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Phoenix Children's

Case Studies in Pediatric Audiology: Wins Worth Sharing, in partnership with Phoenix Children's Hospital

Tanner Robinson, AuD
Robert Fanning, AuD
Alissa Nickerson, AuD
Ashley Geske, AuD
Allie Sayer, AuD
Wendy Steuerwald, AuD

Tanner Robinson, AuD, CCC-A

Tanner Robinson, AUD, CCC-A is a clinical audiologist at Phoenix Children's Hospital (PCH) specializing in the diagnosis and management of pediatric hearing loss and auditory processing disorder. He graduated from Idaho State University with a Bachelor's in Communication Sciences and Disorders and from Utah State University with a Doctorate of Audiology and special emphasis in Listening and Spoken Language (LSL). Prior to joining Phoenix Children's Hospital in July 2023, he completed his 4th year externship at LeBonheur Children's Hospital in Memphis, TN. Tanner has special interest in the early diagnosis of hearing loss in infants and supporting parent's through their journey with their children. He is also the lead audiologist of the PCH Auditory Processing Disorder Clinic.

Robert 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.

Alissa Nickerson, AuD, CCC-A

Alissa Nickerson, AuD, CCC-A, is a clinical audiologist at Phoenix Children's specializing in the diagnosis and management of childhood hearing loss. Alissa enjoys working with children of all ages and has a special interest in pediatric amplification. She is a collaborating member of "Stop CMV AZ," an organization which works to educate and advocate for those impacted by CMV in the state of Arizona. Prior to joining the staff at Phoenix Children's, Alissa was employed in private practice in Chicago, Illinois. She has publications in the areas of post-baccalaureate audiology training and amplification profiles of pediatric cochlear implant users.

Ashley Geske, AuD, CCC-A

Ashley Geske, AuD, CCC-A graduated from Arizona State University with a Bachelor of Science in Speech and Hearing Sciences and a clinical Doctorate of Audiology. Prior to joining Phoenix Children's Hospital in August of 2022, she worked at Providence Speech and Hearing Center partnered with the Children's Hospital of Orange County where she worked with pediatric patients in both outpatient and inpatient hospital settings. Her clinical areas of specialty include diagnostics, hearing aids, and cochlear implants.

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.

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..

Disclosures

- **Presenter Disclosure**: Financial: Tanner Robinson is an employee of Phoenix Children's Hospital. Non-financial: Tanner Robinson has no non-financial disclosures. Financial: Robert Fanning is employed by Phoenix Children's Hospital. Non-financial: Robert Fanning has no relevant non-financial relationships to disclose. Financial: Alissa Nickerson is employed by Phoenix Children's Hospital. Non-financial: Alissa Nickerson has no relevant non-financial relationships to disclose. Financial: Ashley Geske is employed by Phoenix Children's Hospital. Non-financial: Ashley Geske has no relevant non-financial relationships to disclose. Financial: Allie Sayer is employed by Phoenix Children's Hospital. Non-financial: Allie Sayer 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.
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- The applicability and accuracy of the course content may vary by the clinical, geographical, and cultural context; you should evaluate the course accordingly.
- As healthcare is rapidly evolving, new findings may change the validity of the information presented in this course. Providers should always consult the latest research and guidelines from professional associations.
- The interventions discussed in this course may be limited in generalizability, requiring practitioners to consider specific individual and population factors.
- The course may not cover all possible risk factors, diagnostic methods, or treatment options, and complex cases may require additional considerations.
- Healthcare providers should apply cultural competence and sensitivity to ensure effective and respectful care based on their patients' diverse needs.
- Providers should always evaluate the limitations of diagnostic and screening tools, considering their potential for false results as well as any ethical implications.
- It is the responsibility of course participants to critically evaluate the evidence from multiple sources to guide professional practice.

Learning Outcomes

After this course, participants will be able to:

- Discuss five complex audiology case studies.
- Explain the importance of patient counseling and education.
- Describe common ramifications of cCMV seen in pediatric audiology patients.

continuED

Phoenix Children's

About us

- PCH is the only free-standing children's hospital in Arizona.
- Our audiology team is comprised of 16 audiologists.
- Our newborn hearing screening team has 24 members.
- Complicated patients from the US Southwest and International patients.
- You will hear thought-provoking case studies from 5 of the audiologists today.

Uncomfortable Counseling Moments

Tanner Robinson, AuD, CCC-A

Case Objectives

- Review a case involving uncomfortable counseling
- Explore potential uncomfortable counseling moments
- Review counseling skills that may help

Trevor's Case History

- 2 months old
- Born at 37 weeks to pregnancy complicated by maternal hypertension
- No NICU, no jaundice
- Maternal side grandfather has unilateral hearing loss from childhood, reportedly due to ear infections
- Referred on AABR once inpatient, twice outpatient

Trevor's Case History (cont.)

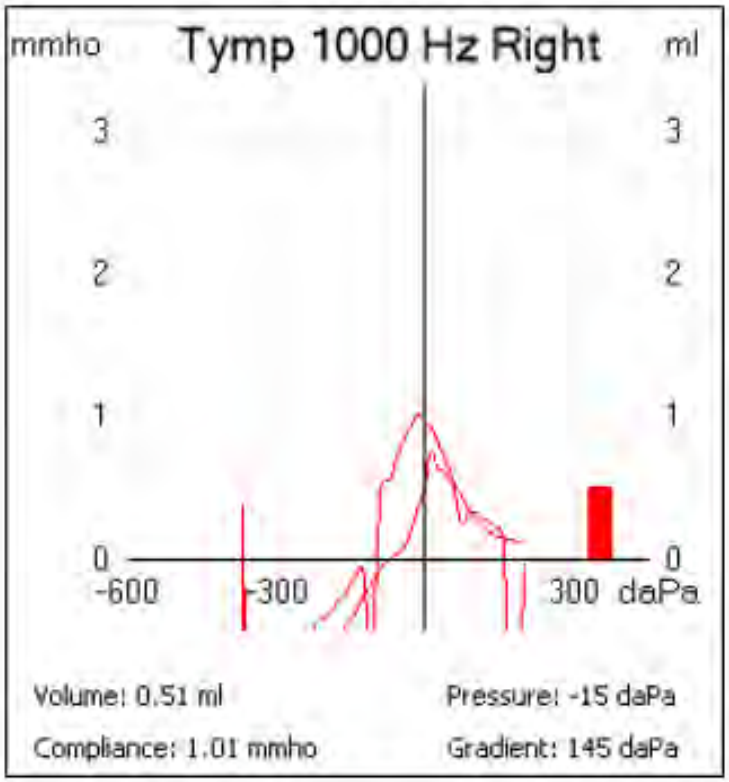
- Diagnostic BAER/ABR testing completed at another facility 5 weeks prior to visit
  - Normal 1000 Hz tympanometry
  - Absent DPOAEs for both ears
  - Mildly elevated thresholds in the right ear, normal thresholds for left ear
    - Bone conduction completed for right ear and results suggest conductive component
- Referred by pediatrician for repeat testing and PCH had earlier availability

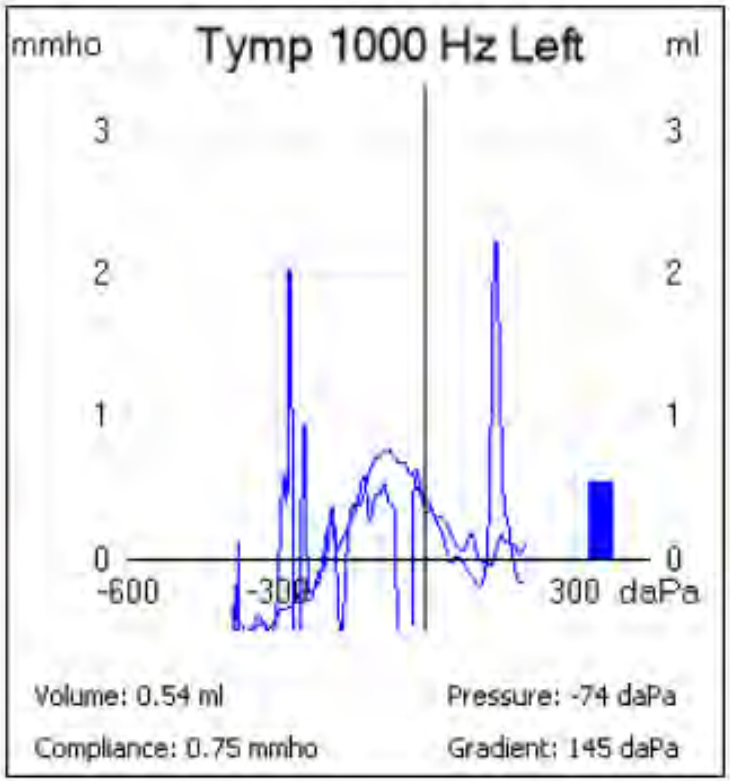
Family's Report

- Family reported they were told hearing was normal for both ears
- Pediatrician visit/additional referral
- Provides good eye contact, does not seem to startle to loud sounds
  - Vacuum, door slams, etc.

Results

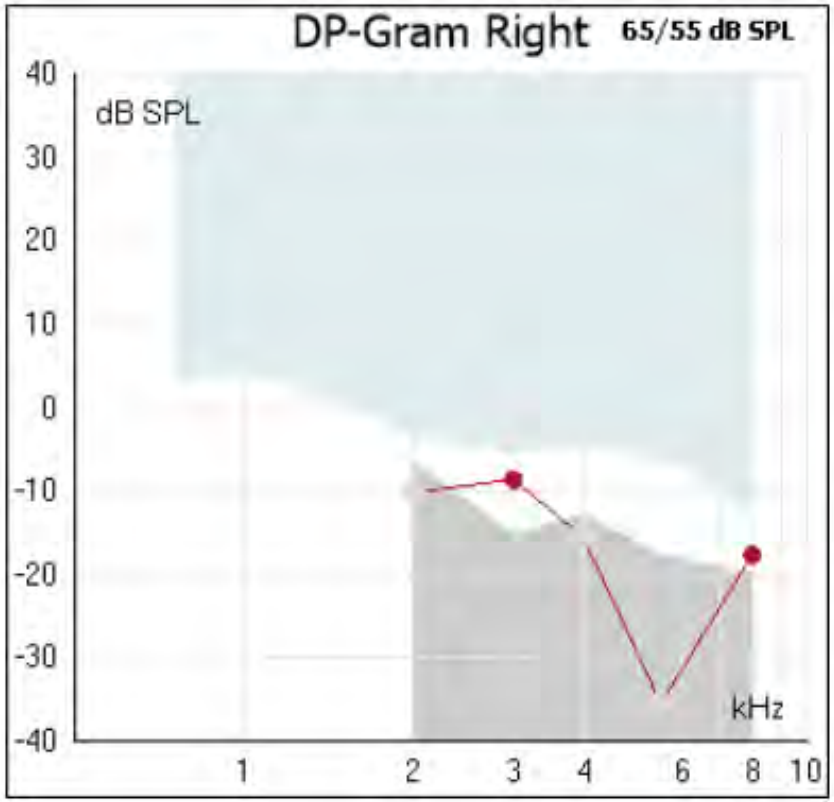
- Tympanometry

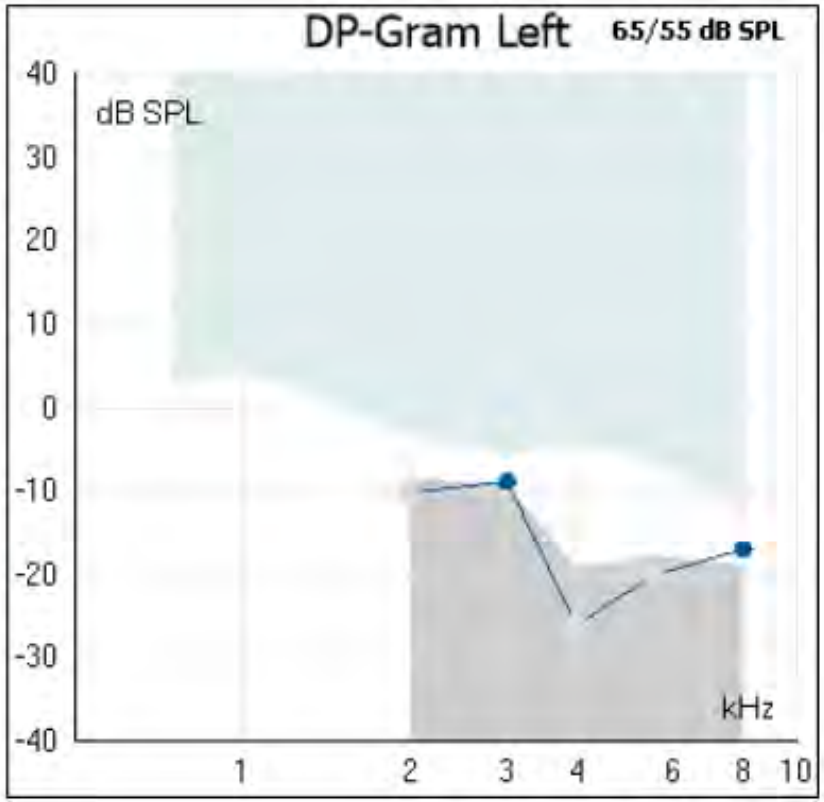




Results

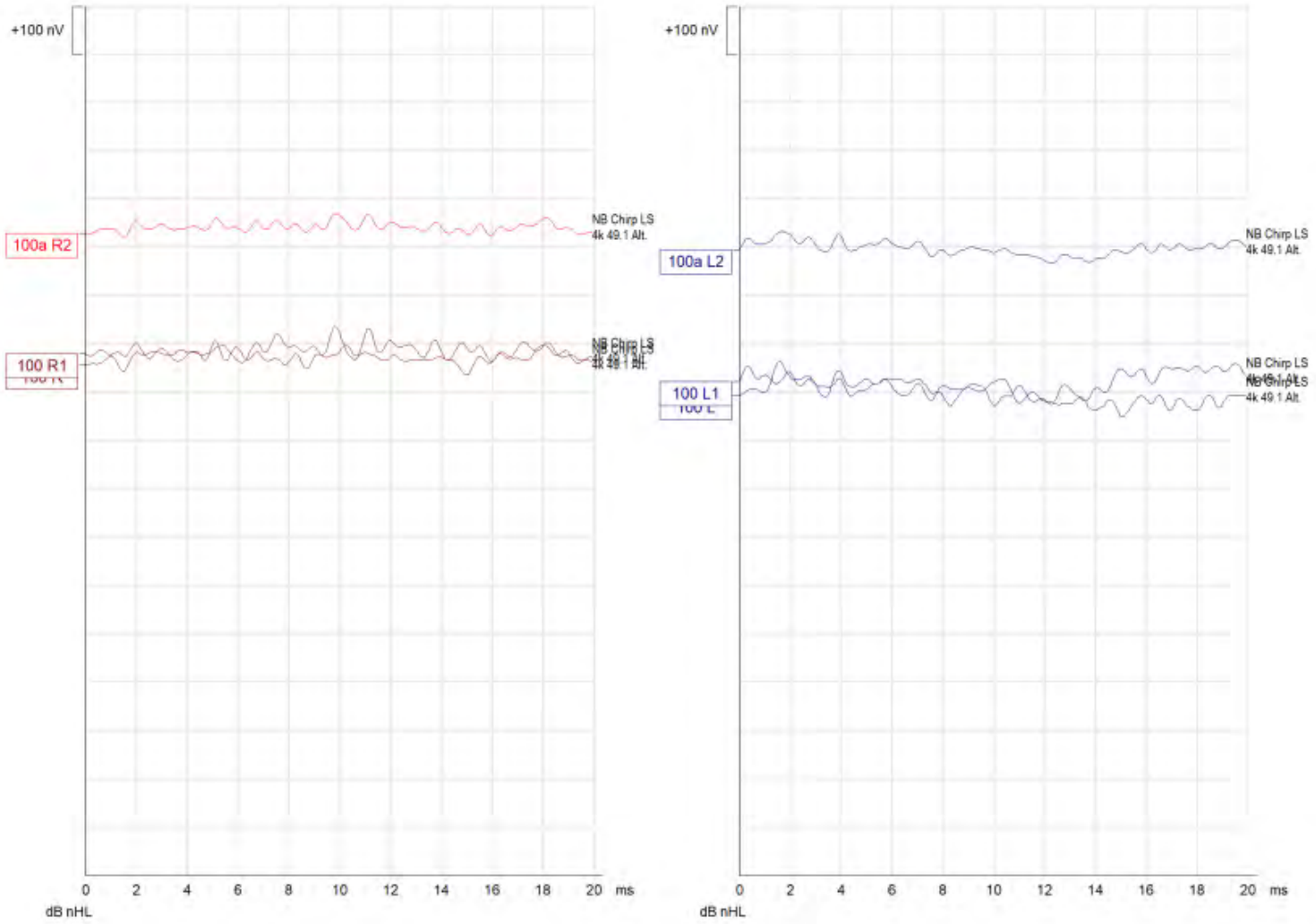
- DPOAEs





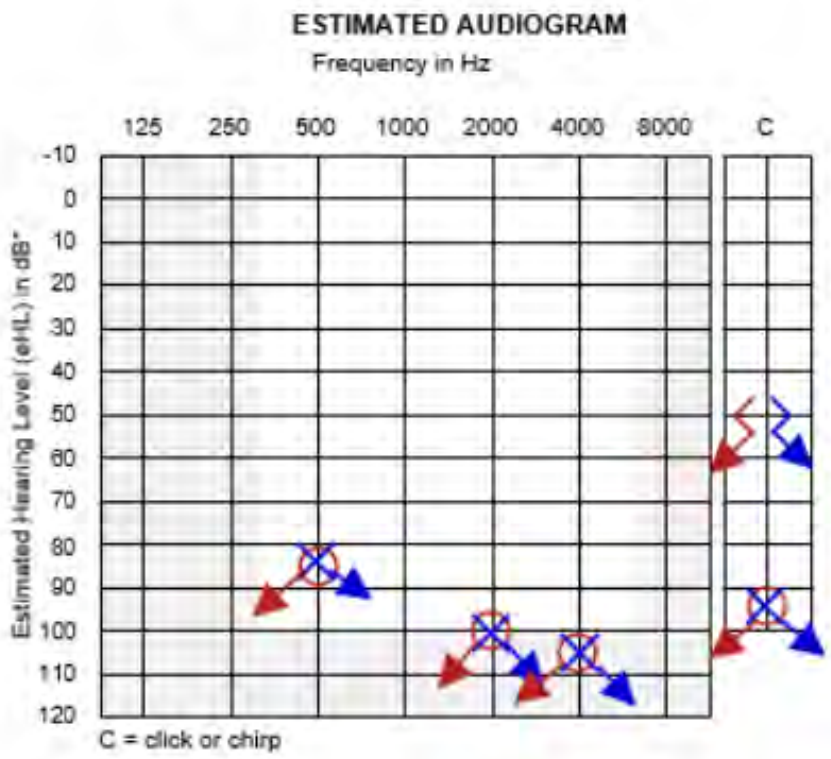
Results

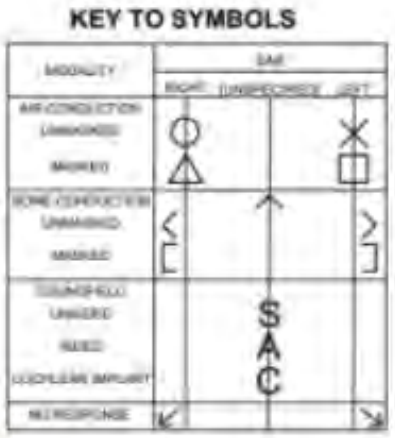
- Wave Example



Results

- Audiogram (eHL)





Uncomfortable Situations

- What makes this uncomfortable or difficult?
  - Provider/Facility bash
  - Different test results
  - How will family react?

Results with Trevor's Family

- What approach to take?
- Initially information based/information "overload"
- Review of previous results and comparing to today's
- Family's reaction, questions and next steps

Robinson et. al.,2023

Potential Uncomfortable Situations

- Different test results from other providers
- Family disagrees with results
- Pseudohypacusis patients
- Others?

Importance of Counseling Skills

- Audiologists know it is important
- Confidence varies
- Influence family reactions/adherence to care
- Ultimately can change outcomes for patients and families
- Family/child centered care

Meibos et. al. 2019

Counseling Tools

- Narrative medicine
- Open ended questions
- Pregnant pause
- Validation and avoiding projection
- Check for understanding
- Informed/shared planning

Narrative Medicine in Audiology

- Narrative Medicine Definition:
  - “Narrative medicine is a pedagogical framework that acknowledges and honors individual and collective narratives in the clinical and academic contexts. In practice, it covers a broad scope of activities from engaging with poetry and prose to reflective writing and engagement with the visual and performing arts. Any medium that tells a story can be used. Narrative medicine aims to address the feelings and experiences that occur when caring for the ill. It validates and bears witness to the experience of the patient while encouraging creativity and self-reflection in the health care practitioner.”

Loy & Kowalsky, 2024

Narrative Medicine in Audiology

- Provides a chance to get information you maybe wouldn't have otherwise
- Example:
  - "Tell me about your last appointment and how you felt afterwards?"

Loy & Kowalsky, 2024

Open Ended Questions

- Self-check of personal bias/assessments
- Avoids projections on a family or patient
- Opens the conversation
- Critical for both case history, during the appointment, and providing results
- Examples:
  - “What questions do you have from today’s appointment?”
  - “What else can I do to support you and your family?”

Pregnant Pause

- Allows times to process
- Avoid projecting providers thoughts/feelings
- Example:
  - WAIT! Even when it may feel uncomfortable ☺

Validating Feelings/Concerns

- Validate feelings family has AFTER asking
- Opportunity to include human connection into healthcare
- Examples:
  - “Feeling upset towards different results is to be expected and very normal.”
  - “This is a lot of information to take in and I understand when you say you feel overwhelmed.”

Check for Understanding

- Provides an opportunity to check in and adjust counseling as needed.
- Examples:
  - “What questions do you have about the results I just went over?”
  - “What do you understand about what your child can and cannot hear based on today’s test results”
  - Questionnaires
    - Scale of Parental Involvement and Self-Efficacy –Revised (SPISE-R)

Informed/Shared Planning

- Often last step, but not a last-ditch effort
- Use the information you gained
- Opportunity to not shame other providers or facilities
- Examples:
  - “Based on today’s information and difference in I would strongly recommend repeat testing

Where is Trevor now?

- Confirmation hearing evaluation – profound in both ears
- Fit with hearing aids with essentially no reaction
- On the path for cochlear implants
- Normal MRI

Closing Thoughts

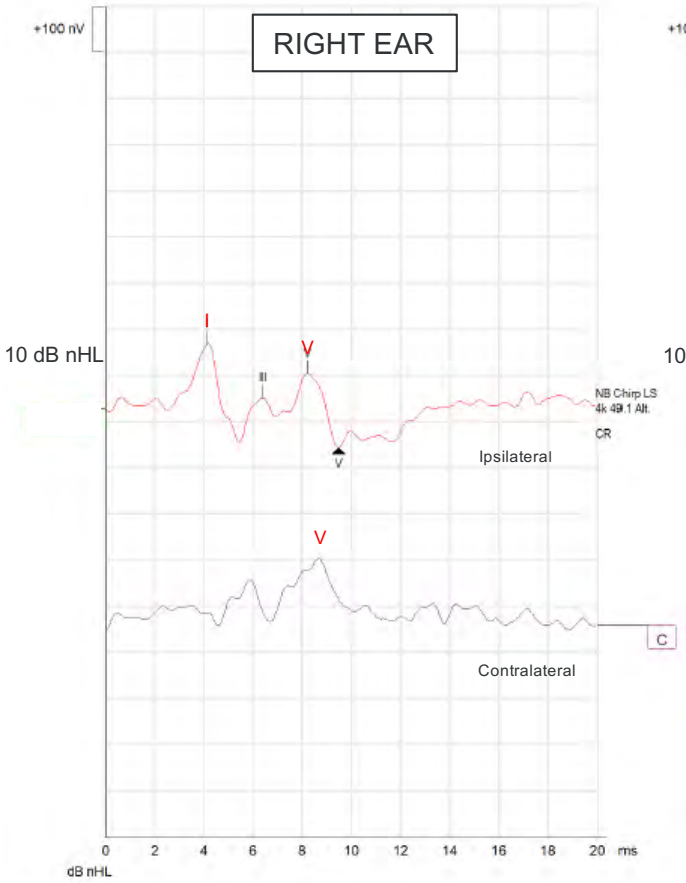
Tanner Robinson, AuD, CCC-A

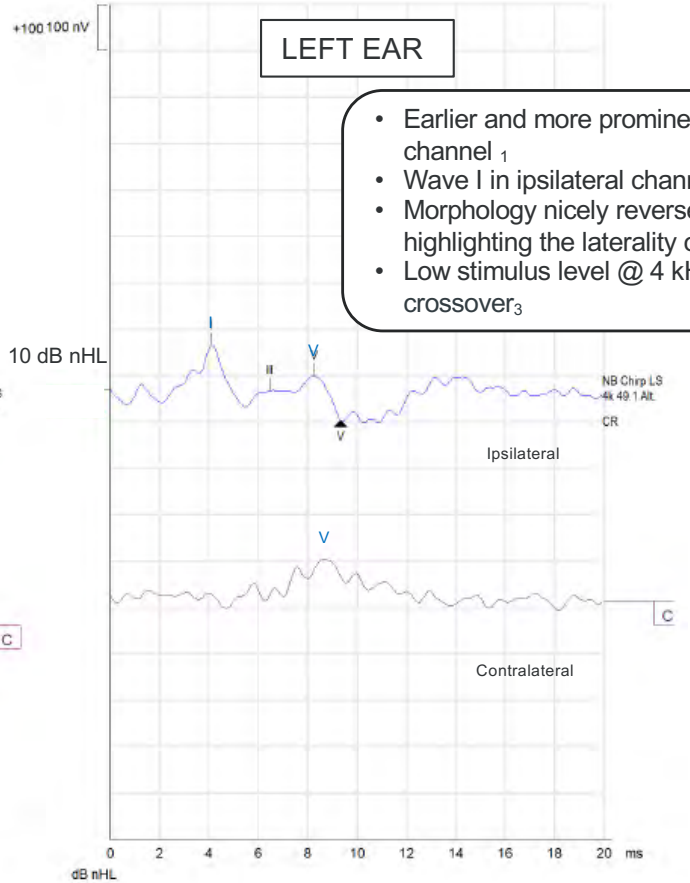
Left, Right, or Wrong?

Wrangling with ABR Laterality

Robert Fanning, AuD

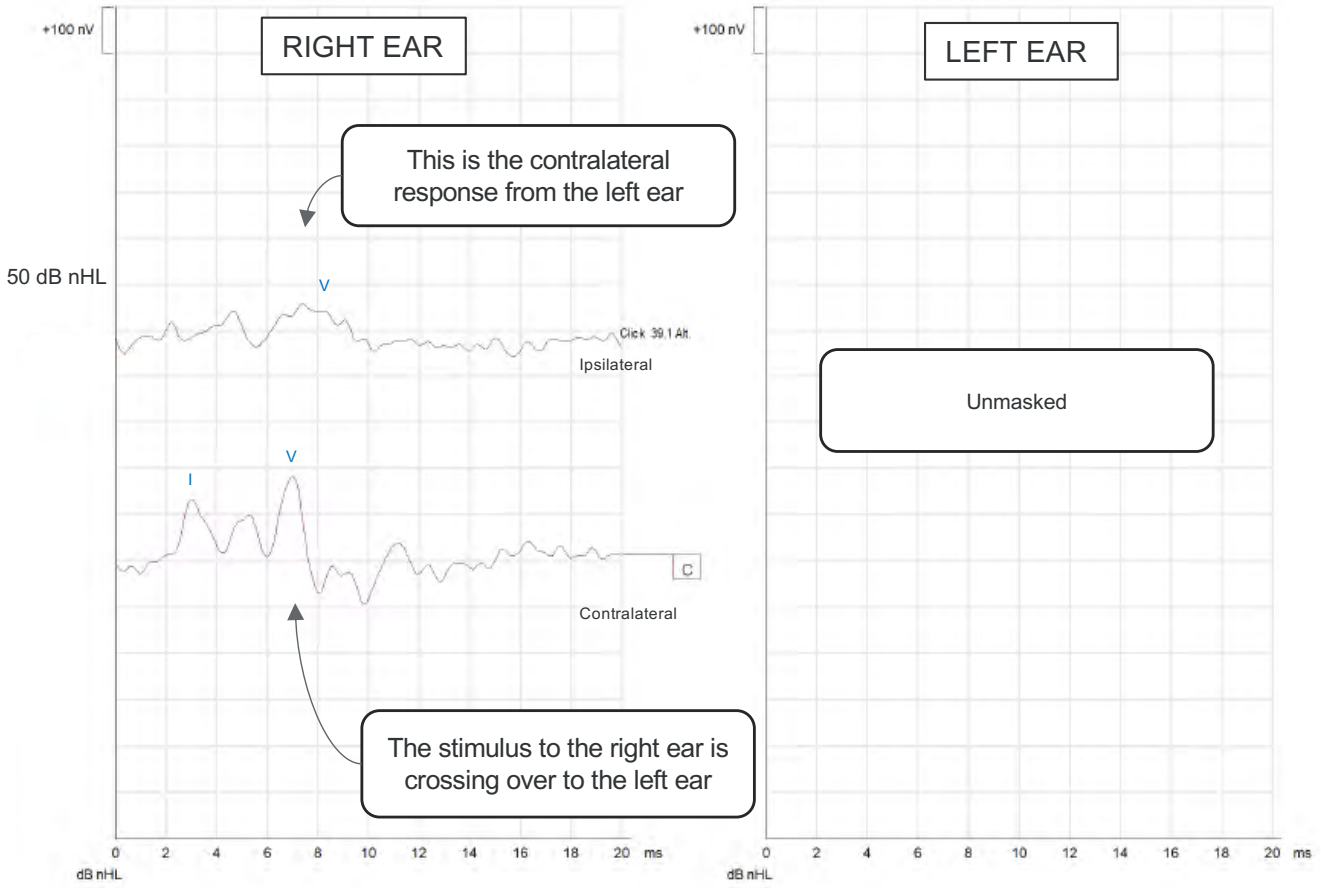
Review of ABR Concepts



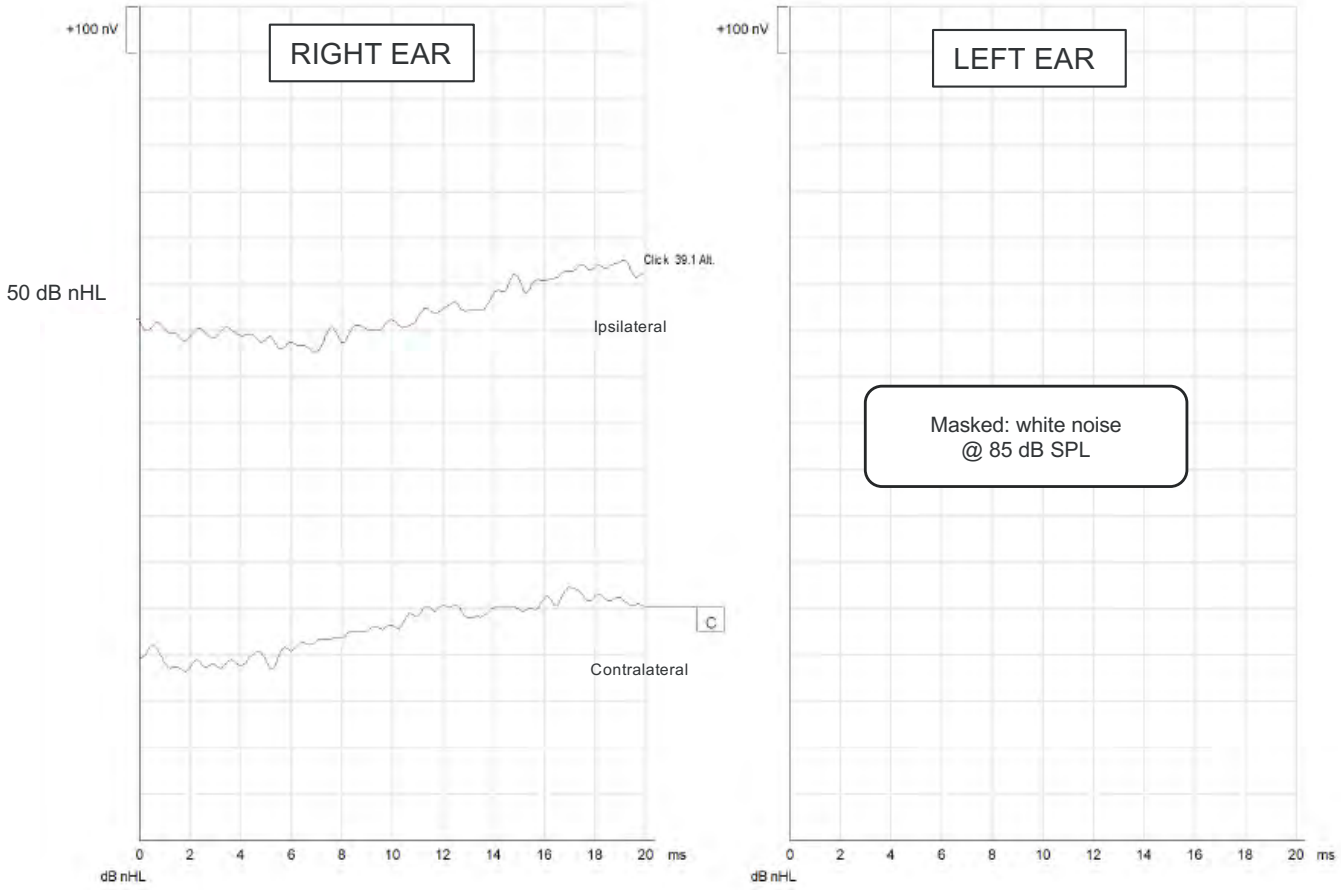


- Earlier and more prominent wave V in ipsilateral channel [1]
- Wave I in ipsilateral channel [2]
- Morphology nicely reverses from RE to LE, highlighting the laterality of the responses.
- Low stimulus level @ 4 kHz ⇒ little risk of crossover[3]

Bilateral conductive hearing loss in a toddler. Bone conduction test with CE Chirps at 4,000 Hz. Masking is not indicated.

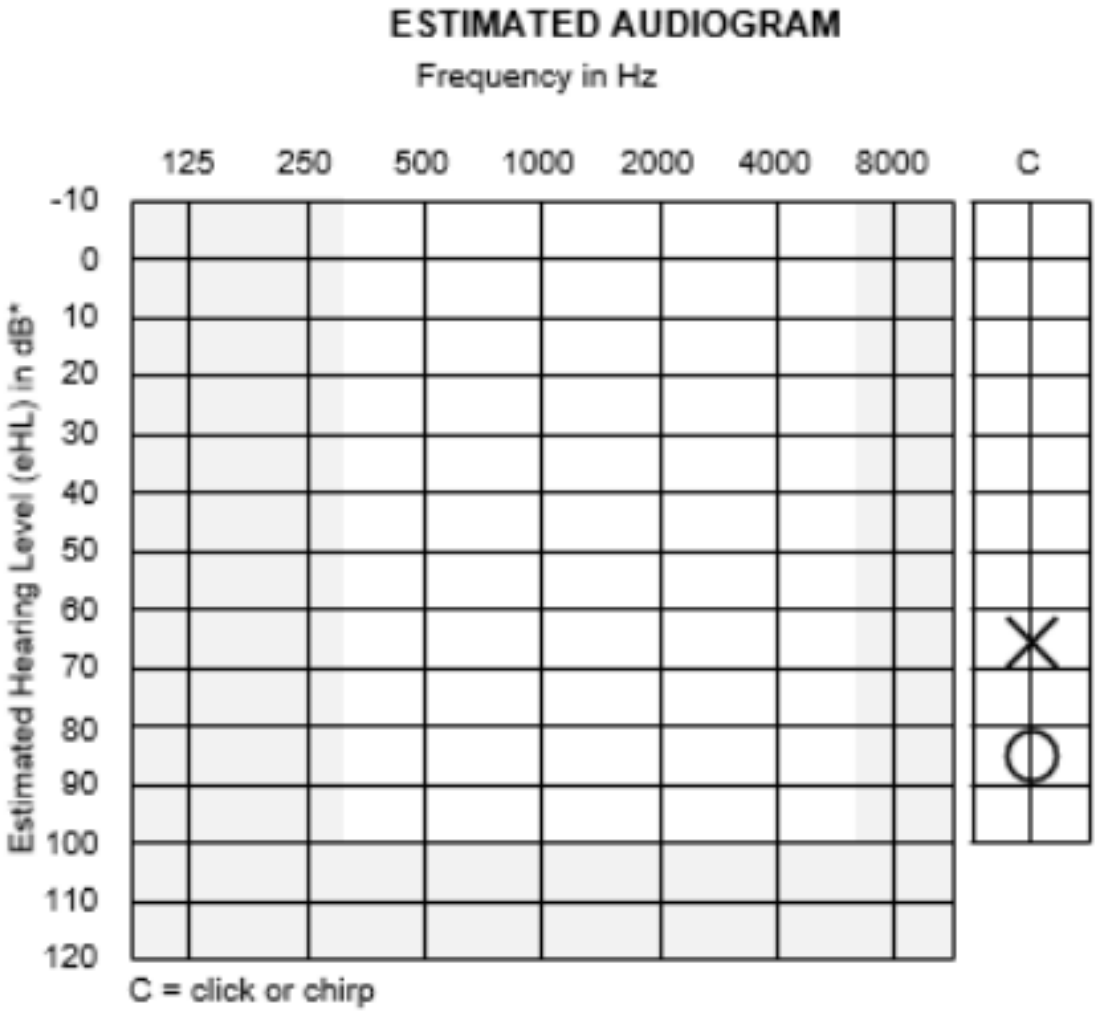


Unilateral profound SNHL/ANSD in the RE. Bone conduction test with click to right mastoid, which is crossing over to left ear. Masking is indicated.

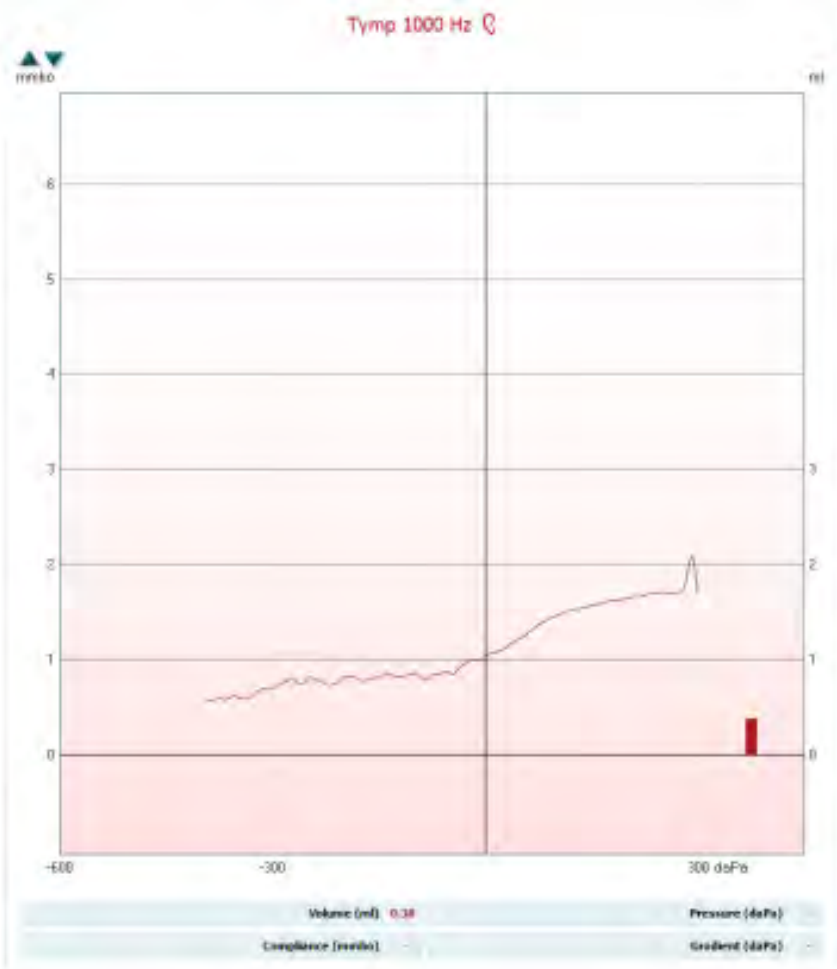


Unilateral profound SNHL/ANSD. Bone conduction test on right ear; left ear effectively masked.

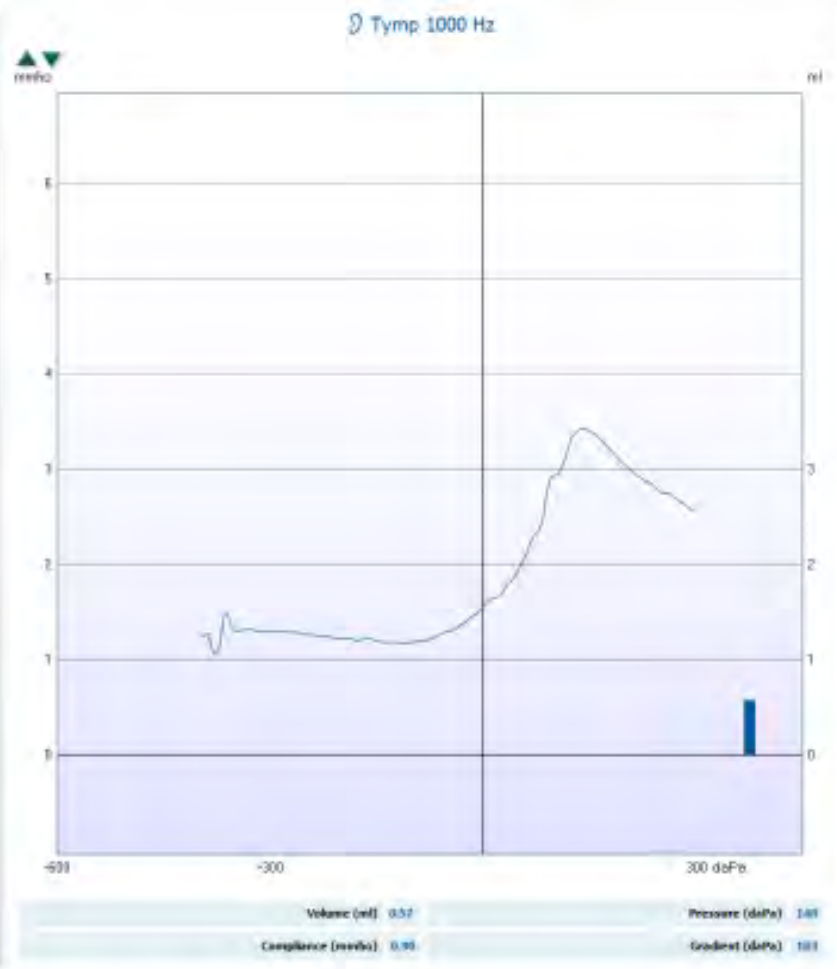
- 6-month-old male born at 32 wga admitted to our NICU
  - 19 weeks corrected age on DOS
- **Multiple congenital anomalies** that include
  - ASD, large VSD
  - Hydrocephalus s/p placement of VP shunt
  - Grade 3 BPD
  - Club feet
  - Genital abnormalities
  - Ventriculomegaly
  - Tracheomalacia,
  - Subglottic stenosis,
  - S/P tracheostomy and Ladd's/G-tube placement
  - S/p VSD closure and flow restrictor removal
  - **Bilateral hearing loss**
    - No screening due to otologic malformations
    - ABR test at bedside in natural sleep showed **severe hearing loss in the right ear** and **moderately-severe hearing loss in the left ear.**


ESTIMATED AUDIOGRAM
Frequency in Hz
125
250
500
1000
2000
4000
8000
C
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
Estimated Hearing Level (eHL) in dB*
C = click or chirp

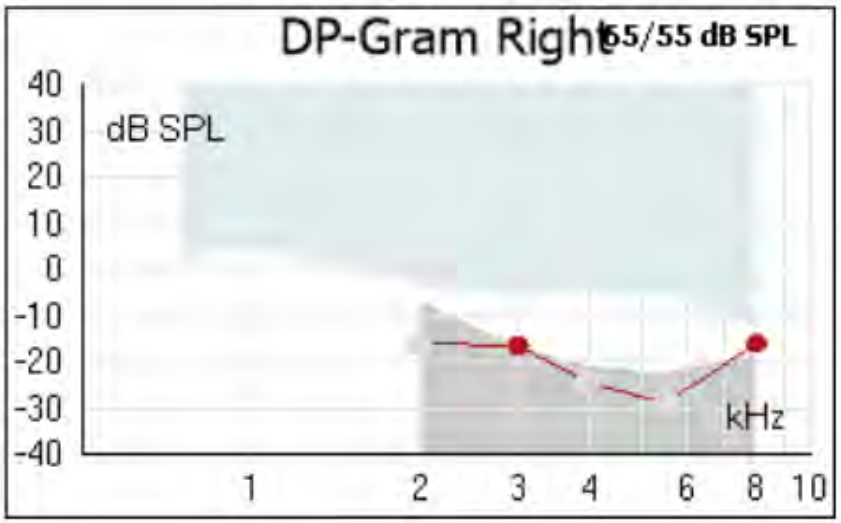
RIGHT EAR



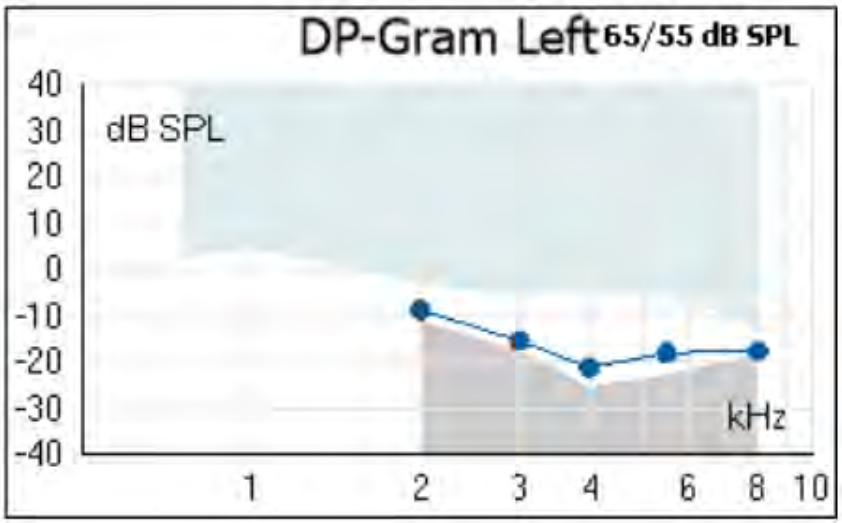
LEFT EAR



RIGHT EAR

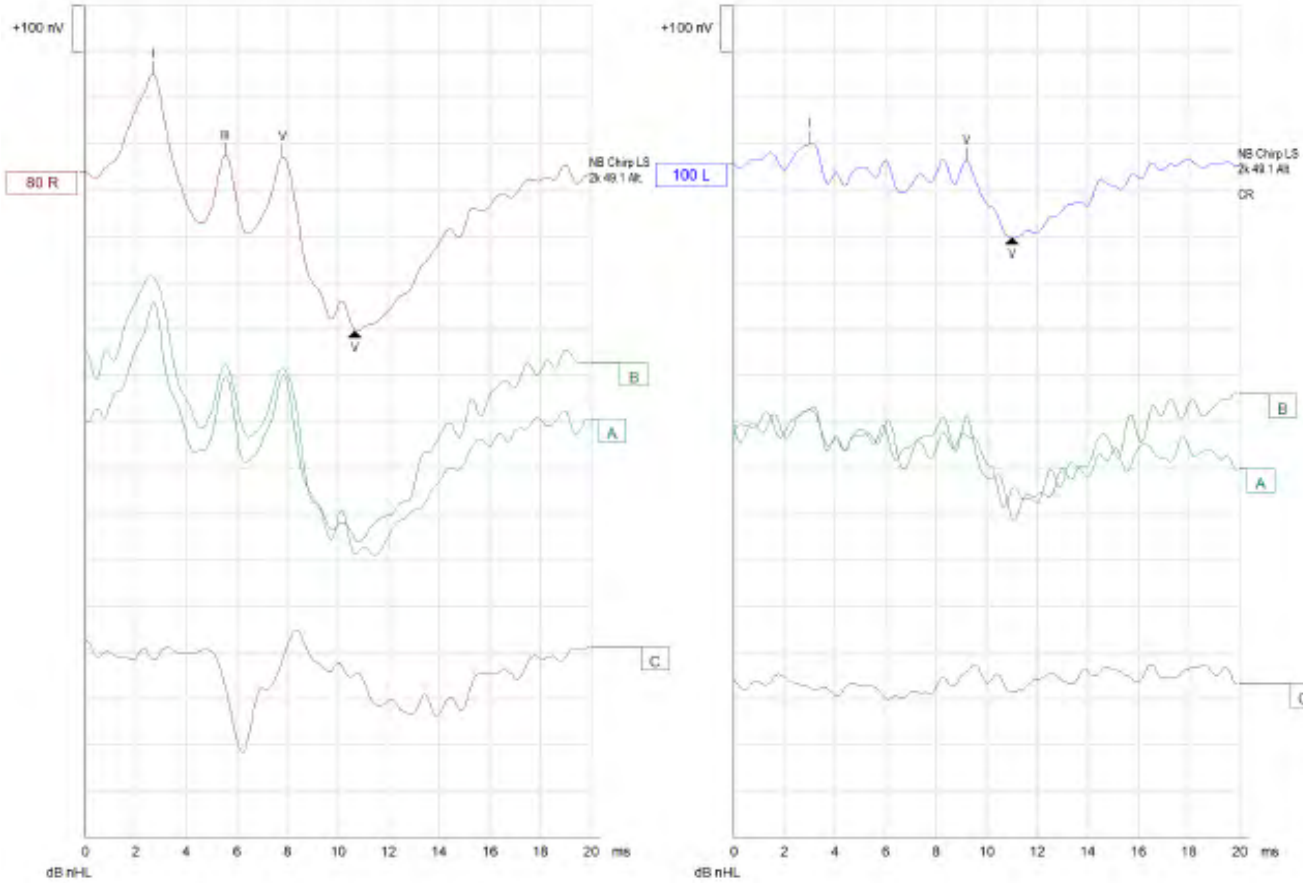


LEFT EAR

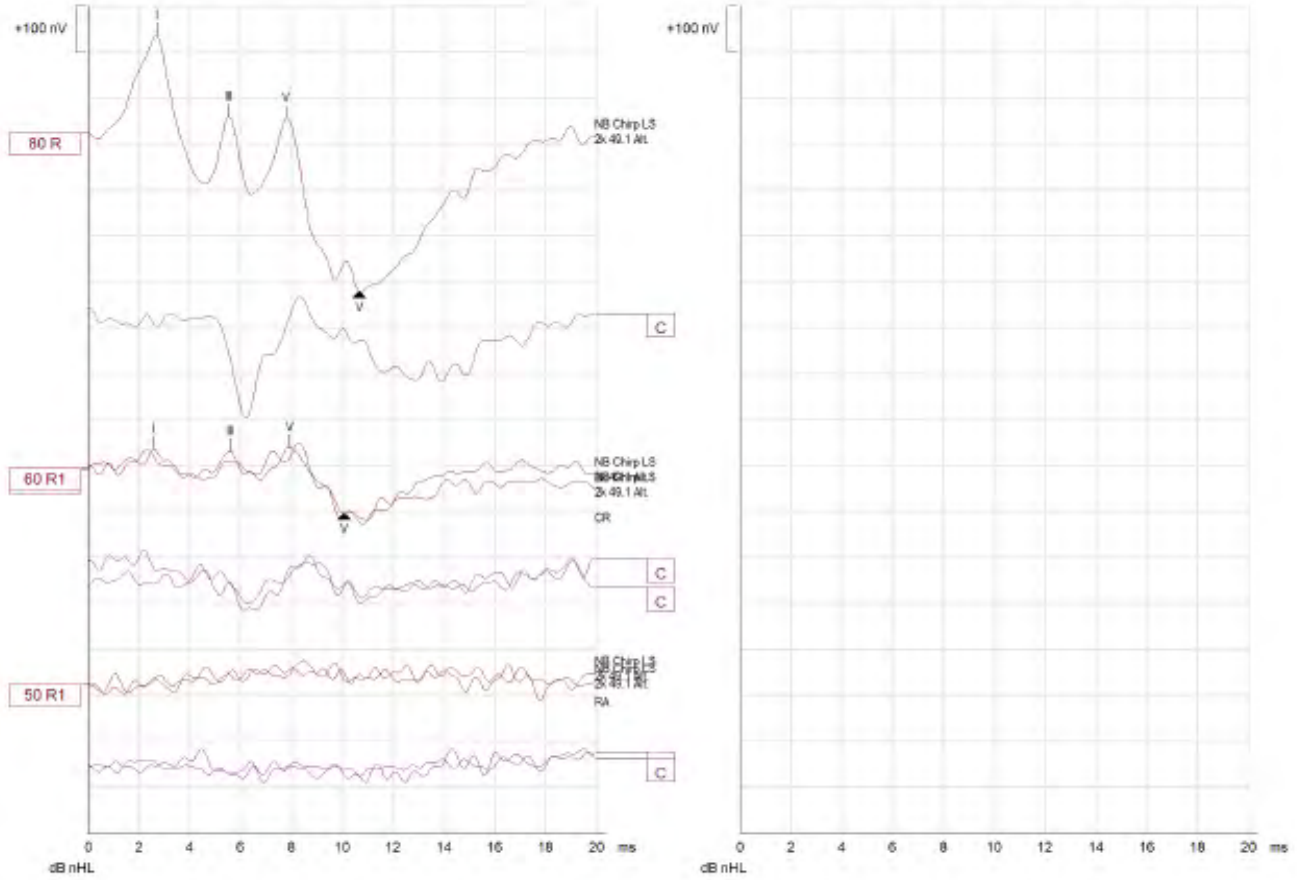


RIGHT EAR

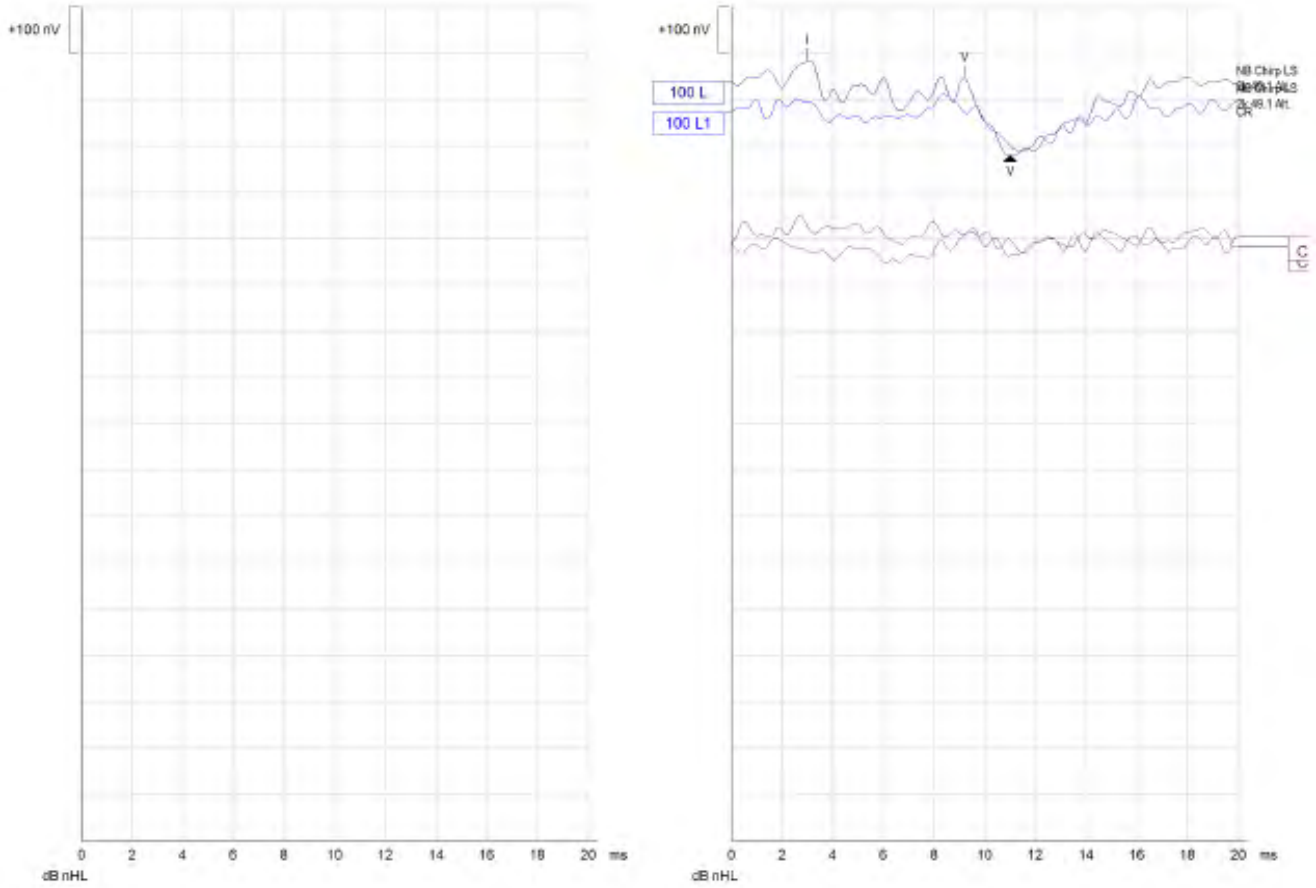
LEFT EAR



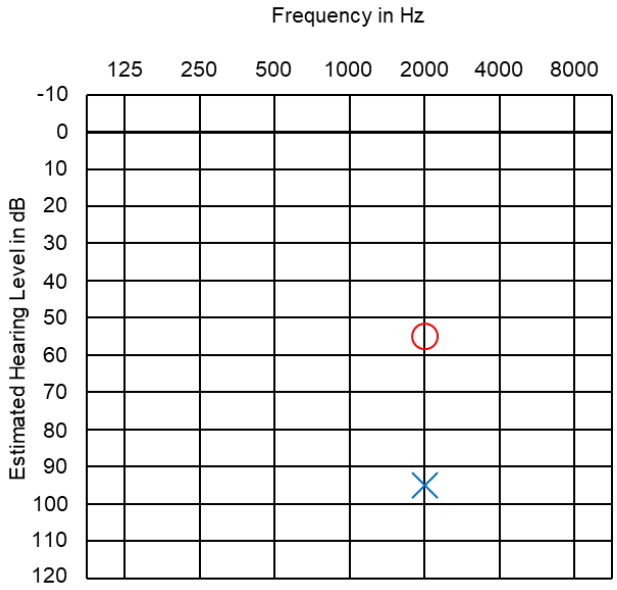
AC 2 kHz NB Chirps



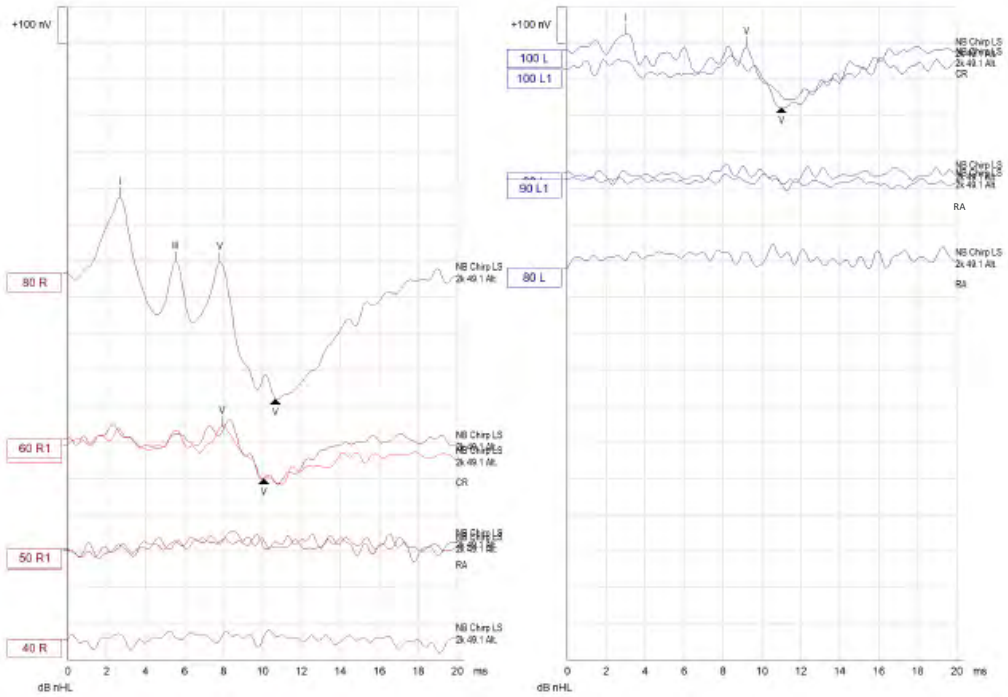
AC 2 kHz NB Chirps

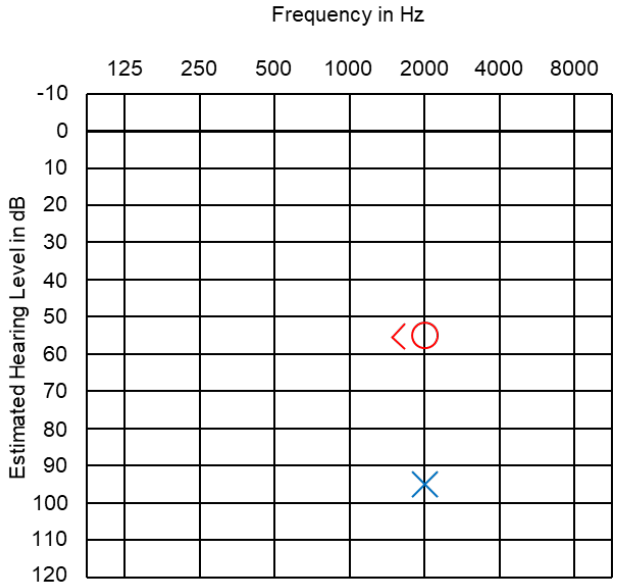


AC 2 kHz NB Chirps

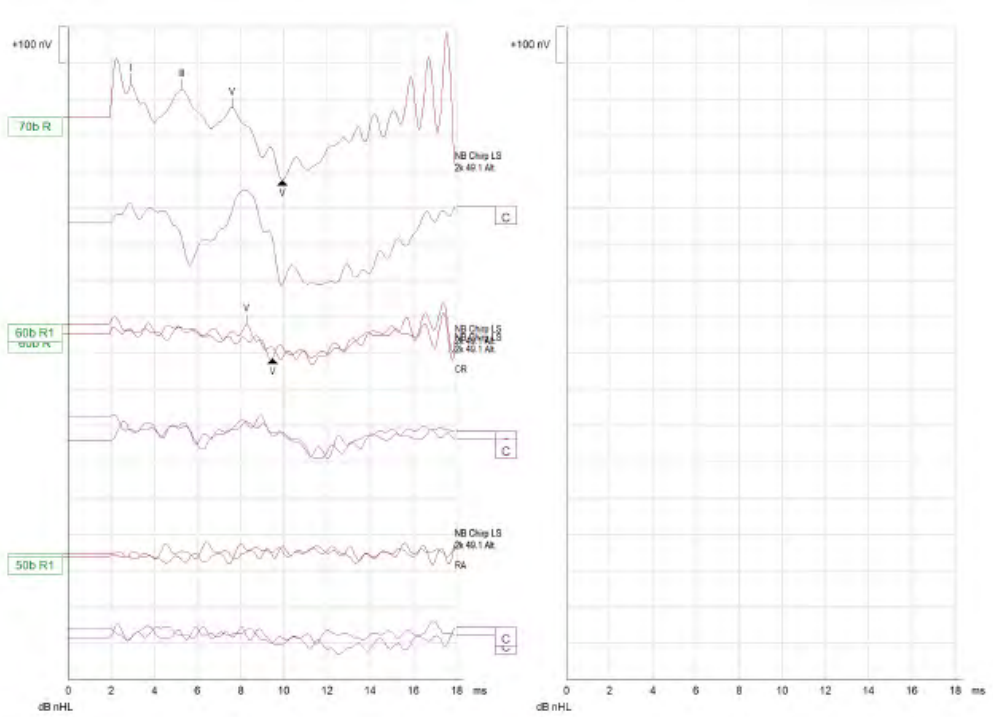


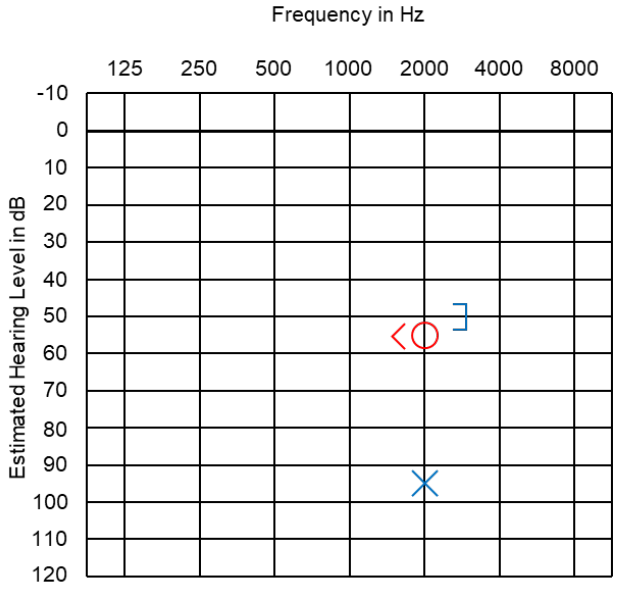
AC NB CE Chirps
2000 Hz



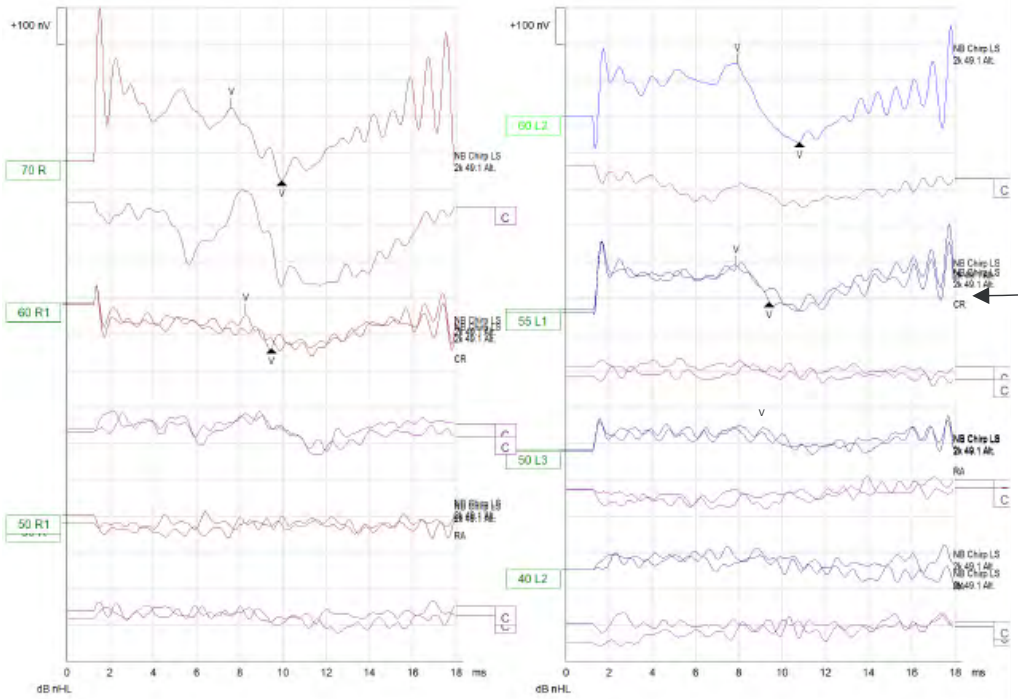


BC NB CE Chirps
2000 Hz

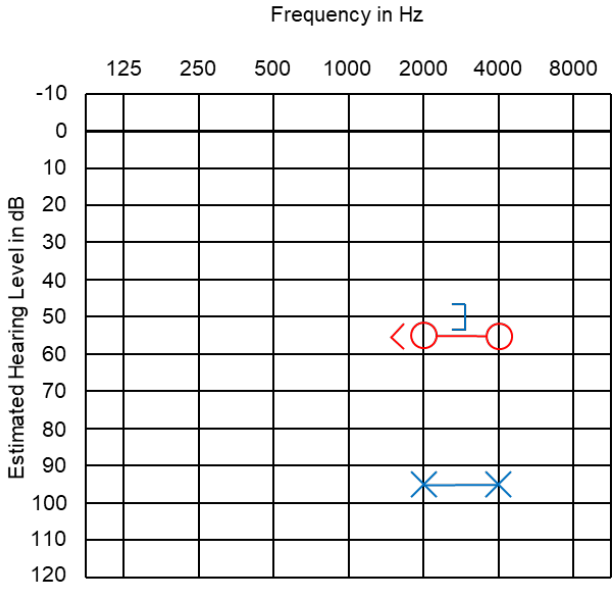




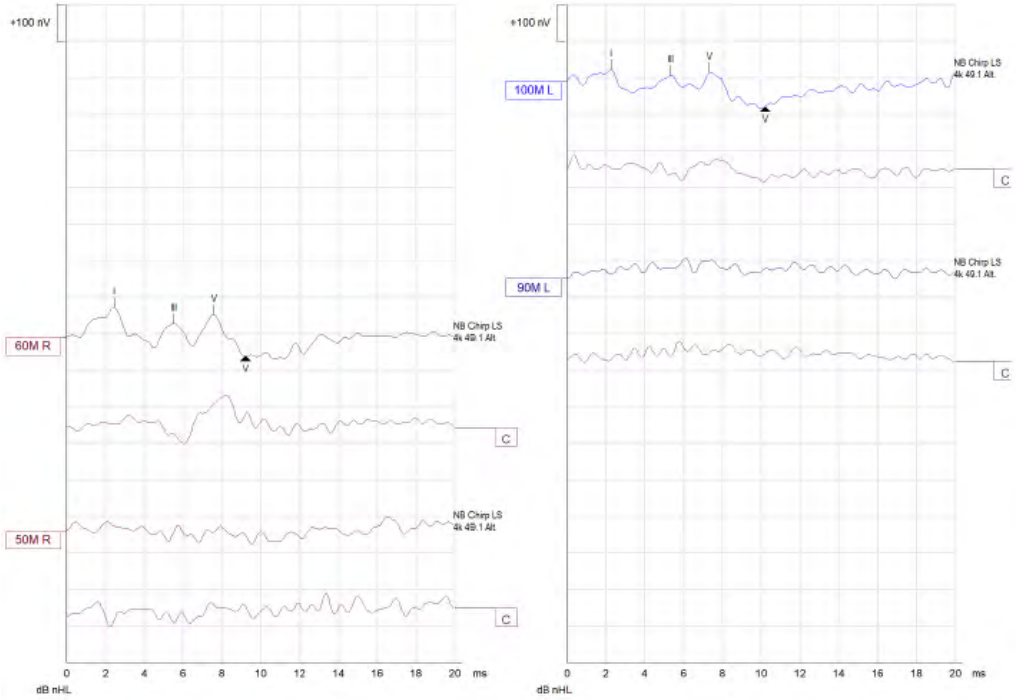
BC 2000 Hz

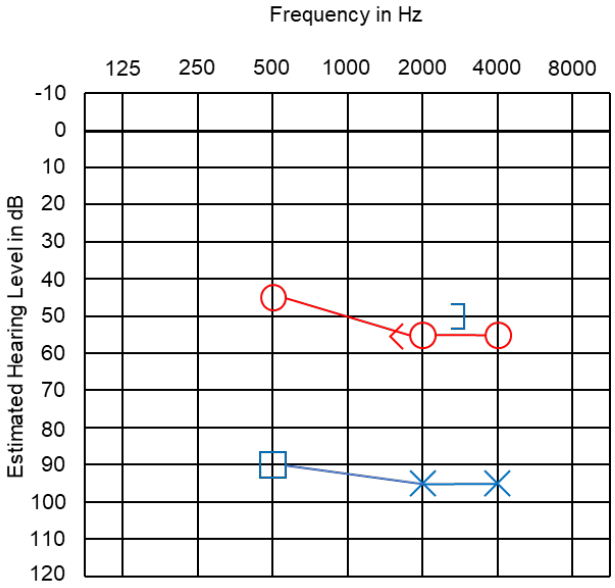


One tracing is masked: 110 dB SPL

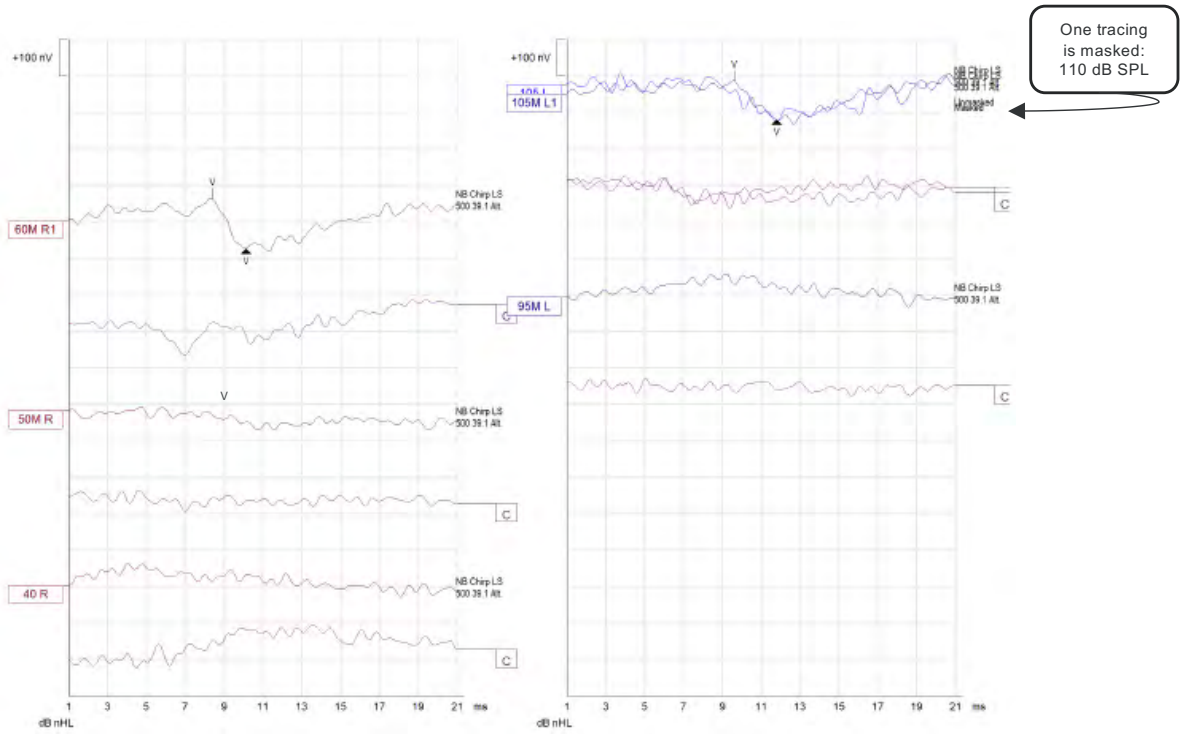

Frequency in Hz
125
250
500
1000
2000
4000
8000
Estimated Hearing Level in dB
-10
0
10
20
30
40
50
60
70
80
90
100
110
120

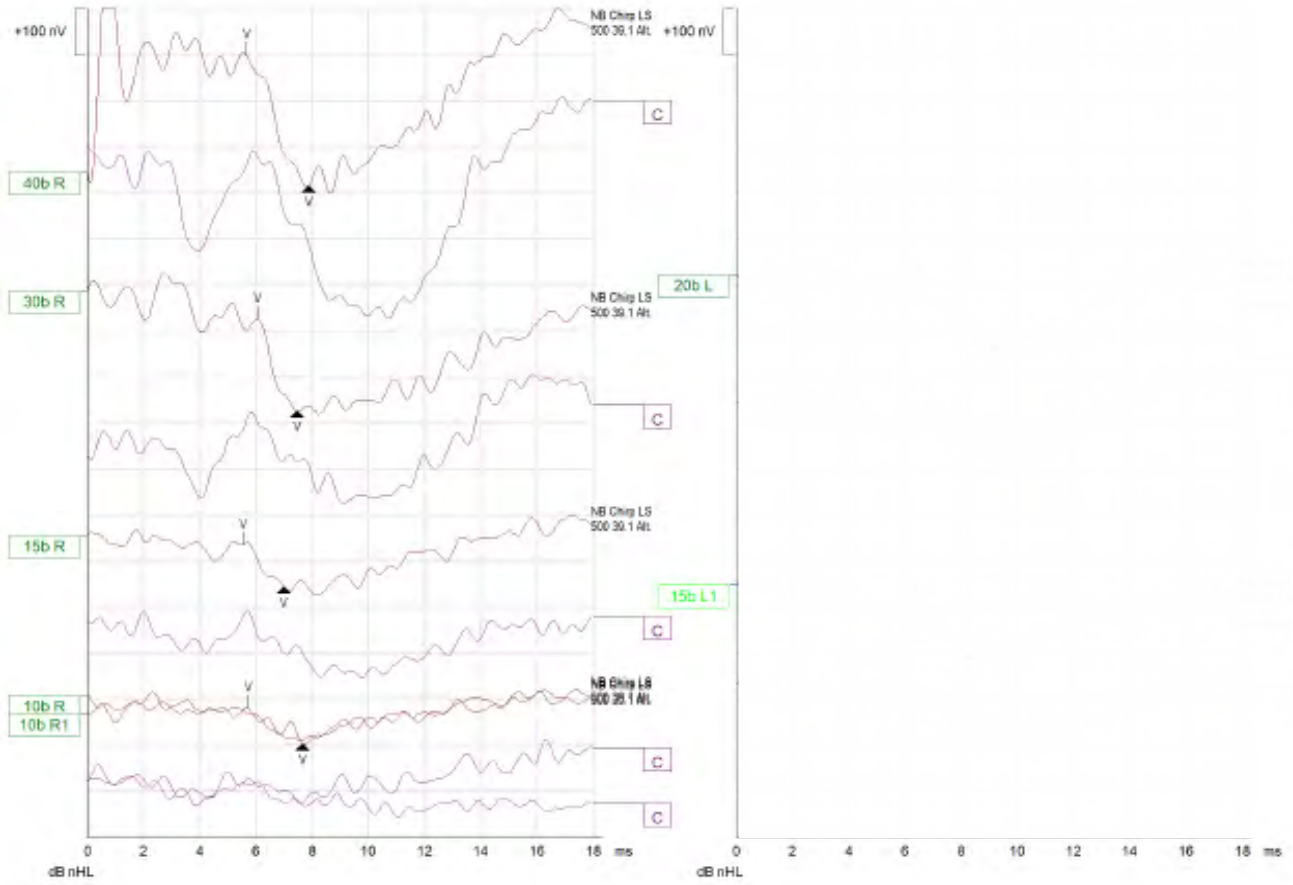
AC 4000 Hz


+100 nV
100M L
90M L
60M R
50M R
NB Chirp LS
4k 49.1 Alt
C
0 2 4 6 8 10 12 14 16 18 20 ms
dB nHL

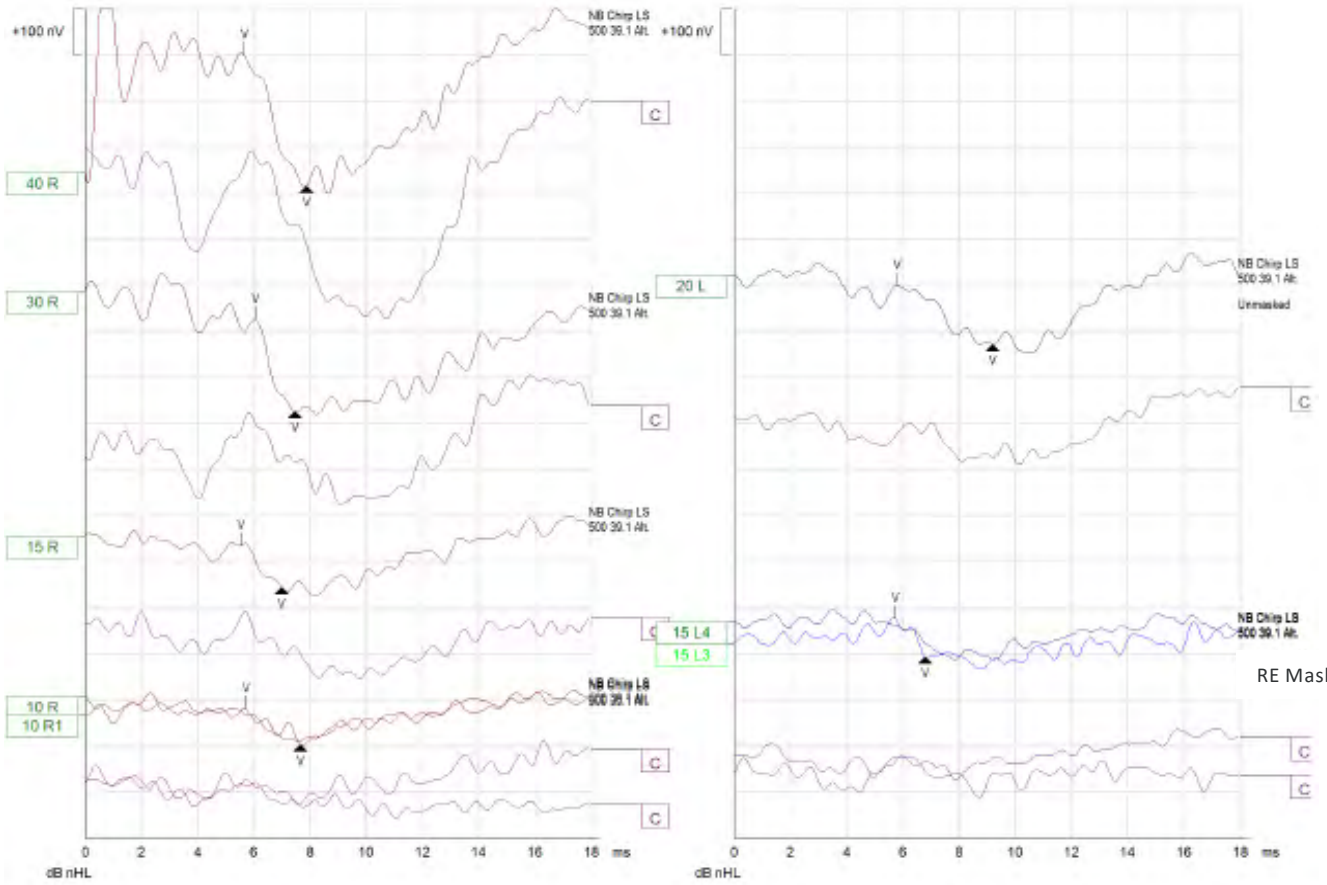

Frequency in Hz
125
250
500
1000
2000
4000
8000
Estimated Hearing Level in dB
-10
0
10
20
30
40
50
60
70
80
90
100
110
120

AC 500 Hz


One tracing is masked: 110 dB SPL
+100 nV
105M L1
60M R1
95M L
50M R
40 R
NB Chirp LS
500 39.1 Alt
C
V
1 3 5 7 9 11 13 15 17 19 21 ms
dB nHL

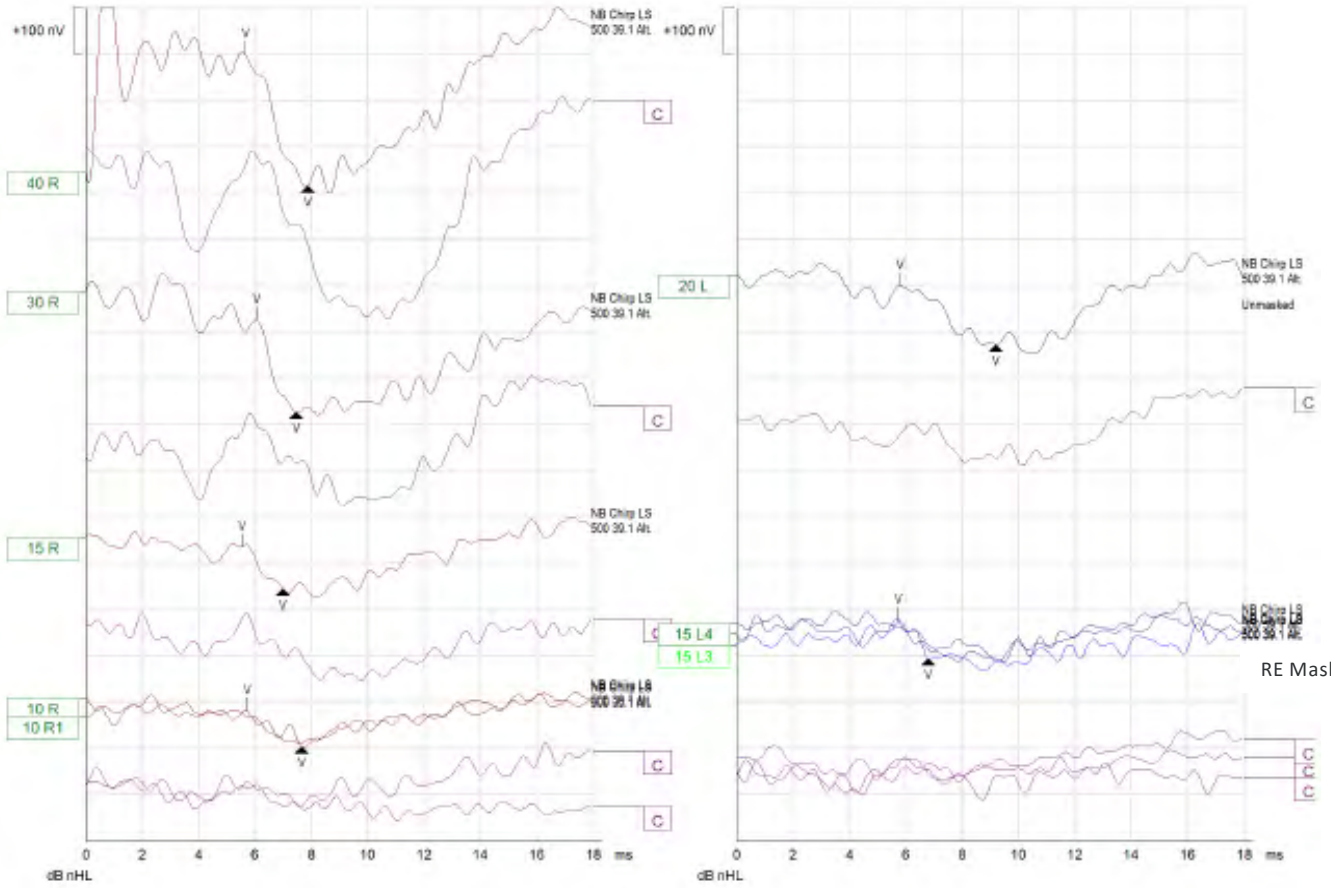


BC 500 Hz NB Chirps



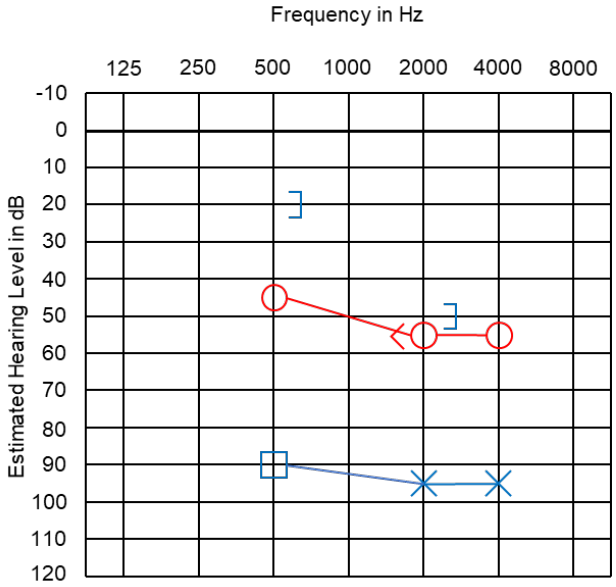
RE Masked @ 100 dB

BC 500 Hz NB Chirps

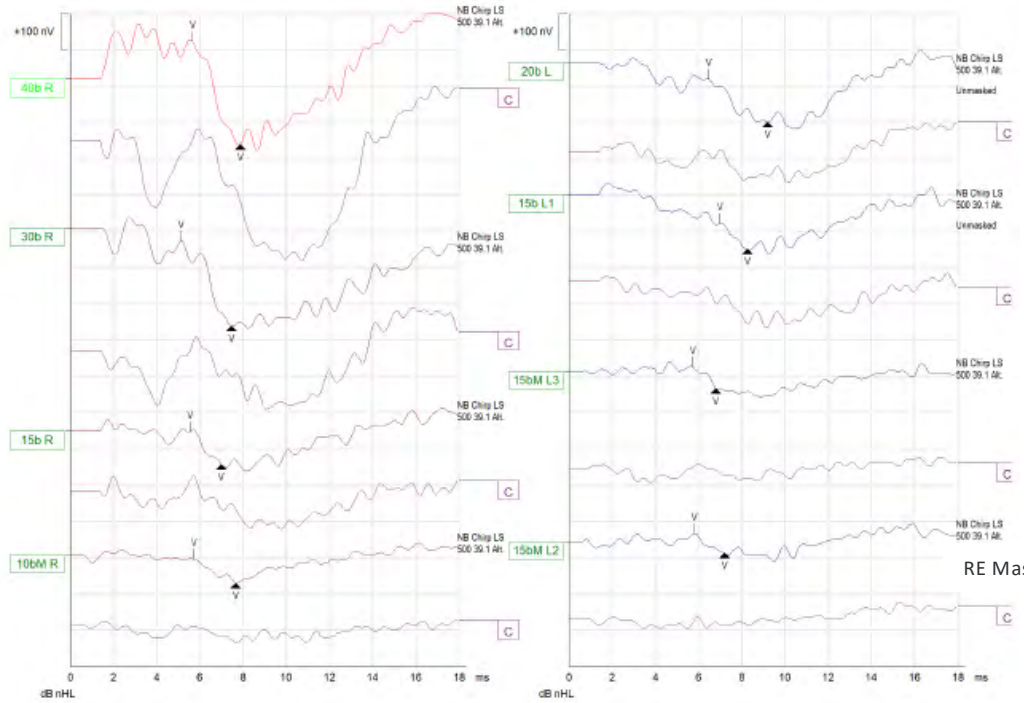


RE Masked @ 100 & 110 dB

BC 500 Hz NB Chirps



BC 500 Hz



RE Masked @ 100 dB

Assessment and Plan

- RE: Primarily SNHL, possibly mixed
- LE: Mixed
- Retest w/in 3 months to extend and confirm
- Hearing aid RE; consider LE
  - Otologic malformations
  - Changes in LE hearing levels?

Summary

- Evaluating both channels can help you determine the laterality of the ABR with and without the use of clinical masking
- Assessing contralateral recordings even when it is not necessary can help guide your evaluations when they become murky.
  - Get in the habit of looking at them even in straightforward cases. It trains your eyes.
  - Print them so you and others can review them later
- Think dynamically. It is sometimes alright to seek clarity in follow up evaluations.

Closing Thoughts

Robert Fanning, AuD

A Frequent Finding: GJB2-Related Hearing Loss

Alissa Nickerson, AuD

Genetics & Childhood Hearing Loss

- Hearing loss impacts 1 / 650 newborns[7]
- Genetics account for more than half of congenital hearing loss[5]
- Of this genetic hearing loss, ~70% is nonsyndromic and ~30% is syndromic[6]
- Majority of nonsyndromic sensorineural hearing loss is autosomal recessive – approximately 80%[6]

Gap Junction Beta 2 (GJB2)

- Changes in GJB2 account for nearly half of non-syndromic, prelingual sensorineural hearing loss in some regions[4]
- The relationship between hearing loss and GJB2 was reported in 1997[2]
- Gene which codes for the gap junction protein Connexin 26[2]
- Variant prevalence differs by population[4]
- Carrier rate higher in some areas, such as the Mediterranean[4]
- There are greater than 150 mutations, polymorphisms, and variations in the GJB2 gene[5]

Common GJB2 Audiologic Findings

- Onset: Prelingual[4]
- Degree: Frequently moderate or greater[4]
  - Although can range from mild to profound[1]
  - Severity related to type of mutation[3]
- Laterality: Often bilateral[4]
- Progression: Generally non-progressive[4]
- Configuration: Flat or slightly down-slanting[4]

Case Study #1

- Born full-term
- Referred on initial NBHS in both ears
- Referred outpatient follow-up NBHS in both ears
- Parents report she startled to only loud sounds
- No prior family hx of hearing loss
- Initial Diagnostic Evaluation at ~6 weeks of age
  - Peaked 1000 Hz tympanograms
  - Absent DPOAEs
  - BAER study suggesting bilateral moderate SNHL

Patient #1 Subsequent Work-Up

- Genetics consult revealed she was heterozygous for the c.35delG variant in the GJB2 gene
- Established with ENT, Ophthalmology, and ST
- Unremarkable MRI of brain and temporal bones
- Overall stable hearing loss, as demonstrated by her annual hearing evaluations
- Excellent hearing aid adoption – BTE device use ~14 hours/day binaurally.
- Listening and spoken language

Patient #1 Recent Audiogram

Case Study #2

- Born full-term
- Referred on initial NBHS in both ears
- Referred outpatient follow-up NBHS in both ears
- Family hx: distant paternal cousin with congenital HL
- External BAER study suggested severe/profound SNHL bilaterally
- Diagnostic BAER study at ~7 weeks of age
  - Peaked 1000 Hz tympanograms
  - Absent DPOAEs
  - BAER study suggested profound SNHL AU

Patient #2 Subsequent Work-Up

- Genetics consult revealed she was heterozygous for two pathogenic variants in the GJB2 gene
- Established with ENT, Ophthalmology, and ST
- Unremarkable MRI of the brain and temporal bones
- Underwent a HA trial
- Underwent bilateral cochlear implantation at ~7 months of age; present CI use ~12 hours/day
- Listening and spoken language

In Conclusion

- GJB2-related hearing loss is common
- Prevalence varies by ethnicity and region
- Hearing loss can range from mild to profound – but is often moderate or greater, impacting both ears

Closing Thoughts

Alissa Nickerson, AuD

Lengthy Journey to a Diagnosis

Ashley Geske AuD CCC-A

History

- History of IUGR, born at 36 weeks and 5 days
- Did not sit up independently until 1 year of age
- Did not walk until 21-22 months
- Occupational and physical therapy through early intervention program
- Evaluated by neurology at 3 years old due to gait concerns
  - Noted to have generalized hypotonia
  - Recommended consult with genetics and to proceed with developmental pediatrics evaluation
- Typical speech development until 2.5 years when she stops learning new words
  - Started speech therapy and made significant progress
- Developmental pediatrics
  - No concerns for autism noted during evaluation

History

- MRI recommended by neurology
  - Results showed Subcortical T2 FLAIR signal abnormality in bilateral temporal lobes, right greater than left. This is nonspecific and may be secondary to prior viral infection/inflammation or metabolic insult. Unremarkable MRI of the entire spine without contrast.
- Neurology
  - Referred to genetics given strong suspicion of underlying genetic condition
  - Ordered chromosome microarray and fragile X testing

Genetics and Neurology Follow Ups

- Genetics
  - Metabolic panel was negative
  - Recommended genedx autism/ID Xpanded panel
    - Results were negative
    - Recommended follow up in 2-3 years.
- Neurology
  - Recommended audiology and ophthalmology consults.

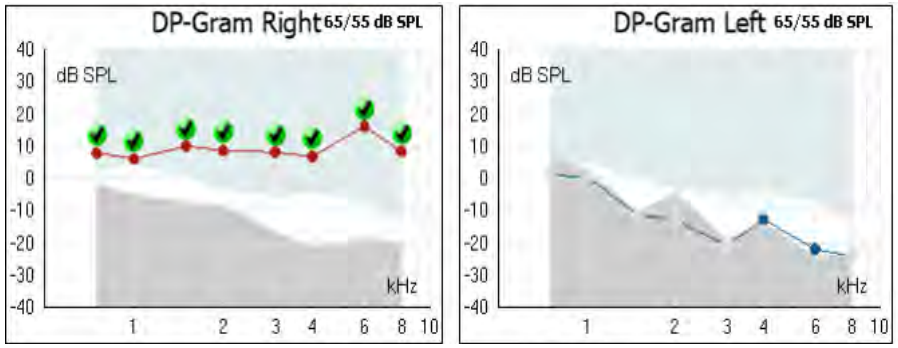
Audiologic Evaluation

- Type A tympanograms
- Present ipsilateral ART at 1000Hz I the right ear and absent ipsilateral ARTs 1000-4000Hz
- OAEs
  - Right: present .75-8kHz
  - Left: absent .75-8kHz

Tympanometry/Immittance Results:

Date	Ear	Probe Tone	Ear Canal Volume (mmhos)	Static Admittance (mmhos)	Peak Pressure (daPa)	Tympanic Width	Tympanogram Type
08-07-2024	Right	226 Hz	0.5	0.22	-10	145	Type A
	Left	226 Hz	0.4	0.22	-2	105	Type A

Acoustic Reflexes Results:

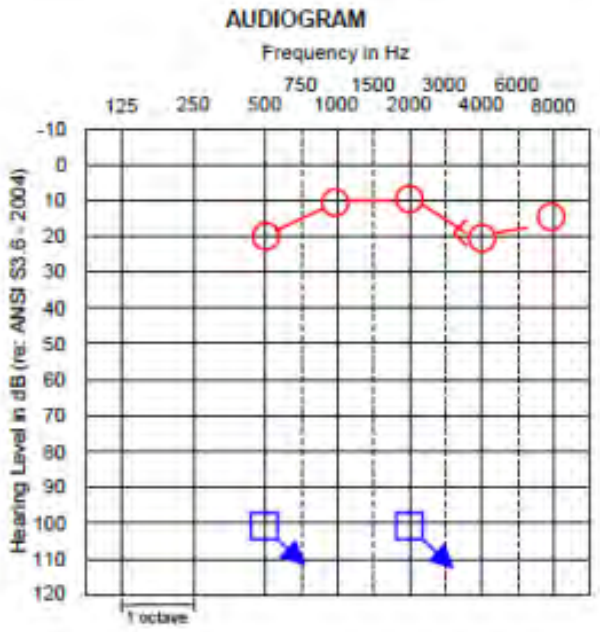


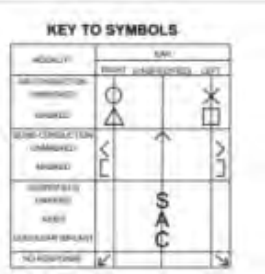
Acoustic Reflex Screening:

	500 Hz	1000 Hz	2000 Hz	4000 Hz	BBN
Right Ear		<100			DNT
Left Ear	DNT	NR	NR	NR	DNT

Audiologic Evaluation

- CPA with insert earphones
- Right: essentially normal hearing sensitivity
- Left: profound hearing loss
- SRT: picture pointing with recorded spondees
  - Right: 15dBHL
  - Left: no response @80dBHL (with contralateral masking)

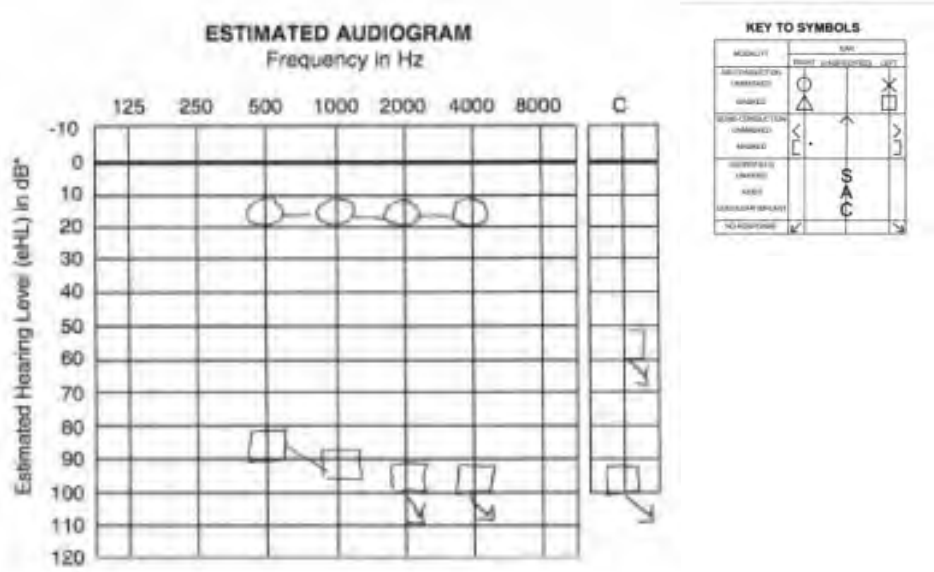




MRI and BAER Study

ENT consult

- MRI recommended if IACs
  - Results showed present cochlear nerves bilaterally.
  - Increased asymmetric bilateral T2/Flair foci throughout the supratentorial brain
  - Similar bilateral temporal subcortical flair hyperintensity
- BAER study
  - Completed in combination with MRI
  - Right: normal hearing sensitivity
  - Left: severe to profound sensorineural hearing loss



Vestibular Evaluation Summary

- Head Thrust Test: **Positive**
  - Catch-up saccades noted, bilaterally.
- Broke Man's Rotary Chair: **Positive**
  - Absence of post-rotary nystagmus towards both directions, consistent with bilateral peripheral vestibular dysfunction
- Standing on Solid Surface with Eyes Open: **Positive**
  - Unable to maintain upright balance without widening stance
- Single Leg Stance: **Positive**
  - Patient fell immediately on attempt of each leg
- (M-CTSIB): **Positive**
  - Fell immediately with both eyes open and eyes closed
- Gait Observation: **Positive**
  - Widens stance to maintain balance while walking. Some veering noted to both sides.
- Tracking: Negative
  - Was able to follow the target with ease.

Vestibular Evaluation Continued

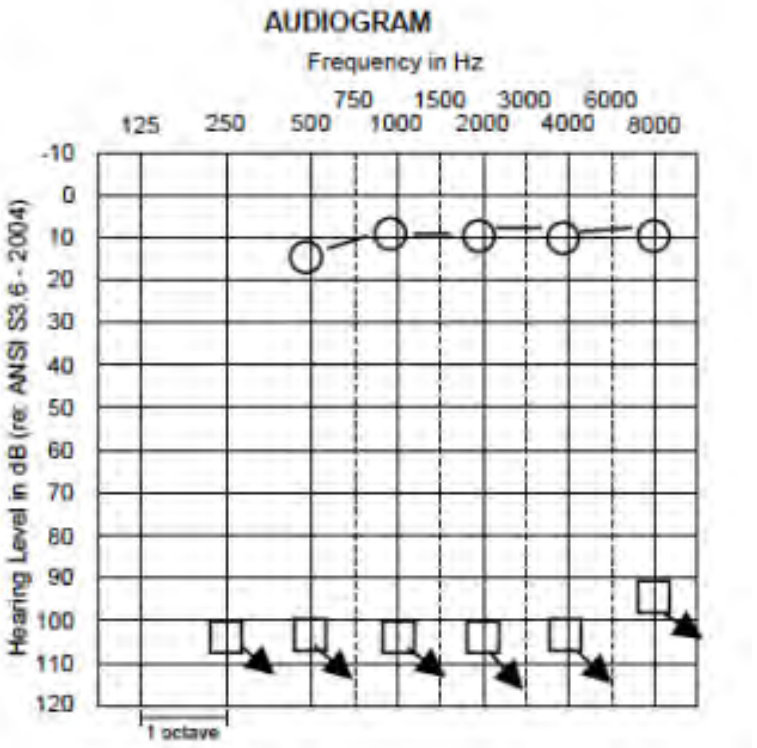
- Cervical Vestibular Evoked Myogenic Potential (cVEMP):
  - Assesses saccule and inferior vestibular nerve. The cVEMP was assessed via BC.
  - Left Ear: Negative
    - Good waveform morphology
  - Right Ear: **Positive**
    - Absent waveforms
    - Suggest abnormal right saccule and/or superior vestibular nerve function
- Occular Vestibular Evoked Myogenic Potential (oVEMP):
  - Assesses the utricle and superior vestibular nerve. The oVEMP was assessed via BC.
  - Both Ear: Negative
    - Normal absolute and inter-ear N1 and P1 latencies and amplitudes for both ears

Suspected Etiology: cCMV

- Characteristics consistent with cCMV:
  - MRI results
    - Subcortical T2 FLAIR signal abnormality in bilateral temporal lobes
  - Rapid hearing loss progression
  - Asymmetric progression of hearing loss
  - Progressive vestibular weakness
  - IUGR
  - Generalized hypotonia

CI Evaluation

- Confirmed Audiometry and completed WRS.
- Attempted reflexes however, patient would not tolerate probe in ear for extended period of time.
- Completed CI discussion with family.
  - Discussed alternative options as well.
- Family and clinician felt CI would be the best option considering her history.
- Did not do CROS, BC hearing aid, or hearing aid trial.



Speech Audiometry:

Date	Side	Test	Presentation Level	Percentage Correct/Threshold	Word List Used	Stimulus
01-22-2025	Right Ear	SRT		15dBHL	Spondees	Recorded
01-22-2025	Left Ear	SAT		>105dBHL	Spondees	Recorded
01-22-2025	Right Ear	WRS	45dBHL	100%	PBK	Recorded
01-22-2025	Left Ear	WRS	105dBHL	0%	PBK	Recorded

CI for SSD

- Reasons for proceeding with implantation.
  - Passed newborn hearing screening and experienced plateau in speech development around 2.5 years old.
    - May correspond with change or progression in hearing in the left ear.
- Possibility her better ear progresses
  - cCMV hearing loss often starts out as unilateral and the cases that progress to bilateral happen at a median of about 4 years.

Surgery and Activation

- Patient underwent surgery for the left ear.
- She was activated and her maps were optimized by 3rd appointment.
- She wears it 9-10 hours a day and tolerates it without issue.
- Time needed to realize full potential of CI

Closing Thoughts

Ashley Geske AuD CCC-A

A Case of Challenging Mixed Hearing Loss

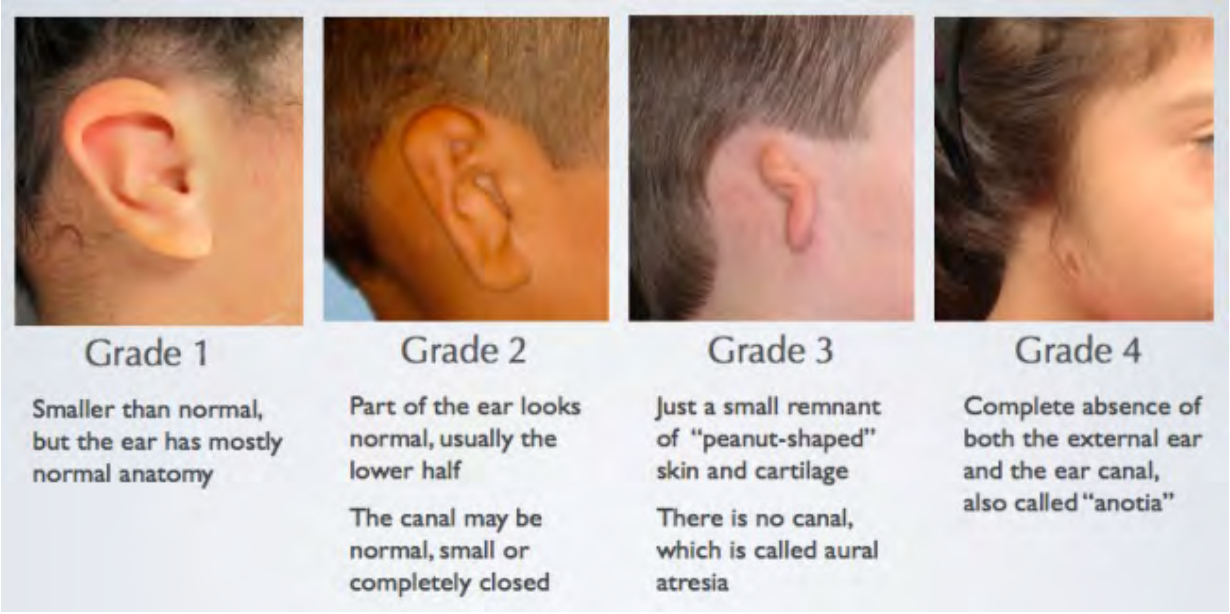
Allie Sayer, AuD, CCC-A

Concepts

- Microtia/Atresia
- Amplification options for mixed hearing loss
- Pediatric cochlear implant candidacy

Microtia/Atresia

- Microtia – congenital malformation of the pinna
- Atresia – congenital malformation/absence of the external auditory canal (EAC)
- Surgical reconstruction



Andrews et. al 2024, Lee et. al 2020; Photo: Dr. Sheryl Lewin, used with permission

Amplification Options for Conductive/Mixed Hearing Loss

- Hearing aids
- Bone-anchored hearing aids (BAHA)
  - Non-surgical
  - Surgical
    - Passive vs. active
- Middle ear implants

Ellsperman et. al 2021, Shohet et. al 2021

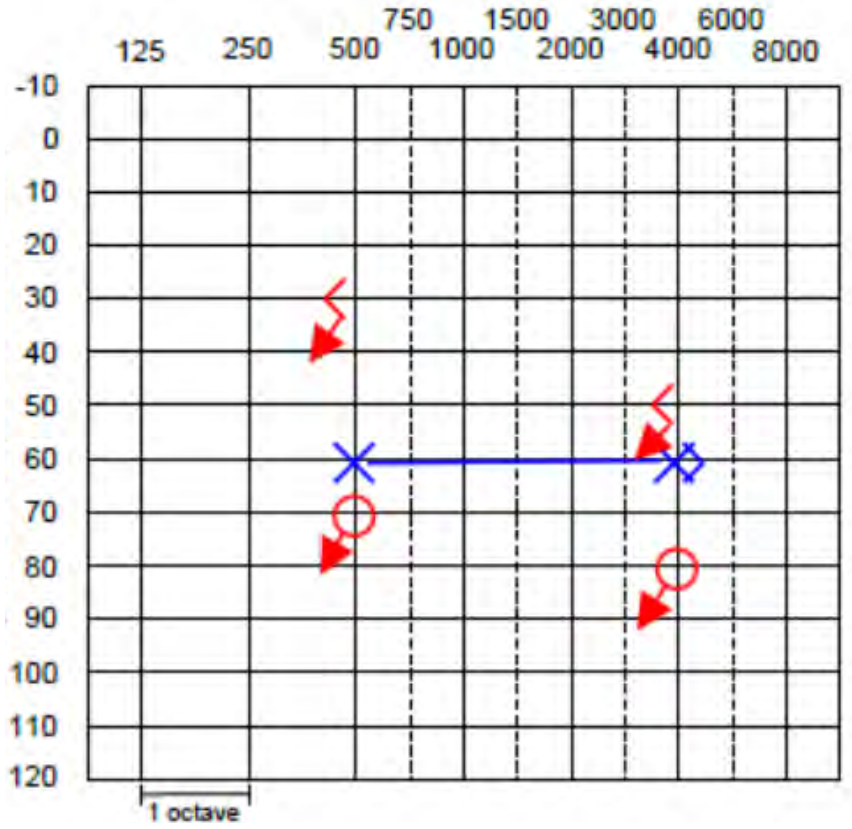
Pediatric Cochlear Implant Candidacy

- FDA Indications
  - Vary by manufacturer
  - General pediatric candidacy:
    - 9-24 months of age: bilateral profound sensorineural hearing loss
    - 2 years+: bilateral severe to profound sensorineural hearing loss
    - Limited benefit from use of hearing aids
- Off-label implantation
  - Asymmetric hearing loss
  - More residual hearing
  - Children <9 months of age

Park et. al 2022, Warner-Czyz et. al 2022

Introducing “Irene”

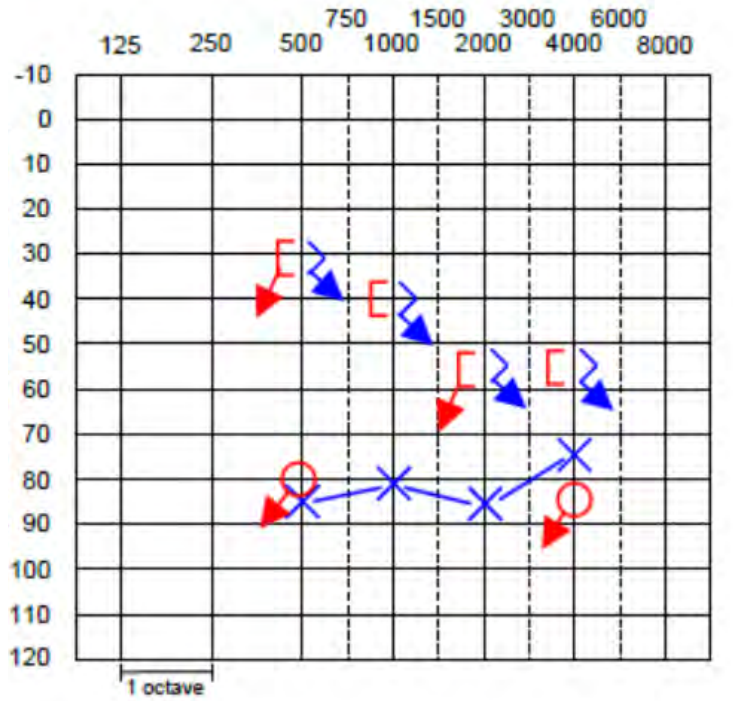
- 6-year-old female
- First seen by PCH audiology at 5 months of age
- Failed NBHS
- MRI ordered by outside provider was normal for inner ear anatomy
- Right grade 4 microtia with aural atresia
- Left grade I microtia
- Initial ABR study:
  - Right: Profound, primarily sensorineural hearing loss; a conductive component is likely due to the malformation of the right ear.
  - Left: Moderately-severe sensorineural hearing loss

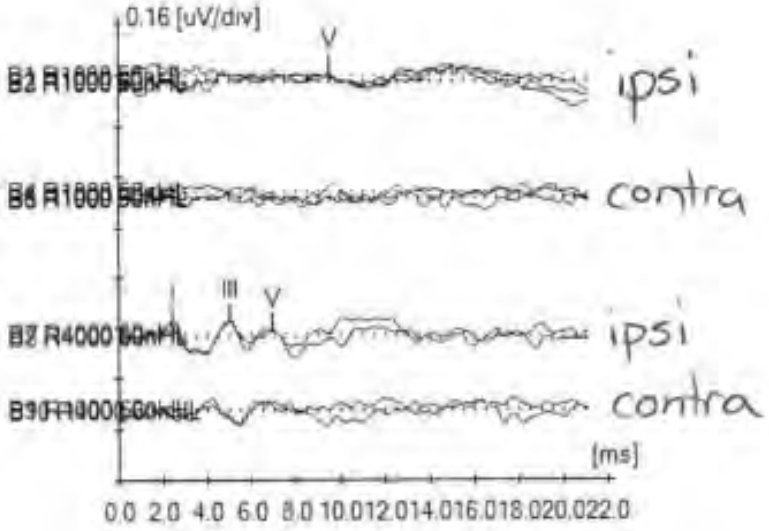


- Established with ENT and plastic surgery
  - Consideration of cochlear implant(s) and impact on microtia reconstruction (around 7-8 years of age)
  - Concern for bilateral facial paralysis noted (reanimation procedures possible around 6-7 years of age)
  - Plan for repeat ABR and imaging under anesthesia at 9 months of age
- Hearing aid consultation and fitting
  - Right Ear: Not a candidate for amplification.
  - Left Ear: Super power BTE recommended and fit
  - Follow-up concerns: constant feedback, minimal improvement in responses to sound

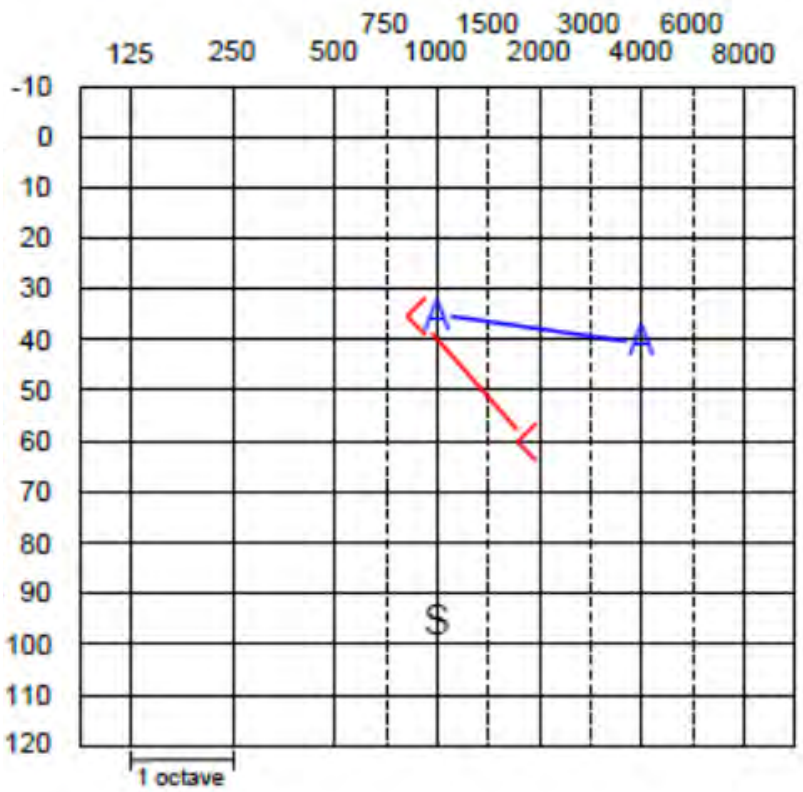
ABR and CT

- ABR
  - Right Ear: Profound mixed hearing loss
  - Left Ear: Severe to profound sensorineural hearing loss
- CT
  - Right aural atresia with small, dysplastic middle ear cavity. Fusion of malleus and incus with dysplastic stapes, which is misaligned with the oval window. Abnormal incudostapedial joint.
  - Abnormal course of the right facial nerve.
  - Left microtia with small, dysplastic middle ear cavity. Fusion of malleus and incus with absent stapes bone. Oval window not identified.

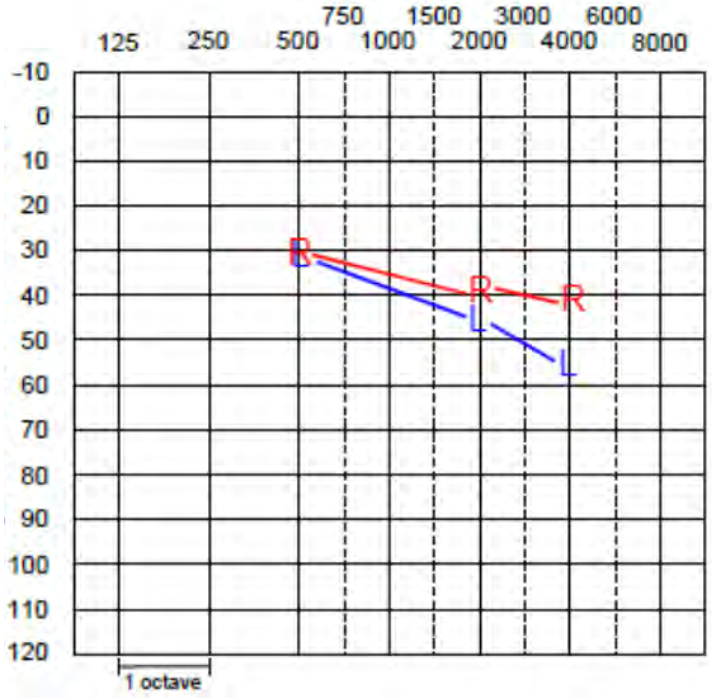




- Initial Cochlear Implant Evaluation
  - SP hearing aid reprogrammed to new ABR – improved response
  - Unaided testing confirmatory of mixed loss in right ear
  - Plan:
    - Pause!
    - May still be a candidate for left CI but further testing needed
    - Right ear needs trial with BAHA before determining CI candidacy
- Right BAHA fitting
  - Fit with super power BAHA on softband
  - New earmold fit for left hearing aid, extended trial recommended due to limited use

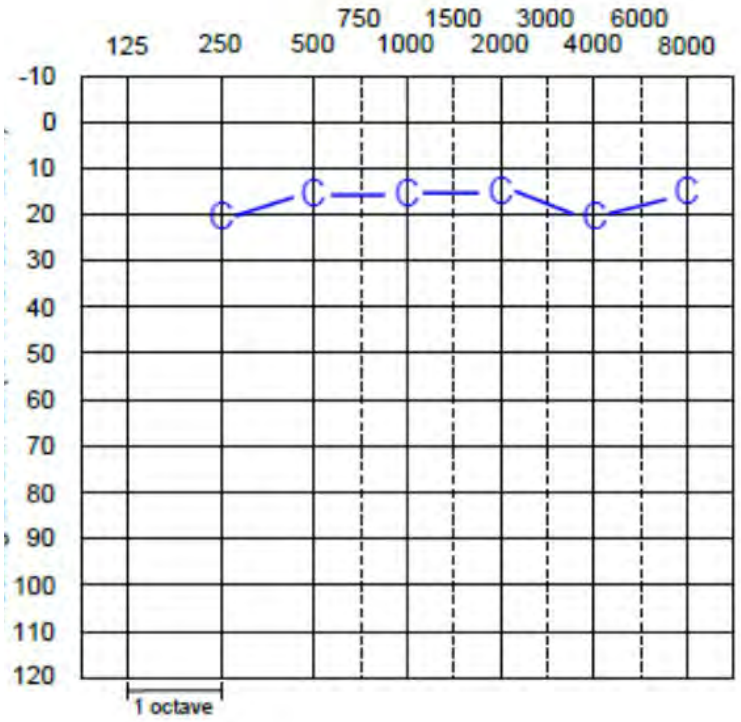


- Follow-up CI evaluation
  - Poor retention of both devices limiting use, BAHA better than hearing aid
  - Datalogging 0.8 hours/day average for hearing aid, 4.6 hours/day for BAHA
  - Responding better to sounds with BAHA than hearing aid by parent report
  - Babbling but no words
  - Patient noticing benefit from BAHA
  - Plan: Left CI!

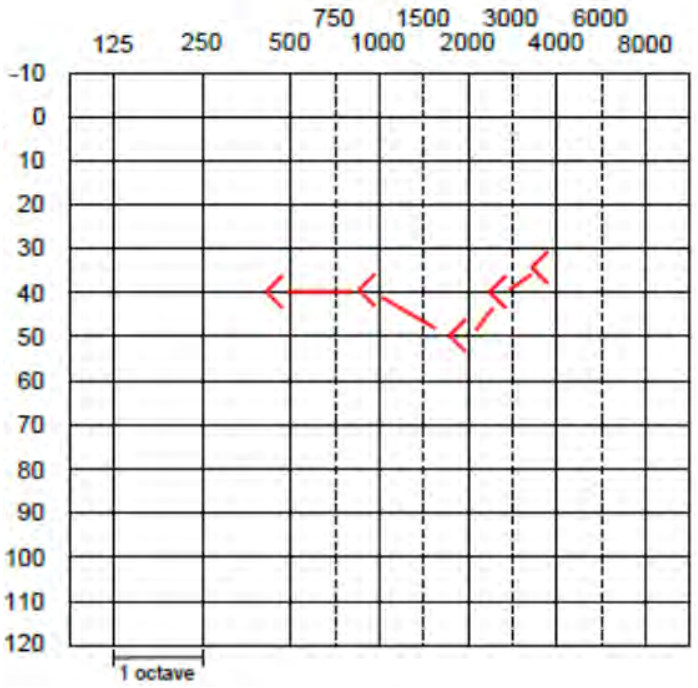


R = Right BAHA Aided Thresholds
L = Left Aided Thresholds

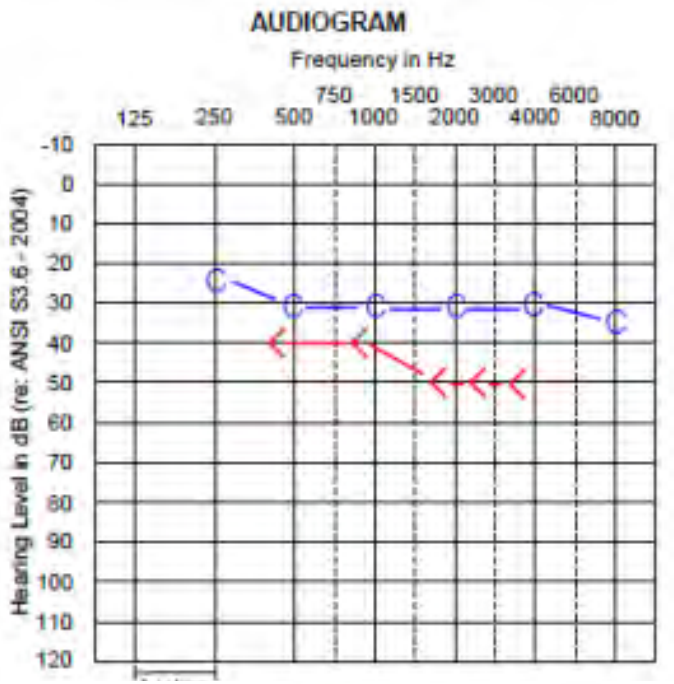
- Left cochlear implantation at 16 months of age
  - normal intraoperative testing
- Activation at 17 months
  - Activation of left cochlear implant
- First follow-up a week later datalogging was improved over hearing aids at 6 hours per day on average
- 3 weeks post activation
  - Improved responsiveness to sounds at home
  - First word!

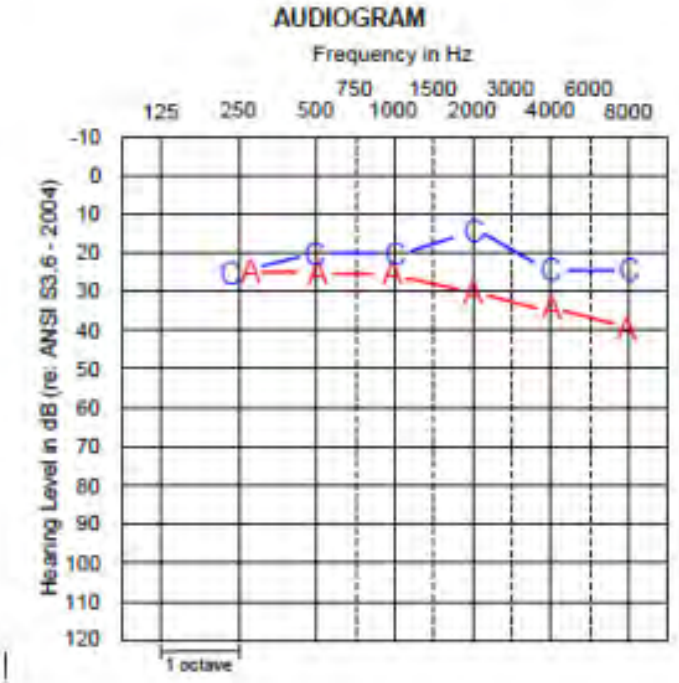


- 8 months post-activation
  - Excellent hearing and understanding reported
  - Limited speech progress due to facial nerve paralysis
    - Communicating with combination of spoken language, AAC, and ASL
  - Family interested in pursuing a sequential implant for the right ear
    - Upset if CI removed, doesn't care if BAHA is on or off
    - Rejecting use of BAHA at home
  - Datalogging for BAHA 9.4 hours/day average, for CI 8.2 hours/day average
  - Improved bone conduction thresholds



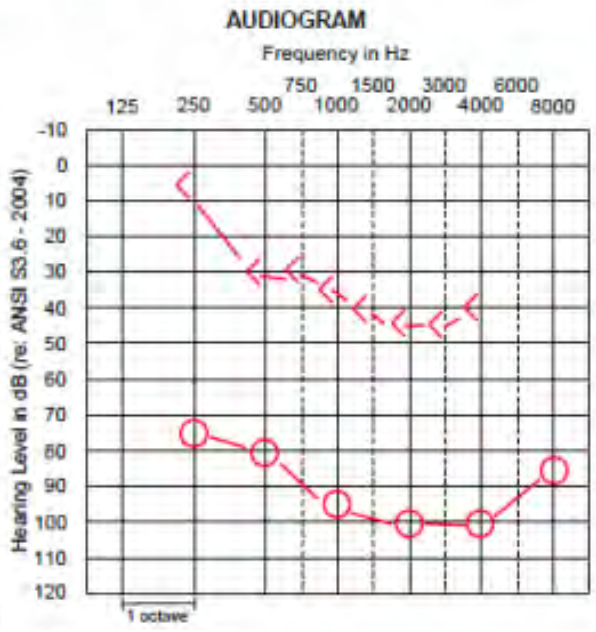
- Return after loss to follow-up
  - Continued preference for CI over BAHA
  - BAHA broken – plan to replace with new model
- Follow up after new BAHA fit
  - Overall doing well with both devices
  - Attending PDSD for pre-k
  - Speech continues to be poor
  - Continued preference for CI over BAHA
    - Irene now expressing that she doesn't like the BAHA



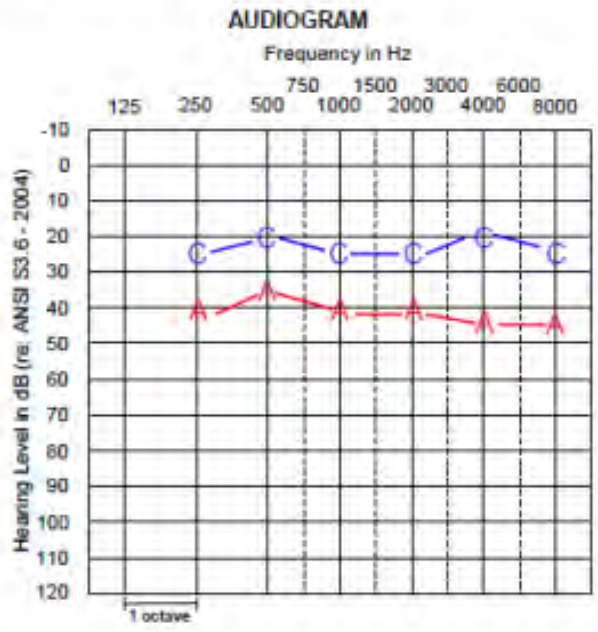


- Most recent visit
  - Continued preference for CI over BAHA
  - Family interested in sequential CI – discussion of implantable bone conduction solution prior to re-evaluating candidacy
    - Considerations for facial re-animation surgery/microtia reconstruction
  - What would it take to consider a cochlear implant for her mixed hearing loss?

Audiogram:



Aided Audiogram:



Veronique et. al, 2009

Closing Thoughts

Allie Sayer, AuD, CCC-A

Questions?

Contact Information

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