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Audiologic Diagnosis of Infants: Expect the Unexpected

**Sound Foundations 2010
November 8th, 2010
Chicago**

**Patricia Roush, AuD
University of North Carolina
School of Medicine
Chapel Hill, North Carolina USA**

Outline

- **Collin: A success story**
- **Diagnostic Challenges**
- **Case Illustrations**

Pediatric Hearing Program

University of North Carolina

Chapel Hill

- Pediatric audiology program within ENT clinic with close collaboration with ENT physician colleagues
- 350 diagnostic ABRs/year
- 100 hearing aid fittings/year
- 115 cochlear implants/year
- Total 1450 infants and children
 - 850 using amplification
 - 600 with cochlear implant
- 200 children with ANSD
- CASTLE Pre-school





**University of North Carolina
Department of Otolaryngology
UNC Hospitals
Chapel Hill, North Carolina**

Pediatric Audiologists:

Nissele Franco, AuD
Paula Johnson, AuD
Corinne Macpherson, AuD
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Patricia Roush, AuD

Otolaryngologists:

Craig Buchman, MD
Oliver Adunka, MD
Carlton Zdanski, MD
Harold Pillsbury, III, MD

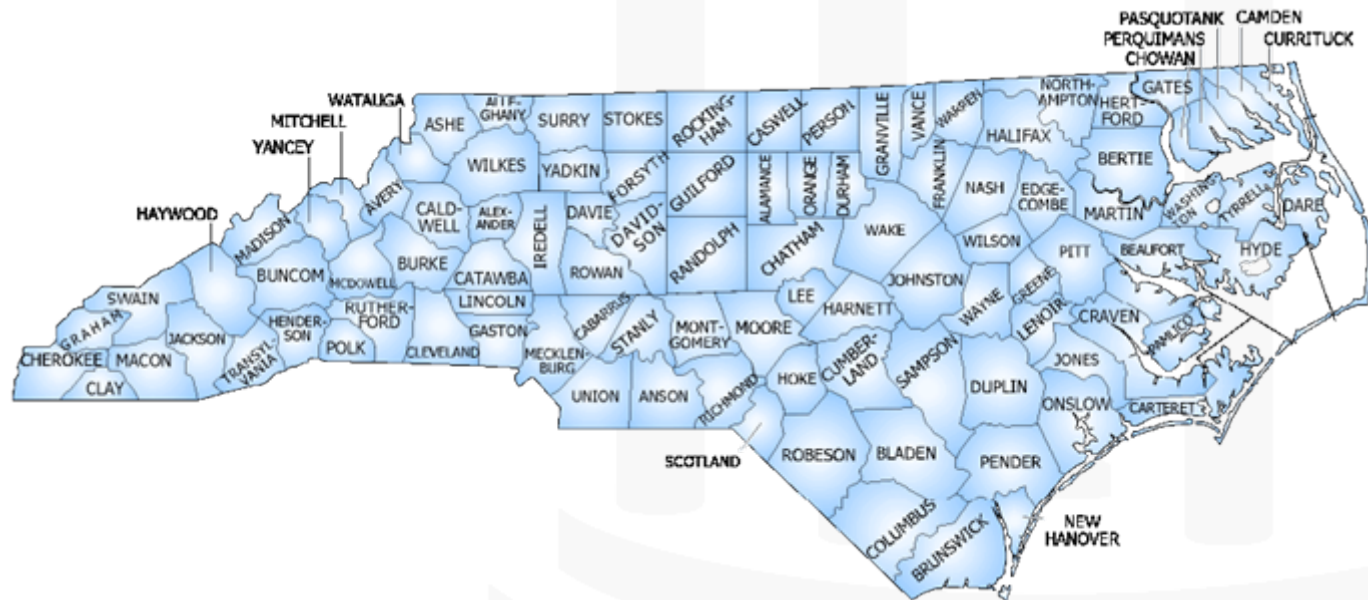
Pediatric CI Audiologists:

Holly Teagle, AuD
Jennifer Woodard AuD
Deborah Hatch, AuD
Lisa Di Maria, AuD



EHDI in North Carolina

- Passed newborn screening legislation in 1999
- Started screening in 2000
- 130,000 births per year
- Screening approximately 98% of infants in 88 hospitals





Management of Hearing Loss in Infants: A Continuum of Care

- Timely referral from NB Screen
- Comprehensive audiologic assessment
- Otologic examination
- Referral for intervention
- Selection of amplification
- Hearing instrument fitting and verification
- Hearing aid orientation
- Behavioral audiometry and readjustment of hearing instruments
- Ongoing audiologic, otologic and intervention services
- Referral for CI when indicated



Collin's Story (with his family's permission!)



A Partnership



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Collin's Story (Current Age: 9 years)

- AABR in outside well-baby nursery
 - » Failed R&L 35dB, bilaterally
- AABR re-screen at 6 days of age
 - » Failed R&L 35dB, bilaterally
- Diagnostic ABR at outside facility:
 - » Clicks: 55 dBnHL-R&L
 - » Referred to UNC for frequency specific ABR and possible hearing aid fitting
- Diagnostic ABR at UNCH at age 6 weeks:
 - » Tone Burst ABR with air conduction and bone conduction confirms mild to moderate SNHL, bilaterally.
 - » Tympanometry: normal (1000Hz probe tone)
 - » Otoacoustic emissions: absent



Age Six Weeks: Ear Impressions



Age 8 Weeks: RECDs Measured Hearing Aids Programmed and Verified



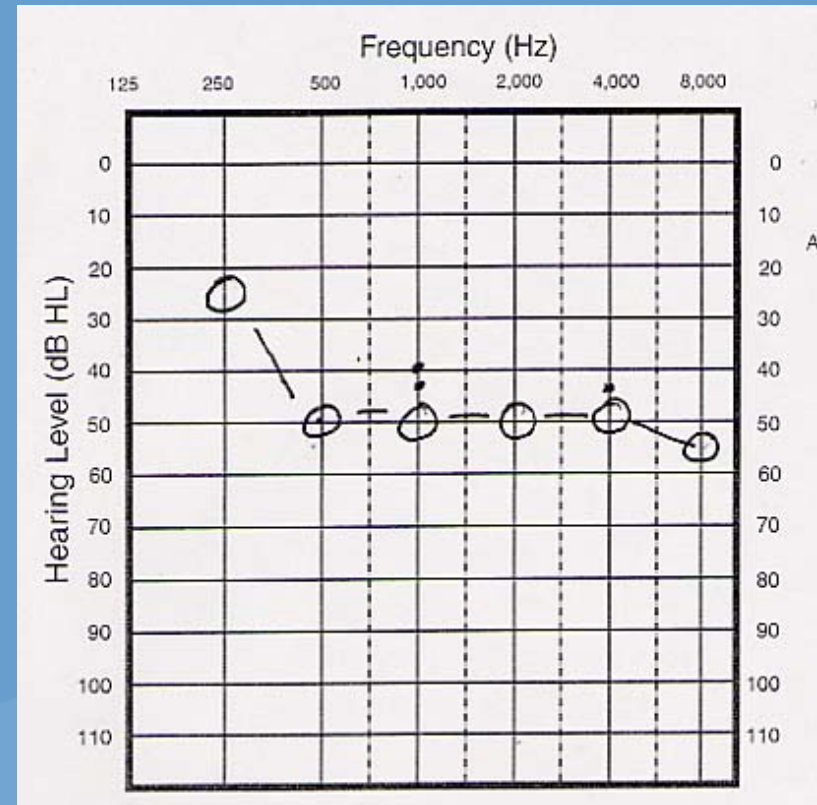
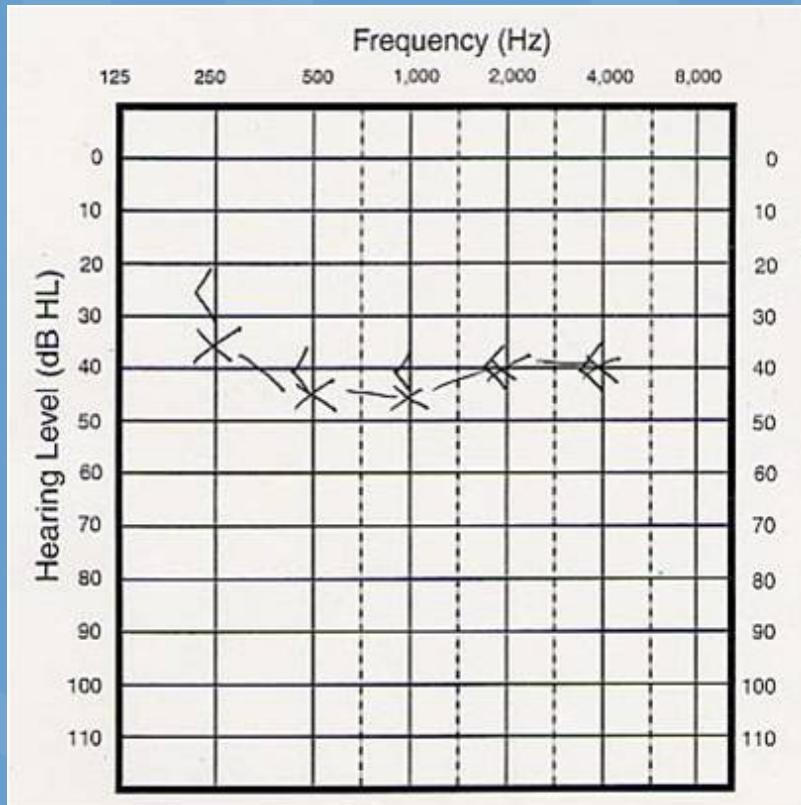
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Left Ear	250	500	1000	2000	4000
HTL	15	20	45	45	45
RECD	7	10	12	11	11



Age 7 months VRA with insert earphones





Twelve Months: FM System Dispensed





Developmental Assessment at age 1 yr, 2 months

	Raw score	Percentile	Standard Score	Age Equivalent
Cognition	88	55	102	1yr., 3 months
Language	61	58	103	1yr., 3 months
Social	70	58	103	1yr., 4 months
Motor	80	58	103	1yr., 4 months
Overall	185	54	103	1yr., 4 months

Calls family members by name

Combines talking & pointing to make wishes known

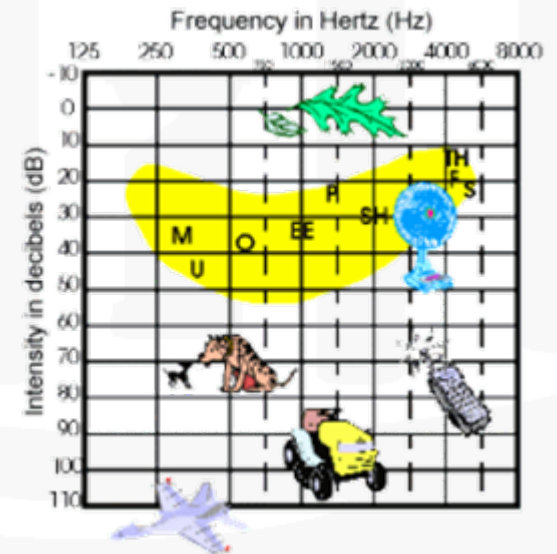
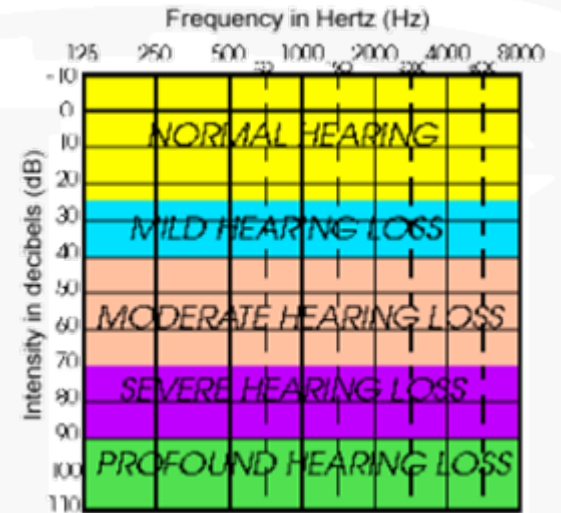
Uses several words others understand

Difficulty maintaining EI services

Brochure Developed by Family for Kindergarten Teachers



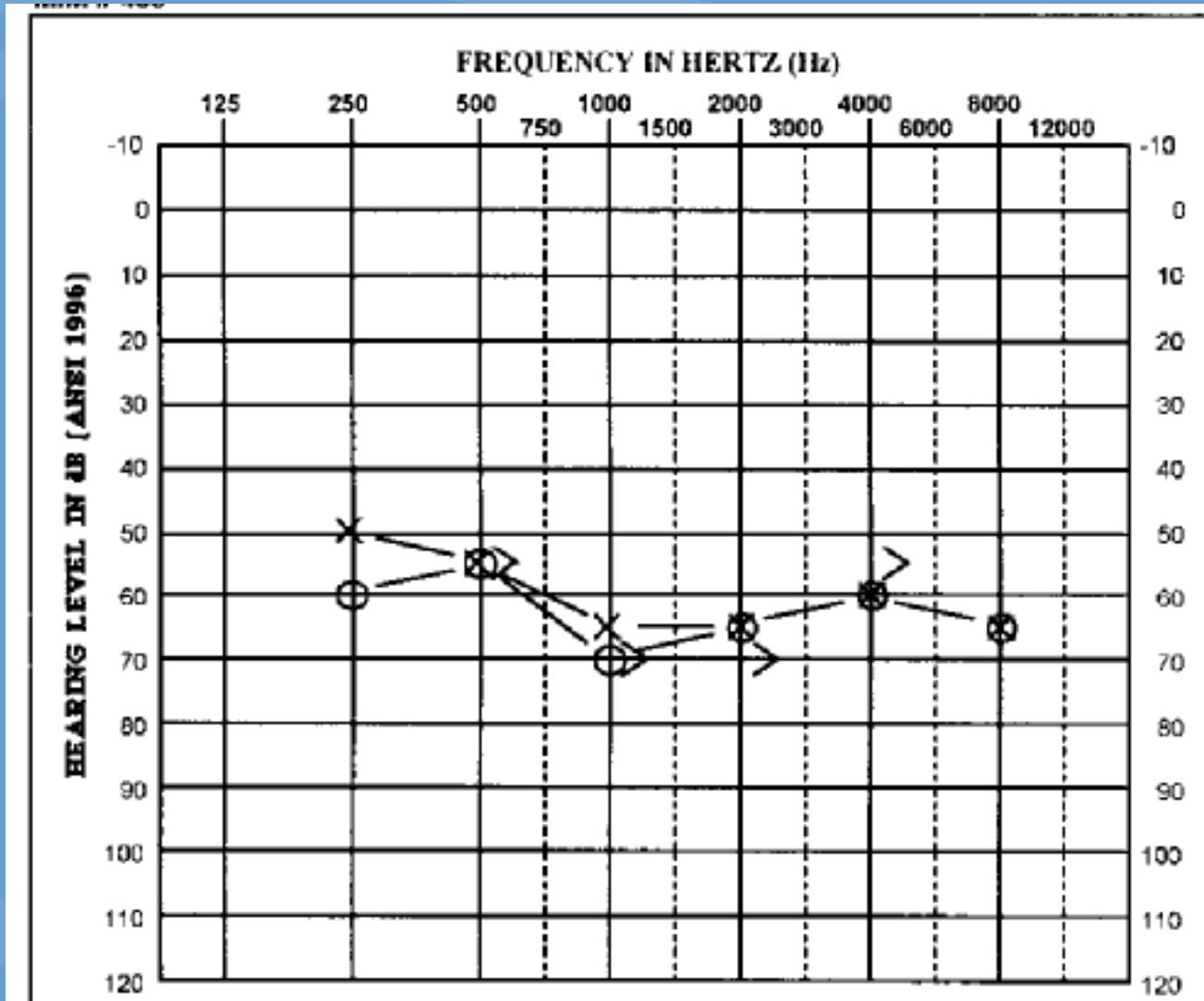
- Hello,
- My name is Collin and I am very excited about being in your class. I am introducing myself to you so you can understand some things about my hearing loss.
-
- MY HEARING LOSS
-
- I was born with a mild to moderate sensorineural (permanent) hearing loss and now I have moderate hearing loss.
-
- I have been wearing hearing aids since the age of 8 weeks.
- I can still hear some without my hearing aids, but I probably would not hear all the sounds. If the need arises for you to talk to me without my hearing aids in, please talk normal volume about 1 to 2 feet from my ears.



Current Audiogram



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Use of FM During Extacurricular Activities



Access to Technology





Collin Speaking to NC State Legislators in Support of a Bill Requiring Insurance Companies to Cover Hearing Aids for Children





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Collin with Governor at Signing of Hearing Aid Insurance Bill





**This may be an exceptional
case...**

**But if we aim high enough we
can work toward making
exceptional the norm**

Factors that may influence outcomes

- Degree of hearing loss
- Age at diagnosis, hearing aid fitting and enrollment in intervention
- Family participation
- Additional medical challenges
- Quality of audiologic intervention
- Quality/quantity of early intervention services
- Availability of funding for needed technologies

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CHAPTER SEVEN

Potential Pitfalls in the Audiological Assessment of Infants and Young Children

Judith S. Gravel

Introduction

Progress made over the last several years in early identification of hearing loss through implementation of newborn hearing screening (NHS) programs has created a unique opportunity for audiologists to provide amplification to children with permanent hearing loss (PHL) very early in life. Along with this long-awaited opportunity comes the daunting responsibility for accurately determining the type, degree and configuration of an infant's hearing loss so that appropriate counseling of parents, fitting of hearing aids, management planning and follow-up can proceed in a timely manner. Also critically important is to confirm the presence of normal hearing in a baby who failed NHS (a false-positive outcome), so that parental anxieties can be alleviated and unnecessary interventions avoided.

Key to this process is the comprehensive audiological assessment. Current otoacoustic, physiological, electrophysiological and behavioral techniques and procedures have provided the pedi-

sound foundation for optimal amplification device provision.

This chapter will address some of the potential pitfalls that audiologists might encounter when assessing the hearing of infants and young children. Three types of errors that could be made in audiological diagnosis will be described. Case studies will illustrate when clinicians might encounter audiological pitfalls, why the pitfalls occur, and comment on how clinicians might avoid these obstacles to accurate, timely and appropriate service provision.

Background

The Year 2000 Joint Committee on Infant Hearing Position Statement Principles and Guidelines (JCIH 2000) state that all infants should have access to hearing screening in the neonatal period regardless of risk status (i.e., universal newborn hearing screening). The goal for infants who fail NHS is to begin



Types of Audiologic Misdiagnosis

- Hearing Loss Identified correctly but incorrect conclusion regarding type or degree of permanent hearing loss (PHL) is made
- False Positive, i.e., error of diagnosing a child with PHL when normal hearing exists
- False Negative, i.e., following the audiological evaluation, child with true PHL is diagnosed with normal hearing (most serious)

~Judy Gravel, Sound Foundations 2001
(Potential Pitfalls in the Audiologic Diagnosis of Infants and Young Children)



Why Should Pediatric Audiologists be Concerned and Continually Vigilant about Misdiagnosis?

- Delay in confirmation of child's true hearing status
- Delay in the referral of a child for a potentially treatable medical condition
- Delay in referral of child and family for beneficial early intervention services
- The provision of inappropriate management/therapies (audiological, medical, surgical, prosthetic, educational, communication)
- Parental anxiety, confusion and loss of confidence in recommendations made by the clinician
- Unnecessary expenditure of resources by the family (e.g. emotional, time, monetary) and by the system

~Judy Gravel, Sound Foundations 2001



Case Illustrations





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CASE #1



Background:

- Full term baby born without complications
- Both parents are deaf
- Newborn Screen (Day 1) with AABR:
 - » Failed right
 - » Passed left
- Repeat Screen 2 weeks of age (AABR):
 - » Failed bilaterally



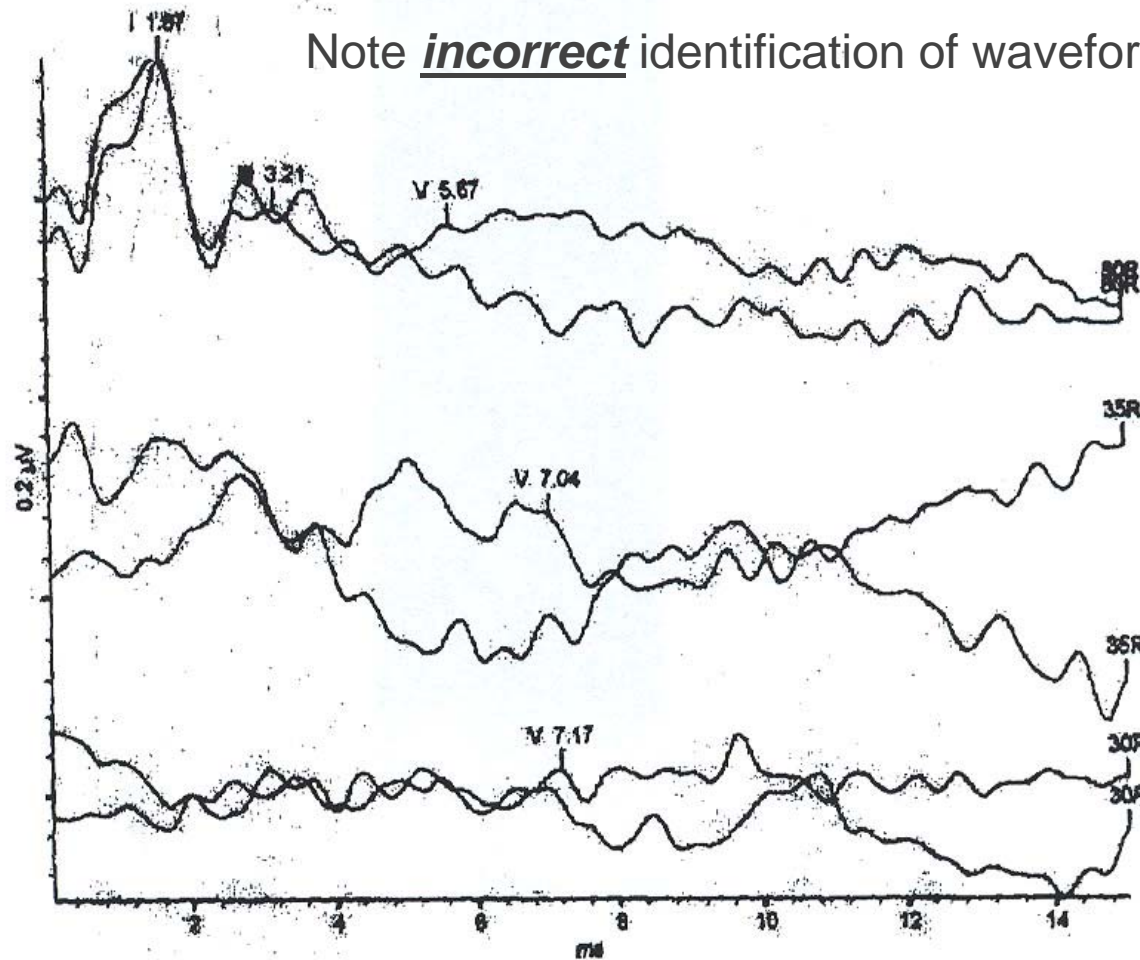
Review of External Records

**First Diagnostic Evaluation at Outside Clinic
Age: 1 month**



ABR Waveforms (Click Stimuli)

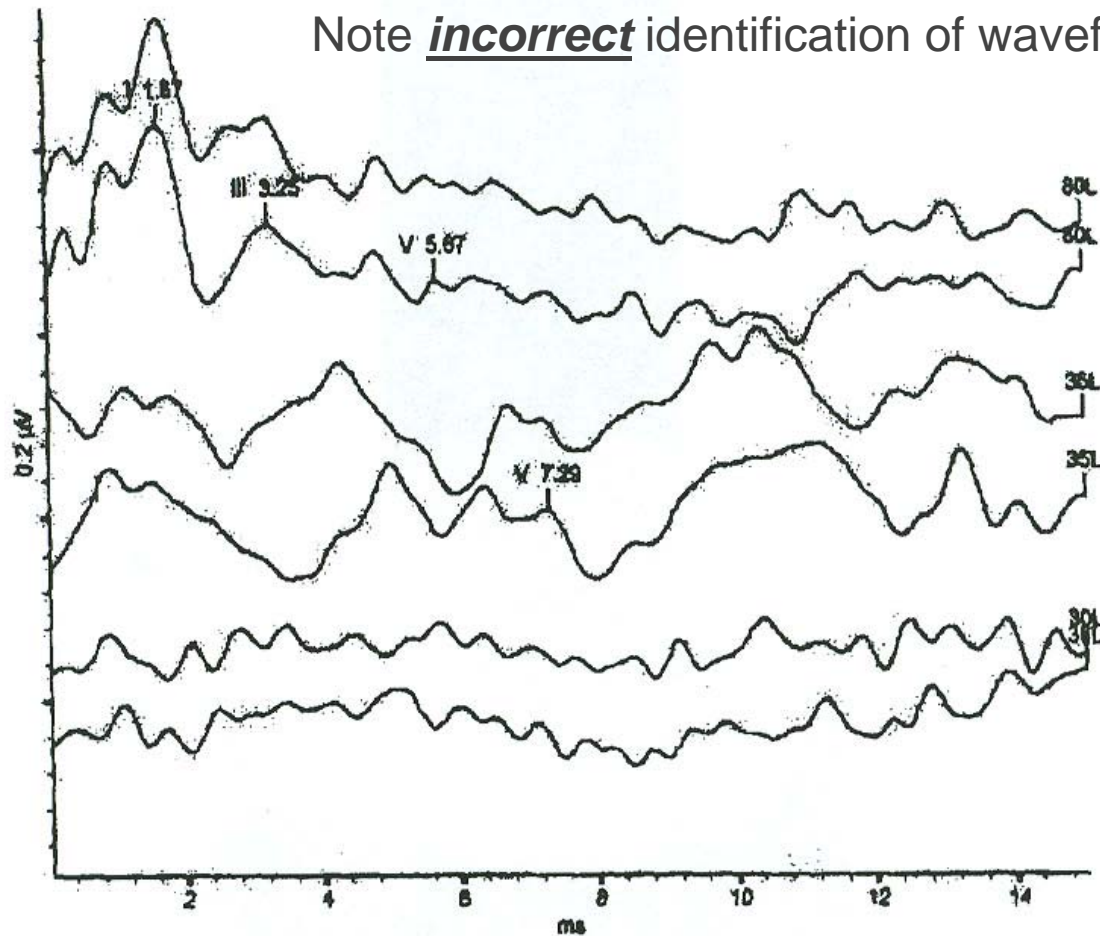
Age: 1 Month





ABR Waveforms (Click Stimuli)

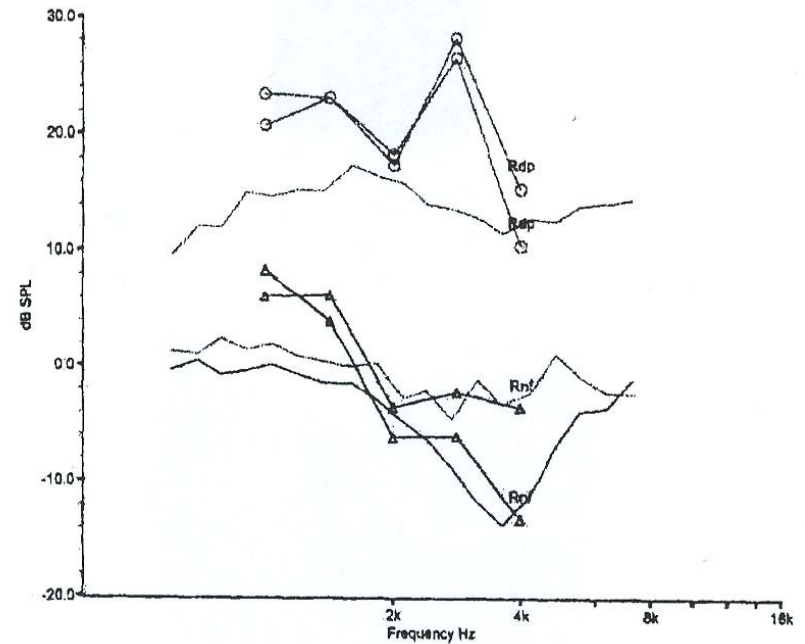
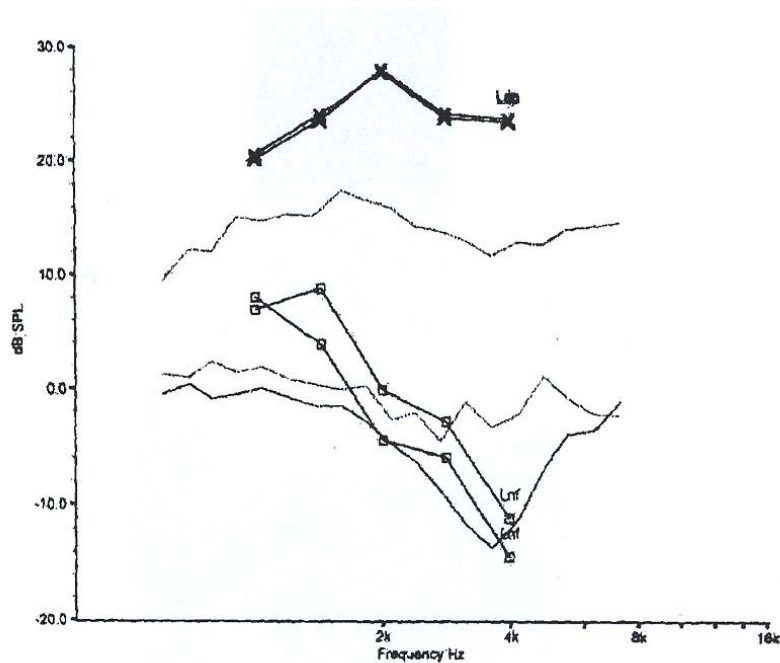
Age: 1 Month





Otoacoustic Emissions (DPOAEs)

Age: 1 month





Report of Diagnostic Testing at ENT Office

Age 1 month:

Test Results:

- *“ABR testing using clicks yielded reliable response down to at least 30dBnHL for the right ear and 35dBnHL for the left ear.”*
- *“500 Hz tone burst produced reliable response down to 35dBnHL for each ear”*
- Tympanometry: Normal(1000Hz probe tone)
- Acoustic reflexes not be done due to lack of seal
- **Robust DPOAEs from 1000-4000 Hz**



Summary of Findings

Age 1 month:

Impressions:

- *“Results of ABR testing rule out a significant hearing loss for each ear at 500Hz and within the 1000-4000Hz region.”*

Recommendations to Family:

- *Return in six months for follow up behavioral assessment*



Second Diagnostic Evaluation

Age: 7 months

(ABR, no behavioral testing)



Follow Up Visit in ENT Office

Age: 7 months

Background:

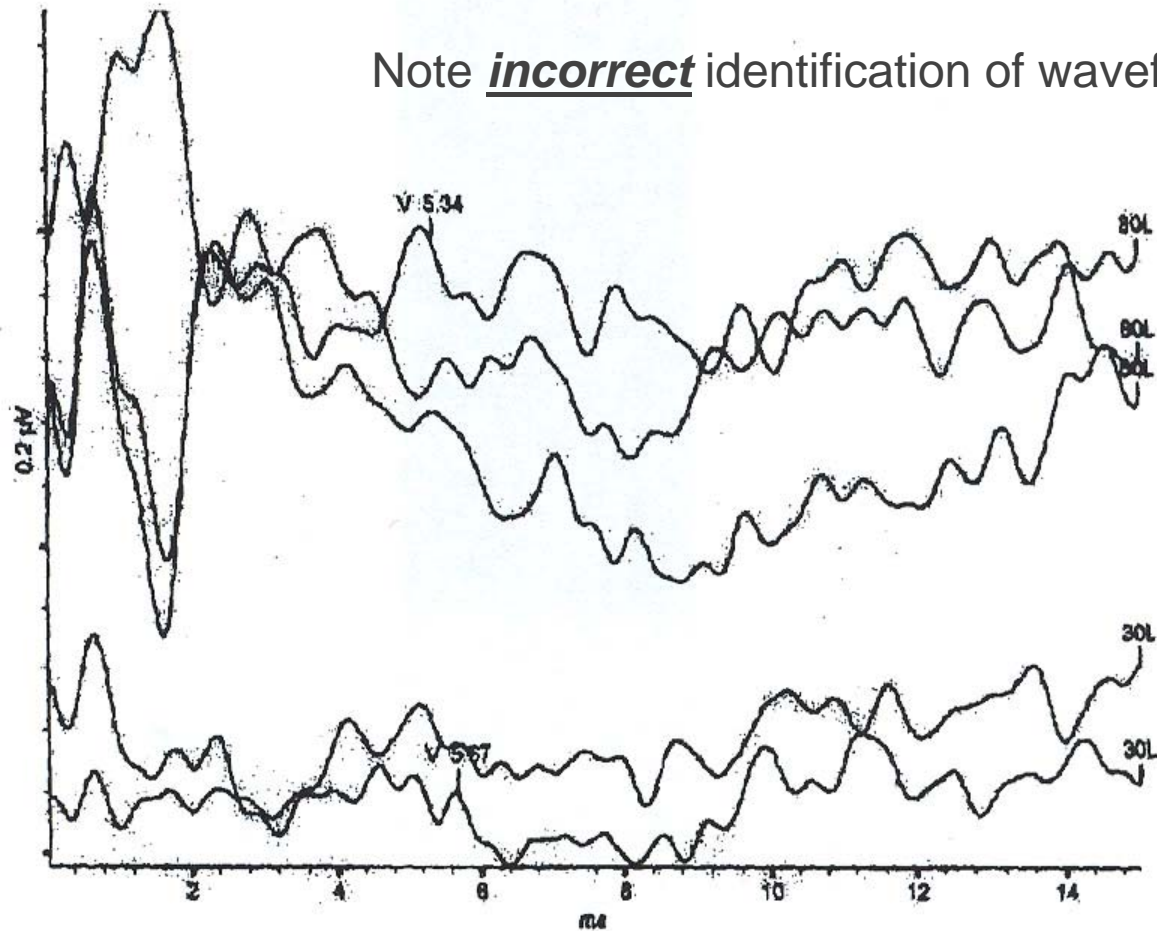
- Parents (both deaf) and maternal grandmother returned for follow up as recommended and expressed concern about the baby's hearing.
- Written report stated:
 - » *“Grandma reported very little speech and language however child's environment essentially silent given that mom and dad are deaf. TV is on mute, telephone ringers are off, etc.”*



Follow Up Visit at ENT Office

Age: 7 months

ABR Waveforms (Click Stimuli)





Follow Up Visit at Local ENT Office

Age: 7 months

Test Results:

- ABR to click stimuli yielded a reliable response for intensity levels down to at least 30dBHL for each ear.
- Tympanometry consistent with normal middle ear function
- **Robust DPOAEs (1000-4000Hz)**

Impressions:

- ***“Results of ABR testing again rule out a significant hearing loss for each ear in the 1000-4000Hz region.”***

Recommendations made to family:

- *“Child has normal hearing”*
- *“Family should be able to stimulate child’s speech and language by talking to him during daily care activities, reading to him, presenting sounds, etc.”*
- *“Return for behavioral audiometry in 3 months”*



Family Decisions

Age: 12 months

- Parents (college educated) concerned child is not developing speech
- Wondering if child is not getting enough exposure to spoken language in their home
- Both parents quit jobs and move 200 miles to live with grandparents



First Contact with UNC

Age: 19 months

- Audiology colleague called to ask if we would see the child in our center for evaluation.
 - » Grandparents contacted him to say their grandson failed his newborn screen but they were told child had normal hearing.
 - » When child did not seem to respond to sound they took him to a another ENT office for behavioral assessment and were told child appears to be deaf
 - » Colleague asked if we would evaluate child in view of varying opinions



Background

- Grandmother contacted us and provided additional background information by telephone
- Requested records of previous testing included ABR reports with waveforms
- Two day appointment scheduled:
 - » Day 1:
 - Behavioral audiometry
 - Acoustic Immittance Measures
 - Otoacoustic Emissions
 - ENT consult
 - » Day 2:
 - Sedated ABR

Behavioral Audiometry

Age: 19 months

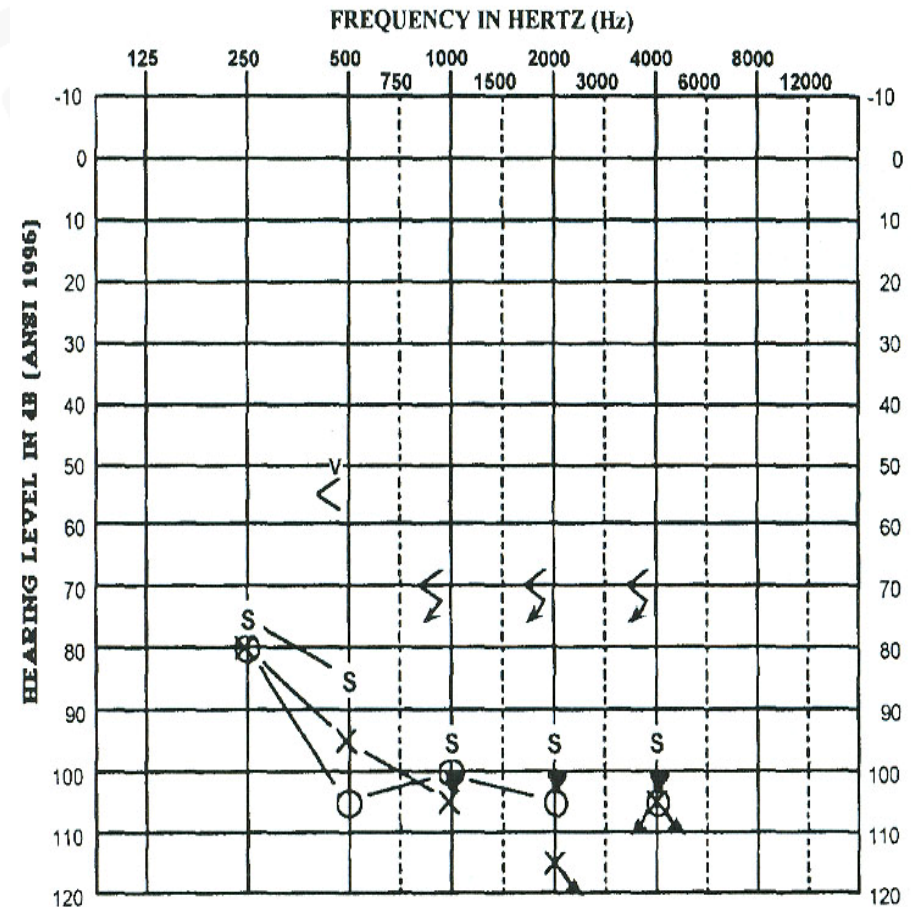
Testing completed with VRA using insert earphones

Results consistent with severe to profound bilateral SNHL

Tympanometry: normal

Communication status:

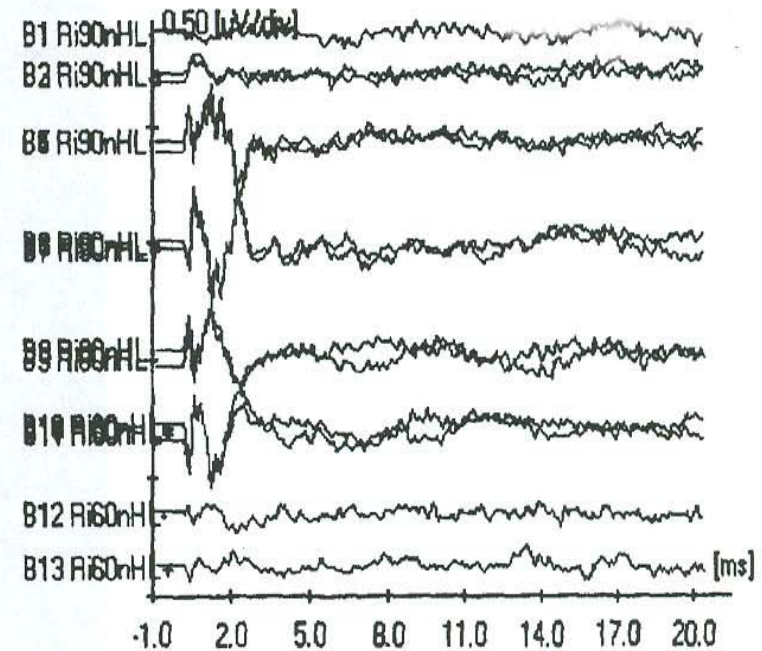
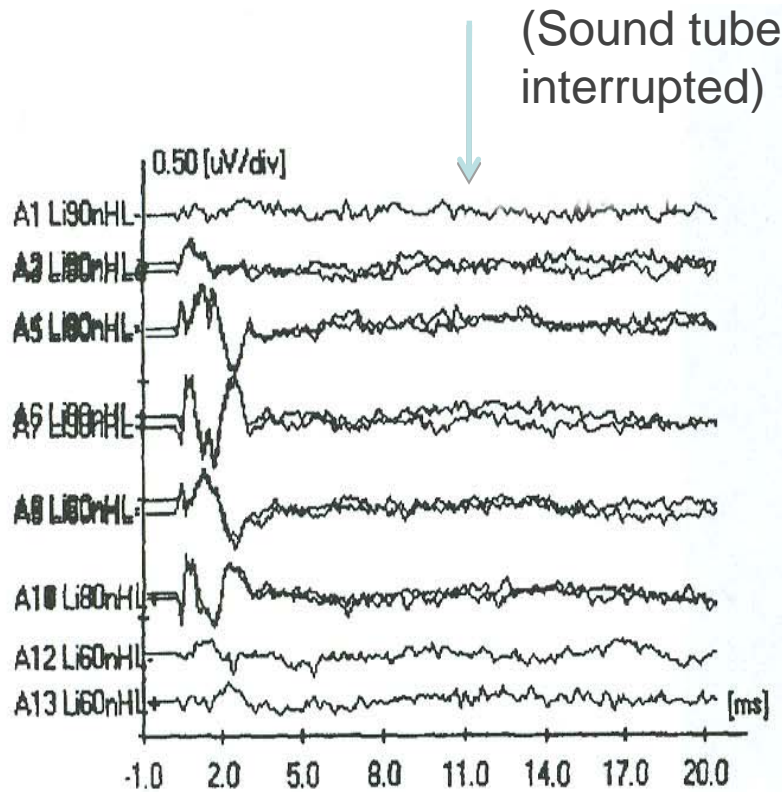
Child understands 25 signs, uses 15 signs, does not use any speech





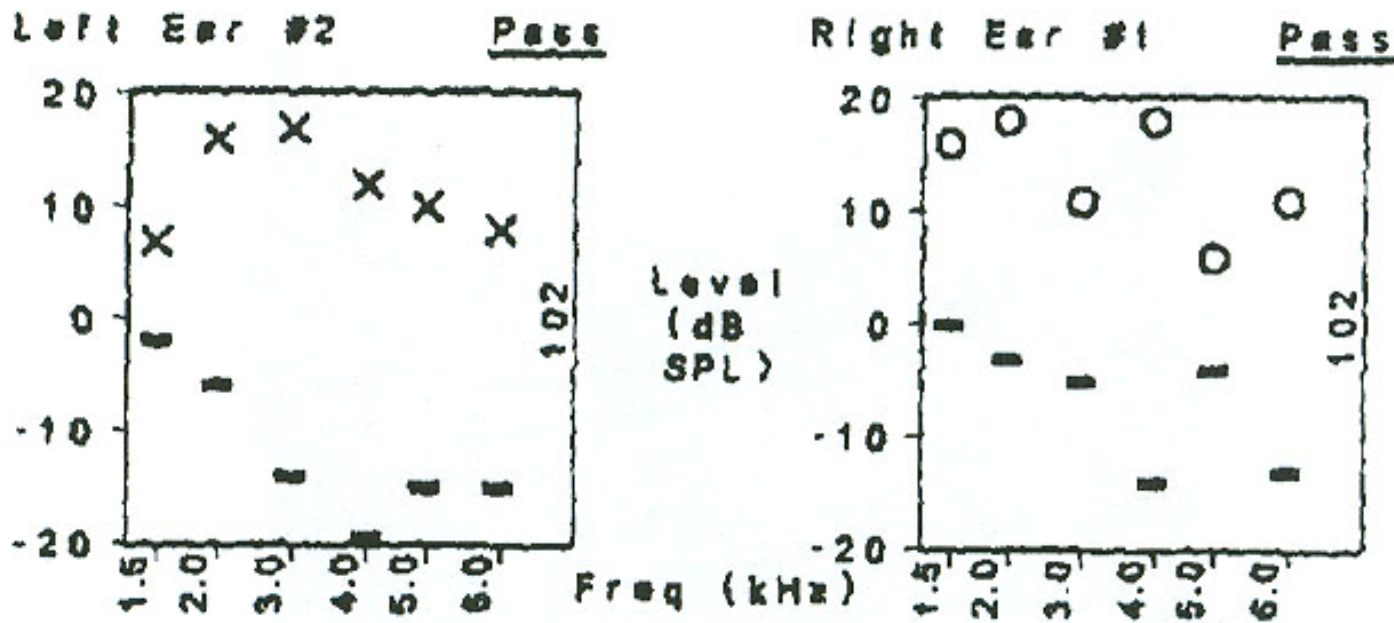
ABR Waveforms (Click Stimuli)

Age: 19 Months



Otoacoustic Emissions (DPOAEs)

Age: 19 months





Summary of Test Results

UNC Hospitals

Age: 19 months

- Ear exam:
 - » normal
- EKG:
 - » normal
- MRI:
 - » normal
- Connexin test:
 - » negative
- Otoferlin test:
 - » *Positive*



Discussion with Family on Day 2

- Child has a severe to profound HL with ANSD pattern (ABR abnormal, OAEs present)
- Discussion of family's communication goals for their child
- Explained spoken language development will be difficult even with consistent hearing aid use in view of severity of hearing loss
- CI likely to provide greater access to auditory information if their goal is for their child to develop speech in addition to his sign language skills
- Family receptive to consideration of CI
- Contact information given for another family with two deaf parents who chose CI for their child



Decisions Family Made

- Age 20 months:
 - » Enrolled child in EI
- Age 21 months:
 - » Hearing aid fitting
 - » Contacted other families whose children have CI
- Age 21 months:
 - » Evaluation with pediatric CI team
 - » Family decided to move forward with CI
- Age 26 months:
 - » Received right CI
 - » Neural response telemetry showed robust responses for electrodes tested
- Mother, who had previously discontinued HA use, interested in resuming HA use



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What went wrong?

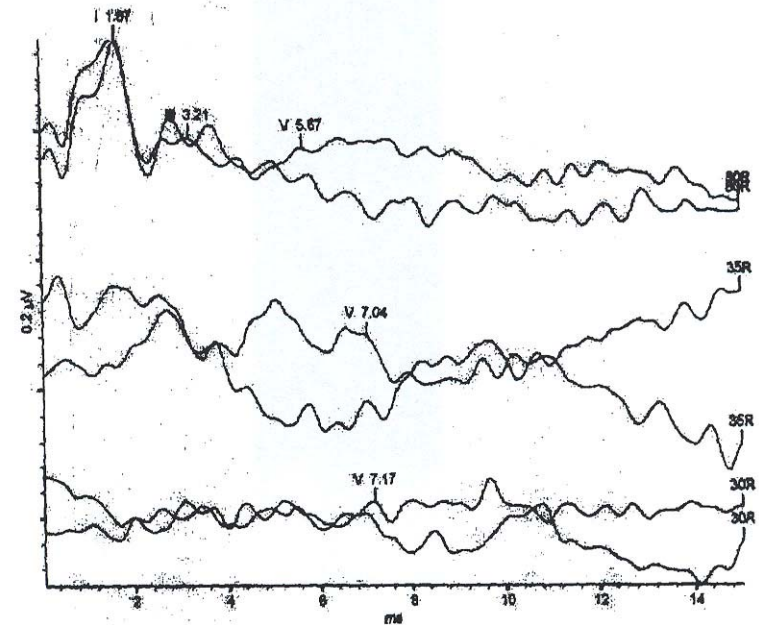
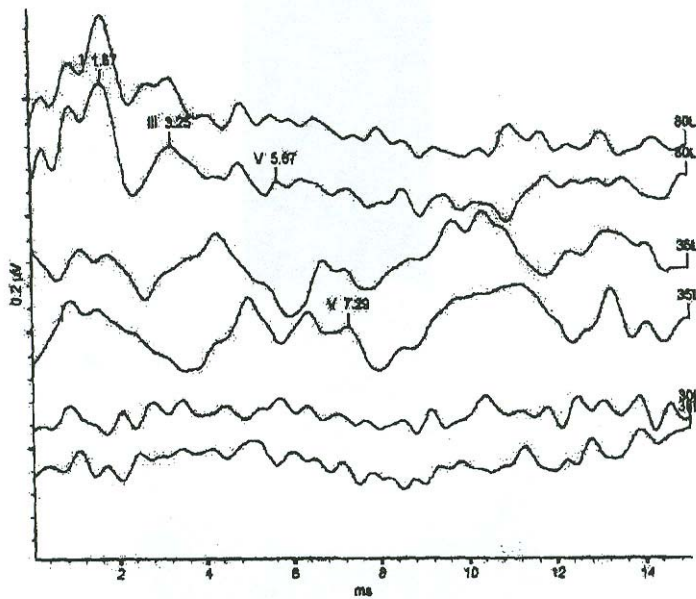
What went wrong?

- Initial Diagnostic Test at one month:
 - » Present OAEs following failed ABR at birth with deaf parents should have raised red flag for possible AN
 - » Poor reproducibility of waveforms
 - » Only rarefaction clicks used without condensation clicks at high intensity level
 - » Despite interpretation of normal hearing, later component waveforms not identified
 - » No shift in latency identified with decreasing intensity levels



Review of ABR #1 (Age 1 month)

Note incorrect identification of waveforms

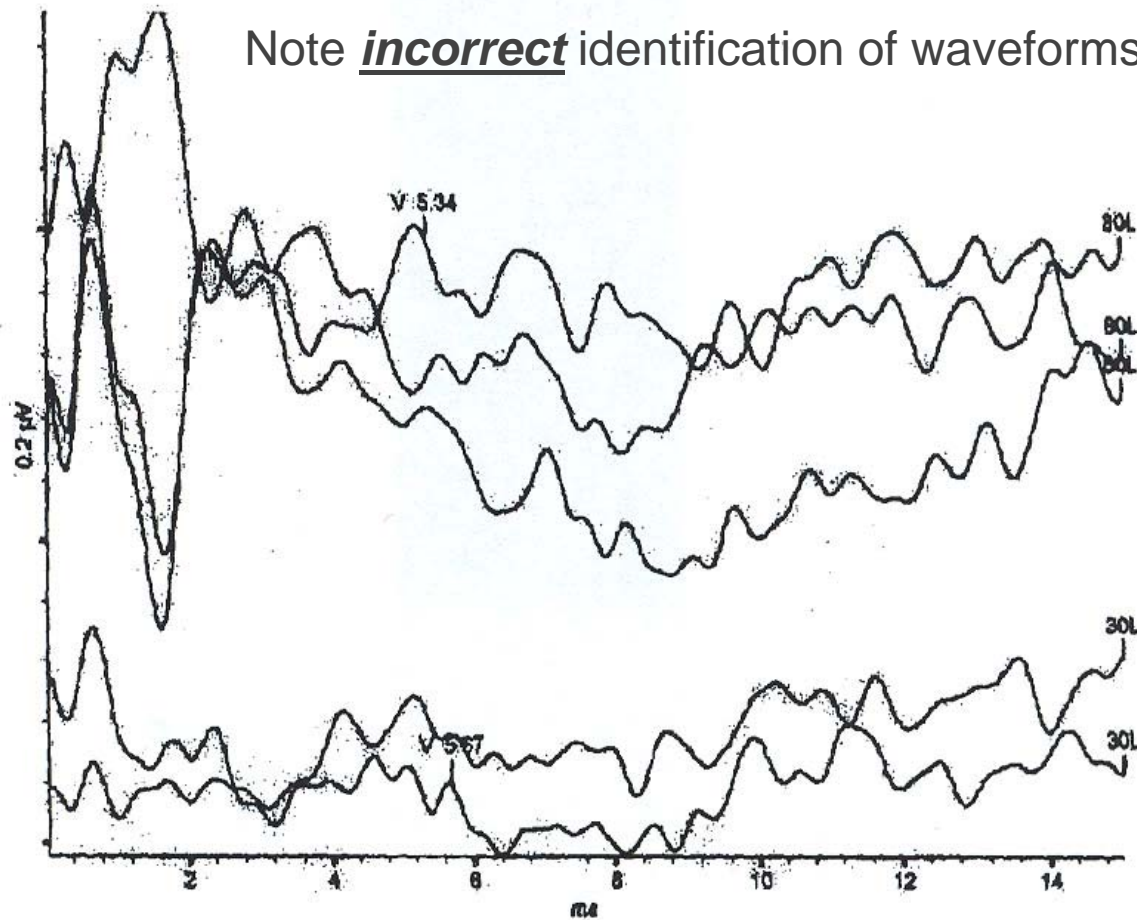


What went wrong?

- Diagnostic Test at 7 months of age:
 - » Both rarefaction and condensation polarities used but CM not identified or commented on (perhaps it was considered stimulus artifact)
 - » Stimulus artifact can be ruled out by completing a run with the sound tube interrupted
 - » Behavioral audiometry using VRA with insert earphones and bone conduction should have been completed to confirm hearing status, particularly in view of family history of deafness.
 - » Family's concern that child not responding to sound were not addressed



Review of ABR #2 Age: 7 months



Conclusions

- **Many children with ANSD pattern are incorrectly diagnosed**
 - » Some diagnosed with profound hearing loss who have lesser degrees of hearing loss, including some with normal hearing sensitivity
 - » Some, as in this case, diagnosed with normal hearing based on present OAEs and poor quality ABR
 - » Important to have a protocol that allows detection of abnormal morphology and presence of CM
- **While ANSD is more common in premature infants, it can also be found in normal, full term infants as in this child with an otoferlin mutation.**



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CASE #2

Background:

- Two year old child aided bilaterally moved into NC from another state.
- Mother brought all records from outside evaluations.
- History:
 - » 26 week preemie born with collapsed lung
 - » Ventilator 4 days, bili lights, oxygen 1 month, 2 blood transfusions
 - » Newborn Screen with AABR:
 - Failed AABR bilaterally

Review of Outside Records:

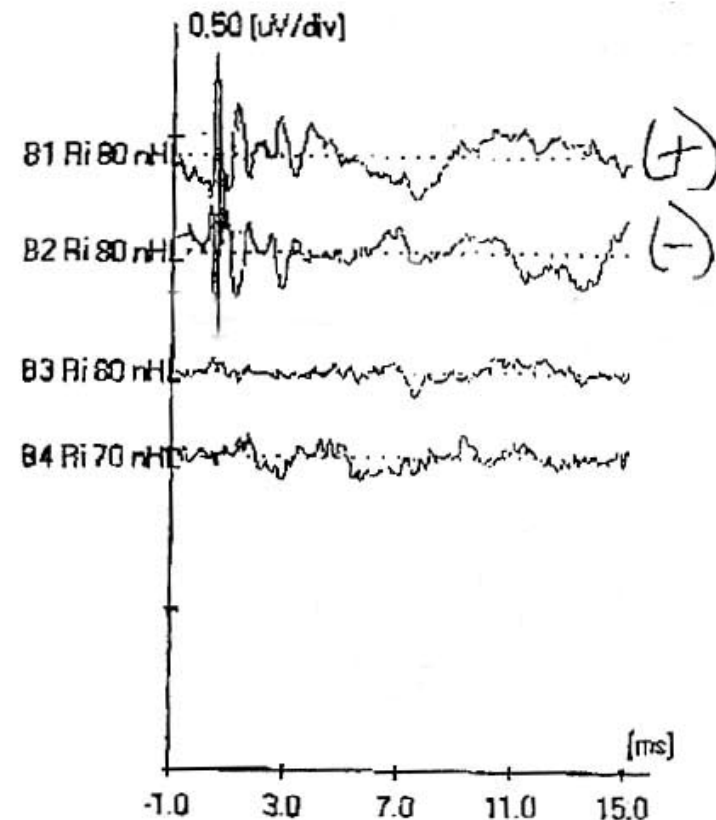
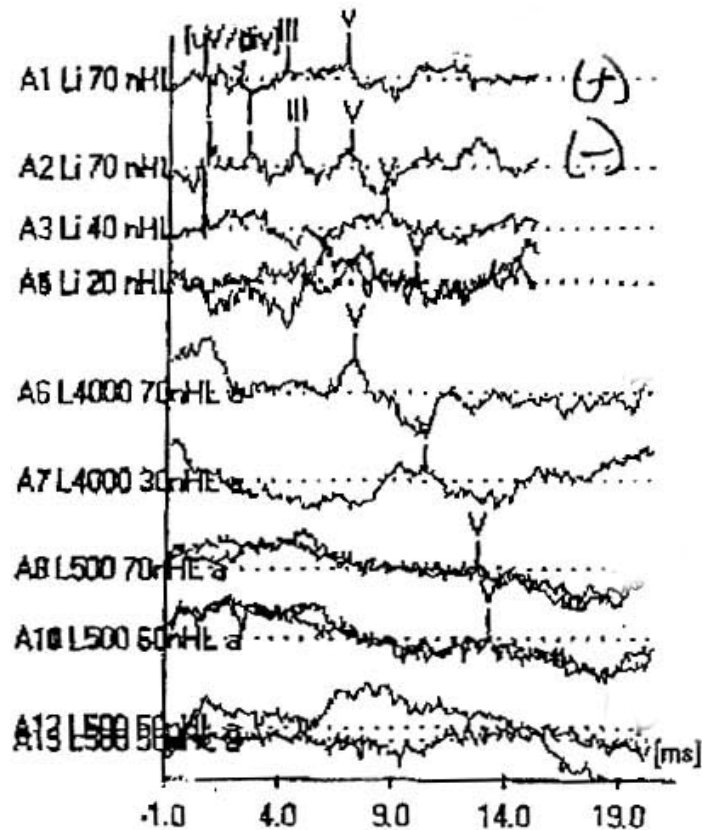
- ABR#1 (Age 1-2 months)
 - » Inconclusive due to movement and poor probe fit
- ABR#2 (Age 3 months)
 - » Clicks and tone bursts
 - » Click max. intensity levels: 80dBnHL RE; 70dBnHL LE
 - » **Summary:**
 - RE: Auditory Neuropathy;
 - LE: Borderline normal to normal hearing
 - “Hearing is adequate for speech and language acquisition”
- ABR#3 (Age 6 months)
 - » Clicks and tone bursts
 - » Click max. intensity levels used: 70dBnHL RE; 85dBnHL LE
 - » **Summary:**
 - Bilateral Auditory Neuropathy
 - *“These results indicate change from previous study”*



Review of ABR #2

Interpretation from Outside Clinic:

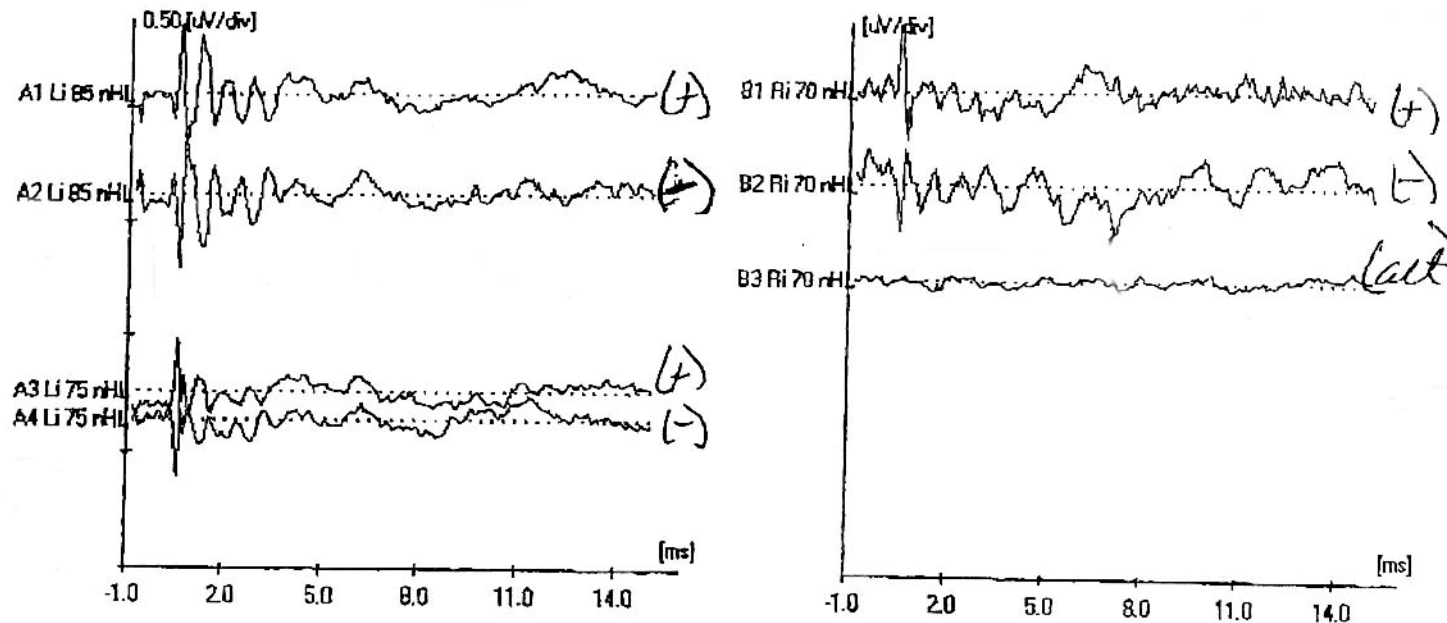
AN R.E.; Borderline Normal to Normal Hearing L.E.





Review of ABR #3

Interpretation: Bilateral Auditory Neuropathy



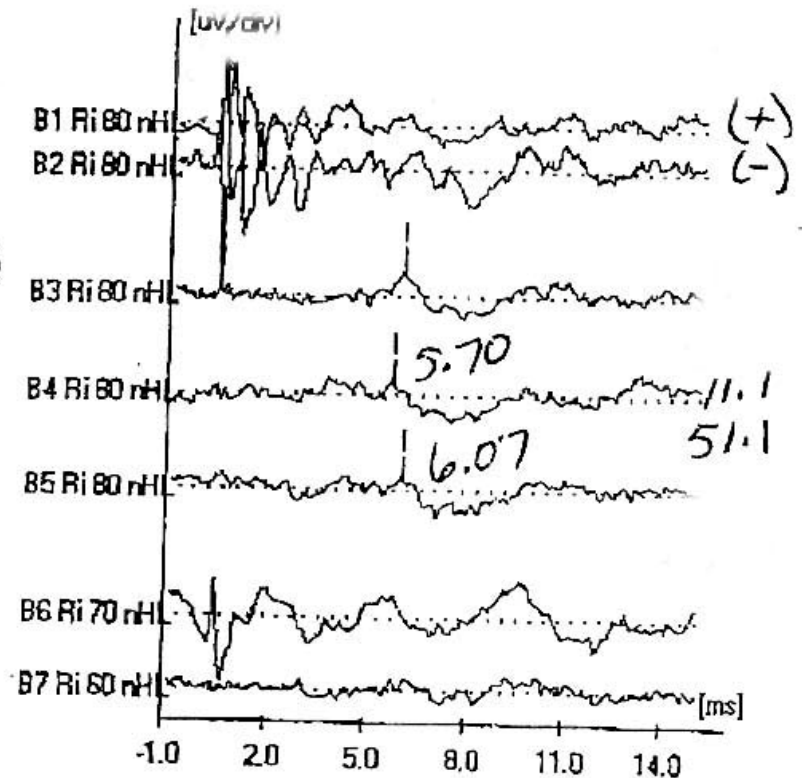
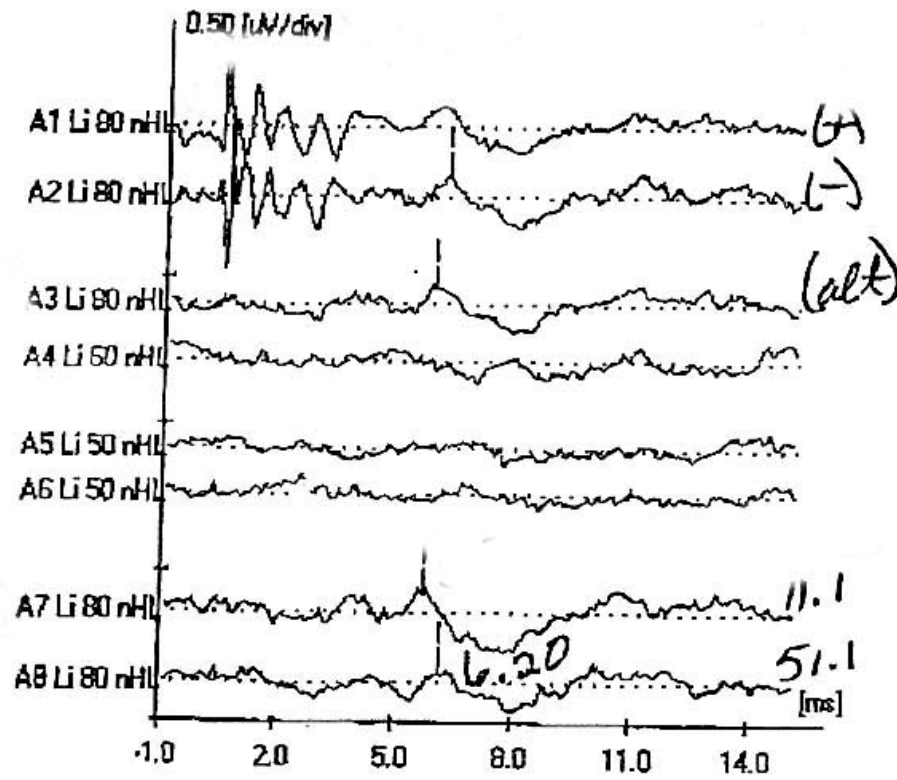
Review of Outside Records (continued)

ABR #4, Age 6 ½ months

- » Clicks and tone bursts
- » Maximum intensity levels for clicks (80dBnHL)
 - **Summary:**
 - “Bilateral Auditory Neuropathy”
 - Clicks show only a CM for RE and LE
 - Tone bursts: Absent RE; mild to moderate loss LE
 - **Recommendations:**
 - Trial with amplification and evaluate for CI candidacy



Review of ABR # 4 from Outside Clinic



Review of Outside Records (continued)

Behavioral Audiometry, Age 7 months

- Sound Field Testing:
 - » Speech awareness at 35dBHL
 - » Startle at 90dBHL
 - » Not responsive to pure tones
 - » OAEs: RE refer, LE pass
 - **Summary:**
 - Results discussed with family
 - Proceed with hearing aid fitting

Hearing Aid Fitting, Age 9 months

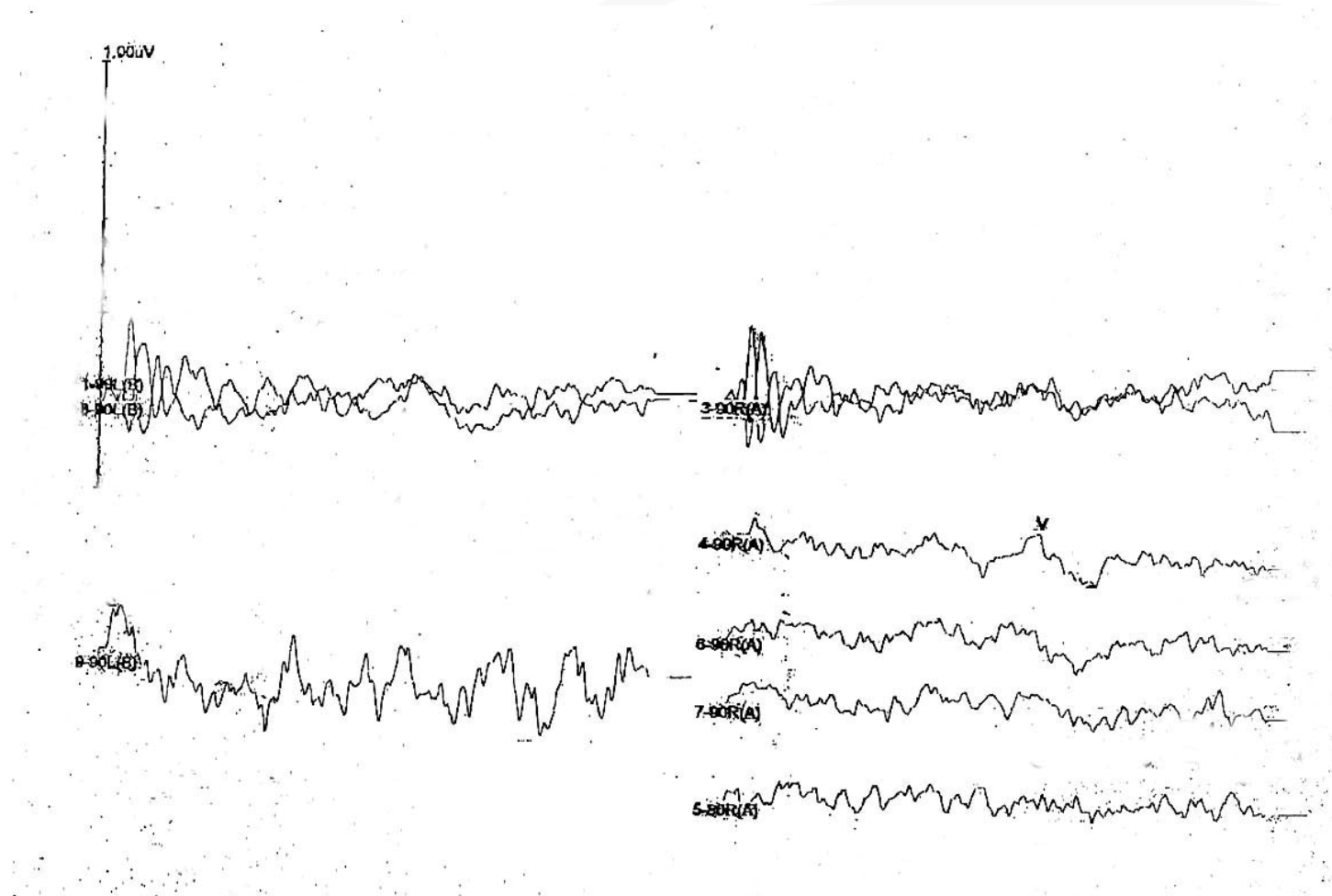
Review of Outside Records (continued)

Behavioral audiometry, 18 months

- Sound Field Test Results:
 - » 500Hz:30dBHL, 2kHz:20dBHL, 4kHz:30dBHL
- Sedated ABR recommended



ABR #5 Age 19 months(Outside Clinic)



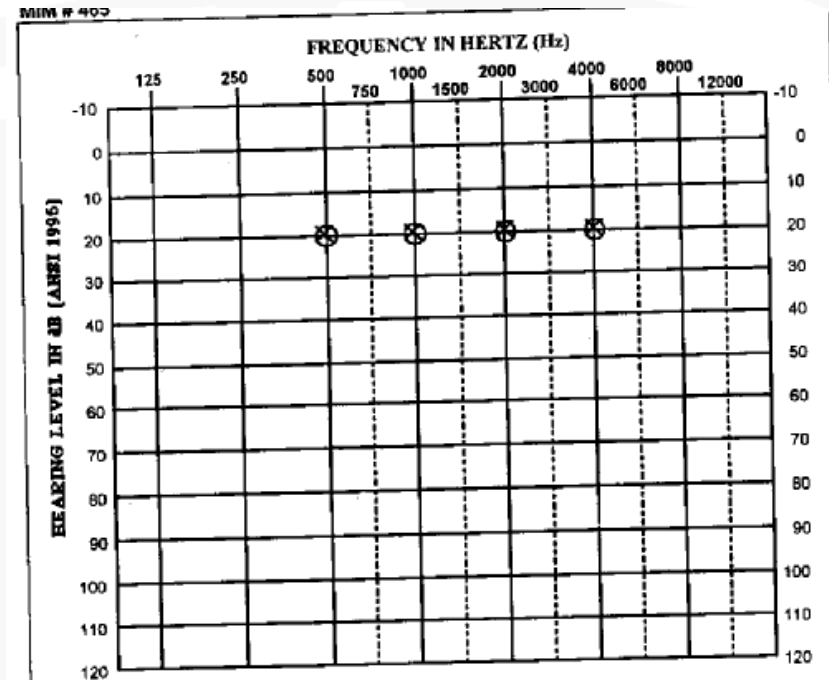
Age 20 months: First UNC Visit

Summary:

- » Behavioral Audiometry:
 - VRA using insert earphones
 - Normal hearing sensitivity bilaterally
- » Tympanometry
 - Type A bilaterally
- » Acoustic Reflexes
 - Absent bilaterally
- » Otoacoustic Emissions
 - Present bilaterally

Recommendations:

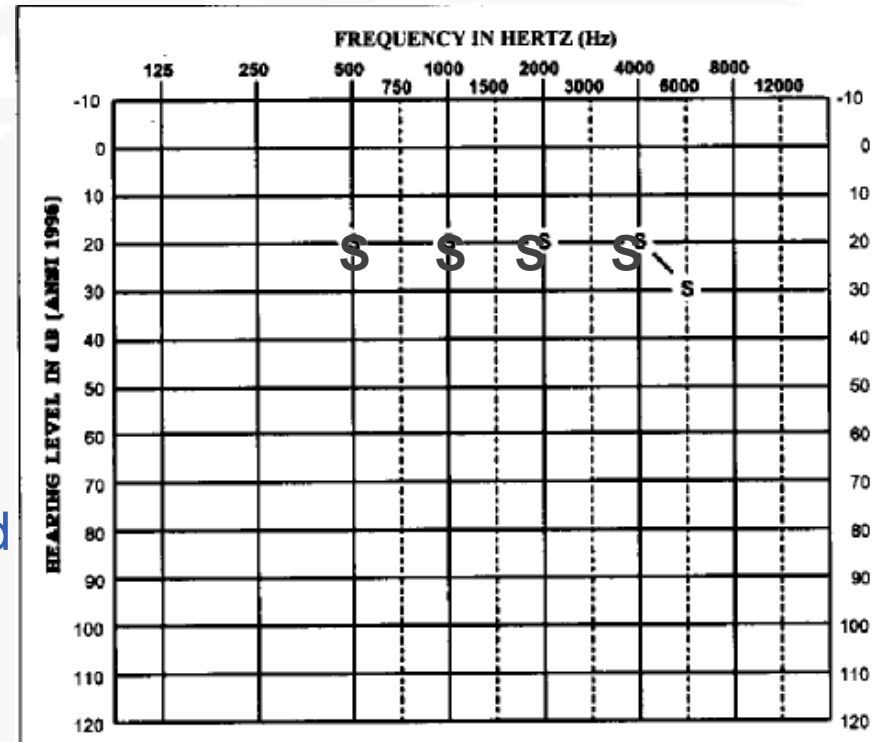
- » Discontinue Hearing Aid Use
- » Enroll in EI to monitor communication status
- » Return in 2 months for otologic exam and to repeat behavioral testing
- » Consider sedated ABR at UNC



Behavioral Audiometry

Age: 26 months

- Child doing well without amplification.
- Mother reports he is comprehending language, speech is still somewhat behind his peers
- Continuing to receive EI services
- Child did not tolerate earphones so testing completed in sound field
- Reliable head turn responses obtained with VRA and responses for at least one ear within normal limits
- Mother met with otologist and after review of 5 previous ABRs, decision made to proceed with UNC ABR to rule out or confirm previous diagnosis



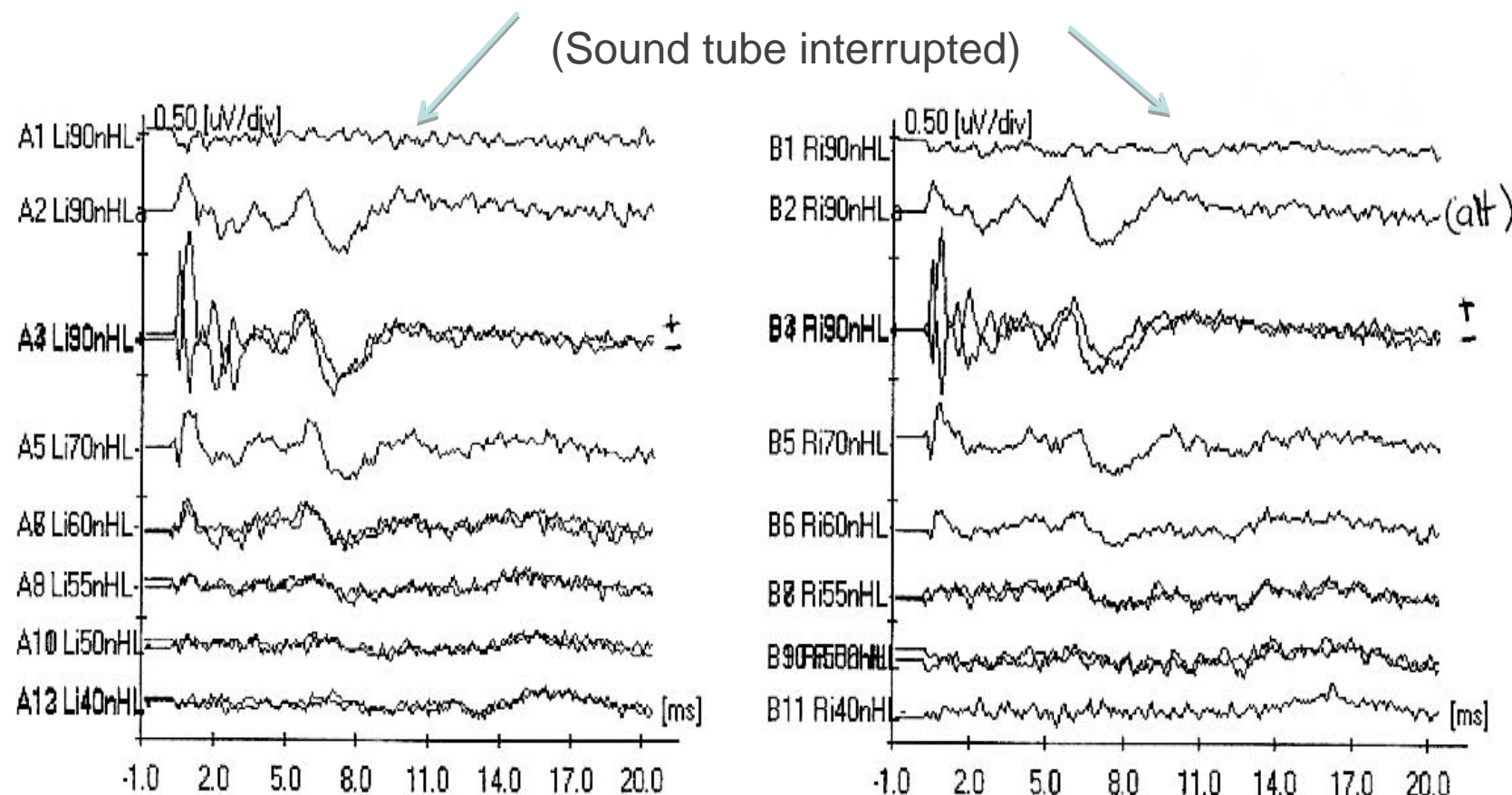
UNC Sedated ABR Results

Age: 2 years, 4 months

Clicks



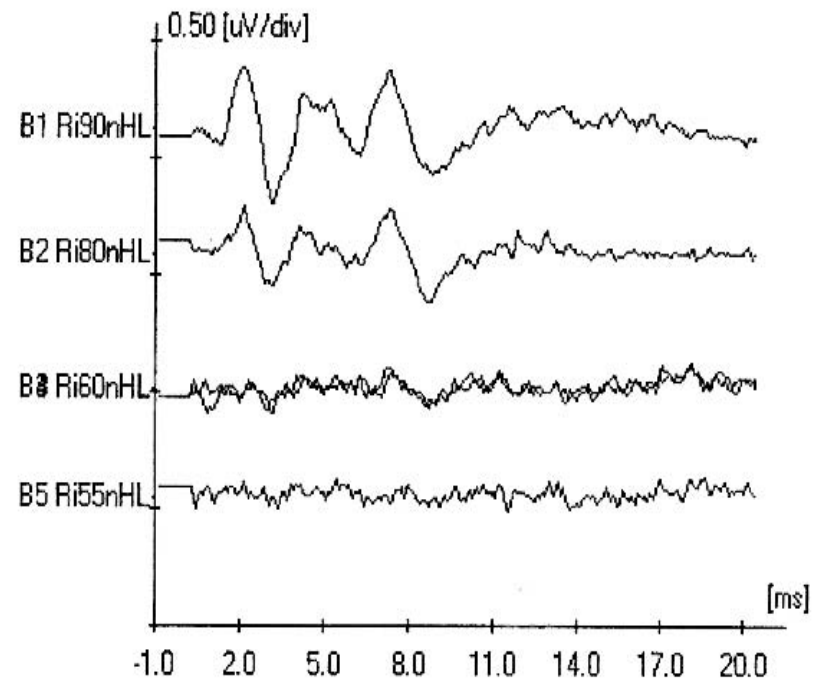
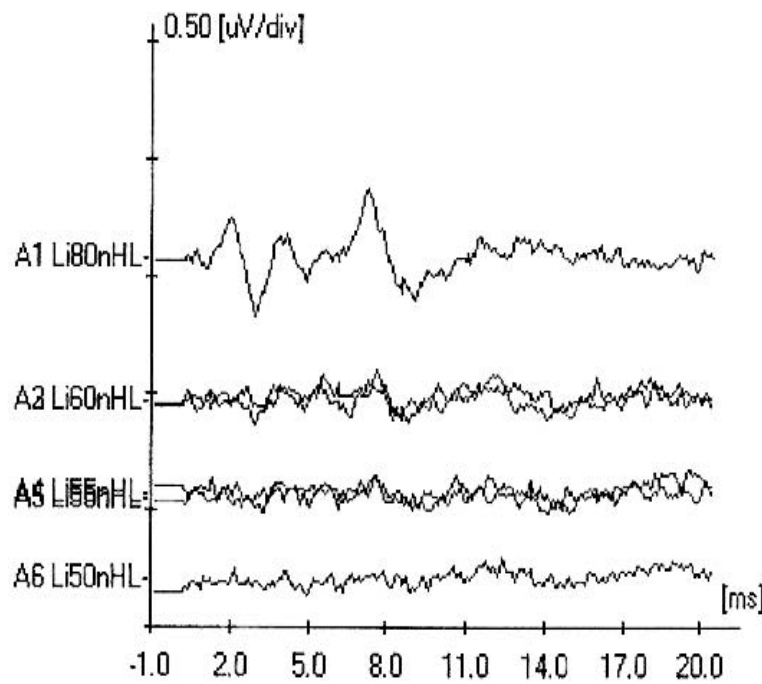
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UNC Sedated ABR Results

Age: 2 years, 4 months
250 Hz Tone Burst



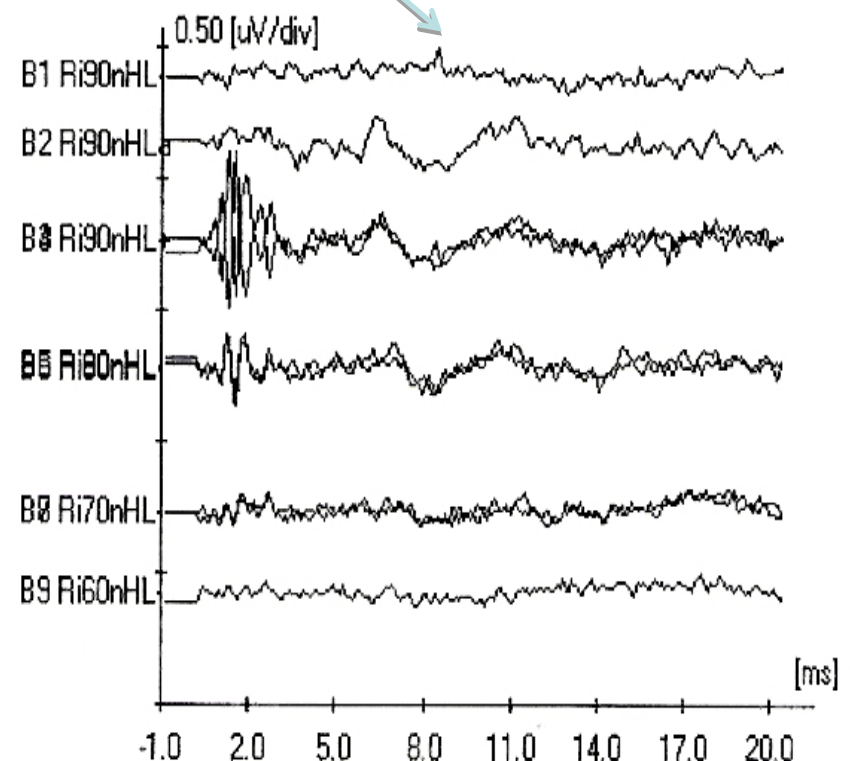
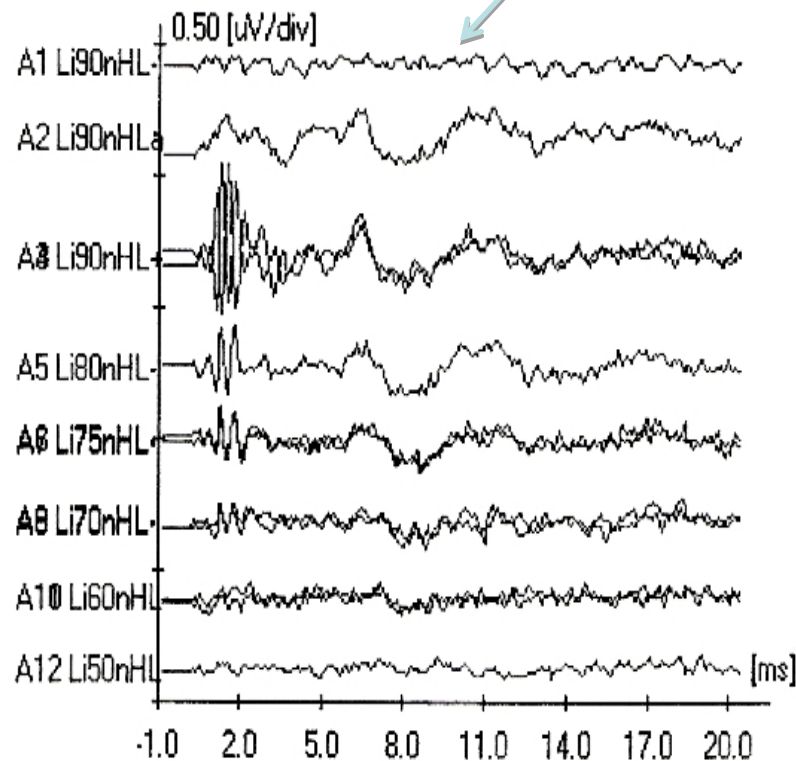
UNC Sedated ABR Results

Age: 2 years, 4 months
2000 Hz Tone Burst



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(Sound tube interrupted)



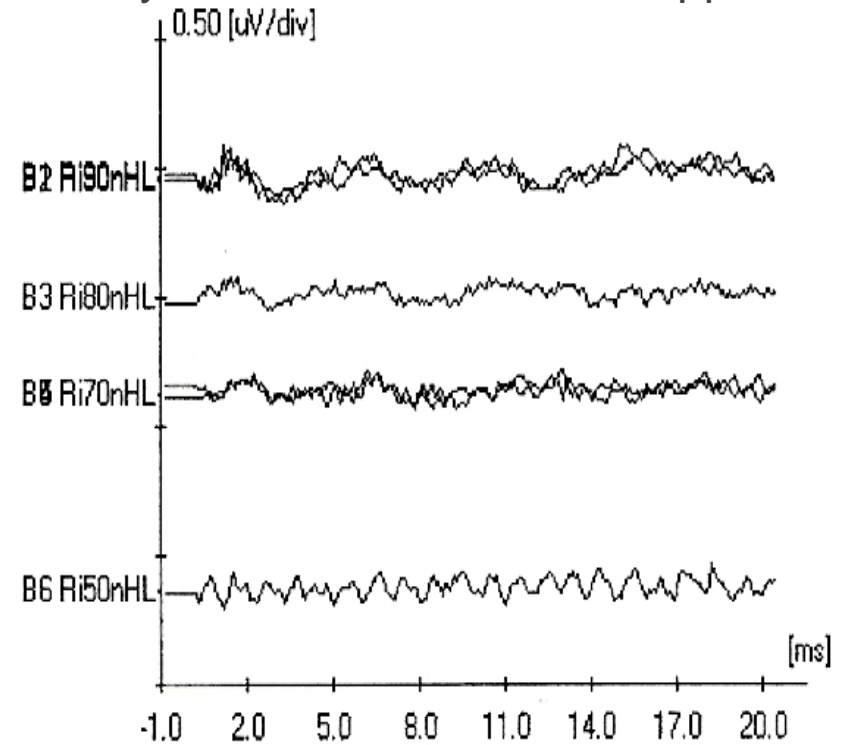
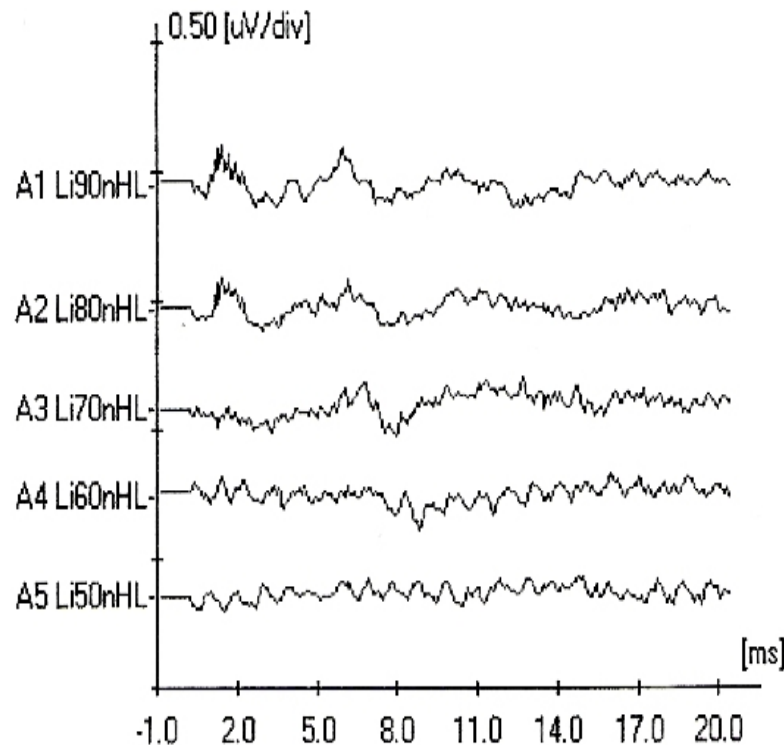
UNC Sedated ABR Results

Age: 2 years, 4 months
4000 Hz Tone Burst



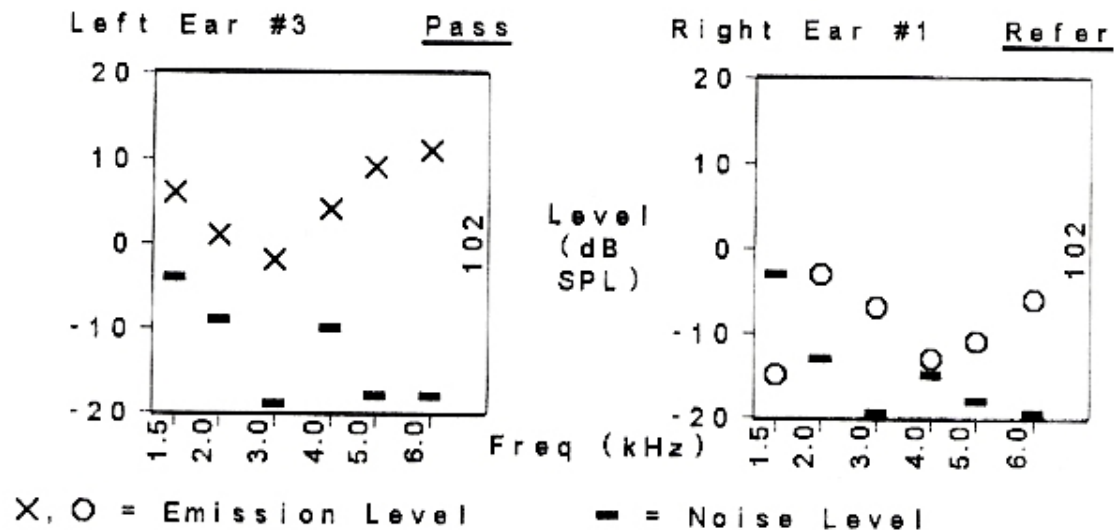
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Note that elevated distal waveforms inconsistent with hearing levels obtained at time of behavioral audiometry even after corrections applied



UNC Otoacoustic Emissions

Age: 2 years, 4 months



1.5	2.0	3.0	4.0	5.0	6.0
6	1	-2	4	9	11
-4	-9	-19	-10	-18	-18
10	10	17	14	27	29

Freq (kHz)	1.5	2.0	3.0	4.0	5.0	6.0
DP	-15	-3	-7	-13	-11	-6
NF	-3	-13	-24	-15	-18	-21
DP-NF	-12	10	17	2	7	15

Screeners: S20001773
Grason-Stadler, Inc. USA

Probe: P19990874
GSI 70 AOAE



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Key Points

- Child now developing speech and comprehending spoken language but...
- In view of abnormal ABR, it will be important to monitor both hearing sensitivity and communication development over time
- Testing with high level (80-90dBnHL) clicks needed to evaluate waveform morphology and rule out auditory neuropathy whenever tone bursts are abnormal
- When high level click ABR is abnormal showing ANSD pattern, ABR can no longer be used to estimate behavioral thresholds and...
- Even when distal waveforms are present, don't assume it's possible to correctly estimate behavioral thresholds
- Critical to use established VRA protocols with insert earphones to obtain behavioral threshold estimates whenever possible
- Multiple ABRs at close intervals probably not indicated



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CASE #3



Case Study: Unilateral

- Newborn Screen with AABR:
 - » Referred on left
 - » Passed on right
- Age 2 months:
 - » Diagnostic ABR: moderate HL left, normal right
- Age 2 ½ months:
 - » Otologic evaluation: MRI, EKG, connexin 26 and CMV testing ordered



Unilateral:Continued

- Age 3 months:
 - » Referred to Beginnings for information and referral to early intervention
- Age 4 months:
 - » MRI: Bilateral enlarged vestibular aqueducts and enlargement of endolymphatic sacs
 - » Otologist advises of risk for progressive hearing loss and avoiding head trauma and refers to neurology and genetics for evaluation

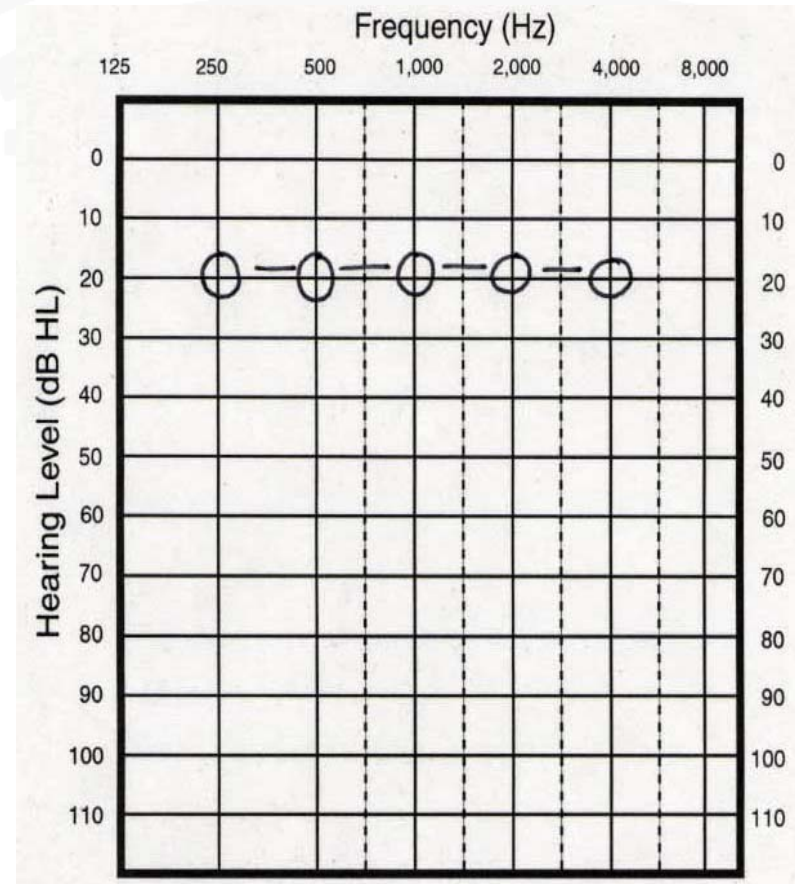


Unilateral: Continued

- 6 months: Genetics consult completed
 - » Most common cause of EVA is alteration of Pendred gene
 - » Several other syndromes can be associated with EVA including branchio-oto-renal syndrome
 - » Will test for Pendred's and if negative will order renal ultrasound
 - » Lab results shows child is connexin 26 negative but has two copies of gene for Pendred's
 - » Recommendation made for pediatrician to periodically monitor thyroid levels

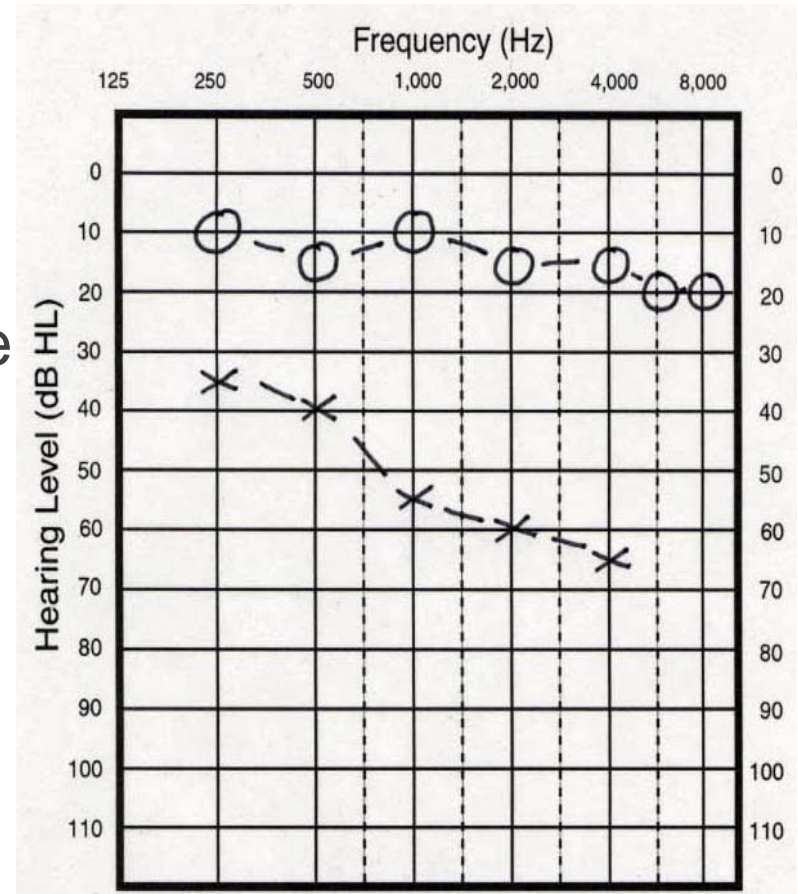
Age: 8 months

- Tympanometry
 - » Right: normal
 - » Left: -275
- Otoacoustic Emissions
 - » Right: Absent above 2000Hz
 - » Left: Absent



Age: 12 months

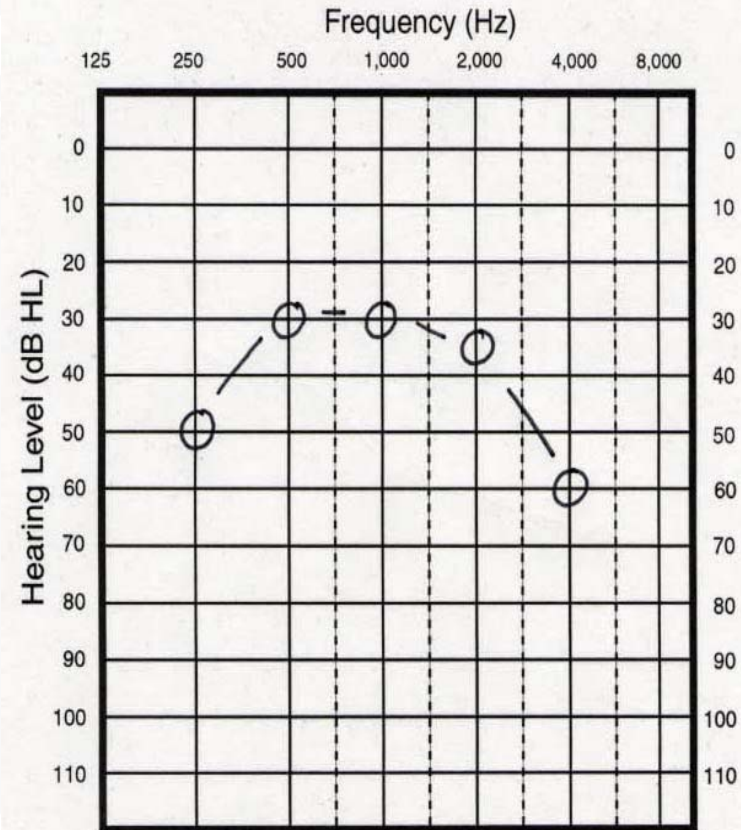
- Right ear:
 - » Normal
- Left ear:
 - » Mild to moderate
- Tympanometry
 - » Right: normal
 - » Left: normal





Age: 17 months

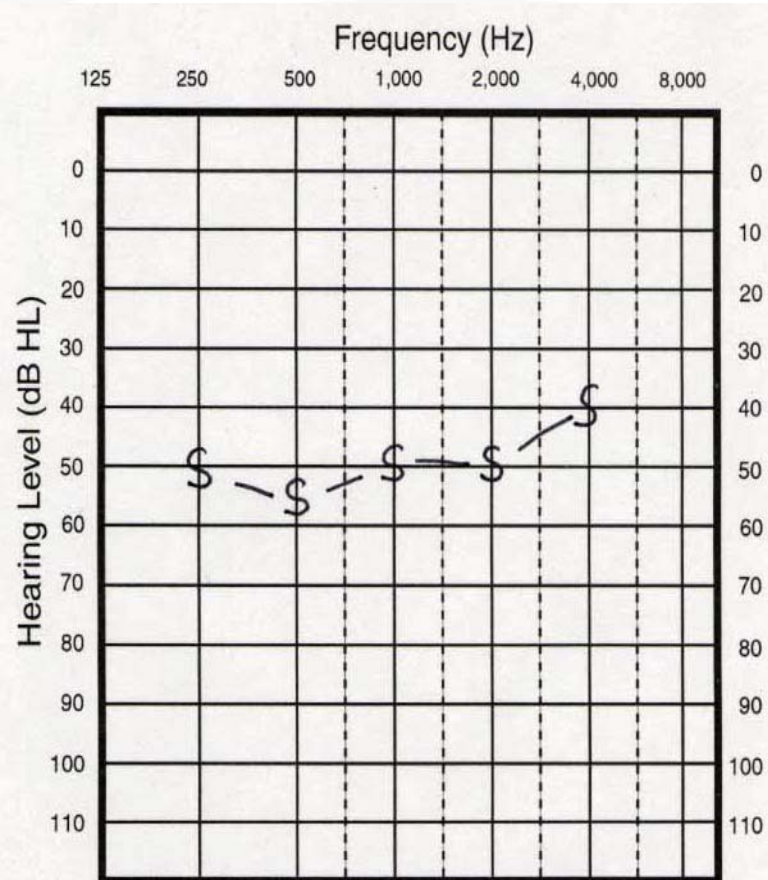
- Difficult to test but right ear responses poorer than expected
- Tympanometry
 - » Right: normal
 - » Left: normal
- Family advised of our concern re progression of HL





Age: 18 months

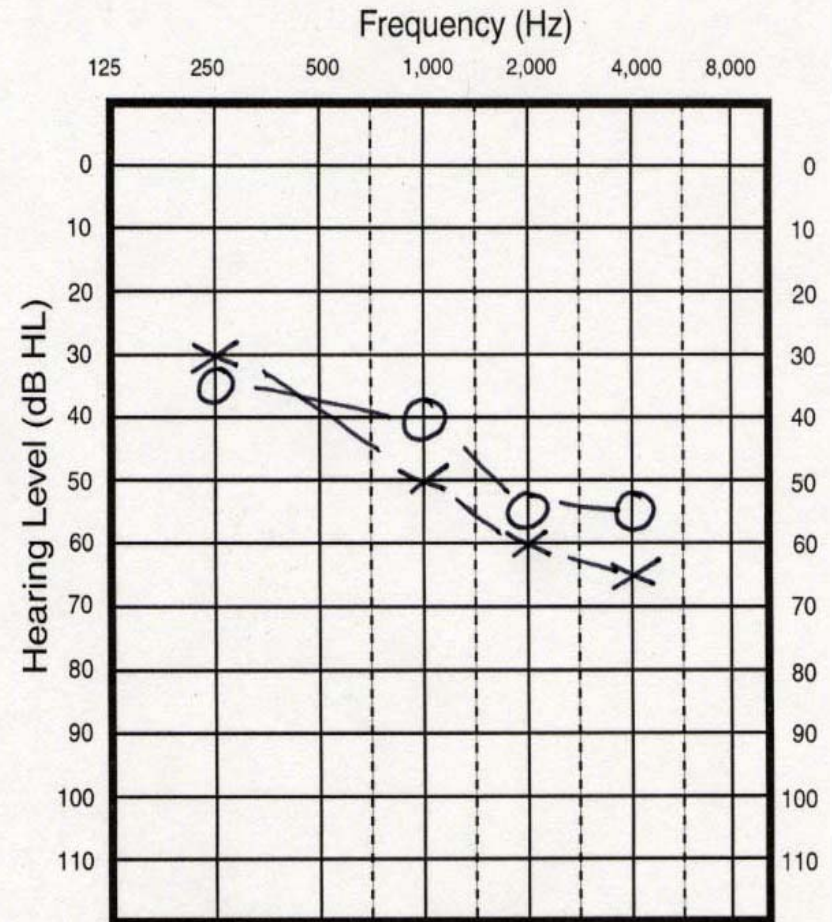
- Child will not tolerate insert earphones
- Unable to rule out hearing loss for “better ear”
- Tympanometry
 - » Right: -225
 - » Left: -190
- Sedated ABR recommended





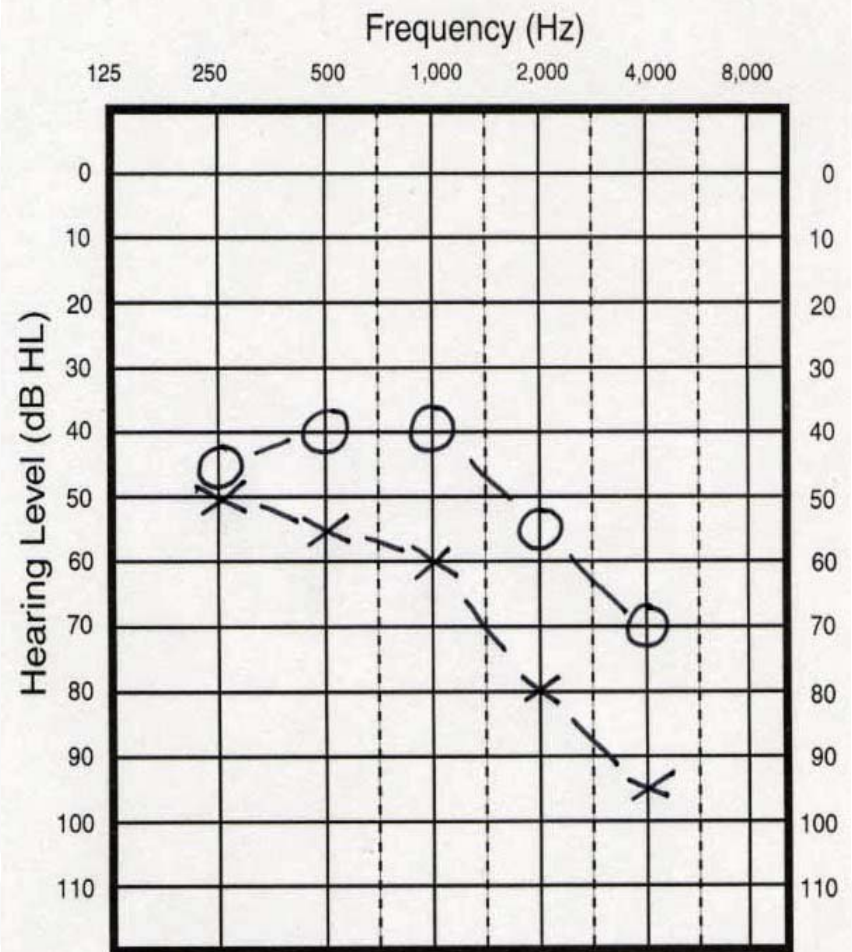
Age 20 months: Estimated Thresholds (eHL) Based on Sedated Tone Burst ABR

- Binaural hearing aids and personal FM dispensed 2 weeks later



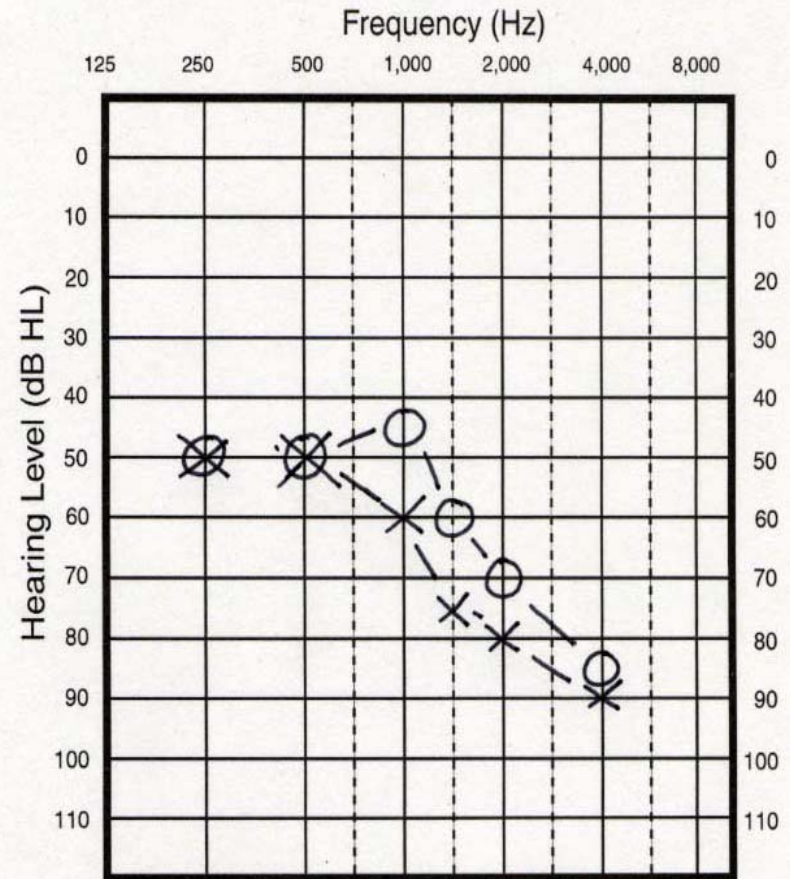
Age: 22 months

- Continued progression of hearing loss noted
- Tympanometry
 - » Right: normal
 - » Left: normal



Age: 23 months

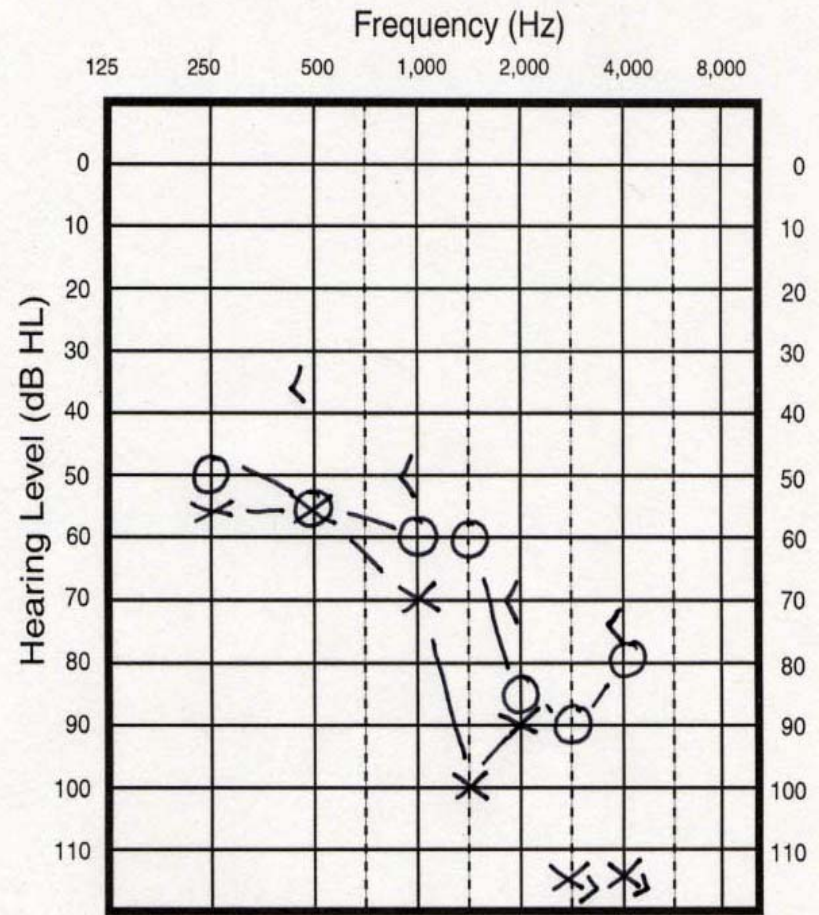
- Play audiometry
- Hearing aids exchanged for model with more power
- Hearing aids programmed for best match to DSL targets





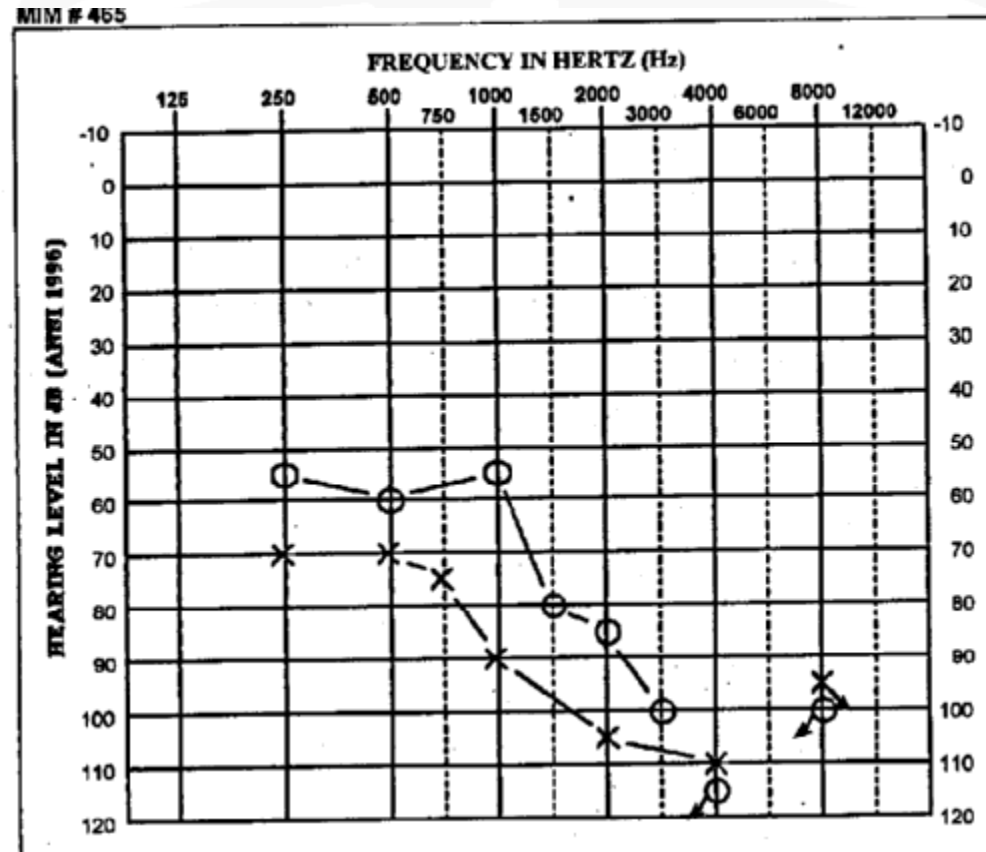
Age: 24 months

- Hearing aids readjusted to better match DSL targets
- Recently fitted with new hearing aids with frequency compression
- Speech and language evaluation scheduled with SLP from CI team to obtain baseline and review current services
- Child will be monitored regularly and referred for CI evaluation if indicated



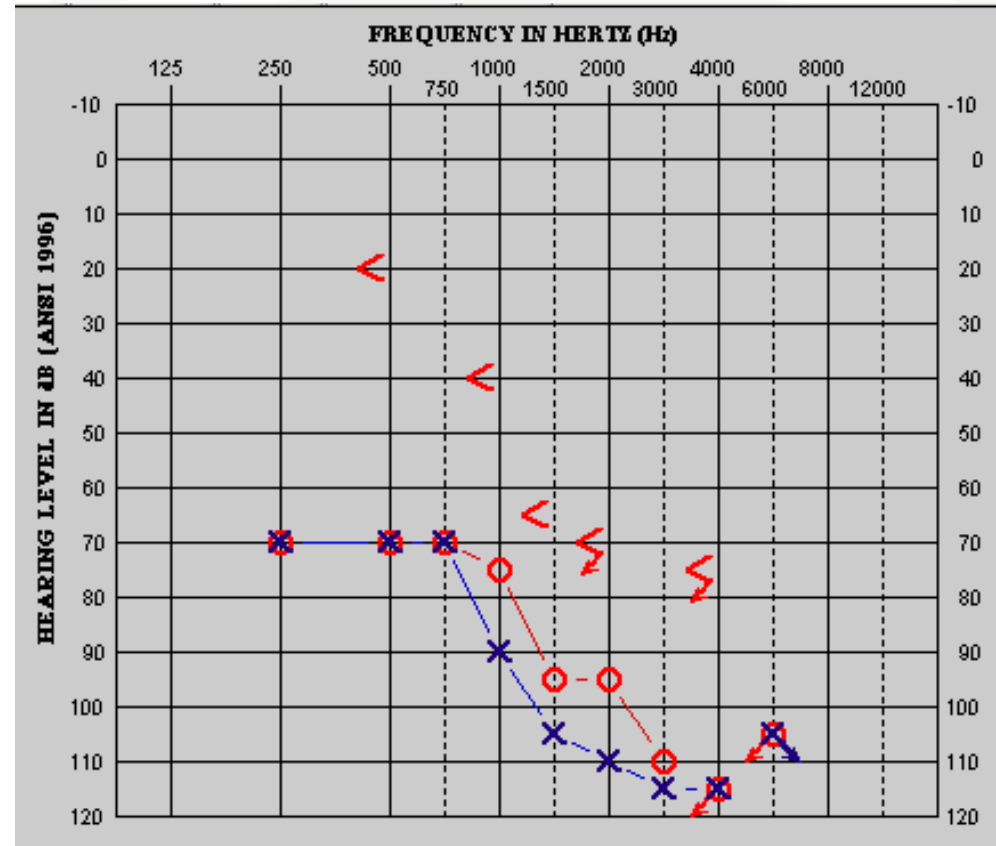
Age: 4 years

- Aided Testing
 - » SRT=25dBHL
- Aided PBK score:
 - » 80% at 55dBHL



Age: 4 years, 11 months

- Limited HA benefit even with FC device
- Aided speech recognition:
» 36% at 55dBHL (PBKs)
- Struggling in pre-school
- After extensive discussion with family, referred to CI team for evaluation





Key Points

- Comprehensive team evaluation useful when working with infant with newly diagnosed HL
 - » Audiology, ENT, Genetics, Early Intervention Specialists, Pediatrics all played role
- ABR used to determine initial thresholds for first hearing aid fitting and to help when results are ambiguous but...
- Behavioral audiometry with VRA to obtain accurate unaided thresholds most useful tool after six months of age in this case
- Evaluation of unaided hearing thresholds combined with use of hearing aid verification measures allowed child to continue to make progress even with progressive changes to hearing



CASE #4



Background

- Full term baby born without complications
- Newborn Screen with AABR and OAE:
 - » Passed right
 - » Failed left
- Diagnostic ABR at 2 months:
 - » Normal right; Moderate SNHL left
 - » DPOAEs present right; absent left
- Repeat Diagnostic ABR at 6 months:
 - » Normal right; Moderate SNHL left
 - » DPOAEs present right; absent left



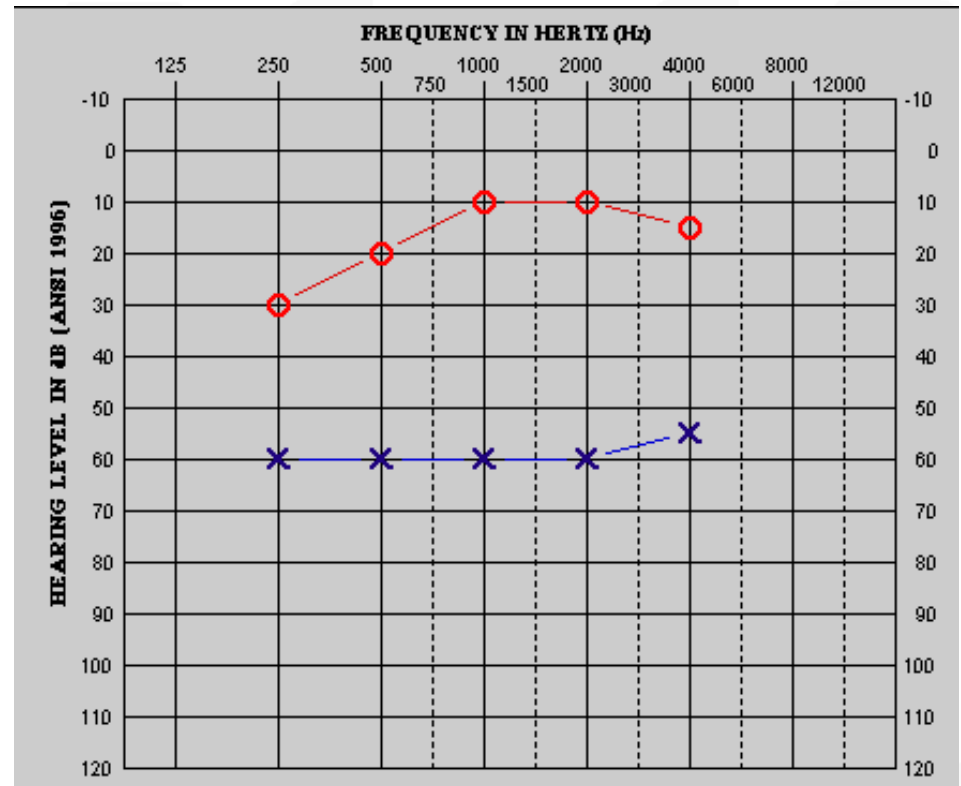
Medical Evaluations

- ENT exam
 - » MRI ordered; normal inner ear morphology
 - » EKG normal
- Genetics evaluation
 - » Connexin negative

Behavioral Audiometry

Age: 12 months

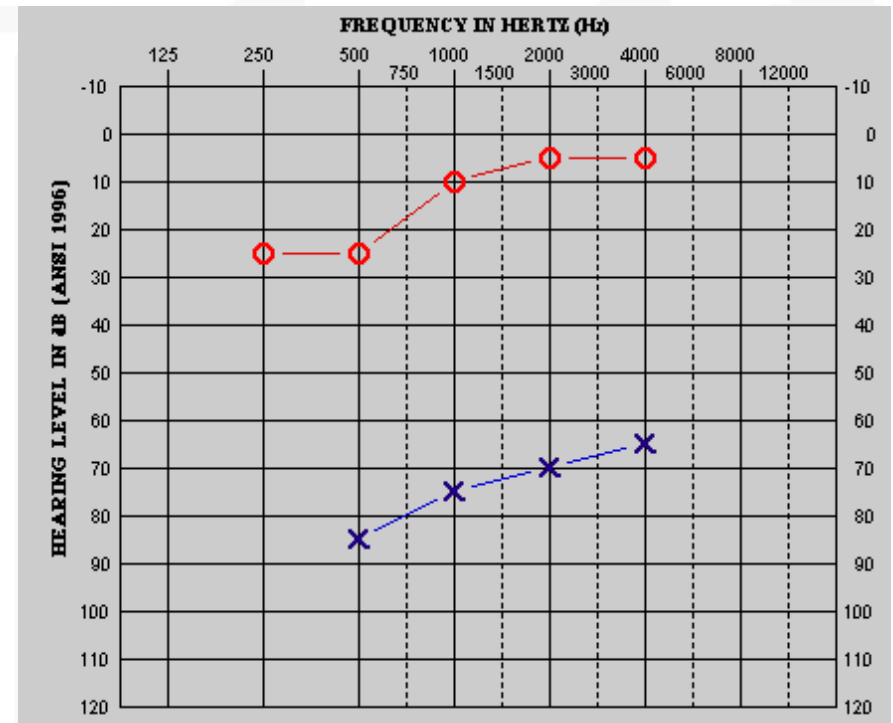
- SAT
 - » Right: 20dBHL
 - » Left: 55dBHL
- Tympanometry
 - » Large physical volumes consistent with patent tubes, bilaterally



Behavioral Audiometry

Age: 26 months

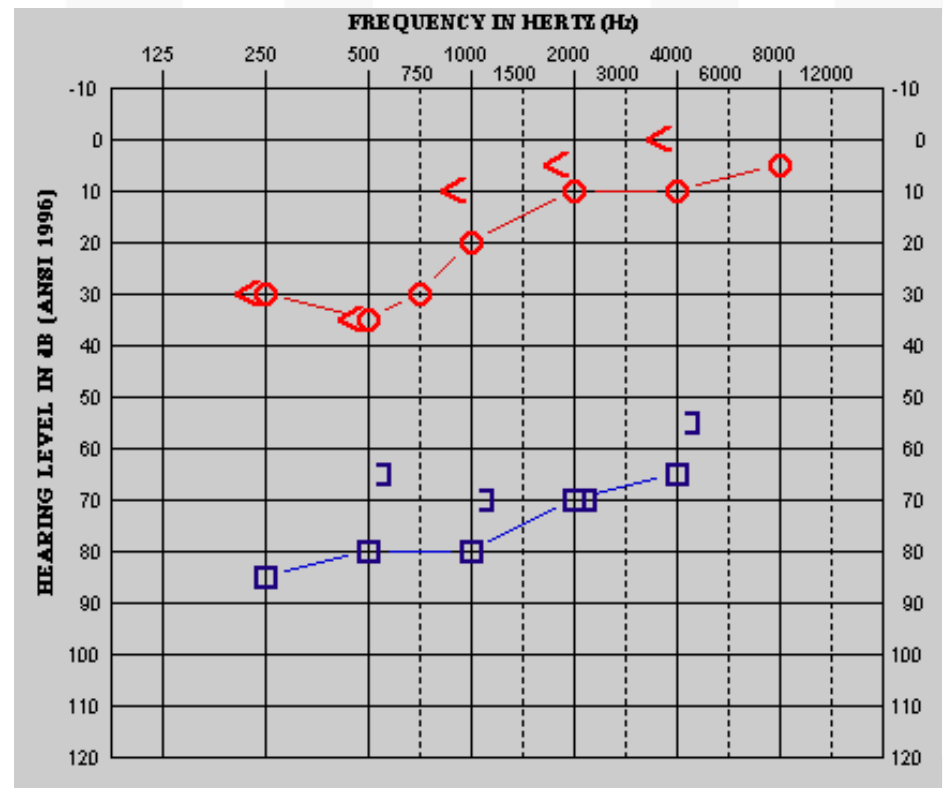
- SRT (Identification of body parts)
 - » Right: 10dBHL
 - » Left: NR at 110dBHL with masking in right
- Tympanometry
 - » Large physical volumes consistent with patent tubes, bilaterally



Behavioral Audiometry

Age: 3 years, 10 months

- **SRT:**
 - » Right: 25dBHL
 - » Left: NR at 110dBHL with masking in right
- **PBK:**
 - » Right: 96% at 65dBHL
 - » Left: 0% at 110dBHL
- **Tympanometry**
 - » Type A
 - (Tubes out)

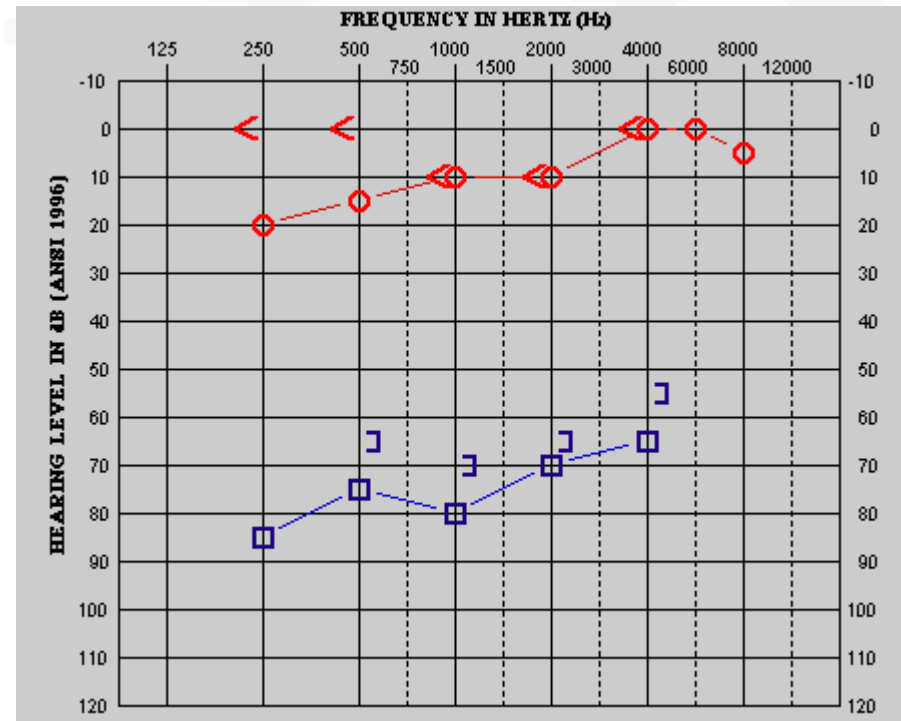




Behavioral Audiometry

Age: 4 years

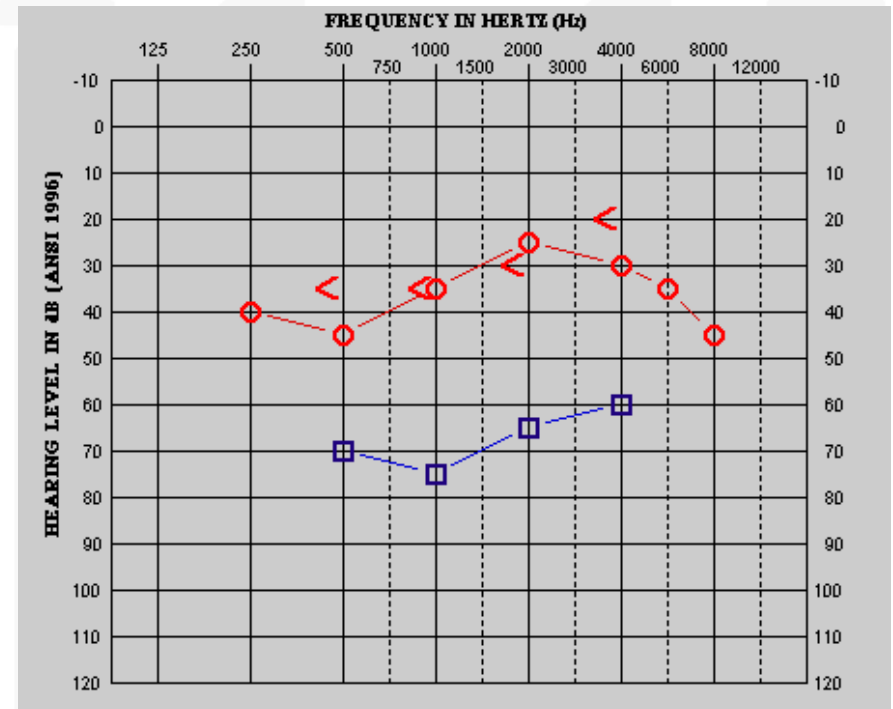
- **SRT:**
 - » Right: 10dBHL
 - » Left: NR at 105dBHL
- **PBK:**
 - » Right: 100% at 50dBHL
 - » Left: CNT



Behavioral Audiometry

Age: 4 1/2

- Hearing aid use discussed with family
- Decision made to proceed with HA for right ear



Parents Express Concern...

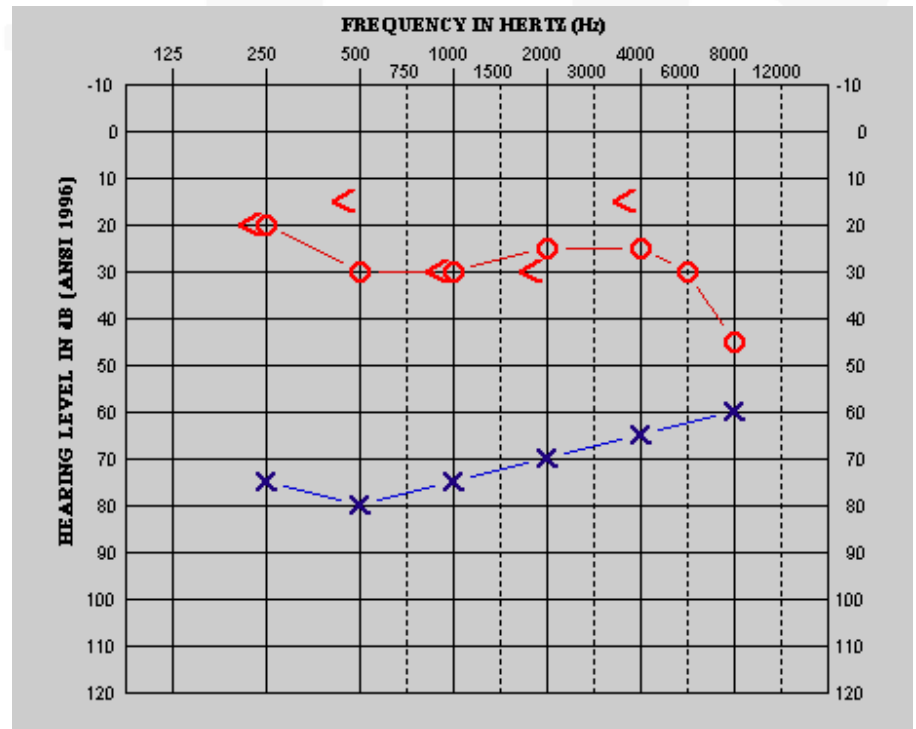
- Child not showing any other neurologic changes but...
- Hearing is fluctuating and progressing
- MRI was normal
- Child returned again to ENT
 - » PCR test for congenital CMV obtained from blood spot taken at time of newborn metabolic screening
 - » Results: **Positive for congenital CMV**



Behavioral Audiometry

Age:6 years

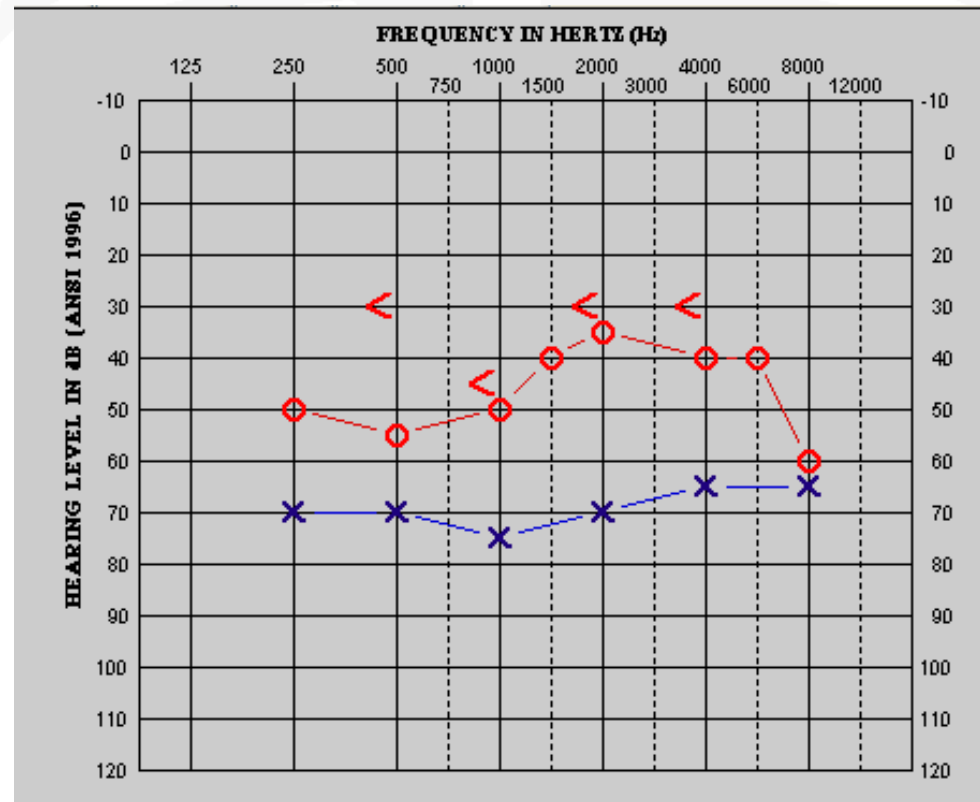
- Continued use of HA in right ear
- FM dispensed for personal and school use



Behavioral Audiometry

Age: 6 ½ years

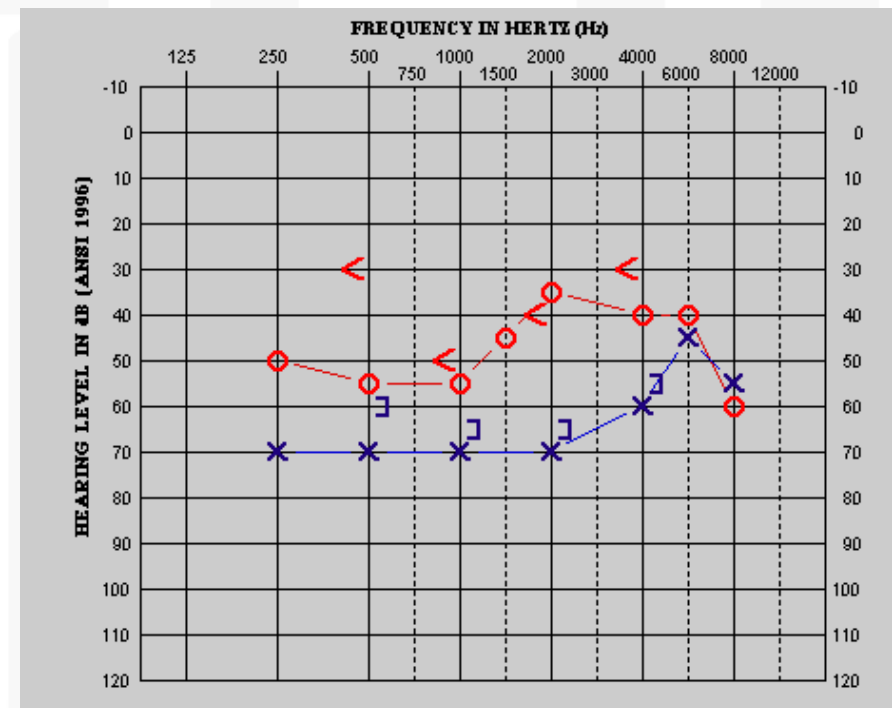
- **SRT:**
 - » Right: 40dBHL
 - » Left: NR at 110dBHL with masking in right
- **PBK:**
 - » Right: 96% at 80dBHL
 - » Left: CNT
- **Tympanometry**
 - » Type A



Behavioral Audiometry

Age: 7 years

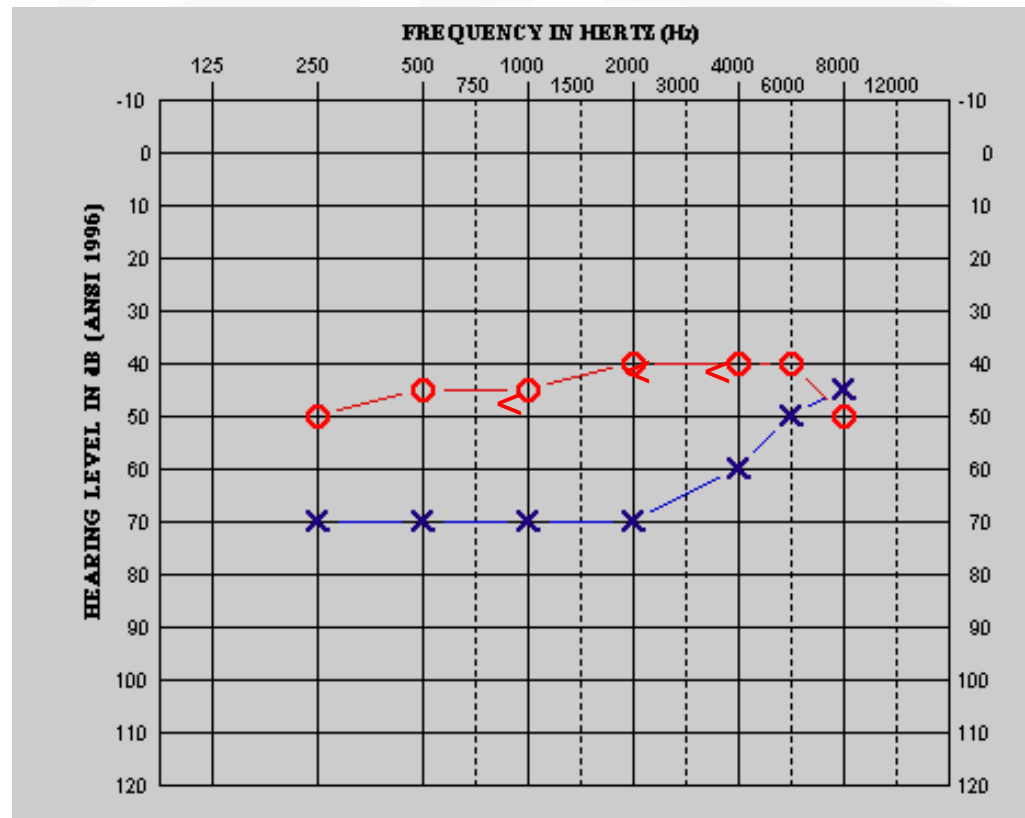
- SRT:
 - » Right: 40dBHL
 - » Left: NR at 110dBHL with masking in right
- PBK:
 - » Right: 96% at 80dBHL
 - » Left: 0% at 100dBHL
- Tympanometry
 - » Type A



Behavioral Audiometry

Age: 7 ½ years

- SRT:
 - » Right: 40dBHL
 - » Left: NR at 110dBHL with masking in right
- PBK:
 - » Right: 100% at 75dBHL
 - » Left: CNT
- Tympanometry
 - » Type A





CMV and Hearing Loss

- Congenital CMV infection is a leading cause of SNHL in children.
- Both asymptomatic and symptomatic children may experience:
 - » Delayed onset of HL
 - » Fluctuating HL
 - » Progressive HL
- Necessitates continued monitoring of hearing status

J Dahle AJ, Fowler KB, Wright, JD Boppana SB, Britt WJ, Pass RF.
J Am Acad Audiol. 2000 May;11(5):283-90

Conclusions

- Two cases of auditory neuropathy spectrum disorder (ANSD)
 - » One with profound bilateral HL who was diagnosed with normal hearing (a false negative)
 - » One with normal hearing sensitivity who was fitted with hearing aids and referred for consideration of CI (a false positive in regards to degree of hearing loss)
- Two case of progressive hearing loss
 - » One with enlarged vestibular aqueduct syndrome
 - » One with asymptomatic congenital CMV infection
- These cases remind us of the need to “expect the unexpected” and to be diligent in following protocols and using evidence-based practice



Conclusions

- Accurate diagnosis is an essential component that provides the foundation for treatment
- Although we are making significant progress, some of the diagnostic errors we made a decade ago still occur
- Management of hearing loss in infancy and early childhood is complex and requires specialized protocols and procedures
- Centers of expertise are needed, especially for young infants and those with complex needs



Thank You!

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