Understanding Auditory Neuropathy: Diagnosis and Management

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Pediatric Audiology Program
University Of North Carolina
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Universal NB screening legislation 1999

Pediatric Audiology and CI Teams

CASTLE pre-school

Total 1400 infants and children

- 800 using amplification
- 600 with cochlear implants
- 180+ with ANSD diagnosis

Outline

- Overview and Definitions
- Variations in Presentation
- Implications for Clinical Practice
- Case Studies

Auditory Neuropathy: A Definition

Clinical syndrome characterized by electrophysiological evidence of normal or near normal cochlear function and absent or abnormal auditory pathway transduction

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Audiologic Findings

- Normal outer hair cell function as measured by present otoacoustic emissions (OAEs) or the presence of a cochlear microphonic (CM).
  - OAEs may be present initially but disappear over time
- Abnormal auditory nerve response as observed by absent or markedly abnormal ABR
- Acoustic reflexes are absent in most cases

Clinical Characteristics Reported

- Pure tone thresholds ranging from normal to profound
- Disproportionately poor speech recognition abilities for the degree of hearing loss
- Difficulty hearing in noise
- Impaired temporal processing
- Hearing fluctuation
- Some individuals with AN have little or no communication difficulties while others are functionally deaf
- Not all individuals diagnosed with AN experience the same problems

(Starr et al 1996, Zeng et al 1999, Kraus et al 2000, Rance et al; 2002; 2004; 2005, Zeng and Liu, 2006)

Auditory Neuropathy: Not a New Disorder

- Term “Auditory Neuropathy” first introduced by Starr et al in 1996
- Not a new disorder
  - Early reports of children with absent ABRs responding to sound
    - Davis and Hirsch, 1979
    - Worthington and Peters, 1980
    - Kraus et al, 1984
- Newer technologies and procedures, in particular OAEs made it possible to conduct differential diagnosis of sensori-neural hearing loss

Starr et al Report 1996

- 10 patients with absent or abnormal ABR with evidence of normal cochlear outer hair cell function
  - Present cochlear microphonic and otoacoustic emissions
- Patients ranged in age from 4-49
- Presented without neurologic involvement at time HL identified
- 8/10 patients subsequently diagnose with other peripheral neuropathies including 3 with Charcot Marie Tooth disease
- Speech recognition scores were poorer than expected for degree of hearing loss
- Results obtained seemed to be characteristic of a “neural hearing loss”

Prevalence

- Disorder initially thought to be rare
- Many published reports since late 90's describing patients with similar audiologic test findings (absent ABR with present CM and/or OAEs)
- Estimates range from 7-10% of children diagnosed with permanent hearing loss

(Rance 2005)

Possible Etiologies and Associations

- Genetic Etiologies:
  - Syndromic:
    - Charcot-Marie-Tooth disease; Friedrich's Ataxia; Hereditary motor and sensory neuropathy (HSMN)
  - Non-syndromic:
    - Recessive genetic mutations: Otoferlin (OTOF), Pejvakin (PJVK)
    - Autosomoal dominant mutations: AUNA1 (onset of auditory symptoms in late teens)
- Perinatal Conditions:
  - Hyperbilirubinemia
  - Hypoxia
  - Low birth weight
  - More common in premature infants

Rance (2005);Rapin & Gravel (2003);Starr et al. (2003); Hayes 2011

Possible Etiologies and Associations (cont.)

- Congenital Conditions:
  - Cochlear Nerve Deficiency
- Infectious Processes
  - Viral Infections (e.g. mumps, meningitis)
- Head injury
  - e.g. Shaken baby syndrome

Rance (2005);Rapin & Gravel (2003);Starr et al. (2003); Hayes 2011

Lessons from the Past

- Kalamazoo (1977)
- ABR/OAE testing unavailable at that time
- 19 year old educated at school for the deaf
- ASL primary mode of communication
- Normal hearing sensitivity by pure tone audiometry
- Central deafness or ?ANSD
- More tools available now but still many questions
- Current Interest in AN

Auditory Neuropathy: Challenges/Questions

- What should we call this disorder?
- How do we accurately diagnose it?
- What should we tell a family to expect following initial diagnosis of AN when infant is only a few weeks of age?
- Do all children who present with audiological findings of AN have the same disorder or to the same degree?
- Can treatment protocols be generalized or should they be individualized?
- Are there clinically available diagnostic tools that allow us to predict benefit from a particular technology?

Auditory Neuropathy: Challenges/Questions

- Will hearing aids be helpful for the short term or the long term?
- What constitutes an adequate trial period with amplification?
- How do we determine who will benefit from hearing aids or cochlear implants?
- Will alternative hearing aid processing strategies result in better performance?
- What communication approach is best?

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Controversy

Exists in almost every aspect of disorder:

- Terminology
- Etiology
- Possible Mechanisms
- Treatment

Terminology

- Starr et al 1996:
  - » Auditory neuropathy
- Berlin et al 2001:
  - » Auditory neuropathy/dys-synchrony
- Starr et al 2004:
  - » Pre-synaptic (Type I):
    - When evidence of hair cell involvement exists
  - » Post-synaptic (Type II):
    - When patient has evidence of auditory nerve involvement

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Terminology

- Gravel and Rapin 2006:
  - Sensory hearing loss (hair cells)
  - Auditory neuropathy (pathology of spiral ganglion cells and VIIIth nerve axons)
  - Central hearing loss (central auditory pathway)
  - Neural conduction disorder (when differentiation cannot be made)
- Gibson et al 2008:
  - Imaging, genetic and electrophysiologic testing should allow us to identify pathologic entities according to site of lesion
  - Blanket terms such as AN/AD may be more misleading then helpful

- *Guidelines Development Conference: Identification of Infants and Children with Auditory Neuropathy*

  Lake Como, Italy, June 19-21, 2008

Found at:

http://www.thechildrenshospital.org/pdf/Guidelines%20for%20Auditory%20Neuropathy%20-%20BDCCH.pdf

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Panel Members

- Gary Rance
- Christine Petit
- Barbara Cone
- Deborah Hayes
- Charles Berlin
- Pat Roush
- Yvonne Sininger
- Jon Shallop
- Kai Uus
- Arne Starr

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Guidelines:
Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder

- Terminology
- Diagnostic Criteria
- Comprehensive Assessments
- Audiological Test Battery
- Amplification Strategies
- Considerations for Cochlear Implantation
- Habilitation for Communication Development
- Screening
- Monitoring Infants with “Transient” ANSD
- Counseling Families of Infants with ANSD

Terminology Considerations

- Same constellation of findings with different sites of lesion:
  - Auditory nerve
  - Synaptic dysfunction at junction of inner hair cell/auditory nerve
  - Myelin disorder
  - Cochlear nerve deficiency (small or absent 8th nerve)
- Panel sought to identify simplified terminology to reflect an *auditory disorder* with a range of presentations secondary to *variety of etiologies*
- *AUDITORY NEUROPATHY SPECTRUM DISORDER*

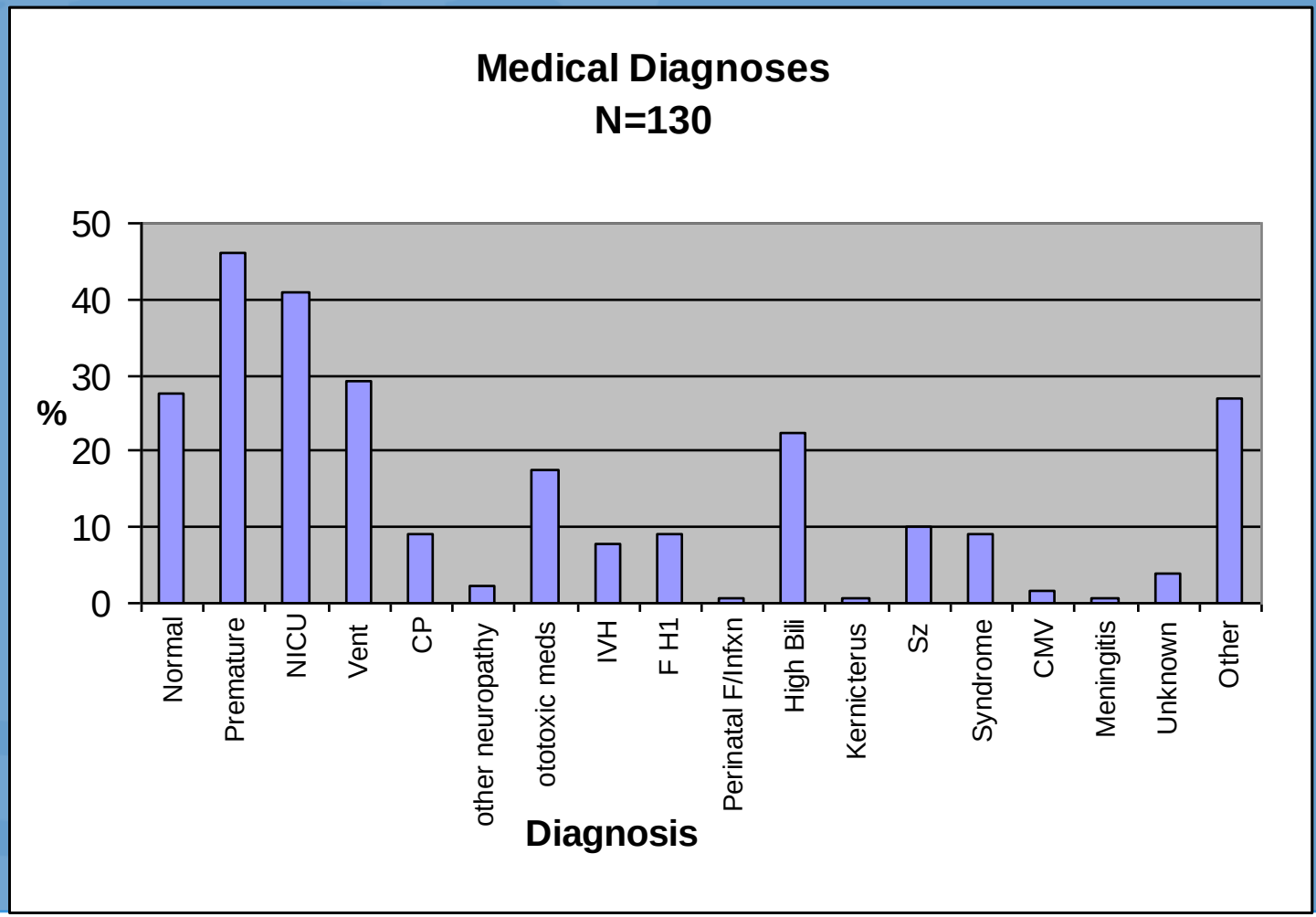
Diagnostic Criteria

- Minimum Test Battery Required for Diagnosis:
  - » Tests of cochlear hair cell (sensory) function:
    - Otoacoustic emissions and/or
    - Cochlear microphonics
- Tests of auditory nerve function:
  - » Click-evoked auditory brainstem response (ABR) to high-level click stimuli

Comprehensive Evaluations Following Diagnosis with ANSD

- Otologic
- Radiologic imaging (MRI/CT)
- Neurologic
- Medical Genetics
- Ophthalmologic
- Pediatric and Developmental Evaluations
- Communication Assessment

Why Comprehensive Medical Evaluation is Important
UNC-Chapel Hill ANSD Children
72% have some positive history)



Otologic Examination

- Medical History
- Ear Exam
- Etiology
- Other associated problems
  - Seizures
  - Motor delays
  - Visual problems
  - Ear canal problems
  - Otitis media
- Radiologic Studies (MRI/CT)
  - Inner ear malformations
  - Cochlear nerve integrity
- Other studies as needed

Recommended Audiologic Test Battery

- Auditory Brainstem Response (ABR)
- Acoustic Immittance Measures
  - » Tympanometry
  - » Acoustic Reflex Testing
- Otoacoustic Emissions Testing
- Behavioral Audiometry
  - » VRA, BOA, play audiometry
- Speech Recognition Testing

Recommended Audiologic Test Battery

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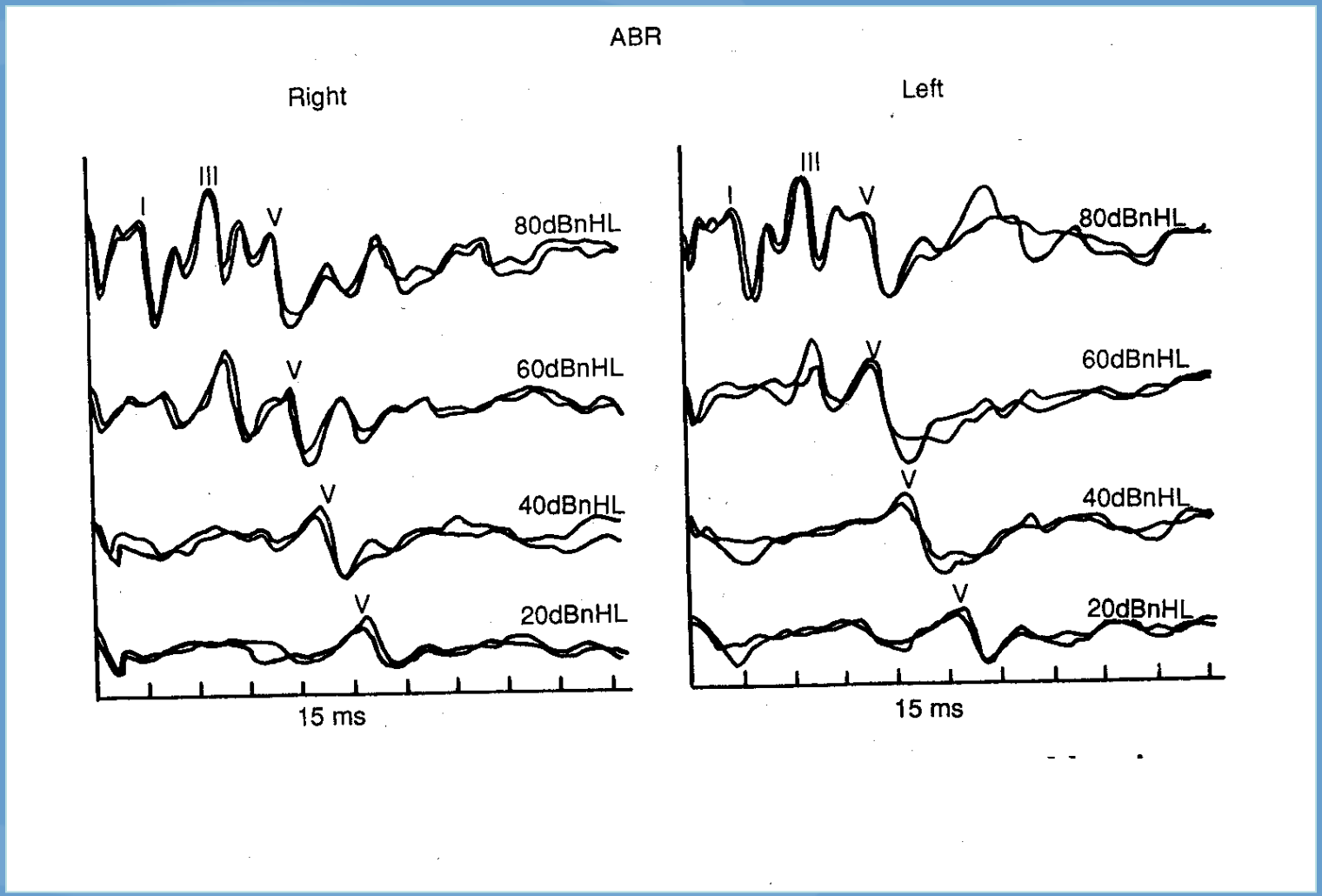
Normal ABR



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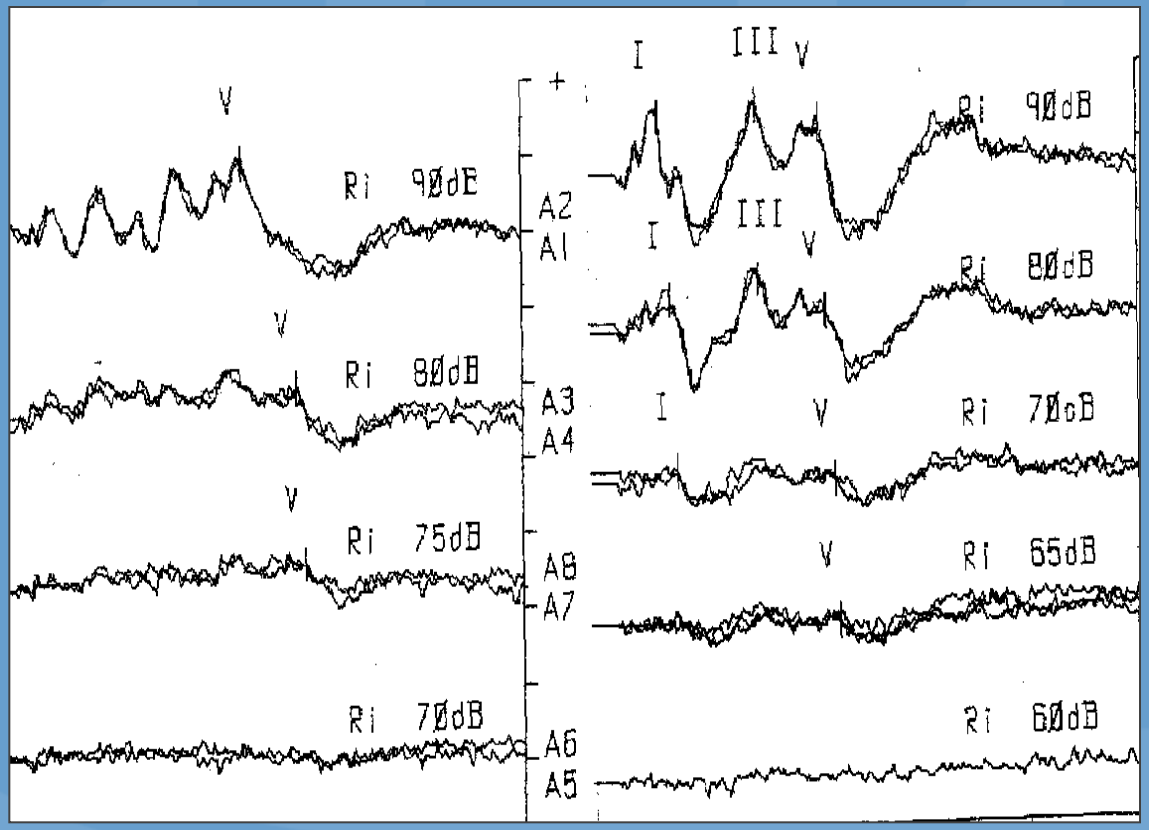
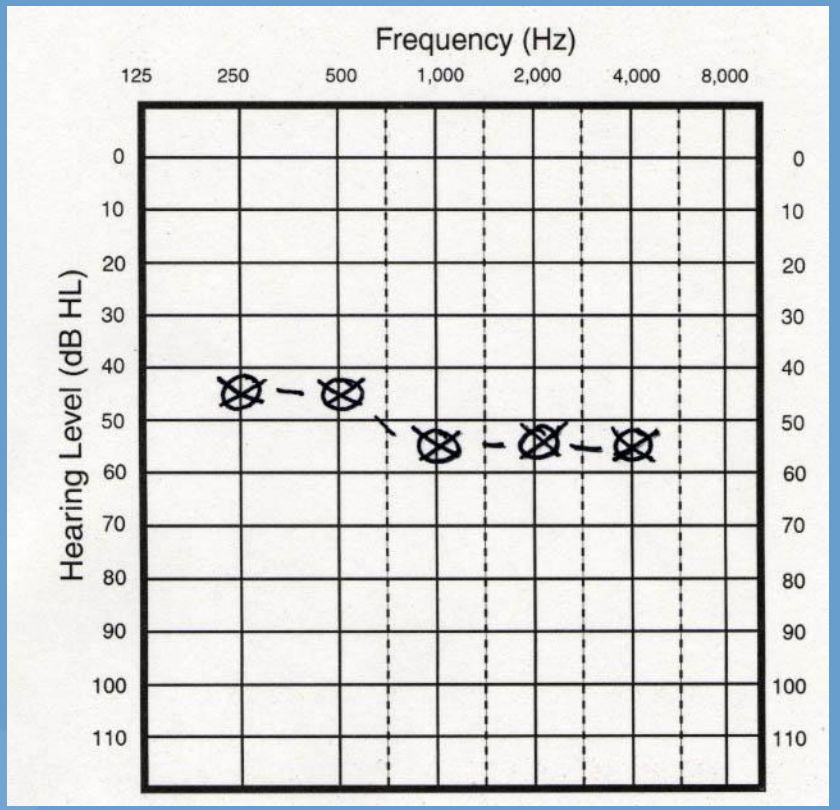
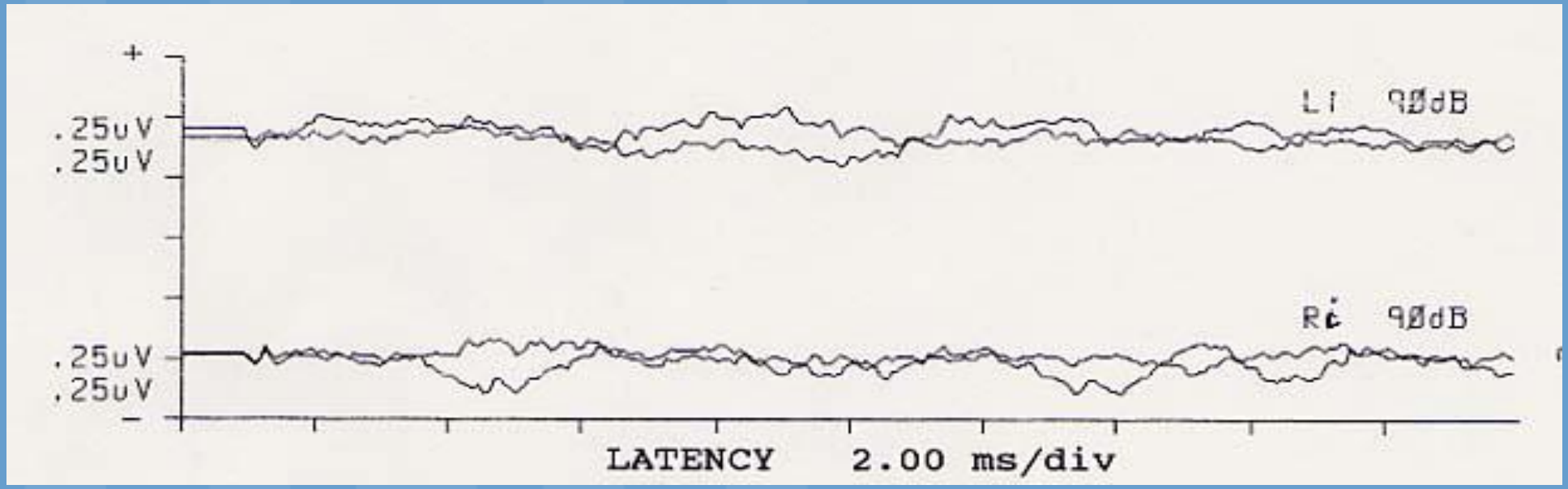
Estimating the Audiogram from ABR





Absent ABR with No Cochlear Microphonic: Child with profound hearing loss



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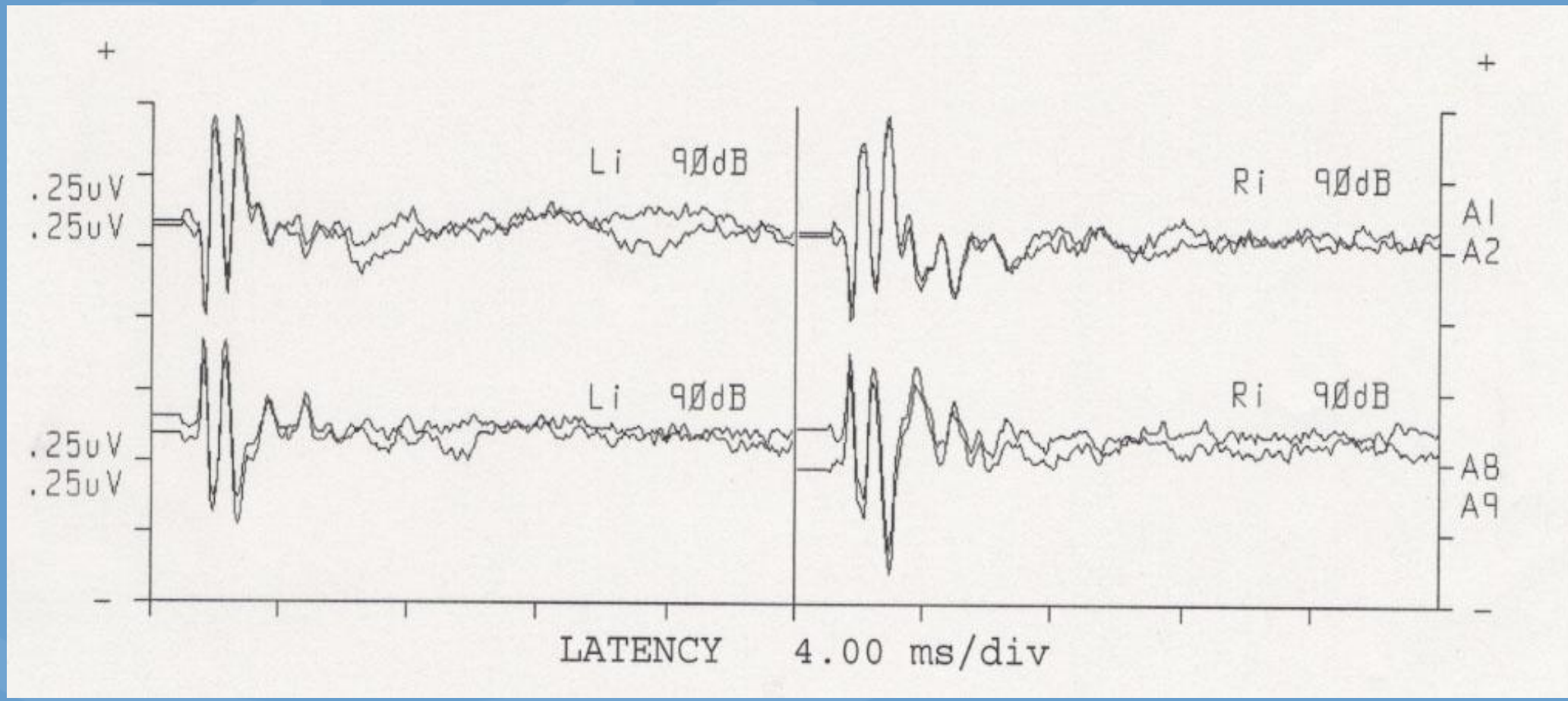
Abnormal ABR with Present CM



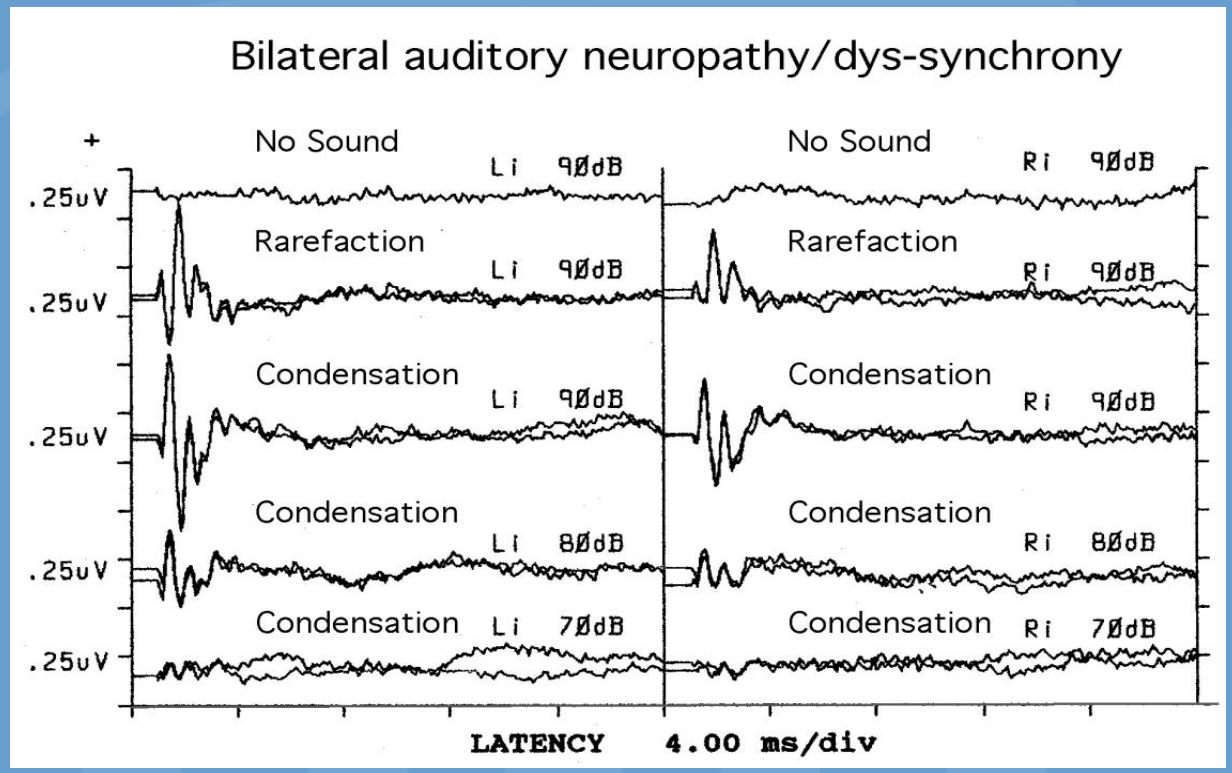
What is a Cochlear Microphonic (CM)?

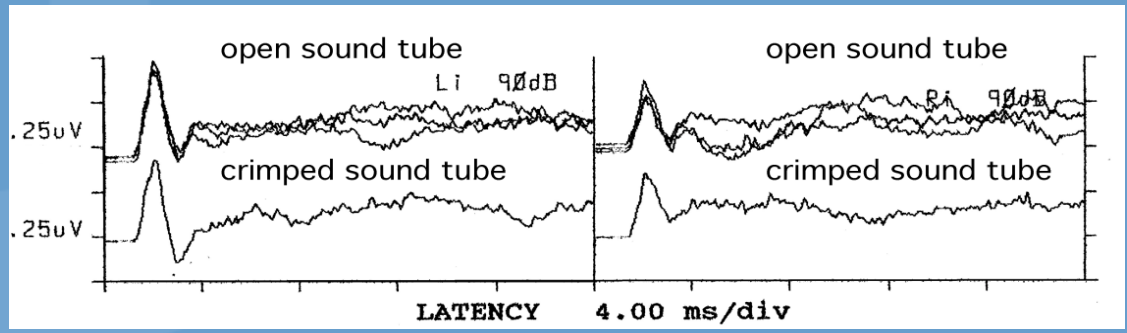
- Pre-neural response (occurs before Wave I in the ABR)
- Unlike the ABR, the CM shows a direct phase relationship to the acoustic wave form. When the polarity of the stimulus is changed there is a reversal of CM waveform
- Considered to have limited clinical use in past; renewed interest in diagnosis of ANSD
- CM can be recorded in normal ears, ears with “typical SNHL” and ears with ANSD
- Significance in ANSD is when CM is present when neural response is absent or markedly abnormal
- Amplitudes larger in patients with CNS problems (Santarelli et al 2006)

ABR Protocol for Evaluating CM

- Must have adequate recording conditions
  - » Infant ready to sleep
  - » Avoid electrodes positioned over transducer
- *Single polarity* clicks at 80 & 90dBnHL with rarefaction and condensation polarities
- Must use insert earphones
- No-sound run with sound tube disconnected or clamped to check for stimulus artifact

CM vs stimulus artifact





Courtesy of John Grose

What's Different?
ANSD vs "typical" SNHL

- Not possible to obtain threshold estimates with ABR
- Difficult to predict severity of disorder at time of diagnosis in infants using currently available tests
- If infant is identified early with ANSD it may be several months before child's detection thresholds can be adequately measured

Auditory Steady State Response (ASSR)

- ASSR responses can be obtained to high signal levels (>80dBHL) with ANSD but responses are elevated even in children who later show normal behavioral audiograms (Attias et al 2006, Rance et al 1998, Rance & Briggs, 2002)
- Therefore, ASSR cannot be used to determine thresholds in ANSD

Cortical Evoked Potentials (CAEPs)

- CAEPs not as reliant on timing as earlier evoked potentials and may be present when ABR is not
  - » Hood, 1998, Rapin and Gravel, 2003
- Unlike ABR must be completed in awake (but quiet) infants
  - » Cone Wesson and Wunderlich, 2003)
- CAEP may be useful tool for some difficult to test patients
  - » Pearce, W, Golding, M, and Dillon, H, Cortical Evoked Potentials in the Assessment of Auditory Neuropathy: Two Case Studies. Journal of the American Academy of Audiology, 2007, 18:380-39
- Further CAEP research needed with normal infants and infants with SNHL and ANSD

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Cortical Evoked Potentials (CAEP)

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Variable Presentations of ANSD Case Examples

Case #1:
Present CM and OAEs

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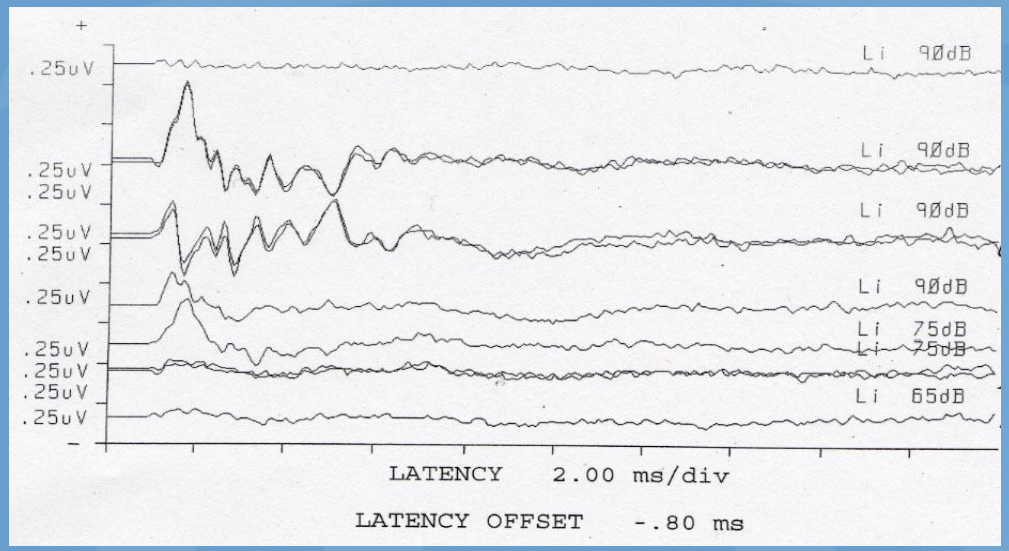
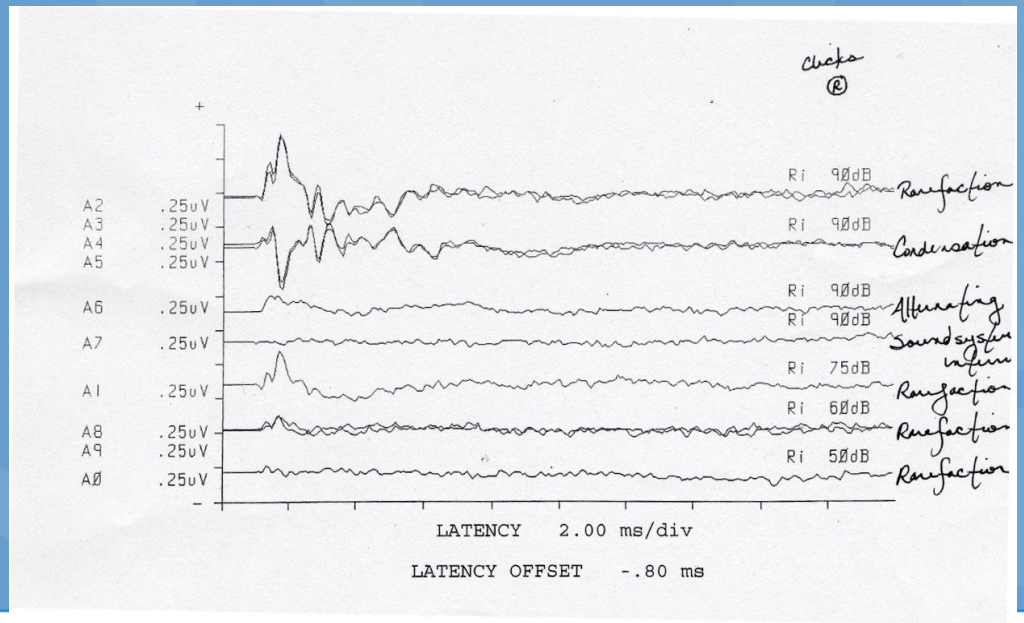
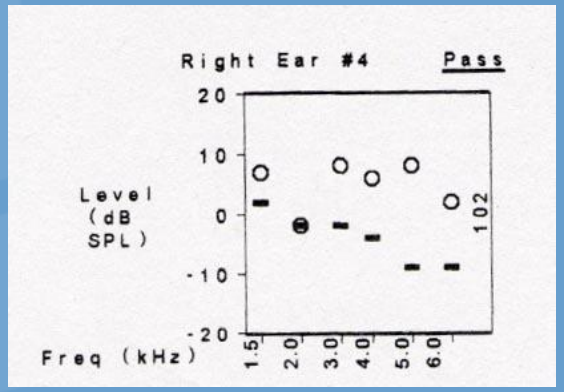
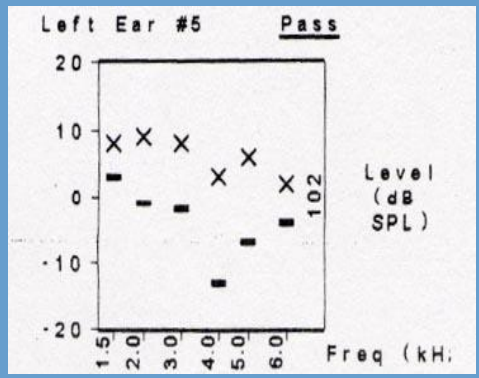




- 24 wk preemie, 940 grams
- NICU 4 months, ventilated
- ABR at 4 and 5 months of age abnormal
- ABR repeated at 18 months- no change





Case #1
Normal thresholds; Present CM and OAEs

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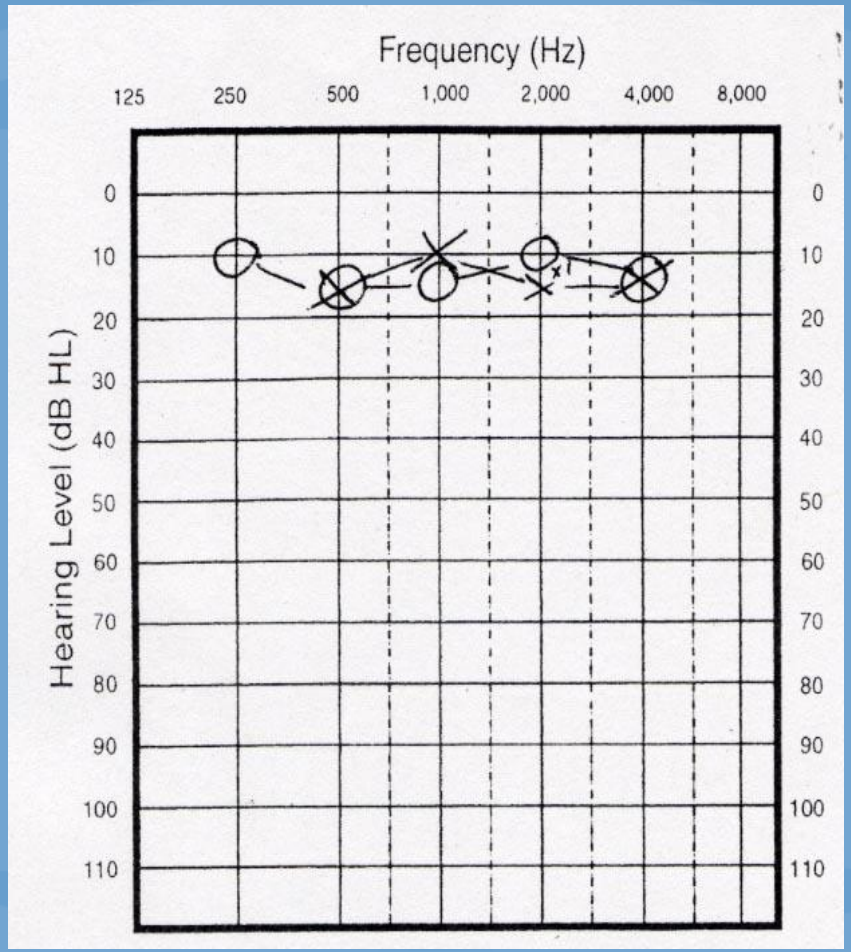
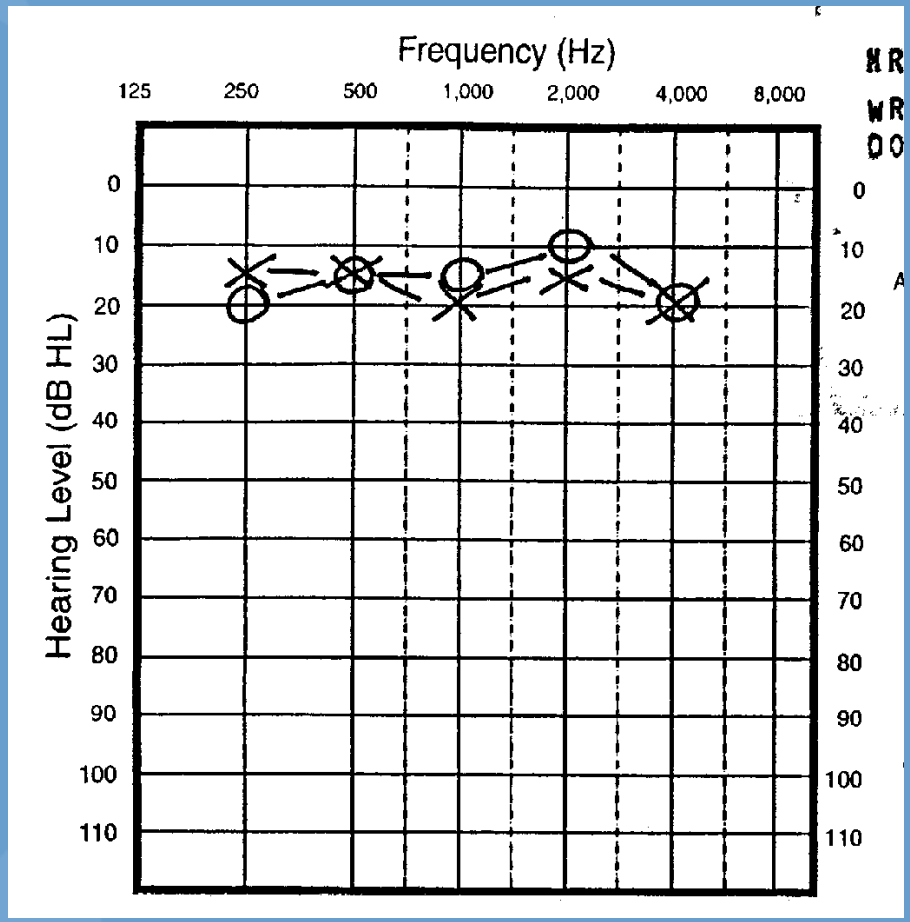


Audiogram at 14 months

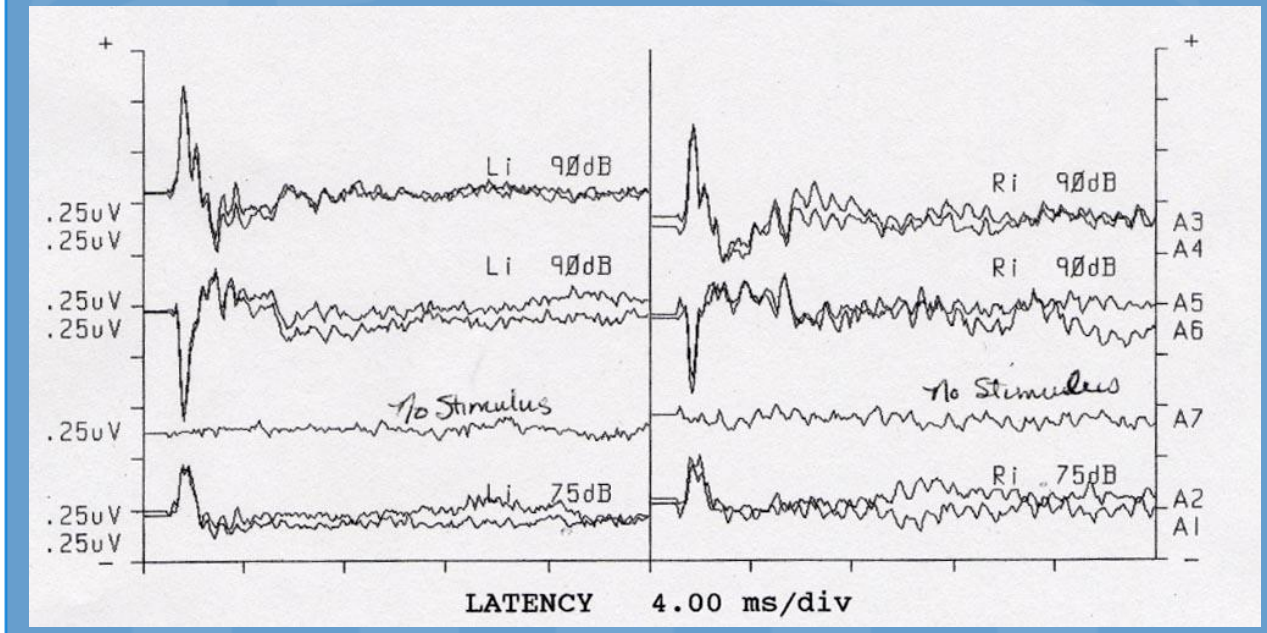


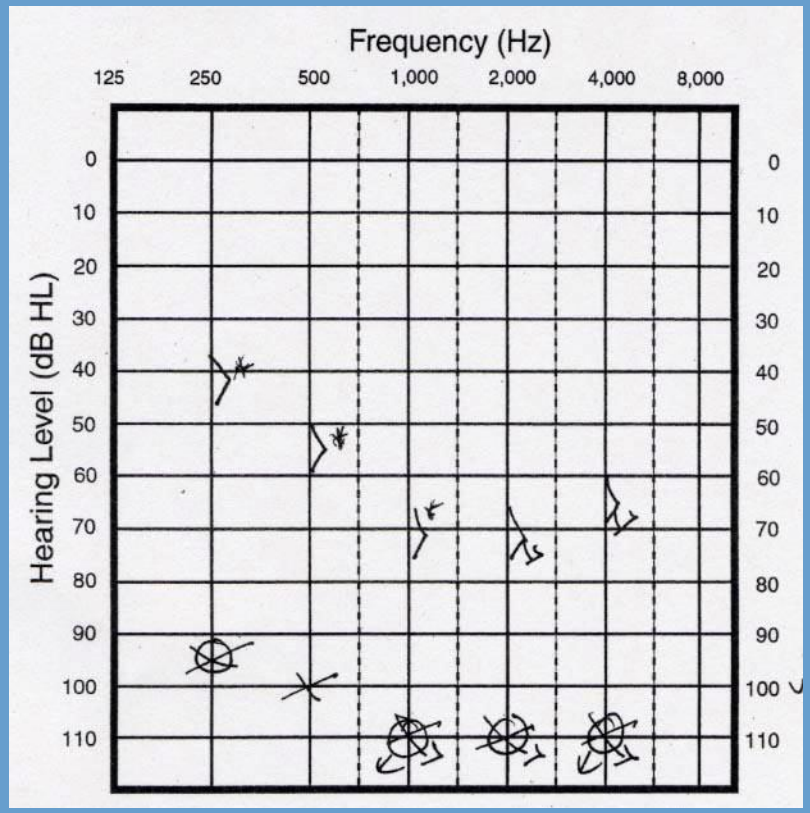
Audiogram at 18 months

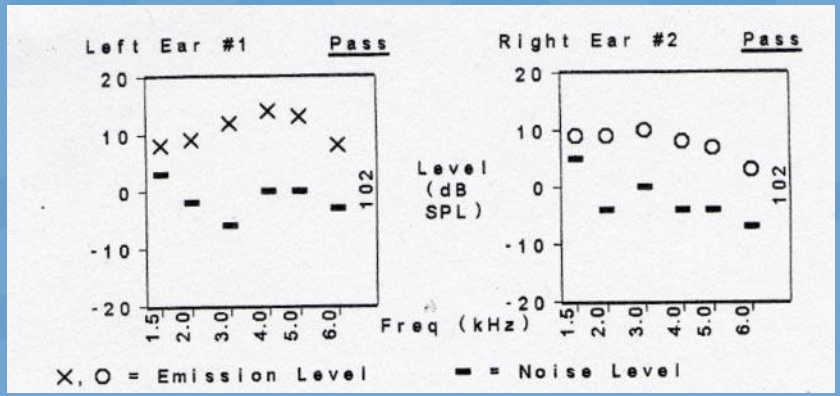
Case #1 (continued)

- Closed set testing at age 2 yrs-11 mos:
  - ESP 12/12 monosyllabic words correct for each ear at 50dBHL
- Open set testing at age 3 yrs-3 mos:
  - PBK words 64% and 72% at 55dBHL (?articulation errors)
- Child participated in 0-3 program
- Speaking in sentences with some speech production issues.
- Current Status: 3 yrs. 9 mos., hearing sensitivity still normal, attending a “Head Start” pre-school. Will continue to monitor hearing, speech and language

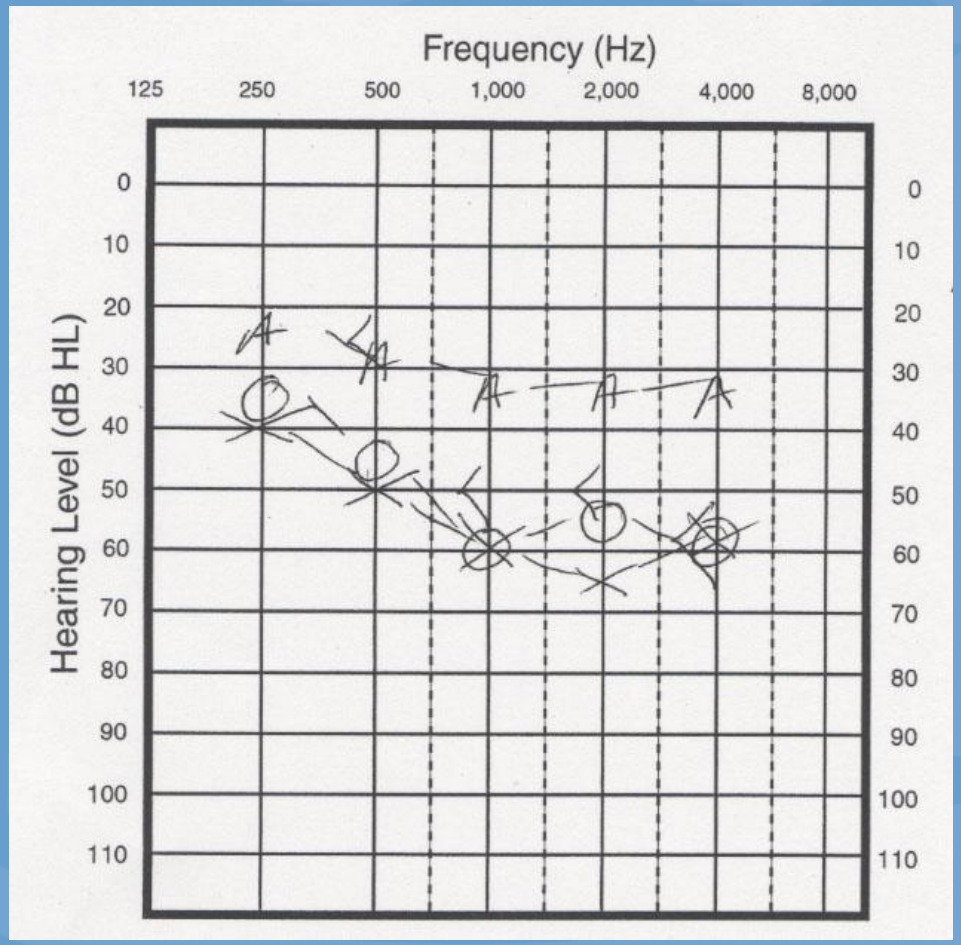
Case #2
Child with Profound Bilateral HL
Present CM and OAEs

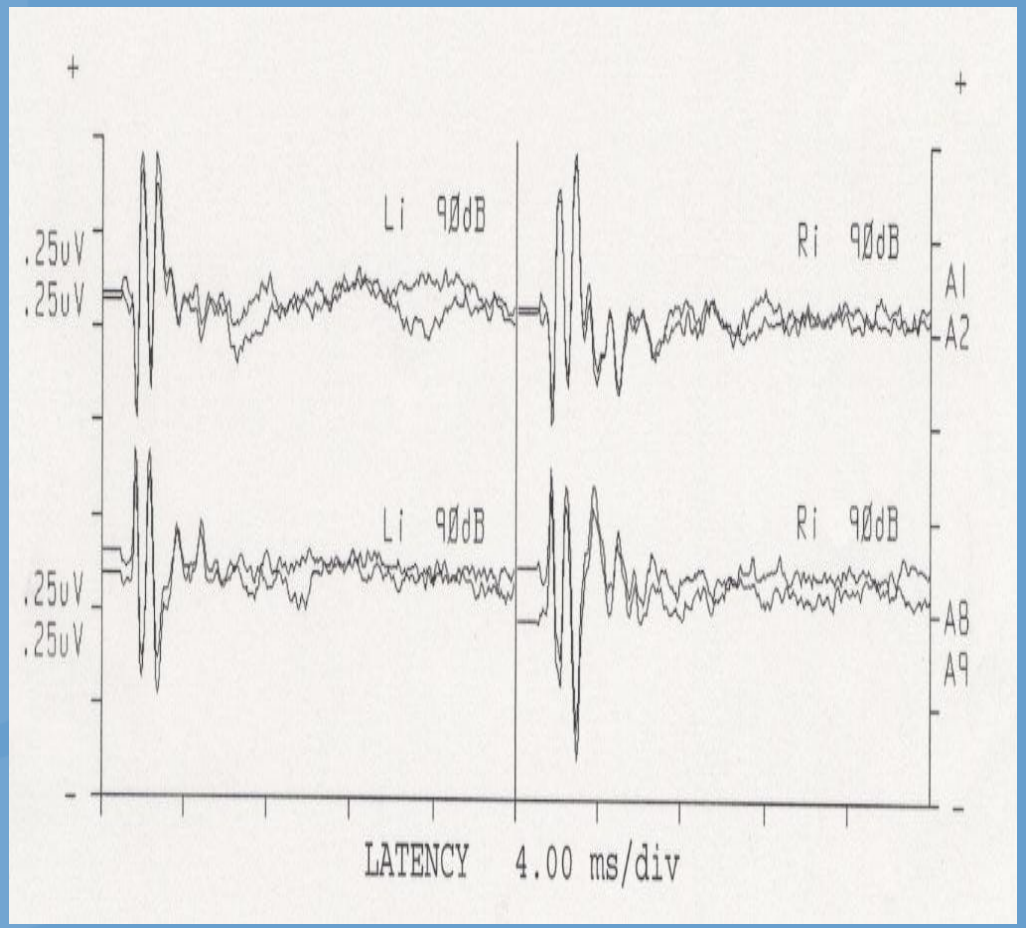






Case #3
Child with “moderate loss”
CM present, absent OAEs





Case #4: Large CM; present OAEs; distal waveforms present

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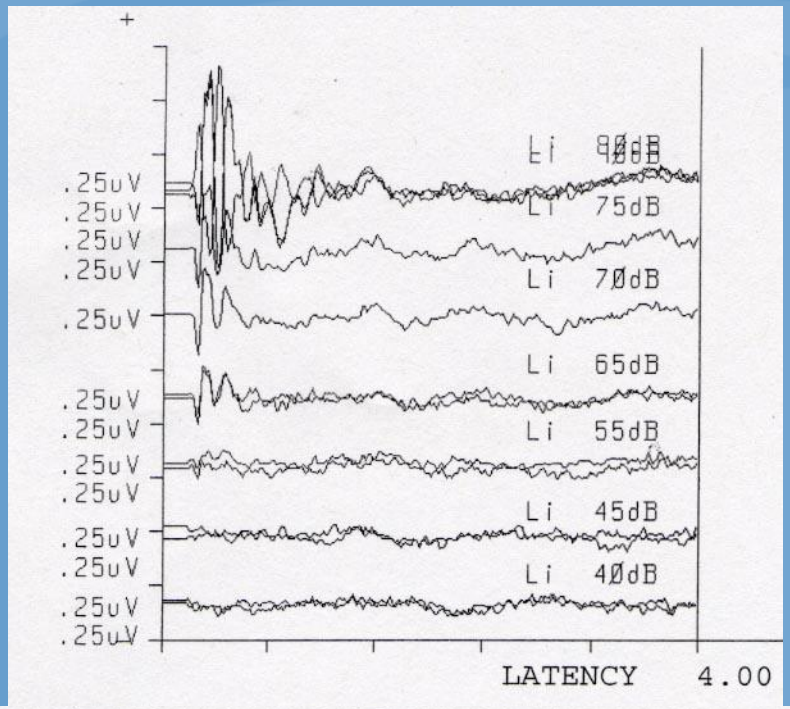
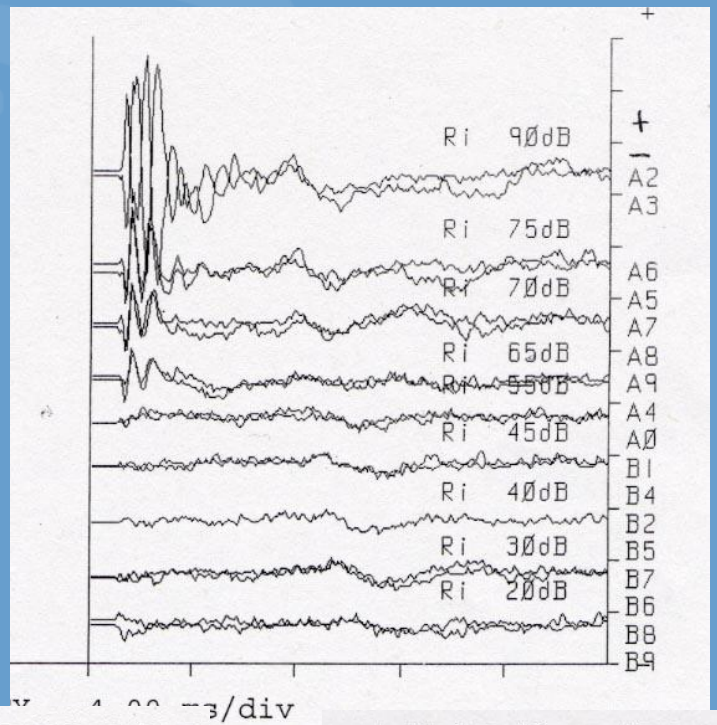
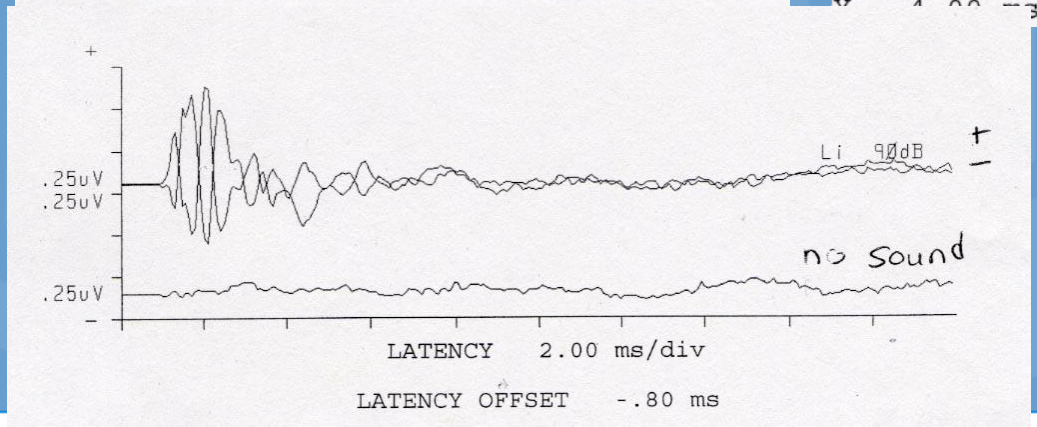
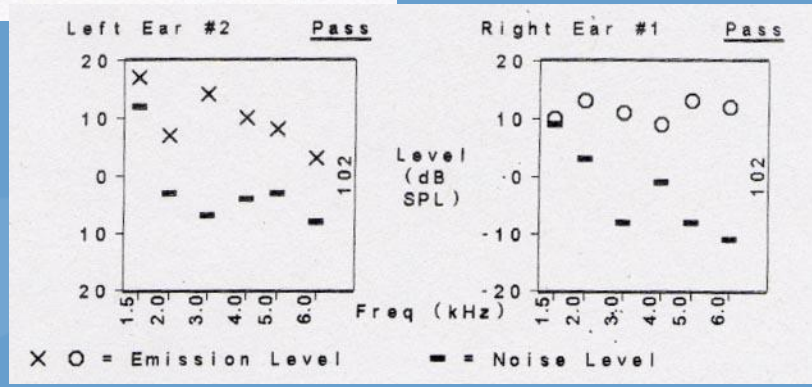


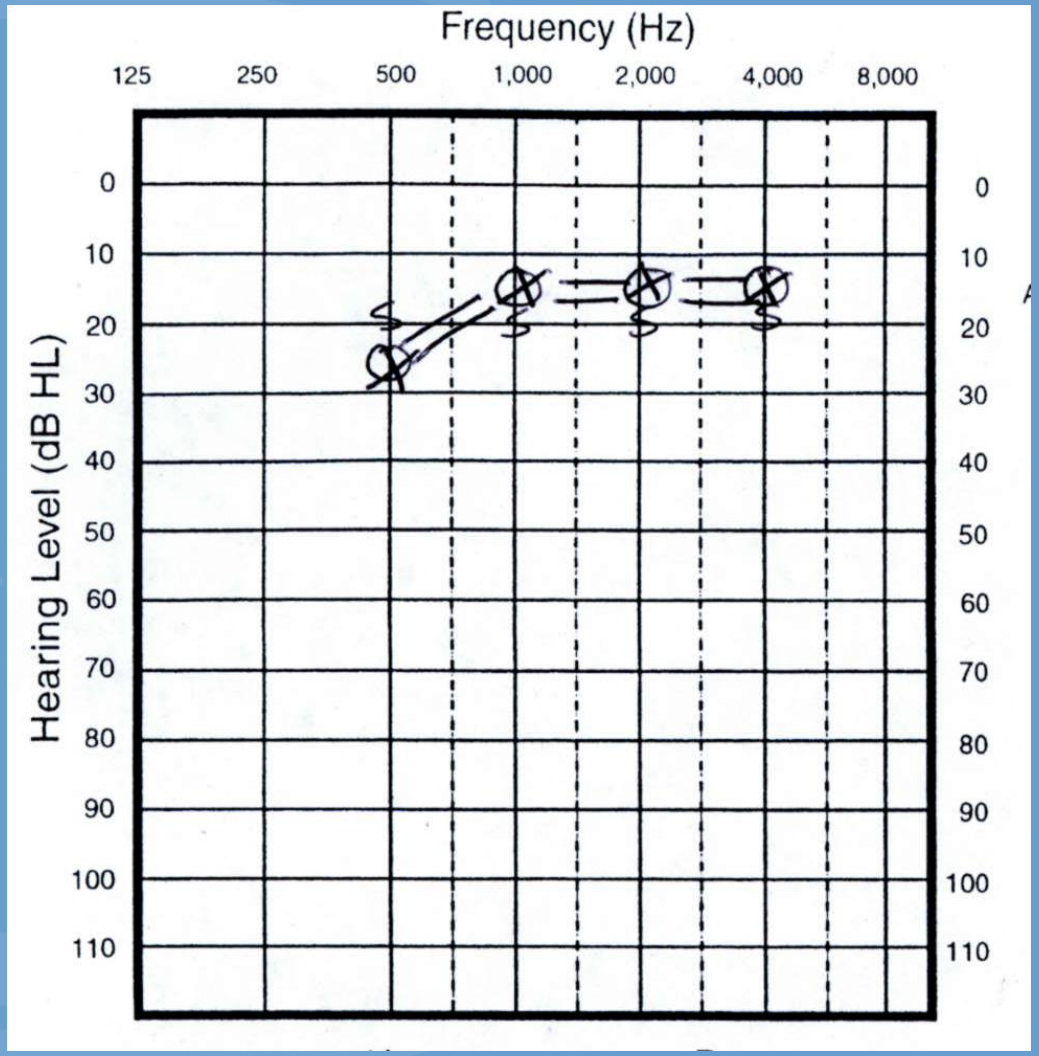






Caution needed when interpreting ABRs that show abnormal waveform morphology at high intensity levels

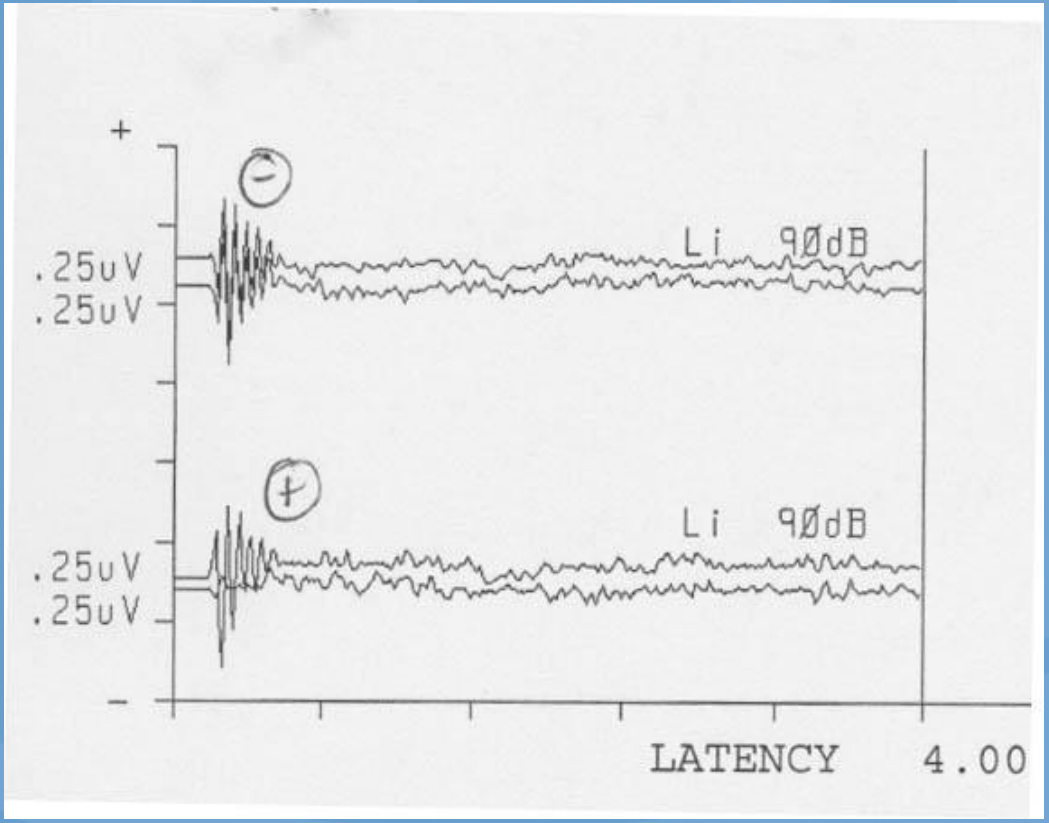
Case #4 (continued)
VRA with insert earphones
Age14 months

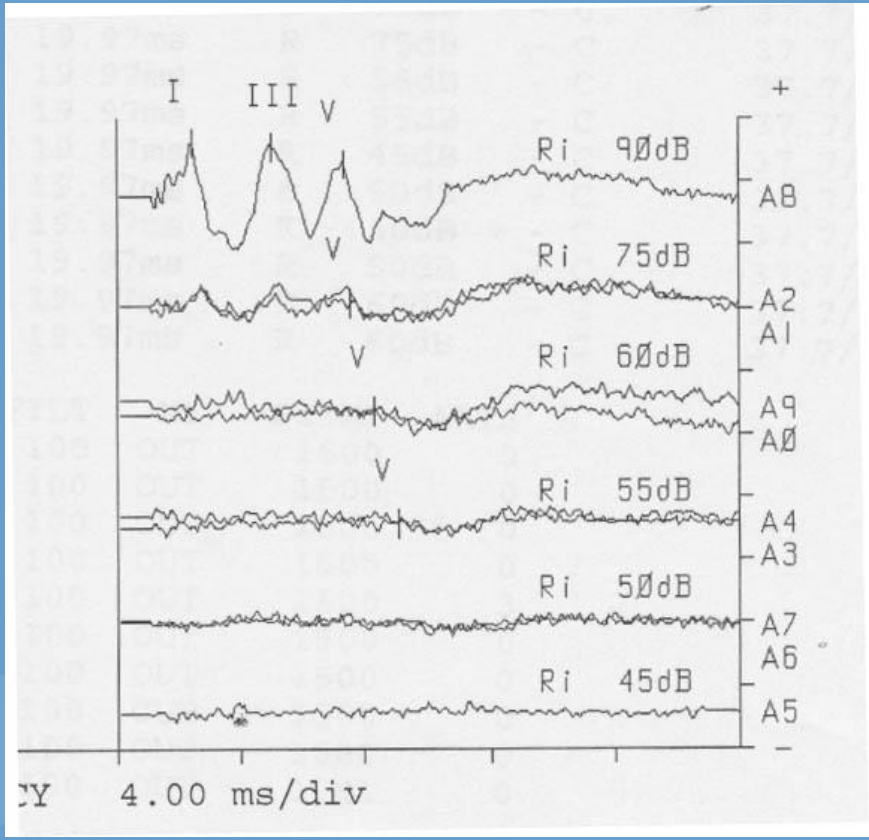


Cochlear Nerve Deficiency (CND)

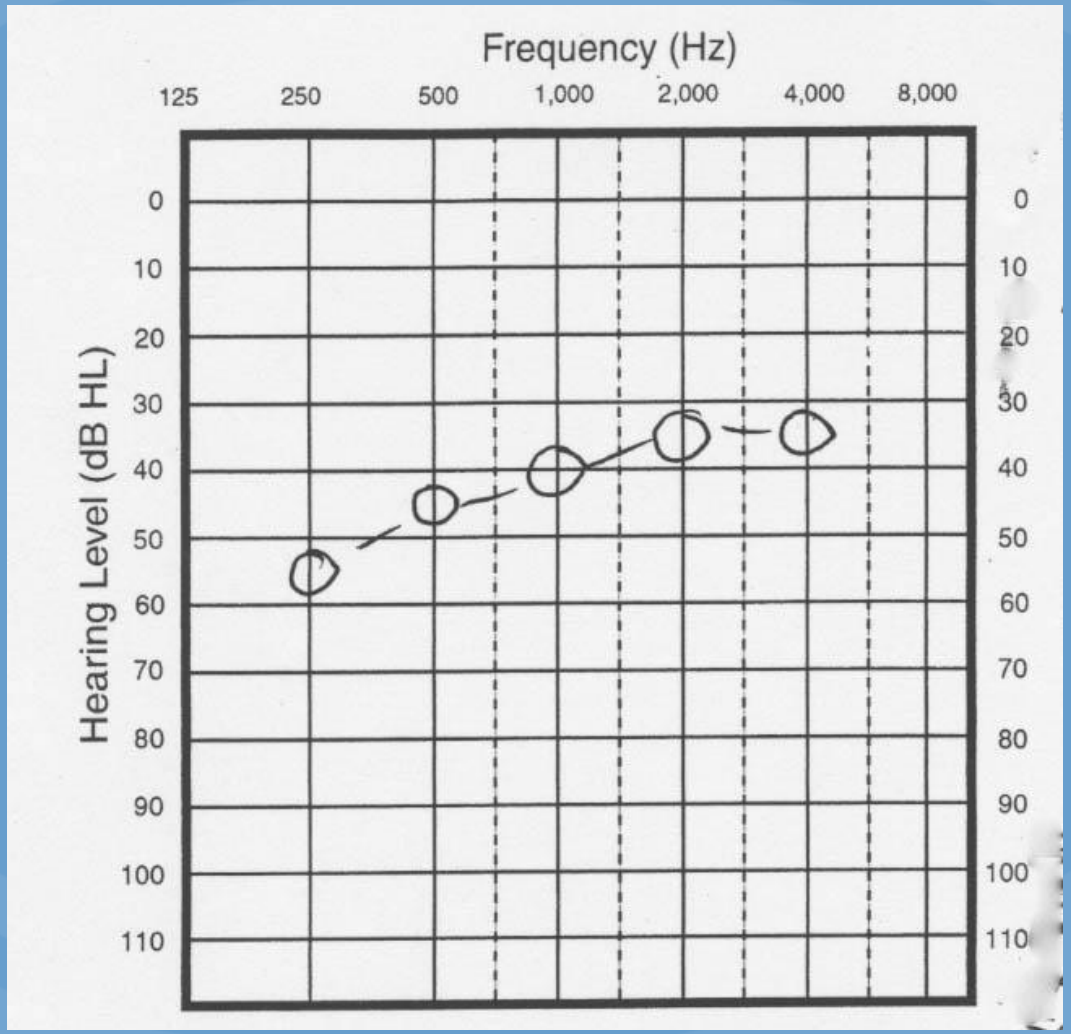
- Small or absent VIII nerve
- Must perform MRI to determine if VIII nerve is small or absent
- Examples of CND showing ANSD pattern on next several slides

Case #5
Child with CHARGE
Right: Moderate loss
Left: Absent cochlear nerve





Case #5 (continued)
Audiogram Age 21 months



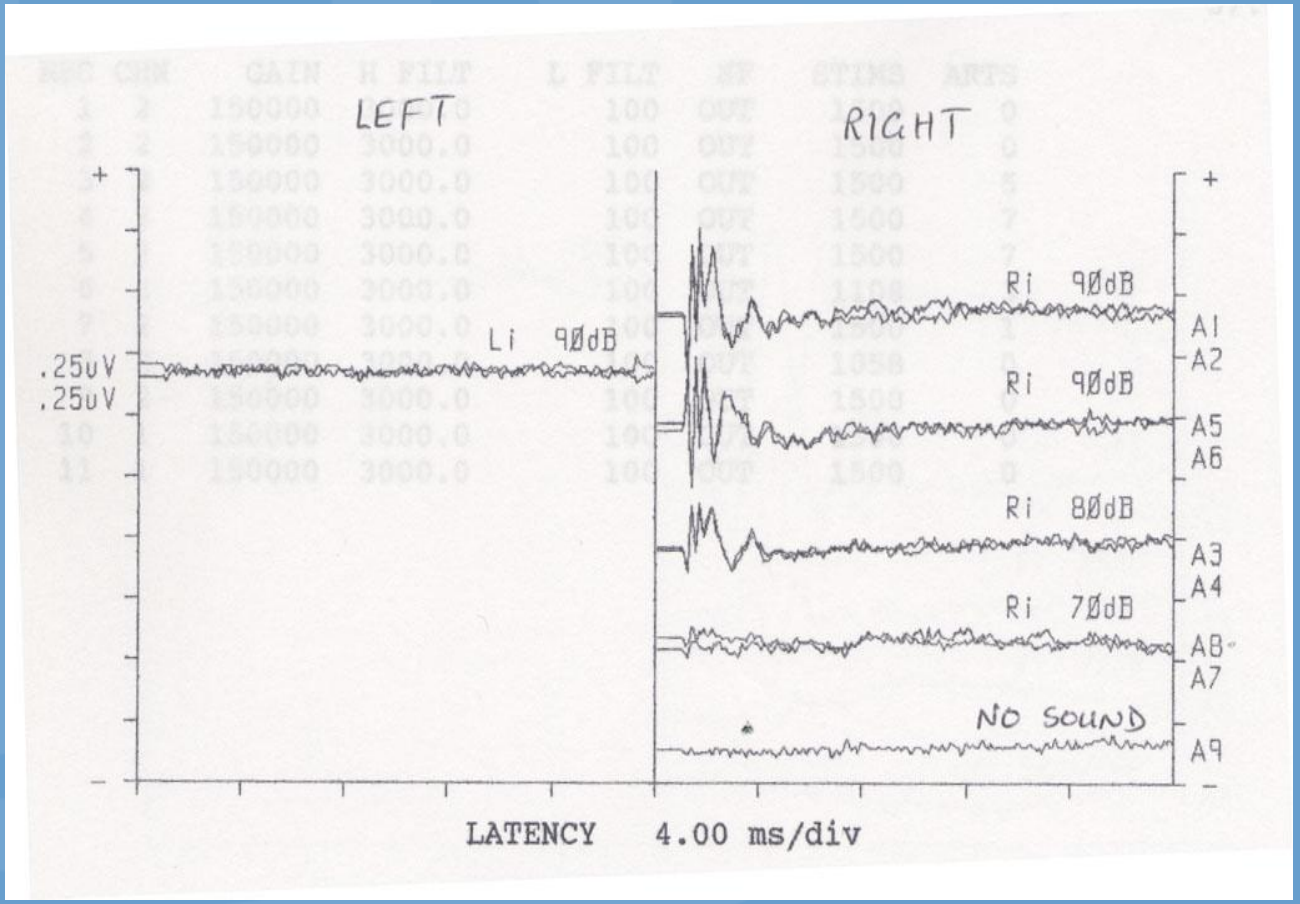
Right ear:

- moderate mixed loss

Left ear:

- Profound SNHL

Case # 6
Bilateral deafness
No VIIIth nerve on right



Case #6
Child with bilateral deafness
No VIIIth nerve on right

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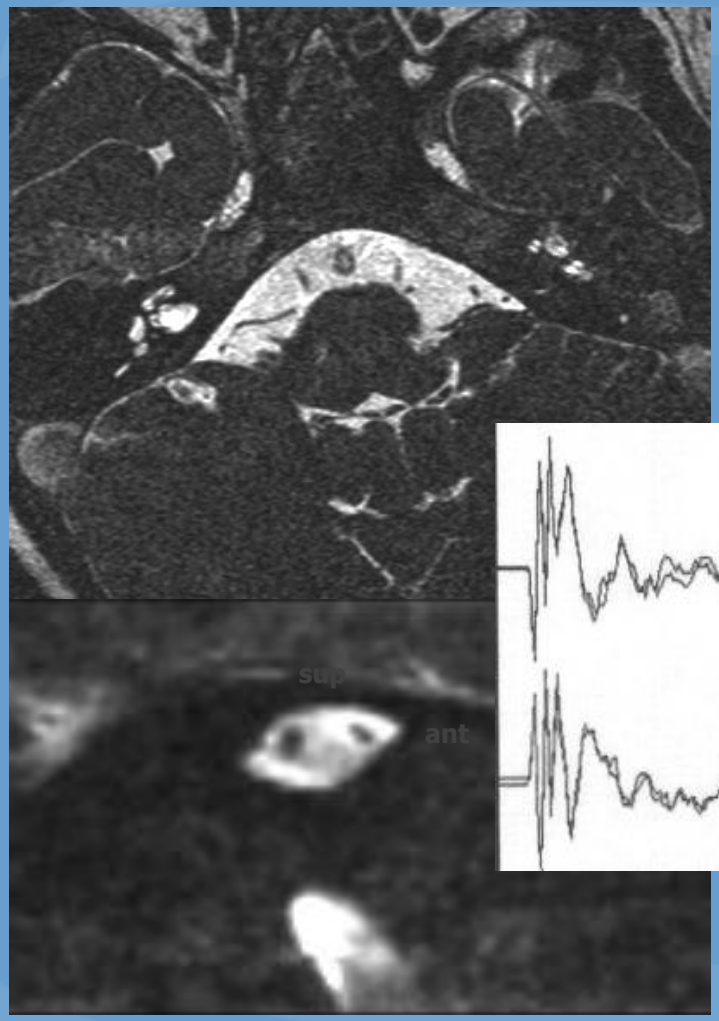

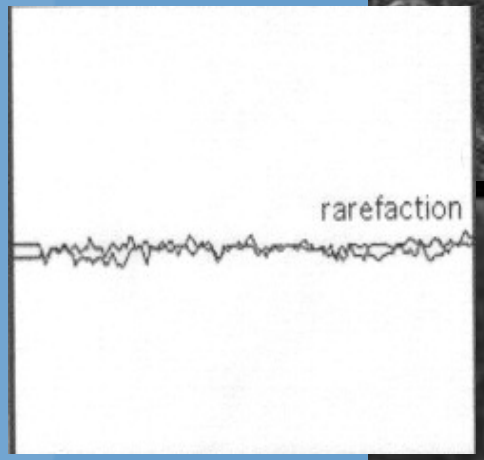

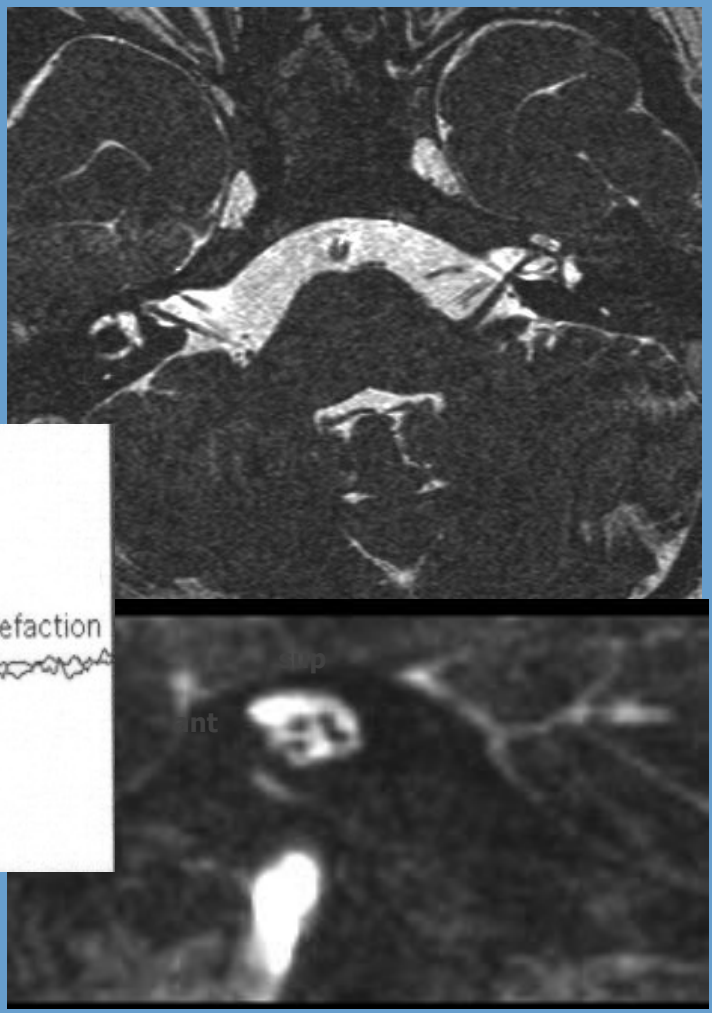

Right Ear

Left Ear

Case #7

- Child presented to clinic with left profound unilateral hearing loss at 4 years of age.
- Born at full term
- Passed newborn hearing screen using OAEs
- No family history of hearing loss

Case # 7

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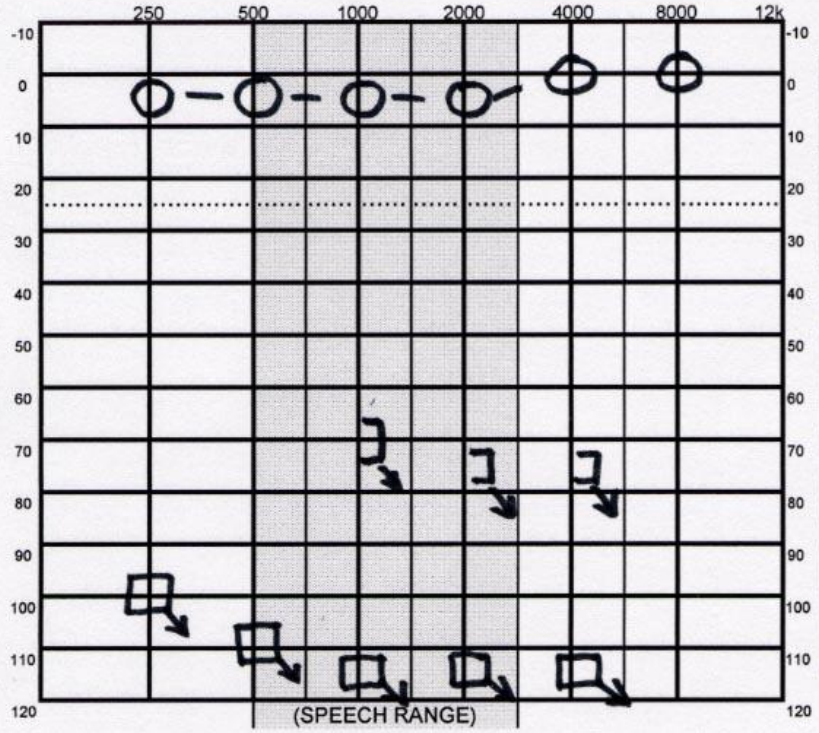
PURE TONE AUDIOMETRY (RE: ANSI 1969)



LEGEND	RIGHT (RED)	LEFT (BLUE)
AIR	O	X
AIR (OPP. EAR MASKED)	Δ	□
BONE	<	>
BONE (OPP. EAR MASKED)	[	]
NO RESPONSE	↓	↓
SOUND FIELD	S	
AIDED	R	L

TEST	
STANDARD	
PLAY	
COR/VRA	
BOA	

SITE	
BOOTH	
OTHER	

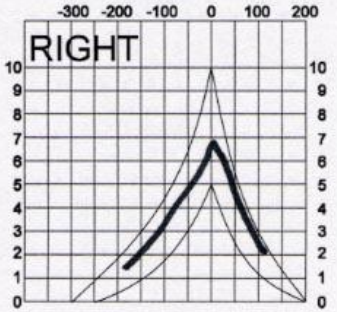
RELIABILITY	
EXCELLENT	
GOOD	
FAIR	
POOR	

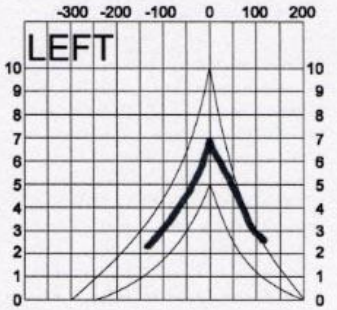
TYMPANOMETRY

CONTRA (Phone Ear)	.5k Hz	1k Hz	2k Hz	4k Hz
RIGHT (AD)				
LEFT (AS)				

IPSI (Probe Ear)				
RIGHT (AD)	85	85	85	
LEFT (AS)	NR	NR	NR	

	REFLEX DECAY @ 1k Hz	STATIC COMPLIANCE
RIGHT (AD)		
LEFT (AS)		





MODE	
STANDARD EARPHONES	
INSERT EARPHONES	
SOUND FIELD TESTING	
ASSISTED EVALUATION	

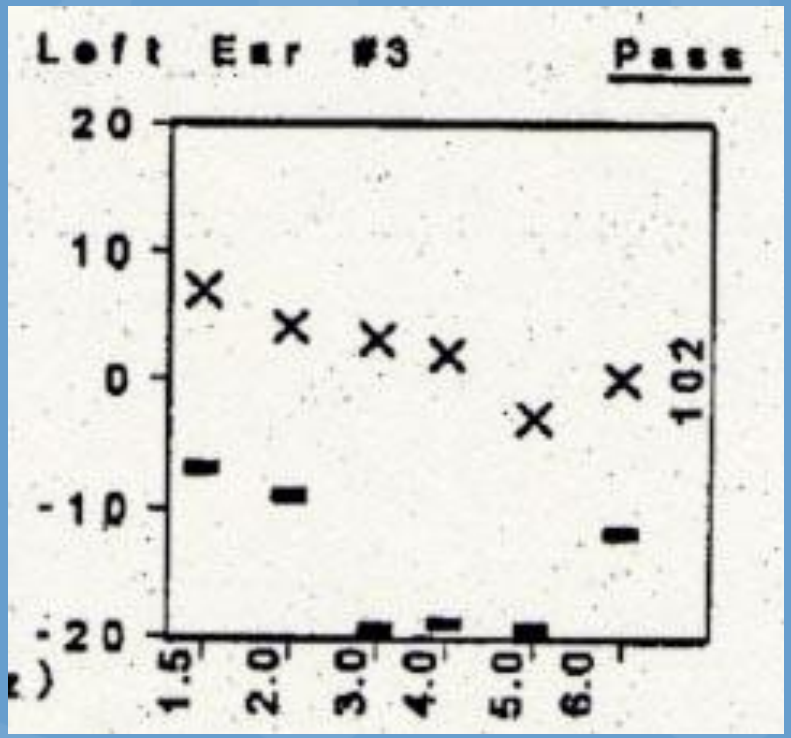
TYMPANOMETER: _____

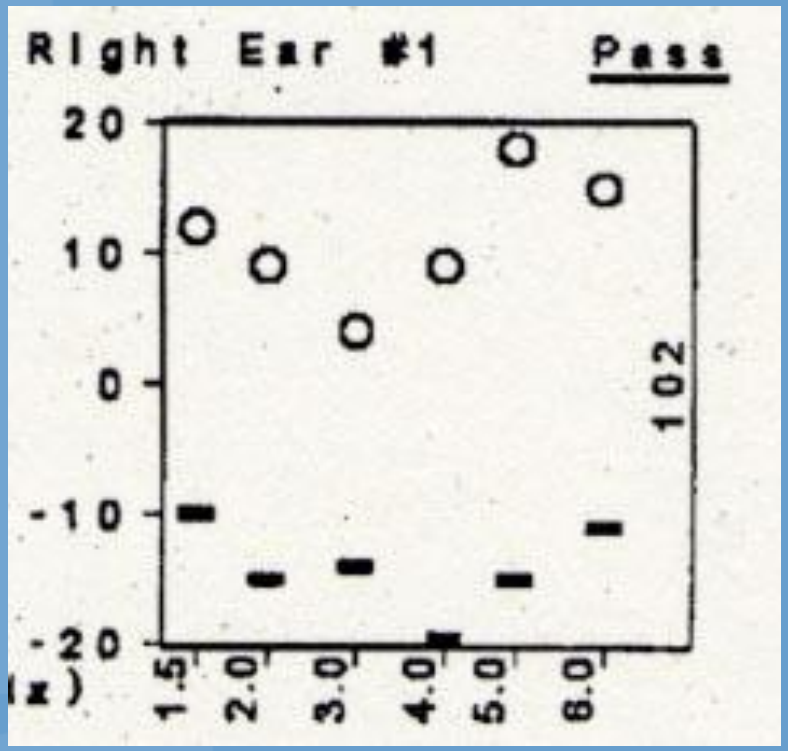
SPEECH AUDIOMETRY

	PTA	SRT/SAT	SPEECH RECOGNITION		SPEECH RECOGNITION		MCL	UCL
RIGHT (AD)		O	% 100	dB HL 50	%	dB HL		
masking								
LEFT (AS)		NR	%	dB HL CNT	%	dB HL		
masking								

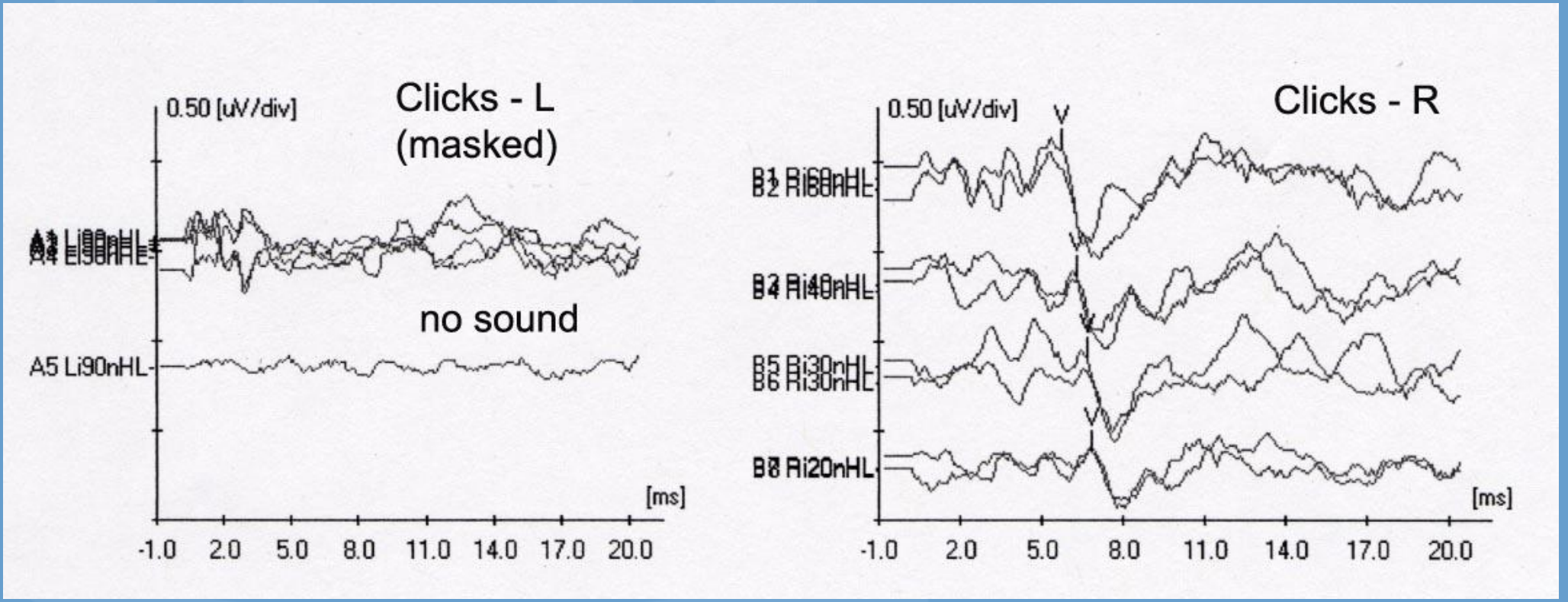
HISTORY: _____

Case #7 OAEs


Left Ear #3
Pass
20
10
0
-10
-20
1.5
2.0
3.0
4.0
5.0
6.0
102


Right Ear #1
Pass
20
10
0
-10
-20
1.5
2.0
3.0
4.0
5.0
6.0
102

Case #7 ABR


0.50 [uV/div]
Clicks - L
(masked)
no sound
A5 Li90nHL
[ms]
-1.0
2.0
5.0
8.0
11.0
14.0
17.0
20.0
0.50 [uV/div]
Clicks - R
V
B5 Ri30nHL
B6 Ri30nHL
B8 Ri20nHL
[ms]
-1.0
2.0
5.0
8.0
11.0
14.0
17.0
20.0

Case #7

- Results of MRI:
  - Cochlear nerve on left smaller than the right
  - Question of left cochlear nerve hypoplasia
- At age 4 child has above average speech and language development
- Will manage as we do other cases with profound unilateral hearing loss.

UNC Children with Characteristics of ANSD and Available MRI (2009)
N=140

- 35/140 (25%) Cochlear Nerve Deficiency (CND) (absent or small cochlear nerve) in one or both ears
  - Unilateral (n=24; 69%)
  - Bilateral (n=11; 31%)

Buchman, C, Roush P, Teagle H, Brown C, Zdanski C, Grose J. Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear Hear*. 2006 Aug;27(4):399-408

Take Home

- Absent 8th Nerve
  - not uncommon
  - can result in auditory neuropathy phenotype
  - commonly has normal internal auditory canal (IAC) morphology
  - commonly has normal labyrinth
- Need MRI instead of CT in all kids
  - with profound hearing loss
  - with auditory neuropathy phenotype
  - Audiological, educational, and medical recommendations will be influenced by these findings e.g. CI or HA candidacy

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Recommended Audiologic Test Battery

- Auditory Brainstem Response (ABR)
- Acoustic Immittance Measures
  - Tympanometry
  - Acoustic Reflex Testing
- Otoacoustic Emissions Testing
- Behavioral Audiometry
  - VRA, BOA, play audiometry
- Speech Recognition Testing

VRA Six Month Old

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Speech Perception Test Battery

- IT-MAIS or MAIS (Parent Questionnaire)
- (Zimmerman-Phillips, et al., 2000; Robbins, et al., 1991)
- Early Speech Perception Test battery (ESP) (Moog and Geers, 1990)
  - Standard
  - Low Verbal
- MLNT/LNT words and phonemes (Kirk, et al, 1995)
- PB-K words and phonemes (Haskins, 1949)
- HINT sentences in quiet and noise conditions

(Use recorded speech materials when possible)

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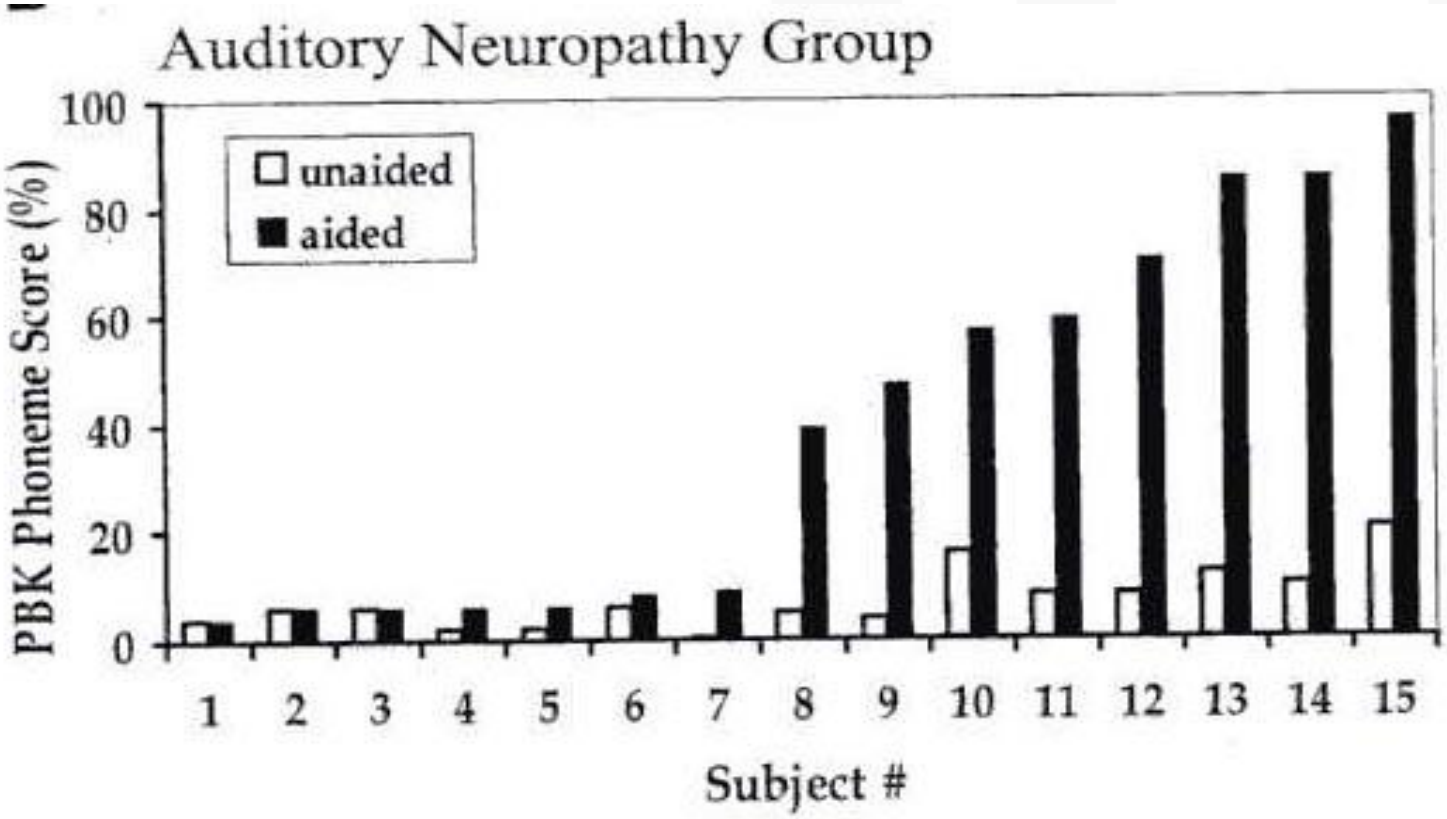
Weighing the Evidence: Hearing Aids, FM and Cochlear Implants

What does existing evidence tell us about clinical management?

Evidence regarding amplification in children with ANSD

- Rance et al 2002
  - Comparison of unaided and aided speech perception abilities in group of 15 children with AN/AD compared to group of children with typical SNHL
  - Results show ~50% of group showed significant open-set speech improvements; ~50% of group showed no open-set speech perception ability.

Hearing Aids in Children with AN/AD: 50% Benefit from Hearing Aids



Rance et al Ear and Hearing 2002

Speech Perception in Noise (Rance et al 2007)

Open and closed-set speech perception assessed in 12 school-age children with ANSD and compared to 20 subjects with SNHL and 25 children with normal hearing.

Results:

- Closed set speech understanding was poorer in presence of noise in the SNHL group than in the normal subjects
- Closed set perception in noise not significantly different for ANSD children and SNHL counterparts but was toward poorer performance in ANSD group
- Open set speech perception was also similar in both the ANSD group and the SNHL groups with trend toward poorer performance in ANSD group

Speech Perception in Noise (Rance et al 2007)

Conclusions:

- Listening in presence of background noise was more difficult for children with ANSD than for children with normal hearing
- Results not consistent across subjects
- Some children with ANSD demonstrated reasonable speech perception abilities at low signal to noise ratios

Summary

- Many children with ANSD have disproportionately poor speech recognition abilities for degree of hearing loss while others perform like their peers with “typical” sensorineural hearing loss.
- Some children with ANSD have greater difficulty in noise while others do not
- Some children with ANSD appear to benefit from amplification and auditory based intervention while others require CI and/or visual supplementation

Summary

- Evidence regarding outcomes from amplification is limited
- Few peer reviewed studies have been published
- Existing literature is based on small number of children
- Many anecdotal reports
- Only a few published studies document use of a prescription-based fitting strategy that ensures audibility of speech signals

Factors that may affect outcomes

For all children benefit from a particular technology will depend on several factors including

- Age at diagnosis and treatment
- Appropriateness of device fitting
- Consistency of use
- Quality of intervention
- Extent of family involvement
- Cognitive abilities of child
- Presence of other medical conditions

Hearing Aid Fitting Protocol for Infants

- Assessment
  - » Behavioral thresholds ***estimated*** based on electrophysiologic measures
- Prescriptive formula selected
  - » e.g. DSL, NAL-NL1
- Program Hearing instrument
  - » Manufacturer's software used
- Verification of Fitting
  - » Alternative to traditional probe microphone measures for use with infants: Real Ear to Coupler Difference measurement (RECD)

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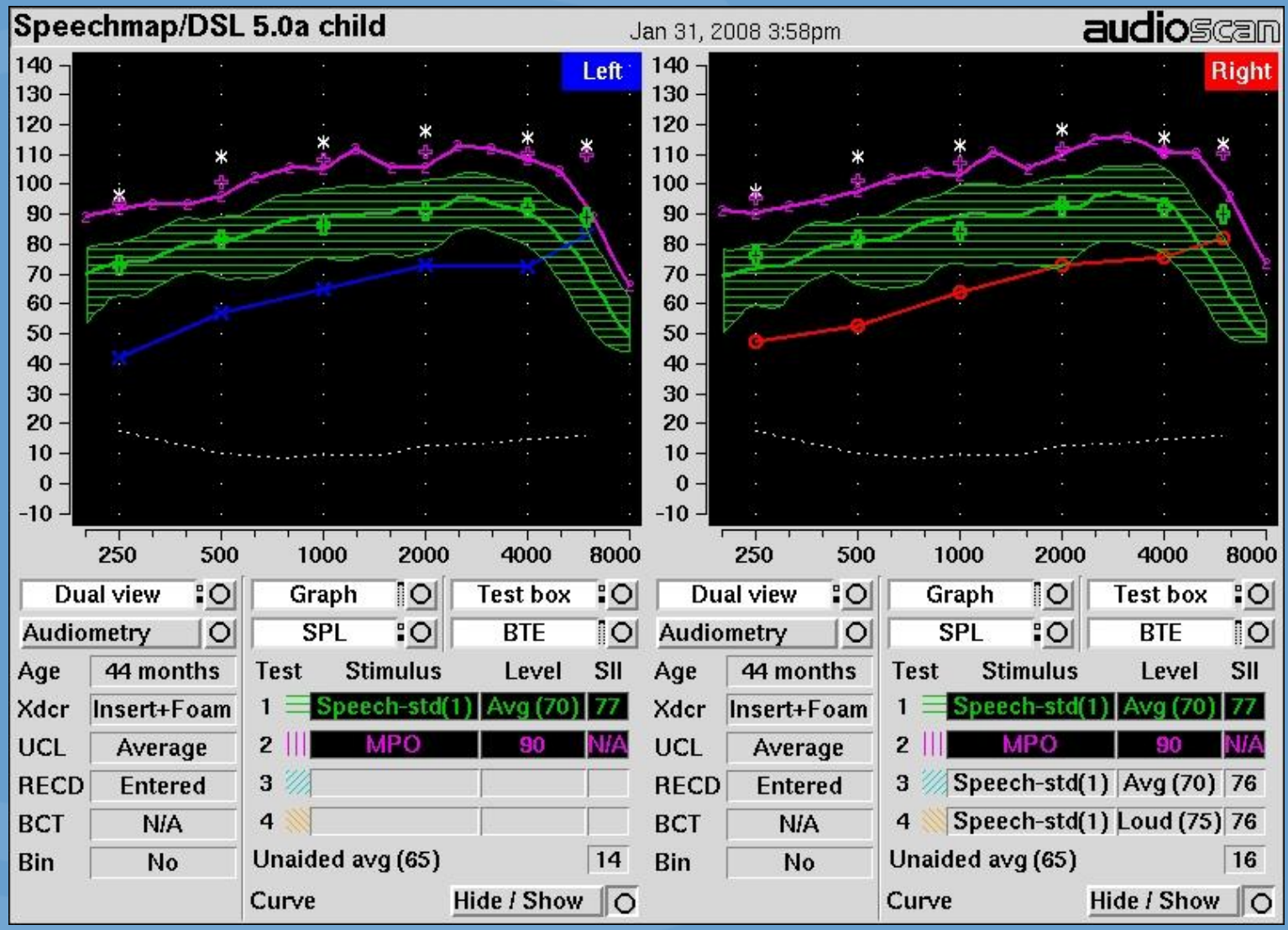
Verifying Audibility of Speech Spectrum



Hearing Aid Fitting in Infants with ANSD: What's different?

- Behavioral thresholds cannot be predicted from ABR or ASSR
- Determination of hearing thresholds is delayed until infant developmentally able to perform task (6-9 months of age for most infants)
- Many children with ANSD are at risk for cognitive impairments resulting in a lengthier and more complicated process of threshold determination
- This results in delays in hearing aid fitting and greater amount of time without adequate audibility of speech signal

Hearing Aid Fitting in Infants with ANSD: What's Different?

- Behavioral pure tone audiogram in ANSD does not have same prognostic value in prediction of aided benefit as with typical SNHL
- Considerable variability in presentation of ANSD even with similar initial test results
  - Difficult to provide prognosis to families of newly diagnosed infants
- Speech recognition testing not possible on children under two years of age making determination of benefit from amplification difficult
  - Also true for young children with severe sensory loss

University of North Carolina-Chapel Hill: ANSD Study

- 165 infants and children with ANSD at UNC
  - 61% males, 39% females
- 10 year prospective study underway to determine characteristics of children with ANSD and benefit from assistive devices
- All patients evaluated by pediatric audiologist and otologist
- Neurology, genetics, ophthalmology consults recommended
- Many in study are very young and we are just beginning to be able to evaluate outcomes

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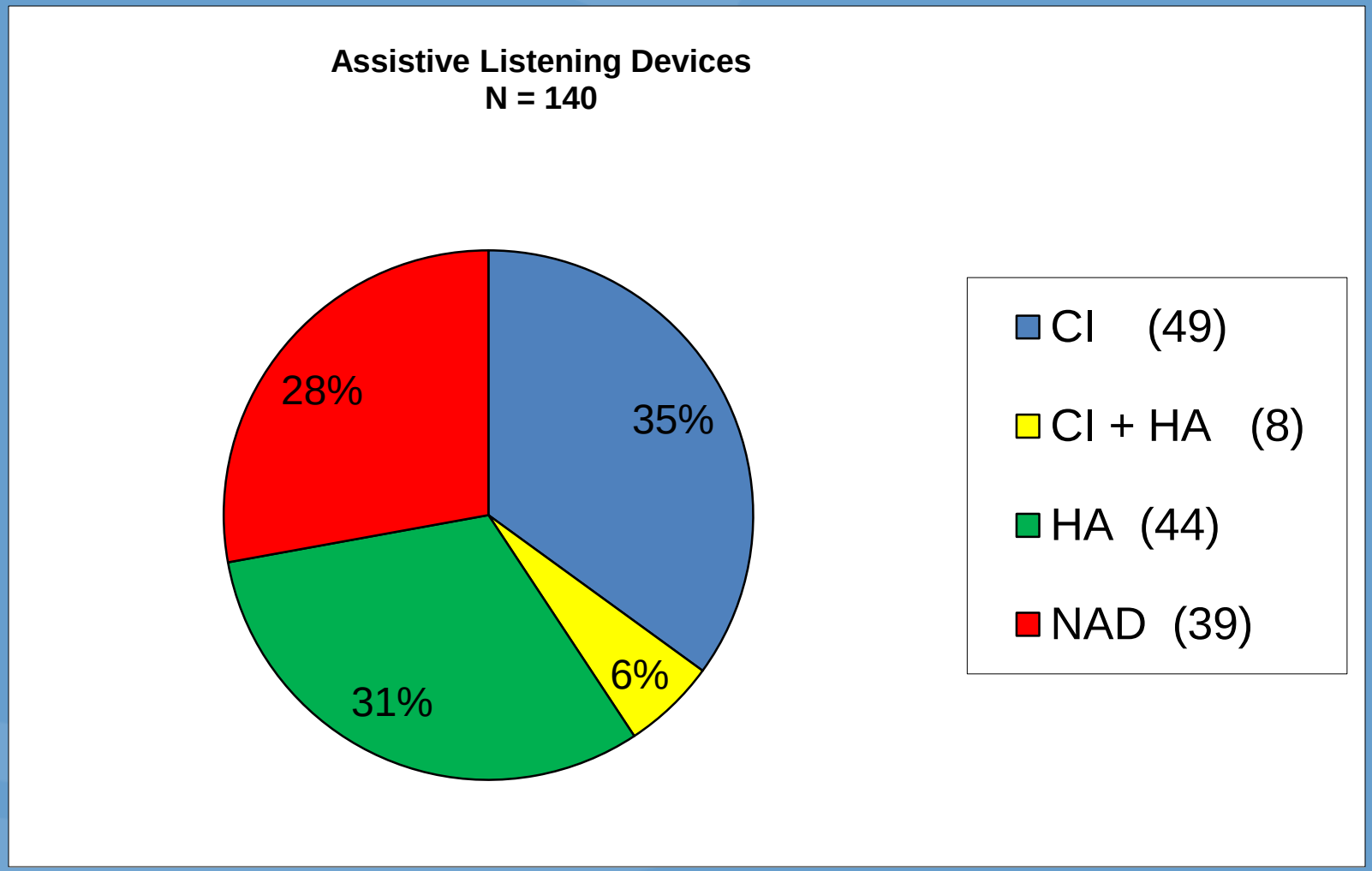
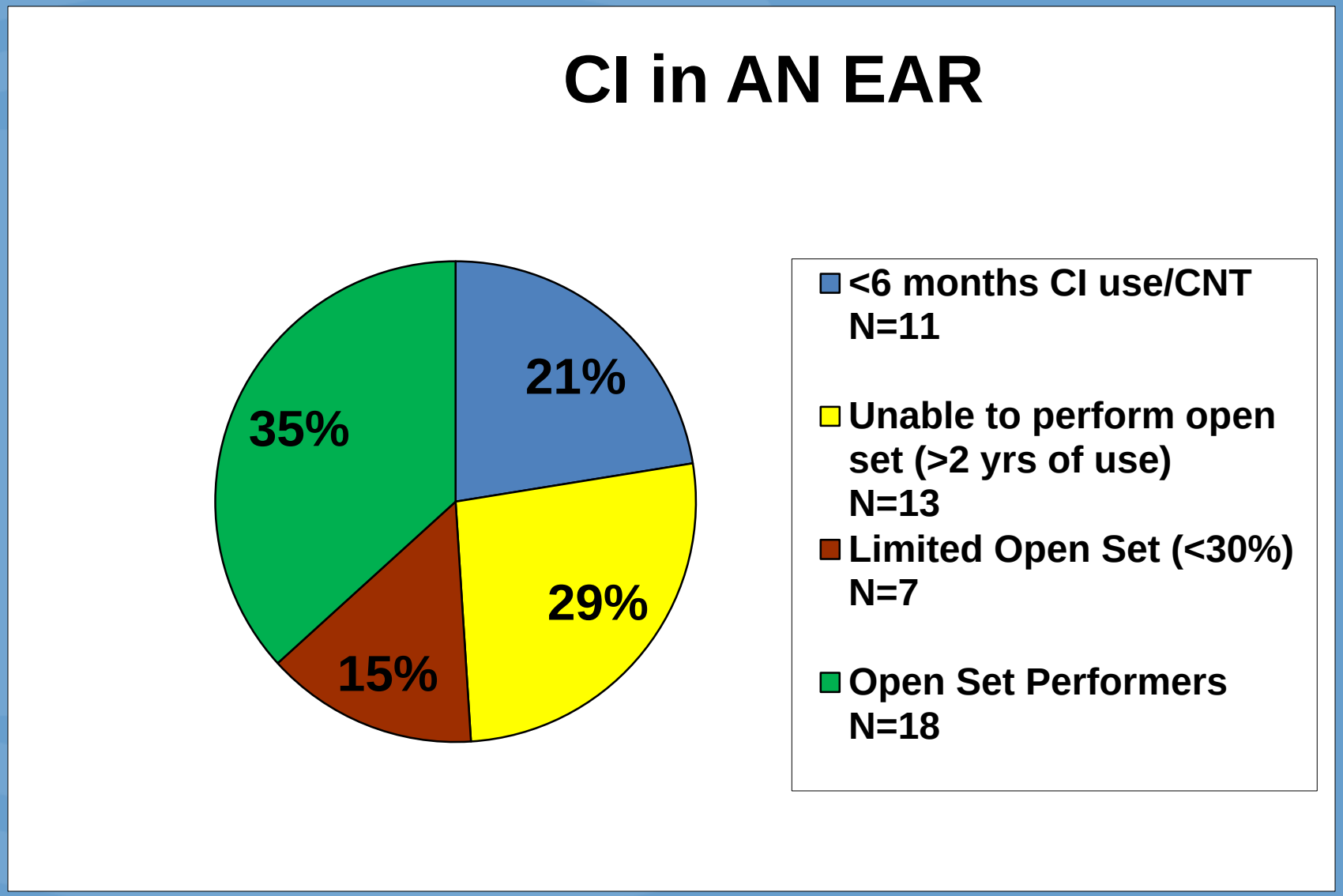
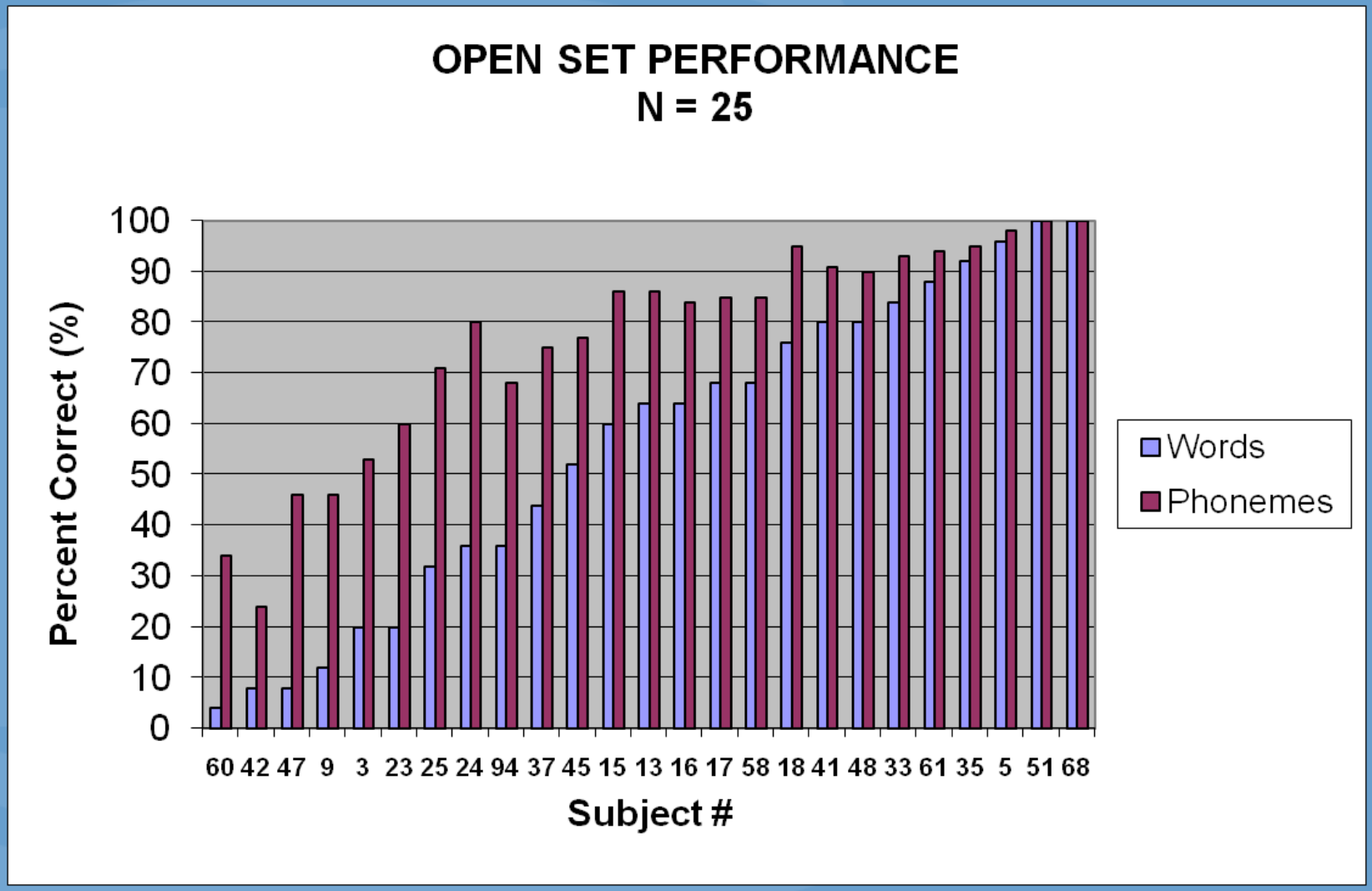
UNC ANSD Children
Assistive Listening Device
N=140



UNC AN Children with CI N=49



UNC AN Children with CI
Open Set Performance N=25



CI Criteria-Children

- Advanced Bionics
  - Children-age 4 or less:
  - Failure to reach auditory milestones or <20% on MLNT at 70 dB SPL
  - Children > age 4: <12% on PBK words or < 30% on open set sentences at 70 dB SPL
- Cochlear Corporation
  - Children-12 months though 17 years
  - Bilateral profound SNHL in children 12 months to 2 years
  - Bilateral severe to profound SNHL in children 2 years and older
  - 30% or less on open set MLNT or LNT
  - 3-month trial with HA if not previously amplified
- Med El
  - Children- 12 months to 17:11 (17 years, 11 months)
  - Profound SNHL specified as 90 at 1K Hz
  - Lack of progress in auditory skills with habilitation and amplification provided for at least 3 months
  - Less than 20% on MLNT or LNT
  - 3-6 month HA trial without previous fitting; waived if ossification

ANSD Guidelines (Como 2008): Recommended Amplification Strategies

- Amplification should be fitted as soon as ear specific elevated pure-tone and speech detection thresholds are demonstrated by conditioned test procedures
- Hearing aid fitting strategies…should follow established guidelines for the fitting of amplification in infants and toddlers
  - e.g. American Academy of Audiology Pediatric Amplification Protocol, 2003
- Since Improvement in auditory function has been reported in some cases, careful monitoring needed to adjust and modify amplification as needed.

ANSD Guidelines (Como 2008): Recommended Amplification Strategies

- Strategies to improve signal-to-noise ratio for children with ANSD should, theoretically improve speech recognition and language learning (Hood et al 2003)
- Trial use of an FM system, especially in structured and spontaneous language-learning activities should be considered.

ANSD Guidelines (Como 2008): Special Considerations for Cochlear Implantation

- Families should be informed that spontaneous improvement in has been reported up to two years. CI should not be considered until test results are stable and demonstrate unequivocal evidence of permanent ANSD. Deferring decision to two years of age may be appropriate.
- Evidence of auditory nerve sufficiency should be obtained prior to surgery using appropriate imaging technology (Buchman et al., 2006)
- Children with ANSD who do not demonstrate good progress in speech recognition and language development should be considered candidates for cochlear implantation *regardless of audiometric thresholds*.

Counseling Families

Counseling in ANSD

- Information provided to families should be based on current evidence and not “hearsay”
- Important that we are confident in our knowledge of disorder or refer to those who are
- While it is more difficult than with non-AN hearing loss to provide “prognosis” for family, there is a lot of useful information that needs to be provided to families at time of diagnosis.
- Families need to be reassured that help is available and be informed of a timeline for the first year following diagnosis

Counseling in ANSD: What Do We Say to Families?

- Child has an auditory disorder
- Difficult to know prognosis
- Degree of deficit may be mild or severe
  - a small number have normal hearing sensitivity
- Results of behavioral testing are necessary before specific recommendations can be made
- Hearing aid use helpful in some cases not in others but we will only know if child is fit appropriately and has consistent use
- Cochlear implantation may be a better option if adequate benefit from HA not received

Counseling in ANSD: What Do We Say to Families?

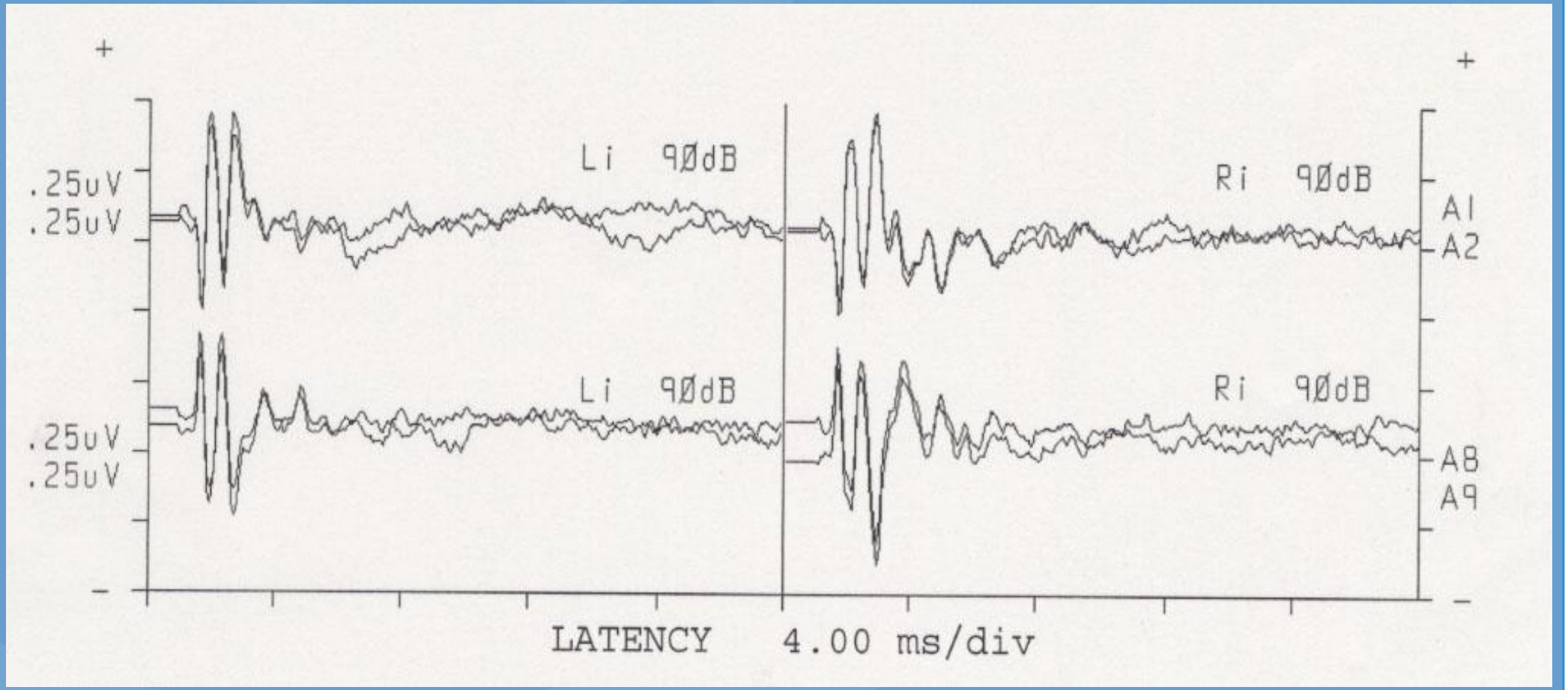
- Frequent follow up visits will be necessary
- Child should be enrolled in early intervention as soon as family is ready
- Most effective communication strategy will need to be determined with input from family, teachers, therapists, and audiologist
- We'll work together as a team to find a solution for their child's hearing disorder

Case Example

Case #1 Background

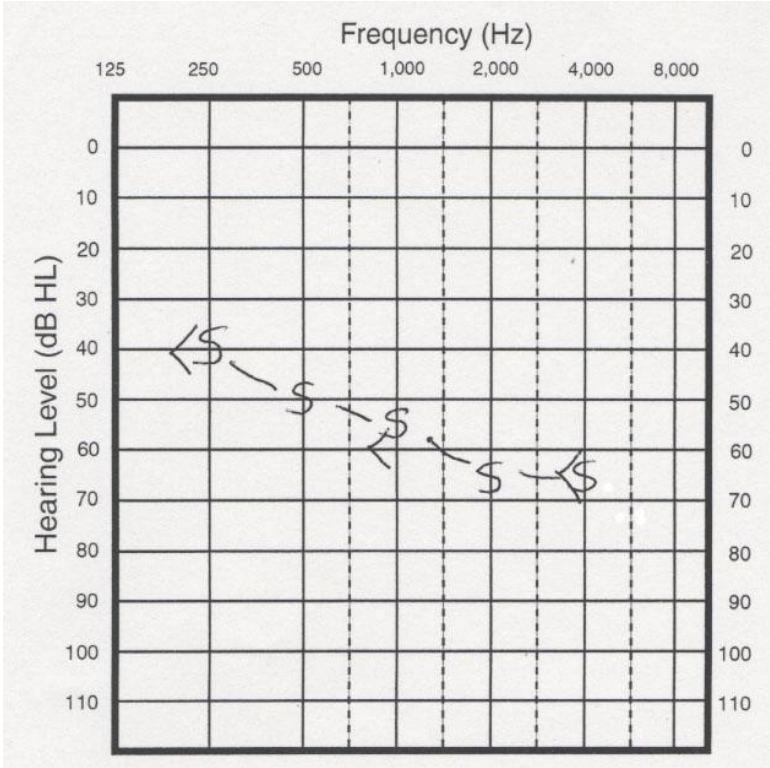
- 25 weeks gestation
- Ventilated for 6 weeks
- Oxygen 3 ½ months
- Hyperbilirubinemia
  - » Treated with lights, exchange transfusion
- Treated with antibiotics and diuretics
- Hospitalized 4 ½ months
- No family history of hearing loss
- Did not pass newborn hearing screen at hospital discharge
- Diagnosed with profound bilateral SNHL and fitted with high gain hearing aids

ABR Obtained at UNC, Age 6 Months (2 1/2 months adjusted age)



Age 10 Months (6 1/2 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



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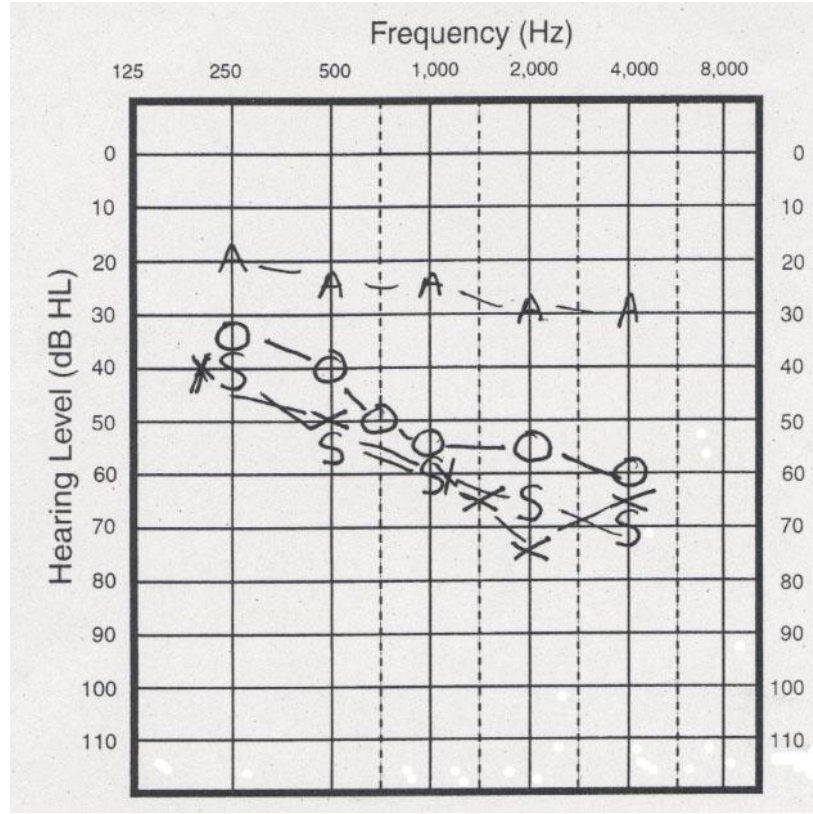
Age 12 Months (8 1/2 Months Adjusted Age): VRA with Insert Earphones Attached to Child's Earmolds

- 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



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VRA with Insert Earphones
Age 24 months
(20 ½ months adjusted age):

Child conditioned for play audiometry procedure but limited attention span

- Results similar to previous audiograms

Tympanometry

- Right: normal
- Left: normal

Communication Status
Age 24 Months (20 1/2 months adjusted age):

- Parental Report:
  - » Child understands several words, using 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

Age 5

- Mainstreamed in kindergarten
  - » Using personal FM in classroom
- Receiving services from auditory verbal therapist and speech and language pathologist
- Functioning in average range in receptive and expressive language development
- Working on articulation errors

Conclusions

- ANSD is more complicated than originally thought and population more heterogeneous
- It's unlikely that a single approach to management will meet the needs of all children.
- Some children will benefit from hearing aids either in the short term or the long term, others will require cochlear implantation.
- Visual methods to support communication may be required for some children even those who have received cochlear implants

Conclusions

- The available clinical evidence does not support withholding audibility from infants with ANSD Although audibility does not ensure good speech recognition, lack of audibility is certain to result in poor speech recognition.
- Important to consider the needs of the whole child, not only the auditory neuropathy diagnosis.
- Important to use team approach to carefully monitor child's progress in meeting communication goals.

Research Needs

- Evidence regarding clinical management and use of amplification is still limited. More research needed especially with infants and young children
- Studies aimed at evaluating hearing aid outcomes should include evidence-based prescriptive hearing aid fitting methods and real-ear verification methods appropriate for use with infants and children.
- Further investigation needed of alternative hearing aid processing strategies; however, non-traditional strategies need to be evaluated in older children and adults before they are used with infants and young children

Research Needs

- Better clinical tools to help determine site of lesion
- Better ways to predict who will benefit from amplification vs cochlear implantation
- Continued research needed into the role of CAEP in evaluation and management
- Research into signal processing strategies that target temporal vs spectral disruptions

Selected References and Resources

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THANK YOU!
Questions??

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