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Estudio de casos de adaptación de audífonos
en niños de 3 a 18 meses

*Case studies of hearing instrument fitting
in infants from 3 to 18 months of age*

Case Studies

Hearing Aid Fitting



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Early Identification, Diagnosis and Treatment of
Deafness in Infants

Madrid

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HEARING CARE FOR INFANTS: WHAT ARE THE ESSENTIAL COMPONENTS?



Screening



Positive outcomes but challenges remain:

- Excessive re-screening
- Delays in referral for diagnostic ABR
- Families falsely reassured at time of screening

Assessment: Electrophysiologic Measures

- ABR
 - Tone burst stimuli used to estimate thresholds for low, mid and high frequencies
 - When ABR shows no response, must use single polarity clicks to rule out auditory neuropathy
 - Air conduction and bone conduction
- Otoacoustic Emissions
- Acoustic immittance measures (1000Hz probe tone for babies <4 months)



Otologic Examination and Follow up

- Medical history
- Otoscopic exam
- Radiological studies
 - Primarily MRI, CT as needed
- EKG
- Laboratory studies
- Referral to Ophthalmology
- Genetic testing offered to family
 - Connexin 26 lab test offered
 - Other genetic lab tests may be ordered

Communication

- Time allowed on same day as ABR to complete ABR and discuss results with family
 - If this is first time family is hearing the news, may be helpful to schedule a return appointment
 - Some families ready to proceed others need more time
 - Important to determine how best to meet families needs
 - “Family friendly” environment can exist even within a medical center
- Reports sent to:
 - Parents
 - Pediatrician/primary care physician
 - Otolaryngologist
 - Early intervention specialist

Hearing Instrument Selection and Ear Impressions

- If family ready to proceed, ear impressions taken
- Hearing instruments selected
- Return appt for hearing instrument fitting two weeks later
- Ideally between 2-3 months of age



Referral for Early Intervention

- Referral to “Beginnings” on day hearing loss diagnosed (www.ncbegin.org)
- Family contacted within one week of diagnosis and home visit from early childhood specialist scheduled
 - Additional information including written materials and video provided to family
- Weekly in home visits with teacher of the HI scheduled as soon as family decides on initial educational approach

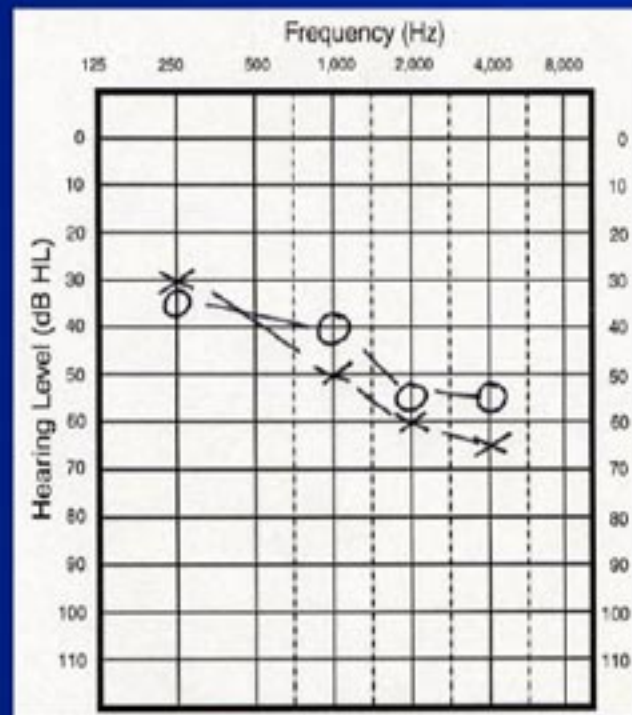
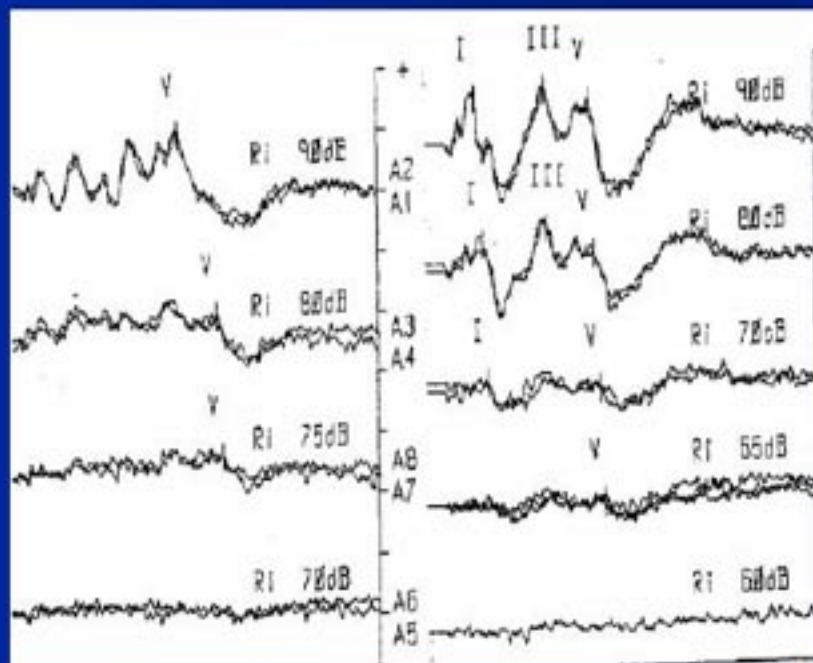


Families

- Important to connect families with other families
- Families offered option of connecting with another family with child similar to their child
- Also provided with information about group meetings (HITCH UP)



Audiogram estimated based on electrophysiologic tests



Infant Hearing Instrument Fitting and Verification UNC Approach

Two hour session consisting of:

1. RECD measurement with child's earmold (may need to use average values if child is too active)
2. Programming of hearing aids
3. Verification measures using DSL prescriptive formula
4. Hearing aid orientation



Hearing Instrument Orientation



- Initial care kit (battery tester, stethoset, air blower, retention strap, dehumidifier)
- Troubleshooting/warranty information
- Importance of volume control and other settings
- Identification of right and left aids
- Need for frequent re-make of earmolds
- Information re: noise issues
- Information re: observation of baby's responsiveness to sound

Hearing Instrument Issues

- **Retention**

- Not problem initially but many report retention problems by 5-6 months. Counseling important
- Retention straps, toupee tape, hats, "Huggie Aids"

- **Acoustic Feedback**

- Frequent re-makes needed in first few months. May need as many as 6 sets of earmolds in first year

- **Moisture**

- Baby's remove and suck on hearing aids
- Baby's perspire, particularly when sleeping
- Use of dehumidifier jars and electronic dehumidifier devices helpful

- **Repairs**

- Need for supply of versatile loaner instruments



Behavioral Audiologic Assessment

- Behavioral testing with VRA beginning at 6-7 months of age
- Goal of behavioral testing: Complete audiogram for each ear (air and bone) by 8-9 months of age.
- Hearing aids readjusted as new threshold information is obtained
- Important to complete behavioral testing early... Babies often more difficult to evaluate in 2nd year than 1st



Early Follow-up



- First follow-up 3-4 weeks following fitting:
 - Questions
 - Wearing time
 - Parent's observations of auditory behavior
 - Status of other medical appointments scheduled
 - Status of early intervention
- Earmold remakes every 4-6 weeks for first few months
 - RECDs measured at time new earmolds are dispensed
 - Monitor middle ear status and repeat OAEs
 - Ask family how they're doing

FM system dispensed at approximately one year of age



FM

Later Follow-up



- Return appointments scheduled
 - Every three months for first 3 years
 - Every 4-6 months after 3 years
- Speech recognition testing completed as soon as child is able
- Regular communication with early intervention specialists

Hearing Aid Selection Considerations

- Small
- BTE
- Electroacoustic flexibility
- Tamper resistant battery door
- Ability to protect volume control and on/off switch
- Feedback management

Hearing Instrument Programming Considerations

Initial fitting for young infants:

- One program using omnidirectional microphone
- “Automatic” switching programs for omni and directional microphones *not used*
- Volume control active but protected with ability for parent to adjust if needed

Case Study #1

“Typical” Hearing Instrument Fitting
for Infant Following Newborn
Hearing Screen

Background



- Full-term baby
- No problems during pregnancy or delivery
- No family history of hearing loss
- Second child

History

- AABR in outside well-baby nursery
 - Referred at 35dB, bilaterally
- AABR re-screen at 6 days of age
 - Referred at 35dB, bilaterally
- First diagnostic ABR at outside facility:
 - Clicks: 55 dB-R&L
- Frequency specific ABR at UNCH at age 6 weeks
 - Threshold estimates based on ABR:
 - 25-30 dB for low frequencies
 - 40-50 dB for high frequencies

Age 6 weeks: Ear Impressions

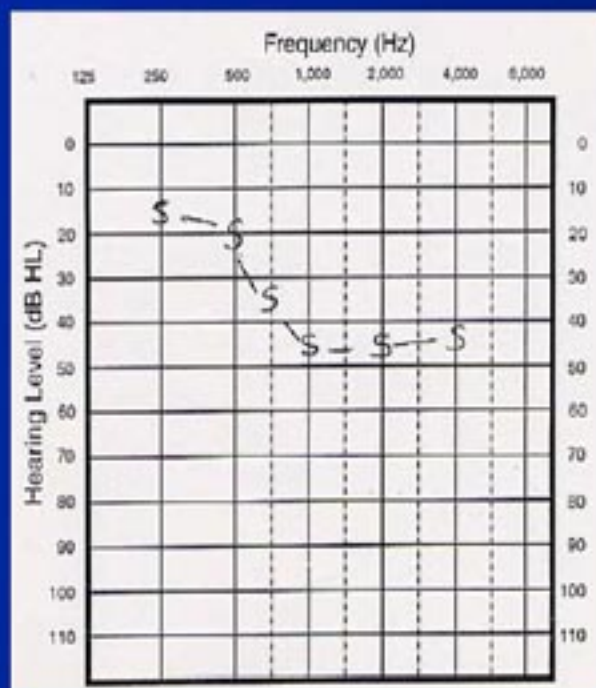


- Results discussed with family and decision made to proceed with amplification
- Ear impressions taken
- Return appt. scheduled
- Referral made to Beginnings and Early Intervention

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

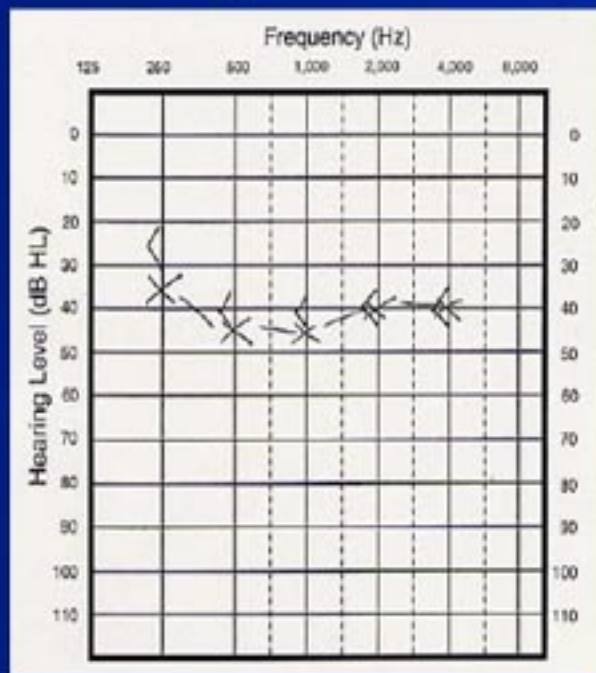
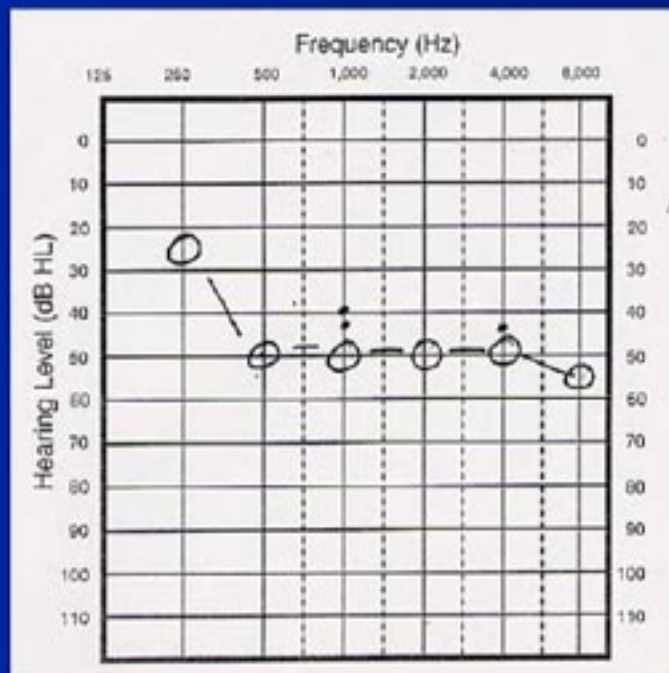
Age 5 mos.

VRA



Age 7 months

VRA with insert earphones and bone conduction testing

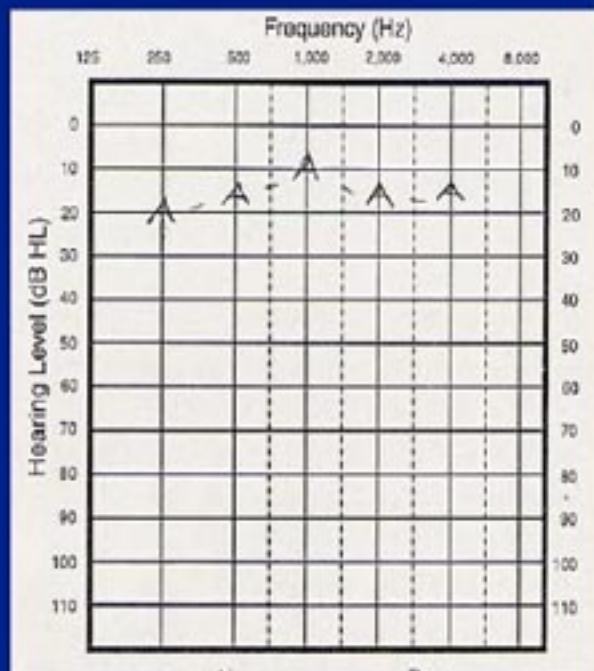


Age 7 months



- Once reliable audiogram obtained for each ear child returns every 3 months for evaluation and hearing aid checks
- RECDs re-measured when new earmolds dispensed
- Child receiving early intervention from teacher of the hearing impaired

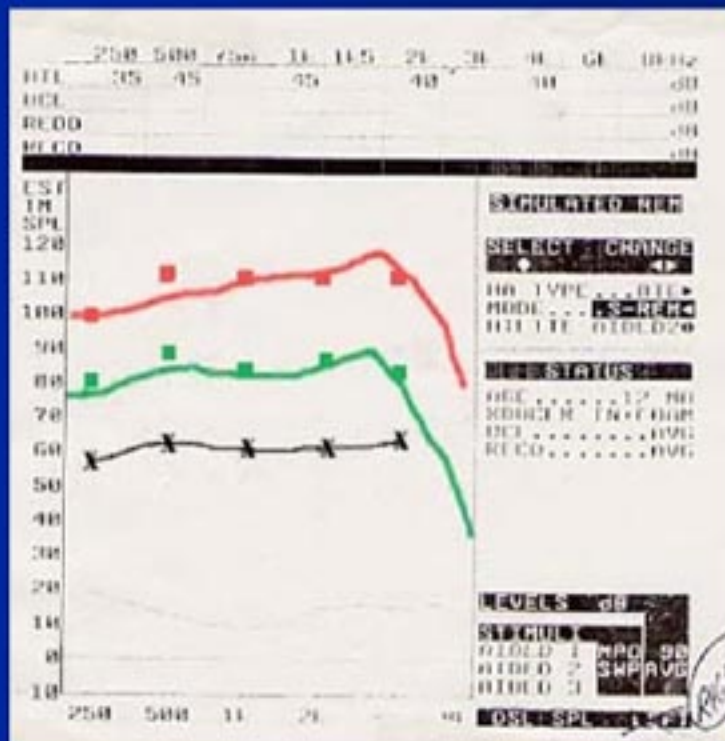
Age 8 ½ mos. Aided Audiogram



Age 12 months

SPL-O-Gram

- FM system dispensed



Developmental Assessment at Age 1 year, 2 months

- Calls family members by name
- Combines talking & pointing to make wishes known
- Uses several words others understand
- Difficulty maintaining EI services



	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

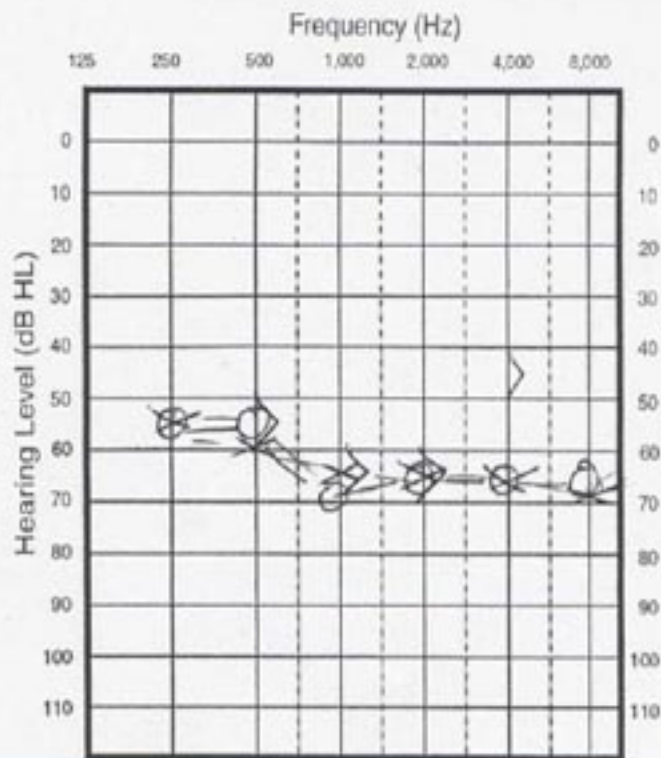
Chronological Age: 4 years 6 months



- Articulation Test:
 - 4 years 9 months
- Vocabulary:
 - 4 -11 to 5-11
 - Receptive vocabulary stopped at the 6-7 year level
- Kindergarten Readiness:
 - 98th percentile

Hearing Evaluation

5 yrs. 5 mos.



- SRTs:
 - Unaided
 - Right: 55dBHL
 - Left: 55dBHL
- Speech Recognition
 - Right: 92%
 - Left: 92%

(Recorded PBK words at 95dBHL)

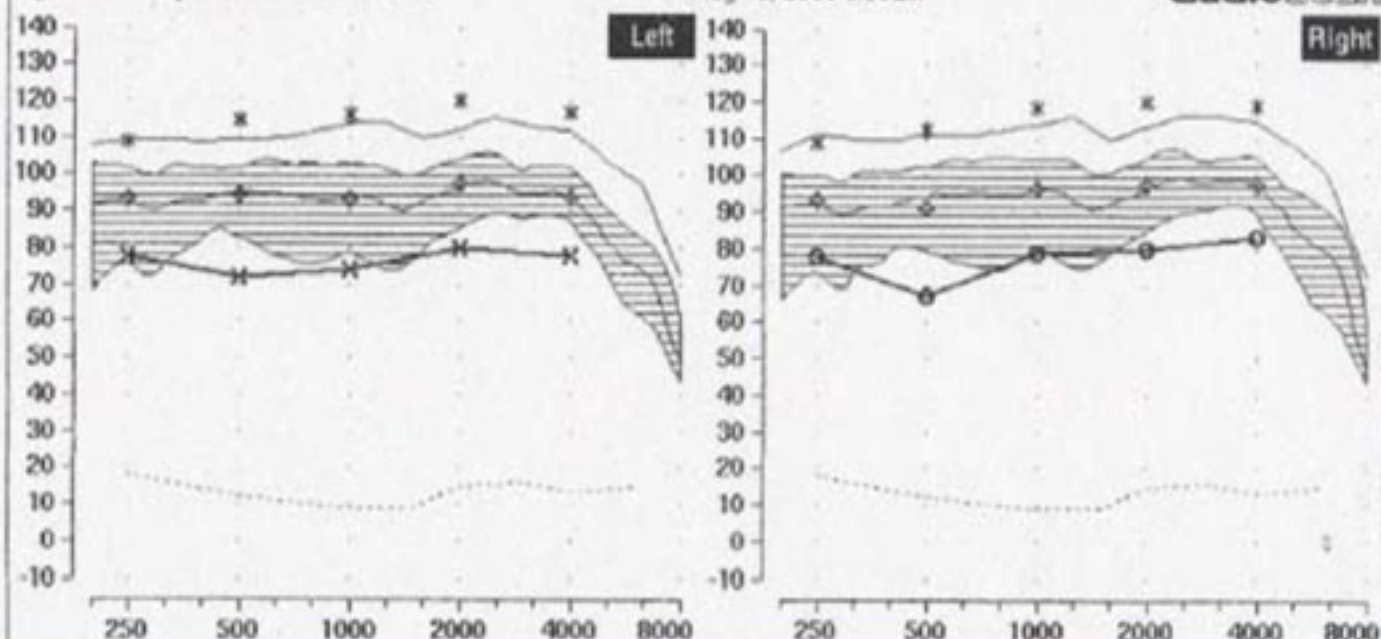
Verification Measures

5 yrs. 5 mos.

Speechmap/DSL - Dual view

Aug 16, 2006 9:06am

audioScan



Current Status



- Five years old
- Speech and language above average
- Doing well in kindergarten
- Enjoying Tai Kwon Do

Case Study #2
Eulalio

Background

- Full-term baby
- Uncomplicated pregnancy and delivery
- No family history of hearing loss
- First child in Spanish speaking home
- Family travels long distance

Screening:

- Screening ABR in hospital nursery:
 - Refer R&L 35dB, bilaterally
- Re-screen ABR post-discharge:
 - Refer R&L 35dB, bilaterally
- Referral made for diagnostic ABR and ENT exam

Diagnostic ABR at Outside Facility

- Diagnosis: "severe to profound bilateral hearing loss"
- Child fitted with binaural, high gain hearing aids at outside facility on basis of click ABR
- Child referred for early intervention services

- **Age 11 months:**

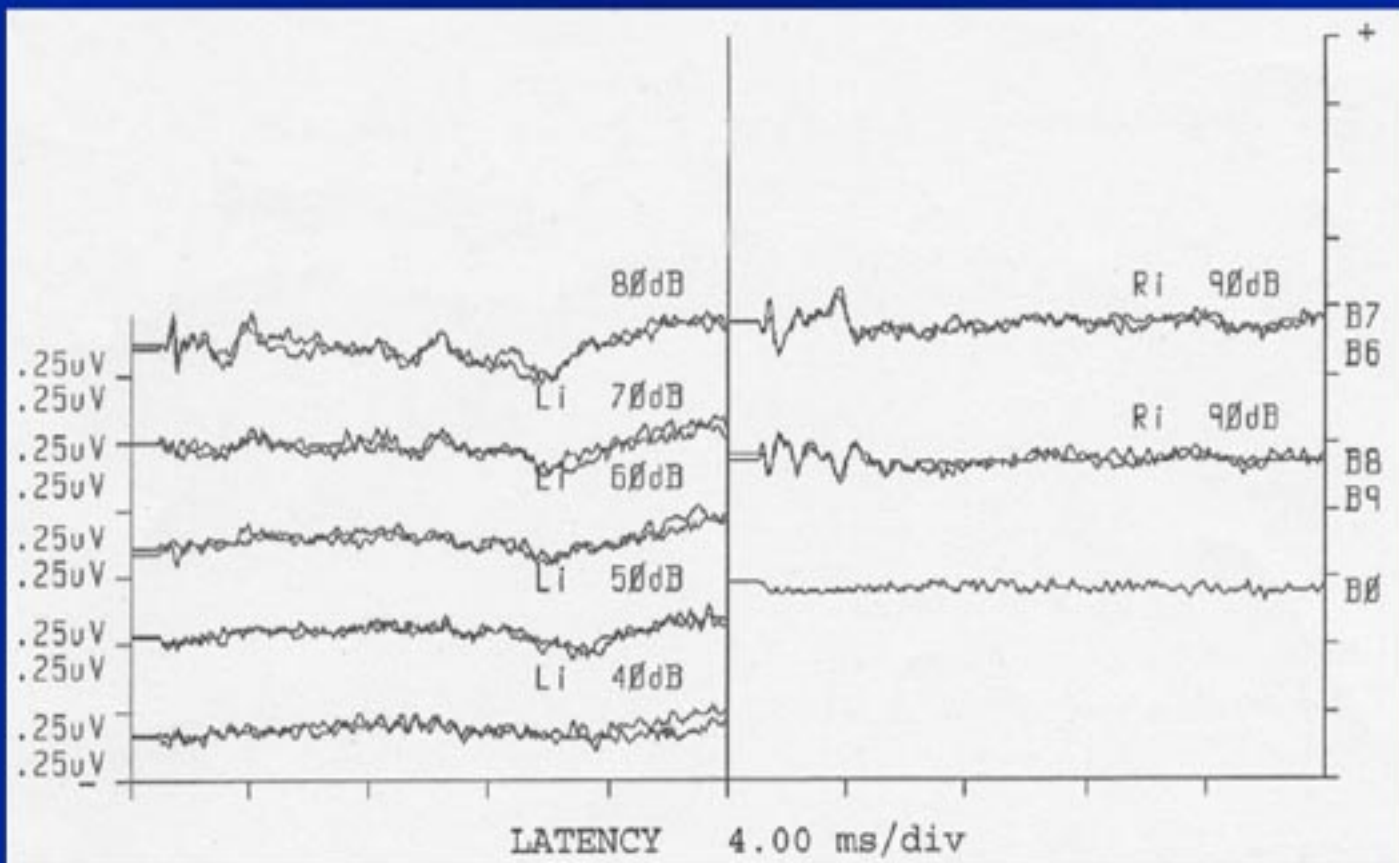
- Referred to UNC pediatric CI team for evaluation
- CI team refers to UNC pediatric audiology team
 - Limited behavioral testing obtained
 - SATs 35dBHL-Left, NR-Right
 - Flat tympanograms bilaterally
 - ENT exam conducted; tubes and ABR in OR recommended

- **Age 12 months:**

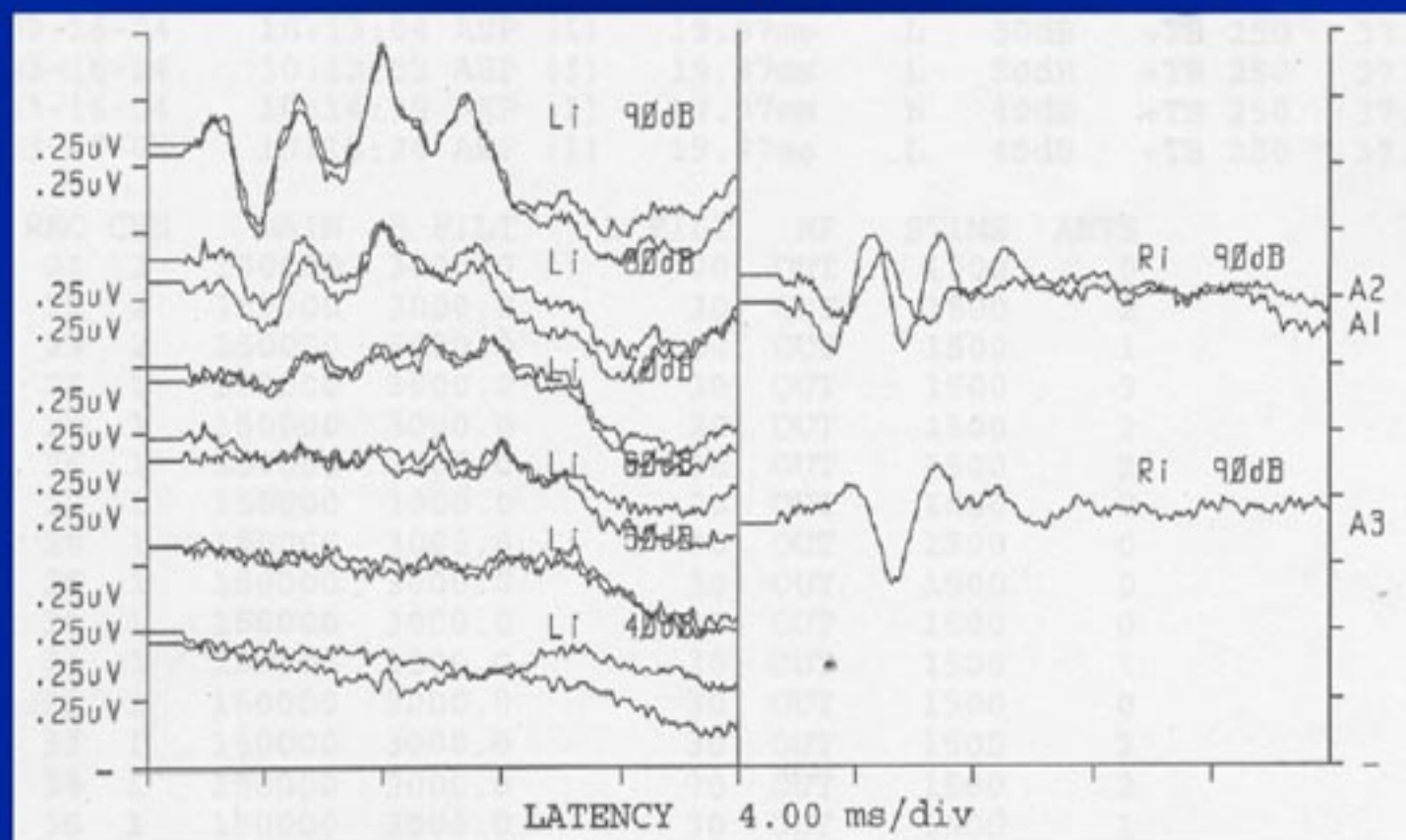
- Tubes placed and ABR and ASSR completed
- Hearing thresholds estimated on basis of ABR/ASSR

ABR (Clicks)

Left ear: 50dBnHL Right Ear: NR at 90dBnHL



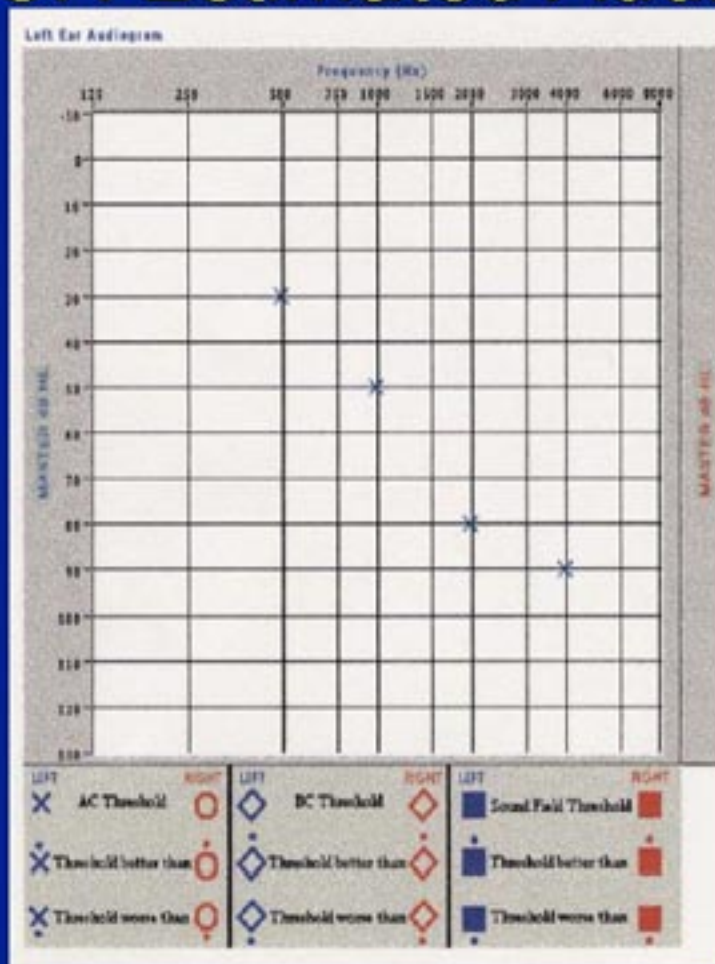
Low frequency tone burst
Left ear: 50dBnHL Right ear: NR at 90dBnHL



ASSR Thresholds

	500Hz	1000 Hz	2000Hz	4000Hz
Right Ear (dBnHL)	NR	NR	NR	NR
Left Ear (dBnHL)	40	60	90	100

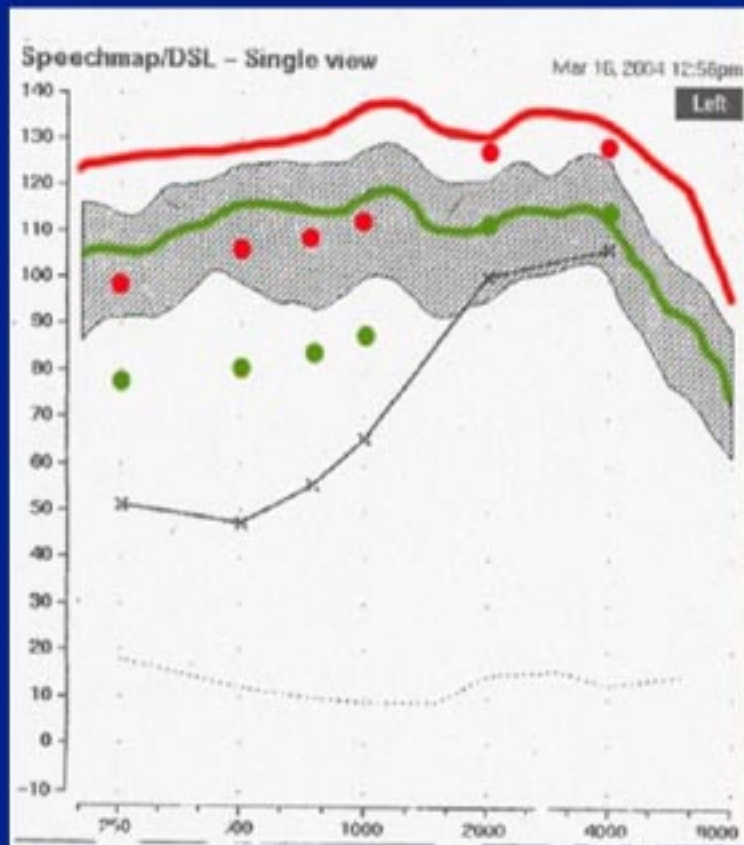
ASSR Estimated Audiogram



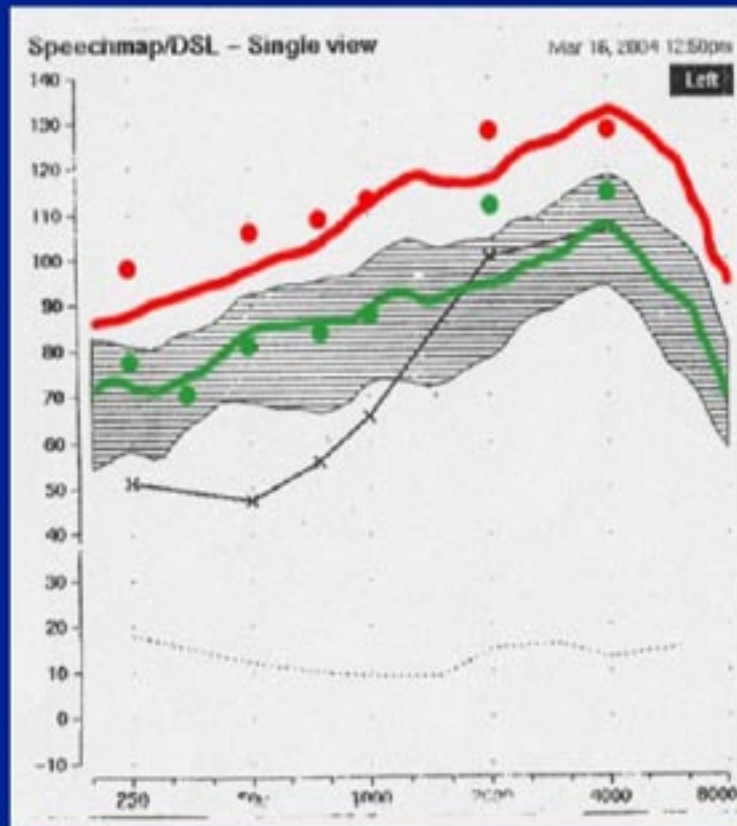
Estimated Thresholds Based on ABR/ASSR in OR

	500Hz	1000 Hz	2000Hz	4000Hz
Right Ear dBHL	>100	>100	>100	>100
Left Ear dBHL	30	50	80	85

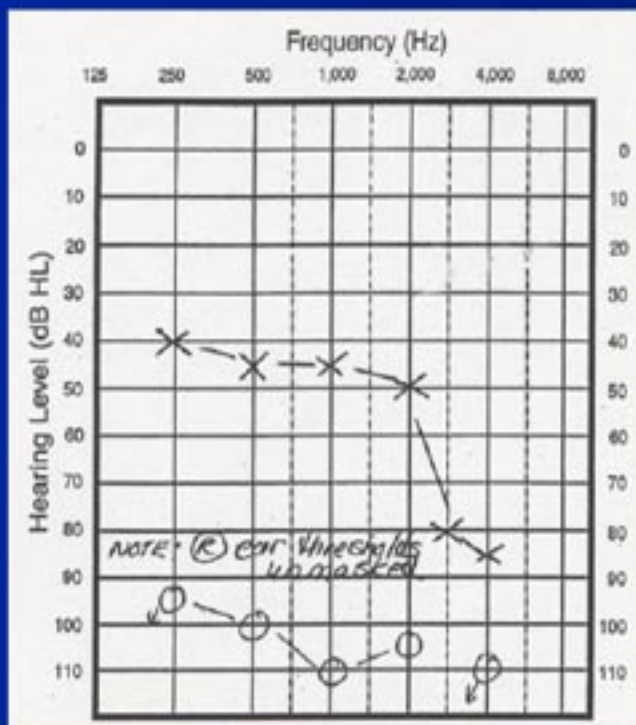
Verification measures for child's own conventional hearing aids using DSL targets on same day as OR testing



Verification measures for loaner programmable hearing aid using DSL targets



Age 13 months: VRA with insert earphones



- SDTs:
 - 95dBHL-R, 50dBHL-L
- Tympanometry
 - Large equivalent volume consistent with patent tubes

Discussion and recommendations for family by
audiologist and otolaryngologist
(via an interpreter)

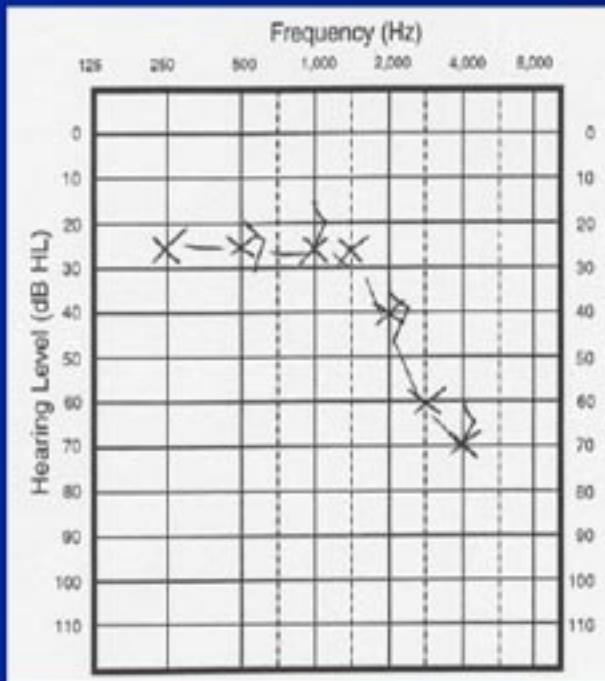
- New digital hearing aid recommended for left ear
- MRI scheduled one month later in view of asymmetric hearing results
- Suggested to family that they return for audiologic follow-up at UNC every three months
- Results communicated to local early intervention specialist and child service coordinator who will assist with scheduling appts.
- Cochlear implantation is not indicated
- With regular hearing instrument use and early intervention services, child expected to make progress in spoken language development

MRI Results:

"MRI of the 8th nerves shows clearly absent 8th nerve on the the right ear with normal inner ear morphology."

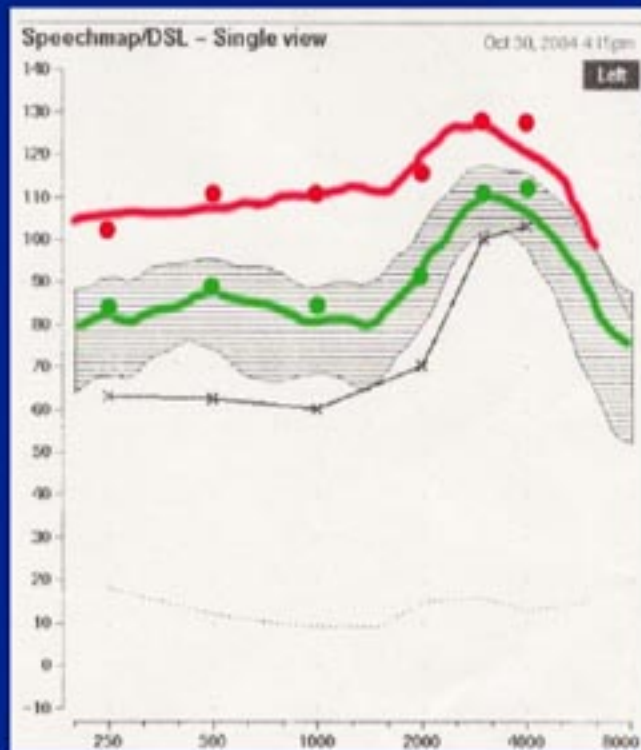
"The left ear has normal inner ear morphology with four nerves present in the internal auditory canal."

Age 18 months: VRA with insert earphone in left ear and bone conduction



- Tympanometry
 - Type C tympanograms; tubes no longer functional
 - 10-20 dB improvement in thresholds may be better due to intact TMs

Verification measures with new digital hearing for left ear dispensed at age 18 months



Key Points

- Early errors in audiologic evaluation can lead to multiple errors in management resulting in “snowball effect”
- Comprehensive physiologic assessment must include frequency-specific tone bursts and not only click stimuli
- Physiologic measures are critical, but including behavioral audiometry in test battery is essential for optimal management
- Middle ear status must be closely monitored and managed
- Collaboration between audiologist, otolaryngologist, and early intervention specialists is essential

Case Study #3

"Blake"

Background

- First child born at 25 weeks gestation
- Ventilated for 6 weeks
- Oxygen 3 ½ months
- Hyperbilirubinemia
 - Treated with lights, exchange transfusion
- Treated with antibiotics and diuretics
- Hospitalized 4 ½ months
- No family history of hearing loss
- Did not pass newborn hearing screen at hospital discharge

Age 6 Weeks:
Hospital #1 First Diagnostic Auditory
Brainstem Response (ABR) Evaluation

- *"ABR consistent with severe to profound sensorineural hearing loss bilaterally"*
- Repeat testing recommended one week later to confirm results

Age 7 Weeks:

Hospital #1 Second Diagnostic ABR

- ABR showed no response in either ear at maximum intensity levels to click stimuli or to tone bursts at 500Hz, 1000Hz, 2000Hz or 4000Hz
- Recommendations:
 - Referred to otologist for medical clearance for hearing aids
 - Fitted with high gain hearing aids following otologic exam

First Contact with UNC Hospital

Age 5 months (2 Months Adjusted Age)

- Mother makes first phone contact with UNC explaining family is moving to our area, requests appointment for ear impressions.
- Mother counseled that repeat ABR needed even though child has already had two ABRs
- Mother concerned that earmolds too small so appointment scheduled prior to ABR to take ear impressions

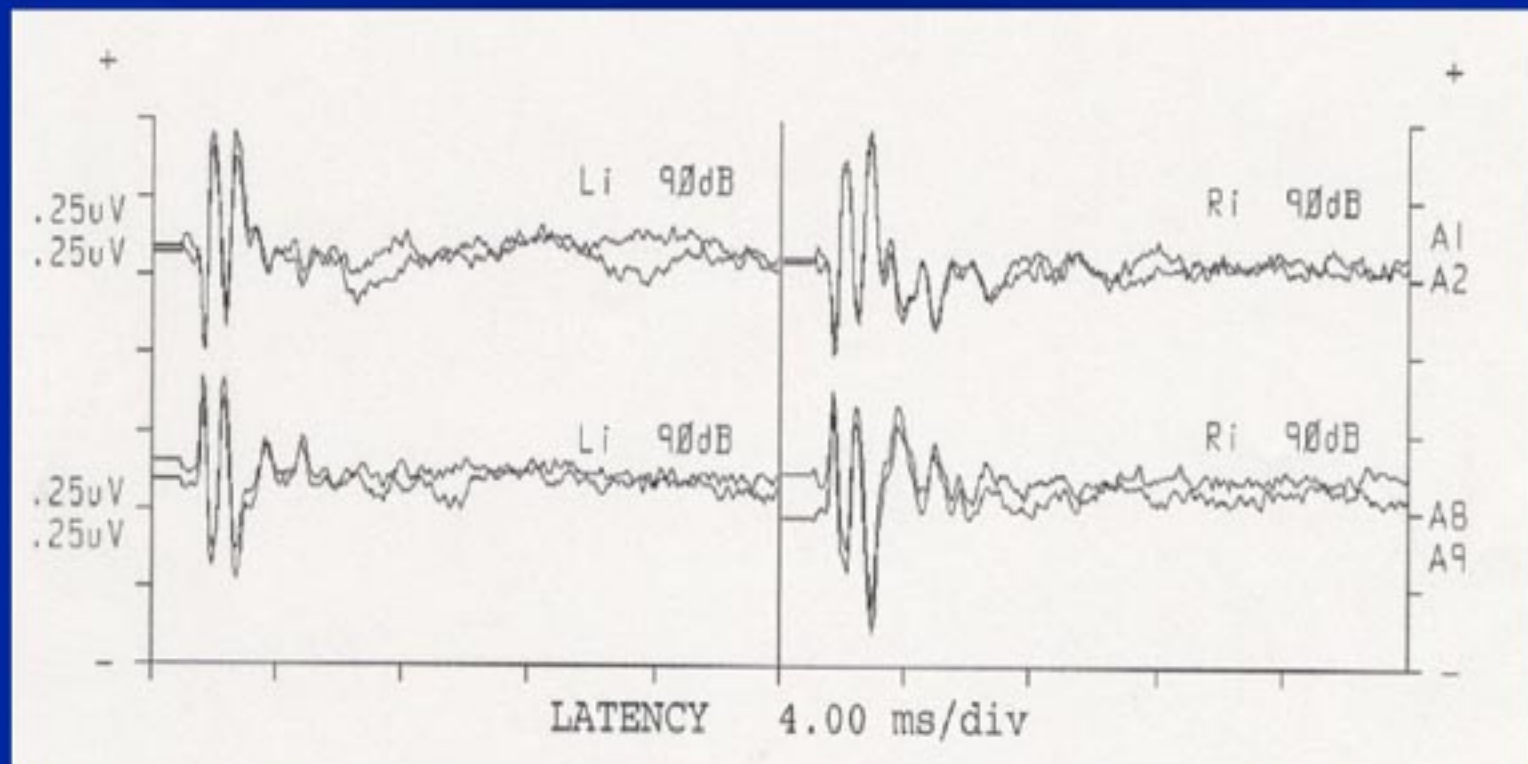
Mother's Comment During Appointment for Ear Impressions:

- *"It's strange, I know he's been diagnosed as being profoundly deaf but the other day I dropped the remote control on the TV and he startled."*

Age 6 Months (3 Months Adjusted Age): UNC Evaluation

- ABR at high intensity level with both rarefaction and condensation clicks shows only a cochlear microphonic with no evidence of neural response
- OAEs absent

ABR Obtained at UNC, Age 6 Months (3 Months Adjusted Age)



Family Meeting to Discuss Test Results:

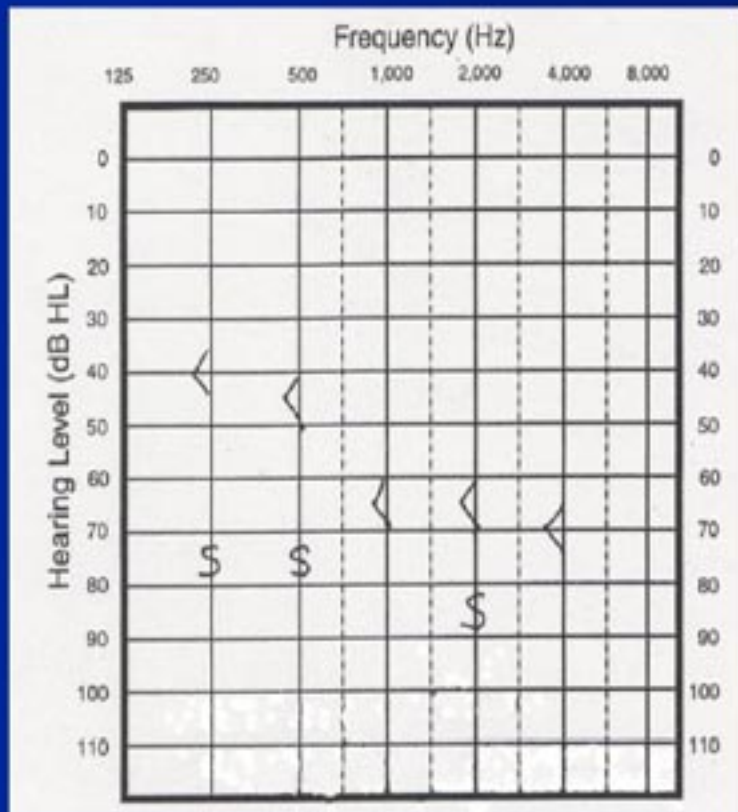
- ABR results consistent with AN/AD
- Explained that in cases of AN/AD ABR is not useful in predicting hearing threshold levels
- Recommended discontinuation of high gain hearing aids until behavioral audiometry can be obtained
- Discussed controversy surrounding treatment options (amplification, communication strategies etc.)
- Scheduled otologic exam
- Referral for early intervention services

Otologic Exam

- Normal otoscopic examination
- Referrals made:
 - MRI → No CNS problems, inner ears normal
 - pediatric neurology → normal
 - ophthalmology → normal
 - genetics → no abnormalities detected

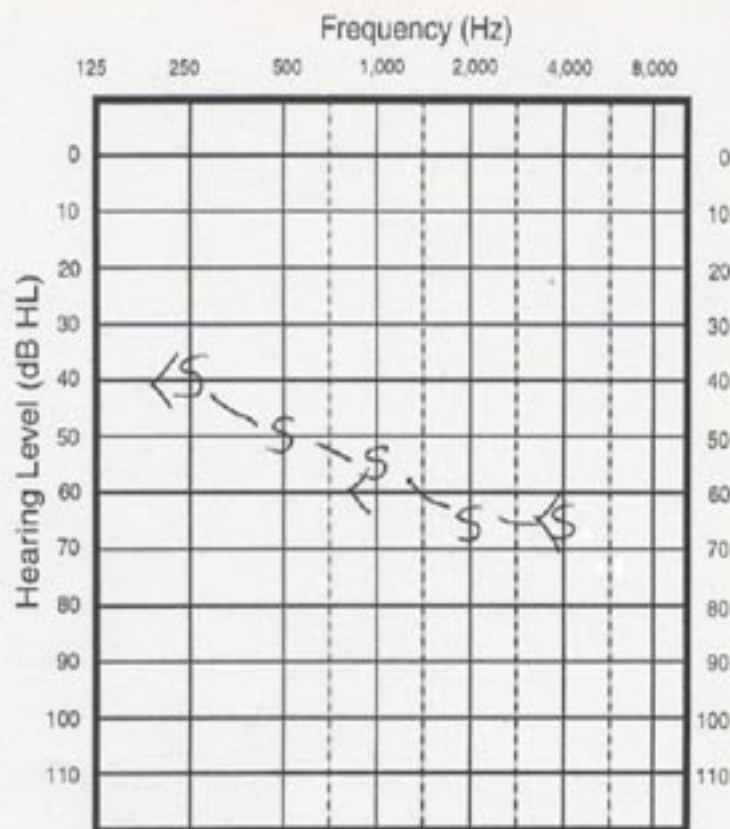
Age 9 Months (6 Months Adjusted Age)

First Attempt at Visual Reinforcement Audiometry (VRA):



- Child beginning to perform VRA task but responses probably "suprathreshold"
- Tympanometry:
 - Right: flat
 - Left: normal
- OAEs:
 - Right: not tested
 - Left: absent

Age 10 Months (7 Months Adjusted Age): Behavioral Audiometry with VRA



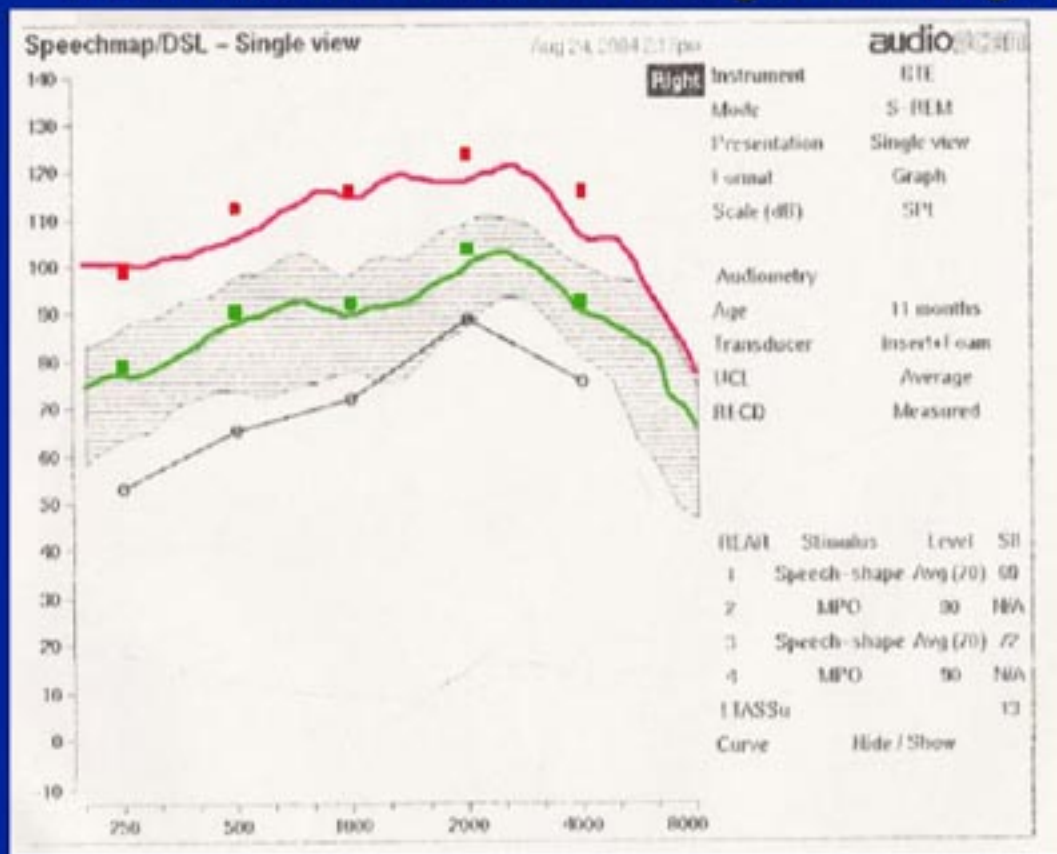
- Sound Field Audiogram:
 - moderate hearing loss for "better ear"
- Bone conduction thresholds confirm sensorineural HL
- Acoustic Immittance:
 - Right: normal
 - Left: normal
- Discussion with family
 - Decision made to proceed with amplification

Hearing Aid and FM Fitting

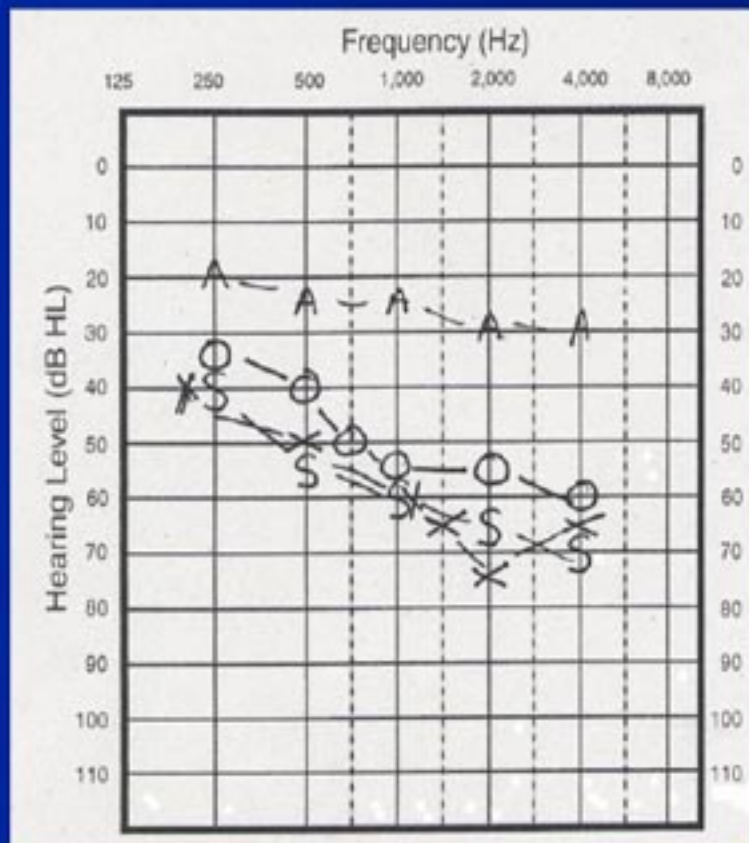
Age 10 ½ Months (7 Months Adjusted Age)

- Child fitted with new binaural digital hearing instruments with ear level FM receivers
 - Appropriate for moderate degree of hearing loss
 - Set to match DSL targets
- Parents counseled to carefully observe child's behavior and notify us if any signs of loudness discomfort
- Return appointment scheduled to continue assessment and obtain individual ear measures

Age 10 ½ Months (7 Months Adjusted Age) Hearing Aid Fitting Verification Measures Using DSL Targets

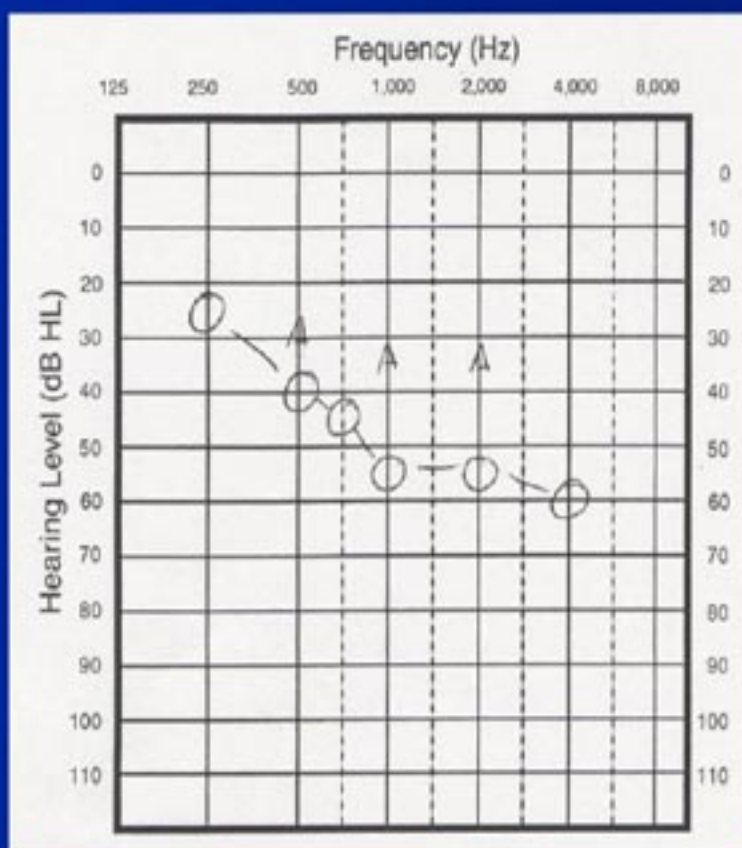


Age 12 Months (9 Months Adjusted Age): VRA with Insert Earphones Attached to Child's Earmolds



- Speech Detection Thresholds:
 - Unaided:
 - Right 40dBHL, Left 45dBHL
 - Aided
 - 20dBHL
- Tympanometry
 - Right: normal
 - Left: normal
- Sound field audiogram (unaided and aided) completed for demonstration to parents
- Parental Report:
 - Child began babbling with consonant sounds in past week: e.g. la,la,la, da,da, da

Age 18 Months (15 Months Adjusted Age): VRA with Insert Earphones



- Tympanometry
 - Right: Type A
 - Left: Type A
- Parental Report:
 - Child now using a few words and beginning to understand simple commands

Age 24 Months (21 Months
Adjusted Age):
VRA with Insert Earphones

- Child conditioned for play audiometry procedure but limited attention span
 - Results similar to previous audiograms
- Tympanometry
 - Right: normal
 - Left: normal



Age 24 Months (21 Months Adjusted Age): Communication Status

- Parental Report:
 - Child has large vocabulary using many two word combinations
 - Comprehension of language seems very good
- Early Speech Perception Test (ESP) administered
 - Aided (auditory only condition) at 50dBHL:
 - Able to accurately identify from closed set of objects for spondee and monosyllabic words

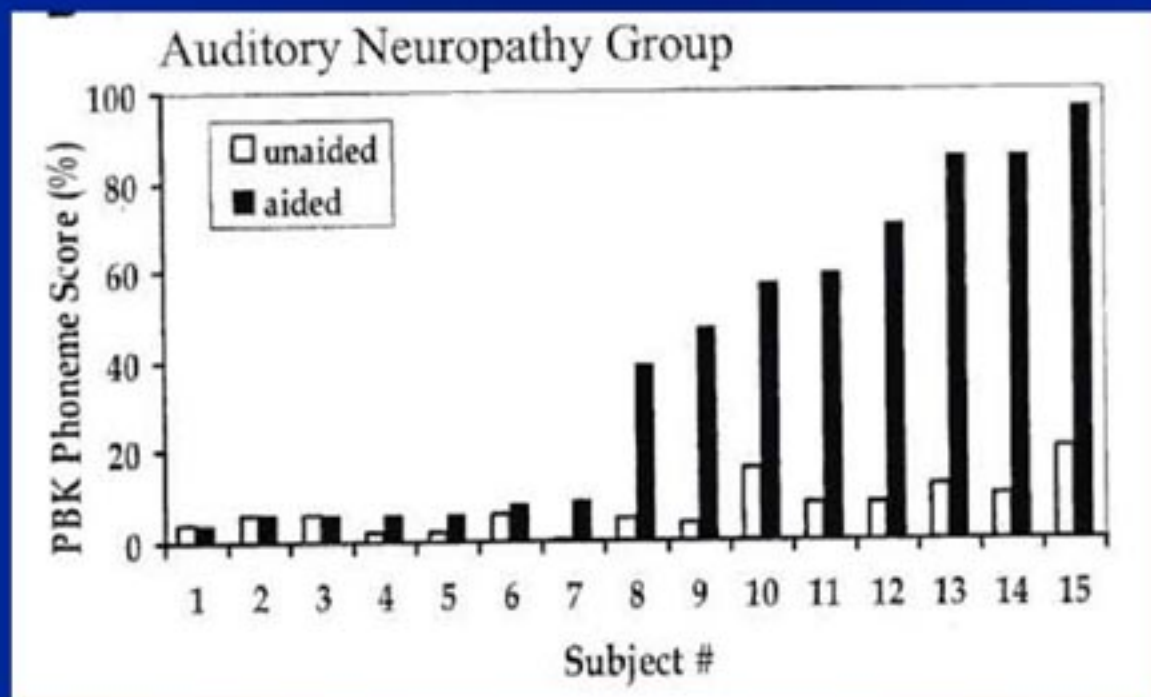
3 years, 9 months

- Continuing to receive services from auditory verbal therapist and speech and language pathologist
- Functioning in average range in receptive and expressive language development
- Working on articulation errors

Key Points

- Physiologic measures critical, but behavioral audiometry essential for optimal management
- Middle ear status must be closely monitored and managed
- Adequate medical work-up essential (e.g. pediatrics, otolaryngology, genetics, ophthalmology, neurology)
- Important to evaluate aided speech perception abilities as soon as possible
- Communication with early intervention specialists familiar with sensory hearing loss and knowledgeable about AN/AD essential

Hearing Aids in Children with AN/AD



Rance et al Ear and Hearing 2002

Case Study # 4

"Autumn"

Background

- Newborn Screen
 - Referred on left
 - Passed on right
- Age 2 months:
 - Diagnostic ABR: moderate HL left, normal right
 - Family chooses to not provide HA on left
- Age 2 ½ months:
 - Otologic evaluation: MRI, EKG, connexin 26 and CMV testing ordered

Background

- Age 3 months:
 - Referred to Beginnings for information and referral to early intervention
- Age 4 months:
 - MRI: Bilateral enlarged vestibular aqueducts, enlargement of endolymphatic sacs and left temporal arachnoid cyst.
 - Otologist advises of risk for progressive hearing loss and avoiding head trauma and refers to neurology and genetics for evaluation
- 5 months: Neurology consult completed
 - Arachnoid cyst likely incidental finding, not of major concern, family advised to follow up in one year

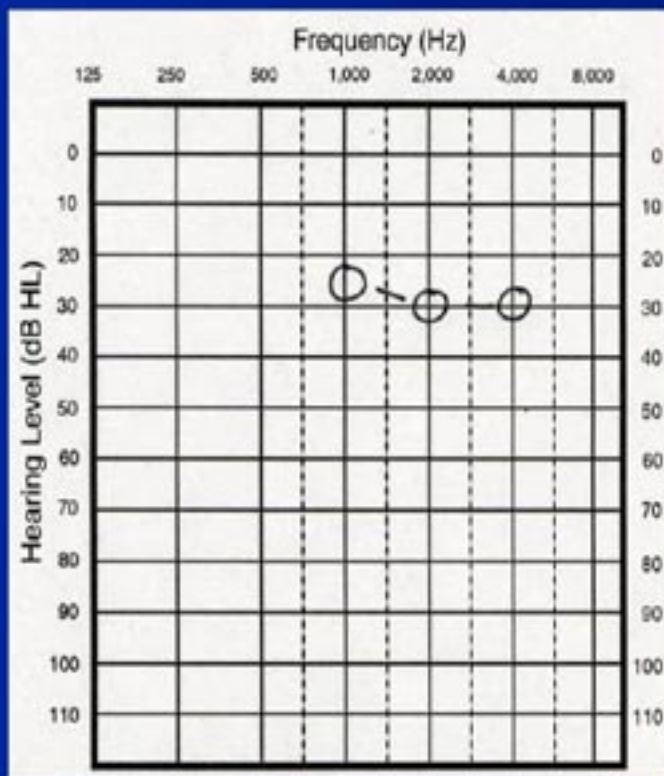
Case 5

(Caleb)

Unilateral: Continued

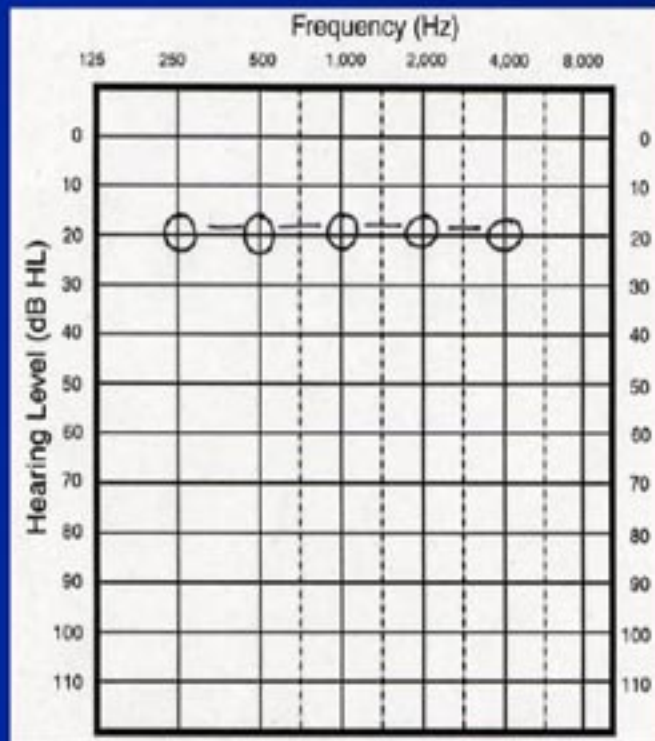
- 6 months: Genetics consult completed
 - Most common cause of EVA is alteration of Pendred gene
 - Several other syndromes can be associated with EVA including branchio-oto-renal syndrome
 - Will test for Pendred's and if negative will order renal ultrasound
 - Lab results shows child is connexin 26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

Age: 7 months:



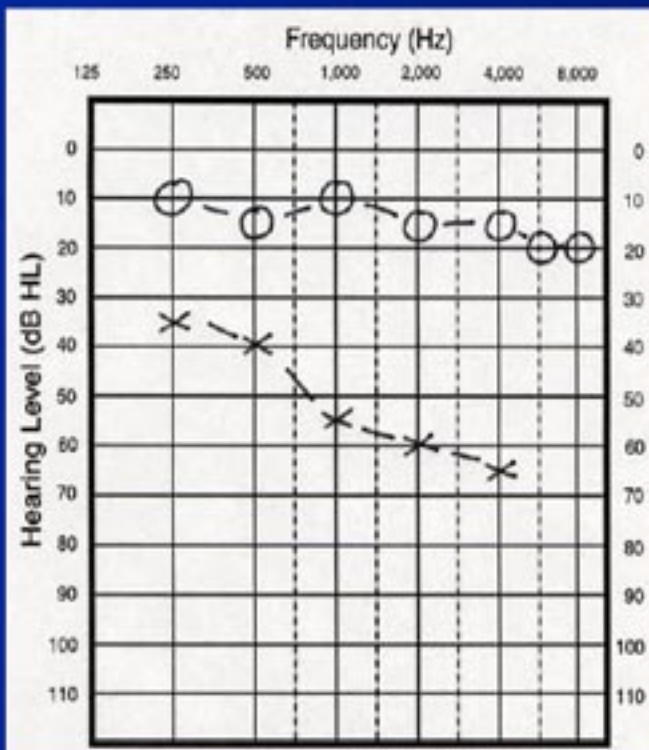
- First attempt at VRA with insert earphone
- Tympanometry
 - Right: normal
 - Left: flat
- Otoacoustic Emissions
 - Right: Absent above 3000Hz
 - Left: Absent

Age: 8 months



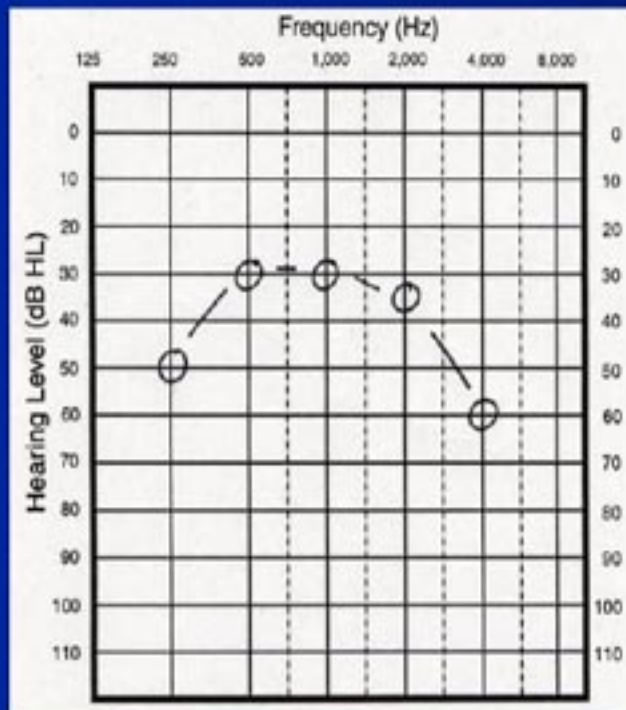
- Tympanometry
 - Right: normal
 - Left: -275
- Otoacoustic Emissions
 - Right: Absent above 2000Hz
 - Left: Absent

Age: 12 months



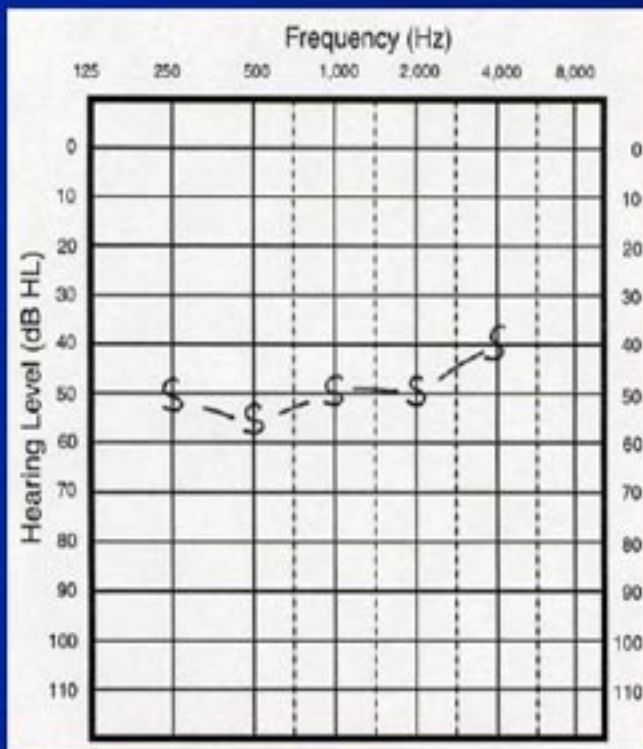
- Right ear:
 - Normal
- Left ear:
 - Mild to moderate
- Tympanometry
 - Right: normal
 - Left: normal

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

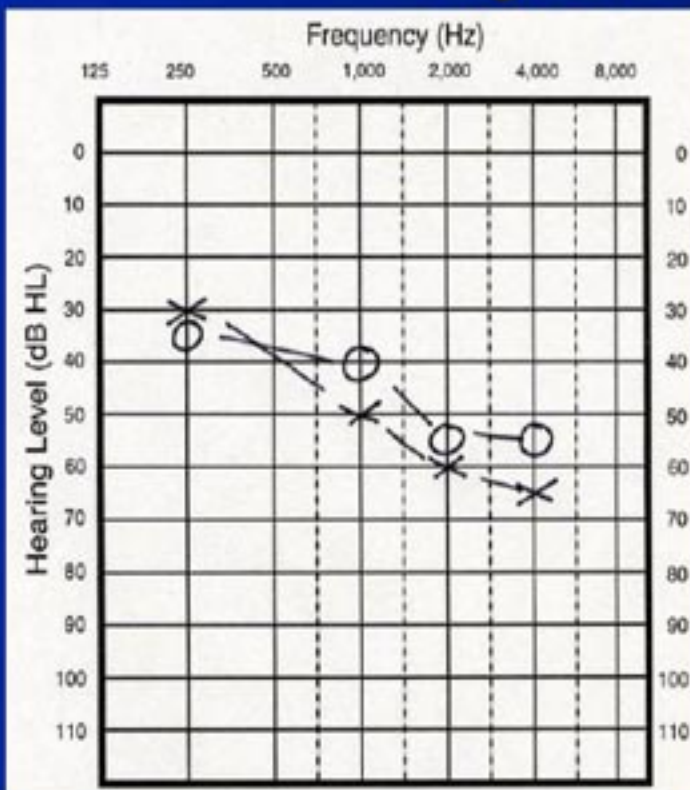
Age: 18 months



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

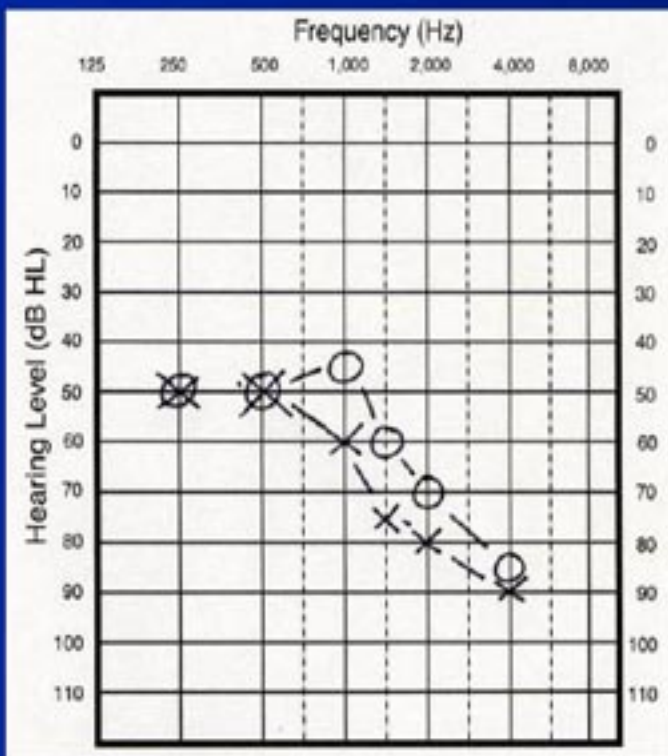
Sedated ABR

Age 20 months



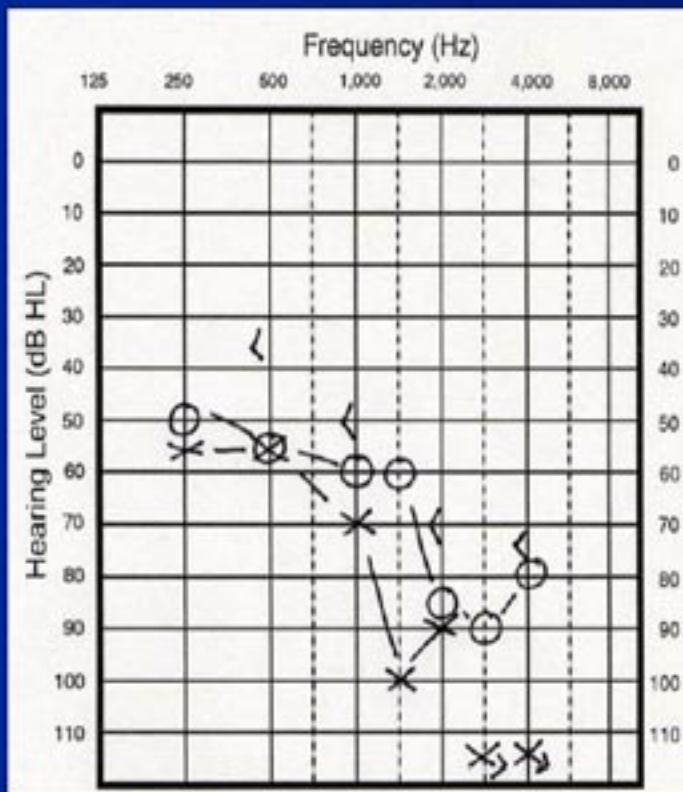
- ABR repeated under sedation
- Estimated thresholds based on tone burst ABR
- Binaural hearing aids and personal FM dispensed 2 weeks later

Age: 23 months



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

Age: 24 months



- Hearing aids readjusted to better match DSL targets
- 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

Key Points

- Comprehensive team evaluation useful when working with infant with newly diagnosed HL
 - Audiology, ENT, Genetics, Early Intervention Specialists, Neurology, 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

Muchos Gracias!!!

Summary of Essential Components of Early Infant Hearing Program

- Early diagnostic ABR used to estimate initial thresholds for hearing aid fitting
- RECDs measured initially and at time of new earmolds
- Hearing aids programmed using manufacturer's software AND.....
- Adequacy of hearing aid setting verified with independent verification system
- Goal is to obtain behavioral audiogram for each ear for 250-4000Hz by 8-9 months of age
- Hearing aids re-programmed if thresholds have changed
- Minimal time spent obtaining aided audiograms
- Evaluate aided speech recognition abilities and speech and language outcomes early

Key Points

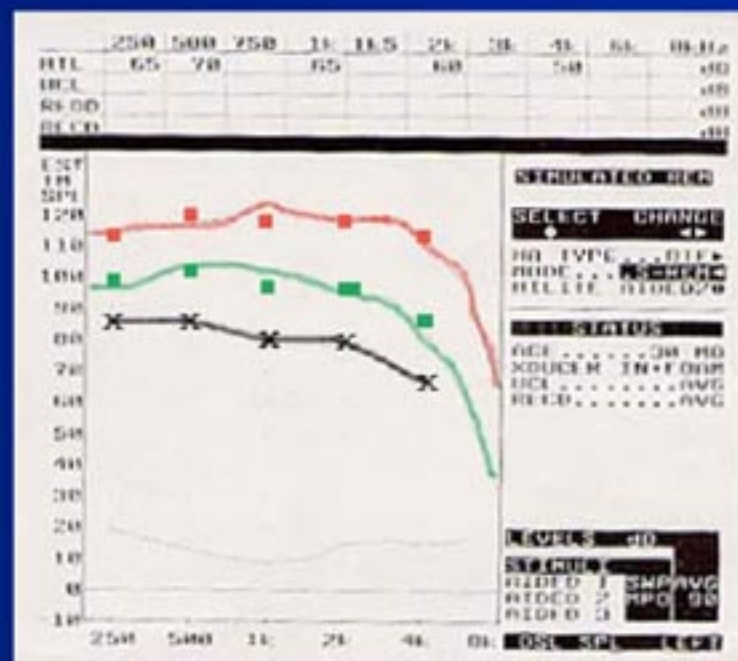
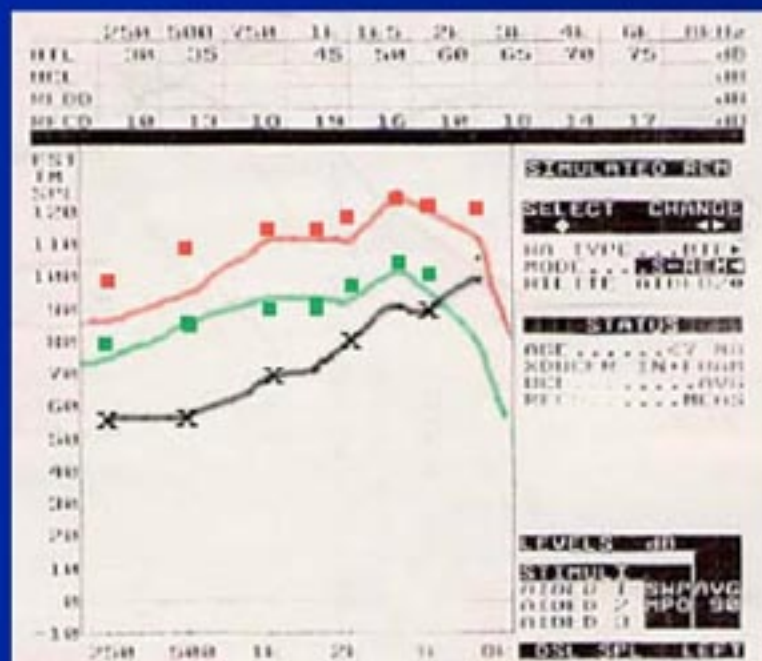
- Middle ear status has significant effect on ABR results
- ABR testing doesn't always give us the "full-picture"
- Air and bone conduction testing with VRA needed
- Important to determine family's preference for timeline re intervention, particularly when multiple medical problems are present
- Cooperation between audiologist and ENT physician essential

Current Status

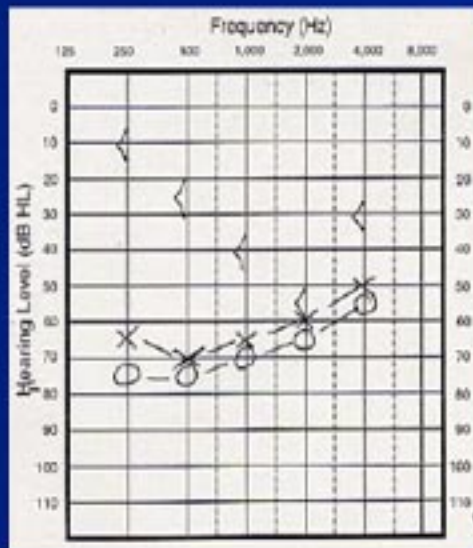


- Imitating 2-3 word phrases
- Beginning to produce some two word phrases independently
- Identifies body parts
- Can identify colors
- Can make “scary face” on demand

SPL-O-Grams with estimated and actual thresholds

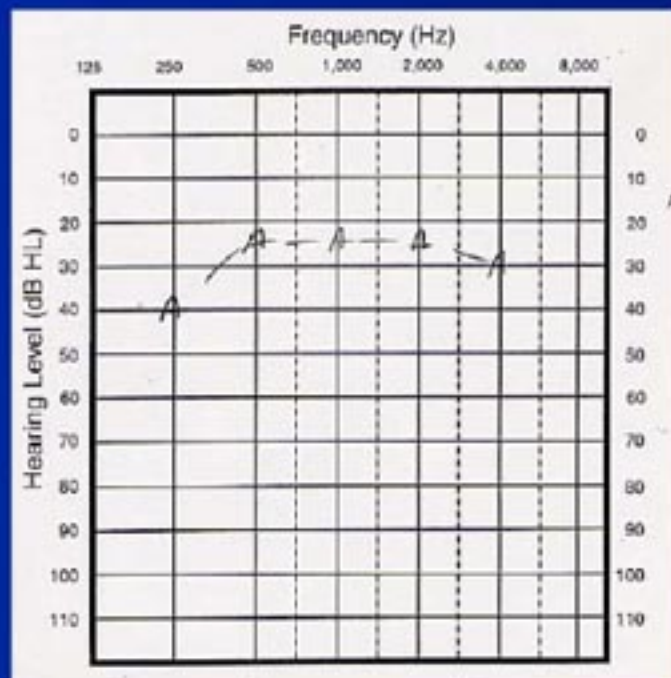


2.5 yrs-Play Audiometry

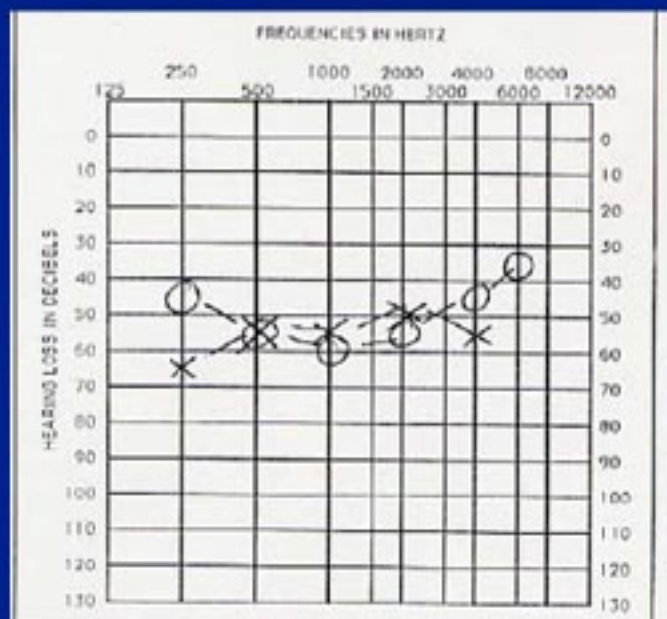


- Tympanometry: large equivalent volume

Aided Audiogram



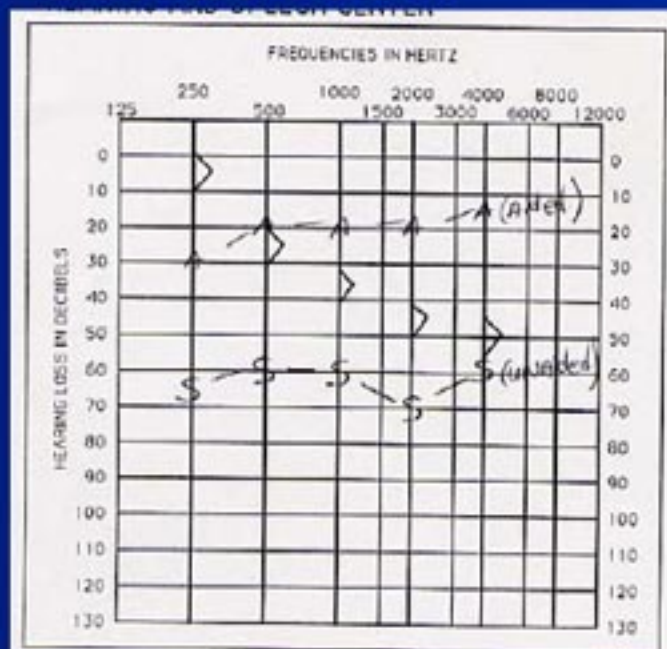
Age 13 mo-VRA with insert earphones



11 mo-VRA in Sound Field

Air & Bone-conduction

- Audiometry consistent with moderate mixed hearing loss

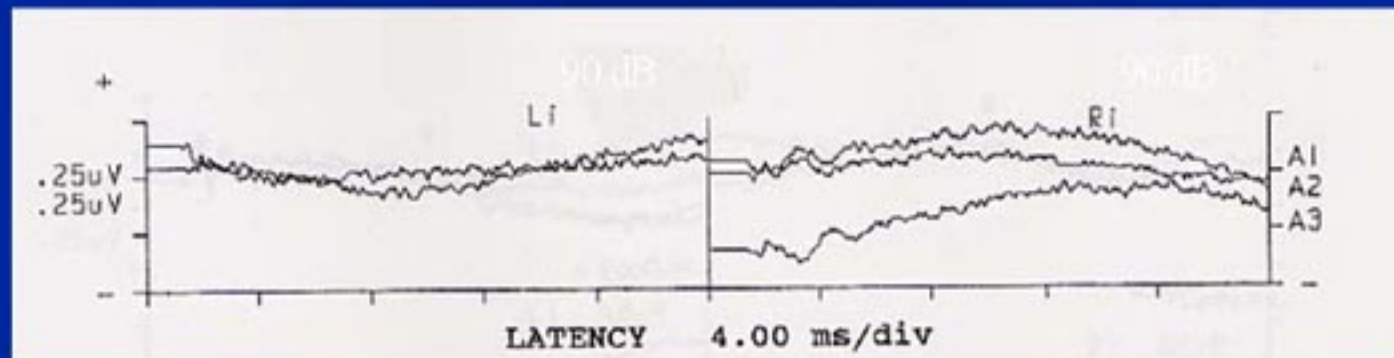


Typical audiometry

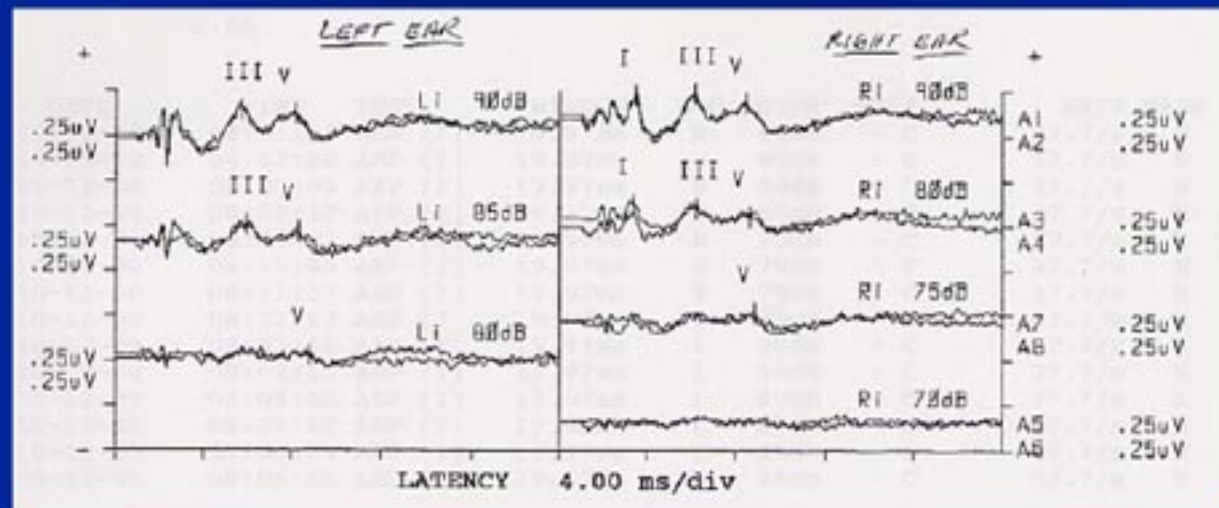
Large equivalent volume

Age 8 mo-OR ABR Post-Tubes

250Hz tone burst



Age 8 mo-OR ABR Post-Tubes Clicks





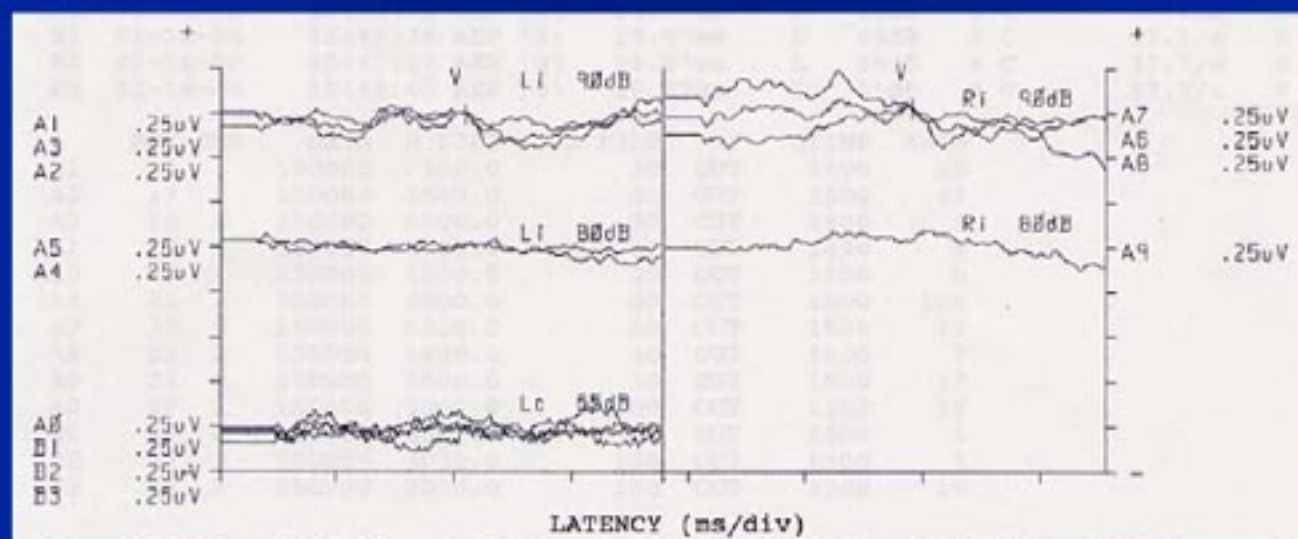
History

- 4 ½ months: Hearing Aid Fitting
- 6 months: "Child beginning to condition for VRA"
- 7 months: Tympanograms flat
- 8 months: Tympanograms flat, tubes recommended by ENT
- 9 months: tubes, ABR in OR

Intervention Plan

- Family counseled re: hearing loss
- Multiple medical problems needing to be addressed but family motivated to address hearing problem
- Return appt. scheduled at age 2 ½ months

Diagnostic ABR in NICU age 2 weeks:
Clicks: 90dB-R&L, No clear responses below 90dB
No response by bone-conduction



Initial Impression: Severe to profound bilateral
sensorineural hearing loss

History

- AABR in NICU day 7:
 - Refer R&L 35dB & 70dB
- Diagnostic ABR NICU day 7:
 - Clicks: NR at 90dB-R, 90dB ?80dB-L
- Diagnostic ABR NICU 2 weeks:
 - Clicks: 90dB-R&L, No clear response below 90dB

Background



- Child with Kniest Dysplasia:
 - Cleft palate
 - Short stature
 - Myopia
 - Respiratory problems
- Followed by multiple medical specialists:
 - Audiology
 - ENT
 - Genetics
 - Ophthalmology
 - Orthopedics
 - Craniofacial Team
 - Pulmonary
 - Physical Therapy