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# Understanding Auditory Neuropathy: Diagnosis and Management

EHDI: Partnering for Progress

Raleigh, NC

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# Outline

- Overview and Definitions
- Protocol for Diagnosis and Management
- Case Studies
- Implications for Clinical Practice

# Auditory Neuropathy: A Definition

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

# 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 (PJKV)
- Autosomal 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

- 
- *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>

# Panel Members

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



## **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

# Comprehensive Evaluations Following Diagnosis with ANSD

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



# 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



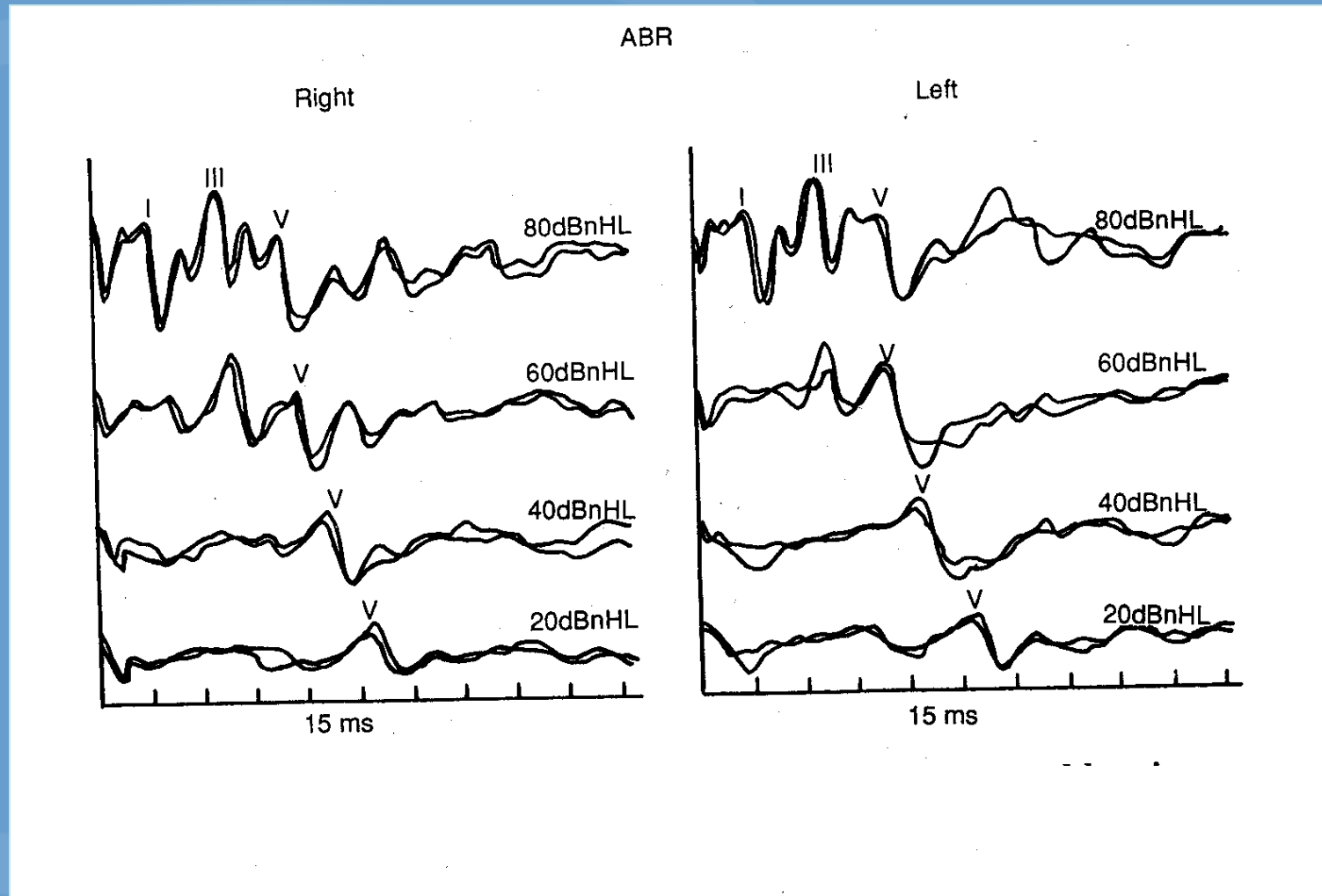
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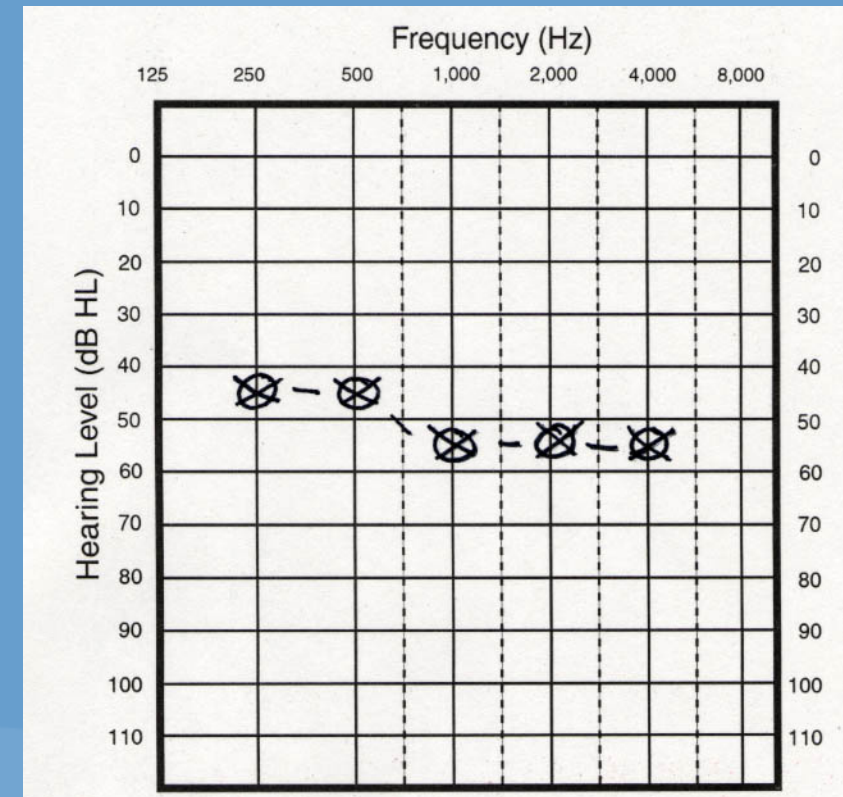
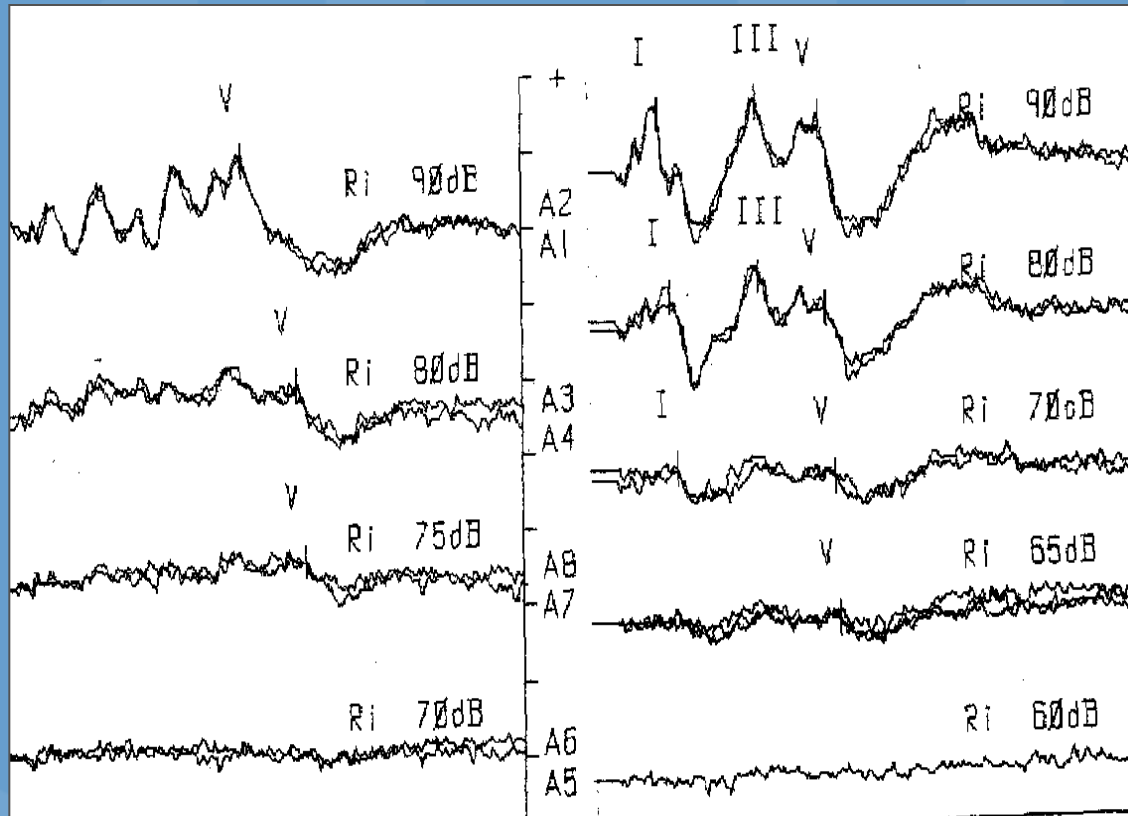


# Normal ABR



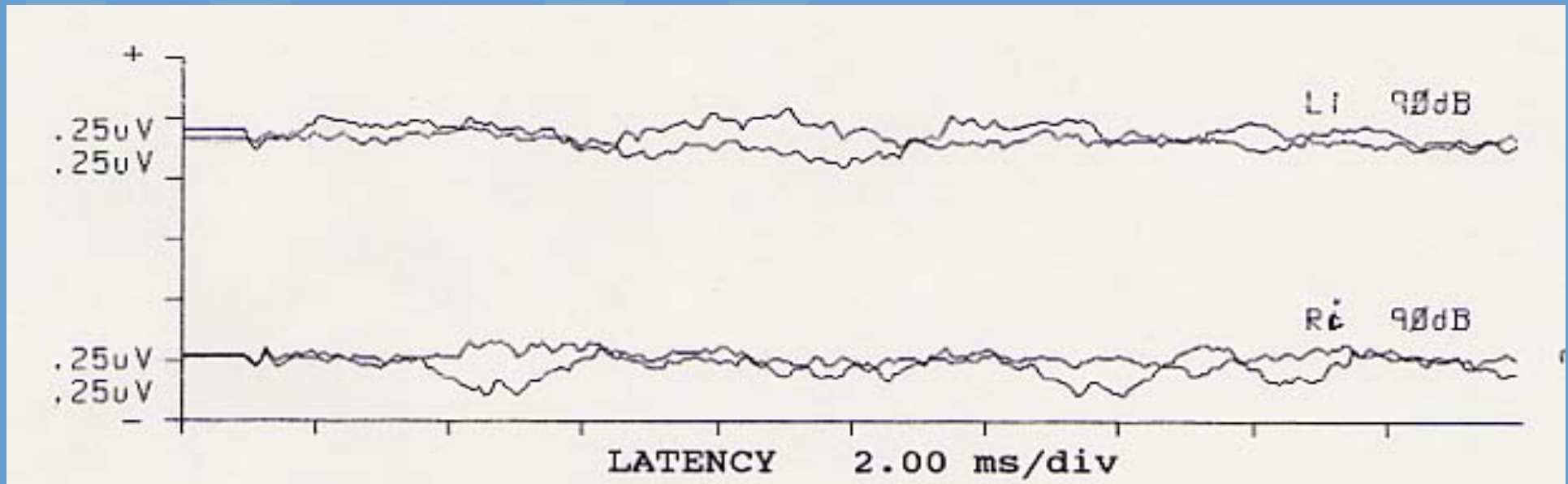


# Estimating the Audiogram from ABR



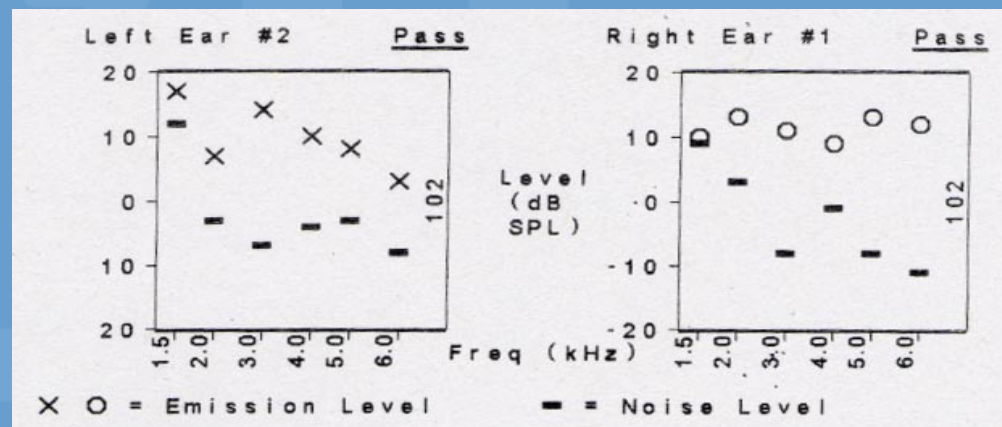
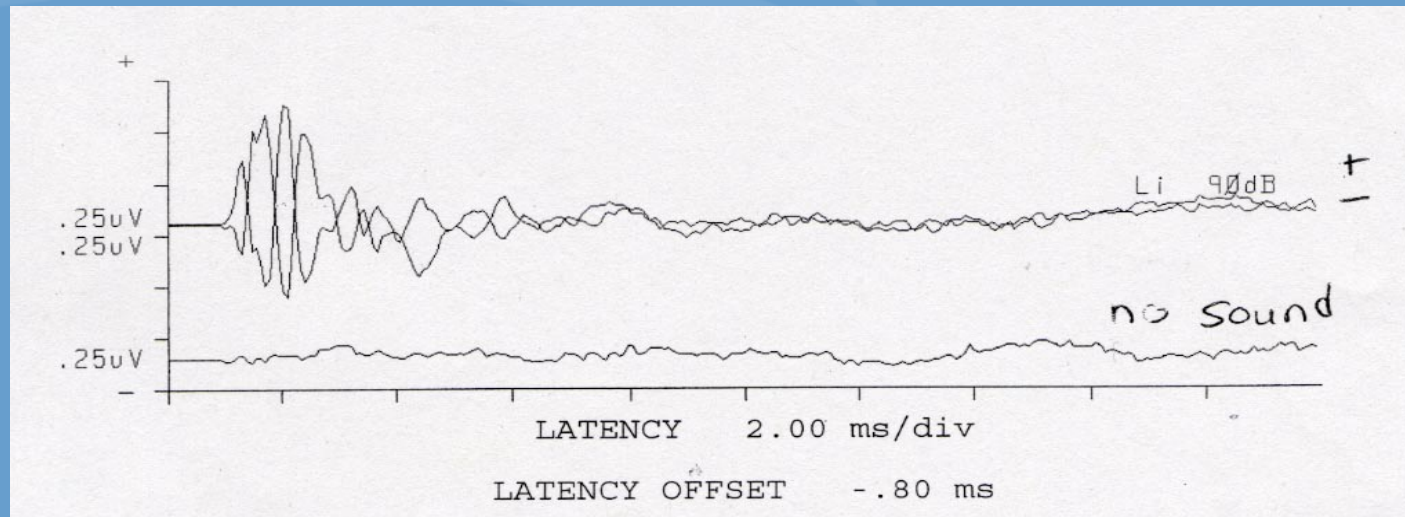


# Absent ABR with No Cochlear Microphonic: Child with profound hearing loss





# 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

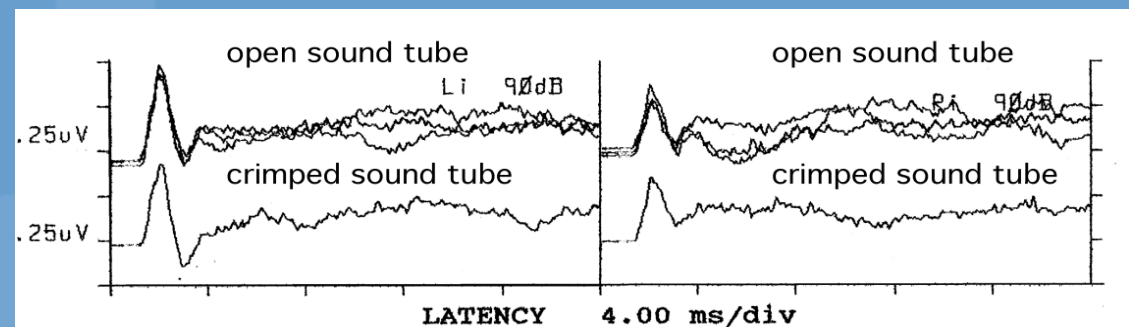
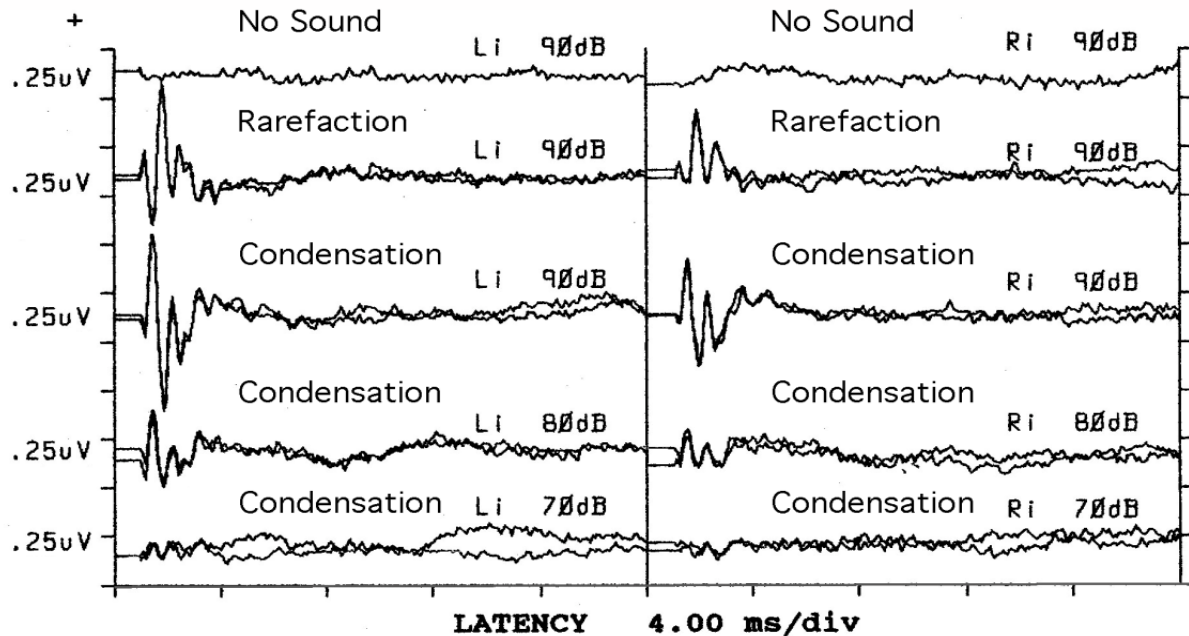
- Must have adequate recording conditions
  - » Infant ready to sleep
  - » Avoid electrodes positioned over transducer
- *Single polarity* clicks at 80 & 90dBnHL with rarefaction and condensation polarities
- Must use insert earphones
- No-sound run with sound tube disconnected or clamped to check for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

# CM vs stimulus artifact



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## Bilateral auditory neuropathy/dys-synchrony



# Auditory Steady State Response (ASSR)

- ASSR responses can be obtained to high signal levels ( $>80\text{dBHL}$ ) 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
- Ongoing electrophysiologic studies at UNC involving CAEP and electrocochleography



# Cortical Evoked Potentials (CAEP)





# 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



# ANSD Protocol for Infants: Behavioral Audiometry

- Behavioral assessment with VRA beginning at 6-7 months (developmental age) with goal of obtaining individual ear measures and bone conduction thresholds by 8-9 months of age
  - » May be difficult (or impossible) with children who have additional developmental or medical challenges
  - » Behavioral Observation Audiometry may be needed



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# VRA Six Month Old







# Hearing Aid Fitting in Infants with ANSD

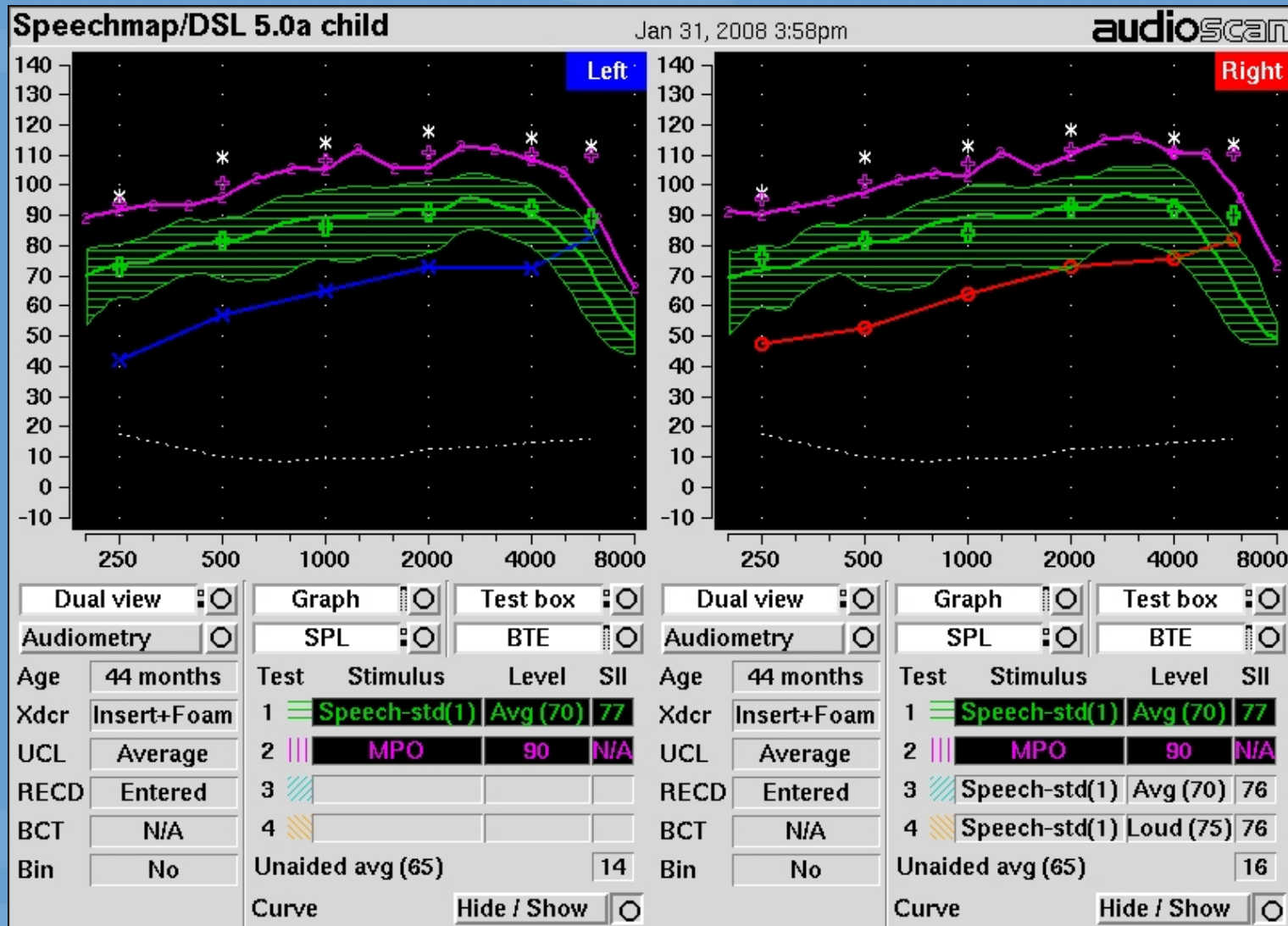
- Behavioral thresholds cannot be predicted from ABR or ASSR
- Determination of hearing thresholds is delayed until infant developmentally able to perform task (6-9 months of age for most typically developing infants)
- Many children with ANSD are at risk for cognitive impairments resulting in a lengthier 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

# 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.



# Verifying Audibility of Speech Spectrum



# 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.

# 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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# 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



## **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*.

# 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



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# Variable Presentations of ANSD

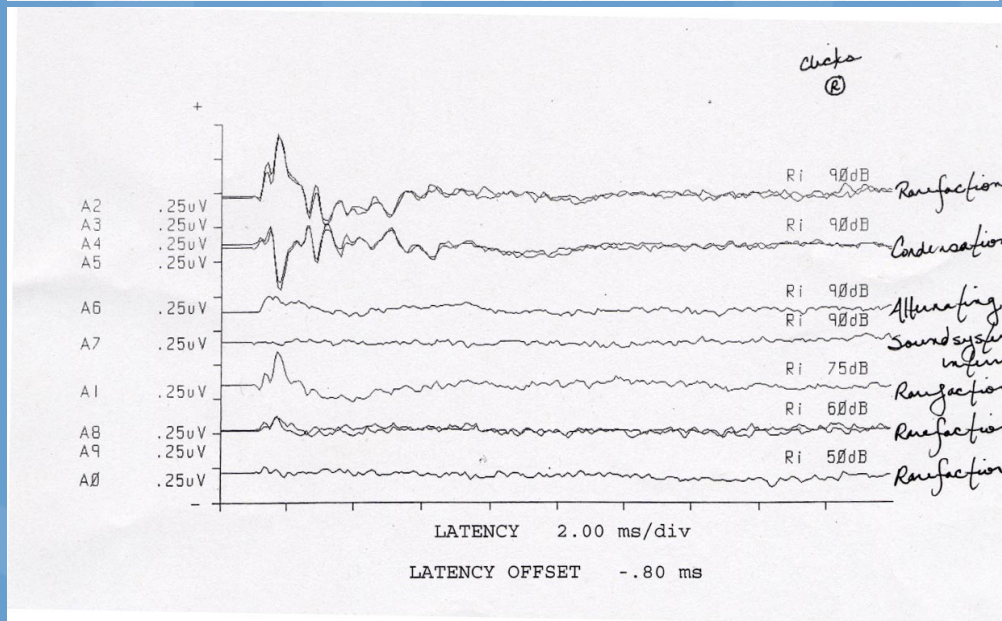
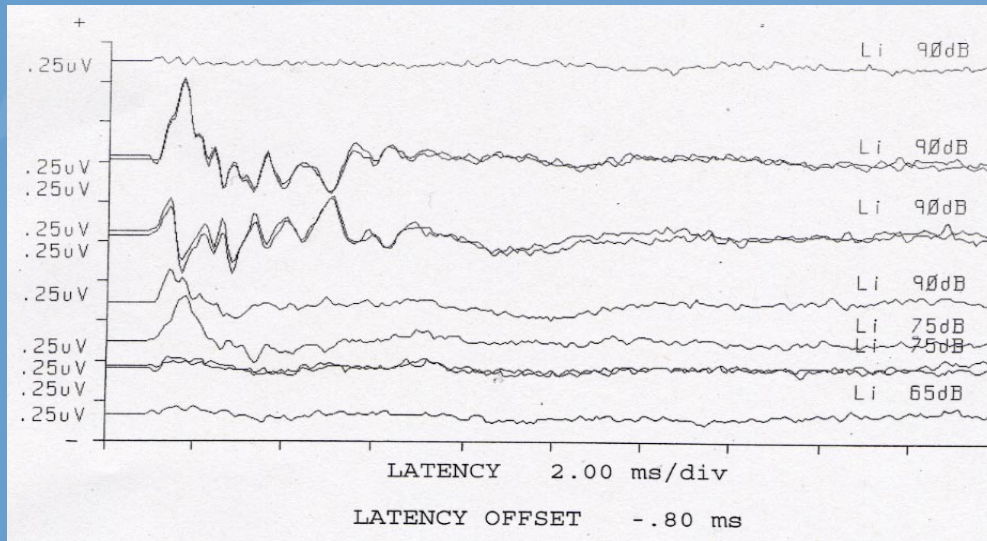
## Case Examples

# Case #1:

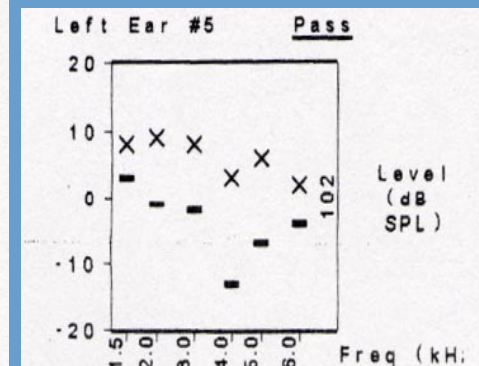
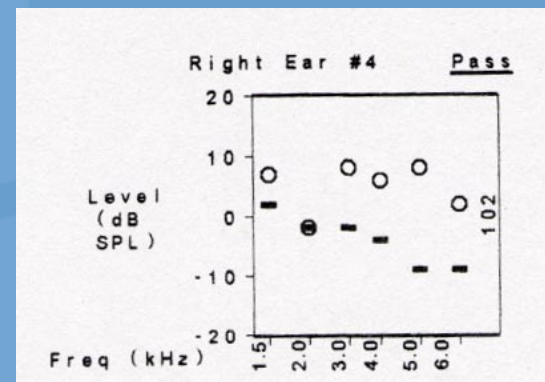
## Present CM and OAEs



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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

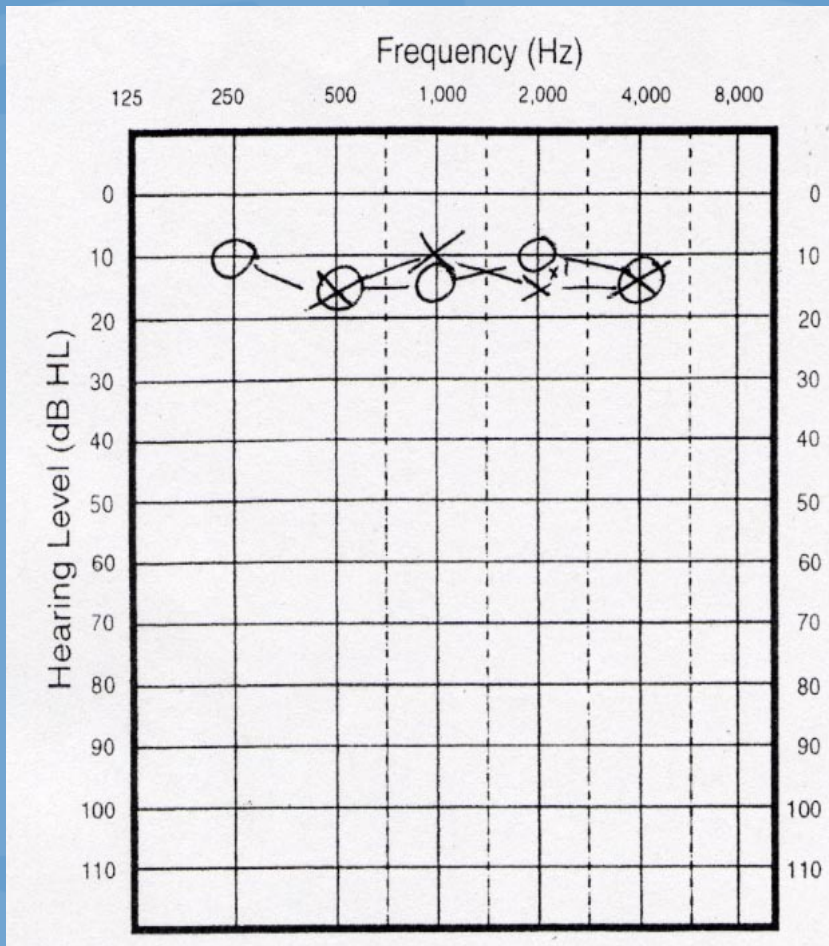


# Case #1

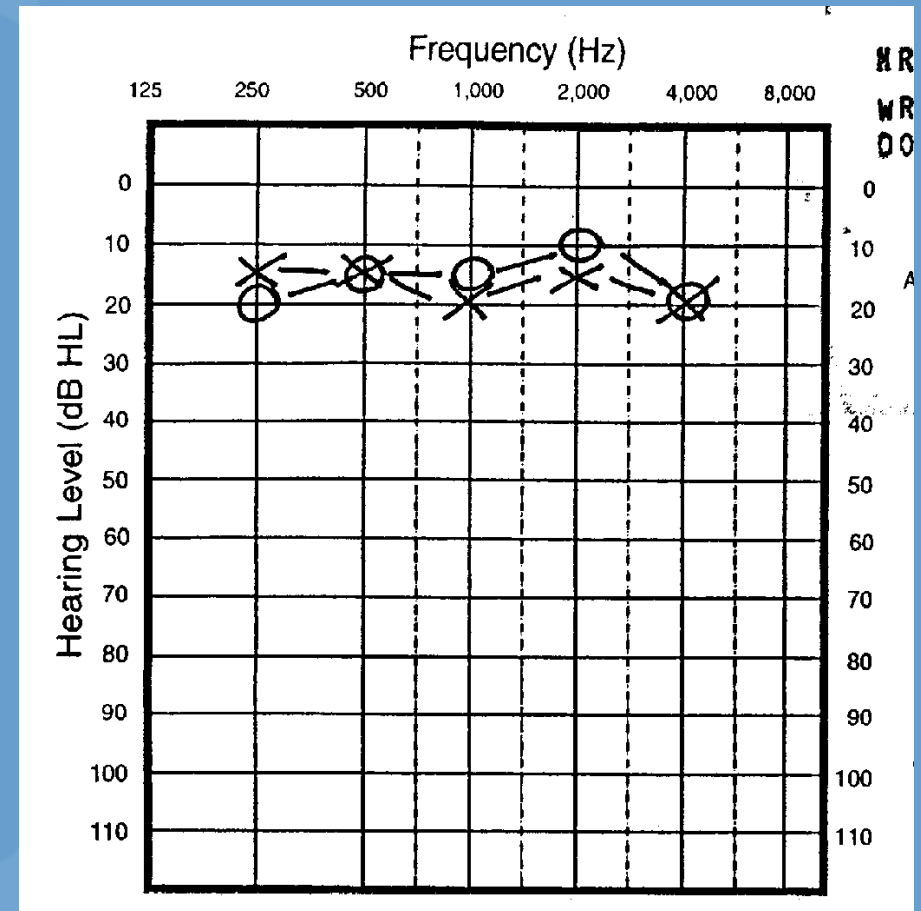
Normal thresholds, Present CM  
and OAEs



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**Audiogram at 14 months**



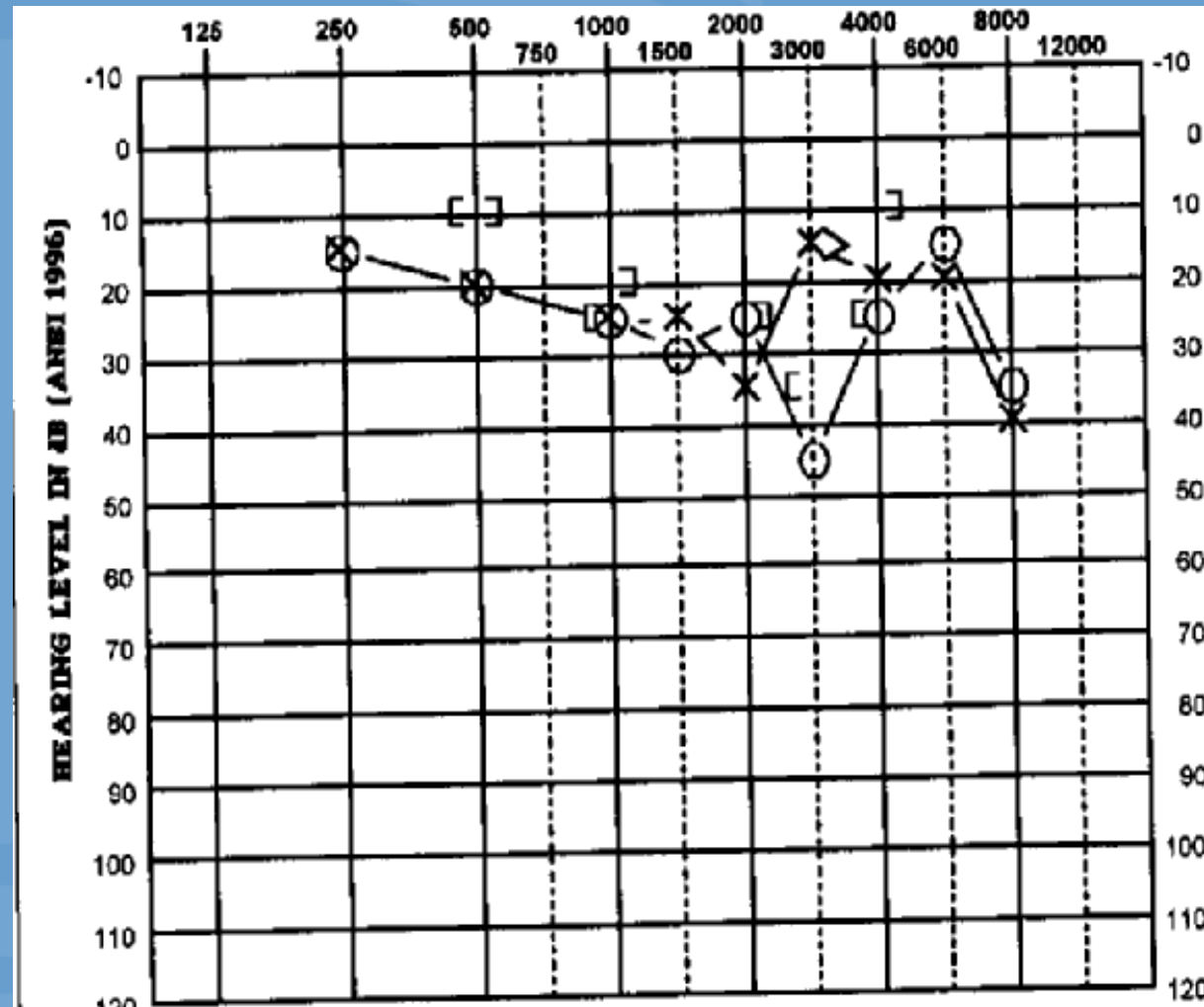
**Audiogram at 18 months**

# Case #1

## Audiogram at Age 5



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# Case #1

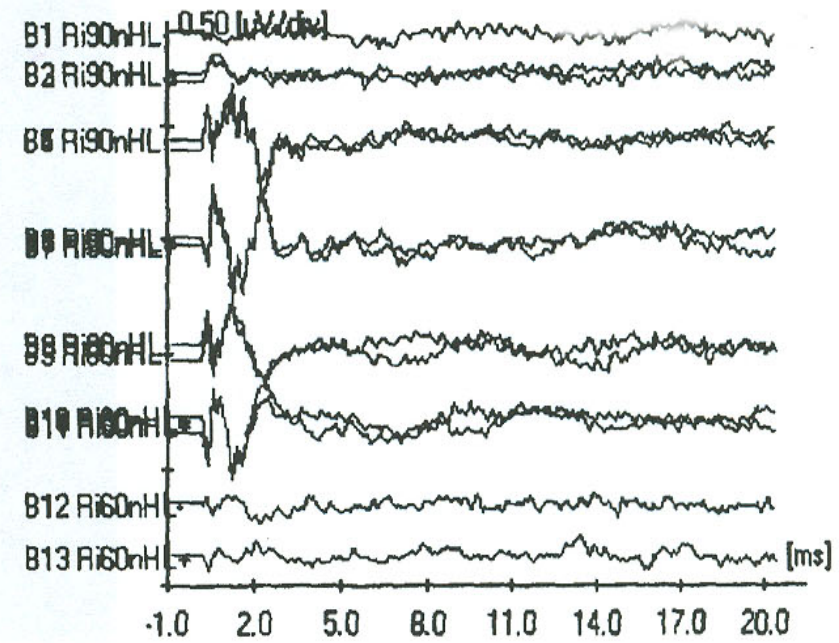
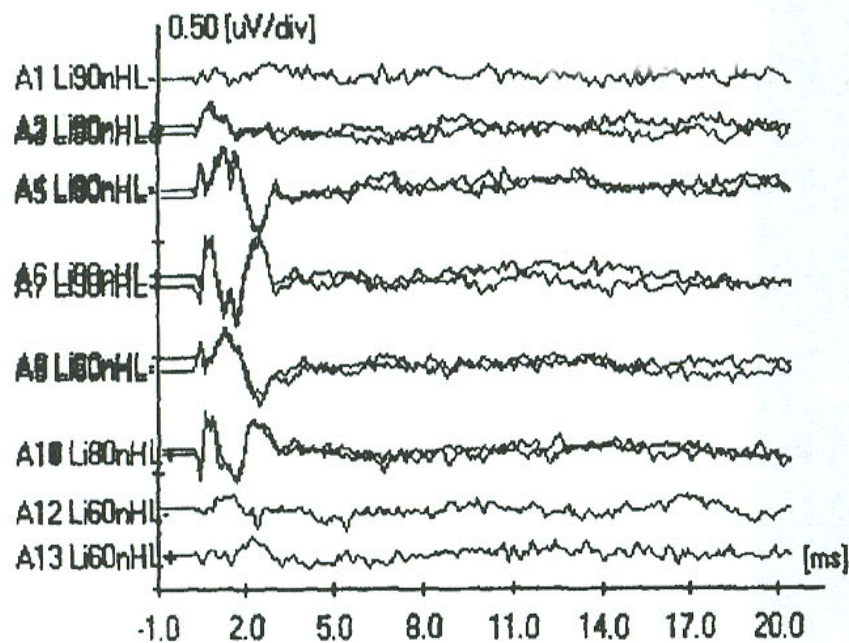
## Speech Perception Test Results

- Age 2 yrs-11 months:
  - ESP monosyllabic word test:
    - 12/12 correct for each ear at 50dBHL
- Age 3 yrs-3 months:
  - PBK words: 64% and 72% at 55dBHL
  - ? Speech production errors
- Age 5 years:
  - PBK words: 80% and 84% at 60dBHL



## Case #2

# Abnormal ABR with Present CM

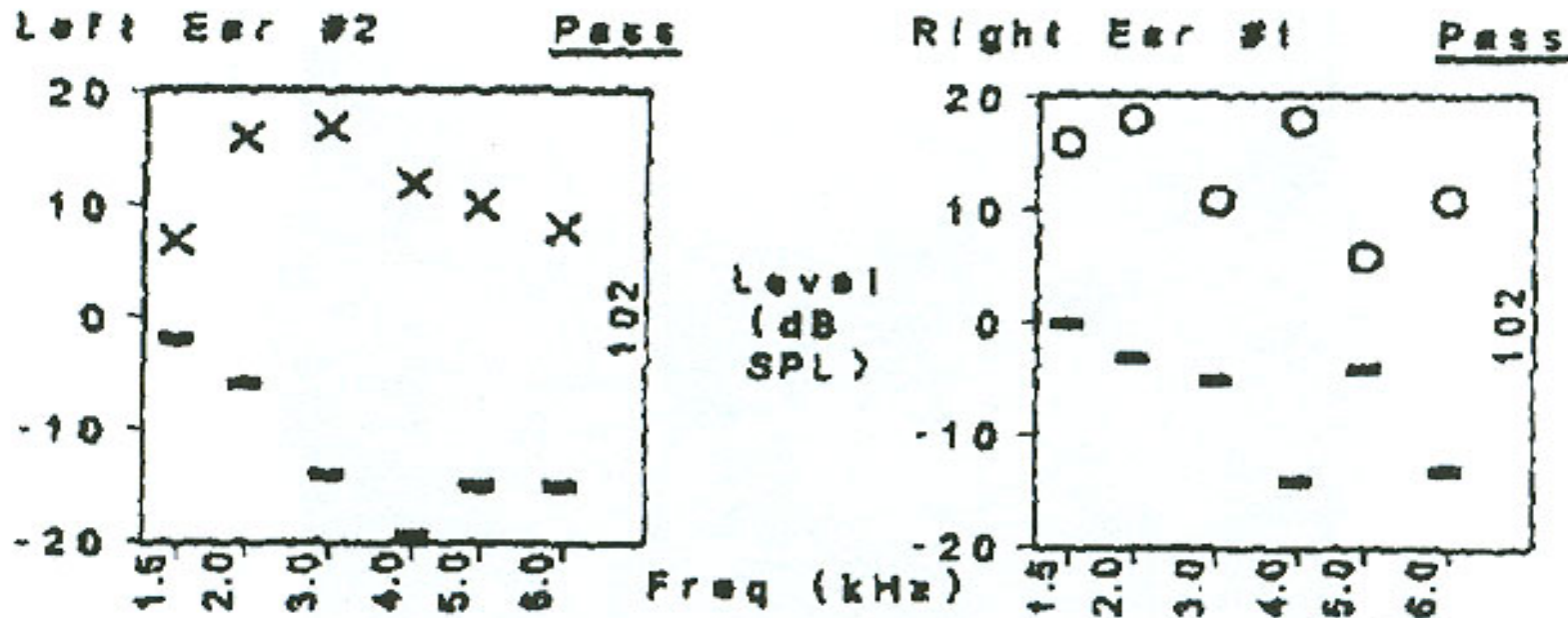






# Case #2

## Present OAEs

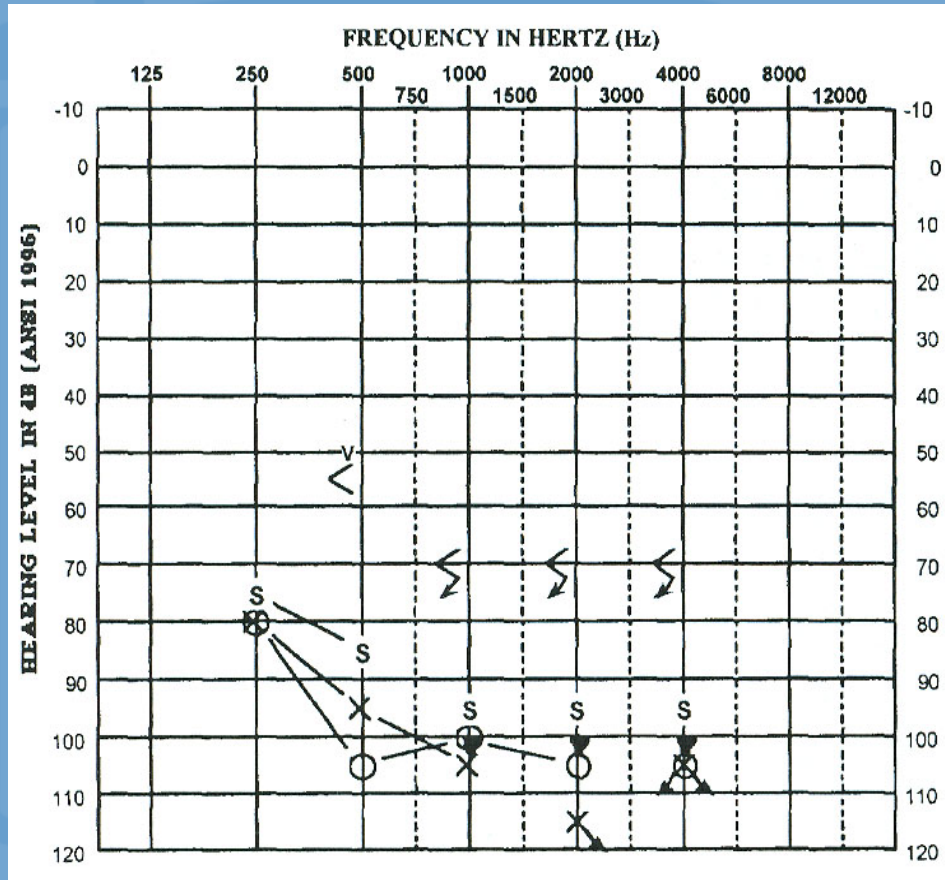


## Case #2

Child with Profound Bilateral HL  
Present CM and OAEs



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Ear exam: *Normal*

EKG: *Normal*

MRI: *Normal*

Connexin test: *Negative*

Otoferlin test: **POSITIVE**

Received CI at 24  
months of age (Late  
diagnosis)

## Case #2 Genetics Report

- Genetic testing showed child to have single disease-causing OTOF mutation.
- Inherited as an autosomal recessive condition
- Parents are likely to be carriers of auditory neuropathy
- Confirmation of their heterozygous carrier status by mutation analysis is recommended
- Recurrence risk for each full sibling is 25%

# Case # 3 Background

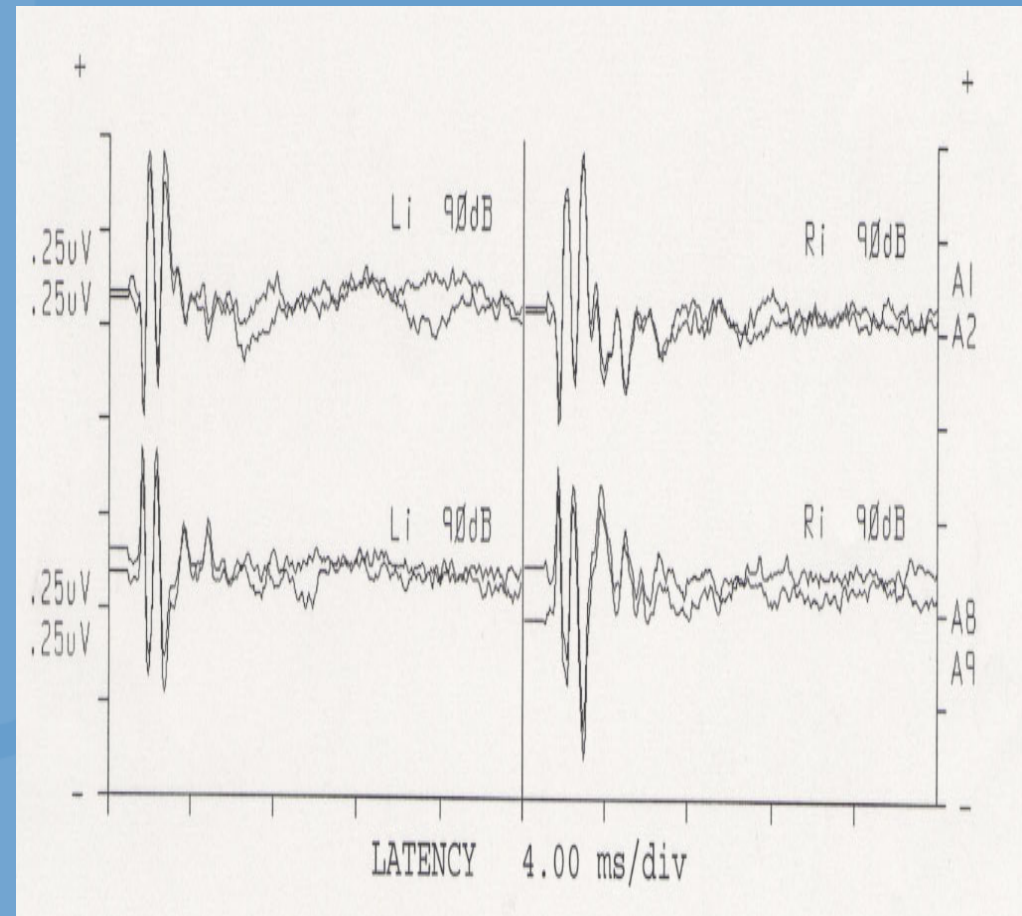
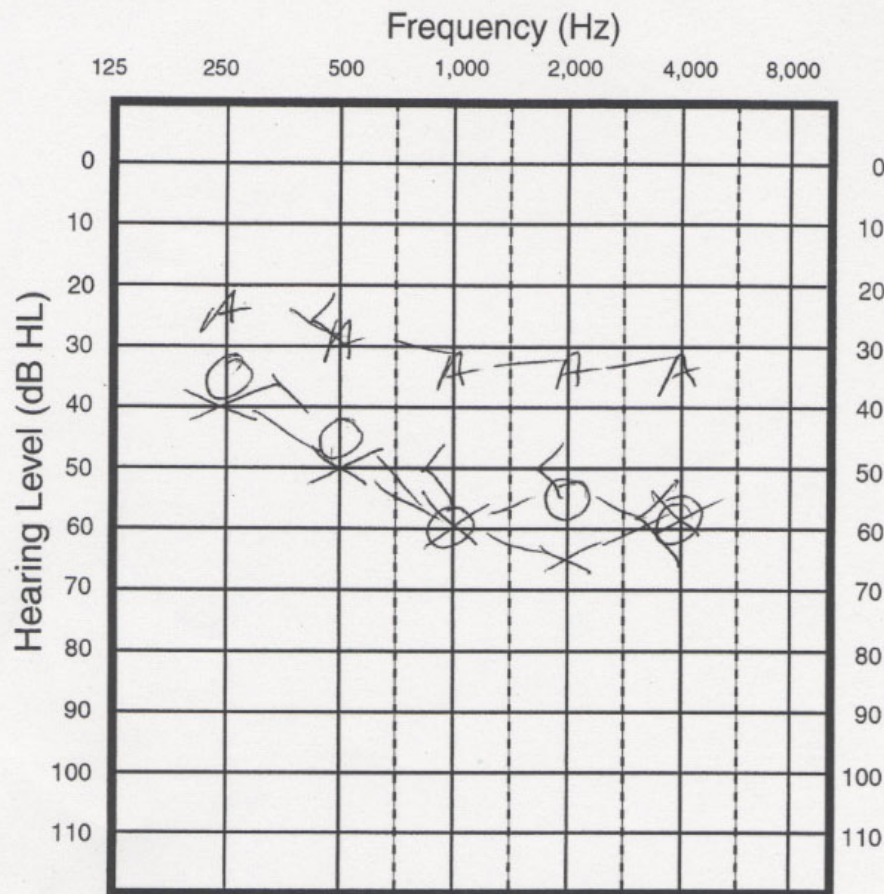
- 25 weeks gestation
- Ventilated for 6 weeks
- Oxygen 3 ½ months
- Hyperbilirubinemia
  - » Treated with lights, exchange transfusion
- Treated with antibiotics and diuretics
- Hospitalized 4 ½ months
- No family history of hearing loss
- Did not pass newborn hearing screen at hospital discharge
- Diagnosed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

# Case #3

Child with “moderate loss”  
CM present, absent OAEs



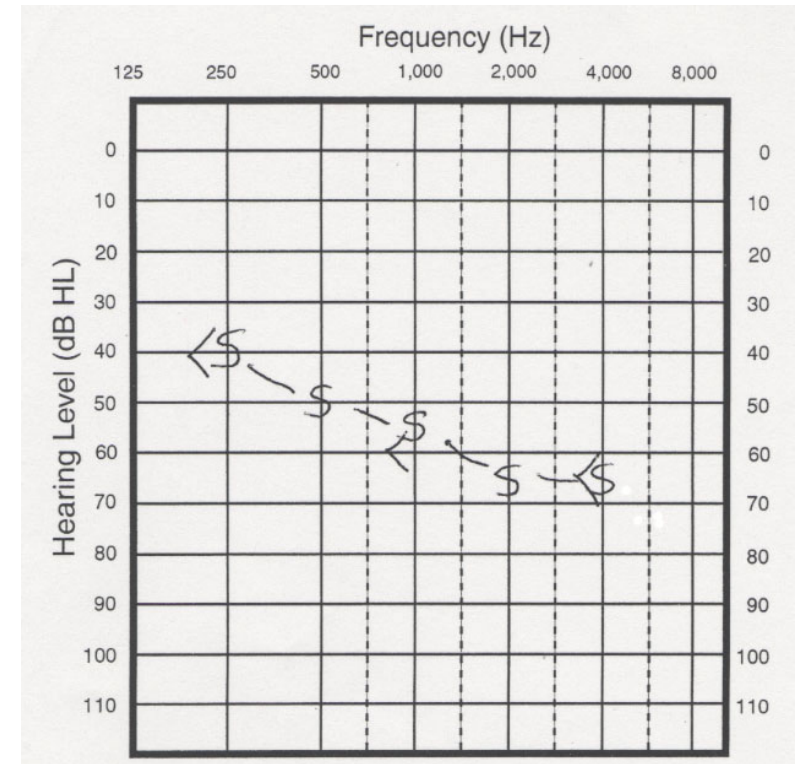
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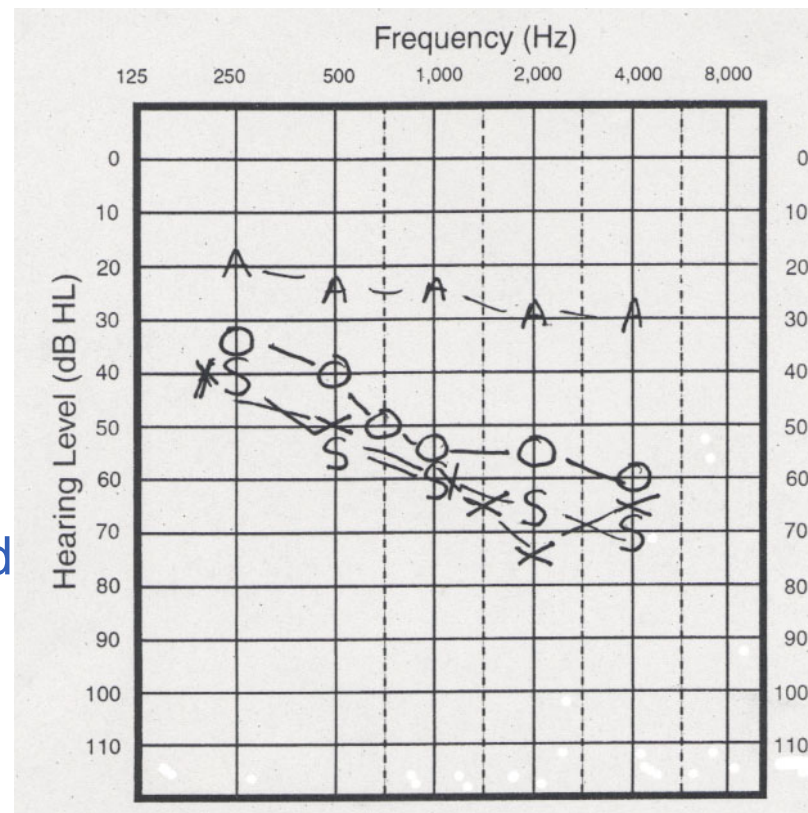
## Age 10 Months (6 1/2 Months Adjusted Age): Behavioral Audiometry with VRA

- 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



## 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





# 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

# Case #3

## Age 8 years

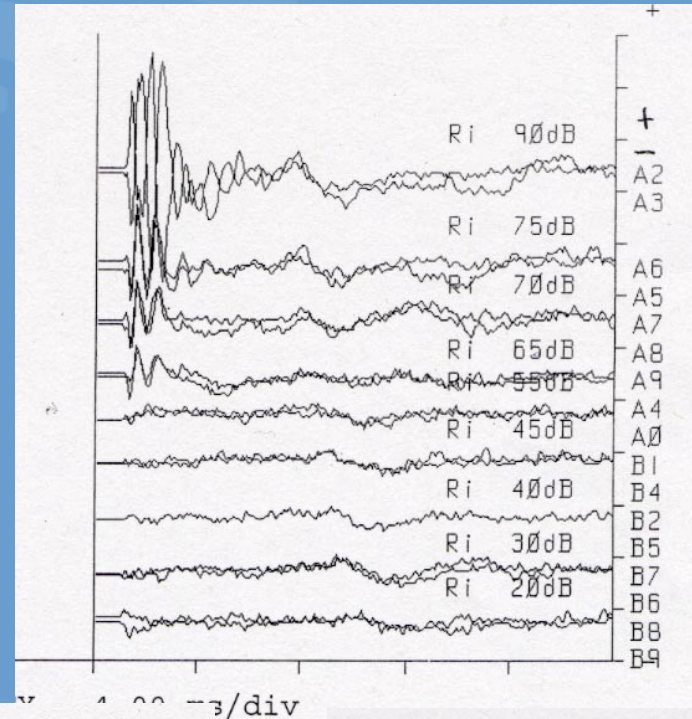
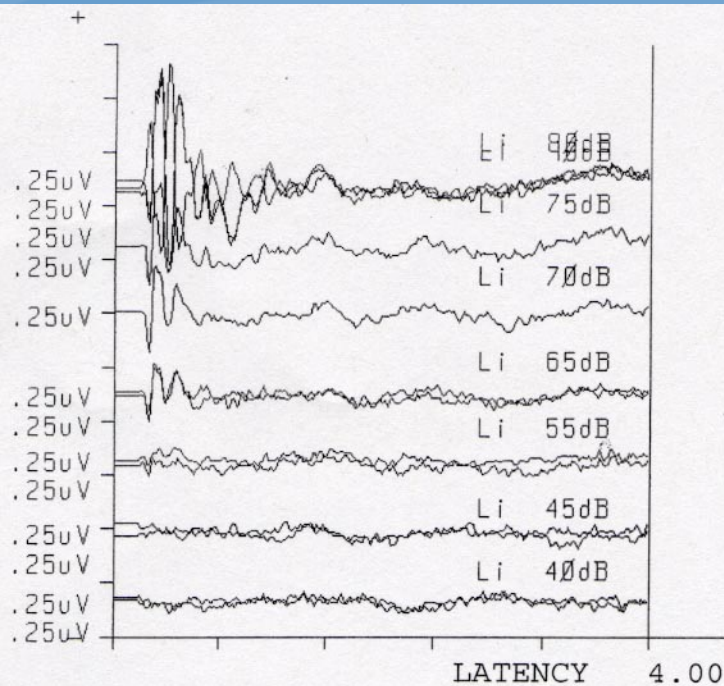


- Mainstreamed in 2<sup>nd</sup> grade
  - » Using personal FM in classroom
- Receiving services from auditory verbal therapist and speech and language pathologist
- Functioning in average range in receptive and expressive language development
- Working on articulation errors

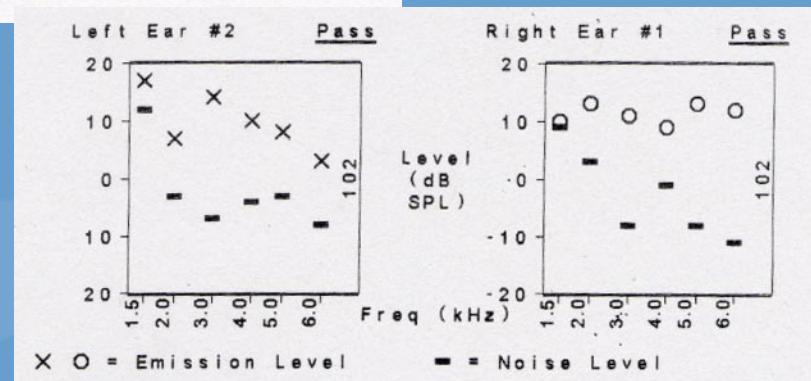
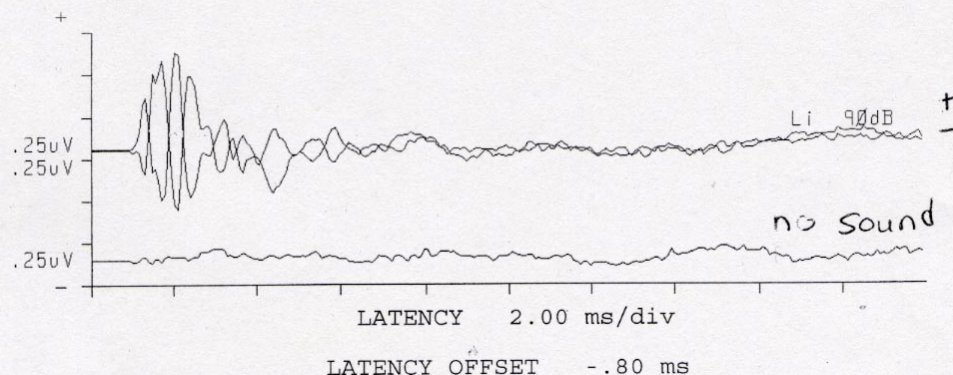
# Case #4: Large CM; Present OAEs; Distal Waveforms Present



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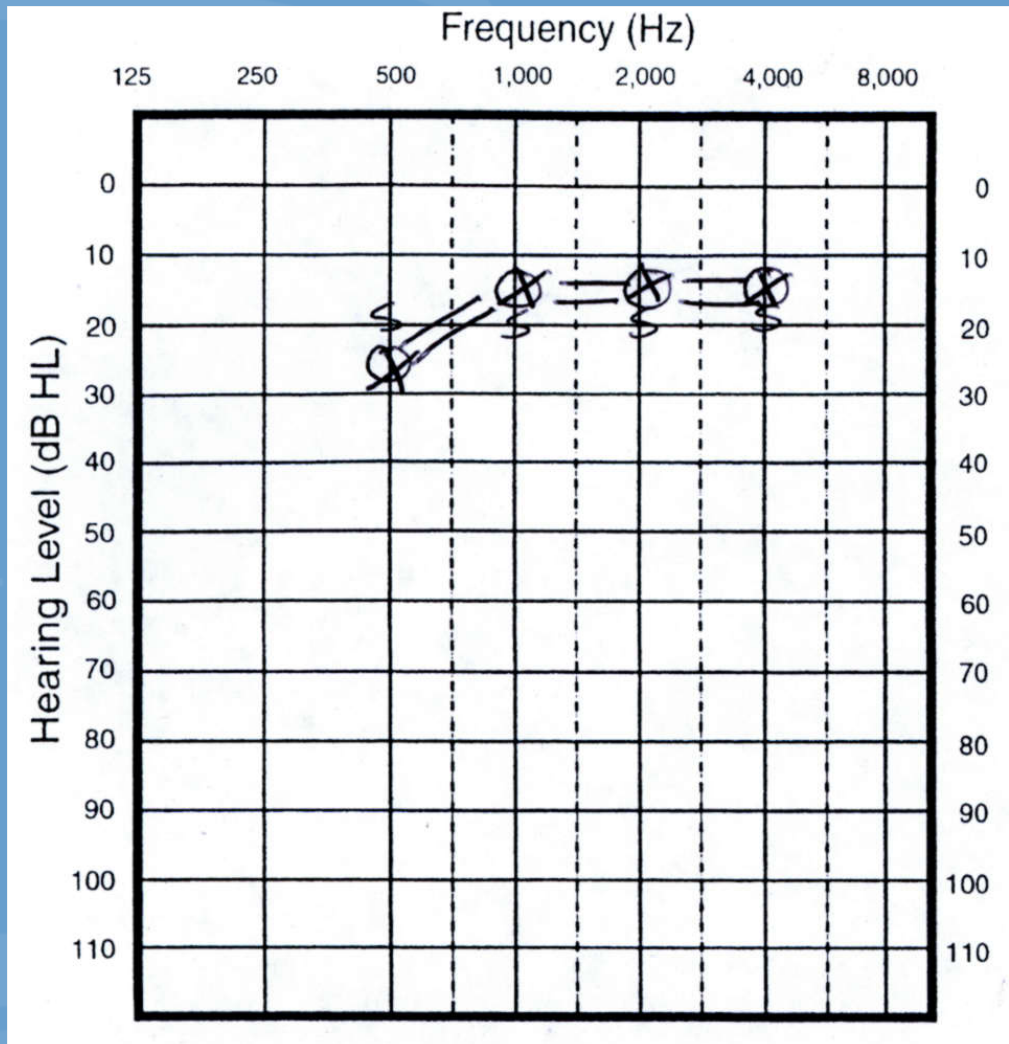
**Caution needed  
when interpreting  
ABRs that show  
abnormal  
waveform  
morphology at  
high intensity  
levels**



# Case #4 (continued)

## VRA with insert earphones

### Age 14 months



## Case #5

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

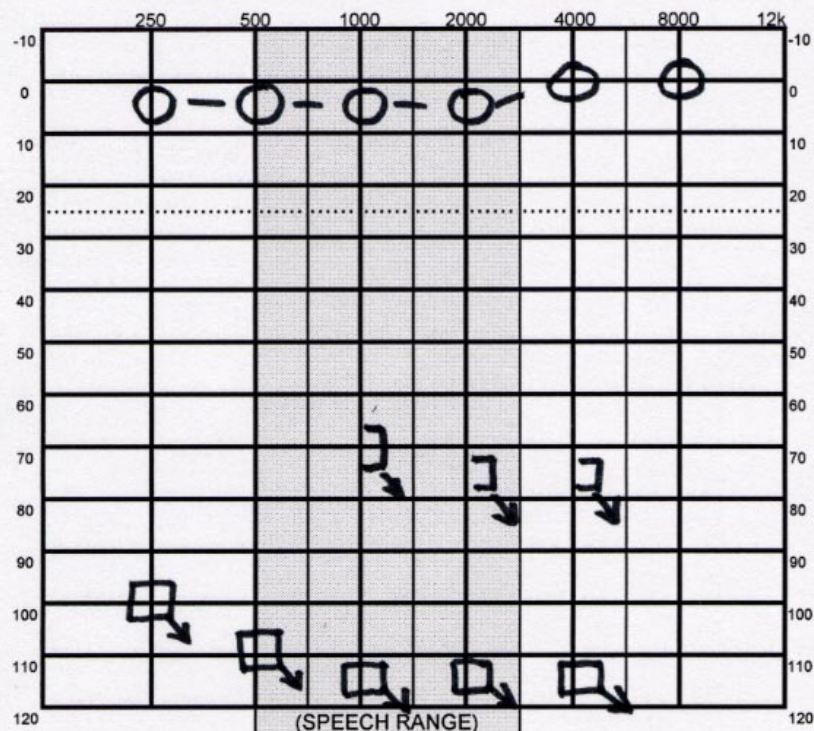


# Case # 5



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## PURE TONE AUDIOMETRY (RE: ANSI 1969)



	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)	.5k Hz	1k Hz	2k Hz	4k Hz
RIGHT (AD)	85	85	85	
LEFT (AS)	NR	NR	NR	

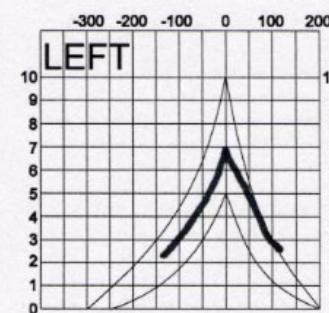
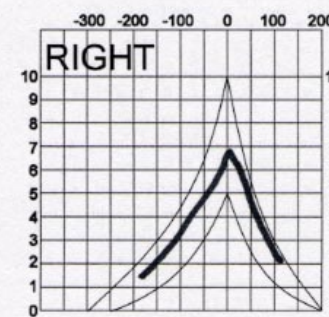
REFLEX DECAY  
@ 1k Hz

STATIC COMPLIANCE

RIGHT (AD)		
LEFT (AS)		

### MODE

STANDARD EARPHONES	
INSERT EARPHONES	
SOUND FIELD TESTING	
ASSISTED EVALUATION	



TYMPANOMETER: \_\_\_\_\_

## SPEECH AUDIOMETRY

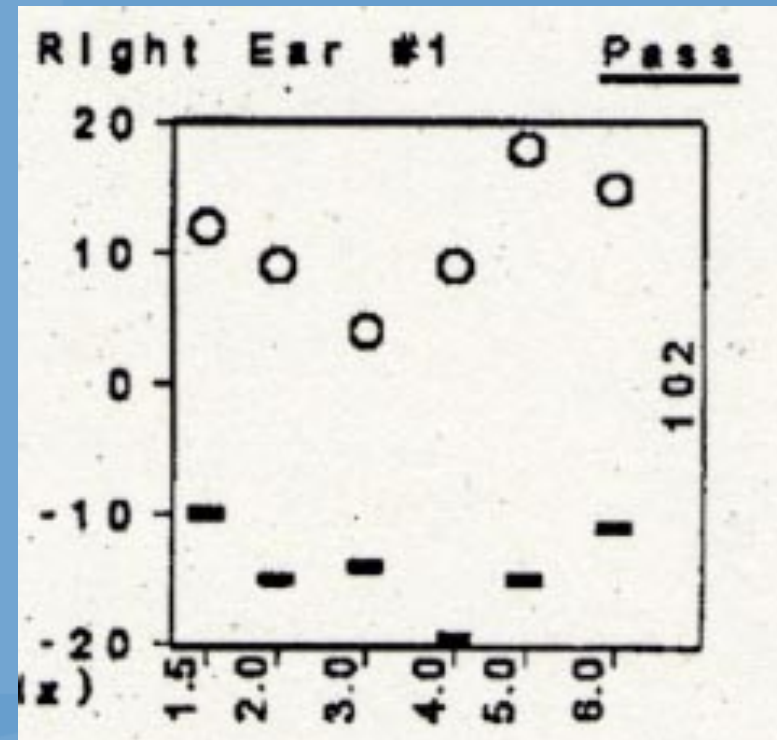
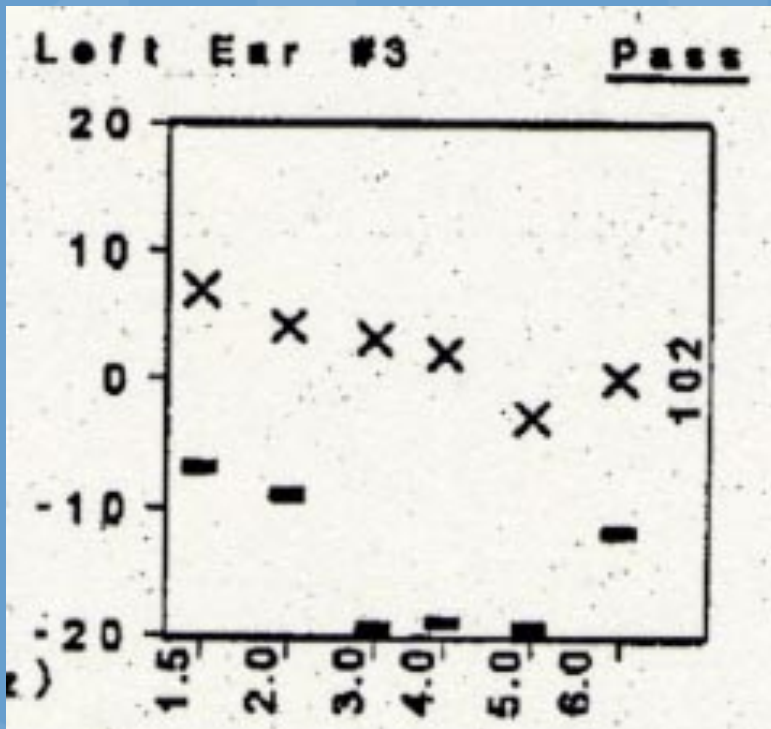
	PTA	SRT/SAT	SPEECH RECOGNITION		SPEECH RECOGNITION		MCL	UCL
RIGHT (AD)		O	%	dB HL	%	dB HL		
masking			100	50				
LEFT (AS)		NR	%	dB HL	%	dB HL		
masking			CNT					

HISTORY: \_\_\_\_\_

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_



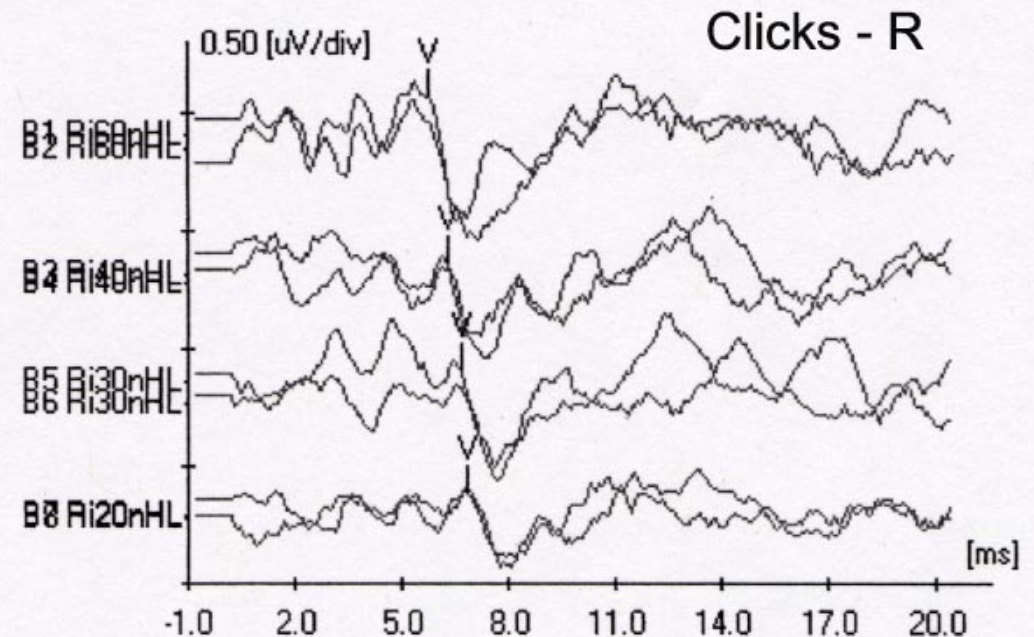
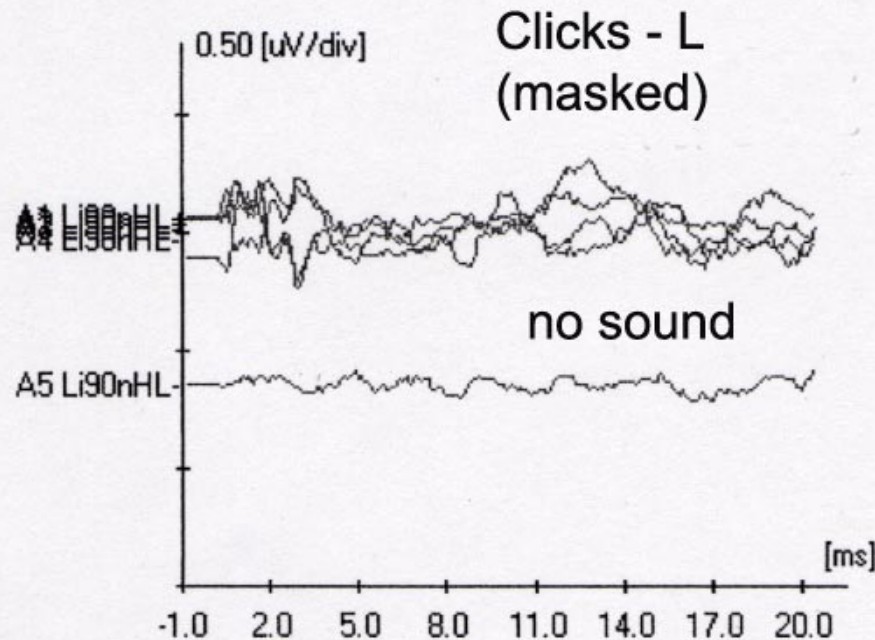
# Case #5 OAEs





# Case #5 ABR

## Completed at Age 4 years





## Case #5

### ➤ Results of MRI:

- » Cochlear nerve on left smaller than the right
- » Question of left cochlear nerve hypoplasia

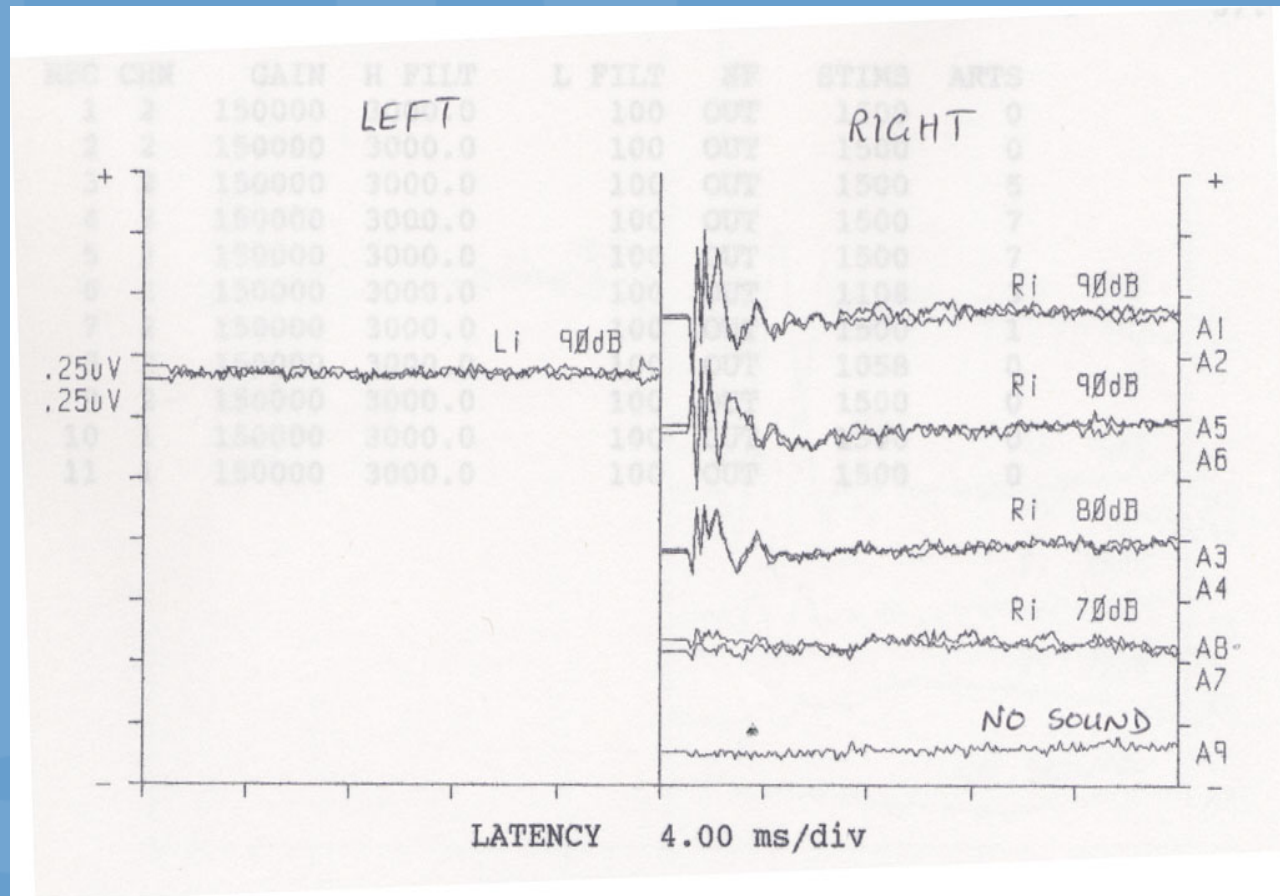
### ➤ At age 7 years child has above average speech and language development, no academic problems

### ➤ Managed as we do other cases with profound unilateral hearing loss.

# Case # 6

## Bilateral deafness

### No VIII<sup>th</sup> nerve on right



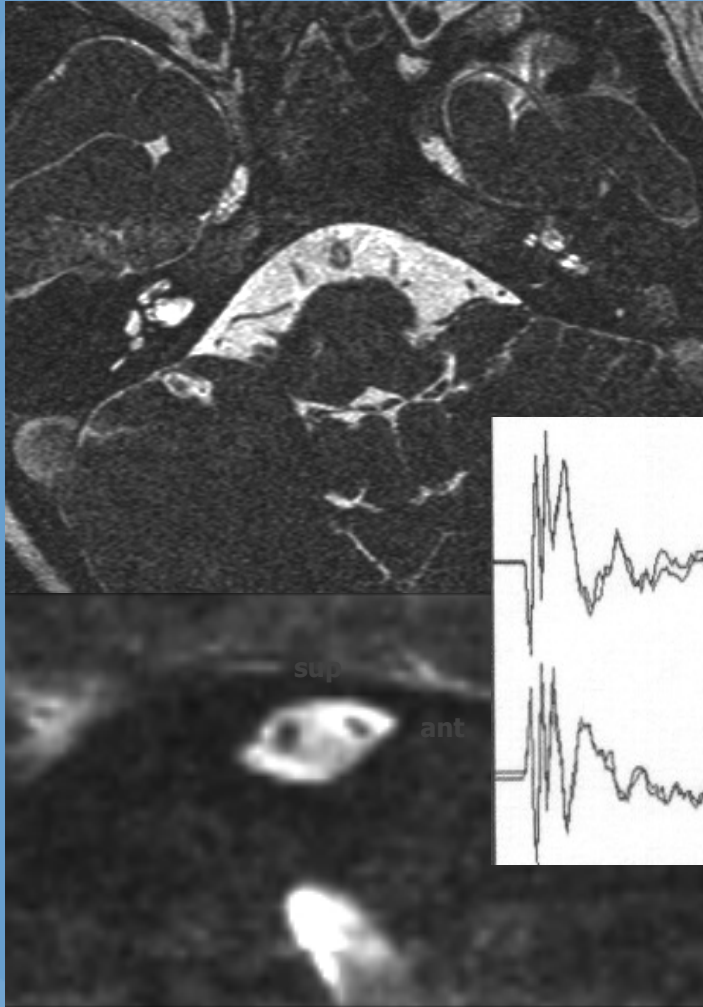
# Case #6

## Child with bilateral deafness

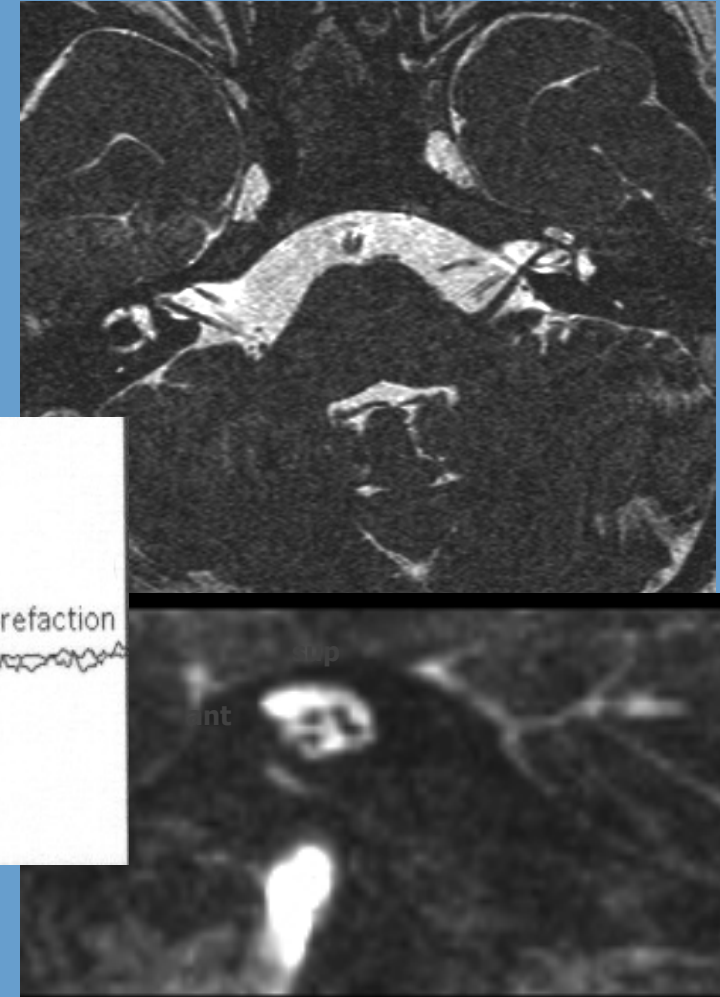
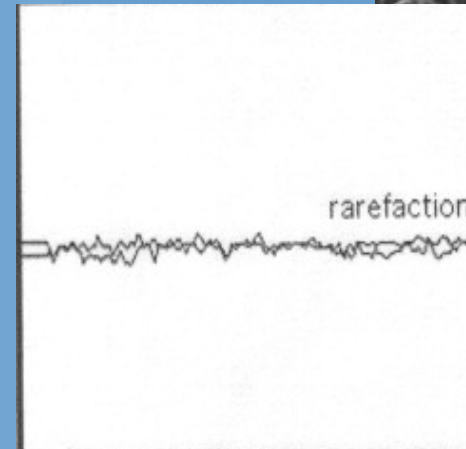
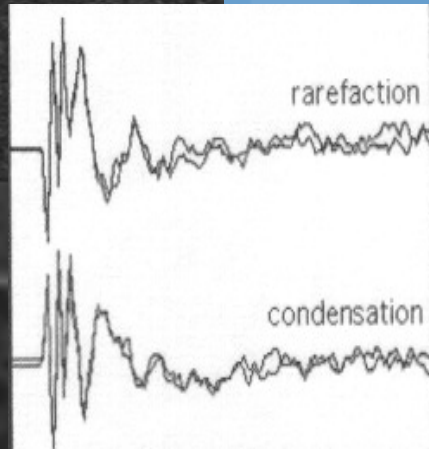
### No VIII<sup>th</sup> nerve on right



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Right Ear



Left Ear

# Cochlear Nerve Deficiency (CND)

- Small or absent VIII nerve
- Must perform MRI to determine if VIII nerve is small or absent
- CT may show normal IAC when cochlear nerve is absent
- In cases when there is question of CND both CT and MRI may be needed

# 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



# Six Cases with ANSD pattern on ABR...Six Different Outcomes

1. Normal hearing sensitivity no device needed, limited services required
2. Child with profound bilateral hearing loss; CI required
3. Child with moderate HL benefitting from amplification
4. Child with AN pattern on ABR but distal waveforms present: normal hearing sensitivity
5. Child with unilateral profound HL and absent cochlear nerve
6. Child with bilateral profound HL and absence of cochlear nerves



# **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

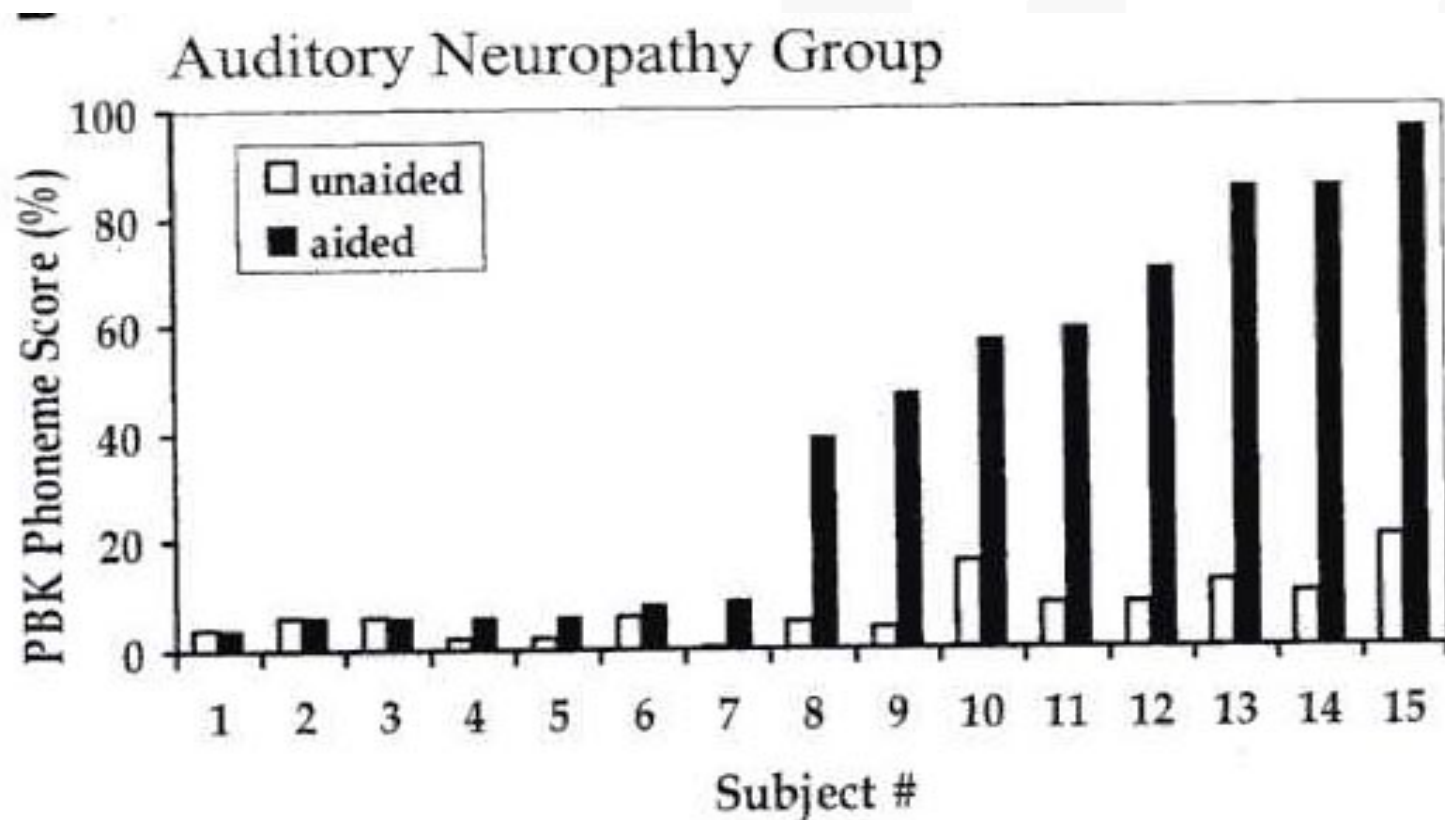
## ➤ Rance et al 2002

- » 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*

# Evidence re Amplification

- 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



# ***Audiological Management of Auditory Neuropathy Spectrum Disorder: A Systematic Review of the Literature***

Roush, P., Frymark, T., Venedictov, R., and Wang., B.  
Am J of Audio 2011, Sept. 22  
( epub ahead of print)



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# Counseling Families

# 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

# 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



# 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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- Rance, G and Barker, E. Speech Perception in Children with Auditory Neuropathy/Dyssynchrony managed with either hearing aids or cochlear implants. *Otology & Neurotology*, 2008, 29:179-182
- Roush, P, Frymark, T., Venedictov, B., and Wang, B. *Audiological Management of Auditory Neuropathy Spectrum Disorder in Children: A Systematic Review of the Literature*. Am J Audiol 2011 Sept 22 (epub ahead of print)
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- Simmons, J. & McCreery, R. (2007, June 19). Auditory neuropathy/dys-synchrony: Trends in assessment and treatment. *The ASHA Leader*, 12(8), 12-15.  
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# THANK YOU! Questions??

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