



*The Audiology Diagnosis  
Process: A look at  
monitoring hearing loss, the  
process of diagnosis and  
sharing results*

*Early Years*

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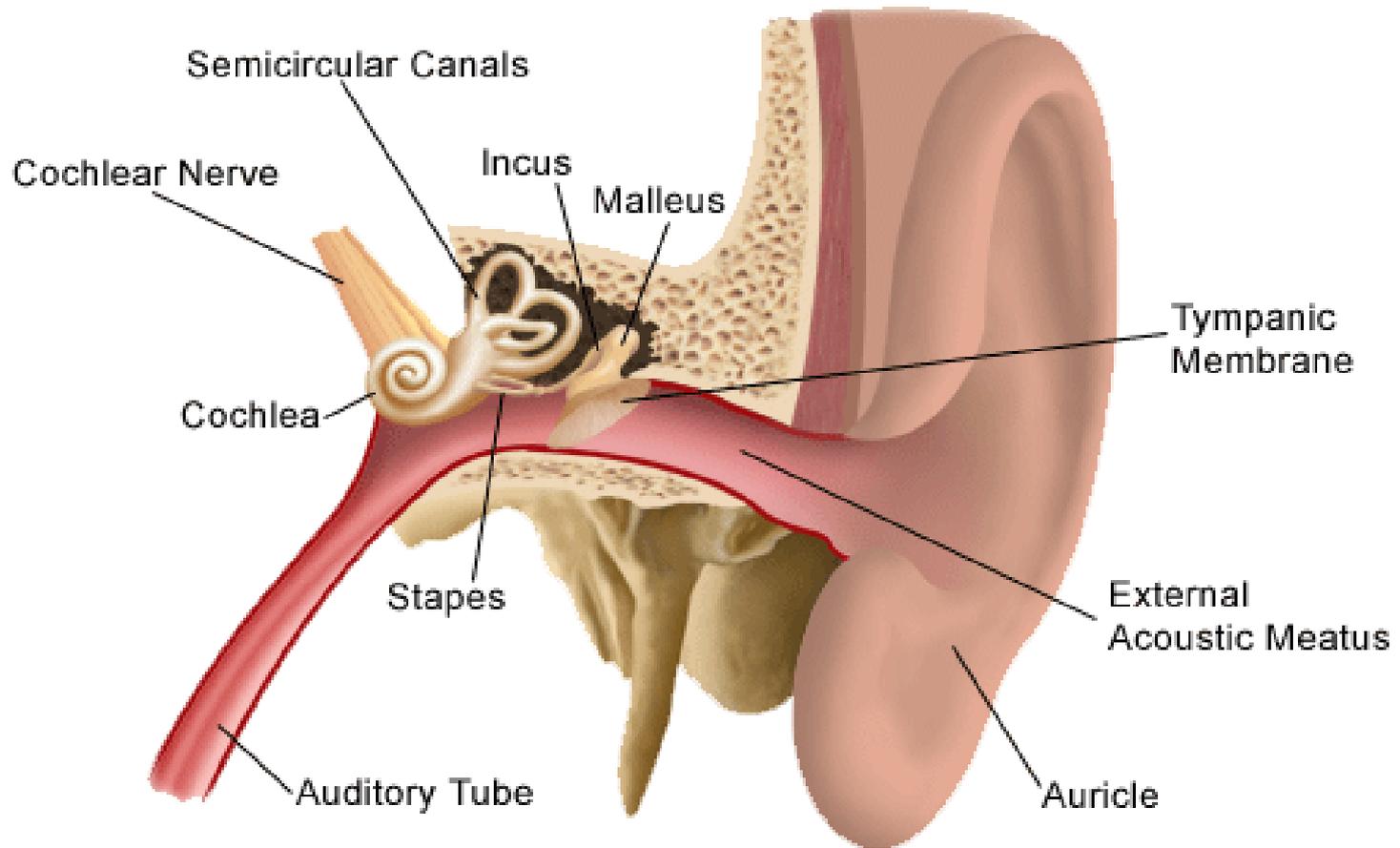
# Who am I?

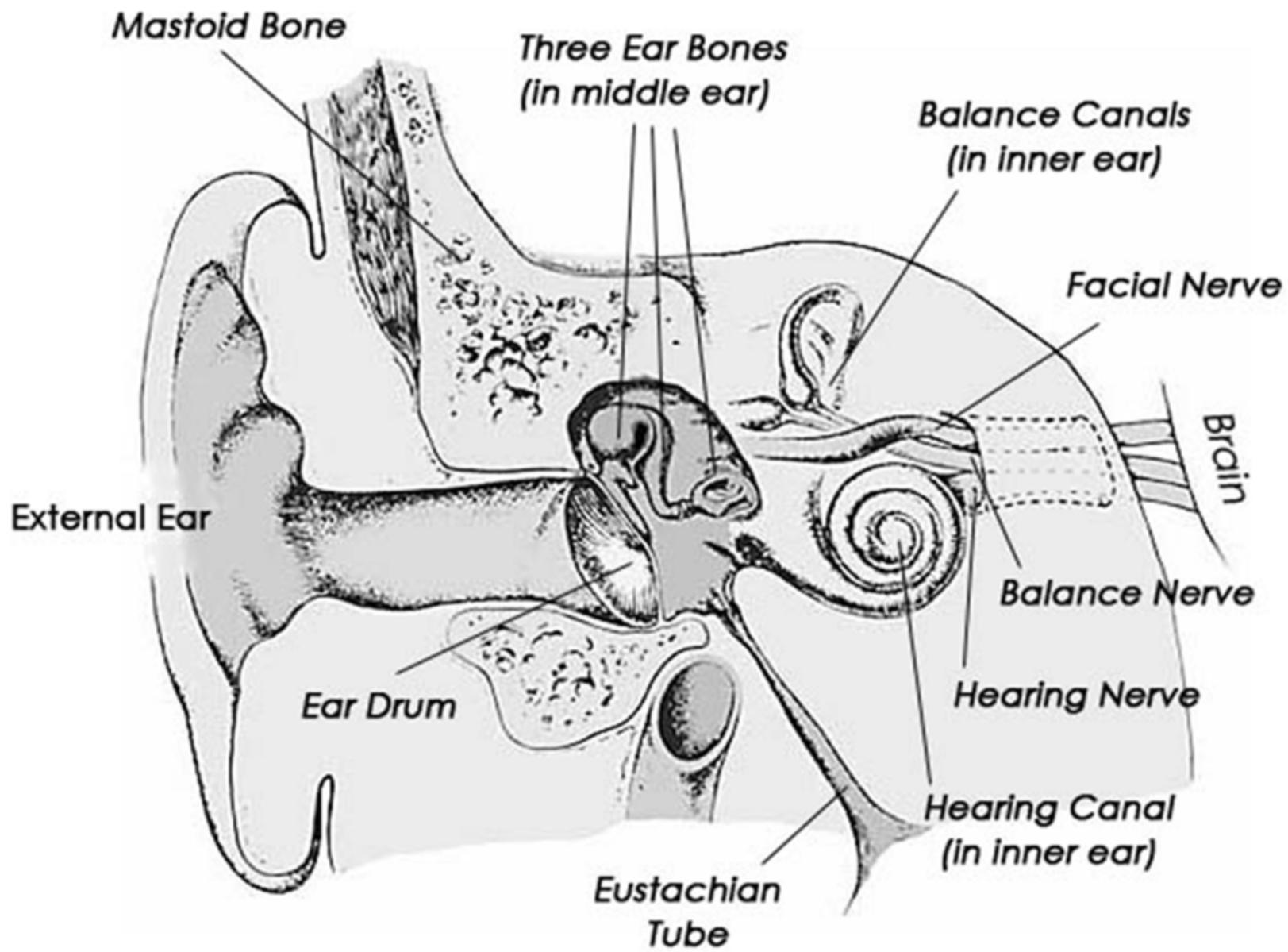
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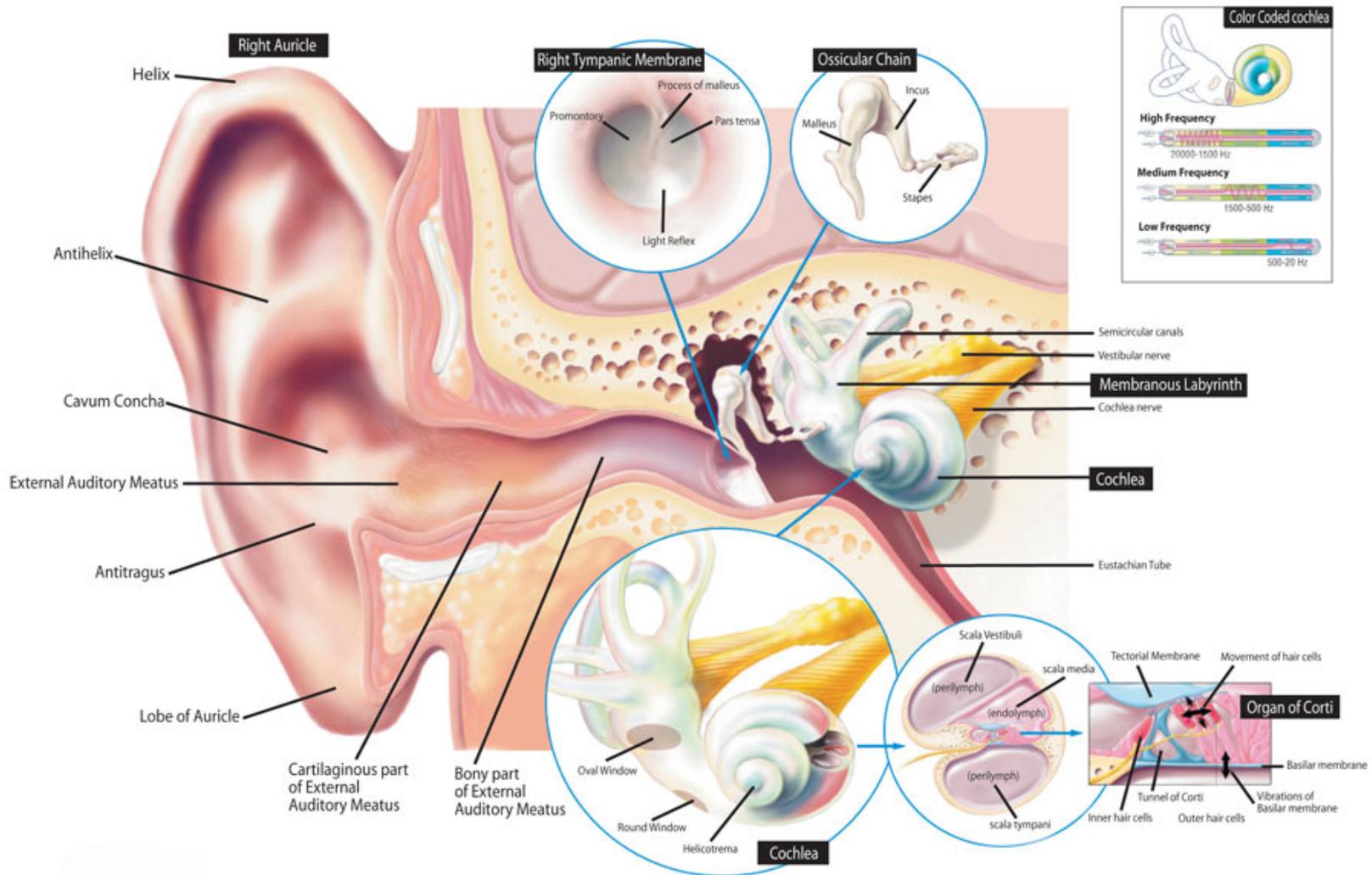
# Objectives

- After this presentation you will be able to:
  - Communicate how we hear to people that are not specialists in hearing
  - Recognize the variety of testing techniques used in diagnosing and managing hearing loss
  - Understand the process for diagnosing/managing a hearing loss
  - Recognize caregiver/parental concern of hearing loss
  - Recognize when it is time to refer or encourage follow up to a Pediatric Audiologist
  - Identify some of the high(er) risk indicators for late onset/progressive hearing loss

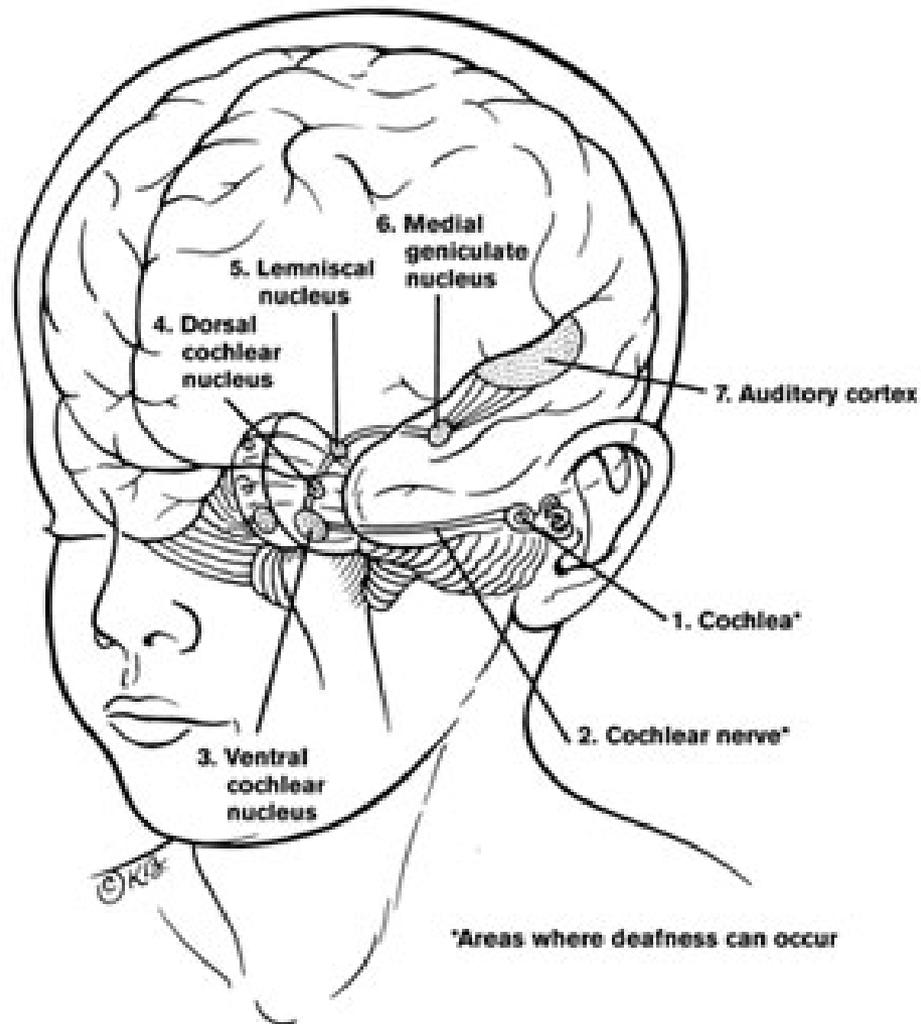
# Ear Anatomy







# How We Hear



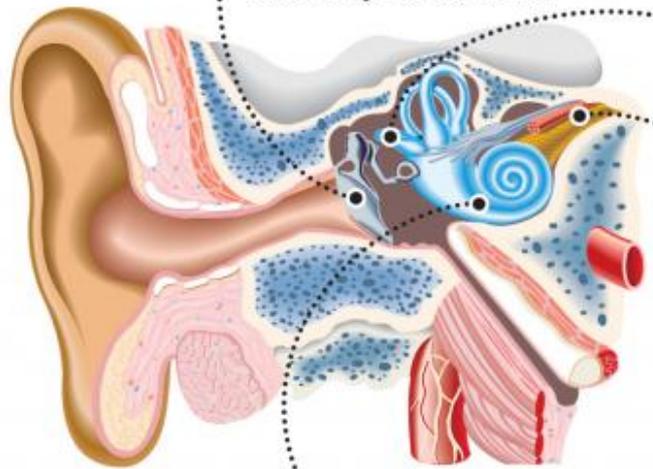
# How We Hear

1.

Sound waves enter your outer ear and travel through the ear canal to your eardrum.

2.

Your eardrum vibrates with the incoming sound and sends the vibrations to three tiny bones in your middle ear.



3.

The bones in your middle ear amplify the sound vibrations and send them to your inner ear, or cochlea. The sound vibrations activate tiny hair cells in the inner ear, which in turn release neurochemical messengers.

4.

Your auditory nerve carries this electrical signal to the brain, which translates it into a sound you can understand.

# Pediatric Audiology

- Multiple objective test measures are used to avoid misdiagnosis and identify site of lesion. One test of hearing is insufficient to define a hearing loss accurately.
- “Cross-check principle” – The results of a single test are cross-checked by an independent test measure. Particularly useful in pediatric behavioral evaluations

# Pediatric Audiology

- Observing auditory behaviors of children does not always yield an accurate description of hearing loss



# Pediatric Audiology

- **Electrophysiologic** screening for hearing loss can be completed within hours of birth (Using screening OAE or AABR)
- **Electrophysiologic** diagnostic tests confirm and define a hearing loss for children too young or uncooperative to obtain behavioral results



# Objective Audiology Tests

Auditory Brainstem Response (ABR)	<ul style="list-style-type: none"><li>• Measures the time it takes for the sound to travel to the lower brainstem. Used for neurodiagnostic evaluation. Also used to obtain ear specific thresholds to specific frequencies or sounds. Requires the child to be sleeping. Not a test of hearing sensitivity, but of auditory function of the cochlea, auditory nerve and lower brainstem.</li></ul>
Otoacoustic Emissions (OAE)	<ul style="list-style-type: none"><li>• Evaluates the outer hair cell function within the cochlear. Not a test of hearing sensitivity, but of cochlear function.</li></ul>
Tympanometry	<ul style="list-style-type: none"><li>• Measuring the child's eardrum movement and middle ear function. Not a test of hearing sensitivity, but of mechanical function of the middle ear space.</li></ul>
Acoustic Reflexes	<ul style="list-style-type: none"><li>• Measuring the child's middle ear muscle reflex to loud sounds. Not a test of hearing sensitivity but of the middle ear space and neural response.</li></ul>

# Pediatric Audiology

- Accurate **behavioral** tests of hearing can begin as young as 4-6 months of age.



# Behavioral Audiology Tests

## Visual Reinforcement Audiometry (VRA)

A test of hearing sensitivity using behavioral responses to sound which are reinforced visually. Test is able to define ear specific thresholds to speech and different frequencies. VRA can begin once a child is old enough to sit unsupported. The technique capitalizes on the natural head turn response young children have when sound is present in their environment. Occasionally, auditory awareness, or the onset response to stimuli needs some training. This may be completed over the course of 2 or 3 visits, or at home with coaching.



# CPA

## Conditioned Play Audiometry (CPA)

Traditional method of obtaining responses to sound using trained play techniques. This method of testing is used for older children, around age 2 ½ -5 or 6. Games and forms of play are used to condition a child to respond to sound. Ear specific threshold information to speech and frequencies can be obtained with this technique.



# What can interfere with test results

- Occlusion of the outer ear canal. Typically this is cerumen, or in infants a few hours old this may be vernix.
- Otoscopy is used to rule out this cause. In the event of cerumen impaction, testing may still be completed to determine the impact of the occlusion on hearing.



# What can interfere with test results



- Middle ear pathology. This could be fluid in the middle ear space, an ear infection, a perforation of the ear drum, malformation, etc.
- Testing is typically still completed to evaluate the impact on hearing, this is important as this information guides the physician and/or surgeon in treatment options.

# The state of the child

- The compliance or cooperativeness of the child plays heavily into the audiologists ability to obtain all the information needed to fully define hearing.



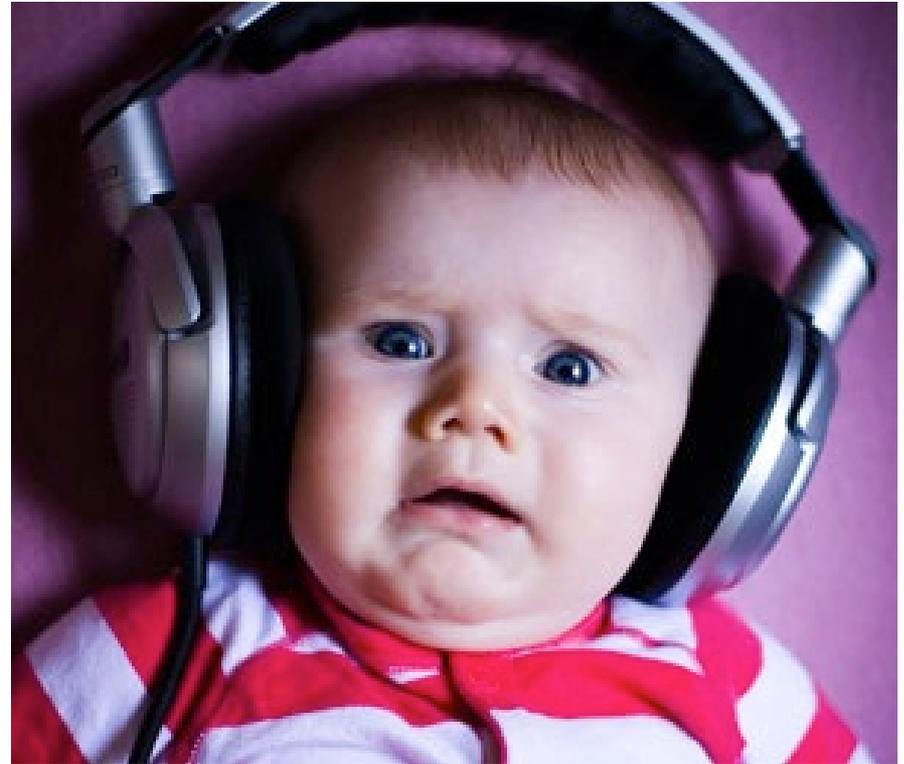
# State of the child

- In cases of electrophysiologic tests, such as ABR, if the child is too alert the EEG activity may be too loud to identify the brainstem response to soft inputs.



# The state of the child

- For behavioral measures, if a child is having a bad day, is tired or scared, testing may not be completed in one visit.



# Hearing Loss

- Types of Hearing Loss

- Conductive
- Sensory
- Mixed (conductive & sensory)
- Neural
- Auditory Neuropathy/Auditory Dysynchrony (AN/AD)
- Congenital/Acquired
- Stable/Progressive

- Degrees of Pediatric Hearing Loss

- Normal hearing (0 to 15 dB HL)
- Minimal/Slight hearing loss (16 to 25 dBHL)
- Mild hearing loss (26 to 40 dB HL)
- Moderate hearing loss (41 to 55 dB HL)
- Moderately-Severe ( 56 to 69 dB HL)
- Severe hearing loss (71 to 90 dB HL)
- Profound hearing loss (greater than 91 dB HL)

- Studies completed in several areas of healthcare looked at how much patients remember of medical/health information given by their physician/healthcare provider.

- (Shapiro et al 1992, Kessels 2003, Geraldo et al 2011, Martin et al 1990, Margolis, 2004)

# What Parents Remember

- 40-80% of medical information provided by their healthcare provider is forgotten immediately following diagnosis.
- About 50% of what is remembered is remembered inaccurately.



# What about the audiogram?

- The audiogram is used by the audiologist to define hearing for other audiologists and other health care professionals.
- The audiogram is a poor tool to use in counseling or explaining hearing loss impact to a family.
- In today's interactions with families, the audiogram is often down played in the explanation of the hearing loss.

# If not the audiogram, then what?

- Audiologists describe hearing loss in children in terms of impact on speech development, ability to hear from distances, and how the specific type of hearing loss, the degree or configuration may impact auditory behavior.
- The audiologist will describe the loss in terms of permanence, fluctuating or temporary and whether it is something that can be medically corrected or would benefit from amplification.

# Informational Counseling



# Interactive Informational Counseling

- Internet tools and resources
- Ear Models and pictures
- Printed materials
- Connections to family support
- Referrals to a network of professionals

# Monitoring hearing

- Diagnosing hearing loss is not an event, it is a process.



# Monitoring Hearing Loss

- Monitoring infants and children with risk indicators for progressive or late onset hearing loss the process will look different depending on the child and the condition being monitored.



# Monitoring Hearing

- Young children with poorly defined hearing or poorly defined conditions may require more frequent monitoring and evaluation early on.
- Monitoring schedules are determined by the managing audiologist in coordination with the family and medical home.

# History of NHS and High Risk

- 1972 JCIH Position Statement established a first attempt at a Newborn Hearing Screening Guideline
  - Recommendation that all infants meeting the high risk criteria be referred for a “hearing evaluation” (behavioral/observational)
- 1982 added some additional High Risk items
  - Also established a guideline to screen by 3 months of age (later than 6 months)
  - Included language to encourage supervision of audiologist
  - Recommended use of behavioral or electrophysiologic evaluation tools

# History of NHS and High Risk

- 1990 JCIH Statement added more high risk criteria and moved to ABR measurement by 6 months of age and eliminated recommendation of observation as a method of evaluating hearing in young infants
- 1994 JCIH Statement pointed out some flaws in using just the high risk register to screen infants as only about 50% of infants with hearing loss have any of the high risks listed (meaning they are born in well baby nursery without any obvious risk concerns for hearing loss)
  - Recommended consideration of Universal Screening

# History of NHS and High Risk

- 2000 JCIH Position Statement recommended Universal Newborn Screening of hearing before discharge
  - Provided guidelines for infants that passed newborn screening but had identified high risk indicators for late onset or progressive hearing loss
    - Every 6 months until age 3 (for ANY risk indicator)

# 2007 JCIH Guidelines

- Expanded definition of targeted hearing loss to include Neural Hearing Loss
  - Infants at risk for Neural loss (ex. NICU) to have AABR not OAE
- Changed language to be less restrictive for monitoring infants that pass NHS but have high risk indicators
  - At least one diagnostic hearing evaluation by a pediatric audiologist by 24-30 months of age.

# High Risk Indicators for Late onset or Progressive Hearing Loss

- High Risk Indicators that may require more frequent monitoring:
  - In-utero infections (CMV, herpes, rubella, toxoplasmosis, syphilis)
  - Culture-positive postnatal infection (meningitis)
  - Syndrome or physical findings associated with a syndrome known to have hearing loss (Down syndrome, Wardenburg's, etc)
  - Craniofacial or temporal bone anomalies
  - Head trauma
  - Hyperbilirubinemia requiring exchange transfusion
  - ECMO assisted ventilation

# High Risk Indicators for Late onset or Progressive Hearing Loss

- Risk indicators for hearing loss that may require less frequent monitoring:
  - Family history of permanent childhood hearing loss
  - NICU stay greater than 5 days
  - Any amount of ototoxic exposure (gentamycin, diuretics, chemotherapeutics)
  - Mechanical ventilation
  - Caregiver concern

# Management of a diagnosed hearing loss

- Management of a diagnosed hearing loss will be specific to the needs of the child and family.
- Family and child centered care are the basis of management decisions
- Auditory habilitation is a critical component of management for families and is a standard recommendation.
- Other Referrals: ITP, ENT, Genetic Counseling/Evaluation, Ophthalmology

# Supporting Early Intervention

- Encourage families that missed the hearing screening, failed the screening or has any high risk for late onset or progressive hearing loss to follow up with a pediatric audiologist
- Encourage families to discuss their communication wants and needs with their audiologist, speech pathologists and other early interventionists.

# Improvement Initiatives

- Improve communication with other professionals.
- Identify existing gaps and work cooperatively to overcome gaps to improve service delivery to families.



# Questions??



# Case #1- Intake history

- Newborn hearing screening- passed in both ears
- High risk factors for late-onset hearing loss: ototoxicity and extended NICU stay
- No family history of hearing loss
- No parental concerns regarding hearing

# Case #1- Assessments

## 9 months old

- Abnormal tympanograms (Type B)
- Abnormal OAEs
- Speech responses:
  - 40 dBHL right ear
  - 35 dBHL left ear
- VRA responses (500-2kHz):
  - 30 to 55 dBHL in both ears
- Dx: Abnormal middle ear function, mild to moderate hearing loss in both ears
- Rec: Medical FU, Reevaluation in 6 weeks

## 11 months old

- Abnormal tympanograms (Type C)
- Absent acoustic reflexes
- VRA responses:
  - Bone conduction 10 dBHL for 500 and 4000 Hz
- Speech responses:
  - 40 dBHL in sound field
- Dx: Abnormal middle ear function, mild hearing loss with normal bone conduction results in at least one ear
- Rec: Medical FU, Reevaluation in 6 weeks.

# Case #1- Assessments

## 1 year old

- Normal tympanograms (Type A)
- Normal OAEs
- Present acoustic reflexes
- Speech responses:
  - 10 dBHL both ears
- VRA responses (500-2kHz):
  - 10-20 dBHL in both ears
  
- Dx: Normal middle ear function and normal hearing sensitivity in both ears
- Rec: Parental and physician monitoring of auditory development

## Case #2- Intake history

- Newborn hearing screening- referred in both ears
- Extended NICU stay with ototoxic medications, mechanical ventilation
- Med Dx: spina bifida, hydrocephaly
- No family history of hearing loss
- No parental concerns regarding hearing

# Case #2- Assessments

## 1 month old

- Abnormal ABR results
  - Mild to severe, bilaterally
- Normal OAEs
- Normal tympanograms
  
- Dx: Auditory Neuropathy/Neural Hearing loss
- Rec: Monitoring every 3 months

## 4 months old

- Abnormal ABR results
  - Slight to mild, bilaterally
- Reduced OAEs
- Abnormal tympanograms
  
- Dx: bilateral middle ear dysfunction, improvements in ABR suggest neural maturation
- Rec: Monitoring every 3 months

# Case #2- Assessments

## 8 months old

- Normal ABR, bilaterally
- Normal OAEs
- Normal tympanograms
- Normal acoustic reflexes
  
- Dx: Normal middle ear function, improvement in ABR continue to suggest neural maturation
- Rec: Monitoring every 3 months

## 1 year old

- Normal OAEs
- Abnormal tympanograms
- Normal speech response: 5 dBHL
- Essentially normal pure tone responses: 20-30 dBHL
  
- Dx: bilateral middle ear dysfunction, mild hearing loss rising to normal hearing sensitivity
- Rec: Monitoring every 3 months

## Case #3 – Intake History

- Newborn Hearing Screening – referred bilaterally
- Full term birth, delivery complications resulting in emergency C-section
- NICU stay for ~35 days
- Mechanical Ventilation and Ototoxic medication administration
- Medical Dx: Congenital Chylothorax; Noonan Syndrome; Plagiocephaly; Torticollis
- No family hx of hearing loss

# Case #3 – Assessments

- **1 month old**
- Abnormal ABR results in St. Al's NICU
- Dx: Severe sensorineural hearing loss left and profound sensorineural hearing loss right
- Rec:
  - Otologic Consultation
  - ABR re-evaluation prior to NICU discharge
  - Ongoing audiological monitoring
  - Binaural amplification following medical clearance
  - Referral for cochlear implant evaluation
  - Genetic counseling
- **2 months old**
- Fit with binaural amplification
- **3 months old**
- Abnormal ABR, Type A tympanograms, Absent Otoacoustic Emissions
- Dx: Severe SNHL left and profound SNHL right
- Rec:
  - Continued binaural amplification trial
  - Cochlear Implant Consultation
- **5 months old**
- Tympanometry abnormal left ear, normal right ear
- Aided Audiometry in Soundfield attempted; no reliable results obtained
- Dx: Abnormal middle ear function left ear
- Rec:
  - Medical follow-up
  - Aided re-evaluation after medical management

# Case #3 Assessments

- **7 months old**
- Cochlear Implant Consultation
- Rec:
  - Continue with binaural amplification use
  - Speech and language therapy
  - Evaluation with Dr. Pickford, CI team psychologist
- **1 Year old**
- Cochlear Implant Surgery: Right Ear, Cochlear Implant Activation 1 month later
- **1 month post-activation**
- Parent Report:
  - Beginning to respond to sound, especially high frequency
  - Likes noise toys, looks up when name called
  - No observed change in vocalizations
- Aided testing: Responds to voice at 50 dBHL and frequency specific stimuli at 30-50 dBHL, inconsistent responses.
- Minimal Response Levels do not equal auditory thresholds

# Case #3 Assessments

- **3 months post-activation**
- Parent Report
  - Responds to name and turns when called
  - “Dances” to music
  - Laughs at certain voices
- Dx: PE tube blocked in right ear, middle ear dysfunction suspected
- Aided testing
  - SAT at 25 dBHL
  - 40 dBHL at 500 Hz, further responses unreliable
- **5 months post-activation**
- Parent Report
  - Loves to “talk” on phone
- Aided testing attempted but patient irritable and unreliable
- No mapping completed
- Currently on agenda for 2<sup>nd</sup> Ear Implantation
- Impedance and NRT completed in surgery
- Mapping schedule recommended includes more frequent appointments near activation graduating to less frequent (ie., every 6 months) intervals upon Map stabilization
- Mapping involves use of objective (impedance, NRT, ESRT) and behavioral (VRA, CPA) techniques to set stimulation levels
- Behavioral testing is used to verify Map settings
- Patients are typically provided with progressive Maps of increased stimulation early on
- Different programs often employ different sound processing strategies after the Map is determined to be stable

# Case #3 – Therapy Services

## SLP Clinic-based

- 7-12 months
  - 1x/month
  - Focus parent education in anticipation of CI
  - Early language development
- 13 months – present
  - 1x/week diagnostic therapy
  - Focus on auditory awareness; language development

## SLP Home based

- 4 months – 15 months
  - - 1x/week
  - Feeding therapy
- 16 months – present
  - 1x/week
  - Language/Communication

## Additional Therapies

- IESDB 1x/week
- PT 1x/week

# Case #3 SLP Assessments

- Baseline - 6 months
- Progress
  - 15 months (HA 1 month)
  - 20 months (HA 6 months)

# Case #3 Auditory Skills

## IT-MAIS

Pre-Implant

0/40

1 month post-implant

5/40

6 months post-implant

16/40

- Alerting to name
- Discriminating familiar voices
- Identifies phone
- Moves to singing
- Associates a few objects with sounds

# Case #3 - Language Skills

	Auditory Skills IT-MAIS	Receptive Language Rosetti	Expressive Language Rosetti
Pre-implant	0/40	No Basal	No Basal
1 month post	5/40	No Basal	0-3 months
6 months post	16/40	3-6 months	0-3 months
	Alerting to names, discriminates familiar voices, identifies phone, moves to singing, sound/object assn.	Emerging Skills at 12 month level Improved attn, engagement, play, motor skills	Emerging Skills At 9 month level Improved motor skills, greater variety of sounds

# Case #4- Intake History

- CI candidacy evaluation: age 7 months
- Newborn Screening – Referred bilaterally (OAE + ABR)
- Risk Indicators:
  - Born premature at 35 weeks, NICU stay: 2 weeks, mechanical ventilation
  - Out of state identification and diagnosis
  - Confirmation of severe bilateral sensorineural hearing loss at age 5 months
  - Hx of chronic OME (PE tubes placed at age 3 months)
  - Recommendations: Hearing aid fitting, SLP evaluation and treatment, audiologic monitoring
- Family status: Mother; age 17 at birth, resides with great grandmother. At birth, deemed high risk social situation; CPS involved

# Case #4 Assessment

- **7 months old**
- Behavioral responses observed in severe range of hearing loss; testing limited due to age
- Fit with binaural amplification
- **9 months old**
- Parent Report: Wearing hearing aids inconsistently
- Unaided SF results: No reliable responses
- Aided SF results: Responded to low frequency stimuli at elevated levels
- IT-Mais Score = 15/40
- Rec:
  - Sedated ABR
  - Increase HA use
- **11 months old**
- Sedated ABR Results: Severe to profound SNHL bilaterally
- **1 year old**
- Obtained left ear Cochlear implant
- **1 month post-activation**
- Speech awareness and cochlear implant responses within mild range
- IT-Mais Score = 29/40

# Case #4 - Assessment

- **4 months post-activation**
- Family Report
  - Adopting CI with good success
  - May pull off processor with loud noise (ie. dogs barking) or when tired and cranky
  - Aware of environmental sounds
  - Increasingly more vocal
  - Regular In-home tx with I/T Program
  - IESDB Outreach Services regularly
  - Using contralateral hearing aid only 2-3 x/week
- CI Aided responses in mild to moderate range
- Middle ear disorder suspected
- Rec:
  - Medical Consultation
  - Increased CI + HA Use
  - Re-eval + Mapping in 2-3 months
  - **6 month post-CI Speech evaluation with Susie Jones, SLP to assess development of auditory skills in relation to cochlear implantation**
- Infant/Toddler Coordinator and IESDB counselor both have expressed concerns regarding wear time and consistent use, particularly with HA
- Request to purchase retention devices to help assist family in increasing CI wear time

# Pediatric Speech/Language Tests

## 0 - 3 years

- Re-evaluate every 3 months, criterion checklists
  - Formal ax every 6 months
- Language ~ Listening ~ Speech ~ Play ~ Cognition
  - Outcomes
    - Tx recs
  - Therapy goals
  - Parent Education
- Ongoing diagnostic therapy

# Pediatric Speech/Language Tests

## 0-3 years

- Language
  - Rosetti Infant-Toddler Language Scale
  - Preschool Language Scale
  - Language sample in play
  - CASLLS
  - CSBS
- Listening
  - IT-MAIS
  - Little Ears
  - Auditory Skills Checklist
  - Early Speech Perception Test
  - CASLLS
- Speech
  - Standardized:
    - Goldman-Fristoe Test of Articulation
    - Arizona Test of Articulation
  - Observation/Parent Report
    - Sound/Syllable Inventory
    - CASLLS
  - Imitative Tasks
- **Integrated Scales of Development**
- Play
  - Rosetti
  - PLS
  - CASLLS
  - CSBS
- Cognition
  - Rosetti
  - PLS
  - CASLLS
  - CSBS