis of the petrous ridge are considered **"transverse"**

"transverse fractures made up approximately 10-30% of all temporal bone fractures"

Fracture of temporal bone - symptoms

- **Hearing Loss & tinnitus**

- 71% of patients with Temporal Bone Trauma have hearing loss
- TM Perforations
- CHL > 40db suspicion for ossicular discontinuity

- **Longitudinal Fractures**

- Conductive or mixed hearing loss
- 80% of CHL resolve spontaneously

- **Transverse Fractures**

- Sensorineural hearing loss
- Less likely to improve

- **Dizziness & Vertigo**

- **CSF Otorrhea and Rhinorrhea**

- **Facial Nerve Injuries**



The background of the slide features a large, faint watermark of the Damascus University logo. The logo is circular, with the university's name in Arabic 'جامعة دمشق' at the top and 'Damascus University' at the bottom. In the center is a stylized yellow and orange emblem resembling a minaret or a sunburst.

Sensory Hearing Disorders

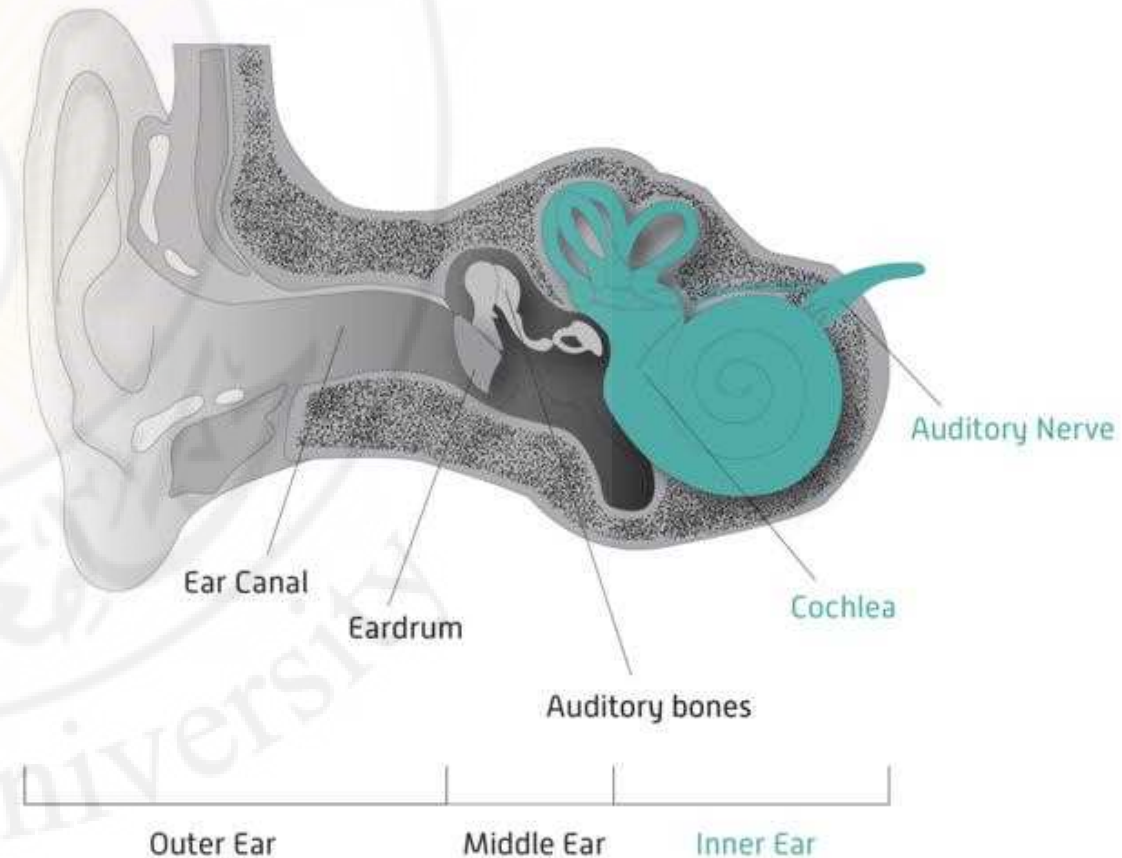
- Congenital and Inherited Sensory Hearing Disorders
- Acquired Sensory Hearing Disorders

Sensory (Cochlear) Disorders: Evaluative Goals

- When an **otologist** refers a patient with a suspected cochlear disorder, the physician is interested in the answers to several questions:
 - Is there a hearing loss and what is the extent of it?
 - Is the loss truly of a cochlear nature or is there also a conductive component?
 - Is the loss truly of a cochlear nature or is it retrocochlear?
 - Is the loss fluctuating or stable?
 - Could the loss be due to a treatable condition such as endolymphatic hydrops or autoimmune disorder?
- For the **audiologist**, there are two primary goals in the assessment of cochlear disorders:
 1. Determine whether a middle-ear disorder is contributing to the problem
 2. Determine the degree and type of hearing loss
 3. Scrutinize the audiologic findings for any evidence of retrocochlear disorder

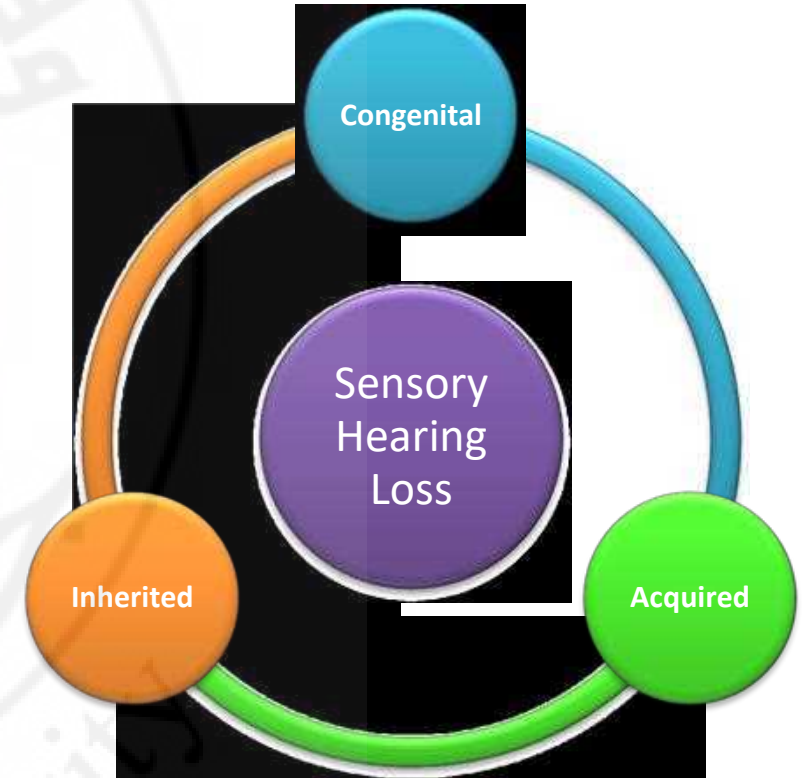
Sensory (Cochlear) Disorders: Test Strategies

- Immittance Audiometry (Tympanometry & Acoustic reflex measures)
- Pure-Tone Audiometry
- Speech Audiometry
- Otoacoustic Emissions
- Auditory Evoked Potentials



Sensory Hearing Disorders

- Sensory hearing disorders result from impaired cochlear function.
- **Congenital disorders** are present at birth and result from structural and functional defects of the cochlea, secondary to embryologic malformation. Congenital disorders are a common form of sensorineural hearing loss in infancy.
- **Inherited hearing disorders** can be present at birth or can manifest in adulthood.
- **Acquired sensory auditory disorders** occur later in life and are caused by excessive noise exposure, trauma, infections, ototoxicity, endolymphatic hydrops, presbycusis, and other influences.

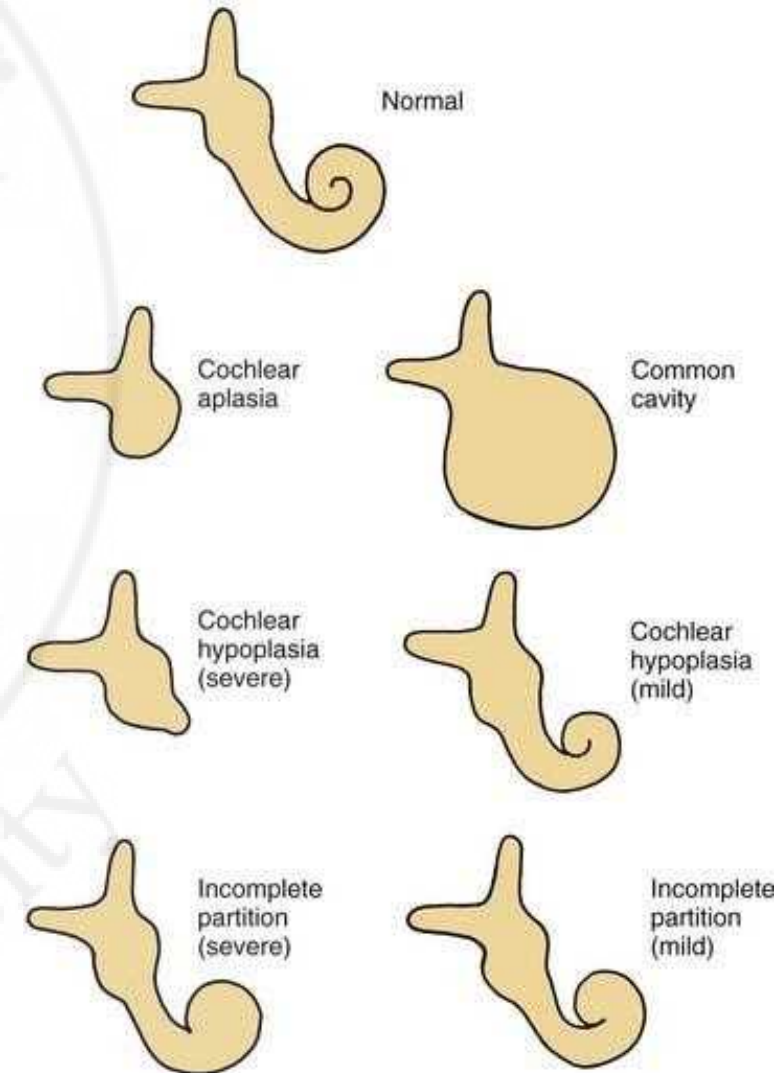


Congenital and Inherited Sensory Hearing Disorders

- Congenital sensory hearing disorders are present at birth. There are a number of causes, both **exogenous and endogenous**, of congenital disorders.
- **Exogenous conditions** are those that are not necessarily intrinsic to the genetic makeup of an individual. The *teratogenic effects* on the developing infant of certain maternal infections or drug ingestion are examples.
- **Endogenous conditions** are those that are *inherited*.
- The actual change in cochlear structure or function may be identical, whether the cause is exogenous or endogenous.
- **In this class**, we will first review the actual **anomalies of the inner ear**, followed by a summary of **teratogenic causes**. Endogenous causes are separated into two categories, those inherited disorders that occur in **syndromes** and those inherited disorders that appear as hearing loss **alone**.

Inner-Ear Anomalies

- Inner-ear malformations occur when development of the membranous and/or bony labyrinth is arrested during fetal development.
- Although in many cases the arrest of development is due to **genetic cause**, some cases are the result of **teratogenic influences** during pregnancy, including viral infections such as rubella, drugs such as thalidomide, and fetal radiation exposure.
- Inner ear malformations can be divided into those in which **both the osseous and membranous labyrinths** are abnormal and those in which **only the membranous labyrinth** is abnormal.
- Anomalies of the bony labyrinth are better understood because they can be readily identified with scanning techniques.



Inner Ear Anomalies

Malformations of both membranous and osseous labyrinths

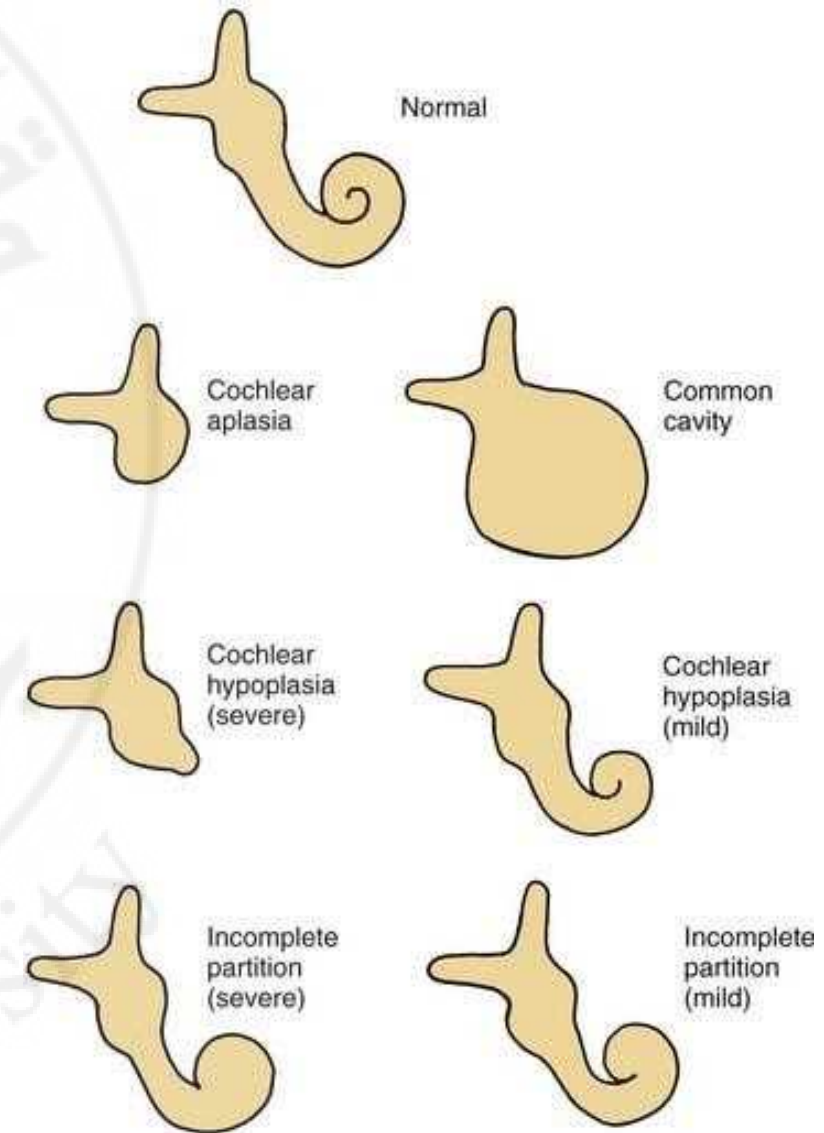
- Complete labyrinthine aplasia (Michel deformity)
- Common-cavity defect
- Cochlear aplasia and hypoplasia
- Mondini defect
- Anomalies of the semicircular canals
- Anomalies of the internal auditory canals
- Anomalies of the cochlear aqueducts
- Large vestibular aqueduct syndrome
- Semicircular canal dehiscence

Malformations limited to the membranous labyrinth

- **Complete membranous labyrinth dysplasia** (Bing-Siebenmann)
- **And two forms of partial dysplasia:**
 - Cochleosaccular dysplasia (Scheibe aplasia)
 - Cochlear basal turn dysplasia (Alexander aplasia)

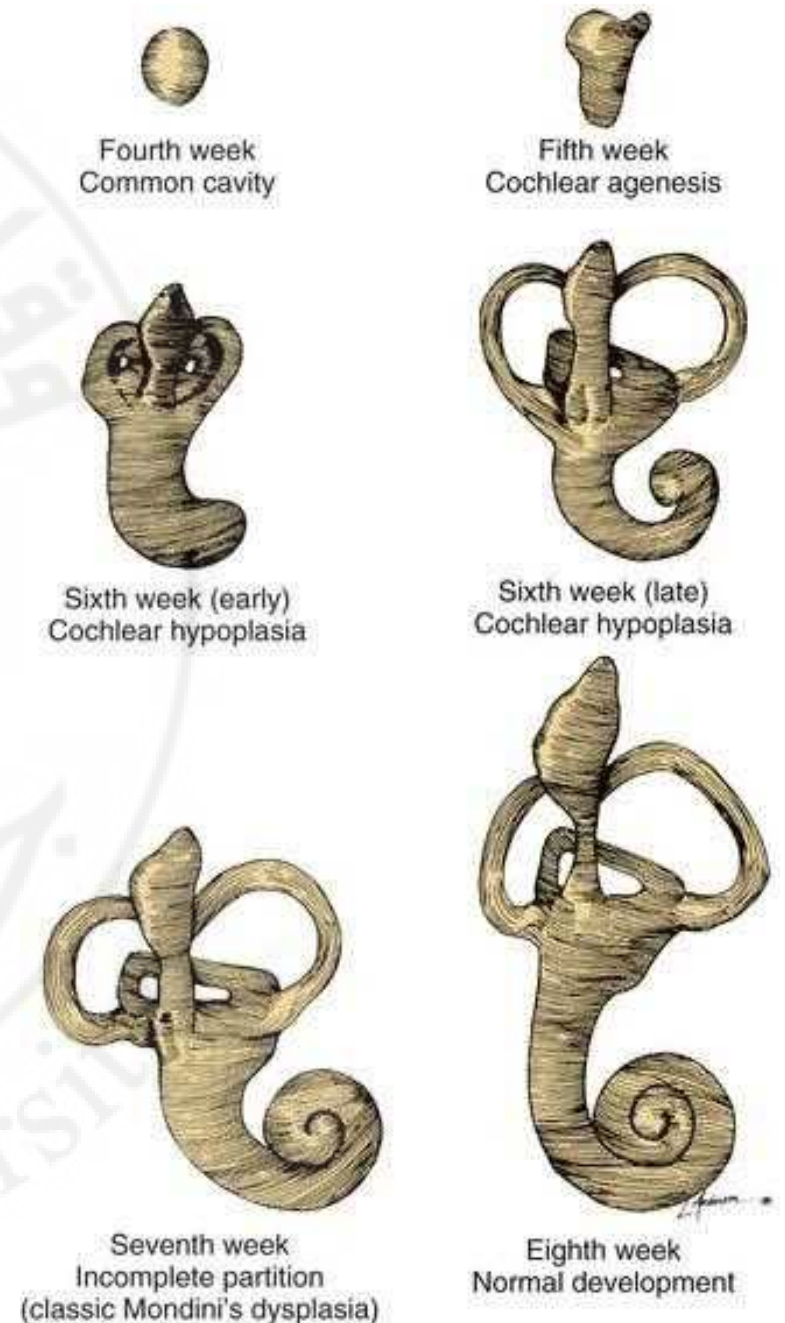
Relative Incidence of Cochlear Malformations

Malformation	Incidence (%)
Incomplete partition (Mondini's dysplasia)	55
Common cavity	26
Cochlear hypoplasia	15
Cochlear aplasia	3
Complete labyrinthine aplasia (Michel's aplasia)	1



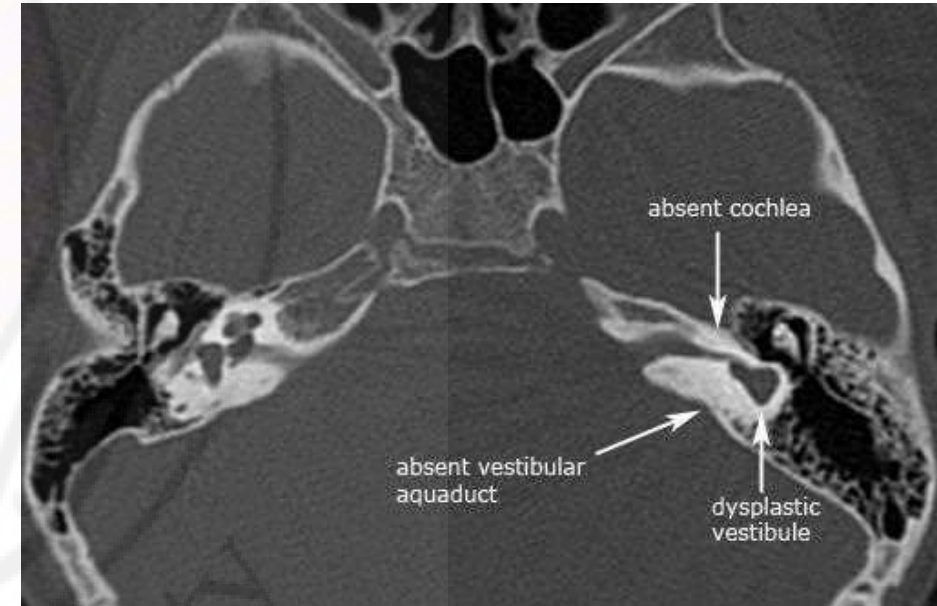
Embryogenesis of cochlear malformations

- Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the **first trimester of pregnancy**.
- This interruption may be either a result of inborn **genetic** error or a consequence of a **teratogenic exposure** during the period of inner ear organogenesis between the fourth and eighth weeks of gestation.
- Genetic errors may be either dominant or recessive and may manifest as sensorineural hearing loss (SNHL) alone or be associated with any of a number of *syndromes*.



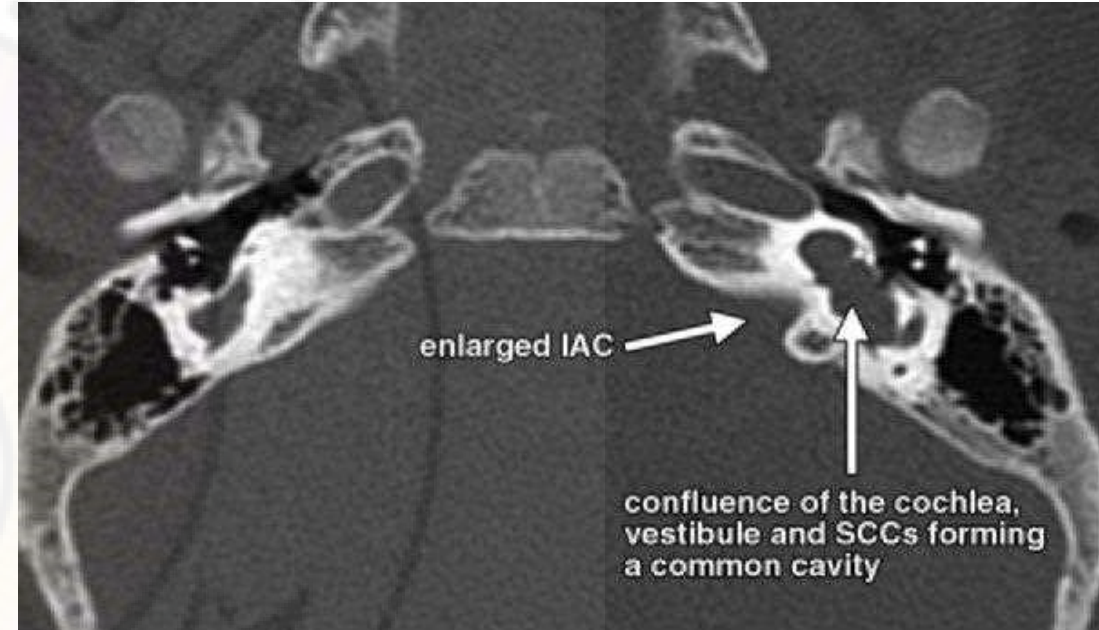
Michel's aplasia

- The worst case is the total absence of any inner ear structures, known as **Michel's aplasia or complete labyrinthine aplasia (CLA)**.
- It is possible for this to occur even with a normal conductive mechanism.
- Michel deformity is a very rare malformation characterized by complete absence of membranous and osseous inner-ear structures, resulting in **total deafness (anacusis)** and delayed gross development of child because of **balance impairment**.
- CT scan or MRI of the inner ear shows no recognizable structure in the inner ear.
- It is generally believed that Michel's anomalies are caused by **autosomal dominant** traits.

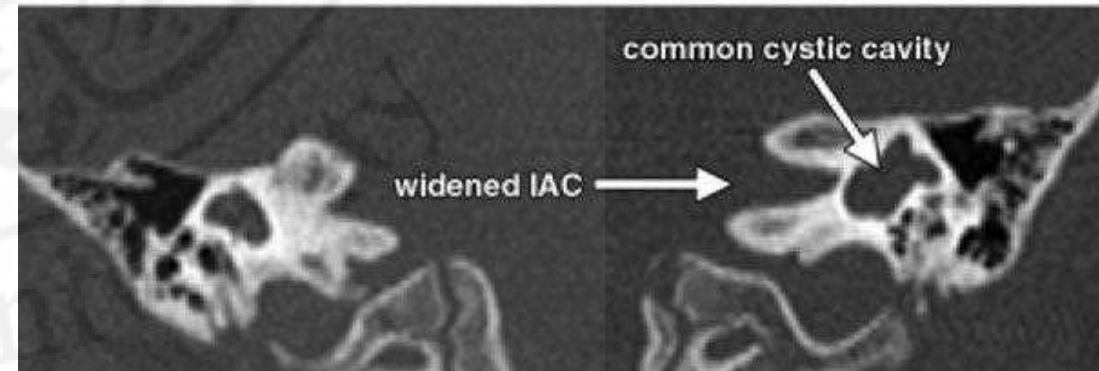


Common-cavity malformation

- The common-cavity malformation is one in which the cochlea is not differentiated from the vestibule, usually resulting in **profound hearing loss**.
- It accounts for about 25% of cochlear malformations.
- **Cochlear implants** are widely used for hearing rehabilitation for common cavity deformities; however, the reported prognosis is poor.

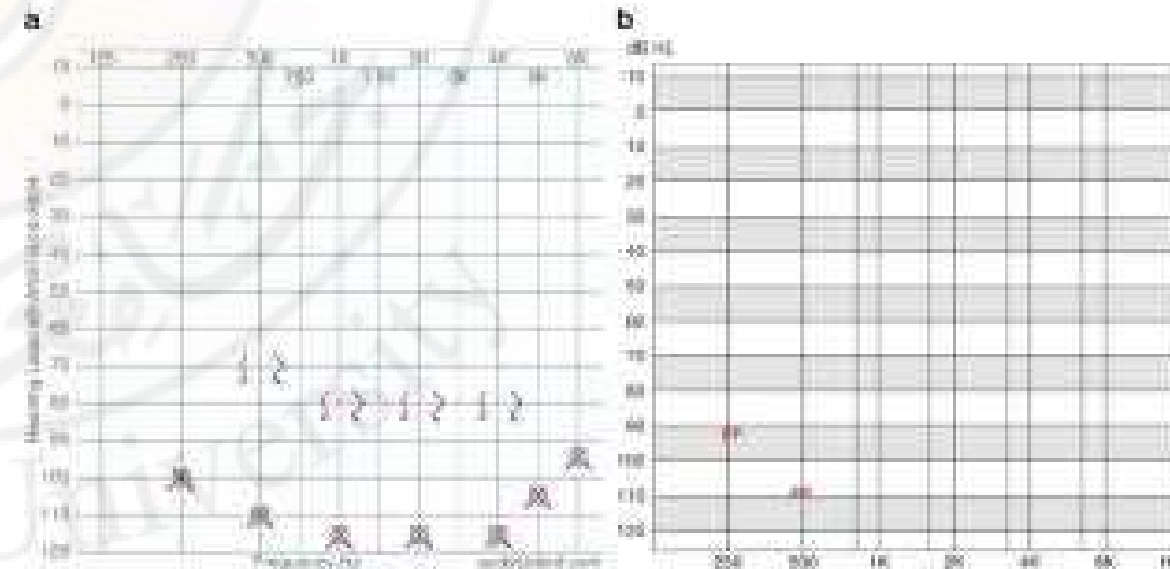


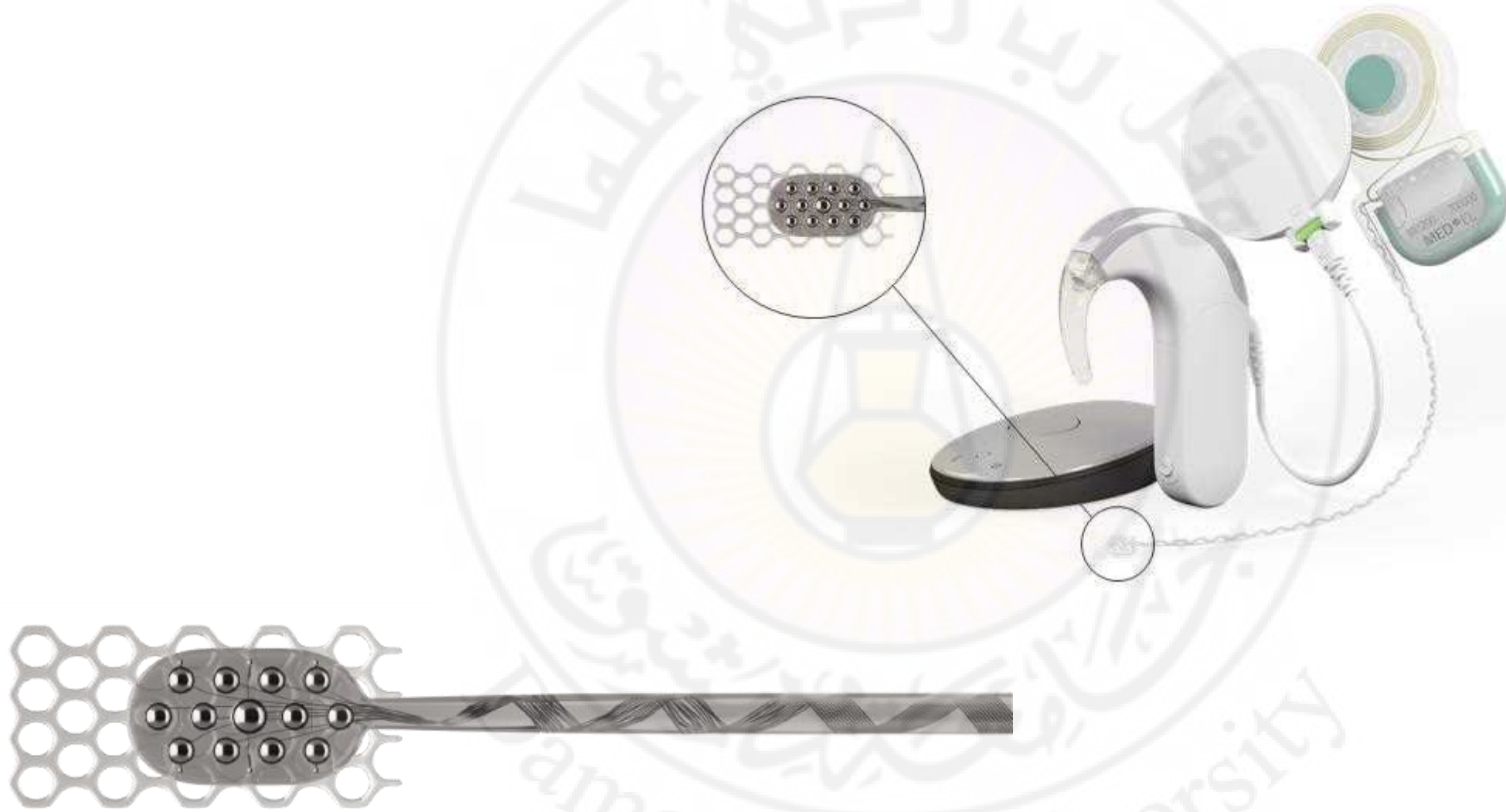
a.



Cochlear aplasia & hypoplasia

- **Cochlear aplasia** is a rare malformation consisting of complete absence of the membranous and osseous cochlea, with no auditory function, but presence of semicircular canals and vestibule.
- Cochlear aplasia is a definite indication for **Auditory Brainstem Implant (ABI)** since there is no inner ear development (No benefit from CI).
- **Cochlear hypoplasia** is a malformation in which less than one full turn of the cochlea is developed.

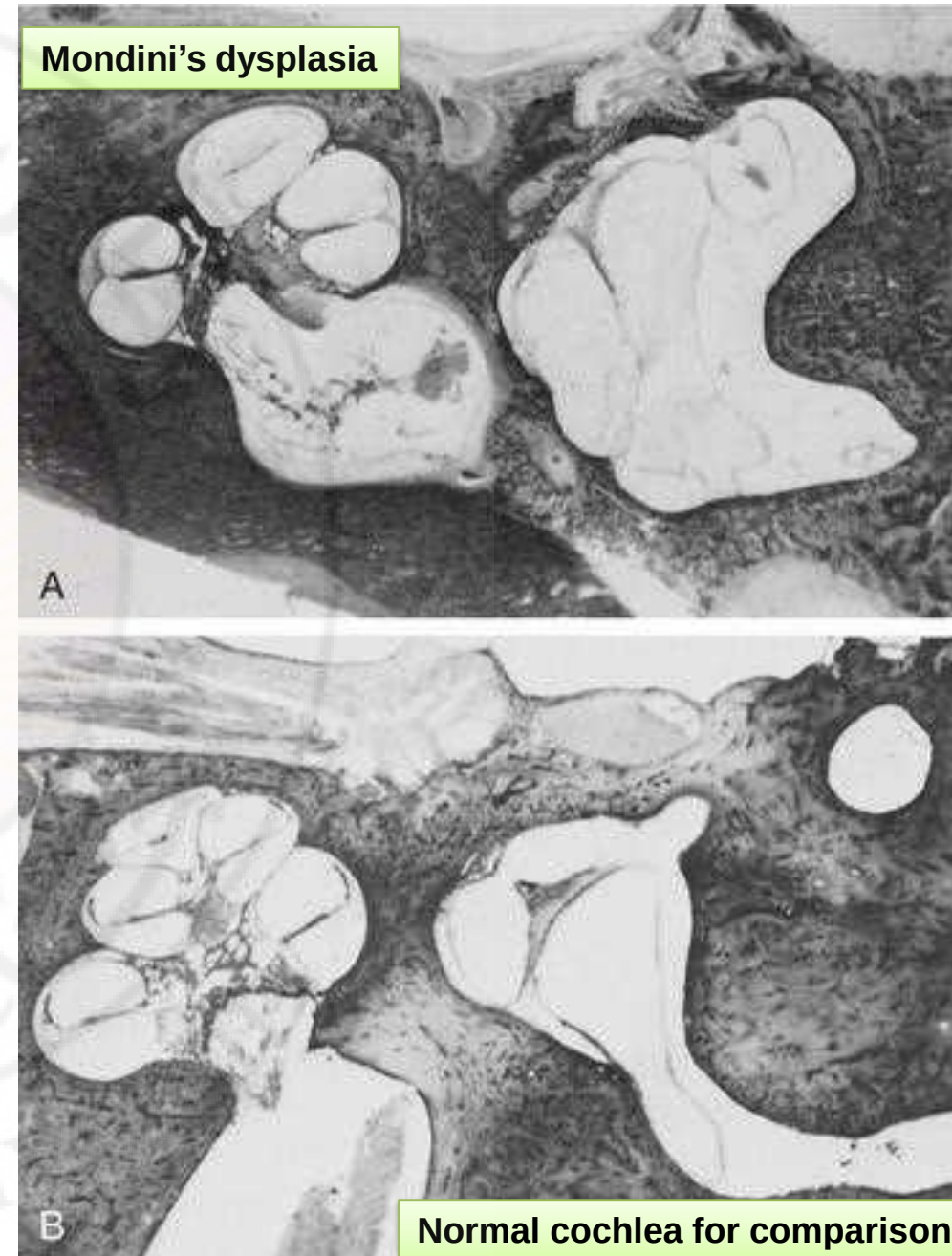




The SYNCHRONY ABI system from MED-EL

Mondini malformation (1)

- Mondini malformation, an incomplete partition of the cochlea, is a relatively common inner-ear malformation (accounts for about 55% of all inner ear malformations), in which the cochlea contains only about **1.5 turns** (instead of the normal two and a half turns), and the osseous spiral lamina is partially or completely absent.
- The resulting hearing loss is highly variable. Nearly all persons with Mondini malformations are completely deaf on the affected side, but high frequency hearing can be present in some.



Mondini malformation (2)

- While the hearing loss in these malformations is probably sensory in origin, the audiograms in these people **sometimes** suggest a *conductive hearing loss* -- this is probably related to relatively better bone conduction in some of these malformations than normal because of lower compliance between the perilymphatic and CSF compartments. This is similar to the situation in superior canal dehiscence.
- It is generally believed that Mondini's anomalies are caused by **autosomal dominant** traits.
- The majority of Mondini cases are **bilateral**. Unilateral Mondini malformations are not generally inherited.

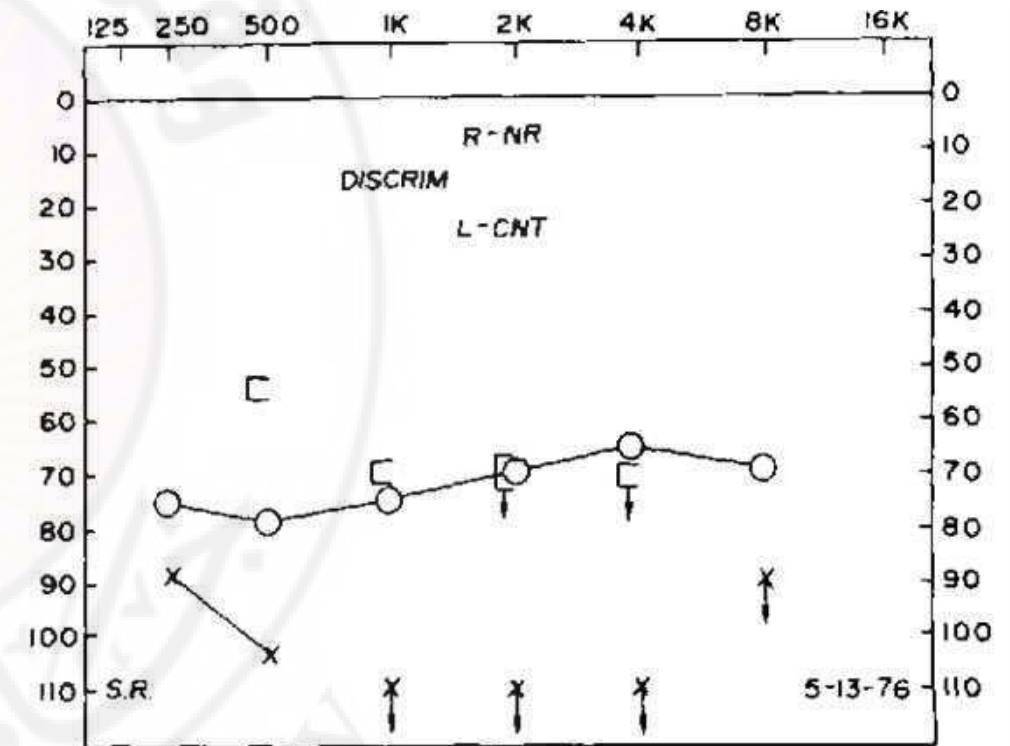
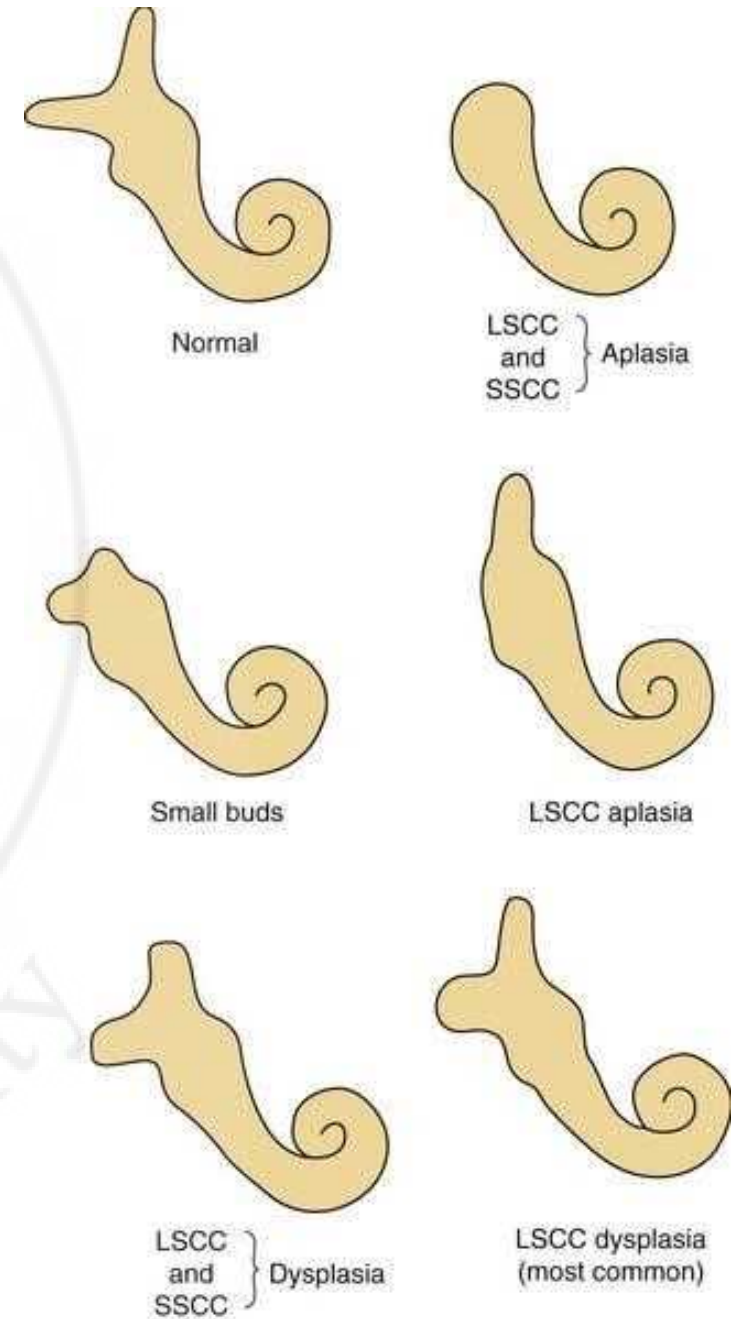


Fig. 9. (Case 6) Audiogram showing profound hearing loss in the left ear caused by Mondini dysplasia. At age 50 the patient suffered a rapidly progressive hearing loss of unknown cause in his right ear.

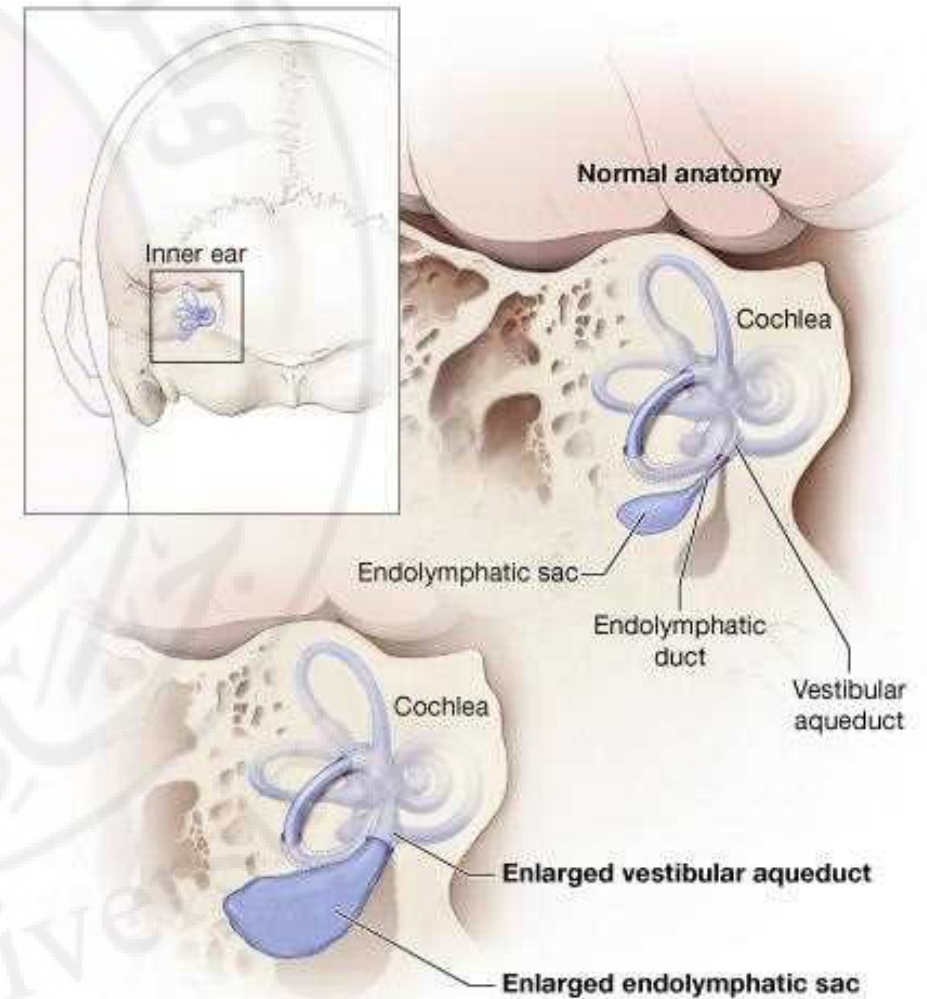
Anomalies of the semicircular canals

- **Dysplasia** of the lateral SCC is a common type of inner ear malformation.
- Approximately 40% of ears with a malformed cochlea will have an accompanying dysplasia of the lateral SCC.
- The lateral SCC is deformed more often than the posterior or superior SCC, apparently because it forms earlier in embryogenesis.
- **Caloric responses** in SCC dysplasia are functionally absent or reduced in most cases, but a few may have normal responsiveness.
- Ears with malformations limited to the vestibular system often have normal or near-normal hearing. When the cochlea also is abnormal, sensory hearing levels tend to be impaired, to a variable degree.
- **SCC aplasia** is only one-fourth as common as SCC dysplasia. It is usually associated with cochlear anomalies. Most cases are syndromic.



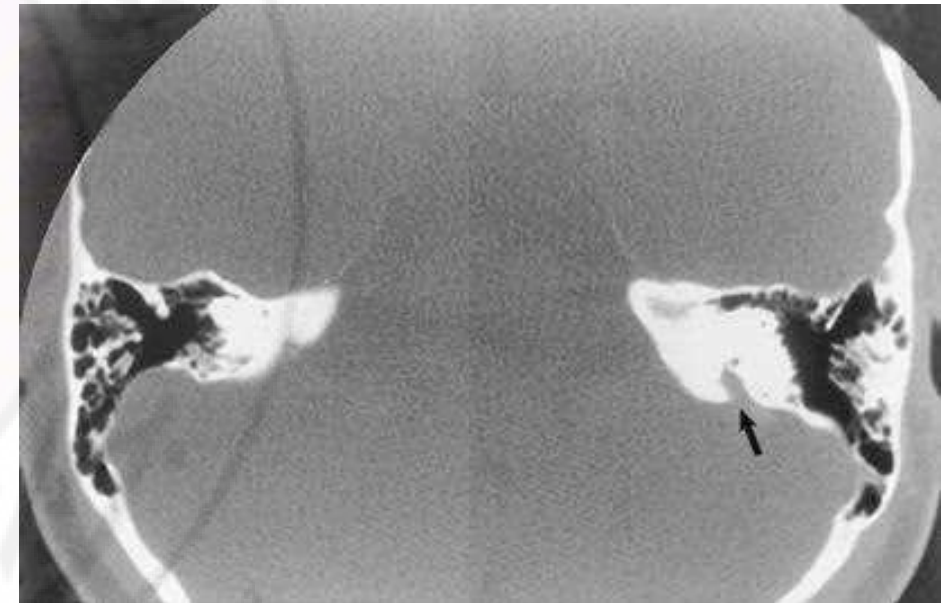
Large Vestibular Aqueduct Syndrome (LVAS/EVA) (1)

- One form of inner ear dysplasia is unique, because it has been associated with **delayed onset of SNHL**.
- An enlarged vestibular aqueduct (EVA) is commonly seen in combination with other inner ear dysplasias, but more recently, it has been noted as an isolated finding in many ears.
- These patients may have any level of hearing, from **normal to a profound loss**. Frequently, **both ears are affected**, and the losses are **asymmetric**.
- **Fluctuation** of hearing is common and usually affects one ear at a time; this may manifest as anacusis in one ear with fluctuation in the other.
- A **conductive component** to the low-frequency hearing loss is often evident, as the EVA may act as a third mobile window.



Large Vestibular Aqueduct Syndrome (LVAS/EVA) (2)

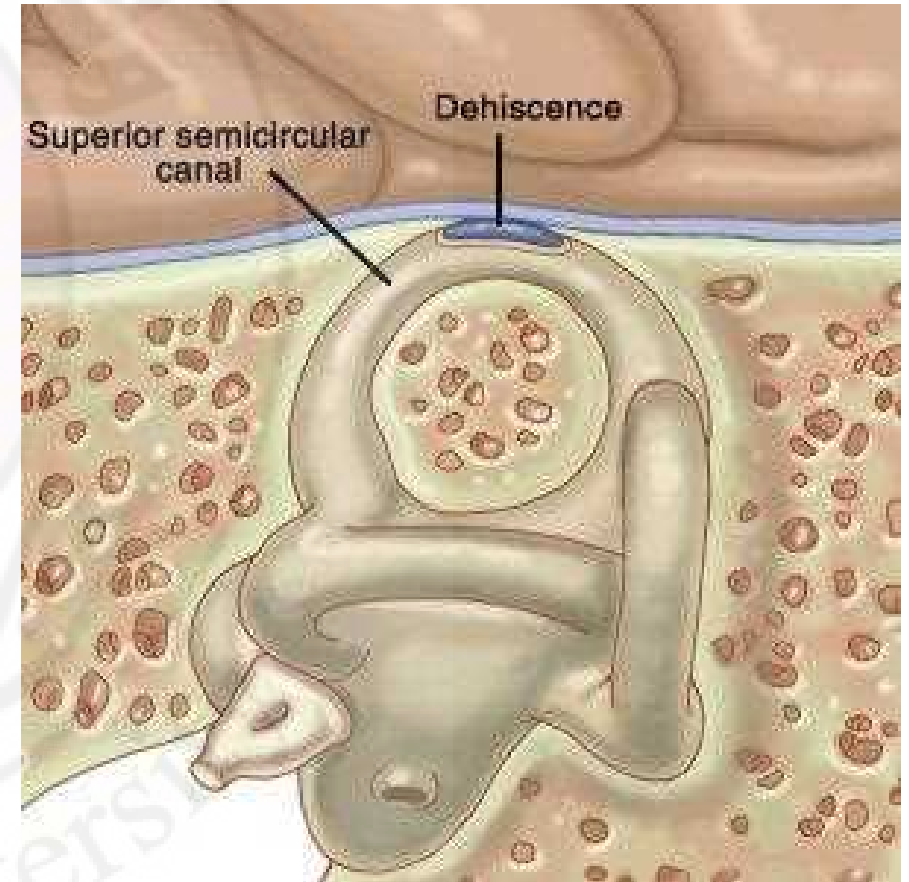
- In patients who have been followed over time, a **progressive** stepwise loss has been noted in many.
- This syndrome has been found to be familial in some cases, and it probably occurs much more commonly than generally appreciated.
- It is seen in isolation, as part of the **Mondini malformation**, and in patients with **branchio-oto-renal syndrome** and **Pendred syndrome**.
- While it may be detected on high resolution volumetric MRI, it is well shown on high-resolution CT imaging of the temporal bone.



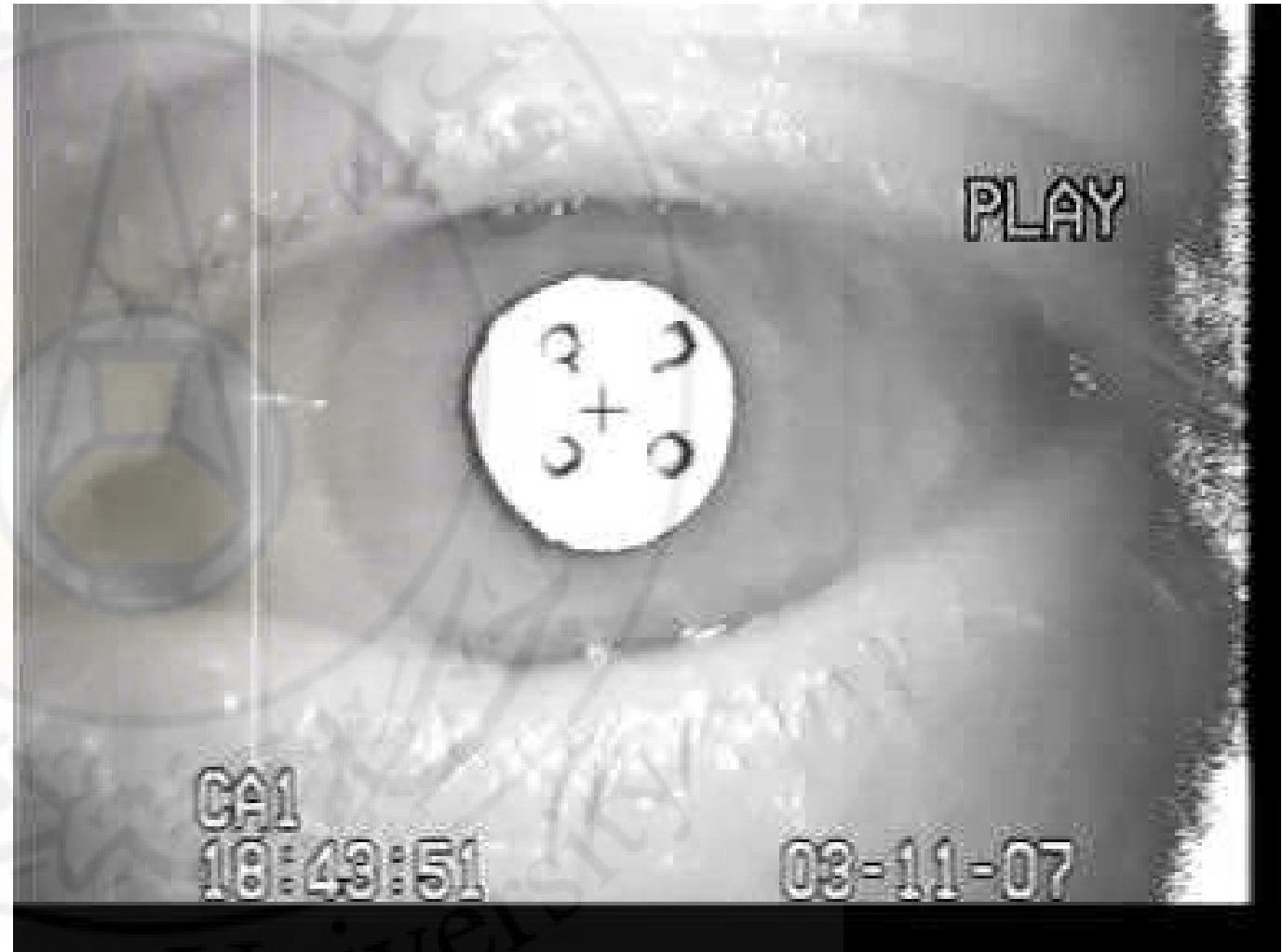
Computed tomography scan of temporal bone shows large vestibular aqueduct syndrome. Arrow indicates the enlarged vestibular aqueduct

Semicircular canal dehiscence (SCD)

- Superior canal dehiscence (SCD) is an inner ear disorder in which a section of the bone enclosing the superior semicircular canal is open or extremely thin (**third mobile window**), resulting in vestibular and or auditory symptoms that are caused by sound and/or pressure.
- The vestibular problems are the most disturbing symptoms of SCD. They include episodes of **vertigo**, **nystagmus**, and **oscillopsia** (an oscillating visual field) that are induced by sound and/or pressure events like nose blowing, straining to lift heavy objects, flying, etc.
- In addition, patients often experience ongoing problems with imbalance or unsteadiness as well tolerating motion.

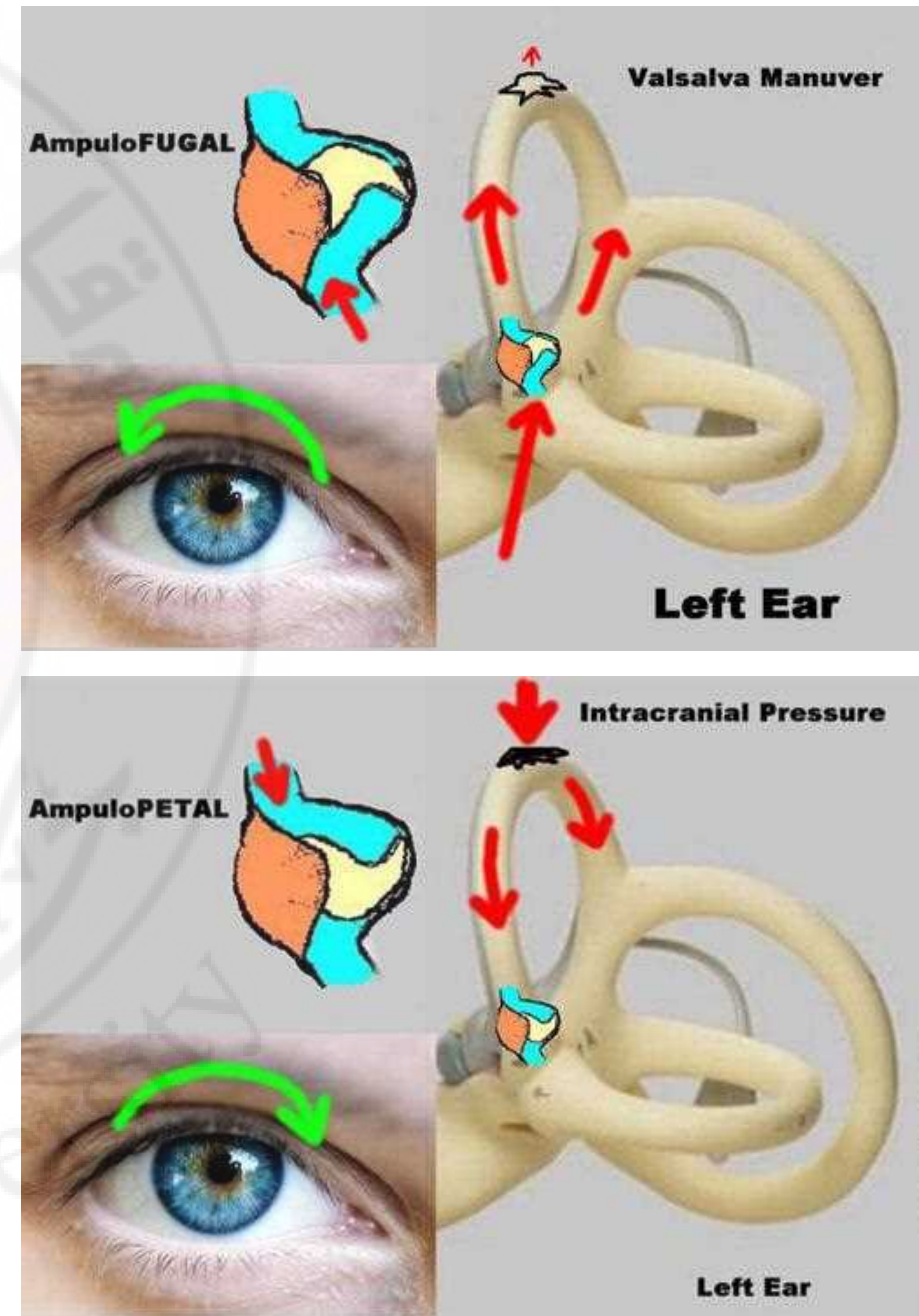


This patient has a dehiscence in the superior semicircular canal on the left. When loud sound is applied to the left ear, the left eye is seen to elevate and rotate towards the midline. When the sound is removed, the eye returns to neutral position. This can also be seen with positive pressure applied to the ear canal.



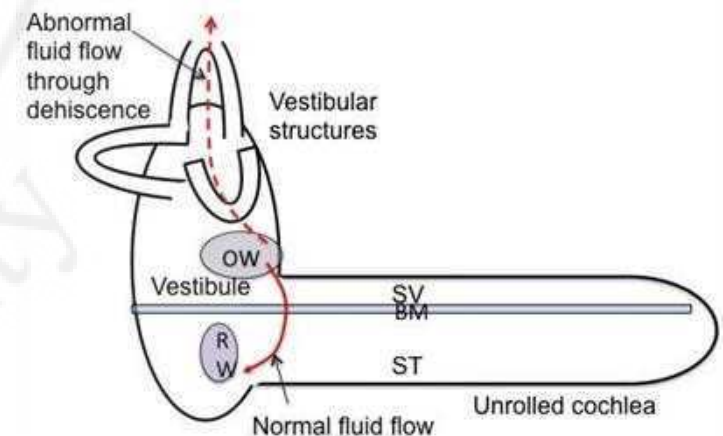
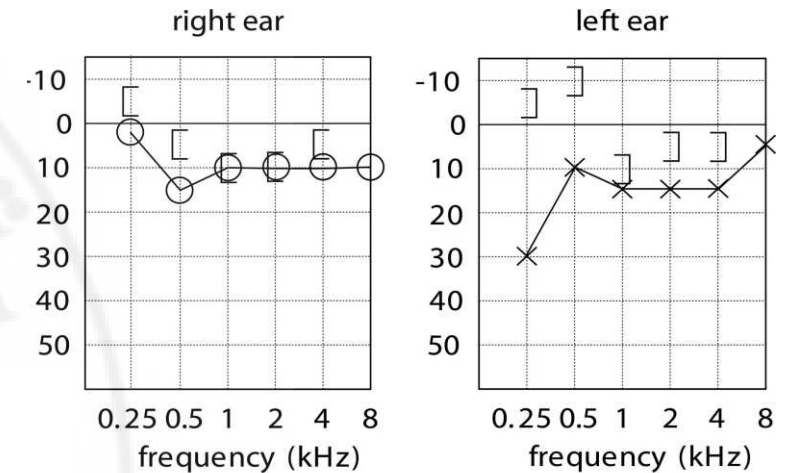
Semicircular canal dehiscence (SCD)

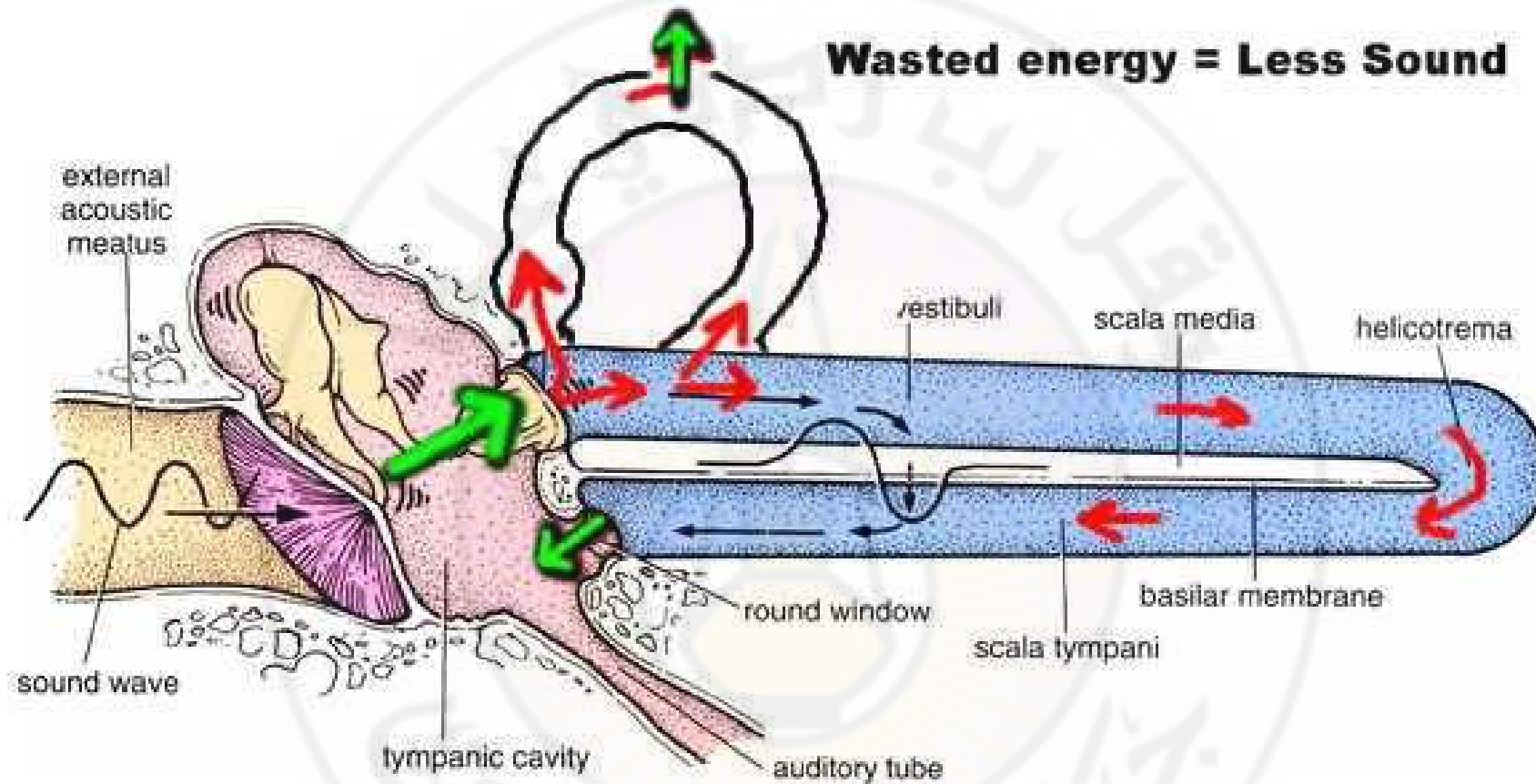
- Under normal conditions, sound pressure by air conduction enters the fluid-filled inner ear via the oval window and exits via the round window.
- In superior canal dehiscence, the deficit of bone overlying the semicircular canal results in a **third mobile window** effect, leading to areas of increased compliance and allowing for the movement of endolymph within the canal in the presence of sound and/or pressure stimuli.
- These stimuli increase the pressure gradient within the membranous labyrinth resulting from the change in compliance at the dehiscence site, with resulting deflection of the superior canal cupula and a subsequent inhibitory or excitatory response from the vestibular nervous system.



Semicircular canal dehiscence (SCD)

- The hearing loss typically associated with SCD is conductive in nature with air-bone-gaps in the low to mid-frequencies, but other kinds of hearing loss are possible.
- The air-bone-gaps involved in SCD often include better-than-expected bone-conduction thresholds (sometimes called **conductive hyperacusis**) in addition to elevated air-conduction thresholds.
- In addition, a notable feature of SCD is the tendency for there to be **normal tympanograms** and **present acoustic reflexes** in spite of the air-bone-gap.
- **Pulsatile tinnitus** and the hyper-perception of the sounds produced by one's own body (**autophony**) are also common.
- A diagnosis of SCD is confirmed with computerized tomography (CT).

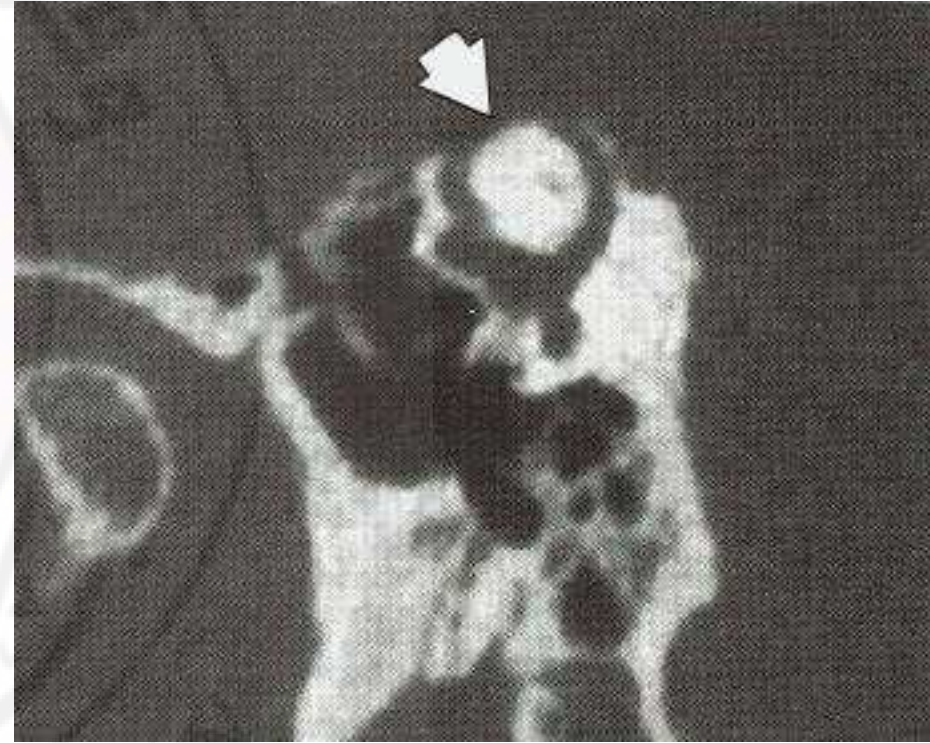




The increased air-bone gap in SCD patients is due to a **“third window”**. This “third window” allows some of the inner ear fluid to move away from the cochlea and into the opening in the superior canal. This, in turn, leads to a decrease in acoustic energy and increased air conduction thresholds.

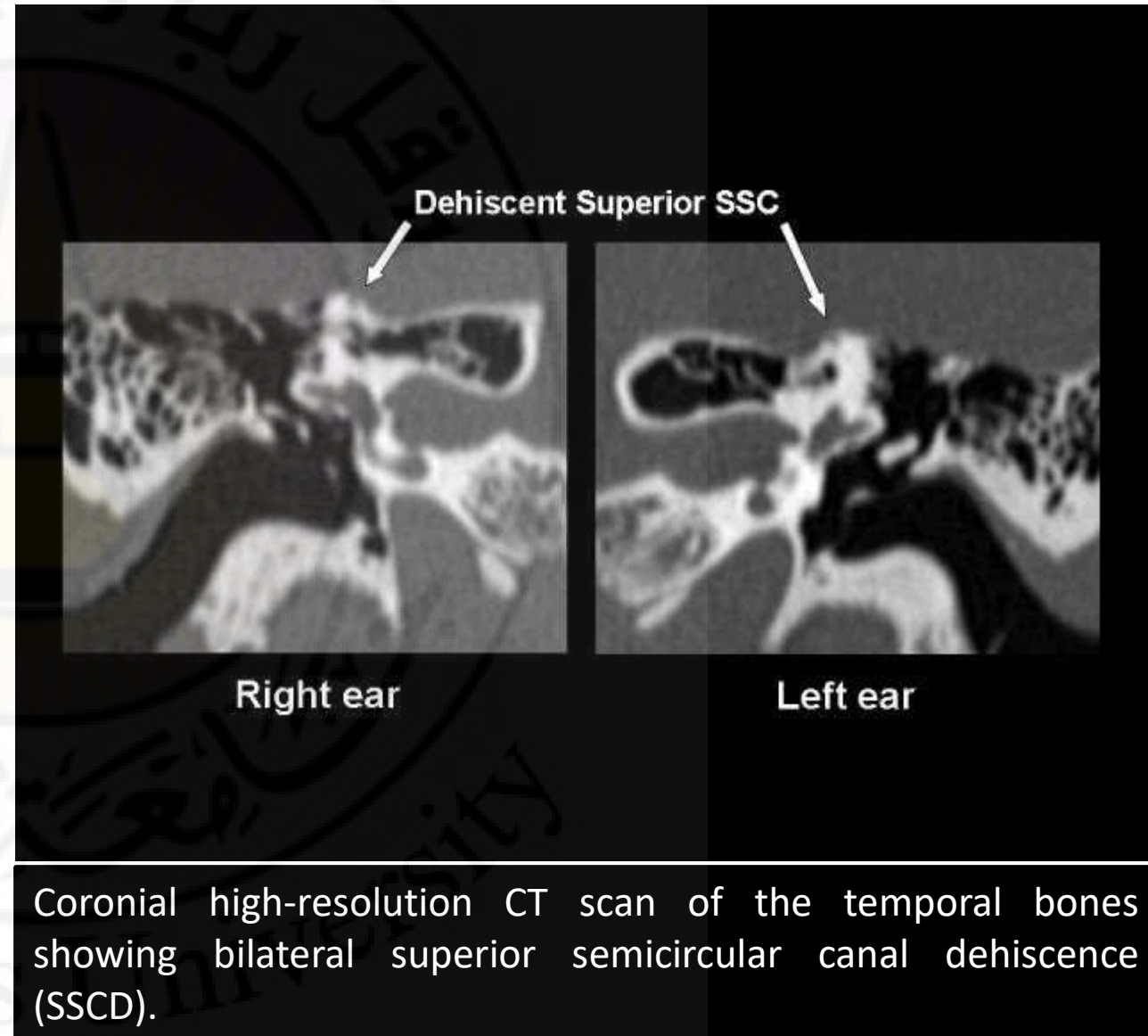
SCD: Common Symptoms

- Increased air-bone gap for low frequencies
- Hyperacusis for bone-conducted sounds
- Fullness sensation in the affected ear
- Pulsatile tinnitus
- Autophony - Hearing one's own speech or other self-generated noises (e.g. heartbeat, chewing, footsteps, creaking joints) loudly
- Pressure and sound induced vertigo (Tullio phenomenon)
- Oscillopsia
- Dizziness/ vertigo/ chronic disequilibrium
- Pulse-related eye movements: often described as blurriness



SCD: Age of Onset

- Symptoms of SCD usually do not occur until adulthood
- Events such as head- or barotraumas may break the fragile bone, causing the dehiscence
- May have a congenital or developmental etiology
- A mutation of the **COCH gene** in a patient with SCD and familial deafness has been reported.



Case Example

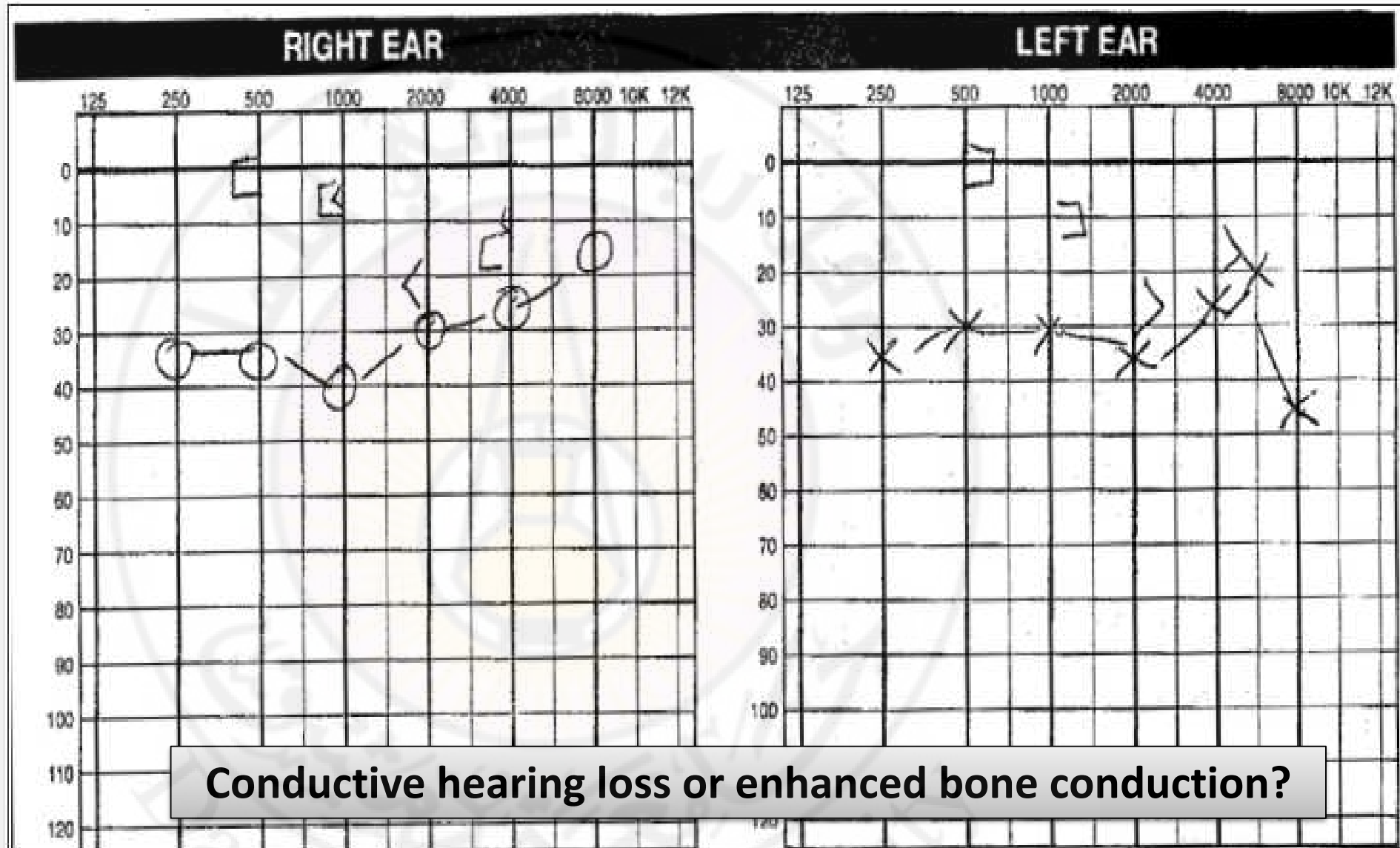


Figure 3b: Conductive hyperacusis in patient with bilateral SCD. This can easily be missed as audiologists just assume that one can't hear better than '0' dB, and don't always test the patient thoroughly. In other words, thresholds can be better than 0.

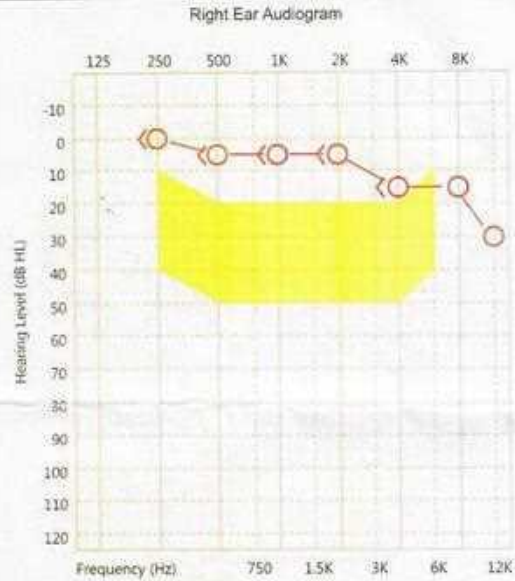
Case Example

First Name: _____
 Name: _____
 Age: 45 years
 Gender: Male

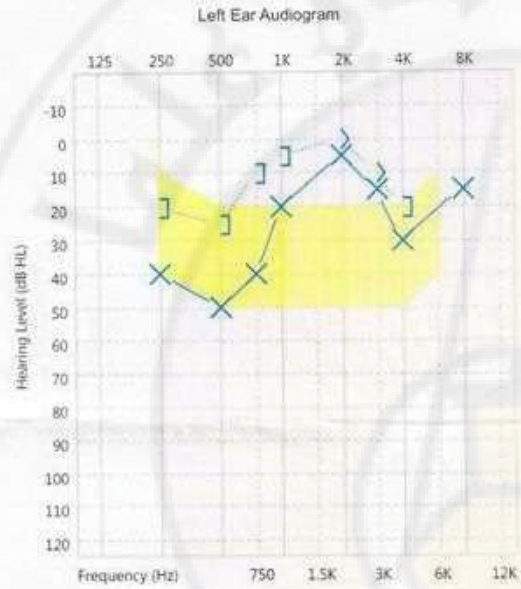
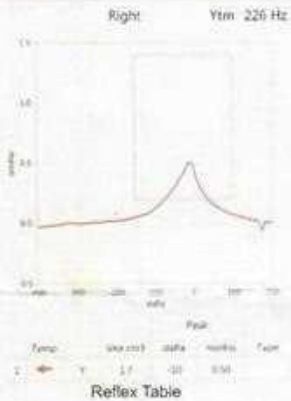


Facility Name
 Address (line 1)
 Address (line 2)
 Phone

8/16/2021 7:23 PM



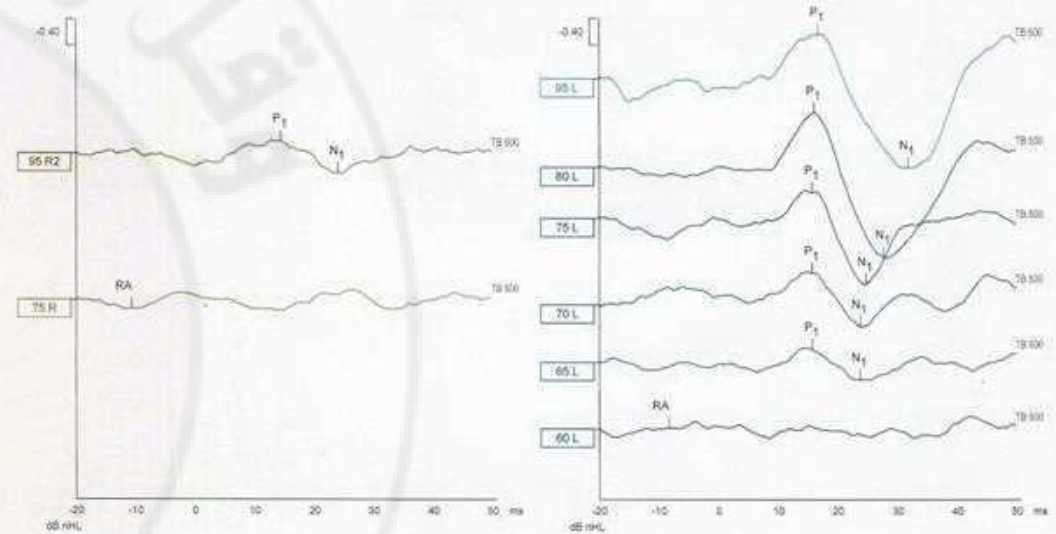
Ear	Test Type	%	dB HL
R	SRT	10	
R	MCL	40	
R	WRS	100	40



Ear	Test Type	%	dB HL
L	SRT	20	
L	MCL	45	
L	WRS	96	45



Address: Day Medical Tower Unit C, 9th Floor., Iran Tehran
 Tel: 021-88205606 Fax: 021-88202934 E-Mail: @tehranearclinic
 Last name: _____ Sex: M ID no: 4000545
 First name(s): _____ Age: 45 years Date of test: 08-16-2021
 Address: _____ Day of birth: 08-04-1976
 Examiner: ADM E-Mail: _____



Reflex Right F:226 Hz P:-24 daPa

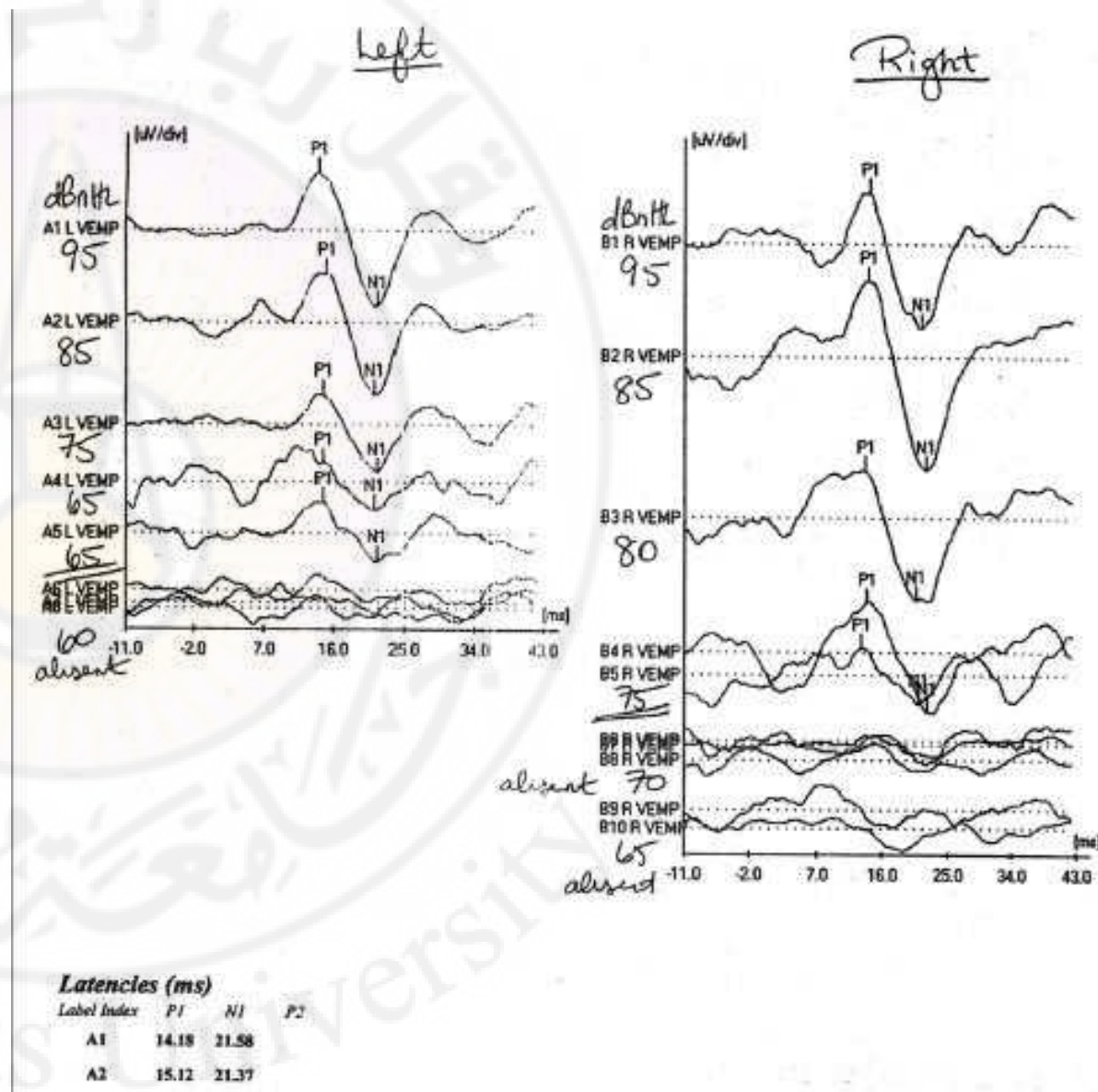
500 Hz I	0.02
1 kHz I	0.06
2 kHz I	0.08
4 kHz I	0.02
500Hz C	
1 kHz C	
2 kHz C	
4 kHz C	

Reflex Left F:226 Hz P:-17 daPa

500 Hz I	0.00	0.00	0.01	0.03
1 kHz I			0.05	
2 kHz I			0.07	
4 kHz I			0.03	
500Hz C				
1 kHz C				
2 kHz C				
4 kHz C				

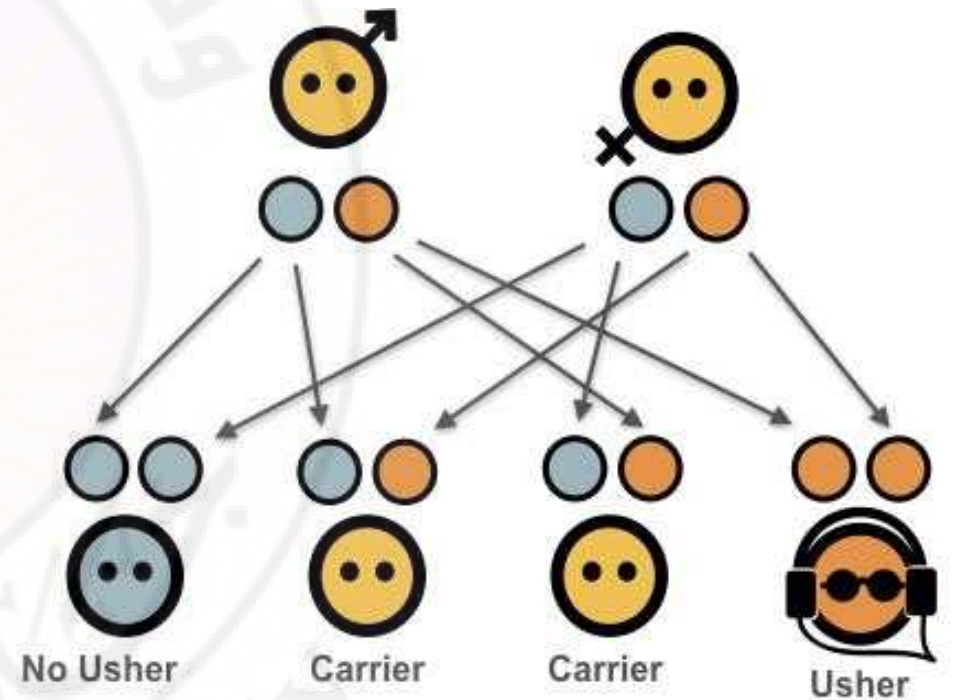
Case Example

cVEMPs in a subject with **bilateral SCD**, right more symptomatic than left. The VEMP response can be seen to persist down to stimulation levels as low as 65 dB HL on the right side, whilst lost past 75 dB on the right.



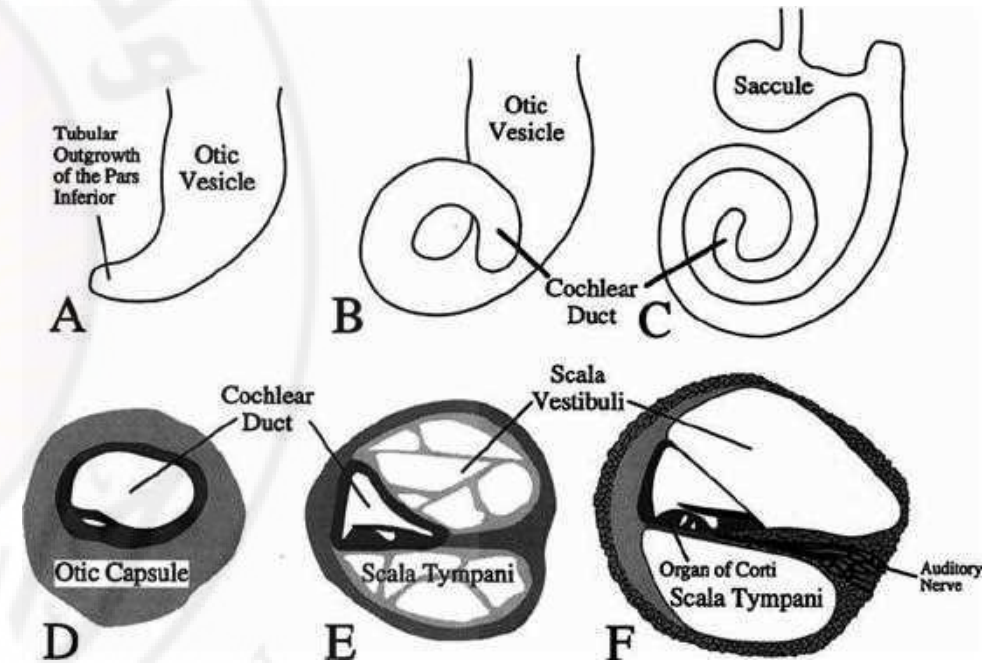
Bing Siebenmann malformation

- The **Bing Siebenmann** malformation is a **rare** malformation resulting in complete lack of development of the membranous labyrinth.
- It has been reported in association with cardioauditory (Jervell and Lange-Nielsen) and Usher syndromes.



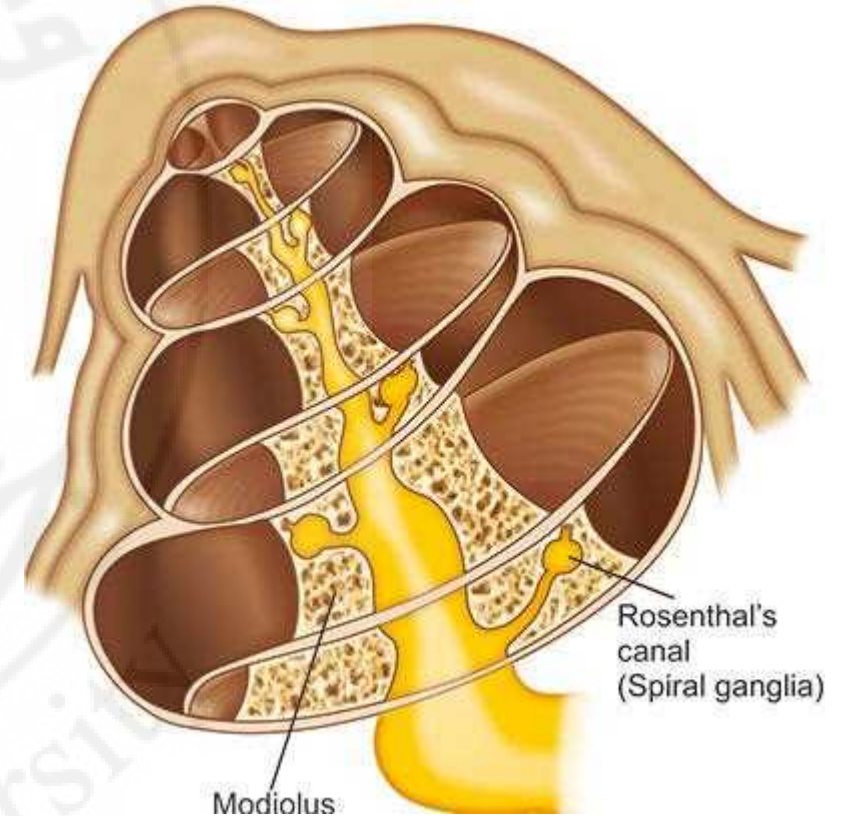
Scheibe aplasia

- **Scheibe aplasia, or cochleosaccular dysplasia**, is a common inner-ear abnormality in which there is failure of the organ of Corti to fully develop.
- In cochleosaccular (Pars inferior) dysplasia, the SCCs and utricle (Pars superior) are normal.
- Auditory neuronal survival is variable but may remain normal into adulthood, at least in some cases.
- Scheibe's aplasia is transmitted by the autosomal recessive route.



Alexander aplasia

- Alexander aplasia is an abnormal development of the basal turn (high-frequency region) of the cochlea, with normal development in the remainder of the cochlea, resulting in low-frequency residual hearing (**familial high-frequency SNHL**).
- Affected persons probably are minimally symptomatic. They would have normal hearing and presumably would have compensated for their congenital vestibular deficit.

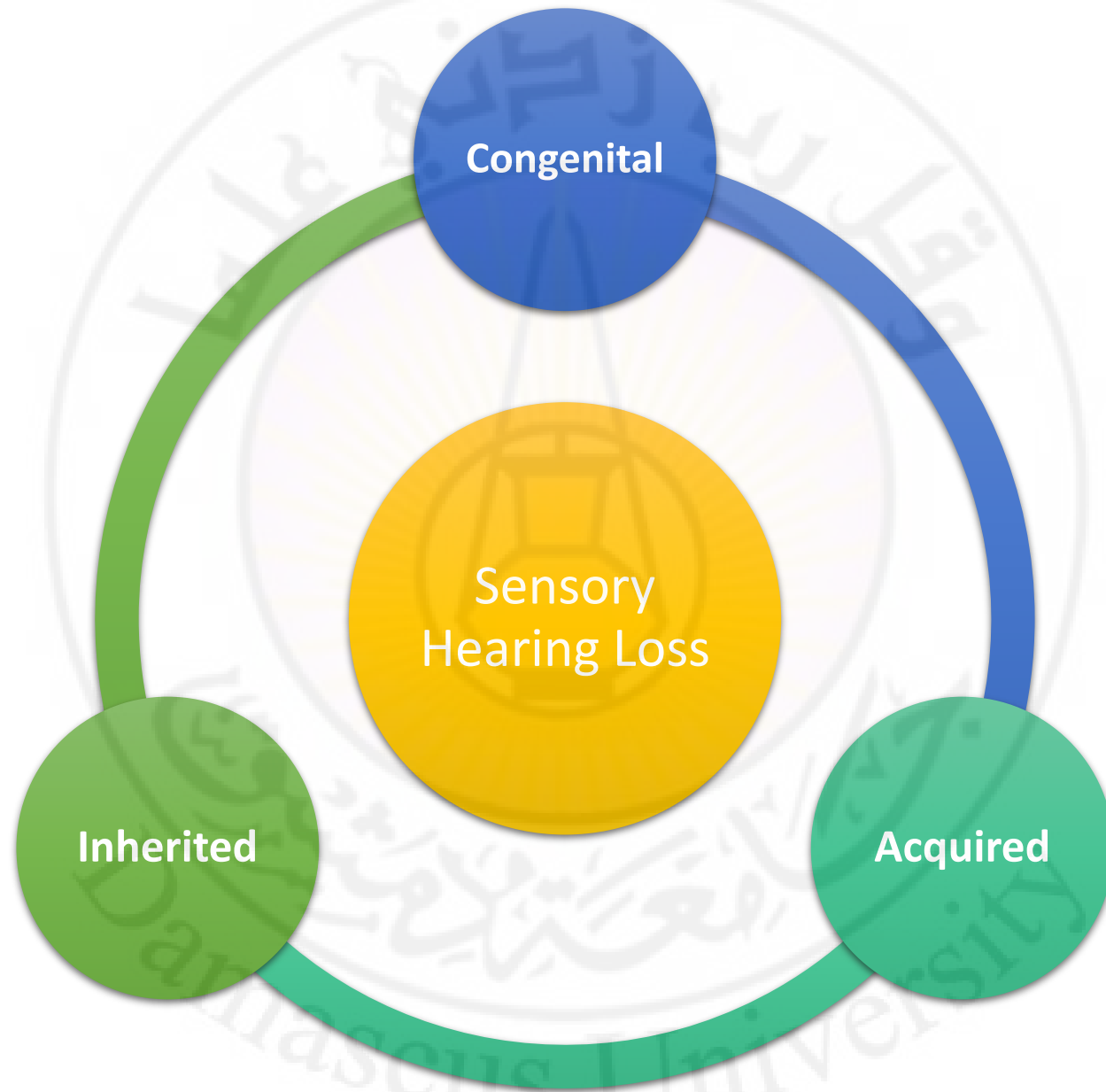




Clinical Audiology

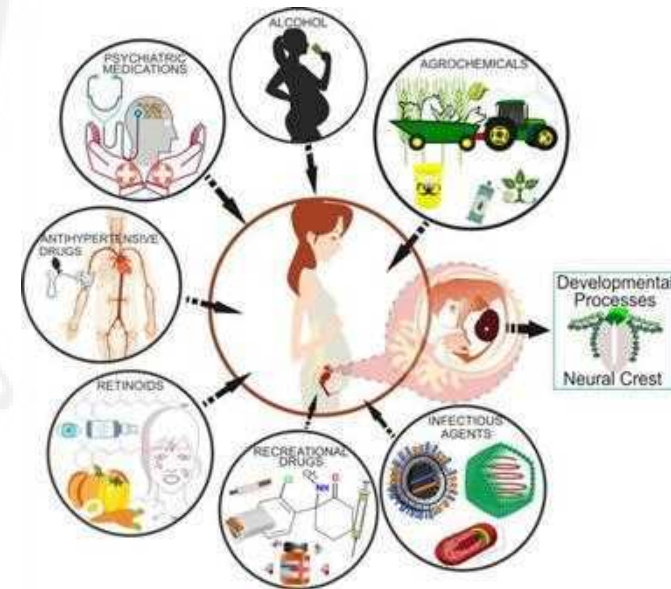
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Teratogenic Factors

- A **teratogen** is anything a person is exposed to or ingests during pregnancy that's known to cause fetal abnormalities. Drugs, medicine, chemicals, certain infections and toxic substances are examples of teratogens.
- Sensorineural hearing loss can result from teratogenic effects of **congenital infections** on a mother during embryologic development of a fetus (**Prenatal factors**).
- Congenital **infections** most commonly associated with sensorineural hearing loss include (TORCH complex):
 - **Cytomegalovirus (CMV)**
 - **Rubella**
 - **Syphilis**
 - **Toxoplasmosis**
 - **Herpes simplex**
 - **Zika virus**
 - **Human immunodeficiency virus (HIV)**
- **And Other Teratogenic Factors (Drugs, maternal diseases, nutritional deficiencies....)**

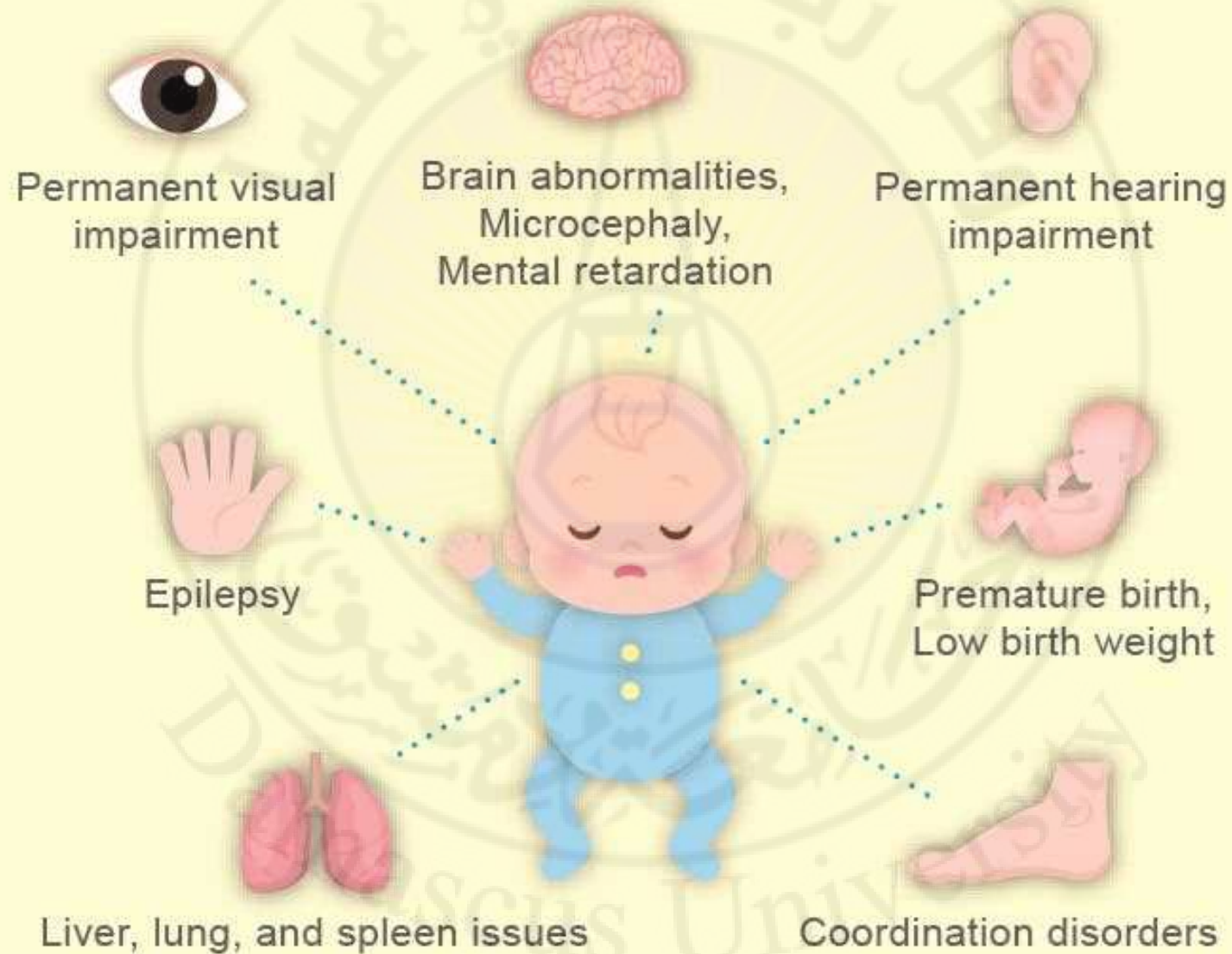


Cytomegalovirus

- Cytomegalovirus (CMV) is the largest known member of the human **herpes** virus family and causes **the most common fetal viral illness**.
- Congenital infection is the result of trans-placental transmission of CMV and is known as cytomegalic inclusion disease.
- Sensorineural hearing loss is the most common clinical finding of congenital CMV, which is estimated to account for ~ 21% of hearing losses at birth and ~ 25% at 4 years of age .
- Other findings include microcephaly, intellectual developmental disorder, cerebral palsy, liver disease, pneumonia, dental defects, visual lesions, and in rare cases death.
- In general, the **hearing loss** varies widely among those born with CMV infections, and may be congenital or delayed in onset, unilateral or bilateral, progressive, fluctuating or stable, and it can range in degree from mild through profound.
- Unfortunately, an effective CMV vaccine is not yet available.



Cytomegalovirus



Auditory assessments of a boy with congenital CMV:

Birth: Passed newborn hearing screening.

Six months: Diagnostic ABR due to risk for progressive loss.

- Left ear: Normal
- Right ear: Mild to moderate loss

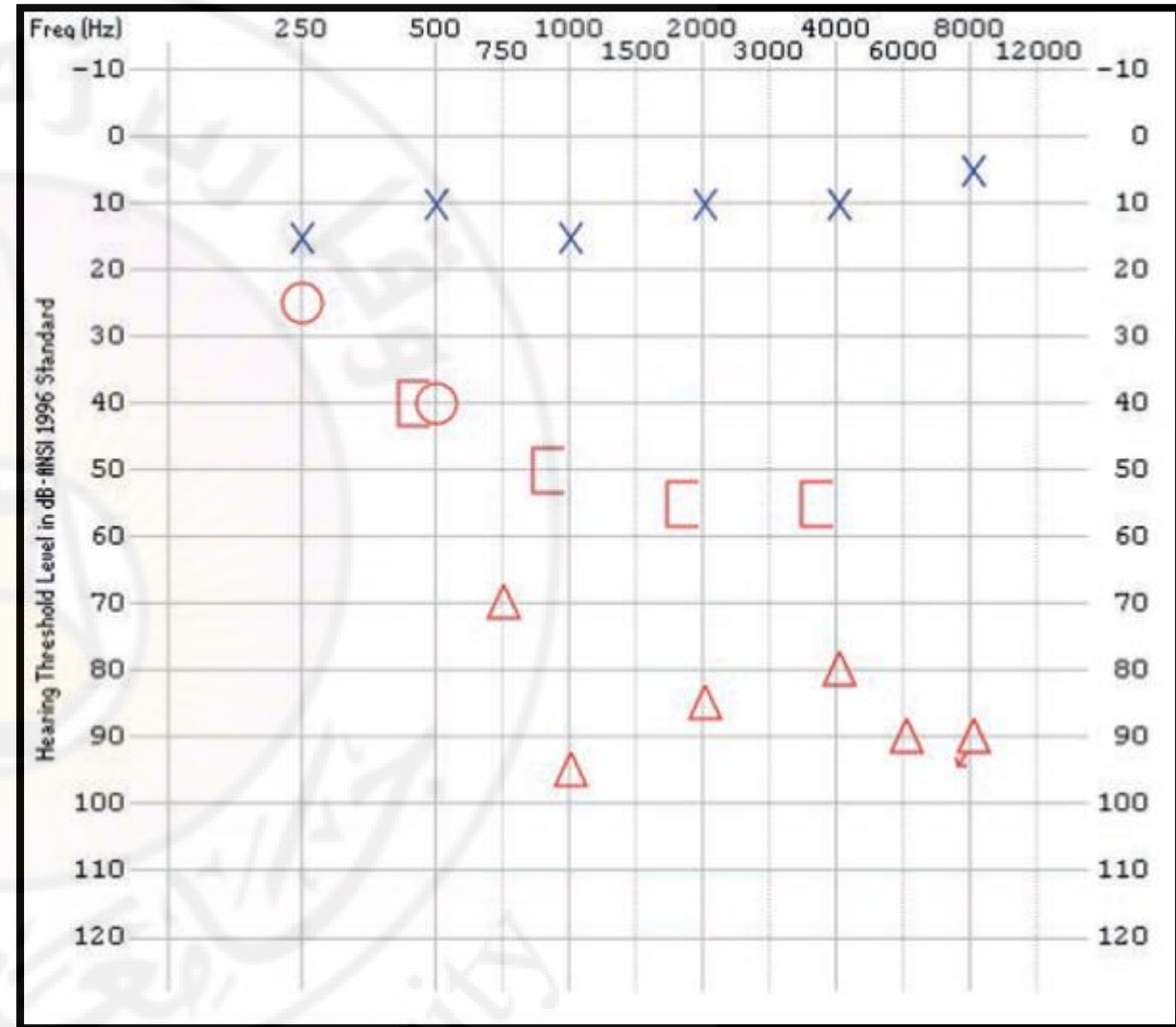
Two years: Follow-up audiology – Normal responses in the sound field and a normal speech awareness threshold was obtained in both ears via insert earphones.

Eight years: A consultation with an otolaryngologist was sought for abnormal gait and balance concerns.

CT scan – Normal temporal bones with long-standing abnormal brain findings characteristic of perinatal CMV infection.

Audiogram

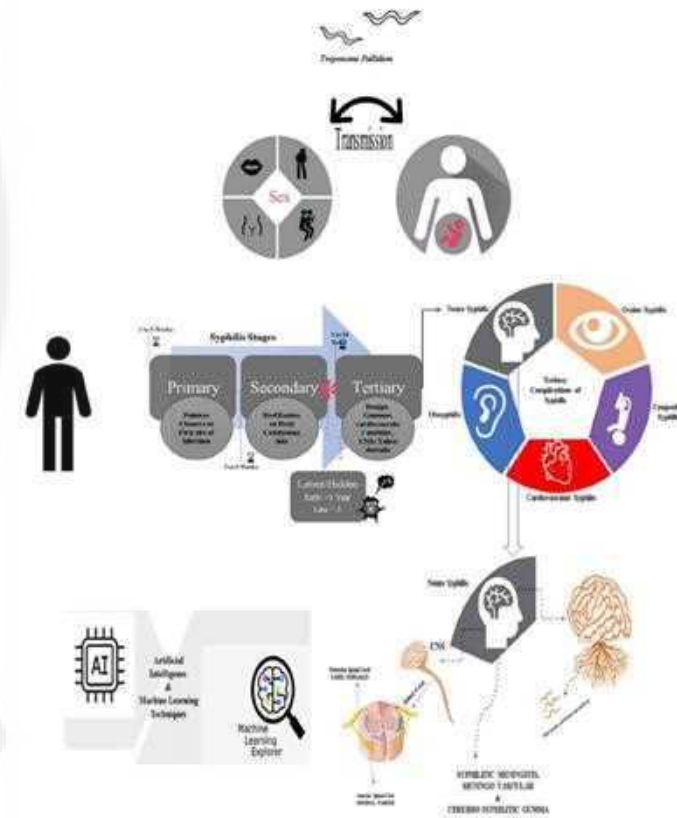
- Right ear: Mild sloping to profound mixed hearing loss
- Left ear: Normal hearing



Audiogram of boy with congenital CMV at 8.7 years of age indicating mild sloping to profound mixed hearing loss in the right ear and normal hearing in the left ear.

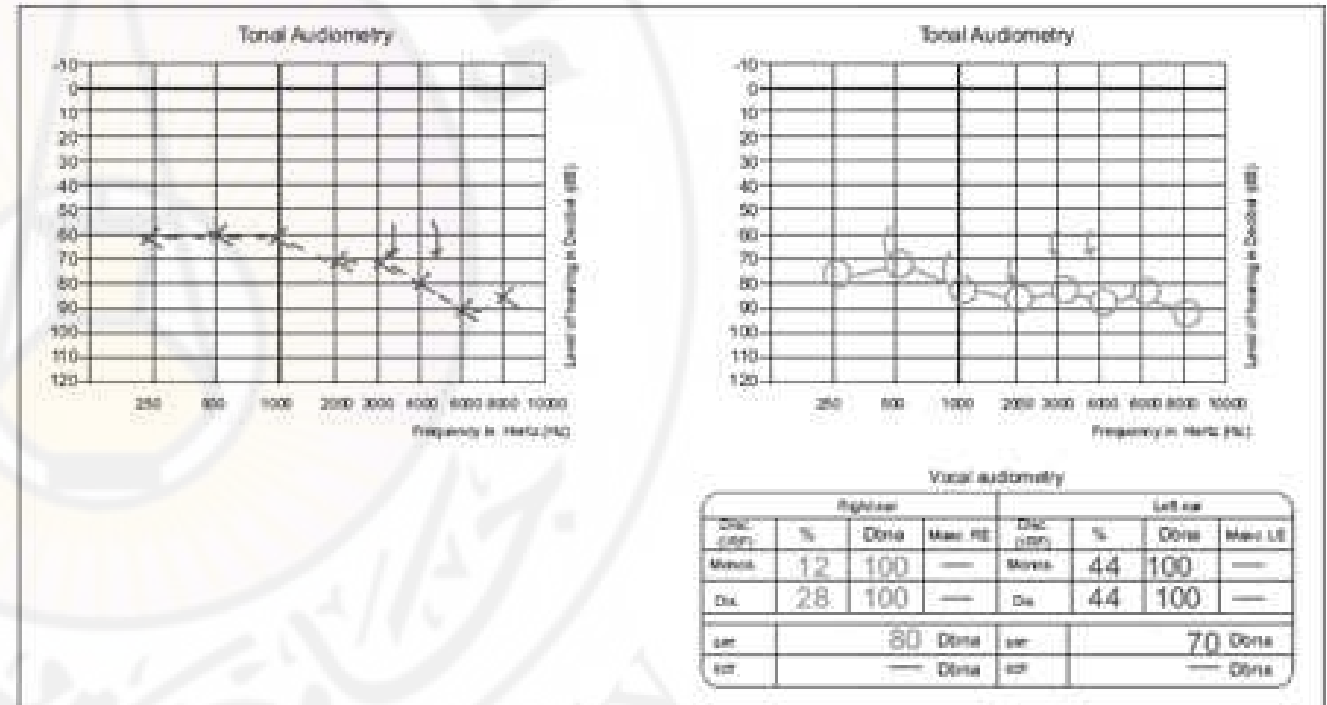
Congenital Syphilis

- Congenital syphilis (**Lues**) is a bacterial infection that is transmitted from infected mother to fetus in utero or through contact with a genital lesion during delivery.
- Congenital syphilis is categorized by its time of occurrence into **early and late** stages.
- The hearing loss can develop at any time during childhood or adulthood, even as late as ~ 60 years old.
- Typical hearing loss is a **rapid symmetric progression** from high-frequency sensory loss to complete bilateral deafness. Hearing loss can also involve neural or conductive components.
- Vestibular function may also be severely affected. The **Hennebert sign**, in which nystagmus is observed as a result of pressure applied to the external auditory canal, is often found in **otosyphilis**.



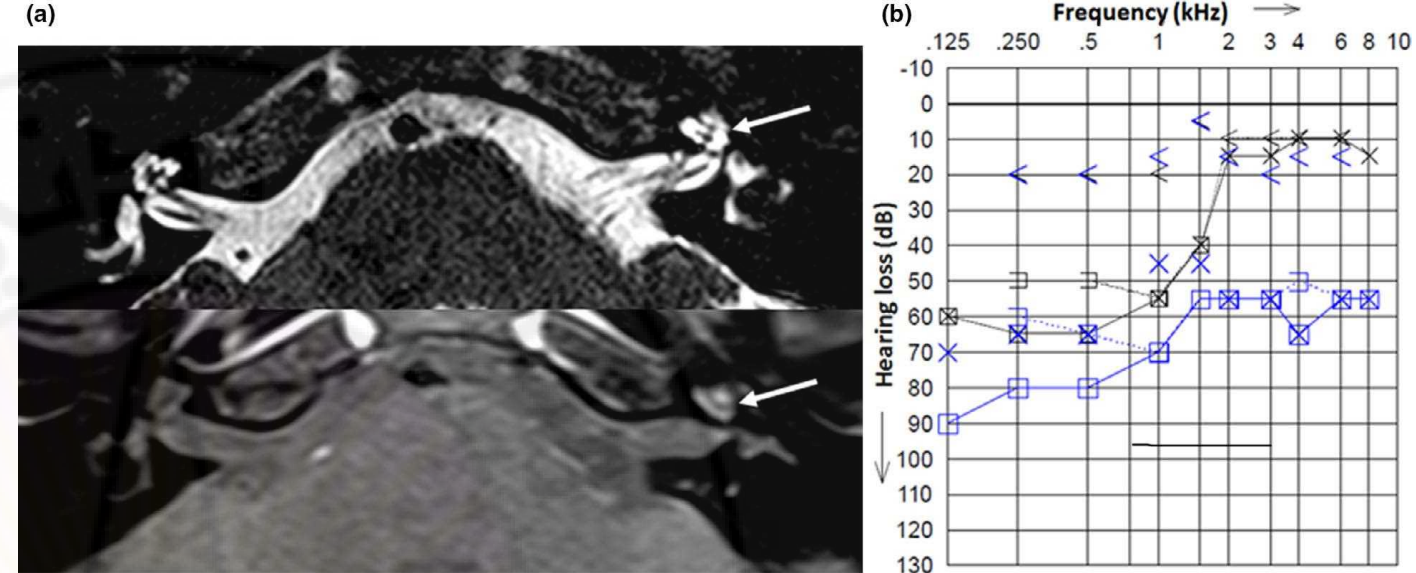
A 64-year old female with **otosyphilis**; complaints of bilateral hearing loss slowly progressive, tinnitus and intermittent unbalance for about 12 years.

The diagnosis of **otosyphilis** was based on the VDRL and FTA Abs results and the audiometric discoveries



MRI, audiograms and laboratory tests of a 31-year-old female patient with **otosyphilis**.

Pure tone audiograms during treatment period (blue line) and after 3 months (black line). While the hearing loss recovered completely in the high frequencies from 2 kHz on, moderate sensorineural hearing loss remained in the low and medium frequencies of up to 65 dB.

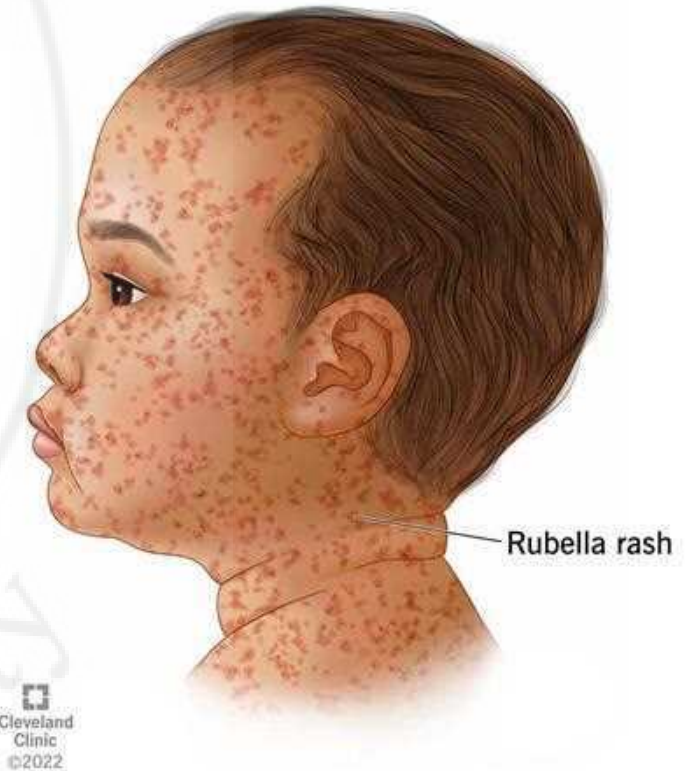


(c)	Variable	Ref.	on pres.	3 d after adm.	1 mo after pres.	3 mo after pres.
Blood						
	TPPA	<80	327680	655360	10240	5120
	RPR AB	<1	16		4	4
Treponema pallidum IgG						
	T.p. IgG p47		246		209	207
	T.p. IgG p44.5	<30	279		228	226
	T.p. IgG p17	<30	246		205	206
	T.p. IgG p15	<30	206		171	171
Treponema pallidum IgM						
	T.p. IgM p47	<50	152		101	68
	T.p. IgM p44.5	<50	<50		<50	<50
	T.p. IgM p17	<50	71		<50	<50
	T.p. IgM p15	<50	<50		<50	<50
CSF						
	cell count	<5,0/μl		21,3/μl		2,0/μl
	lymphocytes			85%		80%
	monocytes			15%		20%
	TPPA			64		8

Rubella (German measles)

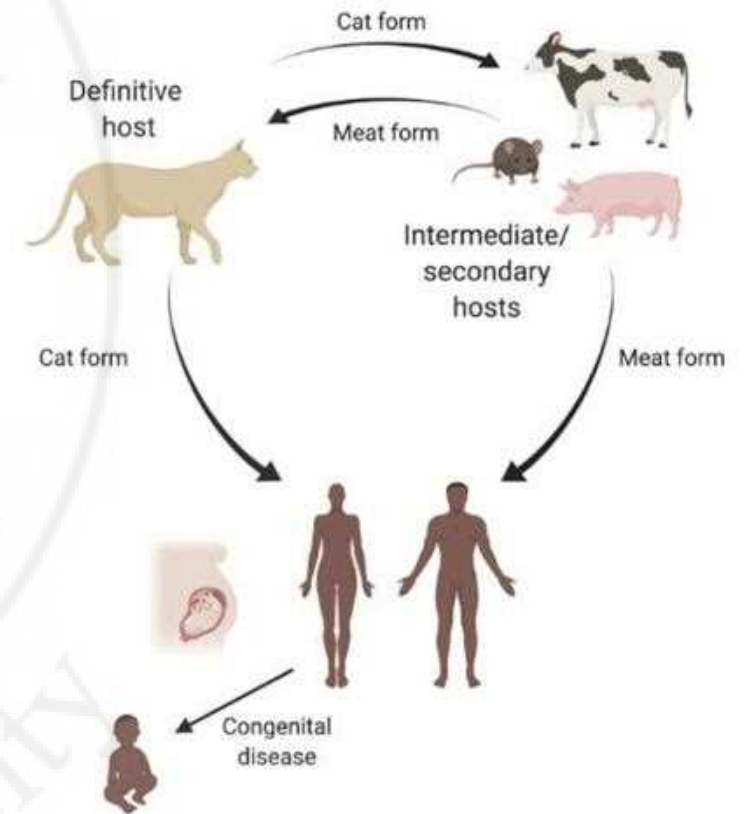
- Maternal rubella occurs when an infected mother transmits the **rubella virus** to the fetus. Expression of symptoms of maternal rubella infection, called **congenital rubella syndrome** or **Gregg syndrome**, includes a wide variety of defects, usually affecting hearing, vision, and heart function, and often involving mental retardation and microcephaly.
- **Hearing loss** is the most common manifestation of congenital rubella, occurring in up to 80% of infected children.
- Hearing loss ranges from unilateral mild loss to bilateral profound deafness; **severe bilateral losses** are more common. Configuration of the hearing loss is usually flat, and hearing sensitivity may be asymmetric.
- Children with normal pure tone thresholds may demonstrate abnormal auditory brainstem responses.

Rubella
German measles



Toxoplasmosis

- Toxoplasmosis is an infection caused by the **parasite** *Toxoplasma gondii*. The parasite can be transmitted to humans by ingestion of raw or inadequately cooked infected meat or foods that have come in contact with infected meat, ingestion of parasites that cats have passed in their feces, or transplacental transmission of the infection from a woman to her fetus.
- Those who are symptomatic often have a classic triad of chorioretinitis, intracranial calcifications, and **hydrocephalus**.
- **Auditory disorder** can occur from mastoid and cochlear inflammation and involvement of the auditory brainstem. The resulting disorder is a sensory or neural hearing loss that may range from mild unilateral loss to bilateral deafness



Herpes simplex

- Herpes simplex is a sexually transmitted viral disease that can be passed from an infected mother to the developing fetus either during pregnancy or during delivery (perinatally).
- Infected infants can be affected by central nervous system problems (e.g., microcephaly and psychomotor disorders), growth deficiencies, retinal dysplasia, and **moderate to severe sensorineural hearing loss** in one or both ears.



Human Immunodeficiency Virus (HIV)

- **HIV** is the virus that causes acquired immunodeficiency syndrome (**AIDS**).
- Congenital HIV infections are increasingly common and can result in substantial neurodevelopmental deficits.
- The prevalence of hearing impairment in HIV-seropositive patients is 14–49%.
- This hearing loss can be due to **secondary** opportunistic infections and ototoxic drugs or can be a **direct effect** of the virus on the central nervous system, peripheral nerves, vestibulocochlear nerve, or cochlea.
- Secondary infection leading to otitis media is the most common presentation in HIV patients and can lead to **conductive, sensorineural, or mixed** hearing loss. However, systemic infections in the form of cytomegalovirus, mumps, measles, Cryptococcus, tuberculosis, or ototoxicity of drugs that are used to treat these infections also contribute prominently in sensorineural hearing loss.



Zika virus

- Zika virus is now considered the newest TORCH Infectious disease, and like the others it can lead to a host of serious disabilities if transmitted to the child.
- It is a type of flavivirus, and is typically transmitted to humans by mosquitoes and ticks. Zika can be transmitted to the fetus prenatally via the placenta, and the most severe outcomes are associated with infections during the **first trimester**.
- The disorders caused by Zika include microcephaly, central nervous damage, subcortical calcifications, congenital joint contractures (arthrogryposis), hypertonia, multiple eye and optic nerve disorders, and sensorineural hearing loss, among others.
- Its estimated that **hearing losses** occur in ~ 21% of babies who had prenatal Zika infections.



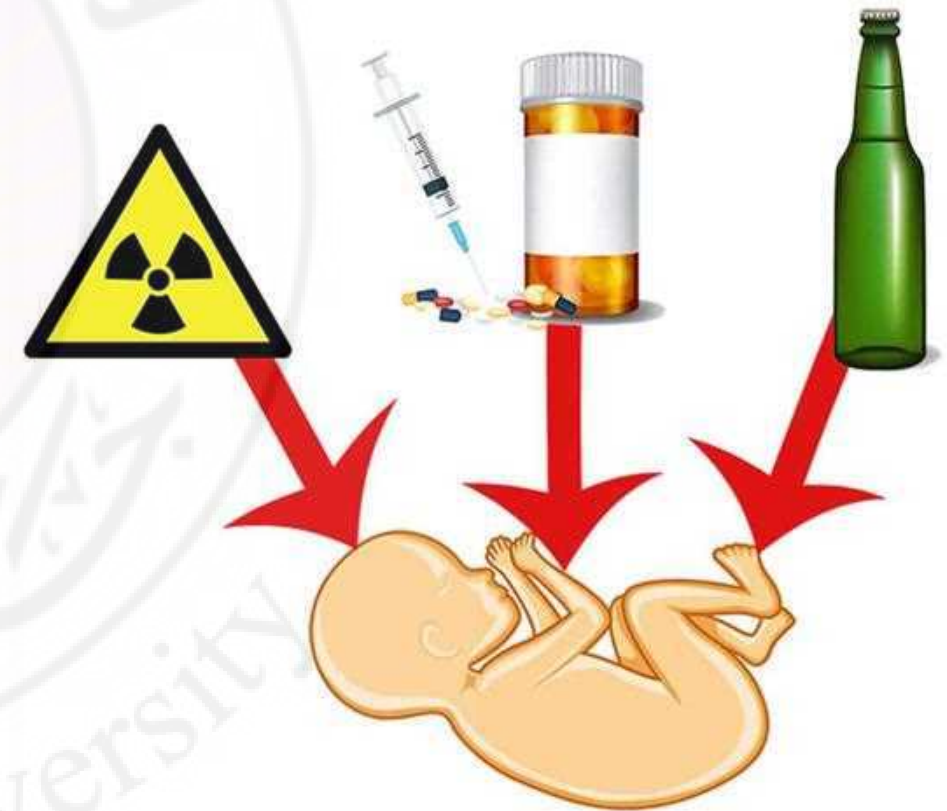
Other Teratogenic Factors

- Maternal infections are not the only adverse influences that can cause or contribute to hearing impairments in the developing fetus.
- Another group of causes includes:

- **Drugs**

- **Maternal environment**

- Maternal diseases
- Chemicals
- Radiations & Heat
- Other agents (Alcohol, Nicotine, ...)



Drugs

- Congenital hearing loss can be caused by the maternal use of certain **medications** that may be passed to the fetus via the placenta.
- The most prominent group of drugs are the **aminoglycoside** antibiotics, which are sometimes essential in the treatment of severe infections.
- Ingestion of some other drugs, especially early in pregnancy, can result in multiple developmental abnormalities, including profound sensorineural hearing loss. These drugs include **accutane, dilantin, quinine, and thalidomide**.

**Aminoglycosides
include: TANGS:**

Tobramycin

Amikacin

Neomycin

Gentamicin

Streptomycin

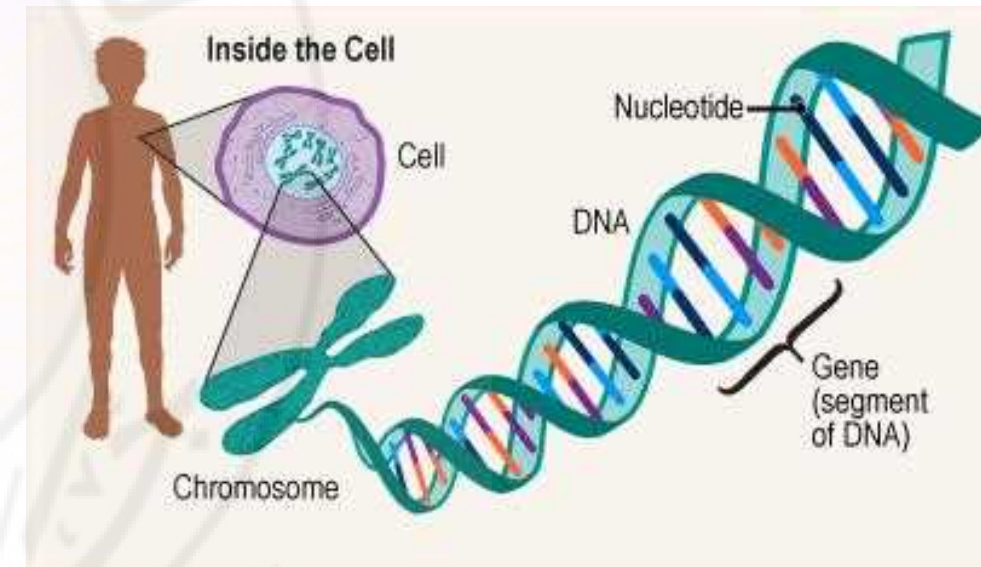
Maternal environment

- Other aspects of the maternal environment that can contribute to congenital hearing impairments and related anomalies include **maternal diseases** such as toxemia and diabetes, nutritional deficiencies and disturbances, Rh-factor incompatibility between the mother and fetus, **exposure to physical agents** such as heat and radiation, and **use of nonmedicinal drugs** and chemicals such as alcohol.



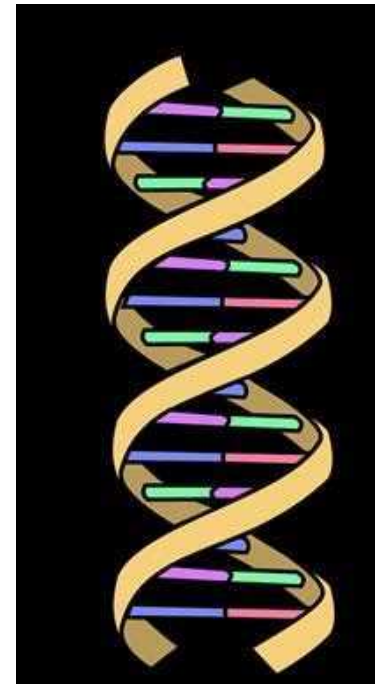
Inherited Sensory Hearing Disorders

- The information needed to make a particular person (the genetic code) exists in the form of deoxyribonucleic acid (**DNA**) molecules packaged as chromosomes in every cell.
- **Genes** are segments of DNA that occur at fixed locations on the chromosomes, and operate as the biological units of inheritance. In other words, hereditary characteristics (traits) are carried by genes, which are arranged along the chromosomes.
- Humans have **46 chromosomes** arranged in 23 pairs.
- There is one pair of sex chromosomes, **XX** for females and **XY** for males, and the other 22 pairs are autosomes, which are the same for both sexes. Each parent contributes half of each pair.



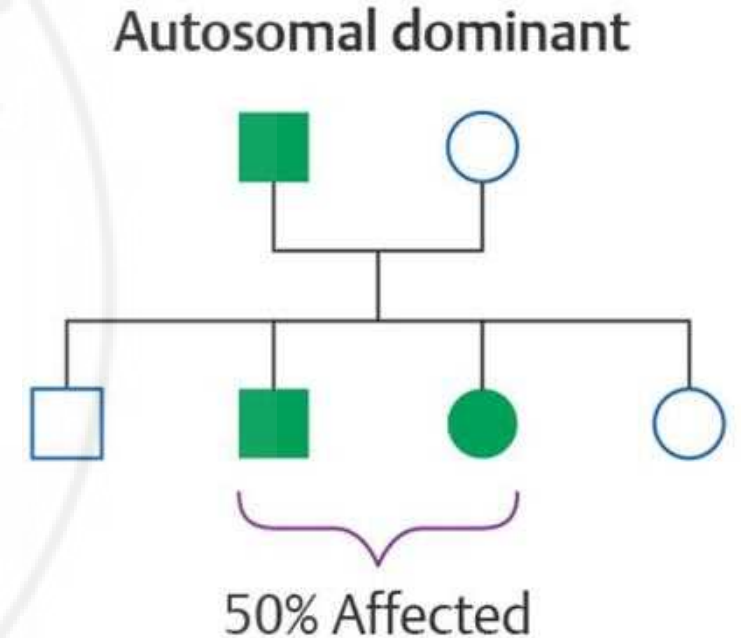
Inherited Sensory Hearing Disorders

- **Hereditary or genetic disorders** are abnormal traits that are transmitted by abnormal genes.
- They account for the largest proportion of **congenital hearing losses**, although exact estimates vary. For example, Morton & Nance (2006) estimated that ~ **68%** of congenital hearing losses are hereditary, and Smith et al (2013) placed the incidence at ~ **50%** of prelingual hearing losses ≥ 40 dB.
- Disorders may be the result of single- or multiple-gene abnormalities, or multifactorial inheritance, which is the combined effect of genetic and environmental factors.
- Hereditary hearing losses can occur alone (**nonsyndromic**) or in combination with other genetic abnormalities (**syndromic**), and may be transmitted by **autosomal dominant, autosomal recessive, and X-linked inheritance, and by mitochondrial mutations**, and are subject to a considerable amount of variability.



Autosomal dominant inheritance

- **Autosomal dominant inheritance** means that only one abnormal gene is needed in order for the trait to appear (hence, it is dominant).
- Suppose one parent is hard-of-hearing due to an autosomal dominant gene and the other parent has normal hearing. It does not make a difference which parent is affected because the sperm and egg each contribute half of every chromosome pair.
- If the mother is affected, then there will be a 50% chance that a given egg will contain the abnormal gene. If the father is affected, then half of his sperm will carry the abnormal gene and half will not. In either case, the chances are 50–50 for any pregnancy to result in a child that is hard-of-hearing.



Circles, females; squares, males; open symbols, individuals free of the abnormal gene; filled symbols, affected individuals with the abnormal gene

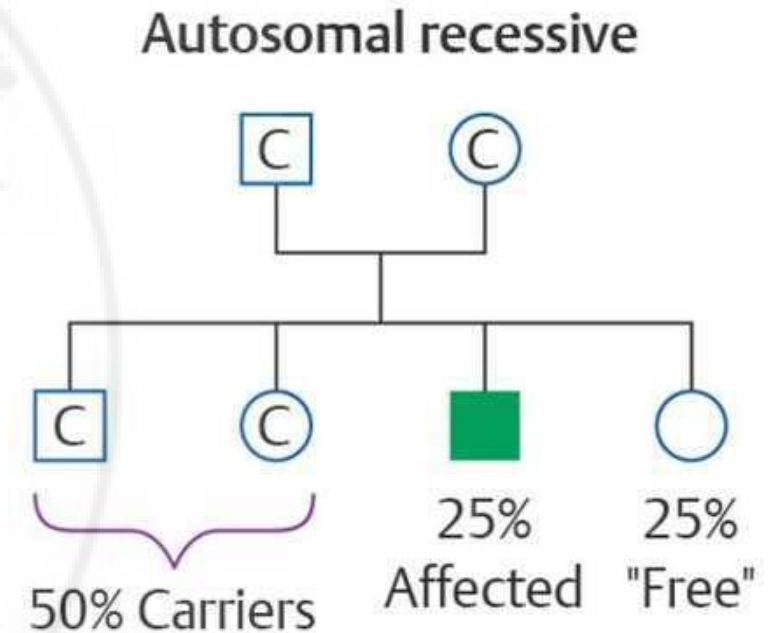
Autosomal recessive inheritance

- The situation is different with **autosomal recessive inheritance** because both genes in the pair must be present for the trait to manifest itself.
- There are three possibilities with respect to the abnormal recessive gene:

(1) One may be altogether free of the abnormal gene.

(2) A person with both abnormal genes will have the disorder and will be able to transmit the disorder to an offspring. Such an individual is a **homozygote** because the same (homo) kind of abnormal gene was contributed by both parents when the fertilized egg (zygote) was produced.

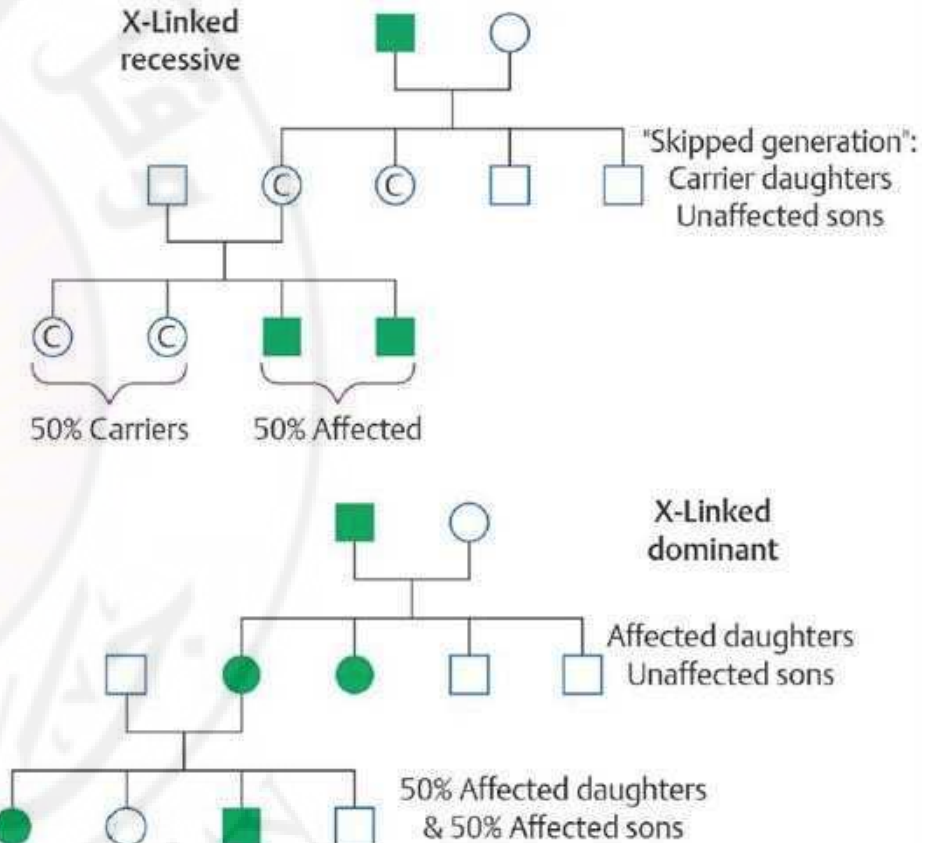
(3) An individual may have only one abnormal recessive gene, while the other one in the pair is normal. This person is a **heterozygote** because the two genes in the pair are different (hetero). This person will not have a hearing loss because they only have one abnormal gene and it is recessive, but they will be a carrier because they can transmit the abnormal gene to their offspring.



Circles, females; squares, males; open symbols, individuals free of the abnormal gene; filled symbols, affected individuals with the abnormal gene; "C," unaffected carriers of the abnormal gene.

X-linked (or sex-linked) inheritance

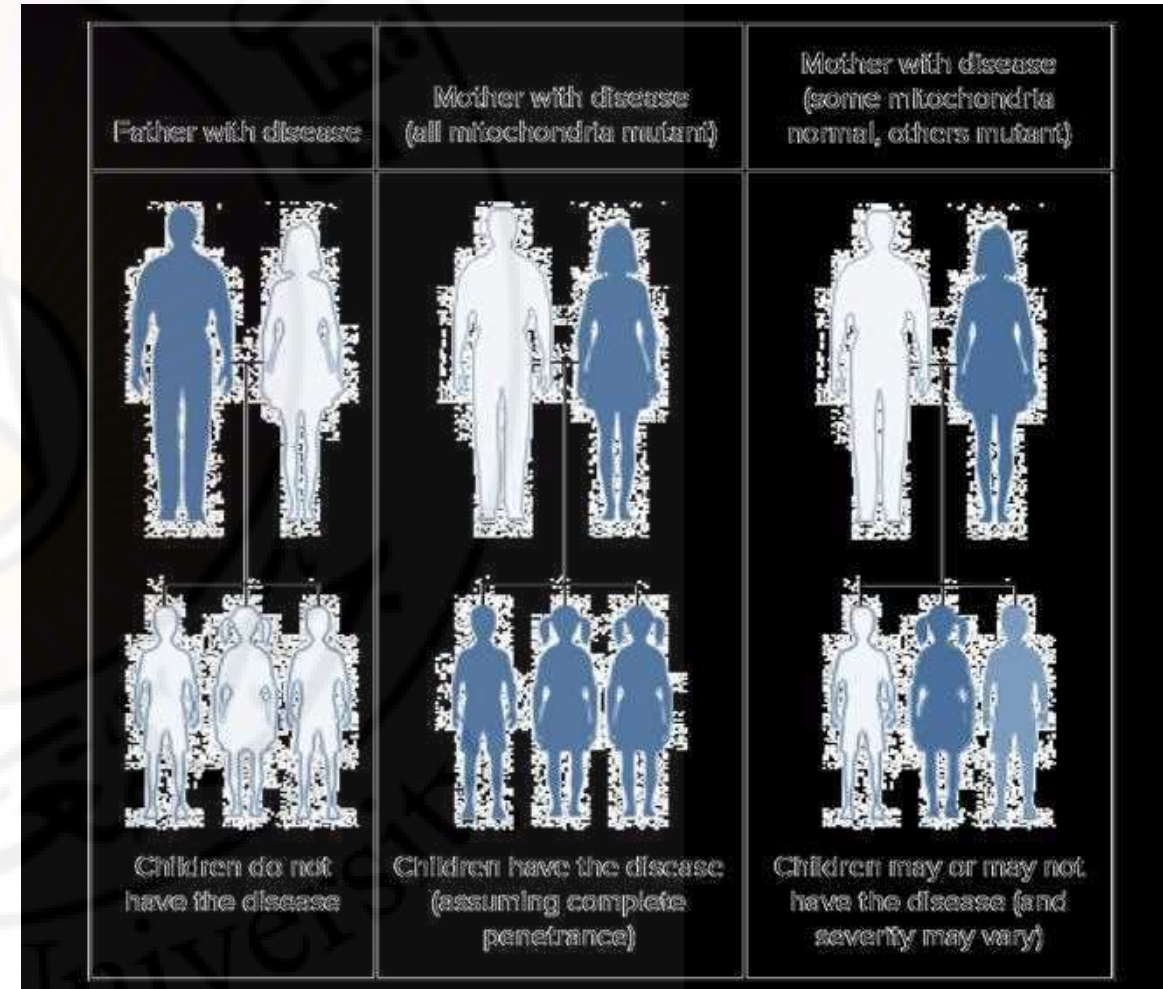
- **X-linked (or sex-linked) inheritance** occurs when the gene is associated with the X chromosome instead of being autosomal.
- With an **X-linked recessive disorder**, an affected male transmits the abnormal gene to all of his daughters, who become unaffected carriers of the trait, but not to his sons. In turn, a female carrier has a 50% chance of having sons who are affected and a 50% chance of having daughters who are unaffected carriers.
- In **X-linked dominant inheritance**, an affected male will have all affected daughters and no affected sons. An affected female has a 50% chance of having affected children, whether they are males or females.



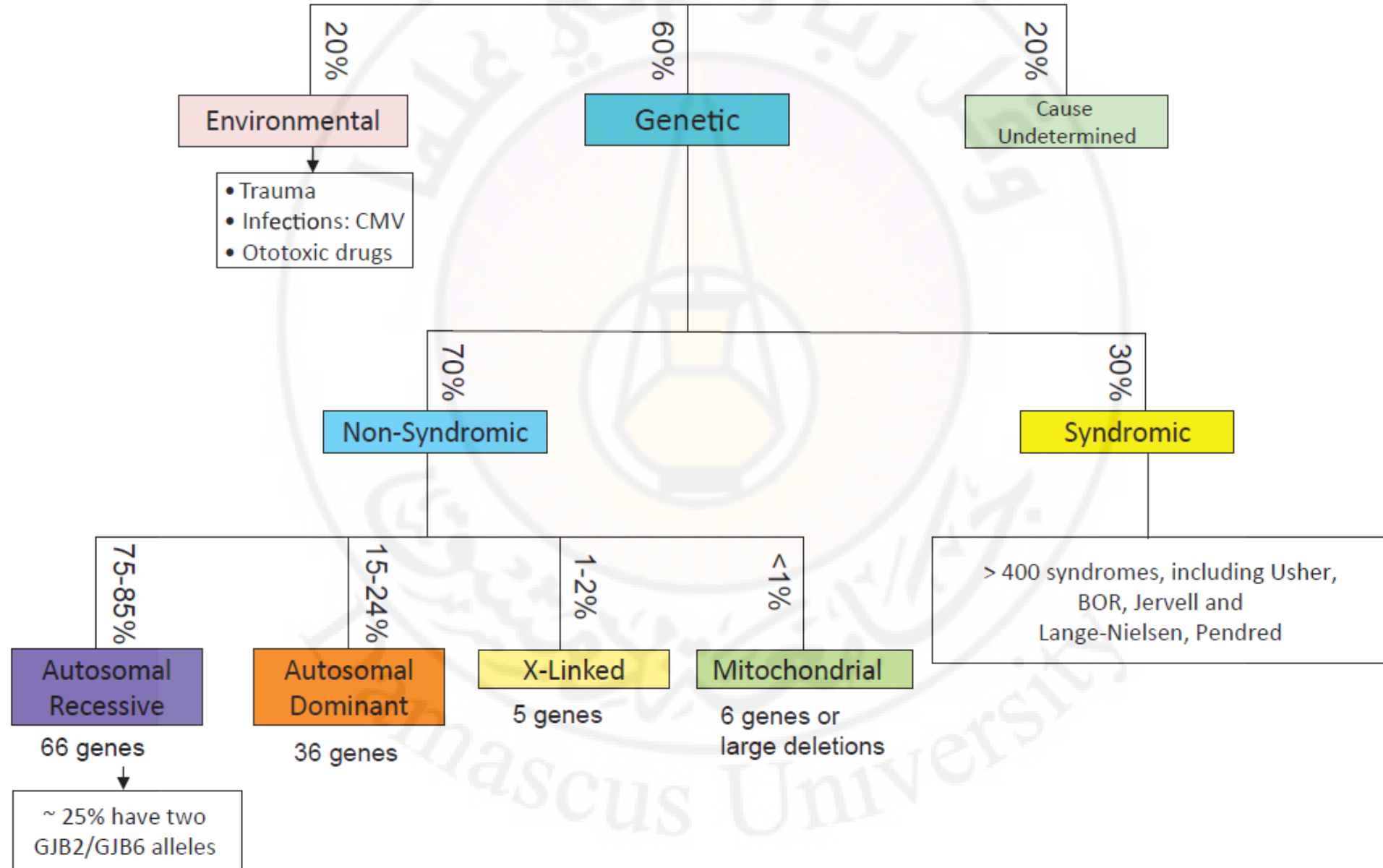
Circles, females; squares, males; open symbols, individuals free of the abnormal gene; filled symbols, affected individuals with the abnormal gene

Mitochondrial inheritance

- In contrast to the DNA in the nucleus, which is inherited from both parents, the genes of the mitochondrial DNA are inherited from the **mother alone**.
- **Mitochondrial mutations** are associated with both syndromic and nonsyndromic deafness, and are often related to aminoglycoside ototoxicity.



Etiologies of Childhood Hearing Loss



Syndromic Hearing Loss

- Approximately **30%** of significant hereditary hearing losses occur in syndromes.
- A **syndrome** is a pattern of abnormalities and/or symptoms that result from the same cause.
- **Auditory disorders** can be found in a great many syndromes, and there are **over 400 syndromes** identified that claim auditory disorder as a component.
- Different syndromes are associated with different kinds of hearing loss; some appear to be *present at birth (congenital)*, whereas others are *delayed in onset*.



Syndrome	Inheritance	Hearing loss	Characteristics
Apert syndrome	Autosomal dominant: FGFR2	Conductive (congenital)	Craniofacial anomalies affecting the ears, stapes fixation, fused fingers and toes, and spina bifida; associated with conductive hearing loss
Alport syndrome	X-linked: ~ 80% COL4A5 Autosomal recessive: ~ 15% COL4A3, COL4A4 Autosomal dominant: ~ 5% single mutation of COL4A3 or COL4A4	Sensorineural (delayed onset, progressive)	Varies by type: renal disease, ocular disorders, blood platelet defect, hearing loss
Autosomal dominant optic atrophy plus (ADOA plus) Syndrome	Autosomal dominant: OPA1	sensorineural (child/adolescent onset, often progressive); ANSD	Variable; usually optic nerve atrophy with visual impairment, reduced pain sensation, decreased tendon reflexes, impaired color vision; developing in adulthood peripheral neuropathy, myopathy, ophthalmoplegia, ataxia, hearing loss, rare
Branchio-oto-renal, syndrome	Autosomal dominant: EYA1, SIX1, SIX5	Sensorineural, conductive, mixed (congenital or delayed onset)	Outer, middle, inner ear deformities (characteristic ear pits), renal disorders, branchial fistulas/cysts, hearing loss
CHARGE syndrome	CHD7 (> 50% of cases)	Mixed, progressive	Coloboma (malformation of iris giving the pupil a "keyhole" appearance, disk, or retina); congenital heart defects; atresia of the choanae (so posterior nasal airway is obstructed); retarded development and/or growth; ear anomalies and/or hearing loss

Syndrome	Inheritance	Hearing loss	Characteristics
CAPOS (cerebellar ataxia, areflexia, pes cavus, optic atrophy, sensorineural hearing loss) syndrome	Autosomal dominant: ATP1A3	Sensorineural progressive; ANSD	Early-childhood onset acute fever-related encephalopathy in recurring episodes, decline over time, results in permanent, progressive, ataxia, areflexia, visual impairment, hearing loss; sometime pes cavus foot deformity (very high arch), nystagmus or strabismus), dysarthria, dysphagia, rare
Charcot-Marie-Tooth disease type 1 (CMT1)	Autosomal dominant: PMP22 (CMT1a), MPZ (CMT1b)	sensorineural; ANSD	Demyelinating peripheral nervous system disorder with weakness/atrophy/sensory loss mainly of distal muscles, starting in legs/feet; onset infancy to ~ 20 years; hearing loss; CMT1a due to PMP22 mutation, most common; CMT1b due to MPZ mutation, less common, more variable
Cowchock syndrome	x-linked recessive: AIFM1	sensorineural; ANSD	Also known as x-linked recessive Charcot- Marie-Tooth disease 4 with or without cerebellar ataxia. Variable onset (infancy to young adult), slowly progressive axonal sensorimotor neuropathy, hearing impairment with ANSD; delayed motor development, peripheral neuropathy and/or cerebellar ataxia, cognitive impairment, hearing loss, rare
Crouzon syndrome	Autosomal dominant: FGFR2	Conductive, mixed (congenital)	Prematurely fused cranial sutures, beakshaped nose, exophthalmos, variable outer/middle ear anomalies, hearing loss
Deafness-dystonia-optic neuropathy (DDON, 2Mohr-Tranebjærg) Syndrome	x-linked recessive: TIMM8A	Sensorineural, progressive; ANSD	Childhood-onset progressive disorders of vision (optic neuropathy), movement (dystonia, ataxia), and sensorineural hearing loss; psychiatric problems; usually affects males

Syndrome	Inheritance	Hearing loss	Characteristics
Down (trisomy 21) syndrome	Improper separation of chromosome pairs, causing an extra copy of chromosome 21	Conductive, mixed	Multisystem disorder; intellectual developmental disorder; characteristic facial features (tilted palpebral fissures, epicanthal folds, broad nasal bridge, flattened profile, open mouth with protruding tongue); outer/middle/inner ear hypoplasias have been reported; muscular hypotonia; short hands with simian line; congenital heart disease; frequent upper respiratory infections; recurrent otitis media, hearing loss
Friedreich's ataxia	Autosomal recessive: FXN	Sensorineural (delayed onset, progressive)	Ataxia, nystagmus, optic atrophy, hearing loss
Klippel-Feil syndrome	Autosomal dominant: GDF3, GDF6	Sensorineural, conductive (congenital)	Fused cervical vertebrae, short neck, decreased head mobility, low occipital hairline, ossicle abnormalities, hearing loss
Hunter syndrome (mucopolysaccharidosis II)	X-linked: IDS	Sensorineural, conductive, mixed (progressive)	Skeletal deformities, dwarfism, coarse facial features, corneal clouding, cardiovascular disorders, intellectual developmental disorder; mucopolysaccharide accumulations in tissues result in progressive characteristics, hearing loss; males affected only
Hurler syndrome (mucopolysaccharidosis I)	Autosomal recessive: IDUA	Sensorineural, conductive, mixed (progressive)	Same features as Hunter syndrome, but may be more severe; affects both sexes

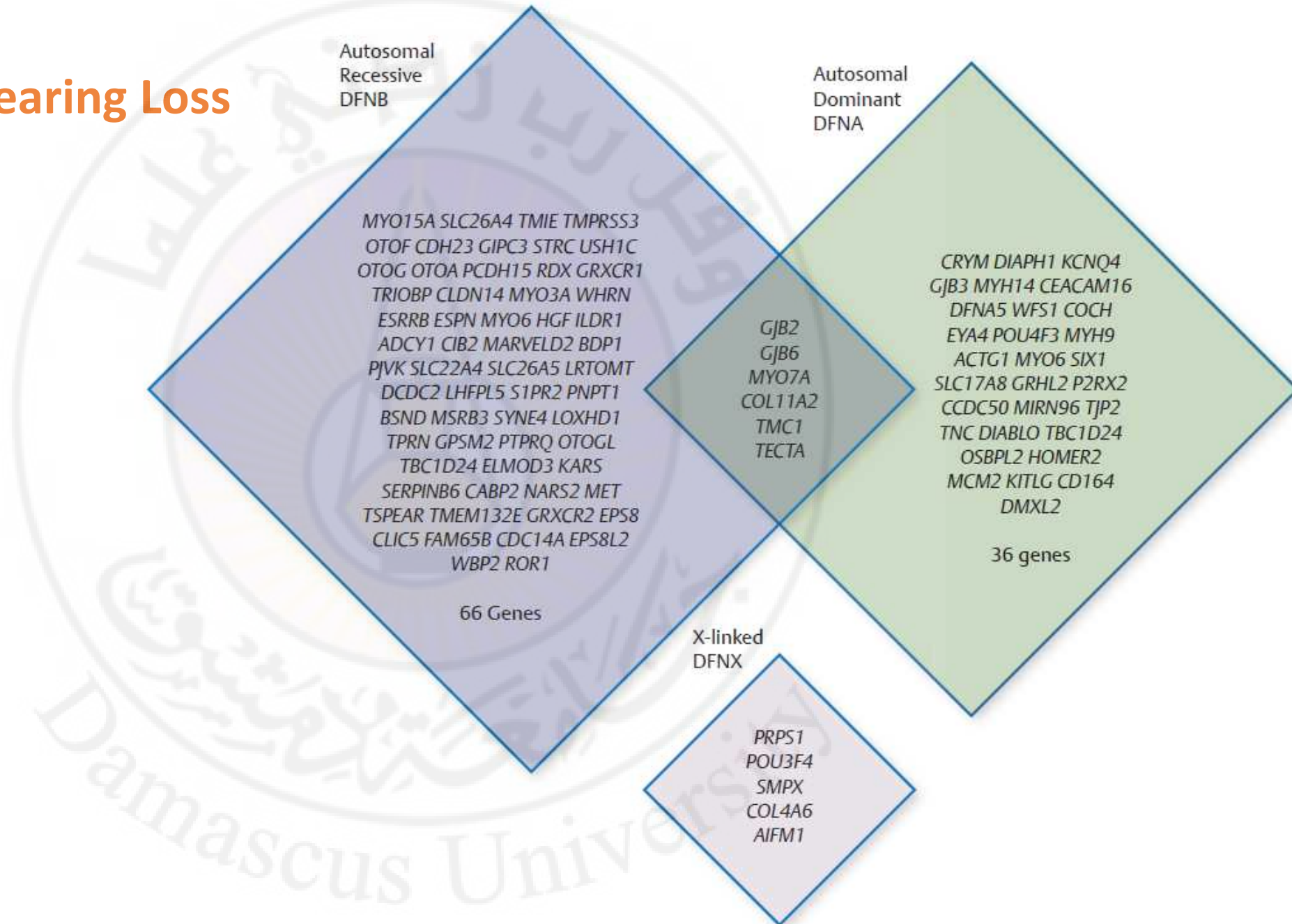
Syndrome	Inheritance	Hearing loss	Characteristics
Jervell and Lange-Nielsen syndrome	Autosomal recessive: KCNE1, KCNQ1	Sensorineural	Cardiac problems, syncope, sudden death, hearing loss
Oculo-auriculo-vertebral spectrum (OAVS; Goldenhar syndrome, hemifacial microstomia, hemifacial microstomia)	Undetermined, heterogeneous	Conductive (congenital)	Facial asymmetry, microtia, atresia, preauricular tags, eye anomalies, oral defects, clubfoot, hemivertebrae, congenital heart disease, abnormal semicircular canals, hearing loss
Osteogenesis imperfecta	Autosomal dominant: COL1A1, COL1A2	Sensorineural, conductive, Mixed (progressive)	Fragile bones, large skull, triangular faces, hemorrhage tendency, stapes fixation, hearing loss
Pendred syndrome	Autosomal recessive: ~ 50% involve SLC26a4; also FOXI1, KCNJ10	Sensorineural (variable, congenital, progressive); ANSD	Hearing loss and thyroid enlargement (goiter), both variable
Stickler/Marshall syndromes	Autosomal dominant: COL2A1, COL11A1 Autosomal recessive: COL9A1, COL9A2, LOXL3 AD/ARc: COL11A2	Sensorineural, conductive, mixed	Three types: severe eye disorders (detached retina, myopia) in types 1 and 2, but not in type 3; small lower jaw, hearing loss

Syndrome	Inheritance	Hearing loss	Characteristics
Treacher Collins syndrome (mandibulofacial dysostosis)	Autosomal dominant: TCOF1 (mainly), POLR1D Autosomal recessive: POLR1C	Conductive, mixed (congenital)	Facial anomalies (depressed zygomatics, eyes slant downward laterally, receding mandible, mouth large and fish-like, dental anomalies, cleft palate); outer and middle ear deformities, hearing loss
Usher Syndrome (Vestibulocerebellar ataxia)	Autosomal recessive: ADGRV1, CDH23, CLRN1, MYO7A, PCDH15, USH1, USH1G, USH2A, WHRN	Sensorineural (congenital in types 1 & 2); ANSD (type 1)	Type 1: retinitis pigmentosa, vestibular dysfunction, profound hearing loss Type 2: retinitis pigmentosa, sloping hearing loss (variable degree) Type 3: retinitis pigmentosa, vestibular dysfunction initially normal hearing or mild loss, then hearing loss progresses
Waardenburg syndrome	Autosomal dominant: KITLG, MITF, PAX3, SNAI2, SOX10 AD/ARc: EDN3, EDNRB	Sensorineural (congenital)	White forelock in hair, prominent root of nose, differently colored eyes, hyperplasia of medial third of eyebrows, medial canthi displaced laterally; inner ear dysplasia, hearing loss Four types identified, differing by abnormalities present and/or genes involved
Williams (Williams-Beuren) syndrome	Spontaneous (sometimes autosomal dominant) chromosome 7 gene deletion(s): CLIP2, ELN, GTF2I, GTF2IRD1, and LIMK1	Sensorineural (typically highfrequency); Hyperacusis	Cardiovascular abnormalities (especially supravalvular aortic stenosis), facial characteristics (e.g., broad forehead, upturned nose, long philtrum, wide mouth, pointed chin), intellectual disorder (but language skills often preserved), impaired visual-spatial skills, hypercalcemia, hypertension, hyperacusis, hearing loss

Non-syndromic Hearing Loss

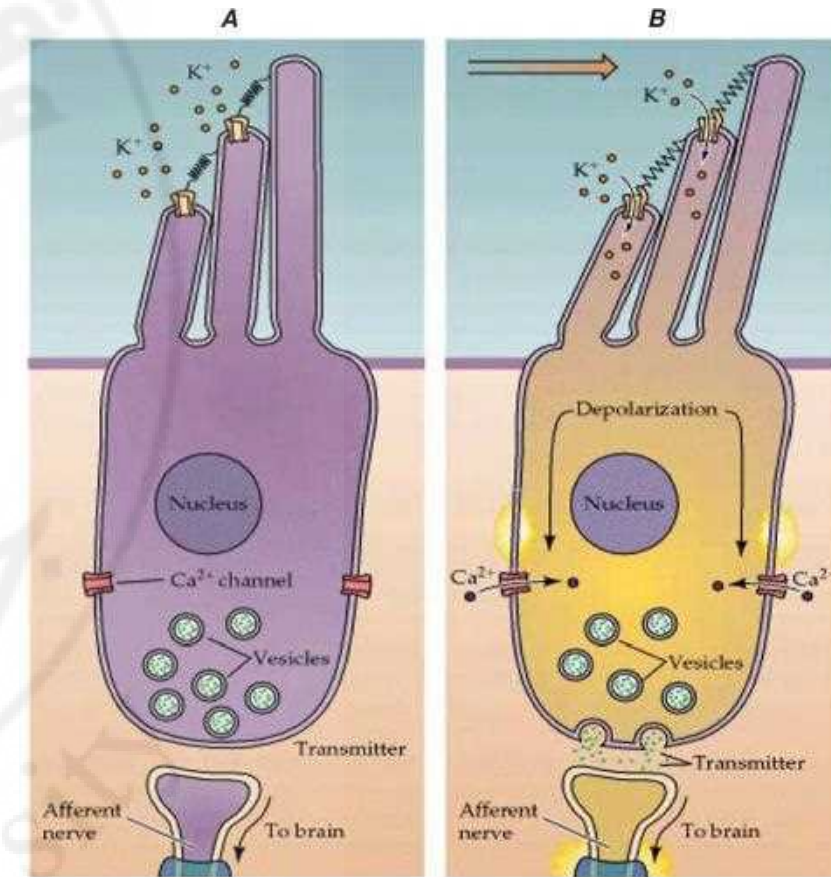
- About **70%** of hereditary hearing losses are nonsyndromic, of which ~ 75 to 85% are autosomal recessive, ~ 15 to 20% are autosomal dominant, and ~ 1 to 2% are X-linked and mitochondrial in origin.
- In fact, nonsyndromic hereditary hearing loss is a **primary cause** of sensorineural hearing loss in infants and young children and probably contributes substantially to much of the acquired hearing loss in aging patients.
- These hearing losses are identified by **DFN** (for “deafness”) followed by **A** for dominant, **B** for recessive, or **X** for X-linked, and then a **number**.
- For example, DFNA1 is an autosomal dominant hearing loss involving the DIAPH1 gene, DFNB21 is an autosomal recessive hearing loss associated with the TECTA gene, and DFNX1 is an X-linked hearing loss related to the PRPS1 gene.

Non-syndromic Hearing Loss



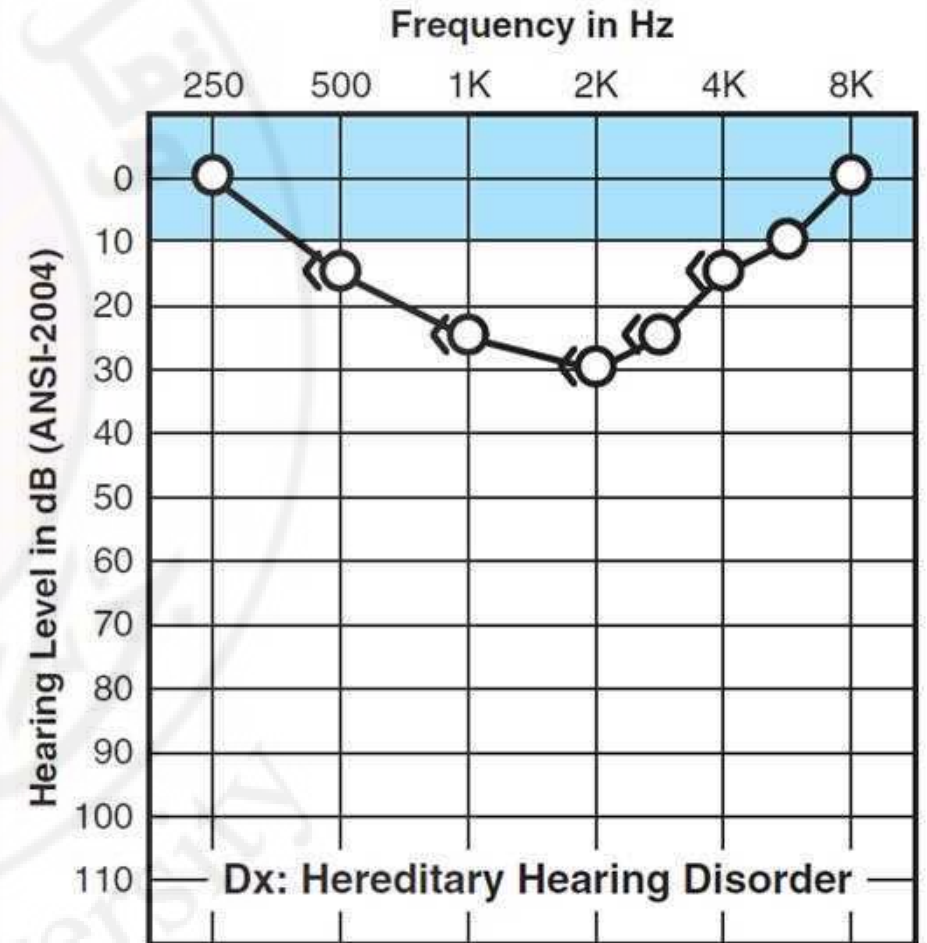
Non-syndromic Hearing Loss

- The most common form of genetic hearing loss is **DFNB1**, which accounts for about half of the autosomal recessive cases.
- Children born with DFNB1 have prelingual bilateral sensorineural hearing losses that can vary in degree of severity from mild to profound, and are nonprogressive. It is caused by mutations of the **GJB2**, which provide for the expression of proteins in the inner ear called **connexin 26 and 30**, respectively.
- These connexins form gap junctions, which are channels that permit substances such as ions to flow across cell membranes, and are thus essential for maintaining the required balance and flow of potassium ions and other functions in the inner ear; however, the exact mechanism of the hearing loss is not fully understood.



Non-syndromic Hearing Loss

- Hereditary hearing disorder may be **present at birth**, or it may have a **delayed onset** and be **progressive** in nature.
- The majority of nonsyndromic hereditary hearing disorders are autosomal recessive, and the hearing loss is predominantly sensorineural in nature.
- Nonsyndromic hearing disorders can be classified in a number of ways:
 - **Dominant hereditary hearing loss**
 - **Dominant progressive hearing loss**
 - **Progressive adult-onset hearing loss**
 - **Recessive hereditary sensorineural hearing loss:**
 - **X-linked hearing disorder**



Top websites on hereditary hearing loss

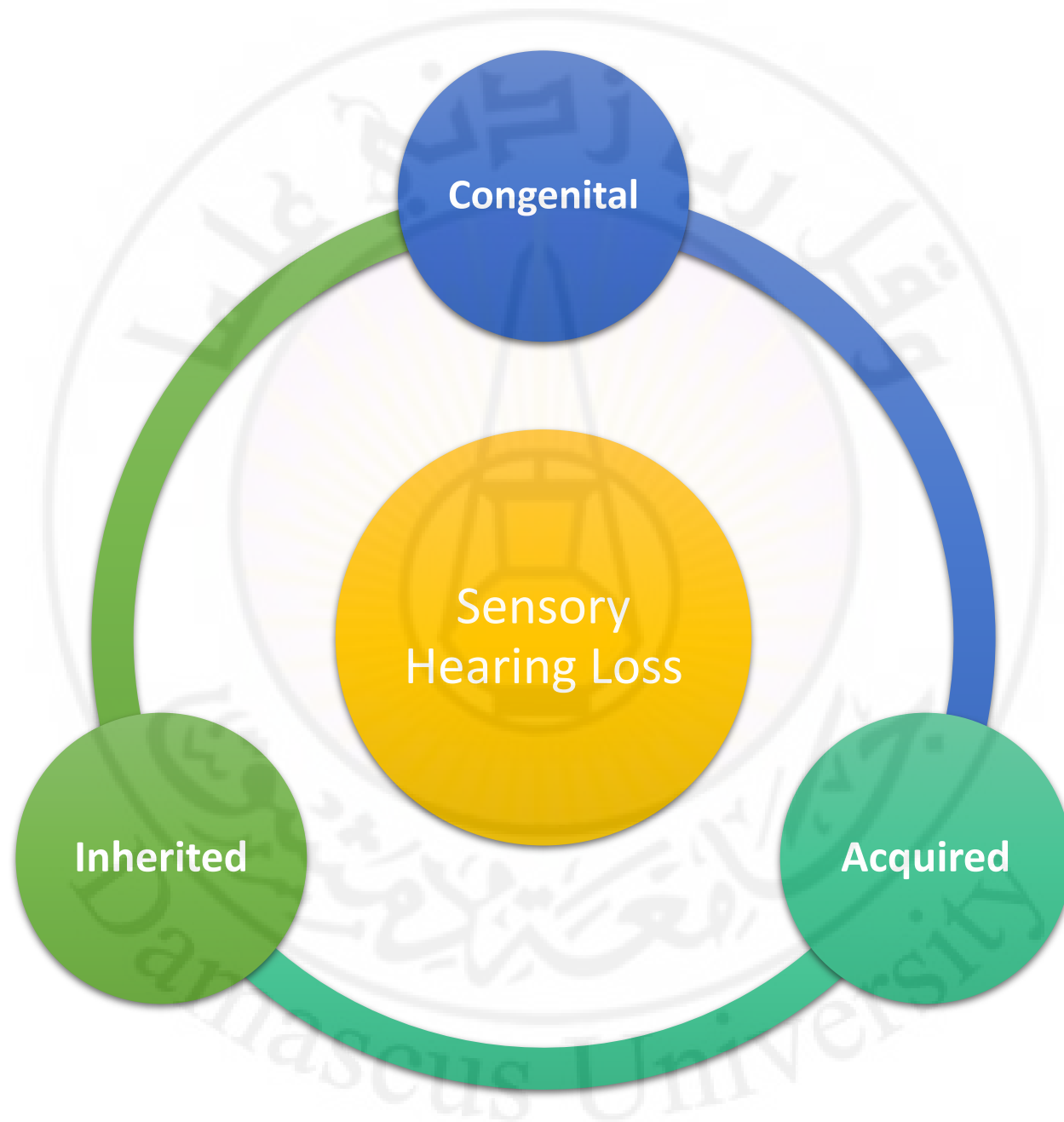
- <http://deafnessvariationdatabase.org>
- <https://hereditaryhearingloss.org>
- <https://ghr.nlm.nih.gov/gene>
- <https://omim.org/>
- <http://www.genecards.org>



Clinical Audiology

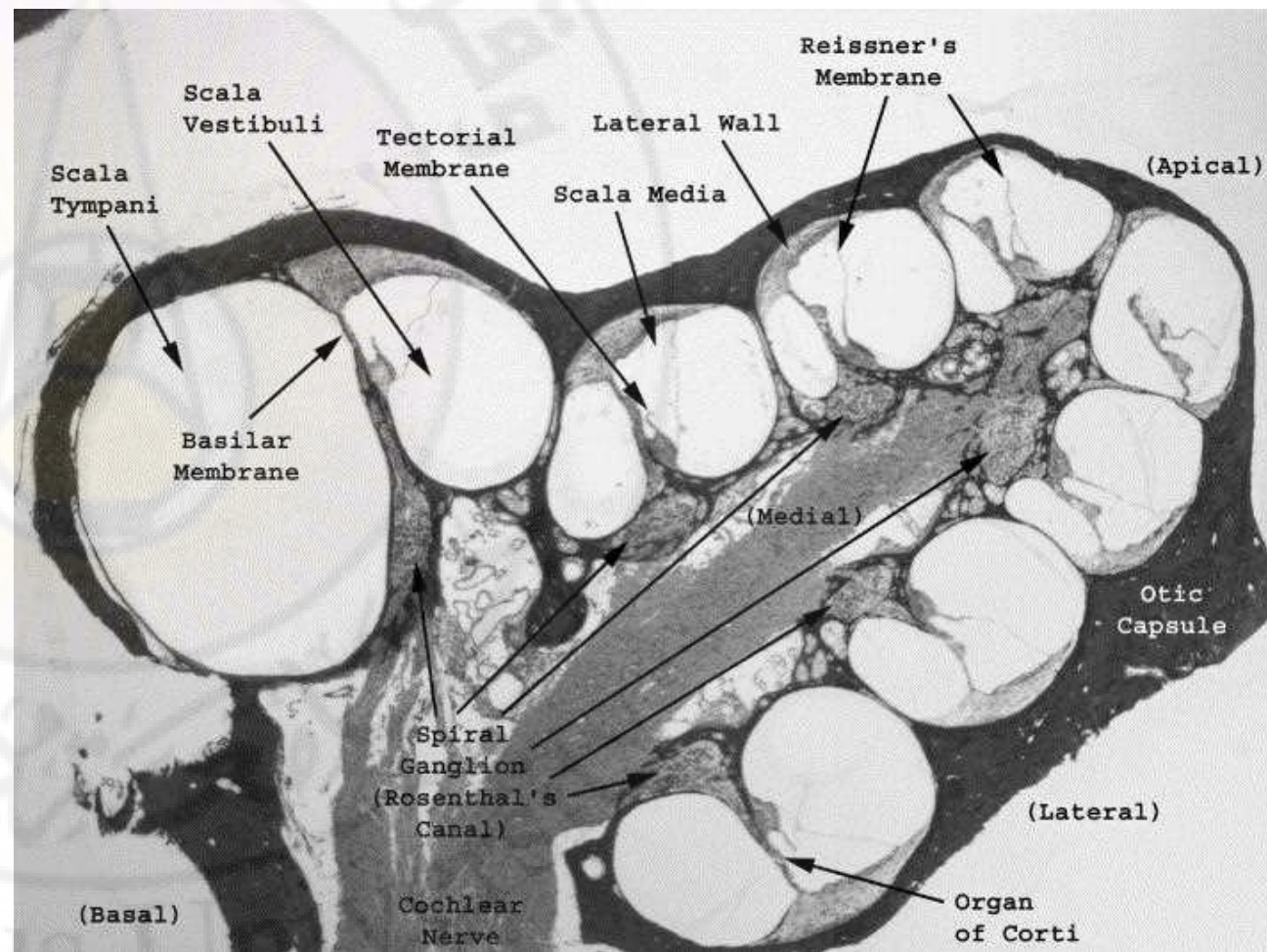
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Acquired Sensory Hearing Disorders

- Perinatal Factors
- Cochlear synaptopathy
- Noise-induced Hearing Loss
- Trauma
- Infections
- Ototoxicity
- Ménière's Disease
- Perilymphatic Fistulas
- Presbycusis
- Autoimmune Inner-Ear Disease
- Cochlear Otosclerosis
- Idiopathic Sudden Sensorineural Hearing Loss
- Cochlear dead regions
- Superior Canal Dehiscence



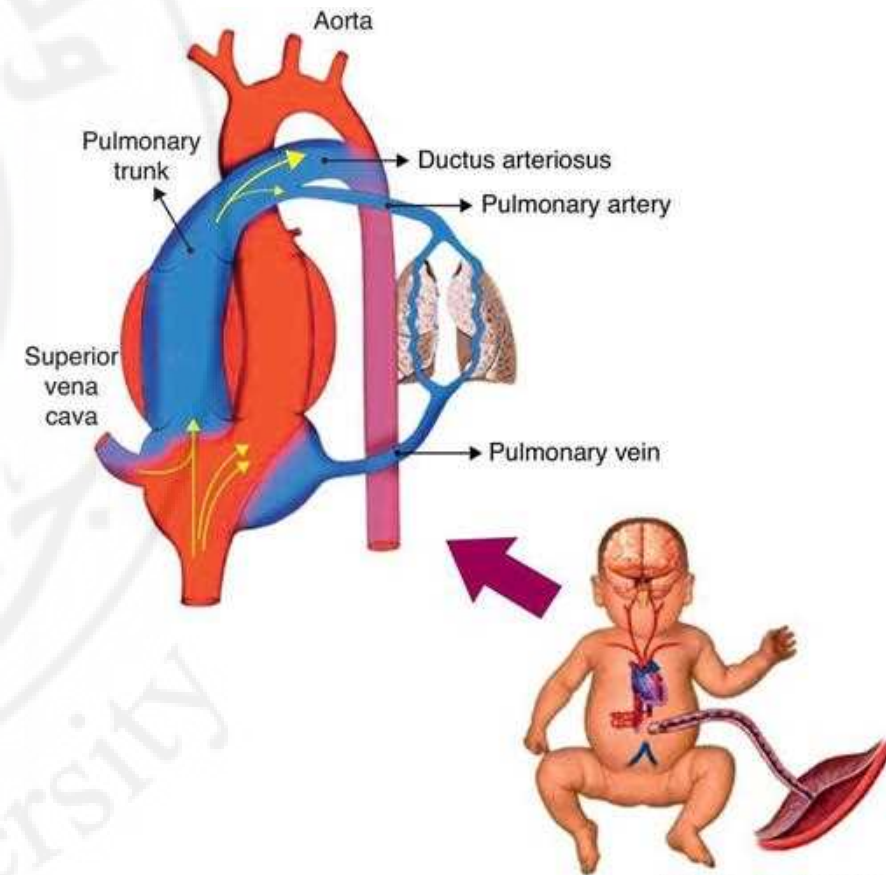
Perinatal Factors

- Adverse factors that occur **just before, during, and immediately after birth** include **compromise of the oxygen supply (asphyxia, anoxia, hypoxia), trauma during labor and/or delivery, hyperbilirubinemia (jaundice) leading to kernicterus, and infections.**
- Infants who have a traumatic perinatal period are at a significant risk for hearing loss.
- The underlying cause of the hearing loss may be unknown, but hearing loss has been associated with a history of low birth weight, hypoxia, hyperbilirubinemia, and exposure to potentially ototoxic drugs.
- Indeed, mere length-of-stay in the **intensive care nursery (ICN)**, with all the various potential adverse influences on the auditory system, has been associated with increased risk for hearing loss.
- Of all factors, one that has been clearly linked to hearing loss is **severe respiratory distress at birth.**



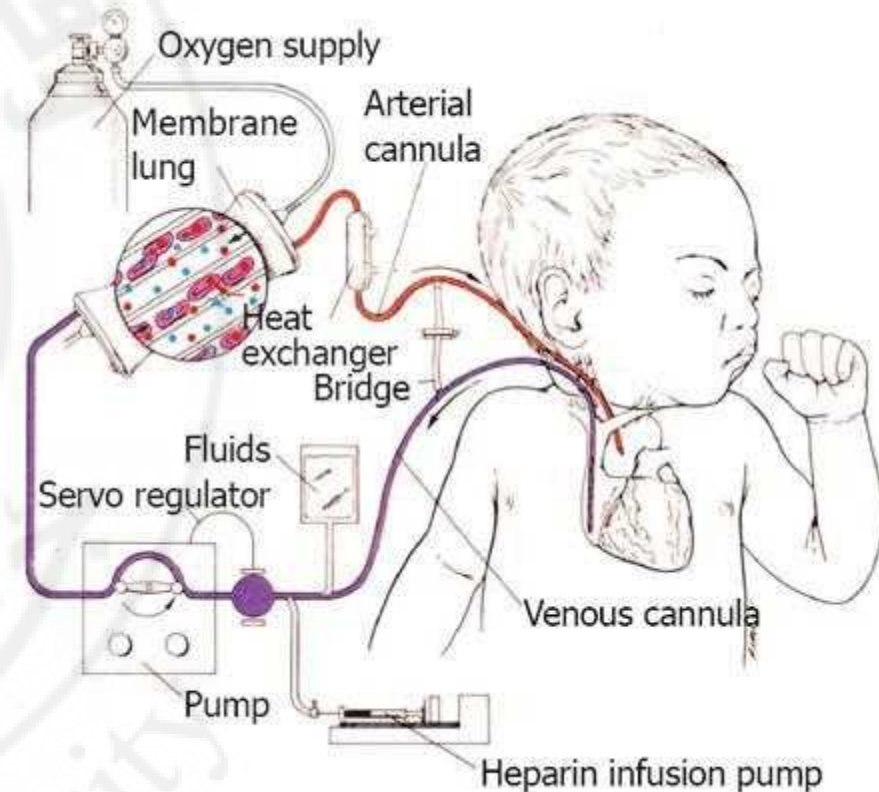
Persistent Pulmonary Hypertension of the Newborn

- **PPHN**, also known as persistent fetal circulation, is a condition wherein the infant's blood flow bypasses the lungs, thereby eliminating oxygen supply to the organs of the body.
- PPHN is associated with perinatal respiratory problems such as ***meconium aspiration*** or ***pneumonia***.
- Sensorineural hearing loss is a common complication of PPHN and has been found in approximately one third of surviving children.
- Hearing loss ranges from high-frequency unilateral loss to severe to profound bilateral loss and is progressive in many cases.



Extracorporeal Membrane Oxygenation

- PPHN is treated by administration of oxygen or oxygen and nitric oxide via a mechanical ventilator.
- **Extracorporeal membrane oxygenation (ECMO)** is a treatment for PPHN that involves diverting blood from the heart and lungs to an external bypass, where oxygen and carbon dioxide are exchanged before the blood reenters the body.
- When ECMO is applied as part of respiratory management, it has been associated with **hearing loss** in up to **75% of cases**.
- Hearing loss is often **progressive**.



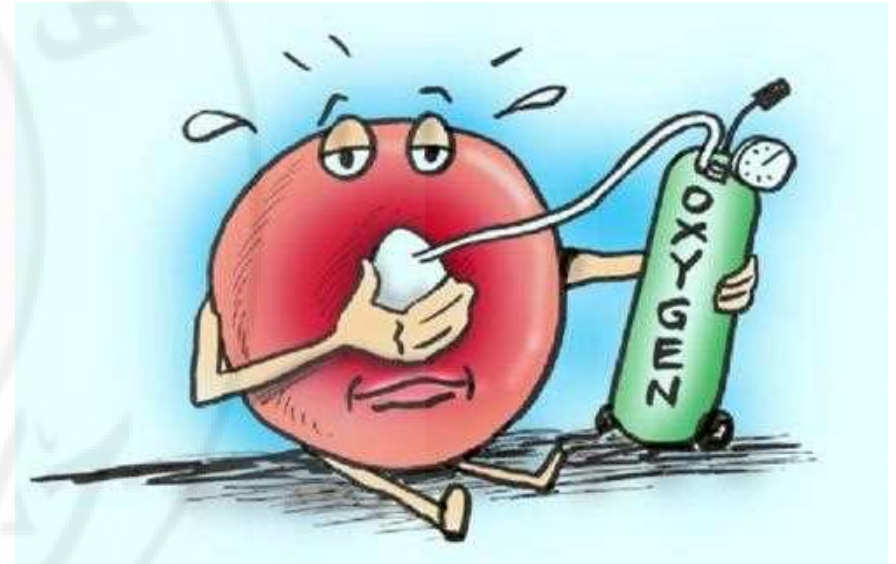
Hearing loss and respiratory distress

- **Prenatal hypoxia** (oxygen deficiency)
- **Ischemia** (blood supply deficiency)
- **Asphyxia** (caused by an injury or obstruction of the airway passages)
- The risk of permanent sensorineural hearing loss is greater for ischemia than for hypoxia; the central nervous system (CNS) is more susceptible to these kinds of insults than is the inner ear; and preterm infants are more susceptible than full-term babies.
- Interestingly, hearing losses are rarely caused by birth asphyxia alone, and hypoxia all by itself appeared to be associated with temporary hearing losses as opposed to permanent ones.



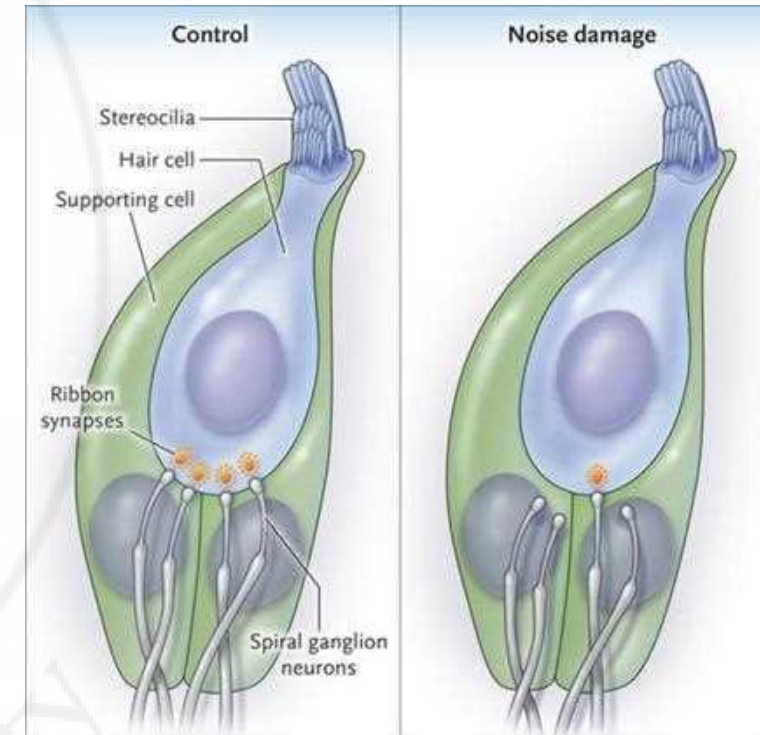
Hypoxia

- Hypoxia is a deficiency in the amount of oxygen in the body.
- Hearing disorder is often associated with hypoxia, although the effects of respiratory distress in neonates are difficult to separate from other possible factors, such as kernicterus, treatment with ototoxic medications, and low birth weight.
- Disorders of auditory neural function are often diffuse, although progressive sensorineural hearing loss can occur as a result.



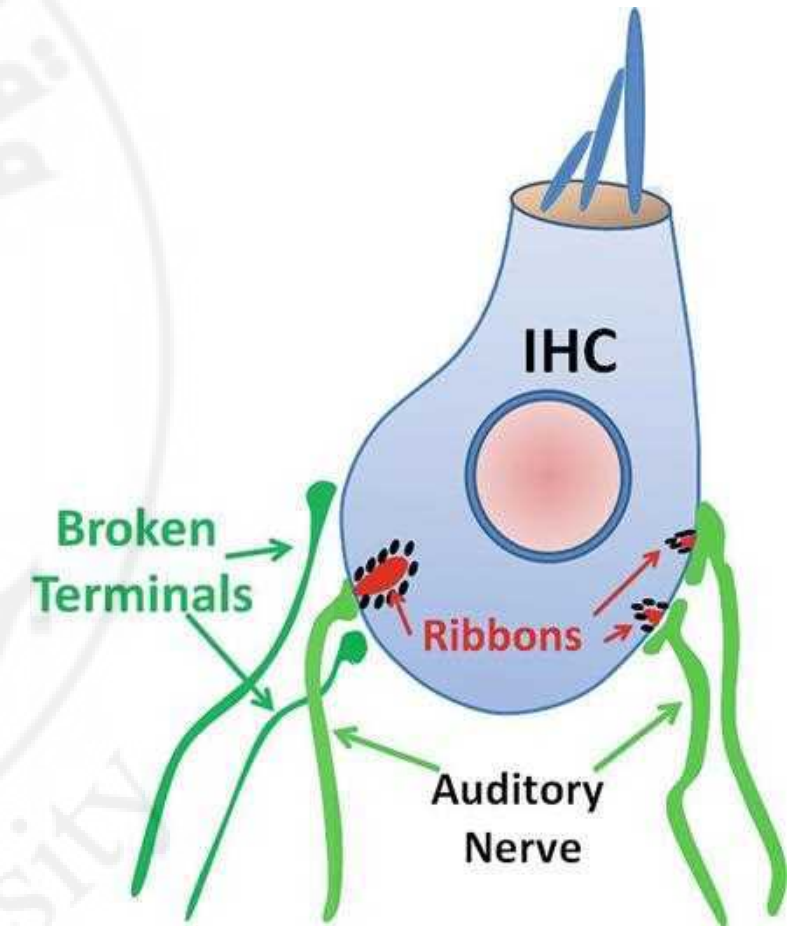
Cochlear Synaptopathy

- **Cochlear synaptopathy** or **auditory synaptopathy** refers to disorders of the structures and/or functions of the synapses between the inner hair cells and their associated auditory nerve fibers.
- It is also known as auditory synaptopathy or, more briefly, **synaptopathy**.
- These problems can be **presynaptic** and/or **postsynaptic** in nature.
- Presynaptic problems involve the structures and processes involved in the release of the neurotransmitter, glutamate, at the base of the inner hair cells, and postsynaptic problems are those involving glutamate receptors of the of the auditory neurons.
- Synaptopathy has been confirmed and described in numerous animal and human studies, and is among the factors involved in auditory neuropathy spectrum disorder, hidden hearing loss, noise-induced hearing loss, and presbycusis.



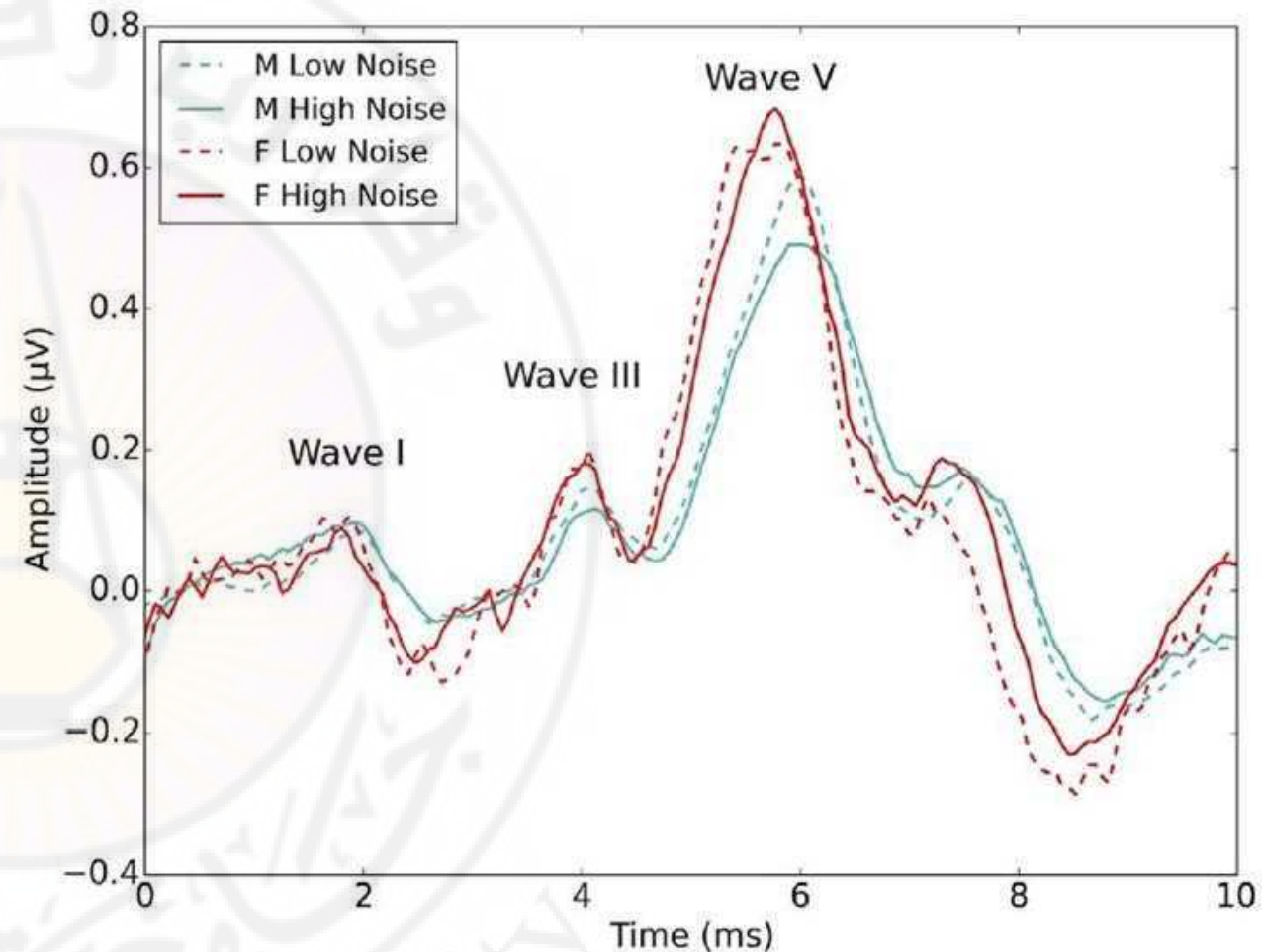
Cochlear Synaptopathy

- The characteristics of synaptopathy include histological, physiological, and perceptual/behavioral features and relationships.
- The histological features relate to synaptic loss or damage.
- Its physiological and perceptual/ behavioral characteristics have included reduced amplitudes for **wave I** of the auditory brainstem response, reduction of the **SP/AP** amplitude ratio, and normal **otoacoustic emissions**, normal **hearing thresholds** at the standard audiometric frequencies, and impaired **speech recognition** performance under difficult listening conditions, such as noise and reverberation.
- However, clear-cut criteria for diagnosing cochlear synaptopathy have not yet been developed.

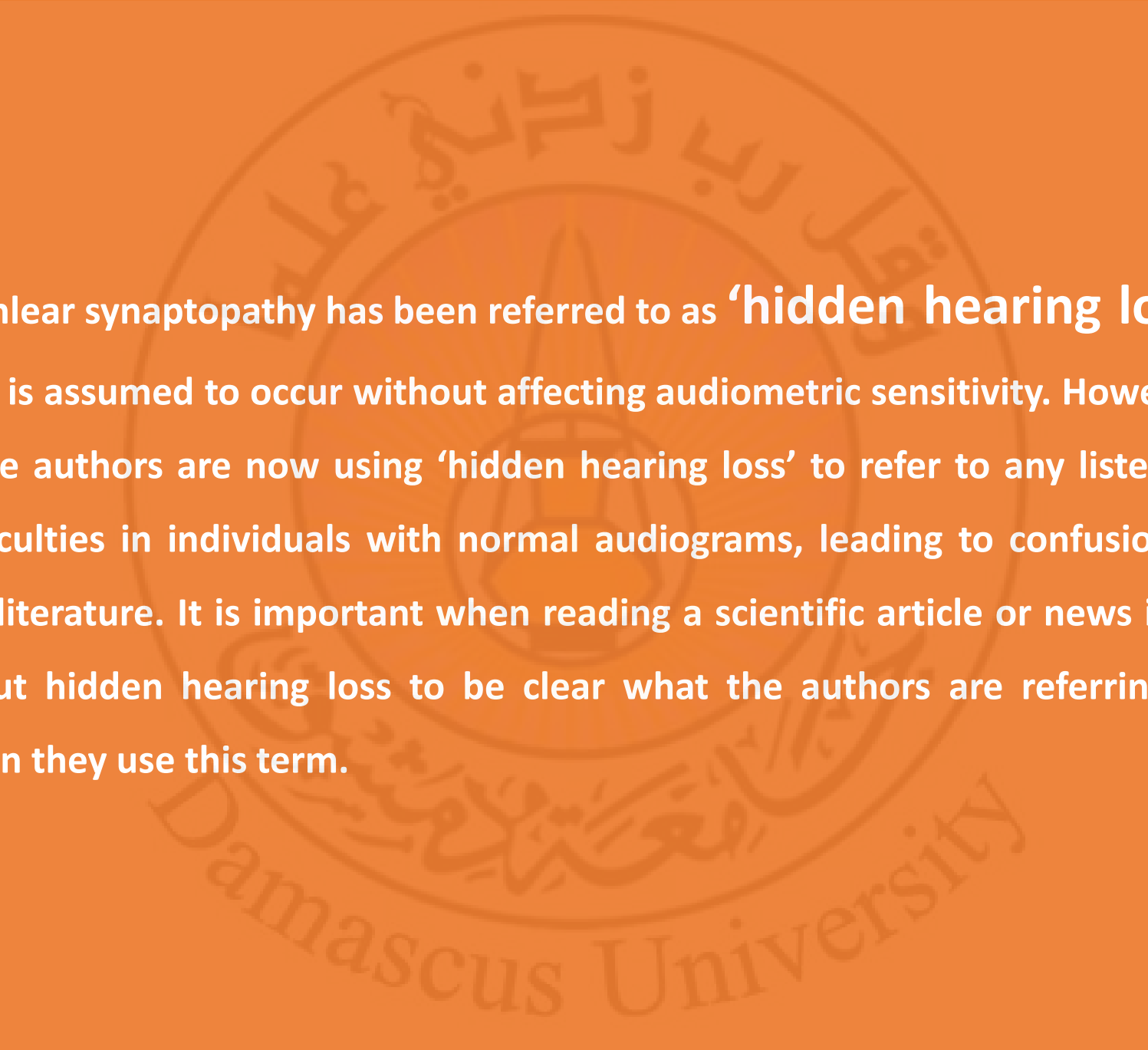


The assumption is that loss of synapses will be reflected by a reduction in wave I, as is the case in the rodent models.

In a large study with 126 young adult participants, with normal audiograms but a wide range of self-reported lifetime noise exposures, authors found no evidence for a reduction in wave I amplitude with increasing noise exposure.



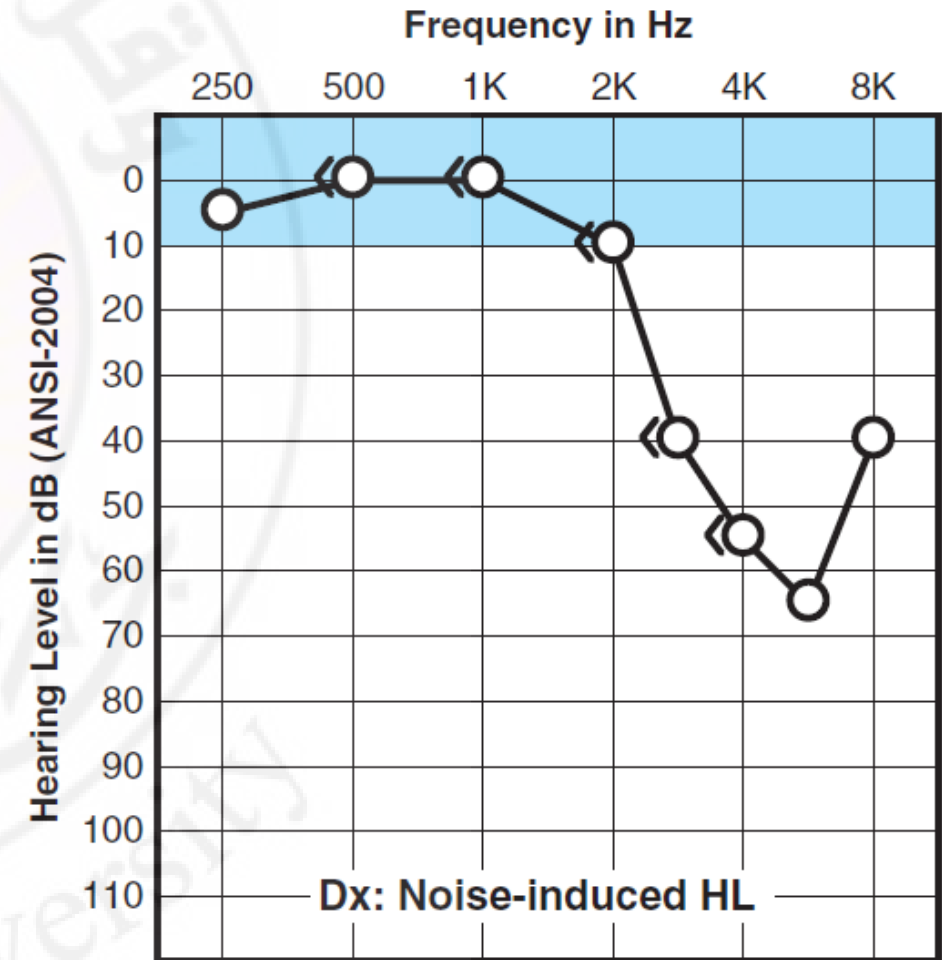
Averaged auditory brainstem responses for young male (M) and female (F) adults with normal hearing, divided into groups with extreme low and high noise exposures.

The background of the slide features a large, faint watermark of the Damascus University logo. The logo is circular, with the university's name in Arabic 'جامعة دمشق' at the top and 'Damascus University' at the bottom. In the center is a stylized emblem. The text of the slide is overlaid on this background.

Cochlear synaptopathy has been referred to as **‘hidden hearing loss’** as it is assumed to occur without affecting audiometric sensitivity. However, some authors are now using ‘hidden hearing loss’ to refer to any listening difficulties in individuals with normal audiograms, leading to confusion in the literature. It is important when reading a scientific article or news item about hidden hearing loss to be clear what the authors are referring to when they use this term.

Noise-Induced Hearing Loss

- High sound levels can produce both temporary and permanent hearing losses due to overstimulation and/or mechanical trauma.
- A sensorineural hearing loss produced by the damaging effects of overstimulation by high sound levels, usually over a long period of time, is called a **noise induced hearing loss (NIHL)**.
- Noise-induced hearing loss (NIHL) is the most common cause of acquired sensorineural hearing loss other than presbycusis.

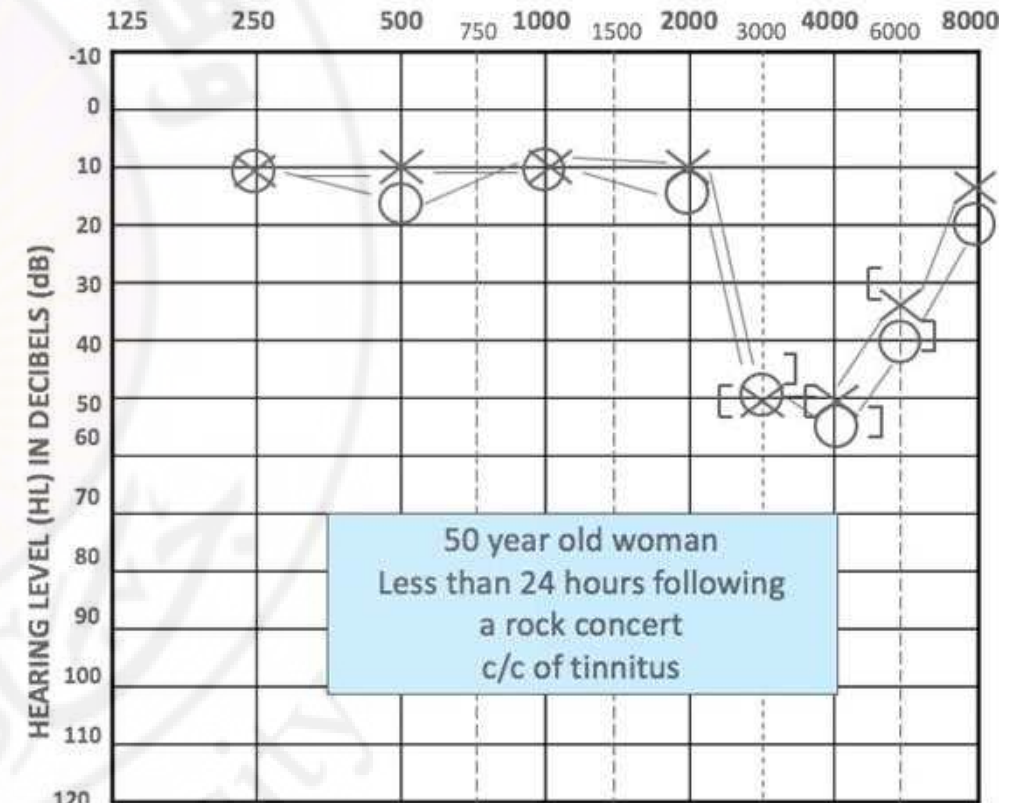


Noise-Induced Hearing Loss

- NIHL can be **temporary** or **permanent**. Exposure to excessive sound results in a change in the threshold of hearing sensitivity or a **threshold shift**.
- If a noise-induced hearing loss is **temporary**, it is referred to as a temporary threshold shift or **TTS**.
- If the hearing loss is **permanent**, it is called a permanent threshold shift or **PTS**.
- TTS is a common occurrence following exposure to loud music at a concert or following exposure to nearby firing of a gun or explosion of fireworks. The experience is usually one of sound seeming to be muffled, often accompanied by **tinnitus**. In TTS, hearing loss will be temporary, and hearing sensitivity will return to normal over time.
- However, if the signal intensity and the duration of exposure were of a sufficient magnitude, the hearing loss will be permanent. PTS occurs as a result of outer hair cell loss in the cochlea due to metabolic changes from repeated exhaustion of the cells.
- Repeated exposure will result in a **progression** of the hearing loss.

NIHL (Chronic Exposure) vs. Acoustic Trauma

- In contrast to NIHL, the term **acoustic trauma** usually refers to the hearing loss produced by extremely intense and impulsive sounds like explosions or gunshots.
- They can mechanically traumatize the eardrum, middle ear, and/or cochlear structures in addition to producing damage by overstimulation, and often from a single insult.
- Acoustic trauma result in necrosis of the outer hair cells (OHC), inner hair cells (IHC), and cause glutamate excitotoxicity of first-order afferent neurons of the spiral ganglion (cochlear synaptopathy).



NIHL (Chronic Exposure) vs. Acoustic Trauma

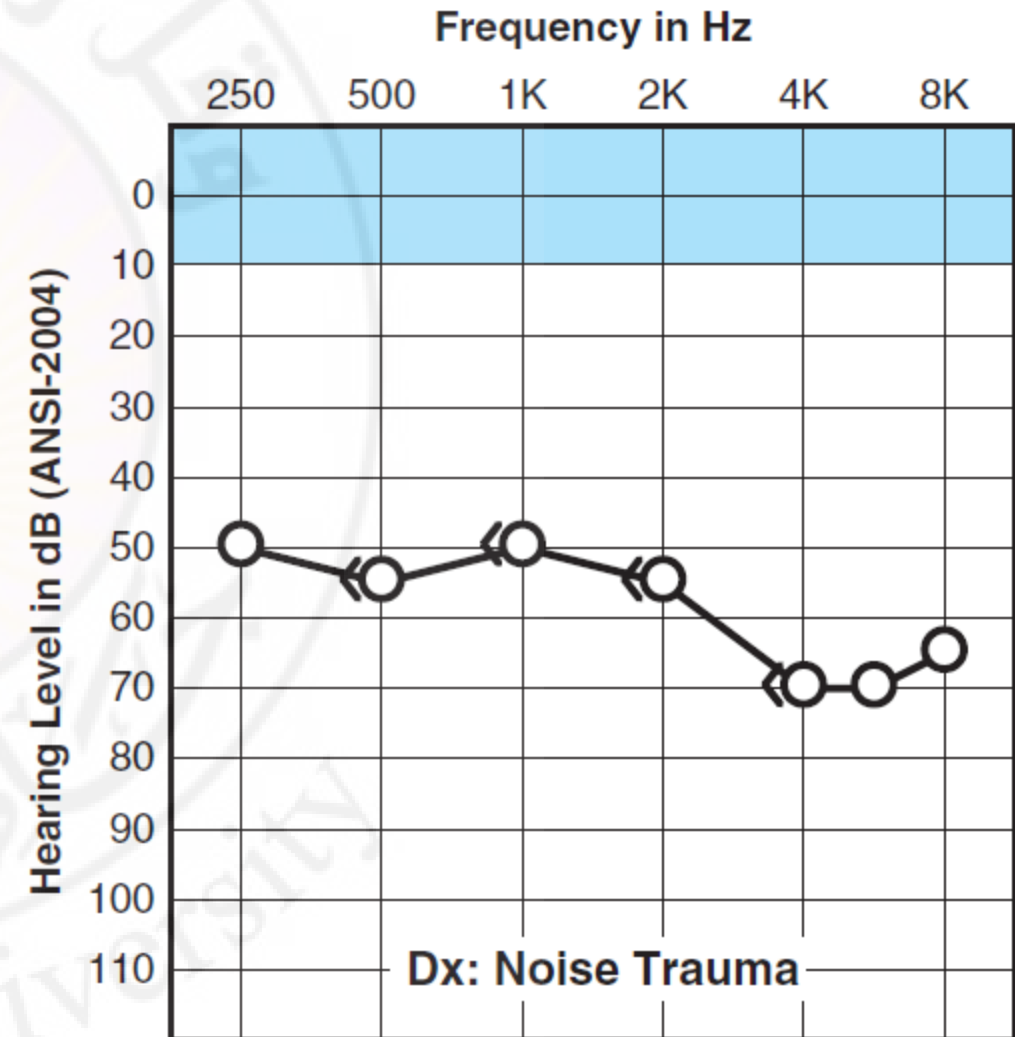
- **Acoustic Trauma** can occur when an impact or impulse sound, like an explosion, occurs abruptly.
- The force of that transient sound exceeds the elastic limit of the tissues. The organ of Corti can be sheared off the basilar membrane when the sound coming through the ear canal, middle ear and cochlea **exceeds 132 dB peak SPL**.
- If the sound is more intense than **184 dB**, the eardrum is ruptured. We see this 184 dB and above with military sound exposures, such as with the explosion of an IED (improvised explosive device).
- When you have a shock wave, you don't just **rupture the eardrum**; you also have **ossicular discontinuities**. You can even end up with traumatic **brain injury**. As a result, you could have an **auditory processing disability**. Lung injuries can develop as well as some injuries to the viscera.



Acoustic Trauma

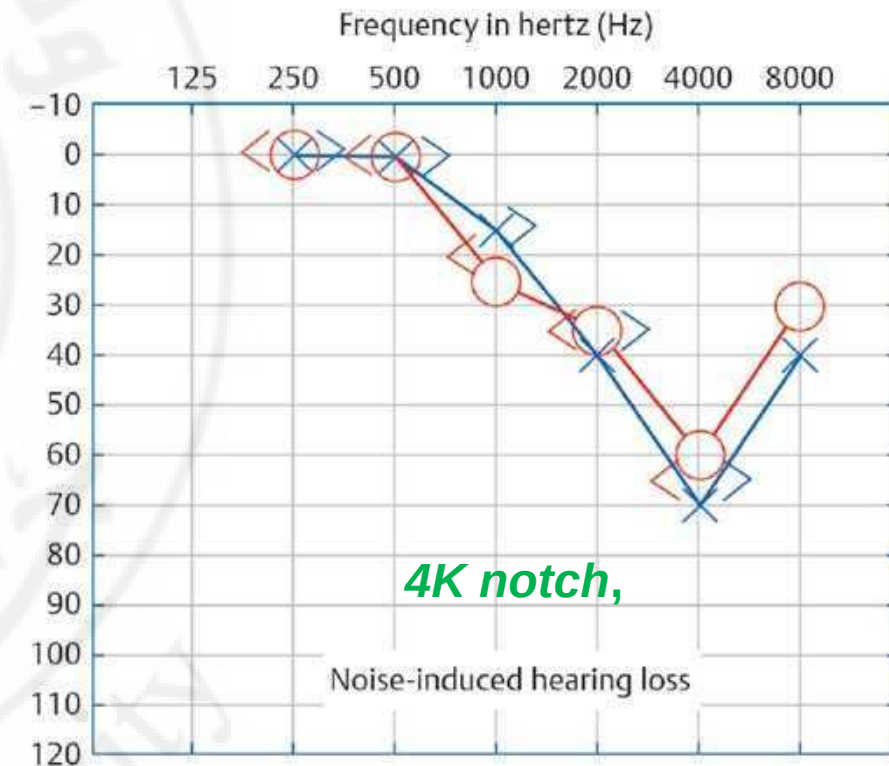
- Hearing loss that occurs from acoustic trauma as a result of a single exposure to excessive sound may resemble the noise-induced hearing loss from prolonged exposure, or it may result in a **flatter** audiometric configuration.

An audiogram representing the effect of a single exposure to an early cordless telephone that rang in close proximity to the user's ear. The level of the sound was estimated to be over **140 dB SPL**, resulting in a relatively flat, moderate sensorineural hearing loss.



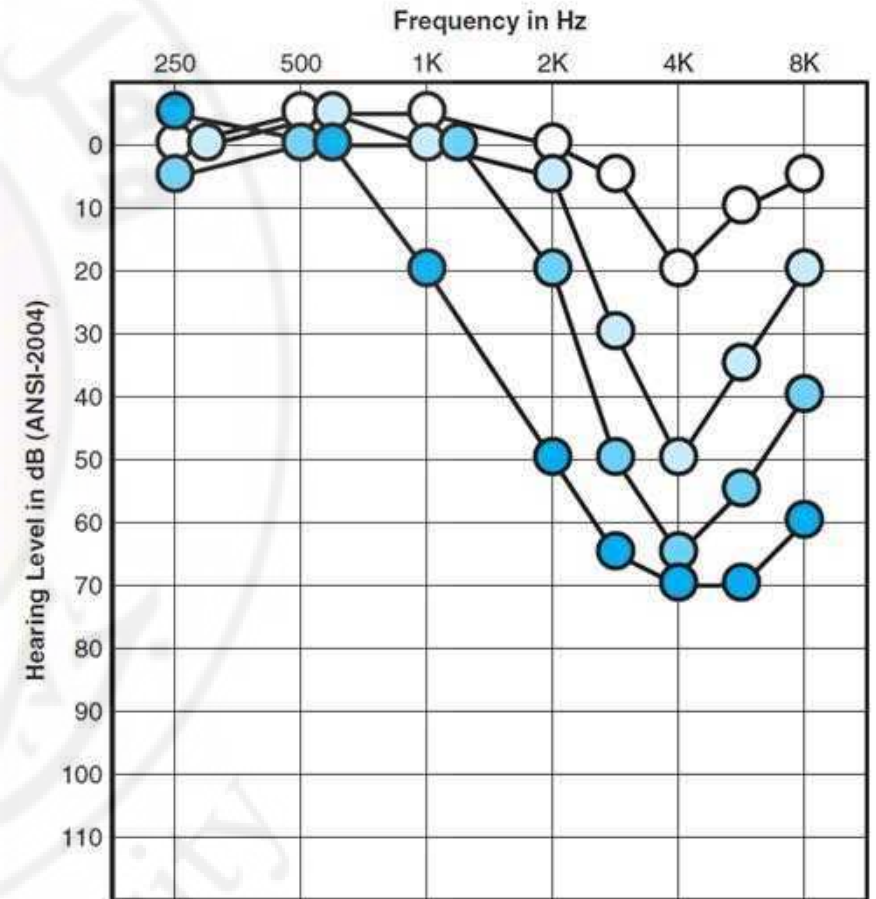
Noise-induced Hearing Loss

- The clinical picture of noise-induced hearing loss is usually associated with a **notch-shaped high frequency sensorineural loss that is worst at 4000 Hz**, although the notch often occurs at 3000 or 6000 Hz as well.
- The reason for the notch in this region is not definitively established. One explanation is that this region is most susceptible to damage due to the biology and mechanics of the cochlea.
- Another rationale is that commonly encountered noises have broad spectra that reach the cochlea with a boost in the 2000 to 4000 Hz region because of the **resonance characteristics** of the outer and middle ear.
- Noise-induced losses tend to be **bilateral** and more or less **symmetrical**; however, there are many exceptions, especially when one ear has been subjected to more noise than the other.



NIHL progression

- Not all “noise-induced” audiograms conform to the idealized **4K notch** picture of NIHL.
- Analyses of the progression of noise-induced hearing losses across many studies have revealed that the general audiometric pattern of noise-induced hearing loss evolves as noise exposure continues over the course of many years.
- The hearing loss typically begins as a notch at 4000 Hz. As noise exposure continues, the notch widens to include a wider range of frequencies, but continues to progress most noticeably at 4000Hz. After perhaps 10 to 15 years of exposure, the progression of the loss at 4000Hz often slows down, and progression now becomes more apparent at other frequencies, such as 2000Hz.



The progression of hearing loss from industrial noise exposure over a period of four decades.

NIHL risk assessment

- There are several important acoustic factors that make sound potentially damaging to the cochlea:
 - The **intensity** of the sound,
 - The **frequency** composition of the sound, and
 - The **duration** of exposure to the sound.
- In general, higher frequency sounds are more damaging than lower frequency sounds.
- Whether or not a particular intensity of sound is damaging to an ear depends on the duration of exposure to that sound. For example, a broad-spectrum noise with an intensity of 100 dBA is not necessarily dangerous if the duration of exposure is below 2 hours per day. However, exposure duration of greater than that can result in permanent damage to the ear (**noise-induced permanent threshold shift (NIPTS)**).



NIHL risk assessment

- Other factors also influence the risk of permanent noise-induced hearing loss.
- For example, some individuals are more susceptible than others, so that safe damage risk criteria for the population in general will not be safe levels for some individuals who are unusually susceptible.
- Also, the damaging effects of a given noise can be exacerbated by simultaneous exposure to certain **ototoxic drugs** and industrial **chemicals**.
- In addition, the amount of hearing loss produced by noise exposure is exacerbated if **vibration** is also present



NIHL damage-risk criteria

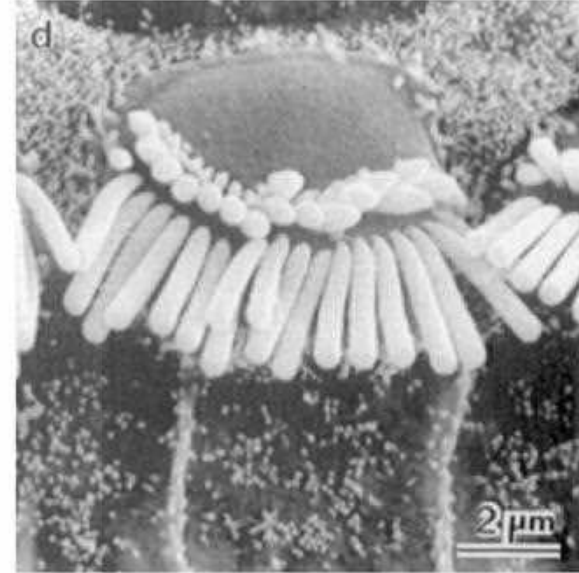
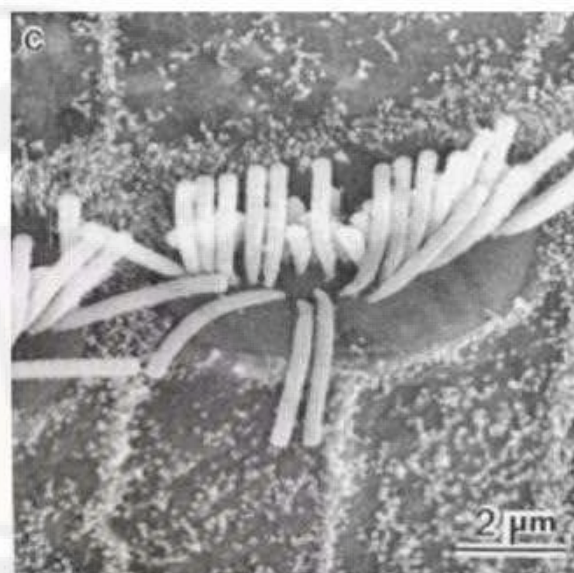
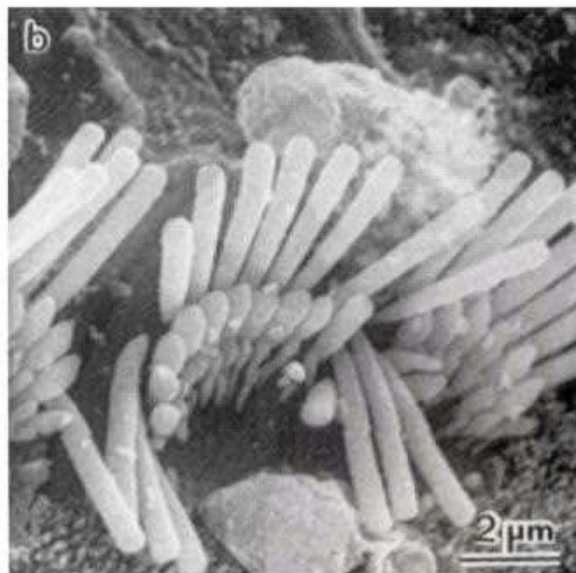
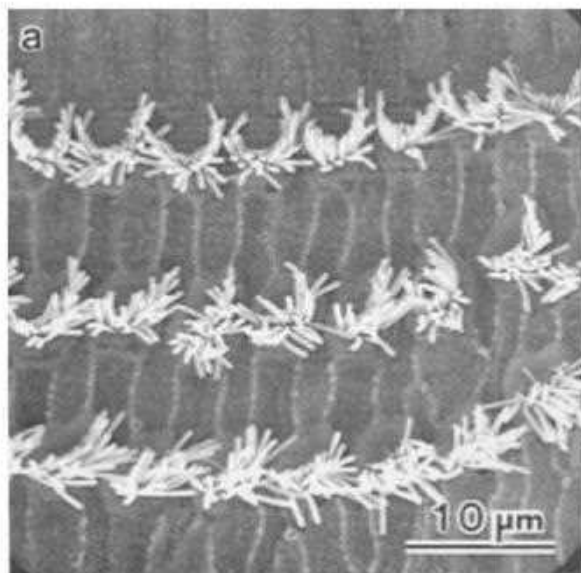
Damage-risk criteria expressed as the maximum permissible noise exposure for a given duration during a workday. Sound level is expressed in dBA, a weighted decibel scale that reduces lower frequencies from the overall decibel measurement.

Duration per Day (in hours)	Sound Level (in dBA)
8.0	90
6.0	92
4.0	95
3.0	97
2.0	100
1.5	102
1.0	105
0.5	110
0.25	115

Note: Criteria based on the U.S. Occupational Safety and Health Act 1983 regulations.

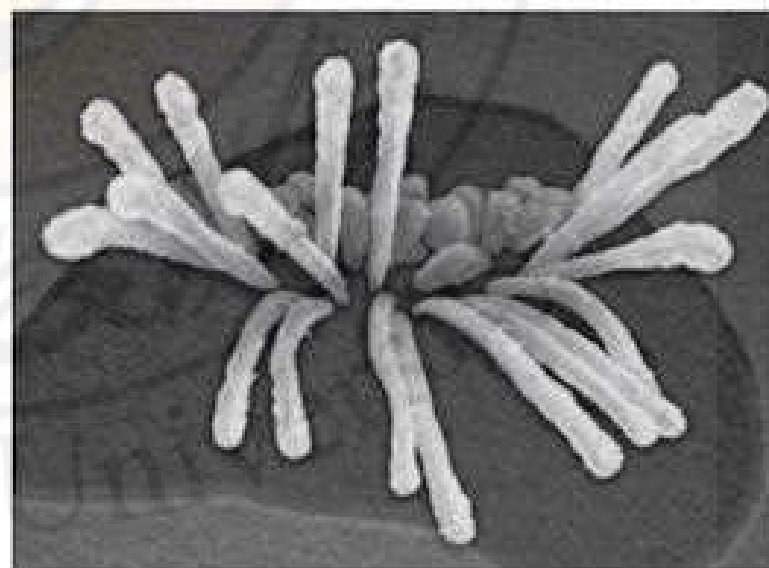
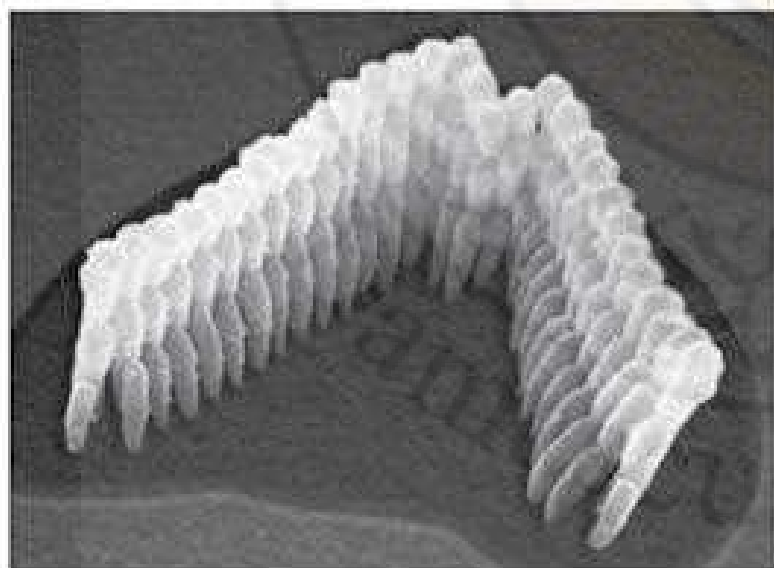
Anatomical and physiological changes caused by NIHL

- The kinds of anatomical and physiological abnormalities caused by noise exposure range from the most subtle disruptions of hair cell metabolic activities and losses of stereocilia rigidity (leading to “floppy cilia”) to the complete degeneration of the organ of Corti and the auditory nerve supply.
- Both outer and inner hair cells are damaged by noise, but outer hair cells are more susceptible.
- Some of the abnormalities include metabolic exhaustion of the hair cells, oxidative stress associated with accumulations of free radicals, structural changes and degeneration of structures within the hair cells, morphological changes of the cilia (so that they become fused and otherwise distorted), ruptures of cell membranes, and complete degeneration and loss of hair cells, neural cells, and supporting cells, as well as damage to auditory nerve fibers and the synapse between the hair cells and auditory neurons.
- In addition, oxidative stress associated with accumulations of free radicals has been identified as a factor in noise-induced hearing loss.-



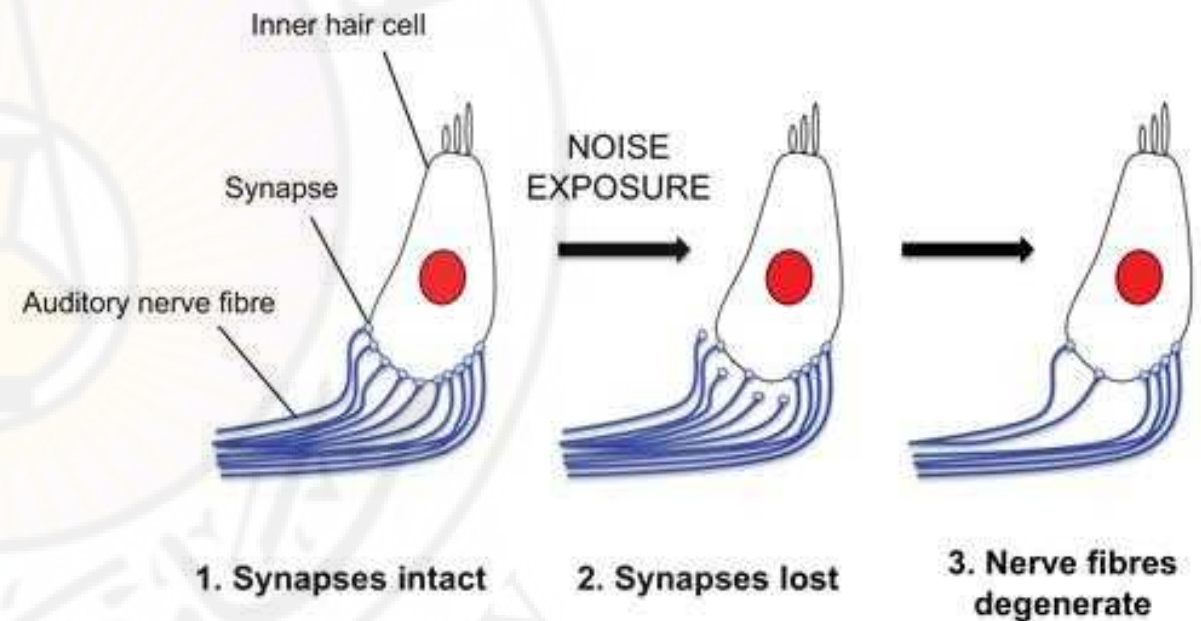
Healthy

Damaged



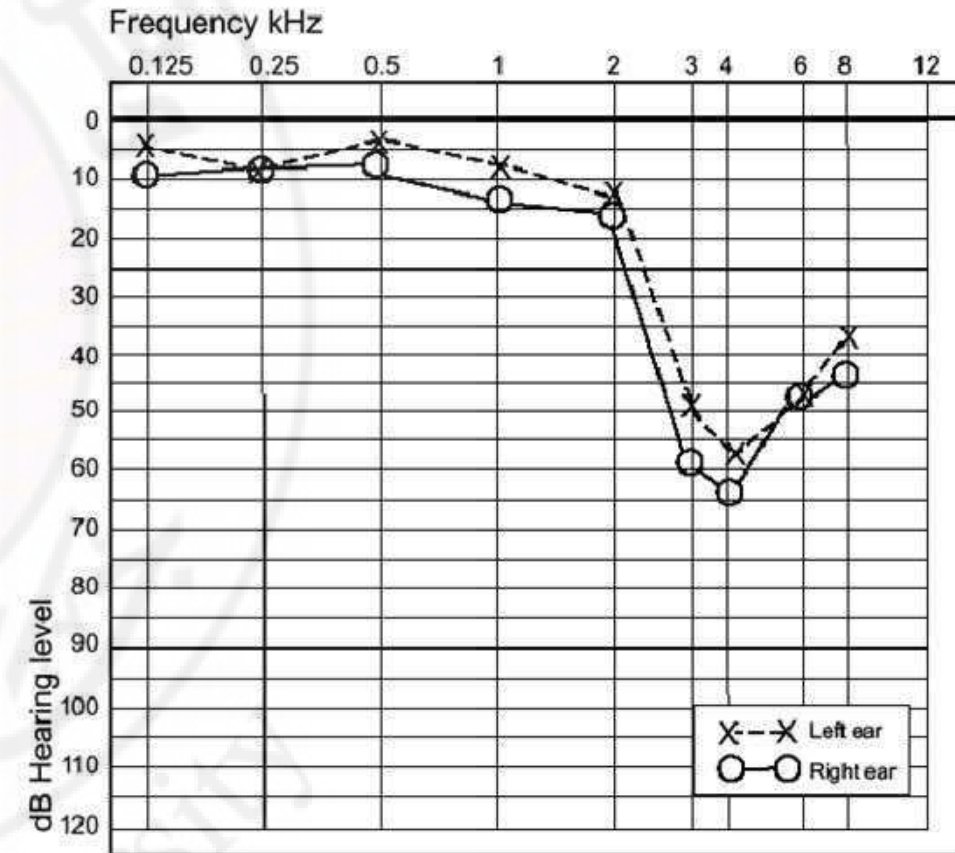
NIHL & synaptopathy

- Noise exposure has also been shown to cause permanent damage to the synapses between the outer hair cells and auditory nerve fibers known as synaptopathy even after the TTS has fully recovered.
- Some authors found that young adults who experience high sound levels in their daily lives had reduced speech recognition in noise and reverberation, elevated thresholds at very high frequencies, and evidence of synaptopathy even though their **clinical audiograms were normal**.



Clinical signs of NIHL

- Noise-induced temporary threshold shift (NITTS) and noise-induced permanent threshold shift (NIPTS) 3-6k Hz.
- Tinnitus (often presents as tone-like hissing)
- Hyperacusis (hypersensitivity to loud sound & Lower LDLs)
- Diplacusis (abnormal pitch perception)
- Suprathreshold speech intelligibility in noise decline



Hearing protection

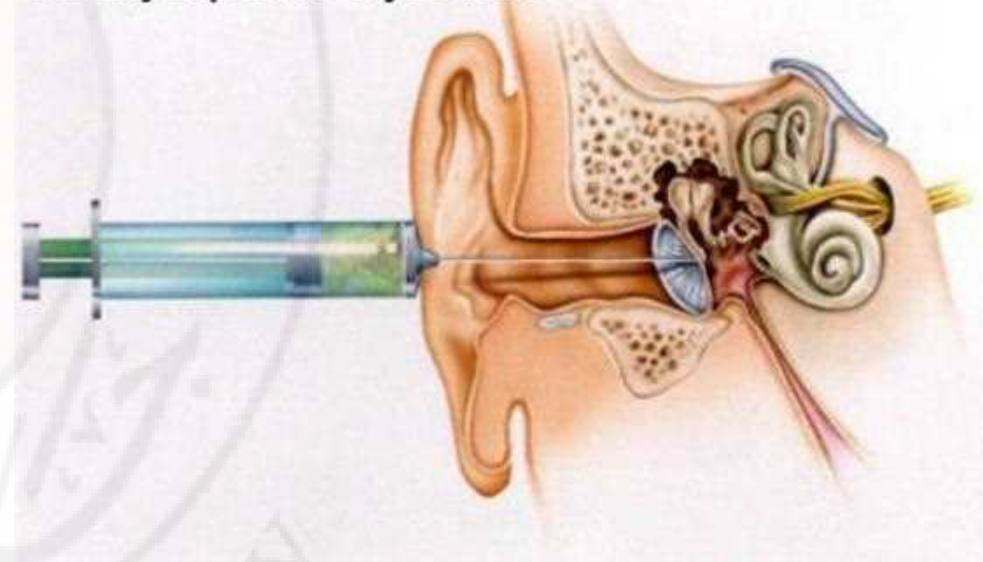
- Passive hearing protection
 - Foam earplugs (non-custom and custom)
- Active hearing protection



Chemicals/Medications for NIHL

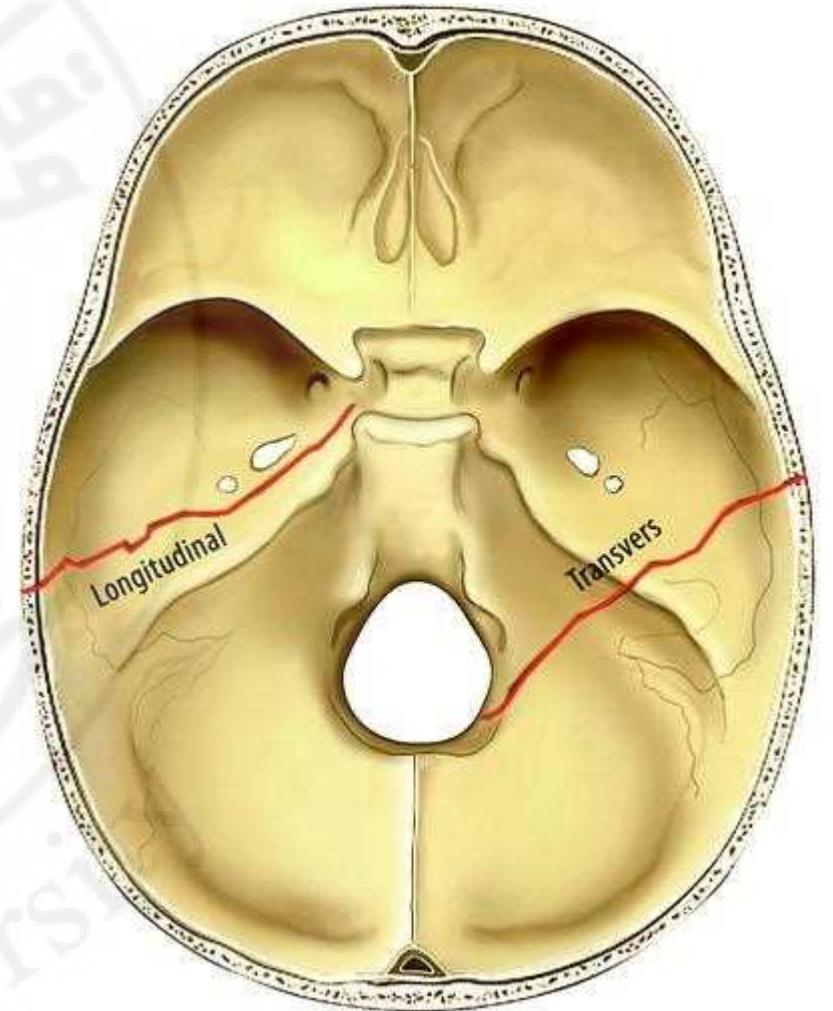
- There is the potential to have restoration in the early stages. Currently, what's being used for people who have suffered an acoustic trauma are **intratympanic steroid injections**.
- Physicians can inject **Prednisone** into the middle ear space within the first 24 to 48 hours. If the mechanism of damage is necrosis, the steroid, as it crosses over the round window into the perilymph you can get a reduction in how much inflammation occurs, which in turn limits how much damage occurs. This option is invasive. This reduces how much damage occurs in the first 48 hours due to the necrotic process.

Intratympanic Injection



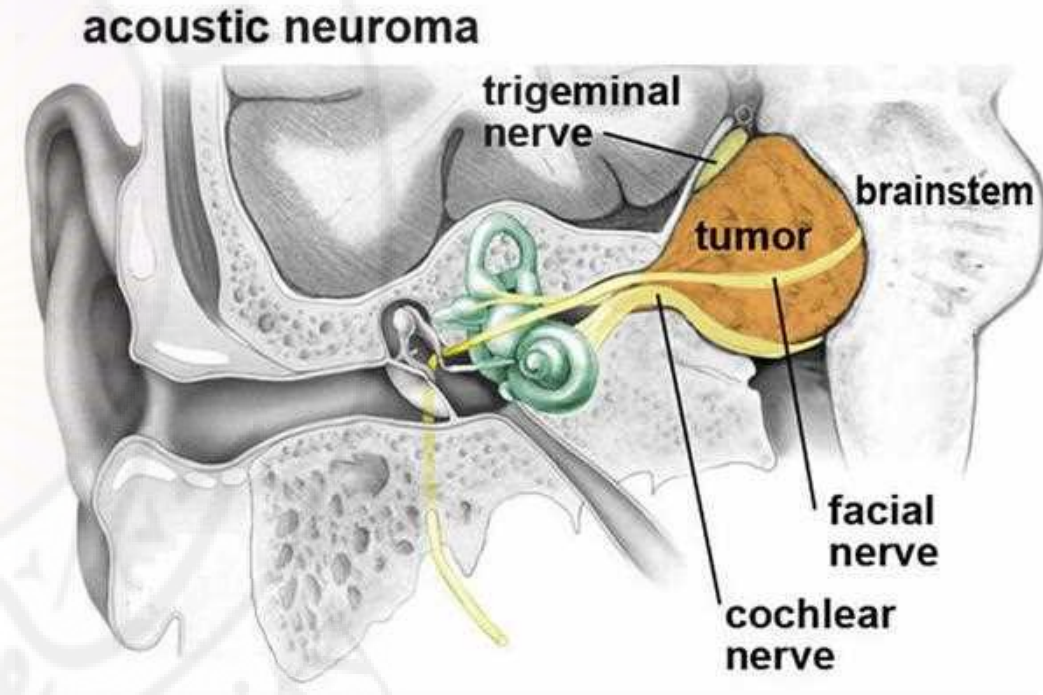
Trauma

- In addition to **acoustic trauma**, other insults to the auditory system can cause significant hearing loss.
- **Physical trauma** that results in a transverse fracture of the temporal bone can cause extensive destruction of the membranous labyrinth. This type of trauma is often caused by a blow to the occipital region of the skull.
- Recall that a **longitudinal fracture** can result in damage to the middle-ear structures, but seldom to the cochlea.
- Conversely, a **transverse fracture** tends to spare the middle ear and damage the cochlea. Depending on the extent of the labyrinthine destruction, the sensorineural hearing loss can be quite severe.



Trauma

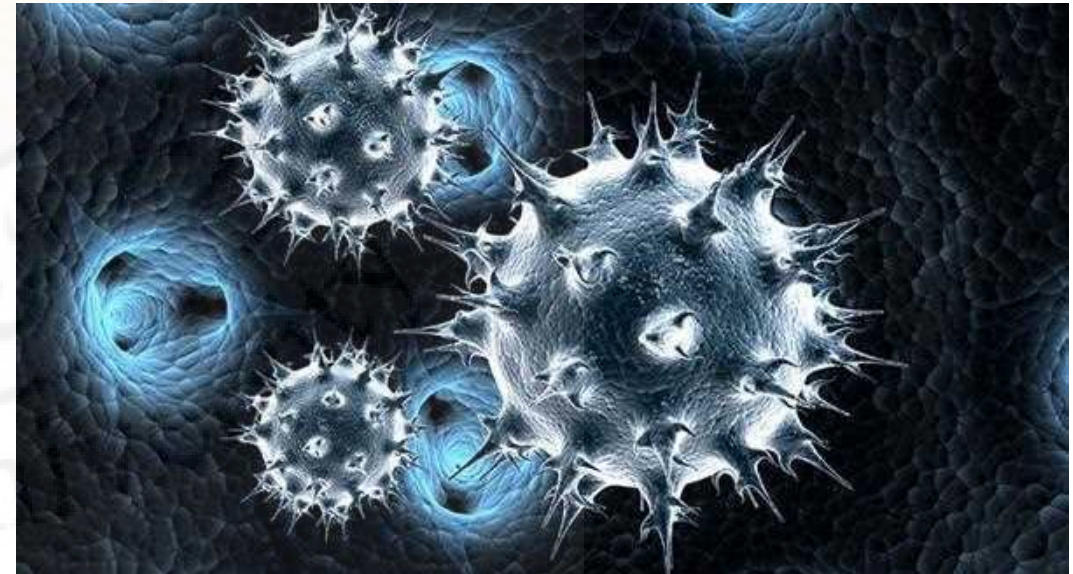
- Another form of trauma occurs as a result of **radionecrosis**, or the death of tissue due to excessive exposure to **radiation**.
- Injury to the auditory system secondary to X-ray irradiation occurs as a result of atrophy of the spiral and annular ligaments causing degeneration of the organ of Corti.
- Hearing loss of this nature is sensorineural, progressive, and usually has a delayed onset.
- X-ray irradiation injury is increasingly common as radio-surgery is used in an effort to irradiate **acoustic tumors**.



Infections

- Sensorineural hearing loss can result from **acquired infections**, the pathogens from which can directly insult the membranous labyrinth of children or adults. Infections from **bacteria, viruses, and fungi** can cause sensorineural hearing loss.

- Labyrinthitis
- Meningitis
- Herpes zoster oticus
- Mumps
- Measles
- Syphilis

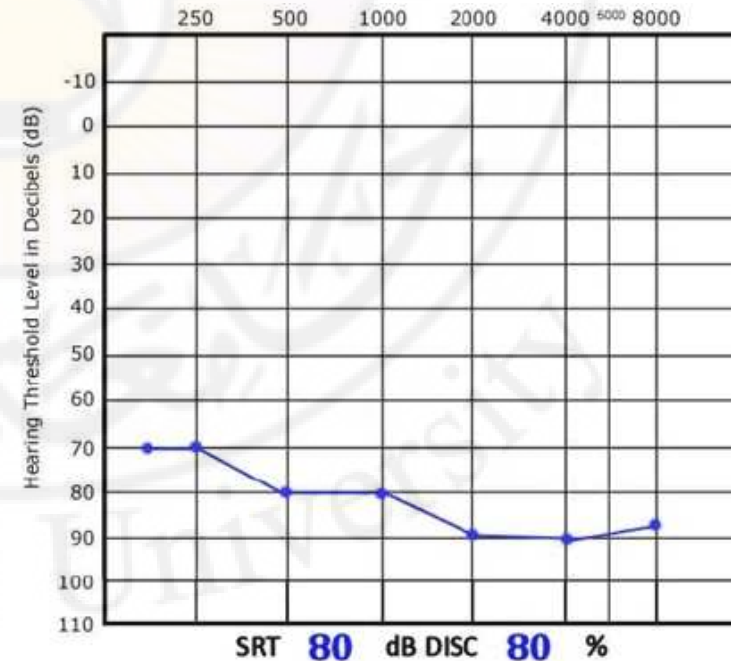
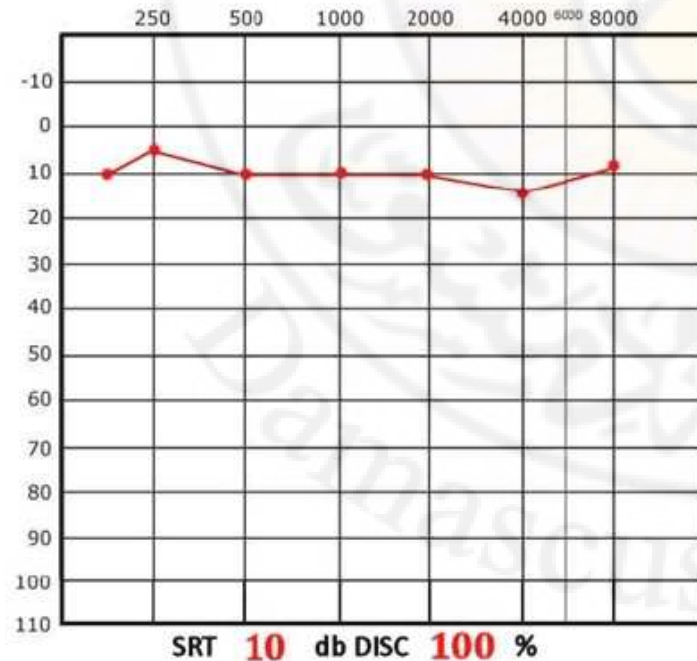


Labyrinthitis

- Bacterial infections can cause inflammation of the membranous labyrinth of the cochlea, or **labyrinthitis**, through several routes.
- **Serous or toxic labyrinthitis** is an inflammation of the labyrinth caused by bacterial contamination of the tissue and fluids, either by invasion through the middle ear (**otogenic**) or via the meninges (**meningogenic**).
- Serous labyrinthitis may be transient, resulting in mild hearing loss and dizziness. In more severe forms, however, it can be toxic to the sensory cells of the cochlea, causing substantial sensorineural hearing loss.
- Serous labyrinthitis sometimes occurs secondary to serous otitis media, presumably from the bacterial toxins traveling through the membranes of the oval or round windows.
- It may also occur secondary to meningitis, with the inflammation traveling along the brain's linings to the membranous labyrinth of the cochlea.

Labyrinthitis

- Bacterial infections can also cause otogenic **suppurative labyrinthitis**. In this type of infection, bacteria invade the cochlea from the temporal bone.
- The pus formation in the infection can cause permanent and severe damage to the cochlear labyrinth, resulting in substantial sensorineural hearing loss.



Meningitis

- **Meningitis** is an inflammation of the membranes that surround the brain and spinal cord caused by bacteria or viruses (and also aseptic).
- The greatest frequency of hearing loss occurs in cases of bacterial meningitis, with estimates ranging from 5 to 35% of cases.
- Other common signs and symptoms of bacterial meningitis include fever, seizures, neck stiffness, and altered mental status. Vestibular function may be compromised as well.
- Hearing disorder ranges from mild to profound sensitivity loss or total deafness and may be progressive. Most involvement is thought to be cochlear, but central auditory involvement may occur in some individuals.
- **Cochlear osteoneogenesis**, or bony growth in the cochlea, may occur following meningitis.



MENINGITIS SYMPTOMS



Fever



Sleepiness



Headache



Light sensitivity



Vomiting



Joints pain



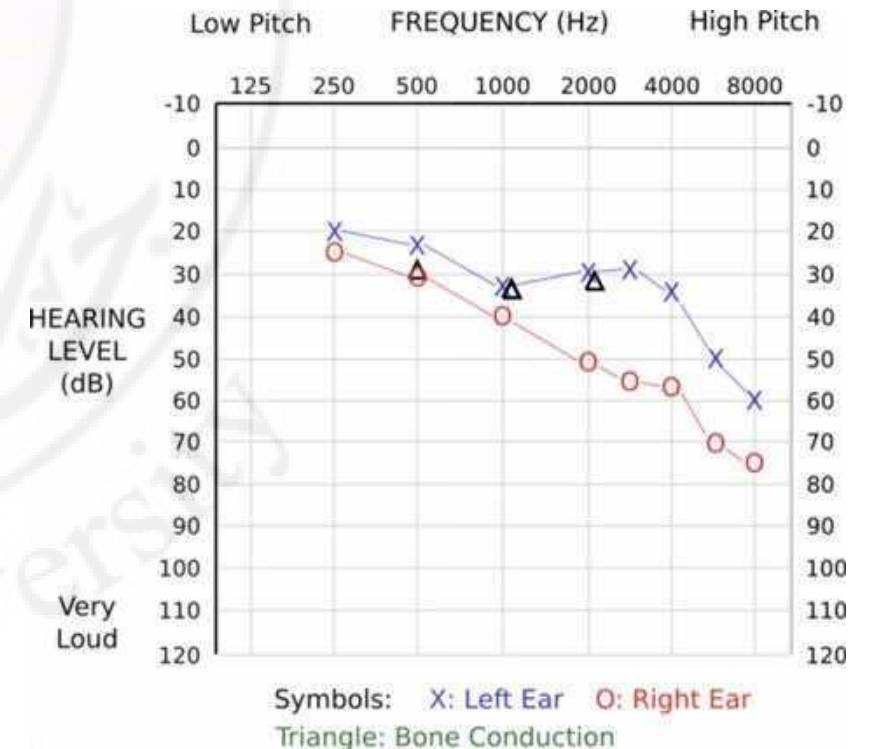
Rash



Seizures

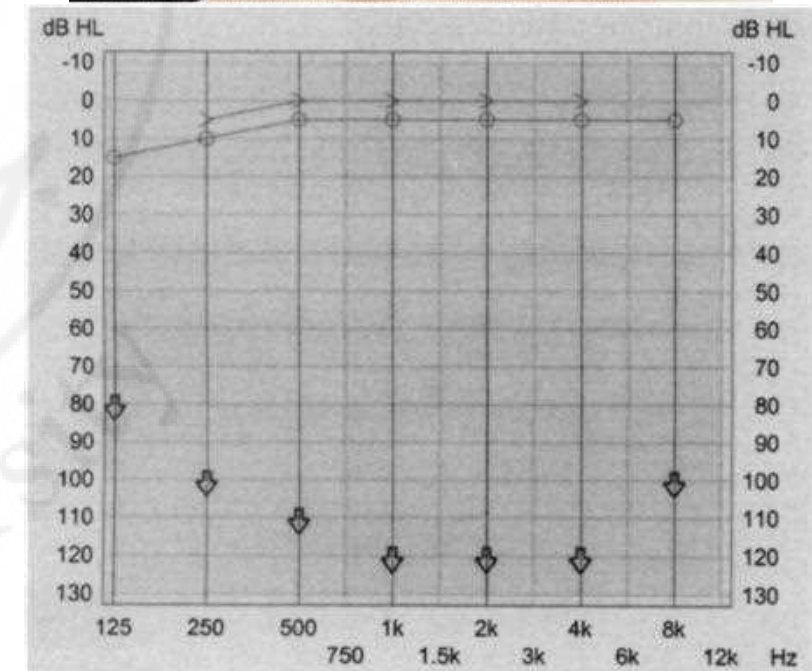
Herpes zoster oticus

- **Herpes zoster oticus, or Ramsay Hunt syndrome**, is caused by a virus that also causes chicken pox.
- The virus, often acquired during childhood, can lie dormant for years in the central nervous system. At some point in time, due to changes in the immune system or to the presence of systemic disease, the virus is reactivated, causing burning pain around the ear, skin eruptions in the external auditory meatus and concha, facial nerve paralysis, dizziness, and sensorineural hearing loss.
- Hearing loss is of varying degree and often has a **high-frequency audiometric configuration**.



Mumps

- **Mumps, or epidemic parotitis**, is an acute systemic viral disease, most often occurring in childhood after the age of 2 years.
- Depending on severity, it usually causes painful swelling of the parotid glands and can cause a number of complications related to encephalitis.
- Despite the systemic nature of the disease, **hearing loss** is peculiar in that it is almost always **unilateral** in nature. Because of this, it often goes undiagnosed until later in life.
- Mumps is probably the *single most common cause* of unilateral sensorineural hearing loss, and the loss is usually **profound**.
- Endolymphatic hydrops (Ménière disease) is also a common manifestation of mumps virus (**Secondary endolymphatic hydrops**).



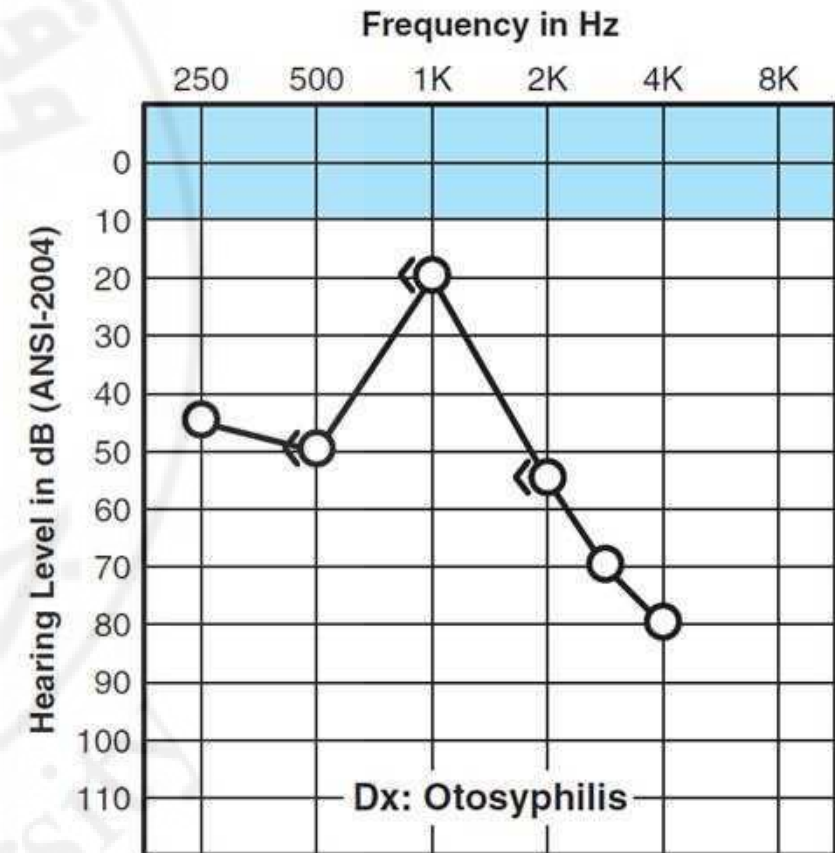
Measles

- Measles is a highly contagious viral illness whose symptoms characteristically include rash, cough, fever, conjunctivitis, photophobia, and Koplik spots (white spots on the membranous surfaces of the mouth).
- **Hearing loss** is a common complication of measles. Sensory hearing loss from measles virus is typically severe, permanent, and bilateral, although milder hearing loss can occur.
- Conductive hearing loss from otitis media is also a common complication of measles that may be due to immunosuppression following viral infection.
- Localized measles virus in the otic capsule is also hypothesized to be a causative factor in later development of otosclerosis.



Syphilis

- Another cause of hearing loss from **acquired** infection is **syphilis**, a venereal disease that can also cause **congenital** hearing loss.
- Syphilis is usually described in terms of clinical stages, from primary infection, through secondary involvement of other organs, to tertiary involvement of the cardiovascular and nervous systems.
- Hearing loss from **otosyphilis** occurs in the secondary or tertiary stages and results from membranous labyrinthitis associated with acute meningitis or osteitis of the temporal bone.
- Hearing loss from syphilis is not unlike that from **Menière's disease**, characterized by fluctuation attacks and progression in severity.





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Ototoxicity

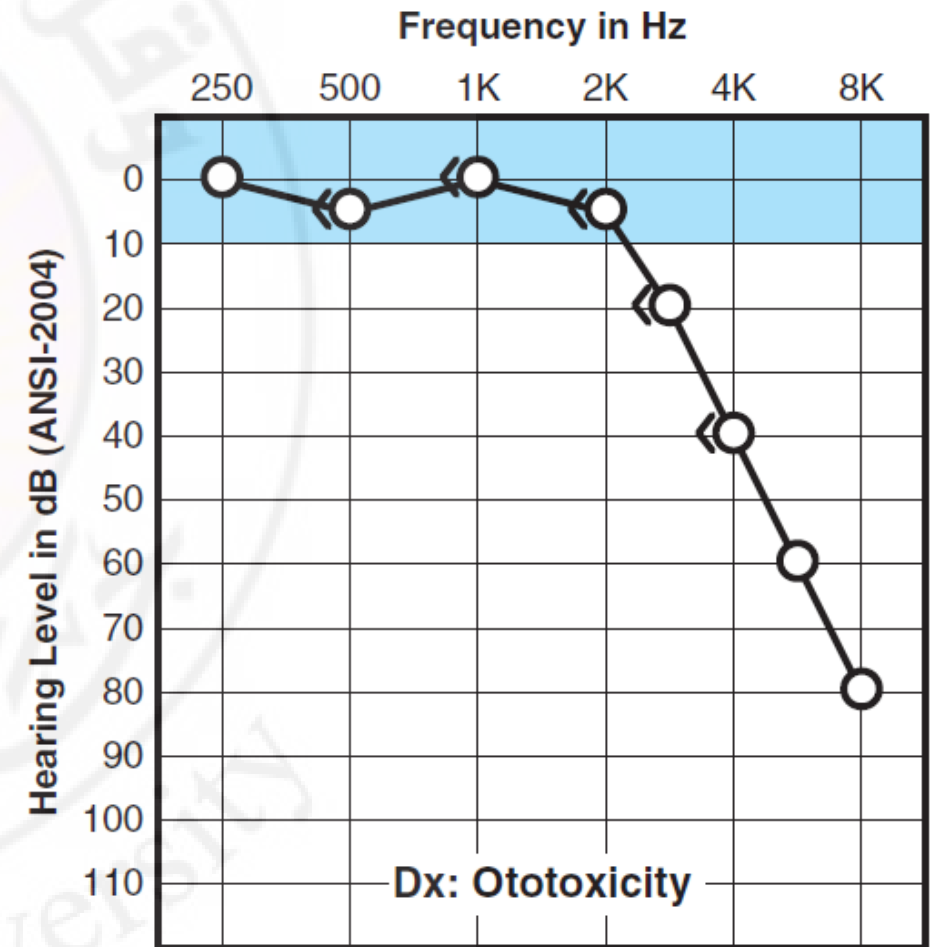
- **Ototoxicity** means damage to the ear due to the toxic effects of various chemical agents, especially drugs, and results in sensorineural hearing loss.
- Ototoxicity can be **acquired** or **congenital**.
- **Acquired ototoxicity** results from the ingestion of certain **drugs** that are administered for medical purposes, such as in the treatment of infections and cancer. Ototoxicity can also result from excessive exposure to certain **environmental toxins**.
- **Congenital ototoxicity**, as discussed earlier, results from the teratogenic effects of drugs administered to the mother during pregnancy.



More than
200
medications
are known to be
OTOTOXIC
OR POISONOUS
TO THE EARS

Ototoxicity: Mechanism for damage

- Hearing loss from ototoxicity is usually **permanent, sensorineural, bilateral, and symmetric**.
- Ototoxic drugs cause hearing losses by affecting the cochlear hair cells and the stria vascularis.
- The mechanism for damage varies depending on the drug, but, in general, hearing loss results initially from damage to the outer hair cells of the cochlea at its **basal end**.
- Thus, the hearing loss typically begins as a high-frequency loss and progresses to lower frequencies with additional drug exposure. **Tinnitus** usually accompanies or precedes the hearing loss, but not always.



Ototoxic agents

- Aminoglycosides
- Antineoplastic & antimitotic drugs
- Antimalarial drugs
- Salicylates
- Loop diuretics
- Industrial solvents & chemicals
- Anti-retroviral drugs

Ototoxic: harmful to hearing



Aminoglycosides

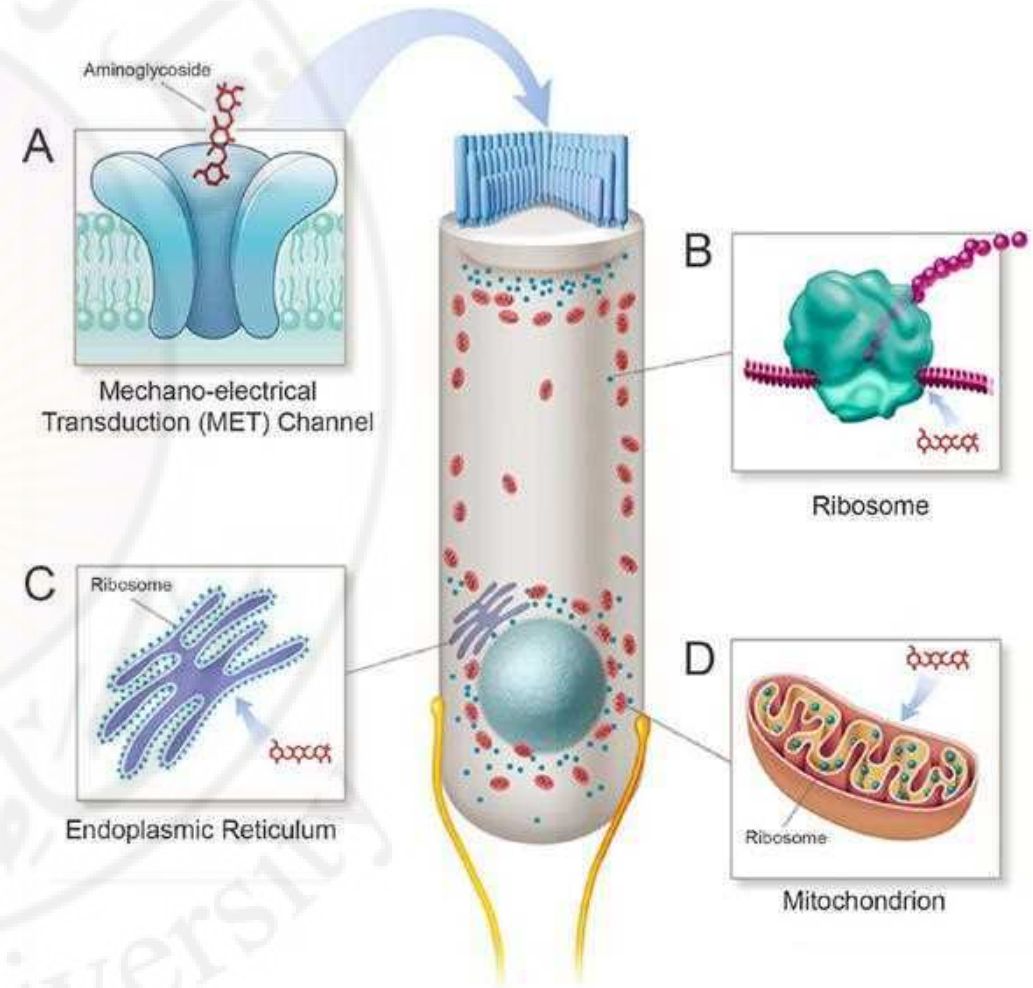
- The **aminoglycosides** are a group of antibiotics that are often ototoxic. They are used primarily against bacterial infections.
- Some of the aminoglycosides have a predilection for hair cells of the cochlea (**cochleotoxic**), while others have a predilection for hair cells of the vestibular end-organs (**vestibulotoxic**).
- Most of these antibiotics can be used in smaller doses to effectively fight infection without causing ototoxicity.
- Sometimes, however, the infections must be treated aggressively with high doses, resulting in significant sensorineural hearing loss.

• Ototoxic antibiotics include:

- Amikacin,
 - Dihydrostreptomycin,
 - Garamycin,
 - Gentamicin,
 - Kanamycin,
 - Neomycin,
 - Netilmicin,
 - Streptomycin,
 - Tobramycin
 - Viomycin
- Other kinds of antibiotics can also be ototoxic, such as capreomycin, vancomycin, and even erythromycin.
 - However, the suffix **mycin** does not necessarily mean a drug is ototoxic for example, clindamycin, Terramycin, and Vibramycin are not ototoxic.

Aminoglycosides

- It is noteworthy that patients are predisposed to aminoglycoside ototoxicity if they have abnormalities of certain mitochondrial genes.
- Patients who receive aminoglycosides are often predisposed to varying degrees of hearing loss if they have mutations of the **MR-RNR1 gene**, and usually have severe to profound losses if they have a mutation of the **MT-CO1 gene**.
- Hearing losses due to aminoglycoside ototoxicity can develop or continue for many months after treatment has ended, depending on the drug.



Antineoplastic & antimitotic drugs

- Other drugs that are ototoxic have been developed in the fight against cancer.
- **Carboplatin**, and **cisplatin (CIS platinum)** are antimitotic and antineoplastic drugs often used in **cancer treatment**.
- It is not unusual for patients who undergo **chemotherapy** regimens that contain either or both of these drugs to develop permanent sensorineural hearing loss.



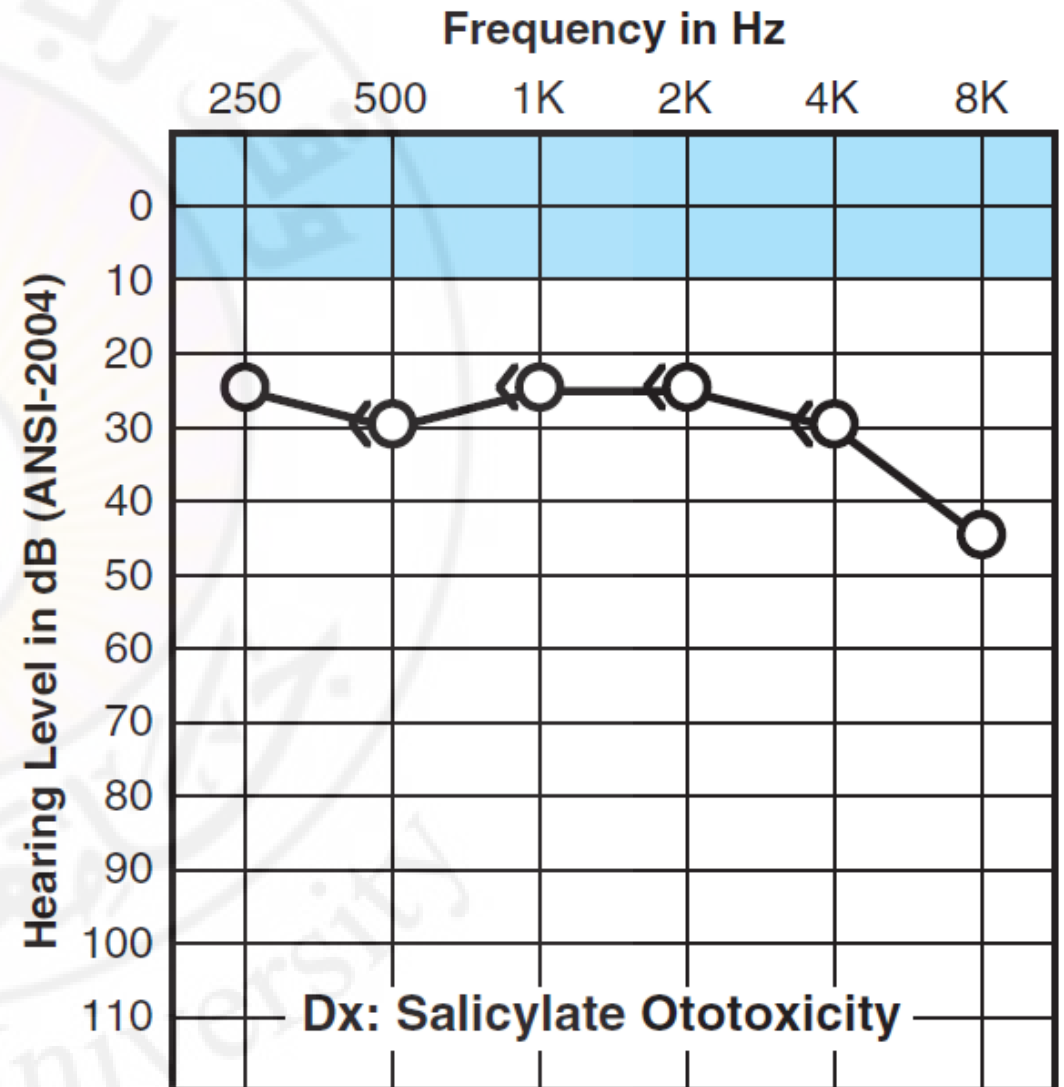
Antimalarial drugs

- Antimalarial drugs, including **chloroquine** and **quinine**, have been associated with ototoxicity.
- Typically, the hearing loss from these drugs is **temporary** (reversible).
- However, in high doses the loss can be permanent.
- **Quinine** is an antimalarial drug that can have a teratogenic effect on the auditory system of the developing embryo.



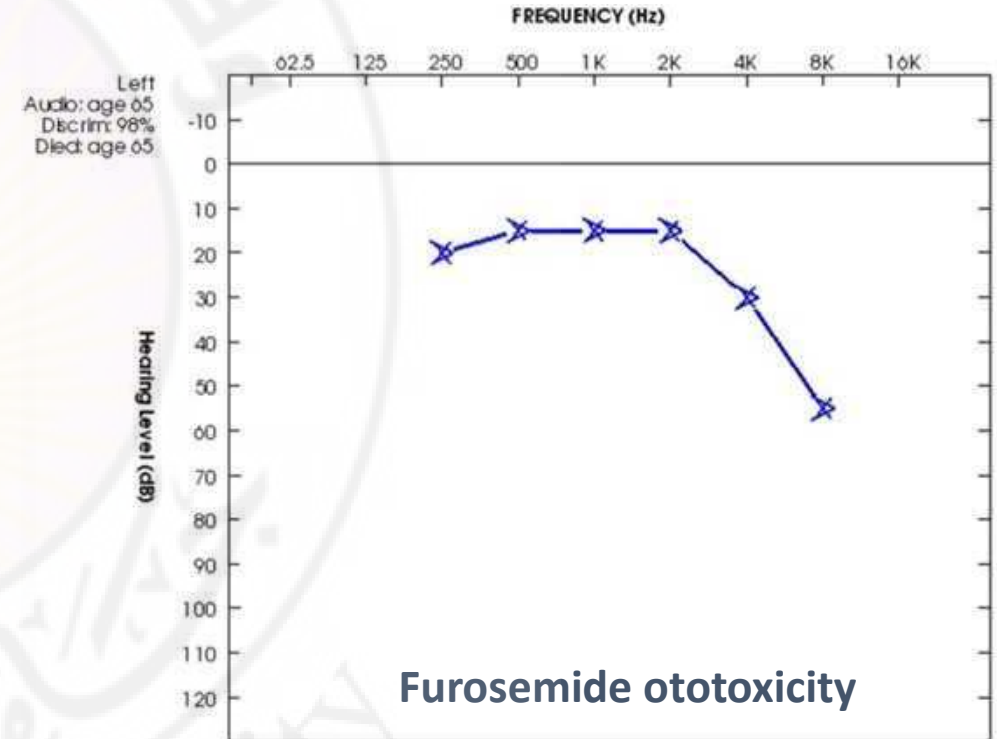
Salicylates

- Drugs known as **salicylates** can also be ototoxic. Salicylates such as **acetylsalicylic acid** and **aspirin** are used as therapeutic agents in the treatment of arthritis and other connective tissue disorders.
- Hearing loss is usually **reversible** and accompanied by **tinnitus**.
- In the case of salicylate intoxication, the hearing loss often has a **flat** rather than a steeply sloping configuration.



Loop diuretics

- Loop diuretics are commonly used in the treatment of congestive heart disease and other edematous disorders.
- Loop diuretics, including **ethacrynic acid (Edecrin)**, **furosemide (Lasix)**, and **bumetanide (Bumex)**, are used to promote the excretion of urine by inhibiting resorption of sodium and water in the kidneys.
- Hearing loss from loop diuretics may be reversible or permanent.



Industrial solvents & chemicals

- Other ototoxic substances include industrial solvents, such as **styrene**, **toluene**, **carbon monoxide**, and **trichlorethylene**, which can be ototoxic if inhaled in high concentrations over extended periods.
- **Potassium bromate**, a chemical neutralizer used in food preservatives and other commercial applications, has also been associated with ototoxicity.
- Other ototoxic **chemicals** include: heavy metals (such as lead and mercury, and probably germanium and tin compounds, as well), tetra chlorocarbon and sulfur compounds, organic phosphates.
- Toxicity from agents such as these also affects other body organs, and exposures to one or more of them are possible in several industries and in homes.



Anti-retroviral drugs

- It is not yet clear whether the anti-retroviral drugs used to treat **human immunodeficiency virus (HIV)** have ototoxic effects.
- Some studies have reported an association between hearing loss and some of these agents, such as HIV. However, this association was not found in a prospective study involving 32 weeks of treatment with antiretroviral drugs.



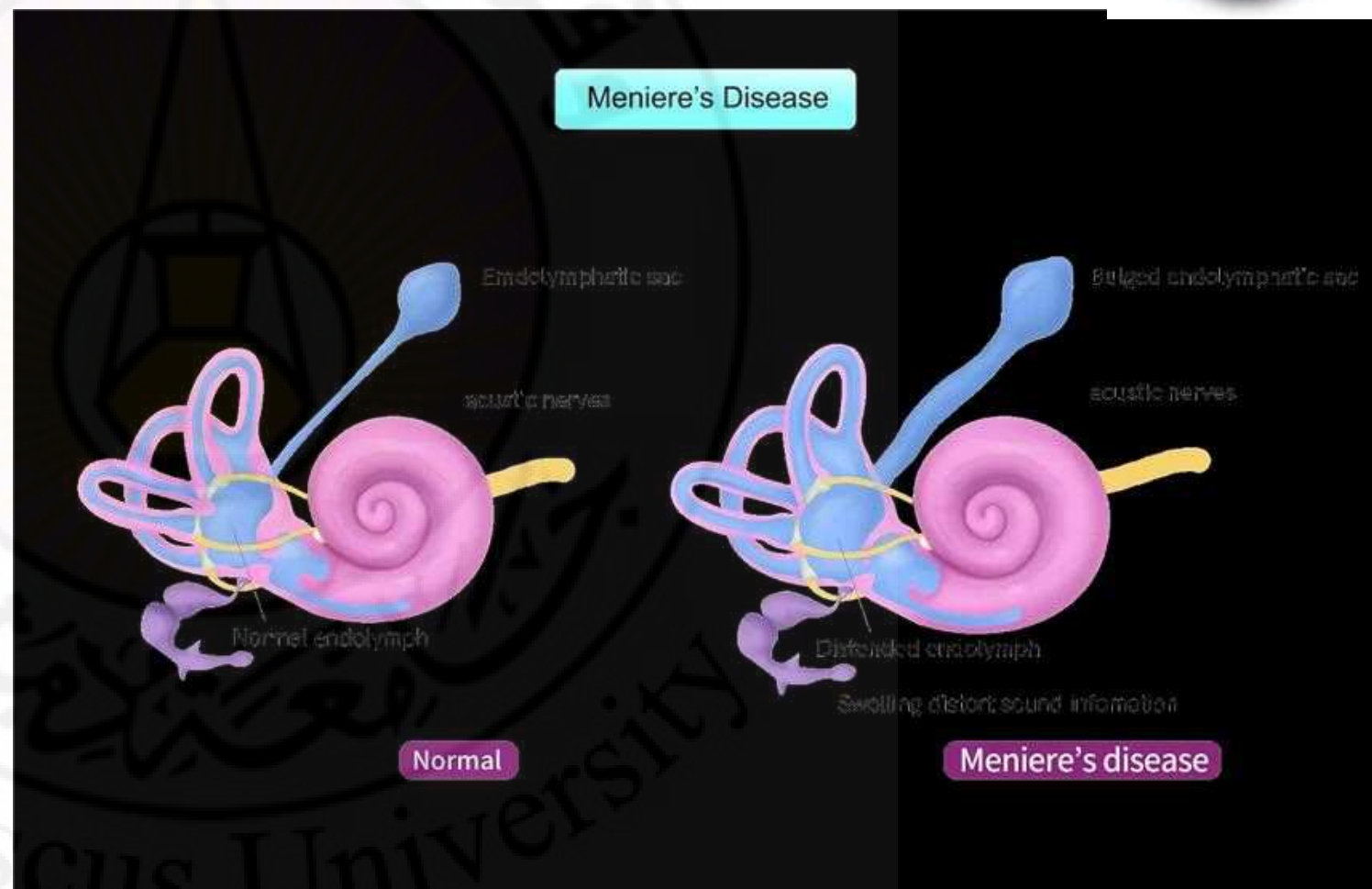
Ototoxicity: minimizing the risk of hearing loss

- The major factors affecting the risk of ototoxicity include **dosage**, **duration** of treatment, the adequacy of the patient's **renal (kidney) functioning**, and whether other potentially ototoxic agents are also being used.
- The ototoxic effects of aminoglycosides are substantially greater when the patient is also receiving diuretics. A further complication is that many ototoxic drugs are also toxic to the kidneys.
- Ototoxic drugs also interact with **noise** to produce greater amounts of hearing loss.
- To minimize the risk of hearing loss, it is important to closely monitor those patients receiving ototoxic drugs for renal functioning, drug levels in their blood, and hearing status throughout the treatment period. A **baseline audiological evaluation** should be obtained as soon as possible.
- Because of the risk of delayed onset and/or progression of hearing loss, **periodic audiological monitoring** should continue for quite some time after treatment is over.



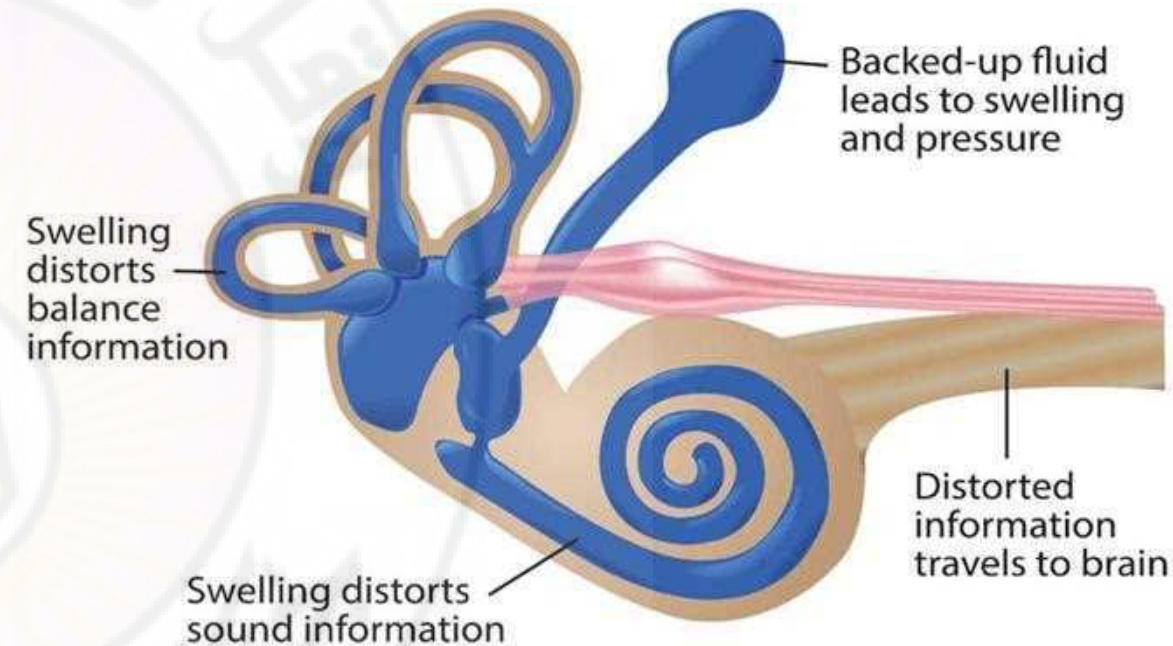
Meniere's Disease

- A common disorder of the cochlea is endolymphatic hydrops. **Endolymphatic hydrops** is a condition resulting from excessive accumulation of endolymph in the membranous labyrinth, causing Reissner's membrane to become distended.
- This excessive accumulation of endolymph often causes **Meniere's disease** or **syndrome**.
- The name of this syndrome is derived from the French scientist, **Prosper Ménière** (1861).



Meniere's Disease: Etiology

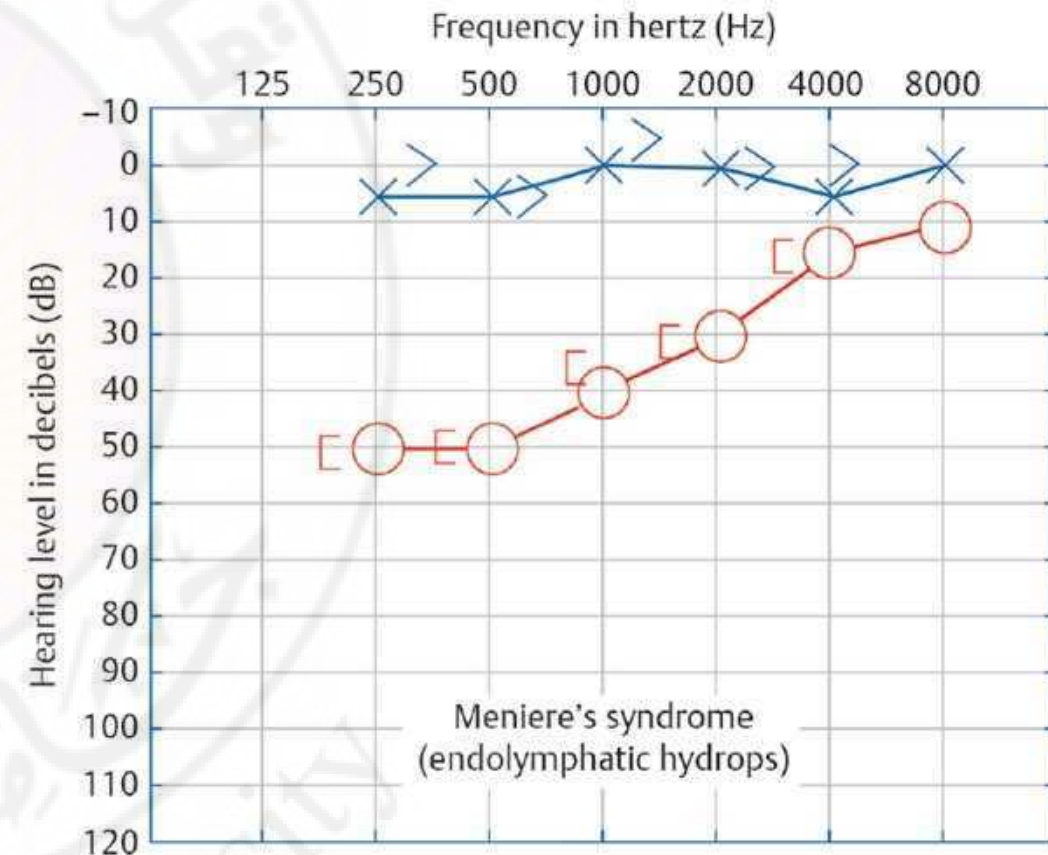
- Many causes have been proposed, and it is generally thought to be associated with problems regulating the production of endolymph or a blockage of the endolymphatic duct and sac, but the etiology of Meniere's disease is not fully established.
- Meniere's disease has been attributed to many different causes, such as food allergies, hypothyroidism, adrenal and pituitary gland insufficiencies, autoimmune disorders, vascular diseases, stenosis of the internal auditory meatus, trauma, syphilis, viral infections, and genetic origins.
- However, Meniere's disease is often viewed as an **idiopathic disorder** because precise etiologies are difficult to establish.



The underlying cause of Ménière's disease is endolymphatic hydrops. The underlying cause of endolymphatic hydrops is most often unknown.

Meniere's Disease: Symptoms

- Clinically, Meniere's disease is characterized by episodic attacks of (1) **vertigo**, (2) **hearing loss**, (3) **tinnitus**, and (4) a feeling of **fullness** or **pressure** in the ear.
- **Vertigo** attacks last for as little as 20 minutes or as long as several hours. The vertigo can be extremely severe and debilitating and is the principal problem for most patients.
- The spells can last from minutes to 2 or 3 hours and often include *unsteadiness* between spells.
- In the early stages of the disease, attacks are dominated by vertigo, and recovery can be complete. In the later stages, attacks are dominated by hearing loss and tinnitus, and permanent, severe hearing loss can occur.



Meniere's Disease: Symptoms

- Episodes of rotatory vertigo are exacerbated by head movements and is often accompanied by nausea, vomiting, diarrhea, and sweating.
- Sudden, unexplained **falls** without loss of consciousness or associated vertigo may also occur in Meniere's disease (reported to occur in 2–6% of patients).
- Tumarkin (1936) attributed these to acute **utrículo-saccular dysfunction** and they have been termed '**otolithic crises of Tumarkin**' or **drop attacks**.
- It is felt that as a consequence of an abrupt change in otolithic input an erroneous vertical gravity reference occurs. Patients often describe being pushed or feeling as if the floor has been pulled out from under them.



Drop Attack in Meniere Disease

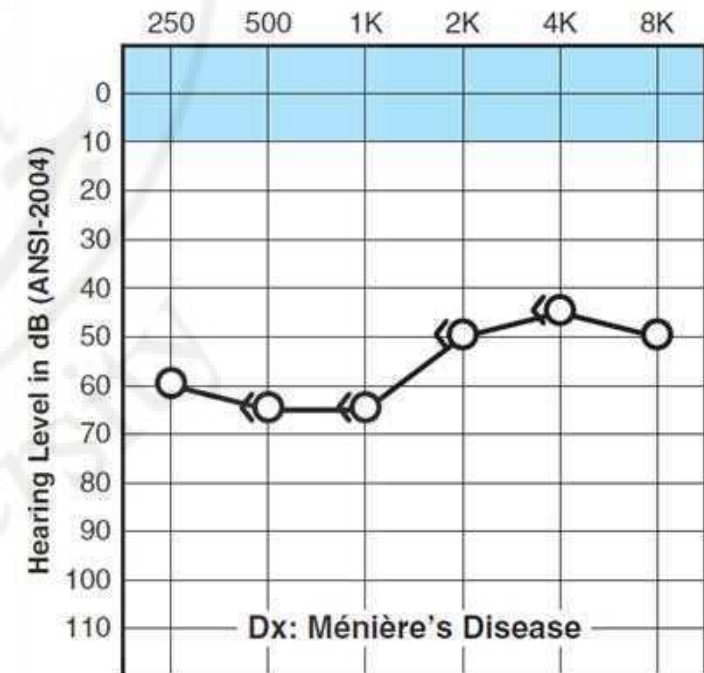
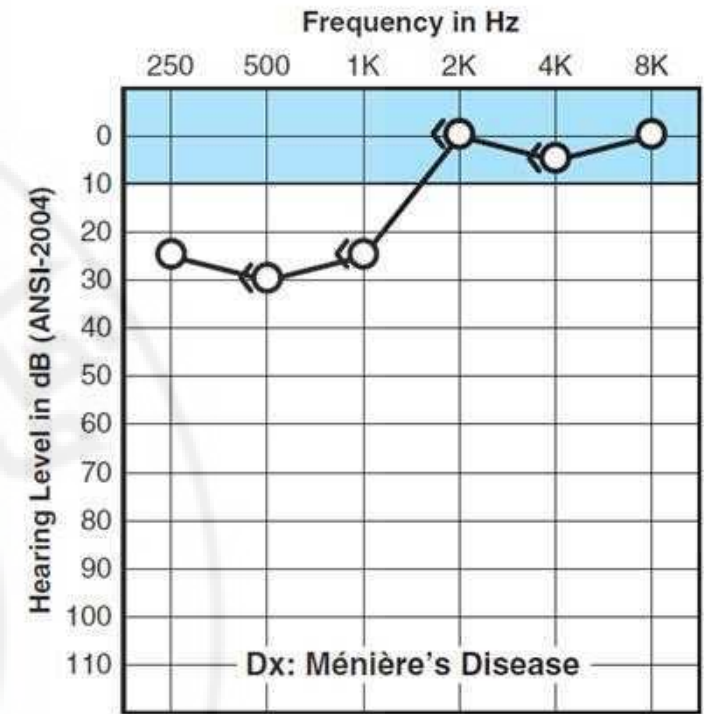
JAMA Neurology



JAMA Network

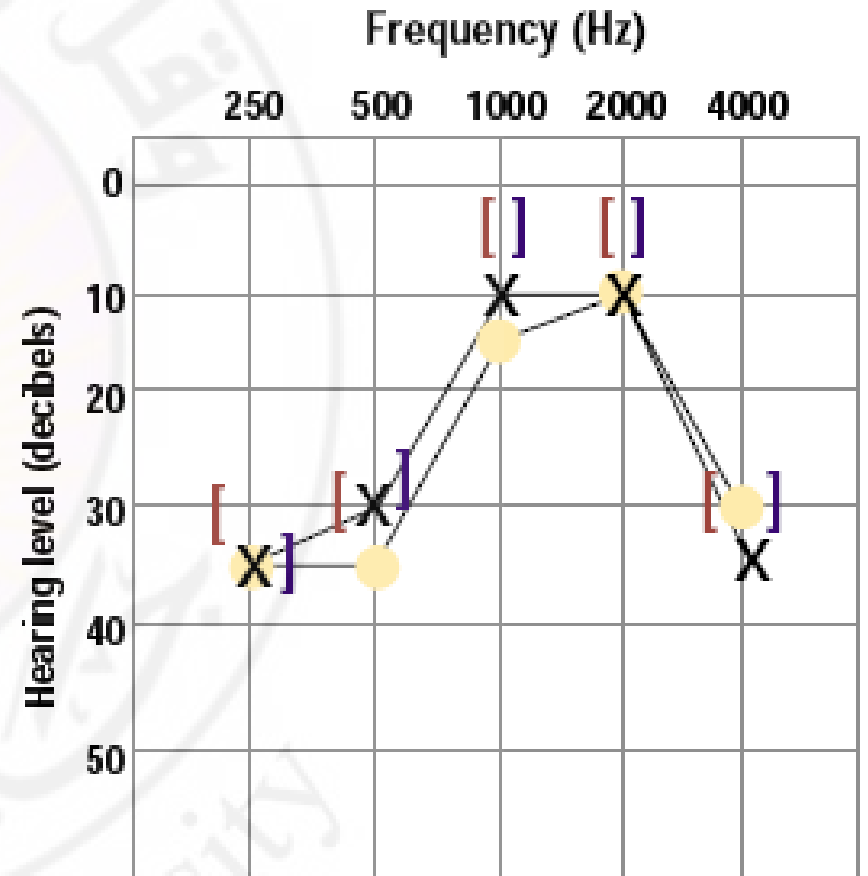
Meniere's Disease: Symptoms

- Hearing loss from Meniere's disease is **most often** unilateral, sensorineural, progressive, and fluctuating.
- In the early stages of the disease, the hearing loss configuration is often **low frequency** in nature.
- After repeated attacks, the loss usually progresses into a **flat**, moderate-to-severe hearing loss.
- One common feature of Meniere's disease is poor speech-recognition ability, much poorer than would be expected from the degree of hearing sensitivity loss.



Meniere's Disease: Symptoms

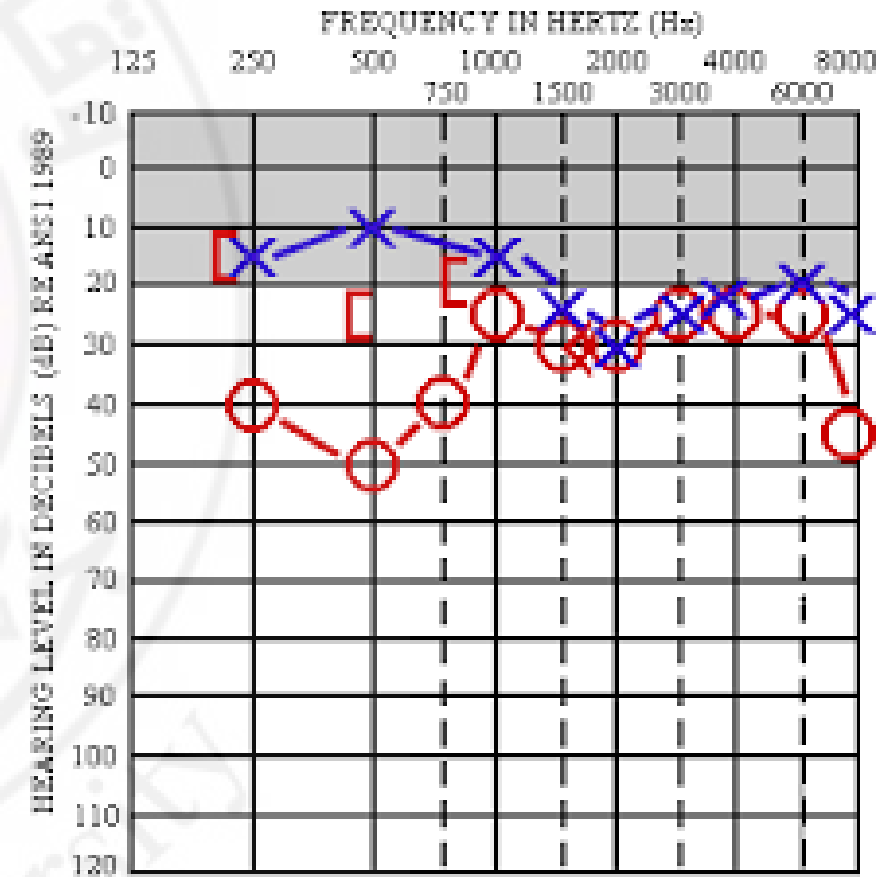
- **Meniere's disease** has also been associated with thresholds that are poorer at the higher and lower frequencies, with relatively better sensitivity at the frequencies between them.
- These **peak-shaped audiograms** can be due to the endolymphatic hydrops itself or to the combination of a low-frequency loss from Meniere's disease with a high-frequency loss from, for example, noise or aging.
- The **tinnitus** is usually described as roaring or as a low-pitch noise, but this can vary.



Peak-shaped audiograms

Meniere's Disease: Symptoms

- Occasionally, the presence of a low-frequency **air-bone gap** without middle ear disease is reported in early MD, probably as a result of the inner ear pressure exerted against the medial surface of the stapedial footplate during bouts of hydrops.
- This conductive component may be a pitfall for patients with MD having a feeling of aural fullness because they may be mistakenly treated for middle ear disease.



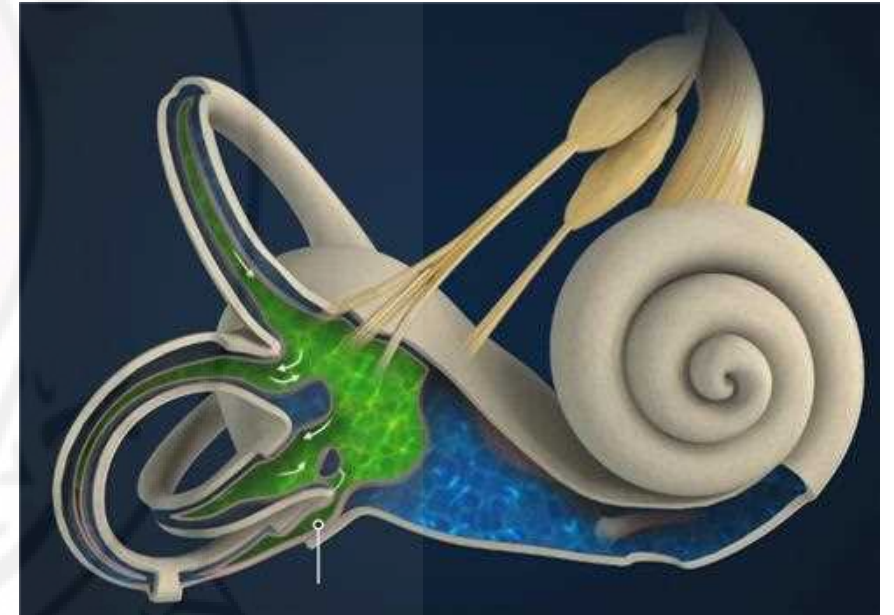
Meniere's Disease: Symptoms

- The impairment **fluctuates** because the hearing loss and tinnitus often improve between attacks, and may even resolve between attacks in the early stages of the disease. Fluctuation is common, but it is not essential for the diagnosis.
- Attacks are often preceded by an **aura** consisting of a sense of fullness in the ear, increasing tinnitus and a decrease in hearing.
- However, the long-term trend of Meniere's disease is in the direction of constant hearing loss and tinnitus that worsens during attacks, and it is possible for the loss eventually to become profound. **Profound hearing loss** in the affected ear(s) occurs in **only 1–2%** of severely affected patients.



Meniere's Disease: Variants

- Although Meniere's disease usually involves both the auditory and vestibular mechanisms, two variant forms exist, so-called **cochlear Meniere's disease** and **vestibular Meniere's disease**.
- In the **cochlear form** of the disease, only the auditory symptoms are present, without vertigo.
- In the **vestibular form**, only the vertiginous episodes are present, without hearing loss.
- One study has shown that 50% of patients presented with vertigo and hearing loss together, 19% with vertigo only, and 26% with hearing loss alone.
- Meniere's disease is **unilateral** in roughly 70 to 85% of the cases, but the incidence of **bilateral** cases increases with the duration of the disease, reaching ~ 40% after 15 years.



Meniere's Disease: Diagnostic Criteria

• Definite MD

- A. Two or more spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours.
- B. Audiometrically documented low- to medium frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during or after one of the episodes of vertigo.
- C. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear.
- D. Not better accounted for by another vestibular diagnosis.

• Probable MD

- A. Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours.
- B. B. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear.
- C. D. Not better accounted for by another vestibular diagnosis.

Meniere's Disease: Diagnostic instrumental support

- **Audiologic tests**

- Pure tone and speech audiometry
- Auditory Brainstem Responses (ABR)
- Traveling-wave velocity (TWV)
- Low-frequency masking (LFM)
- Immittance tests
- Electrocochleography

- **Vestibular tests**

- VNG + Caloric testing
- Vestibular-evoked myogenic potentials
- Video-head impulse test (vHIT)

- **Magnetic resonance Imaging (MRI)**

Test	Possible Common Findings
Caloric	Caloric paresis on the affected side
Cervical Vestibular Evoked Myogenic Potentials	Significant amplitude asymmetry (weaker on the ipsilesional side)
Ocular Vestibular Evoked Myogenic Potential	Significant amplitude asymmetry (weaker on the ipsilesional side)
Video Head Impulse testing	Often normal in the presence of abnormal caloric responses (McCaslin, Rivas, Jacobson, & Bennett, 2015)
Ocular motor testing	Common to see spontaneous nystagmus <i>Acute</i> —spontaneous nystagmus will beat toward the unaffected ear (fast phase) <i>Recovery nystagmus</i> —spontaneous nystagmus will beat toward the affected ear (See Chapter 2 for review)
Audiometry	Low-frequency fluctuating sensorineural hearing loss early in the course of the disease.

Meniere's Disease: Management

- Treatment of MD should be a combination of lifestyle and dietary changes, medical therapy, and invasive therapy.
- Conservative treatment must first be considered and destructive treatment is preferred for patients with irreversible hearing loss.
- Medical treatment generally consists of a low-salt diet, diuretics, and the use of vestibular suppressants and sedatives to control the vertigo. (Corticosteroid injections into the middle ear have also been tried.)
- Medical treatment is effective in controlling the vertigo in ~ 70% of patients, but ~ 30% require more invasive approaches, which include both nondestructive and destructive methods.

Conservative Treatment
Life style chage, Medical Treatment(Duretics and Betahistine),Meniett device

Intratympanic Corsteriods

Dexamethasone and Methylprednisolone

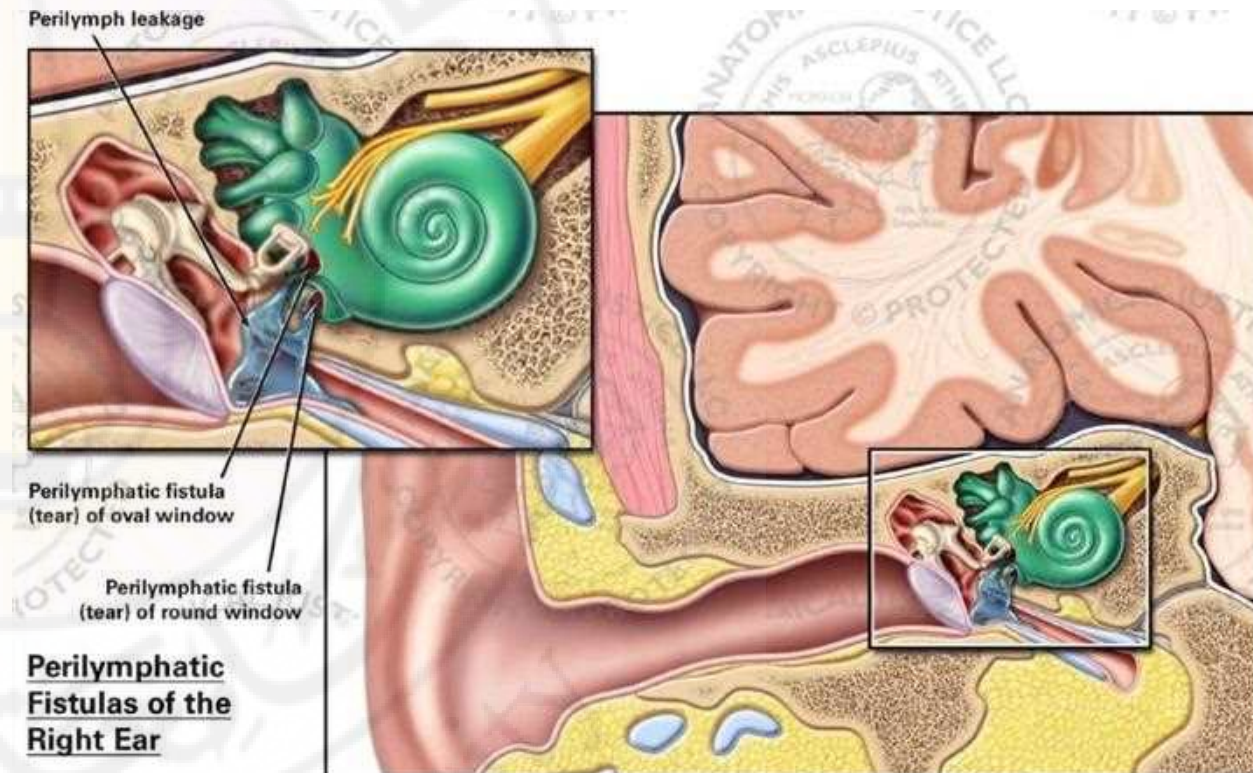
Endolymphatia Sac Surgery
Endolymphatic sac decompression,
Endolymphatic sac drainage

Chemical labyrinthectomy
Intratympanic Gentamicin

Destructive Surgery
Vestibular neurectomy,
Labyrinthectomy

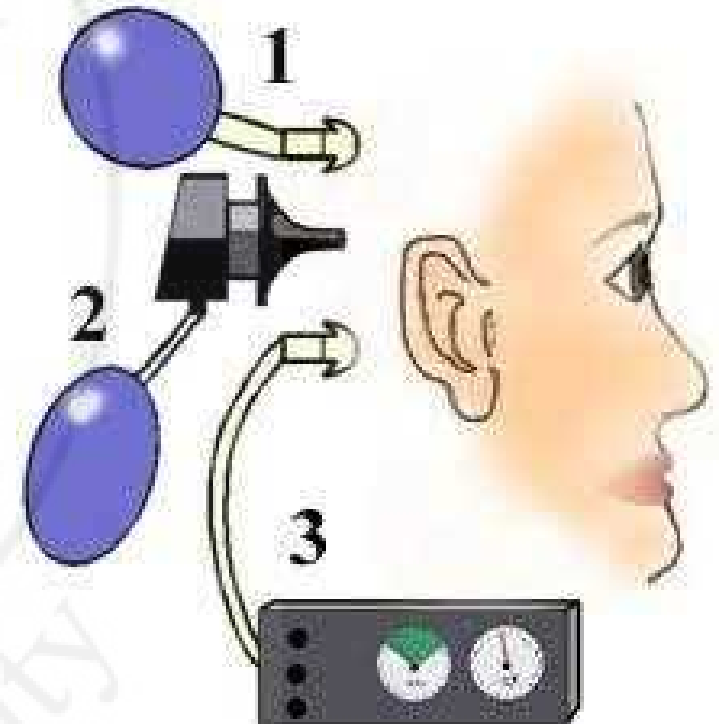
Perilymphatic Fistulas

- A **perilymphatic fistula** is a leak of perilymph from a rupture of the oval and/or round window, and is one of the causes of **sudden hearing loss**. The fistula itself can be quite small and even microscopic.
- Oval and round window fistulas can result from explosions, barotrauma, and increased intracranial pressure due to straining or physical exertion, sneezing, coughing, Valsalva maneuvers, and other precipitating events.
- Fistulas may also be the result of bone erosion due to a cholesteatoma or infection, and of untoward outcomes of stapes surgery.



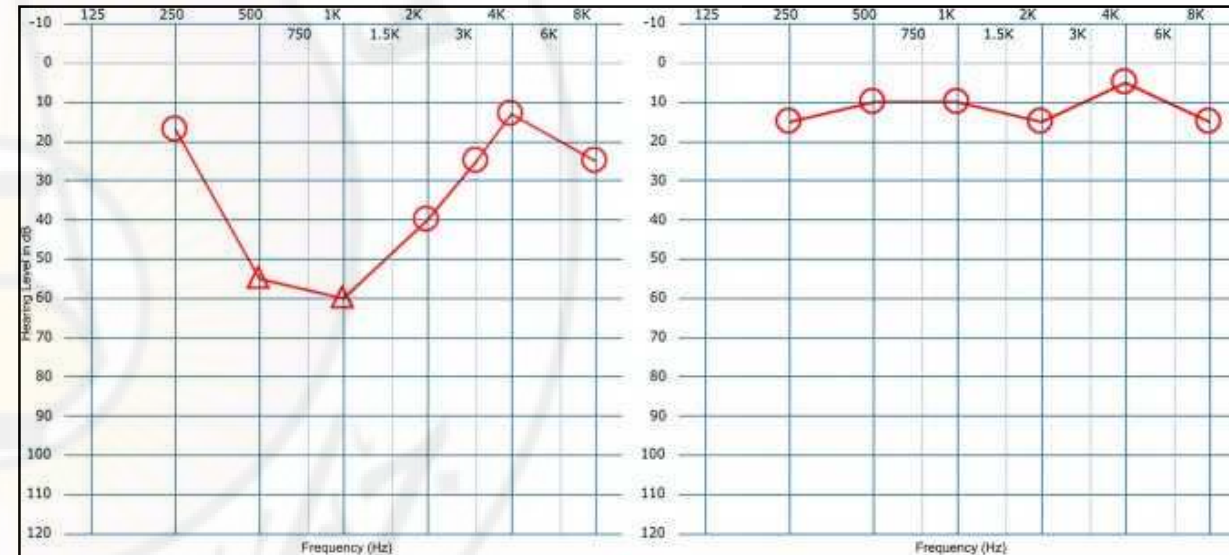
Perilymphatic Fistulas

- The **fistula test** is usually administered to a patient who is suspected of having a perilymphatic fistula.
- Air is pumped into the ear canal by squeezing the bulb of a pneumatic otoscope or using the pressure pump of an acoustic immittance device. The positive pressure is expected to cause a considerable amount of vertigo and nystagmus if the patient has a perilymphatic fistula.
- **Here is why:** The air pressure causes an inward displacement of the tympanic membrane that is transmitted to the stapes footplate, which in turn presses in on the perilymph. There will be an abnormally large amount of fluid displacement because the fistula acts like an open valve. The excessive fluid displacement overstimulates the receptors in the semicircular canals, resulting in vertigo and nystagmus.



Perilymphatic Fistulas: treatment

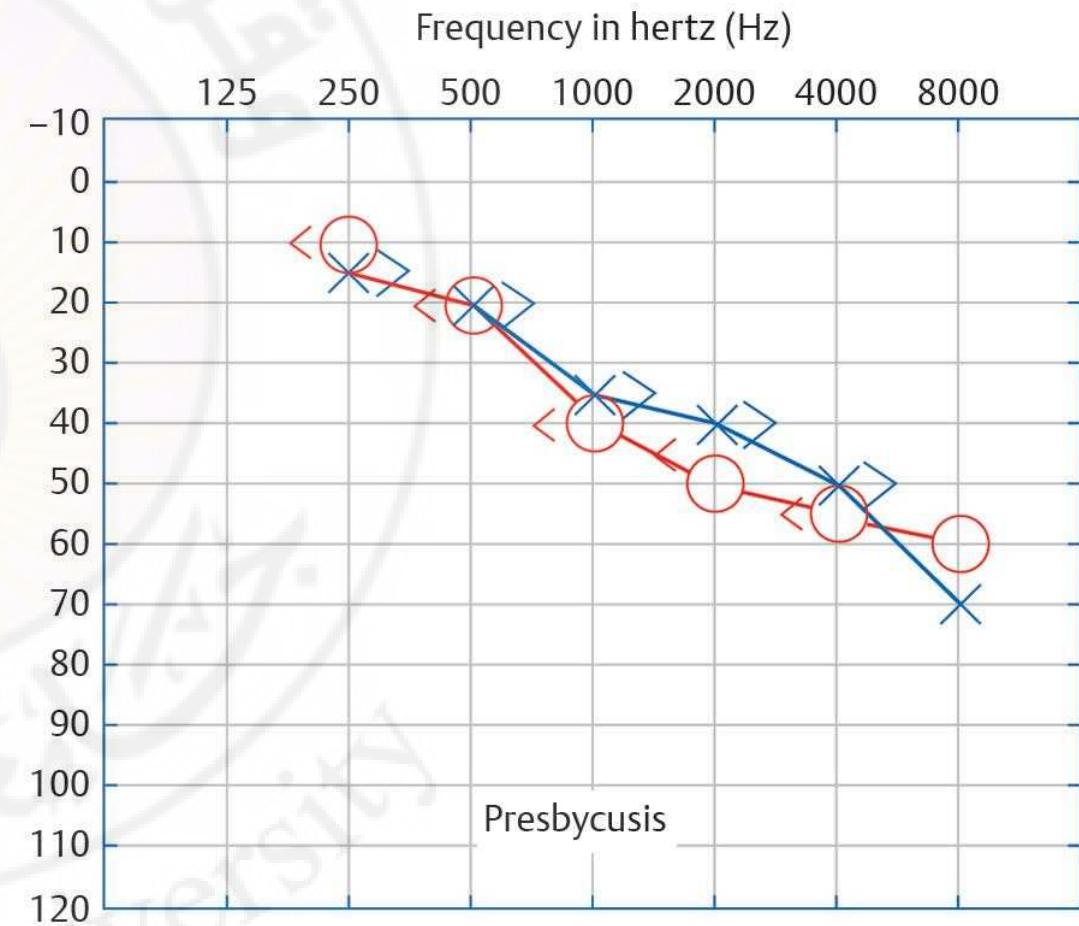
- Once a perilymphatic fistula is diagnosed, the otologist might choose to treat it conservatively, or to perform a surgical exploration of the middle ear (tympanotomy) to find and repair the fistula.
- The **conservative approach** might include bed rest with the head elevated and avoidance of straining.
- The **surgical repair** of a fistula involves patching it with materials such as perichondrium, a vein graft, or fat. Successful surgery can arrest the progression of hearing loss or even improve hearing to some extent if performed soon enough.

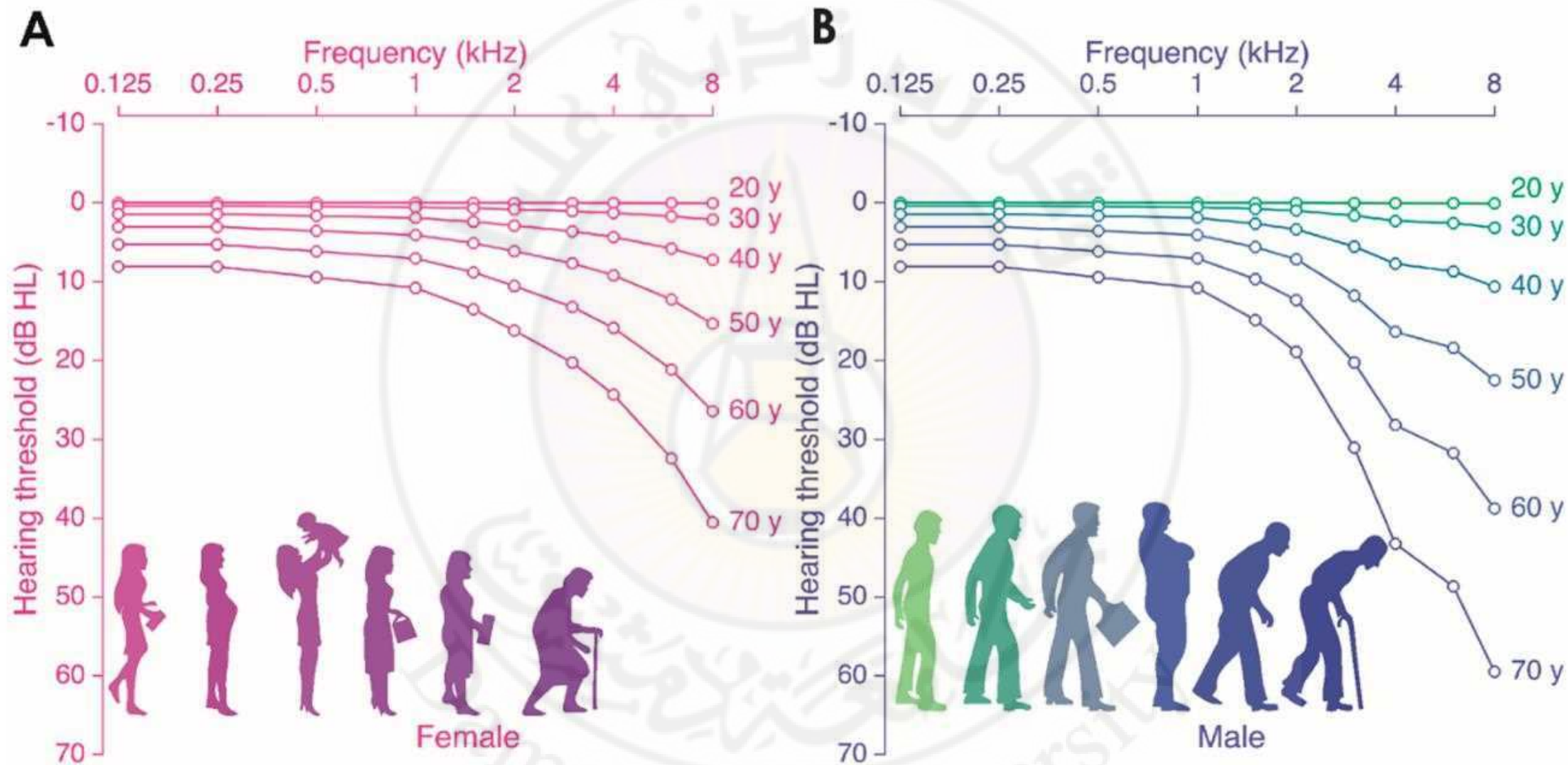


Audiograms of a patient presenting with sudden hearing loss after blowing her nose (left panel) showing improvement of hearing 1 week after a blood patch procedure (right panel).

Presbycusis

- **Presbycusis (or presbycusis)** is a decline in hearing as a part of the aging process.
- As a collective cause, it is the leading contributor to hearing loss in adults.
- Estimates suggest that from 25 to 40% of those over the age of 65 years have some degree of hearing loss. The percentage increases to approximately 90% of those over the age of 90 years.
- Presbycusis is often described in terms of how hearing thresholds change with increasing age.





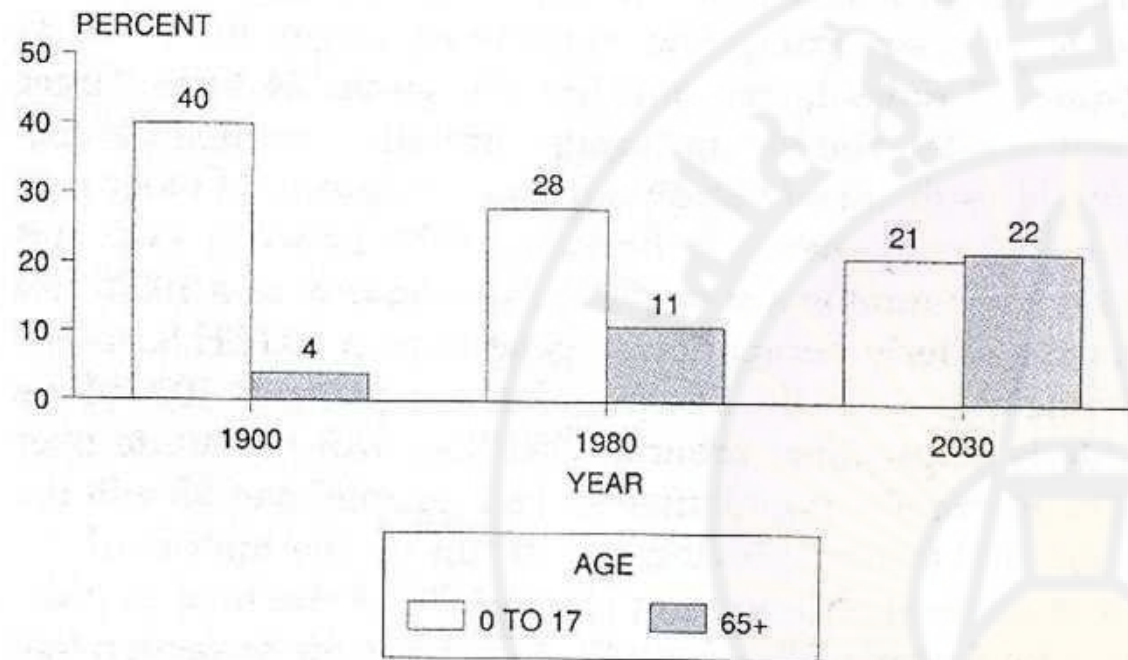
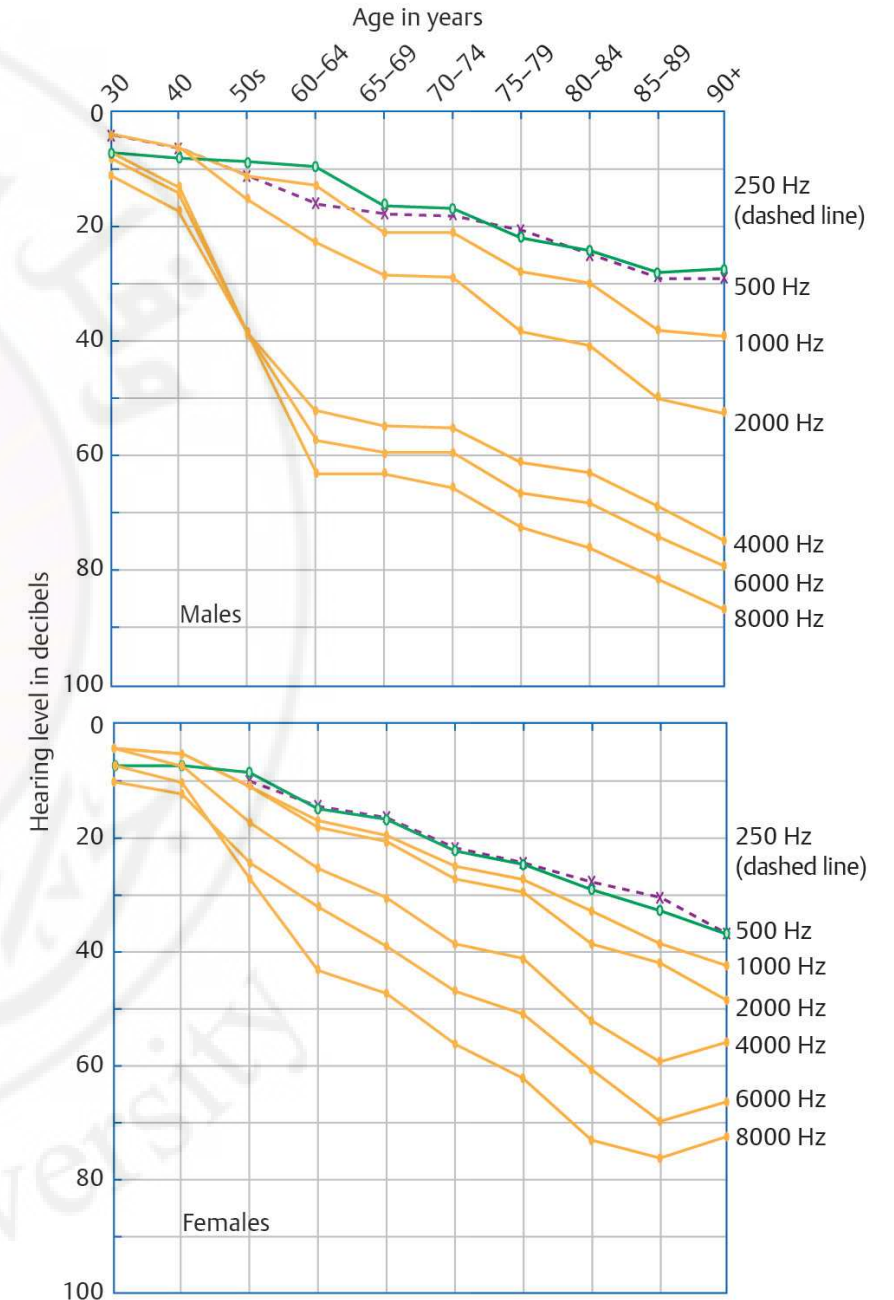


Figure 1-1 Percent of children and elderly in the population: 1900, 1980, and 2030. (Sources: 1900 figures, which exclude Alaska, Hawaii, and Armed Forces overseas: U.S. Bureau of the Census. "Estimates of the Population of the United States, by Single Years of Age, Color, and Sex: 1900 to 1959." *Current Population Reports Series P-25*, no. 311 (July 1965). 1980 and 2030 figures: U.S. Bureau of the Census. "Projections of the Population of the United States, by Age, Sex, and Race: 1988 to 2080," by Gregory Spencer. *Current Population Reports Series P-25*, no. 1018 [January 1989].)



Presbycusis

- There is a general consensus that presbycusis is the result of various kinds of physiological degeneration due to the normal aging process (theoretically “pure presbycusis”) **plus** the accumulated effects of noise exposure, ototoxicity, medical disorders and their treatment, and genetic components.
- Age-related threshold shifts are greater and begin sooner for the **higher frequencies**, suggesting greater involvement toward the base of the cochlea, and **males** are affected more than females, presumably reflecting more exposure to occupational and recreational noises.



Presbycusis

- Structures throughout the auditory system degenerate with age. Changes in cochlear hair cells, the stria vascularis, the spiral ligament, and the cochlear neurons all conspire to create sensorineural hearing loss. Changes in the auditory nerve, brainstem, and cortex conspire to create auditory processing disorder.
- Hair cell loss near of the cochlea produces hearing loss in the high frequencies, which typifies presbycusis.

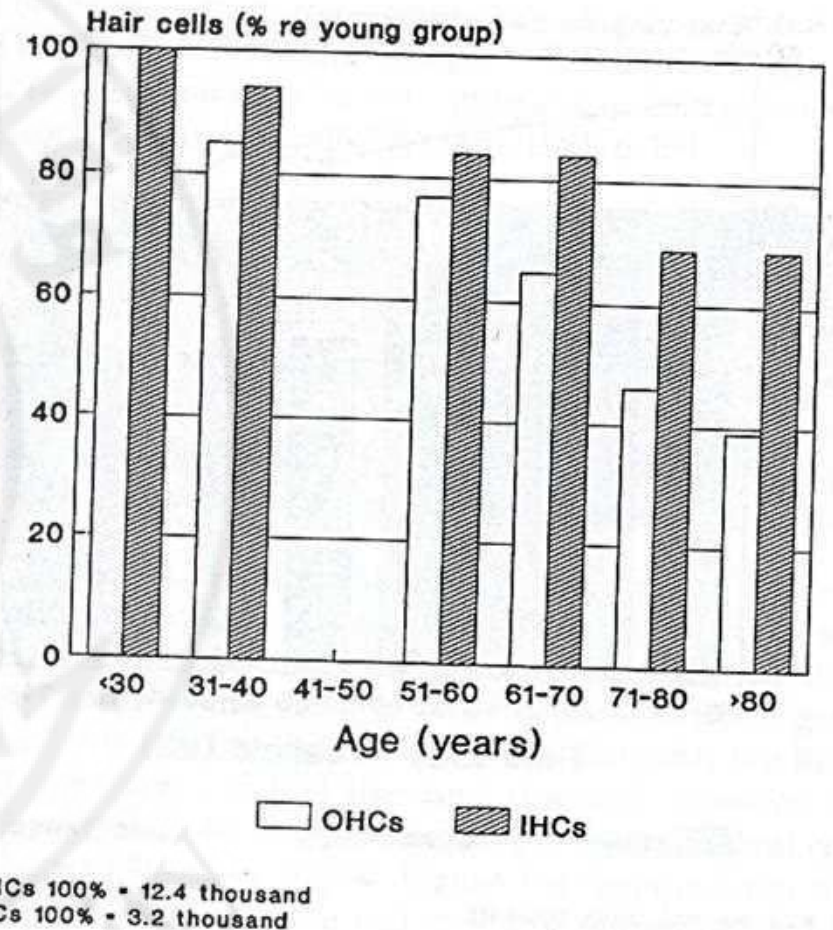


Figure 4-2 Inner and outer hair cell loss as a function of increasing age. (Adapted from Otte, Schuknecht, and Kerr, 1978. Reprinted by permission.)

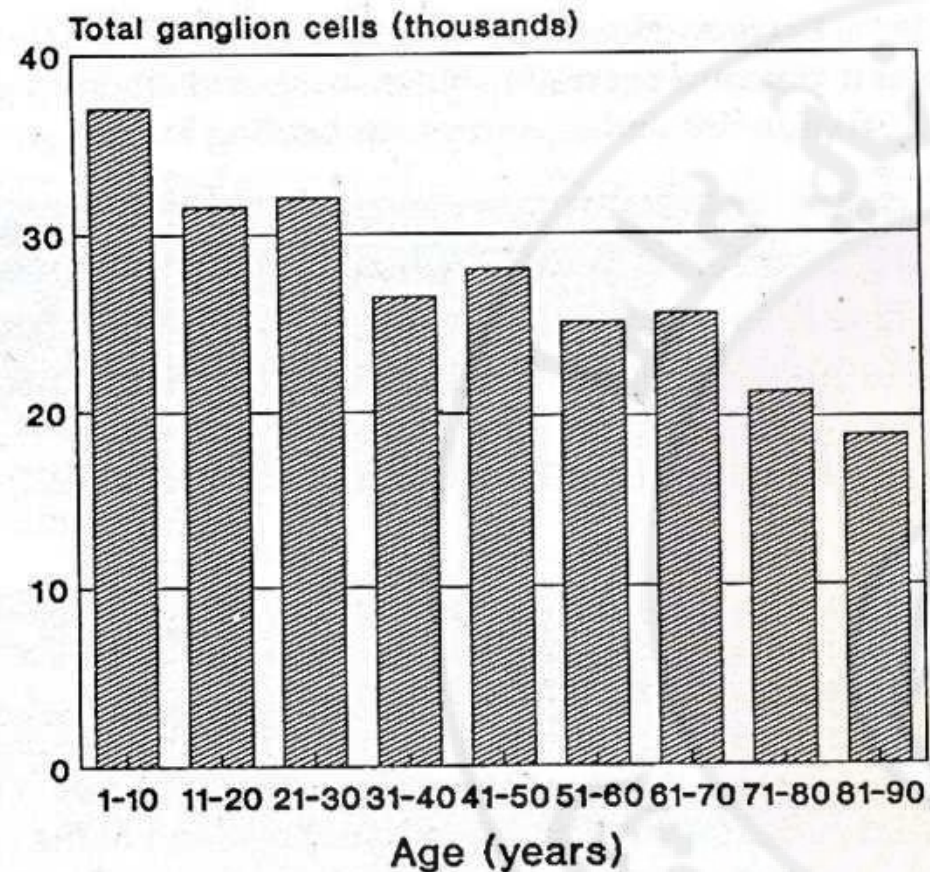


Figure 4-3 Total ganglion cell loss as a function of increasing age. (Adapted from Otte, Schuknecht, and Kerr, 1978. Reprinted by permission.)

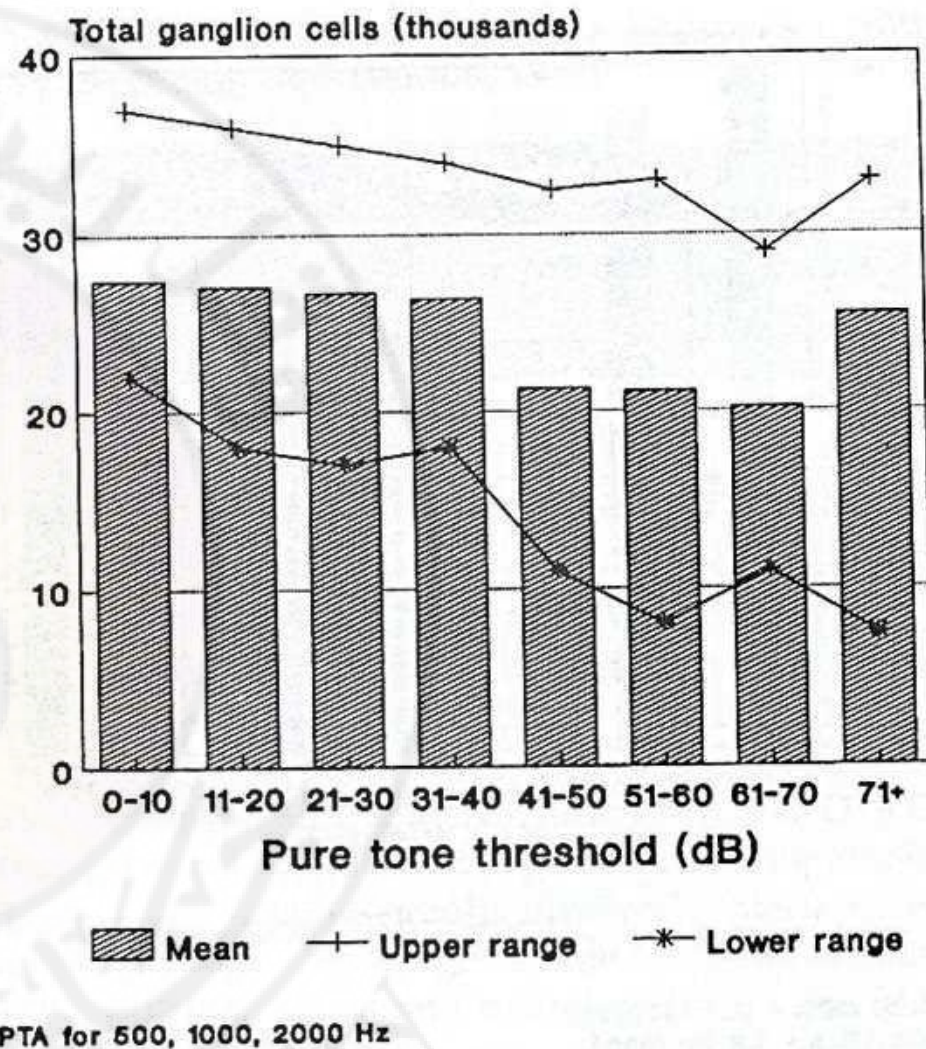
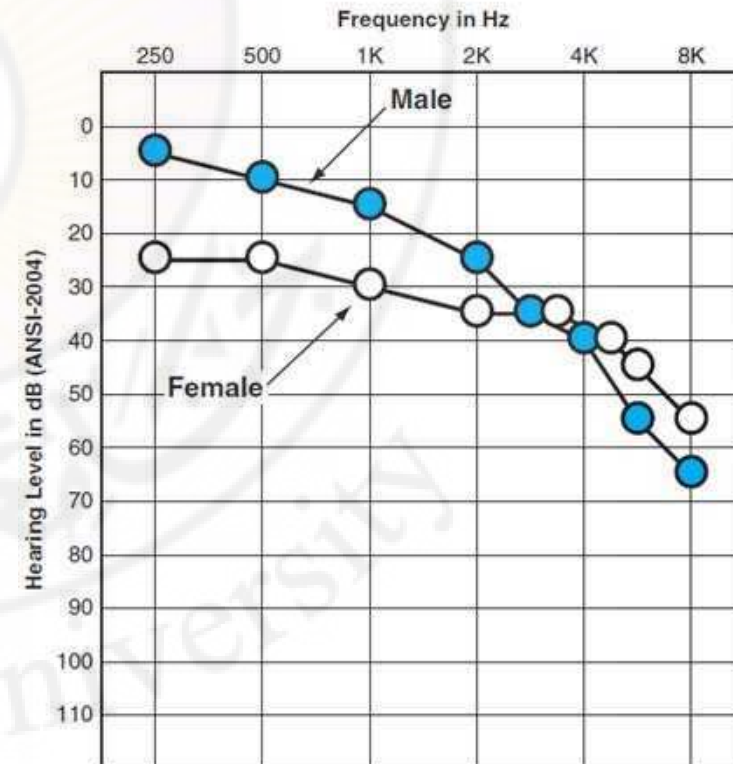
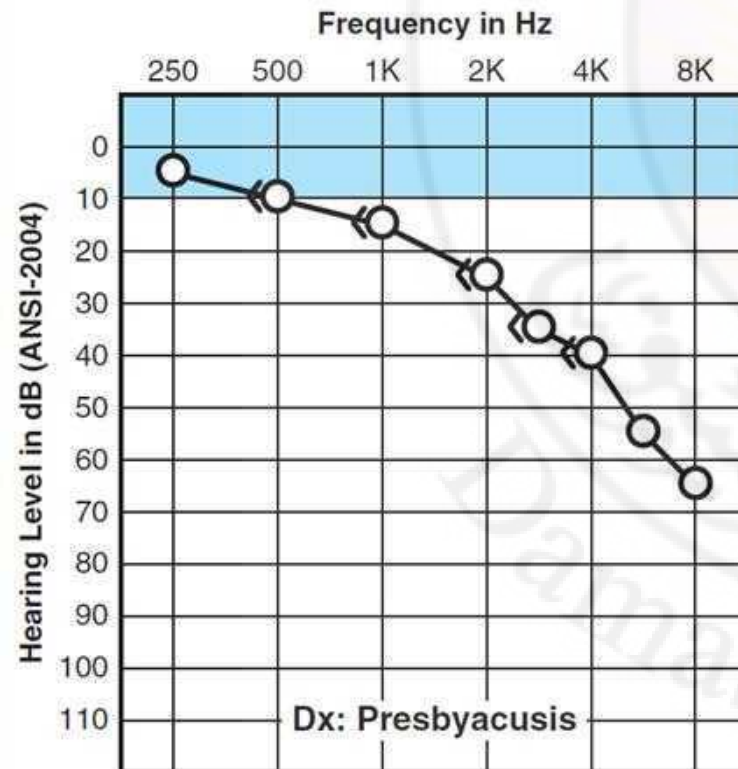


Figure 4-4 Spiral ganglion cell loss as a function of pure-tone average. (Adapted from Otte, Schuknecht, and Kerr, 1978. Reprinted by permission.)

Presbycusis

- Hearing sensitivity loss from presbycusis is bilateral, usually symmetric, progressive, and sensorineural.
- **Men** tend to have more high- frequency hearing loss, and **women** tend to have flatter audiometric configurations. When noise exposure is controlled, the amount of high-frequency hearing loss is similar in the two groups, but women tend to have more low-frequency hearing loss.



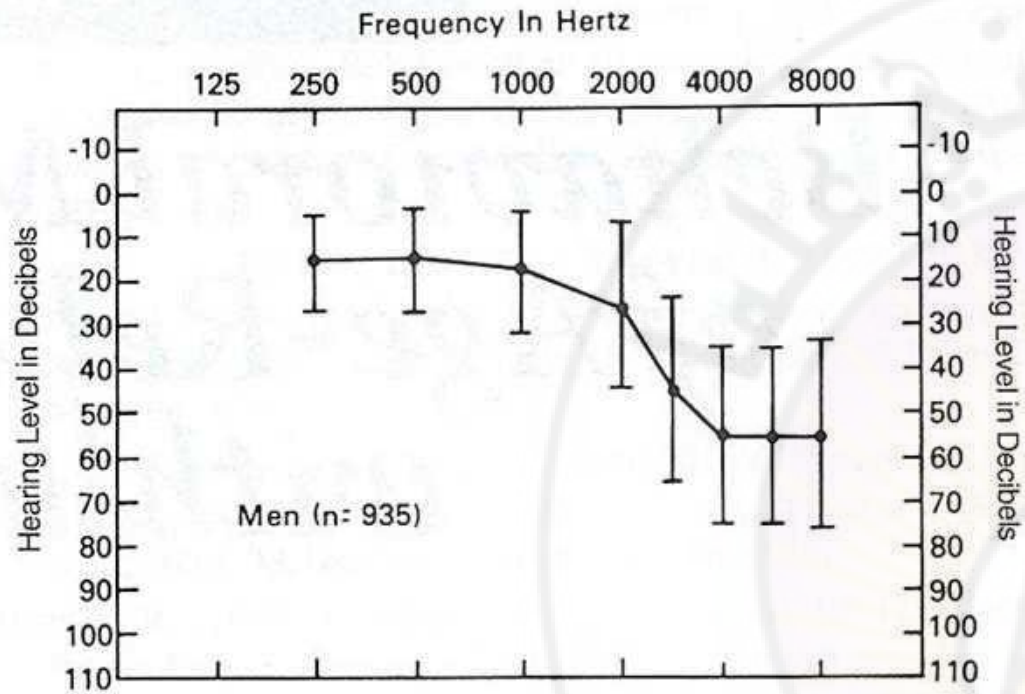


Figure 5-1 Mean pure-tone air-conduction threshold values and standard deviations in the better ear for males. (From Moscicki et al, 1985.)

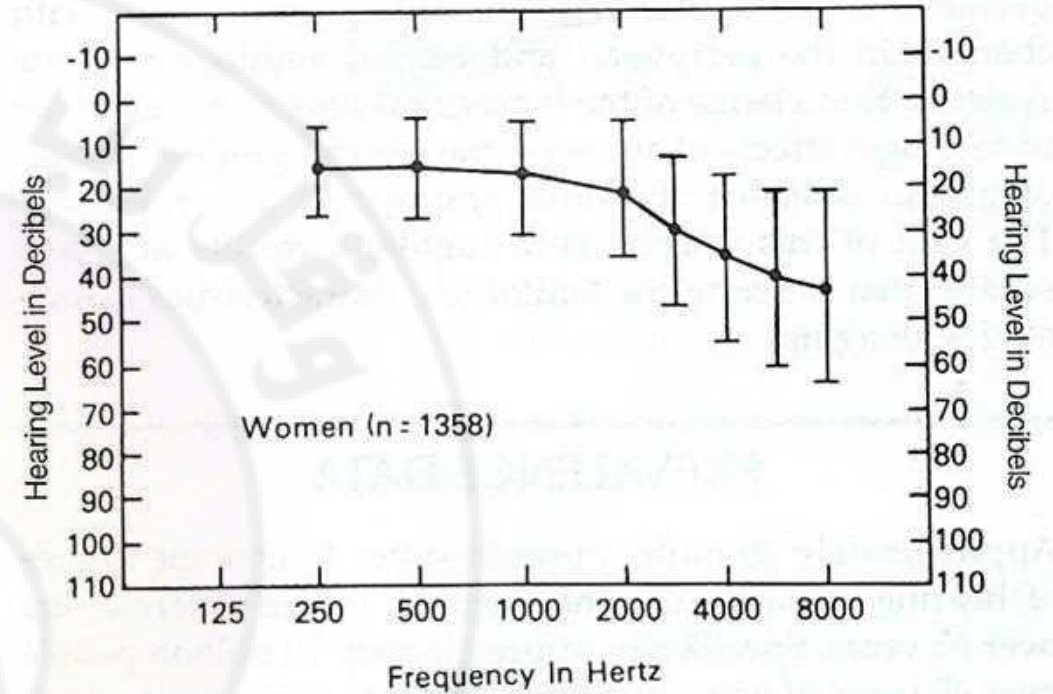


Figure 5-2 Mean pure-tone air-conduction threshold values and standard deviations in the better ear for females (From Moscicki et al, 1985.)



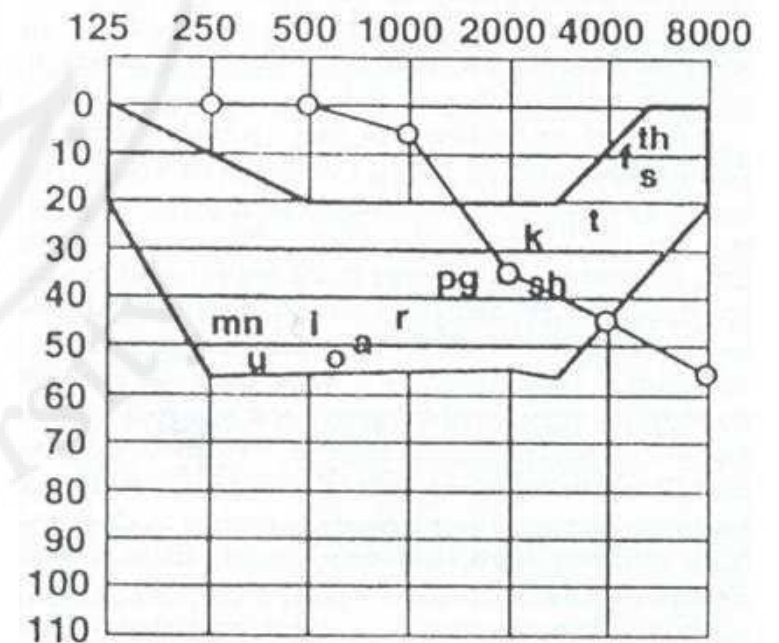
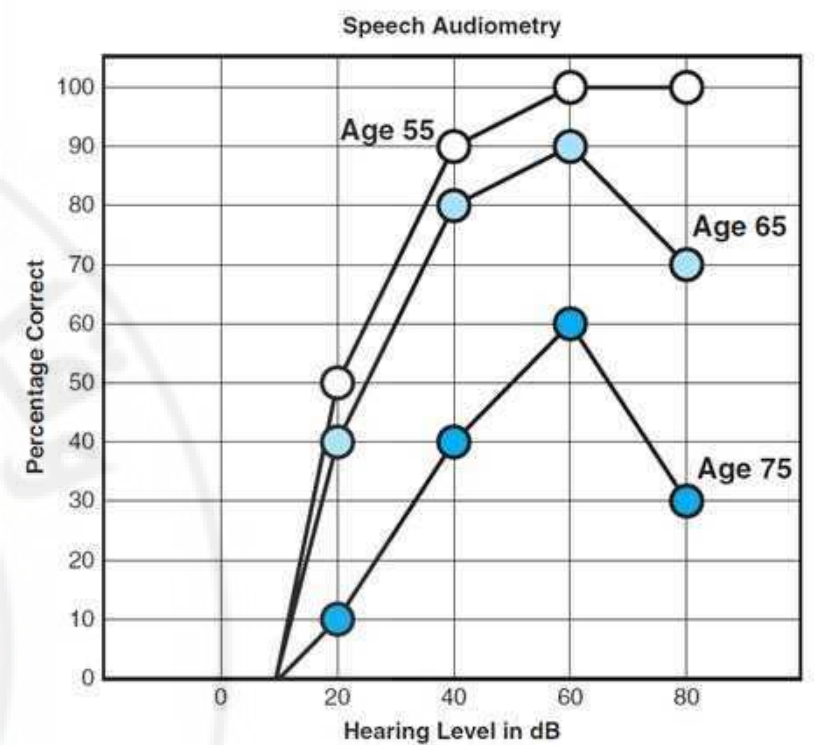
Mild to Moderately severe, Bilateral SNHL
with a Sharply Sloping Configuration.



Mild to Moderate, Gradually Sloping,
bilaterally Symmetric SNHL.

Presbycusis

- **Presbycusis** is also characterized by decline in the perception of speech that has been sensitized in some manner. Decline in **understanding of speech in background competition** or speech that has been temporally altered is consistent with aging changes in the central auditory nervous system.
- **Difficulty with speech recognition** is the most common complaint of presbycusis.



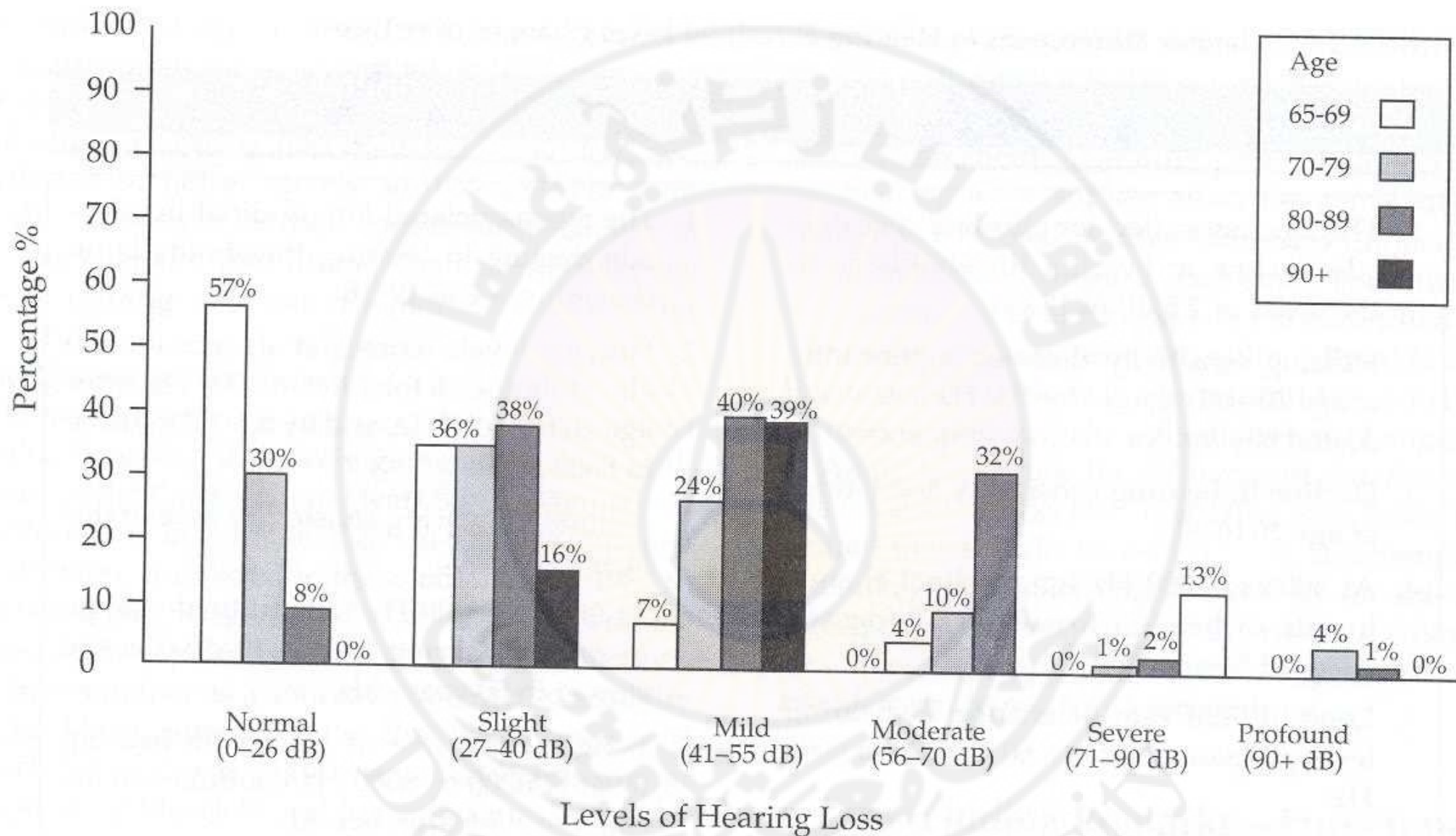


Figure 5-8 Percentage distribution of subjects showing various degrees of hearing loss within various age groups. (Modified from Schow and Nerbonne, 1980, p. 128, Table 3.)

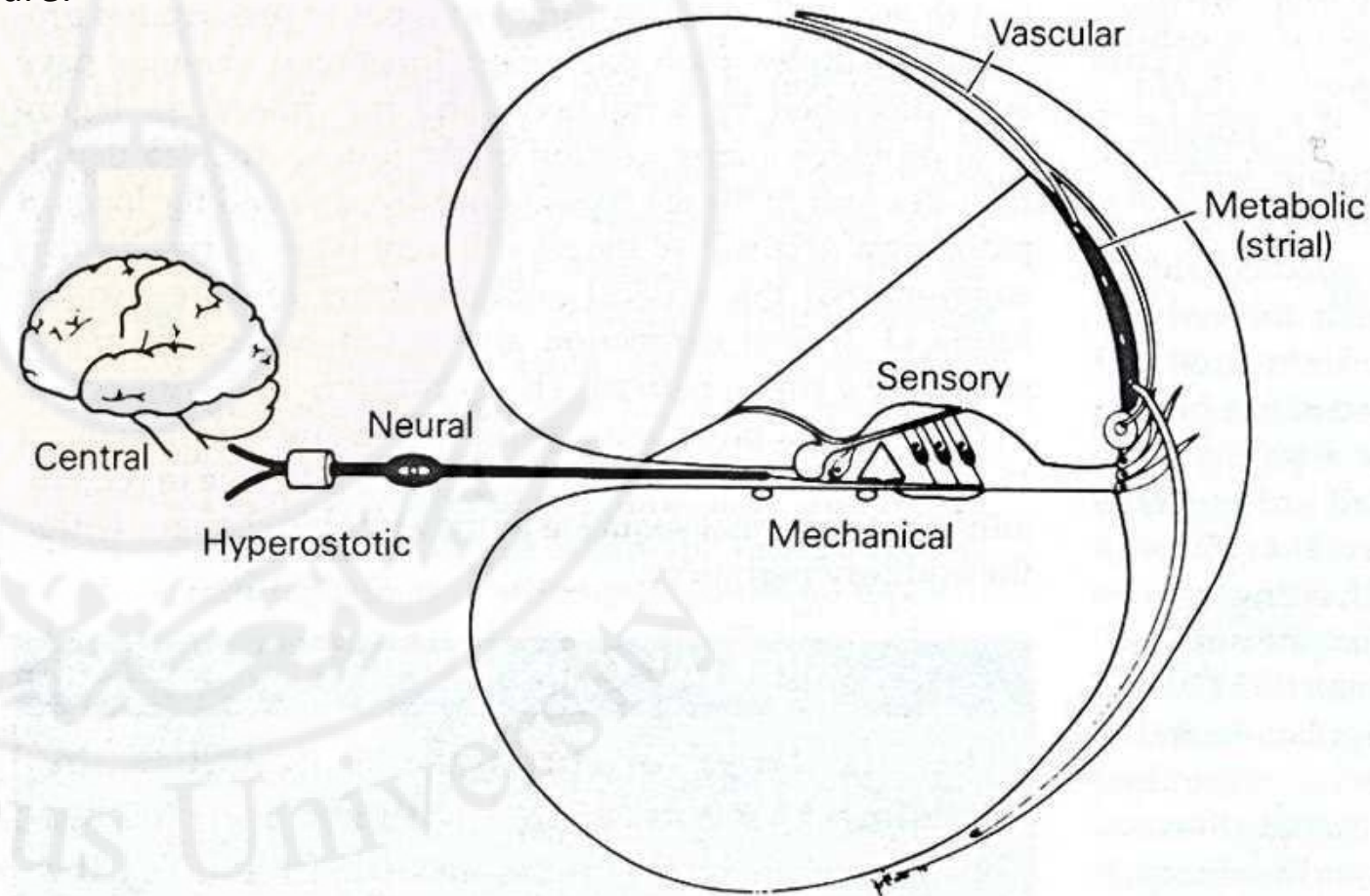
Presbycusis: Histological deterioration

- Classical types of presbycusis based on a combination of clinical and histological findings by **Schuknecht** are:

1. **Sensory presbycusis**
2. **Neural presbycusis**
3. **Metabolic or strial presbycusis**
4. **Mechanical or cochlear conductive presbycusis**
5. **Mixed presbycusis**
6. **indeterminate presbycusis**

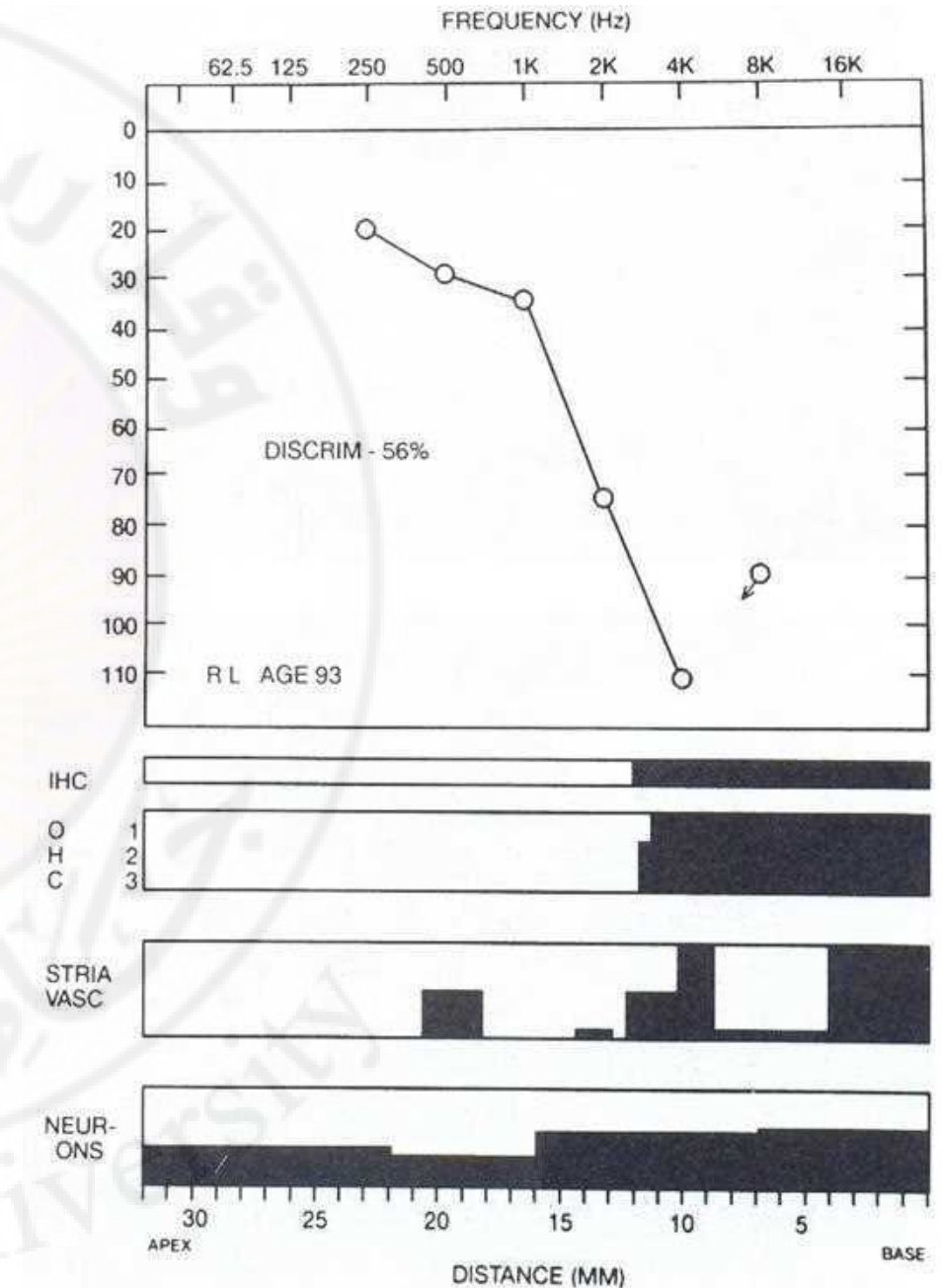
- **Additional forms**

- Vascular presbycusis
- Hyperostotic presbycusis
- Synaptopathic presbycusis
- Central presbycusis



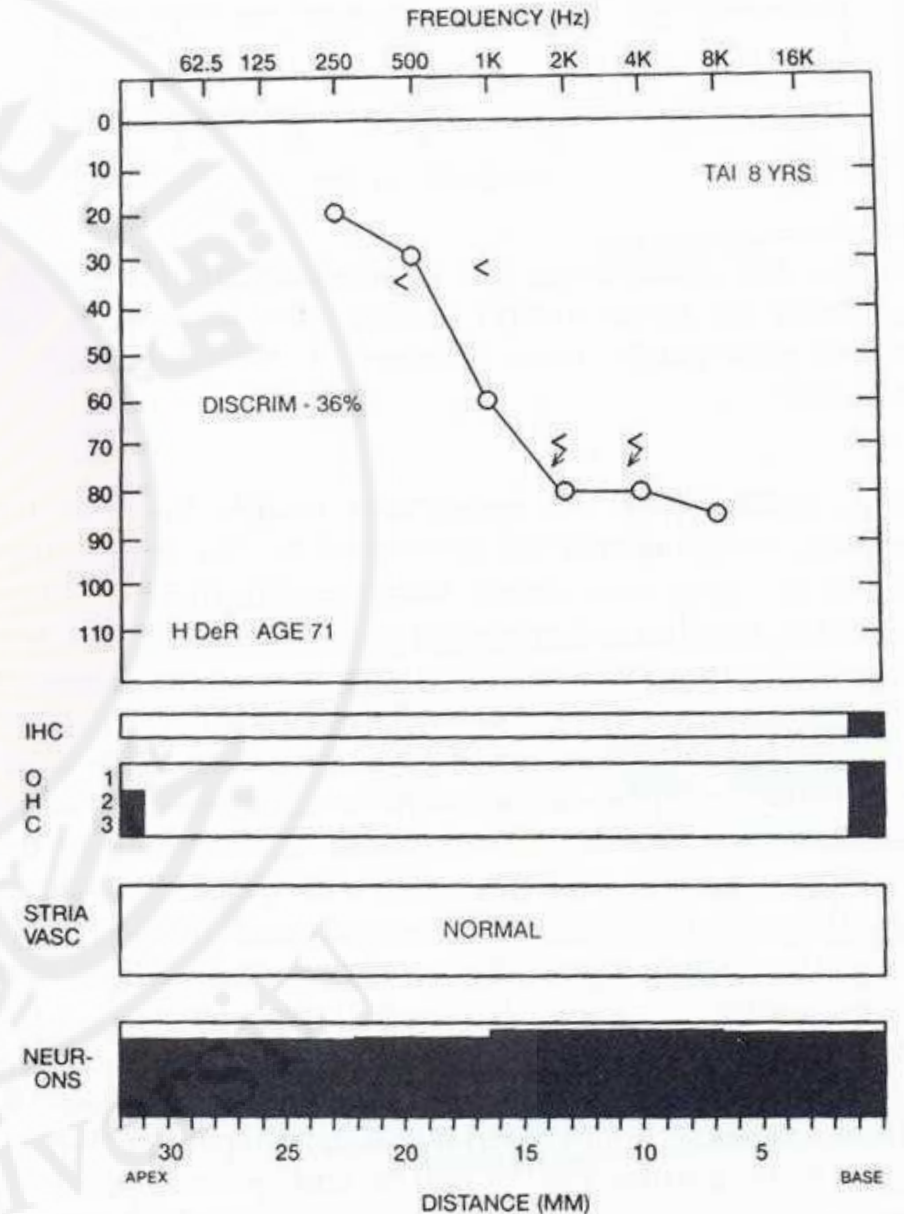
Sensory presbycusis

- **Sensory presbycusis** is due to the degeneration of the sensory hair cells and supporting cells mainly toward the basal turn of the cochlea, and can also include atrophy of the associated neurons as a secondary effect.
- It is associated with a sloping (often steeply) high-frequency sensorineural hearing loss and speech recognition scores that are consistent with the nature of the audiogram.



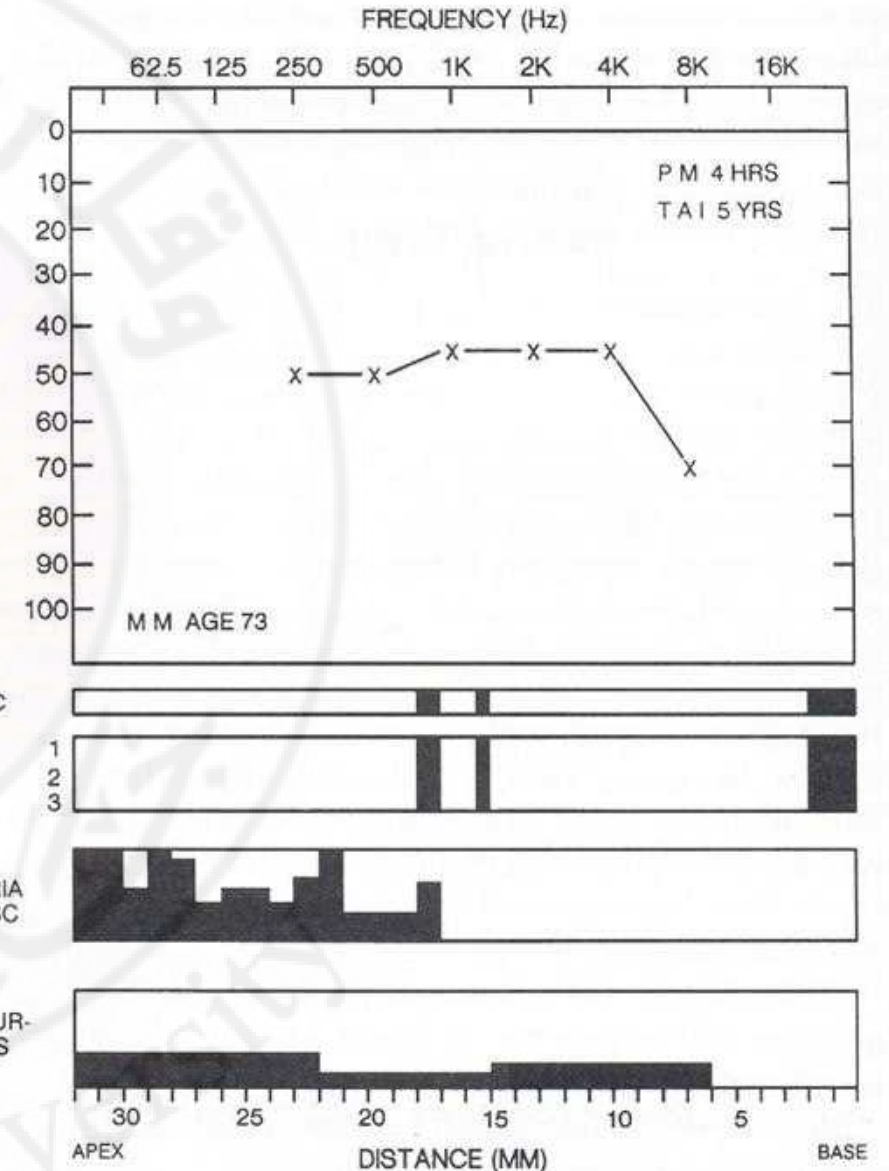
Neural presbycusis

- **Neural presbycusis** is related to primary degeneration of the auditory neurons.
- The reduced neuronal population is characterized clinically by speech recognition scores that are lower than what would be expected from the audiogram.
- Atypically poor speech recognition in presbycusis is sometimes called **phonemic regression**, but this phenomenon appears to be rare.



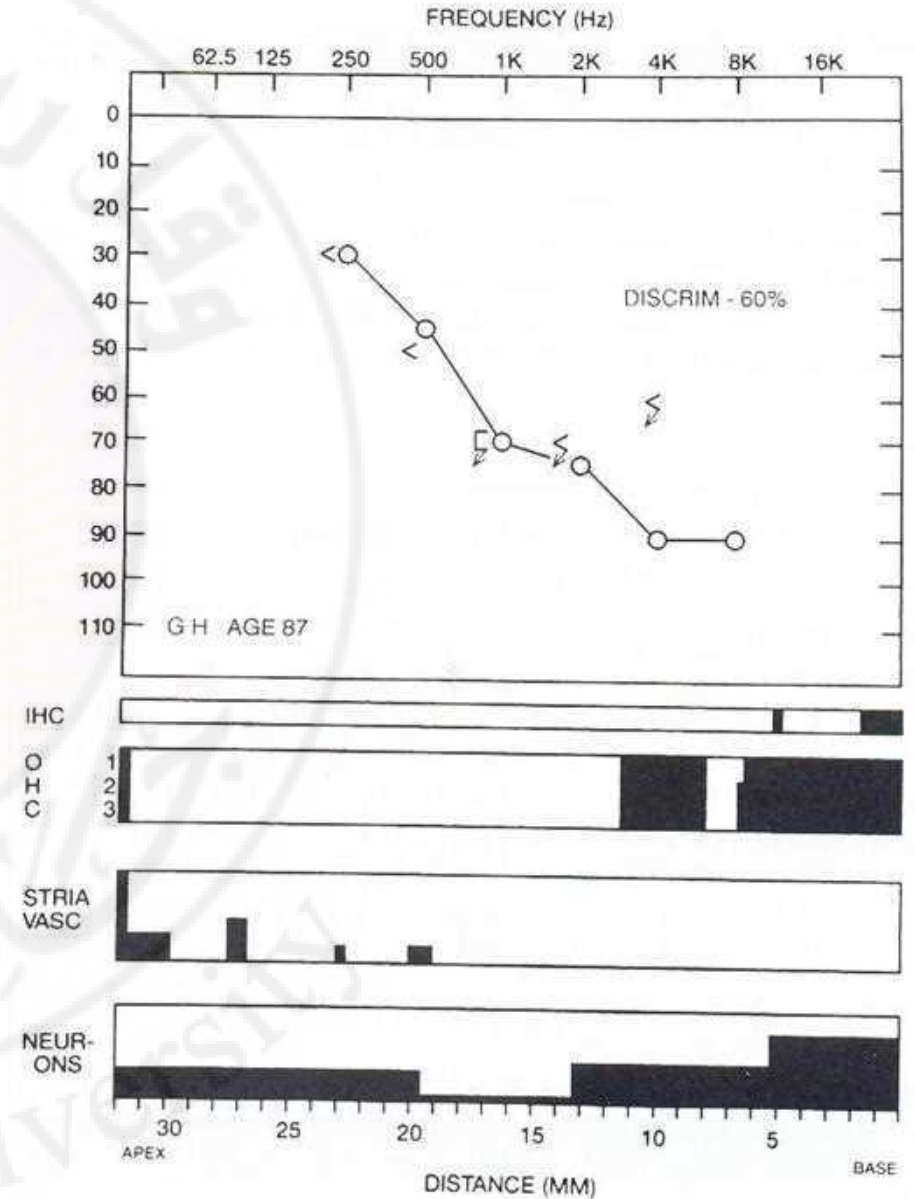
Metabolic or strial presbycusis

- **Metabolic or strial presbycusis** is caused by degeneration of the stria vascularis, causing a relatively **flat** sensorineural hearing loss with speech recognition scores that are in line with the audiogram.



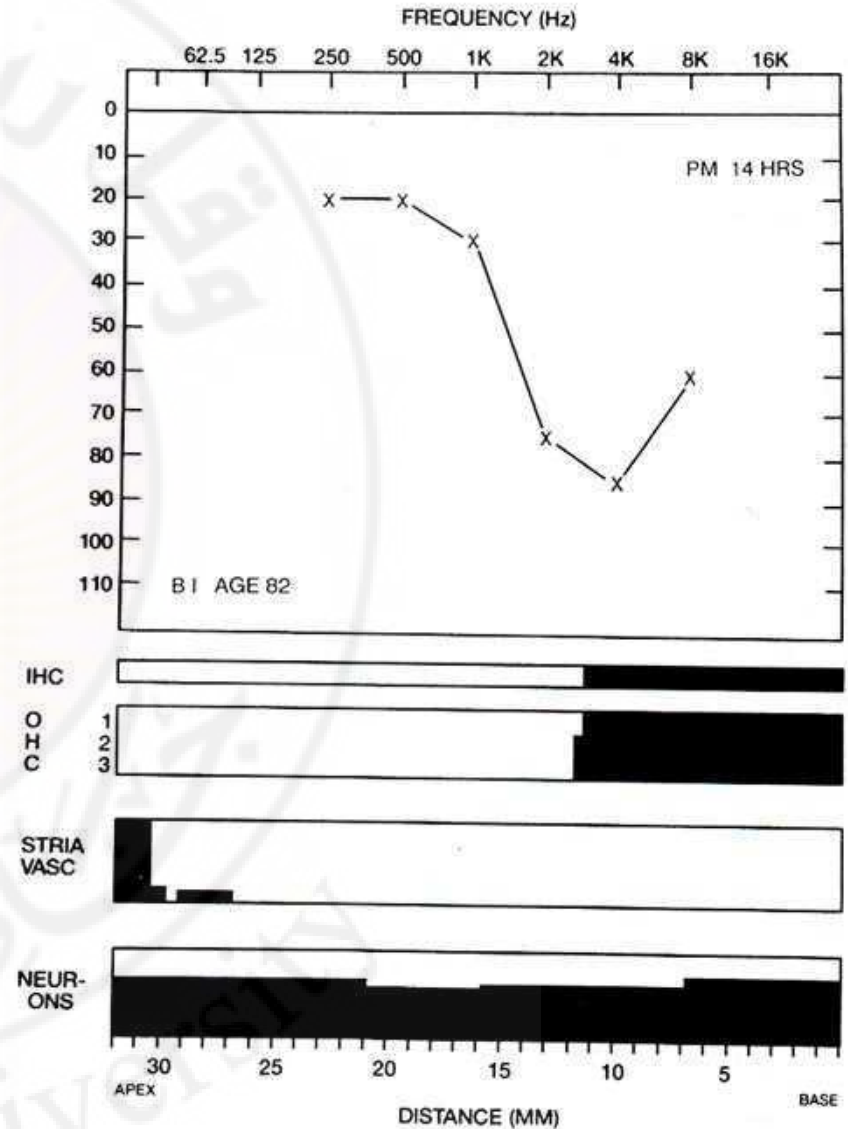
Mechanical or cochlear conductive presbycusis

- **Mechanical or cochlear conductive** presbycusis is attributed to structural changes in the basilar membrane, affecting its mass and stiffness, which would in turn affect the mechanics of the cochlea and produce a sloping sensorineural hearing loss.



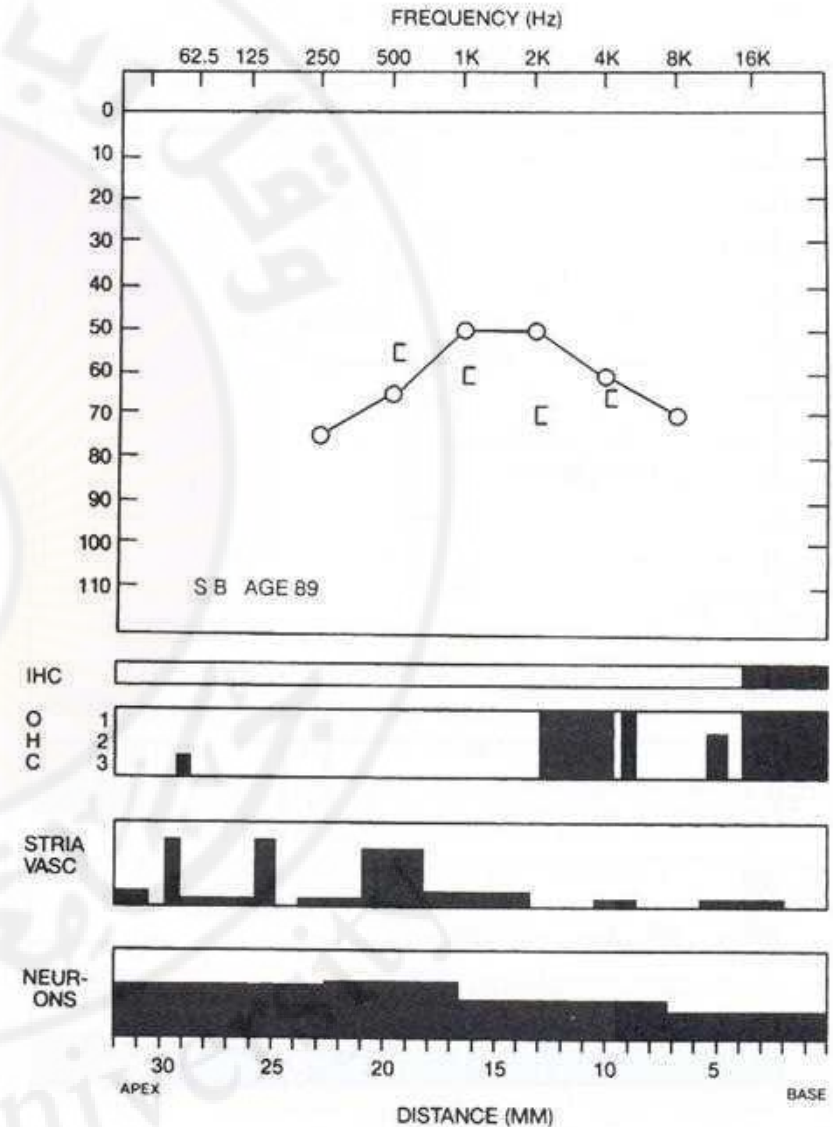
Mixed presbycusis

- When two or more presbycusis conditions are present together (pathological changes in more than one structure).
- Various configurations in audiogram



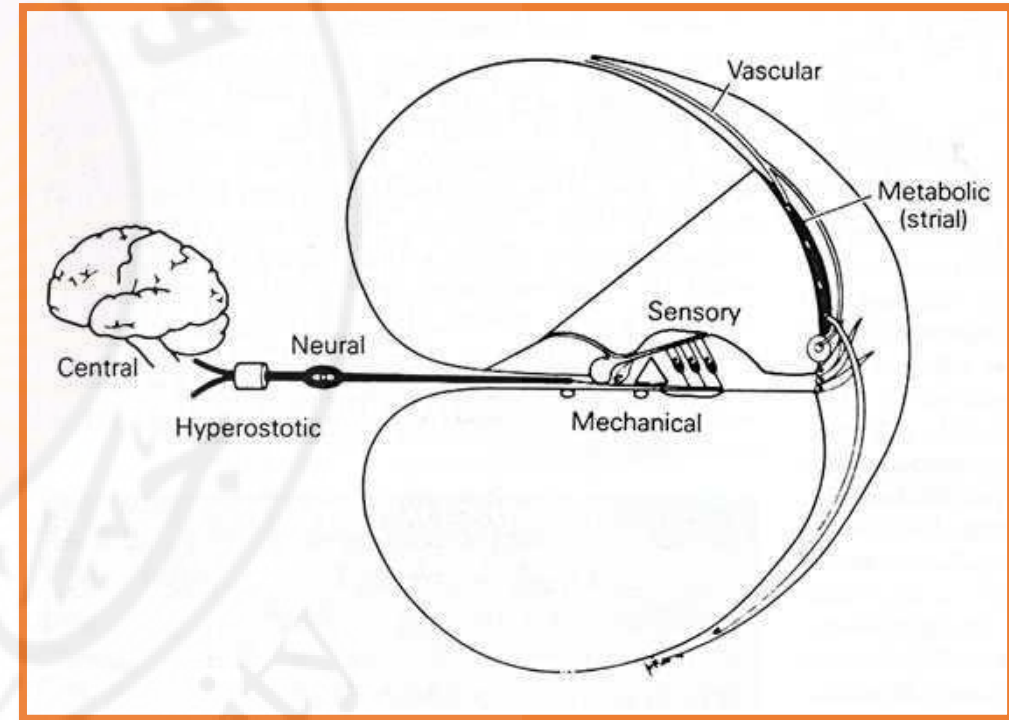
Indeterminate presbycusis

- About 25% of all cases of presbycusis show none of the above characteristics and are classified as indeterminate presbycusis.



Additional forms of presbycusis

- **Vascular presbycusis** is associated with deterioration of the blood supply to the lateral wall of the cochlear duct and the spiral lamina, and may play a part in the deterioration of the stria vascularis and spiral ligament.
- **Hyperostotic presbycusis** occurs when abnormal bone growth (hyperostosis) in the modiolus or internal auditory meatus compresses auditory nerve cells so that they degenerate.
- **Central presbycusis** refers to deficits in auditory perception that are due to age-related changes in the central auditory system.
- **Synaptopathic presbycusis**



Types of presbycusis (based on Schuknecht [27, 28])

Type	Description
Sensory	Caused by damage to outer hair cells at the basal turn of the cochlea (hearing loss in the high frequency range, 5% of cases)
Neural	Degeneration of the ganglion cells, shown by a moderate downward slope of the pure-tone threshold towards high frequency and a sharp drop in speech discrimination compared to the pure-tone threshold (e55). Based on histological data, the loss of 50% or more of 35 500 cochlear neurons is used as the criterion of neural presbycusis. Otte et al. (e56) showed that about 2100 neurons are lost in every decade.
Metabolic	Caused by atrophy of the stria vascularis (slowly progressive hearing loss with flattening of the audiogram and good speech discrimination). The loss of strial tissue results in impaired K^+ recycling, leading in turn to a decrease in endolymphatic potential (EP) (e57).
Cochlear conductive	This is described (hypothetically) as a degenerative change resulting from stiffness of the basilar membrane (e55)
Mixed presbycusis	A combination of the types described above
Indeterminate presbycusis	In about 25% of cases of hearing loss, no association can be shown between the audiometric pattern and pathological changes in the cochlea (e55, e58)



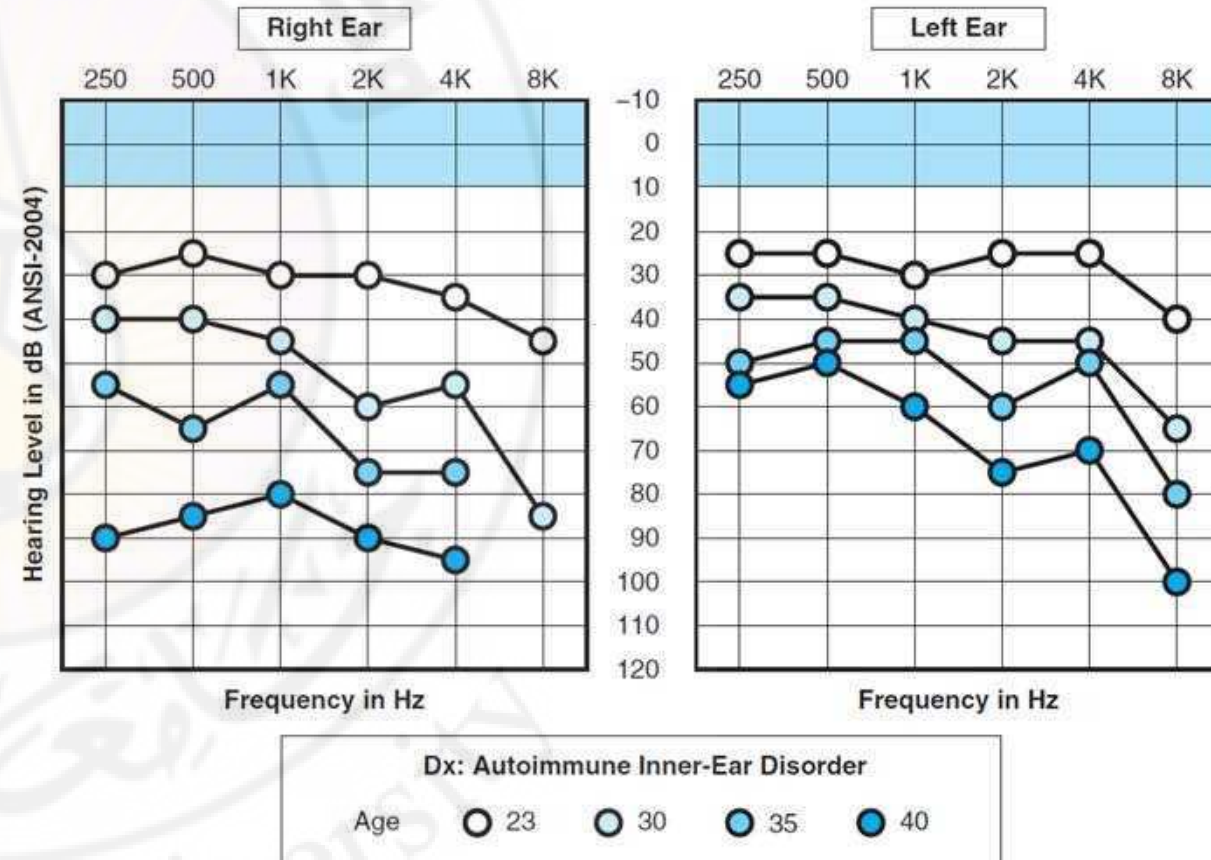
Clinical Audiology

Mohsen Ahadi, Ph.D.

Associate Professor of Audiology
Iran University of Medical Sciences
Rehabilitation Research Center
ahadi.m@iums.ac.ir

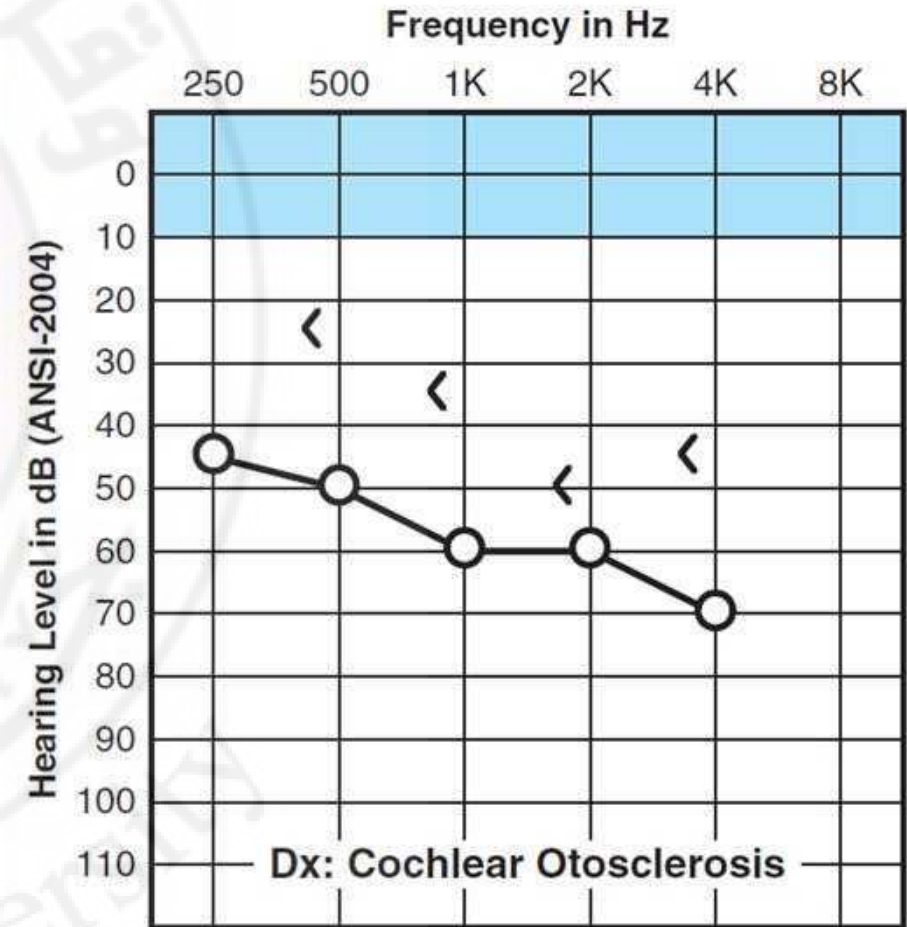
Autoimmune Inner-Ear Disease

- **Autoimmune inner-ear disease (AIED)** is an auditory disorder characterized by bilateral, asymmetric, progressive, sensorineural hearing sensitivity loss in patients who test positively for autoimmune disease.
- It tends to be diagnosed on the basis of exclusion of other causes, but it is increasingly attributed as the causative factor in progressive sensorineural hearing loss.
- The **asymmetry** and **progression** are the signature characteristics of the disorder.
- The hearing sensitivity loss may be responsive to immunosuppressive drugs, such as steroids.



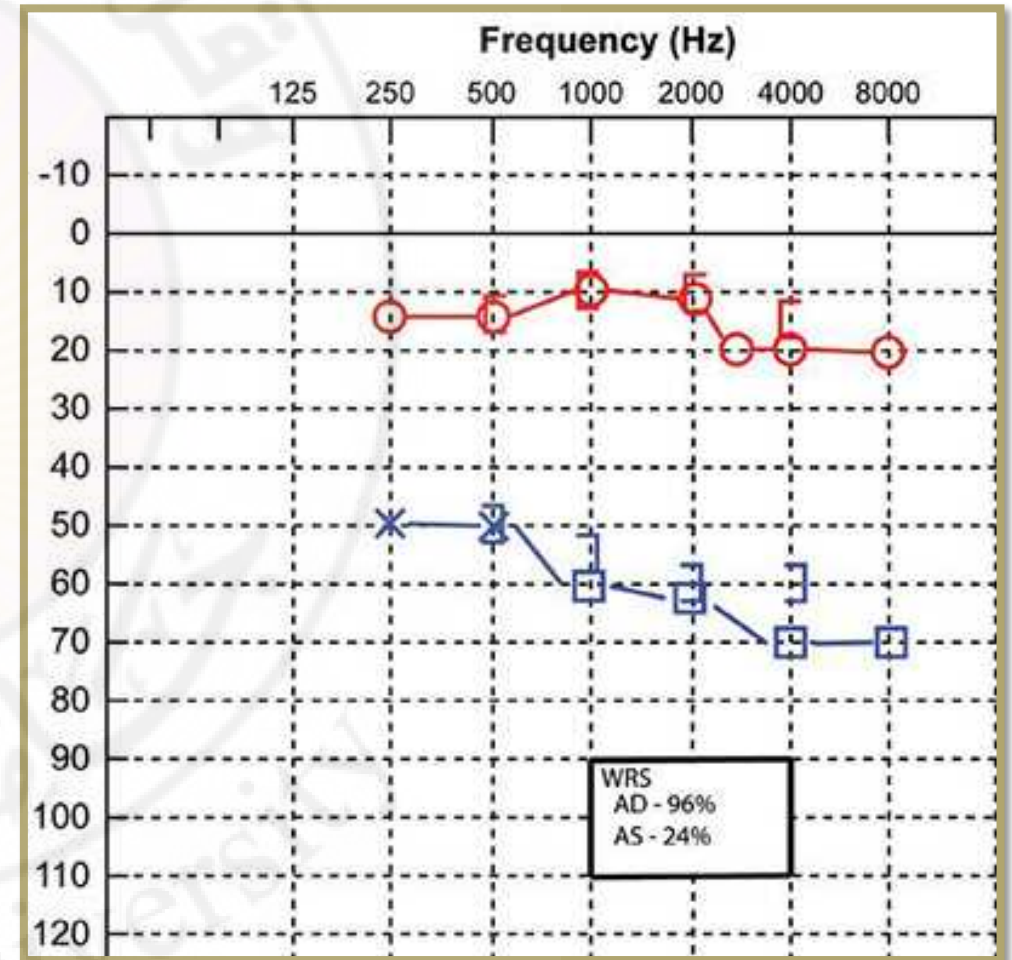
Cochlear Otosclerosis

- The same otosclerosis that can fix the stapes footplate into the oval window can occur within the cochlea and result in sensorineural hearing loss.
- Depending on the extent of cochlear involvement, a sensorineural hearing loss can occur.
- Although there is some debate about whether a sensorineural hearing loss can occur in isolation, it is certainly theoretically possible.
- Nevertheless, cochlear otosclerosis is commonly accompanied by fixation of the stapes, resulting in a **mixed hearing loss**.



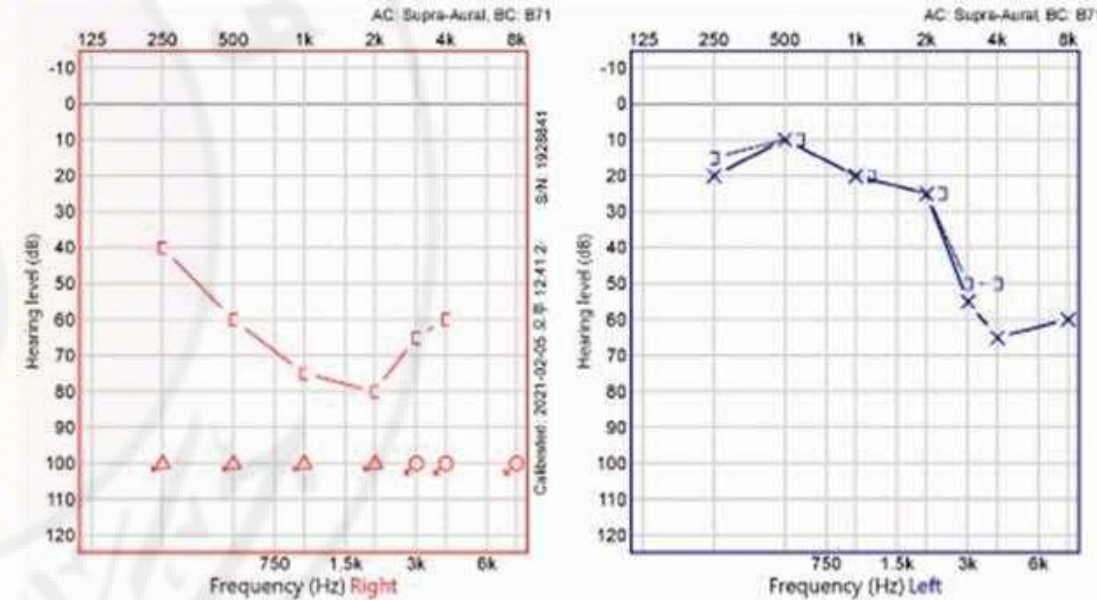
Idiopathic Sudden Sensorineural Hearing Loss

- **Idiopathic sudden hearing loss** is a term that is used to describe a sudden (usually described as developing inside of a 72-hour time frame), often **unilateral**, sensorineural hearing loss.
- Idiopathic sudden hearing loss is often noticed upon awakening and is usually accompanied by **tinnitus**.
- The extent of the sensorineural hearing loss ranges from mild to profound.
- there also may be a feeling of **pressure** or fullness in the affected ear(s).
- **Vertigo** may or may not be present.



Idiopathic SSNHL

- About 66,000 cases per year occur in the United States, with an incidence of somewhere between 5 and 77 per 100,000 and greater risk among **males** and the **elderly**.
- **Reoccurrence rates** have been reported anywhere from < 1% to ~ 47% of patients, and were 5.6% at 2 years and 10.3% in a recent series of cases.
- Partial or full recovery of hearing occurs in approximately 75% of patients.
- The term idiopathic is used because the cause is often unknown, but a wide variety of causes must be considered before arriving at this conclusion.



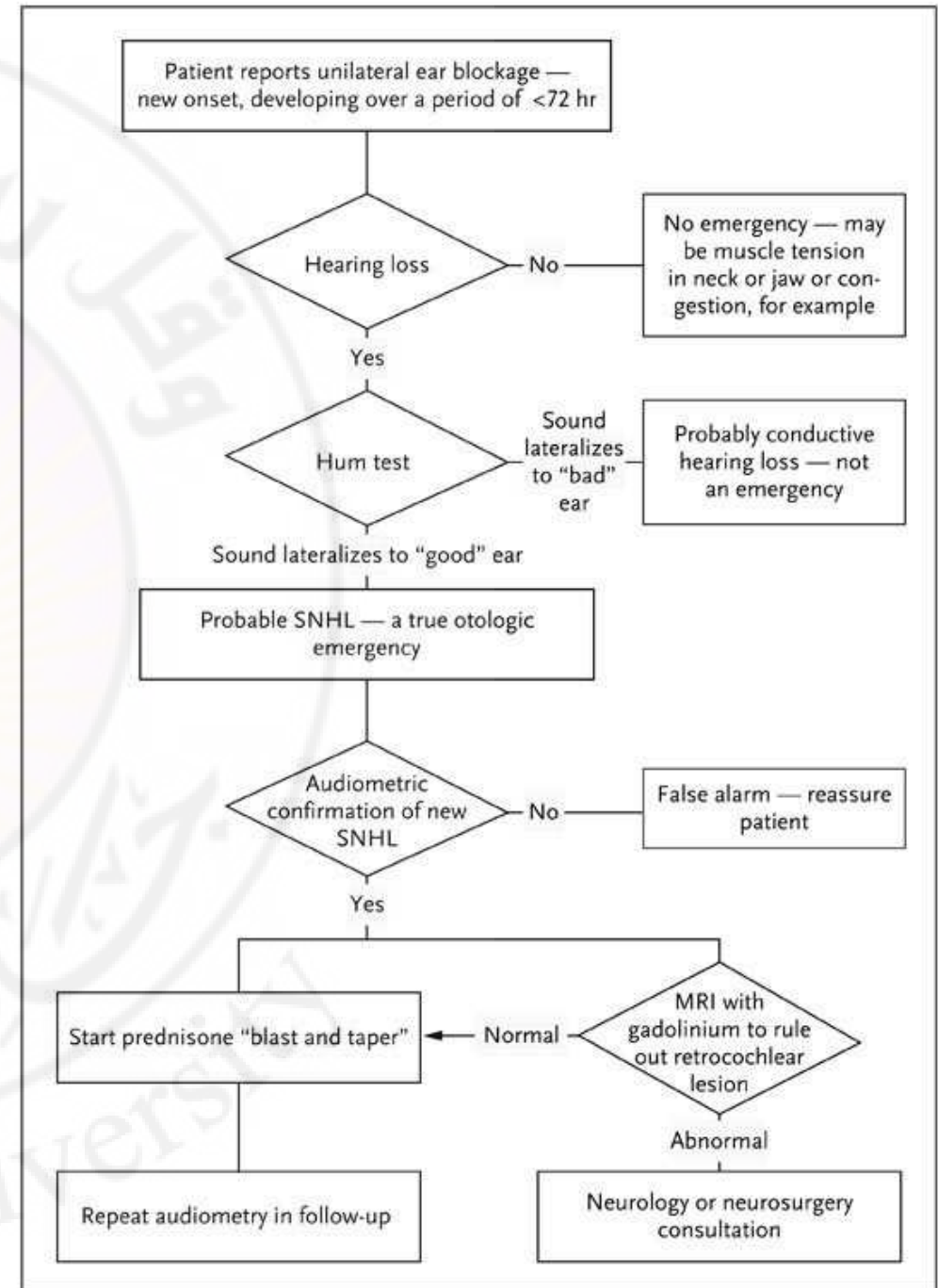
Idiopathic SSNHL: Etiology

- Viral infections
- Autoimmune disorders
- Perilymphatic fistulas
- Interference with the cochlear blood supply due to, for example, vascular disease
- Blood disorders (e.g., sickle cell)
- Embolisms, and vascular spasms
- Perilymph hypertension
- Rupturing of Reissner's membrane, Meniere's disease
- Neurological disorders (e.g., stroke, multiple sclerosis)
- Vestibular schwannoma, and other neoplasms
- Recent head trauma, ear surgery, acoustic trauma, and exposure to ototoxic drugs (including sildenafil and similar medications) should not be overlooked.
- Moreover, it is essential to know that sudden-onset hearing losses also can be caused by conductive disorders (e.g., impacted cerumen, acute Eustachian tube occlusion).



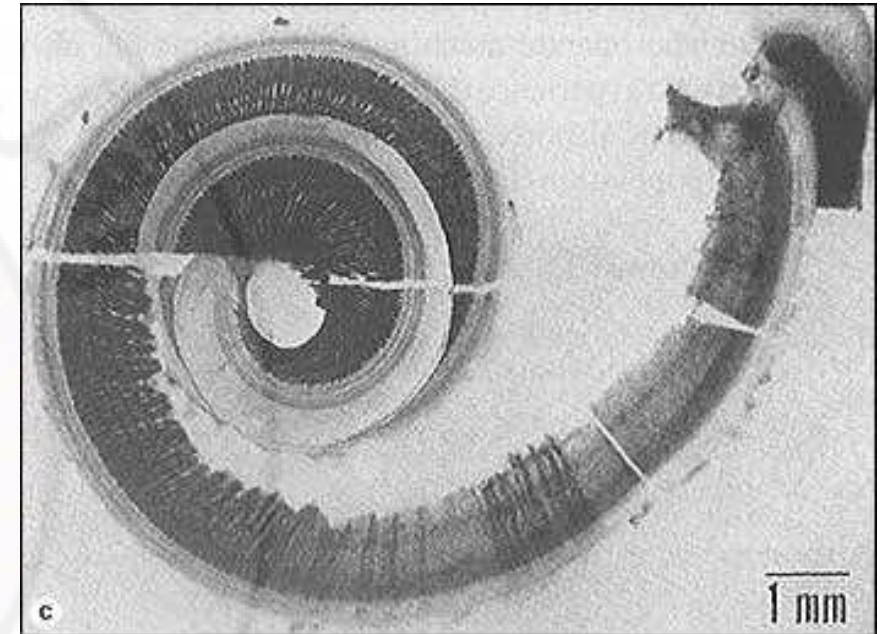
Idiopathic SSNHL

- **Audiological confirmation** of a sensorineural hearing loss is recommended within 14 days of when the symptoms started, and patients with sudden-onset sensorineural losses should be evaluated to rule-out retrocochlear pathology.
- In addition, appropriate evaluations or referrals are in order if there is anything that is not commonly associated with idiopathic sudden hearing loss. These include focal **neurological signs**, as well as sudden hearing losses that are **binaural**, have occurred before, or were preceded by fluctuating hearing loss.
- Once determined to be idiopathic, sudden sensorineural hearing loss is usually treated medically, most commonly with corticosteroids- **“salvage therapy”**.



Cochlear dead regions

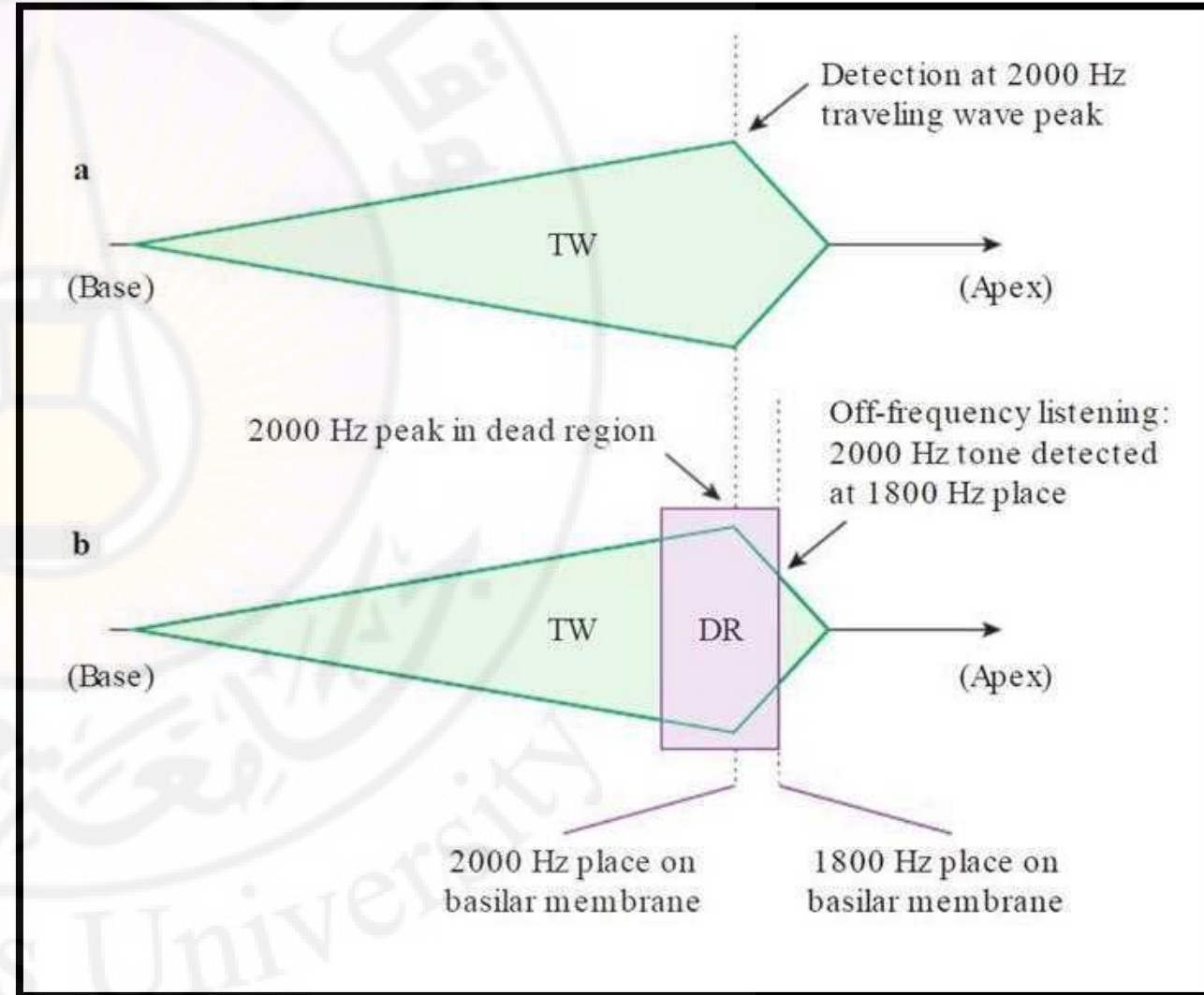
- **Dead regions** are locations along the cochlear spiral without functioning inner hair cells (IHCs) and/or auditory neurons.
- We would expect there to be no response to a test tone that activates such a dead region. Yet it is still possible for the patient to detect it due to **off-frequency listening**, which means that part of the vibration pattern caused by the test tone is picked up by functioning IHCs at nearby locations along the basilar membrane.



Cochlea from a 25-year-old man who had been exposed to gunshots. The dark lines show auditory neurons. There are no neurons coming from the basal part of the cochlea, indicating a dead region.

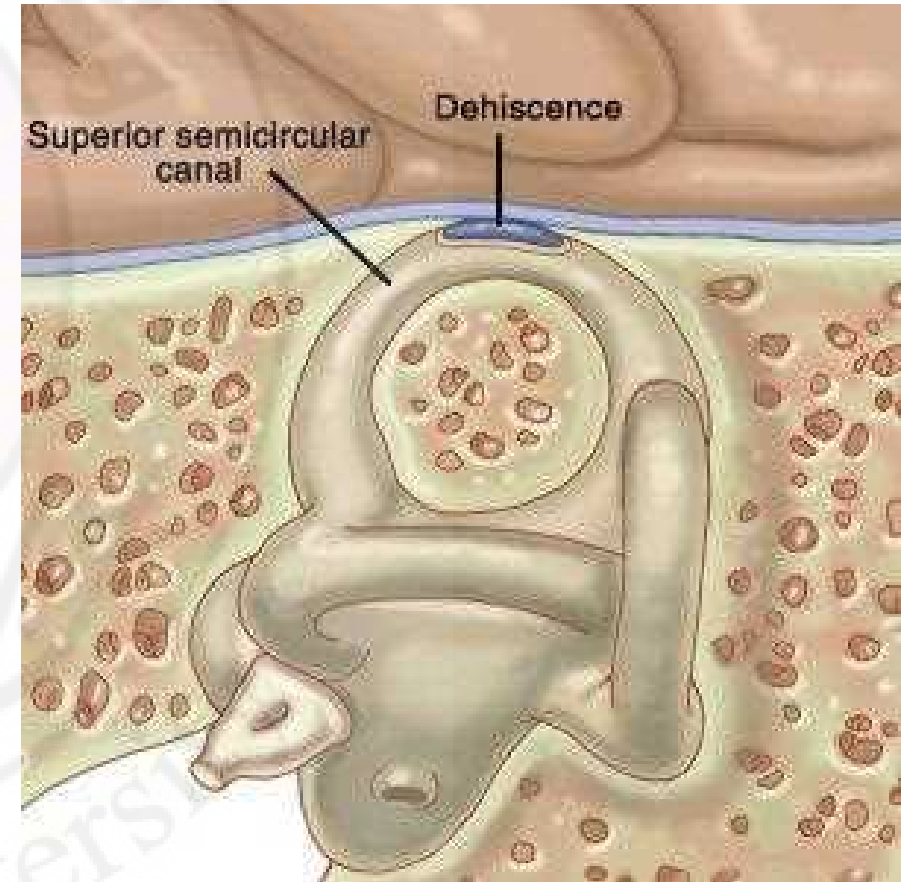
Off-frequency listening

- For example, a patient with a dead zone at the 2000 Hz place might still respond to a 2000 Hz test tone if part of its excitation pattern activates healthy IHCs at the 1800 Hz location.



Semicircular canal dehiscence (SCD)

- Superior canal dehiscence (SCD) is an inner ear disorder in which a section of the bone enclosing the superior semicircular canal is open or extremely thin (**third mobile window**), resulting in vestibular and or auditory symptoms that are caused by sound and/or pressure.
- The vestibular problems are the most disturbing symptoms of SCD. They include episodes of **vertigo, nystagmus, and oscillopsia** (an oscillating visual field) that are induced by sound and/or pressure events like nose blowing, straining to lift heavy objects, flying, etc.
- In addition, patients often experience ongoing problems with imbalance or unsteadiness as well tolerating motion.



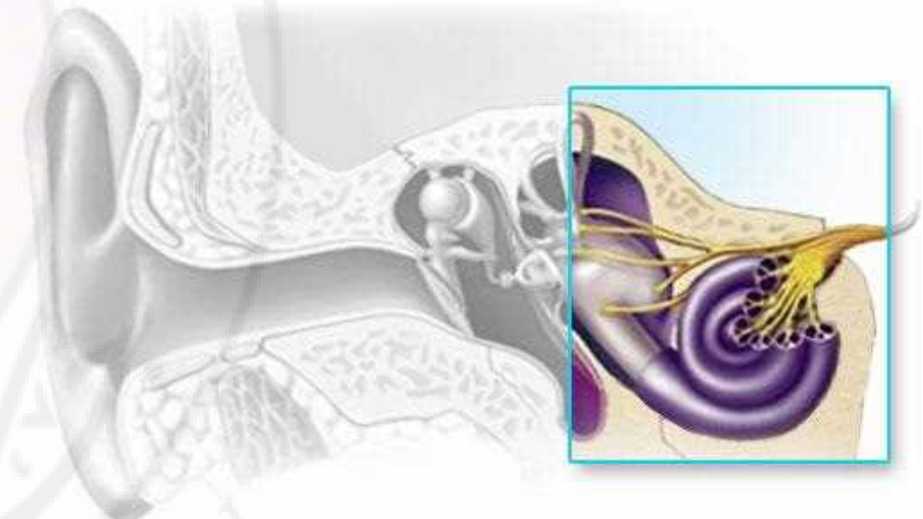
The logo of Damascus University is a circular emblem. It features a central yellow lamp with a flame, set against a purple background. The lamp is surrounded by a circular border containing Arabic text. Below the lamp, the words "Damascus University" are written in English. The entire logo is rendered in a light, faded grey color in the background of the slide.

Neural Hearing Disorders

- 8th nerve disorders
- Brainstem Disorders
- Temporal-Lobe Disorder

Retrocochlear Disorders

- **Retrocochlear disorders** involve the structures that are medial to the cochlea. In the broadest sense, this includes any problem that involves the vestibuloacoustic (eighth cranial) nerve or any part of the central auditory nervous system.
- In practice, however, retrocochlear usually refers to a disorder involving the **eighth cranial nerve and/or the cerebellopontine angle (CPA)**. Disorders are generally classified as **central** when the abnormality is in the central nervous system.
- Particularly when dealing with **brainstem disorders**, the term **extra-axial** is often used to describe a lesion that is located outside the brainstem (e.g., a CPA tumor that is pressing on the brainstem from without), and the term **intra-axial** is used when the lesion is actually inside the brainstem itself.

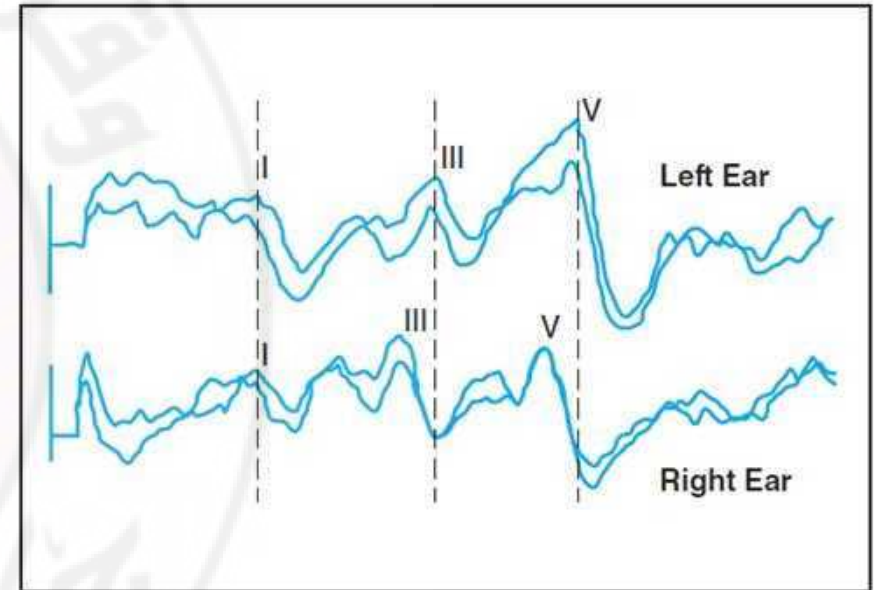


Neural (Retrocochlear) Disorders: Evaluative Goals

- Sometimes the patient history or physical examination will lead the otologist to suspect that a patient might have a **retrocochlear disorder**. In most cases, the physician will be concerned about the presence of a space-occupying lesion on the VIIIth nerve.
- This often occurs when a patient reports the presence of **unilateral hearing loss, unilateral tinnitus, unexplained dizziness**, or other **neurologic symptoms**.
- In such cases, the physician is interested in several aspects of the audiologic evaluation, including:
 - Is there a hearing loss, and what is the extent of it?
 - Is the loss unilateral or asymmetric?
 - Is speech understanding asymmetric or poorer than predicted from the hearing loss?
 - Are acoustic reflexes normal or elevated?
 - Is there other evidence of retrocochlear disorder?

Neural (Retrocochlear) Disorders: Test Strategies

- Immittance Audiometry (Tympanometry & ART)
- Pure-Tone Audiometry
- Speech Audiometry
- Otoacoustic Emissions
- Auditory Evoked Potentials



	Latency Wave V	Interwave Intervals		
		I-III	III-V	I-V
RE	5.9	1.9	2.0	3.9
LE	6.5	2.5	2.0	4.5

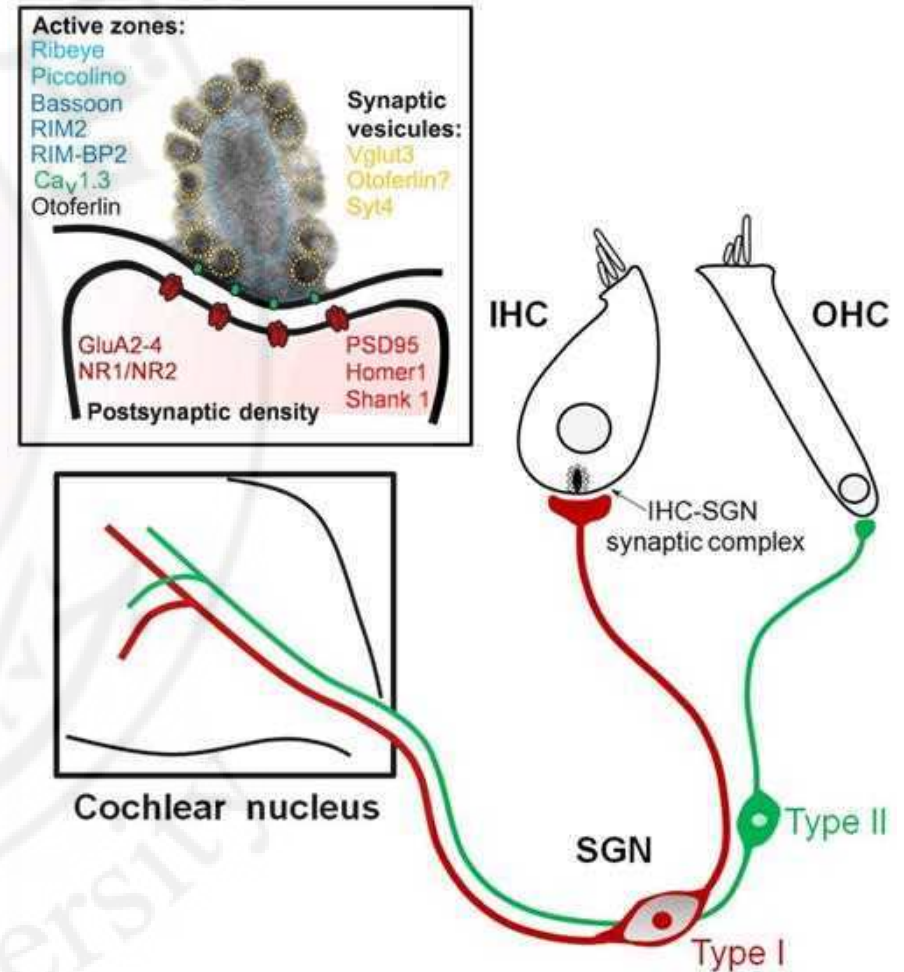
Click Level: 90 dB nHL

Neural Hearing Disorders

- Any disease or disorder process that affects the peripheral and central nervous system can, of course, result in auditory disorder if the auditory nervous system is involved.
- Neoplastic growths on the VIIIth nerve or in the auditory brainstem, cranial nerve neuritis, multiple sclerosis, and brain infarcts can all cause some form of auditory disorder.
- The nature of hearing impairment that accompanies central auditory nervous system disorder varies as a function of location of the disorder.
- A disorder of the **VIIIth nerve** is likely to result in a sensorineural hearing loss with poor speech understanding.
- The likelihood of a hearing sensitivity loss diminishes as the disorder becomes **more central**, so that a brainstem lesion is less likely than an VIIIth nerve lesion to cause a sensitivity loss, and a temporal lobe lesion is quite unlikely to cause such a loss. Similarly, **disorders of speech perception** become more subtle as the disorder becomes more central.

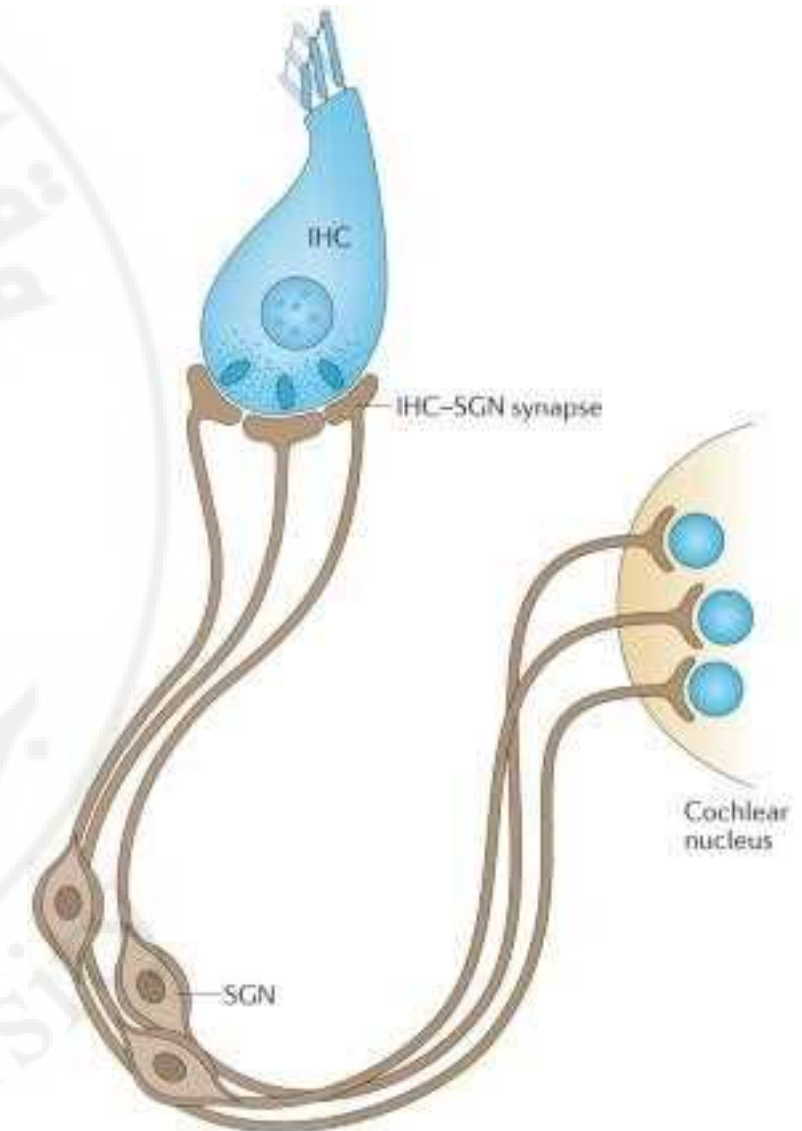
Auditory Neuropathy

- **Auditory neuropathy** is a term that is used to describe a disorder in the synchrony of neural activity of the VIIIth cranial nerve. It is operationally defined based on a constellation of clinical findings that suggest normal functioning of some cochlear structures and abnormal functioning of the VIIIth nerve and brainstem.
- In reality, the term auditory neuropathy as it is defined clinically probably describes two fairly different disorders, one sensory and the other neural:
 - **Auditory neuropathy of sensory origin: AN(S)**
 - **Auditory neuropathy of neural origin: AN(N)**



Auditory neuropathy of sensory origin: AN(S)

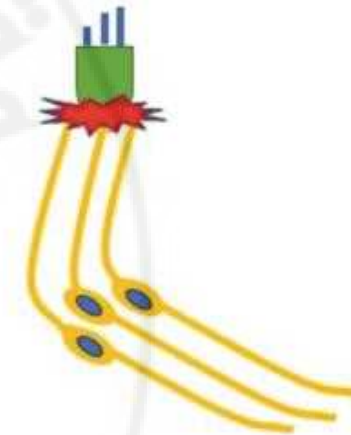
- The auditory neuropathy of sensory origin— AN(S)—is a sensory hearing disorder that represents a transduction problem, with the failure of the cochlea to transmit signals to the auditory nerve.
- The most likely origin of AN(S) is the **inner hair cells** of the cochlea with normally functioning outer hair cells serving little or no purpose.
- The hearing loss from AN(S) acts like any other sensitivity loss in terms of its influence on speech and language acquisition and its amenability to hearing aids and cochlear implants.



Auditory neuropathy of neural origin: AN(N)

- **Auditory neuropathy of neural origin—AN(N)**—was first described as a specific disorder of the auditory nerve that results in a loss of synchrony of neural firing. Because of the nature of the disorder, it is also referred to as **auditory dys-synchrony**.
- The cause of auditory neuropathy is often unknown, although it may be observed in cases of syndromic peripheral pathologies.
- The age of onset is usually before 10 years.
- **Hearing sensitivity loss** ranges from normal to profound and is most often flat or reverse-sloped in configuration.
- The hearing loss often **fluctuates** and may be **progressive**. **Speech perception** is often substantially poorer than what would be expected from the audiogram.

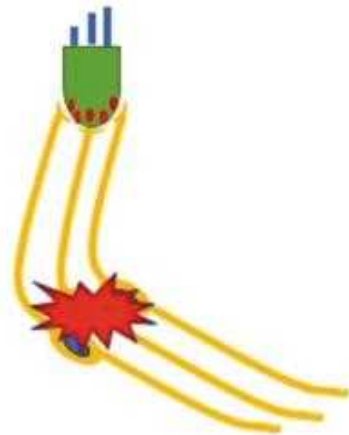
Pre-synaptic type AN



Genetic Hearing Loss

OTOF
DIAPH3
CACNA1D
etc.

Post-synaptic type AN



Genetic Hearing Loss

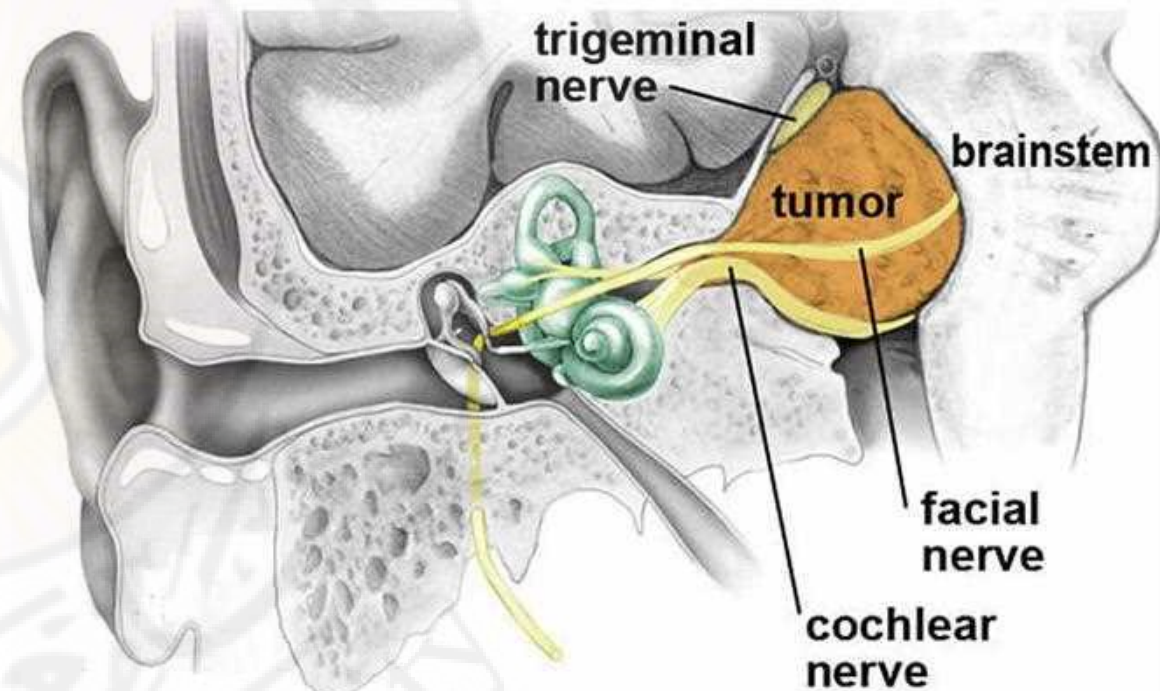
OPA1
MPZ
PMP22
ATP1A3
etc.

Demyelinating Disease

CMT
MS
CIDP

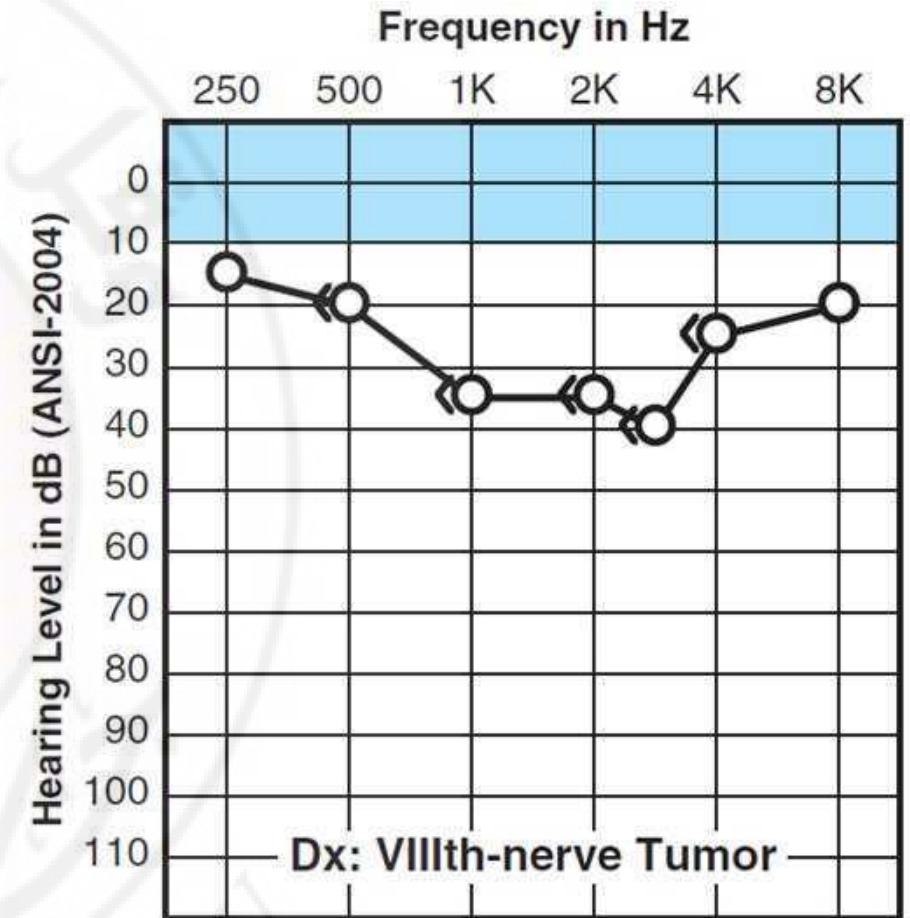
VIIIth Nerve Tumors and Disorders

- The most common neoplastic growth affecting the auditory nerve is called a **cochleovestibular schwannoma**.
- The more **generic terms** acoustic tumor or acoustic neuroma are typically referring to a cochleovestibular schwannoma. Other terms used to describe this tumor are acoustic neurinoma and acoustic neurilemoma.
- A cochleovestibular schwannoma is a **benign, encapsulated** tumor composed of **Schwann cells** that arises from the VIIIth cranial nerve.
- Schwann cells serve to produce and maintain the myelin that ensheathes the **axons** of the VIIIth nerve.

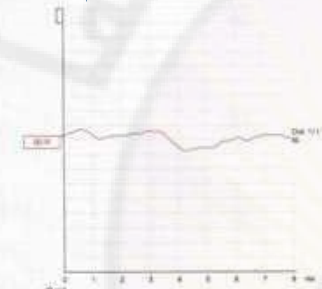


VIIIth Nerve Tumors

- Acoustic tumors are unilateral and most often arise from the vestibular branch of the VIIIth nerve. Thus, they are sometimes referred to as **vestibular schwannomas**.
- The effects of a cochleovestibular schwannoma depend on its **size, location**, and the **extent of the pressure** it places on the VIIIth nerve and brainstem.
- Auditory symptoms may include **tinnitus**, **hearing loss**, and **unsteadiness**.
- Depending on the extent of the tumor's impact, it may cause headache, motor incoordination from cerebellar involvement, and involvement of adjacent cranial nerves (facial weakness & diplopia).



Among the most common symptoms of cochleovestibular schwannoma are unilateral tinnitus and unilateral hearing loss. The hearing loss varies in degree depending on the location and size of the tumor.

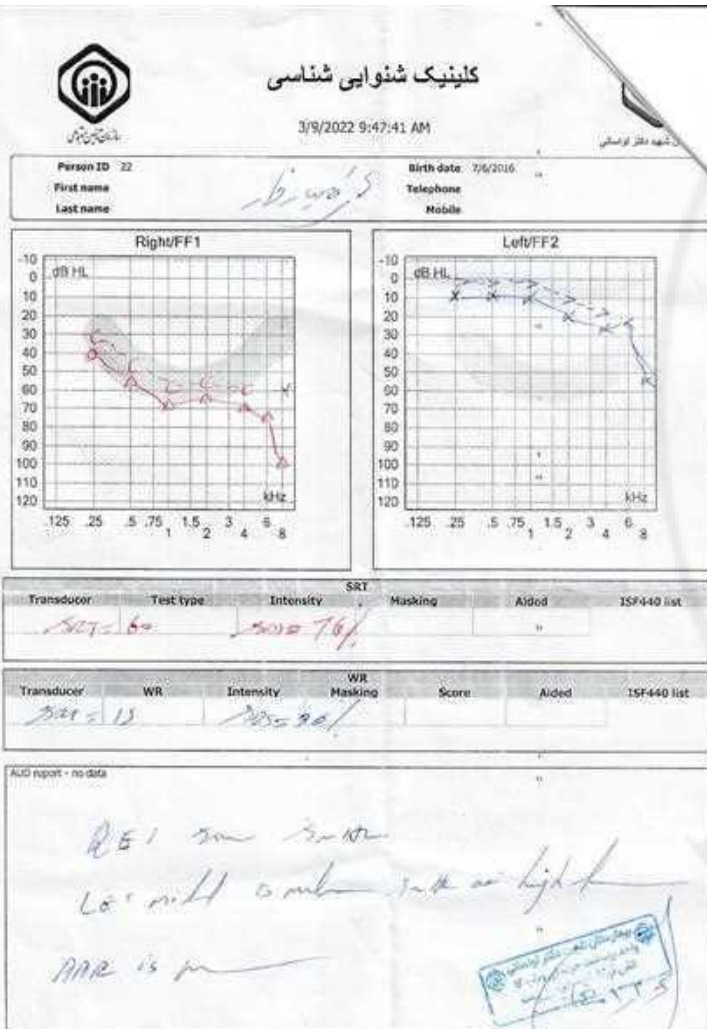


بیت حقوق انسانی

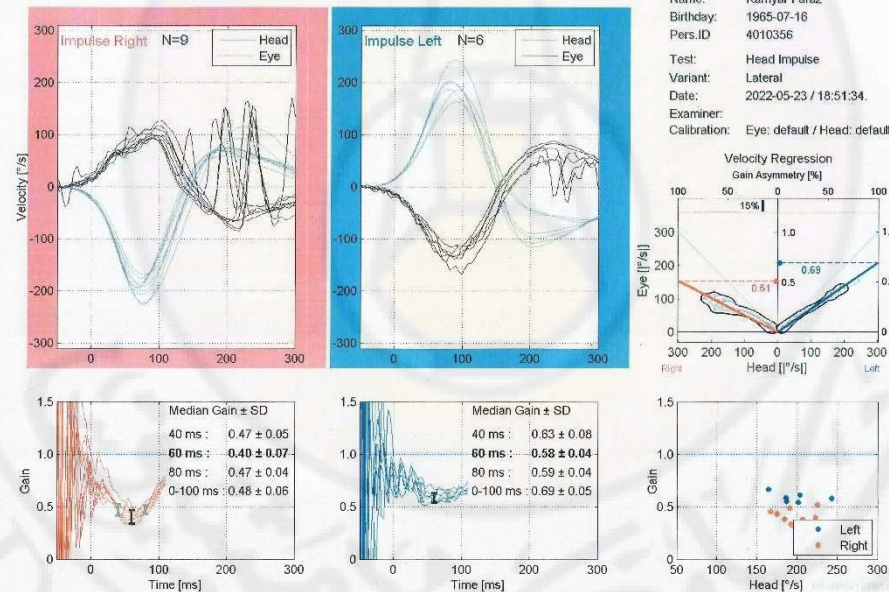
مركز تخصصي اور ادبي قلمخانہ
دکتر سعید علی کاشفی
متخصص اور پروفیسر سولہ گزائی
دانشیار و ریاست علمی دانشگاه علوم پزشکی ایران
تلفون و فکس ۱۰۸۲۲۷

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Case Example



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Unilateral Hearing Loss & Vertigo

ام آر آی ۱/۵ تسلا - سی تی اسکن مولتی اسلایس
آرمانگاه پاتوبیولوژی و تشخیص نسی - سونوگرافی (سی - دایر)
رادیولوژی دیجیتال - ماموگرافی دیجیتال - نواری معز
نواری معص و عسله - سنجش تراکم استخوان - داپلر معز

نام بیمار : آقای کامیار فوار
کد پذیرش : 41021337
تاریخ : 1401/02/15
سن بیمار : 56 سال
همکار گرامی سرکار خانم اجناب آقای دکتر : رضا هدایتی

IAC MRI with and without kontras:

There is a T^* (mm low T) high T^* homogenous intensity enhancing mass with intracanalicular component is seen in right side suggestive for acoustic schwannoma.

The cochlea and semicircular canals appear normal. The mastoid air cells are clear and pneumatized. The tympanic cavity and EAC are normal.

The CP angle areas show normal configuration on each side.

The brain stem also shows normal configuration and signal.

Best Regards
M.SALAH M.D.
Radiologist

درمانگاه تخصصی اعصاب و نورولوژی
دکتر مناصحی
فلوشیپ تشخیص و اپنر و شنوایی
بورد تخصصی رادیولوژی - نظام پزشکی ۱۳۱۹۵

سرپرستی شمالی، مانترا از آپادانا، کوچه ستاکوز، ساختمان ام آر آی، تلفن: ۰۵۰۲۳۲۲۰۰۵۰، فاکس: ۸۸۷۶۱۳۰۷، E-mail: info@sohmr.com

VIIIth Nerve Tumors and Disorders

- One other important form of schwannoma is neurofibromatosis. This tumor disorder has two distinct types.
- **Neurofibromatosis 1 (NF-1)**, also known as **von Recklinghausen's** disease, is an autosomal dominant disease characterized by café-au-lait spots and multiple cutaneous tumors, with associated optic gliomas, peripheral and spinal neurofibromas, and, rarely, acoustic neuromas.
- In contrast, **Neurofibromatosis 2 (NF-2)** is characterized by **bilateral** cochleovestibular schwannomas. The schwannomas are faster growing and more virulent than the unilateral type. This is also an autosomal dominant disease and is associated with other intracranial tumors. Hearing loss in NF-2 is bilateral and often progresses more rapidly.



Diagnostic criteria for neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2): NIH (1991)

NF1 is revealed by the presence of ≥ 2 of the following characteristics:

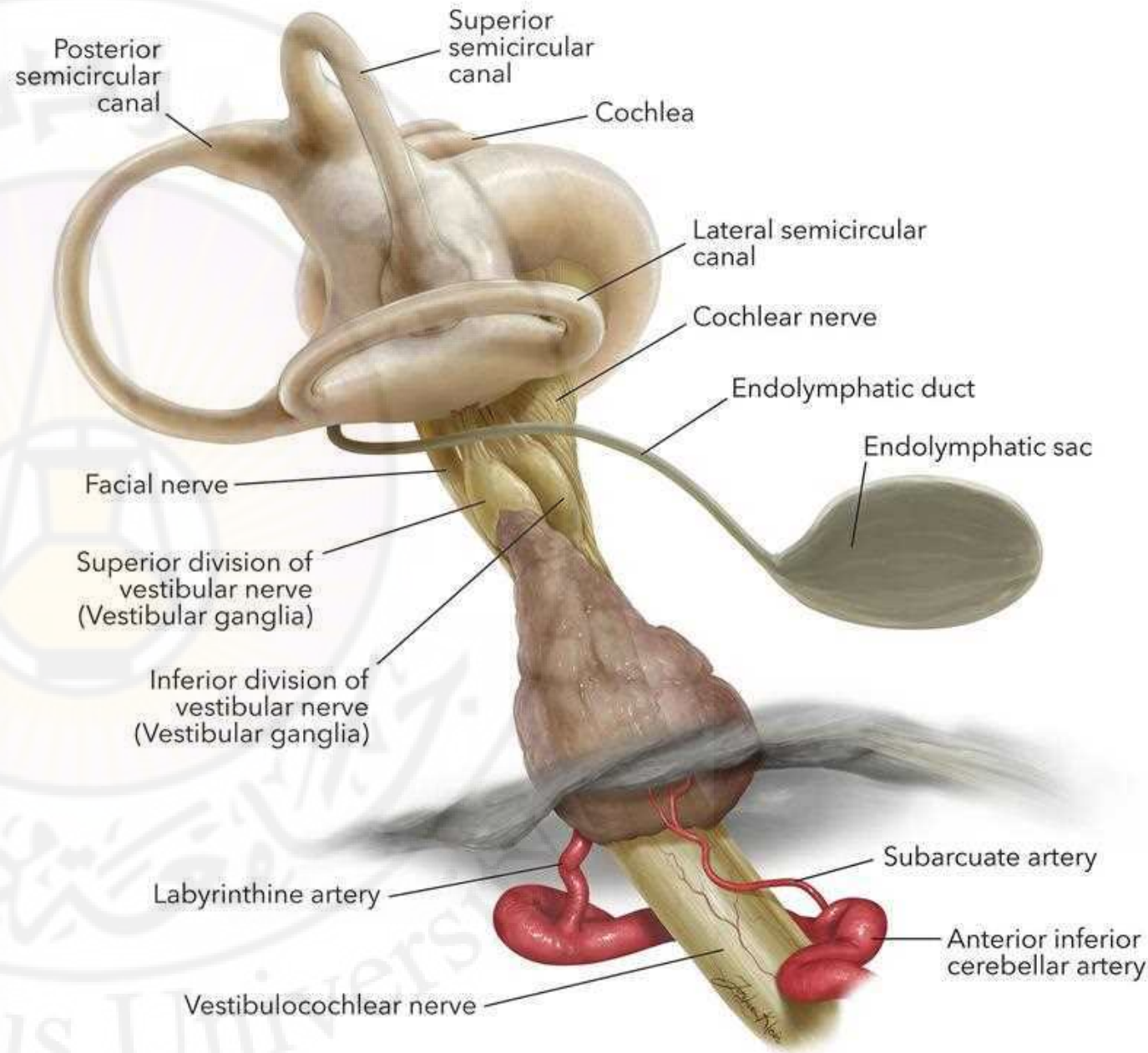
- ≥ 6 café-au-lait spots that are
 - > 5 mm in diameter before puberty
 - ≥ 6 café-au-lait spots that are
 - 5 mm in diameter before puberty
 - 15 mm in diameter after puberty
- 2 neurofibromas of any type, even 1 plexiform fibroma
- Freckling in the axillary (armpit) region or inguinal (groin) region
- Optic glioma
- 2 Lisch nodules (pigmented iris hamartomas)
- Having a parent/sibling/child with NF1 (based on the above criteria)

NF2 is revealed by the presence of any of the following characteristics:

- Bilateral eighth-nerve tumors
 - Having a parent/sibling/child with NF2 *and either a unilateral eighth-nerve tumor*
- or any one of the following:*
- Neurofibroma
 - Meningioma
 - Glioma
 - Schwannoma
 - Posterior capsular cataract or opacity at any early age

VIIIth Nerve Tumors and Disorders

- In addition to cochleovestibular schwannoma, a number of other types of **tumors, cysts, and aneurysms** can affect the VIIIth nerve and the cerebellopontine angle, where the VIIIth nerve enters the brainstem.
- These other neoplastic growths, such as **lipoma** and **meningioma**, occur more rarely than cochleovestibular schwannoma. The effect of these various forms of tumor on hearing is usually indistinguishable.





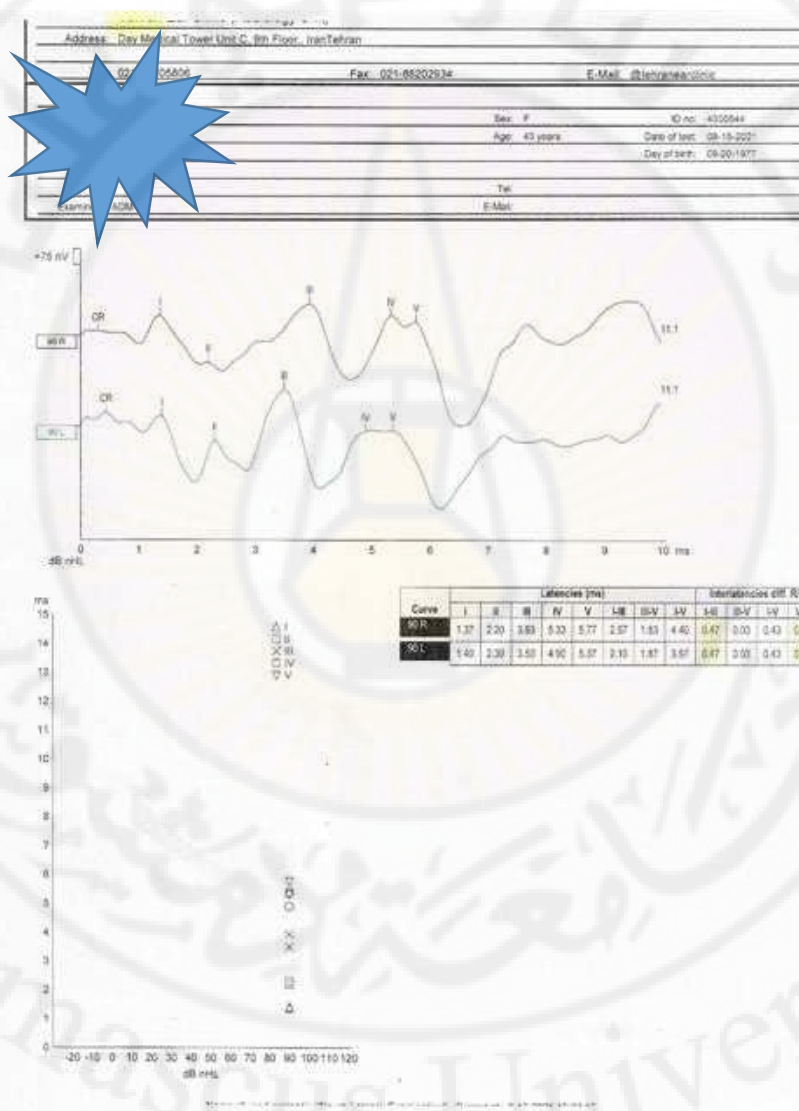
Speech	SDT		SRT		WRS / SRS 1				WRS / SRS 2				MCL	UCL
	dB HL	[m]	dB HL	[m]	%	dB HL	[m]	S/N	%	dB HL	[m]	S/N	dB HL	dB HL
Right			10		100	45							45	
Left			10		100	45							45	
Bin														
Note	1 Live, AC						2							
Aided														
Note	1							2						

Legend		
L	R	Masked
		AC
		BC
		SF
		MCL
		UCL
		NR

PTA AC: 300, 1k, 2k
BC: 500, 1k, 2k

Signed by:

بخاری سہ ماہی و اشاعت پمپستان دی
پمپستان سہ ماہی - ایڈیٹوریل
پمپستان
E. Sajadi-Audiologist



مرکز تصویرنگاری بیمارستان آراد

سی تی انژیوگرافی قلب و تمام بدن با دستگاه موثقی اسلایس 64 - ام آر آی - سی تی اسکن اسپیرال - سونوگرافی (کالر داپلر)
 رگرافی DSA - ماموگرافی - پانورکس - رادیولوژی تشخیصی

تاریخ: 1400/05/17

ام آر ای

ہمارے محترم سرکارِ حائِم اِجْتِہاد اُمّی : صدیق محمد

BRAIN WITH AND WITHOUT CONTRAST MRI/MRS/PERFUSION:

-Technique: Utilizing Siemens Magnetom Aera 1.5 tesla MRI unit without contrast administration multiplanar cross section of different pulse sequences were taken.

-54°43°38mm extraaxial high signal heterogenous mass in right CP-angle with pressure on lateral aspect of right cerebellar hemisphere and right side of middle cerebellar peduncle with internal hemorrhagic component with faint enhancement in post contrast images is detected.

-Another smaller infratentorial lesion (8*7mm) in contact with caudal aspect of left tentorium is noted.

- In MRperfusion study, RCBV in right lesion 2.5-4 with rapid return to base line categorized as high RCBV.

-In MRSpectroscopy with long and short TE, increase in choline, choline/Cr ratio is about 1.5 with obvious alanine peak.

-These findings are suggestive of extraaxial atypical meningioma is the first diagnosis.

SA-TGH

Dr.Taheri
Time: 1400/05/21 09:19

Dr. Adibi

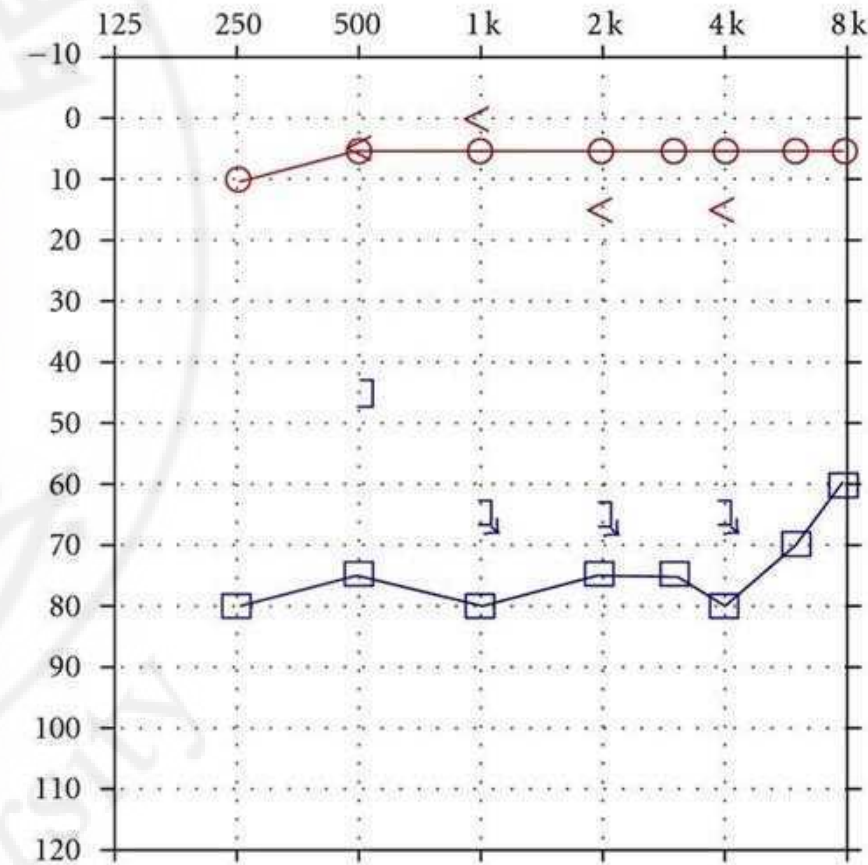
Dr. Taghilo

Dr. Tahmasebi

دکتر آیدین تاشی
پروفیسر، جامعہ اسلامیہ، کراچی
11/1/2017

VIIIth nerve Neuritis

- Not unlike any cranial nerve, the VIIIth nerve can develop **neuritis**, or inflammation of the nerve.
- Although rare, acute cochlear neuritis can occur as a result of a direct viral attack on the cochlear portion of the nerve. This results in degeneration of the cochlear neurons in the ear. Hearing loss is sensorineural and often sudden and severe. It is accompanied by poorer speech understanding than would be expected from the degree of hearing loss.
- One specific form of this disease occurs as a result of **syphilis**. **Meningo-neuro-labyrinthitis** is an inflammation of the membranous labyrinth and VIIIth nerve that occurs as a predominant lesion in early congenital syphilis or in acute attacks of secondary and tertiary syphilis.



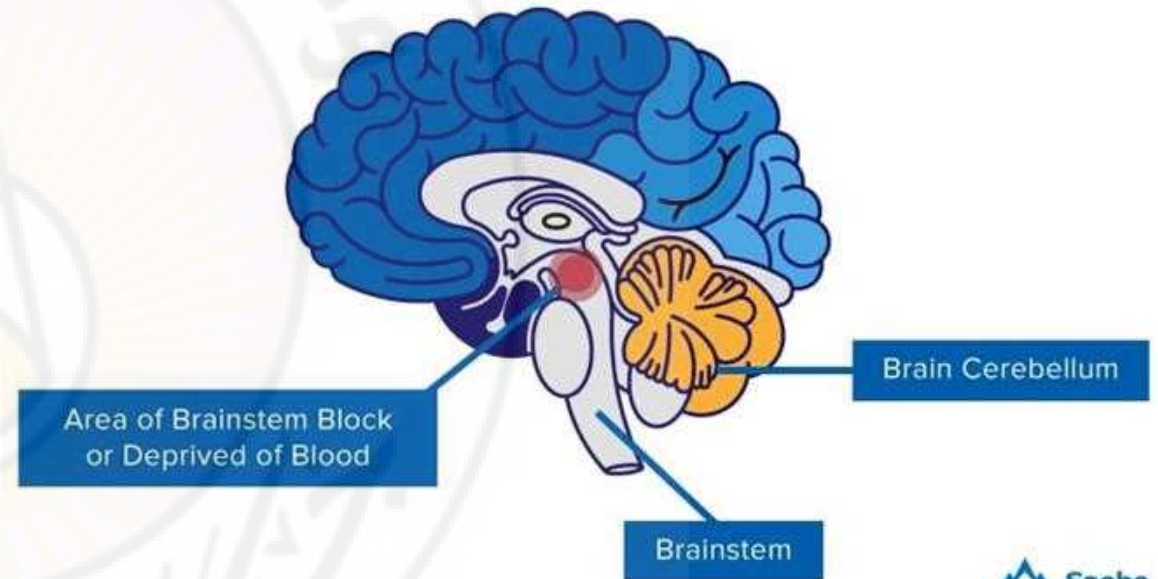
VIIIth nerve & Diabetes mellitus

- **Diabetes mellitus** is a metabolic disorder caused by a deficiency of insulin, with chronic complications including neuropathy and generalized degenerative changes in blood vessels.
- Sensorineural hearing loss was more common among diabetics than age-matched nondiabetic patients, and that worsening of the diabetes was associated with increasing hearing loss.
- **Neuropathies** can involve the central, peripheral, and autonomic nervous systems. When neuropathy from diabetes affects the auditory system, it usually results in vestibular disorder and hearing loss consistent with retrocochlear disorder.



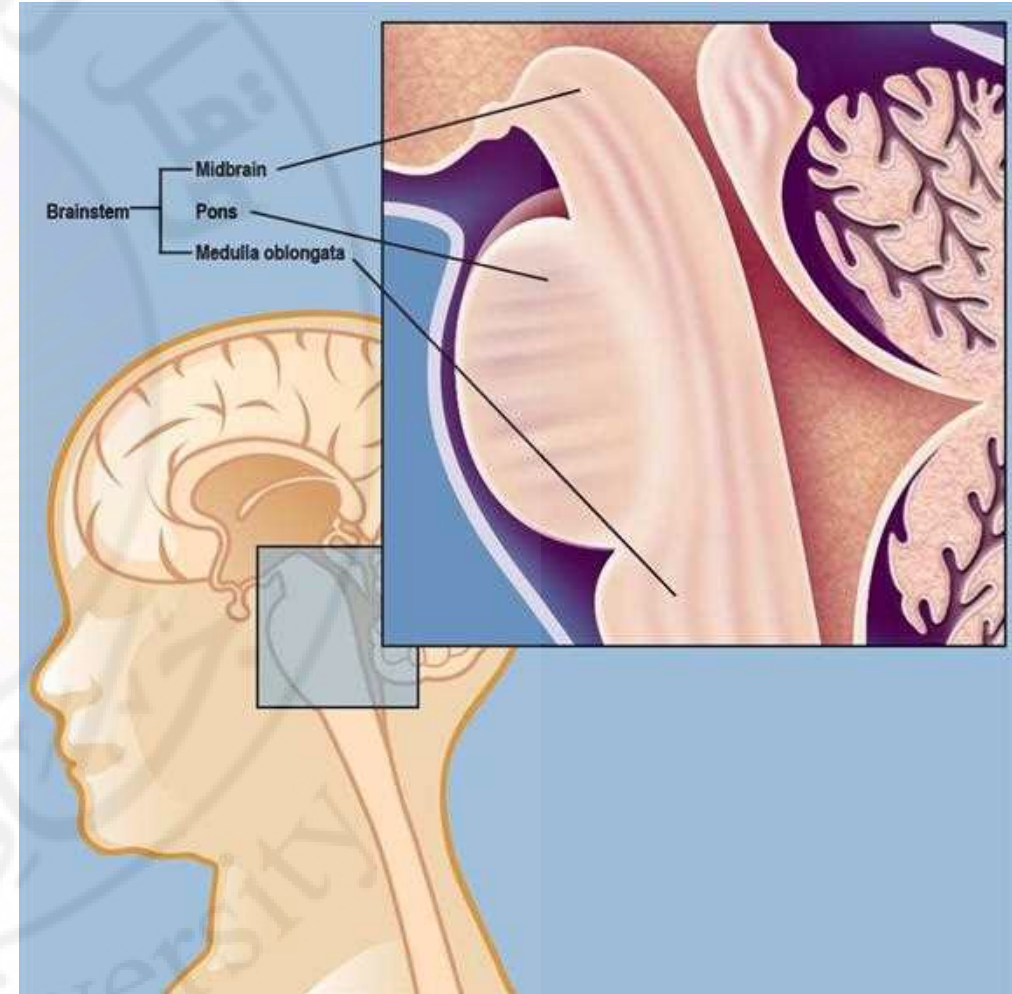
Brainstem Disorders

- Brainstem disorders that affect the auditory system include infarcts, gliomas, and multiple sclerosis.
- **Brainstem infarcts** are localized areas of **ischemia** produced by interruption of the blood supply.
- Auditory disorder varies depending on the site and extent of the disorder.
- Two syndromes related to vascular lesions that include hearing loss are **inferior pontine syndrome** and **lateral inferior pontine syndrome**.



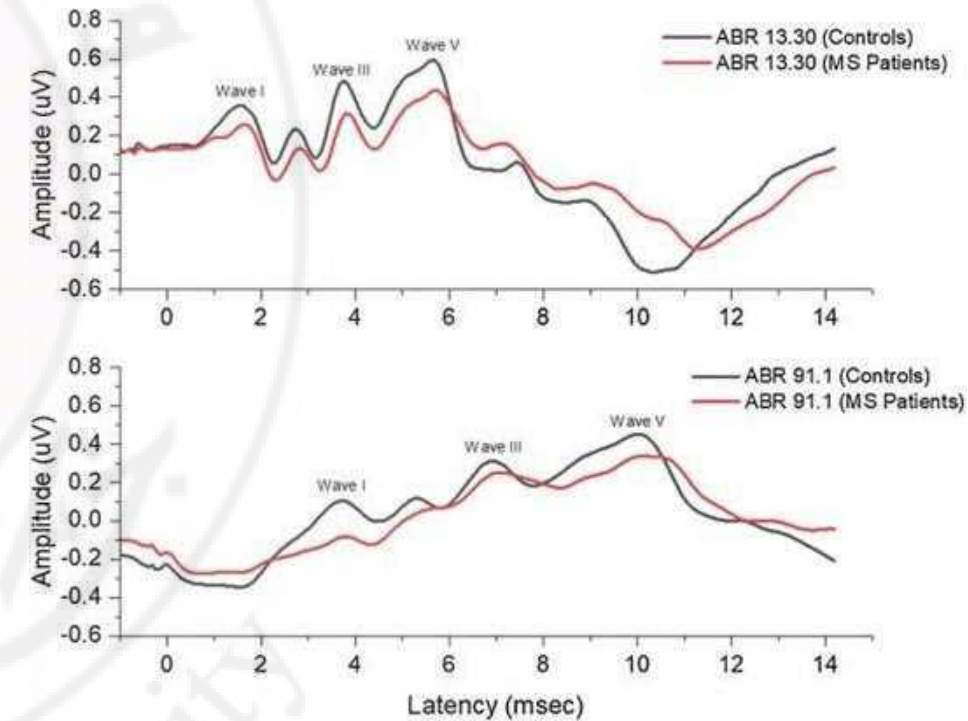
Brainstem Disorders

- A **glioma** is a tumor composed of neuroglia, or supporting cells of the brain.
- It comes in various forms, depending on the types of cells involved, including astrocytomas, ependymomas, glioblastomas, and medulloblastomas.
- Any of these can affect the auditory pathways of the brainstem, resulting in various forms of retrocochlear hearing disorder, including hearing sensitivity loss and speech perception deficits.



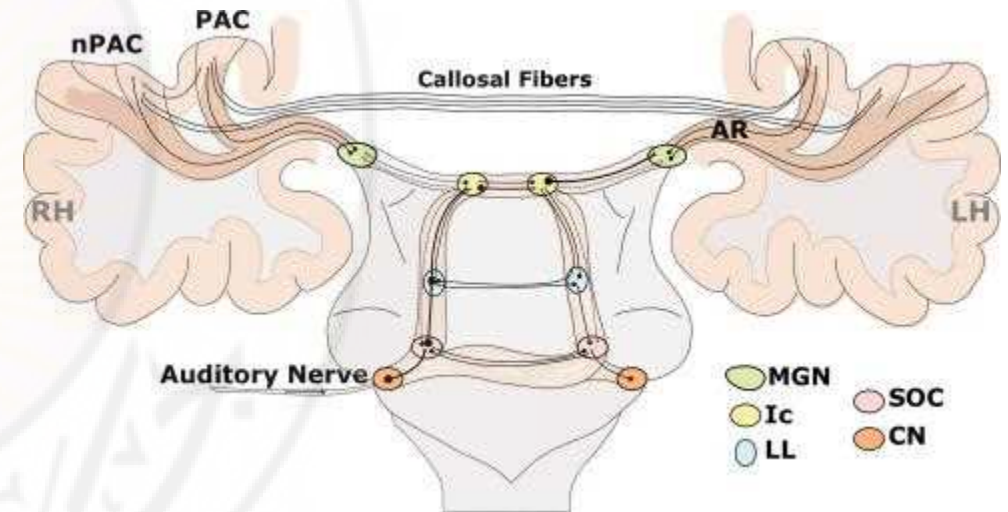
Brainstem Disorders

- **Multiple sclerosis** is a demyelinating disease. It is caused by an autoimmune reaction of the nervous system that results in small scattered areas of demyelination and the development of demyelinated plaques.
- If the demyelination process affects structures of the auditory nervous system, hearing disorder can result.
- There is no characteristic hearing sensitivity loss that emerges as a consequence of the disorder, although all possible configurations have been described.
- **Speech perception deficits** are not uncommon in patients with multiple sclerosis.



Temporal-Lobe Disorder

- **Cerebrovascular accident, or stroke**, is caused by an interruption of blood supply to the brain due to aneurysm, embolism, or clot.
- This results in sudden loss of function related to the damaged portion of the brain. When this occurs in the temporal lobe, audition may be affected, although more typically, **receptive language processing** is affected while hearing perception is relatively spared.
- Indeed, hearing ability is seldom impaired except in the case of **bilateral temporal lobe lesions**. In such cases, **“cortical deafness”** can occur, resulting in symptoms that resemble **auditory agnosia**.



Hidden Hearing Loss (Obscure Auditory Dysfunction)

- **Hidden hearing loss (HHL)**, also known as **obscure auditory dysfunction (OAD)** and **King-Kopetzky syndrome**, describes complaints about hearing difficulty, particularly speech recognition problems in noisy and other difficult listening situations, in spite of essentially normal hearing thresholds and the apparent absence of any known disorders that could explain the problem.
- These complaints may reflect one or a combination of disorders of the peripheral ear, auditory nerve, and/or central auditory system that might be due to the impact of synaptopathy, noise exposure, aging, or frank pathologies, as well as cognitive issues, the effects of changing acoustical environments or communicative demands, linguistic factors, and psychological factors.



QUESTIONS?

