



Pediatric Audiology

Mohsen Ahadi, Ph.D.

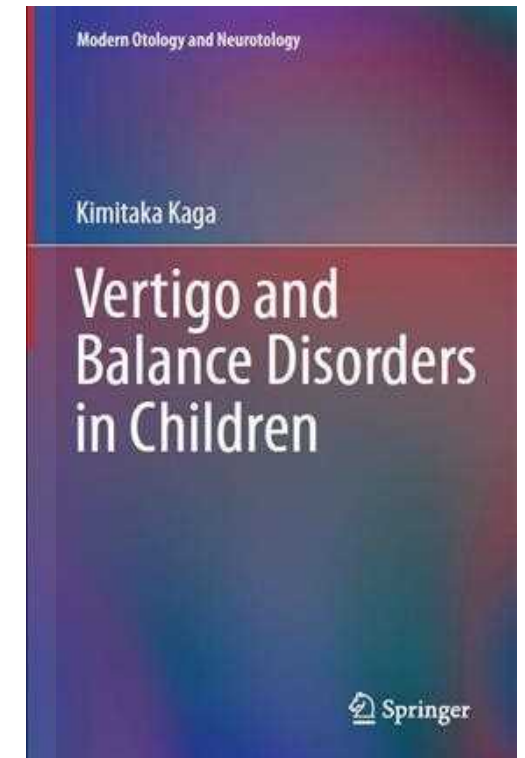
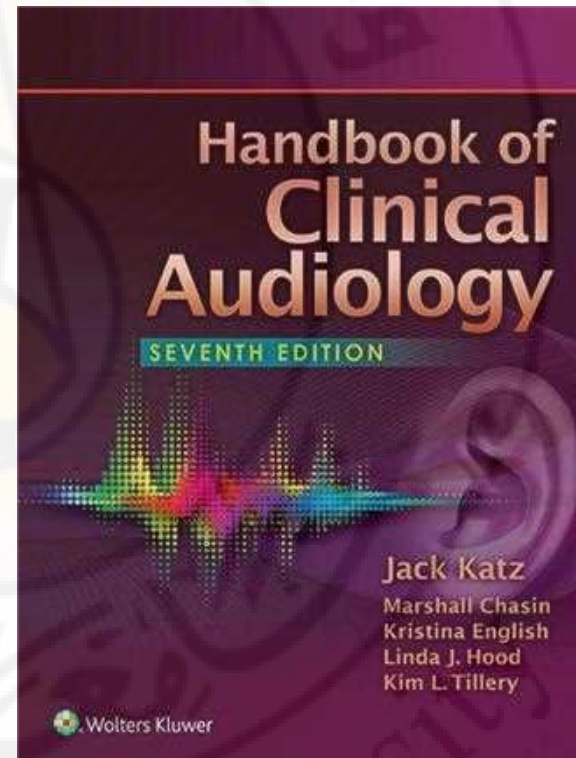
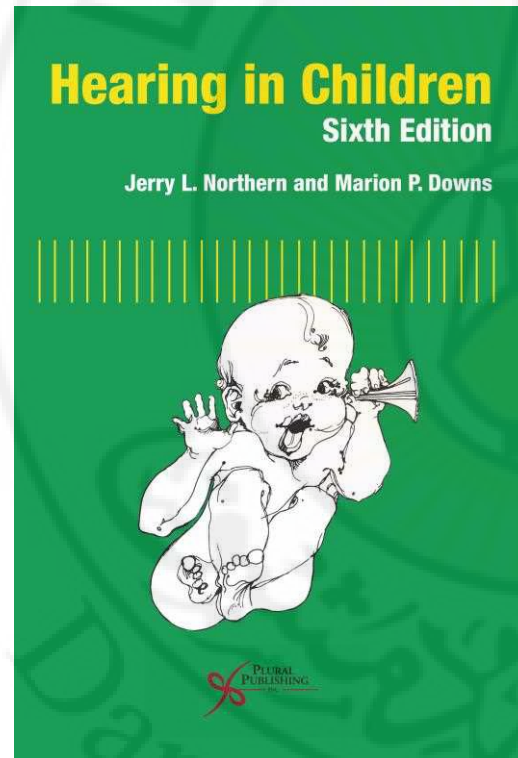
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Main Topics

- Hearing in children
- Hearing loss in children
- Auditory and Speech-Language development
- Hearing disorders in children
- Early intervention and Newborn Hearing Screening
- Behavioral hearing tests in infants and children
- Evaluation of hearing in children with special needs
- Physiologic Hearing Tests
- Vestibular function tests in infants and children



References



E. references

audiologyonline.com

asha.org

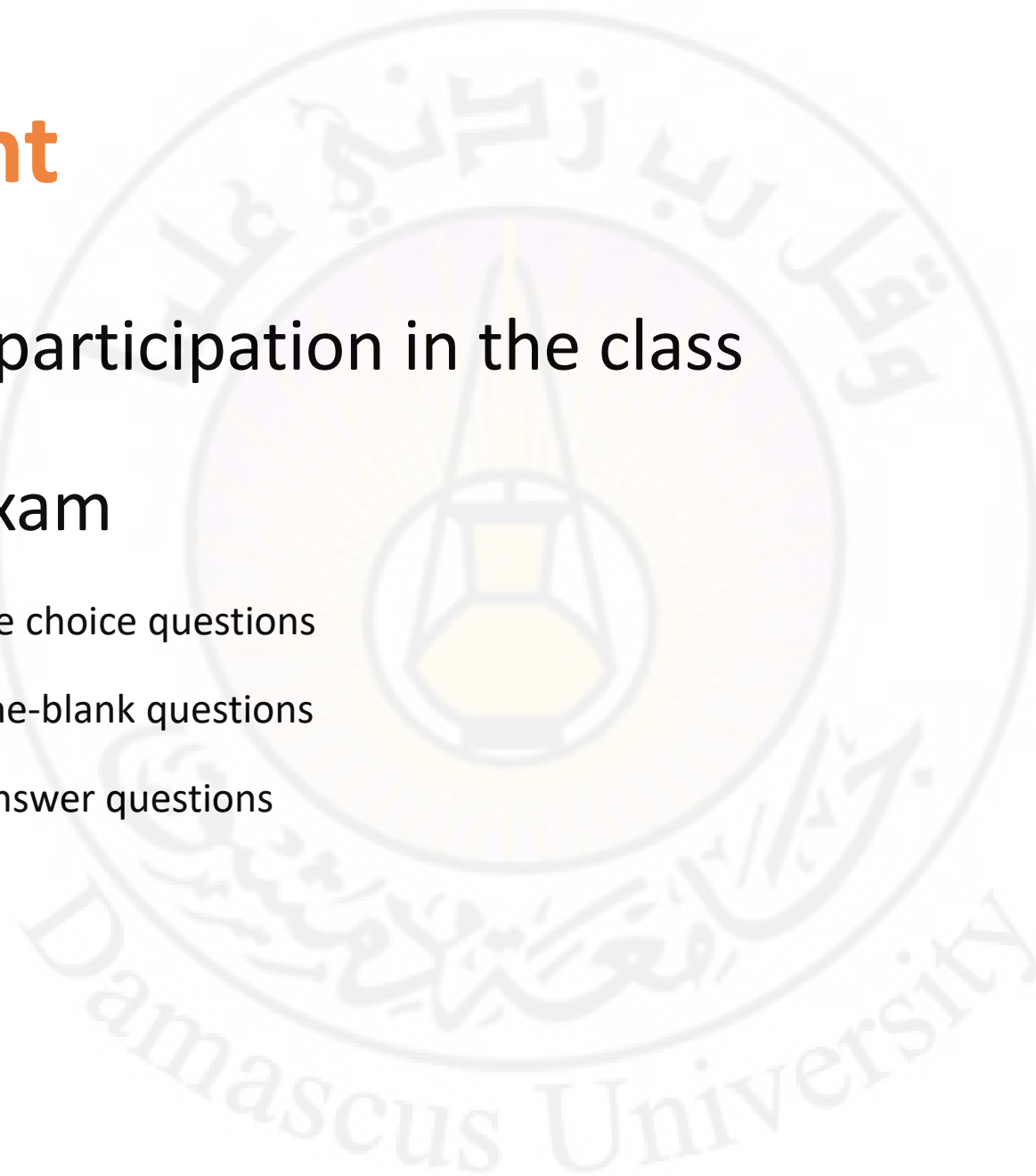
audiology.org

thebsa.org.uk



Assessment

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



1. Hearing in children

Hearing loss the most common birth defect

- Approximately 12,000 new babies with hearing loss are identified every year, according to the National Institute on Deafness and Other Communication Disorders.
- In addition, estimates are that another 4,000 to 6,000 infants and young children between birth and 3 years of age who had passed the newborn screening test acquire **late-onset hearing loss**.
- Therefore, ~ 16,000 to 18,000 new babies and toddlers are identified with hearing loss per year, making hearing loss ***the most common birth defect***.



Hearing loss the most common birth defect

- Of every 1,000 births, two to three children are born with congenital, significant, permanent, bilateral hearing loss.
- Three additional children in 1,000 will acquire deafness in early childhood or by school age.
- Thirty-three infants each day (or approximately 12,000 per year) in the United States are born with permanent hearing loss.
- Infants who spend time in the **intensive care nursery** during the newborn period are at a higher risk for hearing loss with at least **one in 50** showing significant hearing loss.
- Some infants are born with normal hearing but for a variety of reasons may develop **progressive hearing loss** that occurs gradually during preschool and early school years.

Hearing loss the most common birth defect

- It is estimated that 90% of very young children's knowledge is attributed to "incidental reception" of sounds around them. Thus, learning is hindered with even the slightest hearing loss.
- Seventeen of every 1,000 children under the age of 18 have hearing loss.
- There is less than one-half the number of children with severe to profound hearing loss today than two decades ago. But, conversely, there are now more than 10 times the numbers of children with mild to moderate hearing impairments.
- Middle ear infection is the most common infectious disease of childhood and the most common cause of pediatric temporary hearing loss. Nearly all children will develop some period of hearing loss related to ear infections during the period from birth through 10 years of age.
- Ten to 15% of children who receive hearing screening at school fail because they cannot hear within normal limits.

Hearing Loss in Children —A Hidden Disability?

- Hearing loss in children is a silent, developmental, hidden disability.
- Hearing loss is a hidden problem because children, especially infants and toddlers, **cannot tell us** that they are not hearing normally.
- Nearly all their **parents** have normal hearing with little or no knowledge about hearing loss in children.



Primary effects of hearing loss- 1

- **Vocabulary** develops more slowly in children who have hearing loss.
 - These children have little difficulty with concrete words that easily can be matched to objects: family names; specific nouns such as desk, cat, and car; colors; and numbers.
 - They are likely to have more difficulty with **abstract words** such as *before* and *after*, *over* and *under*; concepts such as dreams and imagination and jealousy; and differentiating between items that are similar and/or different.
 - Another area of difficulty is proper utilization of words such as *the*, *a*, *an*, *are*, *have*, *has*, and so forth.

Primary effects of hearing loss- 2

- Children with significant hearing loss understand and express **shorter and simpler** sentences than children who develop with normal hearing.
- Children with hearing loss are more likely to have difficulties with understanding and writing complex sentences. They may not be able to hear word endings such as –s or –ed, which leads to unfortunate and embarrassing misunderstandings of speech, and delay in the proper use of verb tense, pluralization, and possessives as well as nonagreement of subjects and verbs in the same sentence.
- Children with hearing loss often cannot hear **soft speech sounds** such as /s/, /f/, /t/, and /k/, and therefore may not include them in their speech.
- With significant hearing loss, the **vocal quality** of these children may be compromised in terms of prosody, inflection, and rate, making their speech difficult for others to understand. The problem may be that without adequate amplification, the child with hearing loss may not hear his or her own voice as the child speaks, thereby making it difficult for the child to **monitor and correct** his or her own expressive speech and language as it occurs.

Secondary effects of hearing loss

- Hearing loss is indeed a serious problem for children because, if undetected and untreated, it may cause delayed **speech and language** development, **social and emotional** problems, and ultimate **academic** failures.
- Undiscovered hearing loss in children may carry difficulties and **social problems** clear into adulthood.
- Therefore, the secondary effects of hearing loss adversely affect a child's development as much as or more so than the hearing loss itself does.



The importance of Pediatric Audiology

- Recently there has been a surge of technology and information about testing and managing hearing loss in infants and children. The impetus for this surge has been **newborn hearing screening**.
- As a result of identifying and treating hearing loss in neonates, we now are dealing with a vastly different population of children with hearing loss, a population that never existed before.
- With this new population, whose hearing loss is identified at birth, the secondary developmental and communicative deficits of hearing loss that were so common can now be prevented.
- What has happened in the field of hearing loss is revolutionary, and the pediatric audiologist is in the linchpin position.

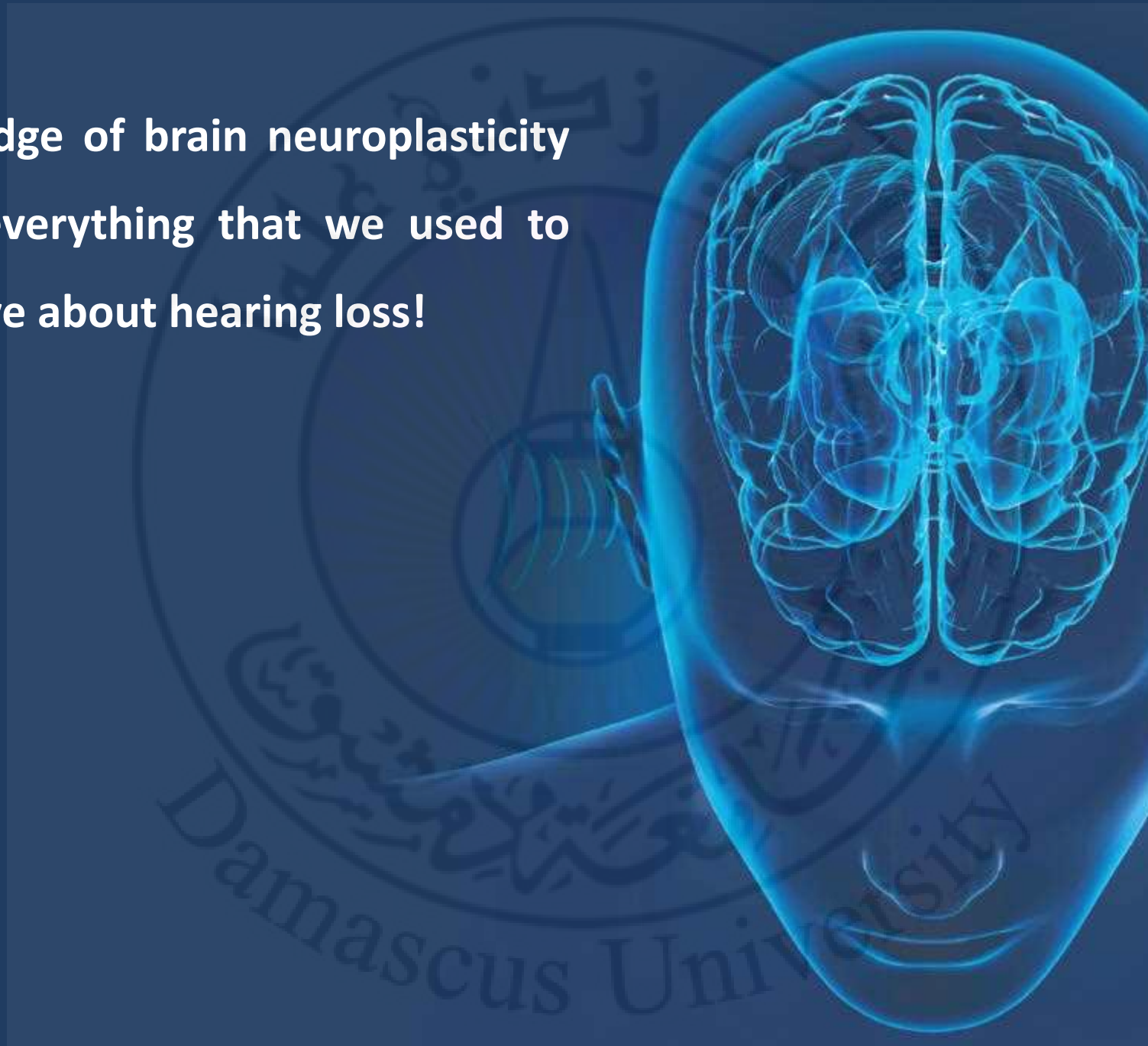
Pediatric Audiology Is Changing!



Pediatric Audiology Is Changing!

- Changes brought about through technology and **early hearing detection and intervention (EHDI)** programs have permitted outcomes of listening and talking only dreamed of a few years ago.
- It is important to realize that the new outcomes available today do not invalidate the treatment decisions made by pediatric audiologists in the past. Audiologists did what was necessary with what was available at the time. For example, until the 1970s, children with bilateral hearing loss were routinely fitted with only one hearing aid.
- With increased knowledge, we can now offer better services. Audiologists today do the best that can be done in today's world.
- Tomorrow's world will bring new possibilities. Our job as pediatric audiologists is to prepare today's babies to be take-charge adults in the world of 2030, 2040, and 2050—not in the world of 1970, 1990, or even 2020. Because information and knowledge are the currencies of today's cultures, listening, speaking, reading, writing, and electronic technologies must be made available to our babies and children to the fullest degree possible.

**Current knowledge of brain neuroplasticity
has changed everything that we used to
know and believe about hearing loss!**



Hearing Occurs in the Brain

- We hear with the **brain**; the ears are the doorway to the brain for sound/auditory information.
- Consequently, **hearing loss is primarily a brain issue, not an ear issue**; the problem with hearing loss is that it keeps sound from reaching the brain.
- The child's listening and spoken language outcomes are not determined by 16,000 (or likely much fewer) hair cells or by 30,000 auditory nerve fibers but by 100 billion neurons in the brain, processing 100 trillion pieces of information per second.



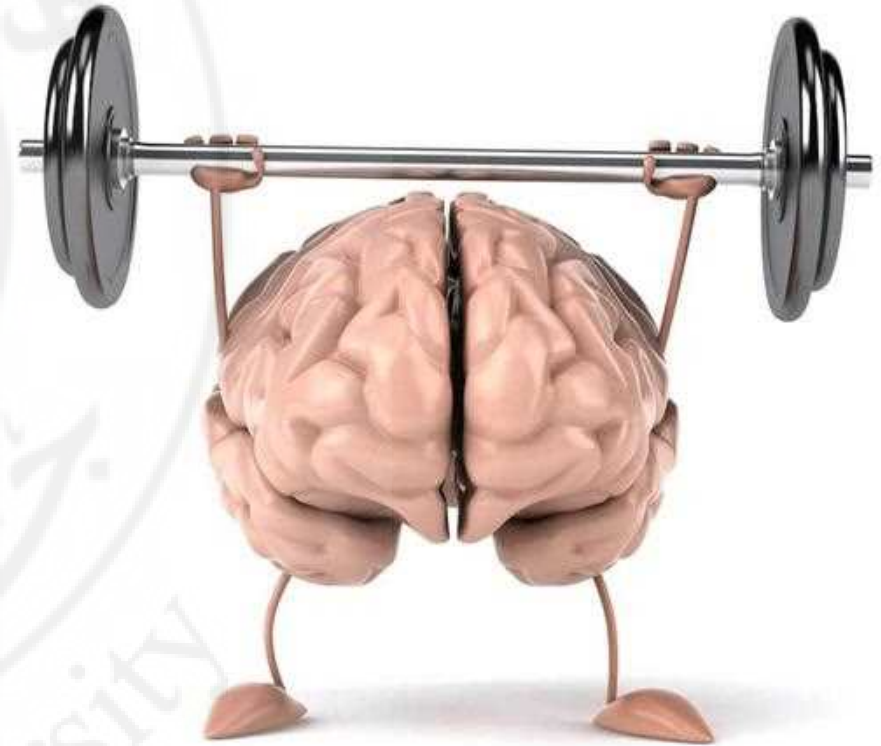
Hearing technologies break the doorway open!

- The purpose of hearing technology is to get auditory information through the doorway (the ear) to the brain. There is no other purpose!
- This **brain access** allows the use of a developmental model of intervention that prevents the negative developmental outcomes of hearing loss that were so common a few years ago.



Neuroplasticity

- Neuroplasticity refers to the brain's ability to reorganize itself, forming new connections and pathways in response to sensory and motor input, learning new skills, or in creating new memories upon which to build additional behaviors.
- This process involves orderly increases in the number and density of neurons, dendrites, synapses, and the proteins that are essential to the survival of nerve cells.



Neuroplasticity

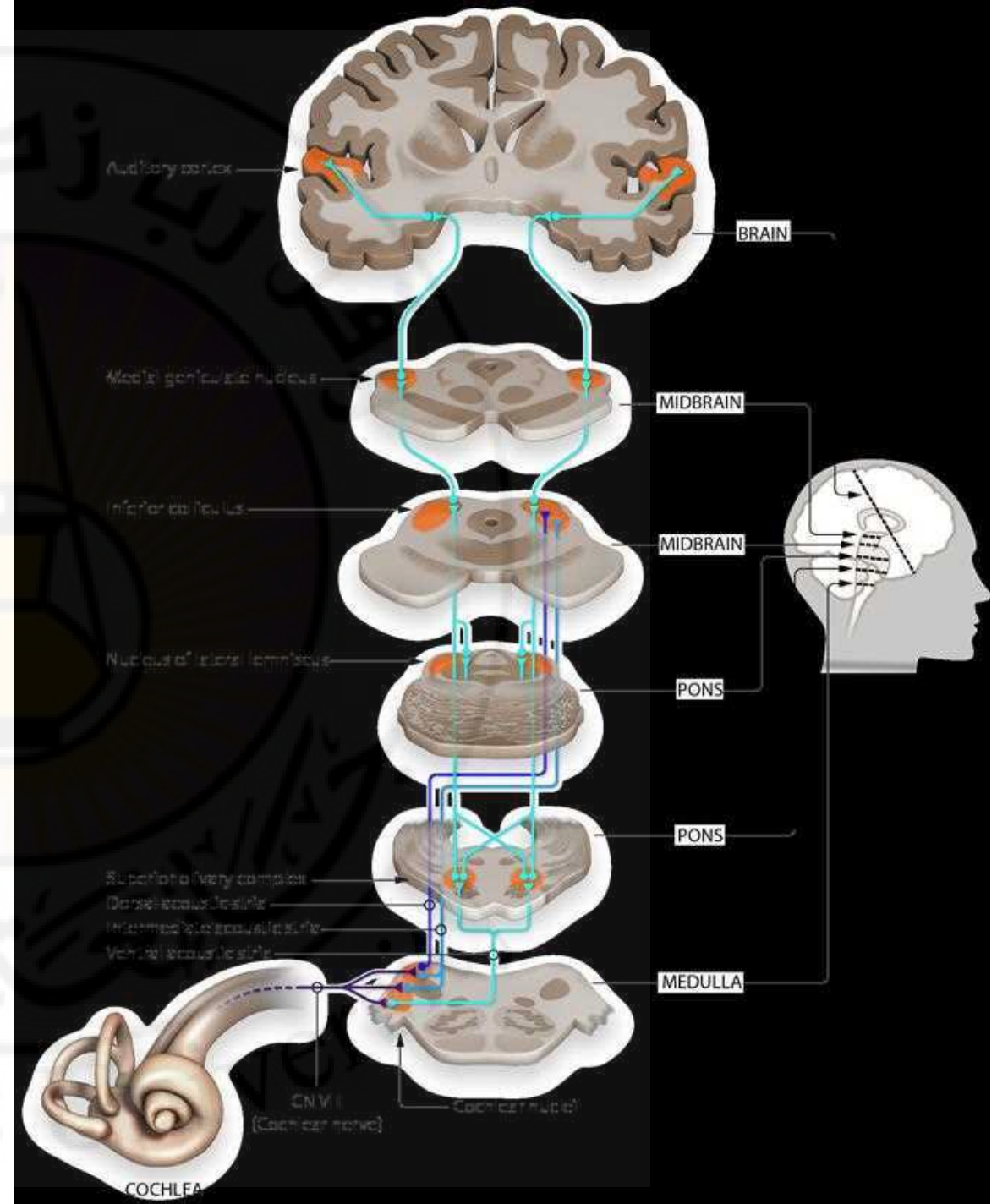
- Studies of brain development show that sensory stimulation of the auditory centers of the brain is critical and, indeed, influences the actual organization of auditory brain pathways.
- The fact is, the brain can organize itself only around the stimuli that it receives.
- If complete acoustic events are received, this is how the brain will be organized. Conversely, if hearing loss filters some or all speech sounds from reaching auditory centers of the brain, the brain will be organized differently.
- When we want to remember (or learn) something we have heard, we must hear it clearly because memory can be only as clear as its original signal.
- Signal enhancement, such as that provided by amplification technology, is really about brain stimulation, with subsequent development of auditory-neural pathways.

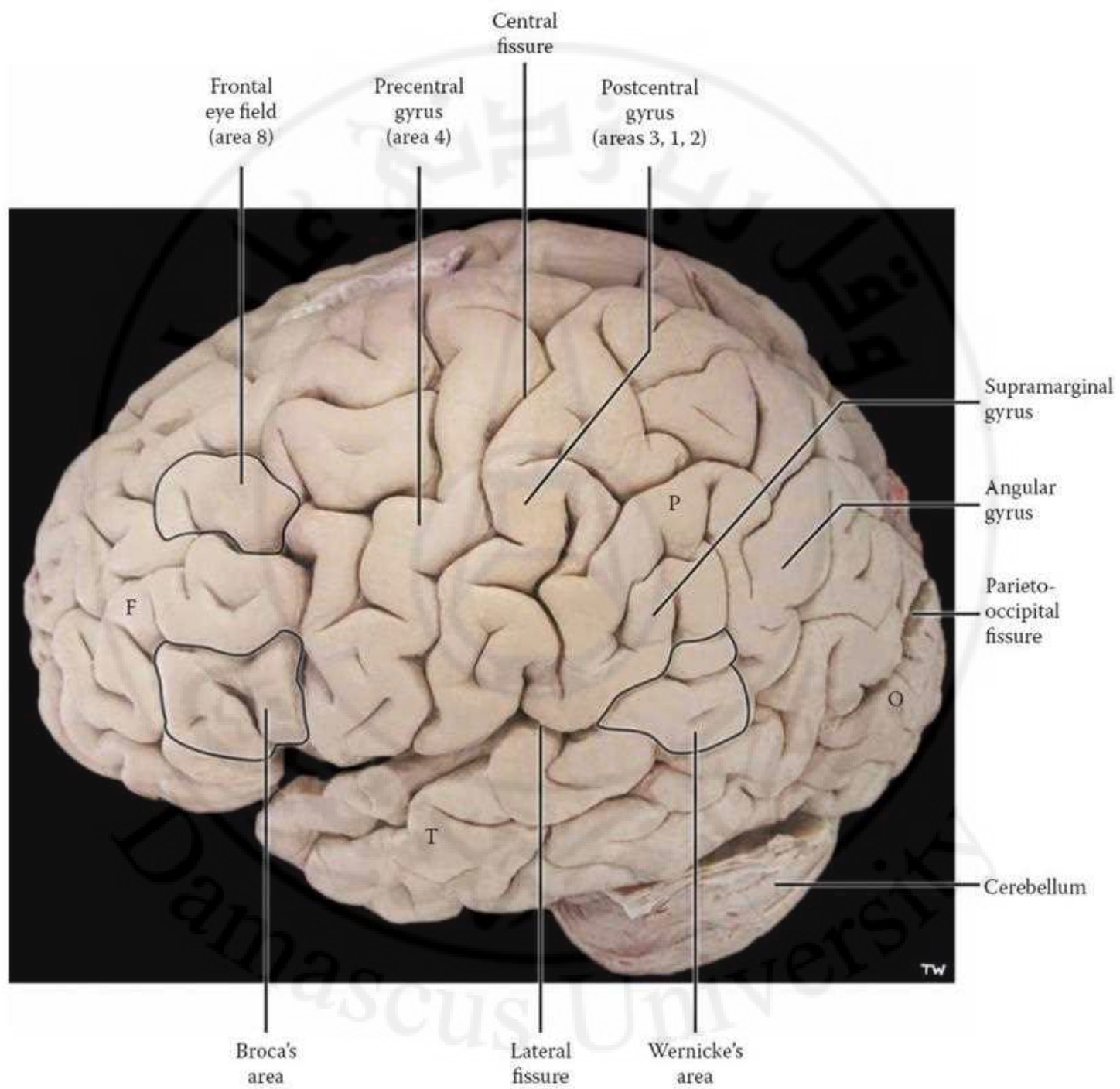


Neuroplasticity

Auditory Cortex

- For persons who have sufficient access to intelligible speech during the first several years of life, the auditory areas of the brain respond robustly when stimulated by speech or other meaningful sounds.





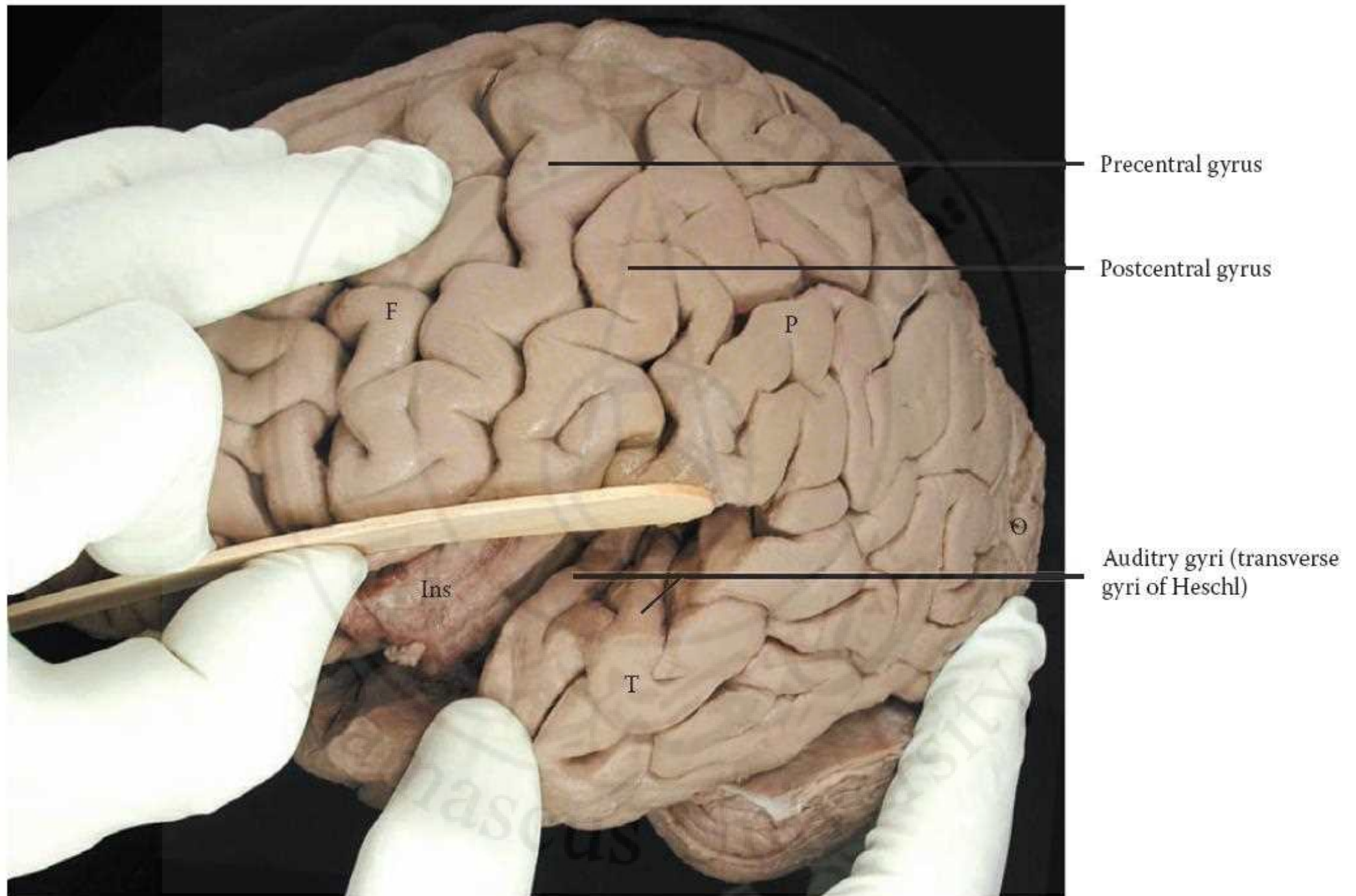


Central
fissure

Auditory gyri
(transverse gyri
of Heschl)

Insula

Lateral
fissure
(opened)



Precentral gyrus

Postcentral gyrus

Auditory gyri (transverse
gyri of Heschl)

F

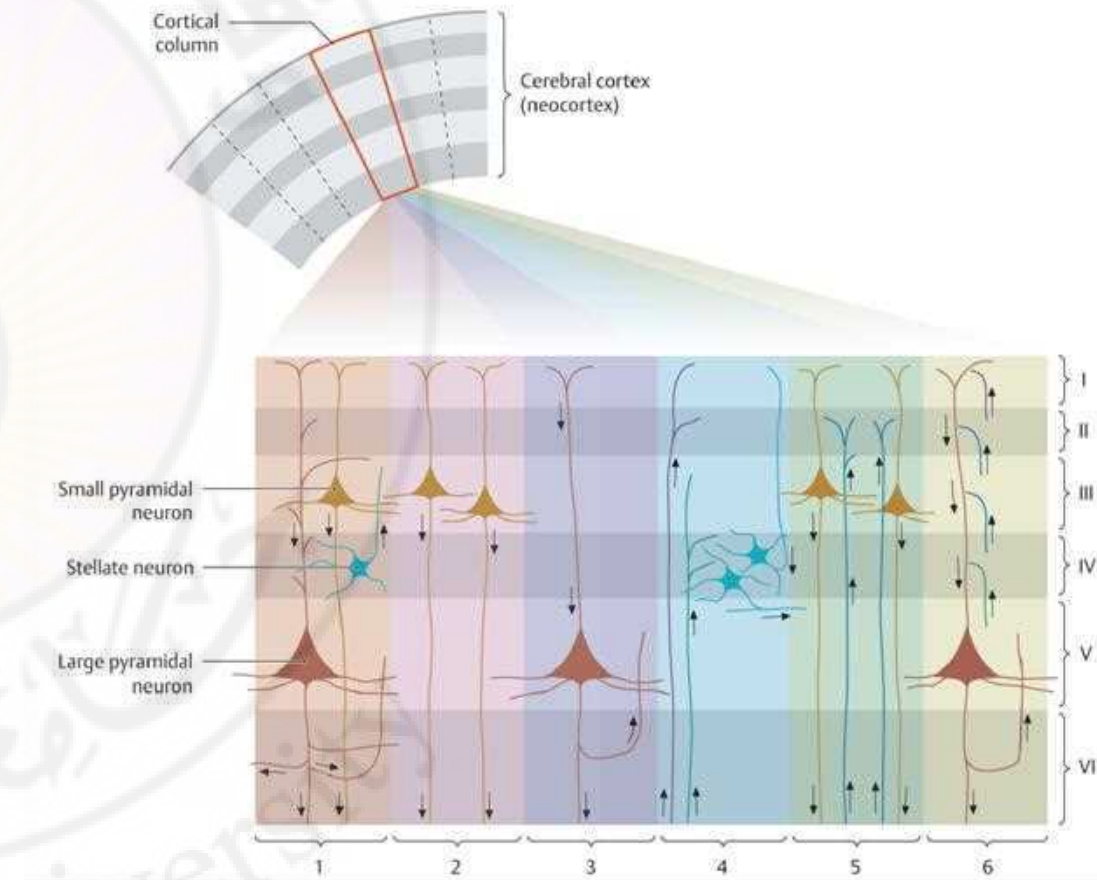
P

Ins

T

Auditory Cortex

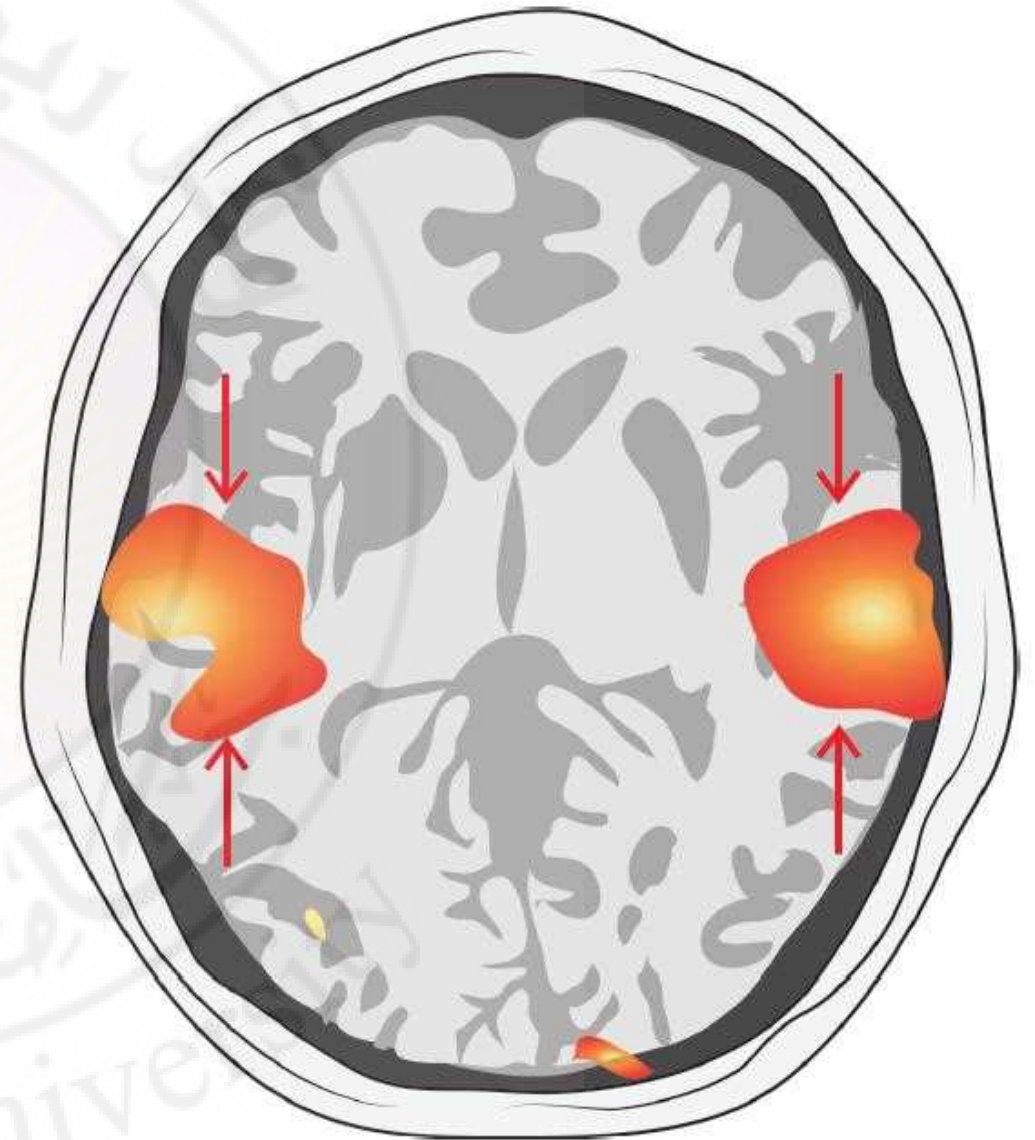
- the auditory cortex, which is **2 to 4 mm in thickness**, comprises **six layers** of neurons.
- The afferent auditory nerve fibers from the medial geniculate body of the auditory thalamus arrive at layer IV, and a good deal of the processing that takes place within the primary auditory cortex occurs in layers I to III, which are referred to as the **supragranular layers**.
- Layers V to VI are known as the **infragranular layers** of the auditory cortex. The infragranular layers accomplish several functions, but most importantly, they serve as the output circuits of the primary auditory cortex.
- More specifically, auditory information from the primary auditory cortex is delivered to other areas of the brain, including to the secondary auditory cortex, from layers V and VI of the primary auditory cortex.



Positron emission tomography (PET) image of areas of the brain that were responsive (arrows) when postlingually deafened adults listened to intelligible speech while using a cochlear implant.

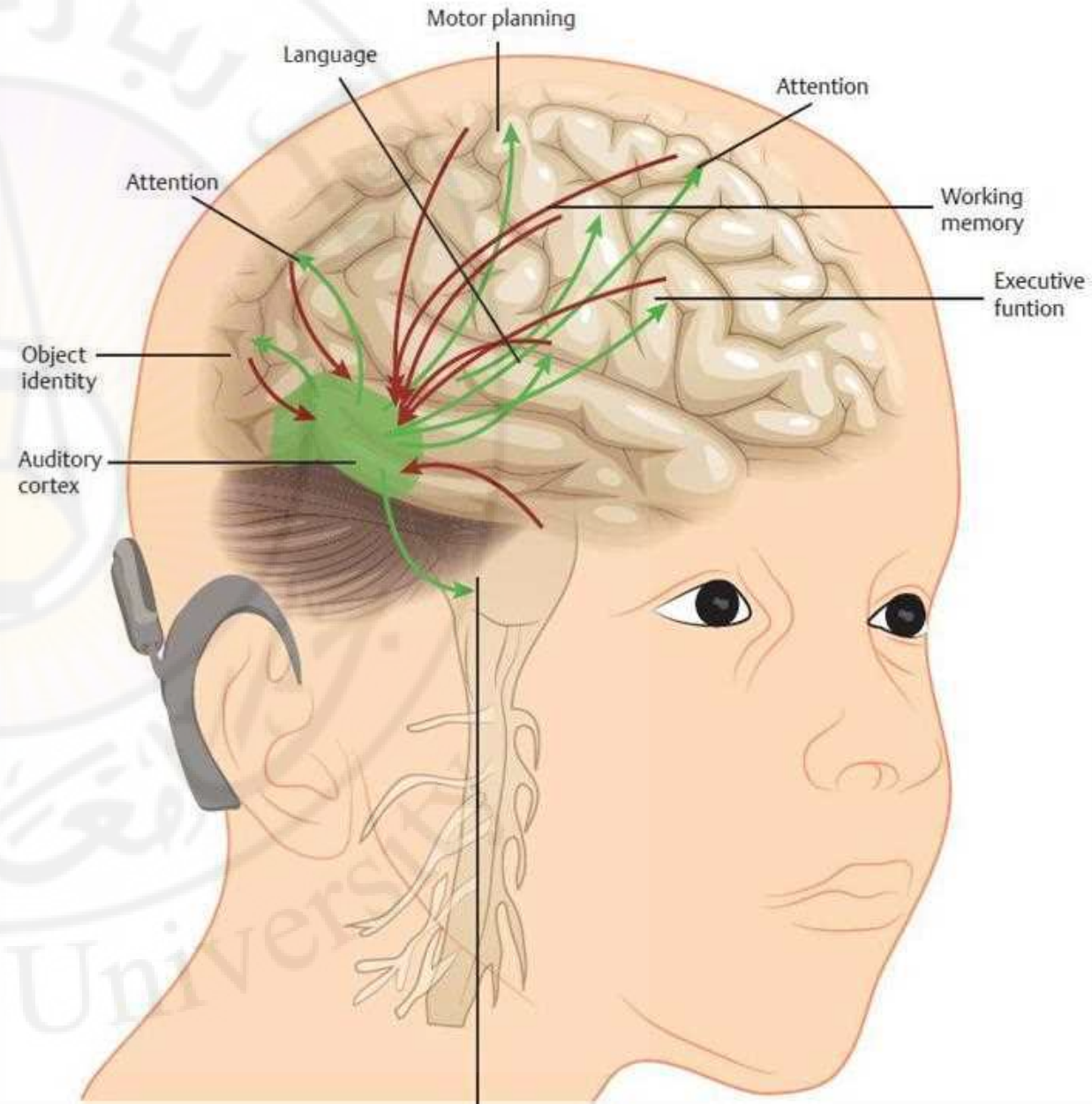
Stimulation from the cochlear implant at one ear in response to the running speech resulted in bilateral activation of both the primary and secondary auditory cortices.

The ipsilateral ascending auditory tracts and the **corpus callosum** serve to facilitate a bilateral response. However, most importantly, the auditory stimulation produces a broad and robust response that spans throughout the primary and secondary auditory cortices.



Connectomes or Neural Networks

- every sound that comes into our minds from our ears is reduced to its own unique pattern of neural activity across the brain.
- In order for our sound-elicited experience not just to result in simple detection but also to possess higher-order meaning and “come to life,” auditory stimulation has to extend beyond the primary auditory cortex to the secondary auditory cortex, where it is delivered to other areas of the brain to form interconnections known as a **neural network or connectome**.
- A connectome is a map of functional neural connections in the brain and may be thought of as the brain’s wiring diagram



Normal neural connectome

- A normal neural connectome is established during the first several years of life through a number of **stimulus-driven** processes.
- First, through a process referred to as **synaptogenesis** (formation of synapses, the structures that carry signals between two communicating neurons), neuron-to-neuron contacts are formed, establishing neural pathways throughout the brain.
- New synapses are created through a process known as **arborization** (from *arbor*, the Latin word for tree), in which the transmitting axons and receiving dendrites of neurons grow and branch out to come into contact with neighboring neurons, with which they can form synapses.
- Synaptic counts reach a maximum **between the first and fourth years of life** and decline thereafter.
- The creation and maintenance of functional synapses is dependent upon **experience**. Synapses across the brain are sustained when they receive consistent and meaningful input from sensory systems or from other areas within the brain. In contrast, synapses that are not used (e.g., stimulated by input from the sensory system) are subjected to **synaptic pruning**, the elimination of synapses in neural pathways that do not receive meaningful stimulation.

Critical period of brain development

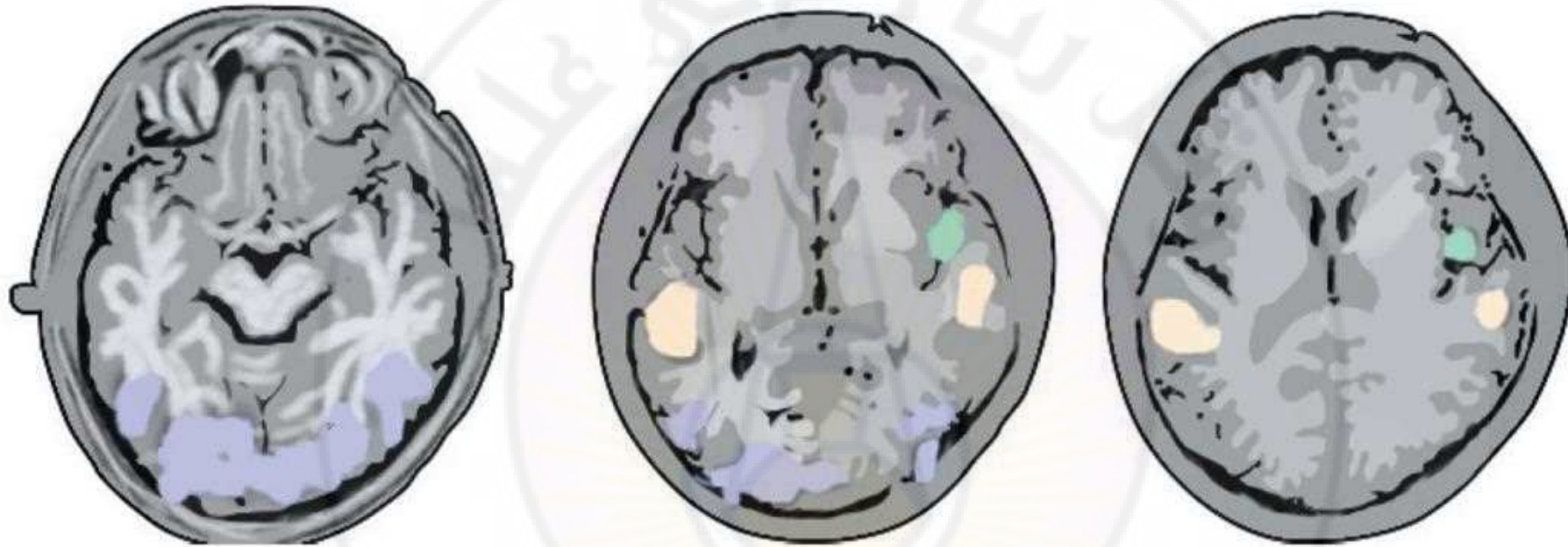
- Although arborization, synaptogenesis, and synaptic pruning occur throughout the lifespan, these phenomena are far more prevalent during the first few years of life (i.e., during the **critical period of development and maturation**) and decline substantially throughout childhood.
- As a result, brain plasticity, or the brain's ability to change and establish new functional connections, is greatest during the early childhood years.
- **Hearing deprivation** during early development prevents functional maturation, delays cortical synaptogenesis, and increases synaptic elimination, ultimately affecting central functions.



Auditory Deprivation

- Although the Primary auditory cortex is committed to audition, and its dedication allows for detection of auditory input from the peripheral auditory system, regardless of an individual's duration of deafness; long duration of deafness most likely results in some reorganization of the primary auditory cortex and some change or reduction in its responsiveness to sound.
- In the absence of sufficient access to intelligible speech from the peripheral auditory system, the secondary auditory cortex is functionally “**decoupled**” from the primary auditory cortex and transforms to assist in the processing of other sensory modalities (Vision or tactile...).
- If auditory input is not delivered to the secondary auditory cortex, then it cannot be distributed to the rest of the brain for higher-order processing that is necessary to convey a meaningful auditory experience. The auditory neural network or connectome is fractured between the primary and secondary auditory cortex.

Auditory Deprivation



Typical response obtained from prelingually deafened adult participants who had no auditory access to intelligible speech and used sign language prior to receiving a cochlear implant (CI) in adulthood, as they listened to running speech while using their cochlear implant. *Blue:* areas activated by visual stimuli (meaningless hand movement); *yellow:* areas activated by sign language; *green:* areas activated by spoken language (CI: left ear).

Critical Period of Auditory Brain Development

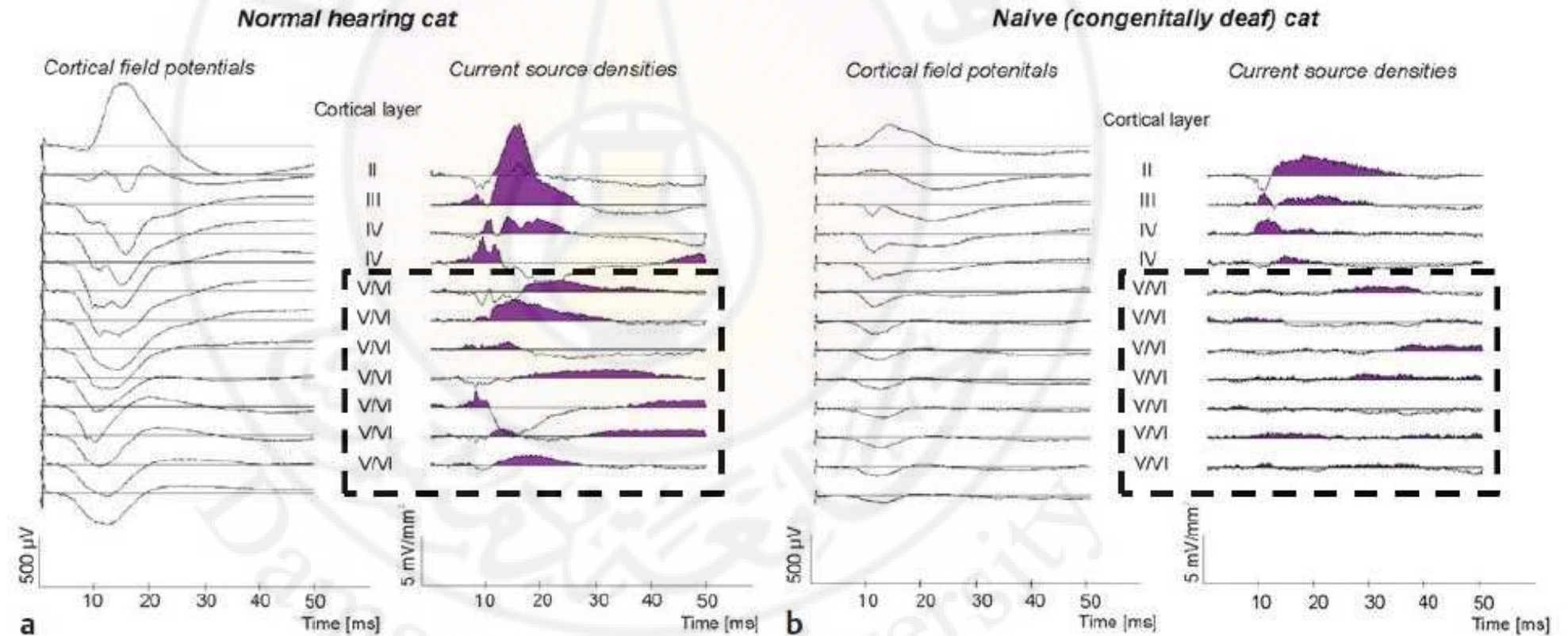


Fig. 1.9 (a) Normal hearing cats and kittens who were implanted during the first 3 months of life, showing normal activity observed at all layers of the primary auditory cortex. (b) White deaf cats who received a cochlear implant after 5 months of age showed normal activity in layers I to IV but reduced or no activity in the infragranular layers (V–VI).

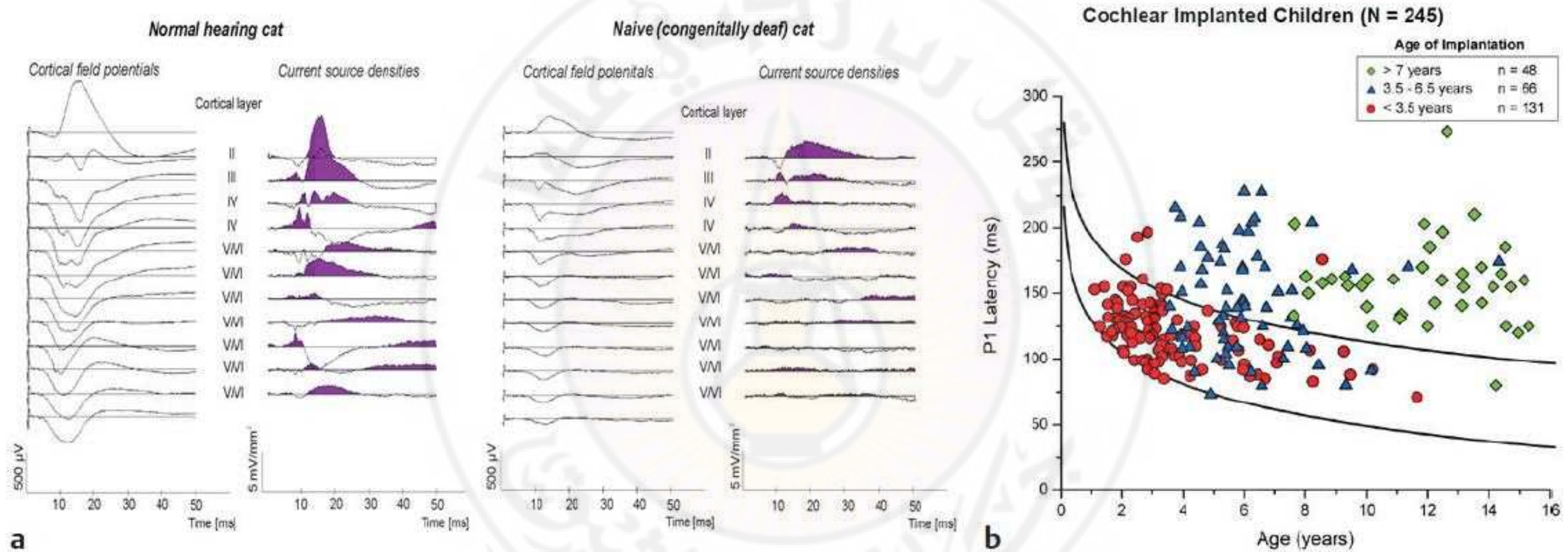


Fig. 1.10 (a) Responses to sound measured in cortical layers of normal-hearing cats (on the left) and congenitally deafened late-implanted cats (on the right).³² **(b)** Latency of the P1 waveform of the cortical auditory evoked potential (P1-CAEP) in subjects who were born deaf and received a cochlear implant at ages ranging from 1 year old to early adulthood. Note that children who received cochlear implants during the first 2 to 3 years of life had normal P1 latencies, while children who received their implants later typically did not.³¹

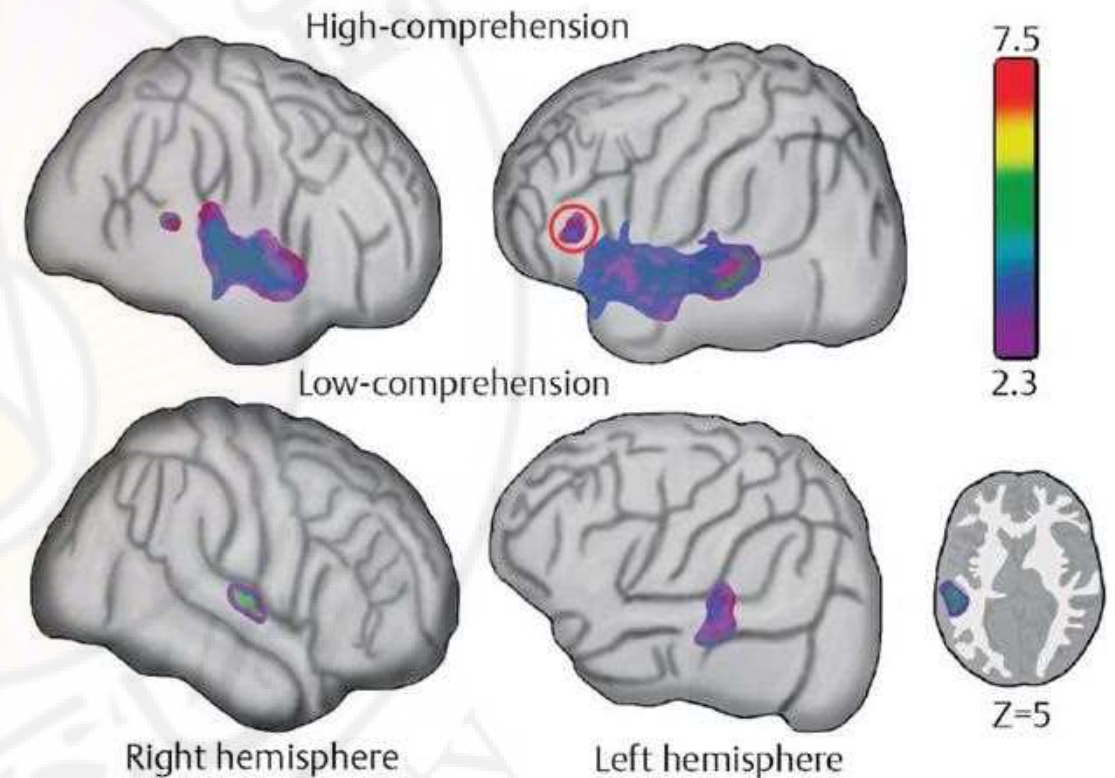
Every day within the critical period is critical!

- Children with hearing loss must be appropriately fitted with hearing technology (e.g., hearing aids, bone conduction devices, cochlear implants, remote microphone systems) **as early as possible** in order to prevent auditory deprivation and irreparable changes to the auditory brain and auditory neural network.



Spoken Language Enrichment: A Necessity for Auditory Brain Development

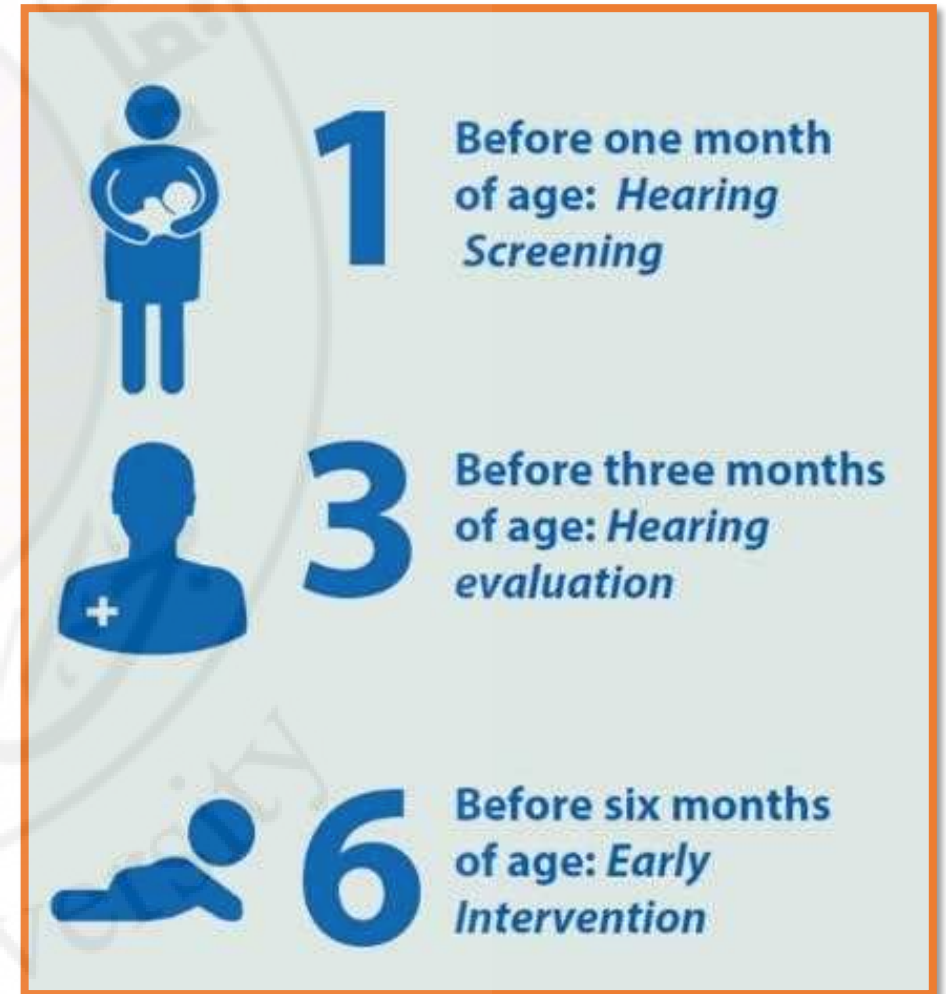
- It is also necessary for child's parents (caregivers) to be equipped with strategies to create an environment that inundates the child with **intelligible speech** and a complex, high-level model of spoken language and abstract language concepts.



PET scan imaging to evaluate activity in the left inferior prefrontal cortex (LIPC) of cochlear implant recipients who listened to running speech. *Top right, circle:* Activation of the LIPC requires exposure to intelligible speech. High performers show activity in LIPC (top right and left), while poor performers do not (bottom right and left).

To achieve age appropriate spoken language outcomes, pediatric audiologists must:

- Offer the climate necessary for the auditory brain to develop as intended
- Identify hearing loss within the first month of life
- Provide the child with the hearing technology he or she needs as soon as possible
- Assist the child's parents (caregivers) in creating an acoustically robust, language-rich listening environment during the first few years of life



Hearing vs. Listening

- There is a distinction between hearing and listening. Hearing is acoustic access to the brain; it includes improving the signal-to-noise ratio by managing the environment and using hearing technology. Listening, on the other hand, is focusing and attending to the acoustic events that are available.

- **Sequencing is important!**

- Hearing must be made available by audiologists *before listening* can be taught by parents, early interventionists, speech-language pathologists, auditory-verbal practitioners, teachers, and, of course, audiologists.
- That is, one can reasonably focus on developing listening skills and strategies only after acoustic events have been made available to the brain— not before.

Hearing	Listening
Hearing is passive	Listening is active
Refers to the act of perceiving a sound through the ear	Refers to the act of making a conscious effort to perceive the sound
Does not require a conscious effort	Require a conscious effort
Involuntary	Voluntary

Even in the presence of significant hearing loss, a child with hearing problems who is identified early in life and enrolled in an appropriate intervention program has the opportunity to hear, learn, and develop similar speech and language skills as children with normal hearing.



1

SCREENING

3

DIAGNOSIS

6

INTERVENTION

What we learn from story of Helen Keller?

- The importance of early hearing to language is demonstrated by the story of the deaf-blind Helen Keller, whose remarkable achievement in mastering language skills has become legend.
- Her proficiency in language can be understood when one realizes that she acquired both deafness and blindness from meningitis in 1882 at 19 months of age.
- **“It seems as if even a short exposure to language, a brief moment during which the curtain has been lifted and oral communication established, is sufficient to give a child some foundation on which much later language may be based.”**



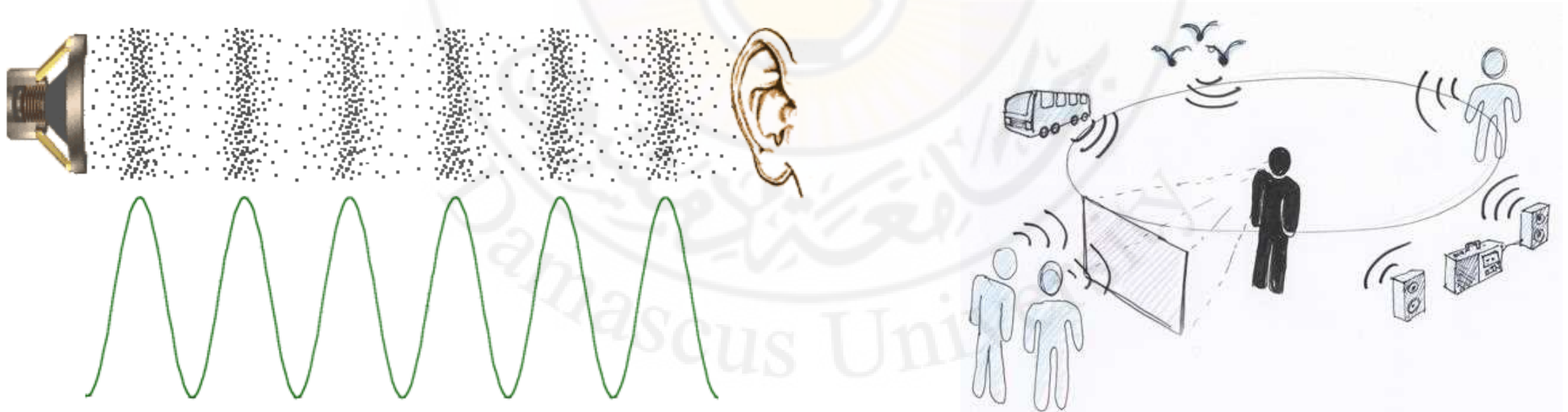


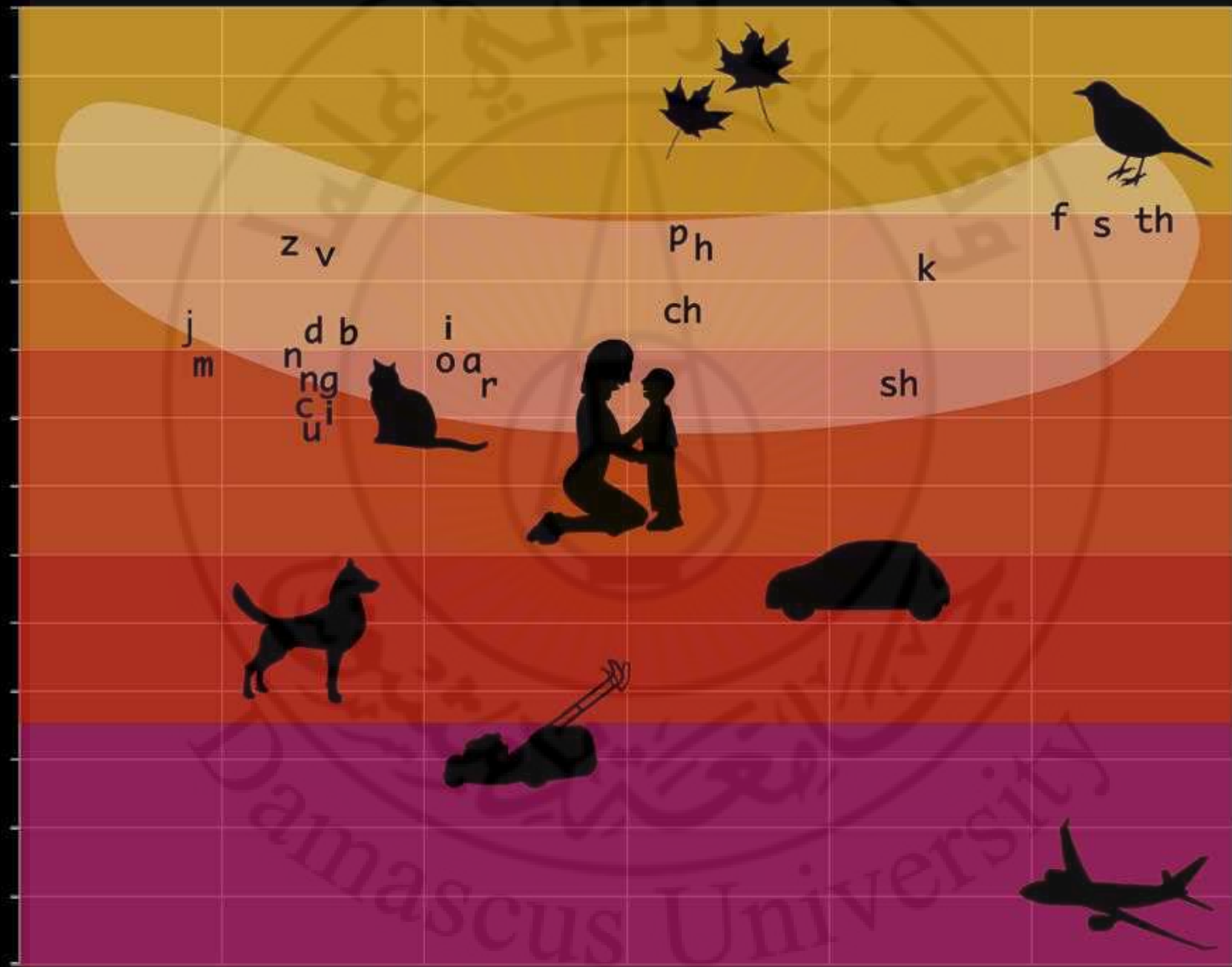
2. Hearing loss in children



The Nature of Sound

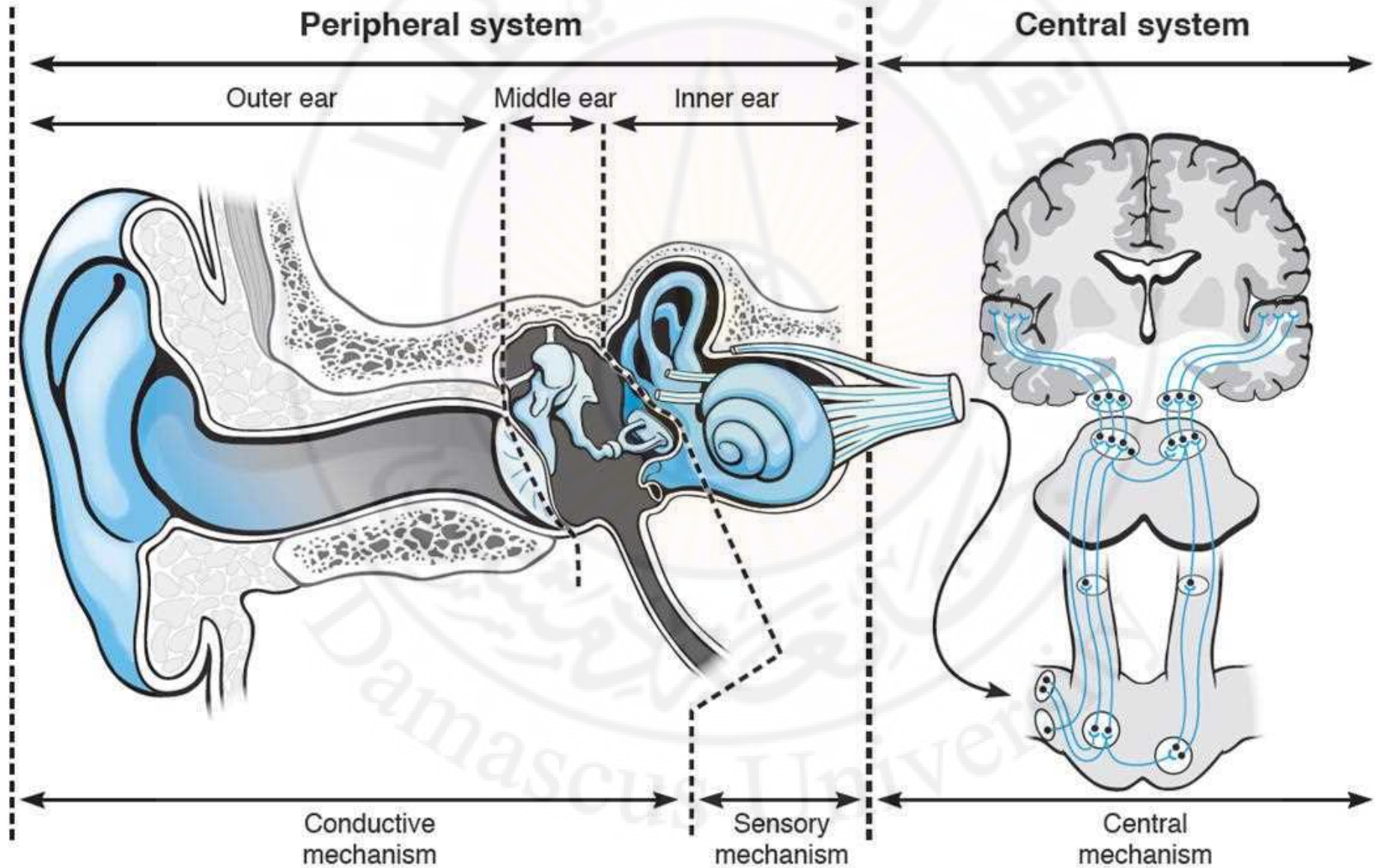
- The physicist views sound as a propagating disturbance in the air initiated by a vibrating source. The physicist studies the generation, transmission, reception, and modification of sound waves as the science of acoustics.
- The psychologist studies the human reaction to sound waves that we know as “hearing.” There is important information contained within both points of view which needs to be acknowledged to understand how we hear and communicate.





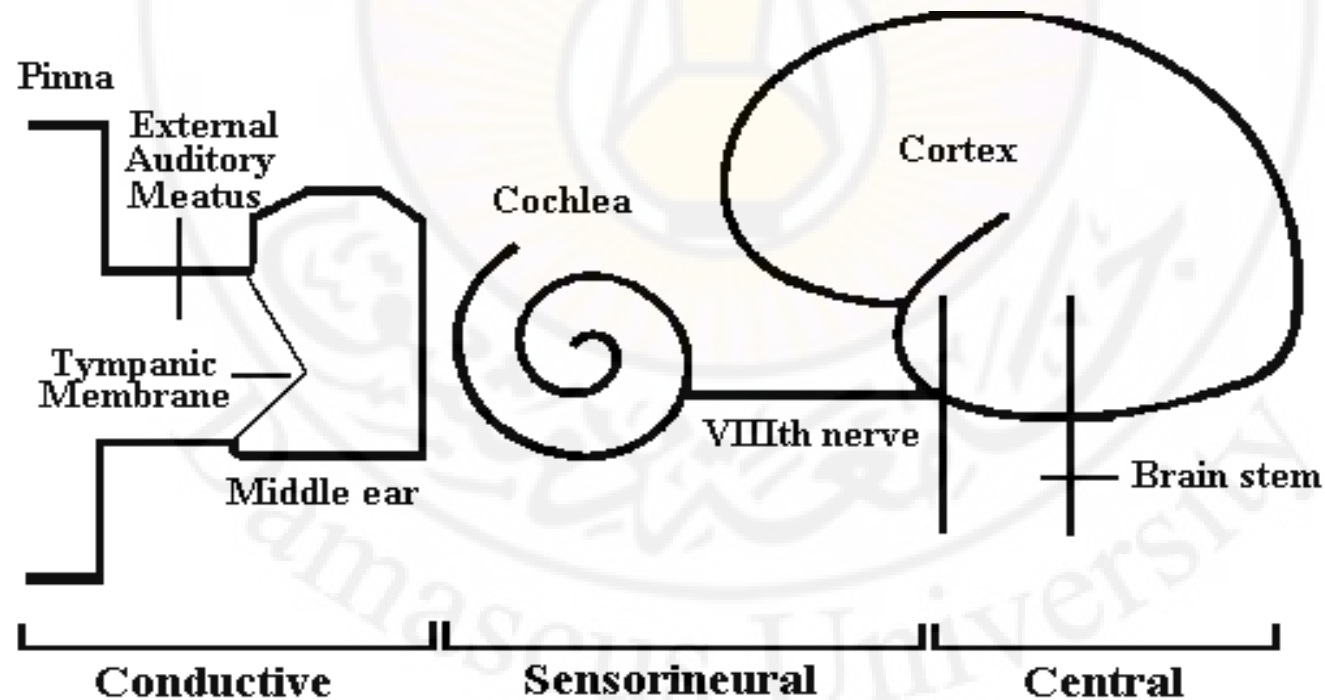
The audibility of speech in regard to the child's hearing loss.

How we hear

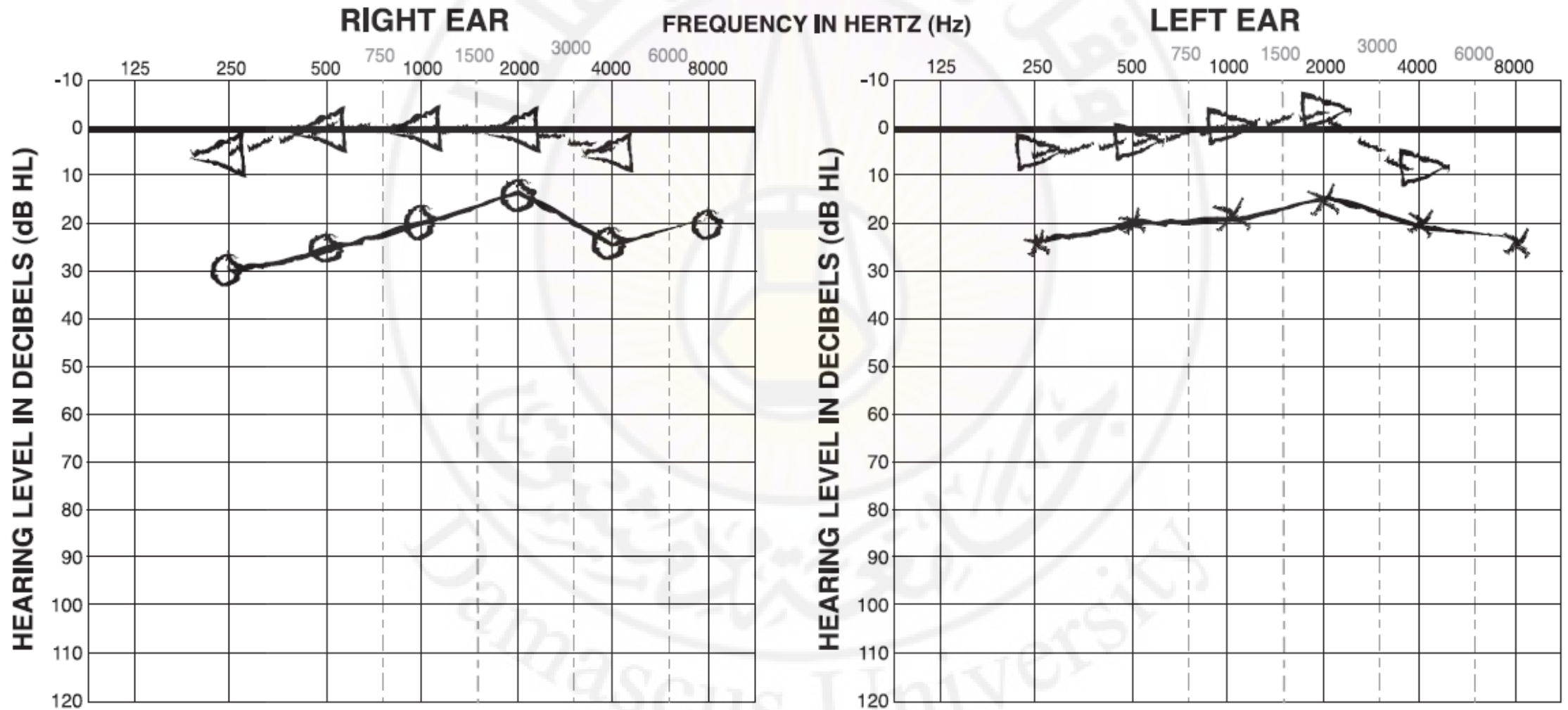


The Nature of Hearing Loss

- Hearing loss can result from a wide spectrum of causes including inherited or congenital problems, infections, diseases, or traumatic situations that affect different portions of the ear and hearing mechanism.
- Audiologists traditionally describe hearing loss in three categories as conductive hearing loss, sensorineural hearing loss, or mixed hearing loss, which is a combination of the two major categories.

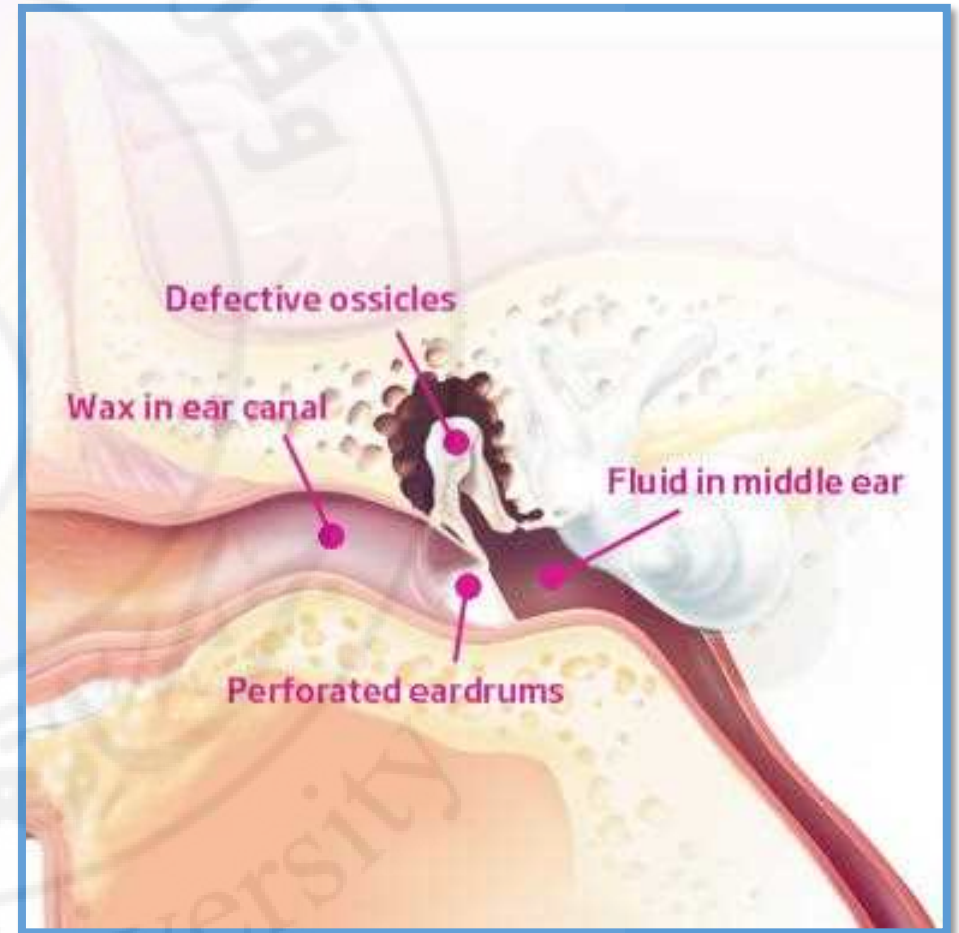


Conductive Hearing Loss



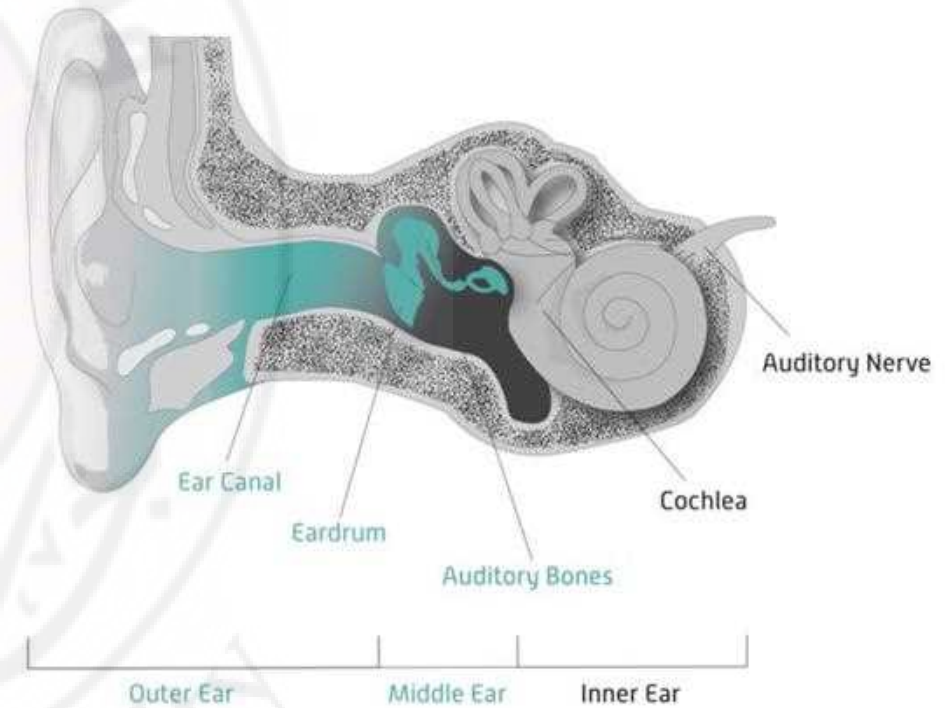
Conductive Hearing Loss

- Conductive hearing loss is the most common cause of hearing problems in children often resulting from ongoing ear infections; and may often be a temporary condition.
- Although the inner ear may function normally in persons with conductive hearing loss, the sound vibration is unable to stimulate the cochlea via the normal air conduction pathway because of blockage in the ear canal, tympanic membrane, or middle ear.



Conductive Hearing Loss

- Conductive hearing loss is usually mild to moderate in degree, may occur in one or both ears at the same time, and may resolve spontaneously over time without treatment.
- However, conductive hearing loss may also persist in many patients leading to the necessity of minor medical or surgical treatment to clear up disease and return the hearing to normal limits.
- More significant conductive hearing loss may be associated with facial or skull malformations that require significant surgical intervention to correct.



Conductive Hearing Loss

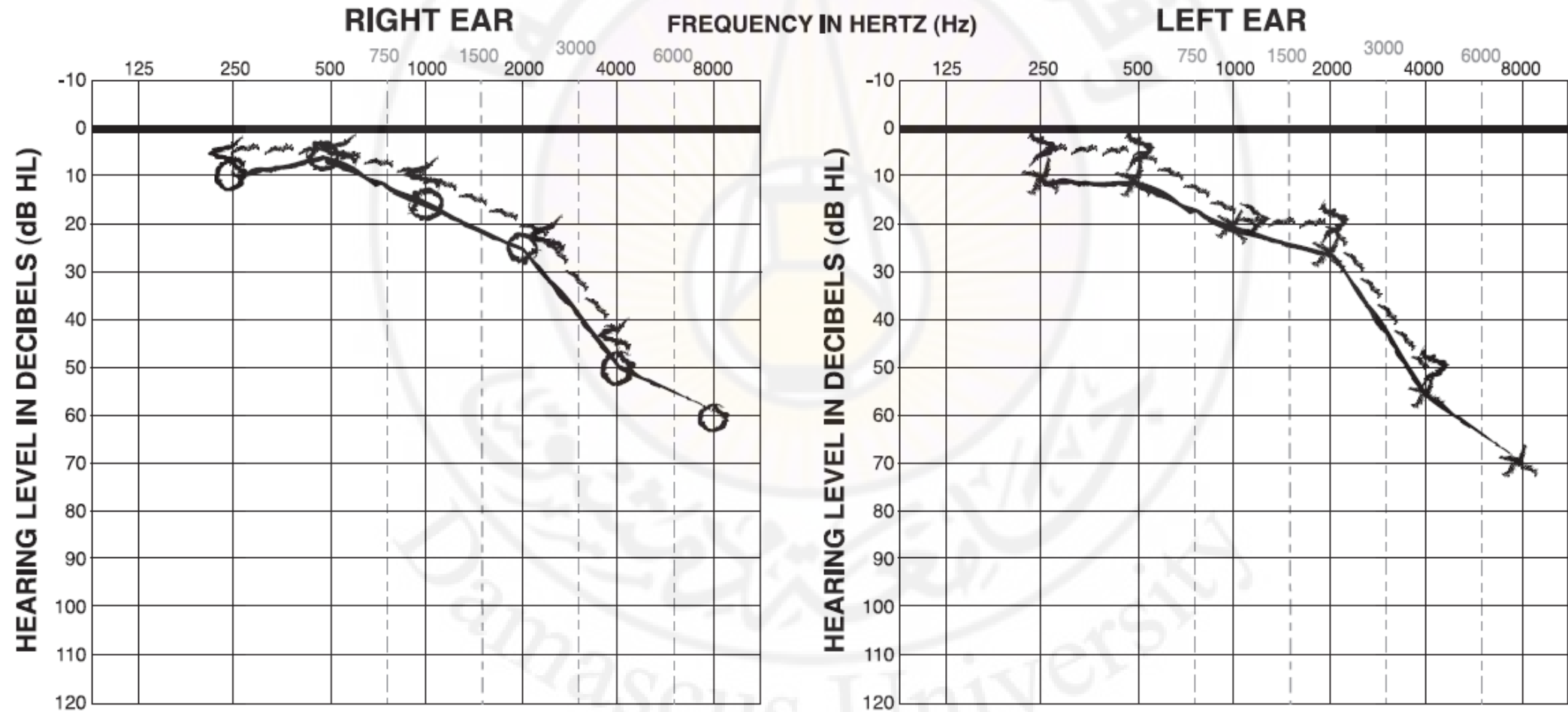
- conductive component to the hearing problem may be present along with permanent sensorineural-type hearing loss. In these patients, the bone conduction threshold values represent “better hearing” than the air conduction threshold measures. Eliminating the conductive component through treatment or spontaneous recovery will result in the improvement of the hearing loss to the levels of permanent hearing loss.
- The most common causes of conductive hearing loss are sterile fluid in the middle ear; ear infections of the ear canal, the middle ear, or both; or foreign objects in the ear canal (i.e., pop beads, broken crayons, insects, etc.) that inadvertently find their way into the ear canal and must be removed.
- Earwax found in the ear canal can be pushed deeper through personal cleaning efforts and should be cleaned out only by someone with proper experience and safe instruments. Public health officials warn against the use of cotton swabs, sometimes known as Q-tips, as a dangerous procedure that deposits shreds of cotton fibers in the ear canal and may actually result in causing more significant problems.

Conductive Hearing Loss

- Conductive hearing losses are the most common type of hearing loss found in children, usually resulting from transient ear infections known technically as “otitis media.”
- Conductive hearing loss can also occur when the ear canal or middle ear air conduction pathway is totally blocked (occluded) with wax or some other physical obstruction, producing a maximal 60 dB HL conductive hearing loss.
- Conductive type hearing loss is often found in individuals with **craniofacial malformations** that might include malformation of the middle ear structures. A
- Although most conductive hearing losses caused by otitis media resolve spontaneously, more severe cases require medical or surgical treatment to return the hearing to normal levels.

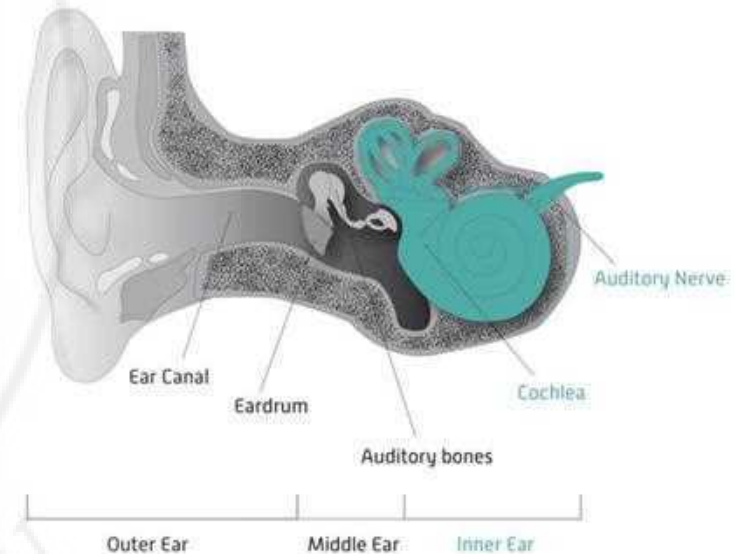


Sensorineural Hearing Loss

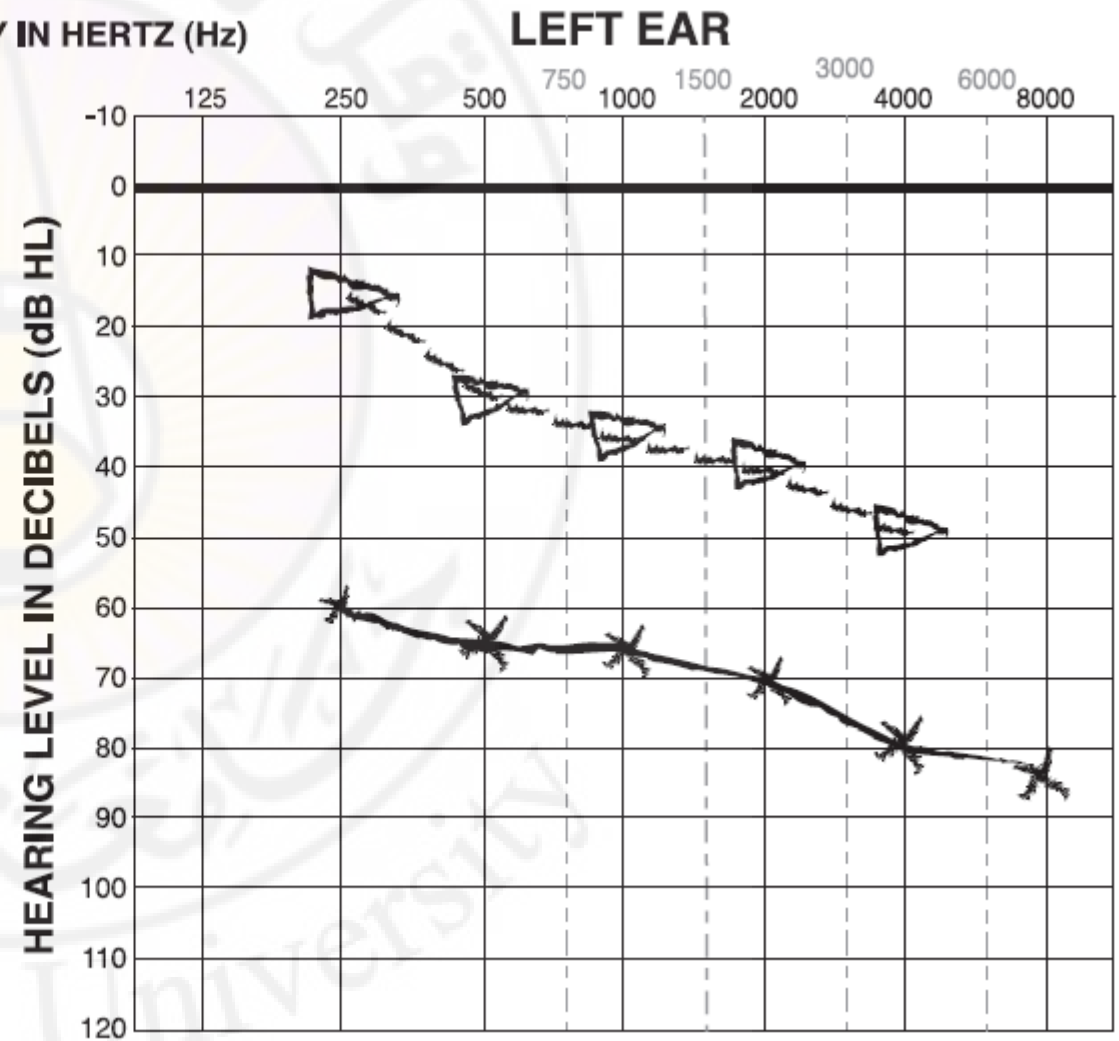
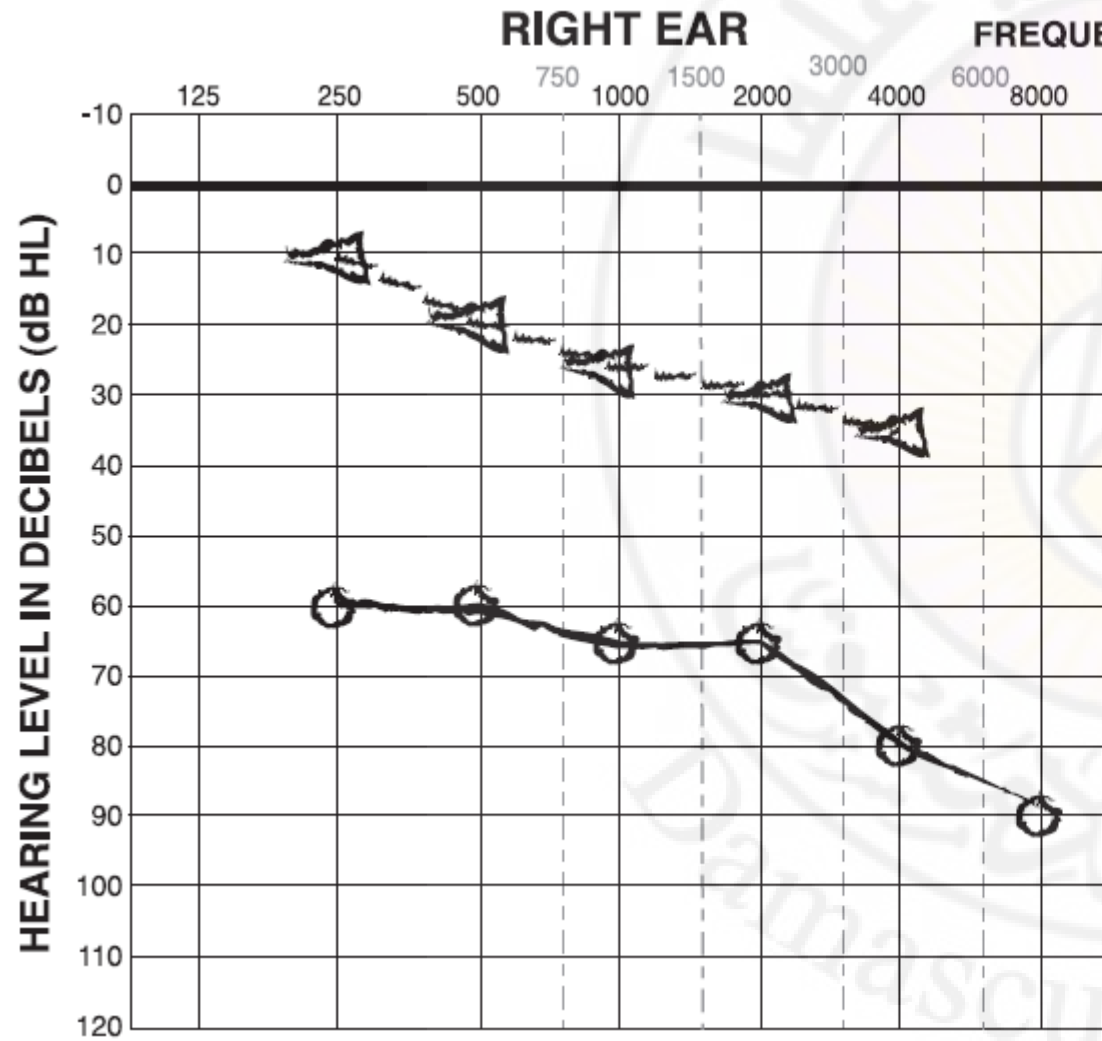


Sensorineural Hearing Loss

- The term sensorineural has long been used to describe permanent hearing loss caused by damage to the cochlear hair cells or auditory nerve fibers leading to the brain.
- Current audiometric testing techniques, such as otoacoustic emissions, electrocochleography, and auditory evoked potentials, can provide an objective means to differentiate between sensory and neural hearing loss.
- Sensorineural hearing loss may be caused by a wide variety of etiologies including bacterial and viral infections; genetic or familial inheritance; disease processes involving the otic capsule, auditory nerve, or membranous labyrinth; metabolic disorders; excessive noise exposure; or toxic pharmacological agents.
- Sensorineural hearing loss is nearly always **permanent and irreversible**.

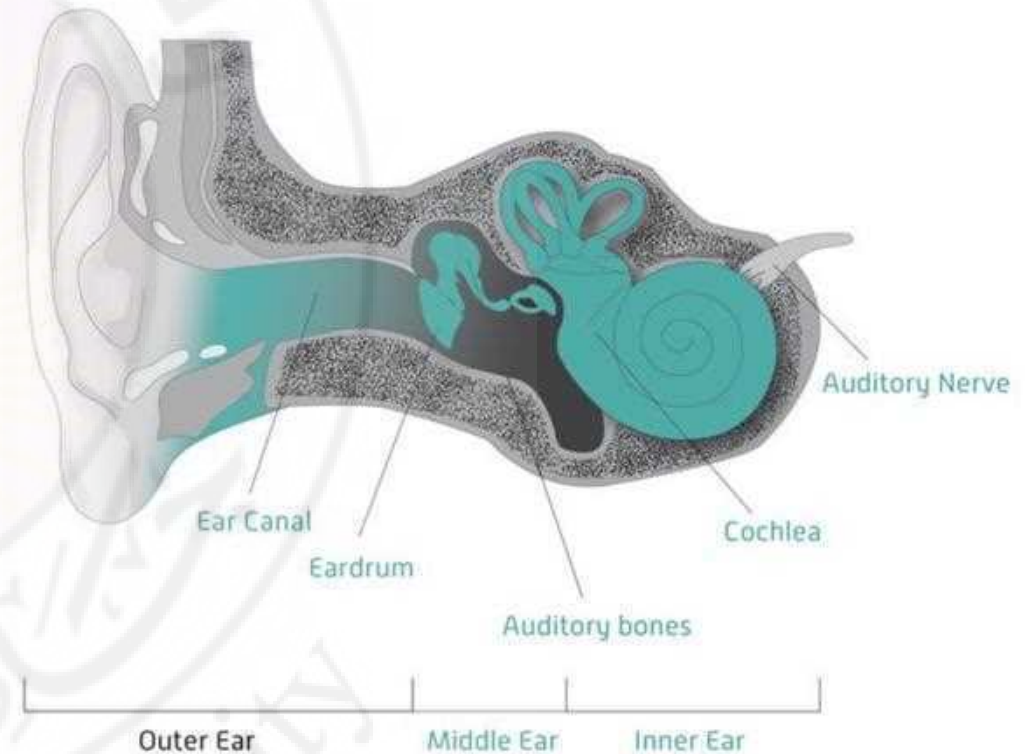


Mixed Hearing Loss

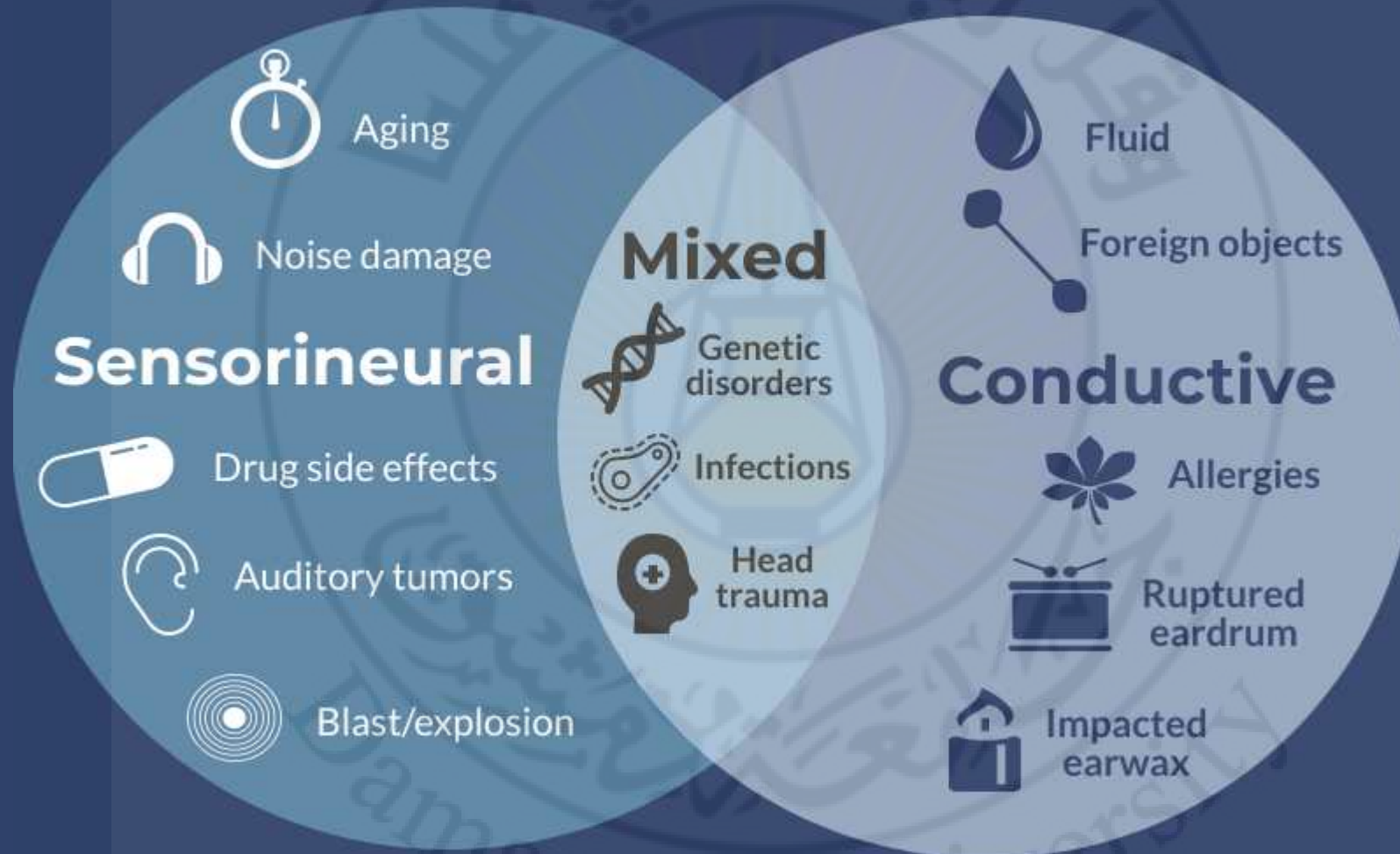


Mixed Hearing Loss

- When both a sensorineural and a conductive loss are present, the result is termed a mixed hearing loss.
- The air conduction and bone conduction threshold differences generally disappear when the conductive portion of the hearing loss is ameliorated.
- The sensorineural part of the mixed hearing loss will typically improve to the level of the previously established bone conduction threshold level.
- In mixed hearing loss, the hearing levels are not likely to return to normal limits due to the presence of sensorineural hearing impairment.

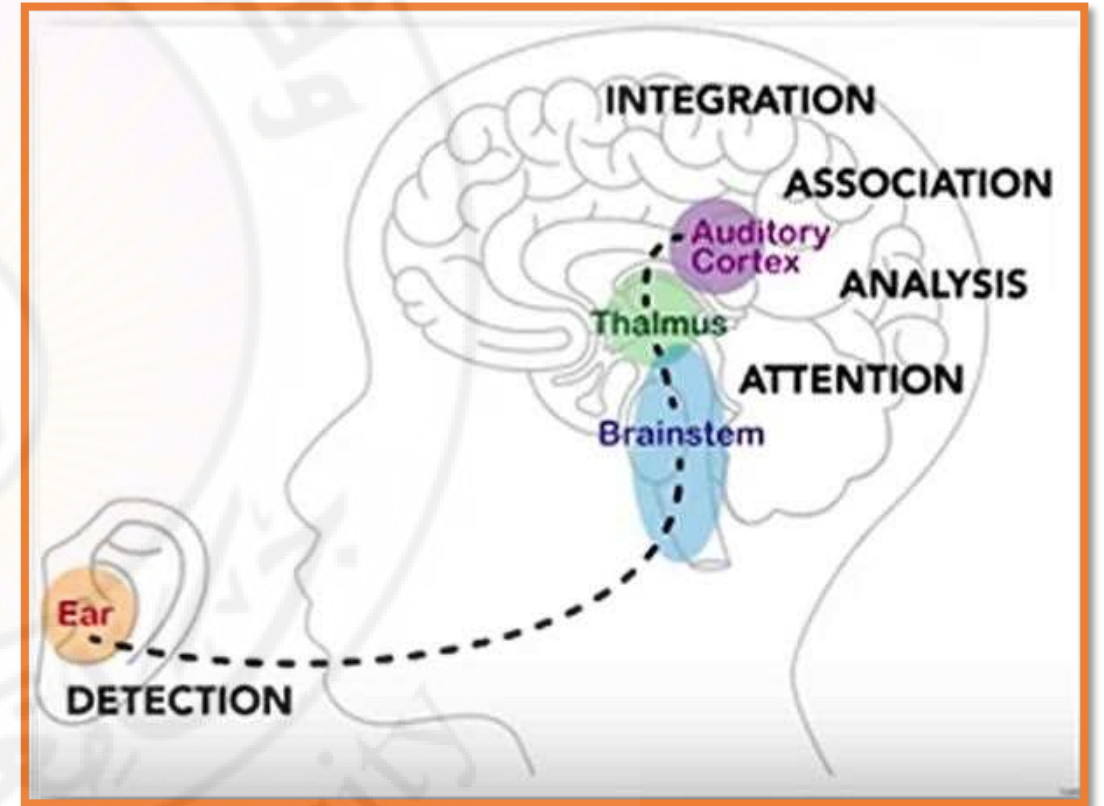


Types of hearing loss



Central Auditory Processing Disorder

- Central auditory processing disorders (CAPDs) refer to difficulties in the processing of auditory information in the central nervous system.
- This deficit is a problem that is likely to be exacerbated in unfavorable acoustic environments as in situations with competing noise background. CAPD may be associated with difficulties in listening, speech understanding, language development, and learning.
- Child may have a normal audiogram but be unable to comprehend complex speech, which in turn, may lead to learning disabilities.



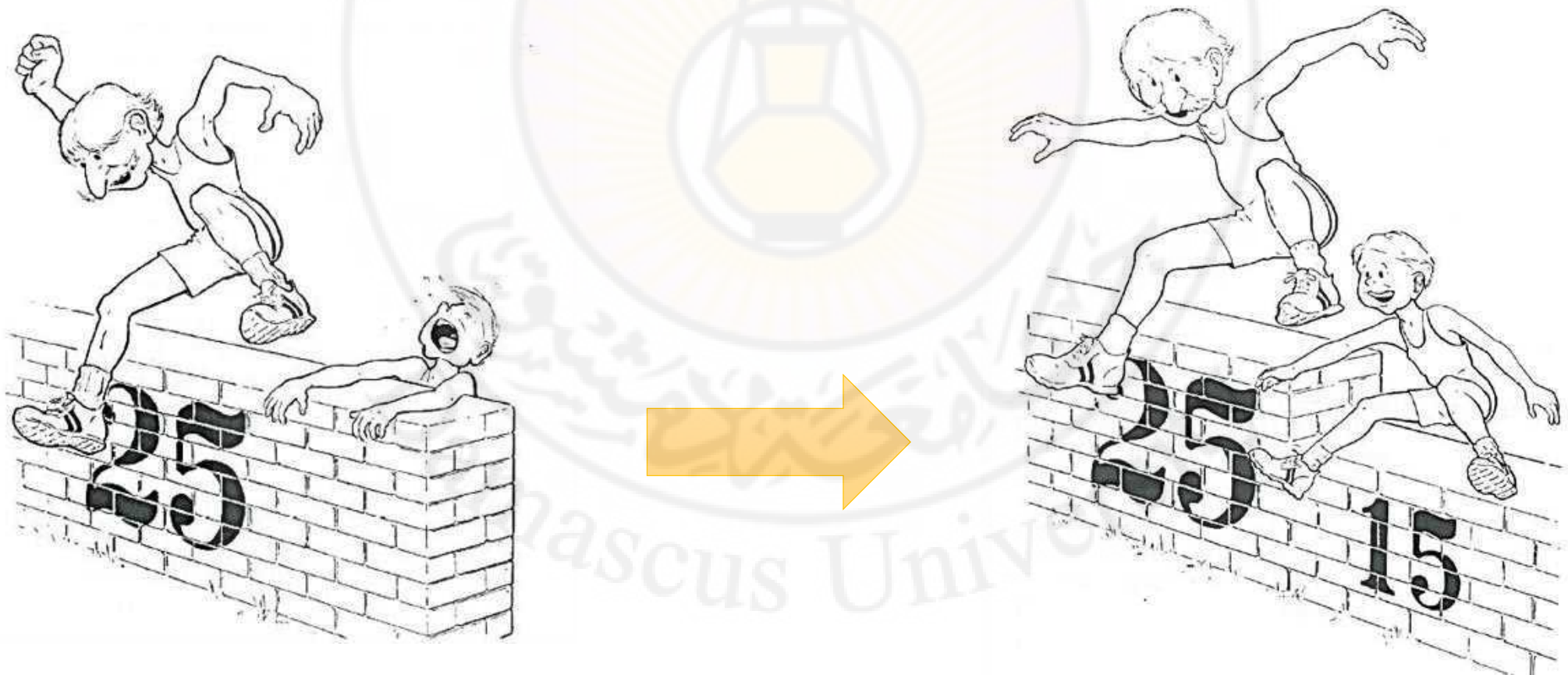
Degree and Severity of Hearing Loss

- complete description of a child's hearing loss should include whether it is **unilateral or bilateral**, the **degree** of auditory impairment, and the **type of loss** (conductive, sensorineural, or mixed).
- *A significant hearing loss in a child is any degree of hearing that reduces the intelligibility of speech to a degree inadequate for accurate interpretation or as to interfere with learning.*

Degree of hearing loss	Hearing loss range (dB HL)
Normal	-10 to 15
Slight	16 to 25
Mild	26 to 40
Moderate	41 to 55
Moderately severe	56 to 70
Severe	71 to 90
Profound	91+

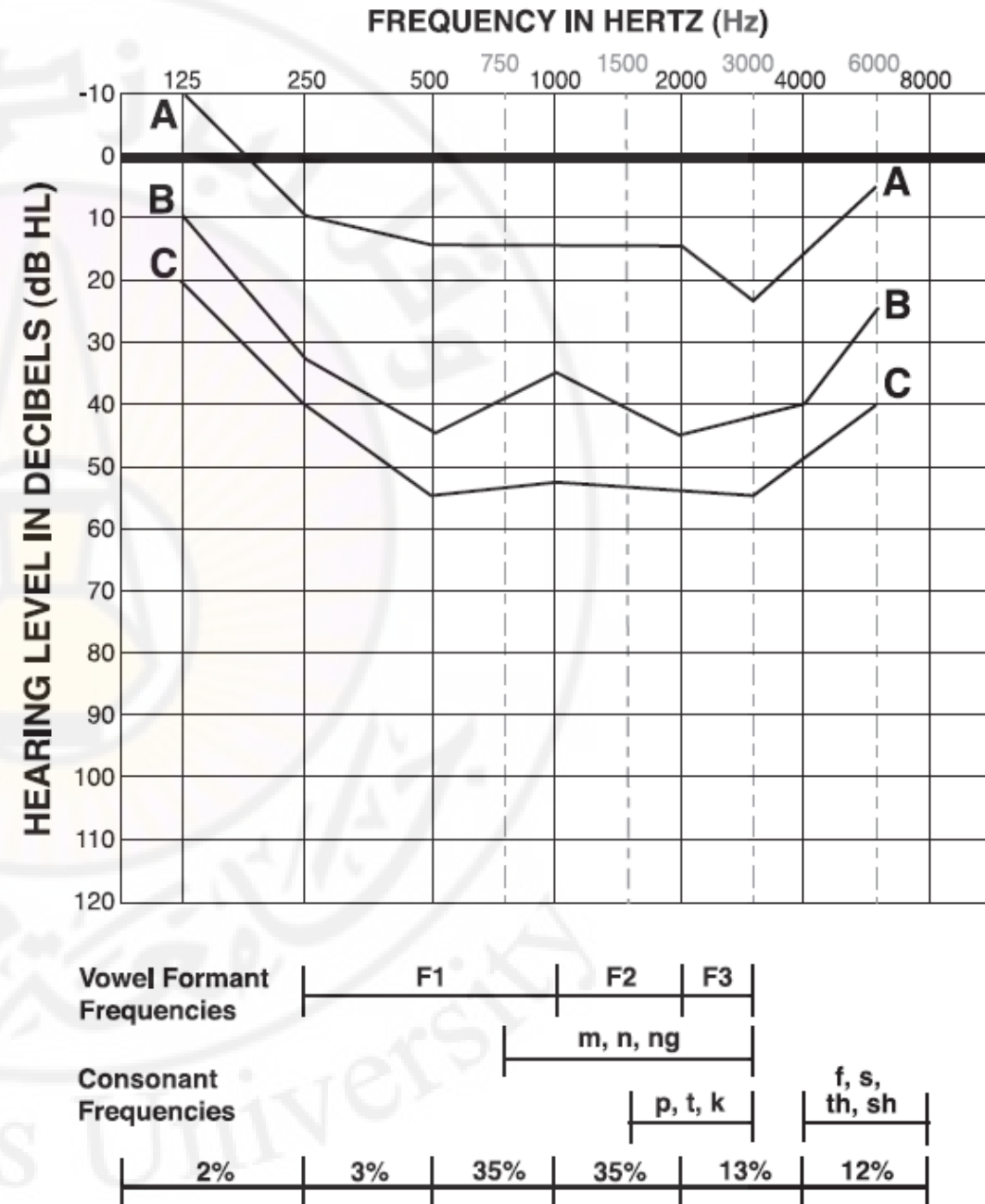
Minimal Hearing Loss in Children

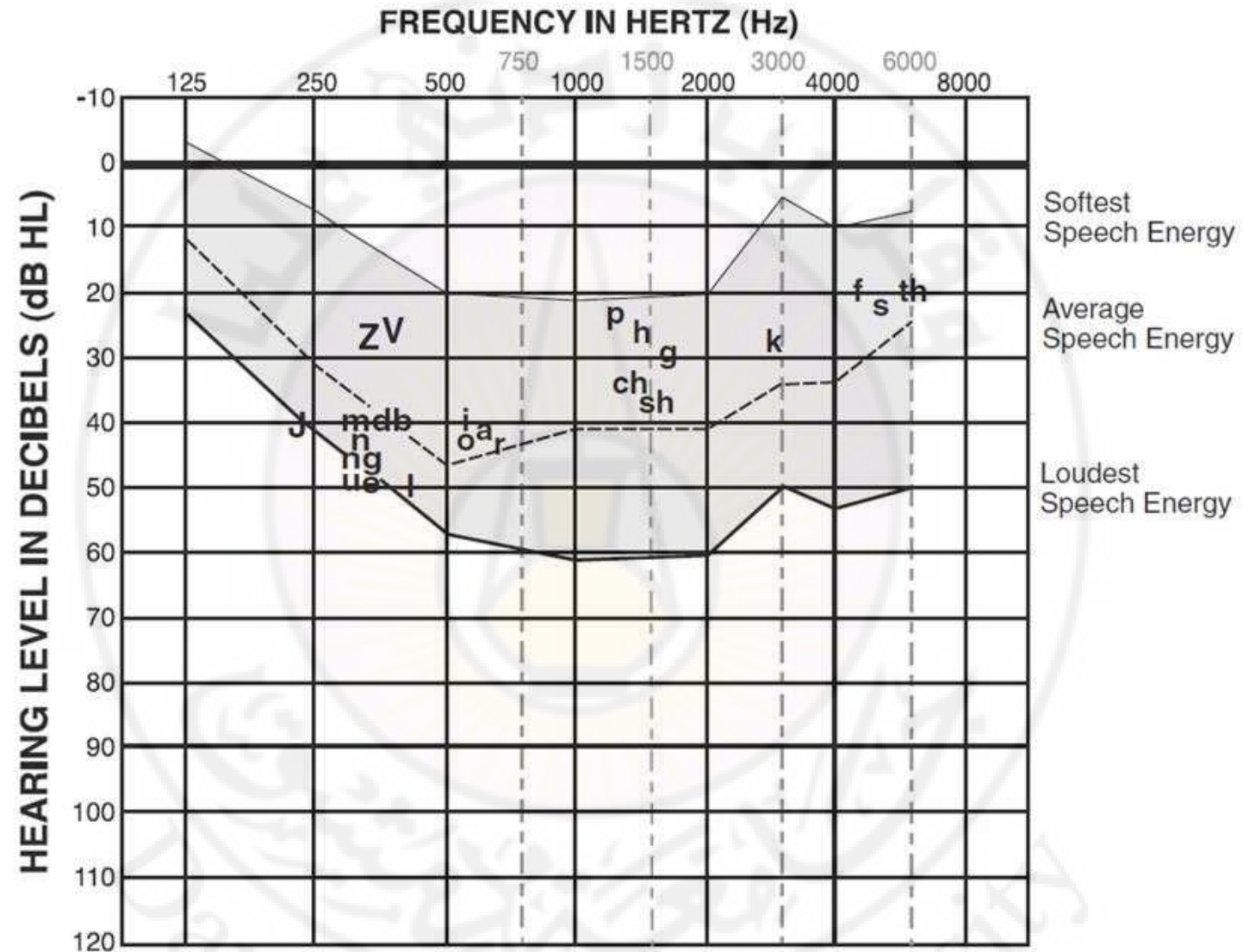
- **15 dB HL** should be considered the lower limit of normal hearing for children and that impairment increases with each decibel of hearing loss greater than 15 dB HL.



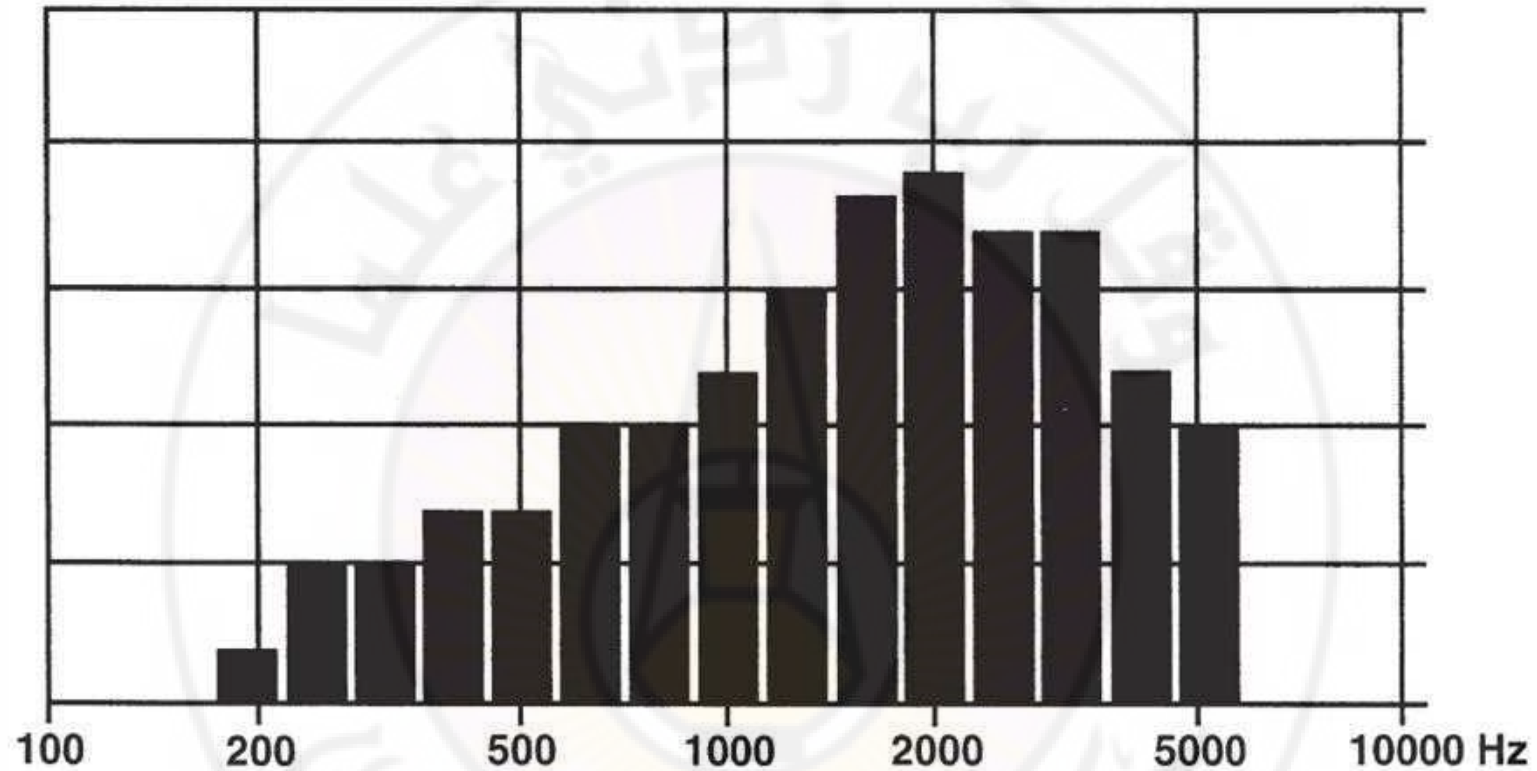
The area between curves A and B represents 80% of normal conversational speech levels. As hearing thresholds fall below curve B, difficulty understanding speech or even isolated words spoken in quiet surroundings are expected.

Conversational speech exceeds curve A 90% of the time, speech exceeds curve B only 50% of the time, and curve C merely 10% of the time.





Average range of speech energy in decibel hearing level (dB HL)



Graph showing the relative importance of individual speech bands for speech intelligibility. nearly 70% of word recognition is determined by speech energy between 500 and 2000 Hz. An additional 25% of word recognition energy occurs by energy falling above 2000 Hz

Effect of Hearing Loss on Speech and Language

- The effects of a hearing loss depend on its:
 - severity;
 - configuration;
 - duration, and stability;
 - The age of the person at onset;
 - The extent and type of early training;
 - The type and timing of amplification;
 - visual, emotional, intellectual and cultural factors;
 - Family support
- Age at identification and intervention are especially important factors in language development. A child who sustains significant hearing loss after acquiring language (3 or 4 years of age) will have a less severe linguistic deficit than the child whose hearing loss is present at birth or develops within the first few months of life.



Effect of Hearing Loss on Speech and Language

Lack of Constancy of Auditory Clues When Acoustic Information Fluctuates.

When a child does not hear speech sounds in the same way from one time to another, there is confusion in abstracting the meanings of words due to inconsistent categorization of speech sounds.

Effect of Hearing Loss on Speech and Language

Confusion of Acoustic Parameters in Rapid Speech.

Even the normal hearing child suffers from variations of speech occurring among speakers and even in the same speaker. Frequency, duration, and intensity vary as a result of differences among speakers of different ages, genders, and personality types. The child with hearing loss will be confused in language learning as a result.

Effect of Hearing Loss on Speech and Language

Confusion in Segmentation and Prosody.

The child with hearing loss may miss linguistic boundaries such as plurals, tenses, intonation, and stress patterns. These factors are requisite to meaningful interpretation of speech.

Effect of Hearing Loss on Speech and Language

Masking of Ambient Noise.

Children with hearing loss who are in classroom settings need a signal-to-noise ratio of at least +15 dB; that is, the speech signal must be 15 dB louder than the background noise for learning to take place. Unfortunately, it is rare in our modern, open, school classrooms for this preferred listening ratio to be present. Obviously, a child with even a minimal hearing loss is significantly handicapped under such poor listening situations.

Effect of Hearing Loss on Speech and Language

Breakdown of Early Ability to Perceive Speech Sounds.

The infant begins to learn to discriminate speech sounds almost immediately after birth. Studies have shown that at between 1 and 4 months of age an infant can discriminate between most of the English speech sound pairs. By 6 months of age, the infant recognizes many of the speech sounds of language and is making ongoing cataloging of speech sounds. If the sounds of speech are not perceived early, due to the presence of a hearing loss, learning can be impeded.

Effect of Hearing Loss on Speech and Language

Breakdown in Early Perceptions of Meanings.

During ordinary speech, the normal listener often misses some unstressed or elided words or sounds but is able to fill in by understanding the context of the message. However, when a hearing loss causes a young child to miss many of these soft or inaudible sounds, there is confusion in word naming, difficulty in developing classes of objects, and misunderstanding of multiple meanings.

Effect of Hearing Loss on Speech and Language

Faulty Abstraction of Grammatical Rules.

When short words are soft or elided, as they often are, it becomes more difficult for a hearing-impaired child to identify the relationships between words and to understand word order.

Effect of Hearing Loss on Speech and Language

Subtle Stress Patterns Missing.

The emotional content of speech, its rhythm, and its intonation are communicated through the low frequencies. When there is low-frequency hearing loss, the emotional content of speech is confused — another condition that impairs the learning of speech and language.

Effects of Hearing Loss

- undetected hearing loss of any degree can have a significant impact on speech, language, cognitive, and psychosocial development.
- We not only need to be concerned with children with moderate to profound hearing loss, the “minimal hearing loss” group also needs our attention.
- The impact of **bilateral mild** and **unilateral hearing loss** on the development of children has only recently been recognized by educators and health care providers. Many children are being identified with unilateral hearing loss from birth, and early bilateral mild hearing loss that previously had not been detected prior to school hearing screening.
- It is no longer reasonable to accept that children with mild and unilateral hearing losses are not at risk for speech and language delays, academic failure, and poor self-esteem. These children, in fact, have difficulties with speech and language development resulting in lower scores in school achievement and more behavioral problems than their peers with normal hearing

Handicapping Effects of Hearing Loss

Average Hearing Level (500–2000 Hz)	Description	Possible Condition	What Can Be Heard Without Amplification	Handicapping Effects (If Not Treated in First Year of Life)	Probable Needs
0–15 dB	Normal range	Conductive hearing losses	All speech sounds	None	None
15–25 dB	Slight hearing loss	Conductive hearing losses, some sensorineural hearing losses	Vowel sounds heard clearly; may miss unvoiced consonant sounds	Mild auditory dysfunction in language learning	Consideration of need for hearing aid; speech-reading, auditory training, speech therapy, preferential seating
25–30 dB	Mild hearing loss	Conductive or sensorineural hearing loss	Only some of speech sounds, the louder voiced sounds	Auditory learning dysfunction, mild language retardation, mild speech problems, inattention	Hearing aid, speechreading, auditory training, speech therapy
30–50 dB	Moderate hearing loss	Conductive hearing loss from chronic middle ear disorders; sensorineural hearing losses	Almost no speech sounds at normal conversational level	Speech problems, language retardation, learning dysfunction, inattention	All of the above plus consideration of special classroom situation

Handicapping Effects of Hearing Loss

Average Hearing Level (500–2000 Hz)	Description	Possible Condition	What Can Be Heard Without Amplification	Handicapping Effects (If Not Treated in First Year of Life)	Probable Needs
50–70 dB	Severe hearing loss	Sensorineural or mixed losses due to a combination of middle ear disease and sensorineural involvement	No speech sounds at normal conversational level	Severe speech problems, language retardation, learning dysfunction, inattention	All of the above plus probable assignment to special classes
70+ dB	Profound hearing loss	Sensorineural or mixed losses due to a combination of middle ear disease and sensorineural involvement	No speech or other sounds	Severe speech problems, language retardation, learning dysfunction, inattention	All of the above plus probable assignment to special classes



World Health
Organization

32

MILLION

CHILDREN LIVE WITH
DISABLING HEARING LOSS

60%

OF CHILDHOOD
HEARING LOSS IS
PREVENTABLE

INFECTIONS
31%



Mumps



Rubella



Meningitis



Measles



Ear
infections

OTHERS*
! 8%



OTOTOXIC
MEDICINES
4%

17%

BIRTH-RELATED CAUSES



Complications
at birth



Prematurity



Low birth
weight

*including prenatal causes, such as substance abuse and metabolic diseases

CONSEQUENCES, IF NOT ADDRESSED



World Health
Organization

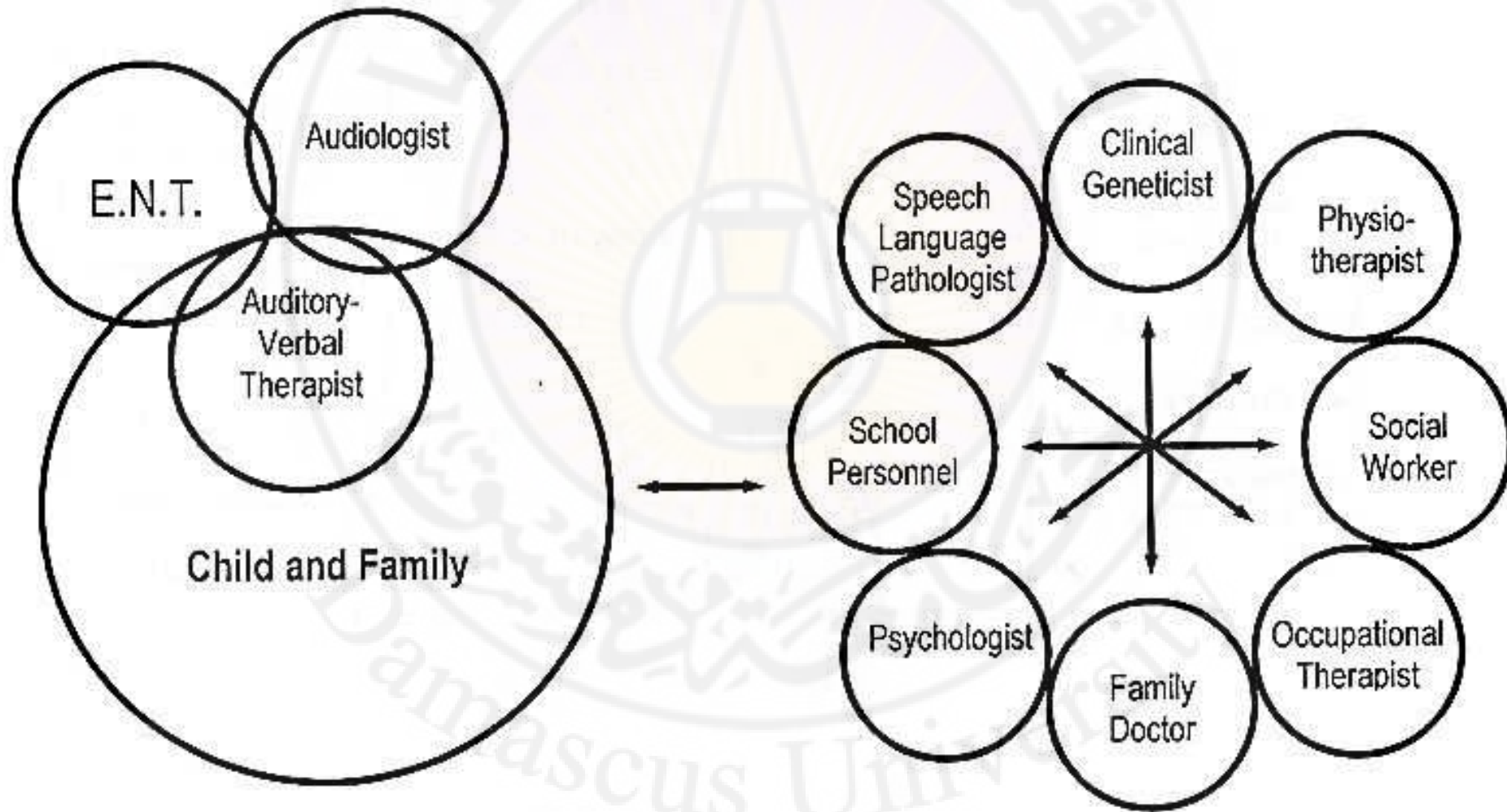


Economic Burden of Hearing Loss and Deafness

- Medical and Audiologic Expenses
- Education and Training Expenses
- Special Living Expenses
- Lifetime Loss of Income Considerations
- Additional Intangible Costs of Hearing Loss and Deafness



Team Management of Children With Hearing Loss



3. Auditory and Speech-Language development

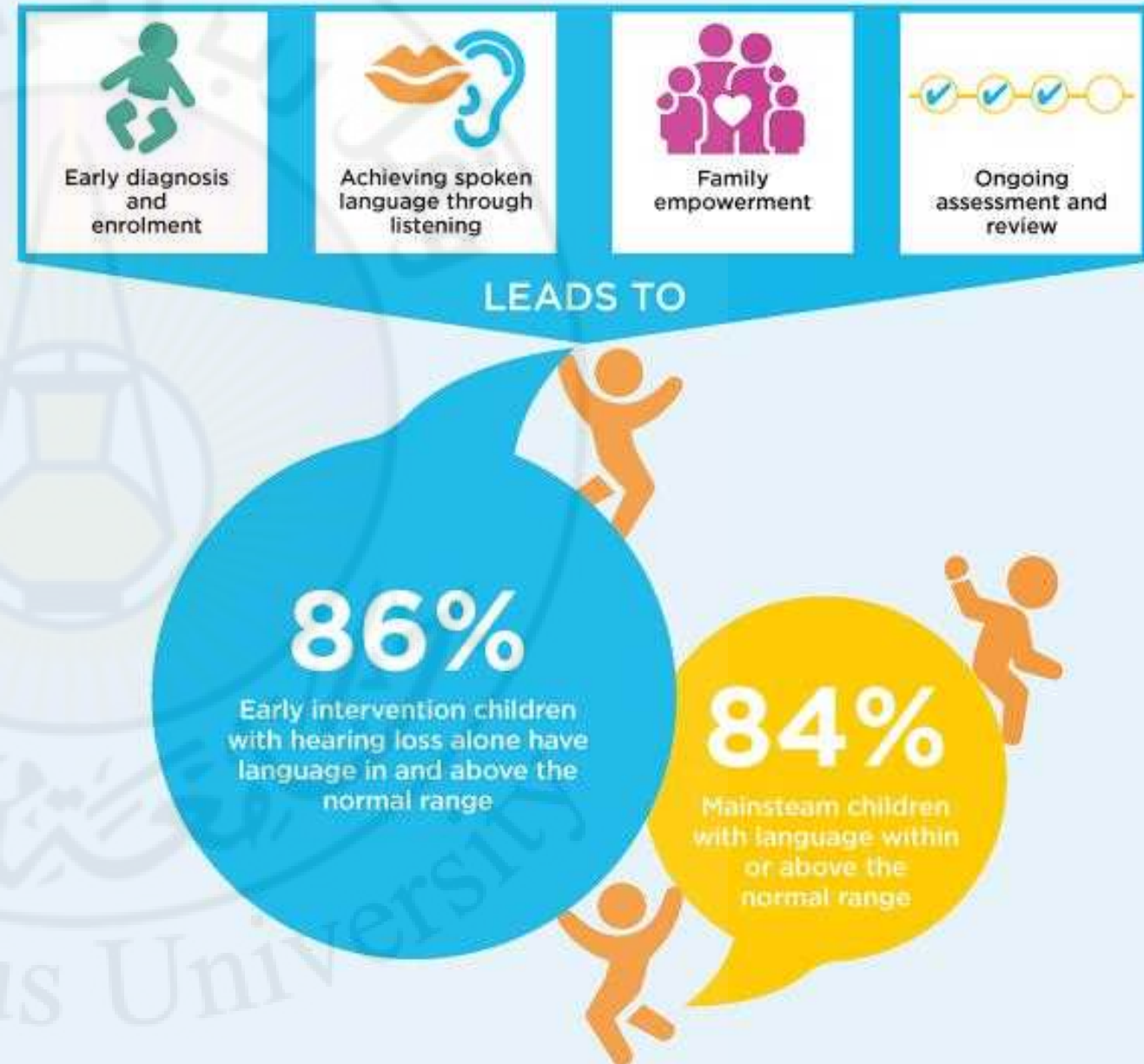
Acquisition of language is highly dependent on an adequately functioning auditory system

- One of the most important aspects in any child's development is the acquisition and production of spoken language.
- Spoken language is the doorway to successful communication and the social interaction that is so important to daily life.
- Language is the key to the doors by which we express our thoughts, needs, and feelings and by which we receive and comprehend the thoughts, needs, and feelings of others.
- Although it is language in a child that opens the door to education, the successful acquisition of language is highly dependent on an adequately functioning auditory system



OUR EARLY INTERVENTION WORKS

- The child with hearing loss does not automatically develop speech.
- The child with hearing loss may be confronted with a life of language difficulties and educational struggles.
- Language develops so rapidly in the first few months of life that the longer an infant's hearing loss goes undetected, the worse the outcome is likely to be; thus, we realize the importance of **early intervention**.



Hearing Development

- It is important for us to know that the auditory development and maturation of a normal-hearing baby follow a standard sequence of behaviors from birth to 12 months that is as regular as clockwork.
- The experiences of sounds and exposure to speech shape the auditory system of infants during their first year of life.
- Although infants are unable to produce recognizable words until about 12 months of age, they quickly develop a remarkable ability to distinguish auditorily among speech sounds.
- Newborns quickly learn to recognize words, phrases, vocal intent, rhythm, and names and to perform sophisticated auditory functions long before they produce their own speech.



Prenatal Hearing

- The auditory system in the fetus is under structural development during the first 20 weeks of gestation, followed by the initial neurosensory maturation.
- Thus, the auditory system becomes functional around **25 weeks' gestation** when the ganglion cells of the spiral nucleus in the cochlea connect inner hair cells to the brainstem and temporal lobe of the cortex.
- Elliot and Elliot (1964) confirmed that the human cochlea shows response to sound after the 20th week of gestation.



Prenatal Hearing

- Bench (1968) reported that for a 72 dB SPL signal there is the least attenuation of sound going into the uterus at 200 Hz (19 dB), slightly more at 500 Hz (24 dB), more at 1000 Hz (38 dB), and the most at 2000 and 4000 Hz (48 dB).
- The frequencies below 1000 Hz contained the frequency response of the maternal voice that may be heard by the fetus from the **fourth month of gestation**.



An illustration of a fetus in the womb, shown in a curled position. The fetus is rendered in shades of red and pink, contrasting with the light pink background of the womb. The womb is depicted as a large, rounded structure within a dark blue silhouette of a pregnant woman. The overall style is simple and illustrative.

Fetus can hear the mother's voice and other voices that were perfectly audible but lacking in tone because the high frequencies were absorbed.

When there is no fetal distress, the fetus reacts to the sound stimulus by a change in heart rate, often associated with movement.

Prenatal Hearing In summary:

- Perceptual learning of the more global, prosodic aspects of language actually commences prior to birth.
- Studies using the sucking and heart rate paradigms show that exposure to sound in utero resulted in a preference of newborn infants for **native-language** over foreign language utterances, for the **mother's voice** over another female's voice, and for **simple stories the mother** read during the last trimester over unfamiliar stories.
- This indicates that the prosodic aspects of human speech, including voice pitch and the stress and intonation characteristics of a particular language and speaker, are transmitted to the fetus and are learnable.
- Understanding the nature of this early phonetic learning and the mechanisms underlying it is one of the key issues in human language development.



Neonatal Hearing Development



The baby will get a shock when he hears a loud sound



Your little one starts imitating sounds like baa, aah, mum etc.



The baby also starts following simple requests of bye-bye, hi



Listens to songs and stories but for a brief time.

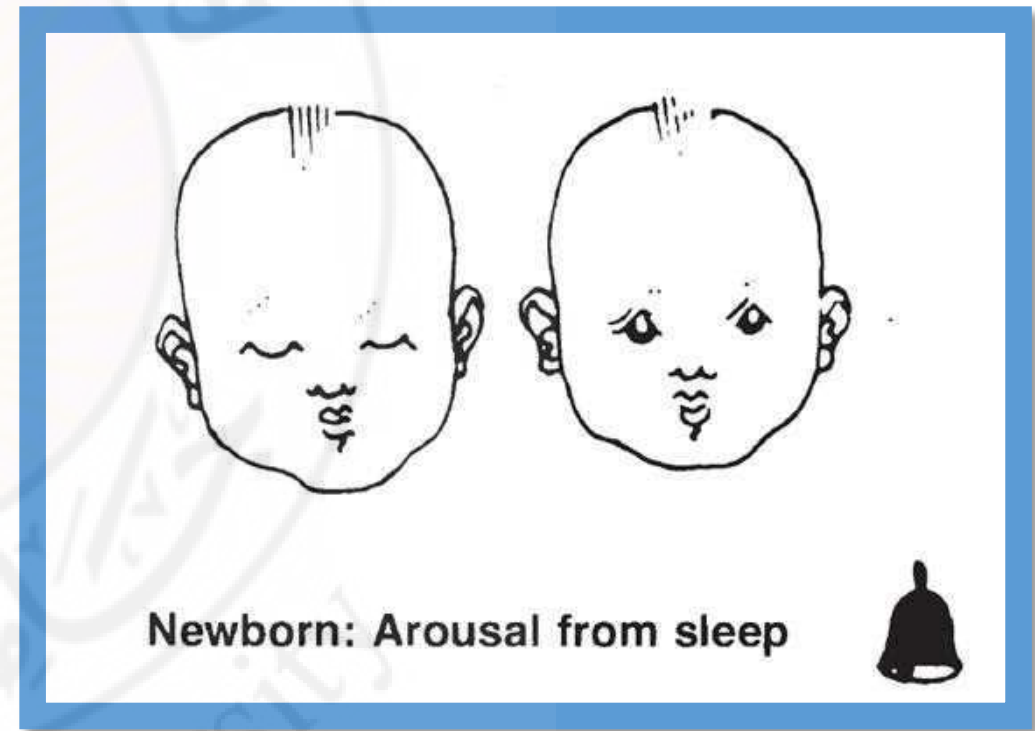
Neonatal Hearing Development

- The normal-hearing, alert infant will respond in the predictable manner in accordance with his or her **mental age**.
- These auditory maturation responses are so reliable that in skillful hands, they can serve as an initial orientation for the audiologist as to what to expect in the more formal hearing evaluation.
- In fact, these auditory development responses are very **age specific**, depending on the maturation of the infant.



Newborn

- if an unexpected noise of 70 dB SPL reaches maximum intensity within a few milliseconds, a newborn infant closes his or her eyes, startles, and shows an increase in heart rate.
- If the same sound reaches its maximal intensity in 2 seconds, the infant opens his or her eyes, looks around, and is likely to show a decrease in heart rate.
- The first reaction is a defensive one; the latter displays interest in the changing environment.
- This easiest auditory reflex response to elicit and observe is **the startle, or Moro, response**.



WHAT IS THE
STARTLE REFLEX?

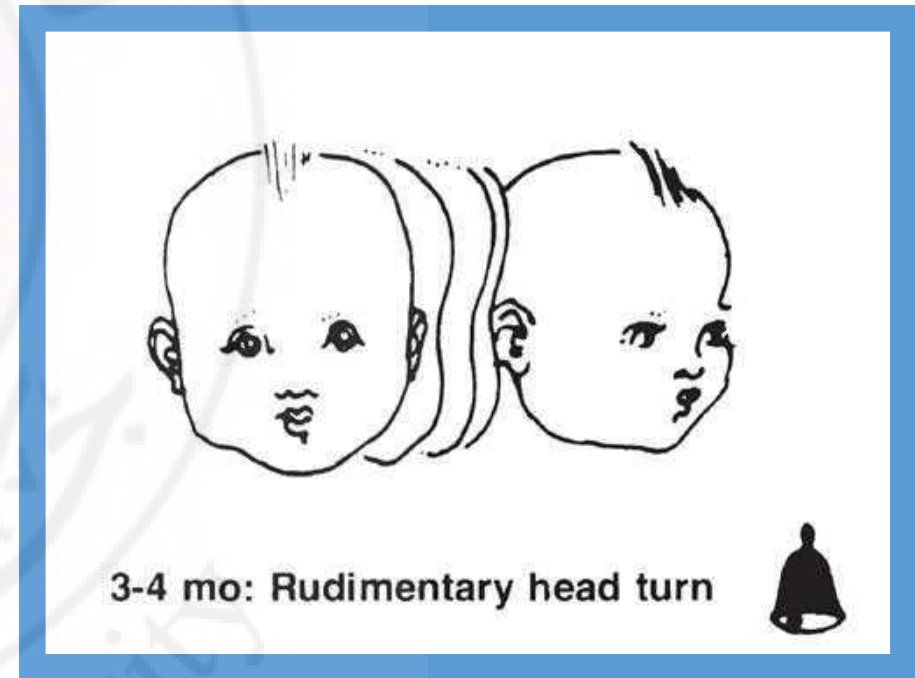
Startle, or Moro, response





Birth to Four Months of Age

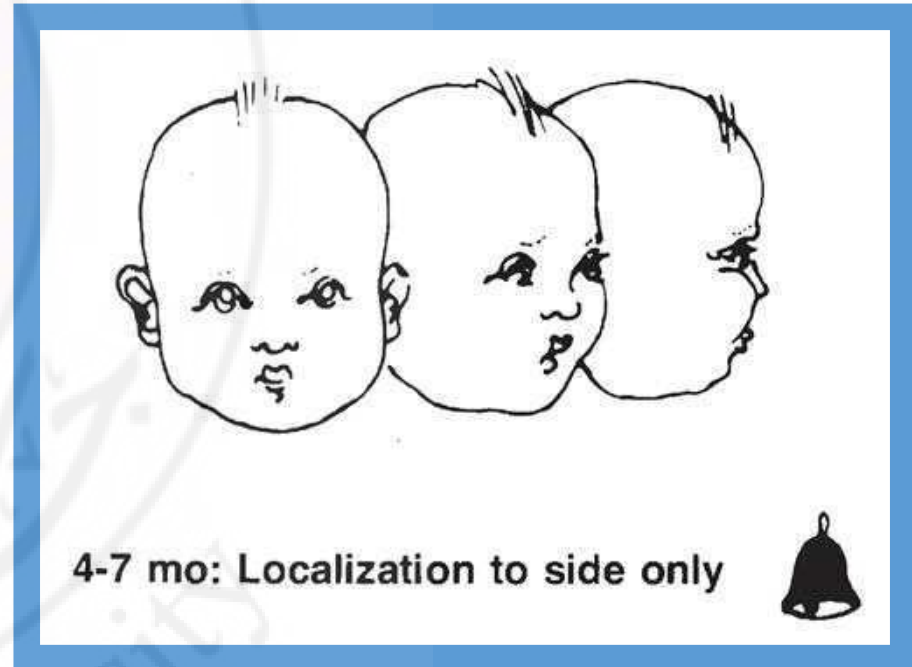
- At this early age, babies may **turn toward their mother's** voice and even **smile** when they hear a new voice. They may be observed suddenly **quieting**, or becoming **still**, at the presentation of a new sound if they are **crying or fussing**.
- However, in general, infant auditory responses are limited and largely reflexive. In a very quiet environment one may only see an eye-blink or eye-widening response to soft sounds from noisemakers or other subtle auditory signals.
- The only reliable behavioral (but reflexive) auditory response at this early age is the Moro startle response or the “surprise” eyeblink to louder sounds.





Four to Seven Months of Age

- By 4 months of age auditory development is noted as the infant begins to turn his or her head toward the sound source in a more consistent, but still slow and wobbly manner.
- By 7 months, the infant's neck muscles will be strong enough to permit a stronger and more direct turn on a **lateral plane** toward the side from which the sound is presented.
- The head turn may not be a direct localization to sounds presented at lower plane levels. When the stimulus sound is presented at a lower level beneath the eyes of the baby, the response may be initially a turn to the appropriate side of the sound and then a slow search downward for the source.





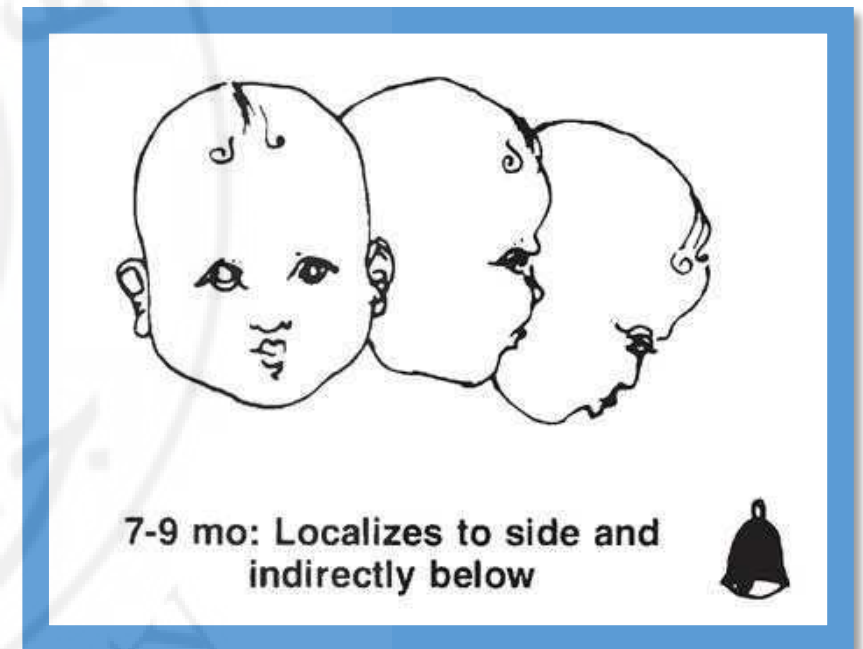
Jyväskylä University



Damascus University

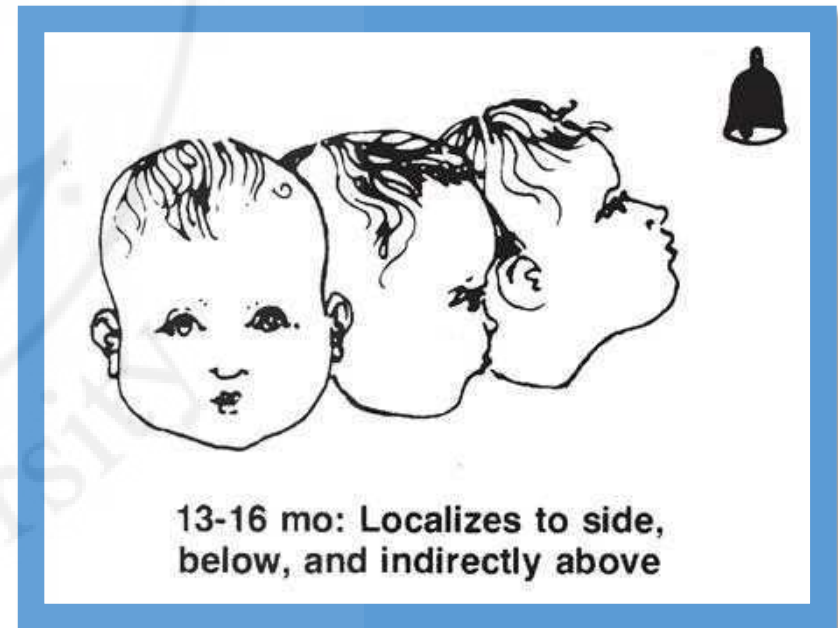
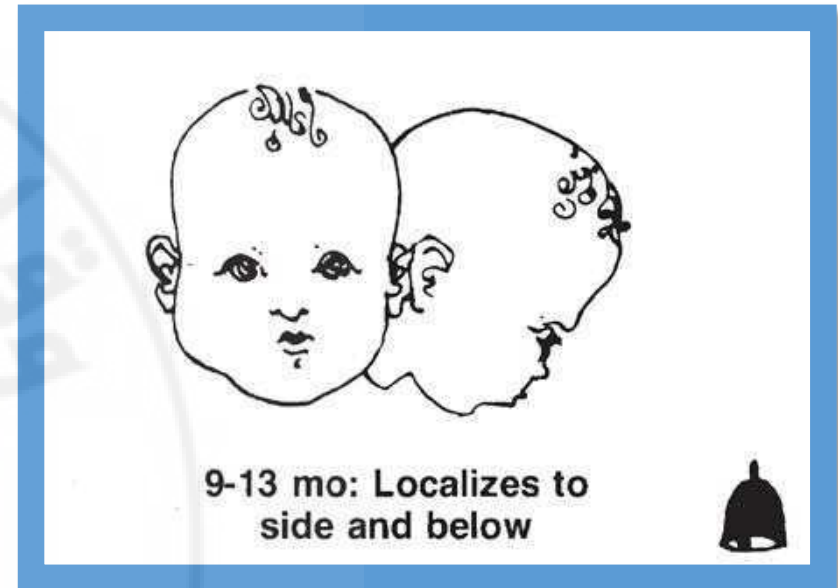
Seven to Nine Months of Age

- Between 7 and 9 months the infant develops the ability to identify the precise location of the sound source with a direct head turn, either on a lateral plane or to a lower level.
- By this age the localization response is brisk and firm. Sound stimuli presented below the child should draw a direct localization down to the sound source.
- However, it is likely that the infant will not yet look directly at a sound on a higher plane (i.e., above eye level), as the ability to localize sound above the head is not demonstrated for another month or two.
- It is a bit remarkable that although the infant can localize sounds to the side or below with briskness and accuracy, they seem unable to localize sounds presented above and over their heads.



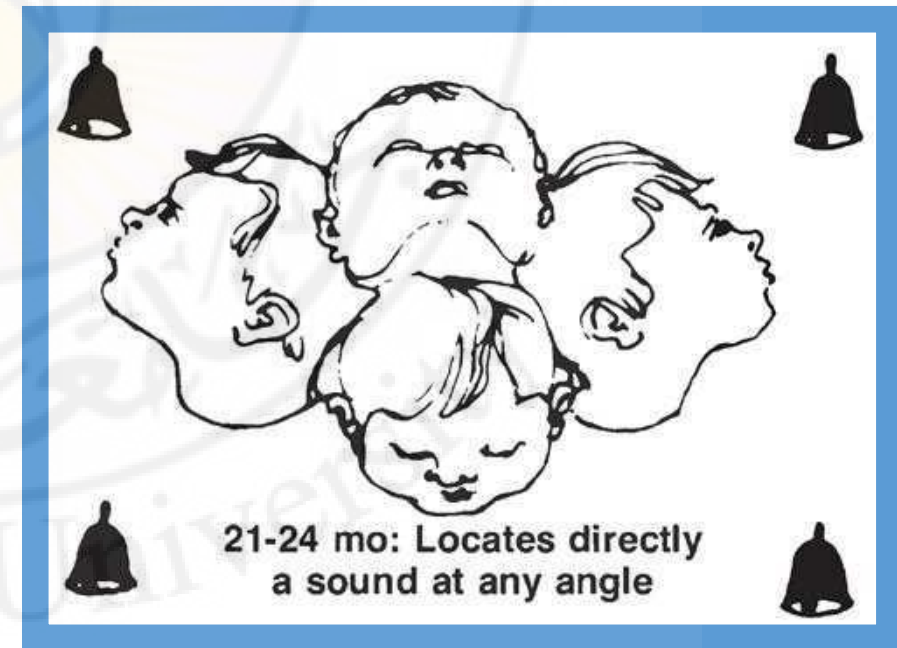
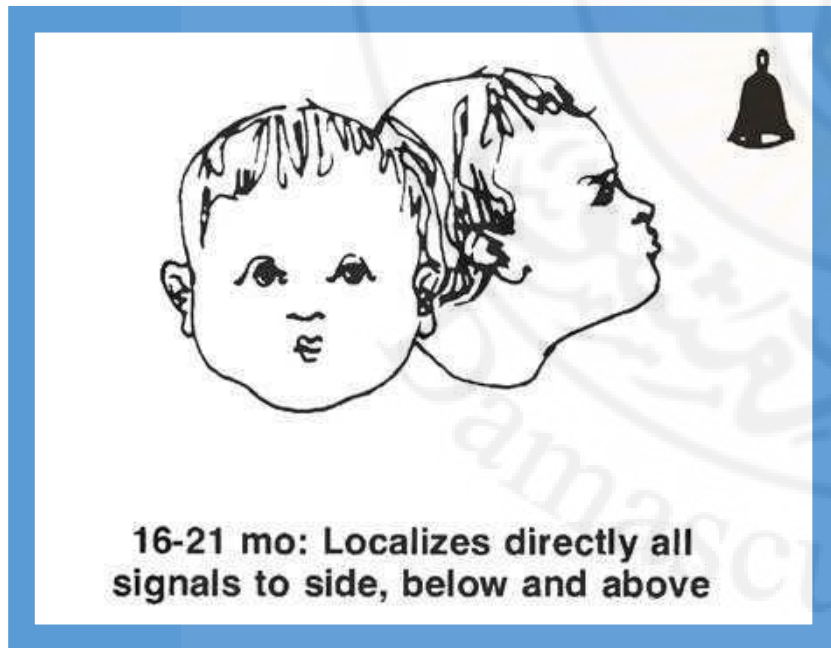
Nine to Thirteen Months of Age

- Certainly, by 12 to 13 months of age, the normal-hearing infant is able to localize sounds briskly and directly in any plane to the side, above, or below eye level.
- Usually, by 12 months of age, the curiosity of the child is full blown and one will see quick localizations to appropriately presented auditory stimulus which will be noted by children with bilaterally normal hearing.
- Full maturation of the child's auditory development has usually been attained by this age.



Thirteen Months and Older

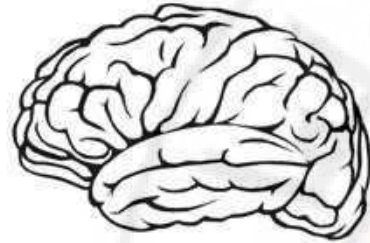
- It is generally easy for the pediatric audiologist to elicit and observe these nonconditioned localizing auditory responses in this older age group.
- For this older age group, reinforced behavioral response techniques such as visual response audiometry or conditioned orientation response audiometry are the required audiologic testing methods.



Development of Oral Communication



Receptive Language



- Receptive language is the ability to understand
- Listening and reading skills are examples of receptive language
- Listens and responds when spoken to
- Following directions
- Understanding the words, sentences, and meaning of what others say or what is read

Expressive Language



- Expressive language is the ability to communicate
- Speaking, gesturing (waving), writing, facial expressions, vocalizations (crying) are all variations of expressive language
- Labeling objects, retelling a story
- Expressive language can be more difficult to develop than receptive skills

Developmental Speech Milestones



The Newborn

- The most common sounds made by the newborn are cries and vowels.
- By the end of the first month, the cries take on meaning and mothers can differentiate anger and pain from tiredness.
- The noncrying sounds include normal phonation but lack resonance.
- Near the end of the first month the infant begins to initiate sounds characterized as “coos” and “gurgles.”
- The newborn can discriminate between different phonemes and different intonational and stress patterns, but this auditory discrimination does not involve sound-meaning relationships.

Two to Three Months of Age

- By 2 months of age the infant has developed muscle control to stop and start oral movements and vocalizations. The infant seems to focus on the production of vowel-like sounds such as “ooh” and “aah.” By 3 months, babbling and laughter are likely to begin.

Four to Six Months of Age

- At this stage, the infant is into strings of sound known as *true babbling* and begins to respond to noisemaking toys.
- There is definitely more control of the tongue.
- The infant may spend long periods experimenting and listening to his or her own vocalizations.
- At about 5 months of age, the infant vocal emissions include consonant-vowel combinations.
- Glottal and labial sounds are heard from the infant by the age of 6 months.

Six to Ten Months

- Babbling slowly decreases at approximately 6 months of age, and during the next few months there is undistinguished progress in vocalizing speech sounds.
- The speech unit becomes the consonant-vowel. During this period, the mother's feedback of the child's sounds provides the groundwork for the first production of words — yet some 6 months in the future.
- As the infant gains increasing control of oral movements and vocalizations, speech production progresses to repetitive syllables such as “bababa” and “dadada.”
- Menyuk (1972) showed that the frequency of consonant appearance in babbling is reflected in the order of speech sound acquisition. With increasing age and vocal experience, the baby's babbling increasingly reflects adult speech in syllable structure and intonation. As mother (or father) imitates the sounds of the infant, and in turn begins to add additional speech improvising, the baby soon begins to imitate the parent's improvisations; the shaping of the infant's speech patterns is underway.
- Comprehension of the sound sequence may precede the imitation; sometimes imitation precedes understanding of the meaning of the sound sequence which the parents learn to associate with specific facial expressions or emotions. By now the infant responds to its own name, plays with voice repetition, understands “no” and “bye-bye,” may be able to repeat “ma-ma” or “da-da,” and listens attentively to music or singing.

Eleven to Eighteen Months

- The child's first meaningful word is usually uttered around the first birthday and is dependent on a full year of attentive listening activities.
- Soon after utterance of the initial word, the child begins to rapidly build a vocabulary.
- By 12 to 14 months the child can follow simple directions; use two to three words in a meaningful phrase; may correctly identify people, body parts, and toys by name; bounce in rhythm to music; and attempt to imitate words.
- By 18 months the child should have at least a 6- to 10-word vocabulary.

Quick Checklist for Speech-Language- Hearing Milestones

Birth to 3 Months

- Startles to loud noises
- Calms to familiar voices
- Makes vowel sounds “ooh” and “ahh”

3 to 6 Months

- Makes a variety of sounds “ba-ba” and “ga-ba”
- Enjoys babbling
- Likes sound-making toys
- Changes voice pitch at will
- Turns eyes and head toward sound

6 to 9 Months

- Responds to own name
- Imitates speech with nonspeech sounds
- Plays with voice repetition, “la-la-la-la”
- Understands “no” and “bye-bye”
- Says “da-da” or “ma-ma”
- Listens attentively to music and singing

9 to 12 Months

- Responds differently to happy or angry talking
- Turns head quickly toward loud or soft sounds
- Jabbers in response to human voice
- Uses two or three simple words correctly
- Gives up toys when asked
- Stops in response to “no”
- Follows simple directions

12 to 18 Months

- Identifies people, body parts, and toys on request
- Turns head briskly to source of sound in all directions
- Can tell you what he or she wants
- Talks in what sounds like sentences
- Gestures with speech appropriately
- Bounces in rhythm with music
- Repeats some words that you say

18 to 24 Months

- Follows simple commands
- Speaks in understandable two-word phrases
- Recognizes sounds in the environment
- Has a vocabulary of 20 words or more

2 Months of Age

Hearing

Have you had any worry about your child's hearing?

☐ Yes ☐ No

When your child is sleeping, does he^a move and begin to wake up to a loud sound?

☐ Yes ☐ No

Development and communication

Does he lift up his head when he is lying on his stomach?

☐ Yes ☐ No

Does your child smile at you when you are looking face to face?

☐ Yes ☐ No

Does he move both hands together in the same way?

☐ Yes ☐ No

Does he look at your face without you making gestures to him?

☐ Yes ☐ No

4 Months of Age

Hearing

Have you had any worry about your child's hearing?

☐ Yes ☐ No

When your child is sleeping, does he move and begin to wake up to a loud sound?

☐ Yes ☐ No

Does he try to turn his head toward an interesting sound or when his name is called?

☐ Yes ☐ No

Development and communication

Does he lift his head up to 90° and look straight ahead?

☐ Yes ☐ No

Does your child touch his hands together and play with them?

☐ Yes ☐ No

Does he laugh and giggle without being tickled or touched?

☐ Yes ☐ No

Does he coo to himself and make noises when he is alone?

☐ Yes ☐ No

6 Months of Age

Hearing

Have you had any worry about your child's hearing?

☐ Yes ☐ No

When your child is sleeping, does he move and begin to wake up to a loud sound?

☐ Yes ☐ No

Does he try to turn his head toward an interesting sound or when his name is called?

☐ Yes ☐ No

Development and communication

Does he lift his head and chest with his arms?

☐ Yes ☐ No

Does he keep his head steady when sitting? Does he roll over in his crib?

☐ Yes ☐ No

Does he reach for objects within his reach and hold them?

☐ Yes ☐ No

Does he see small objects like peas or raisins?

☐ Yes ☐ No

8 Months of Age

Hearing

Have you had any worry about your child's hearing? ☐ Yes ☐ No

When your child is sleeping, does he move and begin to wake up to a loud sound? ☐ Yes ☐ No

Does he try to turn his head toward an interesting sound or when his name is called? ☐ Yes ☐ No

Does he enjoy ringing a bell, playing with a noisy toy, or shaking a rattle? ☐ Yes ☐ No

Development and communication

Does he support most of his weight on his legs? ☐ Yes ☐ No

Can he sit alone without help for 5 minutes? ☐ Yes ☐ No

Can he sit and look for objects that have fallen out of sight? ☐ Yes ☐ No

Can he pick up two objects, one in each hand? ☐ Yes ☐ No

Can he transfer an object from one hand to the other? ☐ Yes ☐ No

Can he feed himself a cracker? ☐ Yes ☐ No

Does he make a number of different sounds and change their pitch? ☐ Yes ☐ No

Does he clap his hands in imitation and make noises at the same time? ☐ Yes ☐ No

10 Months of Age

Hearing

Have you had any worry about your child's hearing? ☐ Yes ☐ No

When your child is sleeping, does he move and begin to wake up to a loud sound? ☐ Yes ☐ No

Does he turn his head toward an interesting sound or when his name is called? ☐ Yes ☐ No

Does he try to imitate you if you make his own sounds? ☐ Yes ☐ No

Development and communication

Does he play peek-a-boo with you? ☐ Yes ☐ No

Can he stand for at least 5 seconds, holding on to a crib or chair? ☐ Yes ☐ No

Does he try to hold a toy when it is pulled away? ☐ Yes ☐ No

Is he shy or afraid of strangers? ☐ Yes ☐ No

12 Months of Age

Hearing

Have you had any worry about your child's hearing?

☐ Yes ☐ No

When your child is sleeping, does he move and begin to wake up to a loud sound?

☐ Yes ☐ No

Does he turn his head toward an interesting sound or when his name is called?

☐ Yes ☐ No

Is he beginning to repeat some of the sounds you make?

☐ Yes ☐ No

Development and communication

Can he pick up tiny objects with his fingers such as a raisin or candy?

☐ Yes ☐ No

Can he get to a sitting position without help?

☐ Yes ☐ No

Does he wave bye-bye or play pat-a-cake when you encourage him?

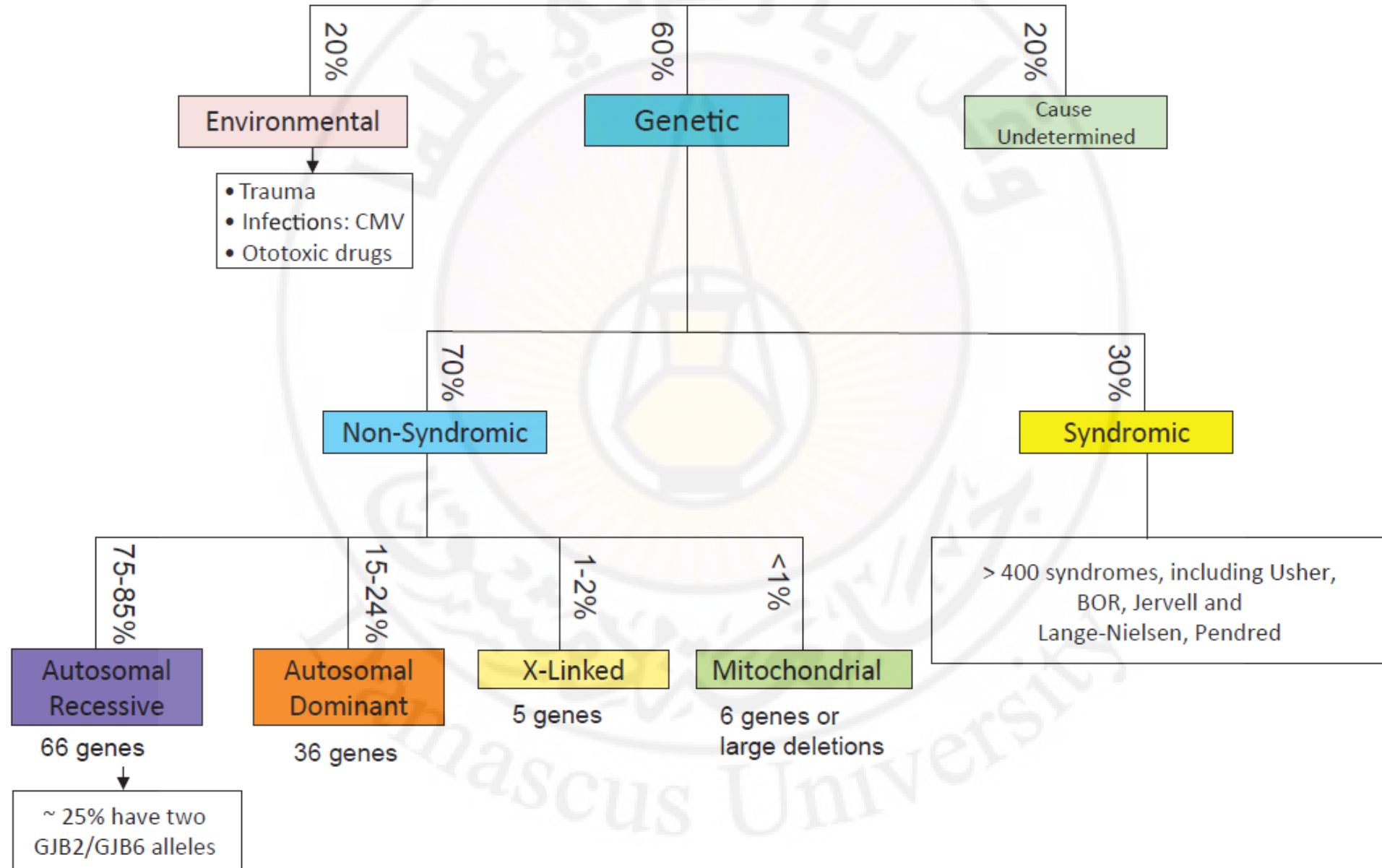
☐ Yes ☐ No

Can he say "mamma" or "dada" appropriately?

☐ Yes ☐ No

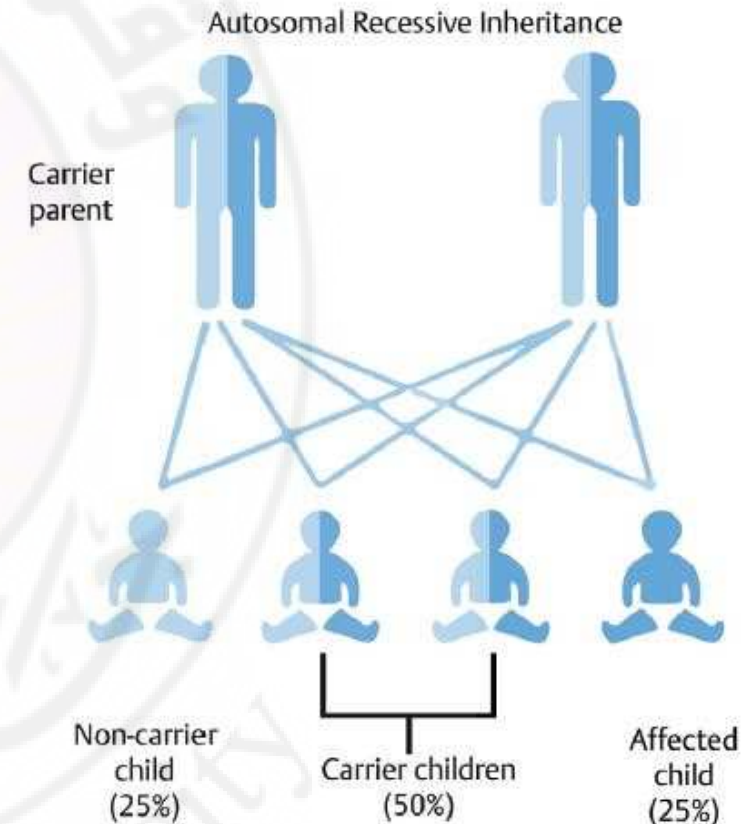
4. Hearing disorders in children

Etiologies of Childhood Hearing Loss



Genetic hearing loss in Children

- Genetics contribute to more than 50% of prelingual hearing loss.
- Over 400 genetic syndromes are associated with hearing loss, and it is important for audiologists to have an awareness of these conditions and how they affect the individual with hearing loss.
- Today, nearly all confirmed hearing loss genes are sequenced simultaneously through a process called next-generation sequencing (NGS).



Genetic hearing loss in Children

- All children identified with permanent hearing loss can benefit from genetics evaluation and testing, although most cases will not present with a family history of hearing loss.
- Today, over 100 hearing loss genes can be tested at one time using NGS.
- A negative result does not rule out a genetic cause. As more genes are discovered and added to the testing panel, the genetic basis may become known.
- In this class, we will consider the **nongenetic causes** of hearing loss in infants and children.



Medical Assessment of Newborn Infants

- All newborn infants receive multiple examinations shortly after birth.
- Typically, the first assessment is completed in the delivery room within moments of birth.
- During the initial hours of life, the infant receives a complete physical examination. In some instances, formal assessment of estimated **gestational age** is also made based on specific physical and neuromuscular characteristics.



Apgar Evaluation

- At birth, babies are rapidly assessed to detect obvious abnormalities and to determine the need for immediate resuscitation.
- Evaluation is completed at 1 minute and again at 5 minutes following birth.
- Both evaluations are based on five standardized observations (**heart rate, respiratory effort, reflex irritability, muscle tone, and color**).
- A rating of 0 to 2 is assigned to each observation. The maximum Apgar score attainable is 10.



Dr. Virginia Apgar

Scoring System for Initial Assessment of Newborn Infants Developed by
Dr. Virginia Apgar (1953)

Observation	Score		
	0	1	2
Heart rate	Absent	Slow (below 100)	Over 100
Respiratory effort	Absent; apnea	Slow, irregular, shallow, gasping	Sustained cry, regular respirations
Reflex irritability	No response	Grimace, frown	Sneeze, cry, active avoidance
Muscle tone	Limp, flaccid good tone	Some flexion of extremities	Active motion,
Color	Cyanotic, extremities pale	Body pink and pale	Completely pink

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 at the top and in English at the bottom. In the center of the logo is a stylized lamp with rays emanating from it.

APGAR SCORE

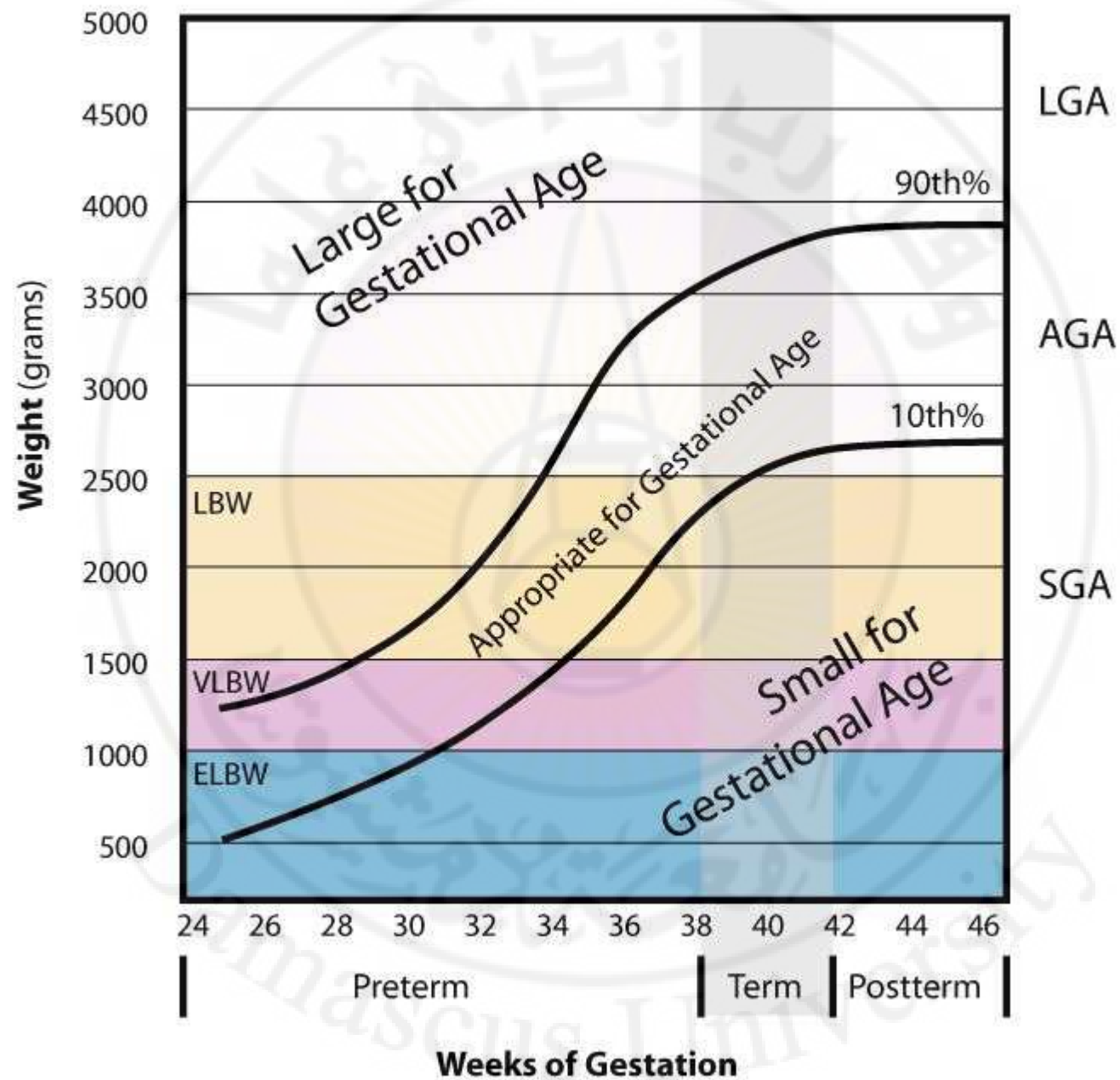
Estimating Gestational Age

- Gestational Age is a common term used during pregnancy to describe how far along the pregnancy is.
- It's **measured from the first day of women's last menstrual period (LMP) to the current date, typically in weeks.**
- Usually, pregnancies last anywhere from 38 to 42 weeks; babies born before the 37-week marker are considered premature.

Description	Gestational Age
Preterm	<36 wk and 6 d
Late preterm	34 wk and 0 d to 36 wk and 6 d
Term	37 wk and 0 d to 40 wk and 6 d
Early term	37 wk and 0 d to 38 wk and 6 d
Postterm	>41 wk

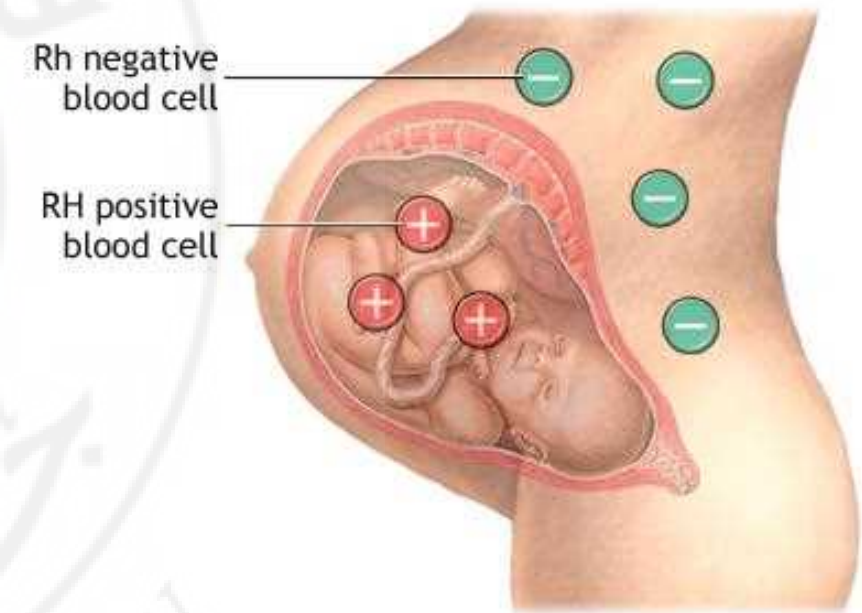
Estimating Gestational Age

- Both birth weight and gestational age are important determinants of the baby's general condition.
- Although gestational age can be estimated from the mother's last menstrual period, this calculation is often inaccurate and unreliable.
- Because certain physical and neuromuscular characteristics emerge in a predictable sequence during fetal development, these characteristics form the basis for an objective estimate of gestational age.
- These physical characteristics include skin appearance, presence of lanugo (fine hair present on the fetus which largely disappears by 40 weeks' gestation), cartilage development in the ear, and appearance of the genitalia. Neuromuscular characteristics include resting posture and flexion and extension of the extremities.
- These features yield an estimate of gestational age which can be compared to birth weight to determine if the infant is small for gestational age (SGA), appropriate for gestational age (AGA), or large for gestational age (LGA).



Rh Incompatibility

- Rh incompatibility is a condition that develops when a pregnant woman has Rh-negative blood and the baby in her womb has Rh-positive blood.
- During pregnancy, red blood cells from the unborn baby can cross into the mother's bloodstream through the placenta. If the mother is Rh-negative, her immune system treats Rh-positive fetal cells as if they were a foreign substance and makes antibodies against the fetal blood cells.
- These anti-Rh antibodies may cross back through the placenta into the developing baby and destroy the baby's circulating red blood cells.



Rh Incompatibility

- When red blood cells are broken down, they make **bilirubin**. This causes an infant to become yellow (**jaundiced**). The level of bilirubin in the infant's bloodstream may range from mild to dangerously high.
- **Clinical symptoms**, when they occur, develop during the immediate neonatal period and include elevated bilirubin, jaundice, and possible brain damage.
- **Kernicterus** is a serious form of brain damage caused by excessive jaundice



Hearing Disorders in Children

- Many pathologic conditions, including disease, trauma, and developmental disturbance, cause hearing disorders during childhood.
- The prevalence of hearing disorders in children is relatively high compared with other childhood disorders.
- Hearing disorders are customarily classified according to the site of the interruption in sound transmission or encoding.

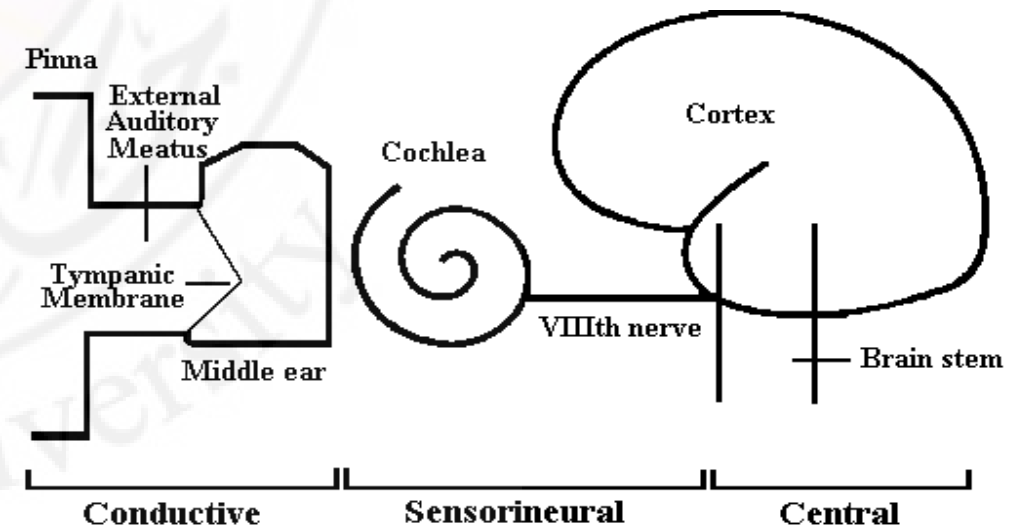
- **Conductive Hearing Disorders**

- Acquired Prenatal Conductive Disorders
 - Acquired Postnatal Conductive Disorders

- **Sensory Hearing Disorders**

- Acquired Prenatal Sensory Disorders
 - Acquired Perinatal and Postnatal Sensory Disorders

- **Neural Hearing Disorders**



Conductive Hearing Disorders

- Conductive hearing disorder results from problems involving structures of the outer and middle ear, including congenital anomalies and otitis media and its complications.
- Conductive hearing disorders in children are most commonly acquired and transient.
- Most respond well to medical management and have negligible impact on long-term auditory function.

Acquired prenatal disorders	Acquired postnatal disorders
Atresia Middle ear anomalies	Otitis media with effusion Tympanic membrane perforation Cholesteatoma Excessive cerumen Otitis externa

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Acquired Prenatal Conductive Disorders

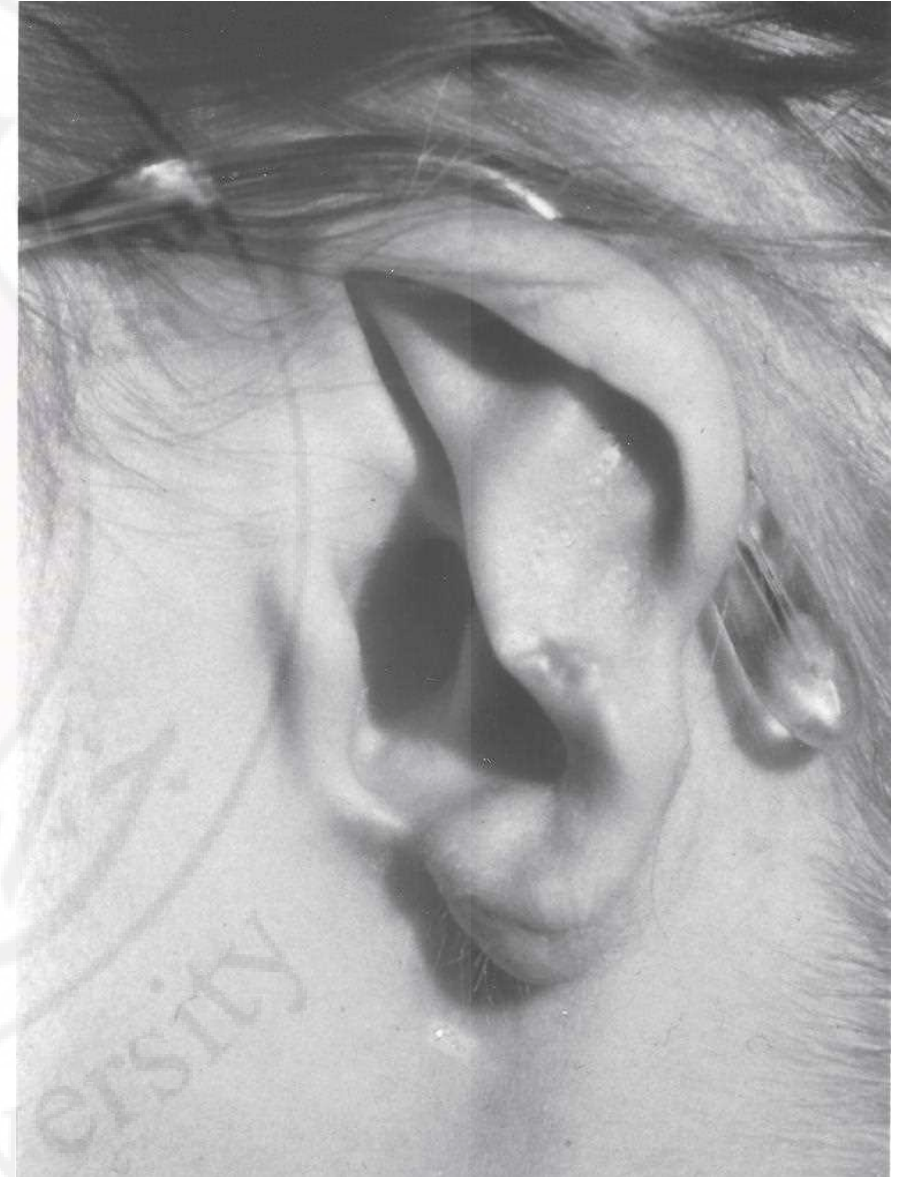
Outer Ear Anomalies

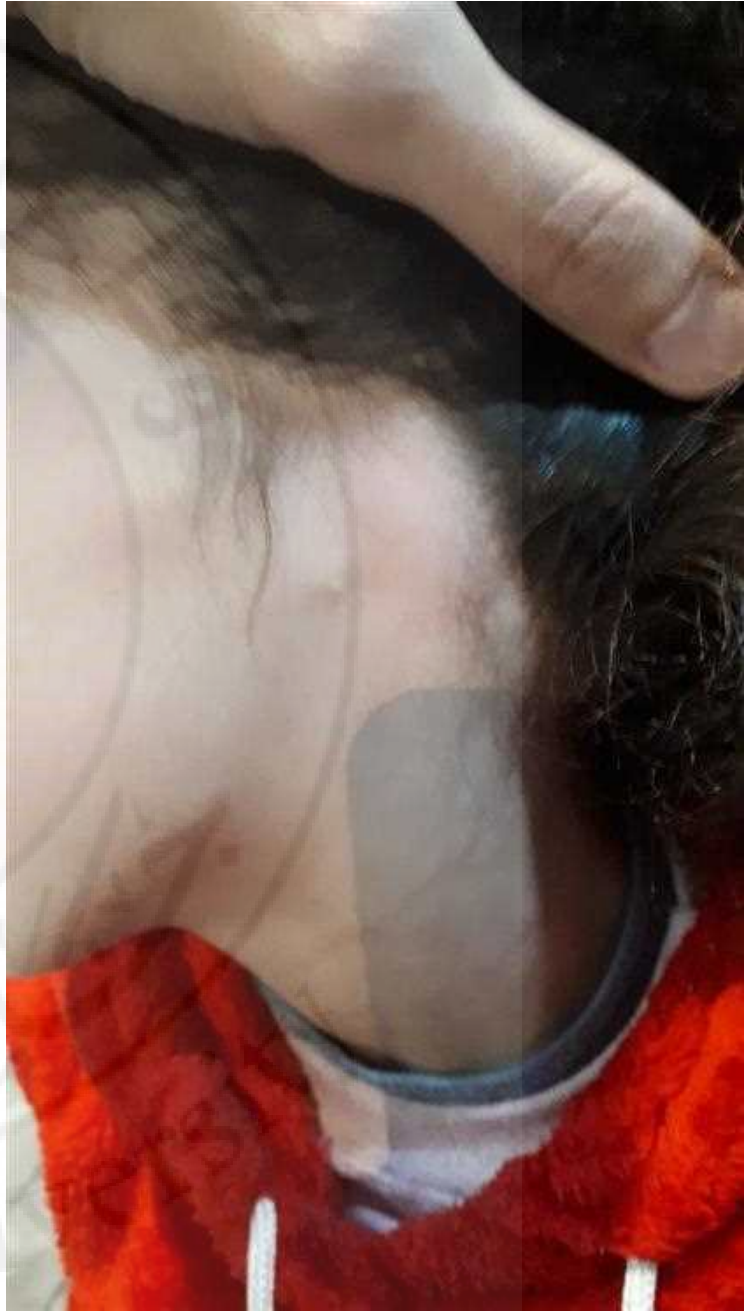
- **Atresia** is the absence of an opening of the ear or external auditory meatus, occurring in ~ 1 in 6,000 births.
- **Bony atresia** is the congenital absence of the ear canal caused by a wall of bone separating the external ear from the middle ear.
- **Membranous atresia** is the absence of a canal caused by a dense soft tissue plug obstructing the canal.
- Atresia is unilateral in ~ 70 to 90% of cases. Atresia can cause maximum conductive hearing loss (~ 60 dB) depending on the density of the blockage



Outer Ear Anomalies

- An abnormally small or malformed ear is known as **microtia**.
- Although microtia does not necessarily cause hearing disorder, it is often associated with abnormalities of the ear canal, including **stenosis** or narrowing of the ear canal.
- **Stenosis** may or may not cause a hearing disorder, but it can cause additional complications, including excessive cerumen accumulation and even cholesteatoma formation.







Lobule type



Small concha type



Concha type



Anotia

**Nagata
classification**

**Firmin
classification**

**Type 1: Lobule
transposition**

**Type 2:
transfixion**

Type 3

**3a: Enough
skin envelope
to cover
construct**

**3b: No skin
envelope**

Middle Ear Anomalies

- Middle ear anomalies are often associated with abnormalities of the ear canal and auricle, although they can occur in isolation.
- Middle ear anomalies include **ossicular dysplasia** (abnormal development of the bones of the middle ear), **fenestral malformations** (malformation of the oval window), and **congenital cholesteatoma**.
- Ossicular dysplasia can result in fixation, deformity, and disarticulation of the bones, especially the incus and stapes.
- In congenital stapes fixation, a fenestral malformation, the stapes footplate is fixed into the bony wall of the cochlea at the oval window. Lack of oval window development is also an example of fenestral malformation.
- Congenital cholesteatoma is a cyst that is present in the middle ear space without any evidence of causative factors such as otitis media.

Causes of Prenatal Outer and Middle Ear Anomalies

- Congenital outer and middle ear anomalies are often a result of **genetic** causes.
- In many cases, these anomalies may be one of a constellation of symptoms occurring as part of a **syndrome** or sequence.
- Often, prenatal outer and middle ear anomalies may occur with other **craniofacial anomalies** and/or sensorineural hearing loss.

- Treacher Collins syndrome
- Branchio-oculo-facial (BOF) syndrome
- Hutchinson-Gilford progeria syndrome
- Down syndrome
- Townes-Brocks syndrome
- lacrimo-auriculo-dento-digital (LADD) syndrome
- Oculoauriculovertebral dysplasia (Goldenhar syndrome)
- Klippel-Feil syndrome
- Cornelia de Lange syndrome
- CHARGE (coloboma of the eye, heart anomaly, choanal atresia, retardation, genital and ear anomalies) syndrome

Treacher Collins syndrome



Branchio-oculo-facial (BOF) syndrome



Hutchinson-Gilford progeria syndrome



Down syndrome



Townes-Brocks syndrome



lacrimo-auriculo-dento-digital (LADD) syndrome



Oculoauriculovertebral dysplasia (Goldenhar syndrome)



Klippel-Feil syndrome



Cornelia de Lange syndrome



CHARGE syndrome

(Coloboma of the eye, Heart anomaly, Choanal atresia, Retardation, Genital and Ear anomalies)

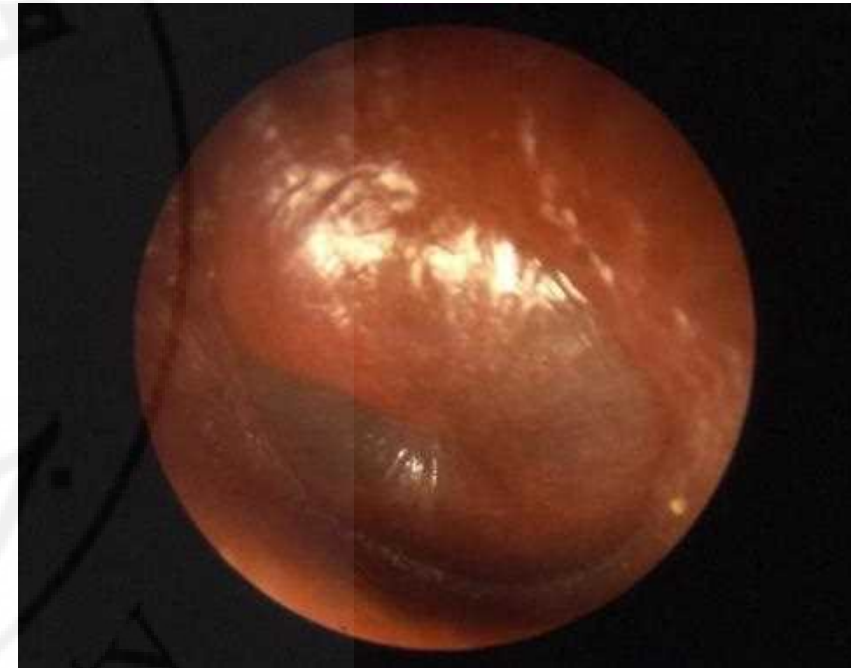


The logo of Damascus University is a circular emblem. It features a central yellow sunburst or star-like symbol. The top half of the circle contains Arabic text in a stylized font. The bottom half of the circle contains the text "Damascus University" in English. The entire logo is rendered in a light gray, semi-transparent style.

Acquired Postnatal Conductive Disorders

Otitis Media with Effusion

- **Otitis media** is a general term to describe inflammation of the middle ear mucous membrane and tympanic membrane.
- Otitis media is the most common diagnosis in patients who make office visits to physicians.
- Estimates are that 76 to 95% of all children have one episode of otitis media by 6 years of age.
- The prevalence is highest during the first 2 years and declines with age.
- The most commonly associated microbial contributions to otitis media are *Haemophilus influenzae* and *Streptococcus pneumoniae*, with *Moraxella catarrhalis* and others contributing to a lesser extent (Vaccination).



CAUSES OF ACUTE OTITIS MEDIA

BACTERIAL

- Streptococcus pneumoniae
- Haemophilus influenzae

VIRAL

- RSV
- Rhinovirus
- Adenovirus
- Many others

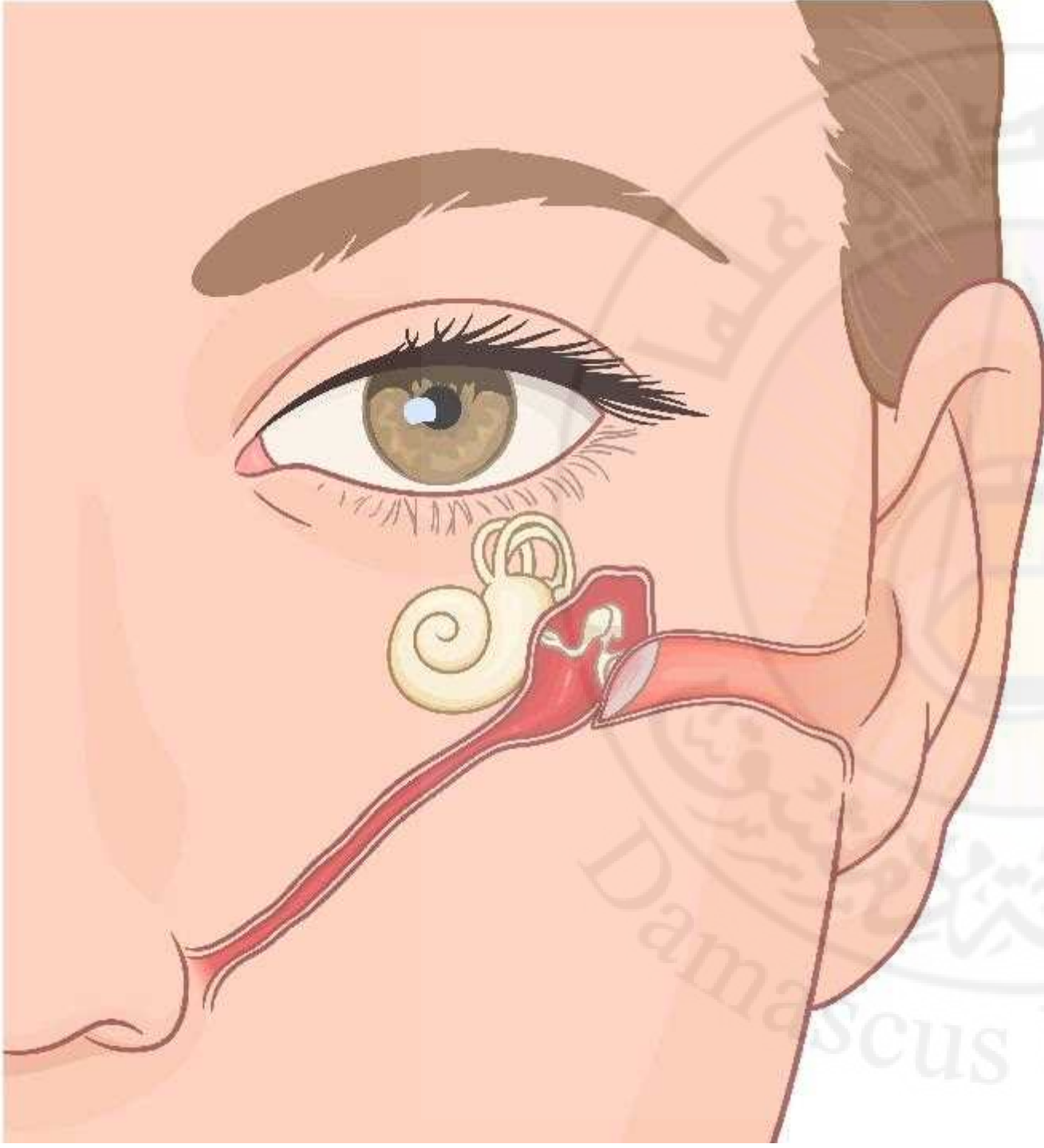
Risk factors for otitis media include:

- young age
- Native American or Inuit heritage
- Down syndrome
- human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS)
- anatomic defects such as cleft palate
- exposure to smoke in the household
- male sex
- crowded living conditions
- poor sanitation (such as clean drinking water)
- inadequate medical care
- eating in prone position
- Obesity
- day care
- There is also increasing evidence of genetic susceptibility to otitis media



Pathophysiology

- Otitis media is typically caused by **eustachian tube dysfunction** secondary to upper respiratory tract infection.
- Swelling of the nasopharynx results in failure of the eustachian tube to protect, clear, and equalize the pressure of the middle ear space, permitting reflux of infectious secretions from the nasopharynx into the middle ear.



Terminology

- Otitis media is usually defined by the presence or absence of **effusion** in the middle ear space, the type of effusion, and the time course of the disorder.
- **Otitis media with effusion (OME)** is the common term used to describe the disorder.
- The fluid may be referred to as **serous** (thin, watery, sterile), **suppurative** (containing pus), **purulent** (suppurative), **mucoïd** (thick, viscid), or **sanguineous** (containing blood).
- **Adhesive otitis media** involves severe retraction of the tympanic membrane into the middle ear space.



Terminology

- **Acute otitis media with effusion** is the term describing rapid onset of symptoms of middle ear inflammation, including redness of the tympanic membrane and otalgia. It is usually referred to as **acute** if it lasts shorter than 3 weeks or as **subacute** if it lasts shorter than 3 months.
- **Recurrent acute OME** consists of repeated episodes of acute otitis media with normal middle ear examinations between episodes.
- **Persistent middle ear effusion** is an asymptomatic effusion that persists following treatment for acute OME.
- The term **chronic OME** is used to describe a condition that lasts longer than 3 months.
- **Chronic suppurative otitis media** is a chronic infection of the middle ear and mastoid air cells with a perforation of the tympanic membrane and otorrhea.

Symptoms

- Middle ear effusion is signaled by bulging and limited mobility of the tympanic membrane, otorrhea, or a fluid level behind the tympanic membrane.
- Other nonspecific symptoms in an infant or toddler may include fever, excessive crying, or pulling on the ears.
- Vertigo, dizziness, and imbalance are also common in children with otitis media.





Hearing loss



Ear fullness

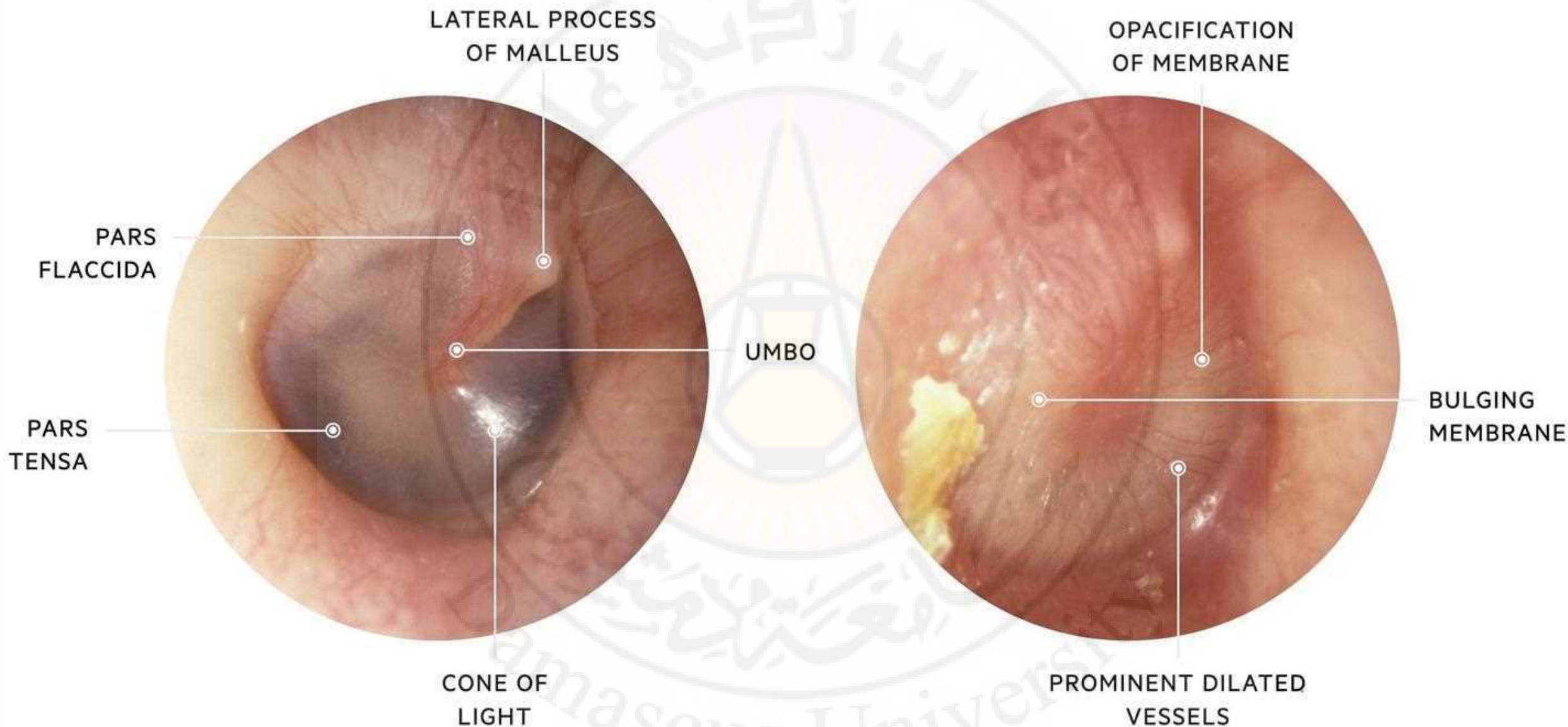


Pain

Change in
behavior

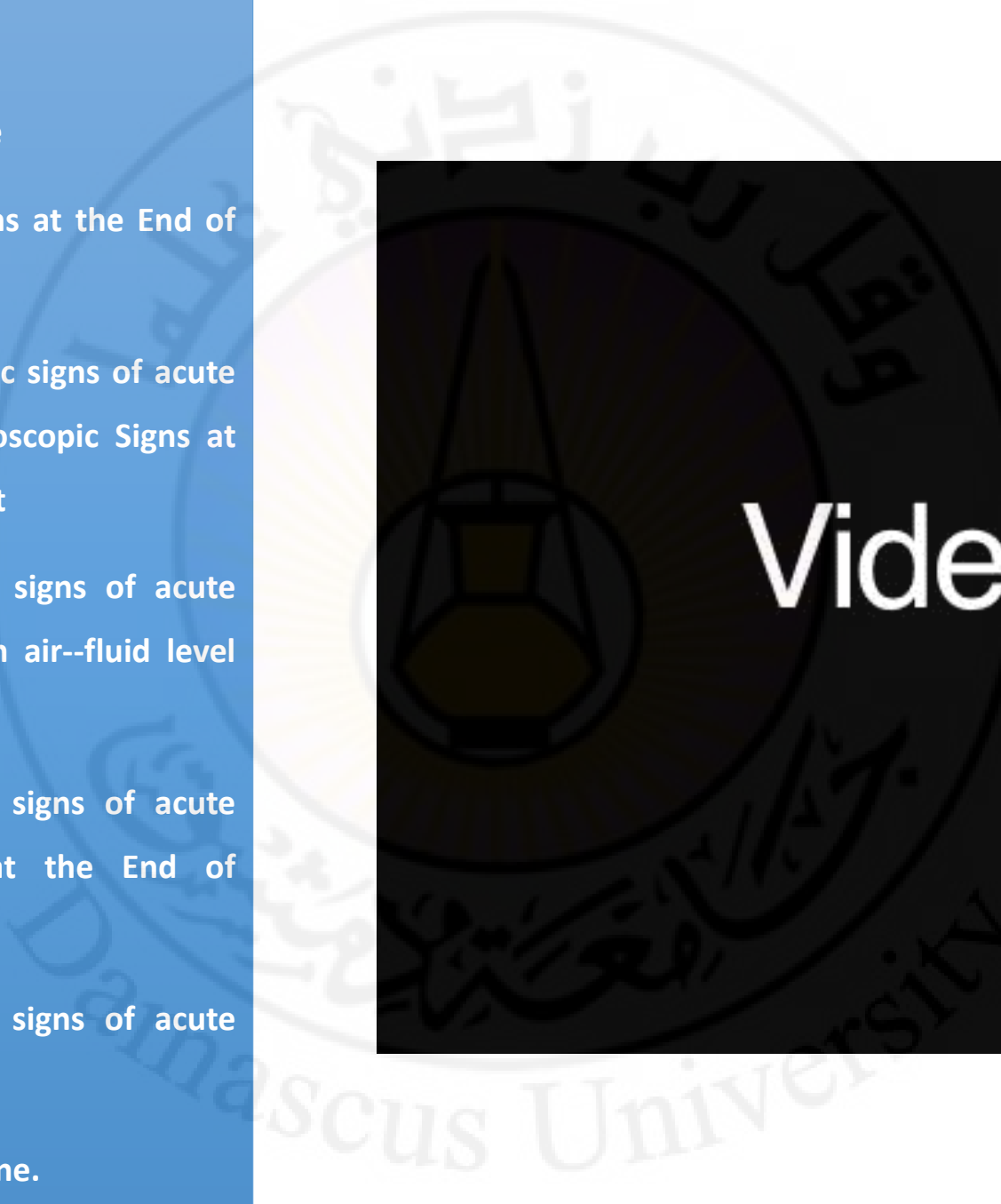
Child pulling
at their ear





Otoscopy is key to diagnosing AOM.

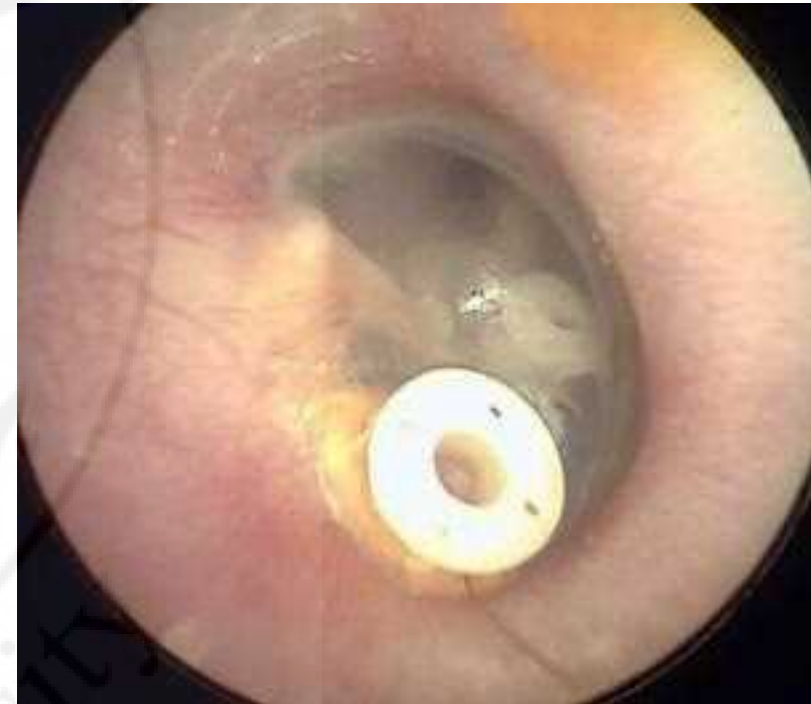
1. Air-fluid level
2. Full, bulging tympanic membrane
3. Bulla formation; Otoloscopic Signs at the End of Treatment: Complete Resolution
4. Complete resolution of otoscopic signs of acute otitis media; middle-ear fluid; Otoloscopic Signs at the End of Treatment: Improvement
5. Partial resolution of otoscopic signs of acute otitis media; middle-ear fluid with air--fluid level remains
6. No improvement in otoscopic signs of acute otitis media; Otoloscopic Signs at the End of Treatment: Failure
7. No improvement in otoscopic signs of acute otitis media
8. Perforation of tympanic membrane.

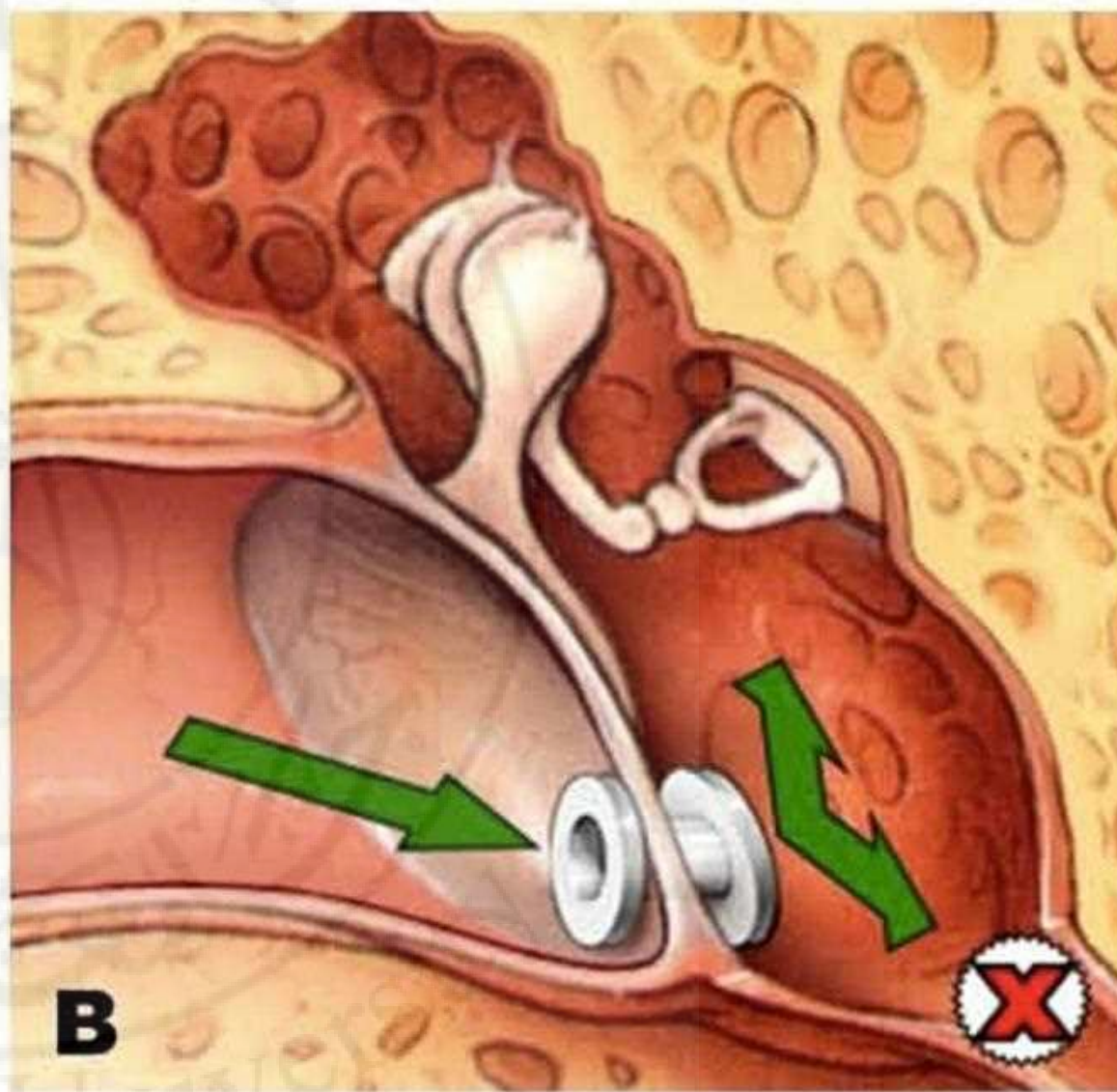


Video 1

Management of otitis media

- OME may resolve spontaneously or may require treatment with antibiotics or pressure equalization tubes (or ventilation tubes / VT), with varying effects on recurrence rates. Hearing loss fluctuates with the presence or absence of fluid.
- **Untreated OME** can lead to several complications, including cholesterol granuloma, adhesive otitis media, facial paralysis, labyrinthitis, acute mastoiditis, petrositis, meningitis, sigmoid sinus thrombosis, extradural abscess, brain abscess, otic hydrocephalus, and sensorineural hearing loss. In rare cases otitis media can result in death.
- Among the *more common complications* are tympanic membrane perforation, tympanosclerosis, and cholesteatoma.





Complications of Otitis Media with Effusion: Tympanic membrane perforation

- The tympanic membrane may become perforated as a result of OME because of increased pressure from fluid in the middle ear space, or it may be perforated from barotrauma, trauma, myringotomy, or tympanostomy tube placement.
- Although perforations- generally heal spontaneously, they may require surgical intervention to repair the damaged membrane.
- Drainage from perforation may cause a secondary infection of the external auditory canal or auricle.
- A perforation may or may not result in hearing loss, depending on its **size and location**.



Complications of Otitis Media with Effusion: Tympanosclerosis

- Tympanosclerosis is a degeneration of collagenous fibrous tissues of the tympanic membrane and is a common sequela of OME, occurring in ~ 10% of cases.
- Calcification or ossification may occur and spread to the ossicles in rare cases.
- Tympanosclerosis can often be observed as a horseshoe-shaped plaque on the tympanic membrane.
- Tympanosclerosis is not associated with significant hearing loss unless it also involves the ossicular chain.



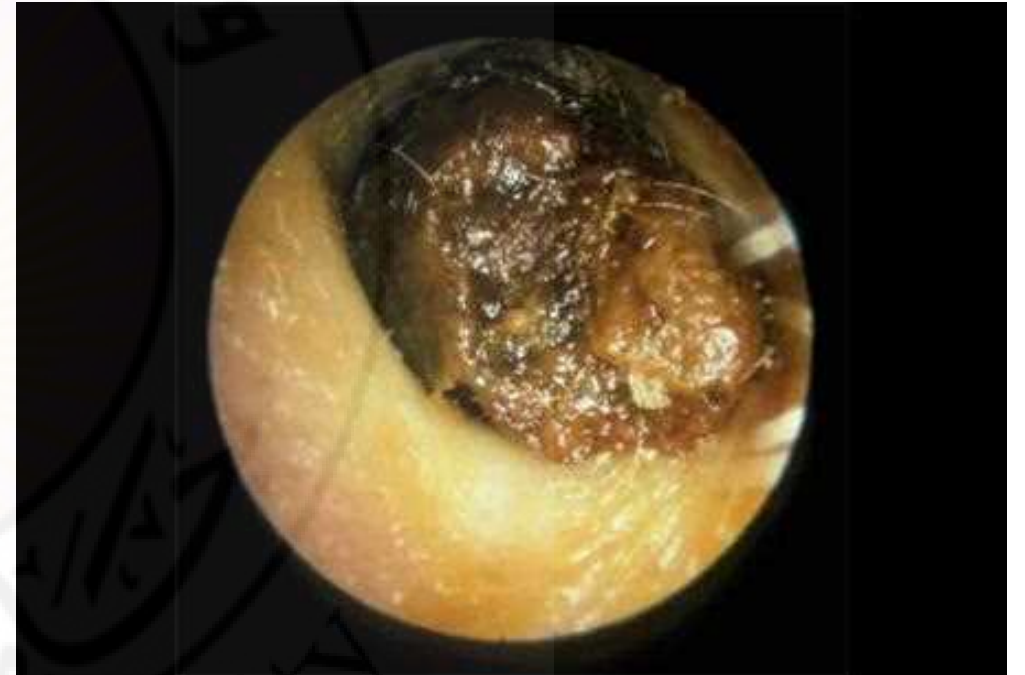
Complications of Otitis Media with Effusion: Cholesteatoma

- Cholesteatomas are cysts that contain keratinizing squamous epithelium and are found in the middle ear, mastoid, external auditory canal, or petrous bone.
- **Congenital cholesteatomas** are present behind an intact tympanic membrane with no history of significant otitis media or eustachian tube dysfunction.
- **Acquired cholesteatomas** are more common and are usually a consequence of chronic otitis media.
- Cholesteatomas can be destructive, as they grow and compete for space with the normal structures of the areas they occupy. Conductive hearing loss varies as a function of the structures involved. Allowed to grow unchecked, a cholesteatoma can erode the middle ear ossicles and cause ossicular discontinuity, creating additional conductive hearing loss. In advanced cases, sensorineural hearing loss can also occur because of cochlear erosion.



Excessive Cerumen

- Excessive cerumen (ear wax) in the ear canal occurs in ~ 10% of children, and it may have an even greater incidence, up to 28 to 36%, in children with developmental delays.
- A high-frequency conductive hearing loss can occur when the ear canal is 80 to 95% occluded.
- A low-frequency conductive loss occurs with total occlusion of the ear canal.



Otitis Externa

- **Otitis externa** is the broad term for inflammation or infection of the external auditory canal and auricle.
- Otitis externa rarely causes hearing disorder, except in cases where it causes stenosis of the external auditory meatus.
- **Acute diffuse otitis externa**, also known as **swimmer's ear**, is one example of otitis externa. It is a *bacterial infection* that causes itching, tenderness, and pain and may include hearing loss and aural fullness as the external auditory canal decreases in size with swelling.
- Complications of otitis externa may include ear canal stenosis, myringitis, and tympanic membrane perforation.



Sensory Hearing Disorders

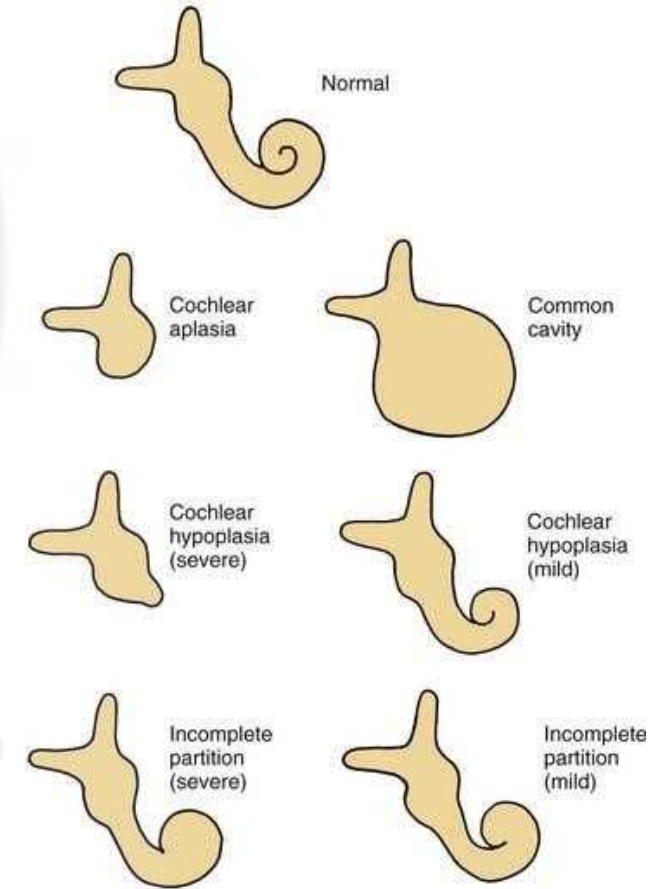
Acquired prenatal disorders	Acquired perinatal and postnatal disorders
Inner ear anomalies Cytomegalovirus Syphilis Rubella Toxoplasmosis	Persistent pulmonary hypertension of the newborn (PPHN)/extracorporeal membrane oxygenation (ECMO) Meningitis Autoimmune inner ear disease Mumps Measles Hemorrhagic fever Ototoxicity

The logo of Damascus University is a circular emblem. It features a central yellow lamp with a flame, set against a background of radiating lines. The lamp is enclosed within a circle, which is itself within a larger circle. The top half of the outer circle contains Arabic text, and the bottom half contains the English text "Damascus University".

Acquired Prenatal Sensory Disorders

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 **genetic**, 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.
- In recent years, there has been greater success with cochlear implantation in cases of deafness caused by these deformities, despite the unusual anatomy.



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

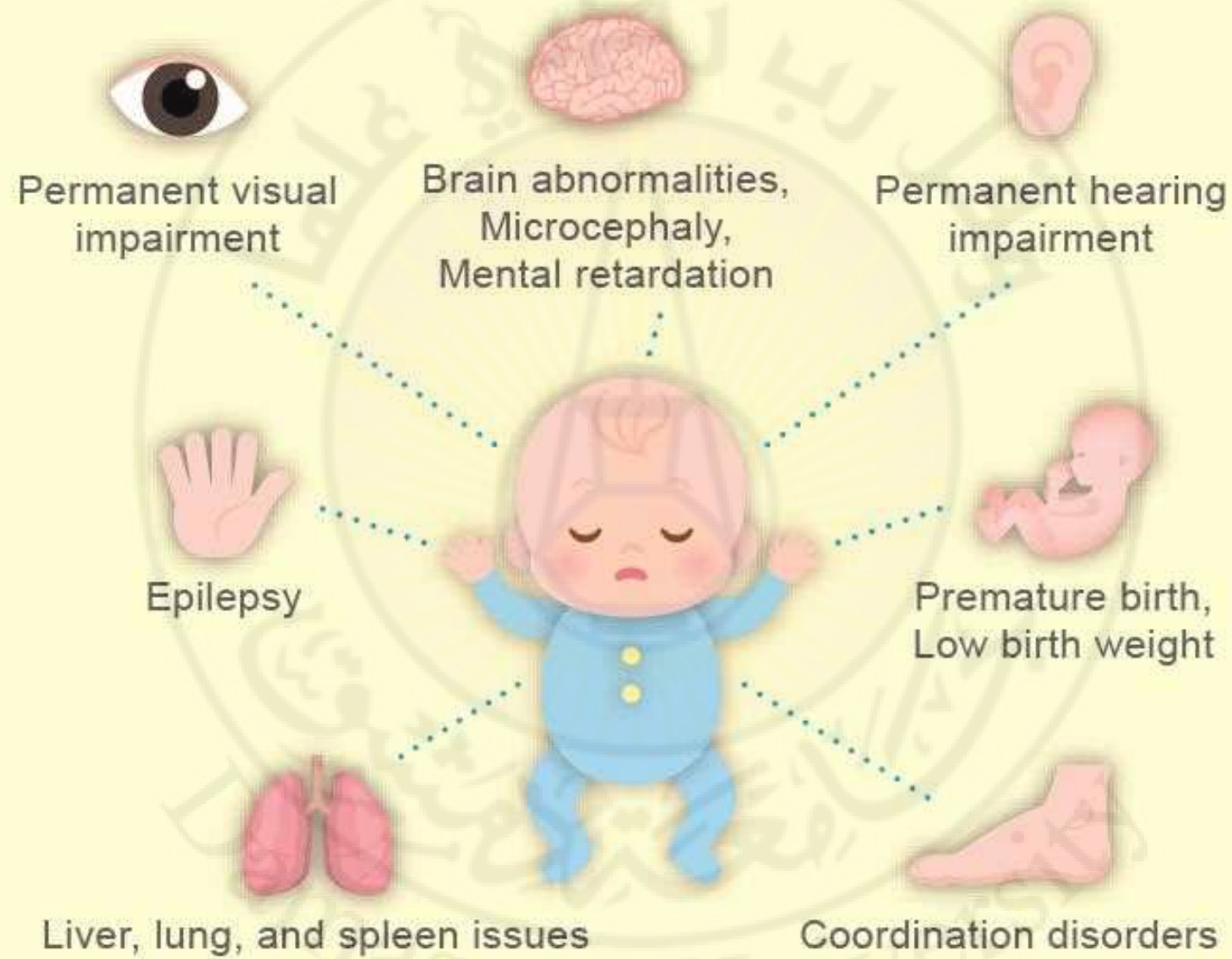
- Complete membranous labyrinth dysplasia (Bing-Siebenmann)
- And two forms of partial dysplasia:
 - Cochleosaccular dysplasia (Scheibe aplasia)
 - Cochlear basal turn dysplasia (Alexander aplasia)

Cytomegalovirus

- Cytomegalovirus (CMV) is the largest known member of the human herpesvirus family and causes the most common fetal viral illness.
- Congenital infection is the result of transplacental transmission of CMV and is known as cytomegalic inclusion disease.
- Sensorineural hearing loss is the most common clinical finding of congenital CMV. Other findings include microcephaly, petechiae (small purple spots on a body surface), intrauterine growth retardation, enlargement of the liver and spleen, and chorioretinitis (inflammation of the choroid and retina).
- Hearing loss is of variable severity and can be bilateral or unilateral. Threshold fluctuations are common (more than 20% of cases), and hearing loss is often progressive (more than 15%).

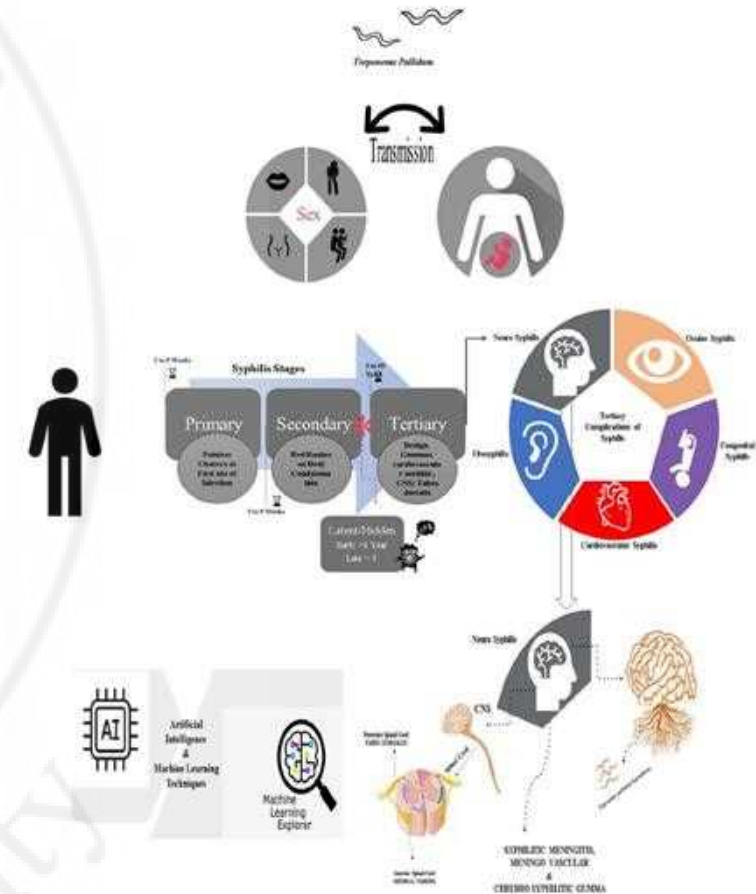


Cytomegalovirus



Congenital Syphilis

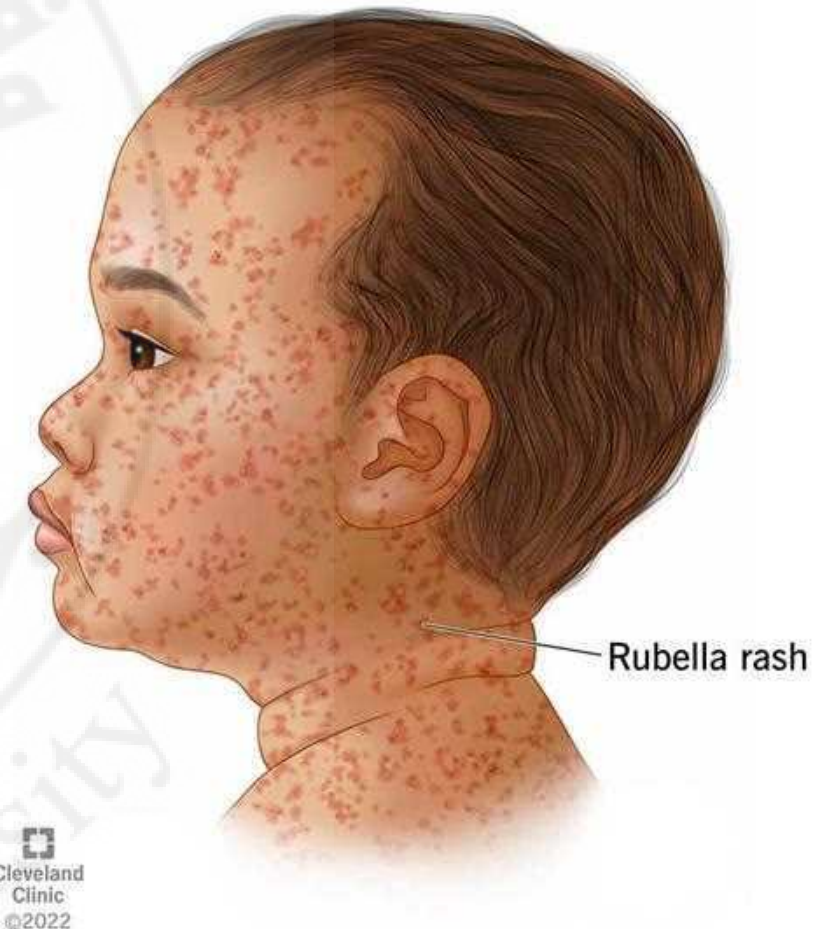
- Congenital syphilis is a bacterial infection that is transmitted from mother to fetus in utero or through contact with a genital lesion during delivery.
- Congenital syphilis is categorized by its time of occurrence into early and late stages.
- Typical hearing loss is a rapid symmetric progression 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**.



Maternal Rubella

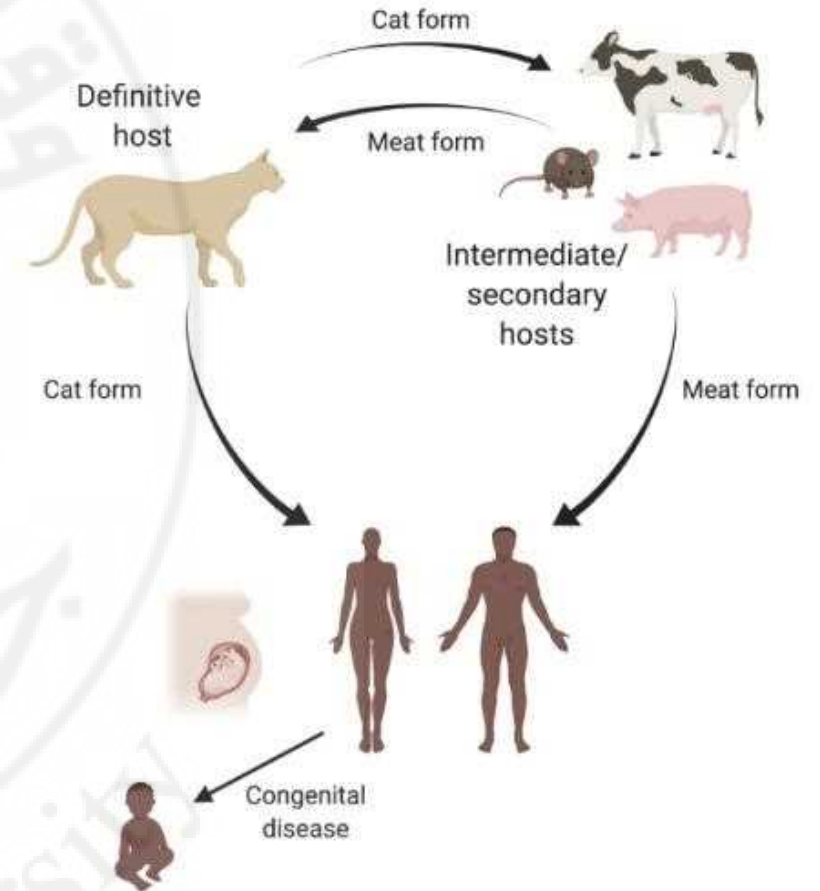
- Maternal rubella occurs when an infected mother transmits the rubella virus to the fetus. Expression of symptoms of maternal rubella infection, called congenital rubella syndrome or Gregg syndrome, includes a wide variety of defects, usually affecting hearing, vision, and heart function, and often 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

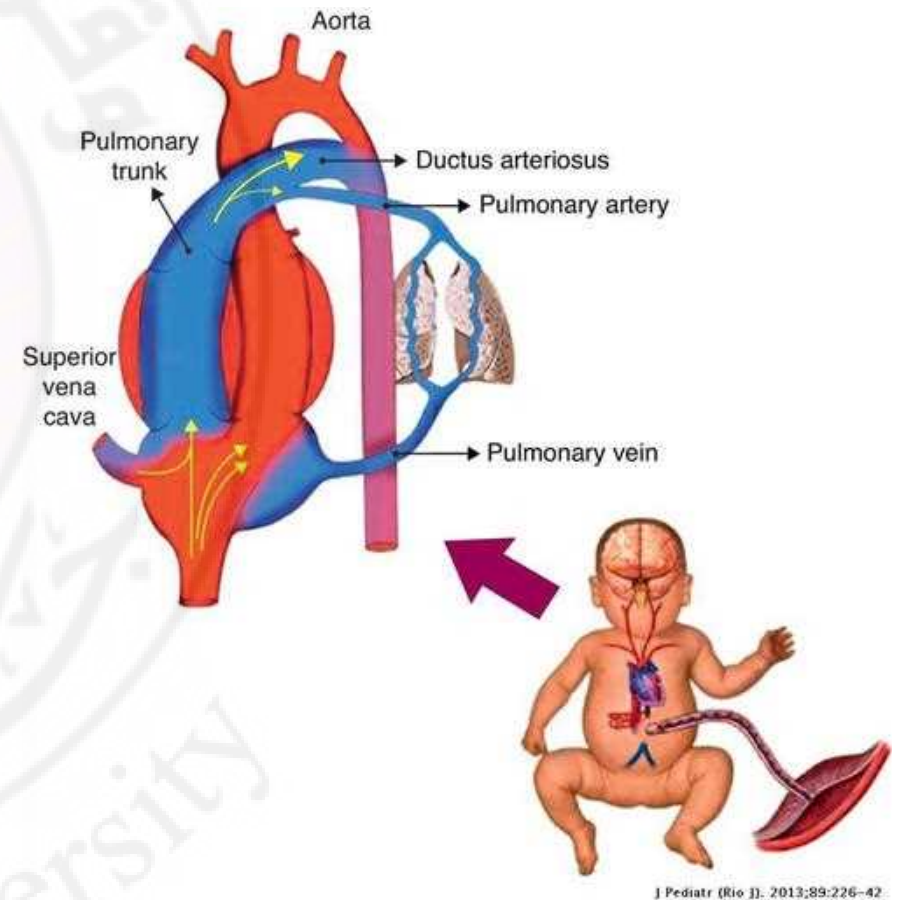


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Acquired Perinatal and Postnatal Sensory Disorders

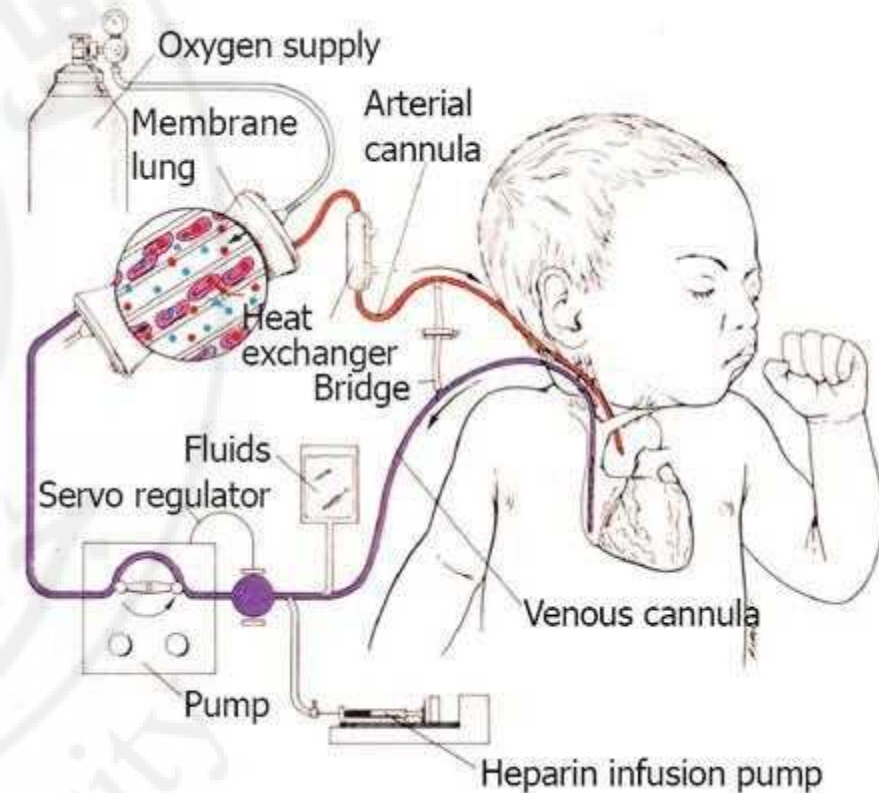
Persistent Pulmonary Hypertension of the Newborn

- **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 35% 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.



Meningitis

- Meningitis is an inflammation of the membranes that surround the brain and spinal cord.
- There are three types of meningitis: bacterial, aseptic, and viral.
- The greatest frequency of hearing loss occurs in cases of bacterial meningitis; estimates range 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, may be unilateral or bilateral, and may be progressive.
- Most involvement is thought to be cochlear, but central auditory involvement may occur in some individuals.
- Hearing loss is usually permanent.
- **Cochlear osteoneogenesis**, or bony growth in the cochlea, may occur following meningitis, complicating possible cochlear implantation.

MENINGITIS SYMPTOMS



Fever



Sleepiness



Headache



Light sensitivity



Vomiting



Joints pain



Rash



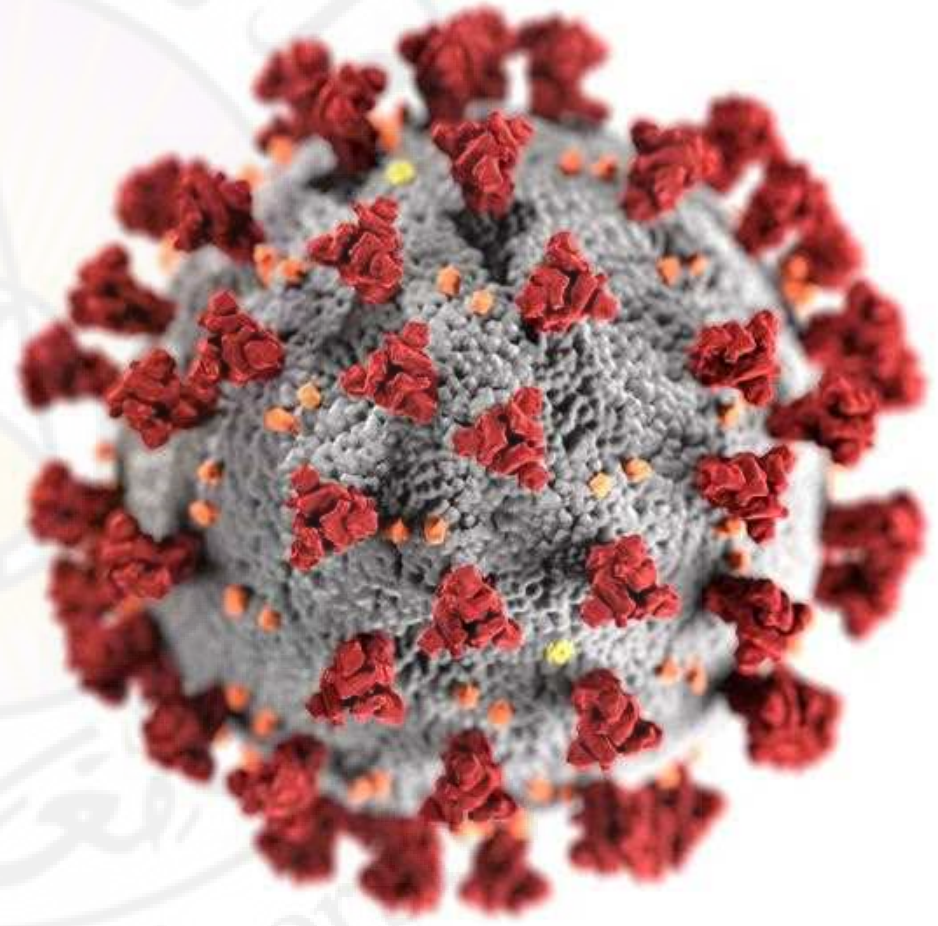
Seizures

Autoimmune Inner Ear Disease

- **Autoimmune inner ear disease (AIED)** is a syndrome of potentially reversible, bilateral, rapidly progressive, and often fluctuating sensory hearing loss that may be associated with vestibular symptoms mimicking Ménière disease.
- Symptoms are thought to be consequences of immune-mediated inflammation in the inner ear.
- The disorder may be specific to the ear or may occur as a manifestation of a systemic immune-mediated inflammatory disorder, such as rheumatoid arthritis, systemic lupus erythematosus, inflammatory bowel disease, polyarteritis nodosa, or Cogan syndrome.
- Hearing sensitivity is generally responsive to immunosuppressive drugs such as steroids.
- AIED is associated with sensory hearing loss that is generally bilateral, asymmetric, and rapidly progressive

Viral Infections

- Mumps
- Measles
- Lassa
- Ebola
- Zika
- **Covid-19 !**



Mumps

- Mumps is a viral infection that attacks a variety of organs, especially the salivary glands.
- Mumps has historically been the most common cause associated with unilateral acquired sensorineural hearing loss in children.
- The typical pattern of sensory hearing loss is unilateral and profound, with sudden onset.
- Endolymphatic hydrops (Ménière disease) is also a common manifestation of mumps virus.



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.



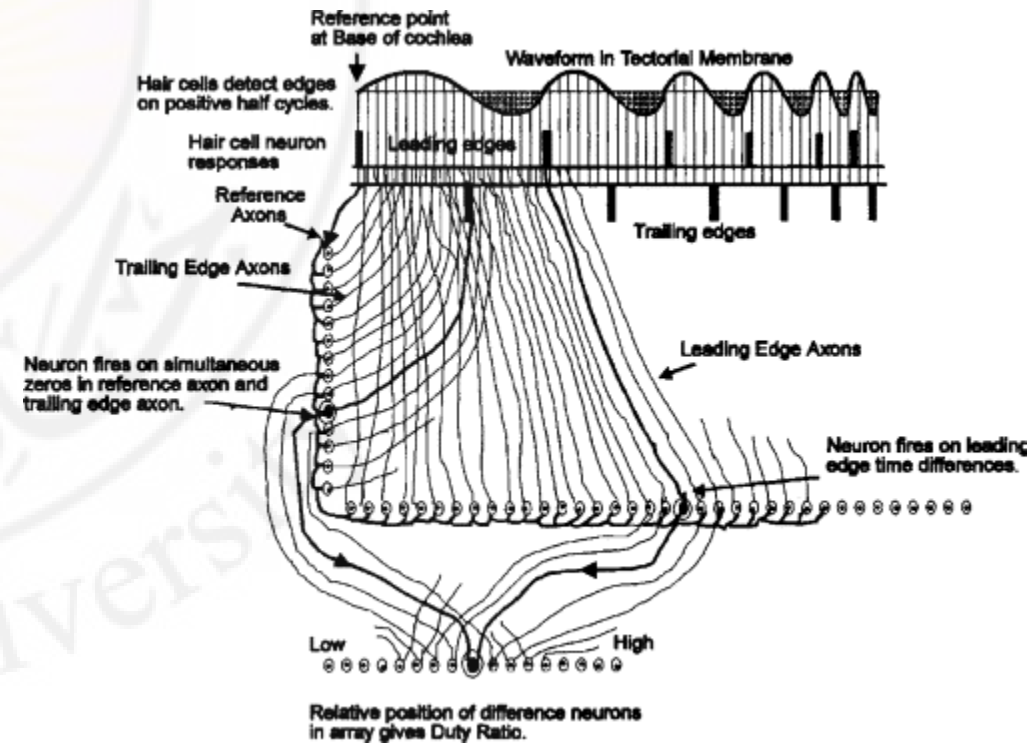
Ototoxicity

- **Aminoglycoside** antibiotics (streptomycin, gentamicin, dihydrostreptomycin, neomycin, kanamycin, erythromycin, and vancomycin), **loop diuretics** (furosemide), **antineoplastic** agents (cisplatin), **salicylates**, and **antimalarial** drugs (quinine) can all damage the cochlea.
- Children and neonates are considered to be at lower risk for ototoxicity than adults in most cases. One exception is **cisplatin-induced hearing loss**, which has a higher incidence and increased severity in children, causing hearing loss in ~ 60% of pediatric cases.



Neural Hearing Disorders

- Auditory Neuropathy Spectrum Disorder
- Neoplasm
- Hydrocephalus
- Hypoxia
- Hyperbilirubinemia



The logo of Damascus University is a circular emblem. It features a central yellow lamp with a flame, set against a background of radiating lines. The lamp is enclosed within a circle, which is itself within a larger circle. The top half of the outer circle contains Arabic text, and the bottom half contains the English text "Damascus University".

Acquired Prenatal Sensory Disorders

Auditory Neuropathy Spectrum Disorder (ANSD)

Scand Audiol 25 1996

Auditory Nerve Disease of Both Ears Revealed by Auditory Brainstem Responses, Electrocochleography and Otoacoustic Emissions

Kimitaka Kaga¹, Masaichi Nakamura¹, Masanobu Shinogami¹, Toshihiro Tsuzuku², Katsushi Yamada² and Mitsuko Shindo³

¹Department of Otolaryngology, University of Tokyo, Bunkyo-ku, Tokyo, Japan; ²Department of Otolaryngology, Teikyo University School of Medicine, Itabashi-ku, Tokyo, Japan; ³Division of Speech Pathology, Hiroshima Prefectural College of Health and Welfare, Mihara, Hiroshima Prefecture 723, Japan

Brain (1996), **119**, 741–753

Auditory neuropathy

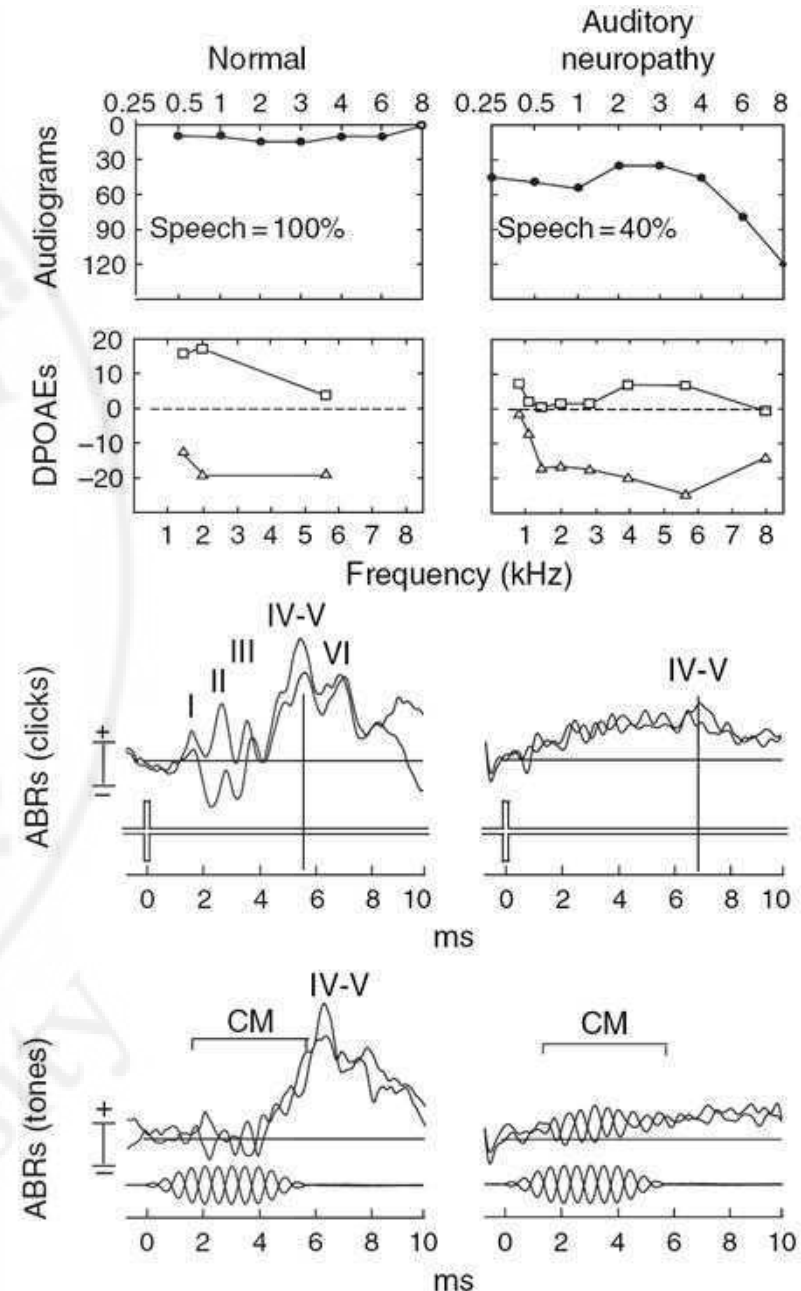
Arnold Starr,¹ Terence W. Picton,⁴ Yvonne Sininger,² Linda J. Hood³ and Charles I. Berlin³

¹Department of Neurology, University of California, Irvine, the ²House Ear Institute, Los Angeles, the ³Kresge Hearing Research Laboratory, Louisiana State University Medical Center, New Orleans, USA and ⁴Rotman Research Institute of Baycrest Centre, University of Toronto, Toronto, Canada

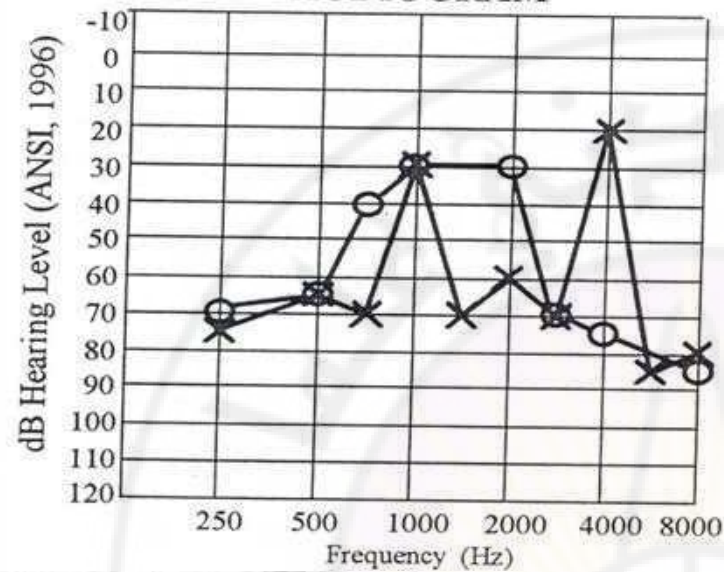
Correspondence to: Arnold Starr, Department Neurology, University of California Irvine, Irvine, CA 92717, USA

Auditory Neuropathy Spectrum Disorder (ANSD)

- *Auditory neuropathy spectrum disorder (ANSD)* is a term that is used to describe disorders that are operationally defined based on a constellation of clinical findings.
- In **older children**, auditory neuropathy is defined by an absent auditory brainstem response (ABR), poor speech perception, varying levels of hearing sensitivity loss, absence of acoustic reflexes, and a preservation of some cochlear function as evidenced by the preservation of OAEs or cochlear microphonics.
- In **infants**, auditory neuropathy is defined by absent ABR and preserved OAEs or cochlear microphonics.



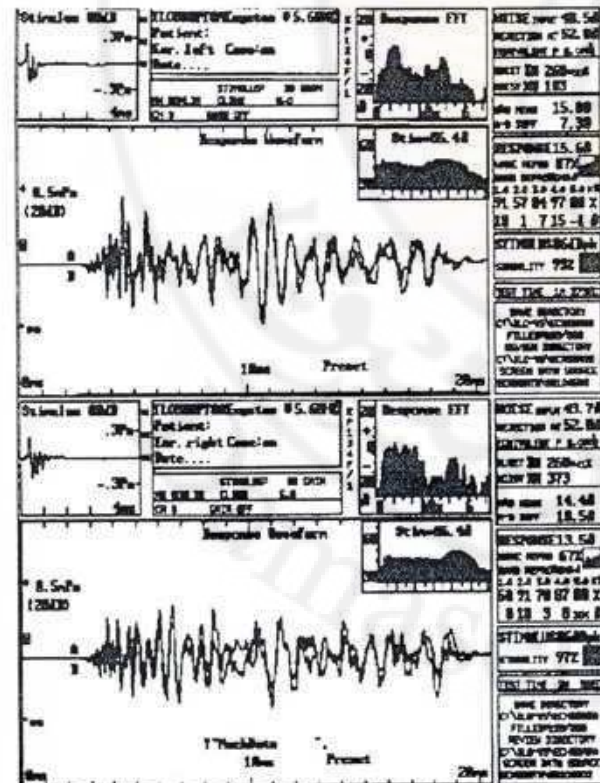
AUDIOGRAM



Speech Awareness Threshold (SAT) -
 Right Ear = 20 dBHL
 Left Ear = 25 dBHL
 Speech Discrimination -
 Right Ear = 28%
 Left Ear = 8%
 Tympanometry = WNL
 Acoustic Reflex Threshold = Absent

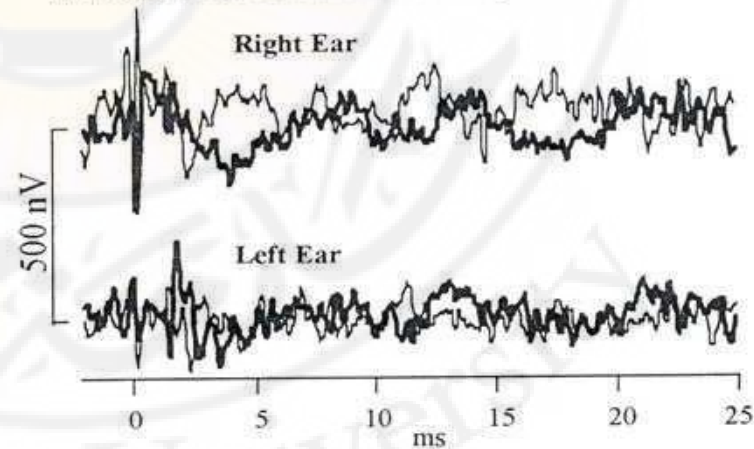
○ Right Ear
 × Left Ear

Transient-Evoked Otoacoustic Emission



Auditory Brainstem Response

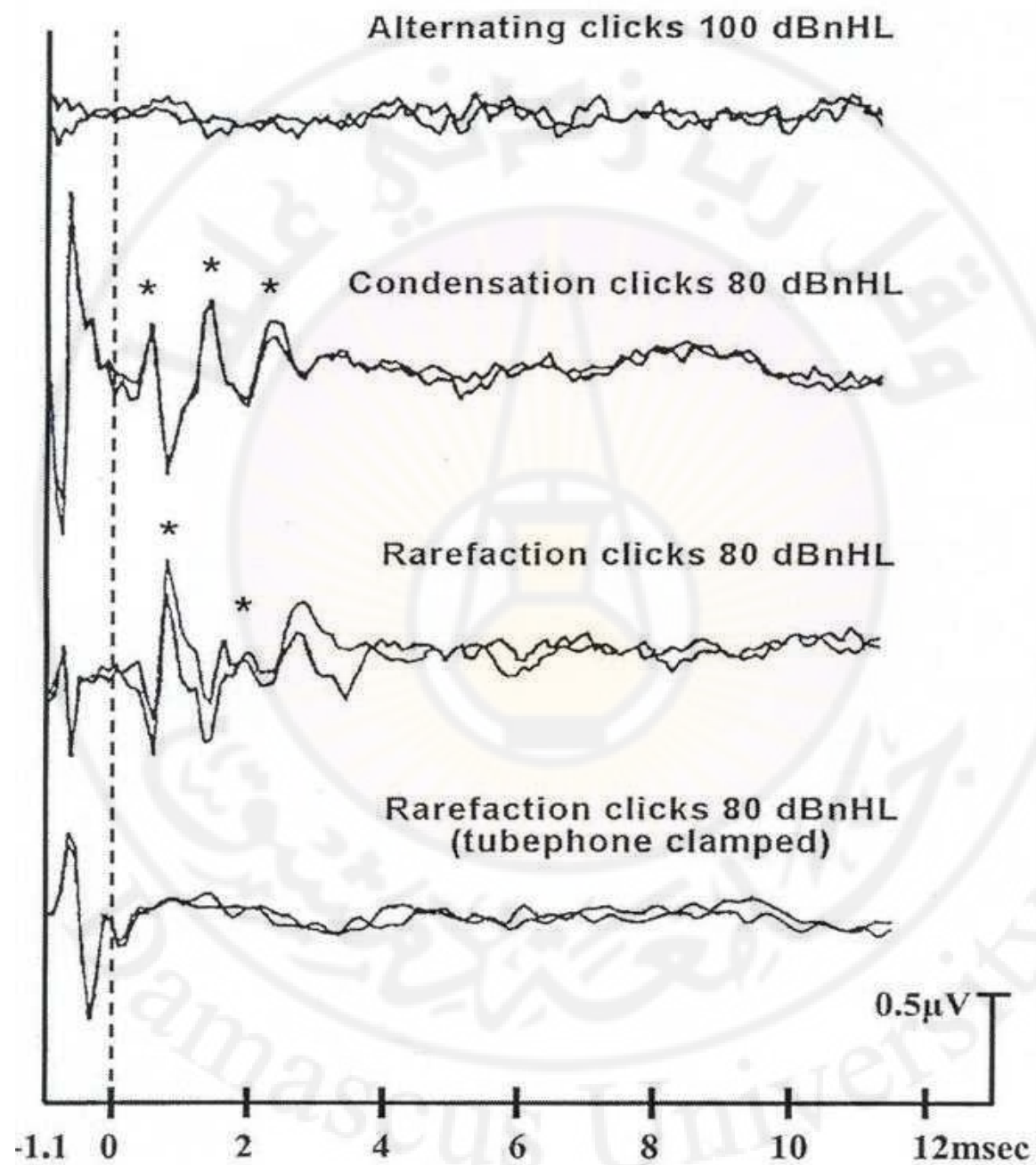
Click Stimulus 25/second 80dBnHL Insert Earphones
 Rarefaction and Condensation Overlaid



	<u>Anatomy</u>	<u>Dysfunction</u>	<u>Location</u>	<u>Auditory Test</u>
SENSORY CELLS	<i>IHC</i> - (auditory receptors) - each synapses with myelinated dendrites of multiple Type I spiral ganglion neurons (receive a few OCB axons)	HAIR CELL DEATH/ DAMAGE	IHC	OAE? CM?
	<i>OHC</i> - (modulating) - groups synapse with unmyelinated dendrites of one Type II spiral ganglion neuron (innervated by OCB axons)		OHC	OAE CM
SYNAPSE	Hair cells with dendrites of the spiral ganglion neurons	SYNAPTIC BLOCK	Synapse	No specific test
SPIRAL GANGLION NEURONS	<i>Type I</i> (first order auditory neurons) myelinated dendrites synapse with IHC; axons project to brainstem CN	NEURONOPATHY (ganglionopathy)	Spiral ganglion	No specific test
	<i>Type II</i> (sensory neurons) small unmyelinated dendrites synapse with OHC; physiologic properties unknown; axons project to brainstem CN			

It is becoming apparent that the term *auditory neuropathy*, as it is defined clinically, may represent at least two fairly distinct disorders, one preneural or sensory and the other neural.

VIII th NERVE	Afferent auditory axons (95% = <i>Auditory Type I</i>): myelinated axons; synapse with neurons of the CN (5% = <i>Modulating Type II</i>): scarce unmyelinated axons; synapse with neurons of the CN	NEUROPATHY Demyelinating Axonal Mixed	<i>Auditory Type I</i>	ABR Wave I ABR Wave II MEMR (acoustic)
	Efferent OCB axons (auditory) Axons from SOC via Medial OC (myelinated); target: OHC Sparse lateral OC (unmyelinated); target: dendrites of Type I SGN		OCB	OAE suppression (OHC activity modified though efferent MOC connections)
	Afferent vestibular axons Myelinated vestibular axons (first order neurons) synapse with 2 nd -order vestibular neurons in BS		Vestibular	Vestibular tests
BRAINSTEM PATHWAY	Cochlear nucleus (second order auditory neurons) Superior olivary complex Inferior colliculus	BRAINSTEM DISORDER	Brainstem	ABR Wave III ABR Wave V MEMR (acoustic & tactile)
THALAMUS	Medial geniculate body	THALAMO-CORTICAL DISORDER	Thalamo-cortex	MLR MMN
CORTEX	Primary auditory cortex (Heschl's gyrus) Secondary auditory cortex (superior temporal gyrus)		Cerebral cortex	CAEP late obligatory event related potentials



Neoplasm

- Unlike those in adults, tumors of the posterior fossa in children are less likely to be acoustic schwannoma and more likely to be intrinsic tumors, such as gliomas and ependymomas.
- These benign tumors of the cerebellopontine angle affect the auditory system when they impinge on the eighth cranial nerve.
- The most common form of acoustic tumor in children is that found in association with neurofibromatosis type 2 (NF2).
- **NF2** is characterized by **bilateral cochleovestibular schwannomas** that are faster-growing and more virulent than the unilateral type. This is an autosomal dominant disease and is associated with other intracranial tumors. Hearing loss in NF2 is not particularly different from that due to a unilateral type of schwannoma, except that it is bilateral and often progresses more rapidly.

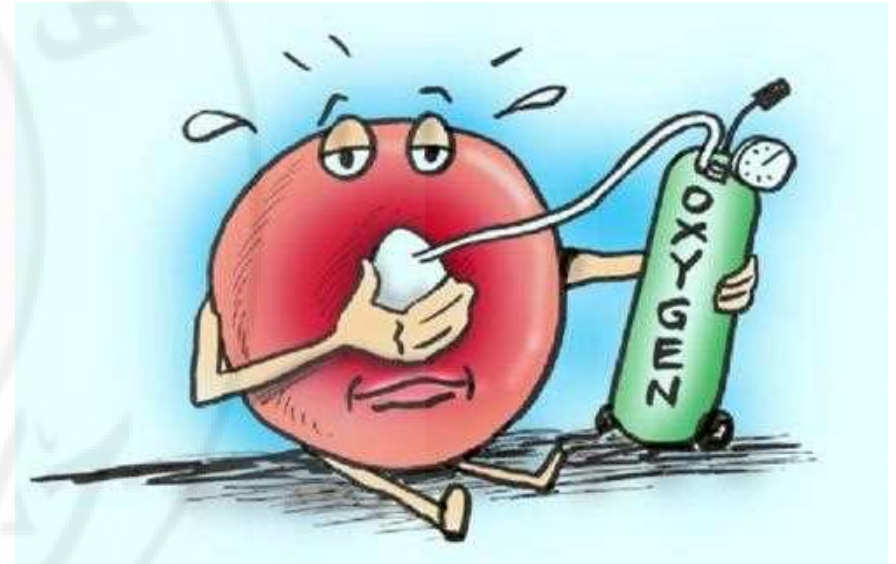
Hydrocephalus

- The cause of neural hearing disorders in children has also been attributed to more diffuse sources, including hydrocephalus, hypoxia, and hyperbilirubinemia.
- In hydrocephalus, the most common finding is one of neuromaturational delay of the auditory system as measured on the ABR.
- This is usually due to enlarged ventricles and is not associated with permanent changes in auditory function.



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.



Hyperbilirubinemia

- Hyperbilirubinemia is an excess of bilirubin in the blood that can be caused by many factors.
- Hyperbilirubinemia is associated with auditory neuropathy and other neural hearing disorders.
- The clinical spectrum of bilirubin-induced auditory toxicity ranges from transient auditory dysfunction to permanent sensory hearing loss when bilirubin levels are high.
- Audiometric findings include predominantly high-frequency bilateral and symmetric hearing loss with recruitment and abnormal loudness growth.
- Auditory nerve and generalized auditory brainstem dysfunction may occur. Other, more diffuse APDs have also been associated with hyperbilirubinemia.



5. Newborn Hearing Screening & EHDI

Definition

- Newborn hearing screening is the standard of care in hospitals worldwide.
- The primary purpose of newborn hearing screening is to identify newborns who are likely to have hearing loss and who require further evaluation.
- A secondary objective is to identify newborns with medical conditions that can cause late-onset hearing loss and to establish a plan for continued monitoring of their hearing status.
- Screening programs target **permanent childhood hearing loss (PCHL)** irrespective of type.

History

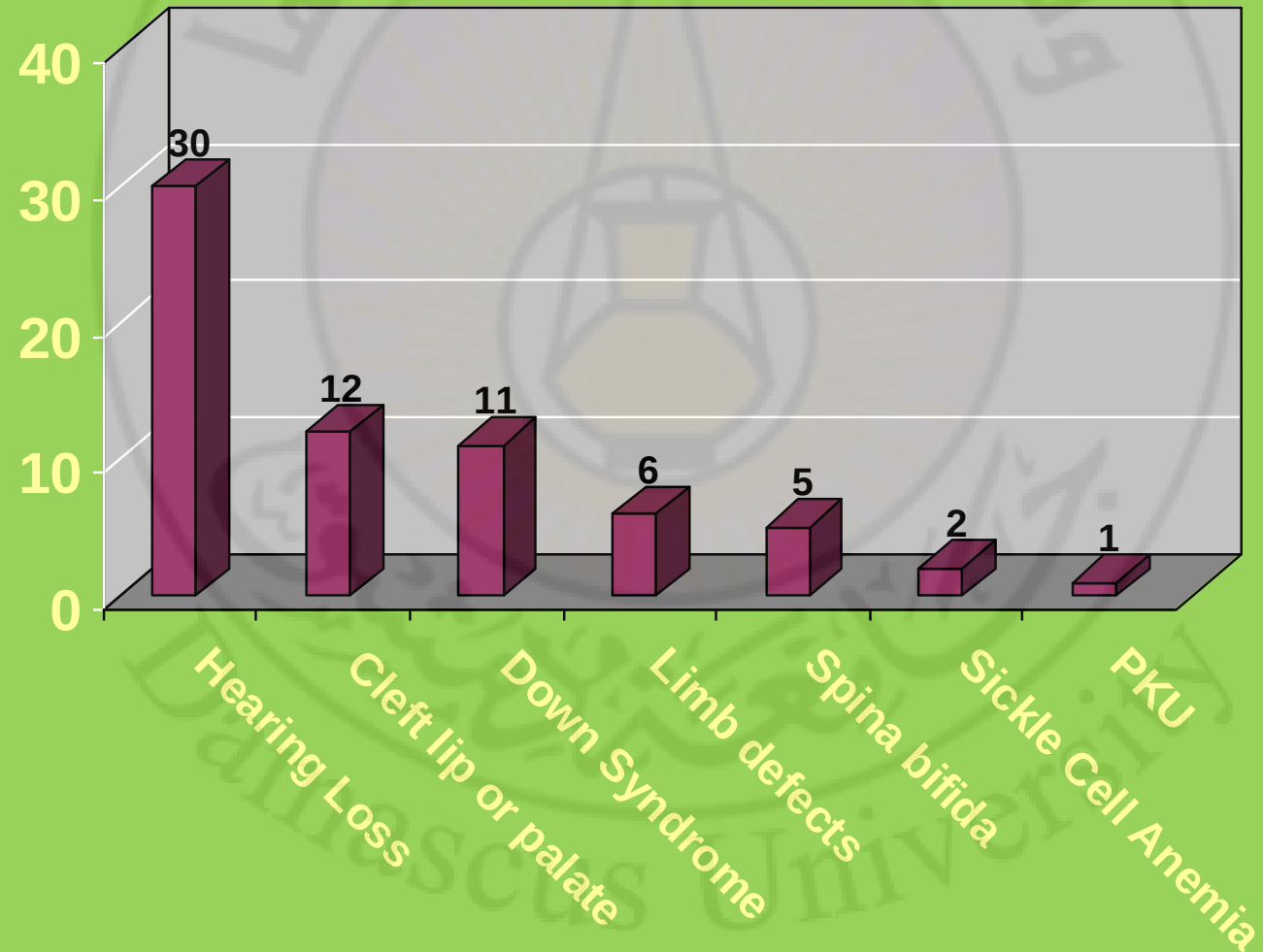
- Historically: Children who are deaf or hard of hearing (DHH) were not identified until 2-3 years of age
- JCIH established in 1969
- 1st Position Statement was in 1971; 1 page document
- Late 1980s: Recognition of new technologies and opportunities for early diagnosis and prompt intervention
- 1990s: Newborn Hearing Screening Programs emerged



Why is Early Identification of Hearing Loss so Important?

- Hearing loss is the most frequent birth defect.

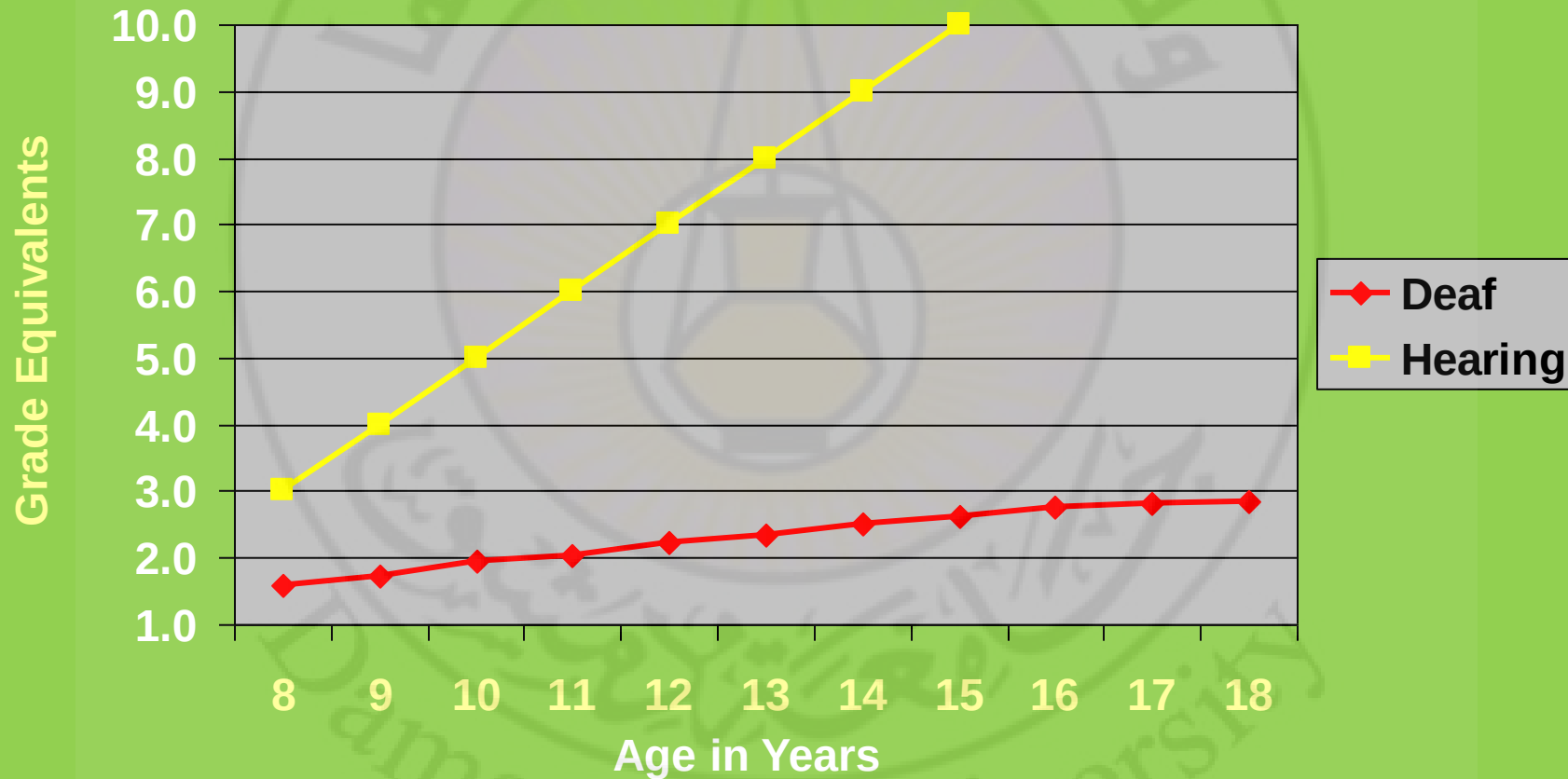
Incidence per 10,000 of Congenital Defects/Diseases



Why is Early Identification of Hearing Loss so Important?

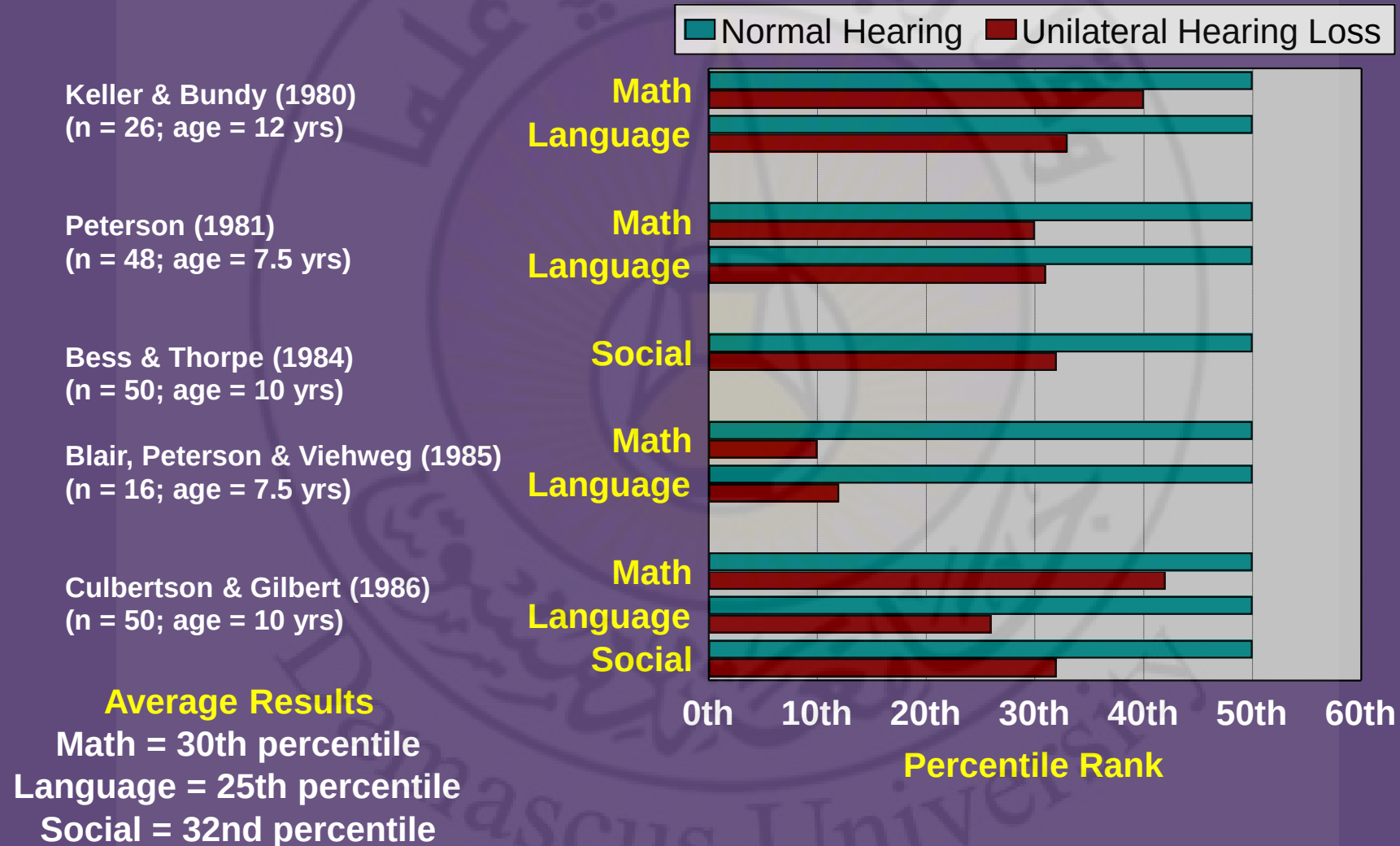
- Hearing loss is the most frequent birth defect.
- Undetected hearing loss has serious negative consequences.

Reading Comprehension Scores of Hearing and Deaf Students



Schildroth, A. N., & Karchmer, M. A. (1986). Deaf children in America, San Diego: College Hill Press.

Effects of Unilateral Hearing Loss



Why is Early Identification of Hearing Loss so Important?

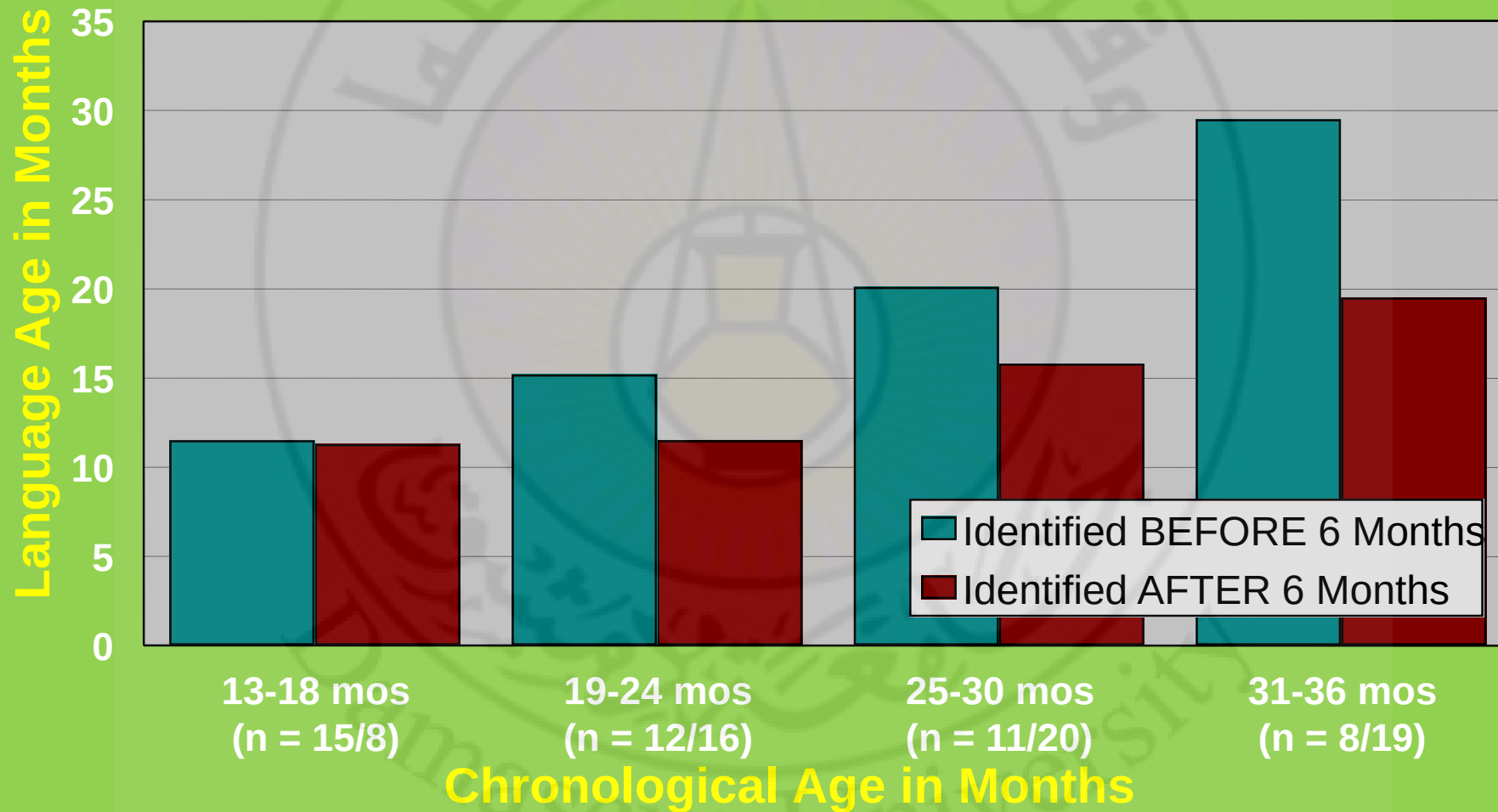
- Hearing loss is the most frequent birth defect.
- Undetected hearing loss has serious negative consequences.
- There are dramatic benefits associated with early identification of hearing loss.

Yoshinaga-Itano, et al., 1996

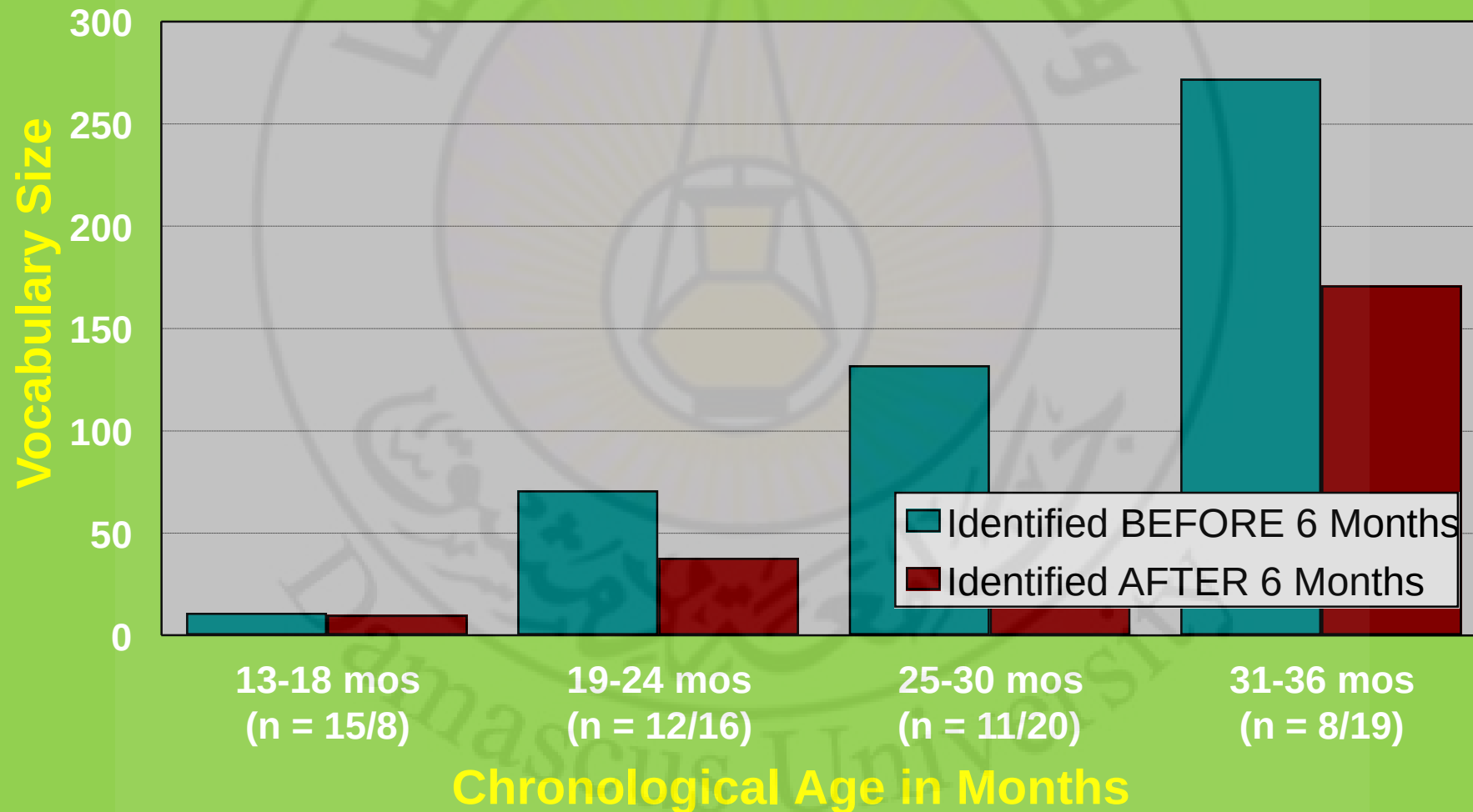
- ⌚ Compared language abilities of hearing-impaired children identified before 6 months of age ($n = 46$) with similar children identified after 6 months of age ($n = 63$).
- ⌚ All children had bilateral hearing loss ranging from mild to profound, and normally-hearing parents.
- ⌚ Language abilities measured by parent report using the Minnesota Child Development Inventory (expressive and comprehension scales) and the MacArthur Communicative Developmental Inventories (vocabulary).
- ⌚ Cross-sectional assessment with children categorized in 4 different age groups.

Yoshinaga-Itano, C., Sedey, A., Apuzzo, M., Carey, A., Day, D., & Coulter, D. (July 1996). The effect of early identification on the development of deaf and hard-of-hearing infants and toddlers. Paper presented at the Joint Committee on Infant Hearing Meeting, Austin, TX.

Expressive Language Scores for Hearing Impaired Children Identified Before and After 6 Months of Age

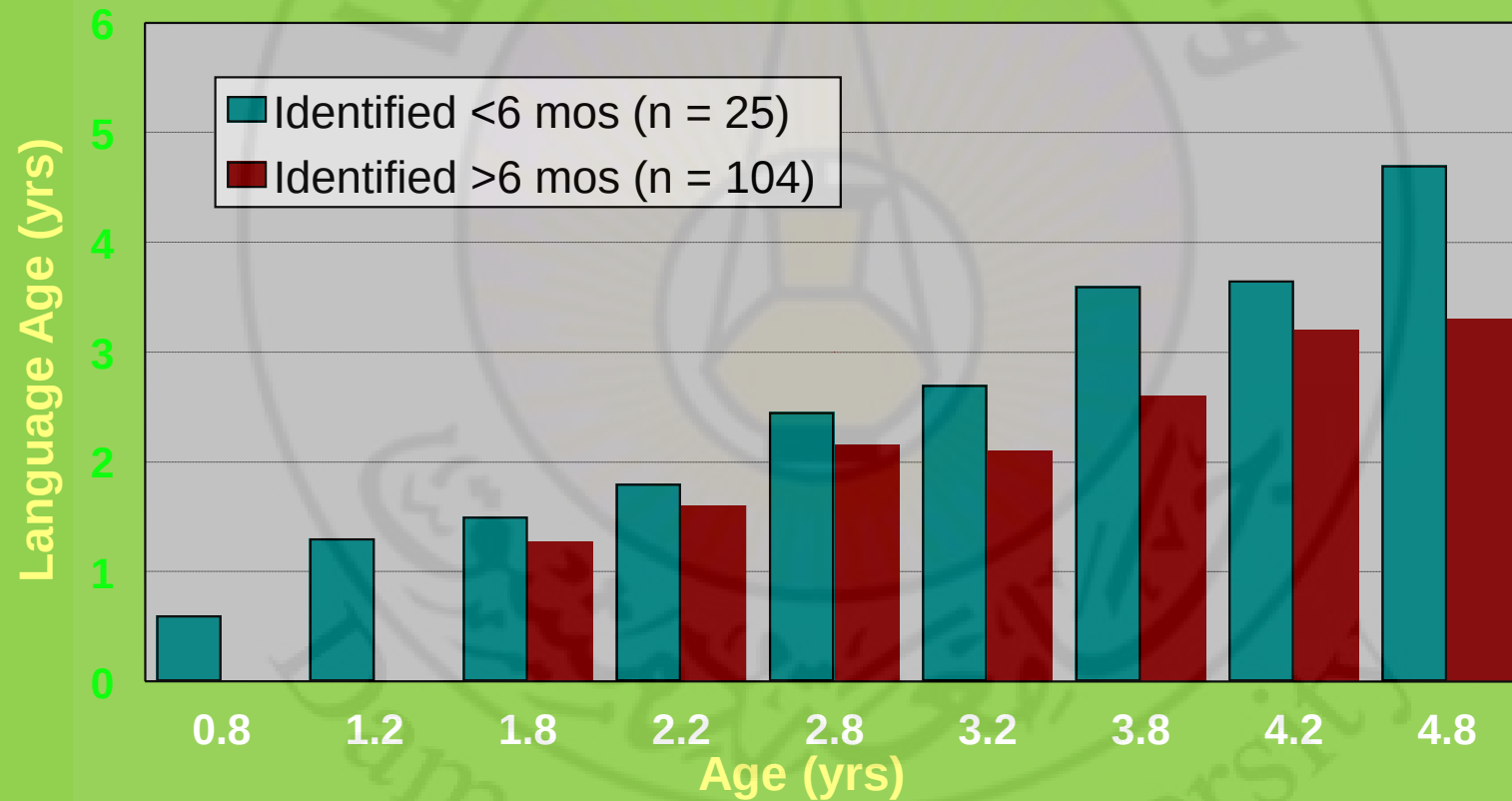


Vocabulary Size for Hearing Impaired Children Identified Before and After 6 Months of Age



Boys Town National Research Hospital Study of Earlier vs. Later

- ① 129 deaf and hard-of-hearing children assessed 2x each year.
- ① Assessments done by trained diagnostician as normal part of early intervention program.

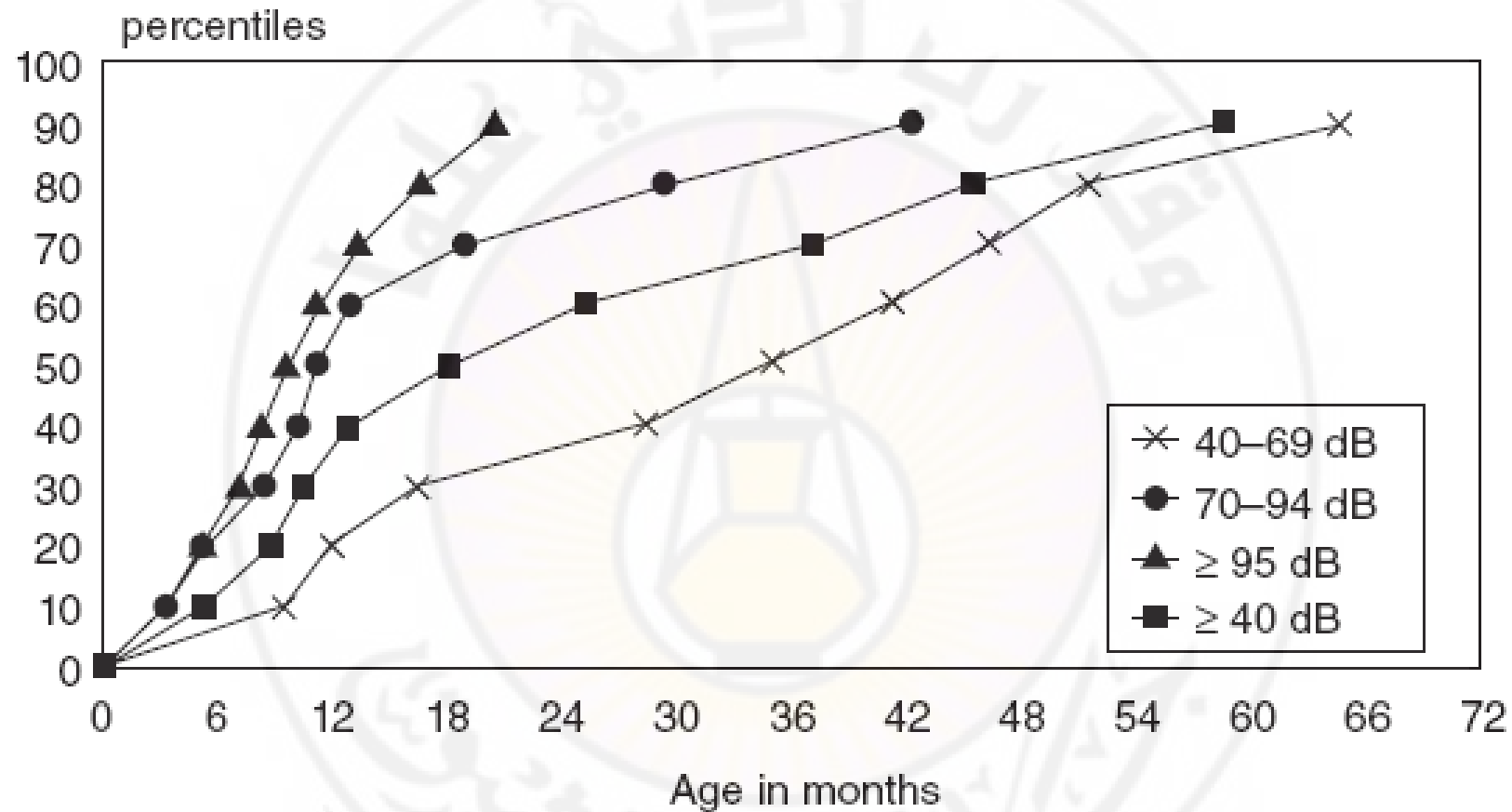


Moeller, M.P. (1997). Personal communication moeller@boystown.org

Introduction

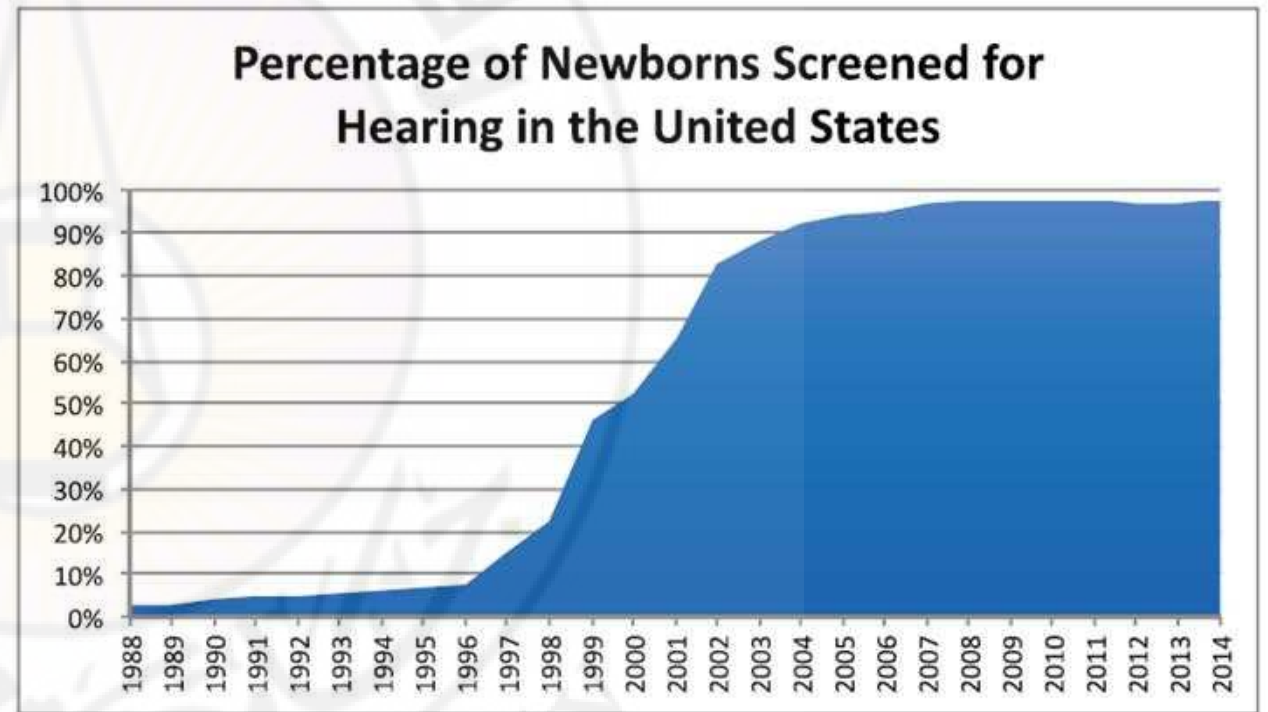
- In the absence of an effective hearing screen, the detection of **permanent childhood hearing impairment** can be delayed by as much as 2 to 3 years.
 - (**PCHI**; average hearing levels (0.5, 1, 2 and 4 kHz) measured at ≥ 40 dB nHL in the better ear)
- Even with screening programmes in place, considerable delays can occur.
- The lack of an effective screen has limited our knowledge of **the age of onset** of hearing impairment in early life, as most studies have had to rely mainly on retrospective analysis of case studies. These studies have indicated that the majority of PCHI is probably present at birth.





Age of confirmation of permanent hearing impairment ($N = 309$). Source: Fortnum and Davis 1997.

- Virtually all newborns in the United States are screened for hearing loss before they are a month old.
- Because of federally funded initiatives, research and technology advances, and legislative mandates in 43 states, newborn hearing screening has become the standard of care in the United States.



Principles Of Screening

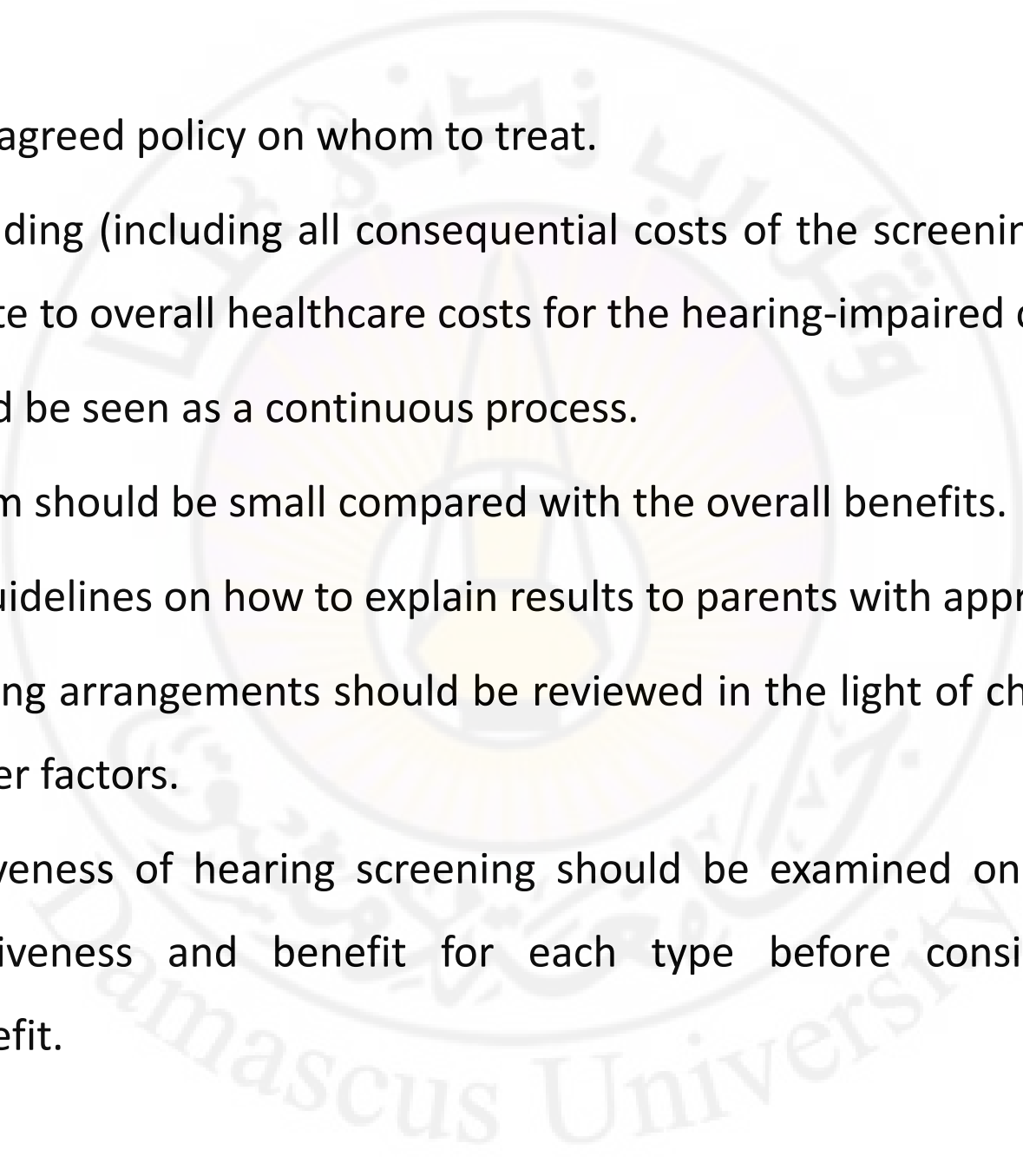
Principles of screening can be stated as follows:



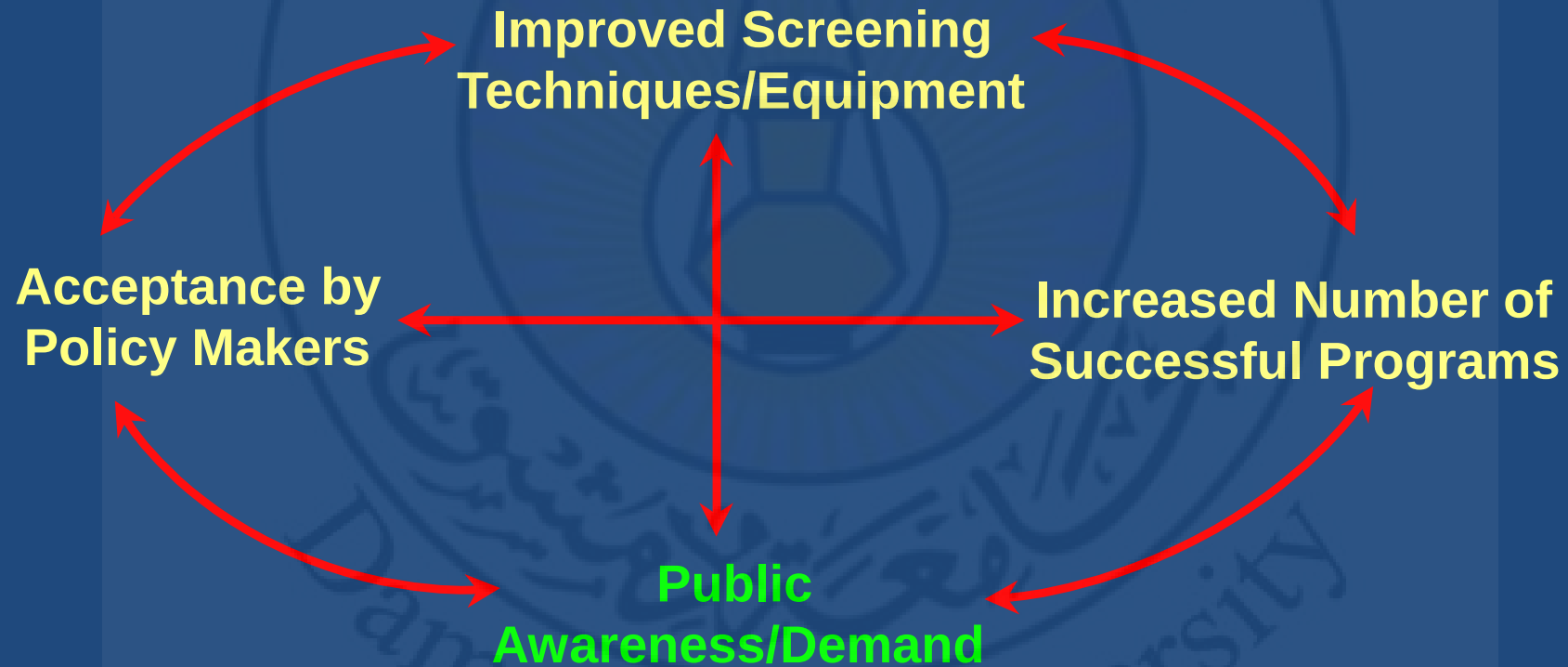
Principles

1. The condition (hearing impairment) should be an important health problem.
2. There should be an accepted treatment, i.e. an acceptable means of habilitation for those identified by the screen.
3. Facilities for assessment, diagnosis and treatment should be available.
4. The hearing impairment should be recognizable at an early stage.
5. There should be a suitable test for use as the screen.
6. The test should be acceptable to the parents and to the child.
7. The natural history of the condition should be known and understood.



- 
8. There should be an agreed policy on whom to treat.
 9. The cost of case finding (including all consequential costs of the screening programme) should not be disproportionate to overall healthcare costs for the hearing-impaired child.
 10. Case finding should be seen as a continuous process.
 11. The incidental harm should be small compared with the overall benefits.
 12. There should be guidelines on how to explain results to parents with appropriate support.
 13. All hearing screening arrangements should be reviewed in the light of changes in demography, epidemiology and other factors.
 14. Costs and effectiveness of hearing screening should be examined on a case type basis to maximize the effectiveness and benefit for each type before considering overall costs, effectiveness and benefit.

Why is Implementation of Newborn Hearing Screening Accelerating?



NHS or EHDI?

- As a result of work done by JCIH and the Centers for Disease Control and Prevention (CDC), most people stopped using the phrase “universal newborn hearing screening” and began referring to “**early hearing detection and intervention**” (EHDI) programs.
- The change in how these programs are described is important, because it emphasizes that to identify and serve infants and young children who are deaf or hard of hearing, successful screening programs must be coupled with *timely diagnosis; appropriate medical, audiology, and educational services; coordination with the child’s primary health care provider (often referred to as the child’s medical home); and tracking and data management systems.*





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ASHA
American
Speech-Language-Hearing
Association



Voting Organizations

Risk Factors for Early Childhood Hearing Loss:

	Risk Factor Classification	Recommended Diagnostic Follow-up	Monitoring Frequency
	Perinatal		
1	Family history* of early, progressive, or delayed onset permanent childhood hearing loss	by 9 months	Based on etiology of family hearing loss and caregiver concern
2	Neonatal intensive care of more than 5 days	by 9 months	As per concerns of on-going surveillance of hearing skills and speech milestones
3	Hyperbilirubinemia with exchange transfusion regardless of length of stay	by 9 months	
4	Aminoglycoside administration for more than 5 days**	by 9 months	
5	Asphyxia or Hypoxic Ischemic Encephalopathy	by 9 months	
6	Extracorporeal membrane oxygenation (ECMO)*	No later than 3 months after occurrence	Every 12 months to school age or at shorter intervals based on concerns of parent or provider
7	In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis	by 9 months	As per concerns of on-going surveillance
	In utero infection with cytomegalovirus (CMV)*	No later than 3 months after occurrence	Every 12 months to age 3 or at shorter intervals based on parent/provider concerns
	Mother + Zika and infant with <u>no</u> laboratory evidence & no clinical findings	standard	As per AAP (2017) Periodicity schedule
	Mother + Zika and infant with laboratory evidence of Zika + clinical findings Mother + Zika and infant with laboratory evidence of Zika - clinical findings	AABR by 1 month AABR by 1 month	ABR by 4-6 months or VRA by 9 months ABR by 4-6 months Monitor as per AAP (2017) Periodicity schedule (Adebanjo et al., 2017)

Risk Factors for Early Childhood Hearing Loss:

8	Certain birth conditions or findings: • Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia • Congenital microcephaly, congenital or acquired hydrocephalus • Temporal bone abnormalities	by 9 months	As per concerns of on-going surveillance of hearing skills and speech milestones
9	Over 400 syndromes have been identified with atypical hearing thresholds***. For more information, visit the Hereditary Hearing Loss website (Van Camp & Smith, 2016)	by 9 months	According to natural history of syndrome or concerns
	Perinatal or Postnatal		
10	Culture-positive infections associated with sensorineural hearing loss***, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis	No later than 3 months after occurrence	Every 12 months to school age or at shorter intervals based on concerns of parent or provider
11	Events associated with hearing loss: • Significant head trauma especially basal skull/temporal bone fractures • Chemotherapy	No later than 3 months after occurrence	According to findings and or continued concerns
12	Caregiver concern**** regarding hearing, speech, language, developmental delay and or developmental regression	Immediate referral	According to findings and or continued concerns

Note. AAP = American Academy of Pediatrics; ABR = auditory brainstem response; AABR = automated auditory brainstem response.

* Infants at increased risk of delayed onset or progressive hearing loss

**Infants with toxic levels or with a known genetic susceptibility remain at risk

***Syndromes (Van Camp & Smith, 2016)

****Parental/caregiver concern should always prompt further evaluation.

EHDI 1-3-6 Goals

1. All infants should undergo hearing screening prior to discharge from the birth hospital and no later than one month of age, using physiologic measures with objective determination of outcome.
2. All infants whose initial birth-screen and any subsequent rescreening warrant additional testing should have appropriate audiologic evaluation to confirm the infant's hearing status no later than 3 months of age.
3. A concurrent or immediate comprehensive otologic evaluation should occur for infants who are confirmed to be deaf or hard of hearing.
4. All infants who are deaf or hard of hearing in one or both ears should be referred immediately to early intervention in order to receive targeted and appropriate services.

1

SCREENING

3

DIAGNOSIS

6

INTERVENTION

EHDI 1-3-6 Goals

5. A simplified, coordinated point of entry into an intervention system appropriate for identified children is optimal.
6. Early intervention services should be offered through an approach that reflects the family's preferences and goals for their child, and should begin as soon as possible after diagnosis but no later than six months of age.
7. The child and family should have immediate access, through their audiologist, to high-quality, well-fitted, and optimized hearing aid technology. Access should also be assured, depending on the child's needs, to cochlear implants (CI), hearing assistive technologies, and visual alerting and informational devices.
8. The EHDI system should be family-centered with infant and family rights and privacy guaranteed through informed and shared decision-making, and family consent in accordance with state and federal guidelines.

EHDI 1-3-6 Goals

9. Families should have access to information about all resources and programs for intervention, and support and counseling regarding the child's educational and communication/language needs.
10. All infants and children, regardless of newborn hearing screening outcome, should be monitored within the medical home according to the periodicity tables regarding their communication development.
11. Professionals with appropriate training should provide ongoing surveillance of communication development to all children with or without risk indicators.
12. Appropriate interdisciplinary early intervention programs for identified infants and their families should be provided by professionals knowledgeable about the needs and requirements of children who are deaf or hard of hearing (JCIH, 2013).
13. Early intervention programs should recognize evidence-based practices and build on strengths, informed choices, language traditions, and cultural beliefs of families they serve.
14. EHDI information systems should be designed and implemented to interface with clinical electronic health records and population-based information systems to allow the exchange of electronic health information for the purposes of outcome measurement, quality improvement, and reporting the effectiveness of EHDI services for the patient/family within the medical home, healthcare community, state, and federal levels.

JCIH 2007

EHDI Benchmarks

JCIH 2019

1 month

Hearing Screening
Outpatient Rescreen
(if needed)

1 month

3 months

Diagnostic Hearing Evaluation
by Audiologist
Medical and Otologic Evaluation

2 months

6 months

Early Intervention
Enrollment in Part C Services

3 months

Programs that currently meet the 1-3-6 benchmark goals should strive for a 1-2-3 timeline to further promote early access to optimal language and learning.

Three Key Components of Early Hearing Detection & Intervention Programs



Damascus University

Technology

- Auditory brainstem response (ABR) and otoacoustic emissions (OAEs) are appropriate physiologic measures for screening the newborn population. Both are **noninvasive** and available in **automated versions** that are easily utilized by trained hospital staff.
- Both ABR and OAE technologies will miss **delayed-onset hearing loss, mild hearing loss**, or hearing loss that is present only at **isolated frequencies**.
- Both ABR and OAE responses are affected by outer or middle ear dysfunction. When a transient middle ear condition is present, both technologies will likely result in the newborn not passing the screening.
- Both OAE and ABR screening reflect physiologic processes within the auditory system and identify hearing loss most accurately from 2kHz to 4kHz.



Technology

- Automated technologies (those that determine pass/refer) do not require interpretation.
- Automated screening equipment often has test parameters set by the manufacturer, and, therefore, different equipment may yield different screening results.
- Even if diagnostic (i.e., nonautomated) technology is used, and an audiologist is interpreting the results, procedures in the nursery are limited to screening (pass/refer).
- Equipment that is used should be calibrated and maintained according to the manufacturer's specifications.



Auditory Brainstem Response (ABR)

- The ABR is recorded using surface electrodes that are attached to the infant's head.
- Click stimuli are presented through insert or muff-style earphones that are worn on both ears.
- ABR measurements are sensitive to neural auditory disorders (i.e., auditory neuropathy). *Auditory neuropathy* is a neural hearing loss that leaves cochlear (outer hair cell) function intact.
- ABR is more prevalent in the NICU population than in the well-baby nursery population.
- ABR screening is less sensitive to outer ear debris than OAE screening, resulting in lower referral rates.



Auditory Brainstem Response (ABR)

- **Stimuli:** Most automated equipment presents click stimuli at 35 dB nHL at a rate of 30-37 clicks per second. If the equipment allows a choice of stimulus levels, the screening program audiologist can adjust to lower click levels.
- **Response Criteria:** For automated ABR (AABR), manufacturers use their own proprietary stopping rule, based on a template comparison or statistical algorithms. Most instrumentation does not allow operators to change the stopping rule criteria.
- **Procedure:** The infant should be asleep or resting quietly for the test and positioned to reduce muscle artifact. The screener visually inspects the outer part of the ear canal to ensure that the canal is clear of debris and places the transducer. Both ears are screened during each session.



Otoacoustic Emissions (OAE)

- OAEs – either transient-evoked OAEs (TEOAEs) or distortion product OAEs (DPOAEs) – are measured using a sensitive probe microphone inserted into the infant's ear canal.
- OAEs are a direct measure of outer hair cell and cochlear function in response to acoustic stimulation. They yield an indirect estimate of peripheral hearing sensitivity.
- OAEs are not sensitive to disorders central to the outer hair cells, such as auditory neuropathy.
- OAEs will be absent when there is outer or middle ear dysfunction or debris/blockage in the ear canal.



Otoacoustic Emissions (OAE)

- **Stimuli:** **TEOAEs** use a high-level click, approximately 80 dB pSPL, and a subtraction (*nonlinear*) paradigm to reduce stimulus artifact. **DPOAEs** use mid-level stimuli (f1 primary = 65 dB SPL and f2 primary = 55 dB SPL).
- **Response Criteria:** At least three test frequencies – of 2000 Hz, 3000 Hz, and 4000 Hz – are evaluated during the screening. Usually, signal-to-noise ratios (SNRs) of at least 6 dB are used; however, some manufacturers will set their own SNRs. DPOAE SNRs vary, depending on whether mean SNR is calculated or mean plus 1 or 2 standard deviations of noise. In addition, a minimum absolute DPOAE level of 5 dB SPL is imposed.



Otoacoustic Emissions (OAE)

- **Procedure:** The infant should be asleep or resting quietly for the test and positioned to reduce muscle artifact.
- A snug probe fit is essential for valid and reliable recordings. Ears should be screened one at a time, with the infant placed on his/her side and the ear being screened facing up. The screener visually inspects the outer part of the ear canal to ensure that the canal is patent and clear of debris. Prior to insertion of the probe, a gentle massage of the area below the tragus helps to open a collapsed canal or dislodge debris that may be blocking the canal.
- After stimulus-level requirements have been met, OAEs are collected to meet stopping criteria.
- If OAEs do not appear to be present, the probe is taken out and inspected to determine if the probe is blocked with cerumen or vernix. A blocked probe should be cleaned and reinserted, and the screening should be repeated.



Timing of Screening

- Newborns cared for in the well-baby nursery are screened as close to hospital discharge as possible and prior to 1 month of age.
- NICU newborns are screened when they are ready for discharge and/or when they are medically stable.
- Newborns who have initially passed a hearing screening are rescreened if readmitted to the hospital in the first month of life or if risk factors for hearing loss develop during the infant's hospital stay following the initial screening.
- Regional laws and hospital protocols may vary regarding which hospital is responsible for screening newborns who are transferred from one hospital to another.

Testing Environment

- Screening can be done in a nursery or a quiet room with the infant resting quietly or sleeping. A sound booth is not needed.
- The preferred method for testing is to have the newborn resting quietly in his/her bassinette – although, if needed, the newborn can be held.

Pass/Refer Indications

- A newborn must pass the screening in both ears during one session for the screening to be considered a "pass." Otherwise, the newborn will be referred for rescreening.
- If the newborn does not pass in one ear, both ears must be rescreened.
- If the newborn passes the screening or the rescreening and has no risk factors for late-onset or progressive hearing loss, then the screening is complete.
- If the newborn passes the screening or the rescreening and has risk factors for late-onset or progressive hearing loss, then it will be very important to monitor the newborn's hearing during early childhood.

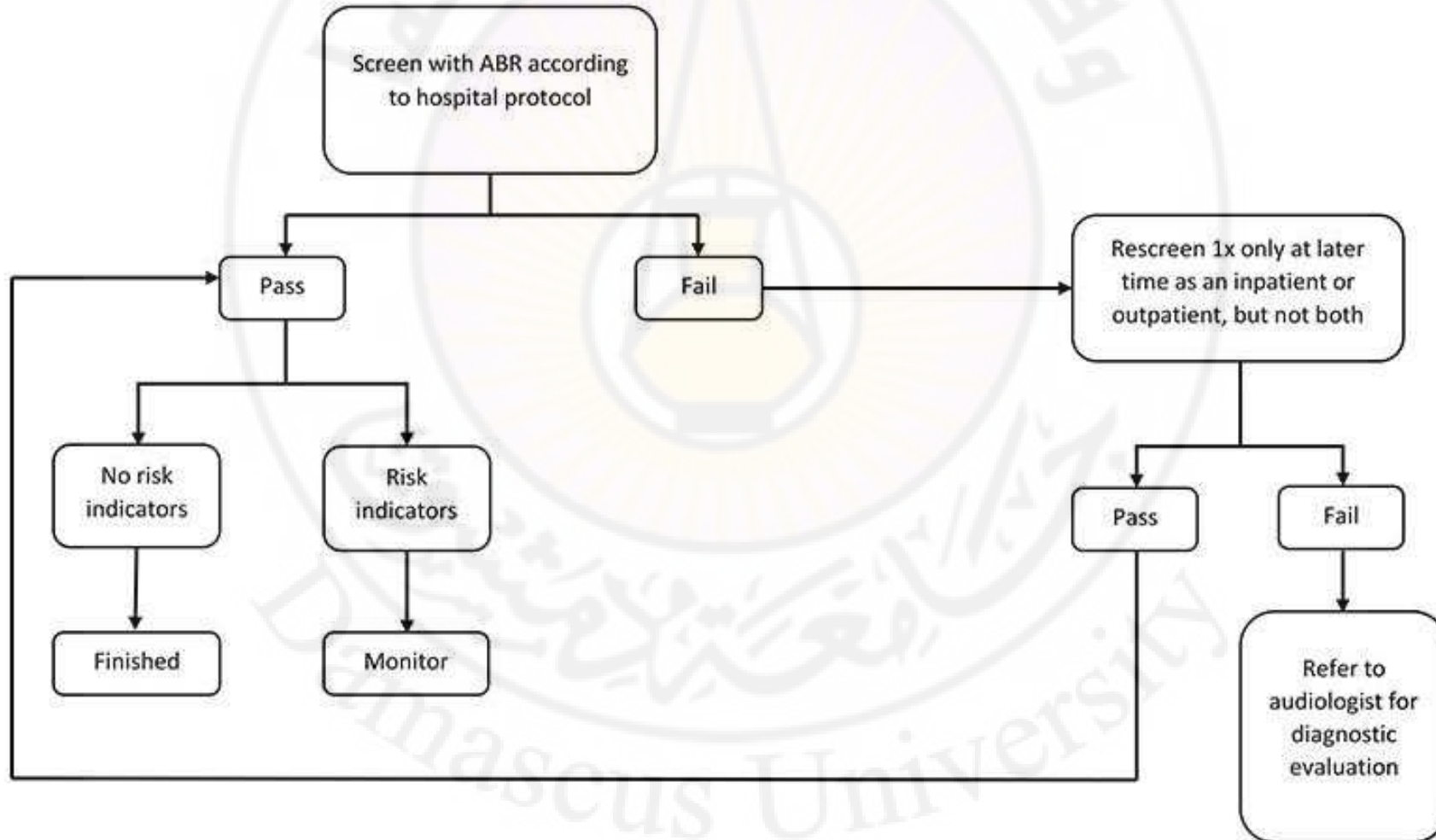
Standard Universal Precautions

- All procedures must ensure the safety of the patient and clinician and adhere to universal health precautions (e.g., prevention of bodily injury and transmission of infectious disease).
- Decontamination, cleaning, disinfection, and sterilization of multiple-use equipment before reuse must be carried out according to facility-specific infection control policies and procedures and according to the manufacturer's instructions.

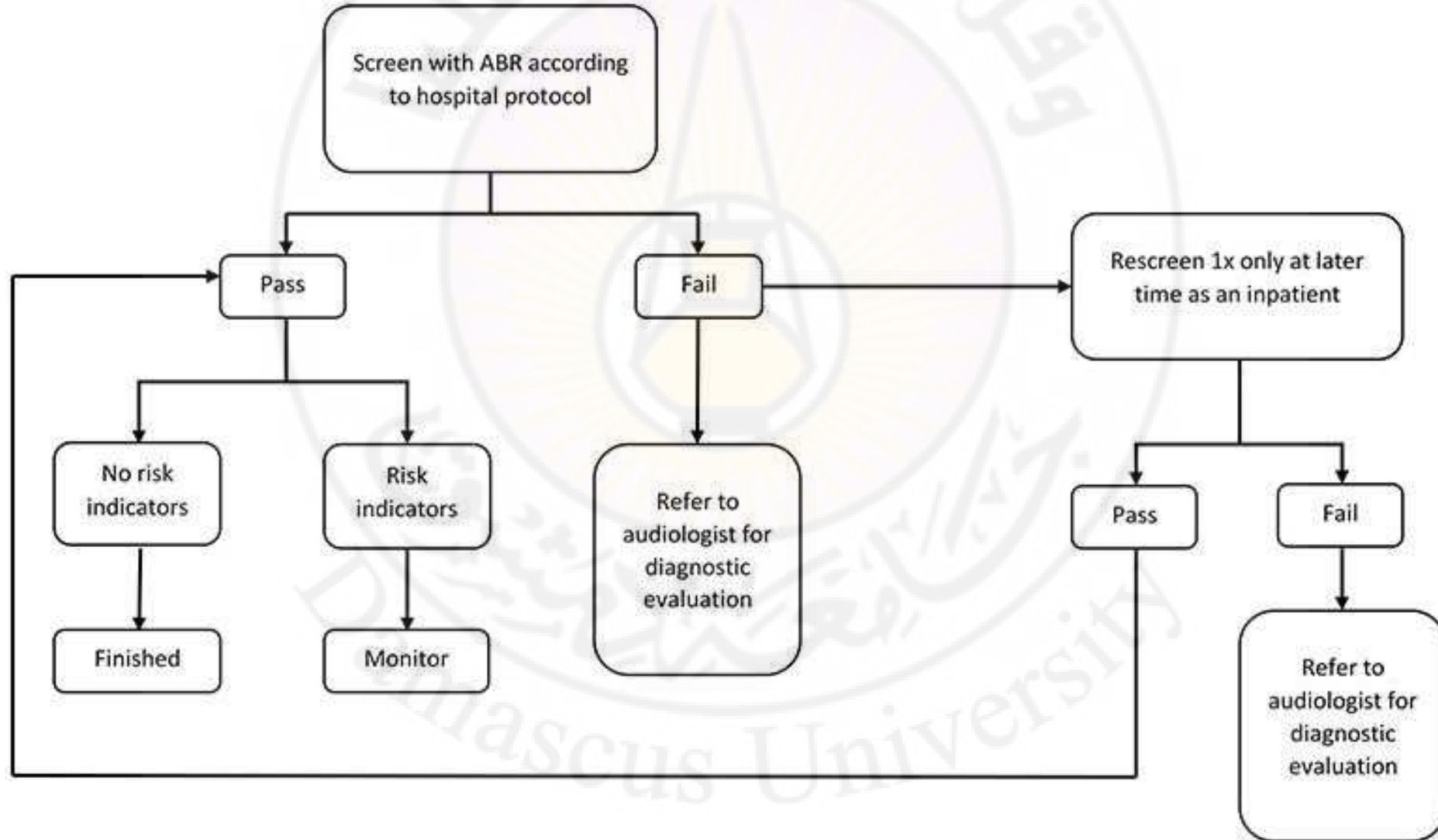
Protocols

- Screening protocols can be broadly classified into four different categories:
 - ABR only
 - OAEs only
 - Two-tier (OAEs with immediate ABR rescreening if OAE is not passed)
 - Two-technology (ABR and OAEs)
- The choice of protocol for a newborn hearing screening program is based on the specific needs of the population (i.e., well-baby nursery or NICU) and requirements of the state or hospital program.

Screening with ABR Only: Well-baby nursery



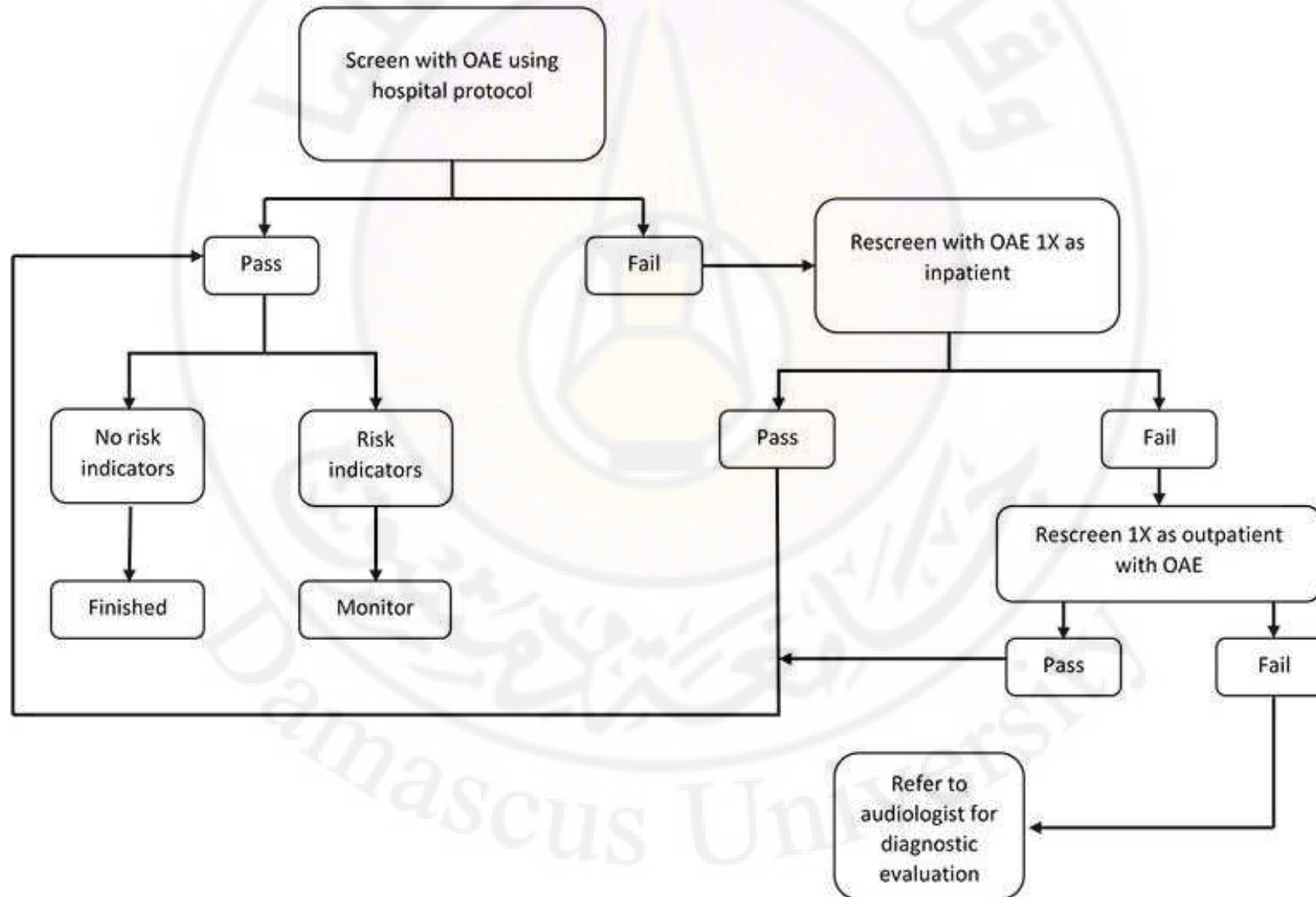
Screening with ABR Only: NICU



Screening with ABR Only: Factors To Consider

- More of the auditory system is accessed with ABR screening than with OAEs, allowing for detection of neurologic involvement.
- ABR screening results are less susceptible to false positives due to ear canal debris than are OAE screenings.
- ABR screening can be less cost effective than an OAE screening because of the higher cost of disposables (electrodes and disposable earphones) and increased personnel time. This cost differential may be balanced with a lower refer rate, resulting in reportedly lower or similar overall program costs.
- ABR test time can be longer than OAE test time because of electrodes application.
- ABR will result in false positives when used with babies who have immature neurological systems that affect the ABR waveform.

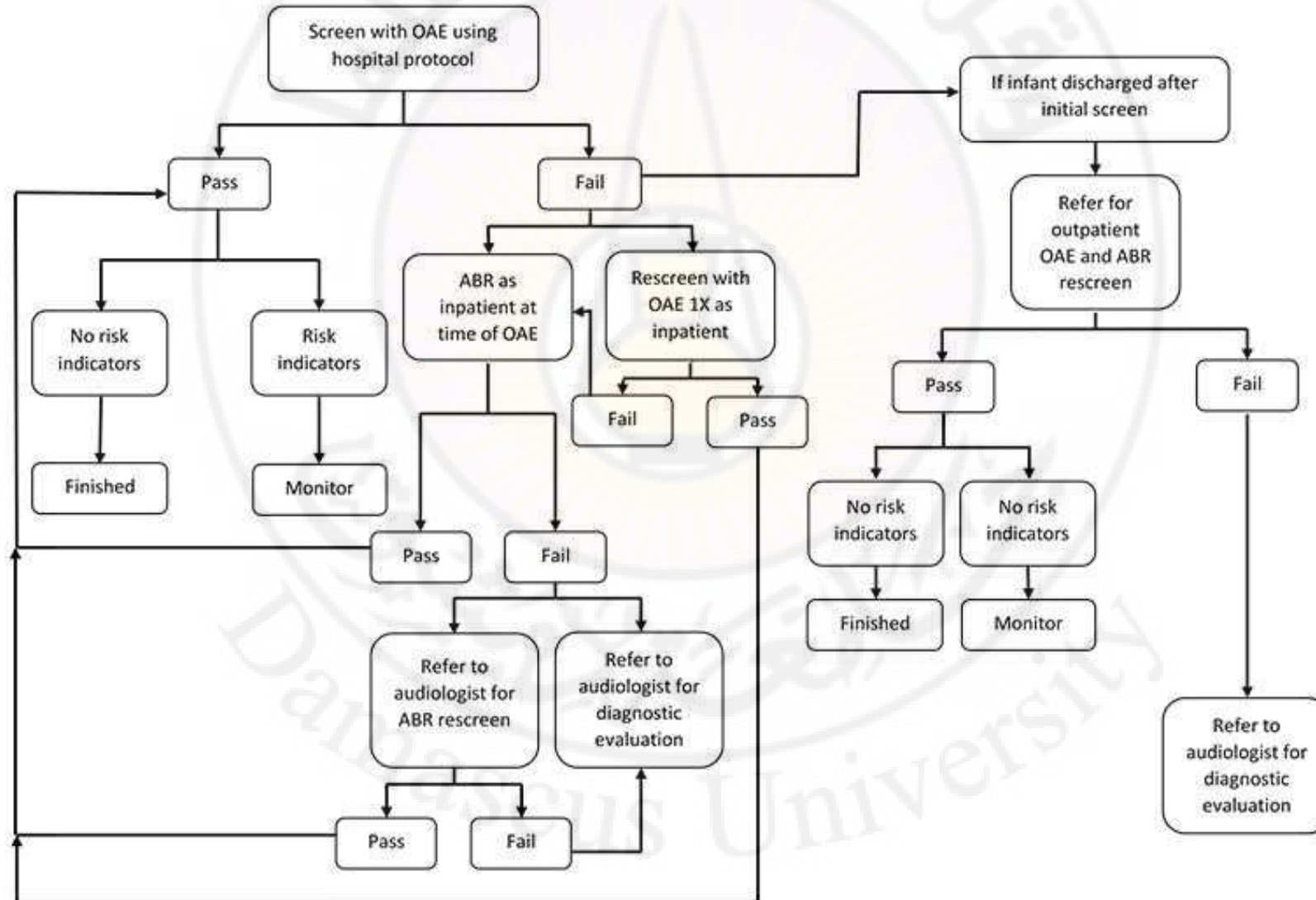
Screening With OAE Only: Well-baby nursery



Screening with OAE Only: Factors To Consider

- OAE disposable supplies are often less expensive than those used with ABR.
- OAE test time has been reported to be shorter than ABR test time.
- OAEs do not detect neural hearing losses.
- OAEs have a higher inpatient refer rate as compared to ABR. Outpatient rescreening is advised for newborns who do not pass inpatient screening before referral to an audiologist for diagnostic testing.
- OAEs may be reduced or absent due to outer ear debris and/or middle ear fluid common in the newborn population, resulting in higher refer rates. That is, OAEs have a higher false positive rate than ABR.

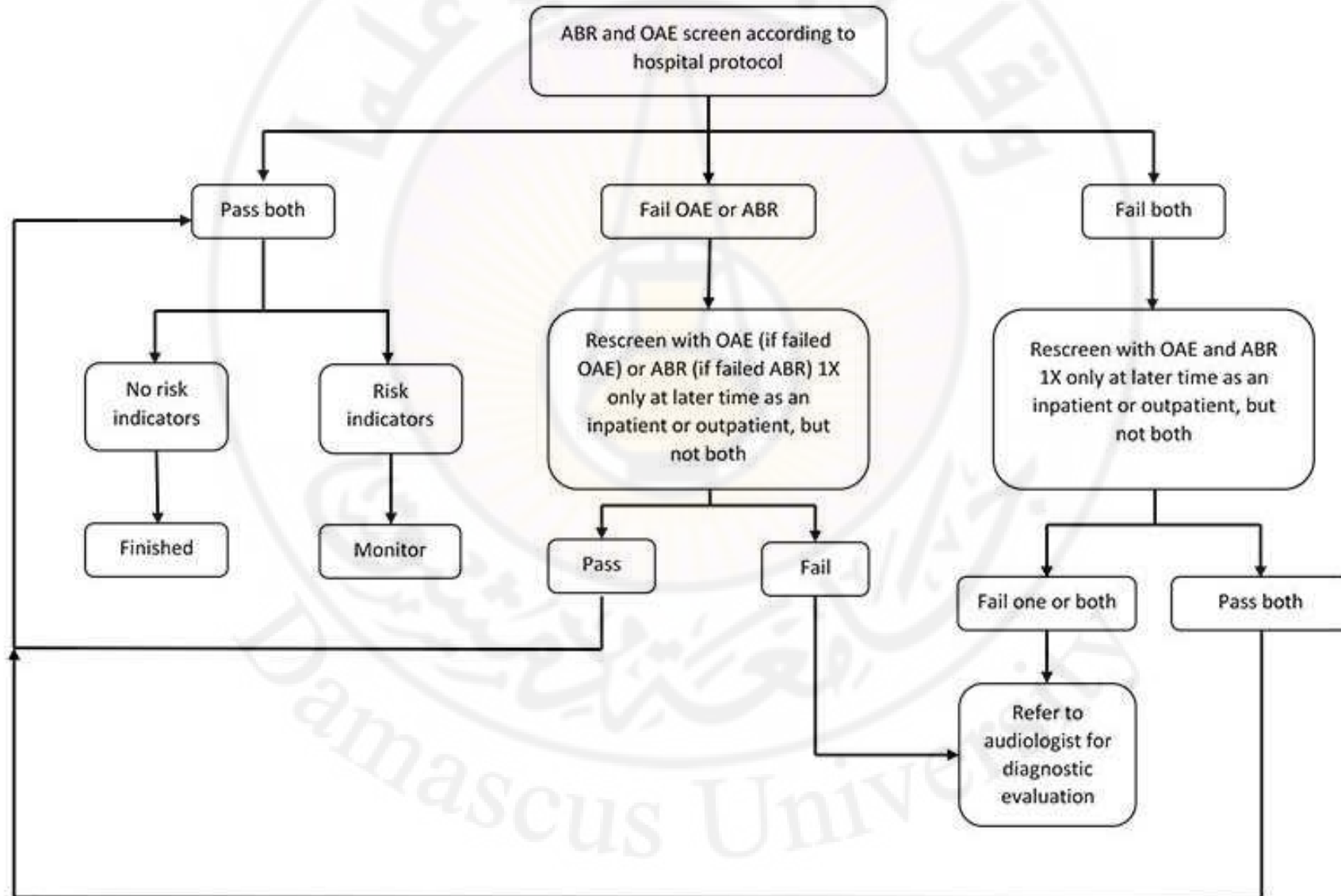
Two-Tier Screening: OAE Followed by ABR if the OAE is Not Passed



Two-tier protocol: Factors To Consider

- The number of referrals for diagnostic testing will be lower in a two-tier protocol than in an OAE-only protocol.
- Two types of disposables will be required, and two pieces of equipment may be needed, although some manufacturers have both tests on one piece of equipment.
- Personnel must be trained on two procedures.
- Because tests differ in detecting different degrees of hearing loss, a baby may not pass the OAE screening but pass the ABR screening. As a result, some children with mild hearing loss may be missed.
- A two-tier protocol may miss auditory neuropathy because infants who pass an OAE screening will not be referred for an ABR.

Two-Technology Screening: ABR and OAE



Two-Technology Screening: Factors To Consider

- A two-technology protocol is more likely to detect both mild hearing loss (due to the use of OAE technology) and neural hearing loss (due to the use of ABR technology).
- Two pieces of equipment or a device that includes both tests must be purchased and maintained, as well as two types of disposables.
- Personnel must be trained on both technologies.
- Additional screening time is required because two procedures are completed.
- Refer rates may be higher because two screening tests must be passed.

Detection of Late-Onset Hearing Loss

- The onset of hearing loss can occur at any time in a child's life.
- The JCIH recommends that the pediatric primary care provider regularly monitor all infants and children for hearing loss and communication development.
- This allows for the identification of children with either missed newborn, late-onset, or progressive hearing loss, regardless of the presence or absence of high-risk indicators at birth.
- Developmental milestones, hearing skills, and speech and language milestones should be monitored in all children.



In summary:

Passing a screening does not mean that a child has normal hearing across the frequency range. Because minimal and frequency-specific hearing losses are not targeted by newborn hearing screening programs, newborns with these losses may pass a hearing screening. Because these losses have the potential to interfere with the speech, language, and psychoeducational development of children, monitoring of hearing, speech, and language milestones throughout childhood is essential.

Counseling and Education

- Providing appropriate counseling and education is an important part of the newborn hearing screening process.
- Parent/guardian education regarding newborn hearing screening begins before an infant is screened.
- If information about the screening process is provided only after a mother is admitted to the hospital to deliver her child, she may not have the level of concentration needed to consider the information being presented.



Communicating Screening Results

- Where possible, **audiologists** are the professionals who communicate with the family when an infant has not passed a hearing screening.
- When communicating screening results, consider that:
 - communication is confidential and should be presented in a family-focused manner;
 - information should be delivered in a clear and concise manner that avoids technical jargon;
 - information should be provided in the family's preferred mode of communication and language;
 - findings should be presented in a positive manner, emphasizing the importance of appropriate follow-up; and
 - parents/guardians should be given the opportunity and encouraged to ask questions.



Communicating Screening Results

- If an infant is referred for further testing, the audiologist provides the information in such a way that the family feels supported yet clearly understands the importance of following up with further audiologic testing.
- Families of newborns who **pass** the hearing screening should be counseled about the need for ongoing surveillance and periodic monitoring.



**Blindness separates people from things.
Deafness separates people from people.**

--- Helen Keller



7. Hearing Test Protocols for Children



Hearing assessment in infants and children

- Pediatric audiologic assessments involve the selection of **developmentally appropriate protocols** that include the cross-check principle.
- Before testing, the child's cognitive age and physical status must be determined.
- A **case history** contributes valuable diagnostic information, provides an opportunity to observe the child, and allows a rapport to be established between the audiologist and the family.



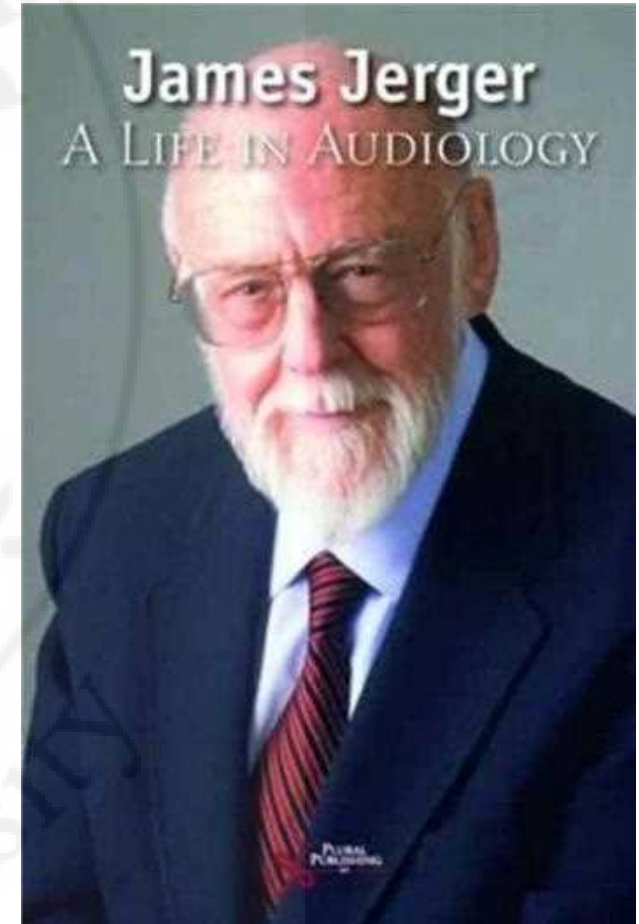
Main purposes:

1. To obtain a measure of **peripheral hearing sensitivity** that rules out or confirms hearing loss as a cause of the baby's or child's problem.
2. To confirm the status of the baby's or child's **middle ear**;
3. To assess auditory functioning using **speech perception** measures when possible;
4. To observe and interpret the baby's or child's **auditory behaviors**.



The Cross-Check Principle for Test Batteries

- The **cross-check principle**, originally described by Jerger and Hayes, posits that several appropriate behavioral and electrophysiologic tests must be used to determine the extent of a child's auditory function.
- A **test battery approach** furnishes detailed information, avoids drawing conclusions from a single test, allows for the identification of multiple pathologies, and provides a comprehensive foundation for observing a child's auditory behaviors.



Pediatric Audiologic Test Protocols

- The American Speech-Language-Hearing Association (ASHA) and the American Academy of Audiology (AAA) recommend the following test protocols according to the chronological/developmental age of the child:



ASHA
American
Speech-Language-Hearing
Association

- **Birth through 6 months of age**
- **Six months through 24 months of age**
- **Twenty-five months through 60 months of age**



Birth through 6 months of age

- The testing of infants or children should rely primarily on physiologic measures of auditory function, such as auditory brainstem response (ABR) and auditory steady-state responses (ASSR) using *frequency-specific stimuli* to estimate the audiogram.
- The **tone burst ABR** is the current gold standard measure of choice for the estimation of hearing thresholds in infants from birth to 6 months of age.
- In addition, otoacoustic emissions (OAE) and acoustic **immittance measures** should be used to supplement ABR and ASSR results.
- **Case history**, parent/caregiver report, behavioral observation of the infant's responses to a variety of sounds, developmental screening, and functional auditory assessments should also be performed.

Six months through 24 months of age

- At these ages, **behavioral assessments** should be performed first, with visual reinforcement audiometry (**VRA**) being the behavioral test of choice.
- **OAEs** may be used in routine assessments, but ABRs should be conducted when behavioral audiometric tests are unreliable, ear-specific thresholds cannot be obtained, behavioral results are inconclusive, or auditory neuropathy is suspected.
- Developmental screening and **functional auditory assessments** also should be performed.

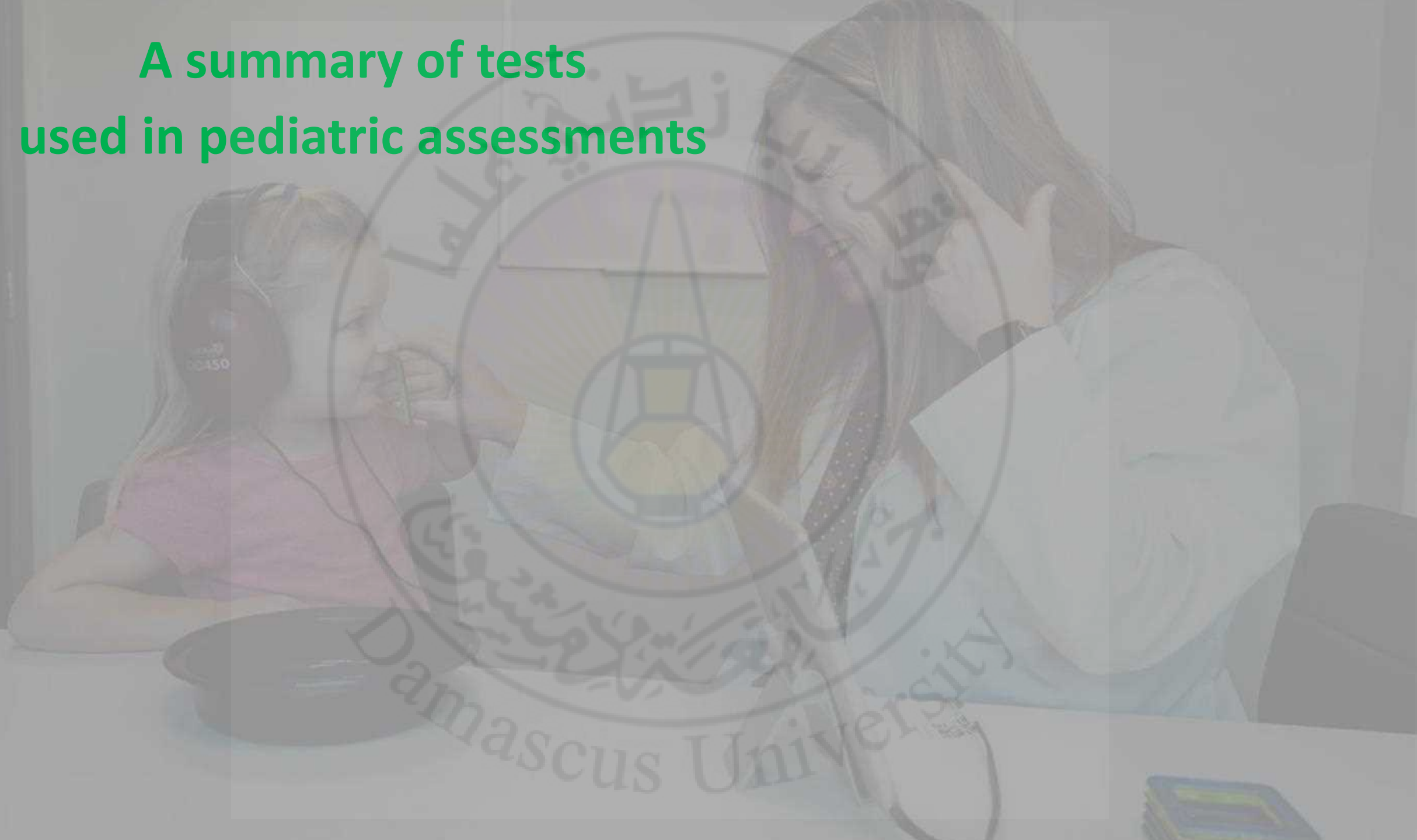


Twenty-five months through 60 months of age

- Behavioral tests (**VRA** or conditioned play audiometry [**CPA**]) and acoustic **immittance** tests are usually sufficient.
- OAEs, speech perception tests, and developmental screening and functional auditory assessments also can be used routinely to assess this age group.



A summary of tests used in pediatric assessments



Behavioral Audiologic Tests Need to Be Included in the Evaluation of All Infants and Children

- The information obtained from **behavioral testing** is useful.
- The behavioral audiogram provides valuable information not available from **electrophysiologic testing** and should be attempted on infants and children of any age or developmental status.
- **Functional auditory assessments** (e.g., questionnaires) completed by the child's caregivers also provide valuable information regarding the child's auditory responsiveness.



Test	Expected infant/ child response	Cognitive age range	Benefits	Challenges
Behavioral observation Audiometry (BOA)	Change in sucking in response to auditory stimulus; Other behavioral changes are not accepted because they usually indicate suprathreshold response.	Birth–6 months	• Enables the audiologist to obtain valuable behavioral responses in infants; part of the cross-check principle • Can be conducted in sound fields, with earphones or with bone oscillator, hearing aids, or cochlear implants • Enables accurate fitting of technology because minimal response levels (MRLs) can be obtained	• Requires careful observation of infant sucking by the audiologist • Cannot be used with infants who do not suck (e.g., infants who use feeding tubes) • Can be performed only when the infant is in a calm awake or light sleep state • BOA not generally accepted in the audiology community because audiologists typically have not been trained to use a sucking response paradigm

Test	Expected infant/ child response	Cognitive age range	Benefits	Challenges
Visual Reinforcement audiometry (VRA)	Conditioned head turn to a visual reinforcer, usually a lighted animated toy	5–36 months	• Enables the audiologist to obtain valuable behavioral responses in infants and young children; part of the crosscheck principle • More responses possible per test session because responses are conditioned • Can be conducted in soundfield, with earphones or with bone oscillator, hearing aids, or cochlear implants • Enables accurate fitting of technology because MRLs can be obtained • The state of the infant or child less problematic than in BOA because the child can be more easily involved in the task	• Obtaining individual ear data when child will not accept earphones

Test	Expected infant/ child response	Cognitive age range	Benefits	Challenges
Conditioned Play audiometry (CPA)	Motor act by child in response to hearing a sound (e.g., the listen and drop task)	30 months to 5 years	- Accurate responses possible at threshold level - Can be conducted in soundfield or with earphones, with bone oscillator, hearing aids, or cochlear implants	Keeping the child entertained and involved long enough to obtain all the necessary information
Immittance	None	All	Provides information about middle ear functioning and about intactness of the auditory system reflex arc	Necessity for the child to sit still, not speaking or moving, during the test Battery
Transient otoacoustic emissions (TOAE)	None	All	- Measures outer hair cell function - Presence of emissions indicates no greater than a mild hearing loss - Contributes to evaluation of the overall function of the auditory system	Cannot rule out mild hearing loss
Auditory brainstem response (ABR)	None	All	- Tonal ABR to provide frequency-specific threshold information - Click ABR to provide information about the intactness of the auditory pathways, including measures contributing to the diagnosis of auditory neuropathy	- Necessity for the infant or child to be asleep, sedated, or very still for the duration of testing. - Not a direct measure of hearing and not a substitute for behavioral audiologic testing

Necessary Steps for Selecting the Appropriate Test Protocol

- **Determine the child's cognitive age from:**
 - Case history
 - Reports from other evaluations
 - Infant developmental screening scales
- **Evaluate the child's physical status in terms of:**
 - Upper-torso control
 - Head and neck control
 - Vision
 - Ability to manipulate toys
- **Choose the test room setup:**
 - One room with one audiologist
 - Two rooms with two audiologists, or one audiologist and one test assistant
 - Two rooms with one audiologist and one parent who also functions as a test assistant



Determining the child's cognitive age

- It is critical to know the cognitive age of the child to select the appropriate test protocol and obtain reliable results. *Although many children have compatible cognitive and chronologic ages, some do not.*
 - **BOA**: Birth to 6 months cognitive age
 - **VRA**: 6 months to 36 months cognitive age
 - **CPA**: 30 to 36 months and older cognitive age
- Case history
 - Speech, language, and motor skills
 - Reports and scales from other clinicians, including speech-language pathologists and pediatricians,
 - Experience spending time with young children
 - Developmental milestones for children

Evaluating the child's physical status

- the child's physical condition needs to be evaluated to be certain that the child is capable of performing the test tasks:
- **BOA:** Does the child possess a *sucking response* (e.g., does the child breastfeed or suck on a bottle or pacifier)? If yes, the audiologist can implement the BOA procedure.
- **VRA:** Requires the child to have *vision* good enough to see the reinforcing toy and *neck control* sufficient to turn and look for the reinforcing toy.
- **CPA:** Requires that the child perform a motor task in response to the presentation of a sound. The ability to accomplish this task is limited only by the creativity of the audiologist. A child who cannot hold a toy and drop it in a bucket, for example, may be able to blink, move a finger, or push a button.



Test Room setup: Two-Room

- Audiologist and audiometer in one room and the child, parent, and audiology assistant in the other.
- The audiologist and the audiology assistant must have a full view of the child.
- The audiologist, who is presenting the test stimuli, needs to be able to observe the child's behavioral state to know when to present stimuli and when not to, and both testers need to be able to judge the presence or absence of a response.
- It is also important that the two testers be able to communicate.





Test Room setup: One-Room

- The advantage of a one-room test setup is that testing can be accomplished with only one audiologist, who now performs both the tester and test assistant roles, thus having more control of the test situation.
- To accomplish this type of testing, the audiologist places the audiometer in the test room where the child will be.





Case History

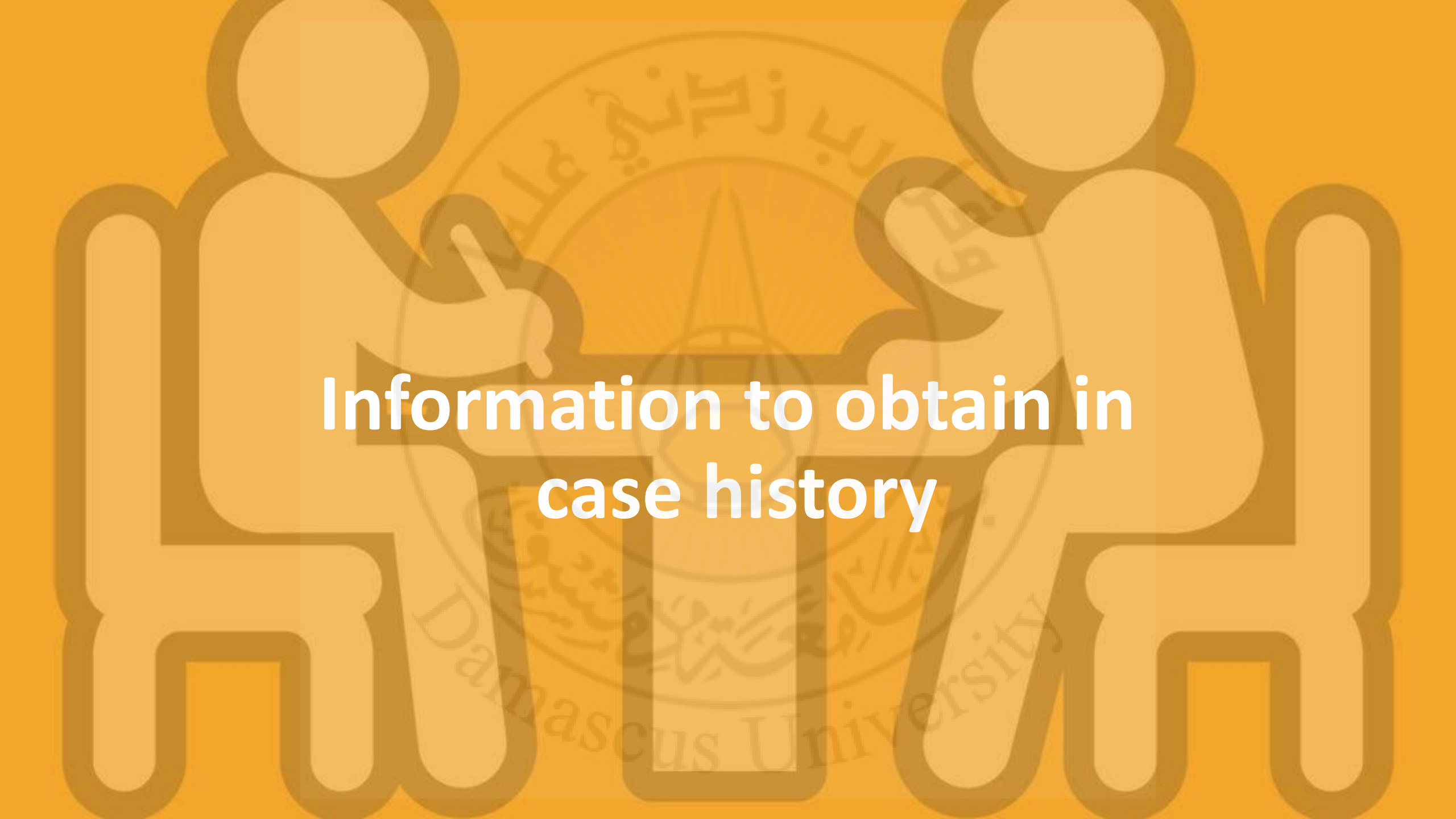
- Obtaining a case history enables the audiologist to learn about the child and to understand the parents' concerns and assessment expectations.
- History taking also facilitates the development of a rapport between the audiologist and the family that will be invaluable when counseling about test results.
- Advance information by questionnaires & forms
- Interview at the center (Formal vs. informal)



Case History

- A complete history covers **several content areas**, and depending on the reason for the evaluation, emphasizes different segments of information.
- It may be helpful to try to estimate the audiogram from history information, from observation of the child, and from listening to the child's utterances, before beginning the test.
- Doing so over a period of time will improve the audiologist's ability to take a history and make accurate observations of a child's auditory status.



The background is a solid orange color. In the center, there is a faint, large circular seal of Damascus University. The seal contains Arabic text at the top and bottom, and a central emblem. Overlaid on the seal are two stylized, light-orange human figures. The figure on the left is pointing its right index finger towards the center. The figure on the right is pointing its left index finger towards the center. The text "Information to obtain in case history" is written in white, bold, sans-serif font across the middle of the image, partially overlapping the figures and the seal.

Information to obtain in case history

Birth and prenatal history

- Previous pregnancies
- Illnesses during the pregnancy, including the week of pregnancy an illness occurred
- RH incompatibility, ABO blood incompatibility
- Medications, drugs (legal and illegal) taken during the pregnancy
- Complications during the pregnancy
- Length of the pregnancy
- Delivery: caesarean section or vaginal
- Birth weight
- Complications at birth: anoxia, jaundice, Apgar scores, breech, other
- Length of hospitalization

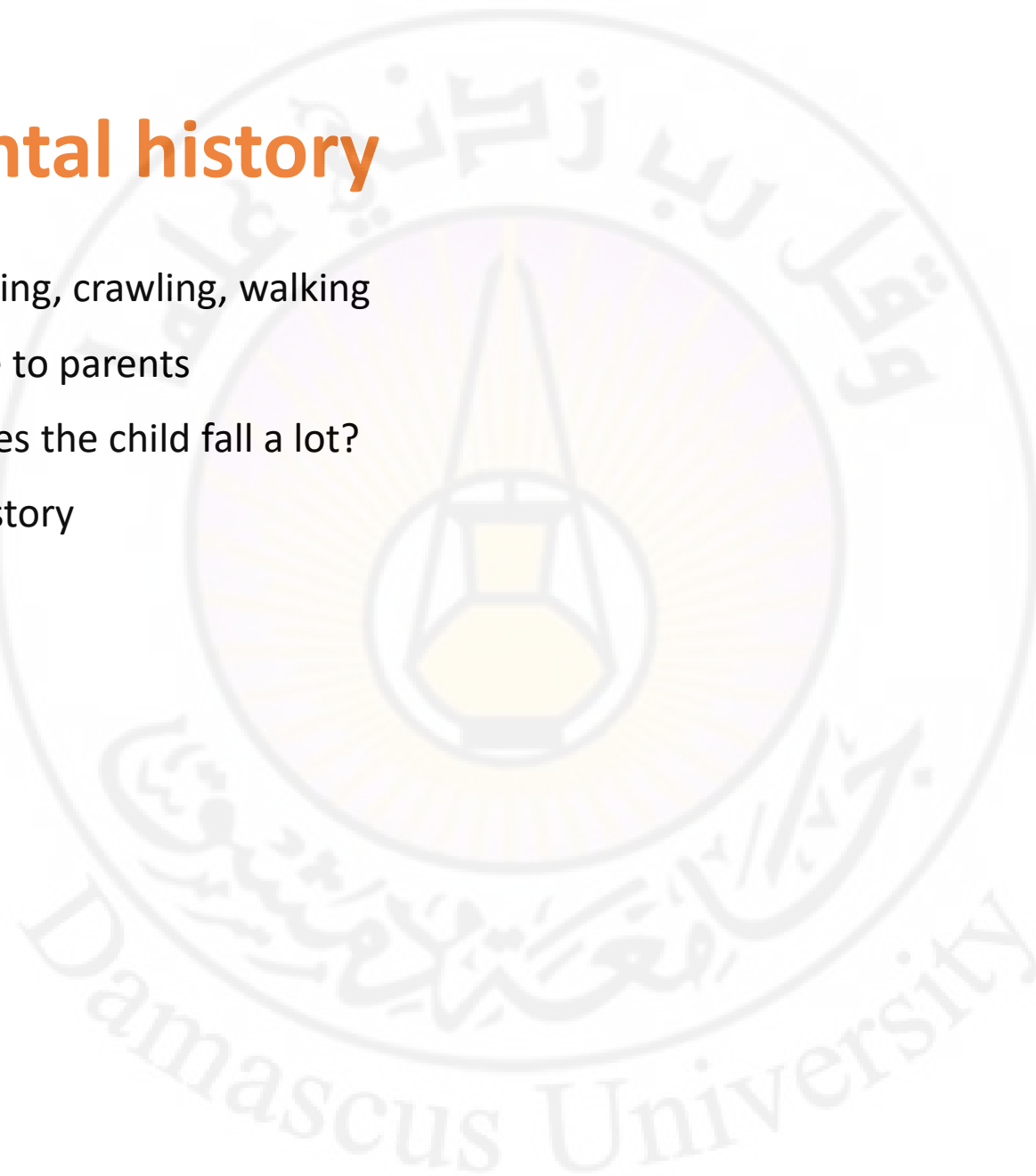
Health history

- Colds, allergy, ear infections
- High fevers
- Immunizations
- Meningitis
- Other viruses (mumps, cytomegalovirus)
- Immunization history; reaction to immunizations
- Drugs taken regularly and drug reactions
- Feeding or swallowing problems
- Seizures
- Head injury



Developmental history

- Motor milestones: sitting, crawling, walking
- Age of visual response to parents
- Is walking clumsy? Does the child fall a lot?
- Feeding and eating history
- Age of toilet training



Communication history: Hearing

- Parents' thoughts of child's hearing
- Sounds to which child responds
- Does the child distinguish between sounds (phone, doorbell)?
- Does the child want TV/CD/DVD/computer loud?
- Does hearing fluctuate? Under what conditions?
- Are sounds comfortable? What sound? Under what conditions?
- Amplification history:
 - Does the child wear a hearing aid and/or a cochlear implant?
 - Name and model number of the instrument(s)? Which ears?
 - Does the child wear an FM system? Name and model number of the instrument? Which ears?
 - Who recommended the devices?
 - When were they acquired?
 - When does the child wear them? (e.g., all day? only at school?)

Communication history: Speech and language

- Age of babbling, first word, phrases, sentences
- Does the child understand verbal requests with, without visual cues?
- How does the child communicate his/her needs? Voice? Gesture? Sign?
- Has there been a change in the child's speech and language?
- Did the child speak and then stop?

Educational history

- Current school
- Type of educational program
- Previous school placements
- Reasons for change in school placement
- Special services received in school
- Describe educational problems or concerns

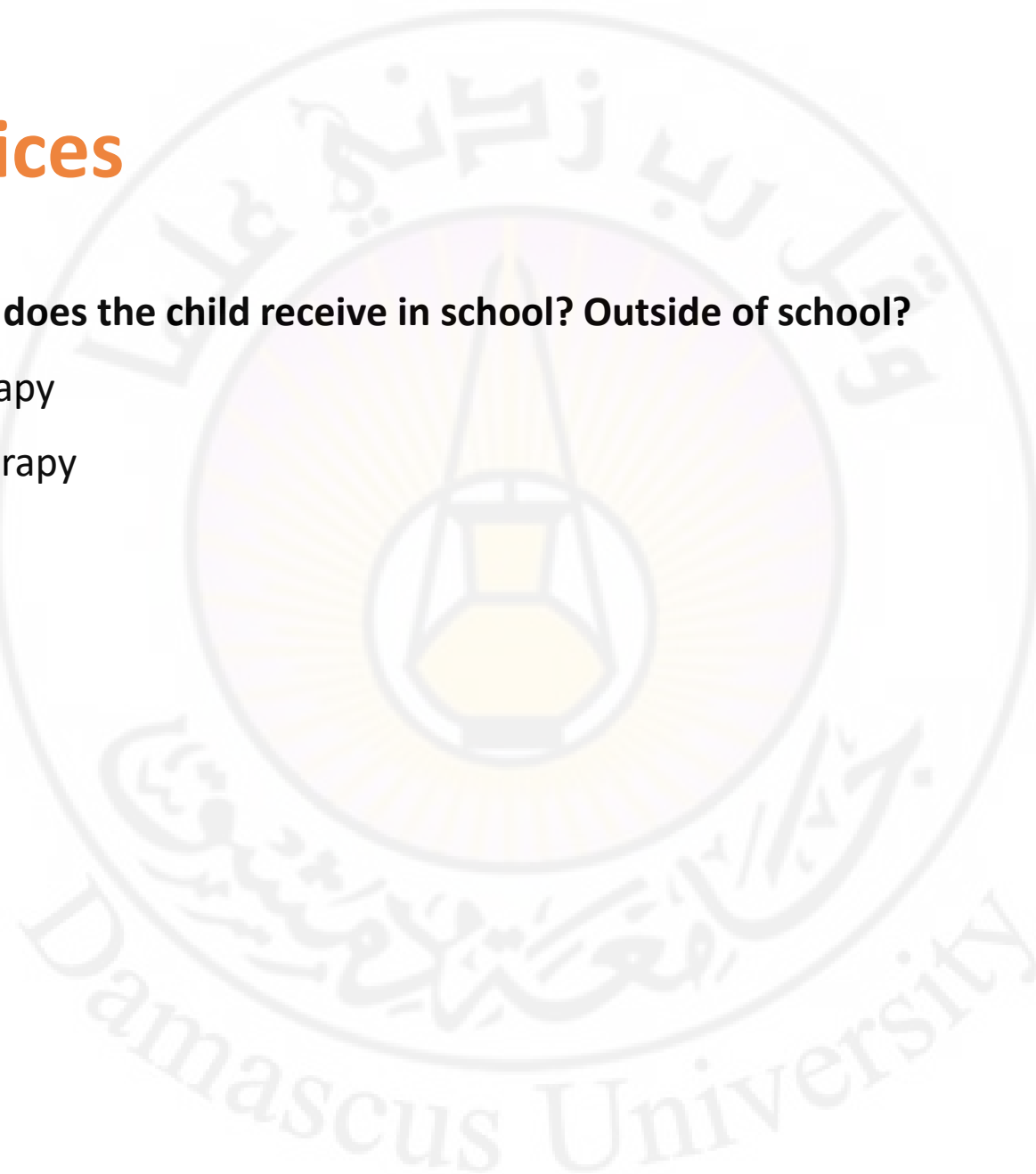


Social history

- When did the child feed himself? Dress himself?
- Does she play with other children?
- What toys or objects does the child like to play with?
- Does the child have any behavior problems?
- How does the child get along with other children? Adults? Family?
- Have there been any changes in the child's behavior?
- Does the child respond to others? Make eye contact?

Special services

- **What special services does the child receive in school? Outside of school?**
- Speech-language therapy
- Hearing (auditory) therapy
- Occupational therapy
- Physical therapy
- Psychological services
- Educational tutoring
- Other



Other evaluations

- **What other evaluations has the child had (evaluator, dates, and results)?**
- Audiologic
- Speech-language
- Hearing (auditory) therapy
- Occupational therapy
- Physical therapy
- Psychological
- Educational
- Pediatric
- Otolaryngologic
- Neurologic
- Psychiatric

Screening Questionnaire for Parents

I. Chief complaint _____	
When was the problem first noted? _____	
Extent of problem _____	
Previous examinations and evaluations _____	
II. Prenatal history _____	
Exposure to viral diseases during pregnancy? _____	
Which viral disorder? _____	
During which pregnancy month? _____	
Drugs during pregnancy? _____	
Trauma during pregnancy? _____	
III. Birth history _____	
Gestation age at birth _____	
Birth weight _____	Bilirubin level high? _____
Asphyxia? _____	Meningitis? _____
IV. Family history _____	
Childhood deafness in family? _____	
Relationship to patient _____	
Birth defect or abnormalities _____	
In any other relatives? _____	
V. Developmental history _____	
Age of first smile response? _____	
Age when sat up alone? _____	
Age when first crawled? _____	
Age of "stranger anxiety"? _____	
Age of walking? _____	
VI. Physical history _____	
Cleft lip or palate _____	Submucous cleft _____
Low-set ears _____	Poorly formed ears _____
High fevers with illness _____	Seizures _____
Ear infections _____	How many? _____
Previous treatment for ear conditions? _____	
VII. What do you (parents) really think caused this hearing problem? _____	
Name of child's pediatrician _____	
Names of other physicians who have seen this child _____	

Functional Auditory Assessments

- An important part of a basic test battery for an infant or child of any age is an evaluation of auditory function.
- Numerous tests and surveys have been developed for this purpose.
- Functional auditory assessments are typically accomplished by having the teacher, student, or parent complete a questionnaire before and after the use of a hearing aid, cochlear implant, personal remote microphone (RM), or soundfield system or the delivery of therapy or educational services.
- Most important, functional auditory assessments can monitor the child's auditory progress over time by repeating assessments periodically

Measurement tool	Authors	Age range	Purpose
Auditory Behavior in Everyday Life (ABEL)	Purdy et al 2002 ⁵	Children 2–12 years	Twenty-four-item questionnaire with three subscales (aural-oral, auditory awareness, social/conversation skills) that evaluates auditory behavior in everyday life
Children's Home Inventory for Listening Difficulties (CHILD)	Anderson and Smaldino 2011 ⁶	Children 3–12 years	Parent and self-report versions that assess listening skills in 15 natural situations
Children's Outcome Worksheet (COW)	Williams 2003 ⁷	Children 4–12 years	Teacher, parent, and child rating scales of classroom and home listening situations with amplification device; to specify five situations where improved hearing is desired
Early Listening Function (ELF)	Anderson 1989, 2000 ⁸	Infants and toddlers; 5 months–3 years	Parent observational rating scale of structured listening activities conducted over time to record distance learning
Functional Auditory Performance Indicators (FÁPI)	Stredler-Brown and Johnson 2004 ⁹	Infants through school age	Parent or interventionist assessment of functional auditory skills over time
Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS)	Robbins et al 1991 ¹⁰	Infant-toddler and older child versions	Structured parent interview scale designed to assess spontaneous auditory behaviors in everyday listening situations
Listening Inventory for Education—Revised (LIFE-R)	Anderson et al 1998, 2000 ¹¹	6 years and above	Student and teacher rating scales designed to assess listening difficulty in the classroom
LittleEARS	Kuhn-Inacker et al 2003 ¹²	Birth and up	Questionnaire for the parent with 35 age-dependent questions
Meaningful Auditory Integration Scale (MAIS)	Robbins et al 1991 ¹⁰	Children 3 to 4 years and up	Parental interview with 10 questions that evaluates meaningful use of sound in everyday situations; attachment with hearing instrument, ability to alert to sound, ability to attach meaning to sound
Parents' Evaluation of Aural/Oral Performance of Children (PEACH)	Ching and Hill 2005 ¹³	Preschool to 7 years	Interview with parent with 15 questions targeting the child's everyday environment. Includes scoring for five subscales (use, quiet, noise, telephone, environment)
Preschool Screening Instrument for Targeting Educational Risk (Preschool SIFTER)	Anderson et al 2017 ¹⁴	3 to 6 years	Questionnaire with 15 items completed by the teacher that identifies children at risk for educational failure; has five subscales (academics, attention, communication, class participation, behavior)
Screening Inventory for Targeting Educational Risk (Elementary SIFTER, Secondary SIFTER)	Anderson et al 2017 ¹⁴	6 years through secondary school	Teacher questionnaire designed to target academic risk behaviors in children with hearing problems; has five subscales (academics, attention, communication, class participation, behavior)

8. Behavioral hearing tests in infants and children

Behavioral hearing tests

- Behavioral evaluation is the **gold standard** for evaluation of hearing in infants and children.
- Behavioral audiologic testing is the protocol that provides a direct measure of hearing.
- Behavioral testing is used to determine degree and type of hearing loss and can also be used to monitor performance with technology.
- **BOA**: Birth to 6 months cognitive age
- **VRA**: 6 months to 36 months cognitive age
- **CPA**: 30 months and older cognitive age



Behavioral or Nonbehavioral tests?

- **Nonbehavioral tests**, such as ABR, ASSR, and OAEs, are frequently used to estimate hearing sensitivity in infants. These tests are an important part of the audiology practice and are the gold standard for testing infants. They are, however, not tests of hearing. ABR, ASSR, and OAE measures provide information about **the integrity of specific sites** within the auditory system.
- **Behavioral tests** measure the response of the entire auditory system, from the outer ear through the cerebral cortex. Behavioral tests permit measurement of what an infant or child actually perceives, measure functional hearing abilities not available in any other way, and are a critical component of a complete audiologic evaluation.



Behavioral or Nonbehavioral tests?

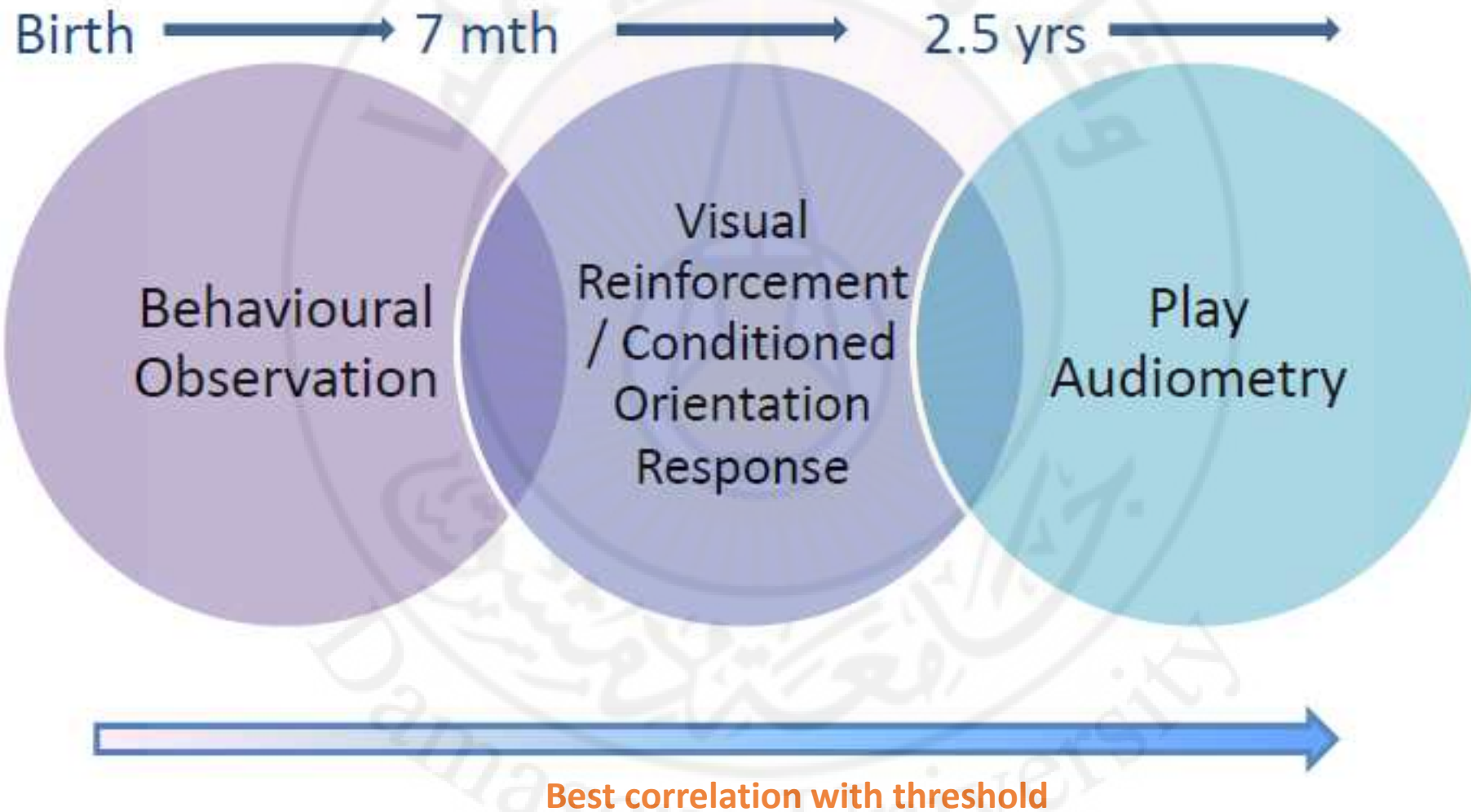
- **Behavioral testing** allows the **parents** to participate in testing by permitting them to observe when the infant is responding to a sound and when he is not. If parents are provided with information about what to observe, they can be active participants in testing, a process that facilitates acceptance and understanding of hearing loss and motivates families to proceed with treatment.
- Critically important, behavioral testing provides information about what an infant can hear **when wearing technology**.
- The necessity for **cross-checking** physiologic results with behavioral data by using a battery of tests to determine hearing sensitivity.



Selecting the Appropriate Test Protocol

- **Determine the child's cognitive age from:**
 - Case history
 - Reports from other evaluations
 - Infant developmental screening scales
- **Evaluate the child's physical status in terms of:**
 - Upper-torso control
 - Head and neck control
 - Vision
 - Ability to manipulate toys
- **Choose the test room setup:**
 - One room with one audiologist
 - Two rooms with two audiologists, or one audiologist and one test assistant
 - Two rooms with one audiologist and one parent who also functions as a test assistant





Diagnostic Audiologic Evaluation

- The primary goal of an audiologic evaluation of an infant or young child is usually to determine whether the child has sufficient hearing (auditory brain access) to develop speech and language.
- A complete diagnostic evaluation includes frequency and ear-specific threshold information throughout the frequency range, using both air and bone conduction procedures.
- It is critical that audiologists have skills to test infants and children using both electrophysiologic and behavioral techniques from birth through childhood.



The Basics of BOA:

Birth to 6 Months

What is Behavioral Observation testing (BOA)?

- BOA involves watching the baby's responses to relatively sudden and intense stimulus sounds presented in the sound field, such as a speech signal (e.g., "bah-bah-bah"), warble tones, narrow band noises, and various hand-held noise-makers.
- **Behavioral observation** is preferred over the older term behavior observation audiometry, because it helps provide insight about the development of global auditory skills but does not actually involve hearing assessment.



Observing subtle unconditioned changes in behavior in response to sound!

Who Is a Candidate for BOA?

- Infants from birth to 6 months cognitive age who are capable of sucking.
- Older infants who, because of **developmental delay**, are cognitively less than 6 months of age
- In children with auditory neuropathy spectrum disorder (**ANSD**), BOA is helpful in trying to determine auditory response levels, since ABR cannot be used as a predictor of hearing sensitivity with this population.



What Is Being Observed?

- Cessation or initiation of **sucking** is a reliable response for observing auditory behavior in infants younger than 6 months. Some infants will start sucking when a sound is presented, others will cease sucking, and some will do both.
- Many behaviors have been used to assess hearing in infants (arousal, limb movement, respiration changes, facial grimace, eye blink), but these behaviors have not proven to be sufficiently repeatable, and, more importantly, they have not been good indicators of threshold.



Maximizing Observation of the Sucking Response

- Sucking can be observed with a bottle, at the breast, or on a pacifier. The family should be bring the infant to the evaluation session hungry so that she will be ready to suck.
- The infant needs to be as comfortable as possible during testing.
- If an infant is very hungry, it is best to allow her a little time to eat to enable her to get over that initial extreme hunger before beginning testing.
- After the infant is finished eating, testing can frequently continue by observing sucking with a pacifier.



Positioning the Infant

- The infant needs to be resting in a comfortable position with full support of the head and torso, and with the mouth visible to the testers.
- The infant may be held or placed in an infant seat. The advantage of an infant seat is that the infant will not be receiving any “signals” from the mother when she hears the sound. If the infant is being held, the mother or other person holding the infant should be very carefully instructed about the need to remain silent and still throughout testing to avoid interfering with testing.





Positioning the infant for testing using a bottle



Positioning the infant for testing using a pacifier



Positioning the infant for testing at the breast.



Positioning the infant for behavioral observation audiometry.

The Role of the Parents

- The parents cannot be relied upon as the primary observers.
- Parents are, however, very valuable in helping the testers to understand the baby and in assisting in making the baby comfortable.
- At least one parent should be in the test room whenever possible to assist in understanding the test protocols and test results.
- Parent observation of how the baby responds will be helpful in helping parents understand test results



Box 7.1 Behavioral Observation Test Protocol

1. Bring infant into test room in hungry state.
2. Seat infant so torso is supported and infant is not fidgety, and so tester(s) can easily see the infant's mouth.
3. Monitor infant state during testing and stop if infant becomes fidgety.
4. Instruct parents not to respond to test stimuli or responses from the child.
5. Test assistant will keep infant centered, observe responses, and monitor parents' behavior.
6. Begin testing in soundfield.
7. Begin testing with a stimulus that is slightly above estimated threshold.
8. Test one low (500 Hz) and one high (2,000 Hz) frequency initially and select additional frequencies to test depending on initial responses.
9. Reduce thresholds in 10-dB steps and increase in 5- to 10-dB steps to bracket threshold. Record a response after three reversals.
10. Take breaks as needed to calm the infant and increase usable test time.
11. If soundfield testing indicates a hearing loss, test bone conduction.
12. If infant is still responding, or at the next test session, test with insert earphones.
13. Test with technology as needed.





FILMIGO

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 at the top and in English at the bottom. In the center is a stylized sun or starburst design. The text "Behavioural Tests" is superimposed on this background.

Behavioural Tests

Infant Thresholds

- Because behavioral test protocols for infants frequently did not reveal threshold responses at soft levels, some audiologists have proposed that responses at 60 to 70 dB sound pressure level (SPL) be interpreted as normal hearing for very young infants.
 - **Minimum Response Level (MRL) is not a threshold!**

Age	MRL (noisemakers)	MRL Warble tones
0-6 weeks	50-70 dB SPL	75 dB HL
6 weeks – 4 mths	50-60 dB SPL	70 dB HL
4-7 mths	40-50 dB SPL	50 dB HL
7-9 mths	30-40 dB SPL	45 dB HL

All infant responses must be time-locked with stimulus presentation and repeatable prior to concluding they are reliable.

Responses to sounds from developmentally normal infants at various ages, and typical stimulus levels expected to elicit these responses in behavioral observation audiometry^a

Age range	Types of behavioral responses	Warble Tones (dB HL)	Speech ^b (dB HL)	Noisemakers (dB SPL, approximate)
0 to 6 weeks	Eye blink Eye widening Startle Arousal/stirring from sleep	78	40–60	50–70
6 weeks to 4 months	Eye blink Eye widening Eye shift Quieting Rudimentary head turn starts by 4 months	70	47	50–60
4 to 7 months	Head turn laterally toward sound Listening attitude	51	21	40–50
7 to 9 months	Directly localizes to side Indirectly localizes below ear level	45	15	30–40
9 to 13 months	Directly localizes to side and below ear level Indirectly localizes above ear level	38	8	25–35
13 to 16 months	Directly localizes to side/below/above ear level	32	5	25–30
16 to 21 months	Directly localizes to side/below/above ear level	25	5	25
21 to 24 months	Directly localizes to side/below/above ear level	26	3	25

^aAdapted from Northern JL, Downs MP. 1991. Hearing in Children, 4th edition. Baltimore, MD: Williams and Wilkins, with permission.

^bA startle response to speech is typically expected at 65 dB HL for all of the age groups shown.

The Basics of VRA:

6 to 36 Months

Visual Reinforcement Audiometry (VRA)

- VRA test involve training the infant to make a **conditioned head turn** in response to a test stimulus.
- Most infants will turn toward the sound source a few times, but the head-turning behavior will habituate to repeated stimuli if not reinforced.
- Fortunately, this head-turning behavior can be shaped using an operant discrimination procedure that permits obtaining numerous responses to auditory stimuli.
- The sound stimulus is used to cue the child to seek the **visual reinforcement**.
- Use of a positive reinforcement, such as a lighted toy or short video clip, will increase the number of responses before the responses are extinguished.



VRA method

- The audiologist presents a stimulus. If the child detects the stimulus, she will turn toward it. The audiologist then activates a reinforcer. After a few repetitions, the child learns to seek the reinforcer when she hears the sound.
- The lowest intensity level at which a reliable response is obtained is considered the **Minimum Response Level (MRL)** and can be frequency- and ear- specific.



Objective of VRA

- The objective of VRA assessment is to establish MRLs which are required to be obtained at a minimum of three frequencies (500, 2000 and 4000 Hz) per ear by air conduction (AC), and of at least one frequency by bone conduction (BC) if AC MRL >25 dB HL.
- Further frequencies shall be included over time at the Audiologist's discretion.
- Hearing sensitivity is considered to be within normal limits if the MRL is 25 dB HL or less.



Target Population for VRA

- Infants aged 6 months developmental age up to approximately 24-36 months of age who possess sufficient vision to see reinforcers and sufficient trunk and neck control to turn towards reinforcers are considered candidates for VRA.
- At 8 to 12 months of age, 95% of infants could perform the test reliably.
- VRA may be successful with children with developmental or cognitive delay when the children reach a mental age of 10 months.



Visual Reinforcers



VRA toys



Video VRA











Positioning in VRA Testing

- A critical factor for obtaining reliable VRA thresholds is the ability to keep the infant's or child's attention focused at **midline** in a position that easily permits a head turn.
- Proper positioning is critical. The child needs to be seated comfortably so that the upper body is steady and allows the infant or child to turn easily to look at the toy.
- The child should not be leaning over trying to get something from the floor, trying to maintain balance, or looking for something or someone seated behind.



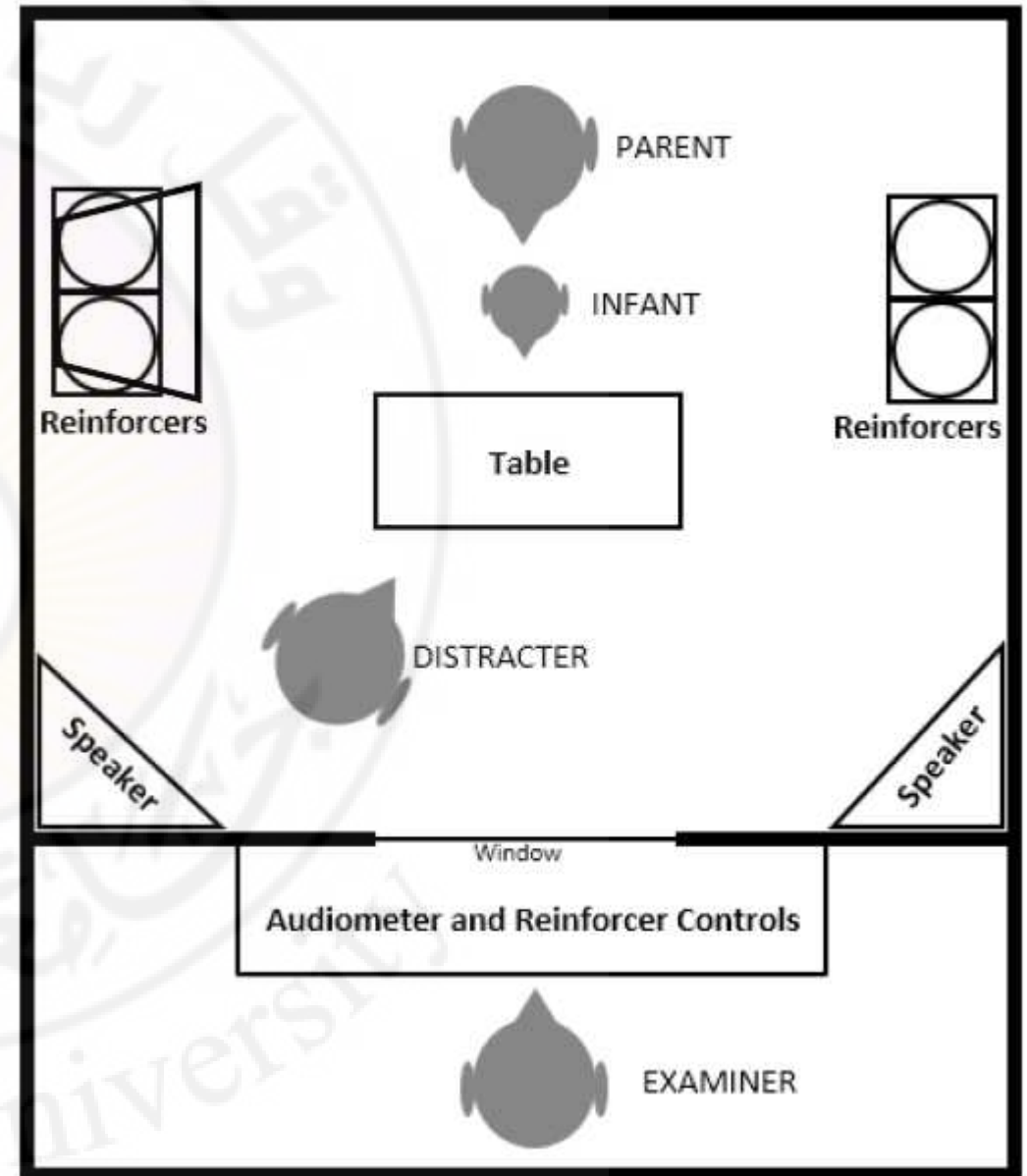
Stimulus Transducers

- Insert earphones (ER-3A)
 - If the child being tested has been fitted with hearing aids, his/her own earmolds, attached to the insert tubing, may be used instead of the foam tip.
- Supra-aural earphones (TDH 49/50)
- Circumaural headphones (HDA-200)
- Bone conduction oscillator (B-71 & B-81)
- Loudspeakers



VRA setup

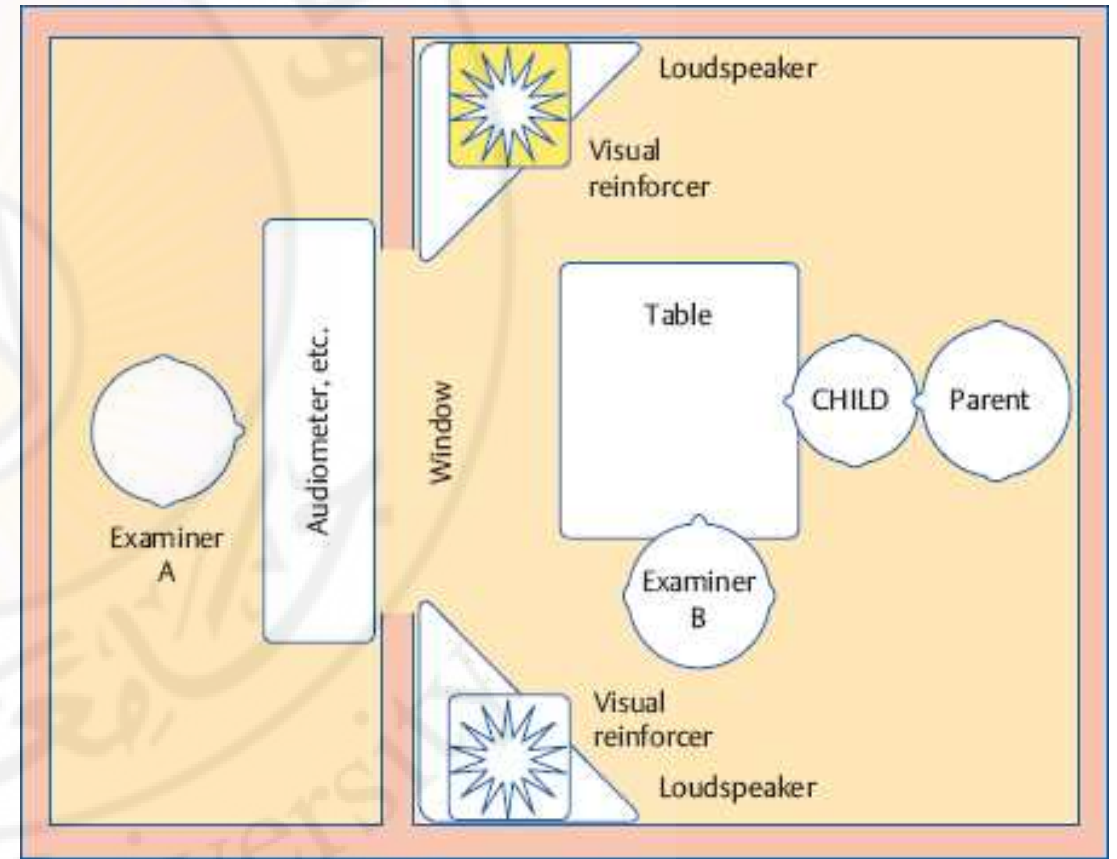
- The child should be positioned so that the VRA reinforcer is located $\pm 90^\circ$ to the position of the child so that the child must make a full head turn in order to see the reinforcer.
- The location of the reinforcers at $\pm 90^\circ$ is important because it requires the child to make a full head turn to the left or right, which reduces the uncertainty of whether a response has occurred that might exist if the reinforcers are located in the frontal plane.





VRA setup-2

- The loudspeakers and reinforcers can be placed at 45° angles, but 90° angles have been recommended so that the child's head turn will be clearly observable.



Use of an Assistant



The use of an experienced assistant is helpful during pediatric hearing evaluations

Distractors

- A large supply of different toys is needed to distract the child and/or for play tasks, to keep the infant focused straight ahead so that a clear head turn can be observed.
- These should be toys that can be manipulated quietly, can be effectively cleaned and/or disinfected as necessary, and are not similar to the reinforcement toys.
- Toys for VRA distraction should be significantly less interesting and appealing to the child than the reinforcement toys.



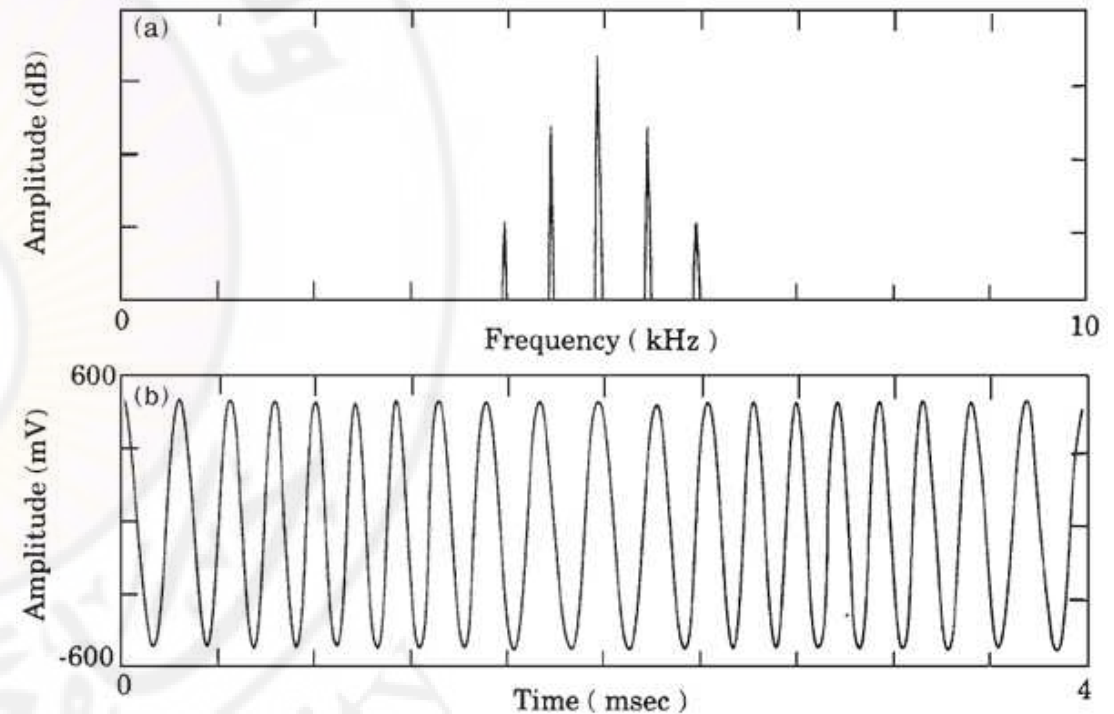
Single Tester VRA

- In situations where a testing assistant is not available, the Audiologist can act as both the examiner and distractor.
- For this to be achieved, it is usually recommended that the tester, the child, the audiometer, and the reinforcer controls should be in the same test room, or in a setup that permits adequate equipment control and observation and reinforcement of responses.



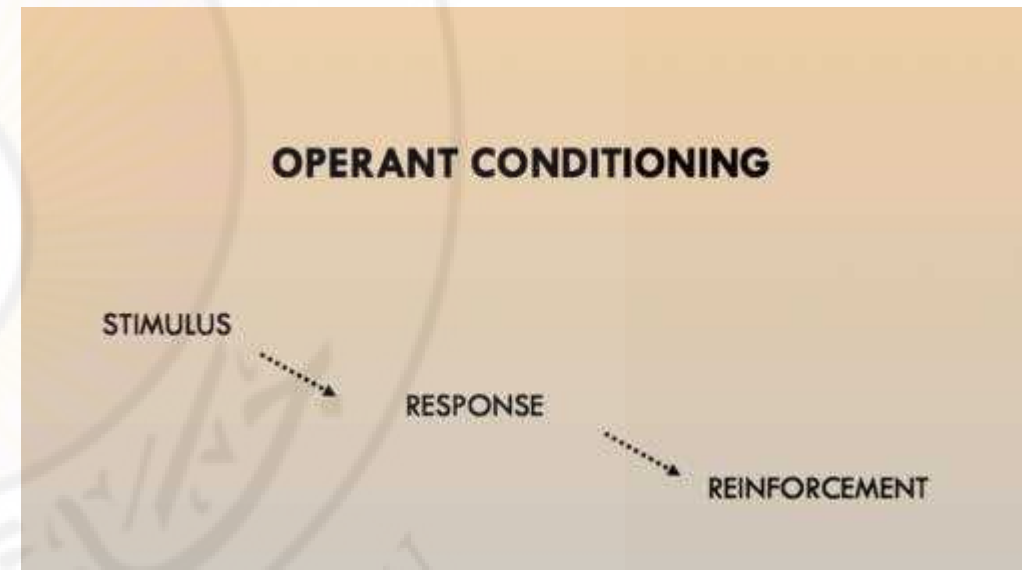
Stimuli

- Pure tones should not be used in sound field due to standing waves; warble tones (frequency modulated tone) are appropriate for use in sound field.
- Clinicians may use pure tones (if using insert phones).
- The narrowband noises were originally developed as masking signals, and are not recommended for thresholds estimation because they may underestimate audiometric slope.
- F.R.E.S.H. noise (Otometrics products)
- Pediatric noise (GSI & Interacoustics products)



Box 7.2 Protocol for Visual Reinforcement Audiometry

1. Seat child in high chair, in a child's chair, or on a parent's lap.
2. The test assistant or parent keeps child's attention focused to the front using quiet toys.
3. The auditory stimulus is presented at a comfortably loud level above expected threshold. The conditioning/reinforcing toy is turned on, and if the child does not turn, the test assistant calls attention to the toy. The auditory stimulus and the conditioning toy are kept on together for 3 to 4 seconds.
4. Step 3 is repeated until the child consistently turns to the auditory stimulus.
5. When the child is conditioned to respond, the auditory stimulus is presented without turning on the conditioning/reinforcing toy. If the child turns toward the sound, the reinforcing toy is turned on and conditioning is complete.
6. Testing proceeds, obtaining thresholds for one low-frequency (500 Hz) and one high-frequency (2,000 Hz) stimulus. The stimulus intensity is decreased until the child stops responding, and is then increased to bracket the threshold. Three responses at the same intensity are sufficient.
7. Additional frequencies to be tested will be determined by the responses to the initial frequencies tested.
8. Testing proceeds using insert earphones, bone vibrator, and technology (e.g., hearing aids and FM systems).
9. The reinforcing toy is turned on only when the child makes a conditioned head turn in response to a sound. When in doubt, do not turn on the reinforcer.



Condition at 70 dB HL

Paired trial 2x

Probe trial:

Present stim
only & look for
a head turn.

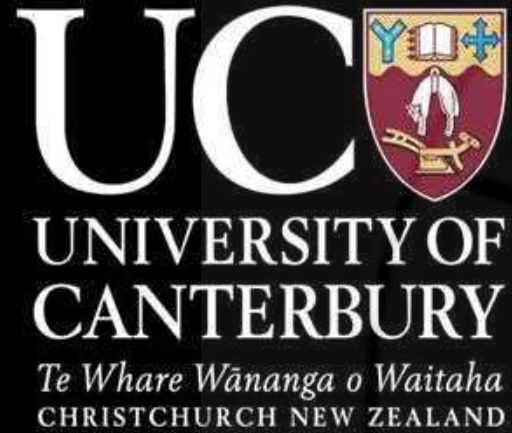
Max 2 trials at 90 dB
HL. If no response,
troubleshoot procedure.

No turn?
Listening
check, then go
to 90 dB HL.

Turn?
Reinforce. Do
it again.

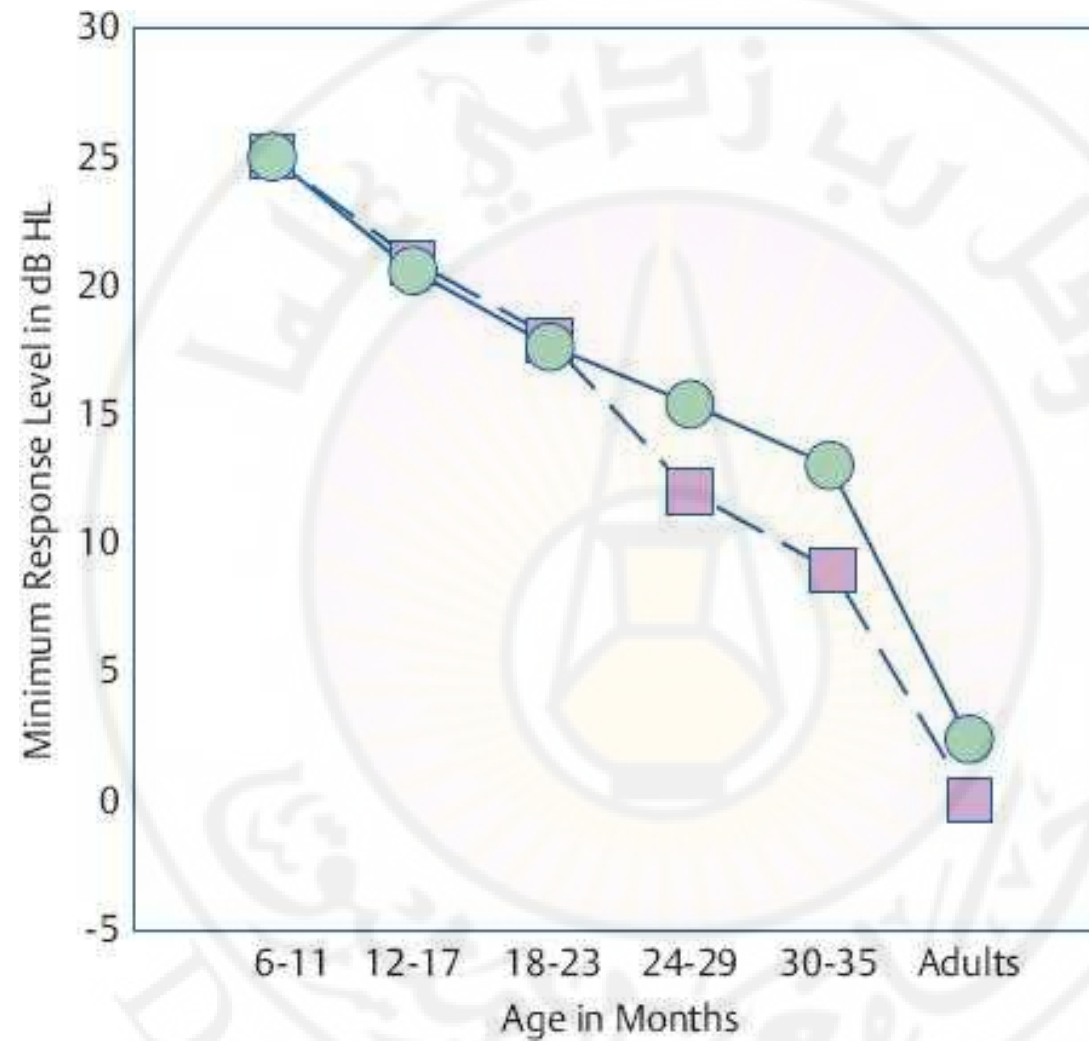
Turn?
Reinforce. Go
to Test
Protocol.

No Turn? Go to
90..



Department of Communication Disorders





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Conditioned Orientation Reflex (COR) Audiometry

- COR uses the same conditioning techniques as VRA. Sound may be presented from either the right or the left loudspeaker, but the child will be reinforced only when turning to the correct side.
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Tangible reinforcement operant conditioning audiometry (TROCA)

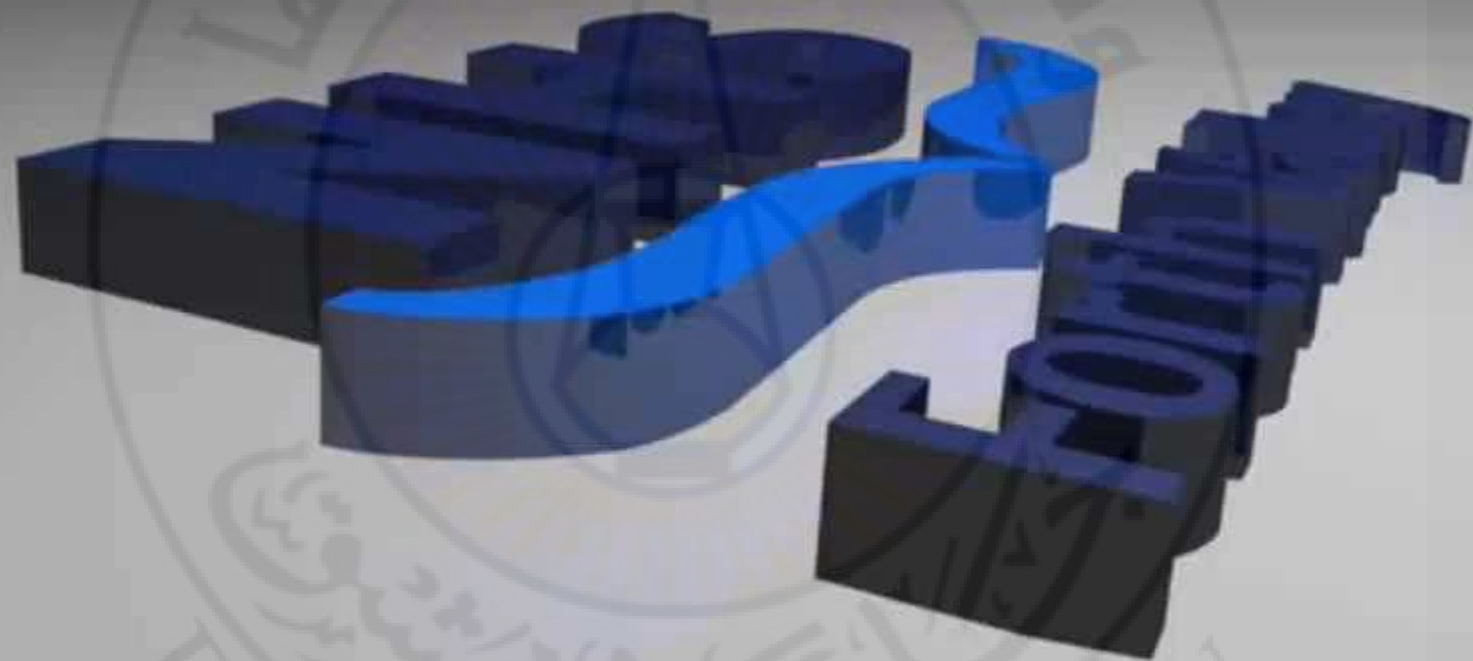
- TROCA is a highly structured testing approach originally described for use with difficult-to-test patients, such as those with developmental disabilities.
- Upon hearing a tone (or other test signal), the child is required to push a response button or to make another simple but specific motor response within his range of neuromotor capabilities.
- Correct responses are reinforced by the delivery of a **tangible reward**, which might be cereal, candy, tokens, or other small trinkets.



visual reinforcement operant conditioning audiometry (VROCA)

- In VROCA, the child is required to **press a response button** (which is a large, bright box) instead of turning toward the loudspeaker, after which a visual reinforcer is presented.
- The visual reinforcer itself is the same kind used in visual reinforcement audiometry.
- When VROCA is employed with sound field testing, the **loudspeaker is kept in front of the child** so he is not distracted from the response box, as opposed to being placed off to one side as is done for a head turning response.





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The Basics of Play Audiometry (CPA): 30 Months and Older

Play audiometry

- Once children reach a cognitive age of ~ 24 to 30 months, they can begin to cooperate voluntarily in hearing testing.
- By this age, children can be taught to drop a toy in a bucket, build a tower, or put a ring on a ring stand when they hear a sound.
- The challenging task for the pediatric audiologist is to find ways to keep the young child entertained for a long enough time to complete the hearing test.
- Conditioned play should be fun. The audiologist needs to be cheerful, toys need to be interesting, and they need to be varied frequently.



Target Population for CPA

- Young children typically aged from 24 to 60 months and/or older children with developmental delay may be candidates for audiometric assessment using CPA.
- A combination of VRA and CPA techniques may be used for younger children transitioning between these two testing techniques.



Testing Assistant

- Use of a test assistant during CPA is not required. If an assistant is used, s/he may act as the play partner during CPA testing while the Audiologist presents the stimuli and records the child's responses.
- Audiologist can act as both the examiner and play partner. For this to be achieved, the audiometer and play activity must be in the same test room as the child, or in a two-room setup that permits the tester to see and interact with the child, and judge if the child has responded or not.



Box 7.3 Test Protocol for Conditioned Play Audiometry

1. Set the child in a high chair or at a children's table so the child is comfortably seated.
2. Select a toy that will be enjoyable for the child and within the child's skill range.
3. Begin using a test stimulus that you expect the child to be able to hear.
4. Begin by demonstrating the task. The audiology assistant holds the toy to her ear. When she hears the sound, she says "I hear that" and drops the toy into the bucket.
5. After a few presentations, the child is given the toy and the audiology assistant holds the toy to the child's ear. When the sound is heard, the audiology assistant helps the child drop the toy into the bucket.
6. Care must be taken to encourage the child to drop the toy in the bucket only when you are certain the child heard the sound.
7. This is repeated until the child is able to perform the task without assistance.
8. Once the child is conditioned and performing reliably, testing can begin.
9. If the child appears to be bored, change toys to increase interest.
10. Testing can be accomplished by air and bone conduction and with hearing aids, cochlear implants, osseointegrated implants, and FM systems.



Test Protocols for Behavioral Evaluation

- Test protocols need to be designed to obtain the most information with the fewest responses.
- The goal of the initial audiologic evaluation of an infant is usually to be certain that the infant has sufficient hearing (auditory brain access) to develop speech and language.
- Obtaining one or two **soundfield** thresholds can ensure the child understands the task and gives the audiologist some information about hearing levels.
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- When earphone testing is being attempted, insert earphones are the earphones of choice for infants.
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- The bone vibrator should be held in place with either a pediatric-sized headband or a fabric one that goes around the head and across the forehead, using Velcro to secure it in place.
- If a metal headband is used, soft material such as foam or other padding should be used for comfort and to keep the headband from moving.



Stimuli

- The goal of the testing is to obtain frequency-specific test results.
- **Warble tones or narrow bands of noise** will provide this information.
- In addition to tonal thresholds, **speech perception** testing provides critical information.
- **Speech awareness thresholds (SAT)** are critical for infants who cannot perform more advanced speech perception tasks. Thresholds to low-frequency (“ba”), mid-high-frequency (“sh”), and high-frequency (“s”) speech stimuli can be used to confirm warble tone/noise band thresholds.

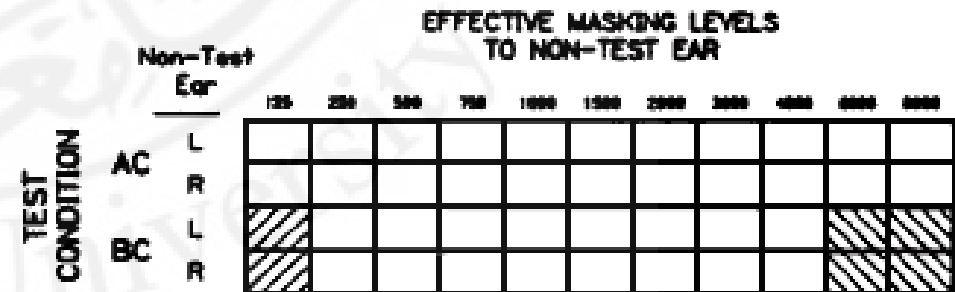
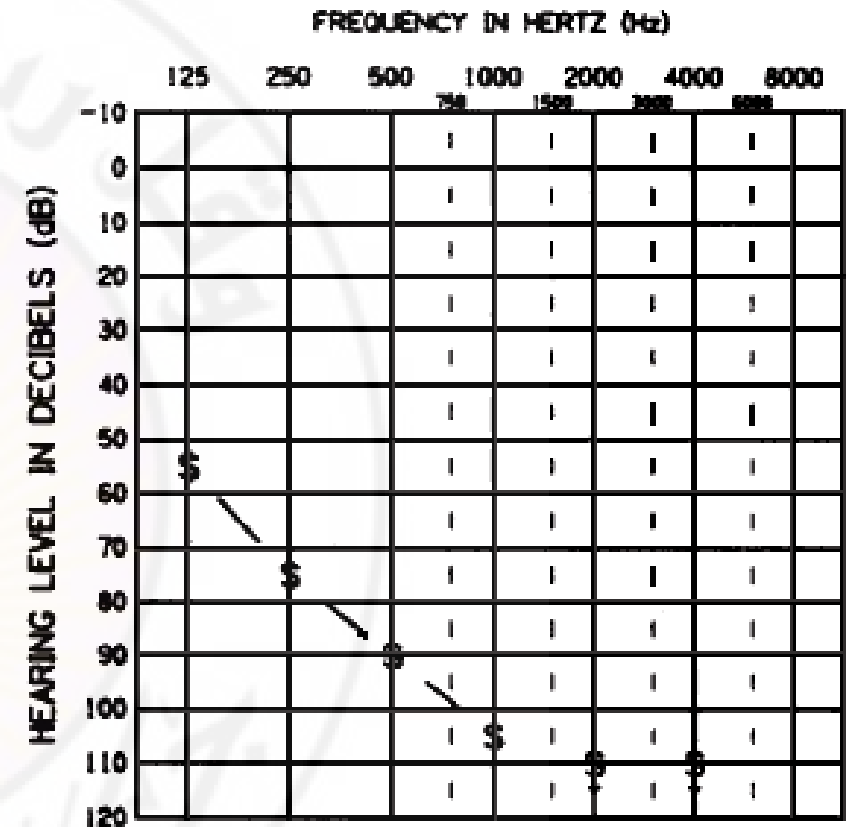


Test Order

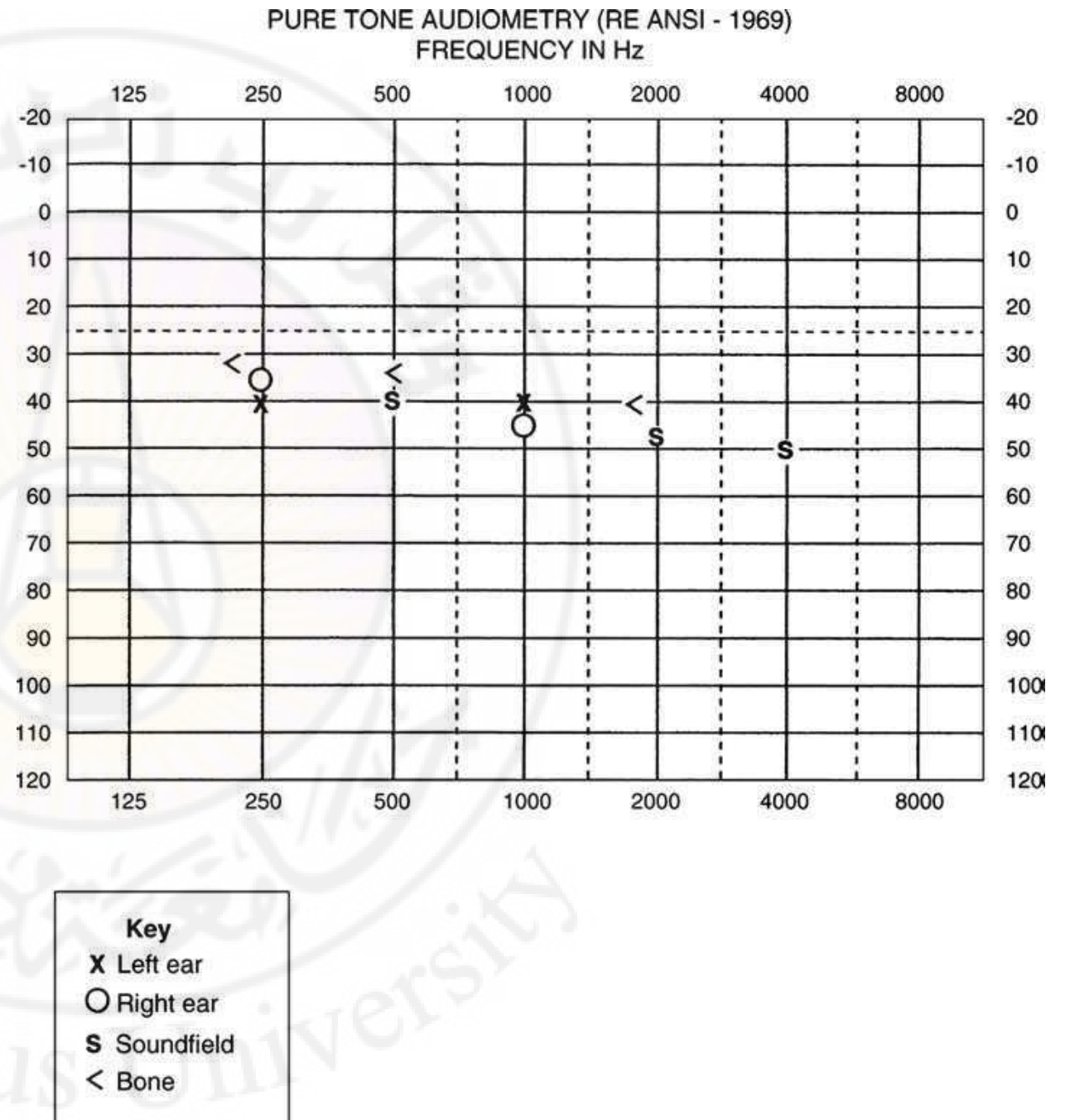
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- The use of symbols for **sound field thresholds**.
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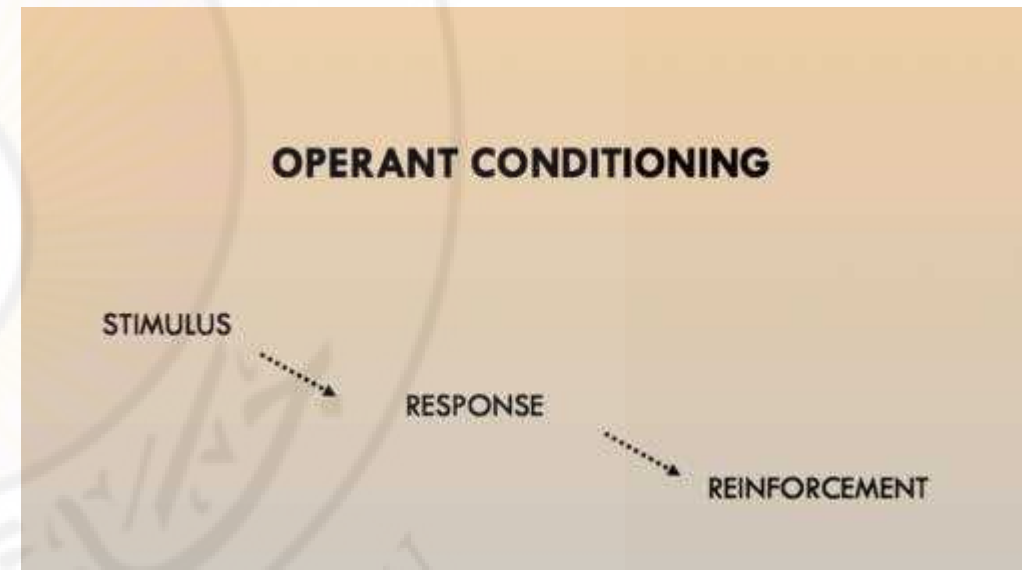


Audiogram with different information from different transducers.



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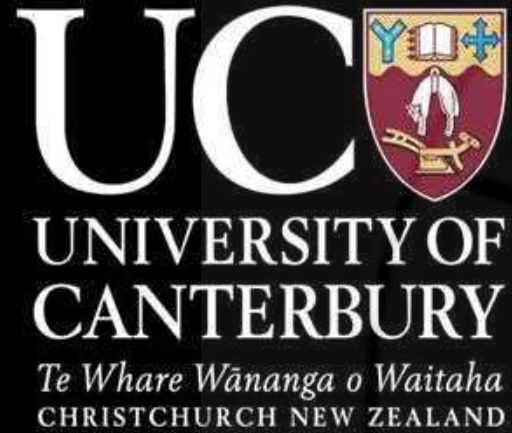
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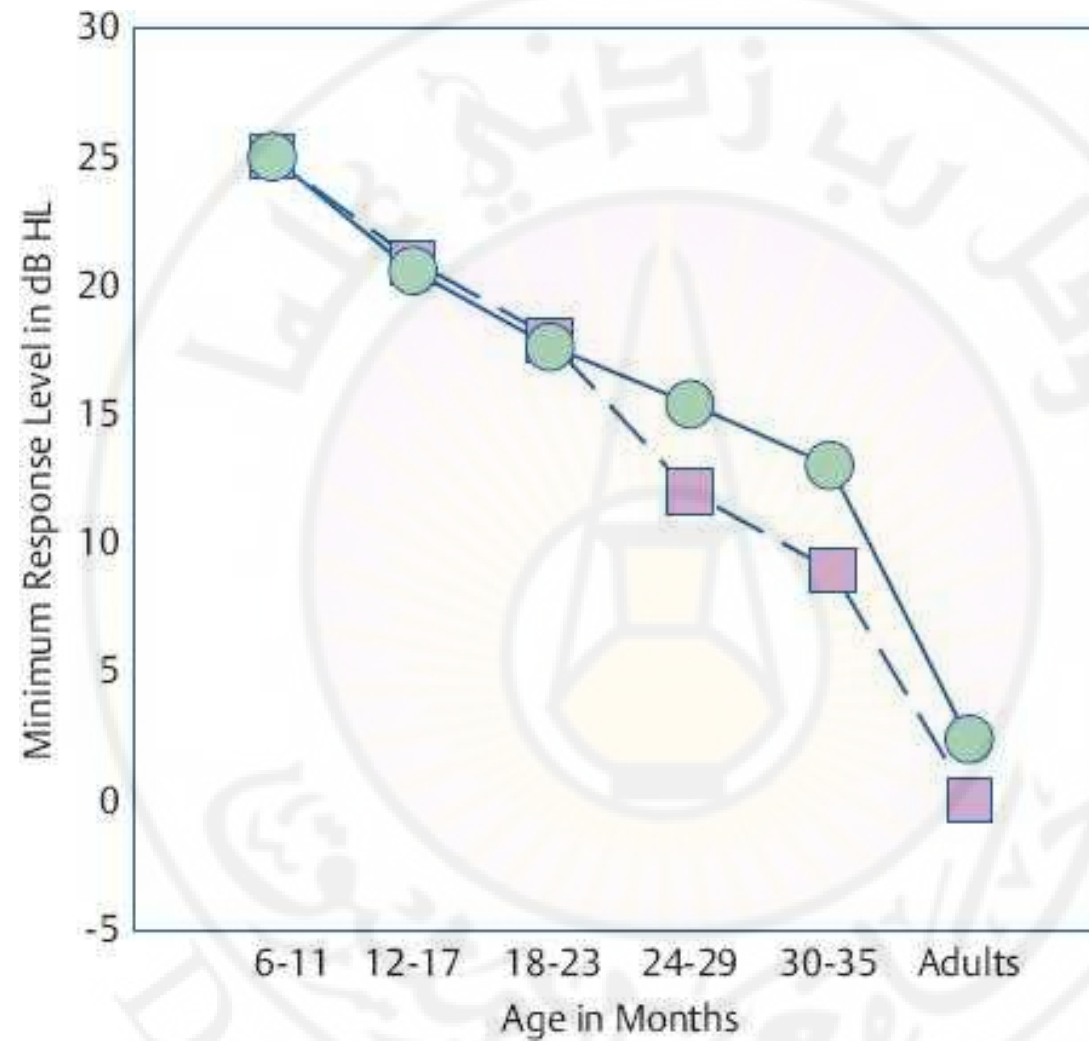
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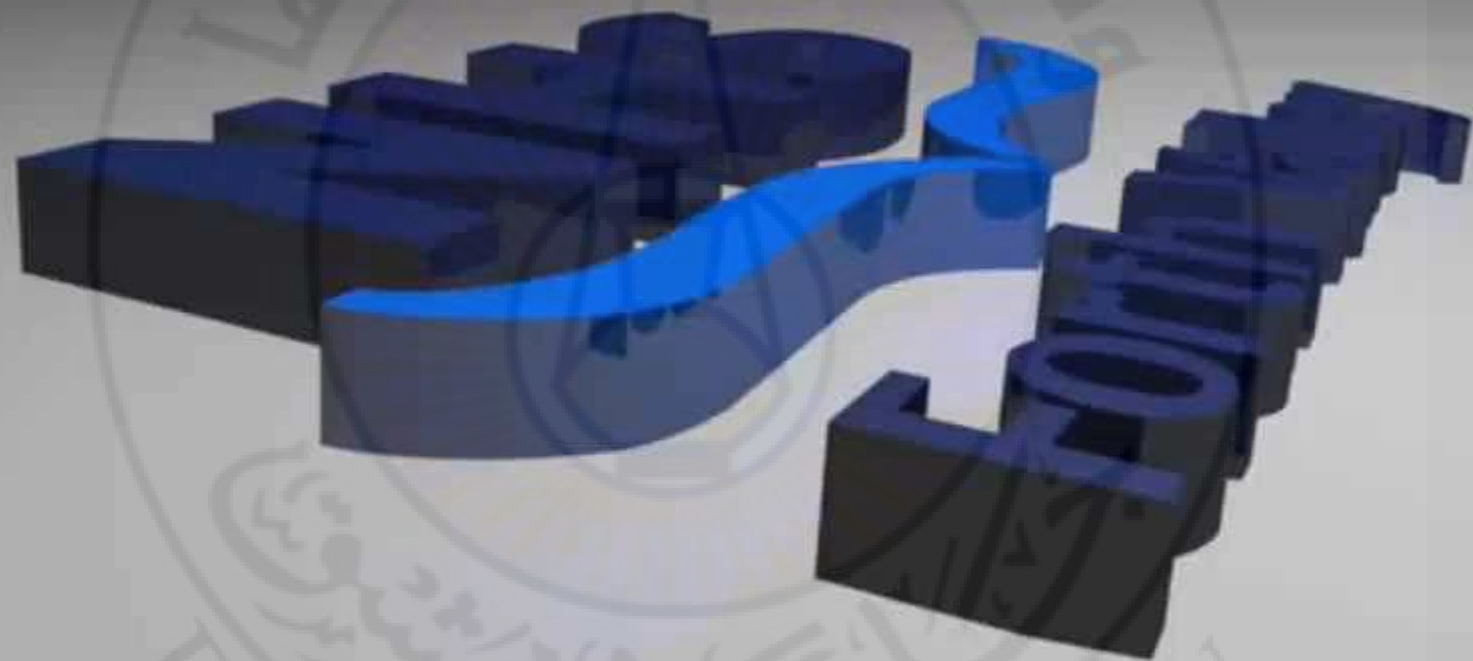
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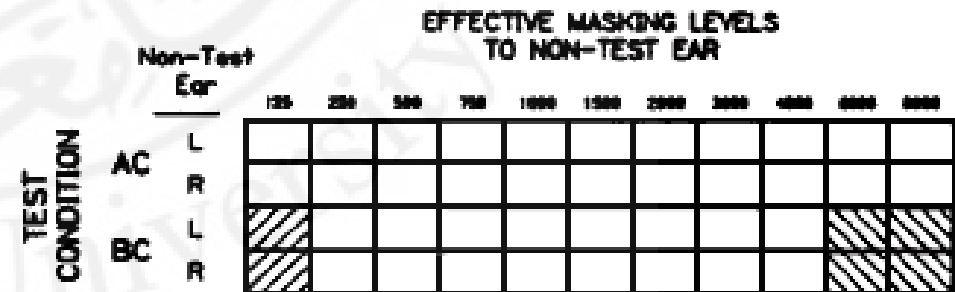
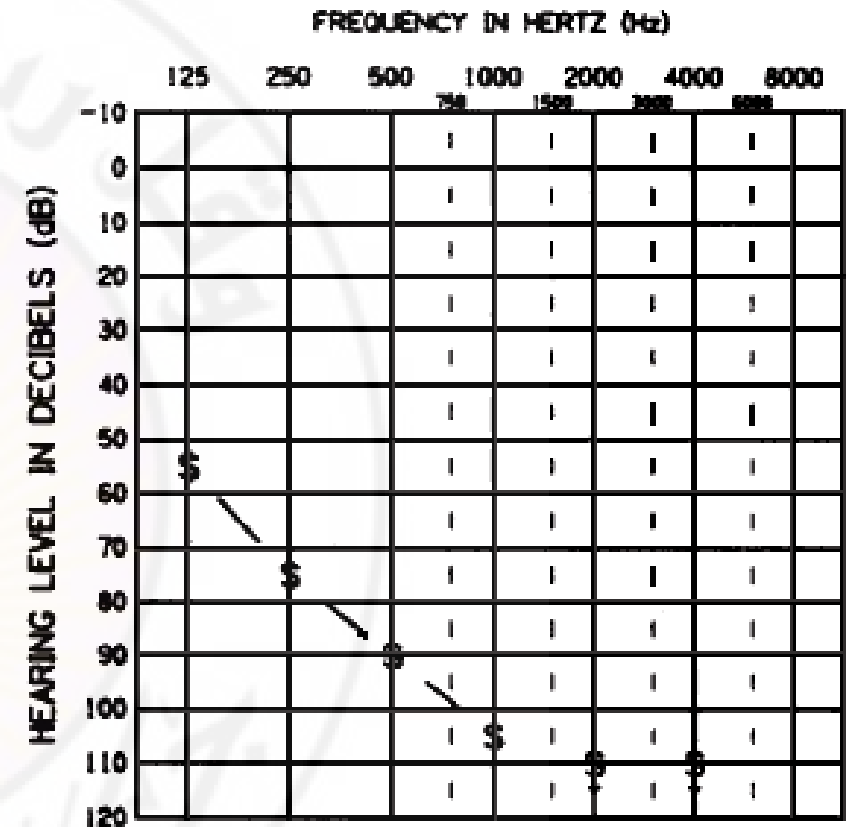


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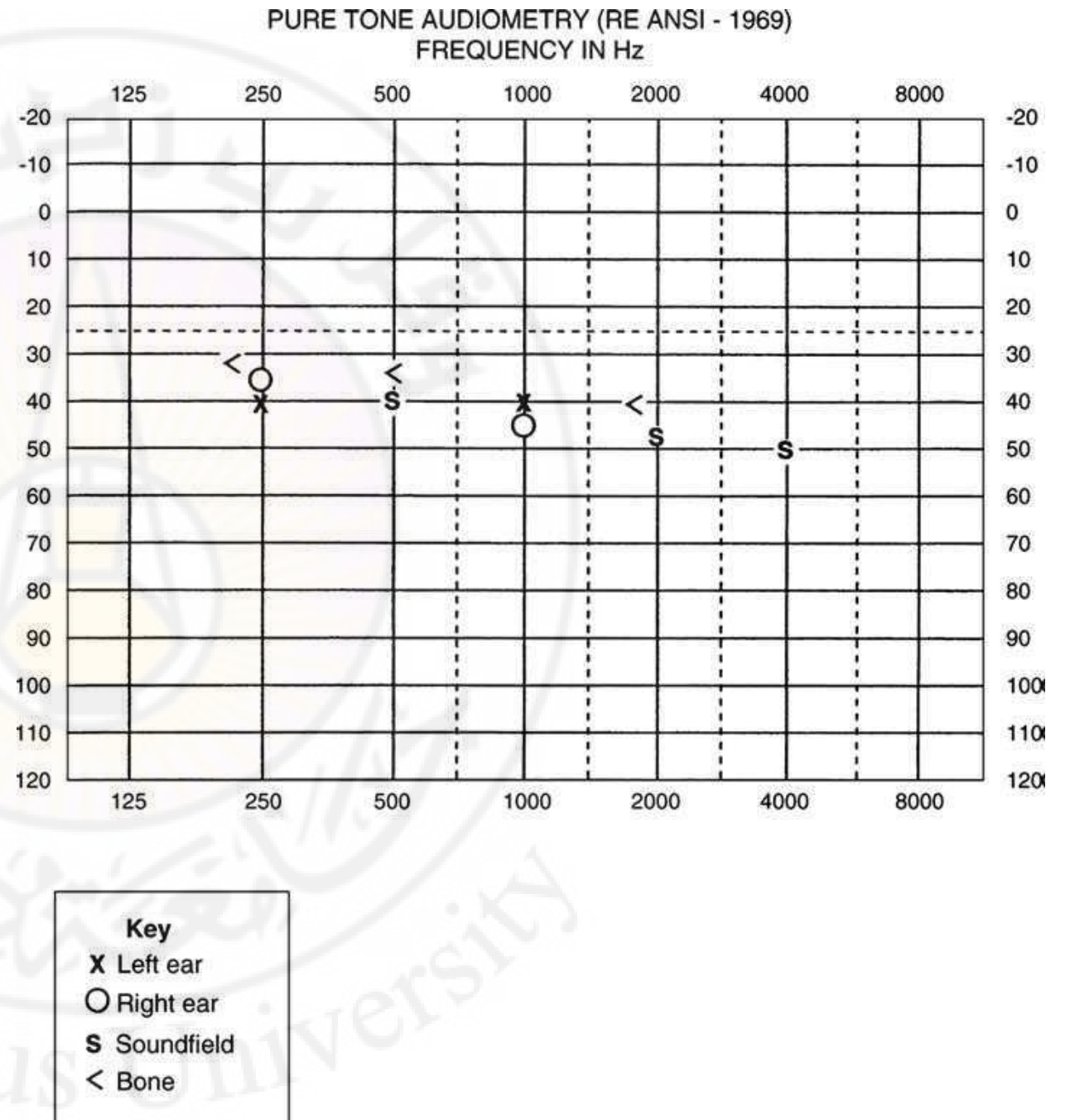
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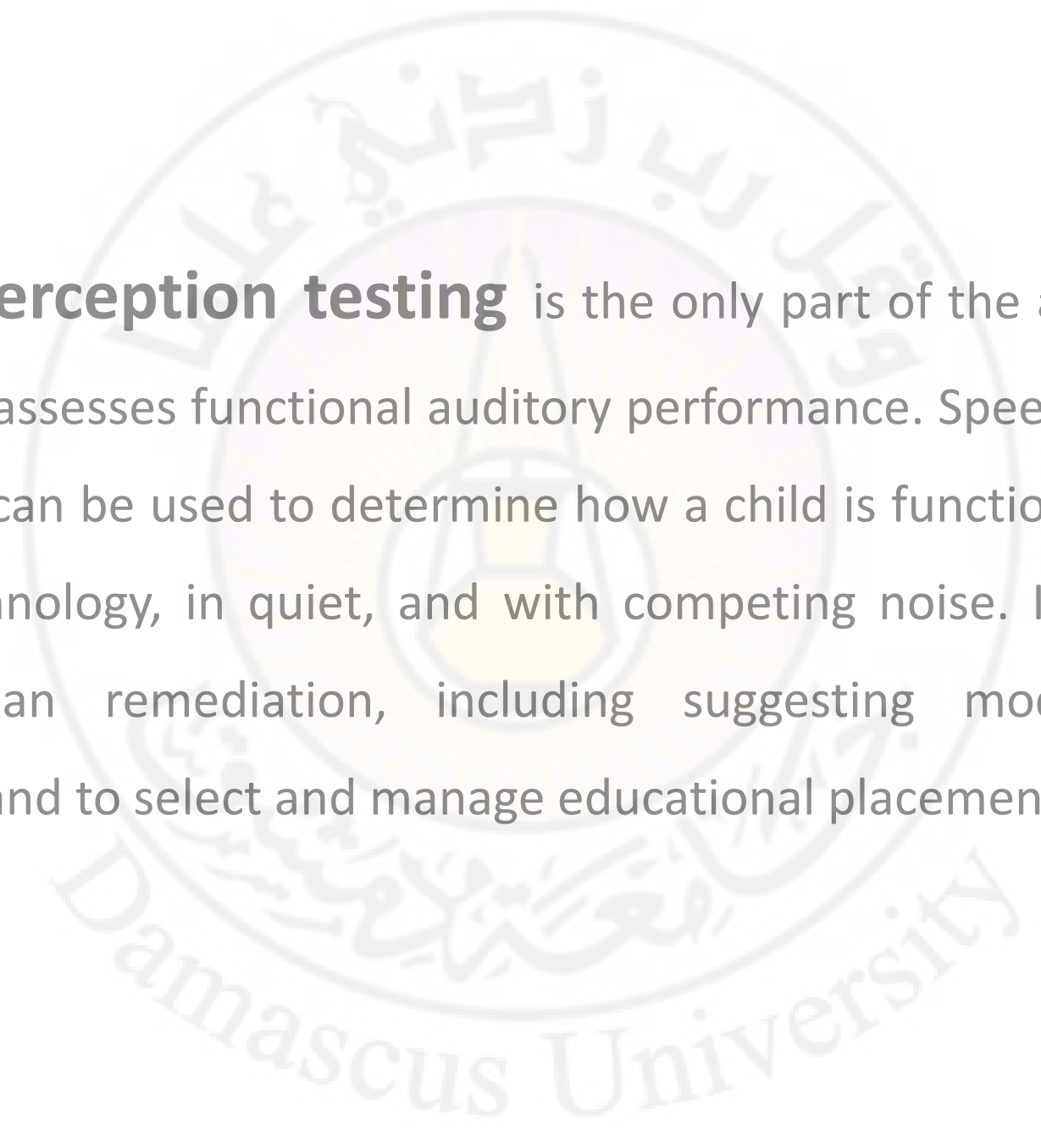


Audiogram with different information from different transducers.



The logo of Damascus University is a circular emblem. It features a central yellow lamp with a flame, set against a background of radiating yellow lines. The lamp is encircled by a purple ring. The entire emblem is surrounded by a larger circle containing Arabic calligraphy at the top and the English text "Damascus University" at the bottom.

Pediatric Speech Audiometry

A large, faint watermark of the Damascus University seal is centered in the background. The seal is circular, featuring a central emblem with a sunburst and a crescent moon, surrounded by Arabic calligraphy. The words "Damascus University" are written in English at the bottom of the seal.

Speech perception testing is the only part of the audiology test battery that assesses functional auditory performance. Speech perception information can be used to determine how a child is functioning with and without technology, in quiet, and with competing noise. It can also be used to plan remediation, including suggesting modifications in technology, and to select and manage educational placement.

AUDITORY HIERARCHY

Comprehension

ability to understand meaning of spoken language.

Identification

ability to identify what has been named or labeled.

Discrimination

ability to determine whether two sounds are the same or different.

Detection

awareness of when a sound is or is not present.

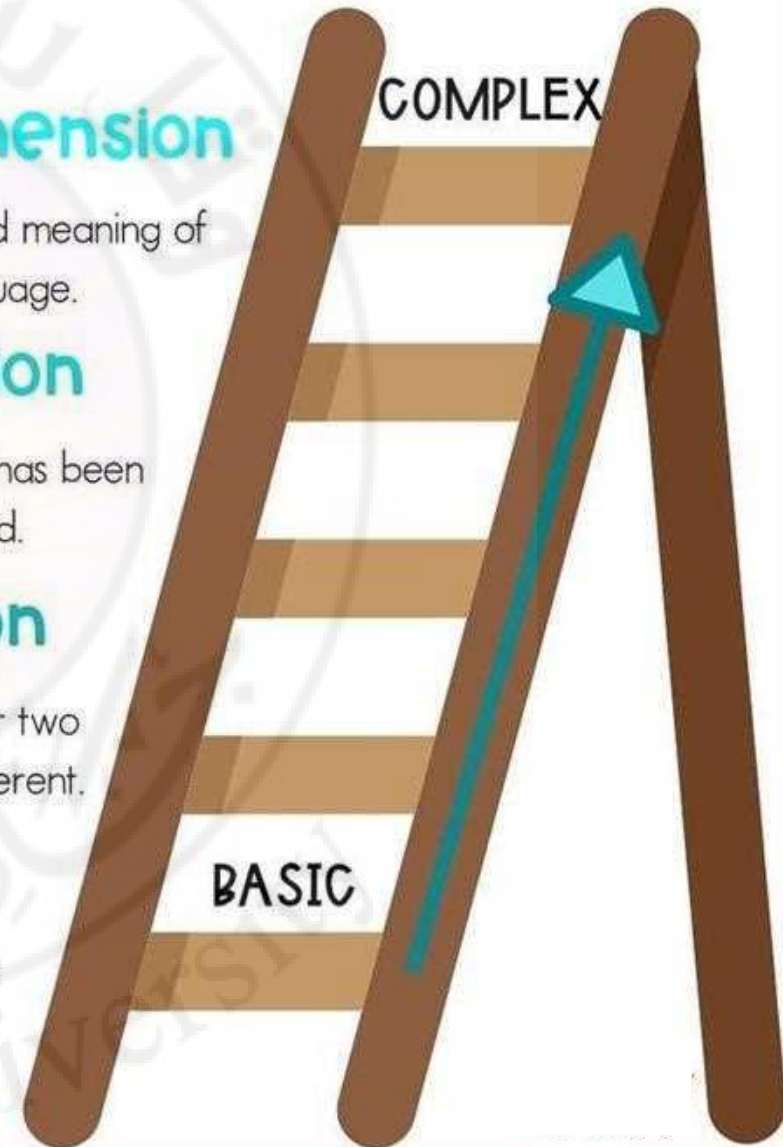
COMPLEX

BASIC

WRS

VRISD

SAT



Pediatric Speech Audiometry

- Speech is the signal of interest to most young children, and they often react positively to speech stimuli in a way that helps audiologists gauge their ability to hear and understand.
 - **Speech awareness threshold (SAT)**
 - **Speech reception threshold (SRT)**
 - **Word recognition scores (WRS)**



Speech awareness threshold (SAT)

- **SAT**, sometimes known as the speech detection threshold (**SDT**) is an informal procedure to establish a minimal response level for awareness of speech.
- The awareness or detection level is recorded as the lowest decibel level at which the child makes any response or acknowledgement to the onset of speech through loudspeakers or insert earphones.
- The SAT response can vary greatly from a surprise eye widening, or a head movement, or a slight head turn toward the speaker, or facial change when the stimulus signal is heard.
- The SAT is typically measured in the soundfield and reflects the level of hearing in the better ear.
- **SAT: Ling six-sound test**





Speech Reception Threshold (SRT)

- **SRT** determines the lowest level at which a person can identify speech stimuli 50% of the time.
- A carefully selected array of **familiar toys**, the names of which approach **spondaic** principles as closely as possible, should be selected. Nonspondaic words and point to body parts task can also be used at softer and softer speech levels until threshold is reached.
- Traditional bracketing procedure 10 dB down/5 dB up).
- Two valid responses on the ascending presentation should be accepted, and the SRT bracketing should then quickly switch to the opposite ear.



Establishing speech reception threshold (SRT)
with a pediatric picture board.

Speech Awareness Threshold 2 - 3 years old



Speech Recognition Threshold
4 - 6 years old



Select Picture Audiometry 3 - 4 years



Word recognition scores (WRS)

- The purpose of evaluating speech perception or speech intelligibility in children is to determine a child's auditory recognition of words presented at **conversational and suprathreshold levels**.
- The results obtained during speech intelligibility measures may actually be more a reflection of the child's interest and motivation for the task than an accurate indication of higher auditory speech recognition abilities.
- There is no standard technique or test of speech intelligibility in children. Although numerous tests have been developed over the years for this purpose.

Score:	General Word Recognition Ability:
90 to 100	Within Normal Limits
75 to 90	Slight Difficulty
60 to 75	Moderate Difficulty
50 to 60	Poor
< 50	Very poor

Test	Materials	Message set; response mode	Task domain	Minimum age (year)
SERT	30 environmental sounds (train, telephone)	Closed; picture identification	Unrestricted: 4 alternatives	3
ANT	Numbers 1 through 5	Closed; picture identification	Restricted: 5 alternatives	3
NU-CHIPS	50 monosyllabic words (food, school)	Closed; picture identification	Unrestricted: 4 alternatives	3
PSI	20 monosyllabic words (dog, spoon)	Closed; picture identification	Restricted: 5 alternatives	3
PSI	10 sentences, 2 syntactic constructions (Show me a bear brushing his teeth. A bear is brushing his teeth.)	Closed; picture identification	Restricted: 5 alternatives	3

Note: *SERT*, Sound Effects Recognition Test (Finitzo-Hieber, Gerling, Matkin, & Cherow-Skalka, 1980); *ANT*, Audio Numbers Test (Erber, 1980); *NU-CHIPS*, Northwestern University Children's Perception of Speech (Elliott & Katz, 1980); *PSI*, Pediatric Speech Intelligibility (Jerger, Hayes, & Jordon, 1980; Jerger, Jerger, & Lewis, 1981). From "Speech Audiometry," by S. Jerger, 1984: in J. Jerger, Ed., *Pediatric Audiology*, San Diego, CA: College-Hill Press.

Recorded versus Monitored Live Voice Testing

- MLV testing is often selected for young children because it is easier for the child and audiologist than recorded testing.
- However, MLV will frequently result in higher scores than recorded testing, thus overestimating the child's actual auditory abilities.
- It is important to use recorded tests whenever the child is capable of performing the task.
- Recorded testing will provide a more accurate representation of auditory performance.

Table 9.3 Word recognition testing for an 8-year-old using MLV and recorded testing demonstrating different results

Conditions	MLV	Recorded
50 dB	100	100
35 dB	100	95%
50 dB, +5 SNR	100	64%
50 dB, 0 SNR	100	50%
35 dB, 0 SNR	88%	32%

Abbreviations: MLV, monitored live voice; SNR, signal-to-noise ratio.

10. Physiologic Hearing Tests



Objective hearing testing

- An **objective** hearing test is one that defines a patient's hearing ability without the patient's active participation in the test.
- A **physiologic** hearing test relies on an autonomic response not based on the child's behavioral action.
- It needs to be made clear that the presence of some physiologic response, seemingly related to the presence of an auditory signal, does not ensure that the child does indeed **"hear."**
- Hearing, in this sense, implies meaningful interpretation of the sound so as to produce thought and language with verbal or nonverbal encoding and decoding.

Tympanometry

Acoustic Reflex

TEOAE

DPOAE

ABR

ASSR

•

•

•

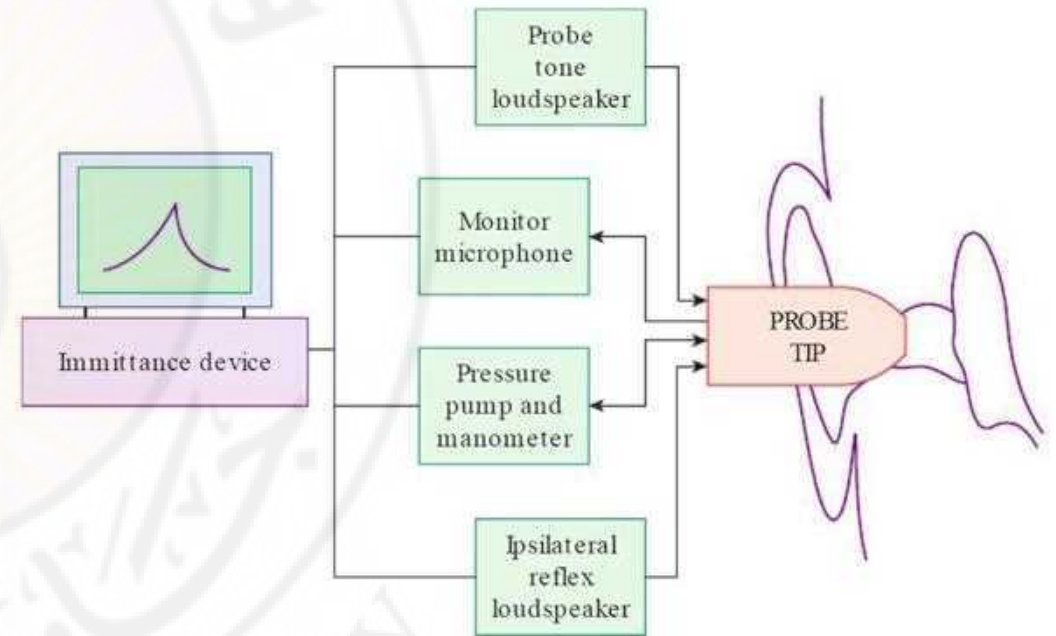
AMLR

ALLR

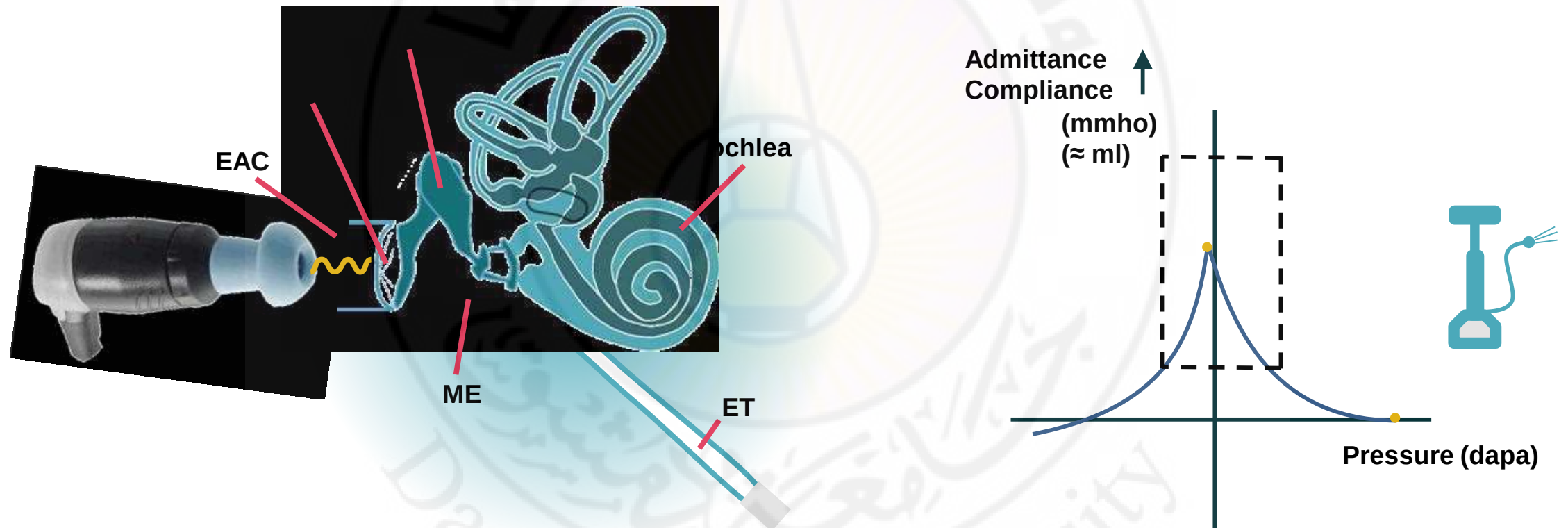
EcochG

Middle Ear Measurement

- **Tympanometry** and **acoustic reflex** testing are the basic components of the middle ear test battery, which provides information about the middle ear, cochlea, auditory nerve (cochlear nerve, cranial nerve VIII), auditory brainstem, and facial nerve (cranial nerve VII).
- Therefore, the middle ear test battery is useful as a **cross-check** with other physiologic and behavioral tests.



Tympanometry is an objective, physiological measure of acoustic admittance of the middle ear as a function of air pressure in a sealed ear canal.



Menu



226Hz

nt session

In ear



226 Hz

START

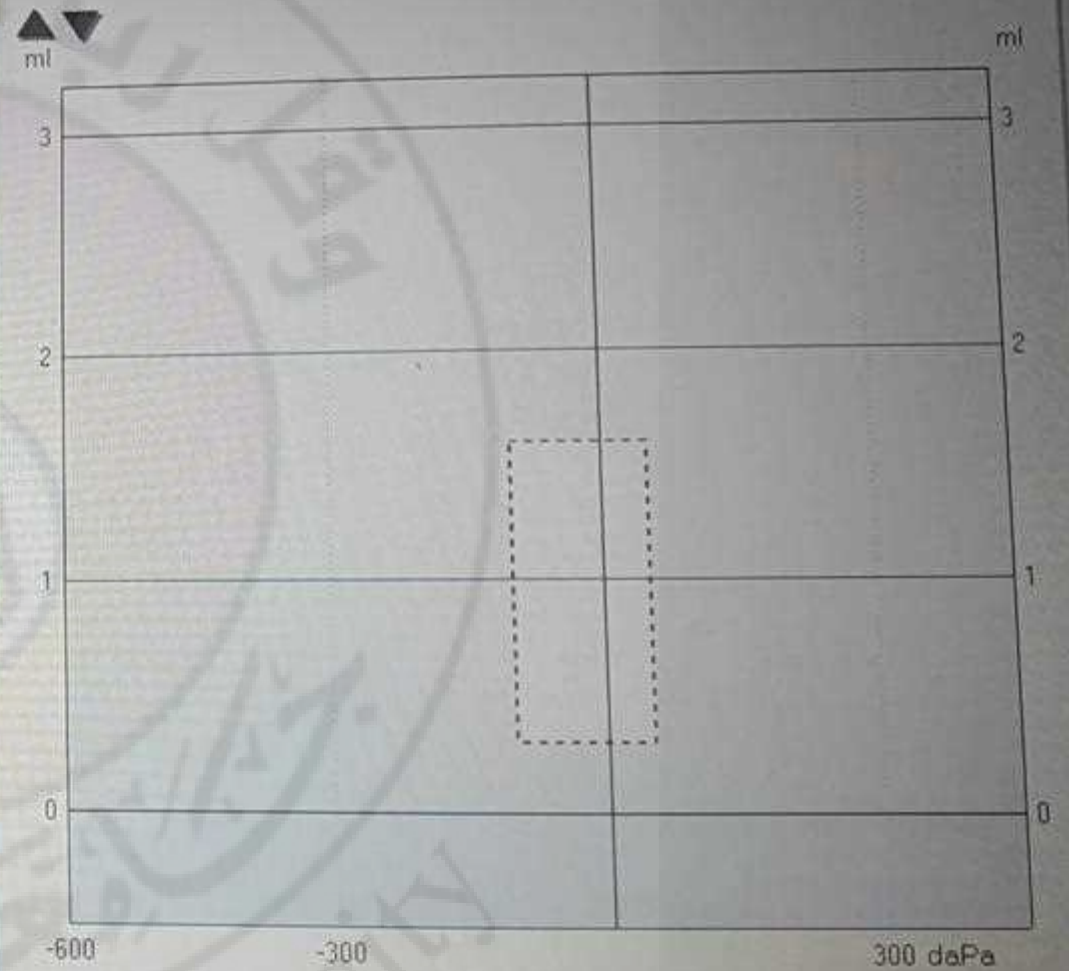
Tymp 226 Hz



Volume (ml)
Compliance (ml)

Pressure (daPa)
Gradient (daPa)

Tymp 226 Hz

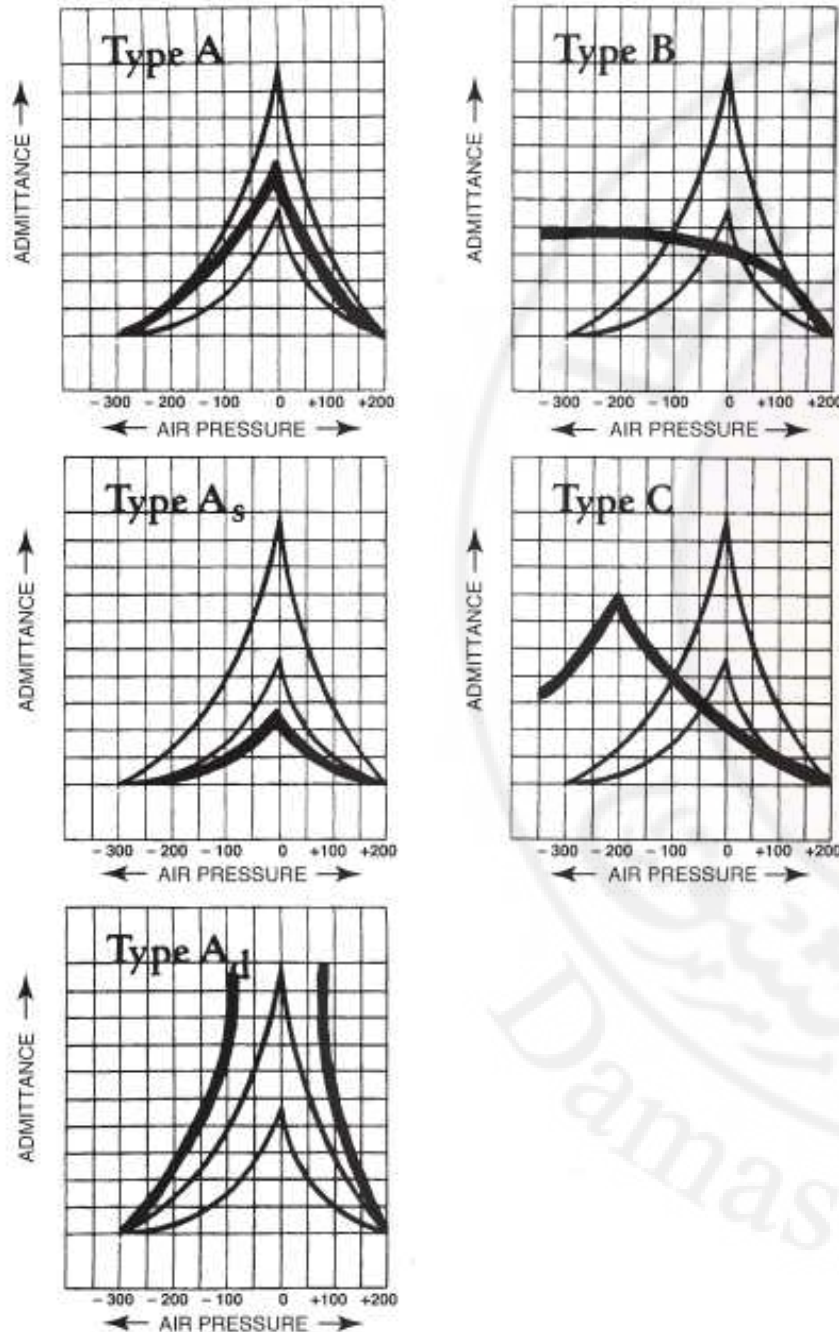


Volume (ml)
Compliance (ml)

Pressure (daPa)
Gradient (daPa)

Qualitative analysis of Tympanograms

Lidén–Jerger Model

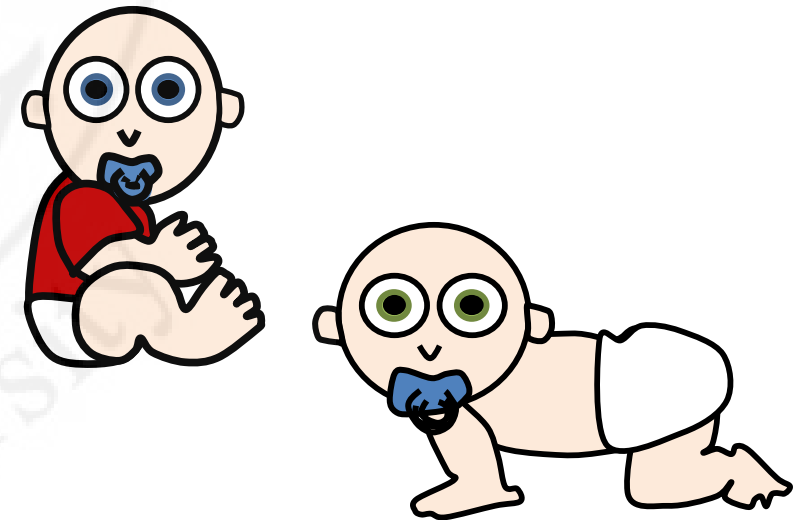


- Lidén–Jerger classification scheme for tympanometric shapes, based on qualitative analysis of the **height** and **location** of the primary peak.

- **A (An)**
- **As**
- **Ad**
- **C**
- **B**

Quantitative analysis of Tympanograms

1. **Equivalent Ear Canal Volume (V_{ea} or V_{ec})**
2. **Peak-Compensated Static Acoustic Admittance (Y_{tm})**
3. **Tympanometric Peak Pressure (TPP)**
4. **Tympanometric Gradient and Width (TW)**



Effects of Middle-Ear Pathologies

	Y	TW	Volume	Mass	Springiness	Resonant Frequency
Middle-ear effusion	Low	Wide	Normal	↑	↓	Low
Monomer or ossicular discontinuity	High	Narrow	Normal	↑	↑	Low
Perforation	Flat or variable	–	High	–	–	–
Tympanosclerosis	Normal to low	Normal	Normal	↑	↓	Low
Cholesteatoma	Low	Wide	Normal	–	↓	Low
Lateral ossicular fixation	Low	Wide	Normal	–	–	High
Medial ossicular fixation [otosclerosis]	Normal	Normal/narrow	Normal	–	↓	Normal to high

Tympanometry and Equivalent Ear Canal Volume (Veq) in Children

Tympanogram	Equivalent ear canal volume in cc ³	Etiology
Type A	0.8–1.0	Normal middle ear
Type B	<0.3	Cerumen or canal wall
	0.8–1.0	Serous otitis; middle ear congenital anomaly
	>2.5	Tympanic membrane perforation; Patent ventilation tube in TM
Type C	0.8–1.0	Inadequate eustachian tube function

Tympanometry in Newborns and Infants



- In neonate ears with confirmed middle-ear disease, 226-Hz tympanograms may not provide accurate diagnostic information.
- tympanometry using a **higher probe tone frequency** (e.g., 1,000 Hz) is more sensitive to middle-ear status, compared with 226-Hz tympanometry, in infants less than 4 to 6 months old.
- These effects are probably related to developmental differences between infant ear canals and middle ears relative to those of older children and adults.
- Anatomical and physical differences in the infant ear, ear canal wall flaccidity, smaller ear canal and middle-ear space, TM thickening, presence of middle-ear fluid and mesenchyme in some ears, and a more horizontal orientation of the TM with respect to the axis of the ear canal, are the most likely contributors.

Normative Data for Tympanometry (226- and 1,000-Hz Probe Tones) in Infants and Children

Study	Age	Probe Frequency [Hz]	Y_{tm} 5–95 Percentiles [mmho]	Tympanic Width [daPa]
Margolis et al. [2003]	Birth to 4 wks	1,000	0.60–4.3 [–400 tail]	NA
Shahnaz et al. [2008]	32 wks gestational age	1,000	0.10–1.50 [+250 tail] 0.53–2.31 [–400 tail]	NA
Kei et al. [2003]	1–6 days	1,000	Right ears 0.39–2.28 [+200 tail]	Right ears 56.6–154
	1–6 days	1,000	Left ears 0.39–1.95 [+200 tail]	Left ears 46.1–144.2
Roush et al. [1995]	6–12 mos	226	0.20–0.50 [+200 tail]	102–234
	12–18 mos	226	0.20–0.60 [+200 tail]	102–204
	18–24 mos	226	0.30–0.70 [+200 tail]	102–204

Reprinted with permission from Hunter and Shahnaz. [2014] Table 8-1, pg. 117.

Frequency range of middle ear assessments

Wideband Tympanometry

Audiometric Frequencies

226 Hz

250 Hz

500 Hz

1000 Hz

2000 Hz

4000 Hz

8000 Hz

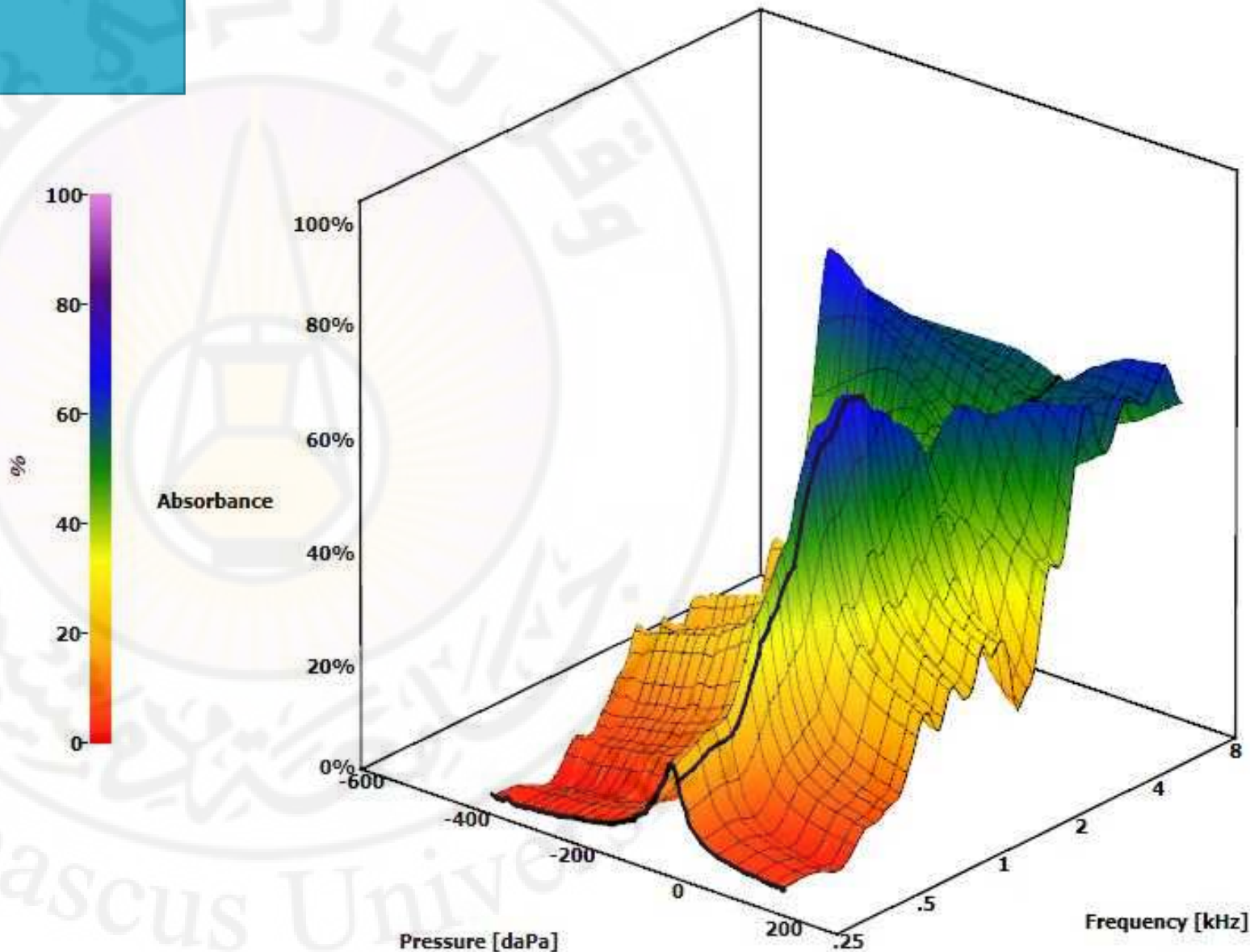
Adults
Tympanometry

Infants
Tympanometry

Wideband Tympanometry

A new dimension in middle ear diagnostics

- Wideband (WB) reflectance and WB absorbance are names for an emerging type of acoustic ear-canal measurement that provides new dimensions for middle-ear assessment across a wide range of frequencies.
- Reflectance measurements are performed over a frequency range from 0.22 kHz up to approximately 8 kHz using a WB stimulus such as a click.



Wideband Tympanometry

Benefits for Children

1

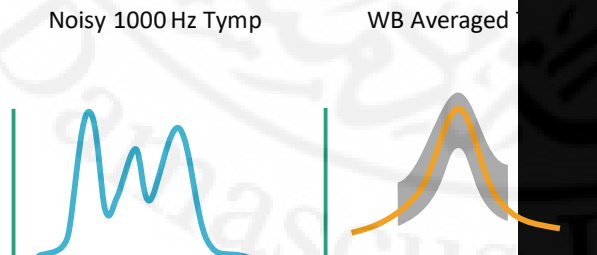
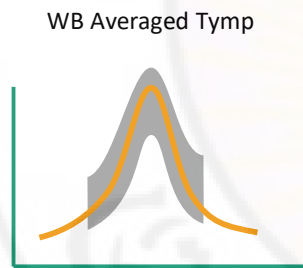
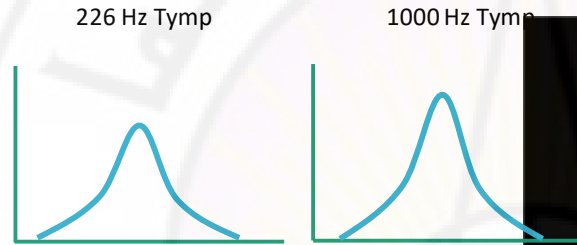
Multiple Tympanograms
Save time with fewer measurements

2

Improved Sensitivity
Improved accuracy over standard tympanometry

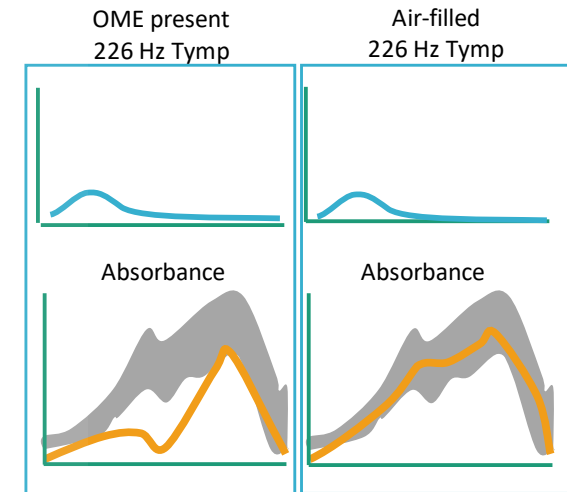
3

Stable and robust results
Results with noisy



4

Know your Type C Tympanogram
When OME is in the middle



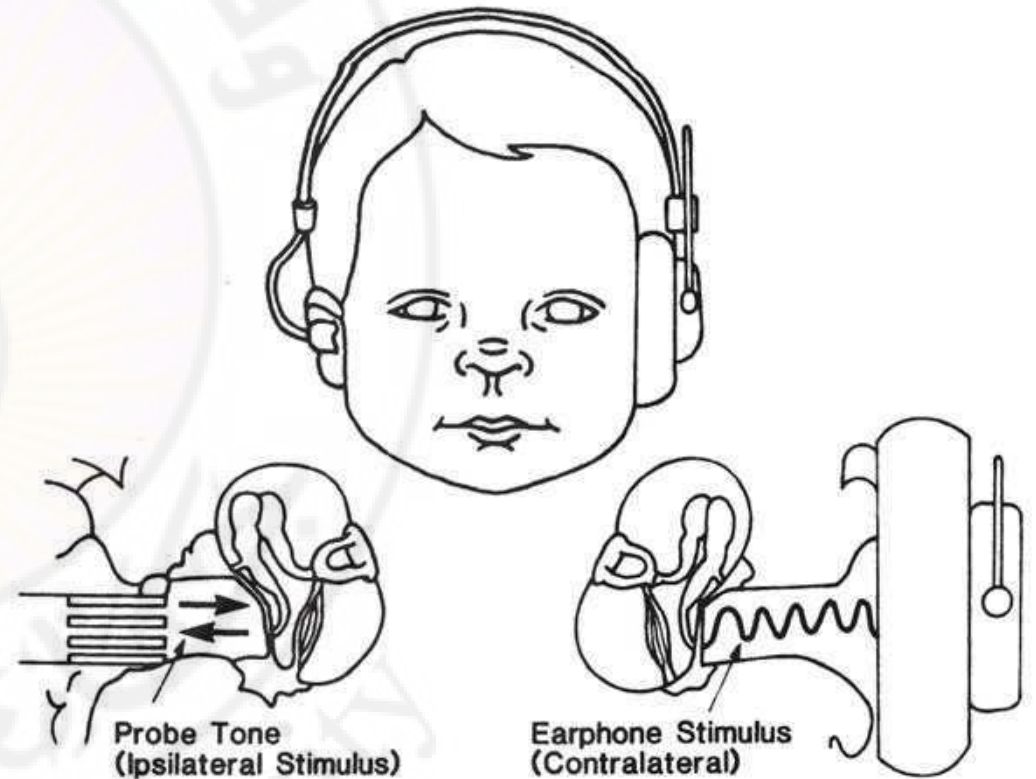
5

Drawing Rainbows
Generates measurement



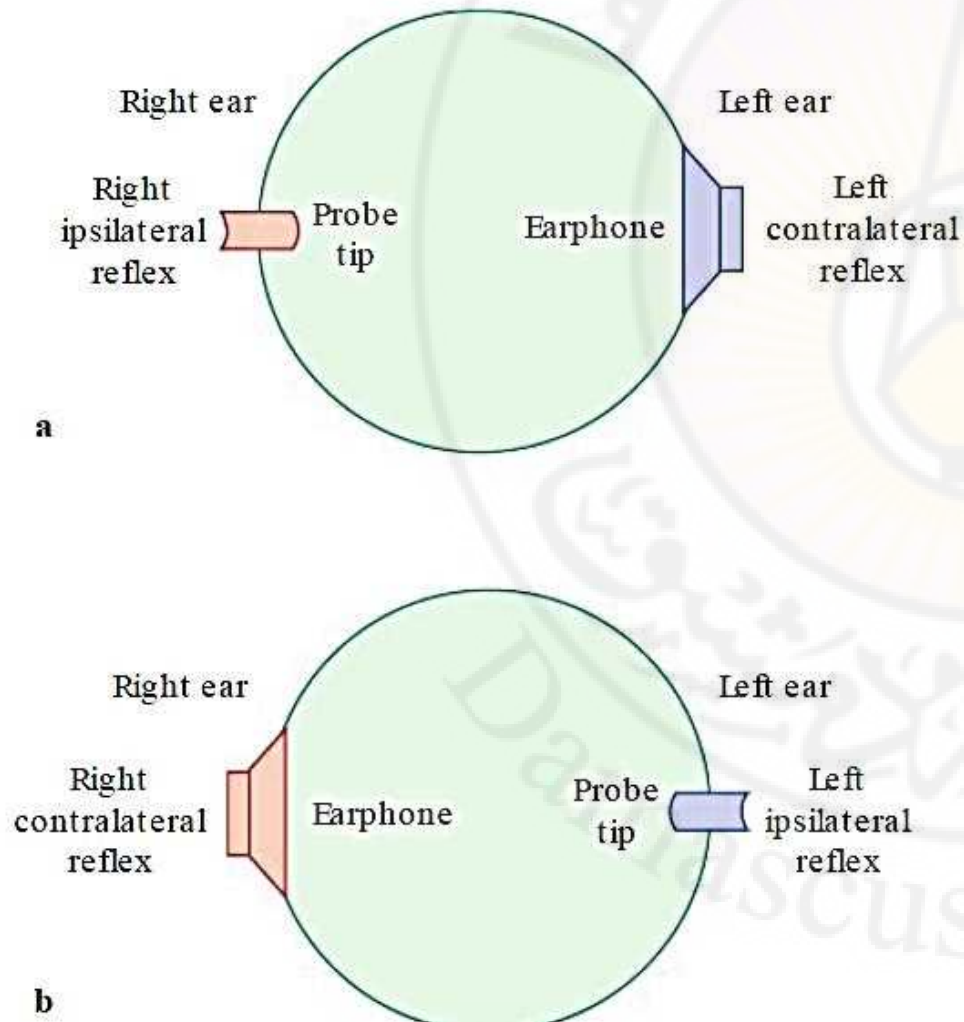
Acoustic reflex testing

- The acoustic reflex test is based on the presentation of a sound of sufficient intensity at which the **stapedial muscles** will involuntarily contract.
- In normal hearing persons a bilateral acoustic muscle reflex can be elicited with pure tone signals between 70 and 100 dB hearing threshold level (HTL).
- The lowest signal intensity capable of eliciting the acoustic reflex is the **acoustic reflex threshold** for the stimulated ear.



Acoustic Reflex test conditions

The ASR test conditions are described based on the ear to which the activator is presented (ANSI, 2012).



a Arrangement in terms of stimulus and probe		
TEST CONDITION	STIMULUS	PROBE
Right contralateral (crossed)	Right ear	Left ear
Left contralateral (crossed)	Left ear	Right ear
Right ipsilateral (uncrossed)	Right ear	Right ear
Left ipsilateral (uncrossed)	Left ear	Left ear

b Arrangement in terms of right ear and left ear		
TEST CONDITION	RIGHT EAR	LEFT EAR
Right contralateral (crossed)	Stimulus	Probe
Left contralateral (crossed)	Probe	Stimulus
Right ipsilateral (uncrossed)	Stimulus & probe	
Left ipsilateral (uncrossed)		Stimulus & probe

Interpreting Crossed and Uncrossed Acoustic Reflexes With the Audiogram to Identify Site of Lesion of Hearing Impairment

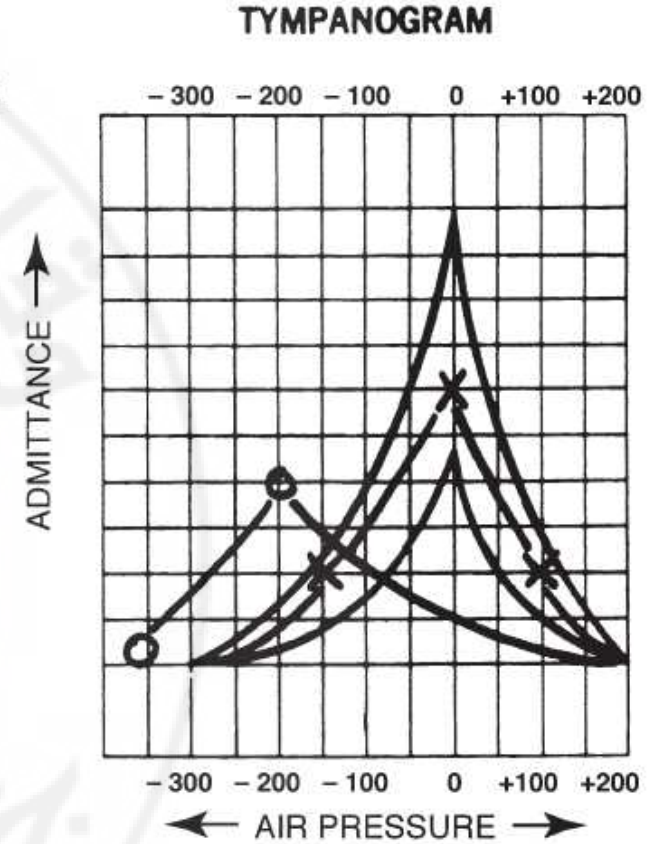
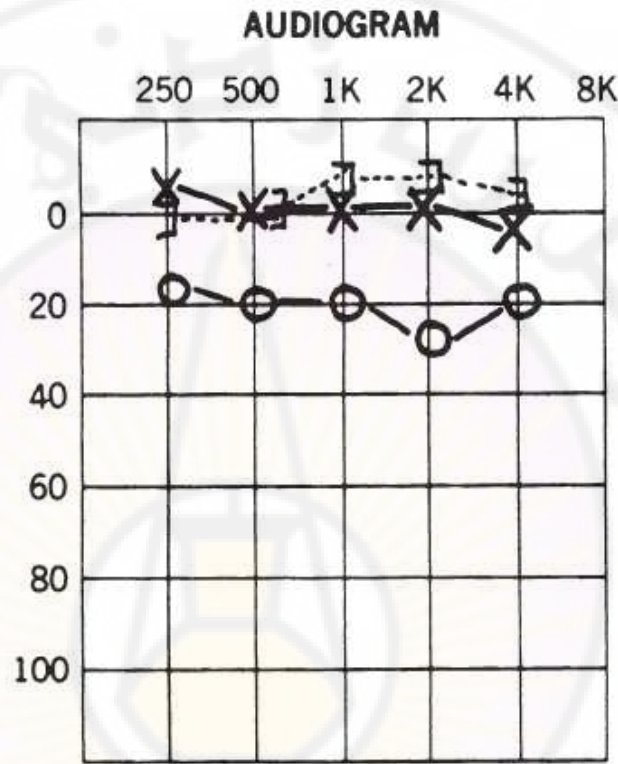
Reflex pattern	Audiogram	Predicted site
Neither crossed or uncrossed can be elicited from either ear	Bilateral air-bone gap	Middle ear
Neither crossed or uncrossed can be elicited from either ear	Bilateral severe sensory loss	Cochlea
Neither crossed or uncrossed can be elicited from either ear	Normal	Brainstem

Use of Immittance to Help Confirm Audiometric Impression in Evaluation of Young Children

Tympanometry	Static compliance	Acoustic reflex	Confirm behavioral audiometric impression
Type A bilaterally	Within normal range bilaterally	Normal bilaterally	Bilateral normal hearing or bilateral mild-to-moderate sensorineural hearing loss or unilateral mild-to-moderate sensorineural hearing loss
Type A in one ear; type B or C in other ear	Normal in A ear, low in B or C ear	Absent bilaterally	Unilateral conductive loss
Type B or C bilaterally	Low bilaterally	Absent bilaterally	Bilateral conductive loss

Source: From "Clinical Experience with Impedance Audiometry," by J. Jerger, 1970, *Archives of Otolaryngology*, 92, pp. 311–324.

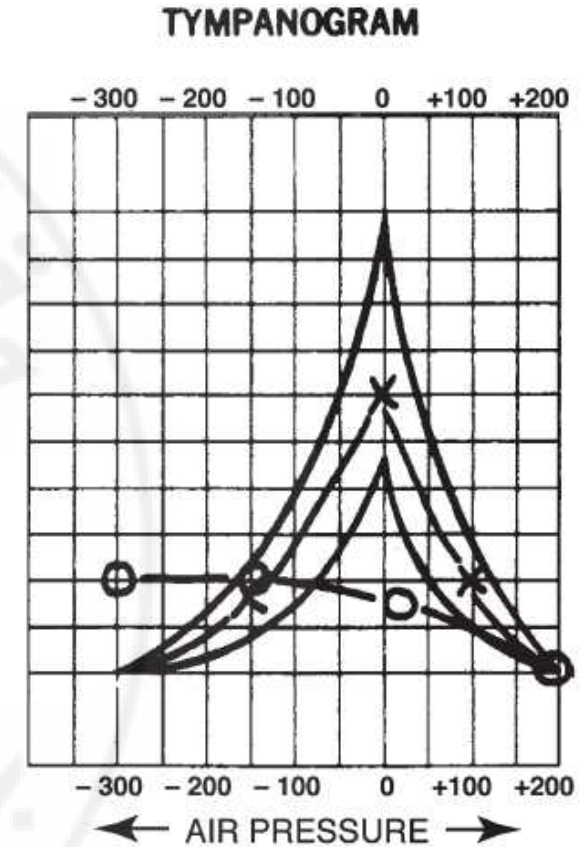
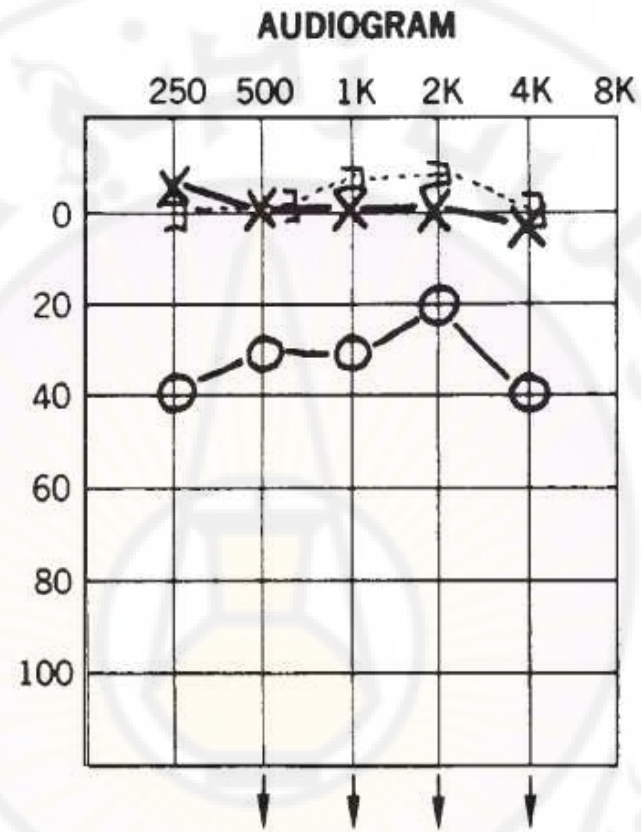
Case 1:



EAR		500	1K	2K
STIM RE	IPSI	a	a	a
	CONTRA	105	100	110
STIM LE	IPSI	100	100	100
	CONTRA	a	a	a

STAPEDIUS REFLEX

Case 2:

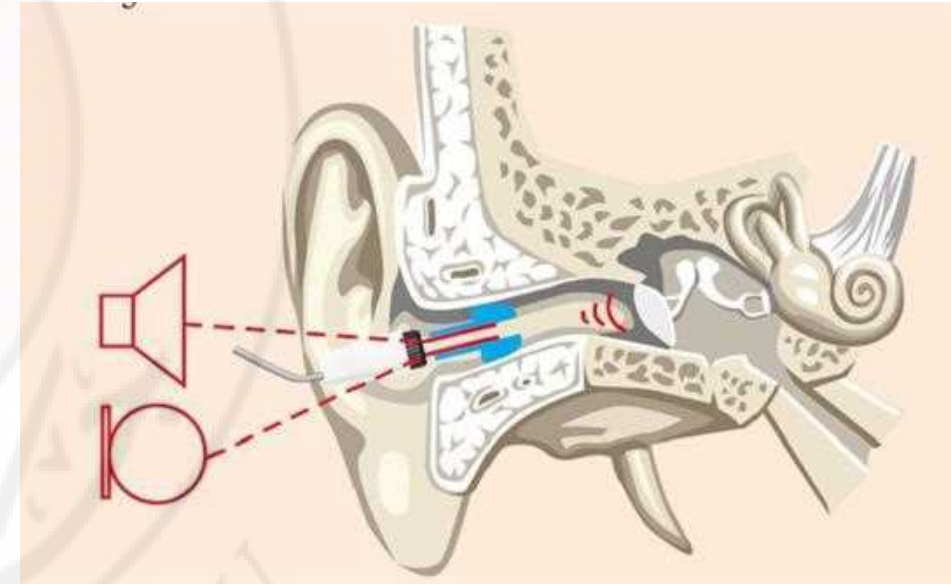


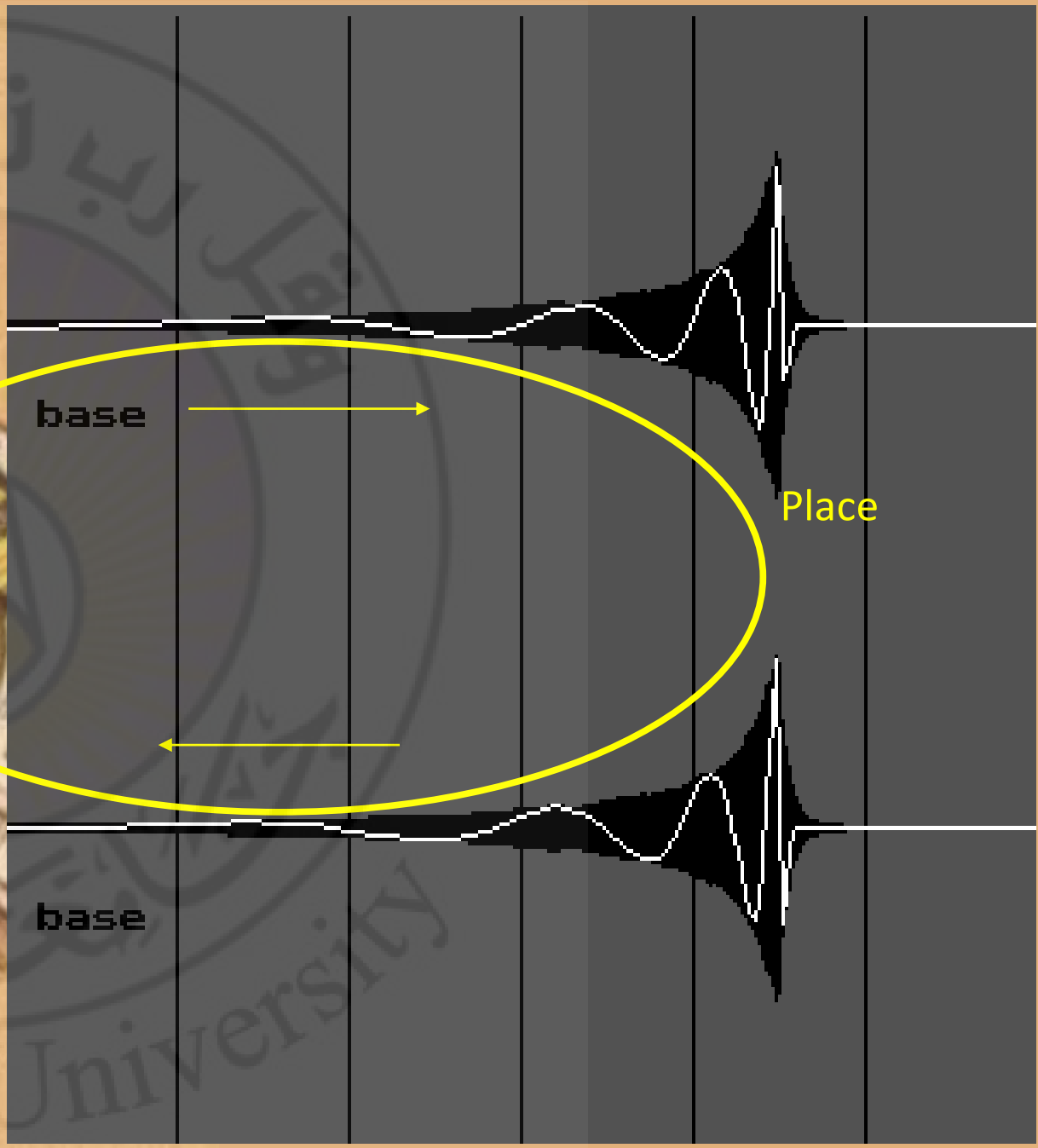
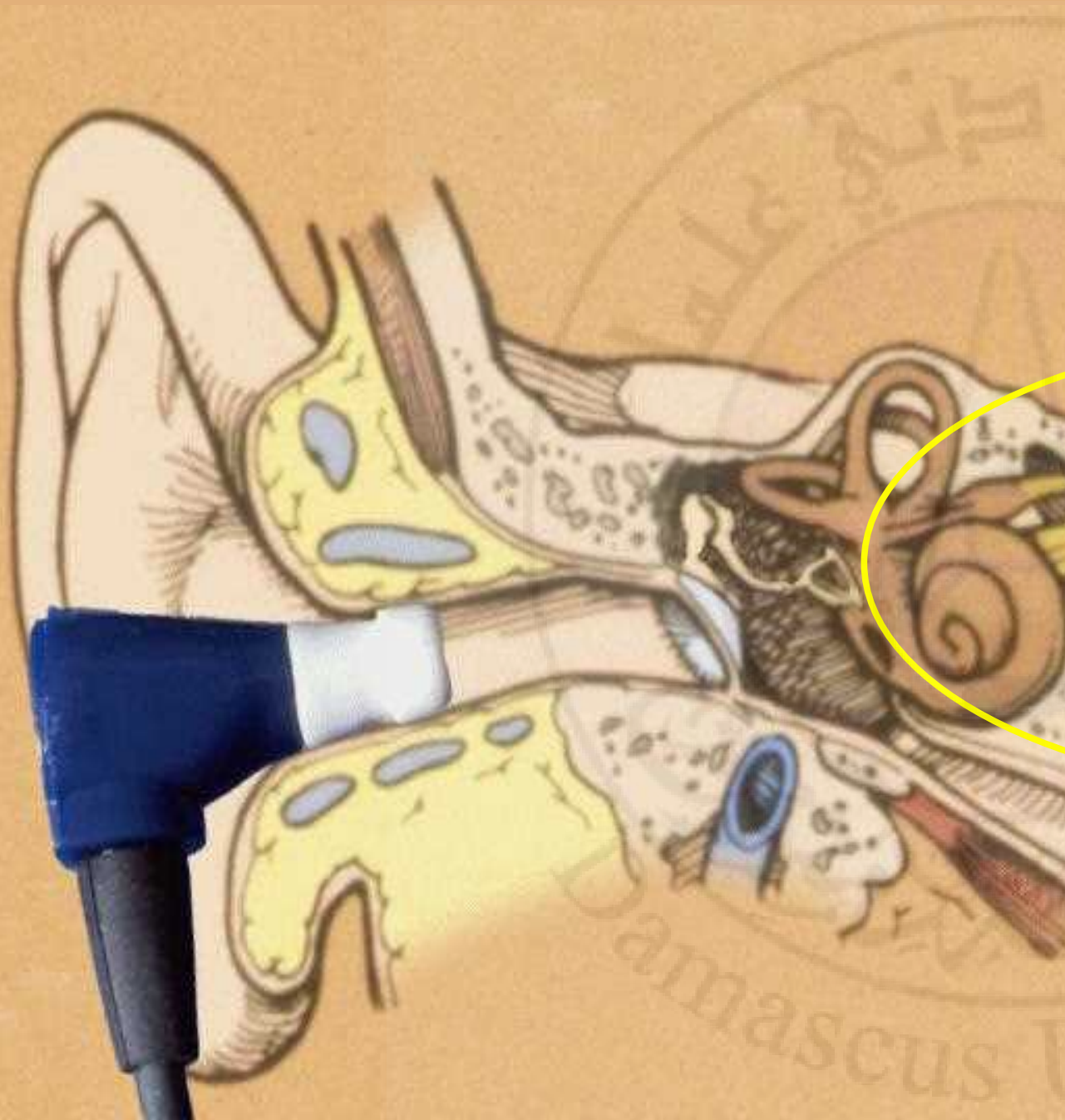
EAR		500	1K	2K
STIM RE	IPSI	a	a	a
	CONTRA	a	a	a
STIM LE	IPSI	100	100	100
	CONTRA	a	a	a

STAPEDIUS REFLEX

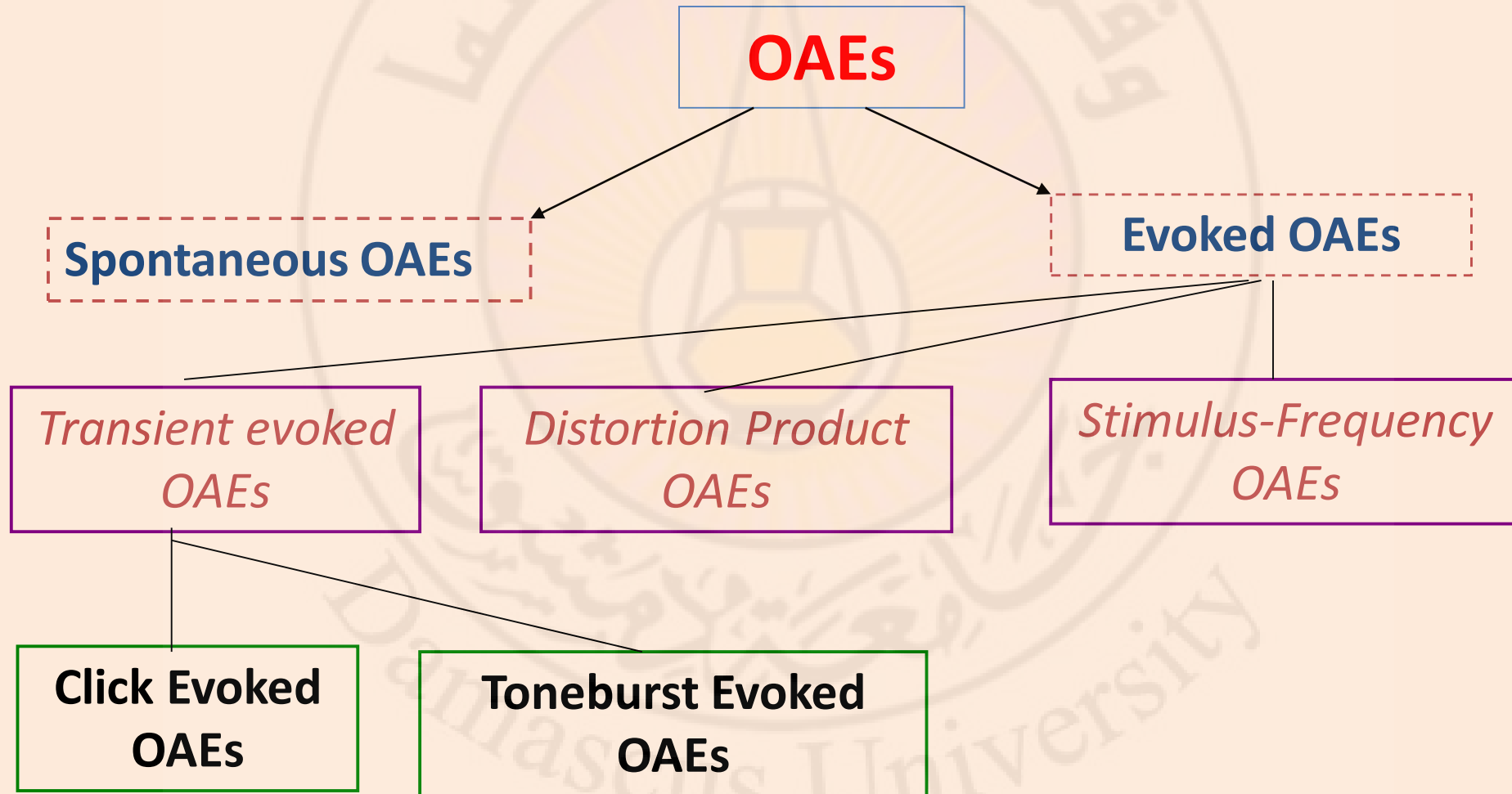
Otoacoustic Emissions

- Otoacoustic emissions are an essential byproduct of the cochlear amplification processes and thus serve as a very sensitive indicator of general cochlear health.
- Otoacoustic emissions can be classified either by the type of stimulus used to record them or by their generation mechanism.
- Although otoacoustic emissions reflect cochlear health, they are significantly influenced by the acoustics of the ear canal as well as forward and reverse transmission through the middle ear.
- Otoacoustic emissions play a critical role in both screening (e.g., newborn, school age) and diagnostic (e.g., ototoxicity) protocols.





Types of OAEs

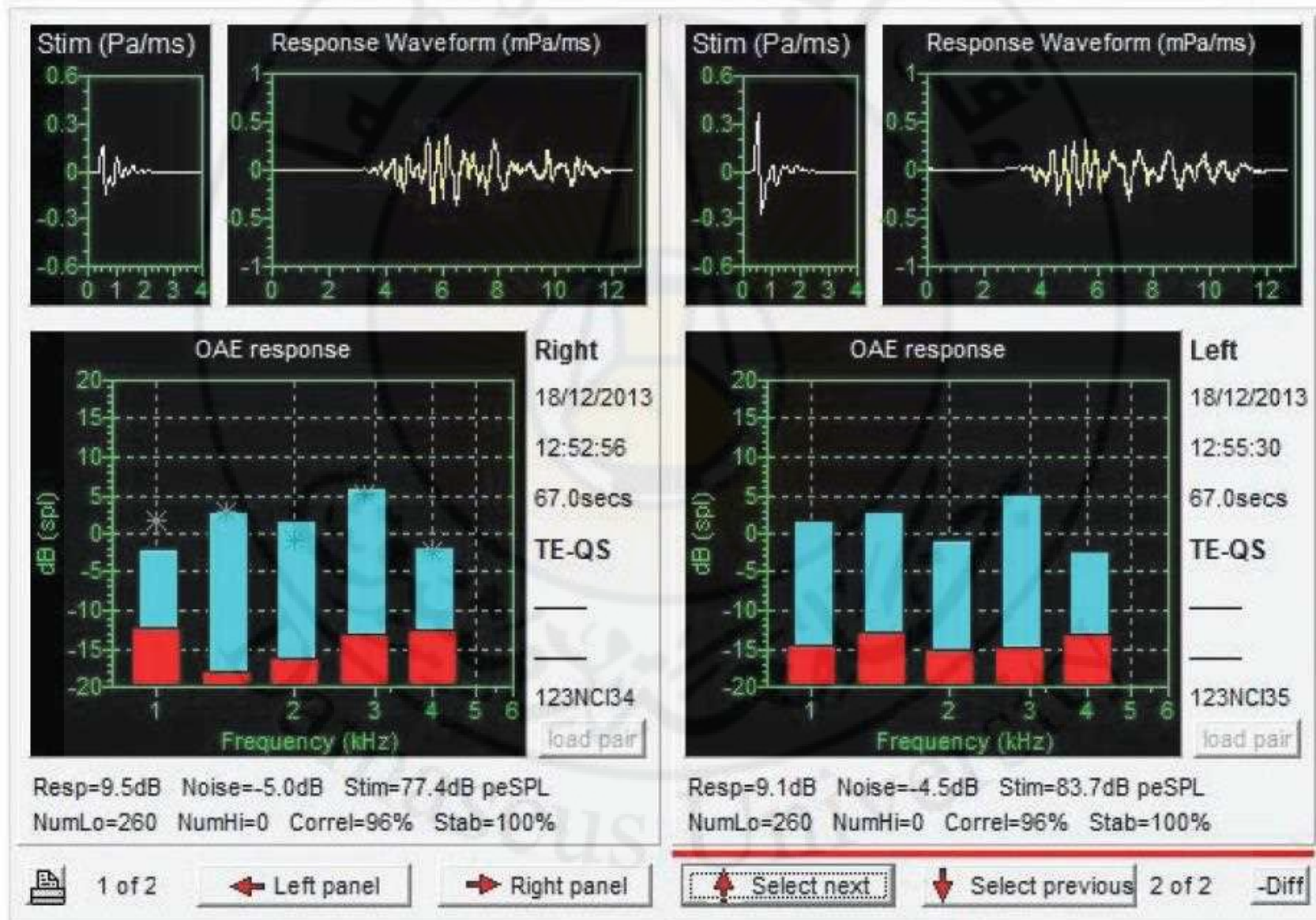


Factors affecting OAEs recording

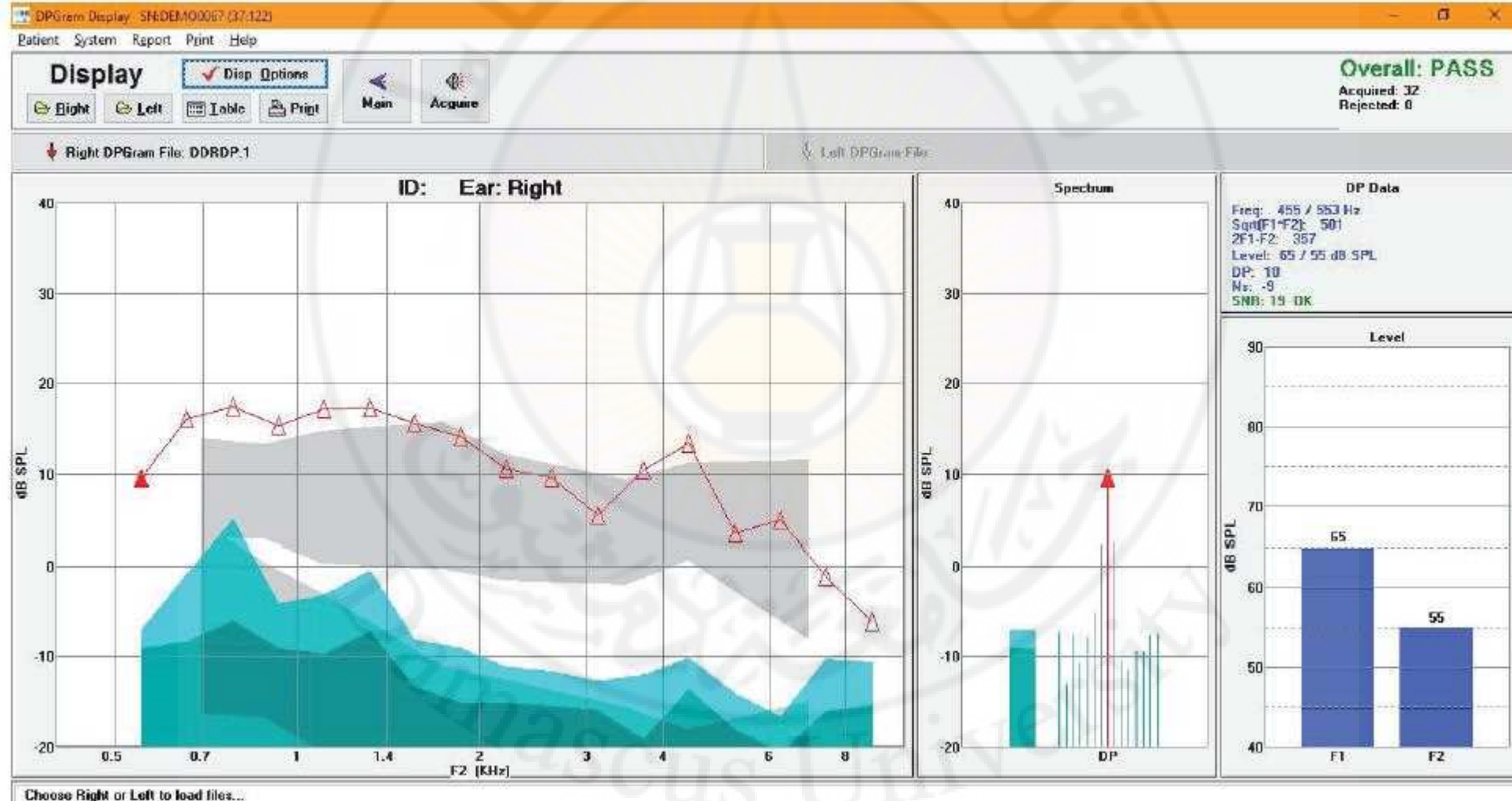
- **Middle ear state**
- **Noise in room**
- **Noise of subject**
- **Debris in EAC**



TEOAE recording



DPOAE recording

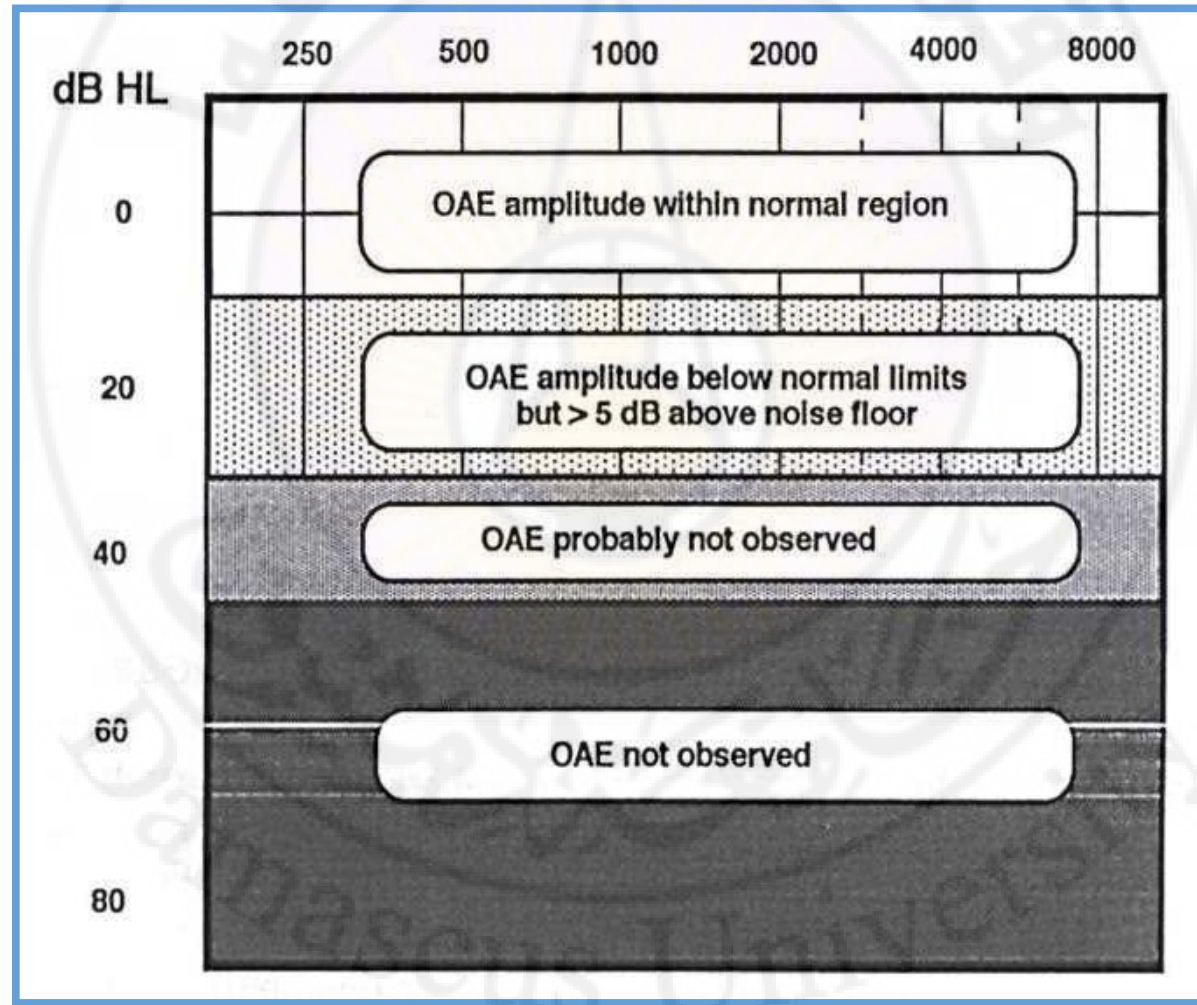


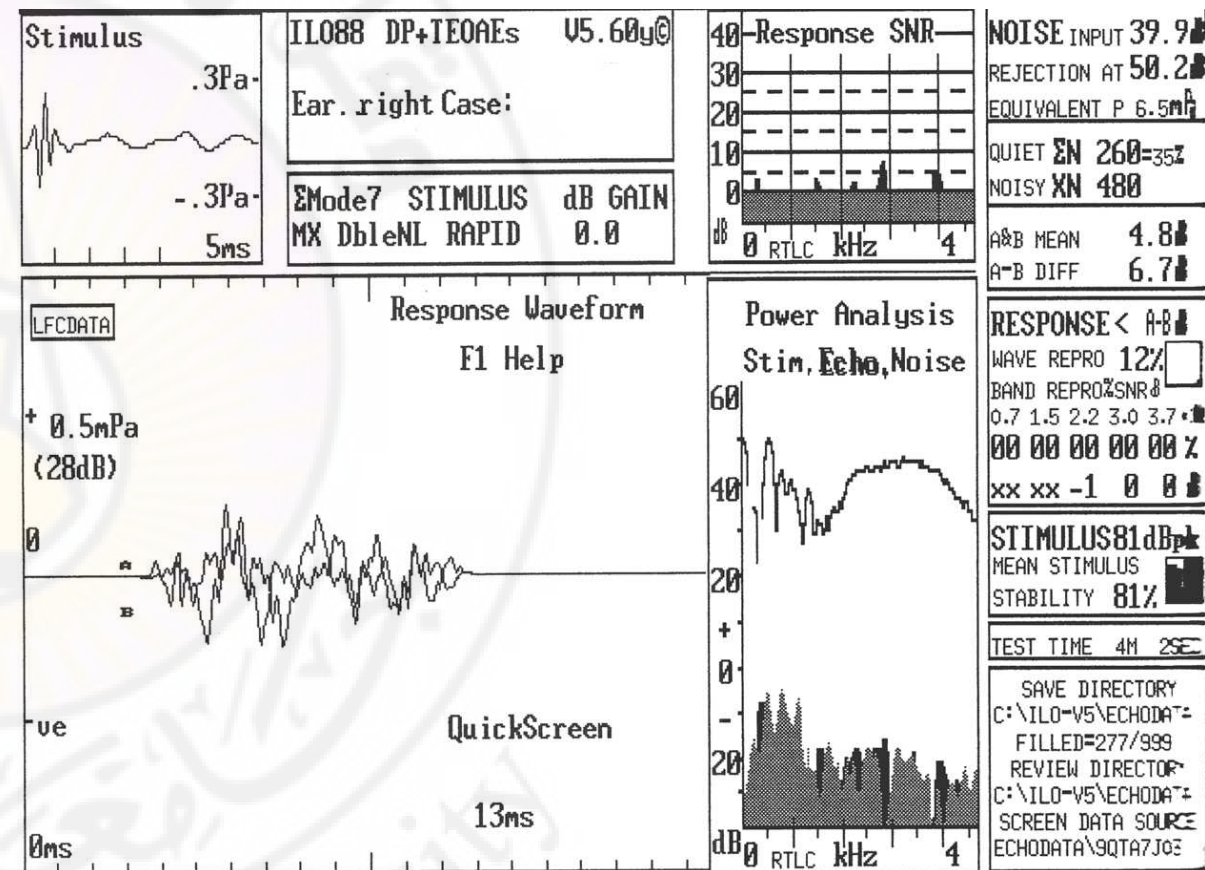
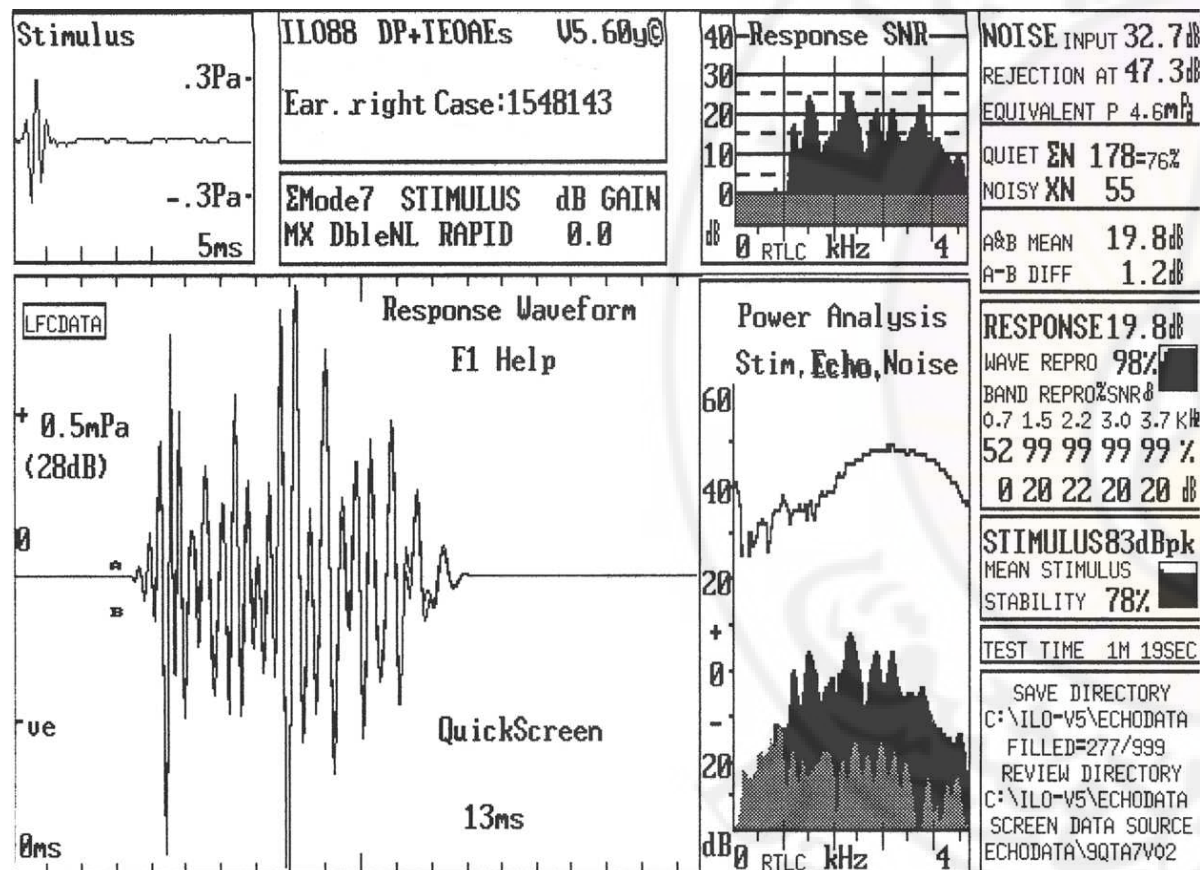
Expected DPOAE levels at various frequencies for term-born and 4.5-week-old infants compared with adults

Age	F2 Frequency (Hz)						
	1,500	2,000	3,000	4,500	6,000	8,000	9,000
Term	15.8 (5.8)	13.6 (5.5)	9.5 (5.9)	9.5 (6.4)	9.6 (5.8)	5.8 (6.8)	5.9 (6.6)
4.5 wks	15.8 (7.1)	14.2 (5.6)	10.5 (9.1)	13.3 (15)	15 (6.4)	9.7 (7.5)	—
Adult	9.9 (4.6)	6 (6)	5.6 (5.8)	5.9 (5.9)	5.1 (5)	-0.3 (6.5)	-4.1 (7.7)
Subject Characteristics							
	N		Sex (F, M)			Ear (L, R)	
Term	118		48, 70			70, 48	
4.5 wks	18		9, 9			7, 11	
Adult	48						

Data are presented from a group of 48 normal-hearing adults between 18 and 35 years of age for comparison.¹¹⁵ The number of subjects in each group is displayed in the bottom half of the table along with the sex and ear distribution.

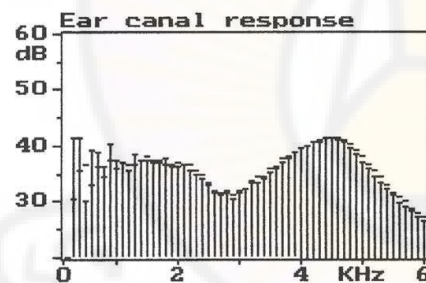
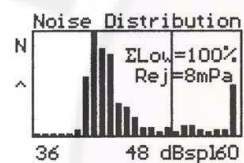
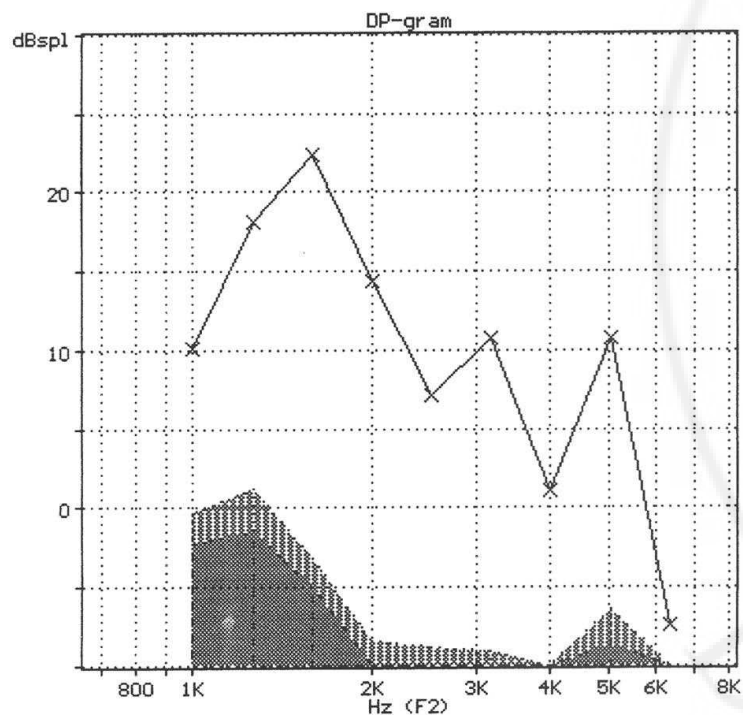
OAEs and Sensory Hearing Loss





F2:F1=1.221 F1=70.0dBspl F2=70.0dBspl Elapsed time =58secs

Recalled -> DP47L001.DPG



Patient record data

Name: dp471

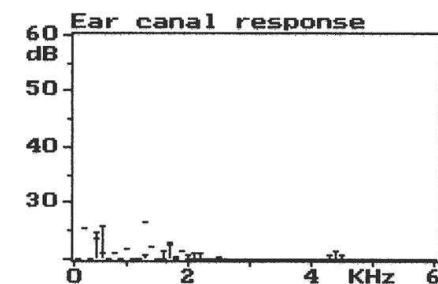
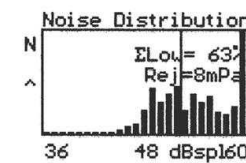
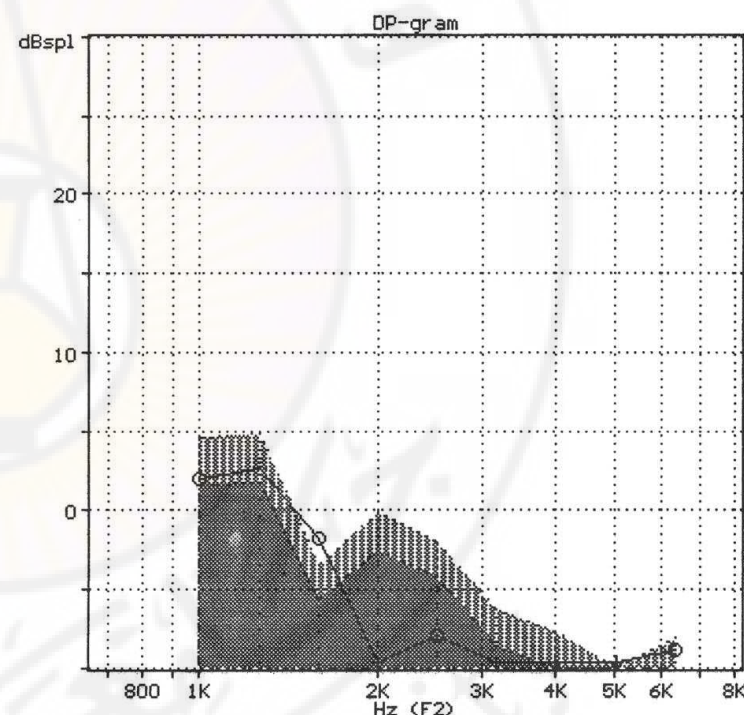
Ear: left

No.:

Note:

F2:F1=1.224 F1=70.0dBspl F2=70.0dBspl Elapsed time =90secs

Recalled -> ERICF004.DPG



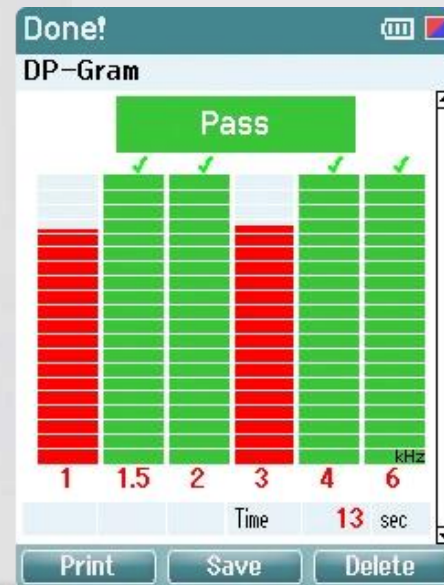
Patient record data

Name:

Ear: right

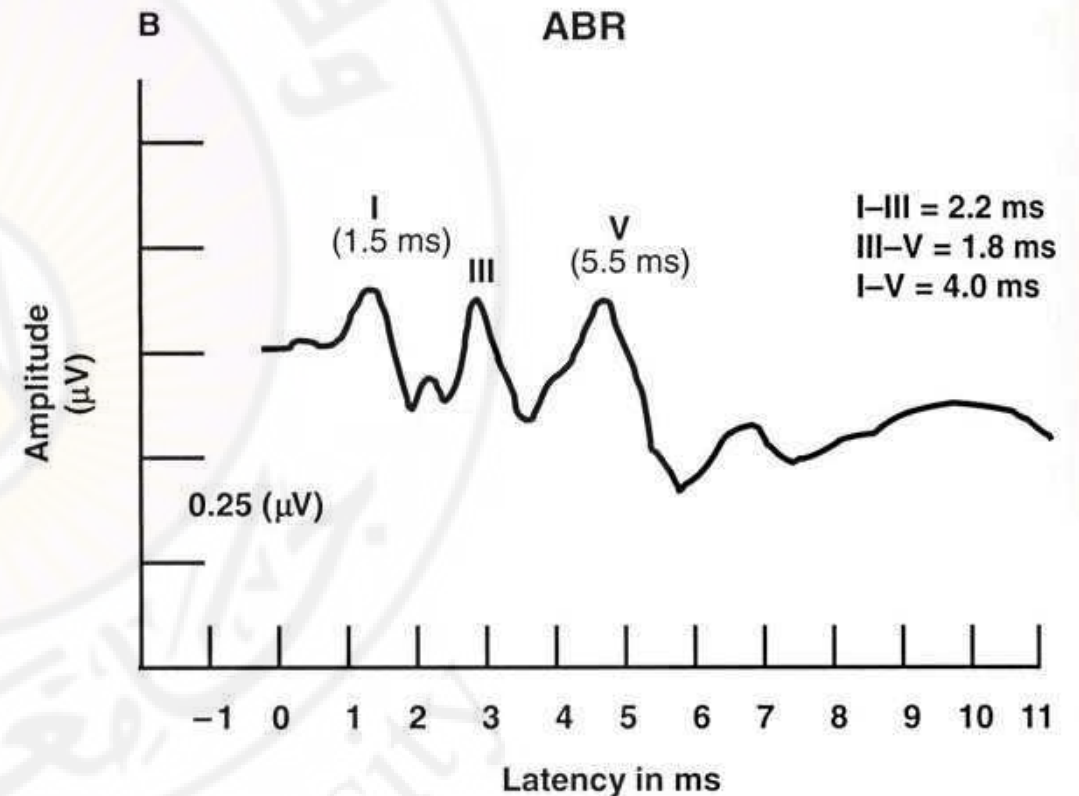
No.:

Note:



ABR

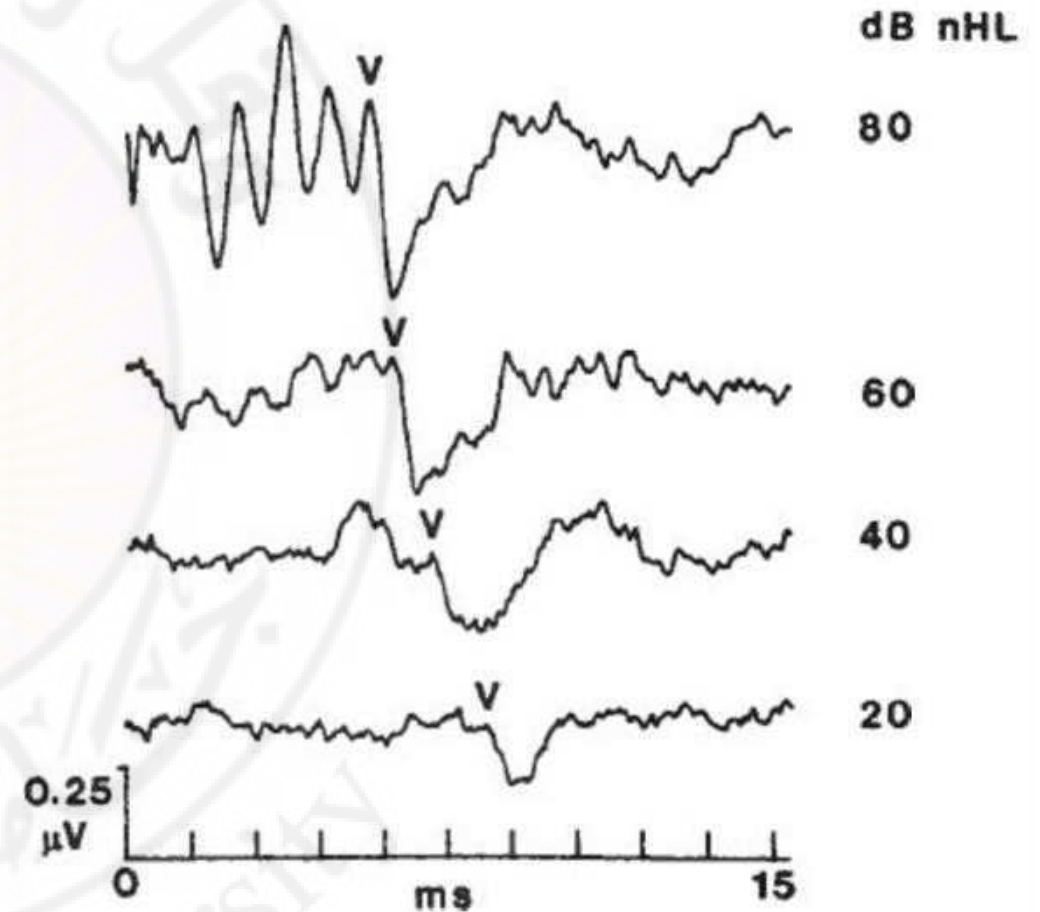
- The ABR includes a series of five to seven peaks arising from auditory nerve and brainstem structures, occurring within 10 ms of the onset of a moderate intensity click stimulus in otologically, audiologically, and neurologically intact adults.
- The ABR is, without question, the most clinically useful AEP at the present time.
- It can be used for estimating hearing threshold and differential diagnosis of peripheral and central abnormalities and for intraoperative monitoring.





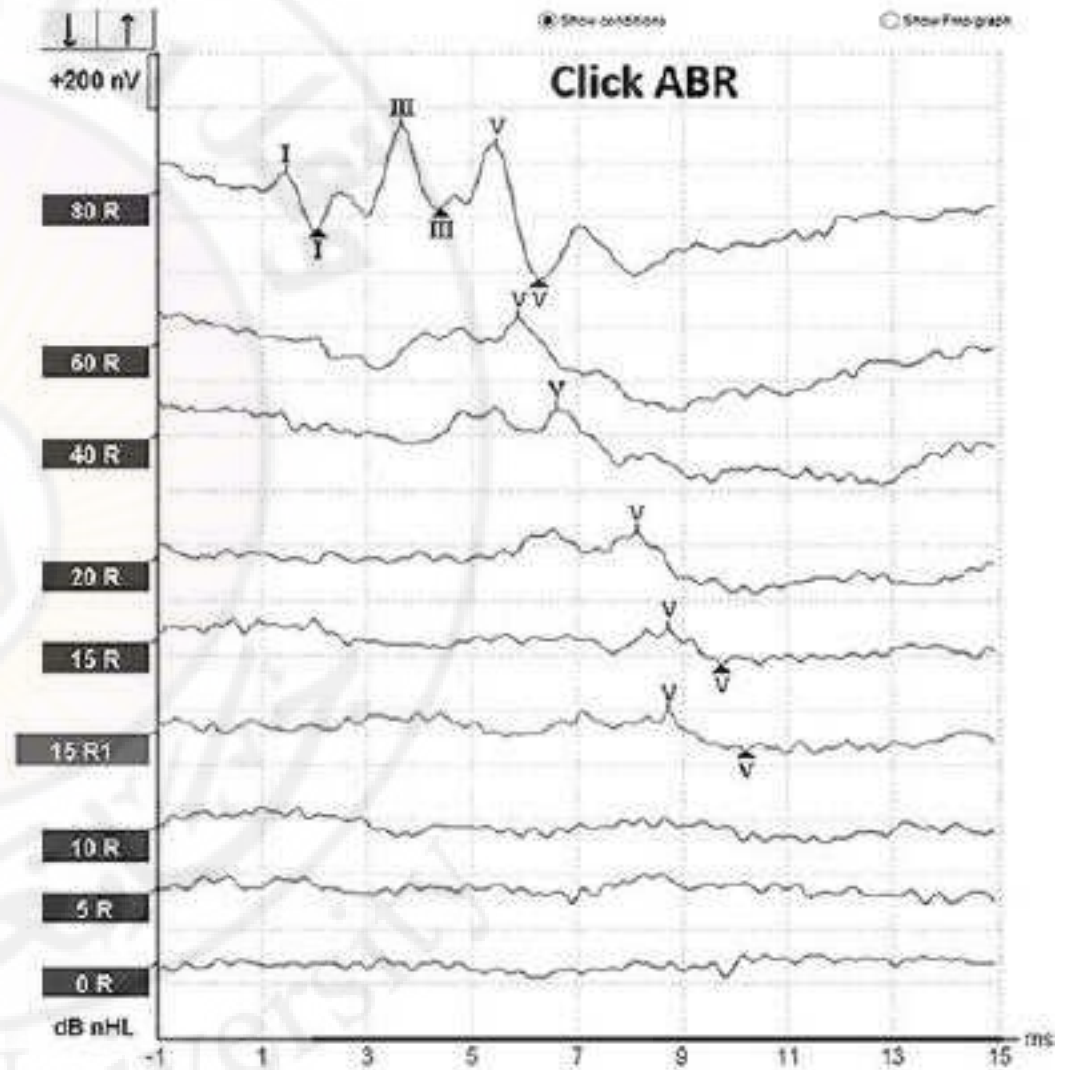
ABR is NOT a test of hearing!

- Although AEPs have proven useful in estimating hearing thresholds, it is important to remember that the ABR and other evoked potentials assess **neural synchrony**, that is, the ability of the peripheral and central nervous system to respond to external stimulation in a synchronous manner.
- For individuals who do display synchronous neural responses, we can find the lowest stimulus intensity level that yields the neural response and relate that to a threshold for hearing.
- **Limitation:** auditory neuropathy/dys-synchrony (ANSD)



ABR threshold estimation

- For assessing the hearing sensitivity, **wave V** of the ABR is used, because it's most robust of the waves and best correlated with behavioral thresholds.
- Wave V can be elicited at intensity levels within 5-20 dB of behavioral threshold for the click.
- If wave V is present at 20 dB nHL, this is considered normal and there is no need to test at lower intensities.



Record Edit... Latency

Latency times

	mS	μV
I		
II		
III		
IV		
V		

X		
Y		
Z		

I-III	
III-V	
I-V	

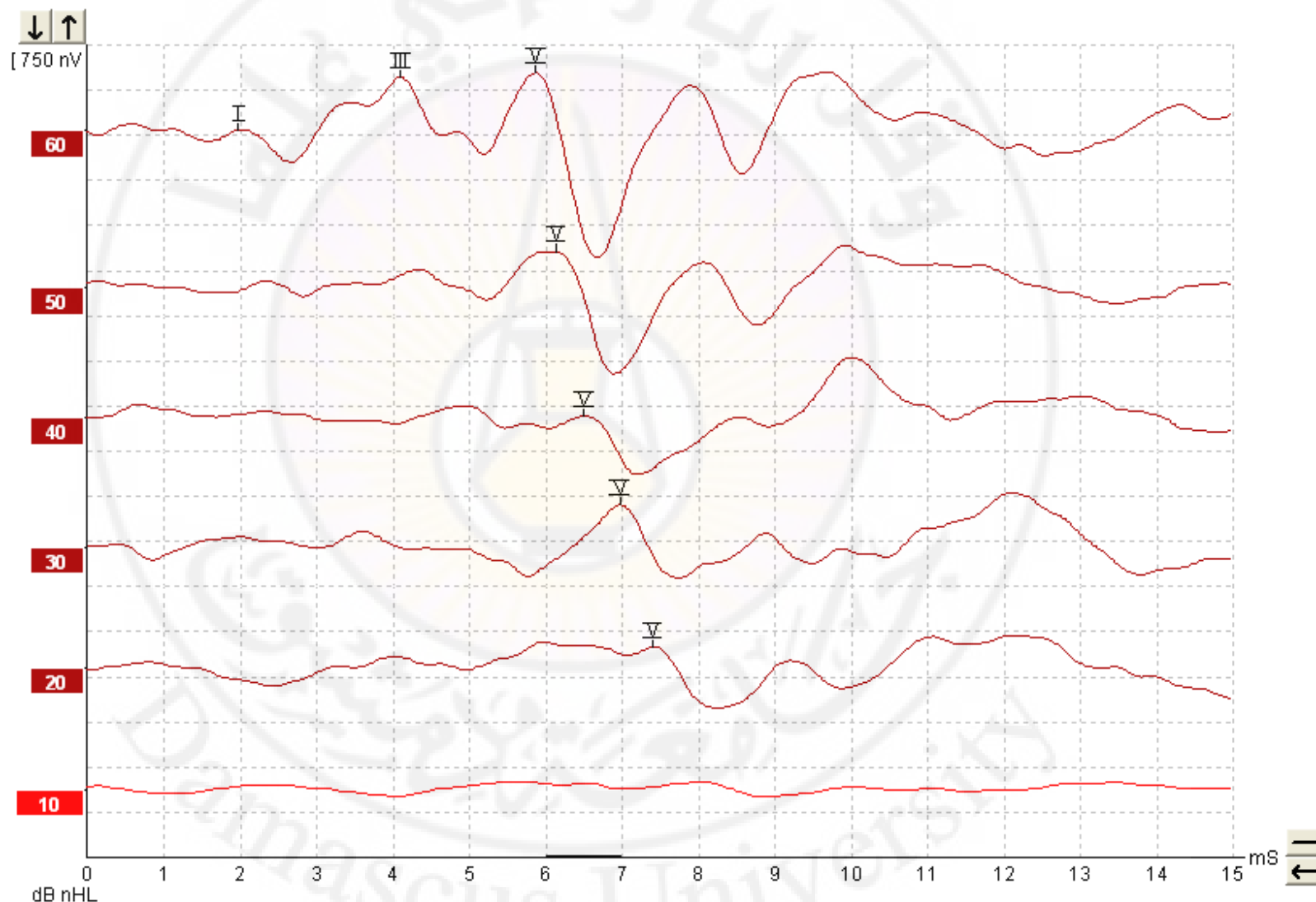
Cursor

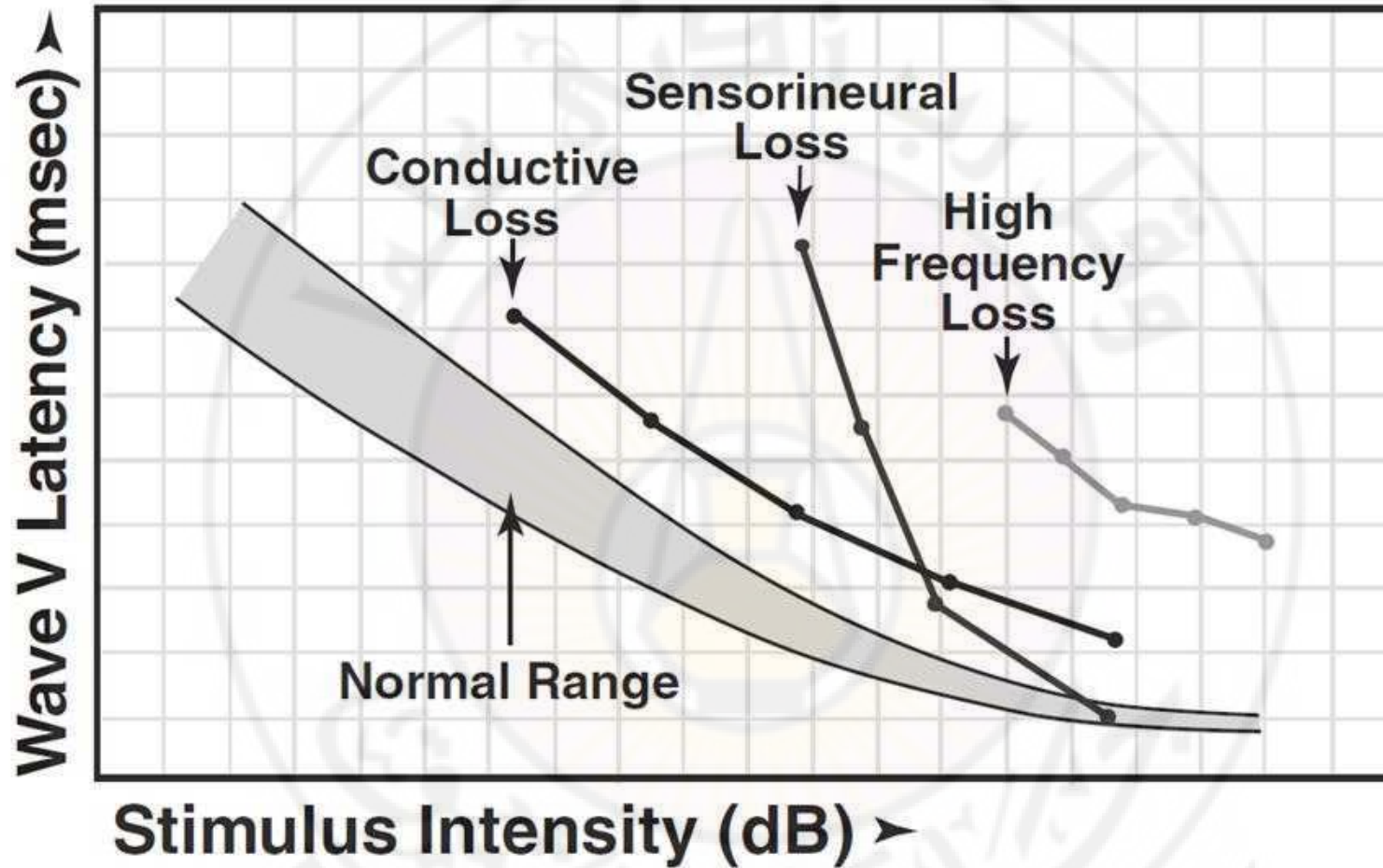
	Fixed	Cursor	Diff.
mS			
μV			

Filter setting

Low pass	High pass
2000 Hz	100 Hz

Measured	2000	Masking	Off	Wave repro.	10 %		
Rejected	1%	Stimuli/Sec	21.1	LP	2000 Hz	HP	100 Hz
Gain	92 dB	Polarity	Burst, 2.0K Rarefaction				

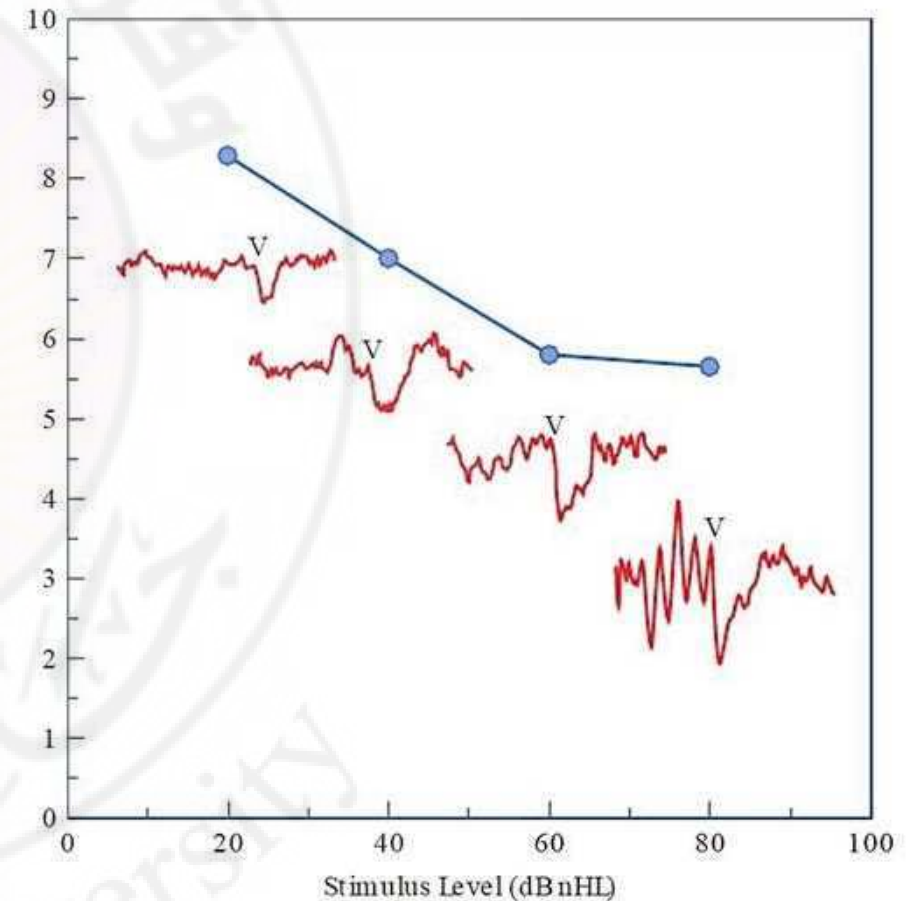




Characteristic latency-intensity functions obtained from normal-hearing patients and patients with various types of hearing loss.

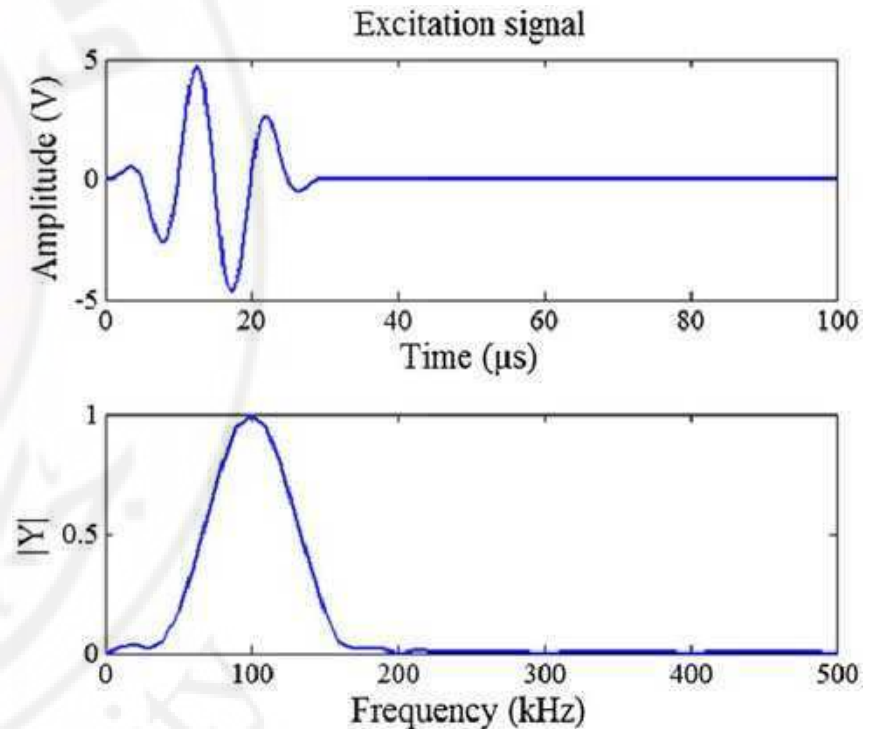
Click ABR agreement with puretone thresholds

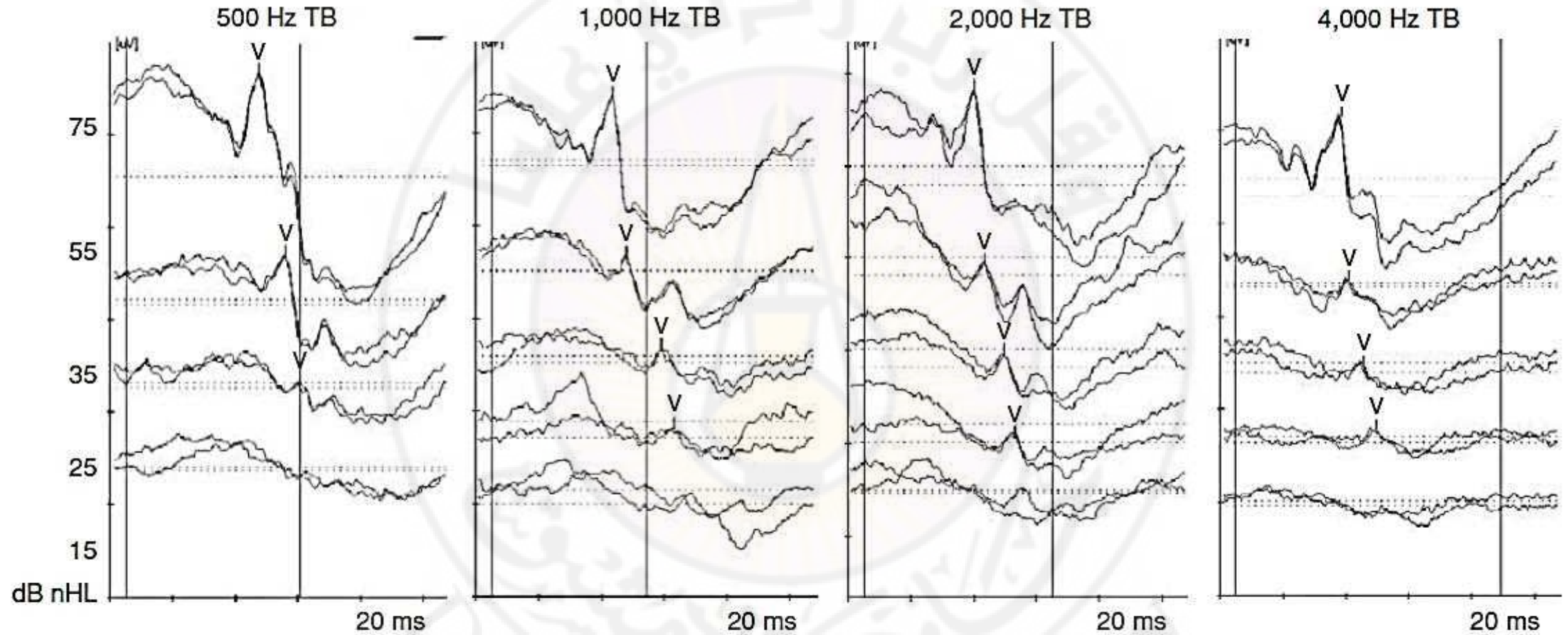
- A 100-microsecond electrical pulse, impressed on an earphone, generates a broadband signal whose primary frequency emphasis is determined by the resonant frequency of the transducer.
- With earphones typically used in clinical evaluation, the maximum energy peaks of these clicks are focused in the frequency region between 1,000 and 4,000 Hz.
- **The greatest agreement with puretone thresholds is in the 2,000- to 4,000-Hz frequency range.**



Frequency Specific Stimuli

- The overall objective for clinical application of ABR in infants is diagnosis and **frequency-specific estimation of hearing loss**, information that is essential for appropriate decisions regarding early intervention.
- Several types of stimuli and recording methods have been proposed to obtain responses from narrower frequency regions.
 - **Tone bursts or tone pips**
 - **Filtered clicks**
 - **Tone bursts in notched noise**
 - **High-pass masking of clicks or tone bursts**
- **Tone bursts** are the most popular option for obtaining frequency-specific responses.

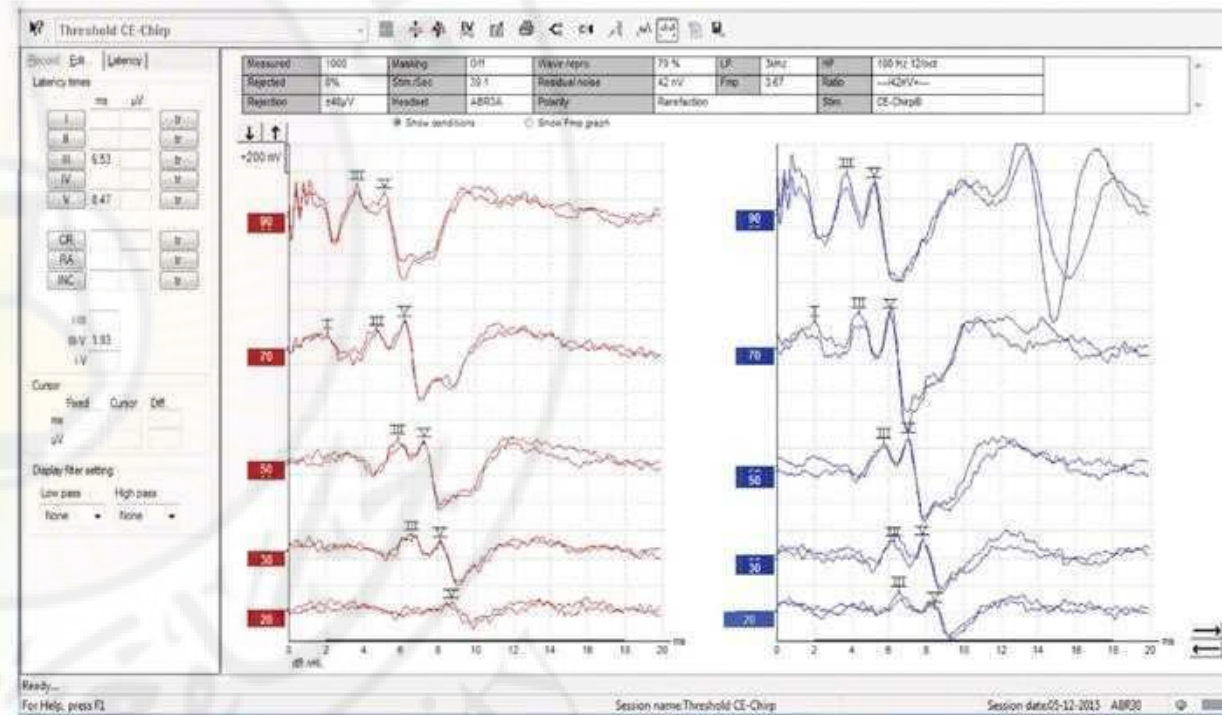
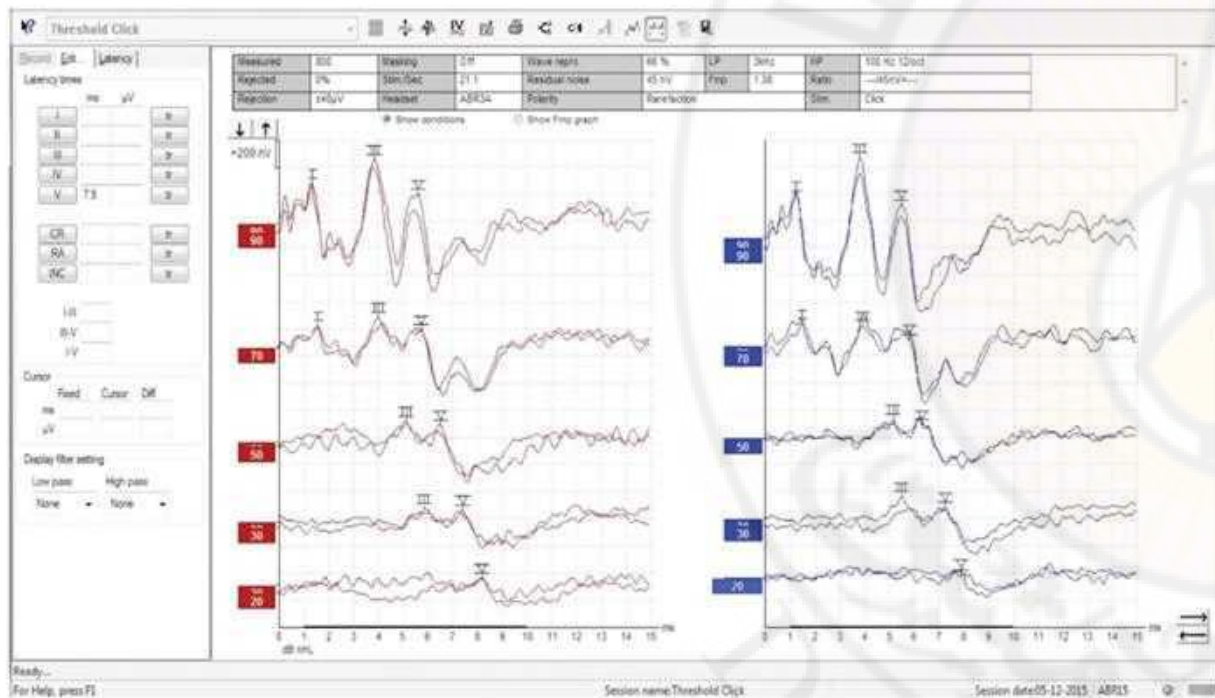




ABRs to tone bursts centered at 500, 1,000, 2,000, and 4,000 Hz recorded from an individual with normal hearing. The latencies of Wave V at 75 dB nHL are 8.53 ms for a 500-Hz tone burst, 7.70 ms for a 1,000-Hz tone burst, 7.03 ms for a 2,000-Hz tone burst, and 6.53 ms for a 4,000-Hz tone burst, demonstrating the latency decrease as center frequency of the tone burst increases.

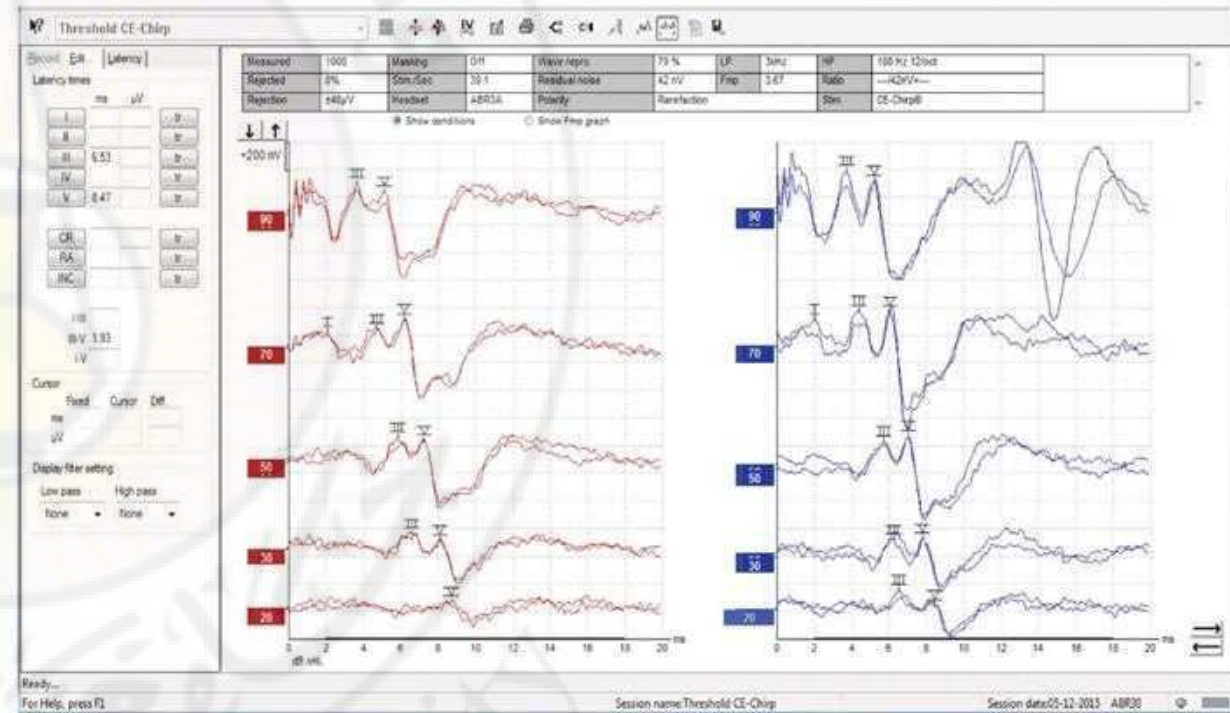
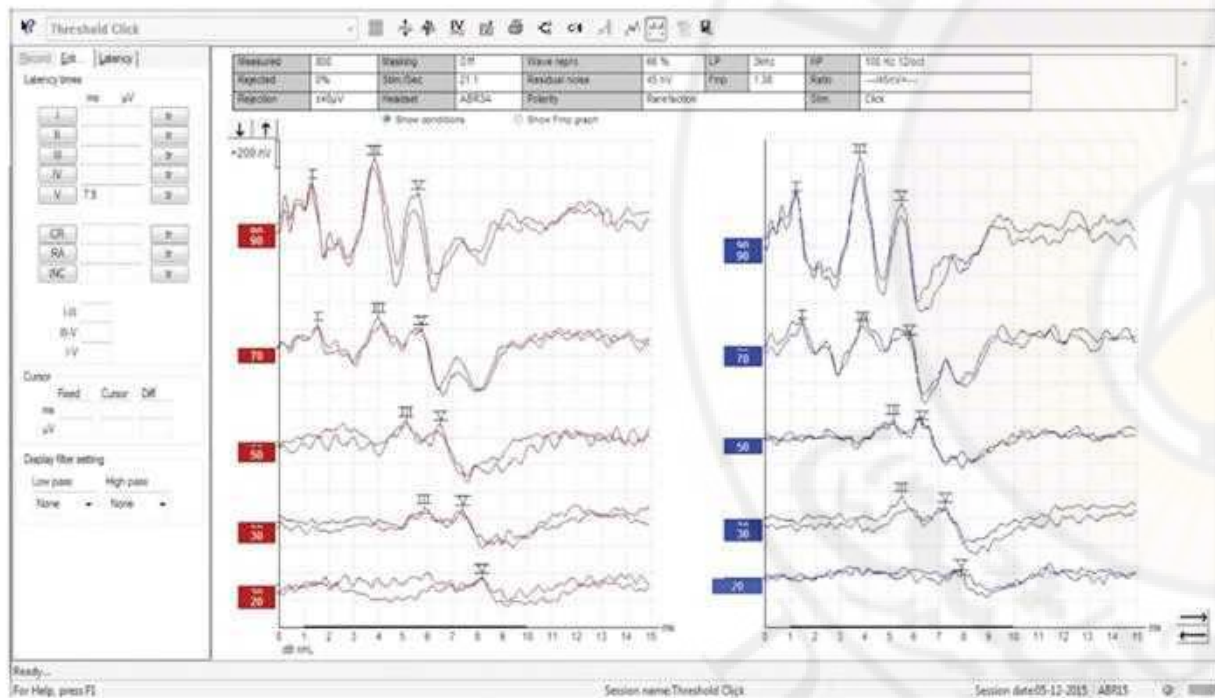
Click

Chirp



Click

Chirp



Estimating the Audiogram

- The ABR threshold, in dB nHL, is not the same as the estimated hearing threshold, in dB eHL.
- the average level at which an ABR is normally seen above the patient's audiogram is **frequency** dependent, with greater differences seen at lower frequencies because of poorer neural synchrony, and also dependent on the type of **stimuli** (Click, tone bursts, chirp), and age related **transducer** offset.

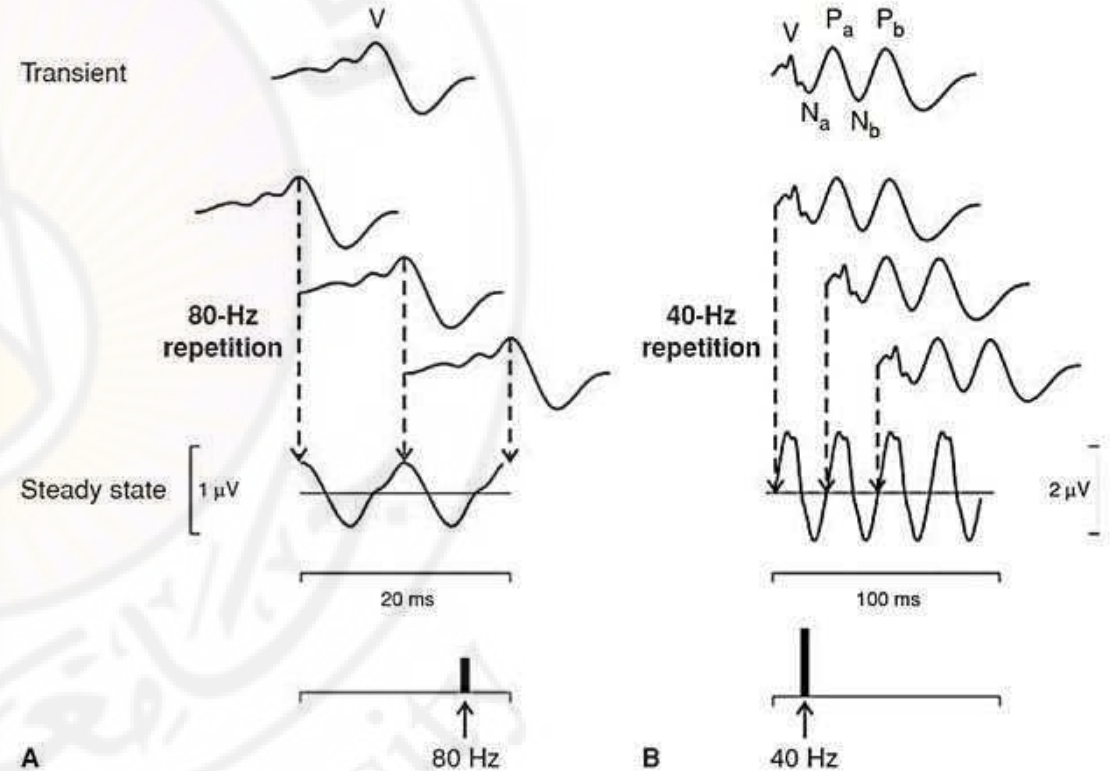
Corrections to be added to ABR thresholds in dB nHL to estimate the hearing level (dB eHL) in babies up to 12 weeks corrected age

Frequency (Hz)	Tone burst ABR				Chirp ABR			
	500	1,000	2,000	4,000	500	1,000	2,000	4,000
Insert phones	-15	-10	-5	0	-10	-5	0	5
Supra-aural earphones	-20	-15	-10	-10	-15	-10	-5	-5
Bone conductor	5	5	-5	0	10	10	0	5

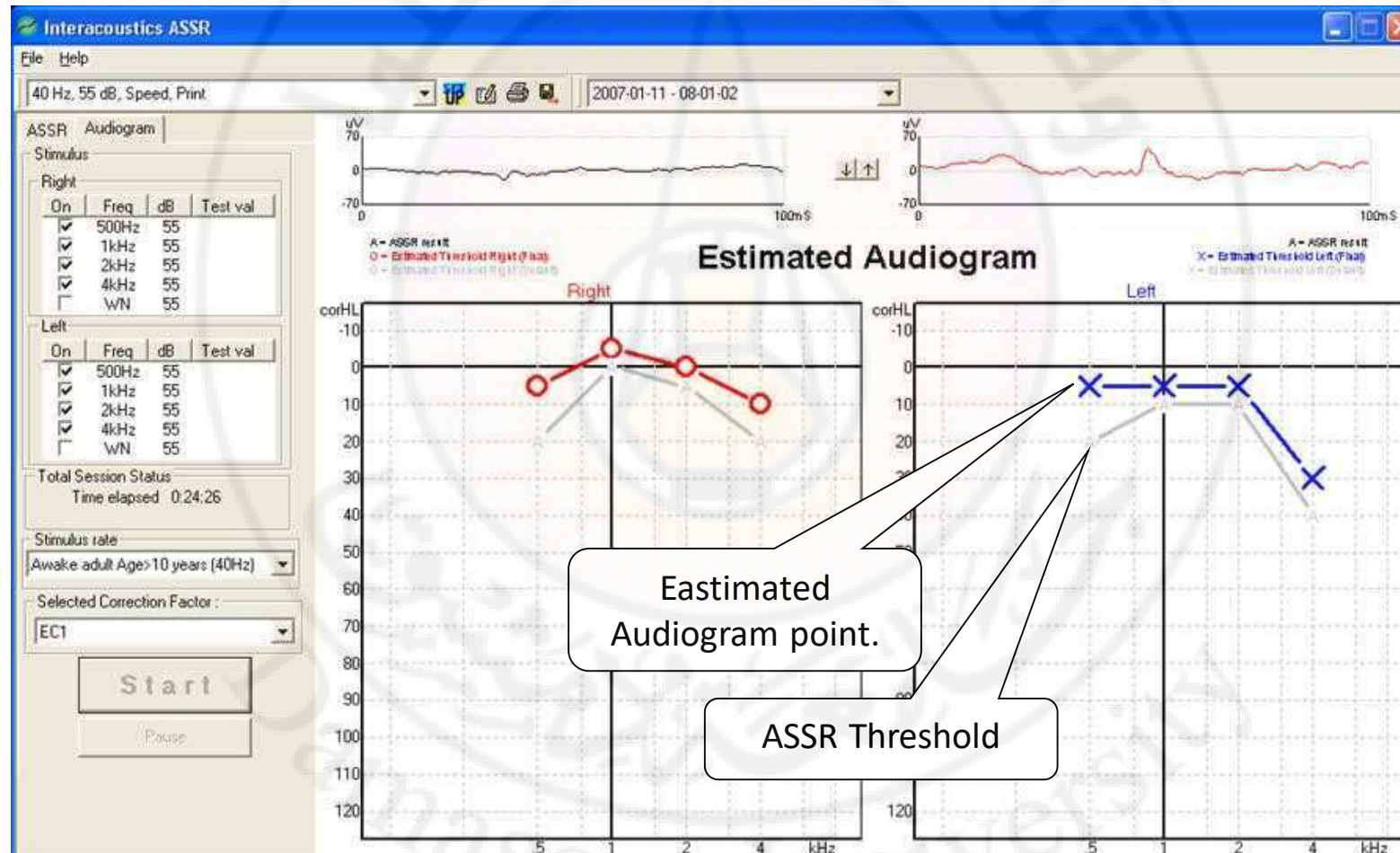
Corrections for older children and for clicks are available from the English NHSP guidance.⁷

Auditory steady-state response (ASSR)

- A relative newcomer to the family of AEP responses is the auditory steady-state response (ASSR).
- This response represents a sustained response and can be elicited by trains of clicks, noisebursts, or tonebursts, as well as amplitude modulated (AM) and/or frequency-modulated (FM) stimuli.



An Estimated Audiogram is Automatically Provided



Thank you!

