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Phoenix Children's

# Pediatric Grand Rounds: Embracing the Unexpected

Deborah Flynn, AuD
Allie Sayer, AuD
Christina Dubas, AuD
Rachel Worcester, AuD
Caroline Sabatino, AuD
Robert Fanning, AuD

Wendy Steuerwald, AuD

# Wendy Steuerwald, AuD, CCC-A

Wendy Steuerwald, AuD, is the Director of Audiology at Phoenix Children's Hospital. She works with hospital leadership and audiologists to create an environment where exceptional patient care is the norm. This includes use of evidence based practice, process improvement initiatives, expansion of services offered, the addition of new locations, and hiring the best audiologists. Wendy's overall focus is to improve the patient's experience within audiology. Professional leadership includes committee work with the American Academy of Audiology, American Speech Language Hearing Association, and multiple state organizations. Wendy also participated in numerous presentations and publications. Local committee work includes participation in the interdisciplinary Stop CMV AZ partnership.

# Deborah Flynn, AuD, CCC-A

Deborah Flynn is the Audiology Lead for Cochlear Implants at Phoenix Children's Hospital. She has extensive experience in identification, intervention, and management of infants, children, and teens with hearing loss. Her involvement in the Audiology Team includes cochlear implants, hearing aids, pediatric audiological evaluations, ototoxic monitoring, evoked potentials, assistive listening technology, and newborn hearing screenings. Her clinic

interests include a multidisciplinary team approach for children with cochlear implants and hearing aids. She presents on various audiological topics throughout Arizona.

She has been an adjunct professor at AT Still University, Arizona School of Health Sciences and enjoys teaching students.

# Allie Sayer, AuD, CCC-A

Allie Sayer, AuD, CCC-A graduated from Arizona State University with a Bachelor of Science degree in Speech and Hearing Sciences; she also graduated from Barrett, the Honors College at ASU after completing an honors thesis studying vocabulary differences in children with mild to moderate hearing loss in comparison to their normal hearing peers. She continued at Arizona State University to obtain her Doctorate in Audiology degree, after completing her hospital-based externship at St. Louis Children's Hospital. Dr. Sayer worked in the private sector with both children and adults before coming to Phoenix Children's Hospital. She is a member of and certified through the American Speech, Language, and Hearing Association. Her clinical areas of specialty include diagnostics, hearing aids, and cochlear implants.

# Christina Dubas, AuD, CCC-A

Christina Dubas, AuD, CCC-A, is a clinical audiologist at Phoenix Children's Hospital where she works with children of all ages in both the inpatient and outpatient hospital setting. She enjoys working with families and medical teams to provide individualized patient care. Her clinical areas of interest include electrophysiological assessment and vestibular assessment. She has publications in the areas of speech-in-noise performance of children and auditory brainstem response.

# Rachel Worcester, AuD, ABA-C

Rachel Worcester, AuD, ABA-C, is a clinical audiologist at Phoenix Children's Hospital (PCH) specializing in the diagnosis and management of childhood hearing loss. She graduated from the Northeast Ohio Au.D. Consortium (NOAC) in 2021 with her Doctorate in Audiology. Prior to joining Phoenix Children's Hospital in September of 2023, she worked at Texas Children's Hospital where she treated pediatric patients in outpatient hospital settings. Rachel has a special interest in vestibular assessment and during her time at PCH she and Dr. Dubas have worked to establish the PCH Balance Clinic.

# Caroline Sabatino, AuD, CCC-A

Caroline Sabatino, AuD, CCC-A graduated from Arizona State University with a Bachelor of Science in Speech and Hearing Sciences. She then went on to work as a Speech-Language Pathologist Assistant at a developmental preschool in Washington Elementary School District. Caroline Sabatino returned to school and in 2016 she graduated from A.T. Still University with her Doctorate in Audiology degree. Her clinical areas of specialty include diagnostics and hearing aids for pediatric patients.  In addition to her position at Phoenix Children's Hospital, she is the resident audiologist at Desert Voices Oral Learning preschool in Phoenix, Arizona where she manages the teacher's hearing assistive technologies and checks the student's hearing aids and cochlear implants. She is a member of, and certified through, the American Speech, Language, and Hearing Association.

# Robert (Bob) Fanning, AuD, CCC-A

Robert Fanning, AuD, CCC-A is a Lead Audiologist at Phoenix Children's. He has been a clinical audiologist for over 25 years and has over 20 years of experience in the diagnosis and treatment of pediatric hearing disorders. He has held volunteer faculty positions as a clinical instructor at Baylor College of Medicine and Arizona State University, has served as a clinical preceptor for students from several training programs, and has given many invited lectures to physicians, advanced practice providers, students, and other learners.

# Disclosures

- **Presenter Disclosure**: Financial: Allie Sayer is employed by Phoenix Children's Hospital. Non-financial: Allie Sayer has no relevant non-financial relationships to disclose. Financial: Deborah Flynn is employed by Phoenix Children's Hospital. Non-financial: Deborah Flynn has no relevant non-financial relationships to disclose. Financial: Wendy Steuerwald is employed by Phoenix Children's Hospital. Non-financial: Wendy Steuerwald has no relevant non-financial relationships to disclose. Financial: Robert (Bob) Fanning is employed by Phoenix Children's Hospital. Non-financial: Robert (Bob) Fanning has no relevant non-financial relationships to disclose. Financial: Christina Dubas is employed by Phoenix Children's Hospital. Non-financial: Christina Dubas has no relevant non-financial relationships to disclose. Financial: Caroline Sabatino is employed by Phoenix Children's Hospital. Non-financial: Caroline Sabatino has no relevant non-financial relationships to disclose. Financial: Rachel Worcester is employed by Phoenix Children's Hospital. Non-financial: Rachel Worcester has no relevant non-financial relationships to disclose.
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# Learning Outcomes

After this course, participants will be able to:

- Discuss the importance of multi-disciplinary teamwork.
- Discuss several complex audiology cases.
- Discuss audiology testing methods used to obtain accurate results with complex patients.

# A Case About Changes in Pediatric Cochlear Implant Guidelines

Case Study: Aiden

Deborah Flynn, AuD, CCC-A

# Current FDA Criteria For Pediatric Cochlear Implants

- Children (2-17 years)
  - Severe to profound sensorineural hearing loss in both ears
  - Limited benefit from binaural amplification
  - Multisyllabic Lexical Neighborhood Test (MLNT) or Lexical Neighborhood Test scores <30%

# Introducing Aiden

- Aiden is an almost 17-year-old male followed on-and-off at our clinic since age 2.
- His history is significant for two failed in-patient newborn hearing screenings, one failed outpatient screening, and a diagnostic ABR at another facility that demonstrated bilateral moderate-to-severe hearing loss.
- He had not been fit with hearing aids when he came to our clinic at the age of 2. Records were unavailable at the time he established with our clinic.

# Diagnostic ABR

- He was initially seen by a colleague, the ABR study confirmed a moderate-to-moderate/severe hearing loss in both ears
- Type A tympanograms, AU
- A month later he was fit with Oticon Safari hearing aids
- 1 year later, his care was transferred to me

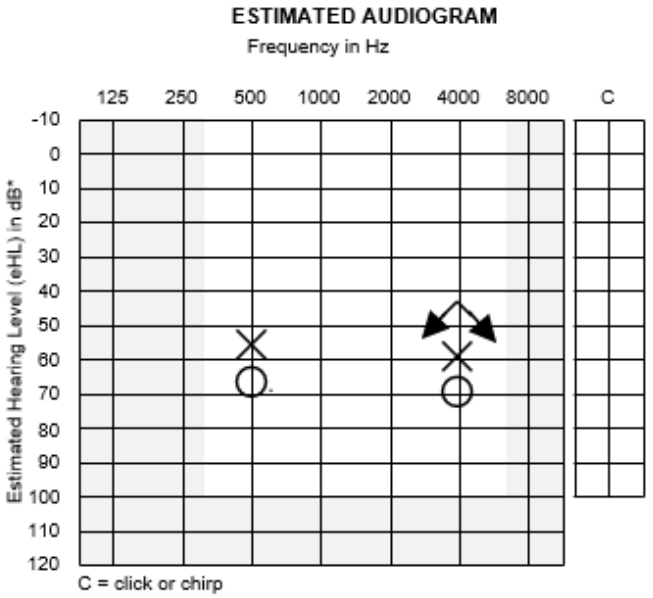



Image: Phoenix Children's Hospital

# Behavioral Testing

- At 3 years 8 months, behavioral testing indicated a moderate hearing loss in the left ear and a moderate/severe hearing loss in the right ear
- Type A tympanograms
- DPOAEs absent, bilaterally
- Speech testing could not be completed due to severe language delay. (bilingual-English/Arabic)
- He attended a developmental preschool program

Audiogram:

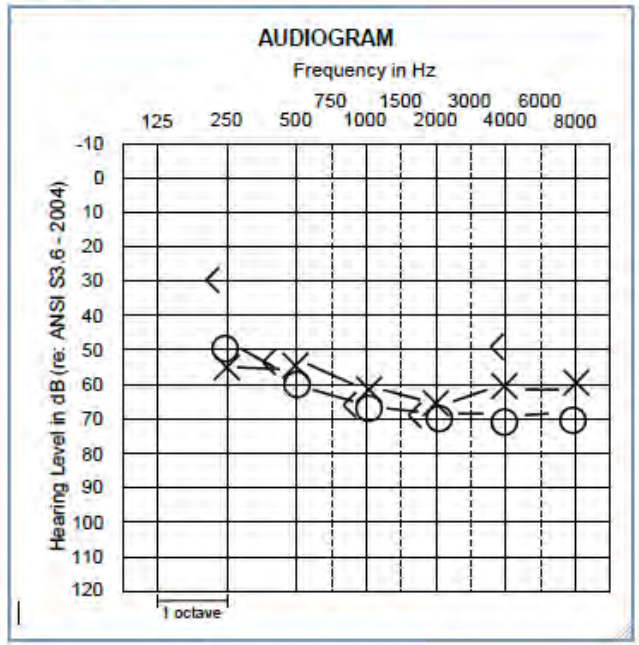


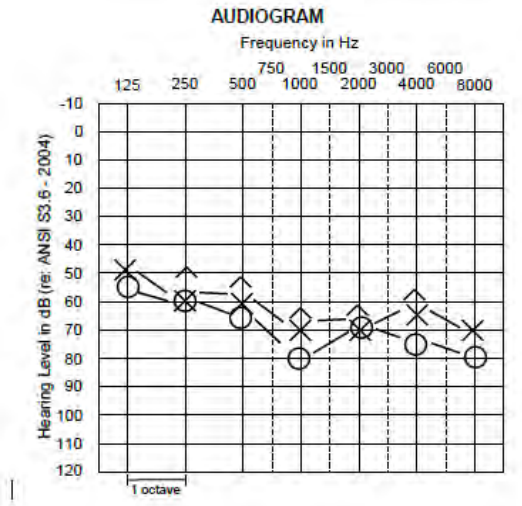

Image: Phoenix Children's Hospital

# Transfer of Care

- Aiden's care was transferred to another clinic due to insurance reasons
- He received services at this clinic for 6 years
- He was fit with Super Power hearing aids initially and later fit with Power hearing aids
- He returned to our clinic in 2018 for a second opinion
- He had Power hearing aids but felt that he could not hear with them and was wearing his previous Super Power hearing aids which were not working well

# Audiological Testing 6.5 Years Later

- At age 11, slight changes in hearing were noted, with results in the moderately/severe to severe hearing loss range, Right>Left.
- His primary complaint was that his hearing aids were not loud enough.
- The Power hearings were reprogrammed but he felt that he could not hear with them and was relying on his old SP hearing aids.




| Side | Device | Presentation Level | dB Type | Percentage Correct (Words) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|
| right | | 100 | HL | 68 | NU6 | Recorded | quiet |
| left | | 100 | HL | 64 | NU6 | Recorded | quiet |
| bilateral | HA | 50 | HL | 64 | WIPI | Recorded | quiet |
| | | | | | | | |

Images: Phoenix Children's Hospital

# Speech and Academics

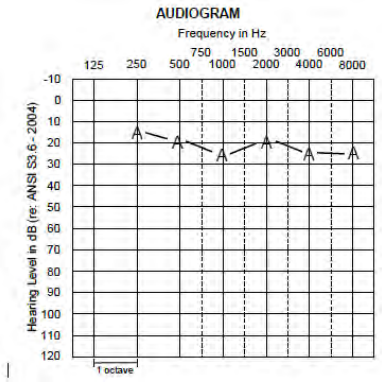
- His speech and language continued to be significantly delayed
- Academically, he was struggling
- His parents switched schools several times in the six years hoping to find a program that could “help him catch-up”
- They tried several public schools, a private school, and a charter school
- He attended private speech therapy and AVT therapy on a weekly basis
- At 11-years-old, he was reading at a 1st grade level
- His articulation was extremely difficult to understand despite full-time hearing aid use

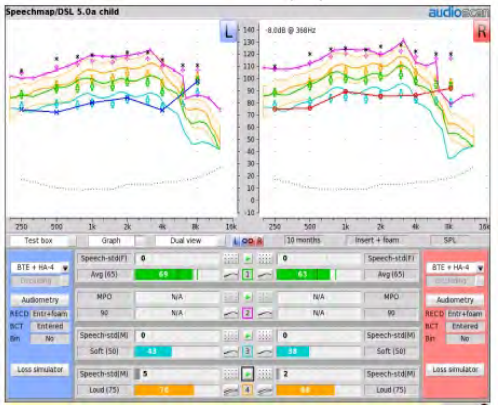
# Speech Testing

- Speech testing was challenging due to severe language delay and articulation errors. At times, he had to use the word in a phrase for clarification or he would show me the word (example: thumb) or explain it.
- Testing was completed with recorded speech and free response but also completed with a WIPI to fully assess auditory function.

# Hearing Aids

- The hearing aid company was contacted and agreed to switch out the Power HAs for Super-Power hearing aids
- Real Ear Measures were used with current thresholds and RECDs to program the SP hearing aids




| Side | Device | Presentation Level | dB Type | Percentage Correct (Words) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|
| bilateral | HA | 50 | HL | 64 | WIPI | Recorded | quiet |
| | | | | | | | |

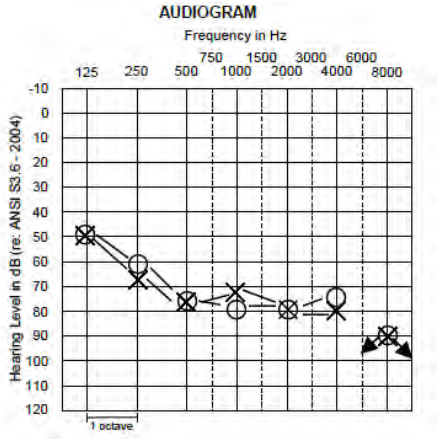
Images: Phoenix Children's Hospital

# Monitoring

- Aiden continued to be monitored every six months as per parent request
- Hearing was stable
- He wore his hearing aids on a daily basis for all waking hours
- He continued to struggle with speech/language and academics. He was making some progress in reading skills but was significantly delayed (3rd grade)
- His mother consulted with several surgeons in the area regarding cochlear implants. Family was told he was not a candidate.

# Two Years Later (age 13)

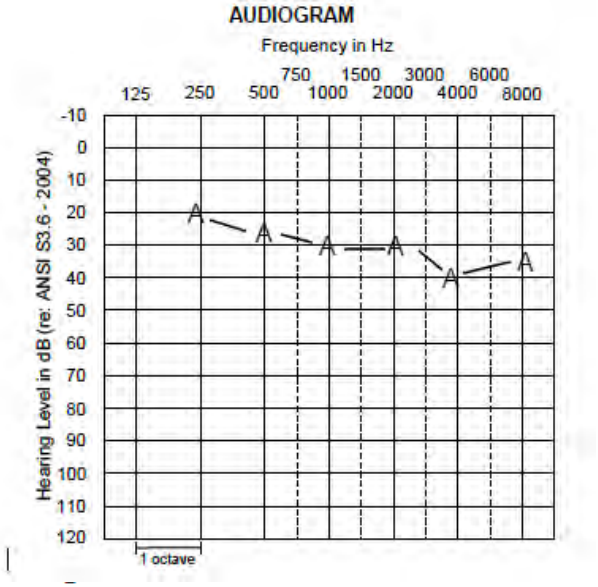
- Aiden returned for bi-annual testing w/ a c/o decreased hearing and HAs not loud enough
- Slight decreased noted
- Speech discrimination ability was very poor
- Academically, he continued to be below grade level

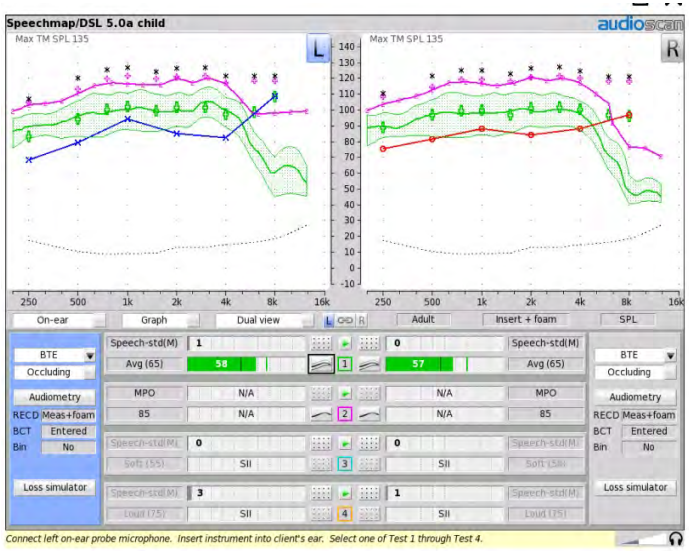



| Side | Presentation Level | dB Type | Percentage Correct (Words) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|
| right | 105 | HL | 44 | NU6 | Recorded | quiet |
| left | 100 | HL | 42 | NU6 | Recorded | quiet |
| | | | | | | |

Images: Phoenix Children's Hospital

# Aided Benefit



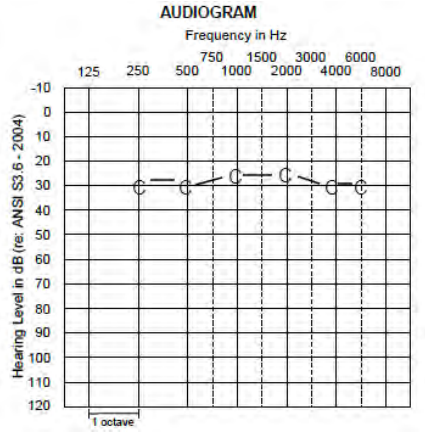




| Side | Presentation Level | dB Type | Percentage Correct (Words) | Percentage Correct (Phonemes) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|
| bilateral | 60 | dBA | 42 | 67 | CNC | Recorded | quiet |
| right | 60 | dBA | 34 | 62 | CNC | Recorded | quiet |
| left | 60 | dBA | 30 | 56 | CNC | Recorded | quiet |
| | | | | | | | |

Images: Phoenix Children's Hospital

# Cochlear Implant

- He was implanted and activated in the right ear in September
- On the day of activation, he had discrimination of basic LING sounds
- Within one month his aided detection levels were in the mild hearing loss range and his WIPI score was 92%
- By December of the same year, his speech discrimination ability improved immensely, and he moved through the PMSTB




| Date | Side | Device | Presentation Level | dB Type | Percentage Correct (Words) | Percentage Correct (Phonemes) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|---|---|
| 10-01-2020 | right | CI | 60 | dBA | 92 | | WIPI | Recorded | quiet |
| | right | CI | 60 | dBA | 76 | | LNT | Recorded | quiet |
| 12-01-2020 | right | CI | 60 | dBA | 76 | 93 | CNC | Recorded | quiet |
| | | | | | | | | | |

Articulation errors still noted

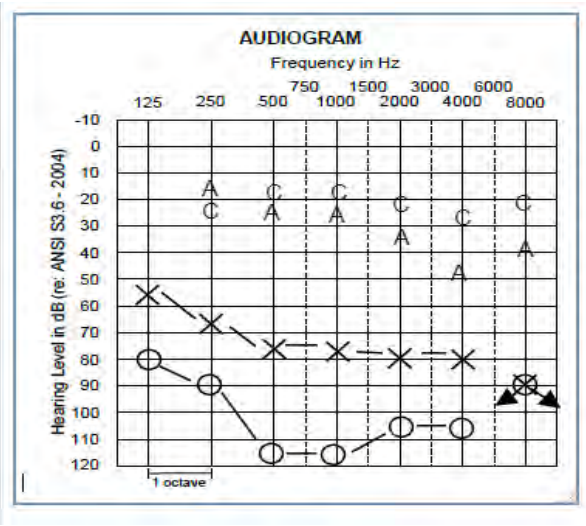
Images: Phoenix Children's Hospital

# One Year Later

- Aiden returned. He and his mother were extremely pleased with his progress
- His family found a private school that had 1:1 tutoring, and he was rapidly making progress in his reading/language skills. In addition, his articulation had improved significantly.
- They inquired about a second cochlear implant
- Aiden indicated that he struggled to hear with his hearing aid alone

# CI Evaluation for Other Ear

- CI Evaluation completed




| Side | Device | Presentation Level | dB Type | Percentage Correct (Words) | Percentage Correct (Phonemes) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|---|
| left | HA | 60 | dBA | 32 | | CNC | Recorded | quiet |
| right | CI | 60 | dBA | 84 | 94 | CNC | Recorded | quiet |
| | | | | | | | | |

Images: Phoenix Children's Hospital

# 2nd CI Progress

- Aiden was as successful with the second implant as the first
- His aided detection improved within one month and his speech discrimination was excellent by 3 months
- His academic performance continued to improve

| Side | Device | Presentation Level | dB Type | Percentage Correct (Words) | Percentage Correct (Phonemes) | Word List | Stimulus | Condition |
|---|---|---|---|---|---|---|---|---|
| left | CI | 60 | dBA | 80 | 93 | CNC | Recorded | quiet |
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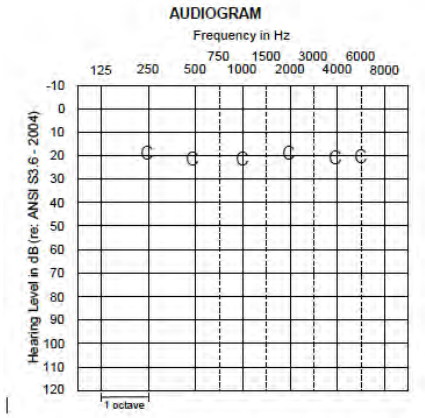



Images: Phoenix Children's Hospital

# Aiden's Academic Performance

- With 1:1 tutoring, ongoing speech therapy and AVT therapy over the past 4 years, Aiden has substantially closed the gap in regard to his academic grade equivalency
- He is currently enrolled in high school and will be taking college courses his senior year
- He plans on attending University after graduation
- He started a successful printing business that now has an online presence

# Changes in Pediatric Cochlear Implant Candidacy

- When Aiden was progressing through this process, he certainly did not meet the FDA criteria, yet was struggling significantly
- At the time, there was limited published consensus on pediatric cochlear implant candidacy; although, publications were starting to emerge
- Since that time, The American Cochlear Implant Alliance (ACIA), has published The “American Cochlear Implant Alliance Task Force Guidelines for Determining Cochlear Implant Candidacy in Children”
- This has significantly improved our CI Team’s ability to make decisions regarding pediatric candidates that fall under the “off-label” category

# Cochlear Implant Assessment

- His mother continued to inquire about cochlear implantation and was pushing for consideration
- A CI evaluation was completed and with hesitancy, the PCH CI team decided to consider a cochlear implant in one ear
- The deciding factor was his poor speech discrimination ability despite adequate fit HAs and poor academic performance

# New ACIA Pediatric CI Guidelines

1. Audiometric Thresholds >70dB HL and/or
2. Word recognition <50% and/or
3. Poor functional performance, limited progress, poor quality of life

- REFER FOR COCHLEAR IMPLANT EVALUATION

# Summary

- Aiden met all of the criteria mentioned in the ACIA guidelines; however, at the time of our decision, we did not have these guidelines
- Aiden significantly benefited from cochlear implants
- When evaluating children, particularly those who are struggling, consider the criteria on the last slide and if they meet any or all of the criteria, consider referring for a cochlear implant evaluation


ACIA Pediatric CI Guidelines for Candidacy
Progressive HL
Non-traditional CI Candidate
Aiden
Poor Aided Speech Discrim
Poor Articulation
Poor Academic Performance

# A Case About Consent and Assent

Case Study: Jake

Allie Sayer, AuD, CCC-A

# Consent and Assent in Pediatric Audiology

- Consent = to give approval, to permit, to agree
  - Proxy Consent/Parental Permission
- Assent = to agree or concur; to approve of something; to give in, yield, or concede
- Dissent = to withhold assent or approval; to differ in opinion
- How do we determine if a patient is able to provide assent?
- How do we balance parental consent and patient assent when making decisions if they are misaligned?

# Introducing Jake

- 16-year-old male
- Diagnosed with autism and bilateral, asymmetric sensorineural hearing loss
- Using a hearing aid for the left ear only
- Highschool student, on an IEP, receiving speech/occupational therapy at school and outside ABA therapy

# History Continued

- Birth history
  - Full term
  - Complicated by meconium aspiration
  - NICU stay where he received IV gentamicin
- Early hearing concerns which were later attributed to autism (diagnosed at age 2)
- No newborn hearing screening
- First diagnosed with sensorineural hearing loss in 2013 at 4 years of age – moderate in the left ear, profound/no response in the right ear Fit with left hearing aid
- CT Scan – normal but “mildly prominent right vestibular aqueduct”
- Progression of loss first noted in January 2018, and further decreases in May 2019

# Outside Records

- Most recent evaluation prior to establishing with our office was in August of 2022
- Reliability for testing was reported as good
- 80% at 90 dB HL

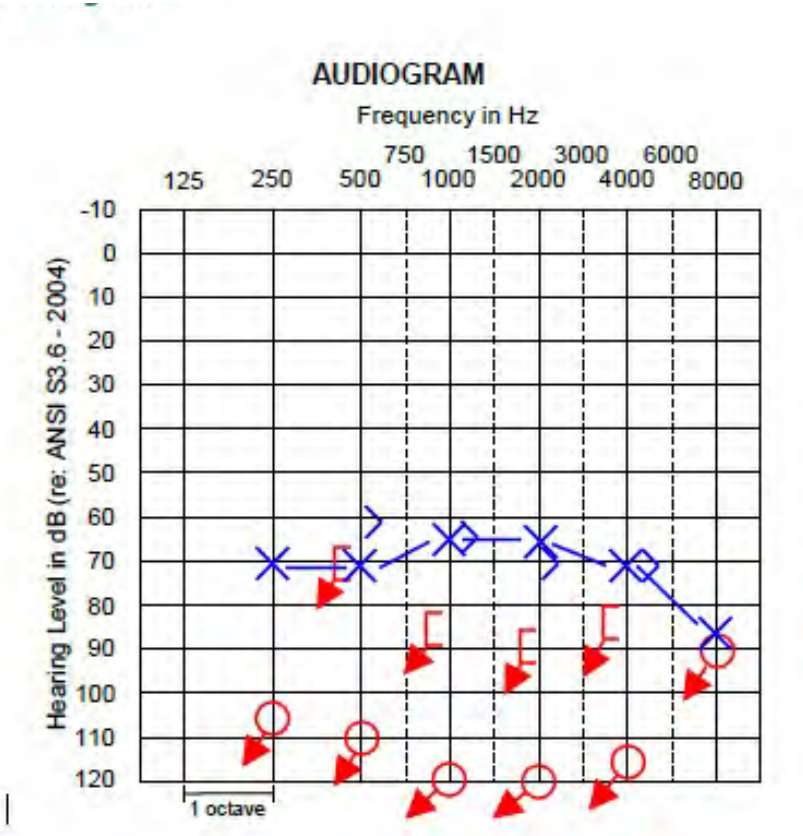



Image: Phoenix Children's Hospital

# March 2023 Results

- Pure tone testing
  - Right ear profound (no response) sensorineural hearing loss
  - Left ear moderately-severe sloping to profound hearing loss
- Speech testing
  - SRT at 65 dB HL
  - WRS
    - @ 95 dB with PBK – 52%
    - @95 with WIPI – 76%

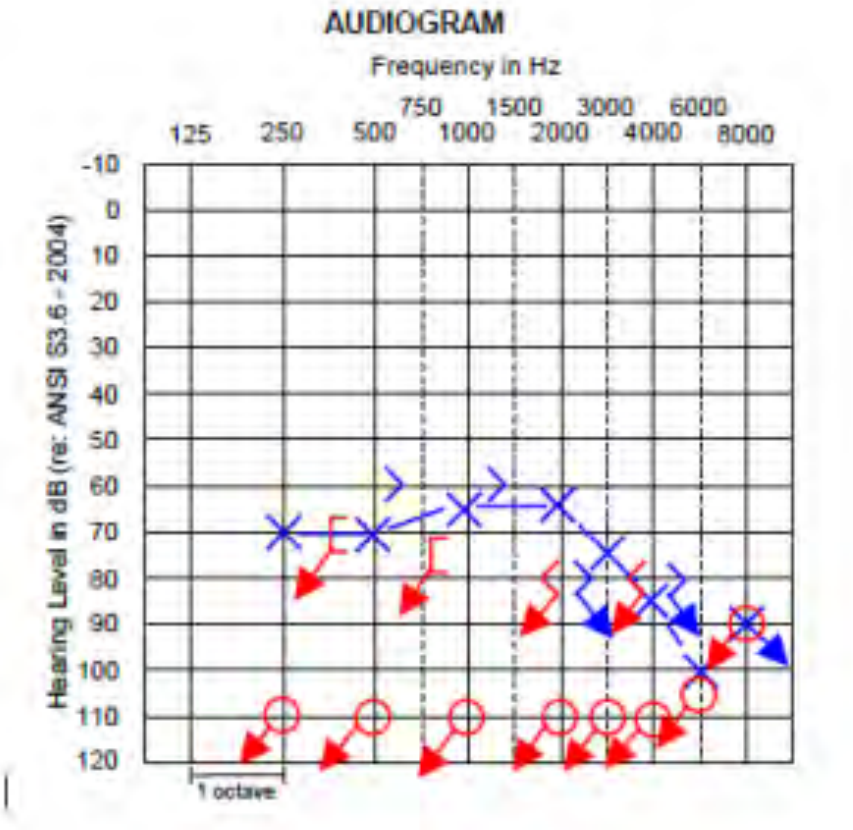



Image: Phoenix Children's Hospital

# Cochlear Implant Team Workup

- Established with ENT/CI surgeon August 2023
  - MRI, speech and language evaluation, CI audiology evaluation, genetics, ophthalmology and psychology consultations ordered
  - Mom requesting evaluation for right cochlear implant

# CI Workup Findings

- Genetics (September 2023)
  - Trio Whole Exome Sequencing with Mitochondrial Analysis, Reflex to Microarray, Reflex to Fragile X labs ordered
  - Mutations in the GJB2, ZEB2, and AUTS2 genes
    - GJB2 result consistent with patient being at least a carrier for autosomal recessive sensorineural hearing loss but cannot establish a molecular diagnosis due to lack of 2nd variant in this gene. Cannot exclude possibility that this result may be associated with autosomal dominant GJB2-related SNHL.
    - Other mutations are not hearing loss related and also not found to be causative of his developmental delays or autism

# CI Workup Findings

- MRI (January 2024)
  - Bilateral perisylvian polymicrogyria (BPP)
  - Unremarkable appearance of the internal auditory canals bilaterally
  - No evidence of cochlear nerve deficiency
  - Upper limit of normal globe size
- Ophthalmology (January 2024)
  - Glasses not needed at this time

# CI Workup Findings

- Speech (February 2024)
  - Severe receptive and expressive language disorder
  - Therapy expectations post-CI for 1x/week or 2x/month, with telehealth acceptable
  - Parent concern for how Jake would adjust to the CI reported
- Psychology (December 2023 and February 2024)
  - Extreme dislike of missing school to attend medical appointments
  - Difficulty adapting to new hearing aid – fixated on finding old one
  - Mom's main concern is for limitation of his future communication abilities with progressive hearing loss and loss of quality of life with his love of music and singing – along with safety concerns if he loses hearing in the left ear

# Unaided Test Results

- Type A tympanograms
- Absent ipsilateral acoustic reflexes 500-4000 Hz bilaterally
- No changes to air-conducted thresholds during conventional audiometry
- Speech testing:
  - Right: No response at max output limit
  - Left: 80 dB SRT and 64% WRS using NU-6

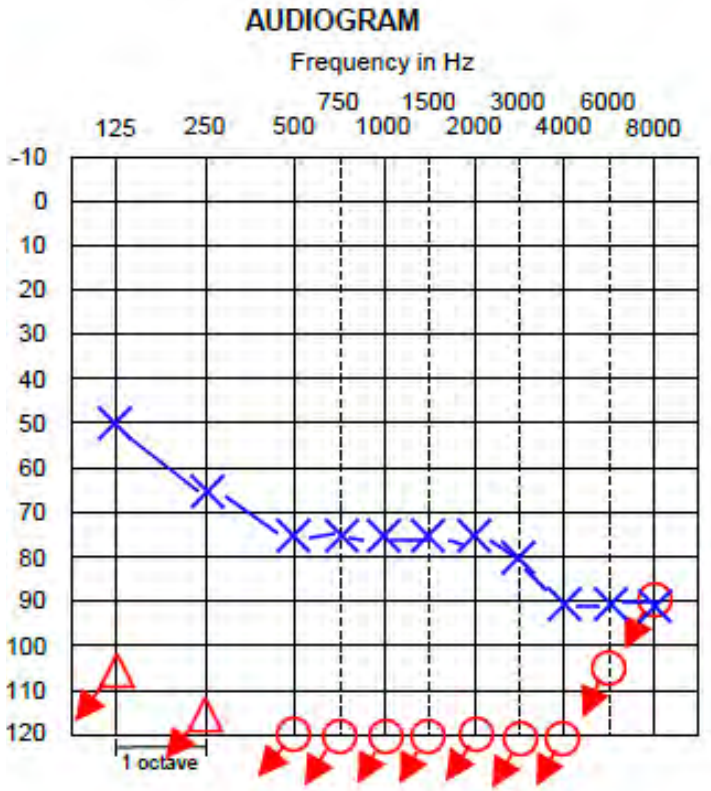



Image: Phoenix Children's Hospital

# Aided Testing

- Aided thresholds for each ear
- Speech testing:
  - Right Ear: NR SRT @ max output, 0% word and phoneme scores on CNC
  - Left Ear: SRT at 25 dB HL, 52% word score and 73% phoneme score on CNC

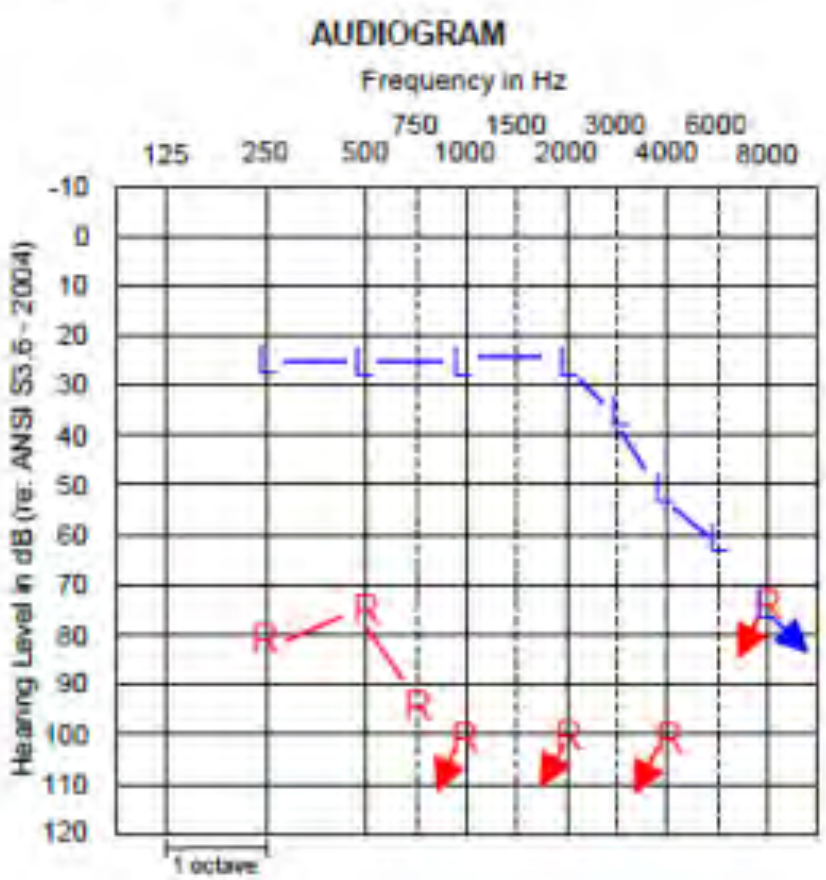



Image: Phoenix Children's Hospital

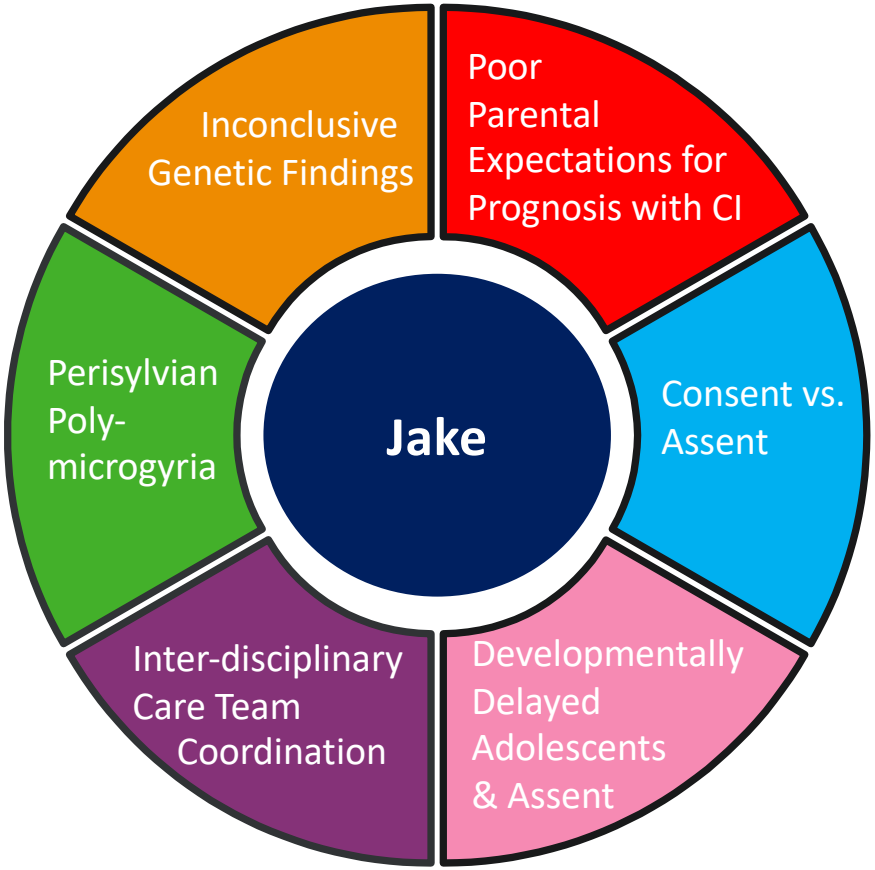
# Counseling and Discussion

- Left ear borderline candidate for implantation, but not recommended at this time
  - PTA 75 dB
  - Overall degree of hearing loss severe
  - Poor word understanding both aided and unaided
- Right ear technically meets FDA criteria for implantation, but not expected to have good outcomes
  - Profound hearing loss with no response at limits
  - No measurable word understanding
  - No measurable benefit from amplification

# Team Review and Decision

- Concerns:
  - Poor prognosis for performance
  - Inconsistent parental report of history and patient tolerance of changes to devices
  - Unclear etiology of hearing loss (genetic testing inconclusive, history of IV gentamicin)
  - Poor parental understanding of expected benefits and limitations of CI use for the right ear
  - Lack of patient understanding and assent for the process
  - Mother's desire to proceed/consent for implant with unrealistic expectations
- Decision: Not a candidate for the right ear. Will be monitored for candidacy in the left ear.

# Learning from Jake



# Anything Goes After Trauma

Case Study: Beth

Christina Dubas, AuD, CCC-A

# Temporal Bone Fractures

- Longitudinal fractures
  - 70 to 80% of temporal bone fractures
  - The fracture line is parallel to the long axis of the petrous temporal bone
  - Conductive hearing loss is common
    - Hemotympanum, ossicular disruption, tympanic membrane perforation
- Transverse fractures
  - Less common
  - The fracture line is perpendicular to the long axis of the petrous temporal bone
  - Sensorineural hearing loss occurs approx. 50% of the time
    - Otic capsule violation, labyrinthine concussions, interruption of the blood supply to the cochlea, damage to the cochlear nerve

# Classification Based on Fracture Line Direction

- Based on research done in the 1950s
- Drawbacks:
  - Temporal bone fractures are more complex (could include both types)
  - The research shows this categorization does not help understand the prognosis of hearing loss and other complications

# Classification Based on Otic Capsule Status

- Otic capsule-sparing
  - Often involves the temporal bone's squamosal portion and external auditory canal, particularly the mastoid air cells, and middle ear
  - Higher occurrence of mixed or conductive hearing loss due to hemotympanum or ossicular chain disruption
- Otic capsule-violating
  - Force is transmitted to the capsule through the foramen magnum and petrous pyramid
  - Strongly associated with CSF leakage, SNHL, and intracranial pathology. Rarely affects the ossicular chain or external auditory canal

# Ossicular Discontinuity

- Connection between some ossicles can be difficult to visualize on imaging with the presence of hemotympanum
- Complete
  - No contact between the disconnected ends
  - Large, flat conductive hearing loss
- Partial
  - Joint contact or part of the ossicle is replaced by soft tissue or opposing ossicles are held in place by the force of one another
  - High-frequency conductive hearing loss is possible

# Introducing Beth

- 15-year-old female
- Admitted to the hospital following a high-speed motor vehicle collision
- Injuries include severe traumatic brain injury, traumatic occlusion of right internal carotid artery and left sided direct carotid-cavernous sinus fistula (CCF), left optic neuropathy (vision loss), bilateral temporal bone fractures and ossicular chain dislocation, right facial paralysis, and right leg paresis.

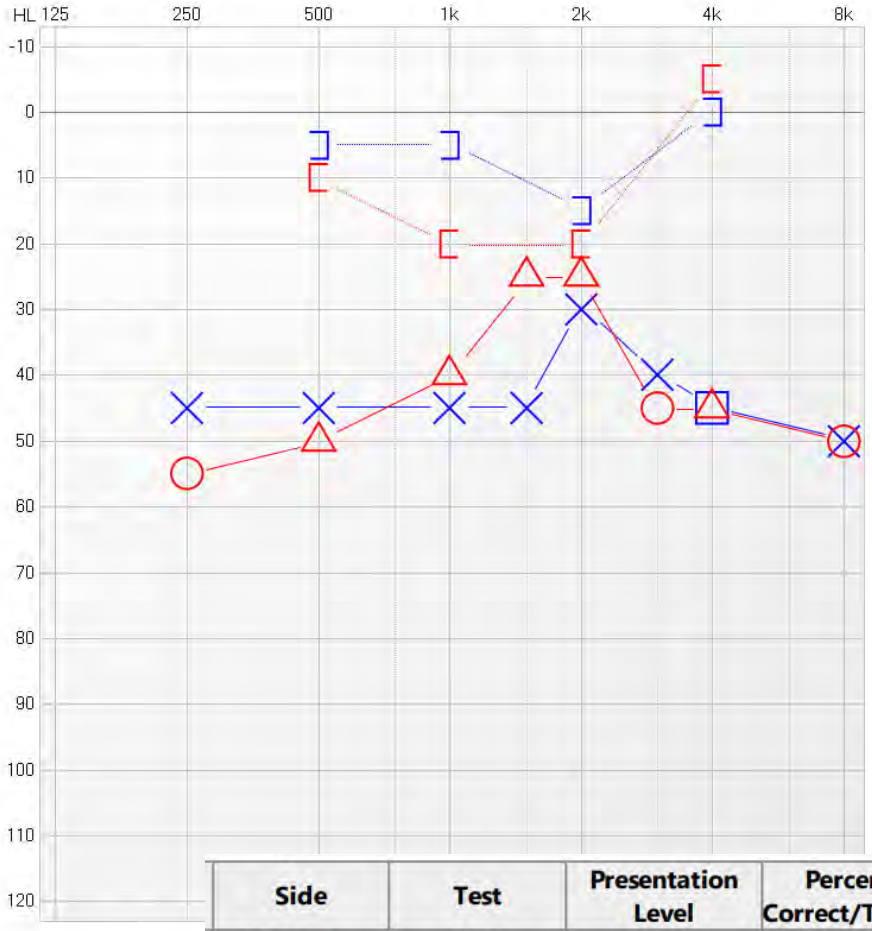
# CT Findings

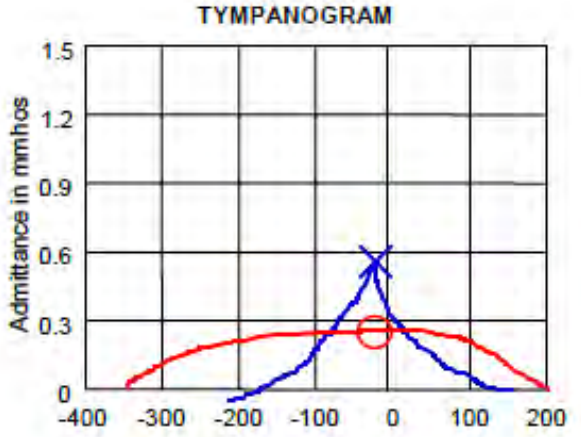
- Right CT Findings
  - Longitudinal and transverse right temporal bone fractures
  - Right middle ear ossicles disrupted and dislocated
- Left CT Findings
  - Longitudinal and transverse left temporal bone fractures
  - Left ossicular dislocation

# Inpatient ENT Evaluation

- Patient was seen by ENT the day prior to audiometric evaluation for bilateral ear canal debridement
  - Persistent, bloody, otorrhea, bilaterally (likely CSF leak)
  - Left TM somewhat thickened
  - Right TM opaque
  - Middle ears difficult to evaluate
- Patient counseled on the severity of hearing loss commonly seen with transverse fractures

# Inpatient Audiometric Presentation




| Right Ear | | Left Ear |
|---|---|---|
| 1.57 | Earcanal (mmhos) | 1.39 |
| 0.27 | Static Admittance (mmhos) | 0.55 |
| -25 | Peak Pressure (daPa) | -17 |
| 349 | Tympanometric Width (daPa) | 157 |

Images: Phoenix Children's Hospital

| Side | Test | Presentation Level | Percentage Correct/Threshold | Choice | Word List Used | Stimulus |
|---|---|---|---|---|---|---|
| Right Ear | SRT | | 40 | HL | Spondees | Recorded |
| Left Ear | SRT | | 35 | HL | Spondees | Recorded |
| Right Ear | WRS | 65m* | 92 | HL | NU6 | Recorded |
| Left Ear | WRS | 70m* | 96 | HL | NU6 | Recorded |

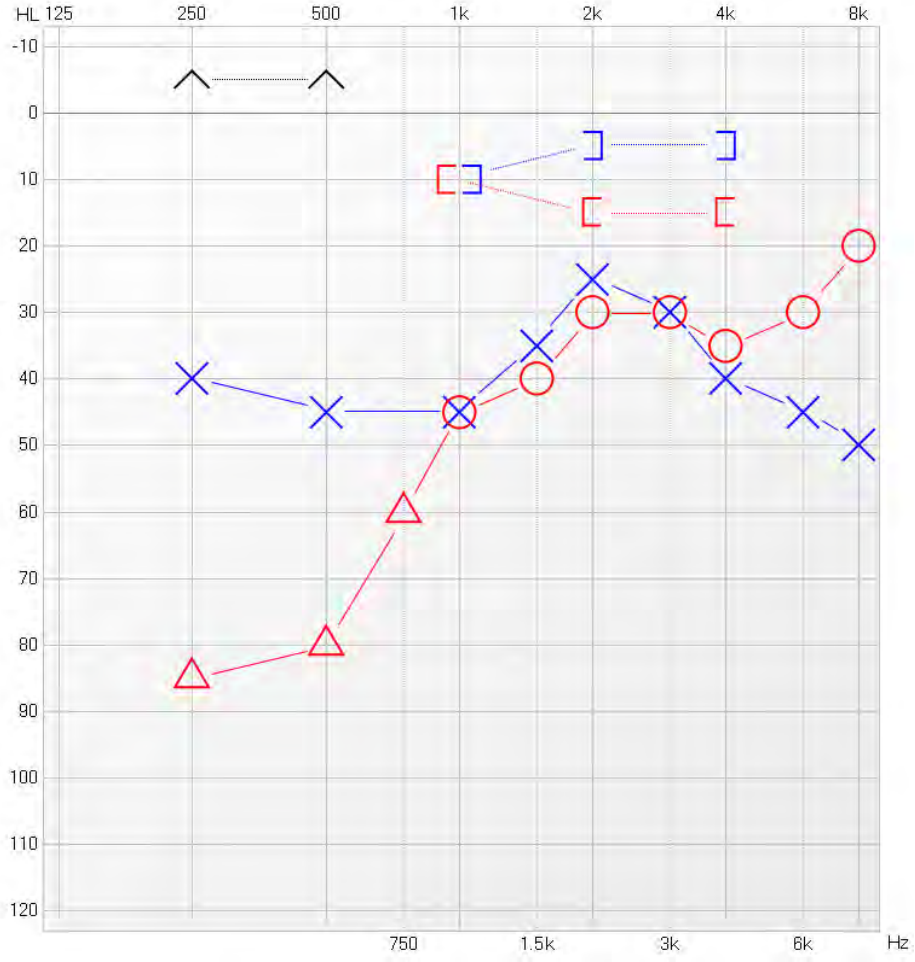
# Inpatient Hearing Aid Fitting

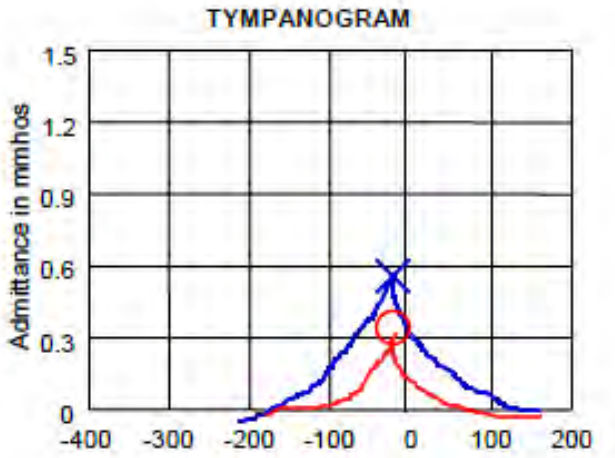
- The day after the evaluation the patient was fit with receiver in the canal hearing aids with non-custom domes from our loaner stock

- The patient was reluctant, and the parents were appreciative

# Follow-Up Audiometric Presentation

Completed at the time of ENT follow-up



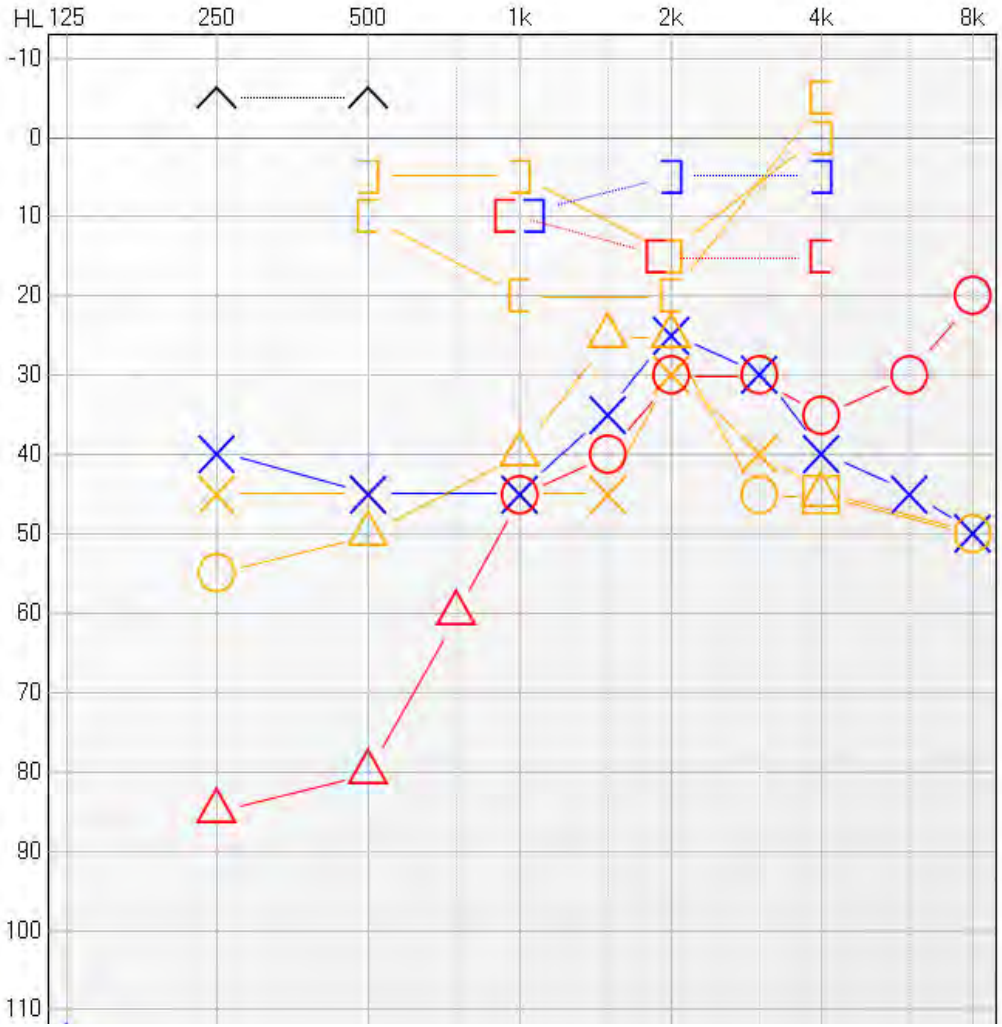




| Right Ear | | Left Ear |
|---|---|---|
| 1.1 | Earcanal (mmhos) | 1.3 |
| 0.27 | Static Admittance (mmhos) | 0.58 |
| -5 | Peak Pressure (daPa) | -8 |
| 25 | Tympanometric Width (daPa) | 34 |

- Masking plateau obtained at all masked frequencies
- Masking dilemma present for low-frequency BC
- SRT obtained at 35 dB HL in both ears with spondees

Images: Phoenix Children's Hospital

# Comparison of Audiograms




Noteworthy Change

- Tilt of right air conduction thresholds

Why did this happen?

- Hemotympanum previously providing support of ossicles?

How are there some thresholds so mild with ossicular dislocation?

**Anything goes after trauma**

Image: Phoenix Children's Hospital

# ENT Recommendations

- May discuss middle ear surgery in the next 6 months
  - More urgent matters to address at this point
- Referred back to Audiology for hearing aid management

# Hearing Aid Programming Adjustments

- A more powerful receiver was trialed on the right device to give more gain to the low-frequencies, but this only increased the SII by 1 point and the patient reported that the sound was "muffled" and there was a lot of interference with her own voice
- The lower strength receiver was kept

# Hearing Aid Follow-Up

## Datalogging:

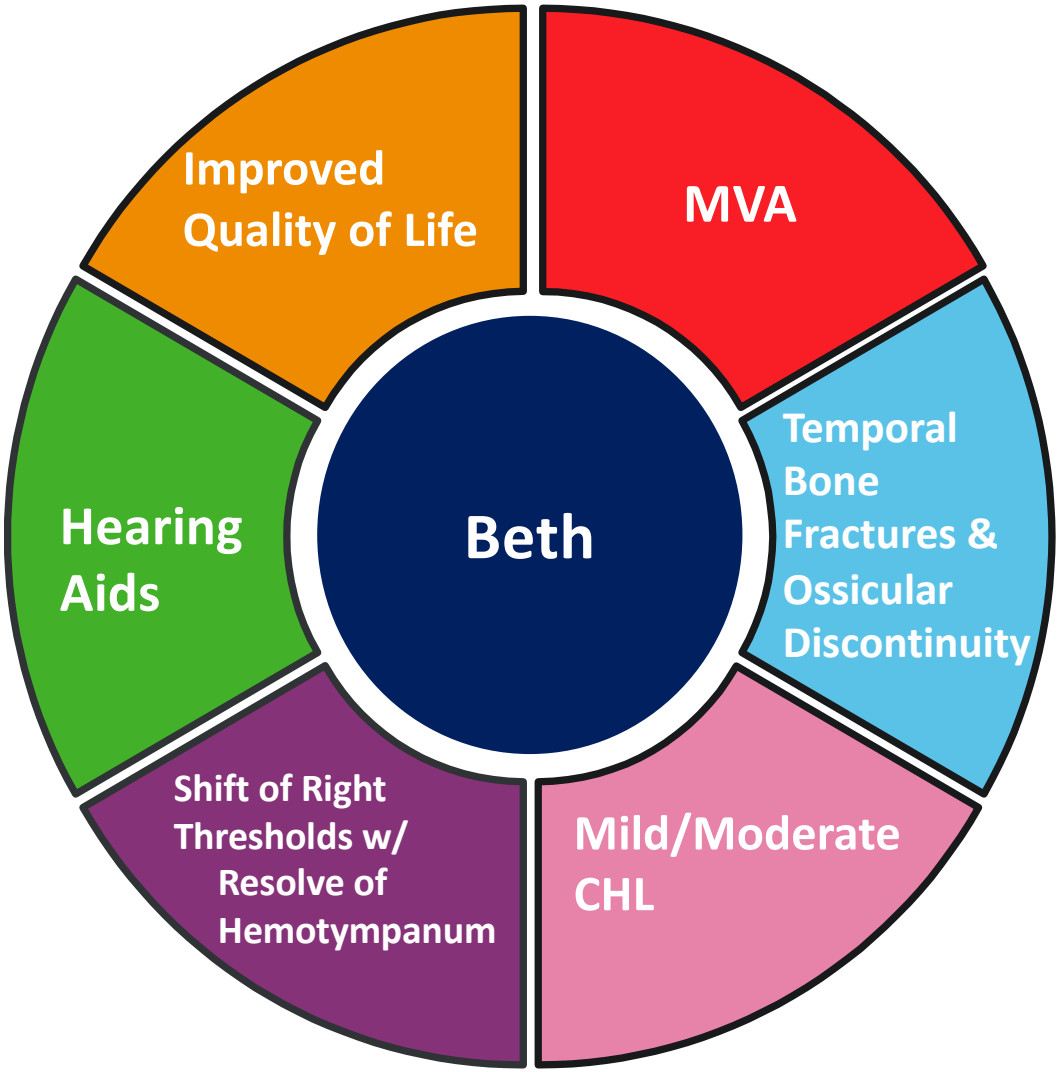
**Datalogging:**

| Date | Side | Hours Per Day |
|---|---|---|
| 05-15-2024 | right | 10.4 |
| 05-15-2024 | left | 11 |
| | | |

Image: Phoenix Children's Hospital

# Currently

- The patient and family are extremely satisfied with the improvement in quality of life with the hearing aids.


Improved Quality of Life
MVA
Temporal Bone Fractures & Ossicular Discontinuity
Mild/Moderate CHL
Shift of Right Thresholds w/ Resolve of Hemotympanum
Hearing Aids
Beth

# Vestibular Testing and Beside Assessment

Rachel Worcester, AuD, ABA-C

# What testing are we able to perform currently at PCH?

- cVEMP
- oVEMP
- Bedside screeners using Frenzel goggles

# What are VEMPs?

- The **vestibular evoked myogenic potential** (VEMP) is a neurophysiological assessment technique used to determine the function of the otolithic organs (utricle and saccule) of the inner ear. It complements the information provided by caloric testing and other forms of inner ear (vestibular apparatus) testing.
- Anatomy review:
  - Saccule is responsible for vertical acceleration in the vertical plane.
    - Going up or down in an elevator
  - Utricle is responsible for horizontal acceleration
    - Driving in a car
  - Once the otolith senses the motion, the brain sends the message to the reflex. The vestibulo-ocular reflex (VOR) is going to maintain visual acuity. The vestibule-spinal reflex (VSR) maintains posture and gait. The vestibulo-colic reflex (VCR) maintains head stability. When the brain receives all this information, it is able to determine what piece the body needs to keep it upright.

# How are VEMPs assessed?

The **cVEMP** tests **saccule and inferior nerve**. It is recorded from the sternocleidomastoid muscle (SCM). It is an inhibitory potential and an ipsilateral response. When a muscle is flexed, there is a split second within that flex where it releases. That provides the waveform or the response from the sound.

The **oVEMP** is primarily a **utricle and superior nerve** response. A little part of this oVEMP response comes from the saccule. This is an excitatory response that we record from the extraocular muscles. What generates that waveform is the person looking up; this a contralateral response. When we stimulate the right, we record the left and vice versa.

# Beside Testing

- Will be dependent on age and ability
- Screenings include:
  - Head thrust test
  - Single leg stance
  - Modified clinical test of sensory interaction on balance (M-CTSIB)
  - Dynamic visual acuity (DVA)
  - Gait observation
  - Tracking
  - ”Broke man's" rotary chair
  - Head righting reflex
  - Positionals
  - Dix-hallpike
  - Etc.

# Head Thrust Test

- Head is tilted 30 degrees downwards and high acceleration, unpredictable head thrusts are delivered to the horizontal canal
  - Cut off: seeing a corrective overt saccade

# Modified clinical test of sensory interaction on balance (MCTSIB)

- Ask child to maintain balance with their arms crossed over their chest for 30 seconds
  - 4 conditions
    - Stable surface, eyes open (Tests all three systems together)
    - Stable surface, eyes closed (Tests somatosensory system & vestibular system)
    - Neurocom foam, eyes open (Tests visual & vestibular system)
    - Neurocom foam, eyes closed (Tests vestibular system alone)
  - Maximum score is 120 seconds: cut off is 100 seconds (about 1 and a half minutes)

# Dynamic Visual Acuity (DVA)

- Dynamic test for the VOR
- Have patient stand 10 feet from Snellen chart or Lea eye chart
    - Obtain static vision acuity
    - Obtain dynamic vision acuity with headshake
    - Increase of greater than 2 lines is abnormal

# Gait Observation

- Have the patient walk in a straight line
  - Swaying and/or veering while walking
  - Falls
  - Abnormal movements with arms or legs

# Tracking

- Have patient follow your finger with their eyes only
  - Looking for nystagmus or abnormal eye movements

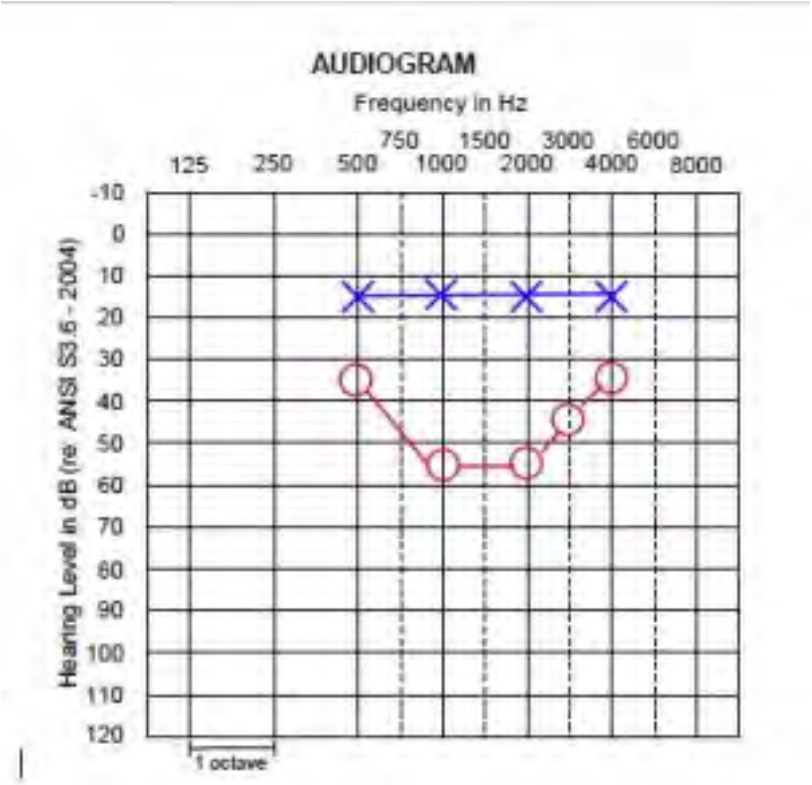
# Case Study

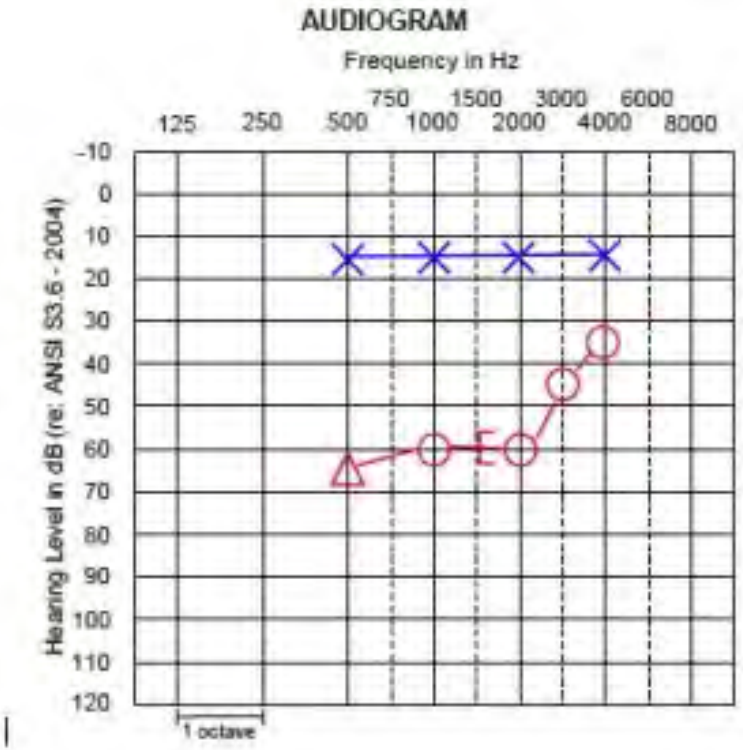
## Introducing Luna

# Luna's Case History

- 4-year-old female
- Medical history of epilepsy
- Initially referred to ENT following an MRI of the brain ordered for epileptic episodes that also revealed abnormal findings of the right inner ear that was found to be enlarged vestibular aqueduct (EVA)
- Failed her initial NBHS in the right ear
- Was seen for an outpatient ABR on 05/21/2019 which revealed normal hearing sensitivity, bilaterally.
- She was then seen for a hearing evaluation following her MRI on 02/12/2024 and 03/26/2024 where testing was suggestive of moderately-severe to moderate sensorineural hearing loss for the right ear and normal hearing sensitivity for the left ear
- Per her mother's report, she has always been clumsy and off-balance
- Gross motor delays in crawling and walking and now having difficulty learning to ride a bike

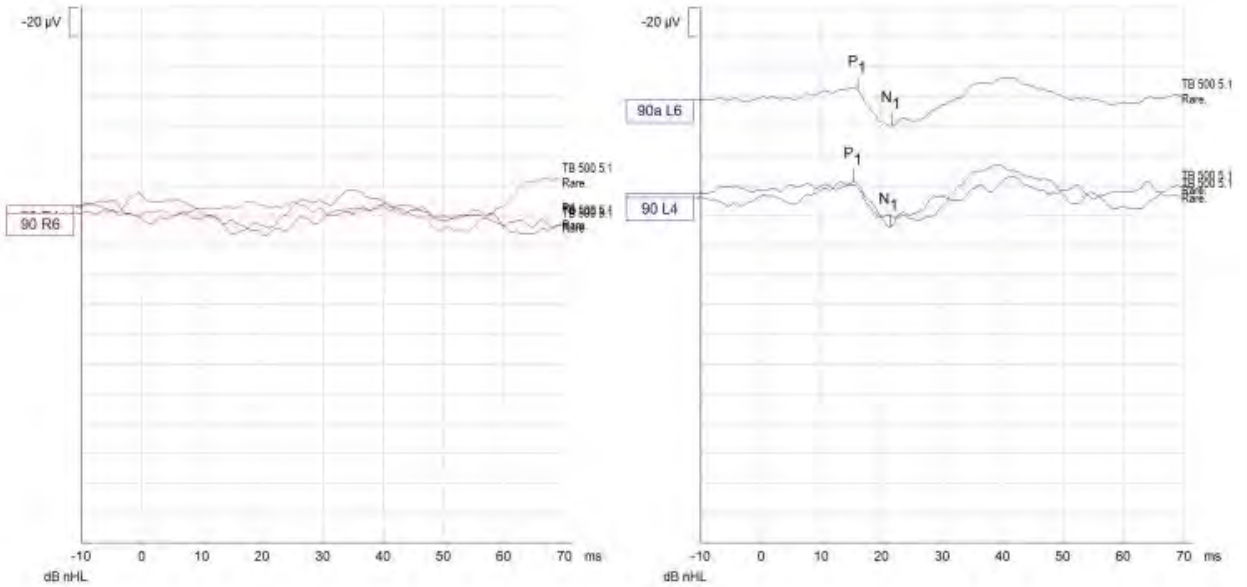
# Audiogram History




Images: Phoenix Children's Hospital

# cVEMP Testing




| Curve | Latencies (ms) | | | | | | |
|---|---|---|---|---|---|---|---|
| | P1 | N1 | P1' | N1' | N1-P1 Lat | N1-P1 Amp | (LA-SA)/(R+L) |
| 90 R | | | | | | | |
| 90 R1 | | | | | | | |
| 90 L3 | | | | | | | |
| 90 L4 | 15.33 | 21.33 | | | 6.00 | 31.15 | |
| 90a L6 | 16.00 | 21.67 | | | 5.67 | 24.98 | |
| 90 R6 | | | | | | | |

Image: Phoenix Children's Hospital

# Bedside Screening Results

Head Thrust Test: Positive - Overt catch up saccades were observed when her head was turned towards the right which is suggestive of abnormal SCC function. *Further testing will be completed when additional equipment (vHIT) is available to expand and confirm upon today's findings.*

Modified clinical test of sensory interaction on balance (M-CTSIB): Positive - Luna was able to maintain balance during conditions such as stable surface with/without eyes closed and on the foam pad with eyes open. She was only about to maintain balance for 20 seconds on foam pad in the eyes closed condition suggesting vestibular involvement.

Dynamic Visual Acuity (DVA): Could not complete due to patient compliance.

Gait Observation: Positive - On a flat surface, Luna was able to walk a majority of the time without stumbling, but appeared to be veering towards the right and did stumble a number of times. While running she fell towards the right side.

Tracking: Negative - No nystagmus was noted and Luna was able to follow the target with ease.

# Vestibular Results Summary

Abnormal cVEMP response for the right ear suggestive of abnormal saccule and inferior nerve function for the right ear

→

Abnormal HIT screening suggestive abnormal horizontal SCC function for the right ear

→

Abnormal M-CTSIB screening suggestive of vestibular weakness

↓

Abnormal gait and frequent falling

→

Results are most consistent with inferior vestibular nerve and/or saccule involvement in the right ear and peripheral vestibular dysfunction

# Next Steps for Luna

- Fit with a hearing aid and audiologic monitoring
- Vestibular rehabilitation with physical therapy team
- Continued vestibular monitoring

# Why screen the vestibular system in our pediatric patients?

| |
|---|
| **Emerging evidence suggests that 15–80% of children with hearing loss have concomitant vestibular impairment** |
| Cochlear implant users with vestibular dysfunction are 8x's more likely to have a device failure |
| Difficulty with reading |
| Delayed gross motor milestones |
| Individuals with EVA as more likely to have vestibular dysfunction which may lead to a progression in their hearing loss |

# Who Is At Risk for Vestibular Dysfunction?

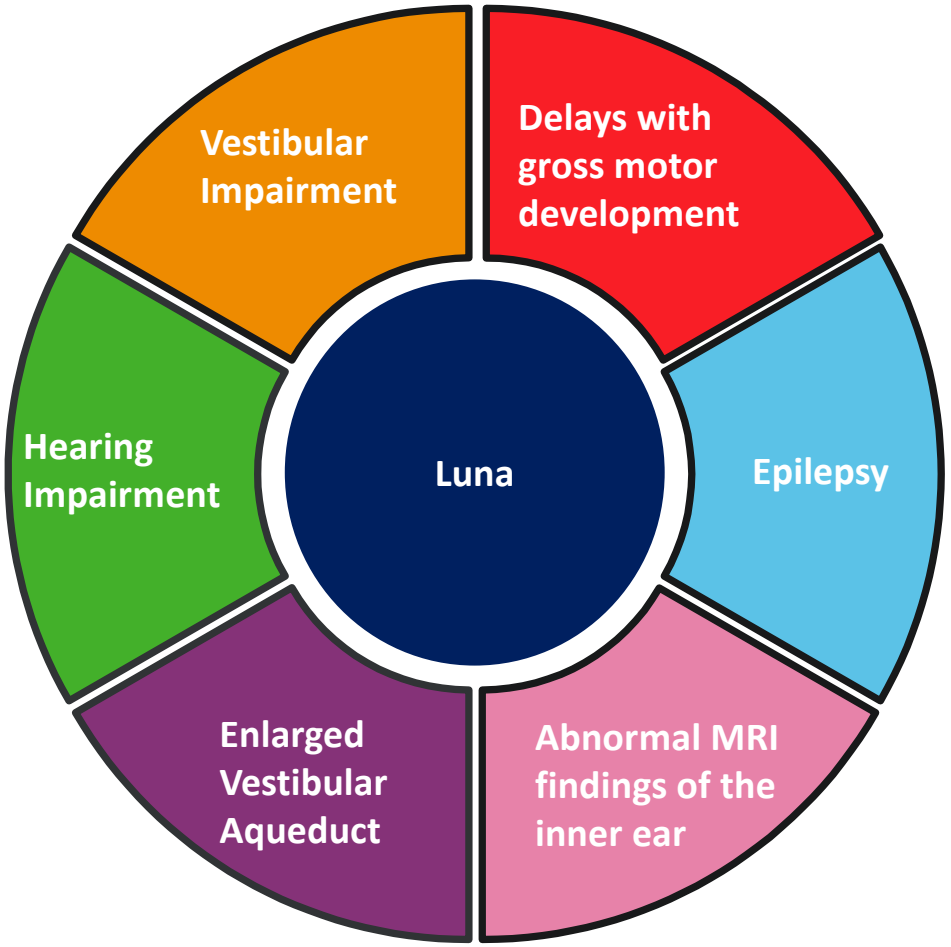
- Severe to profound hearing loss
- Cochlear implant post-op complication
- Otitis media
- Enlarged Vestibular Aqueduct (EVA)
- Migraines
- Labyrinth disorders
- Usher's syndrome
- Meningitis
- Ototoxicity
- Neuropathy
- Waardenburg's syndrome
- Rubella
- Congenital cytomegalovirus (cCMV)
- Anoxia
- Etc.…

# Common Presentations of Vestibular Loss

| | | | |
|---|---|---|---|
| Difficulty standing unsupported | Difficulty standing | Difficulty walking | Problem walking/ navigating in the dark |
| Clumsiness | Problems riding a bike | childhood activities such as playing sports, dancing, | Difficulty reading (skipping words or letters), visual acuity |
| Problems with schoolwork | Delayed gross motor control | Dizziness | Hearing loss |
| | Headaches (family history of migraines) | Torticollis in infancy | |

# Referral Guidelines

| |
|---|
| Children with hearing loss > 66 dB PTA should automatically be referred for testing |
| Children with hearing loss < 66 dB PTA should be referred when there are concerns for dizziness, imbalance, or delayed milestones (siting alone, crawling, walking, learning to ride a bike, etc.) |
| Cochlear implant patient pre- and post-op |
| Individuals with peripheral vestibular loss concerns |
| Inner ear anomalies such as EVA, SSCD, etc. |


Vestibular Impairment
Delays with gross motor development
Hearing Impairment
Luna
Epilepsy
Enlarged Vestibular Aqueduct
Abnormal MRI findings of the inner ear

# A Case of Suspected Branchiootorenal (BOR) Syndrome

Caroline Sabatino, AuD, CCC-A

# Branchiootorenal Spectrum Disorder (BORSD)

- Branchiootorenal (BOR) syndrome
- Branchiootic syndrome (BOS)
  - Two phenotypes only differ by the presence or absence of renal abnormality
- Hearing loss (>90%)

| Type | Severity | Progression |
|---|---|---|
| Mixed (52%) | Mild (27%) | Non-progressive (~70%) |
| Conductive (33%) | Moderate (22%) | Progressive (~30%) |
| Sensorineural (29%) | Severe (33%) | |
| | Profound (16%) | |

| Pinnae | EAC | Middle Ear | Inner Ear |
|---|---|---|---|
| Preauricular pits (82%) | Atresia or stenosis (29%) | Malformation, malposition, dislocation, or fixation of the ossicles | Cochlear hypoplasia |
| Lop ear malformation (36%) | | Reduction in size or malformation of the middle ear space | Enlargement of the cochlear and vestibular aqueducts |
| Preauricular tags (13%) | | | Hypoplasia of the lateral semicircular canal |

# Diagnostic Criteria

- Major and minor diagnostic criteria for branchiootorenal spectrum disorder (BORSD)

| Major Criteria | Minor Criteria |
|---|---|
| Second branchial arch anomalies | External auditory canal anomalies |
| Deafness | Middle ear anomalies |
| Preauricular pits | Inner ear anomalies |
| Auricular malformation | Preauricular tags |
| Renal anomalies | Other: facial asymmetry, palate abnormalities |

# Introducing Marcus

- Referral
- 5 year 4-month-old male
  - Failed OAE
  - Ear pits
  - Autism
  - Possible genetic disorder

# Initial History

- Accompanied by foster mother
- Birth history: unknown
- Medical records from pediatrician
  - Born deaf in right ear, never used hearing aid
  - Autism
- Speech: unclear, uses some sign language
- School: enrolled in Kindergarten
- History of ear infections: yes

# Initial Testing

- Otoscopy: Preauricular pit, ear tag, dull TM
- Tympanometry: Type B, Au
- DPOAE: Absent 750 – 8,000 Hz
- Behavioral testing:
  - Right: Profound SNHL
  - Left: Moderate to moderately-severe mixed hearing loss
- Recommendations
  - ENT consultation
  - Return for re-evaluation and to discuss amplification

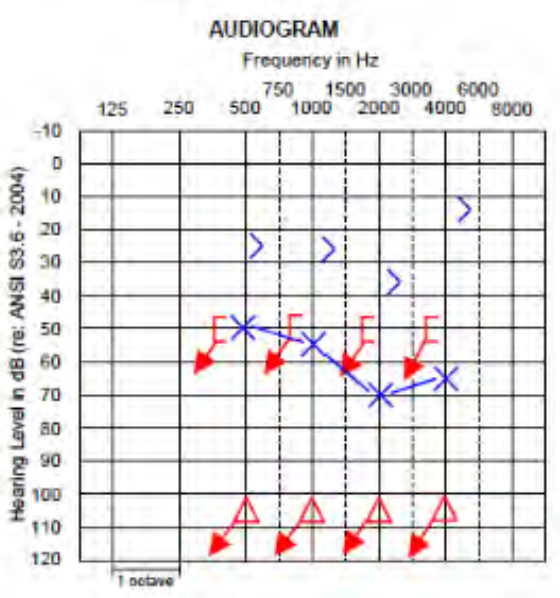



Image: Phoenix Children's Hospital

# ENT

- Bilateral branchial anomalies
  - Drainage from neck pits
- Exam concerning for branchiootoic syndrome (BOS)
- Orders
  - CT/MRI
  - Renal/Neck Ultrasound
- Referrals
  - Genetics
  - Speech
  - Phoenix Children's PCP
  - Fellow ENT
- Cleared for hearing aid use ASAP

# October/November 2023

- Renal Ultrasound
  - Bilateral small for age kidneys with no hydronephrosis or focal renal lesion
- Neck Ultrasound
  - Thyroglossal duct cyst
- MRI
  - Morphology of the cochlea and inner ears is suggestive of branchiootorenal (BOR) syndrome
  - Absent right cochlear aperture and right cochlear nerve
  - Effusions in both mastoids and middle ears
  - Microcephaly
- CT Temporal Bone
  - Irregular appearance of the incus, Au
  - Posterior semicircular canals are not seen, Au

# Repeat Testing and HA Consult

- Transferring to Phoenix Day School for the Deaf
- Repeat testing
  - Tymps: Type B, Au
  - Right: Profound SNHL
  - Left: Moderate to moderately-severe mixed hearing loss
- Consult for left hearing aid
  - Traditional behind-the-ear (BTE) hearing aid
  - Bone anchored hearing device (BAHD)

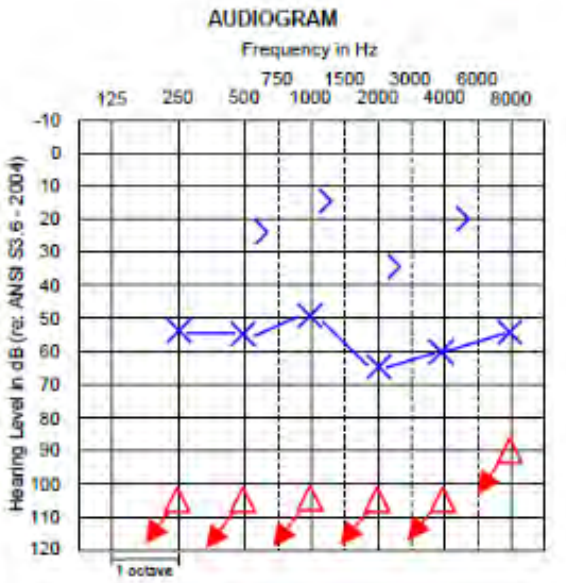



Image: Phoenix Children's Hospital

January 2024

# Hearing Aid Fitting

- BTE hearing aid in the left ear
- Marcus liked the hearing aid
  - Asked for HA for his right ear
- We discussed fitting a Bi-CROS

# Hearing Aid Follow-Up

- Patient chewed the earmold on the drive home from fitting appointment
- Patient chewed the earmold at school and broke it
  - Brainstormed solutions
- Earmold was torn in two locations
  - Hearing aid unusable until he gets a replacement earmold

# Continued Care

- ENT placed ear tubes
  - Marcus was not currently utilizing hearing aid
- Post-operative testing
  - Mild sensorineural hearing loss in the left ear
  - Adjusted the hearing aid
  - Fit with replacement earmold

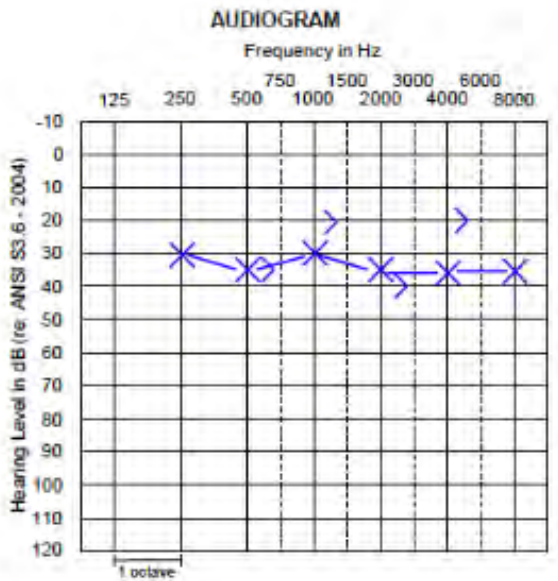
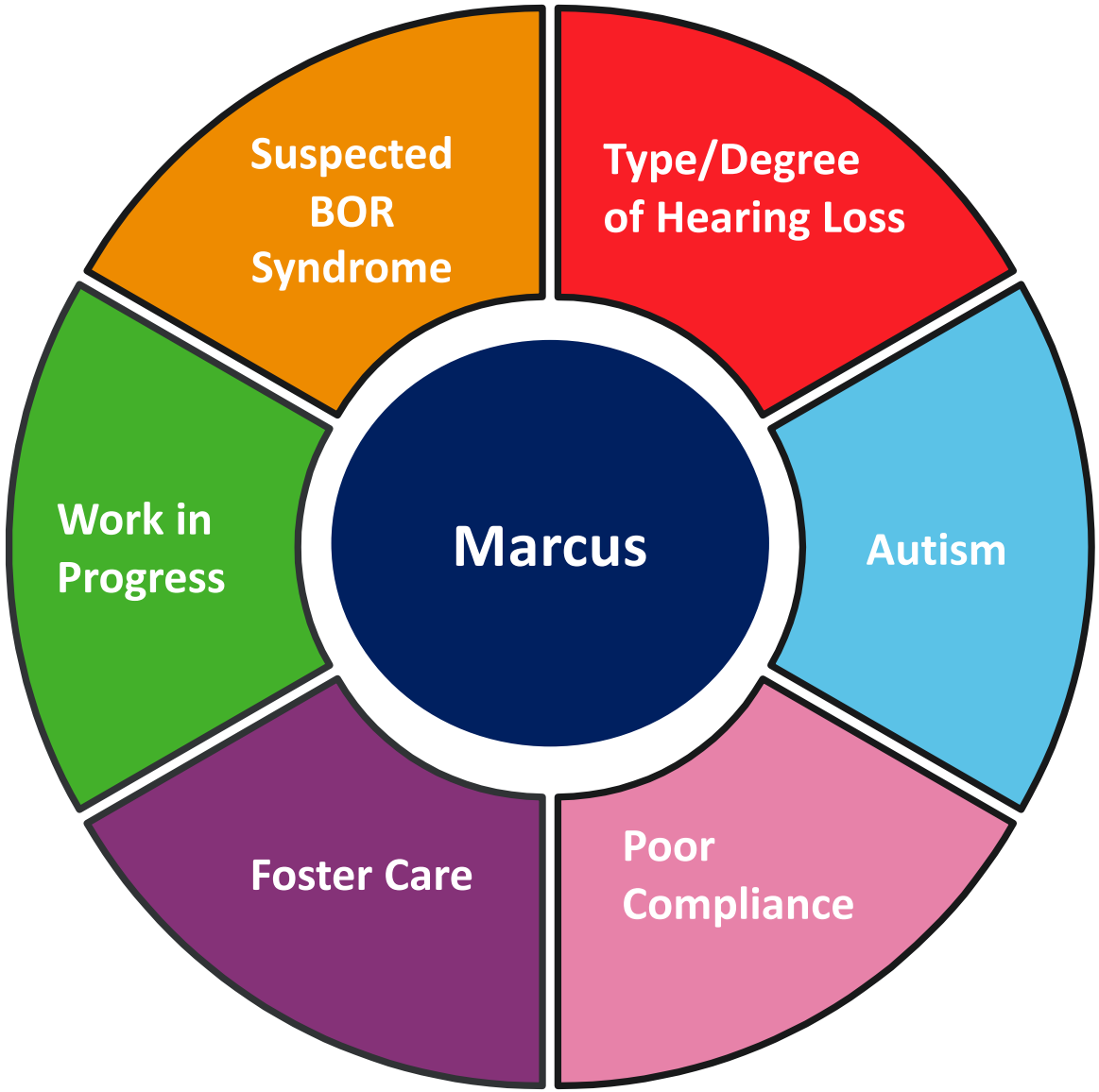



Image: Phoenix Children's Hospital


Suspected BOR Syndrome
Type/Degree of Hearing Loss
Work in Progress
Marcus
Autism
Foster Care
Poor Compliance

# Which Side Are You On?

## A Tricky ABR Test Involving Mixed Hearing Loss

Robert Fanning, AuD, CCC-A

# My Role

- Collaborator
  - We all often participate in the diagnosis and treatment of others' patients due to schedule limitations and our availability at certain locations
  - I am not Judy's managing audiologist, but I have performed two of her ABR tests (4yo; 6yo)
  - My work will provide Judy's managing audiologist the information she needs to effectively care for her

# Meet Judy

- 6 year 10-month-old female
- Trisomy 21
- 2q and 13q deletion
- Autism
- Global developmental delay
  - Nonverbal
  - Some signs
  - AAC device
- Chronic otitis media with effusion
  - Bilateral ear tube placement at age 8 months and replacements at ages 2 and 4 years

# Meet Judy

- Failed UNHS
- ABR test at age 8 months following bilateral placement of ear tubes:
  - Right ear: Mild-to-moderate, primarily sensorineural hearing loss
  - Left ear: Moderate, primarily sensorineural hearing loss
- Results from subsequent ABR tests in early childhood were generally confirmatory
- She does not comply or condition for behavioral audiological evaluations
  - We have mostly relied on ABR tests

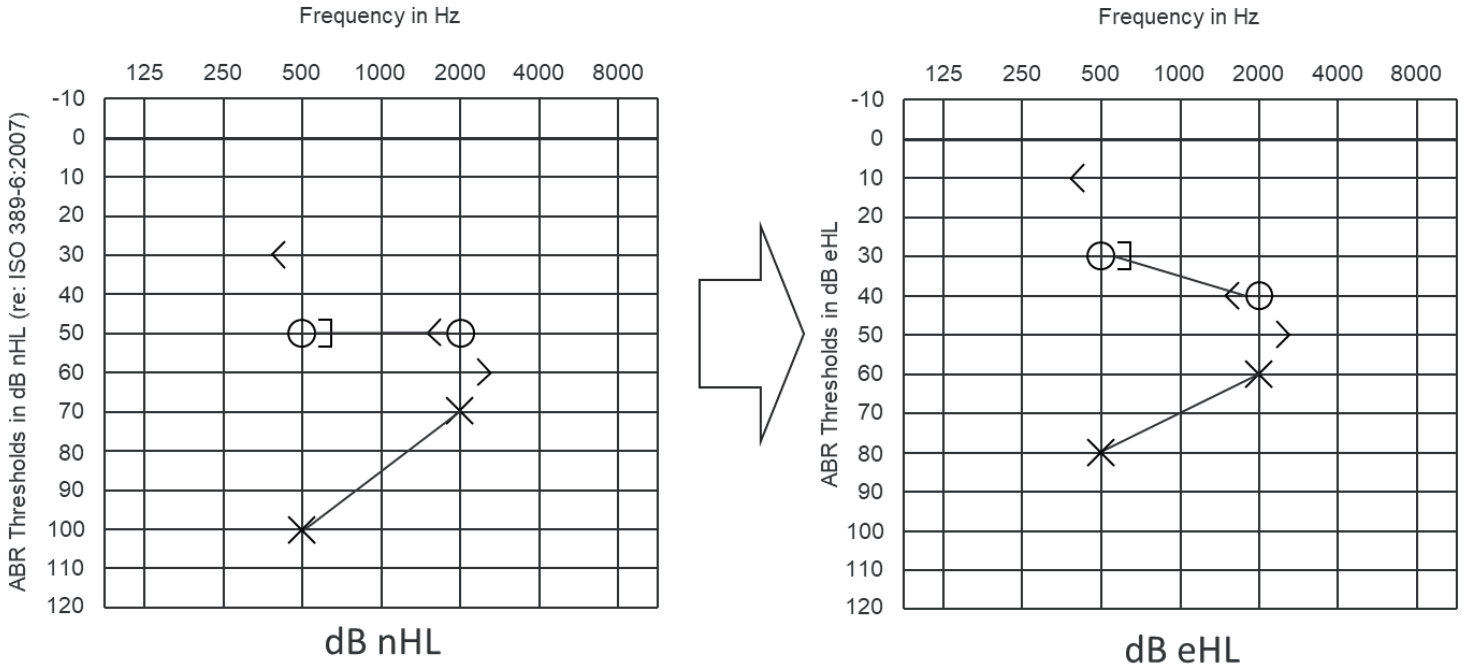
# Meet Judy

- Fit with BTE hearing aids around the age of 10 months at another facility but did not tolerate them
- Fit with one and then two powerful BAHD with a soft band here at the age of 2 years
  - Initially accepted them and appeared to benefit from them
  - Rejected them around the age of 4 years
- Trying traditional hearing aids again since last year with some success

# Review of ABR Concepts: Laterality

- Excluding the non-test ear in ABR studies is just as important as it is in clinical audiometry
- Appreciating differences between ipsilateral and contralateral recordings can help determine laterality of the ABR (i.e., test ear vs. non-test ear) (Stapells & Ruben, 1989)
  - Wave V tends to be later and smaller in contralateral recordings than in ipsilateral ones
- These differences can be especially helpful in cases when masking the non-test ear is not possible
  - Bilateral microtia/atresia
  - Masking dilemmas

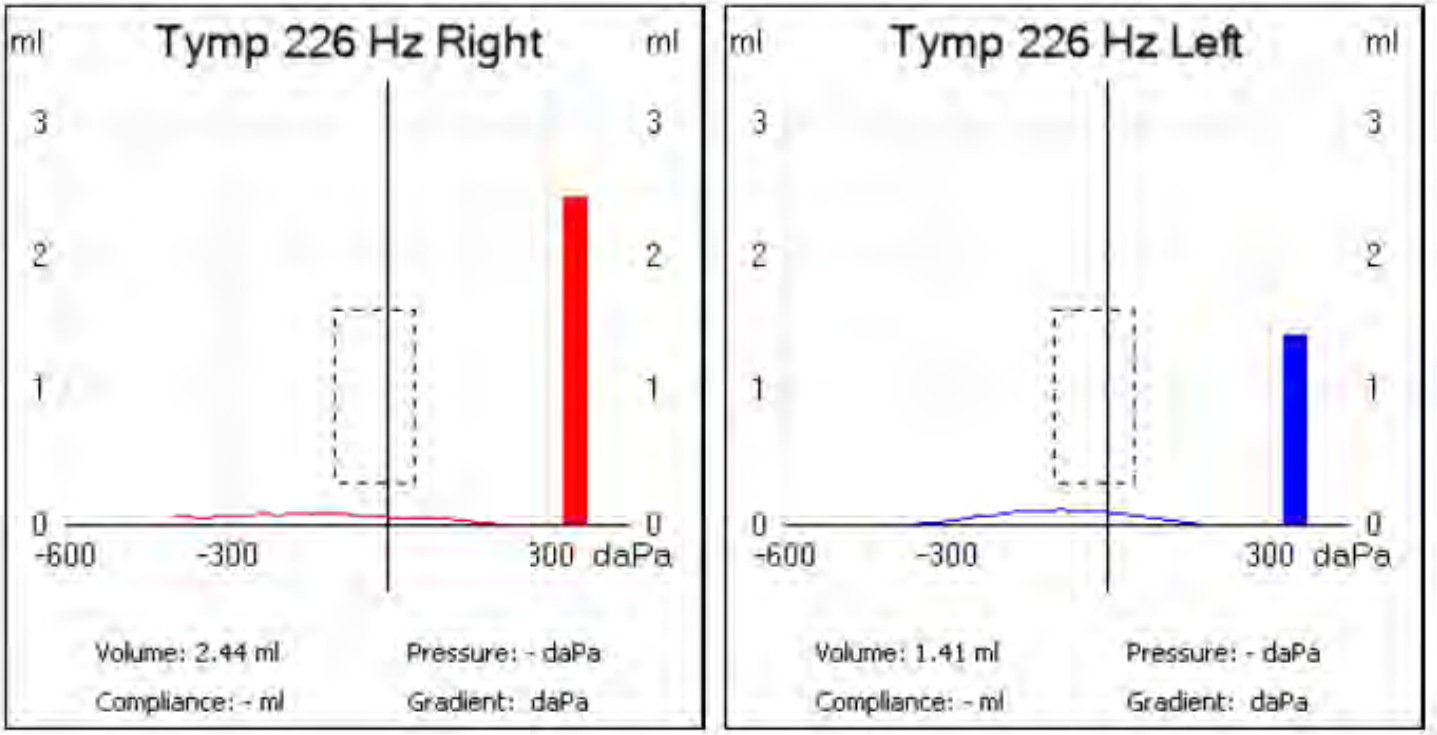
# Results from Judy's Test at 4 Years Old




- Previous tone-burst ABR test at age 4 years: Bilateral, asymmetrical, mixed hearing loss.
- dB nHL converted to dB eHL (Stevens et al, 2013)

Images: Phoenix Children's Hospital

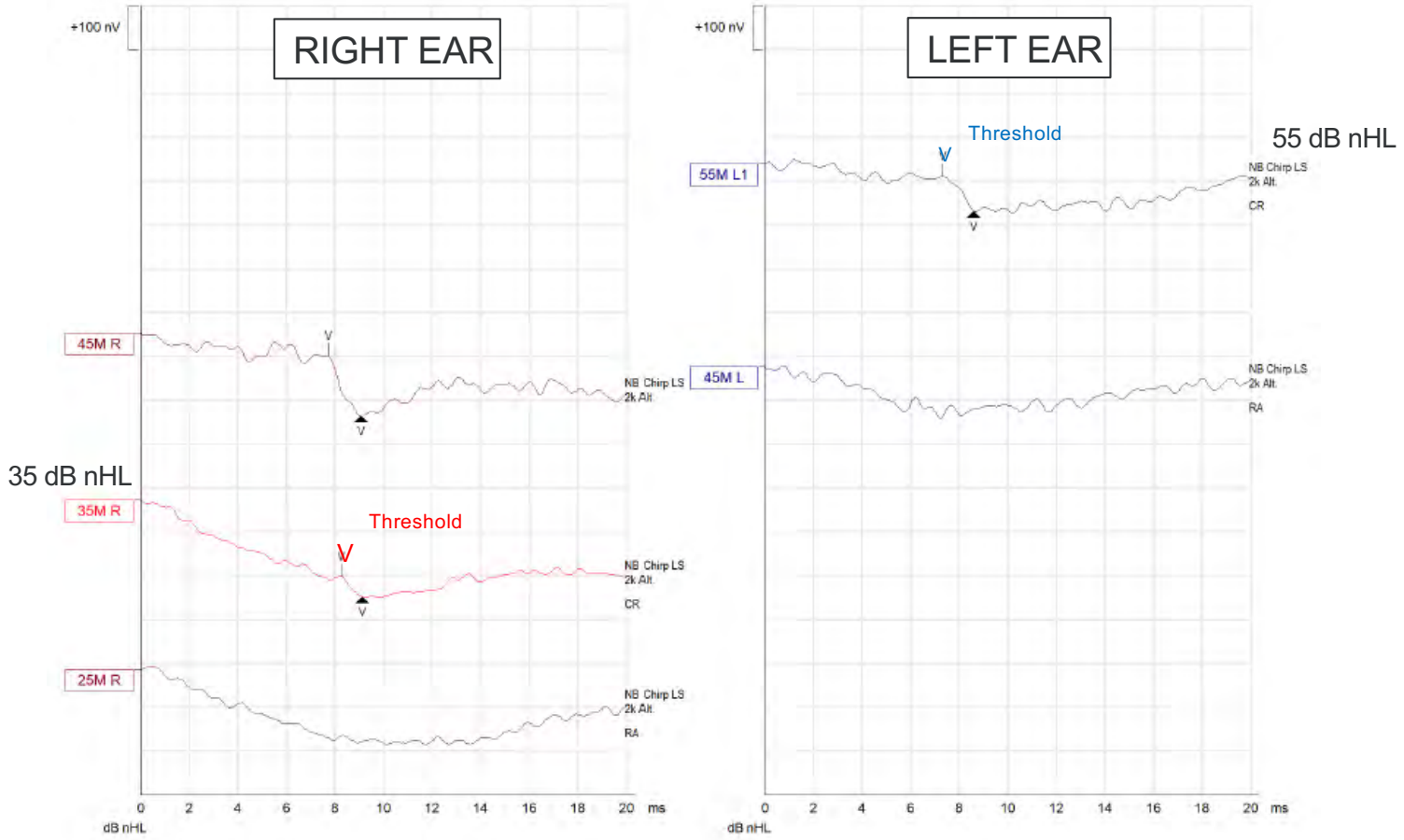
# Evaluation at 6 Years Old




- Otoscopy: bilateral T-tubes in place
- Tympanometry: large volumes

Image: Phoenix Children's Hospital

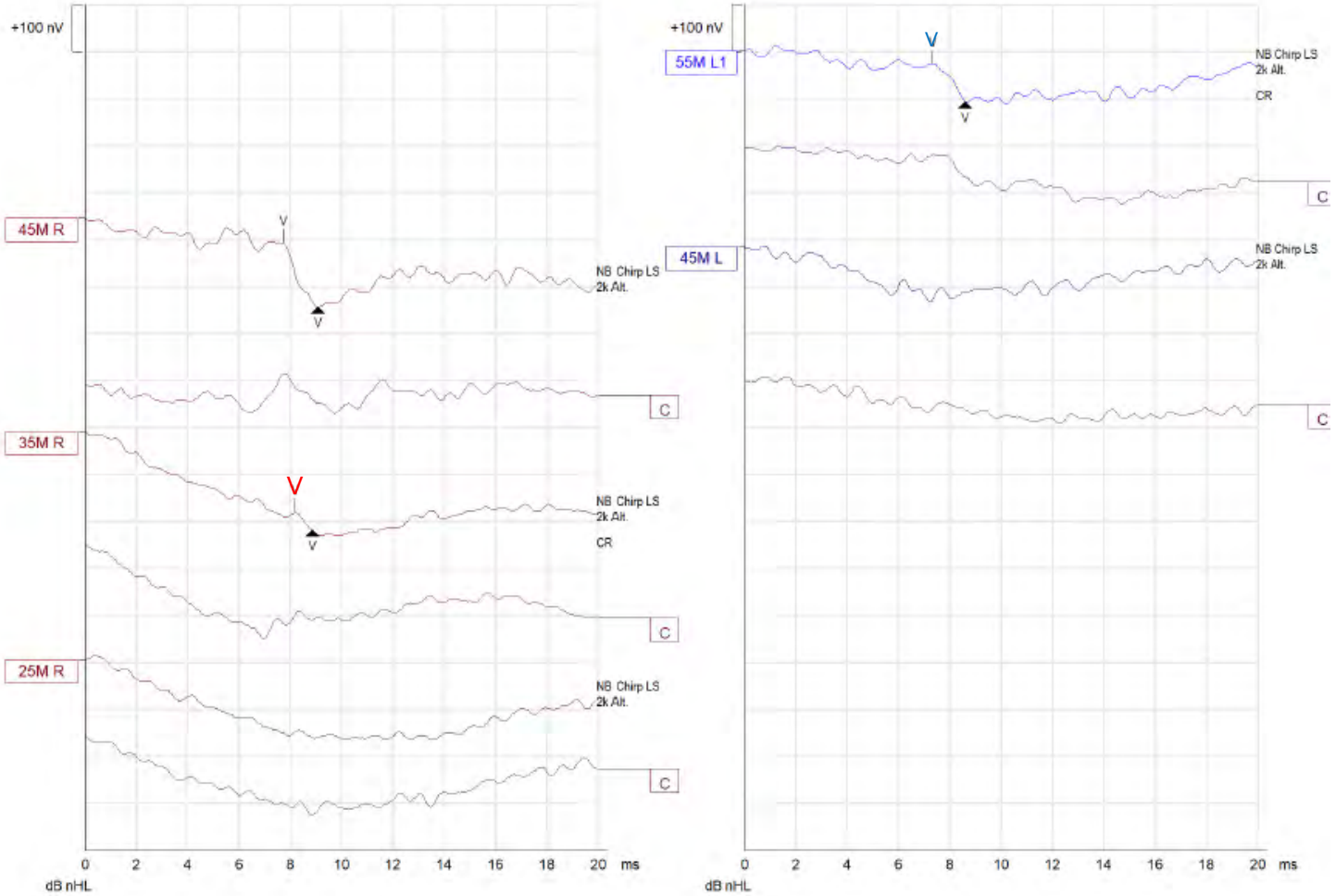
# ABR test: AC 2000 Hz




ABR thresholds to air-conducted narrow-band CE Chirps at 2,000 Hz, ipsilateral recordings only.

Image: Phoenix Children's Hospital

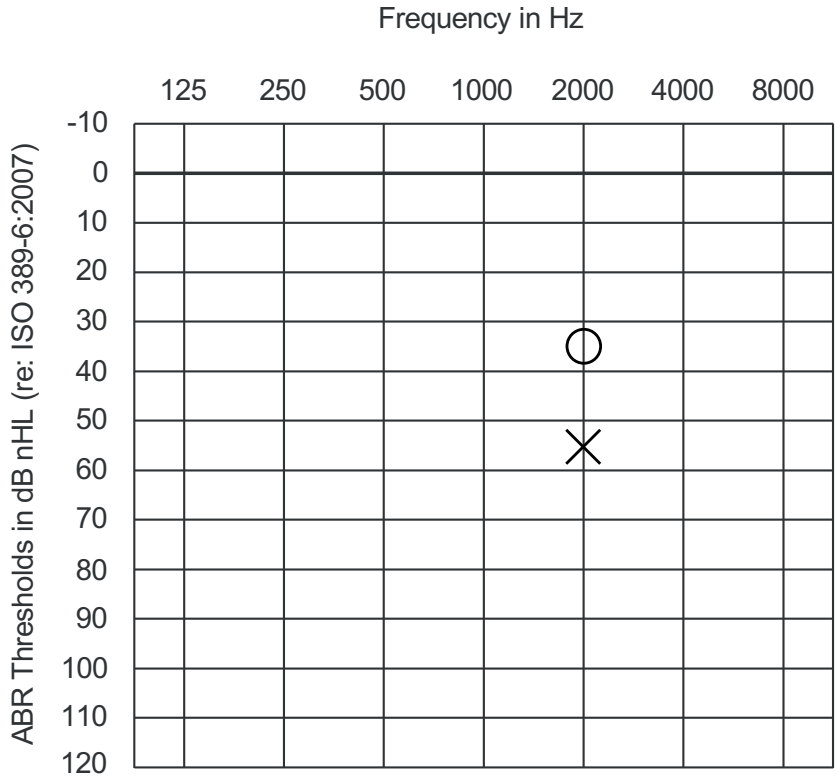
# ABR test: AC 2000 Hz, continued




Comparing the ipsilateral and contralateral channels.
Wave V in the contralateral channels is a little later and smaller.

Image: Phoenix Children's Hospital

# Plotting the Audiogram




ABR thresholds test at 2,000 Hz in dB nHL

Image: Phoenix Children's Hospital

# ABR test: AC 500 Hz

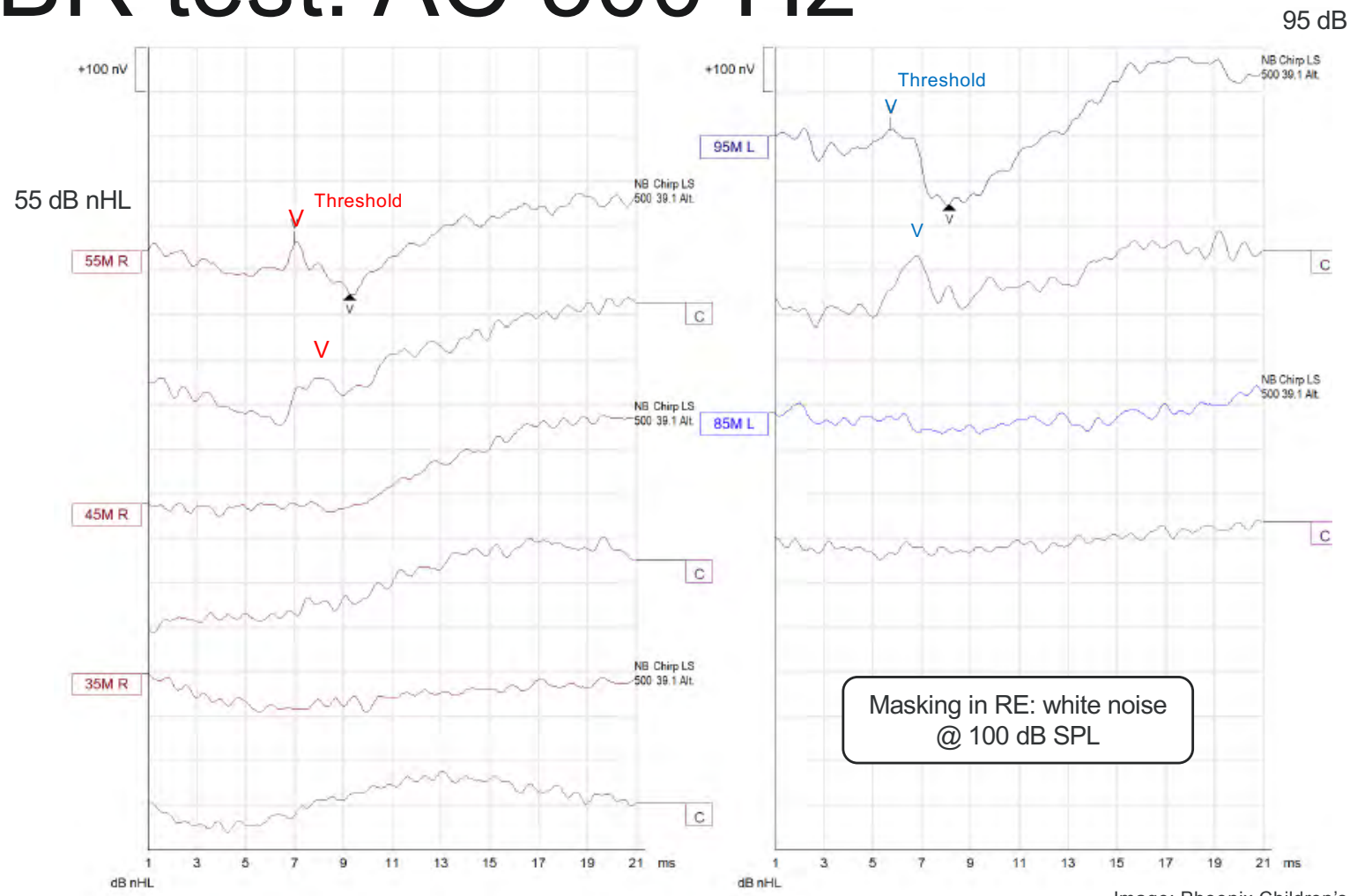
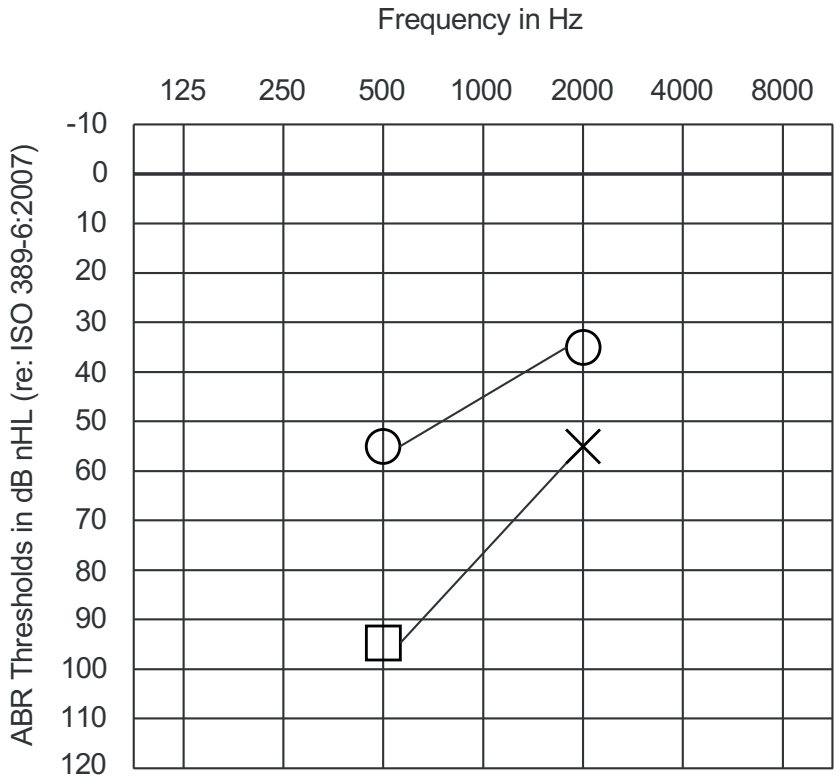



Image: Phoenix Children's Hospital

ABR thresholds to air-conducted narrow-band CE Chirps a 500 Hz.
Wave V in the contralateral channels is a little later and smaller.

# Plotting the Audiogram



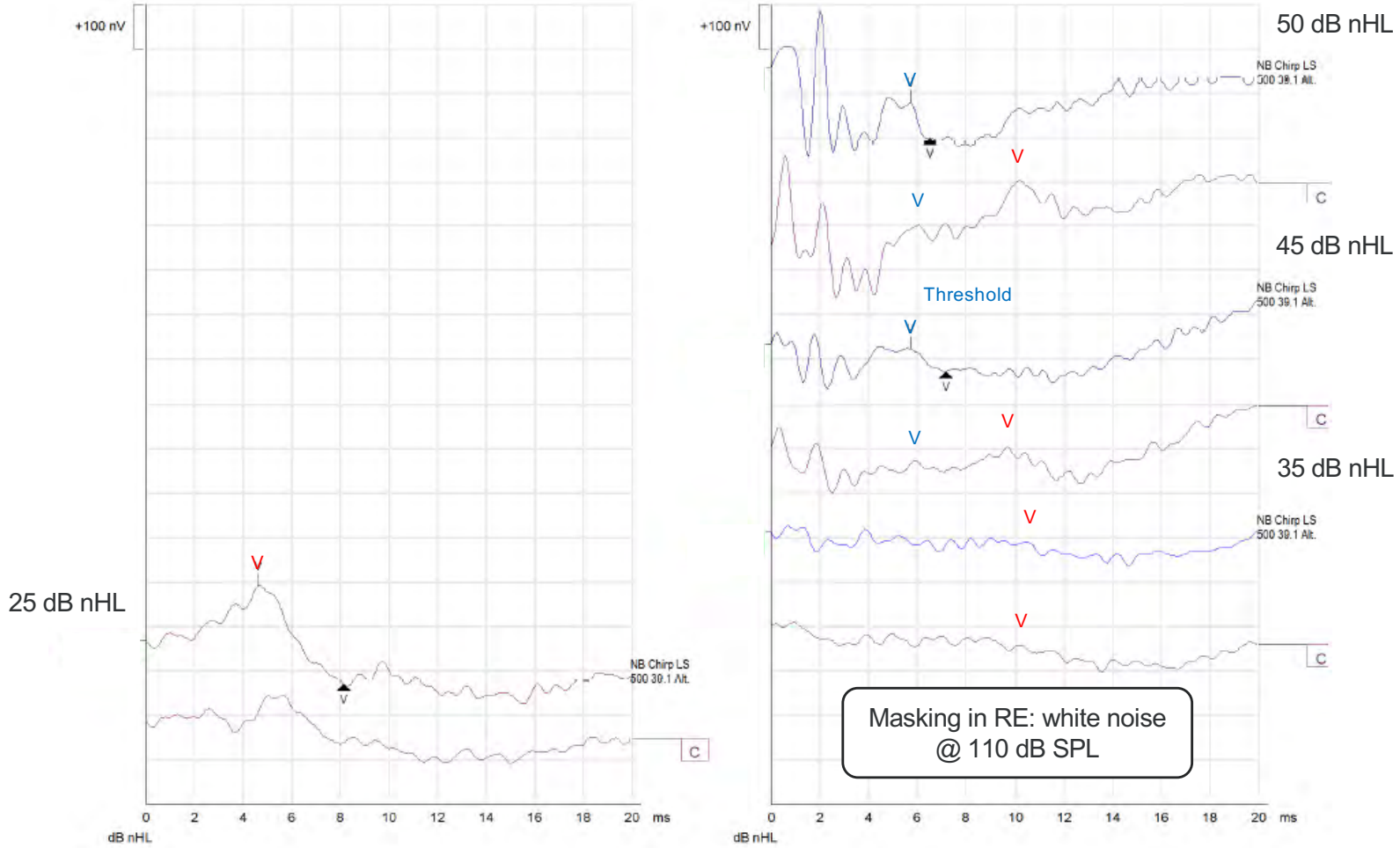

ABR thresholds test at 500 & 2,000 Hz in dB nHL

Image: Phoenix Children's Hospital

# ABR test: BC 500 Hz

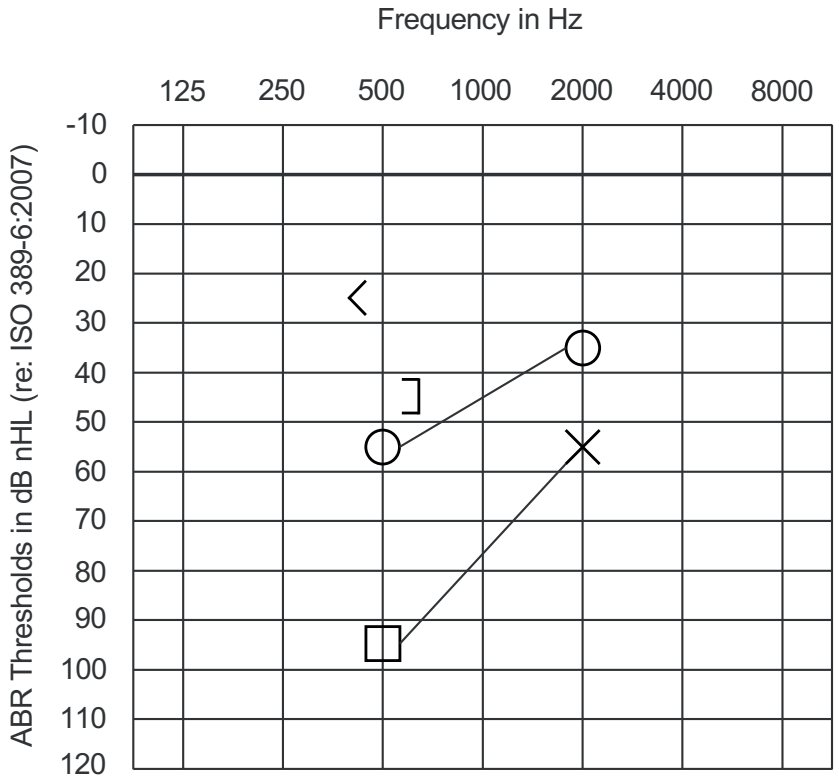

ABR thresholds to bone-conducted narrow-band CE Chirps a 500 Hz.
RE unmasked. Stimulus to LE is also crossing over to RE.

# ABR test: BC 500 Hz



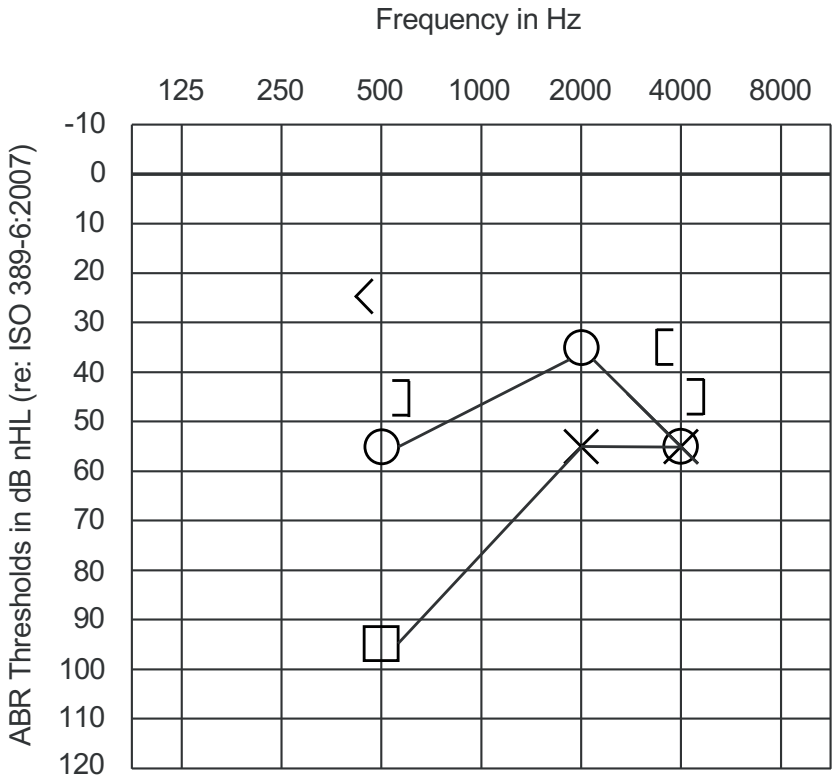

ABR thresholds to bone-conducted narrow-band CE Chirps a 500 Hz.
RE masked.

# Plotting the Audiogram




All thresholds in dB nHL

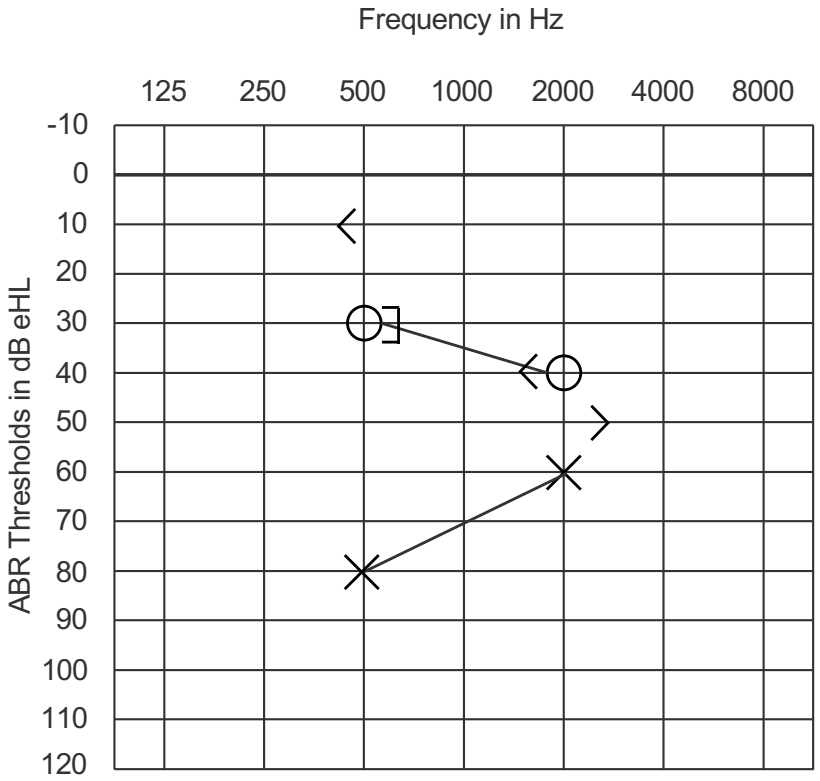
# Final Audiogram



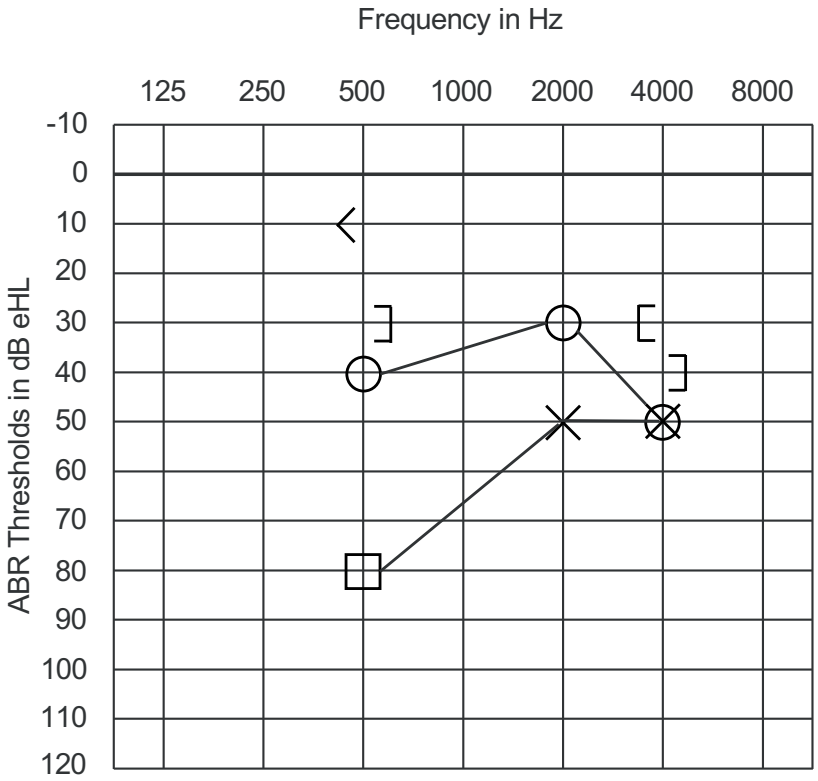

ABR thresholds for narrow band CE Chirp stimuli (dB nHL)

Image: Phoenix Children's Hospital

## 4 years old

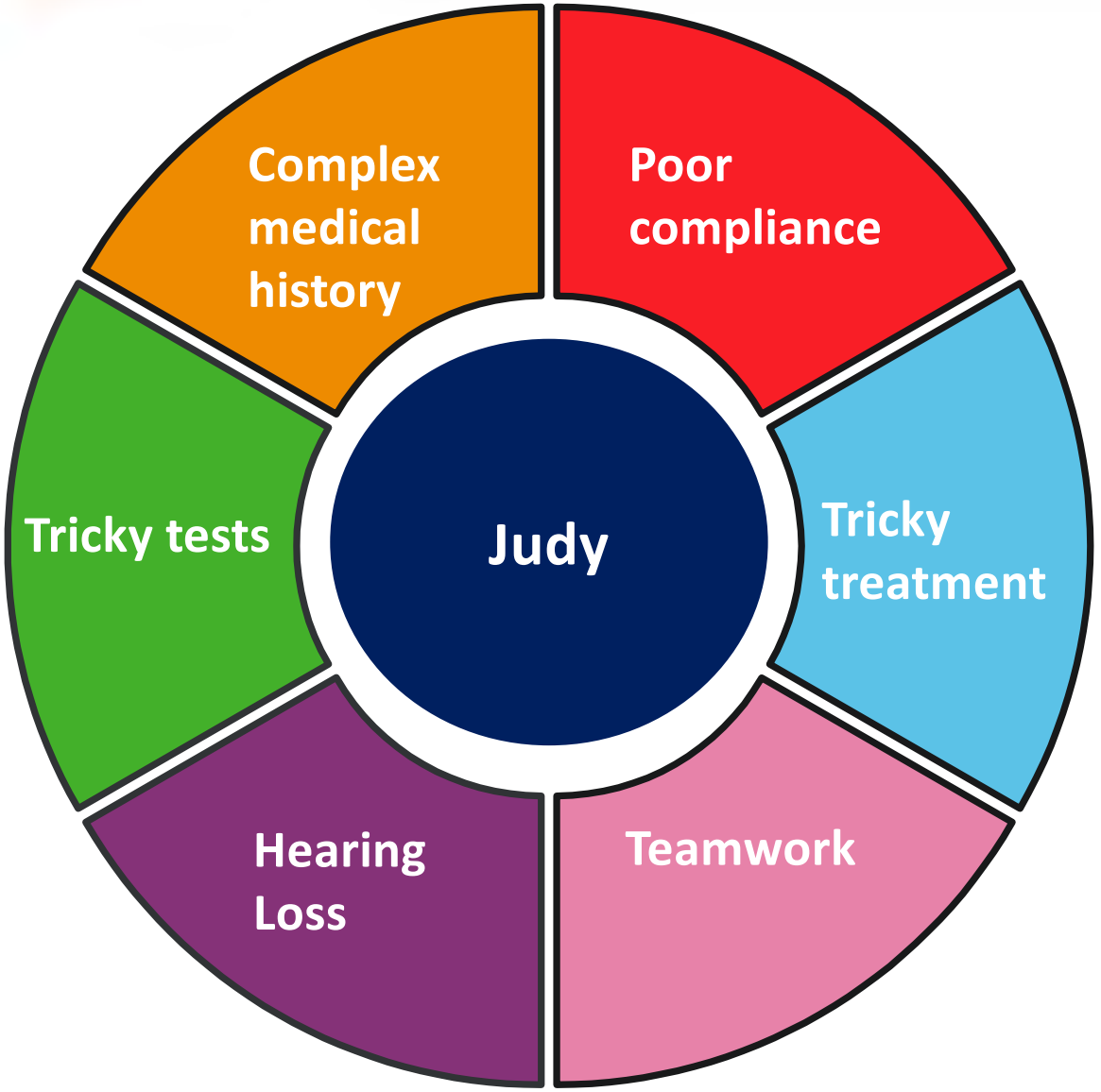



## 6 years old




Comparison of old and new audiograms.
ABR thresholds plotted in dB eHL (Stevens et al, 2013)

Images: Phoenix Children's Hospital


Complex medical history
Poor compliance
Tricky tests
Judy
Tricky treatment
Hearing Loss
Teamwork

# Questions? Please put any questions in the Q&A or send an email after the presentation

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