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Auditory Neuropathy Spectrum Disorder (ANSD)

Language and Literacy Workshop
Greensboro, NC
September 11, 2012

Patricia Roush, AuD
Holly Teagle, AuD

Department of Otolaryngology
University of North Carolina School of Medicine



NC Chapter of AG Bell

- Local, non-profit organization that brings families and professionals together to support children with hearing loss.
- Our community:
 - Families of children with hearing loss
 - Adults with hearing loss
 - Professionals

Why Join The AG Bell Association?

- » **FREE** subscription to *Volta Voices* and weekly e-newsletter
- » **ACCESS** to the Parent Advocacy Training (P.A.T.) program, financial aid and teen programs, local Chapters and professional listings
- » **DISCOUNTS** from the AG Bell bookstore and AG Bell conference and event registrations
- » **SUPPORT** from your local chapter through scholarships , advocacy efforts, and much more



University of North Carolina

Department of Otolaryngology

UNC Hospitals

Pediatric Audiology Program

Audiologists:

Nissele Franco, AuD
Corinne Macpherson, AuD
Sarah Martinho, AuD
Laura Fleenor McCall, AuD
Jill Ritch, AuD
Patricia Roush, AuD

Otolaryngologists:

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



University of North Carolina Department of Otolaryngology

Carolina Children's Communicative
Disorders Program

Center for the Acquisition of Spoken
Language through Listening Enrichment

Audiologists:

Debora Hatch, AuD

Lisa Park, AuD

Jennifer Woodard, AuD

Holly Teagle, AuD

Speech-Language Pathologists:

Hannah Eskridge, AVT

Lillian Henderson, AVT

Sandra Hancock, AVT

Erin Thompson, AVT

Maegan Evans, PhD



University Of North Carolina Chapel Hill, North Carolina

Universal newborn hearing screening legislation 1999

Pediatric Audiology and CI Teams

CASTLE pre-school

Total 1700 infants and children

- » 1000 using amplification
- » 800 with cochlear implants
- » 200+ with ANSD diagnosis

Outline

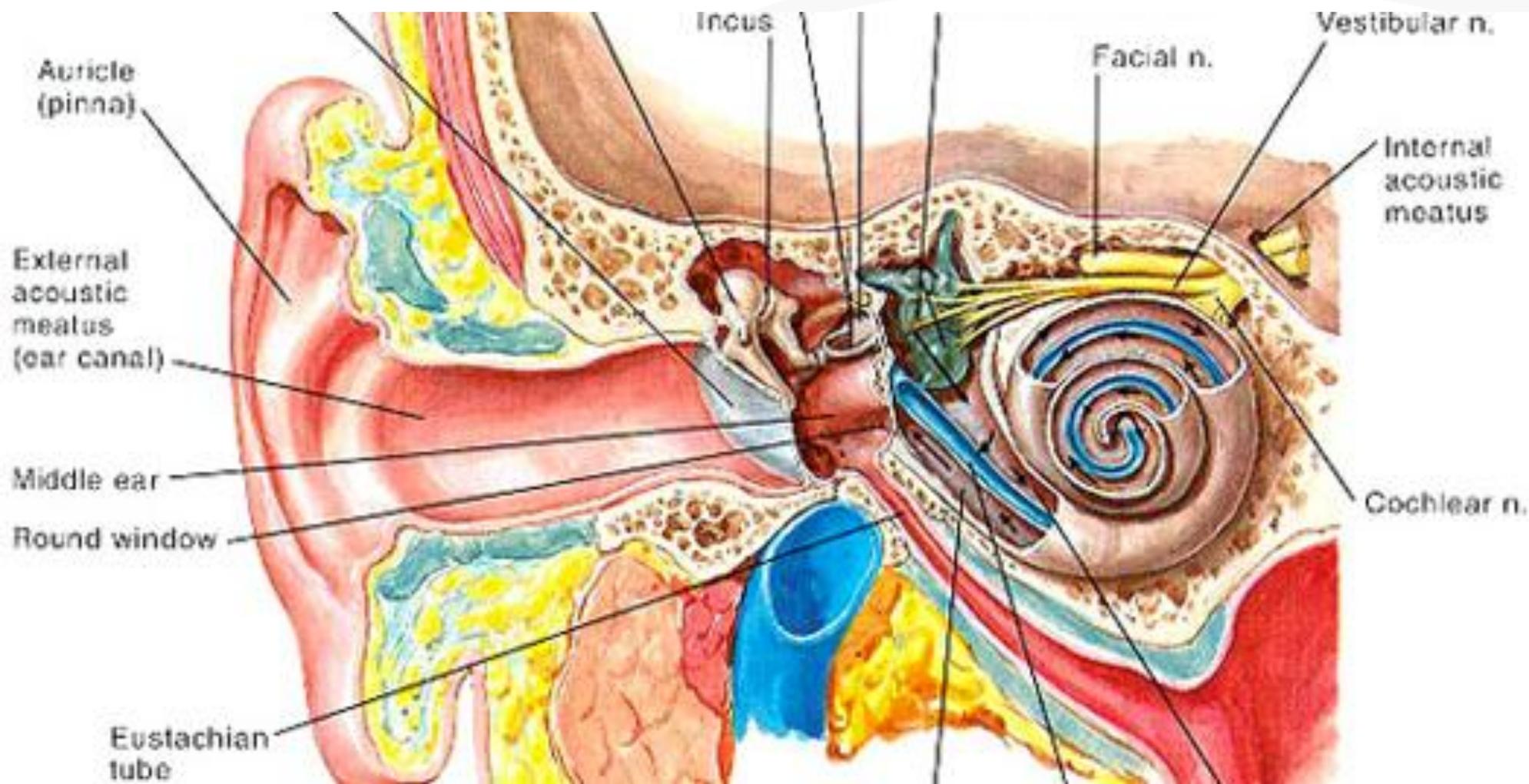
- Overview and Definitions
- Variations in Presentation
- Implications for Clinical Practice
- Case Studies

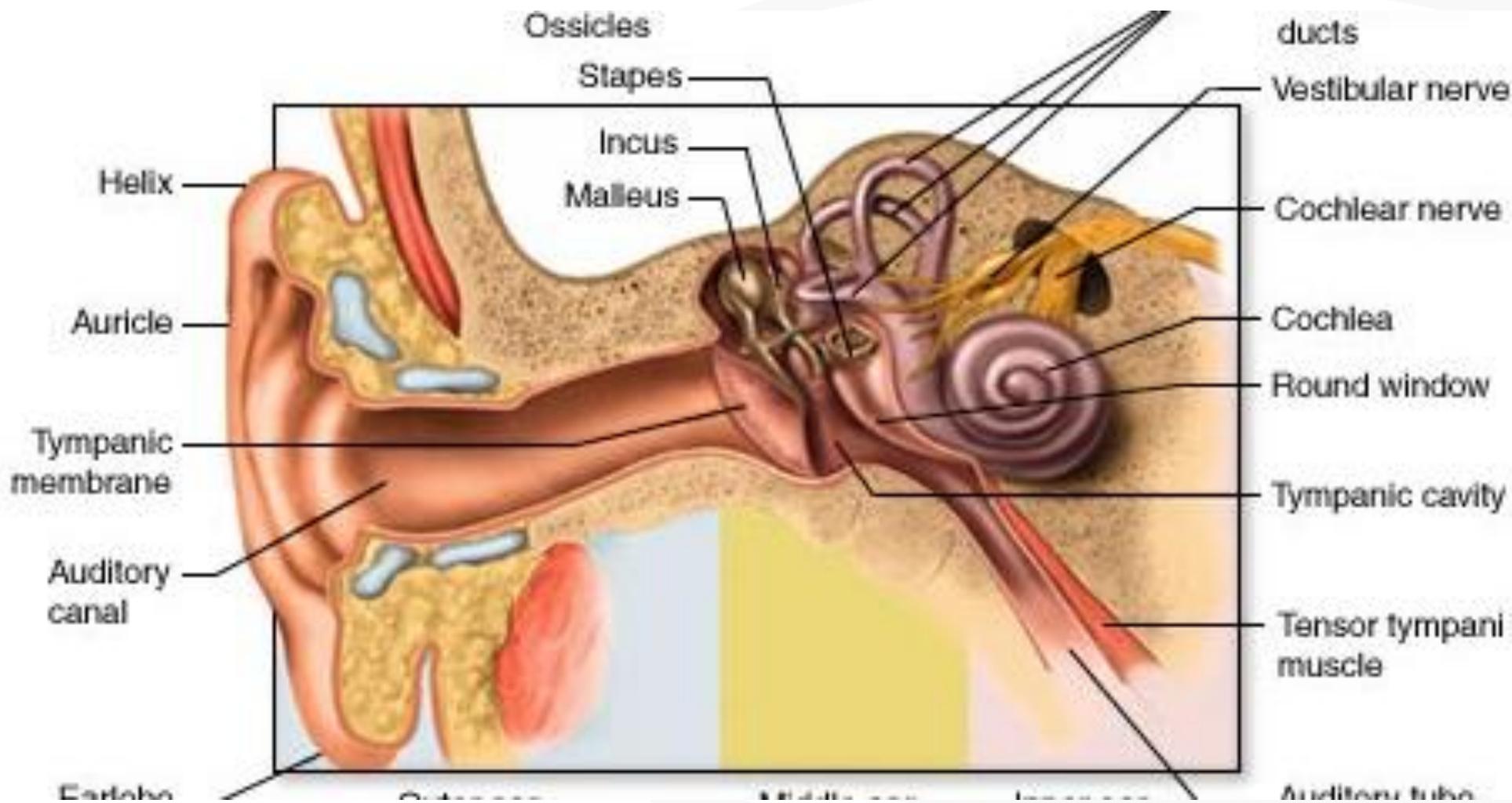
Auditory Neuropathy: A Definition

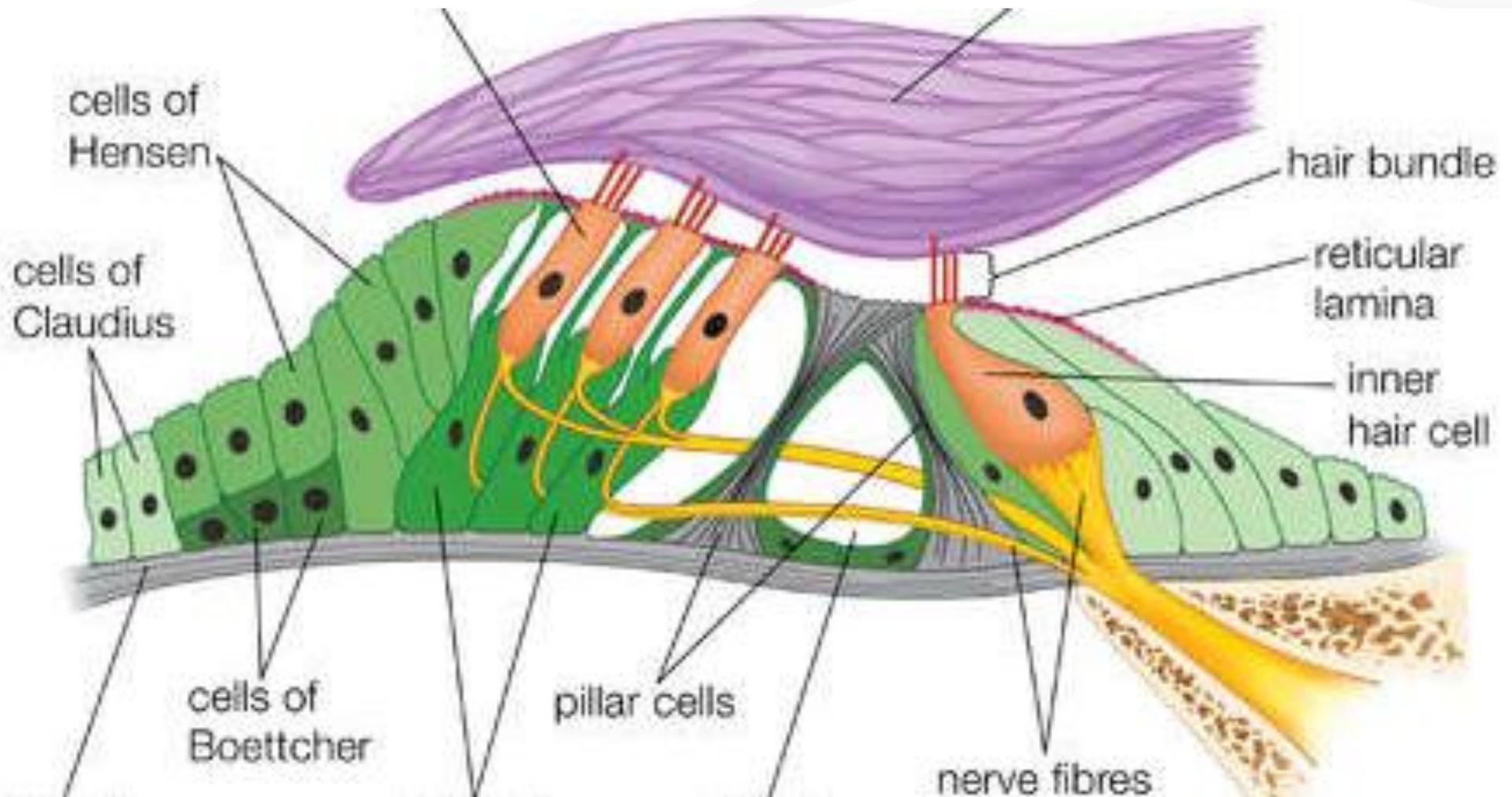
Clinical syndrome characterized by electrophysiological evidence of normal or near normal cochlear function and absent or abnormal auditory pathway transduction

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, or be absent at time of diagnosis
- 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 ANSD experience the same problems or to the same degree**

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

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)

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”

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

- 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?

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

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 than helpful

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

Lake Como, Italy, June 19-21, 2008

Guidelines available at:

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

(Denver Children's Hospital Website)

Panel Members

- Gary Rance
- Christine Petit
- Barbara Cone
- Deborah Hayes
- Charles Berlin
- Patricia Roush
- Yvonne Sininger
- Jon Shallop
- Kai Uus
- Arnold 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

**Guidelines Development Conference:
Identification of Infants and Children with Auditory Neuropathy
Lake Como, Italy, June 19-21, 2008**

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

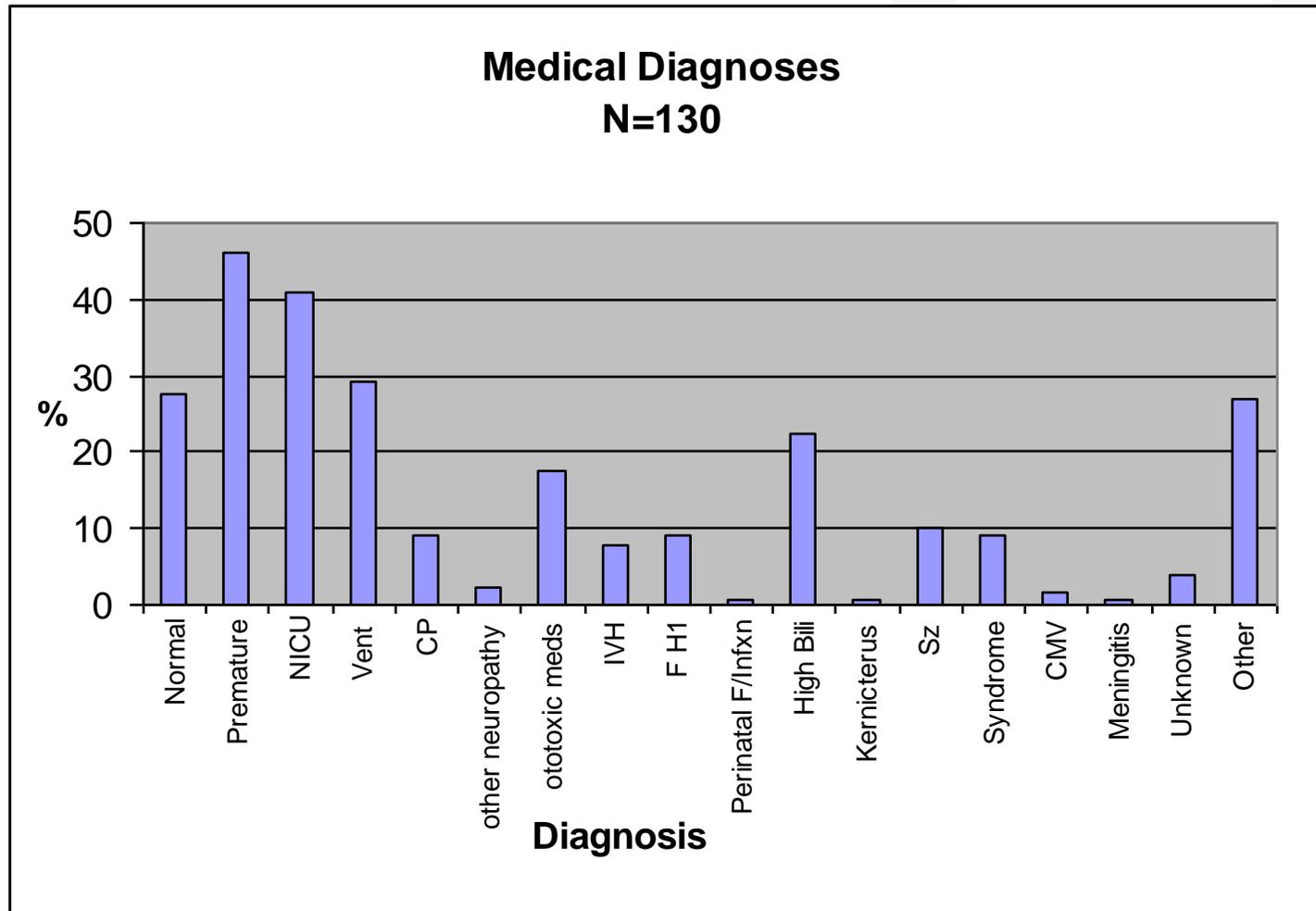
- **Minimum Test Battery Required for Diagnosis:**
 - » Tests of cochlear hair cell (sensory) function:
 - Otoacoustic emissions and/or
 - Cochlear microphonics
- **Tests of auditory nerve function:**
 - » Click-evoked auditory brainstem response (ABR) to high-level click stimuli



Comprehensive Evaluations Following Diagnosis with ANSD

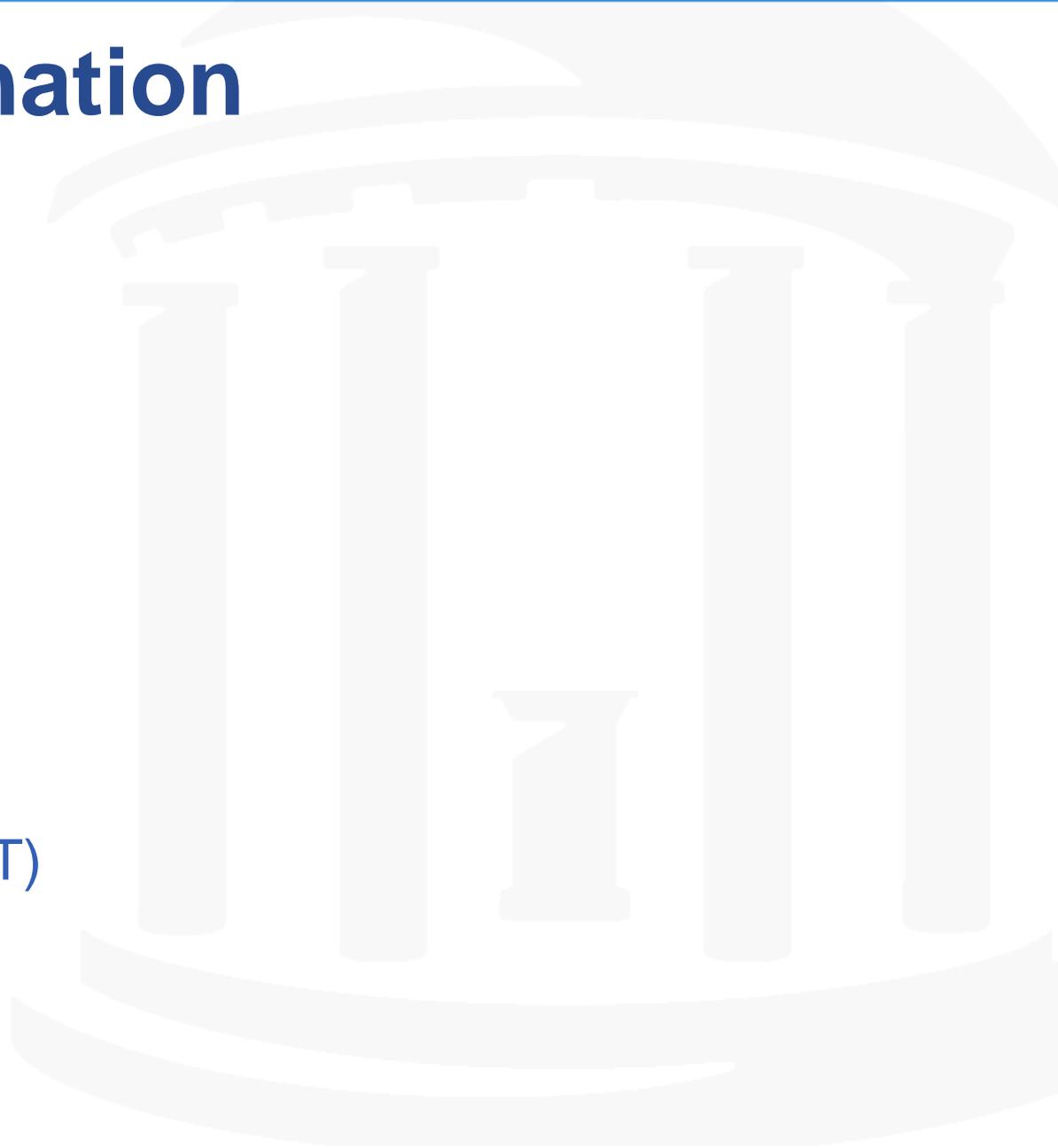
- Otologic
- Radiologic imaging (MRI/CT)
- Neurologic
- Medical Genetics
- Ophthalmologic
- Pediatric and Developmental Evaluations
- Communication Assessment

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





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DIAGNOSIS OF AUDITORY NEUROPATHY

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

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
- Therefore, ASSR cannot be used to predict behavioral thresholds in cases of ANSD

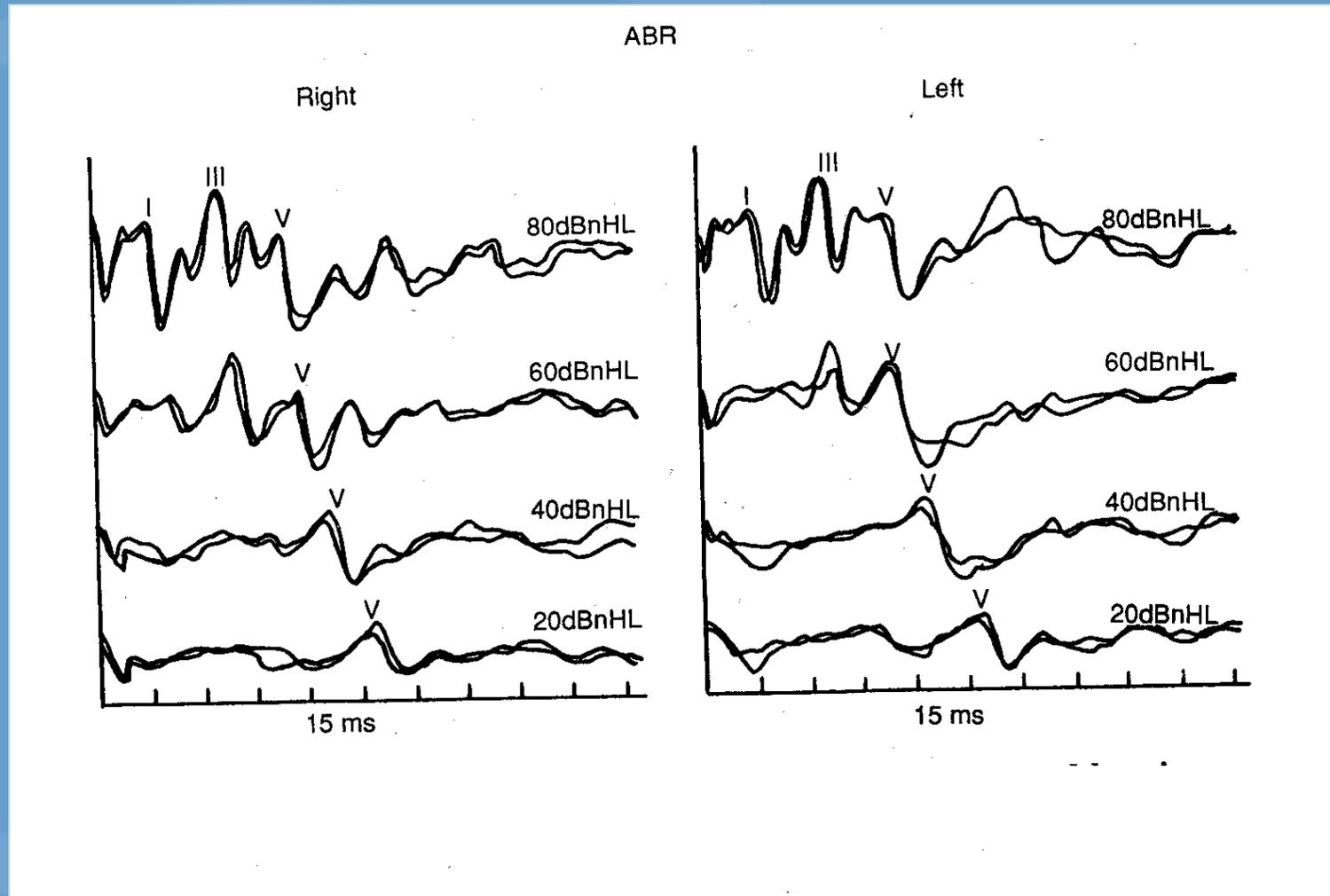
(Jahari et al, 2009, Attias et al 2006, Rance et al 1998, Rance & Briggs, 2002)

Recommended Audiologic Test Battery

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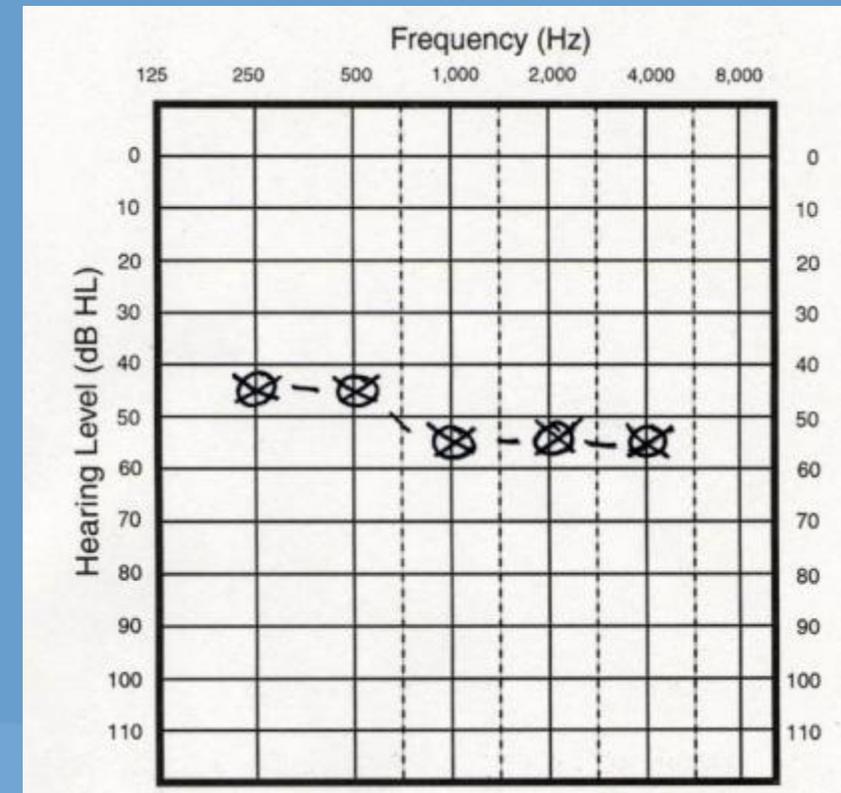
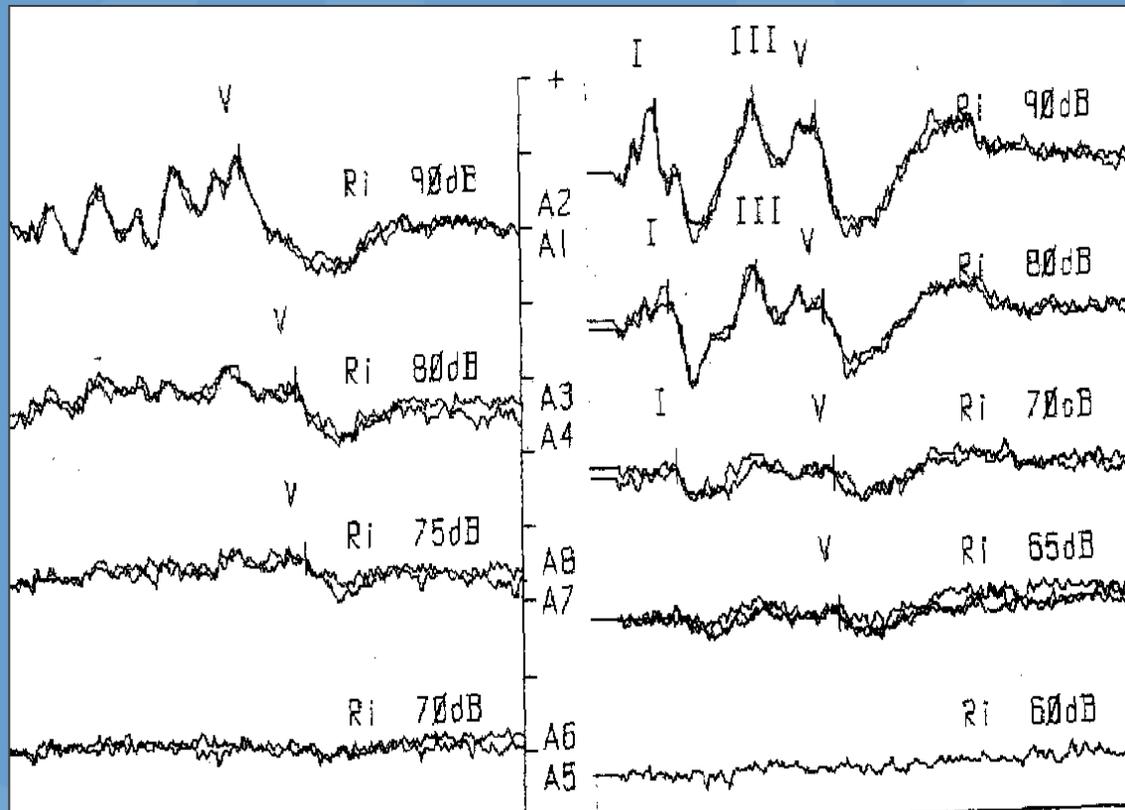


Normal ABR



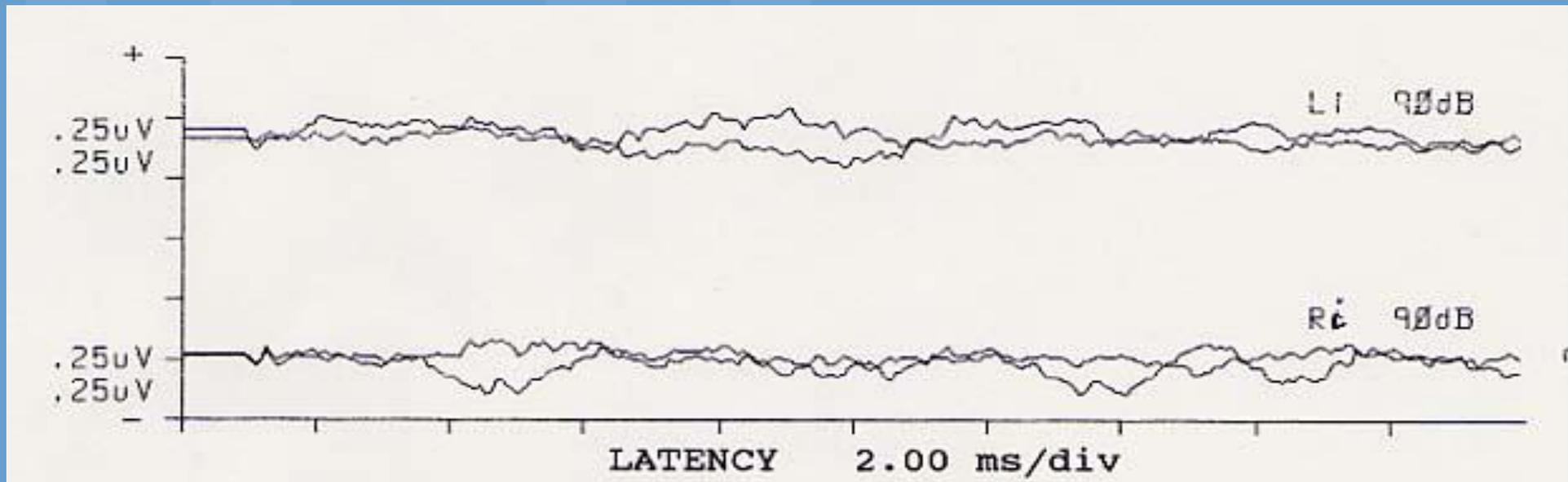


Estimating the Audiogram from ABR in non-AN type hearing loss



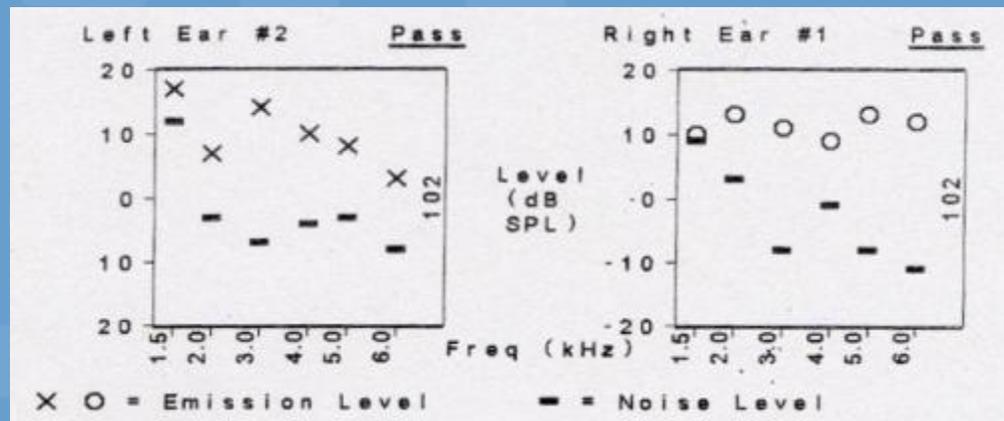
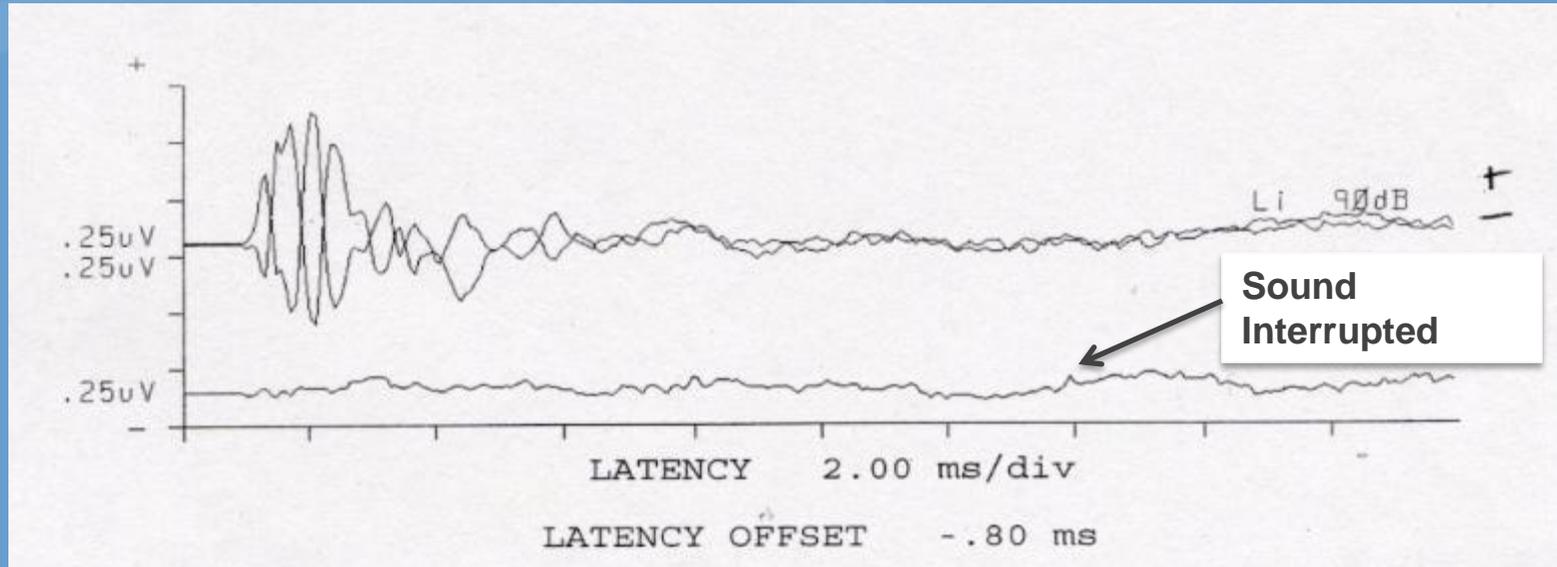


Absent ABR with No Cochlear Microphonic: Child with profound hearing loss



Typical Pattern of ANSD

Abnormal ABR with Present CM

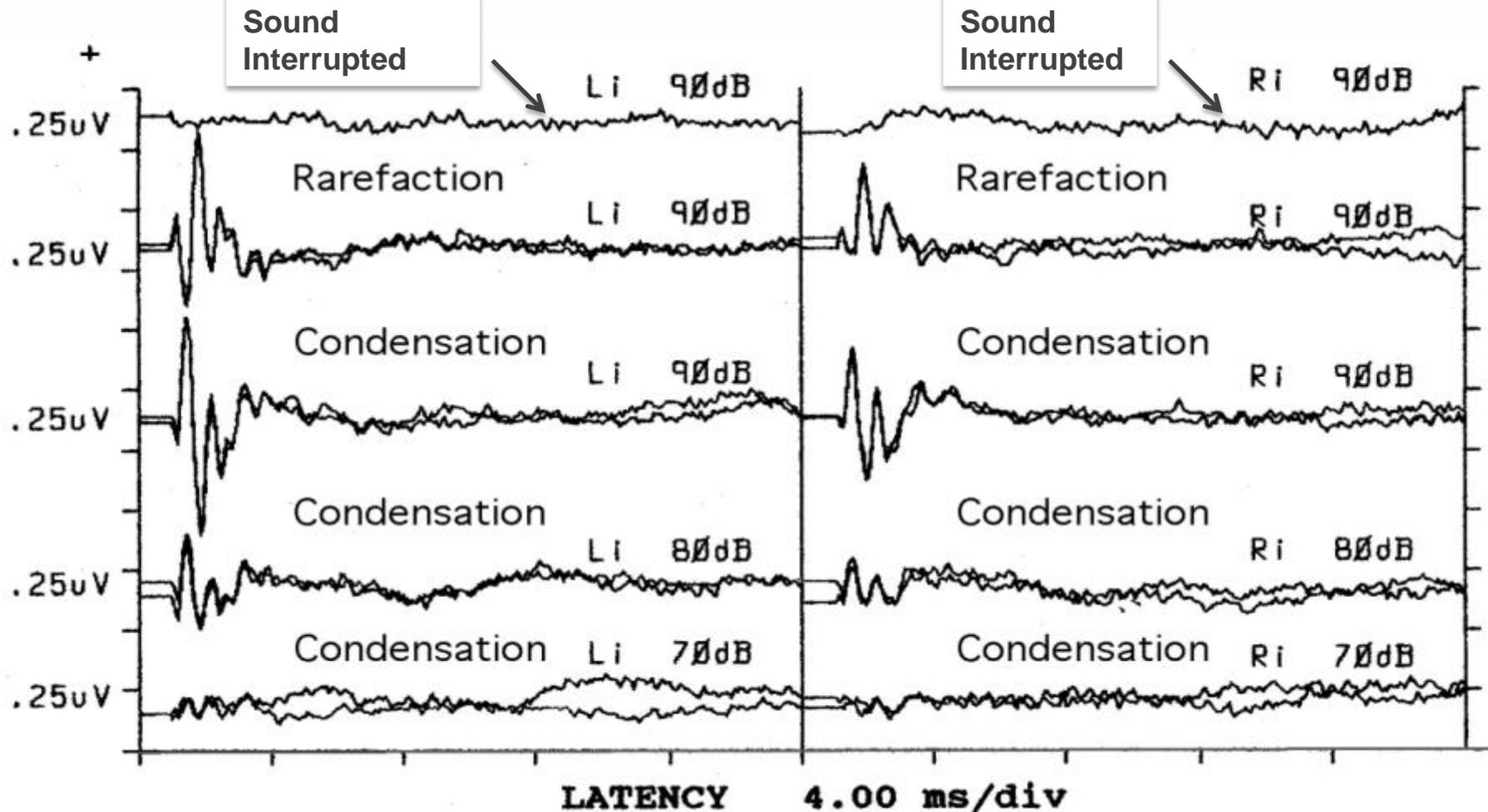


What is a Cochlear Microphonic (CM)?

- 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



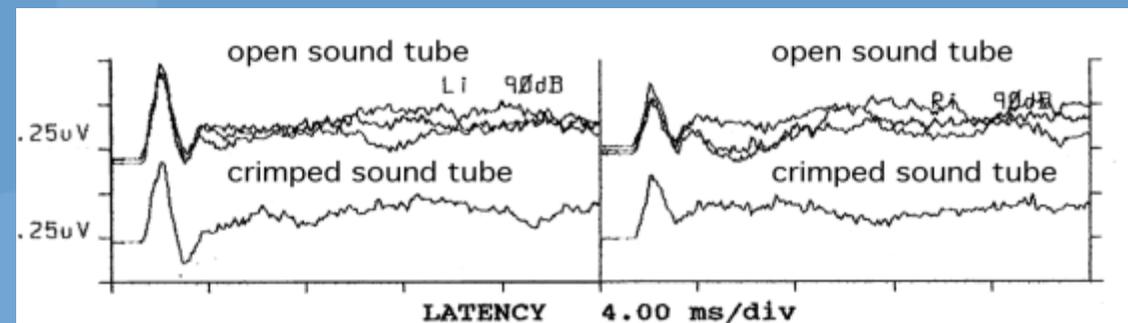
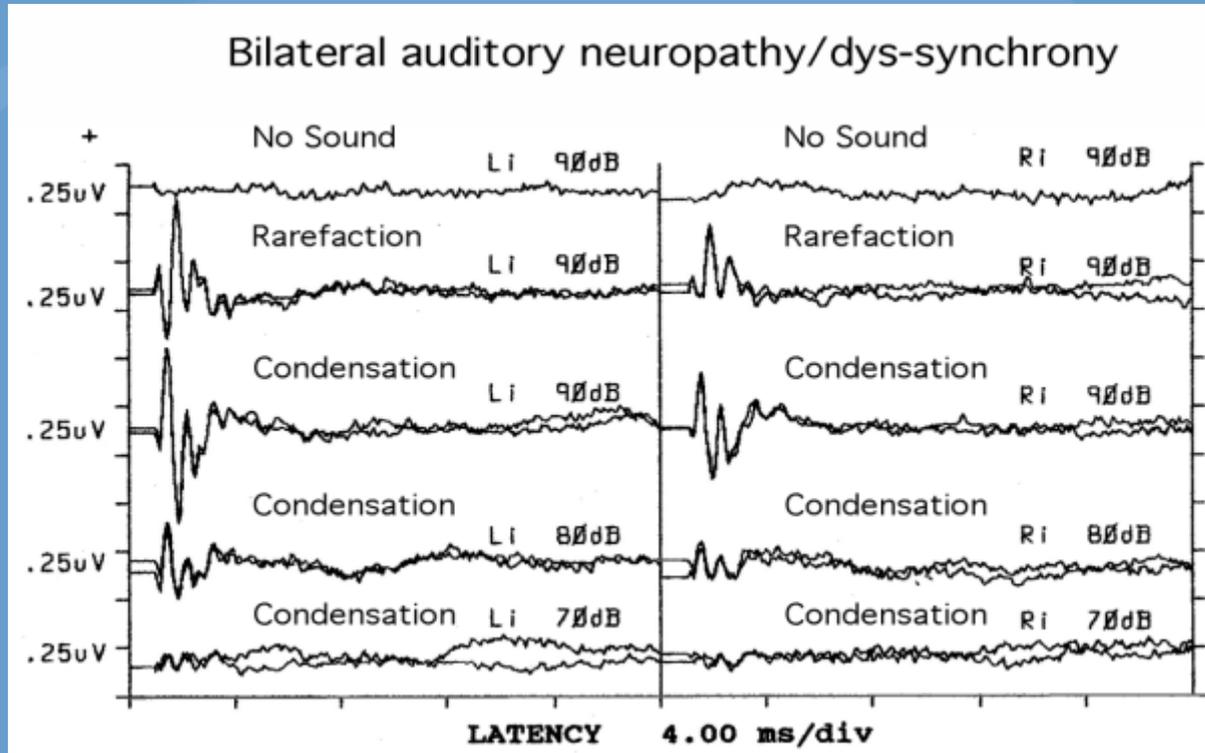
Bilateral auditory neuropathy/dys-synchrony



ABR Protocol for Evaluating CM

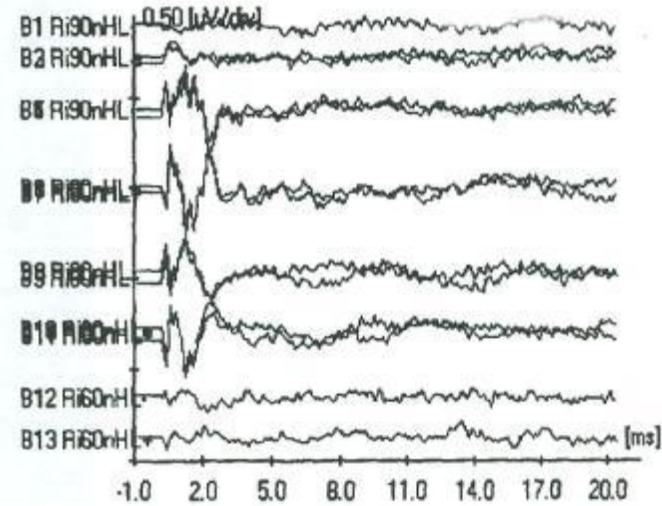
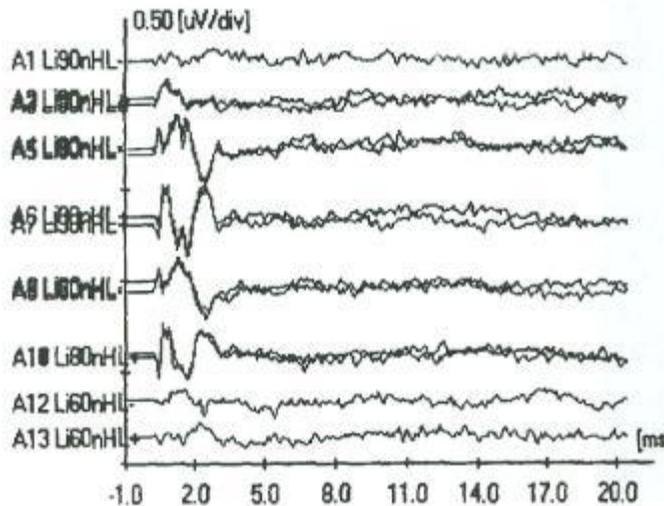
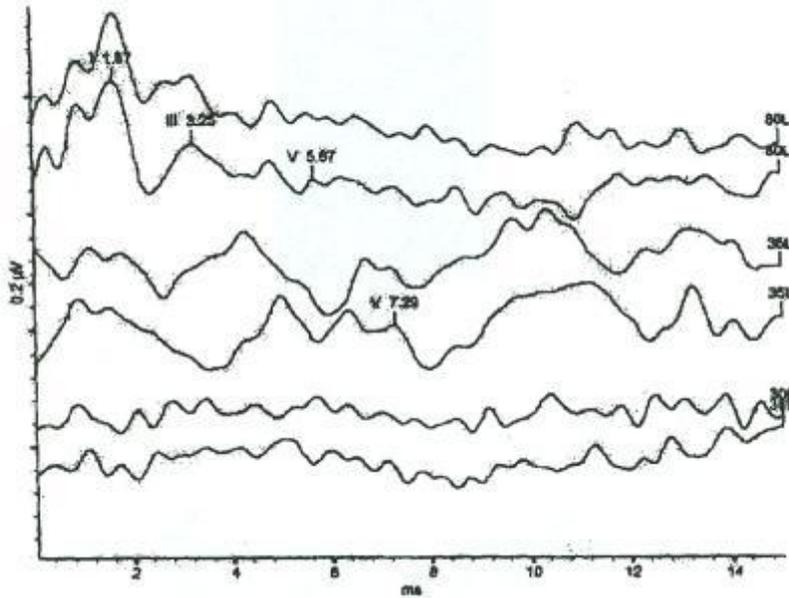
- Must have adequate recording conditions
 - Infant ready to sleep
 - Avoid electrodes positioned over transducer
- *Single polarity* clicks at 80 & 90dBnHL with rarefaction and condensation polarities
- Must use insert earphones
 - Excessive stimulus artifact with standard headphones obscures cochlear microphonic
- Sound interrupted run with stimulus on but sound tube disconnected or clamped to check for stimulus artifact

CM vs stimulus artifact





Example of Child with ANSD
incorrectly diagnosed with
normal hearing
~Note incorrect identification
of waveforms on left



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 internal auditory canal when cochlear nerve is absent
- In cases when there is question of CND both CT and MRI imaging may be needed
- Imaging is especially important when behavioral audiometry shows profound 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

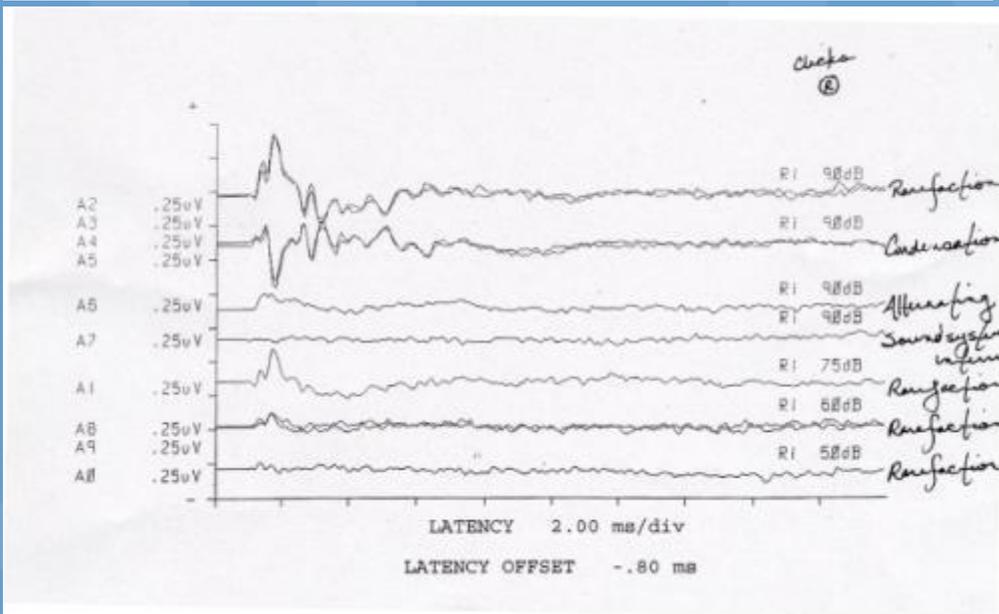
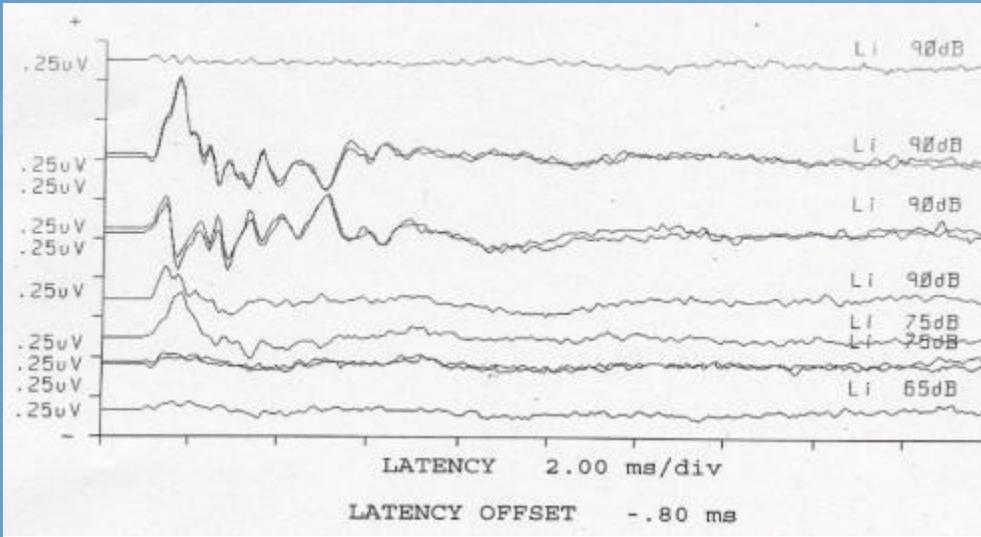
- **Absent 8th Nerve**
 - not uncommon
 - can result in auditory neuropathy phenotype
 - commonly has normal internal auditory canal (IAC) morphology
 - commonly has normal labyrinth
- **Need MRI instead of CT in all kids**
 - with profound hearing loss
 - with auditory neuropathy phenotype
 - Audiological,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



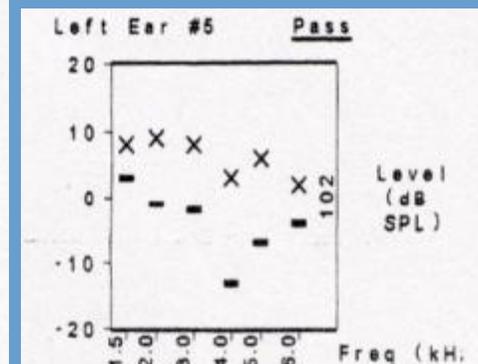
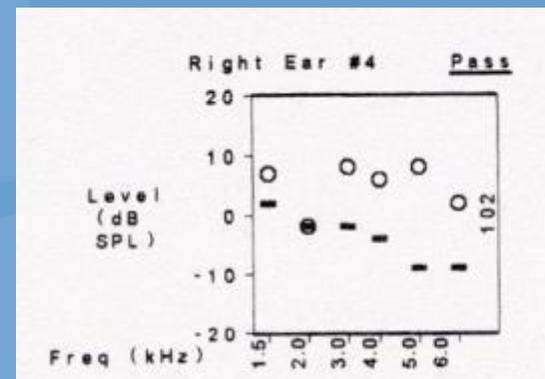
Variable Presentations of ANSD

Case Examples

Case #1: Present CM and OAEs

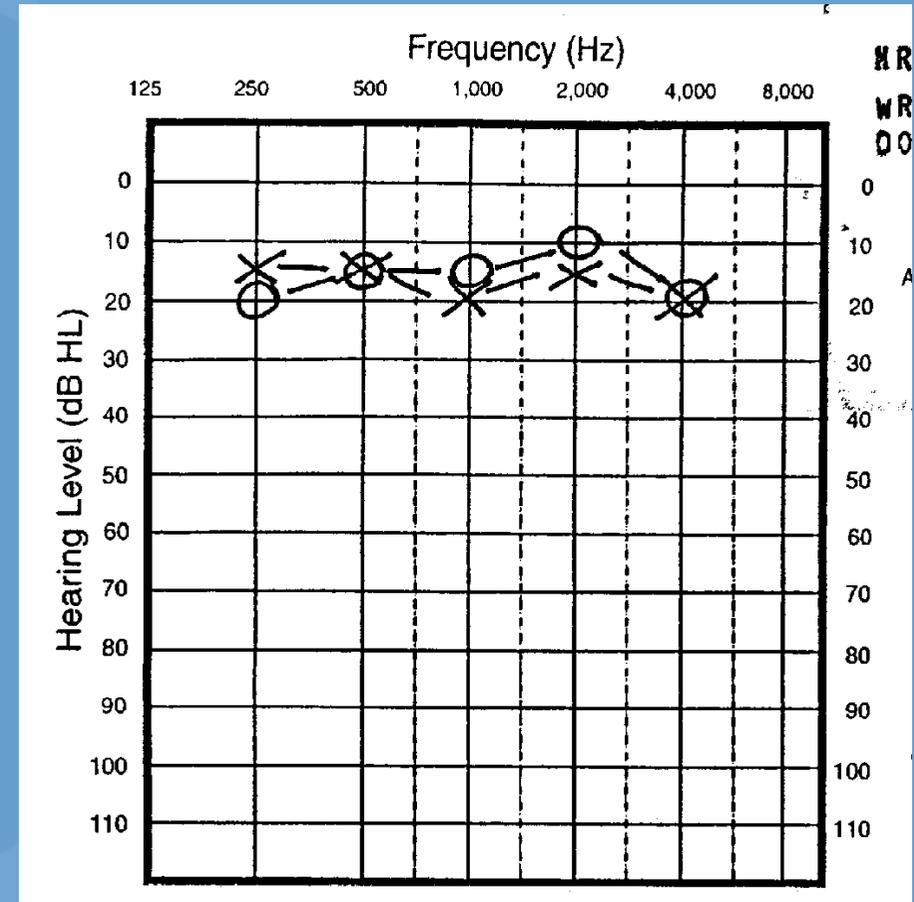
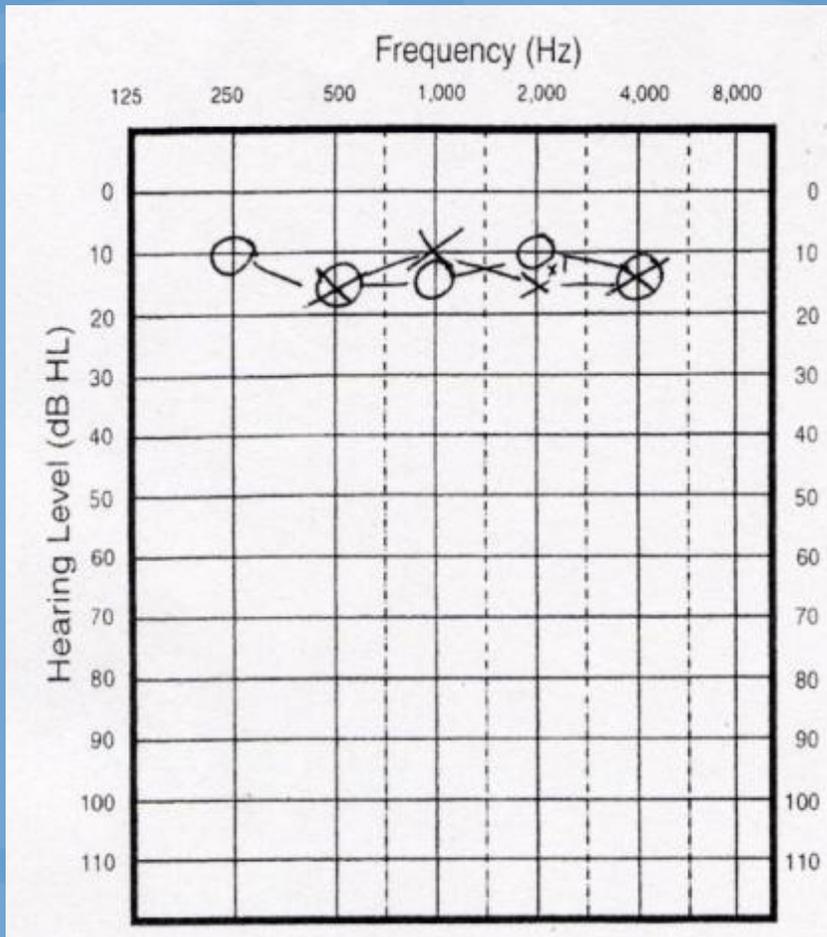


- 24 wk preemie, 940 grams
- NICU 4 months, ventilated
- ABR at 4 and 5 months of age abnormal
- ABR repeated at 18 months-no change



Case #1

Normal thresholds, Present CM and OAEs



Audiogram at 14 months

Audiogram at 18 months

Case #1

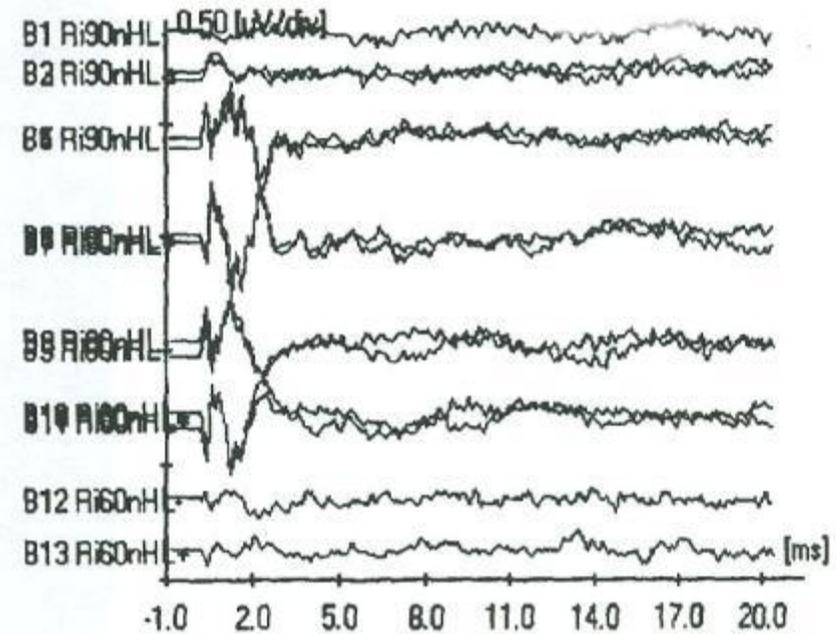
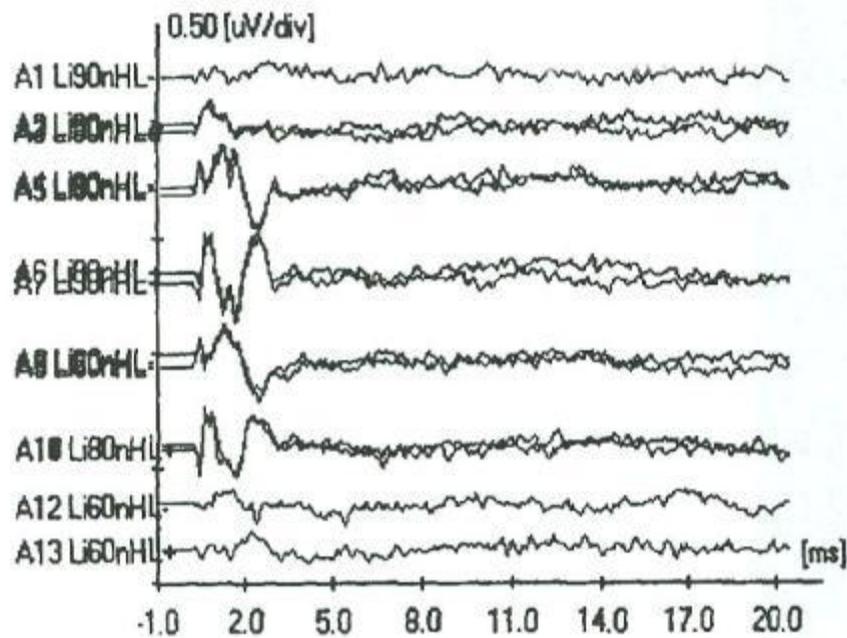
Speech Perception Test Results

- Age 2 yrs-11 months:
 - ESP monosyllabic word test (closed set test of speech perception):
 - 12/12 correct for each ear at 50dBHL
- Age 5 years:
 - PBK words: 80% and 84% at 60dBHL for right and left ears



Case #2

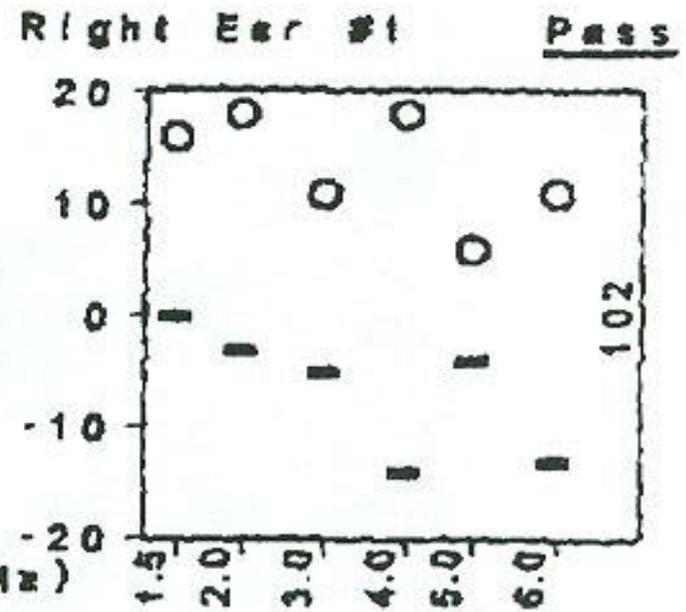
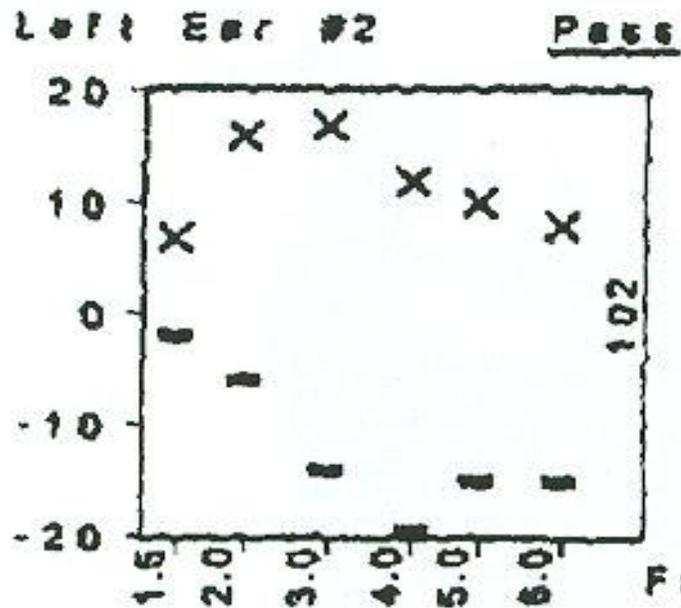
Abnormal ABR with Present CM





Case #2

Present OAEs

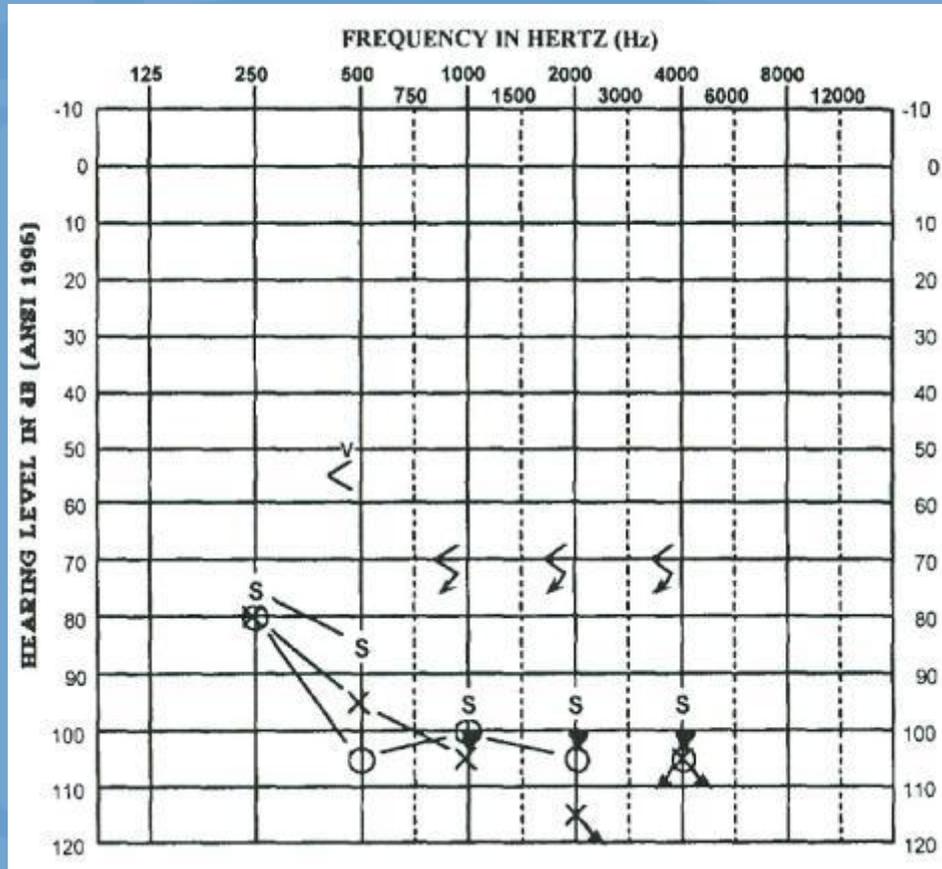


Case #2

Child with Profound Bilateral HL
Present CM and OAEs



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Ear exam: *Normal*

EKG: *Normal*

MRI: *Normal*

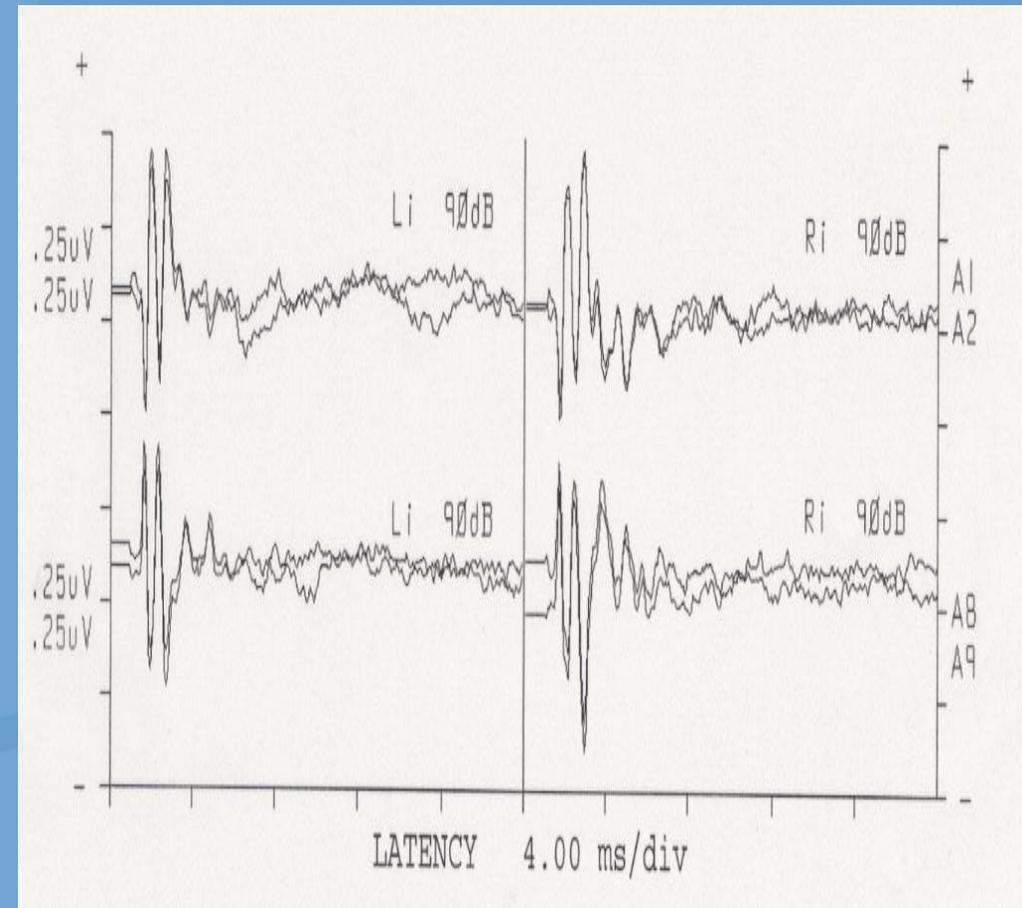
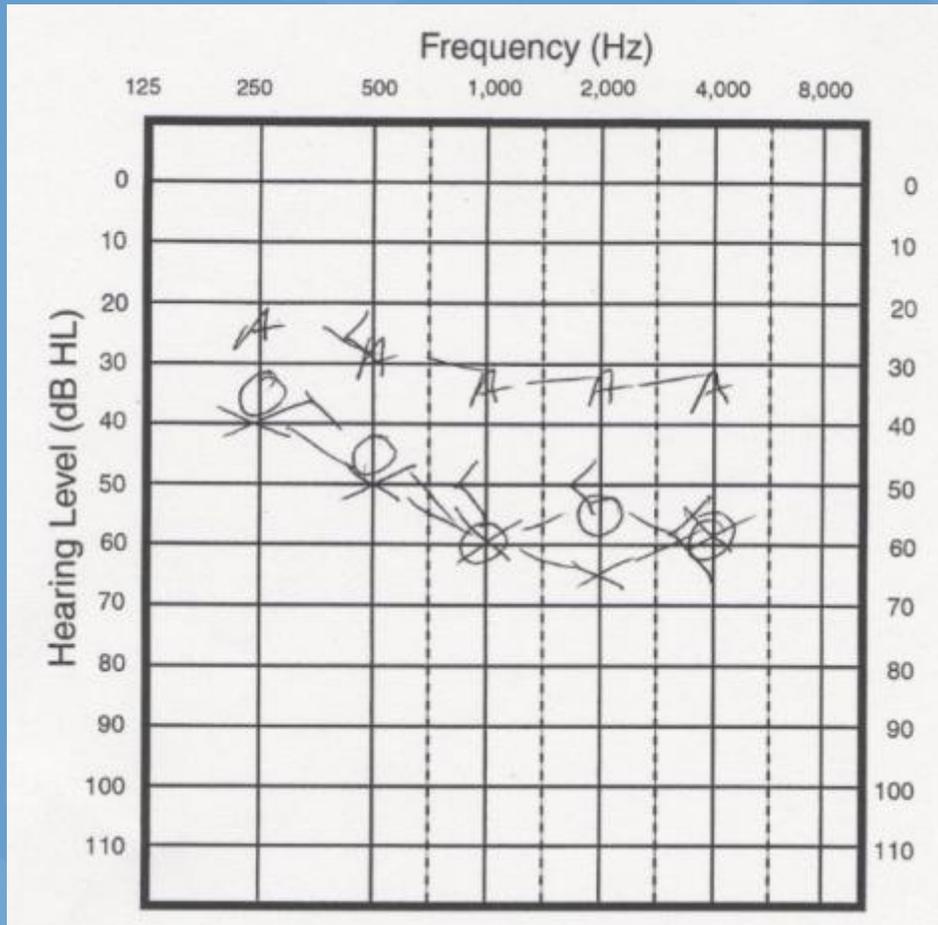
Connexin test: *Negative*

Otoferlin test: **POSITIVE**

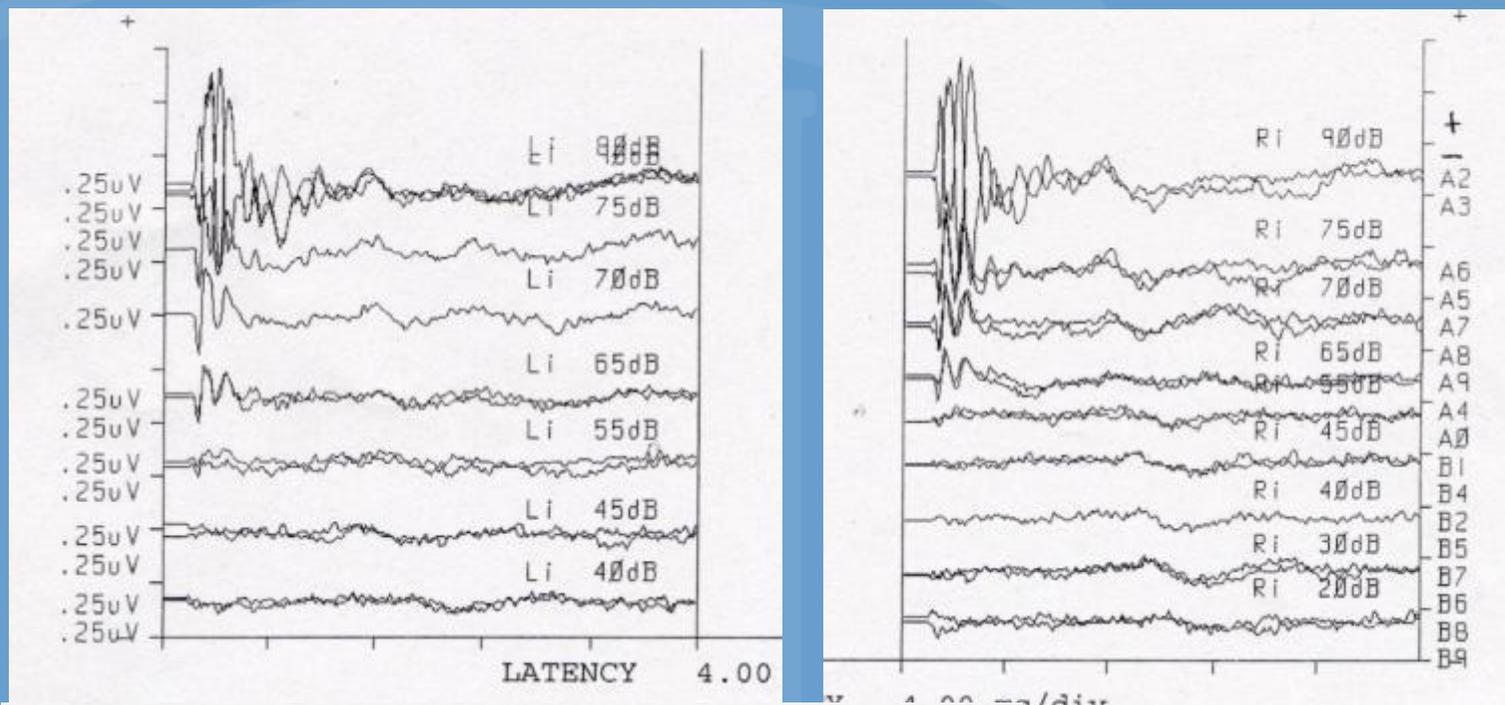
Received CI at 24
months of age (Late
diagnosis)

Case #3

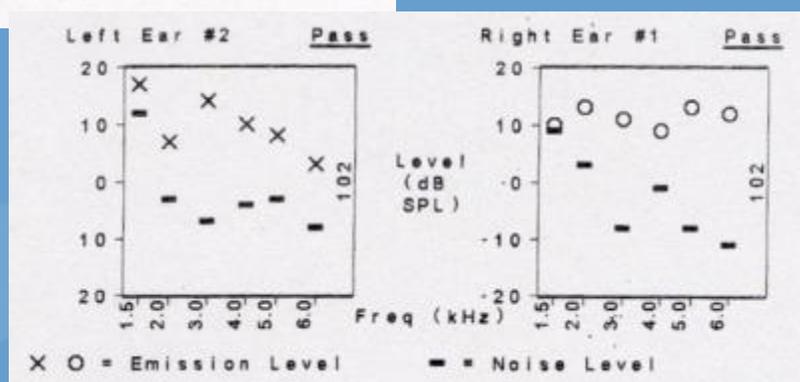
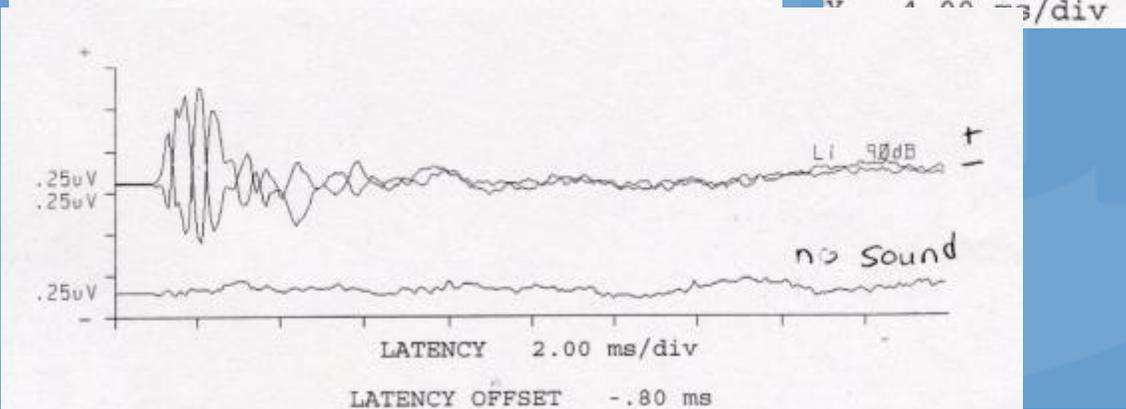
Child with “moderate loss” CM present, absent OAEs



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



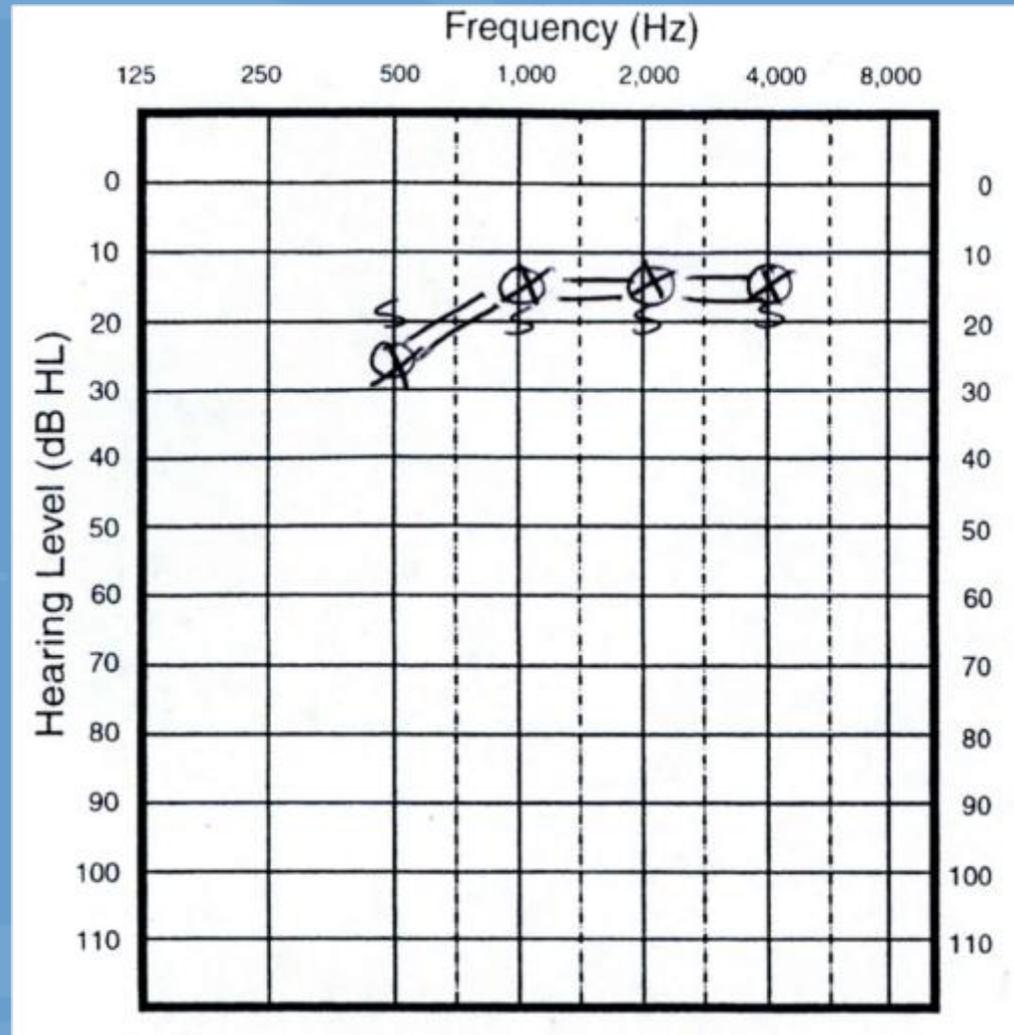
Caution needed when interpreting ABRs that show abnormal waveform morphology at high intensity levels



Case #4 (continued)

VRA with insert earphones

Age 14 months



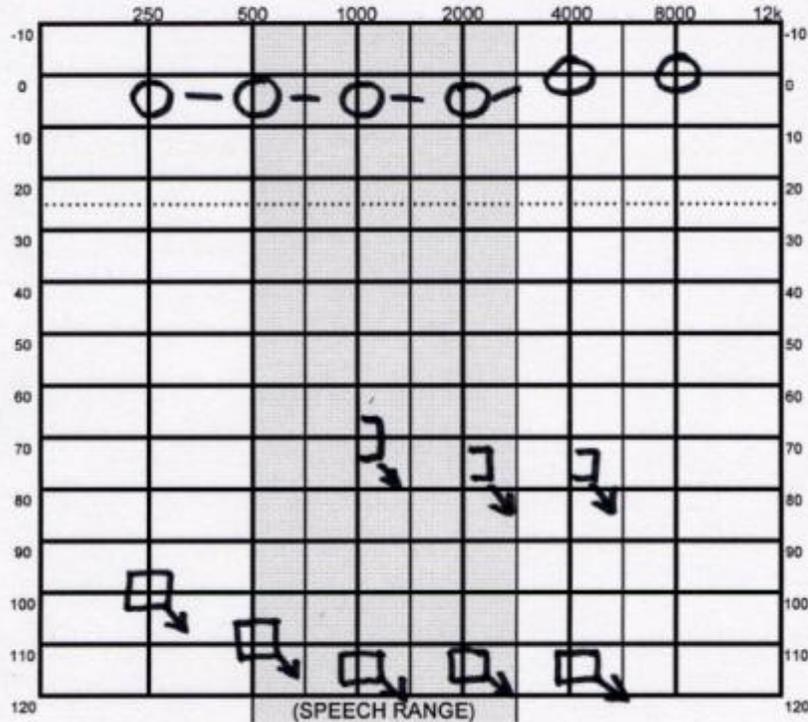
Case #5

- 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



PURE TONE AUDIOMETRY (RE: ANSI 1969)



LEGEND	RIGHT (RED)	LEFT (BLUE)
Air	O	X
Air (Ov. Ear Masked)	Δ	□
Bone	<	>
Bone (Ov. Ear Masked)	[]
No Response	↓	↓
SOUND FIELD	S	
AGED	R	L

TEST	STANDARD
PLAY	
COR/VRA	
BOA	

SITE	BOOTH	OTHER

RELIABILITY	EXCELLENT

TYMPANOMETRY

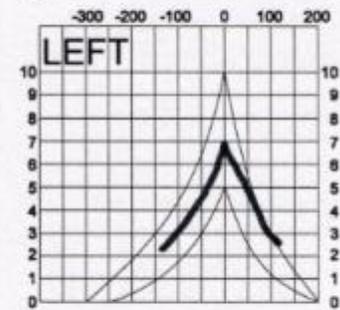
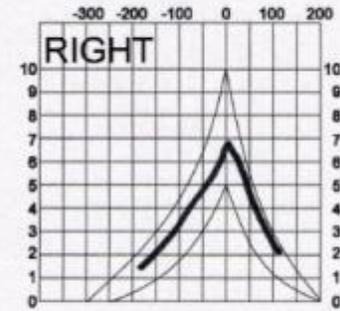
CONTRA (Phone Ear)	.5k Hz	1k Hz	2k Hz	4k Hz
RIGHT (AD)				
LEFT (AS)				

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

TYMPANOMETER: _____



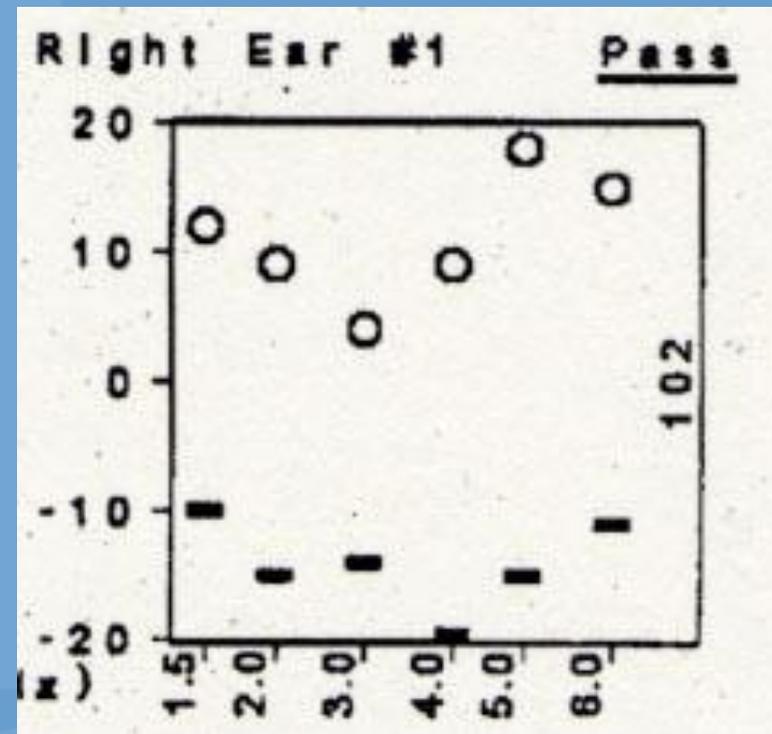
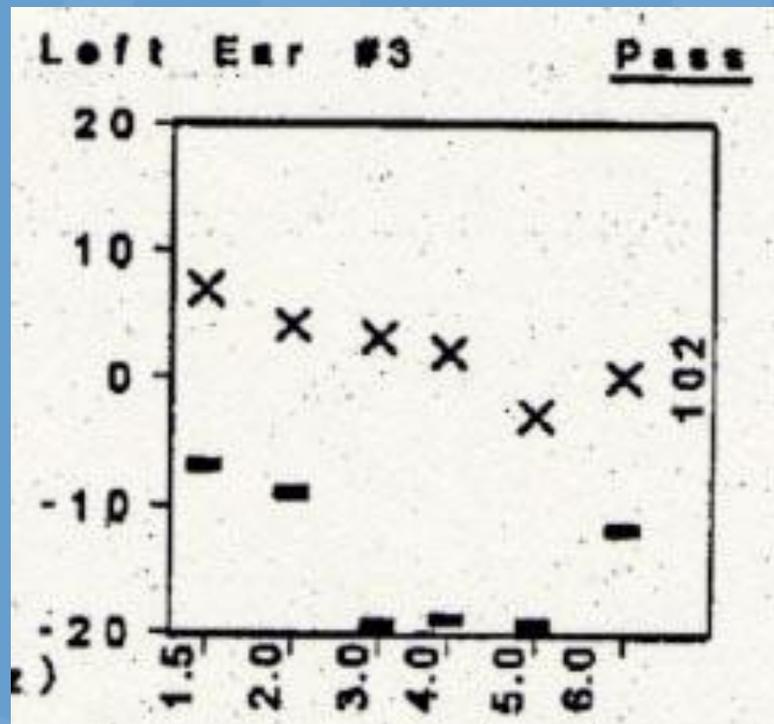
SPEECH AUDIOMETRY

	PTA	SRT/SAT	SPEECH RECOGNITION		SPEECH RECOGNITION		MCL	UCL
			%	dB HL	%	dB HL		
RIGHT (AD)		0	100	50				
masking								
LEFT (AS)		NR		CNT				
masking								

HISTORY: _____



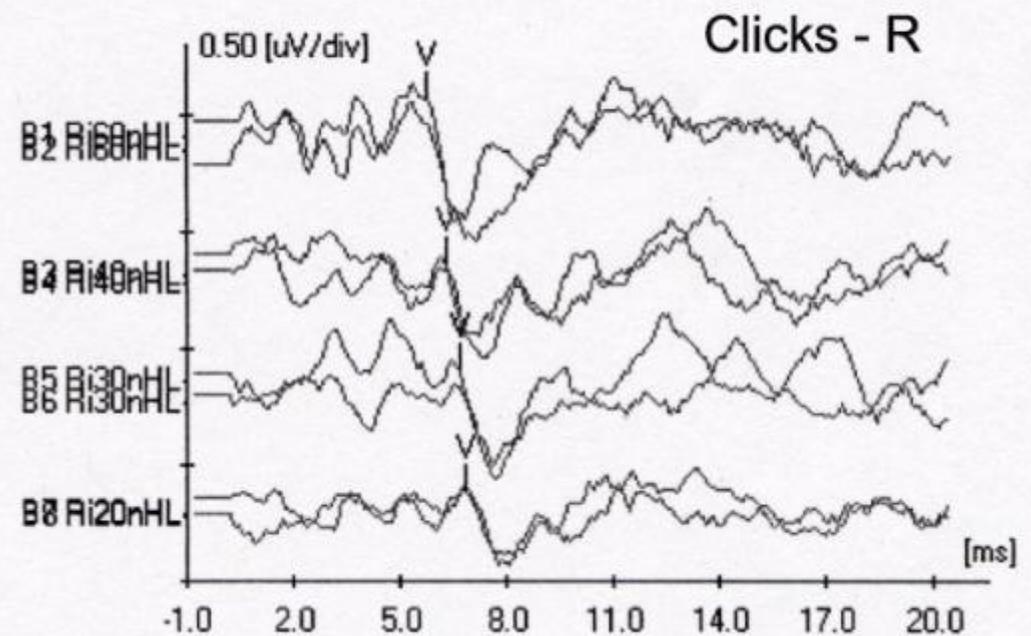
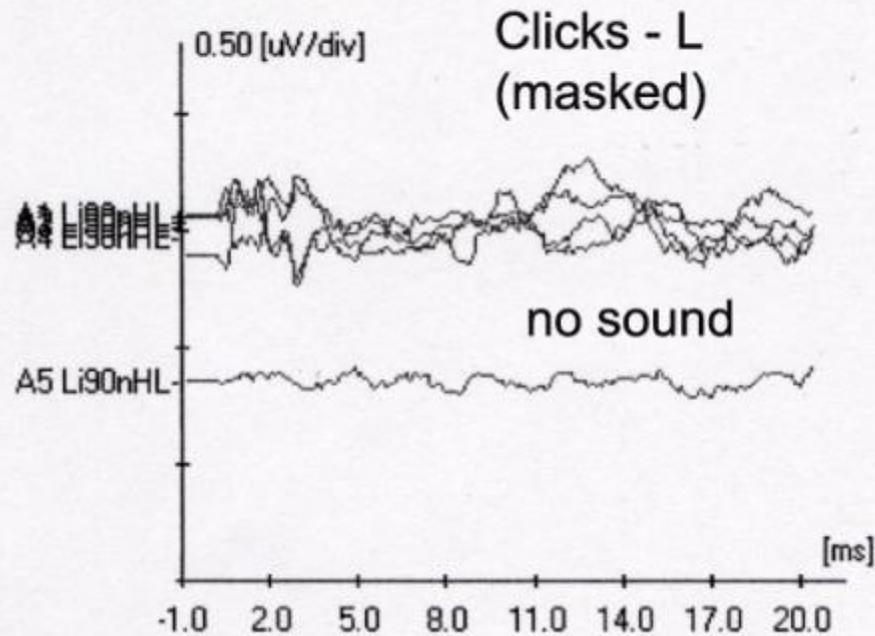
Case #5 OAEs





Case #5

ABR Completed at Age 4 years



Case #5

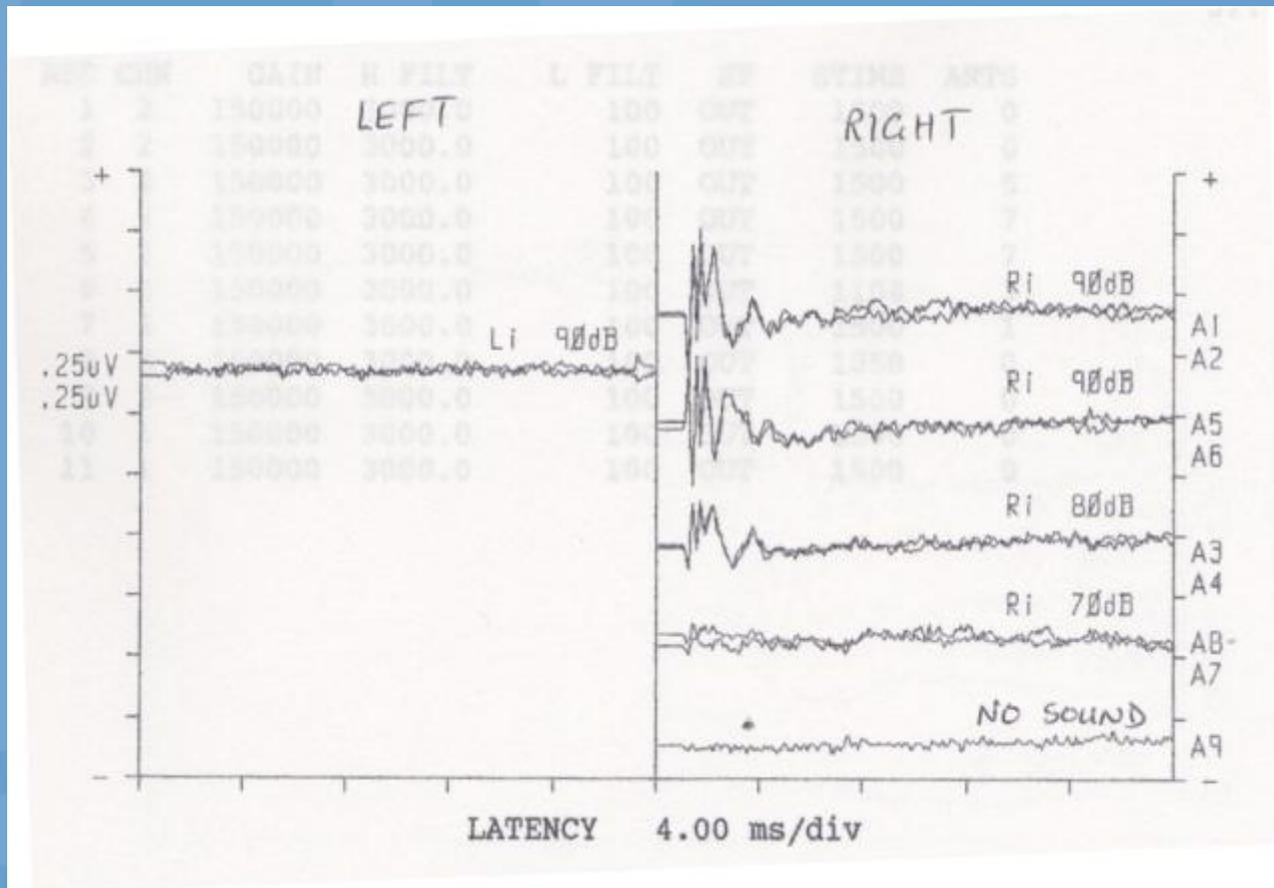
- Results of MRI:
 - » Consistent with small or absent auditory nerve
- At age 7 years child has above average speech and language development, no academic problems
- Managed as we do other cases with profound unilateral hearing loss.



Case # 6

Bilateral deafness

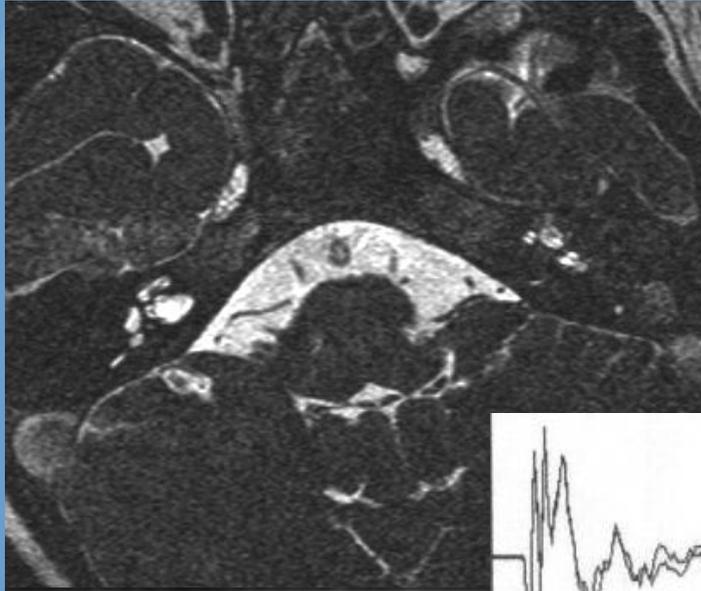
No VIIIth nerve on right



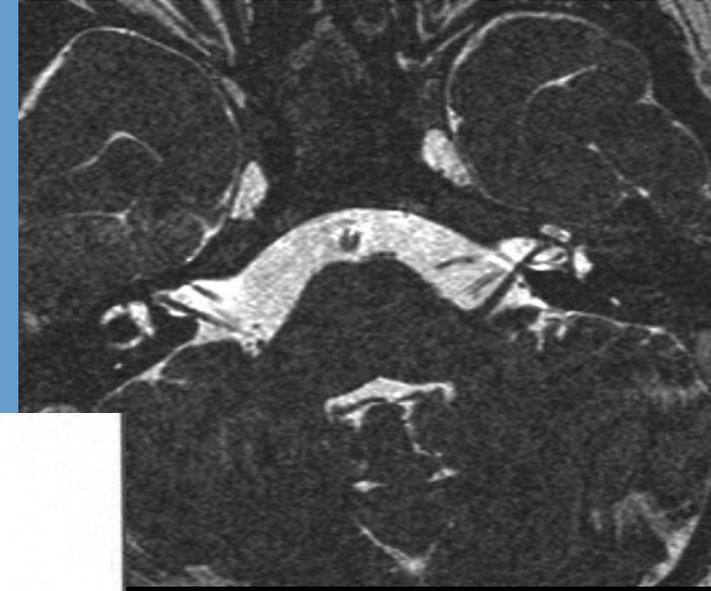
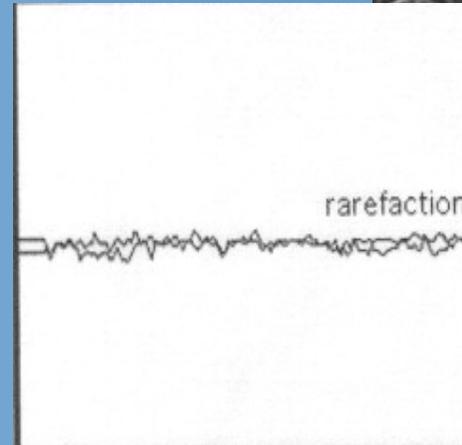
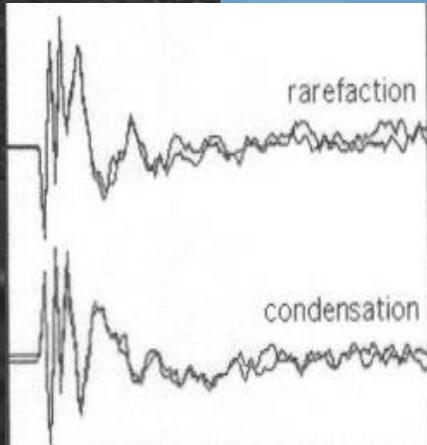
Case #6

Child with bilateral deafness

No VIIIth nerve on right



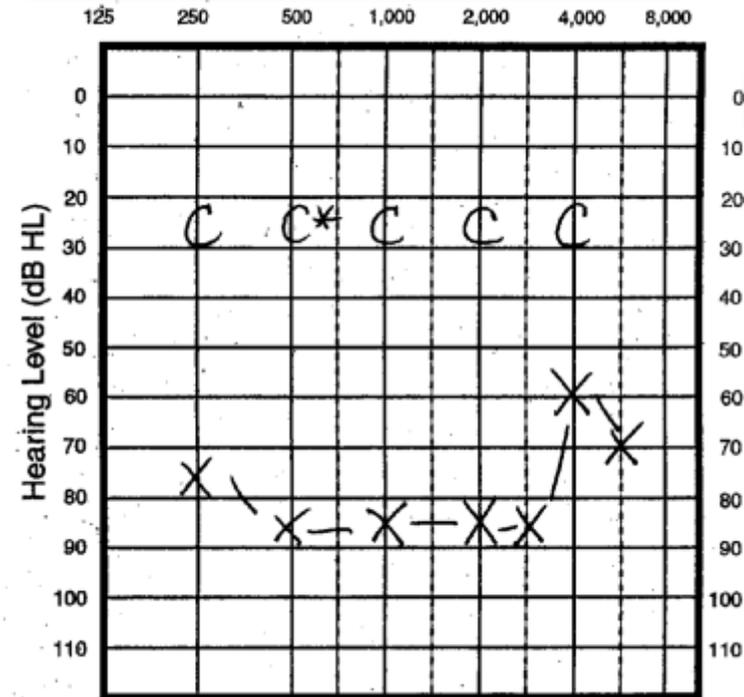
Right Ear



Left Ear

Case #7: 8 year old with ANSD

- Child with progressive neurologic disease
- Speech recognition scores 5 years post CI in right ear:
 - » 6 % words: 38% phonemes
- Recently began wearing HA again in non-CI ear
- Mom reports increased benefit compared to CI alone
- Many additional medical issues:
 - » Ataxic (in wheelchair now)
 - » Optic neuropathy (only sees at close range)
- Probably “true” neuropathy



Seven Cases with ANSD pattern on ABR...Seven Different Presentations

1. Normal hearing sensitivity no device needed, limited services required
2. Child with profound bilateral hearing loss; doing well with CI
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 nerve in one ear
7. Child with CI in one ear HA in other, struggling despite available technologies and interventions

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

ANSD Guidelines (Como 2008): Recommended Amplification Strategies

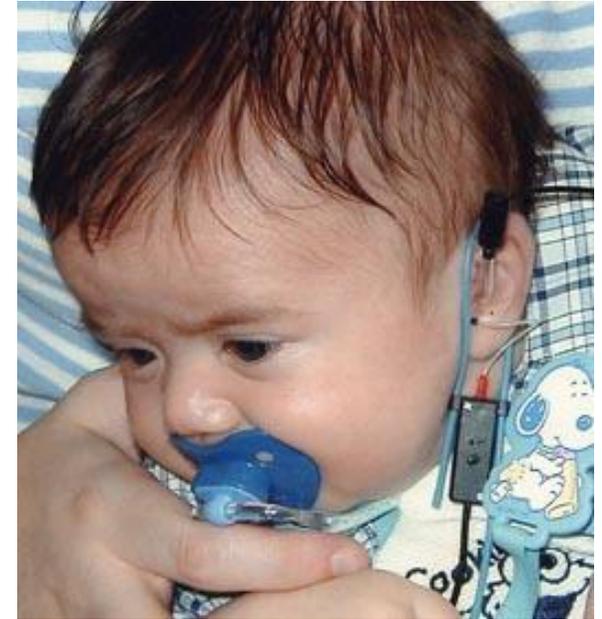
- Amplification should be fitted as soon as ear specific elevated pure-tone and speech detection thresholds are demonstrated by conditioned test procedures
- Hearing aid fitting strategies...should follow established guidelines for the fitting of amplification in infants and toddlers
 - » e.g. American 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.

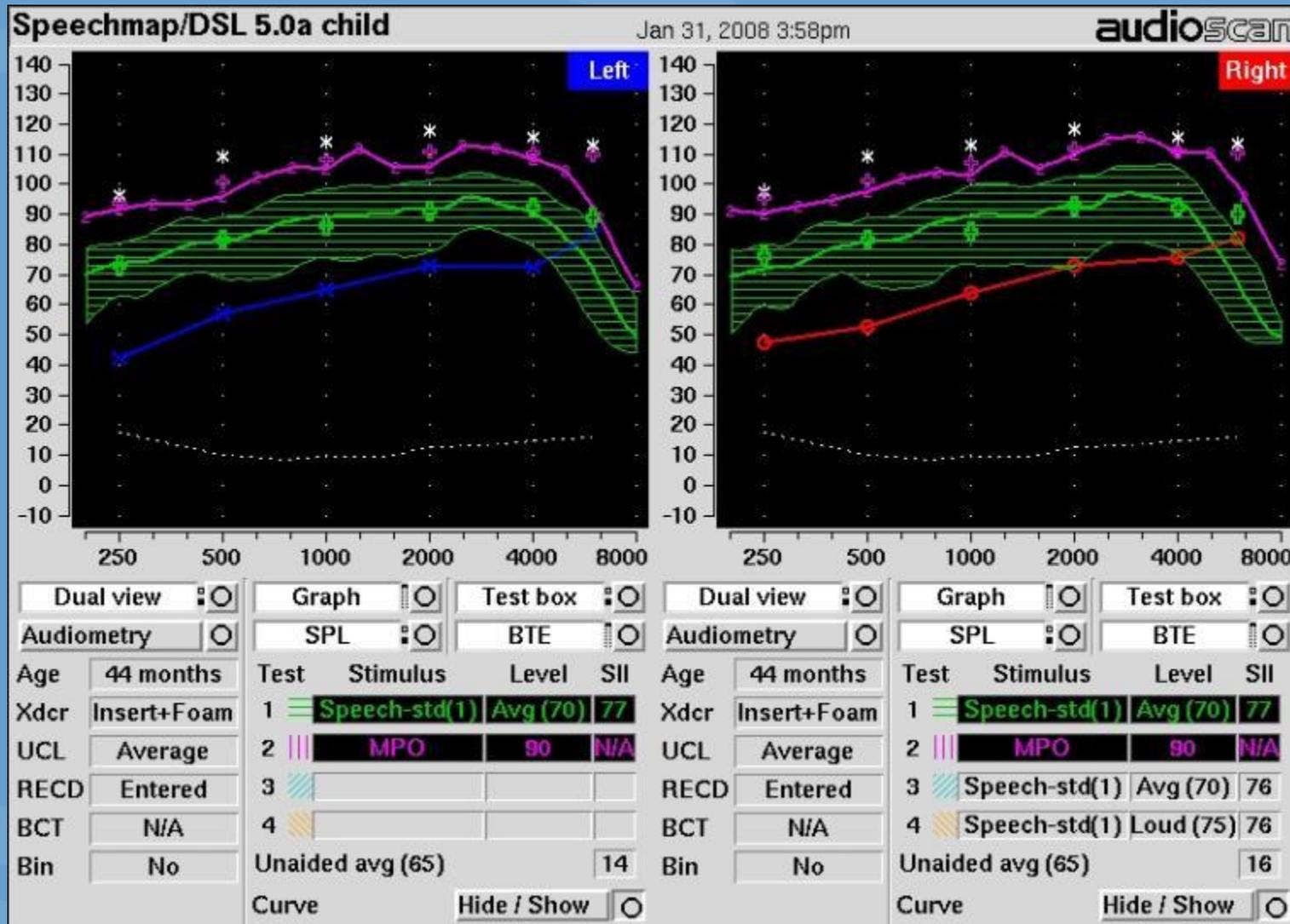
Hearing Aid Fitting Protocol for Infants

- 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)





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

Evaluation of Speech Perception Following Hearing Aid Fitting or Cochlear Implantation

- Parent Questionnaires (e.g. PEACH, IT-MAIS or MAIS)
(Ching and Hill, 2007, Zimmerman-Phillips, et al., 2000; Robbins, et al., 1991)
- Early Speech Perception Test battery (ESP) (Moog and Geers, 1990)
 - » Low Verbal
 - » Standard
- MLNT/LNT words and phonemes (Kirk, et al, 1995)
- PB-K words and phonemes (Haskins, 1949)
- HINT sentences in quiet and noise conditions

Early Speech Perception Test (ESP)



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



Weighing the Evidence: Hearing Aids, FM and Cochlear Implants

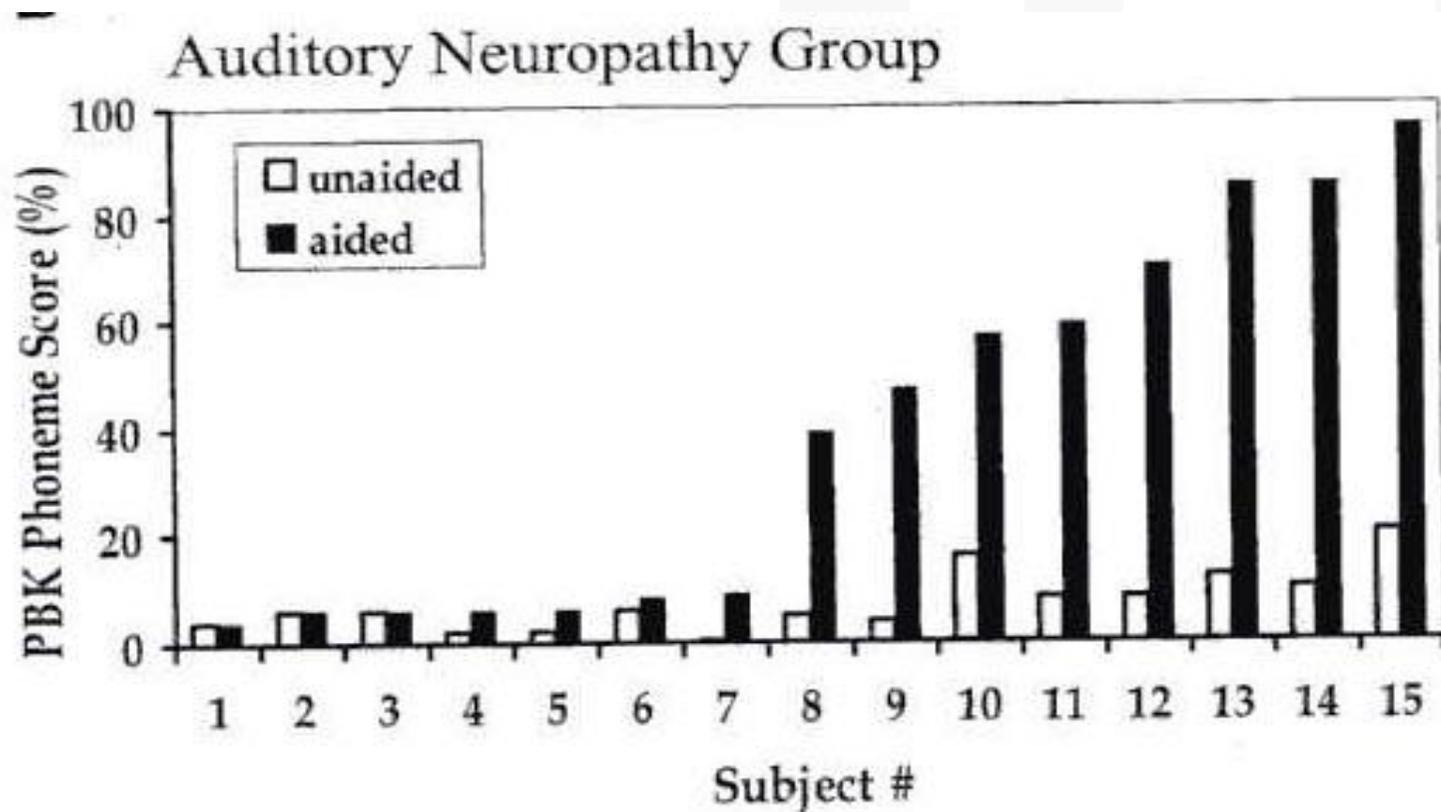
What does existing evidence tell us about clinical management?

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





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

B. Roush, P., Frymark, T., Venedictov, R., and Wang.,

[American Journal of Audiology 2011, Dec. 20\(2\): 159-70](#)

Evidence re Amplification

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



Counseling Families



Counseling in ANSD: What Do We Say to Families?

- Child has an auditory disorder; difficult to know prognosis at time of ABR evaluation
- 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 is helpful in some cases but not in others; benefit can only be determined with appropriate fitting and consistent use
- Cochlear implantation may be a better option if adequate benefit from amplification 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



Part II

What have we learned about ANSD at UNC?

How does this impact cochlear implant patient candidacy considerations and counseling?

How do we manage cochlear implants in children with ANSD?

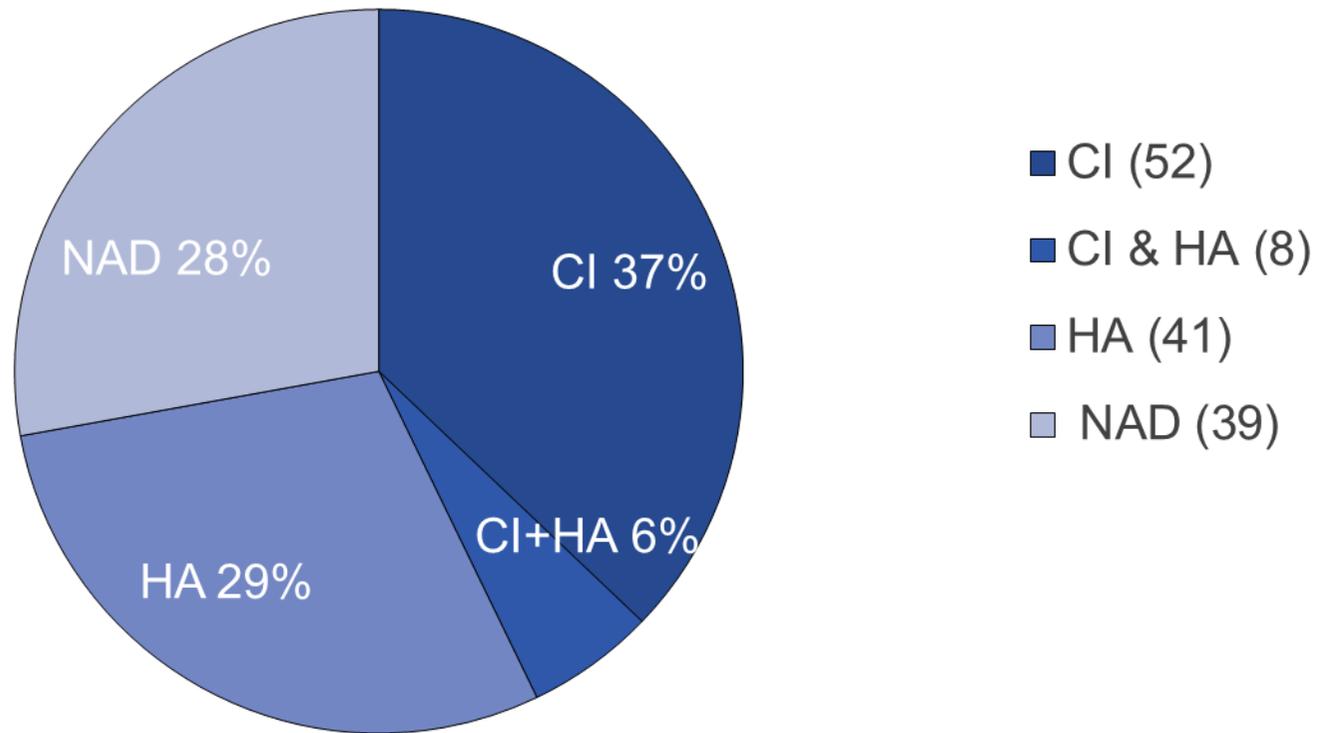
UNC-Chapel Hill : ANSD Study

- 140 infants and children with ANSD at UNC
- All patients evaluated by pediatric audiologist & otologist
- 58 (37%) received cochlear implants
 - » 50 (88%) had bilateral ANSD
 - » 8 (12%) had unilateral ANSD
 - » 52 received CI in ANSD ear
 - » Mean age @ report 88 months (range 14-241 mos)
 - » Mean age @ implant 47 months (range 12-203 mos)
 - » Mean duration of use 41 months (range 6 to 118 mos)

Teagle et al, 2010, Ear & Hearing

UNC-Chapel Hill: ANSD Study

Assistive Listening Device for Children with ANSD (N=140)

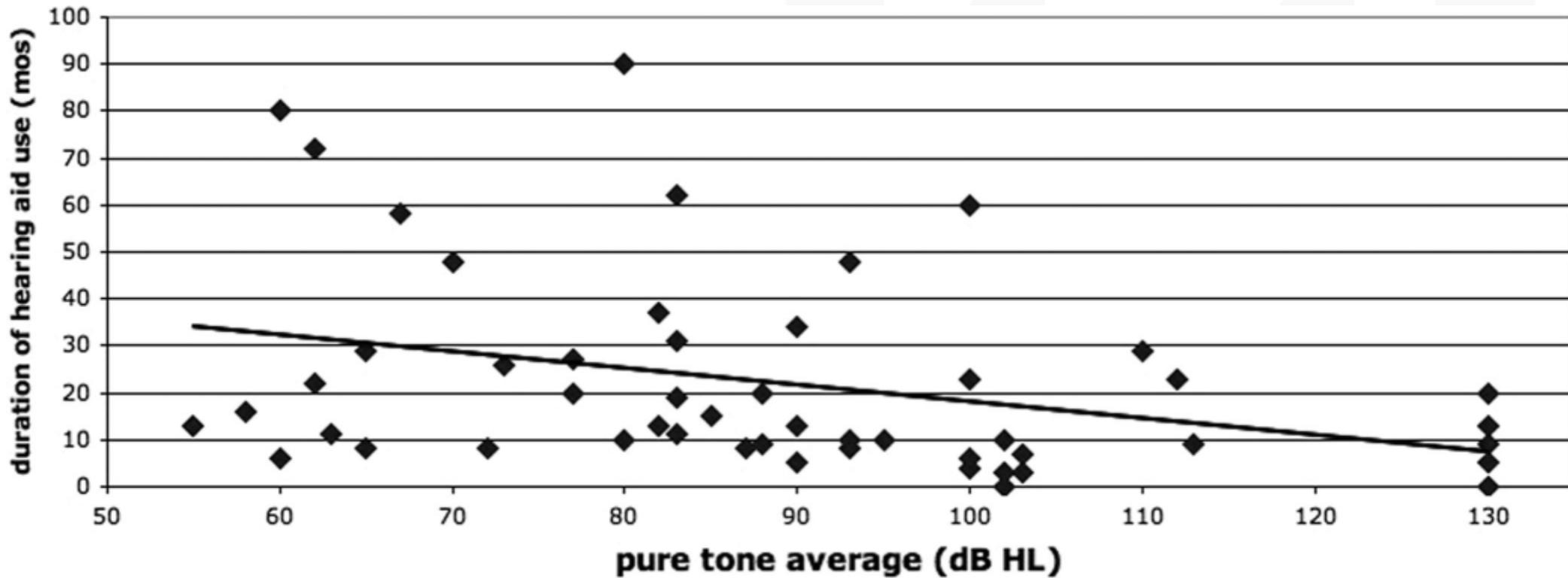


Patient Characteristics

- 42% history of prematurity (<36 weeks gestation)
- Significant co-morbidities
 - » Bronchopulmonary dysplasia (54%)
 - » Necrotizing enterocolitis (18%)
 - » Retinopathy of prematurity (14%)
 - » Intraventricular hemorrhage or hydrocephalus (27%)
 - » Hyperbilirubineamia (17%)
 - » Seizure disorder (12%)
 - » Progressive sensorimotor neuropathy (8%)
- 22% no other medical diagnoses
- 27% positive history for family hearing loss

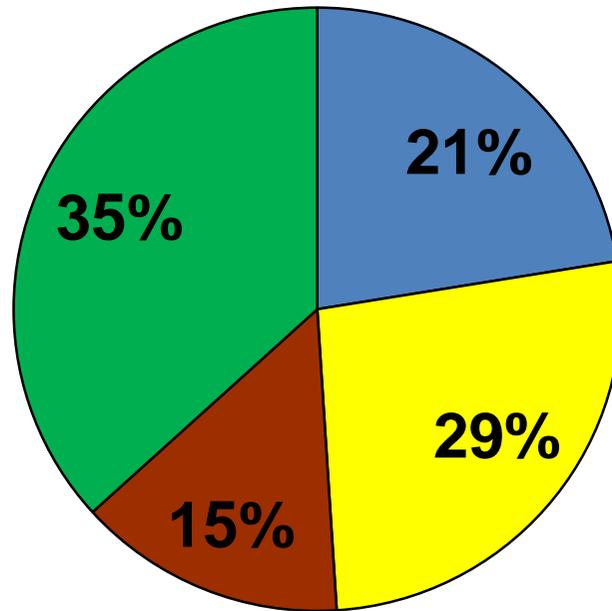


Pre-op best ear PTA for ANSD children receiving CI





CI in AN EAR



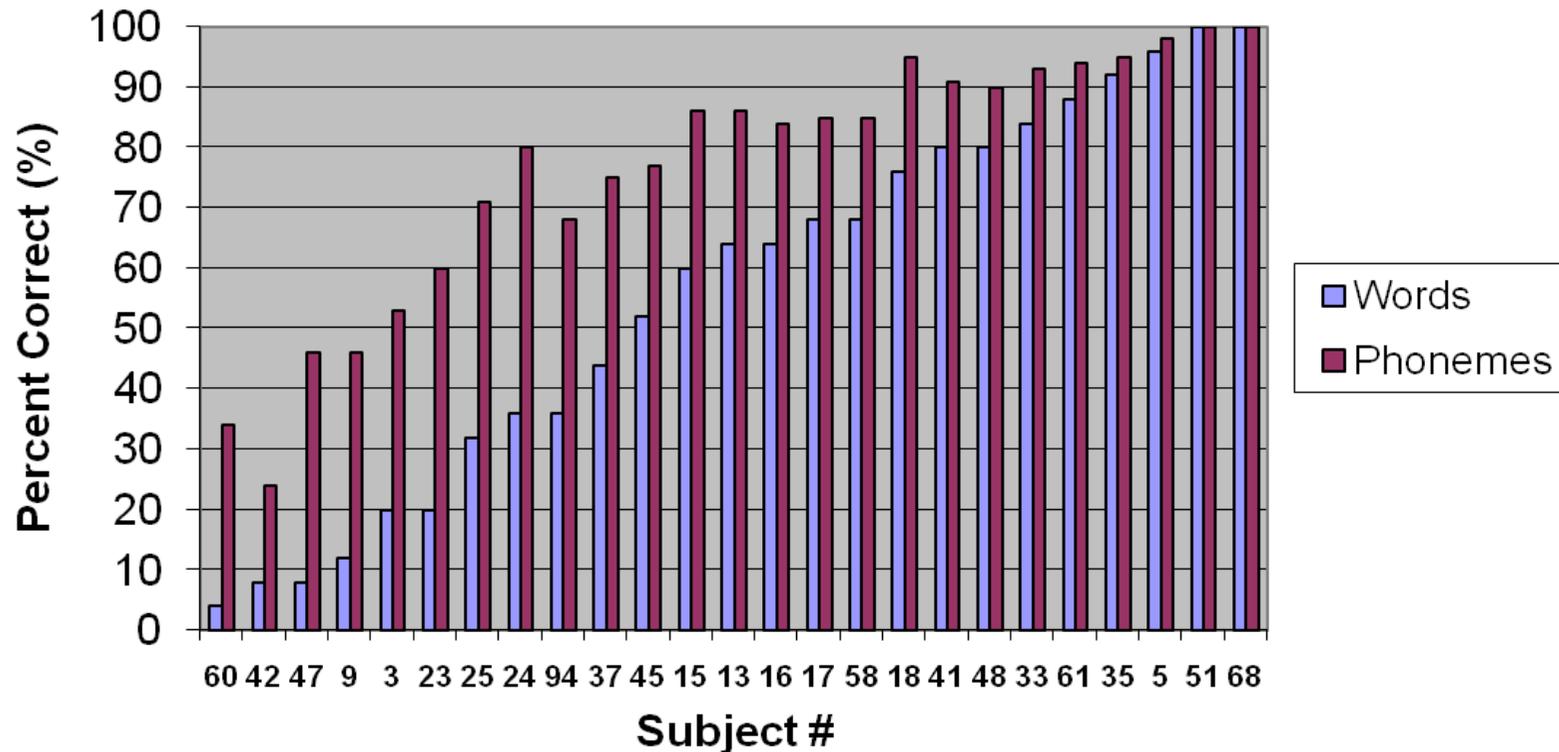
- <6 months CI use/CNT
N=11
- Unable to perform open set (>2 yrs of use)
N=13
- Limited Open Set (<30%)
N=7
- Open Set Performers
N=18

UNC ANSD Children with CI



UNC
SCHOOL OF MEDICINE

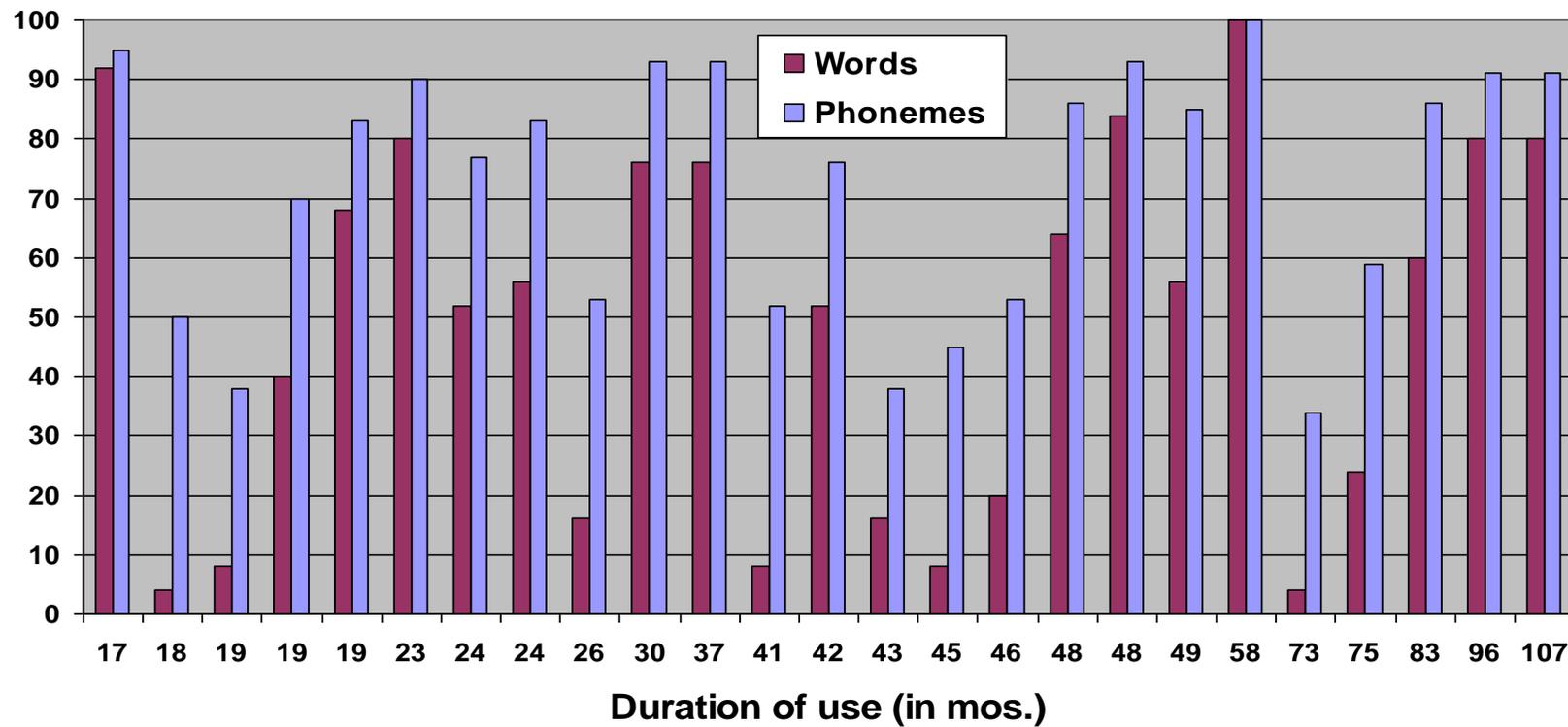
OPEN SET PERFORMANCE N = 25



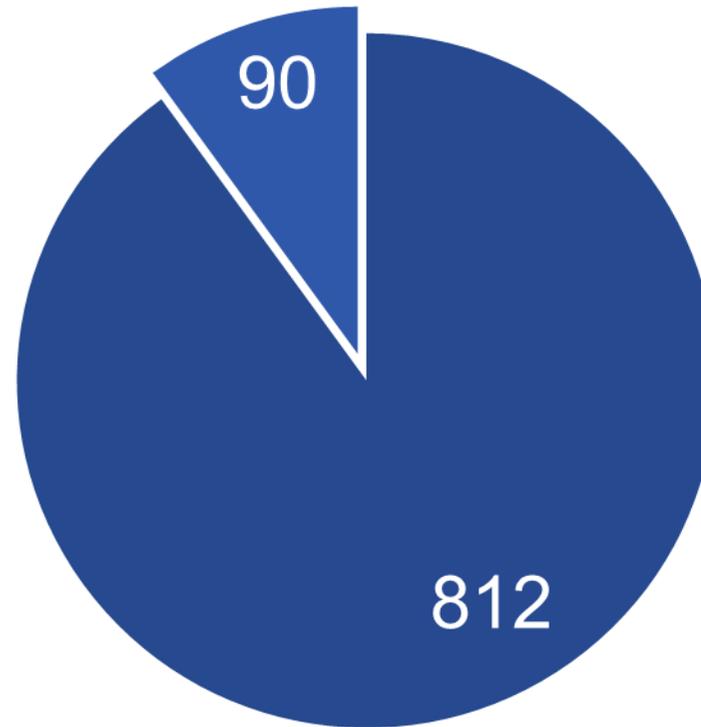
UNC ANSD Children with CI



Open Set Performers (n=25)



UNC Children with Cochlear Implants & ANSD (September 2012)



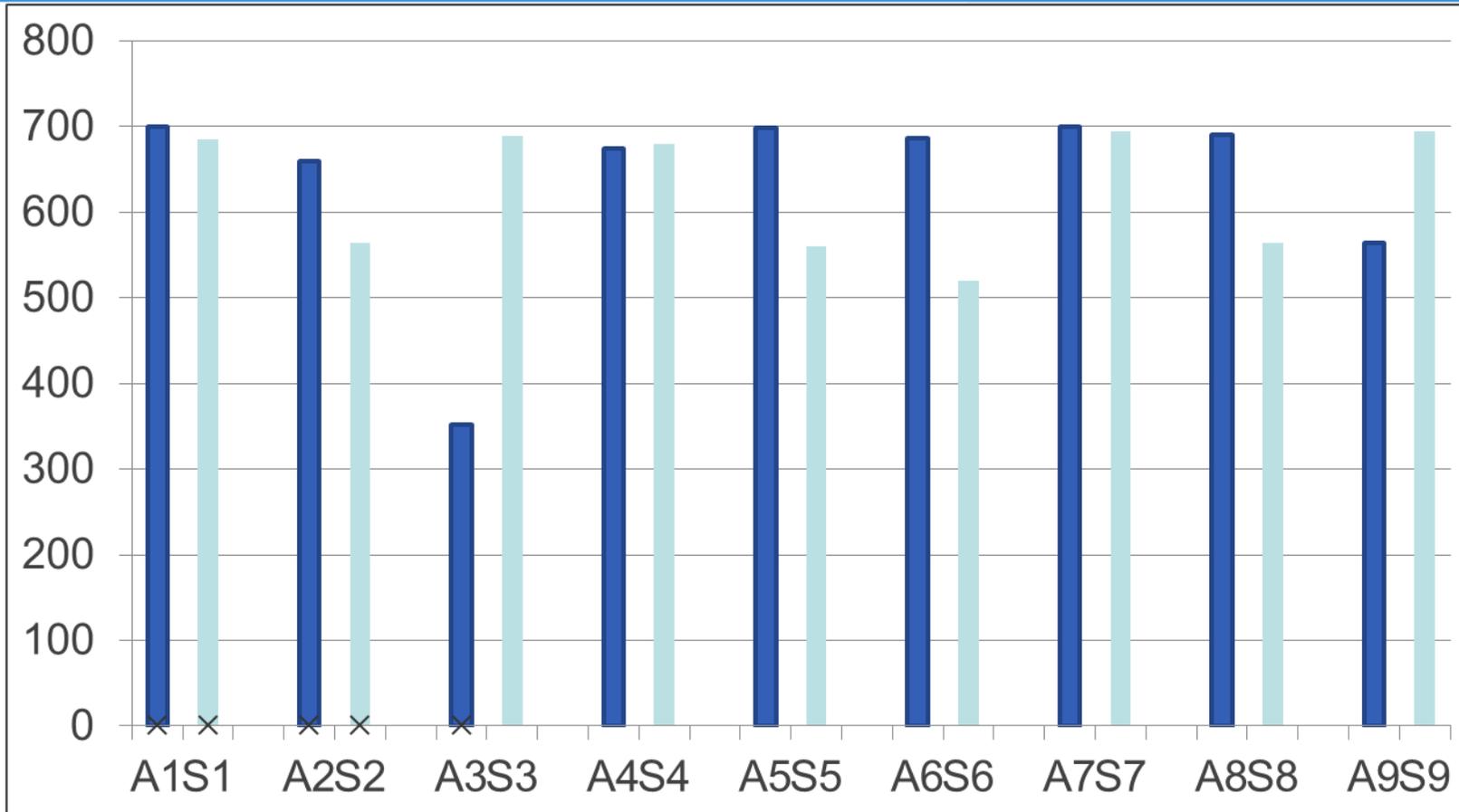
**Approximately 10% identified with ANSD
(Definitive diagnosis of ANSD not obtained
routinely in clinical care until 2000)**

Second look...

Consider only ANSD CI children with no multiple medical diagnoses and match them to children who have SNHL. How do they compare after some duration of CI use?

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) PB-K words and phonemes
(Haskins, 1949)
- MLNT/LNT words and phonemes
(Kirk, et al, 1995)
- HINT sentences in quiet and noise conditions
- SRI-Q - cumulative quotient of test hierarchy. Child must meet a level of performance (70%) before moving on to next level of test. Each level is a 100 point scale.



SRI-Q scores for matched ANSD/SNHL CI Children

Matched for age at implant, first language, communication mode,
uni- or bilateral implant, years of CI experience (3-9),
lack of other medical diagnoses, social/educational status

ANSD
SNHL

64 million dollar question...

- For a child with significant residual hearing, will a CI be better than a hearing aid?
- Given unpredictable outcomes, how do we counsel a family about potential outcomes from CI?
- What additional steps do we take to ensure we are not losing time working towards identifying the best communication mode for each child to reach his or her full cognitive potential?



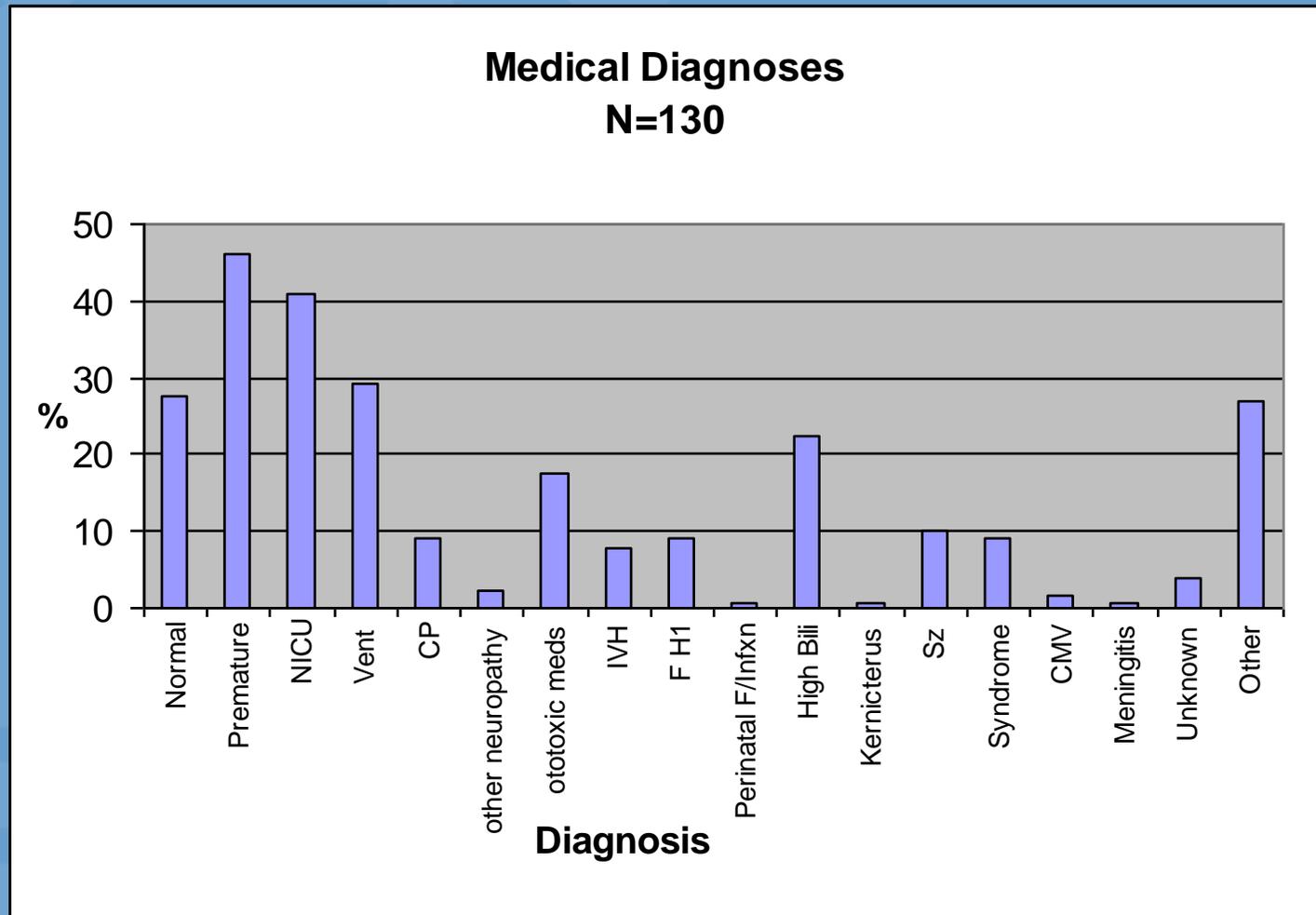
CI Criteria-Children

- **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



Why Comprehensive Medical Evaluation is Important

72% have positive history of other medical diagnoses



ANSD Guidelines (Como 2008): Recommended Amplification Strategies

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- Hearing aid fitting strategies...should follow established guidelines for the fitting of amplification in infants and toddlers
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- 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. (ie, watch and wait)
- Evidence of auditory nerve sufficiency should be obtained prior to surgery using appropriate imaging technology (Buchman et al., 2006) (ie rule out CND and cross fingers)
- 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*. (ie if they are stagnating, just do it)

What other tools can we use to help us predict who will benefit from cochlear implantation? Is there a better mousetrap?

- Maybe.....

Cortical Evoked Potentials (CAEPs)

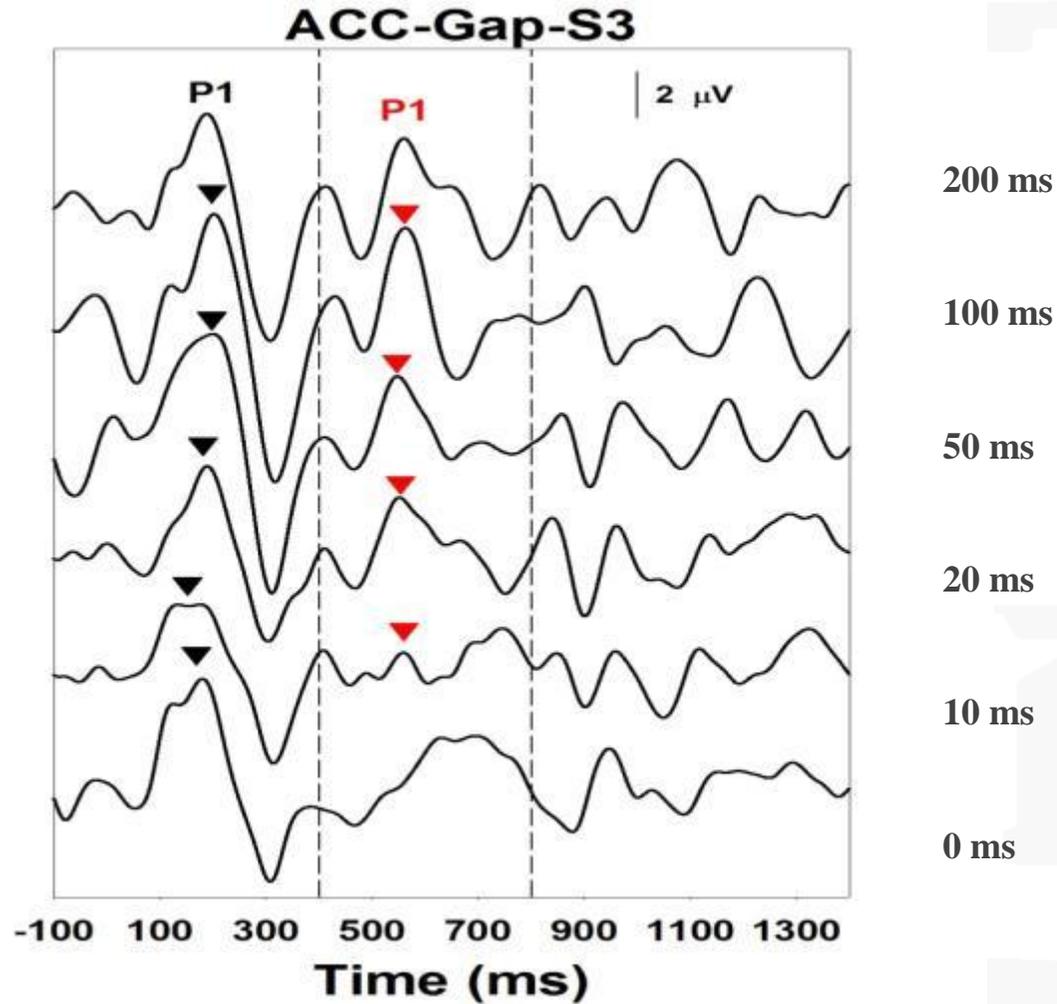
- ABR evaluates outer ear to lower brainstem
- CAEP evaluates outer ear to auditory cortex
- CAEPs not as reliant on timing as earlier evoked potentials and may be present when ABR is not
 - » Hood, 1998, Rapin and Gravel, 2003
- Unlike ABR must be completed in awake (but quiet) infants
 - » Cone Wesson and Wunderlich, 2003)
- Further CAEP research needed with normal infants and infants with SNHL and ANSD



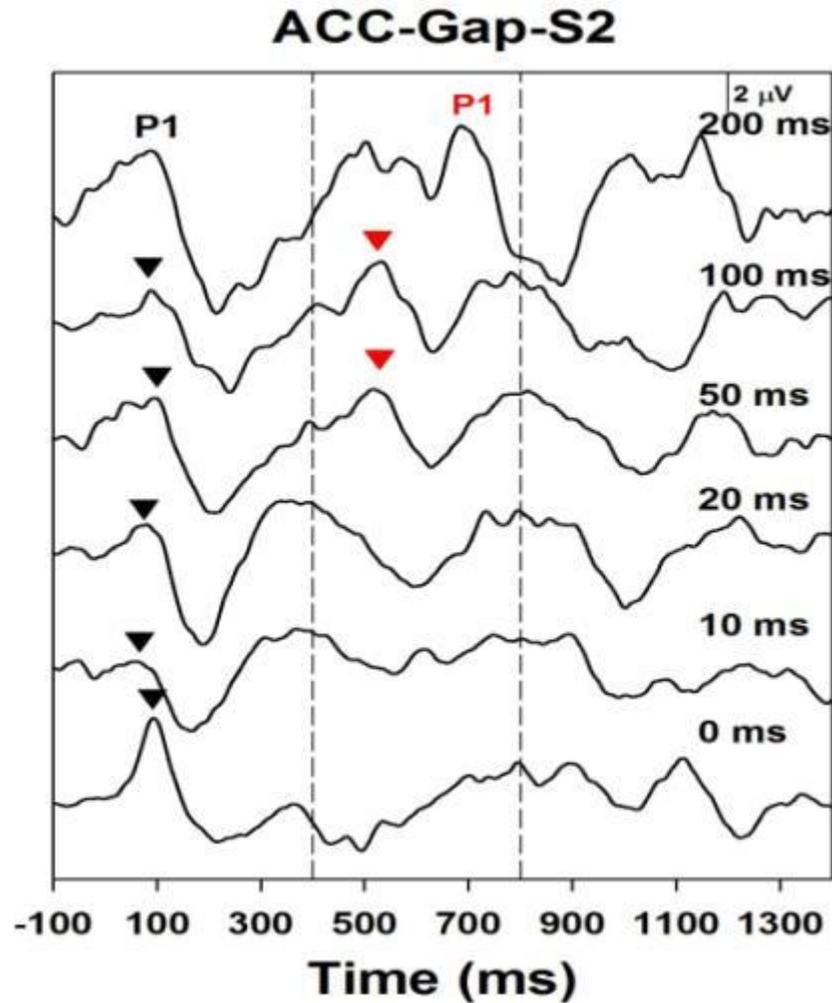
Cortical Evoked Potentials (CAEP)



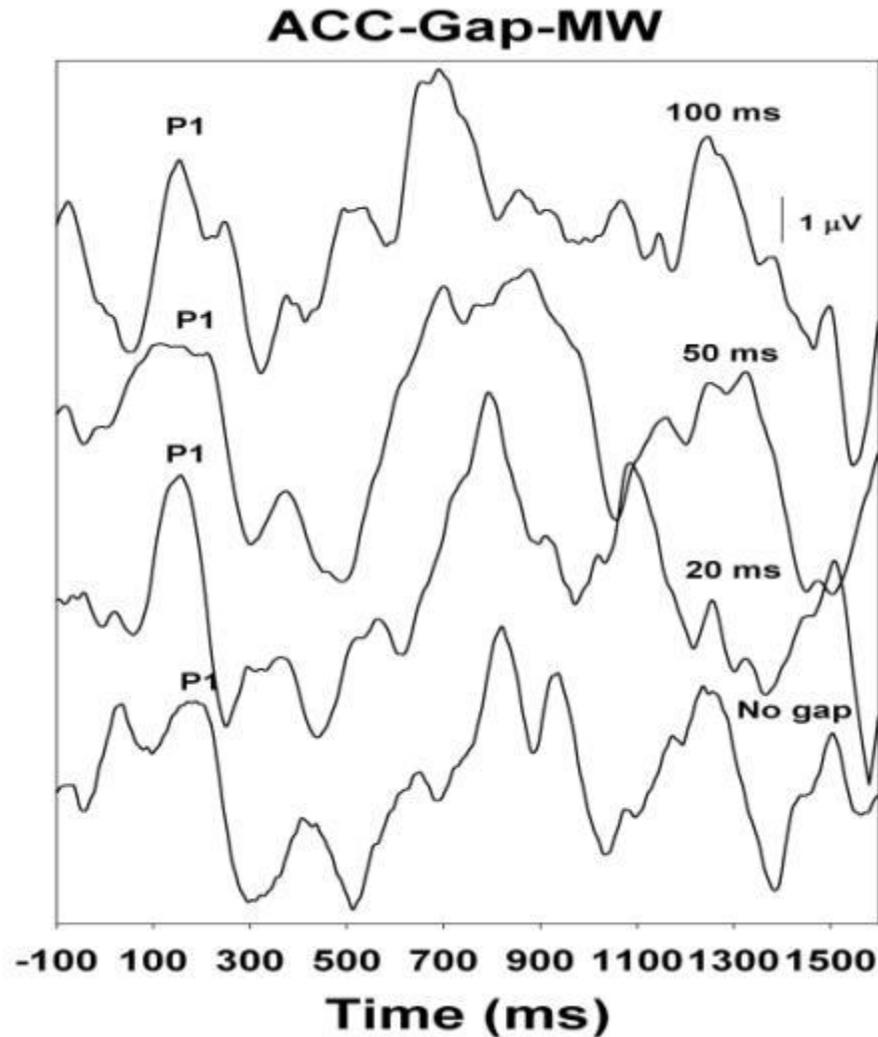
EACC - normal



EACC -60 db HL, ANSD, HA user



EACC – Candidate for cochlear implant



Electrically Evoked Auditory Change Complex in Children with Auditory Neuropathy Spectrum Disorder (ANSD)

Shuman He, PhD; John H. Grose, PhD, Holly FB Teagle,
AuD; Craig A. Buchman, MD

University of North Carolina at Chapel Hill

ANSD Children with CI

- Cochlear implantation has been recommended as a treatment option for ANSD patients.
- Many ANSD children exhibit substantial benefit with their CIs.
- However, a sub-group of ANSD children fails to show significant improvement in speech perception performance.
 - Teagle et al., 2010; Gibson and Sanli, 2007; Miyamoto et al., 1999; Rance et al., 1999, 2002; Buss et al., 2002; Shallop et al., 2001; Madden et al., 2002; Mason et al., 2003.

Research Questions

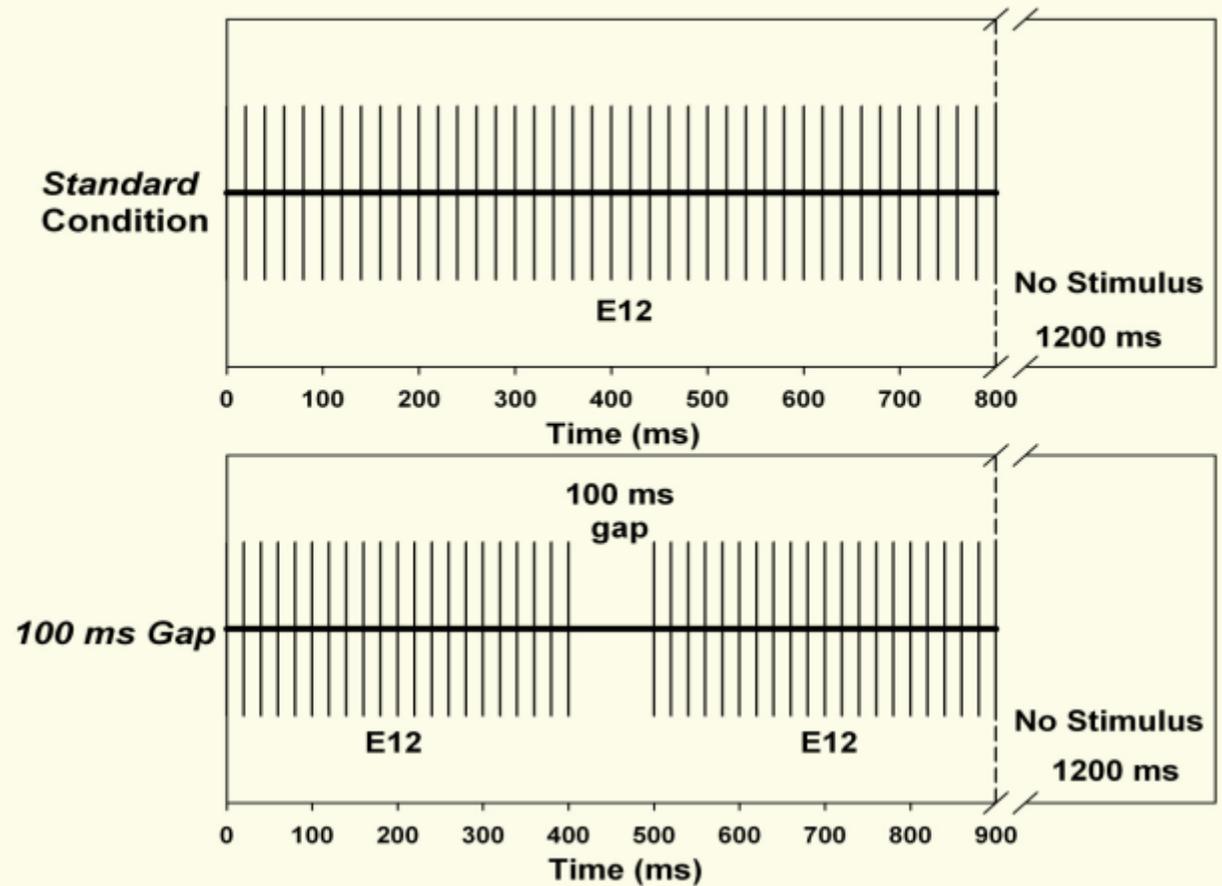
- Do poor performers have temporal processing deficits?
- Do poor performers have spectral resolution deficits?

Methods

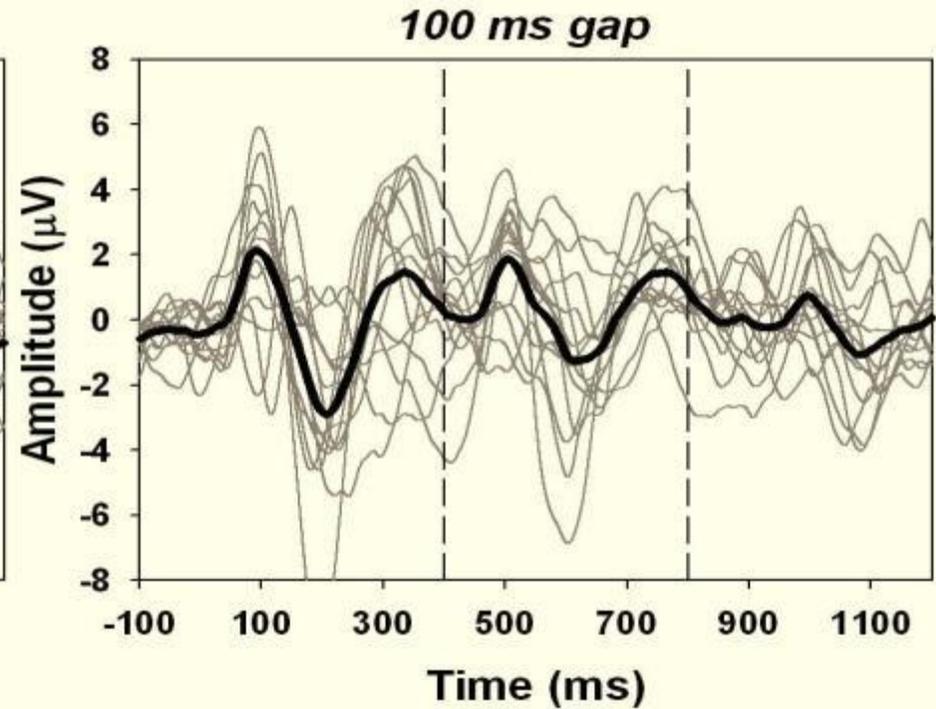
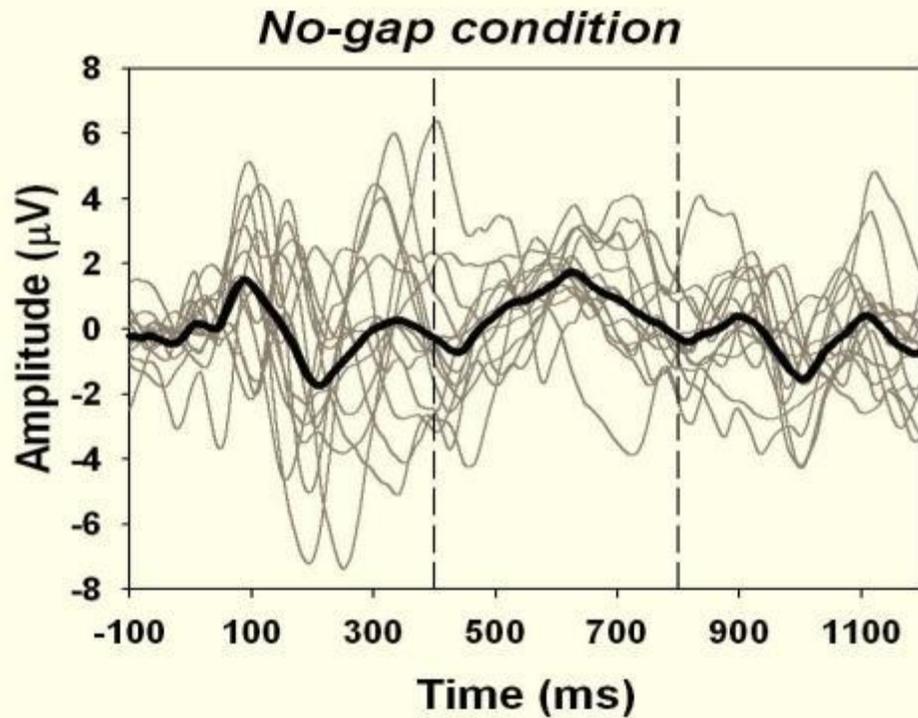
- Subjects:
 - 15 ANSD children with Cochlear Nucleus devices (5.3 to 17.2 yrs; mean: 9.0 yrs).
 - All subjects received their first implants before 4.5 years of age.
 - Good performers: >70% correct on PBK words
- Two stimulating conditions:
 - *Standard condition*
 - *Gap condition*

Methods

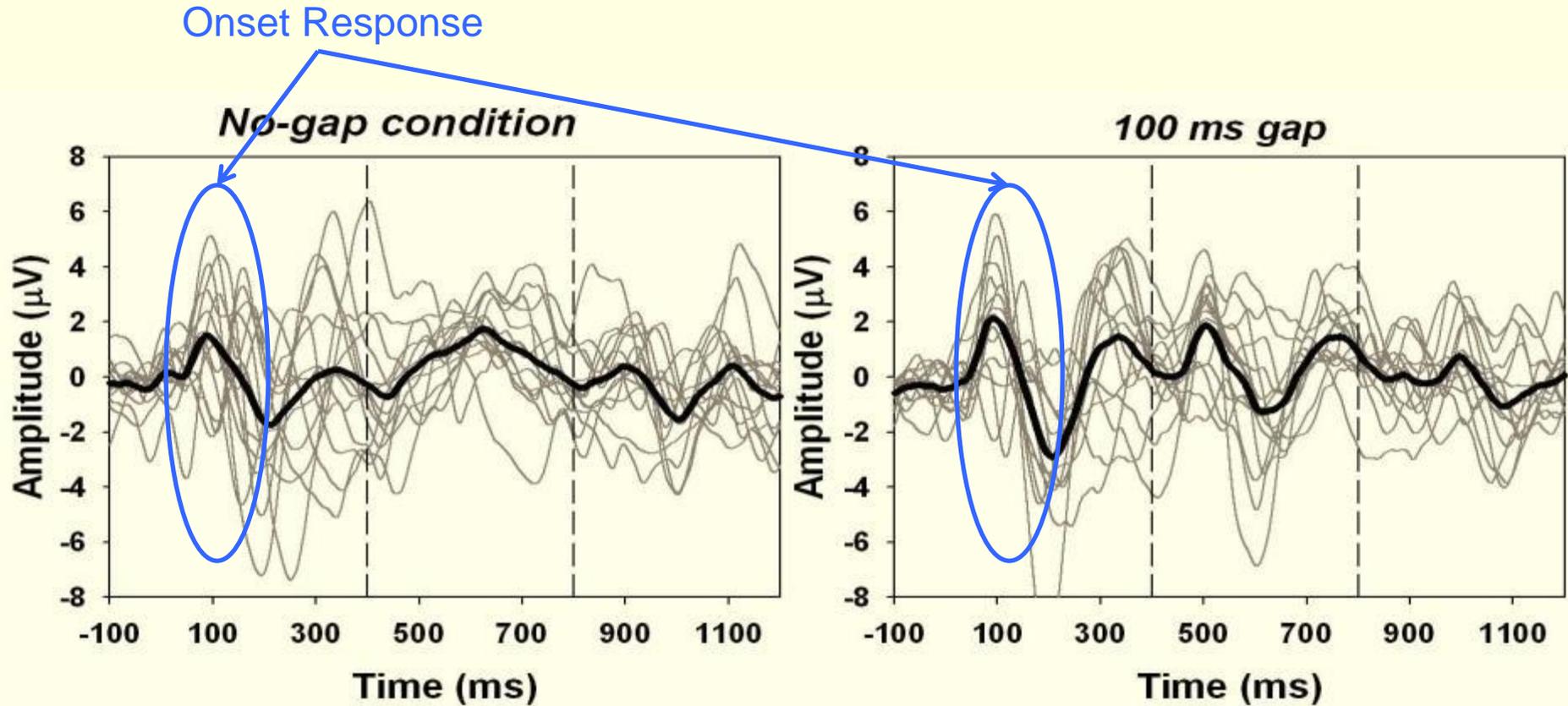
- *Standard condition:*
800-ms biphasic pulse train sent to e12
- *Gap condition:*
 - Two 400-ms stimulus bursts separated by a silent interval (i.e. gap)
 - Gap duration tested: 5, 10, 20, 50 and 100 ms



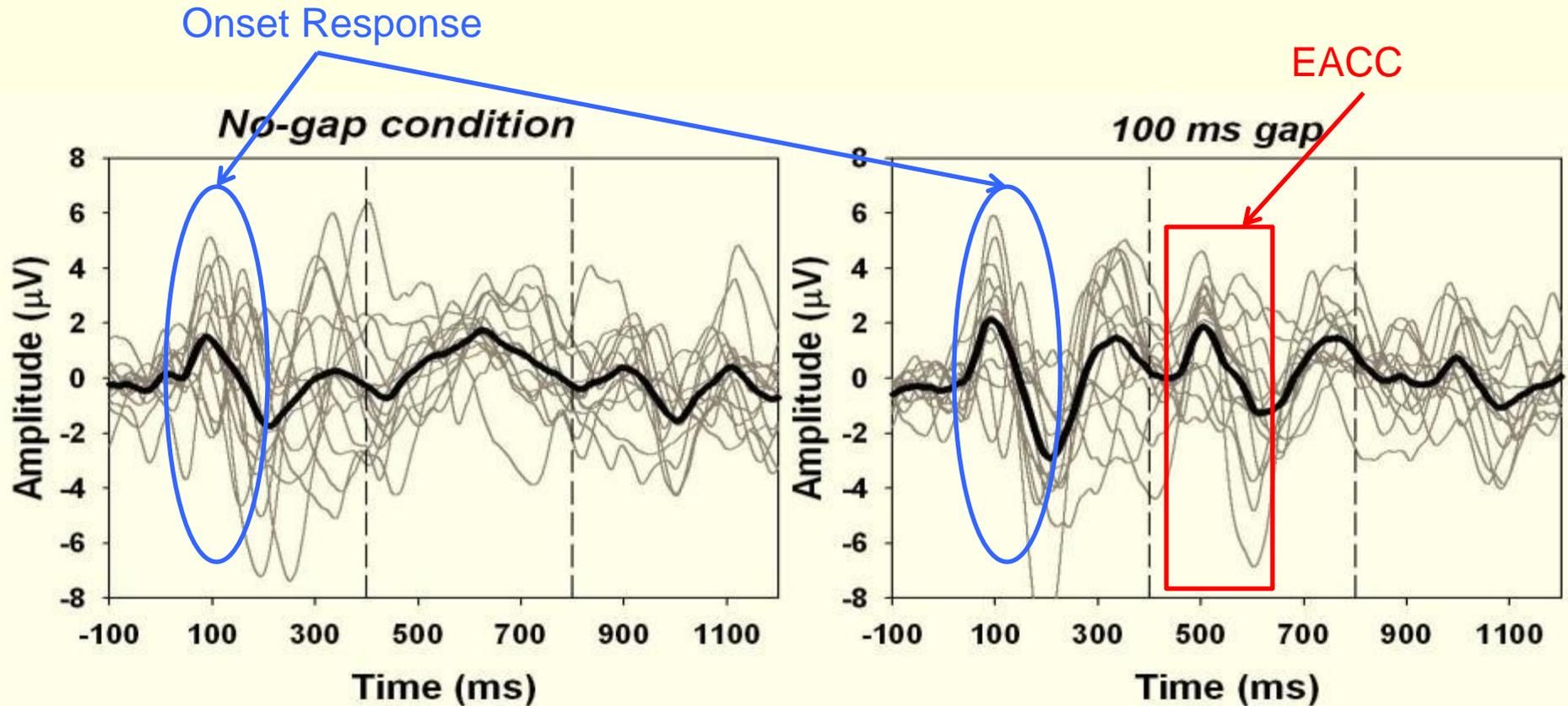
Results: Feasibility



Results: Feasibility



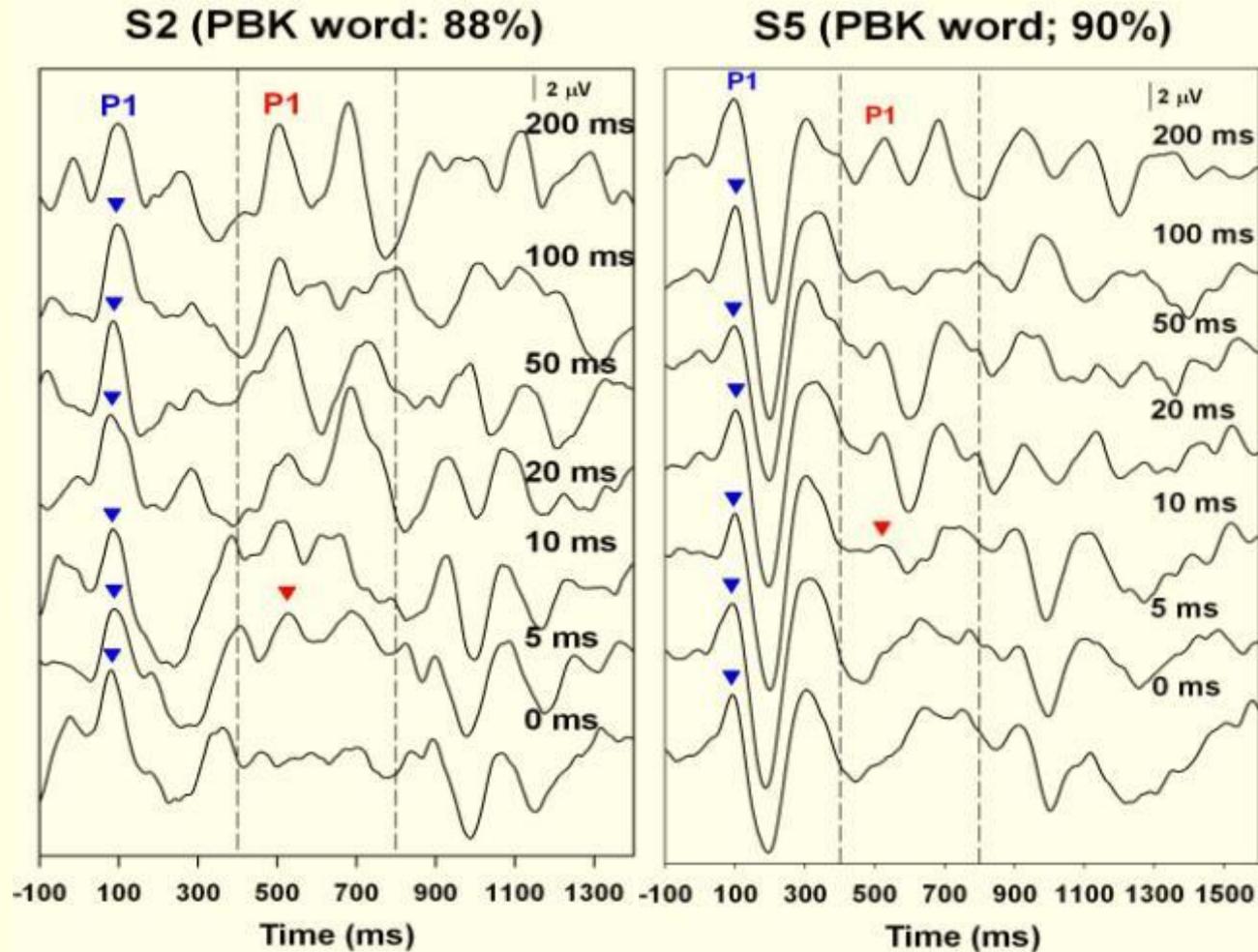
Results: Feasibility



EACC responses were recorded from all ANSD patients using gap stimuli.

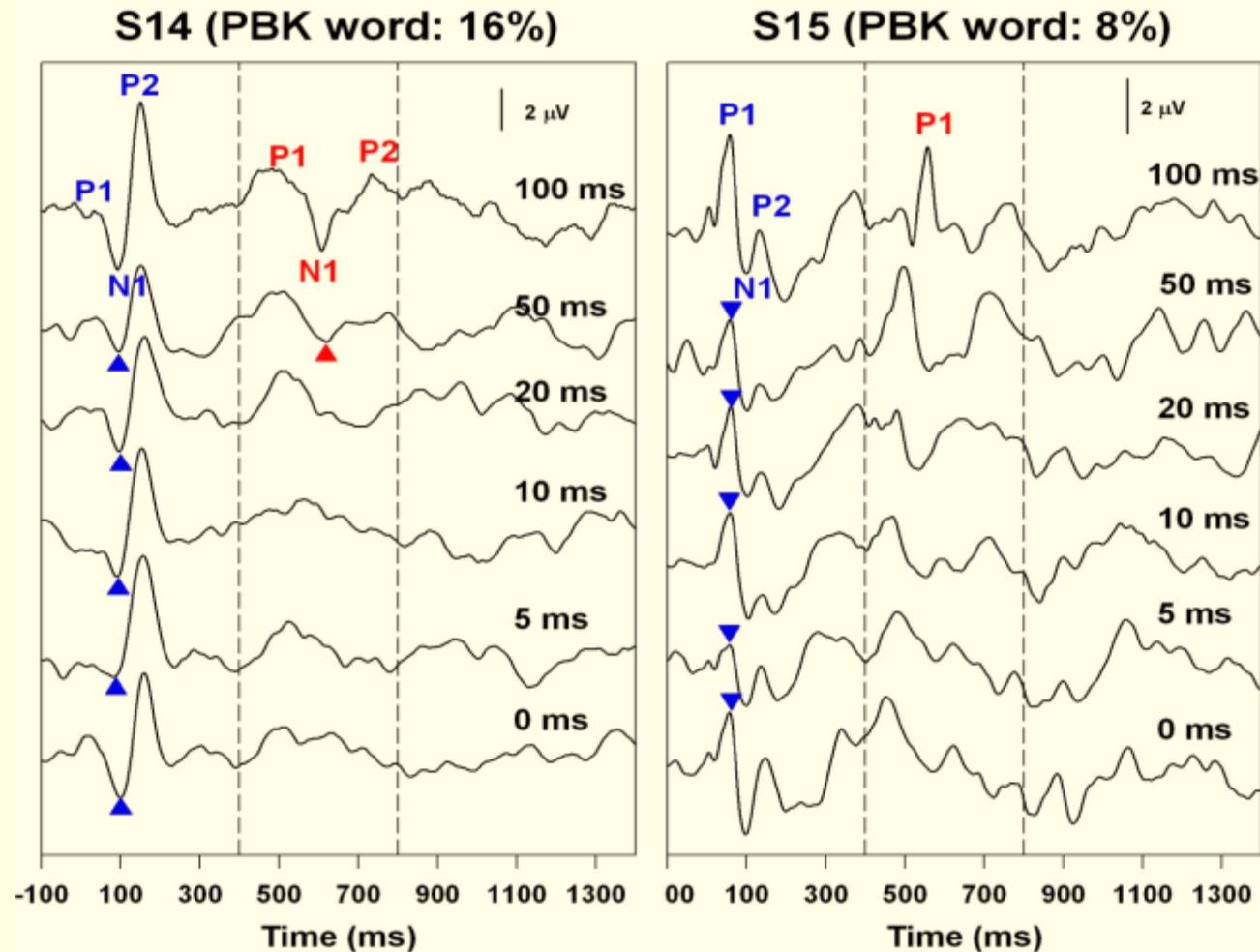
Results: Good Performers

Gap detection threshold (GDT): 5 ms (n=6) or 10 ms (n=4)

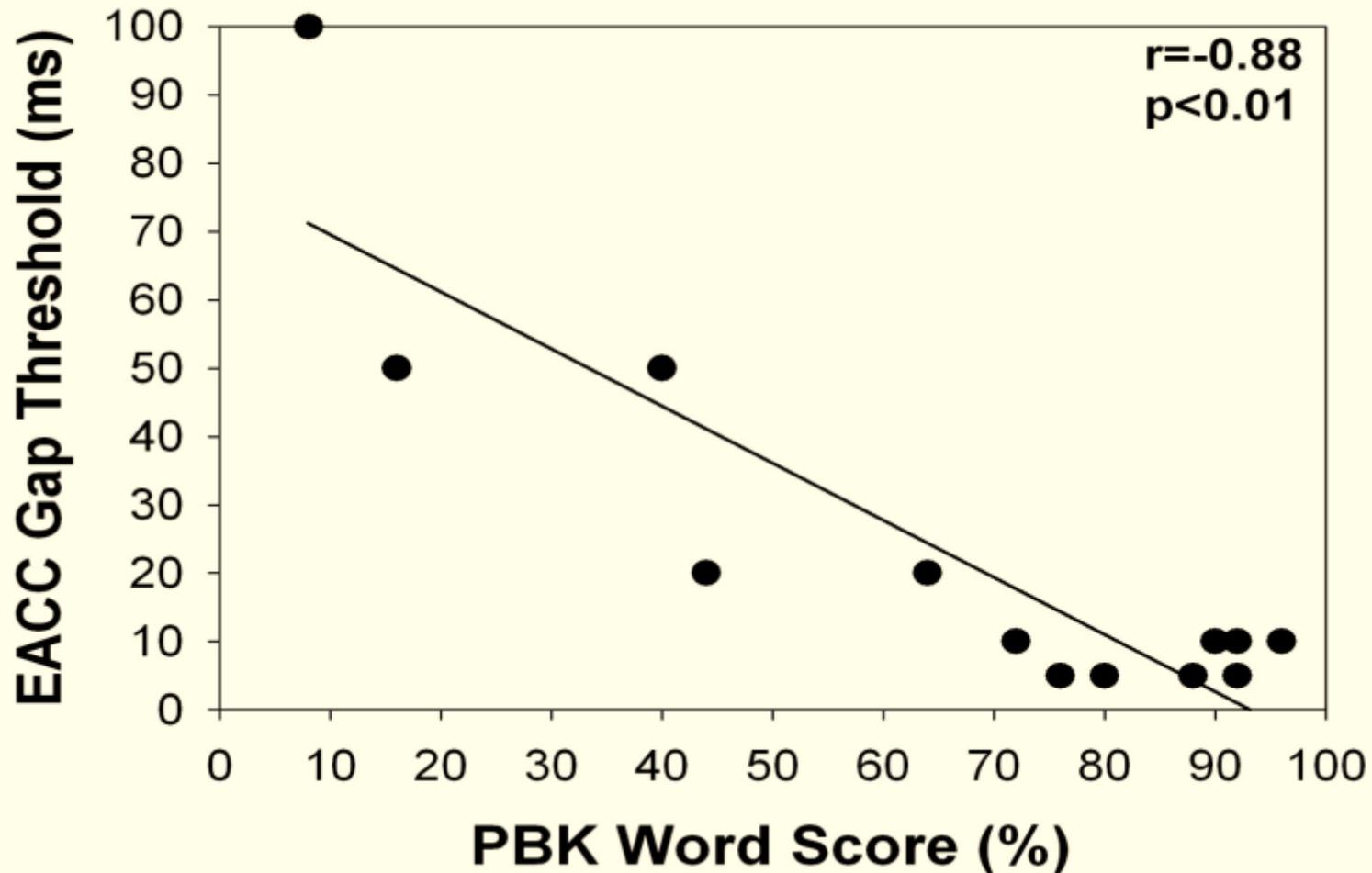


Results: Fair and Poor Performers

GDT: 20 ms (n=2), 50 ms (n=2) or 100 ms (n=1)



Results: Correlation Between the EACC Gap Threshold and PBK Word Score



Research Questions

- Do poor performers have temporal processing deficits?
 - YES.
- Do poor performers have spectral resolution deficits?

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- Do poor performers have temporal processing deficits?
 - YES.
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Methods

■ Subjects

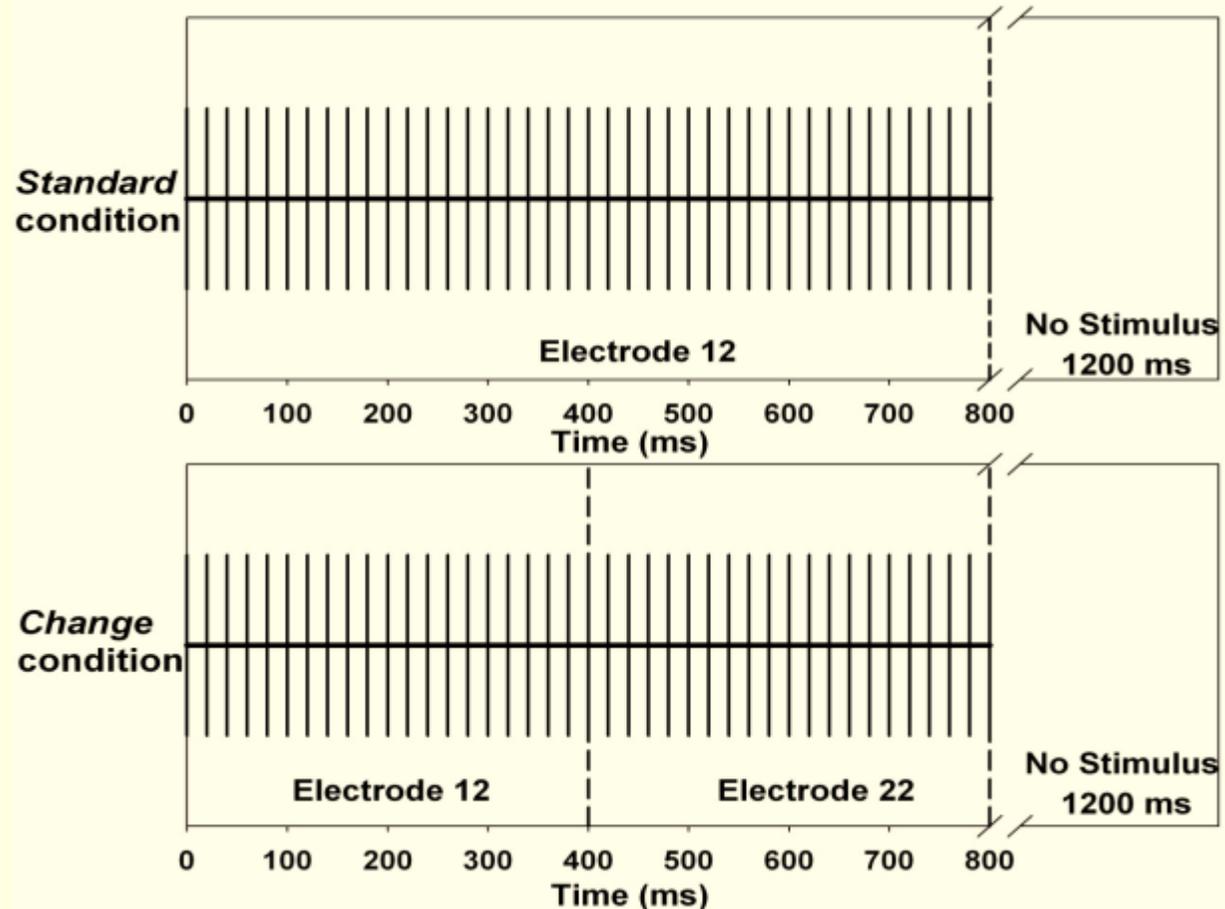
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- All subjects received their first implants before 4.5 yrs of age.
- Good performers: >70% correct on PBK words

■ Two stimulating conditions:

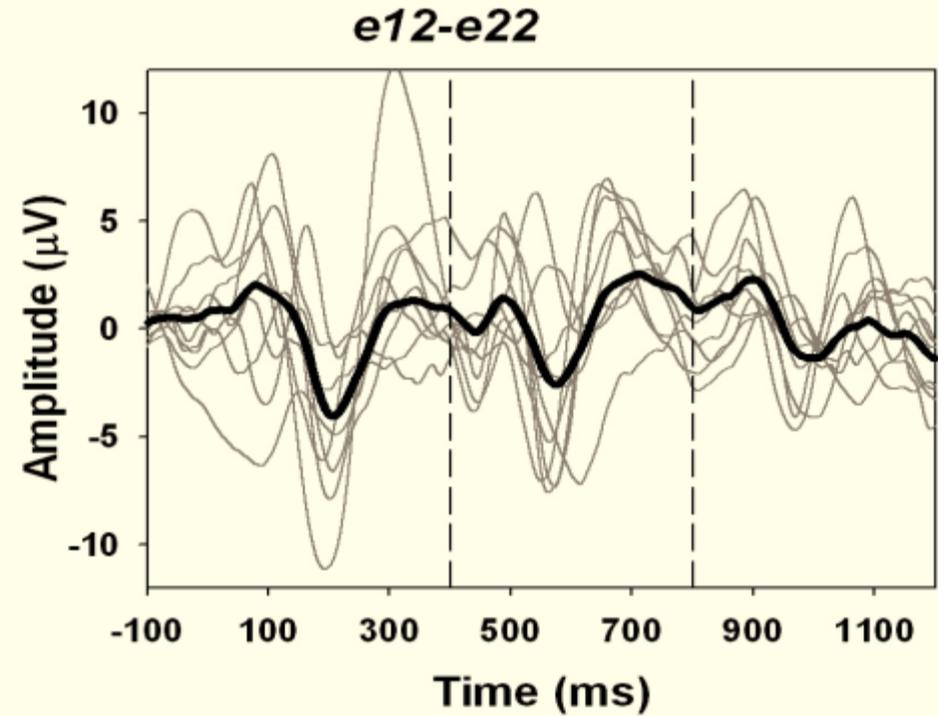
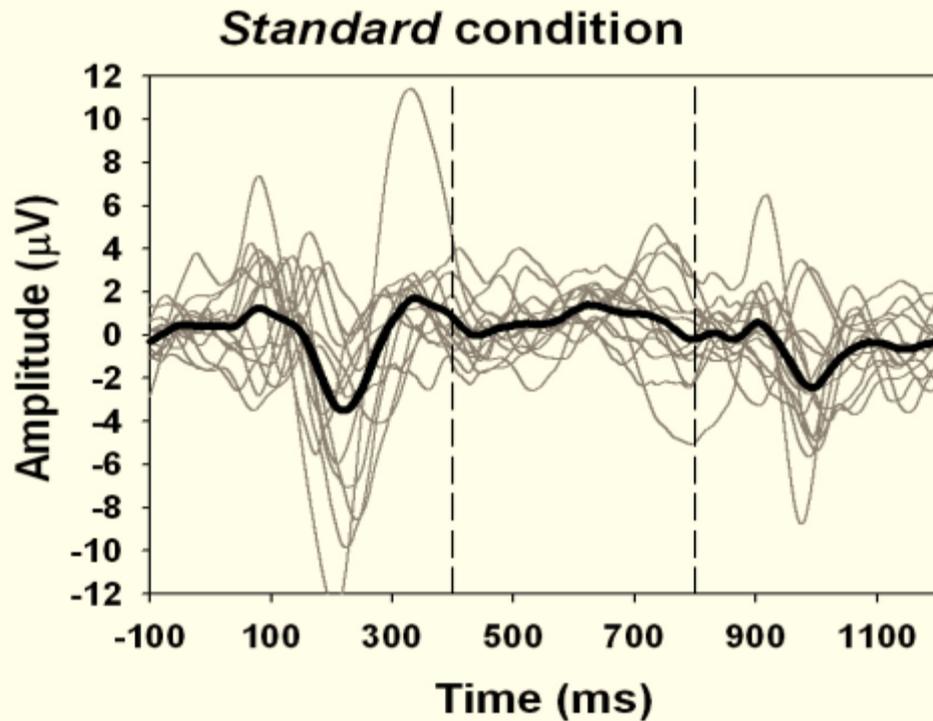
- *Standard condition*
- *Change condition*

Methods

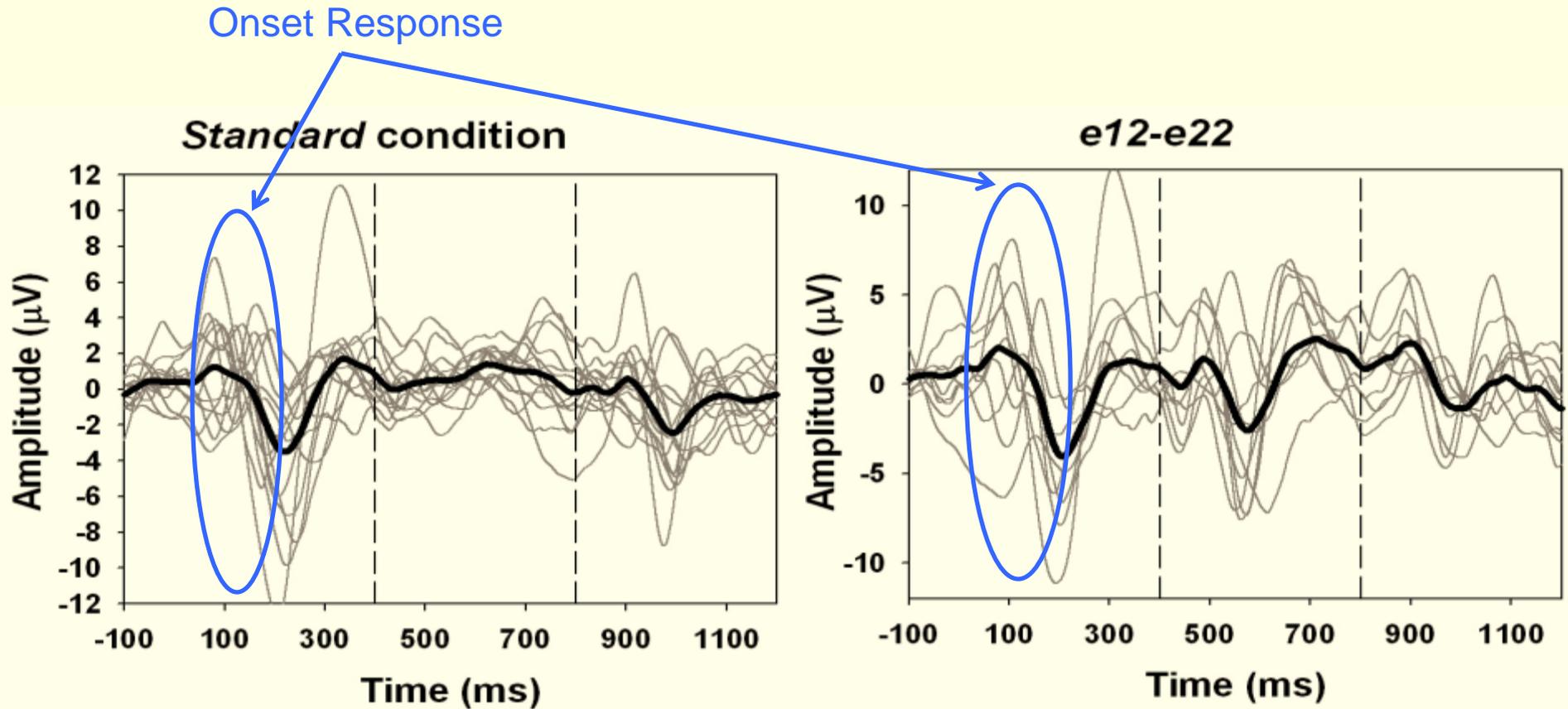
- *Standard condition:*
800-ms biphasic pulse train presented on e12
- *Change condition:*
 - A 800-ms pulse train presented initially on electrode 12, changing to more apical electrodes at 400 ms.
 - Second stimulating electrode: e13-e22.



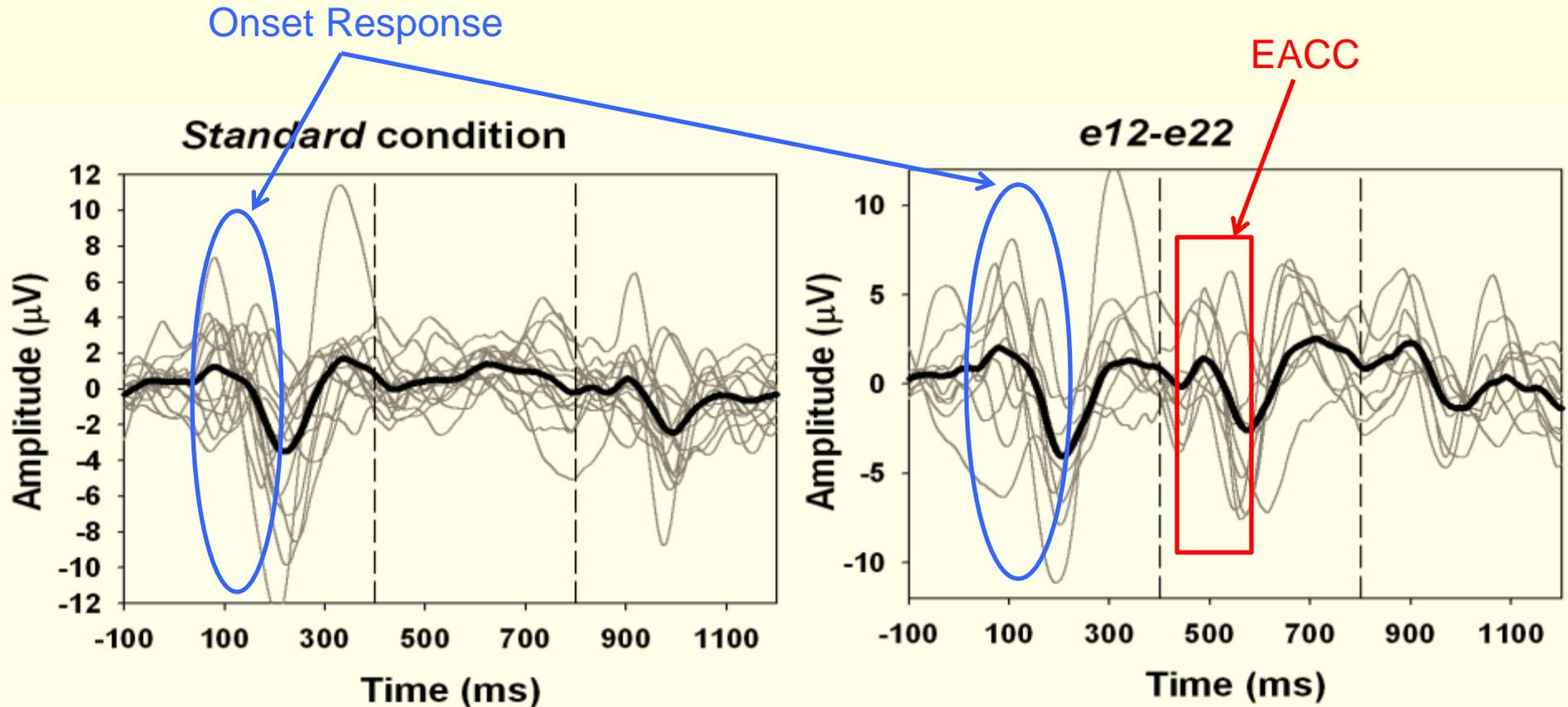
Results: Feasibility



Results: Feasibility

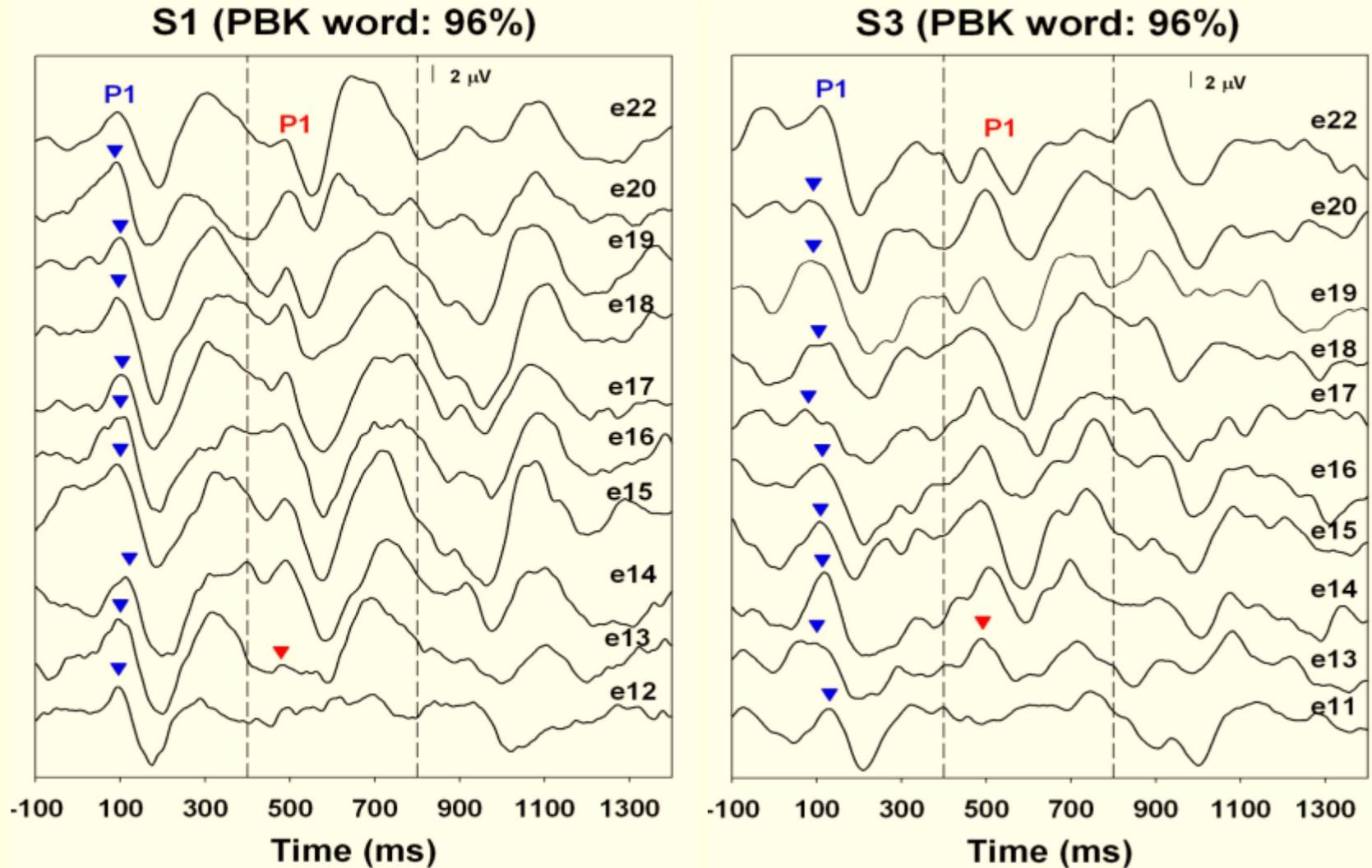


Results: Feasibility

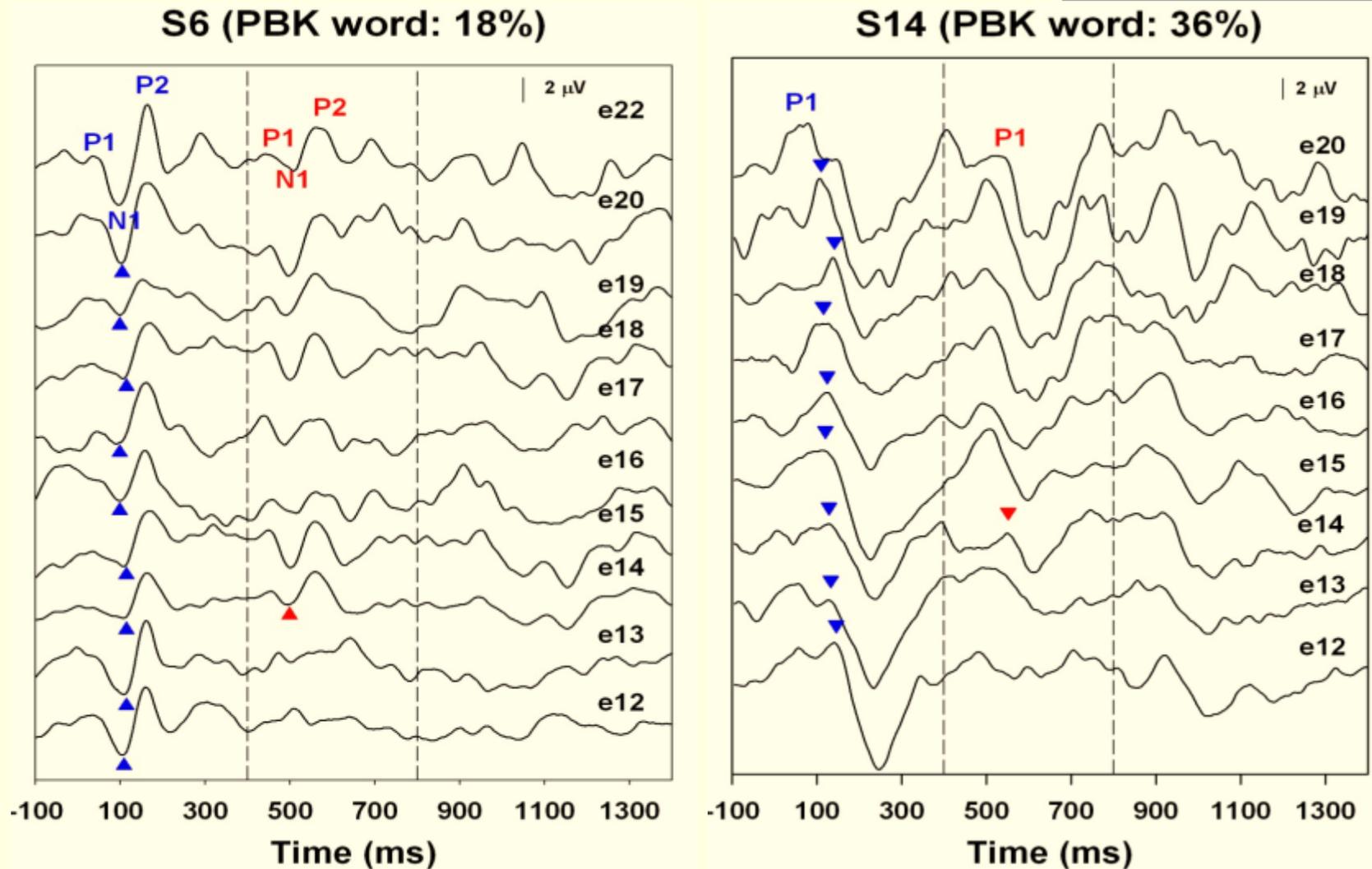


- The EACC in response to changing positions of stimulating electrodes was recorded from all ANSD patients.

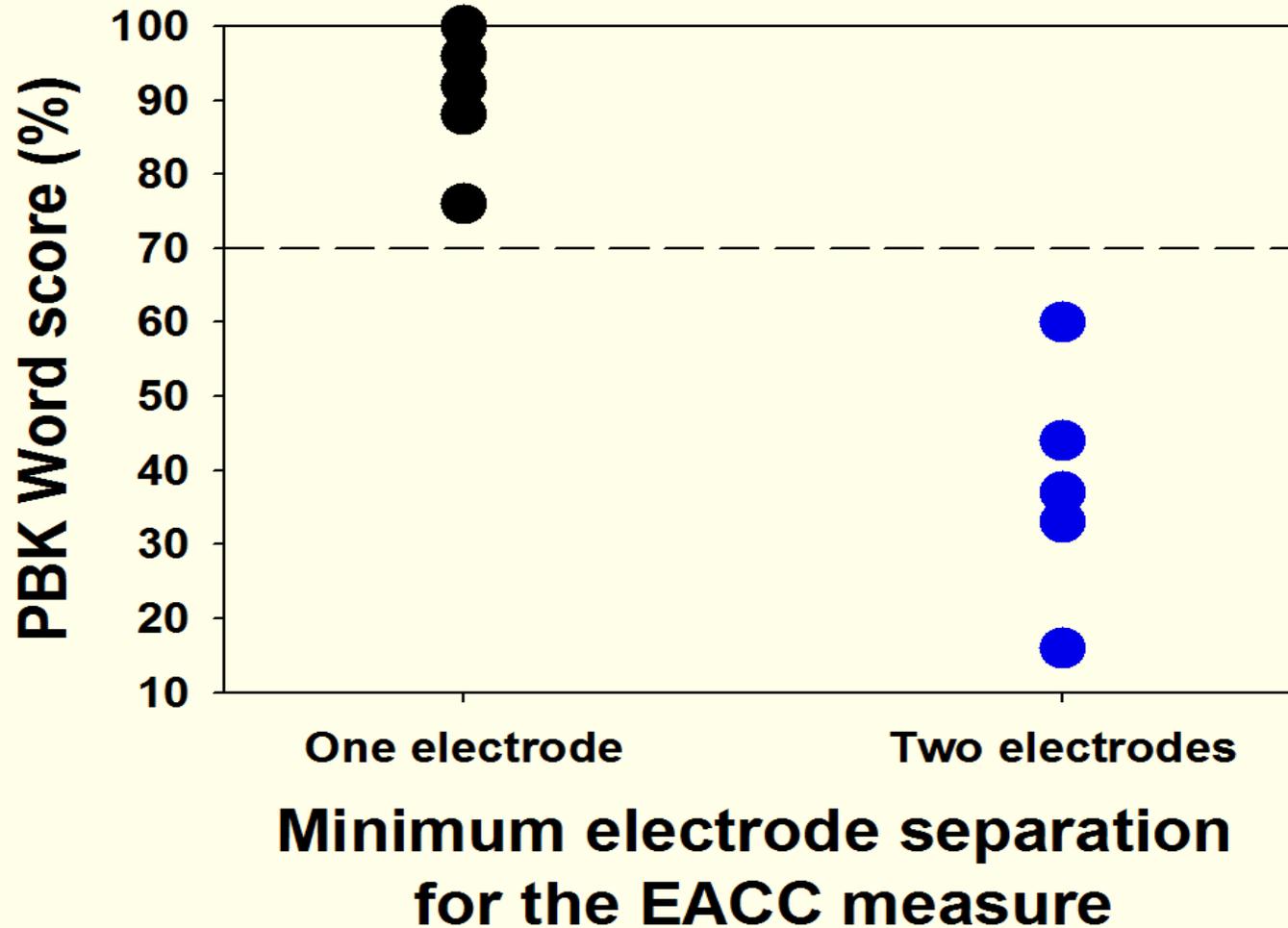
Results: Good Performers



Results: Fair and Poor Performers



Results: EACC thresholds vs PBK Word scores



Research Questions

- Do poor performers have temporal processing deficits?
 - YES.
- Do poor performers have spectral resolution deficits?
 - YES.

Take Home Messages

- Poor performers have temporal processing deficits and/or spectral resolution deficits.
- The EACC can potentially be used as an objective tool to evaluate auditory functions of ANSD patients with CIs.

How do we optimize CI programming for children with ANSD?

- Slow the rate of stimulation
 - » Allow for longer refractory period
- Distance stimulating electrodes
 - » Encourage better spectral resolution
- Widen the pulse width
 - » Excite more neural elements at a time
- Measure loudness growth
 - » Optimize the real dynamic range of neural system
- Pitch rank and eliminate channels that create the same percept
 - » Replicate the tonotopic potential that exists
- Don't employ strategies that create more channels
 - » Decrease the possibility of spectral smearing

All these techniques can be implemented through the programming software but they are **not** the default settings and they take more time.

We are learning more about the electrophysiological profile of children with ANSD and how it relates to hearing performance.

These measures are not standard clinical tools yet.

Given the heterogeneity of the population and the range of outcomes, we should hope for the best but counsel and plan for less than best.

UNC Protocol for Post Implant Evaluation of Children with ANSD/Cochlear Malformations

Pre-operatively, parents counseled on the tentative prognosis for success with an auditory communication approach. If they choose Listening and Spoken Language for their child, the following guidelines will be implemented to monitor progress and make recommendations regarding intervention and communication mode.

Developed in collaboration with many NC speech & hearing professionals

3 Months Post-Initial Stimulation

- Goals for speech processor programming sessions:
 - Child has attended initial stimulation, 2, 5 and 9 week visits
 - Neural response telemetry has been collected using clinic protocol
 - Impedance telemetry is stable
 - Map parameters have been explored
 - Full-time implant use & consistent, appropriate therapy
- Minimum Auditory Skills:
 - Child detects sound, i.e. an audiogram demonstrating sound detection of at least 60 dB HL has been collected,
 - Child has indicated some bonding to or at least acceptance of the device,
 - Parents report some sound awareness
- If the minimum auditory skills have not been achieved,
 - An auditory/ visual system will be recommended to the family.
 - Both options of Cued Speech and Sign Language will be reviewed with the family
 - Information provided regarding increased spoken language and literacy opportunities if they choose Cued Speech.



6 Months Post-Initial Stimulation

- Goals for speech processor programming sessions:
 - Impedance telemetry is stable
 - Map parameters relatively stable
 - Reliable detection audiogram of at least 40 dB HL at octave frequencies
 - Bonding and full acceptance of the device
 - Manufacturer device integrity test completed
 - Verification by surgeon of device placement
 - Full-time implant use & consistent, appropriate therapy

12 Months Post-Initial Stimulation(cont'd)

- Speech-Language Evaluation to document progress
- Minimum Communicative Skills:
 - Half-way through Year 1 of Auditory Learning Guide
 - Imitating supra-segmental qualities and vowel variety
 - Identification of songs/nursery rhymes
 - Comprehension of some phrases
 - Identification of Learning to Listen Sounds
- If the minimum auditory skills have not been achieved, or, if the language gap is too big to close, an auditory/visual system will be recommended to the family. Both options of Cued Speech and Sign Language will be reviewed with the family.

18 Months Post-Initial Stimulation

- If language is not developing at a reasonable rate to make child a functional communicator and able to access academic information to the best of his/her potential, the recommendation will be made that visual support for communication be aggressively implemented.
- **Additional Comments:**
 - Cognitive testing using the Rossetti Playskill subtest/ CSBS may be beneficial to have pre-implant
 - This population of children should be assigned to Early Intervention therapists who are knowledgeable about ANSD and have extensive experience with children with hearing loss .
 - Mentoring of Early Intervention staff is a critical piece currently needed in NC

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
- Results of behavioral response to CI are necessary before communication mode decisions are clear
- Electrophysiological measures are beginning to correlate but we need more information
- Cochlear implant use helpful in some cases not in others but we will only know if child is fit appropriately and has consistent use
- Monitor continuously, adapt and adjust with time

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
- Families need to be reassured that help is available



Part III

Case Studies



Case #1

Child with CI

Case #1 Background

- Fraternal twin
- Born at 27 weeks gestation (1175 gms)
- NICU 3 months
 - » Hyperbilirubinemia
- Later diagnosed with mild spastic CP
- Did not pass NB hearing screen (AABR)
- Diagnostic ABR consistent with ANSD
 - » No response ABR with large CM
 - » Present OAEs
- Behavioral audiometry
 - » Thresholds in moderate range bilaterally



Case #1 Background

- Fitted with binaural hearing aid at 16 months
- Parents and EI specialists did not feel he was making adequate progress
- Received CI at 2 yrs, 8 mos.
- Revision CI at 7 yrs of age following trauma from fall



UNC
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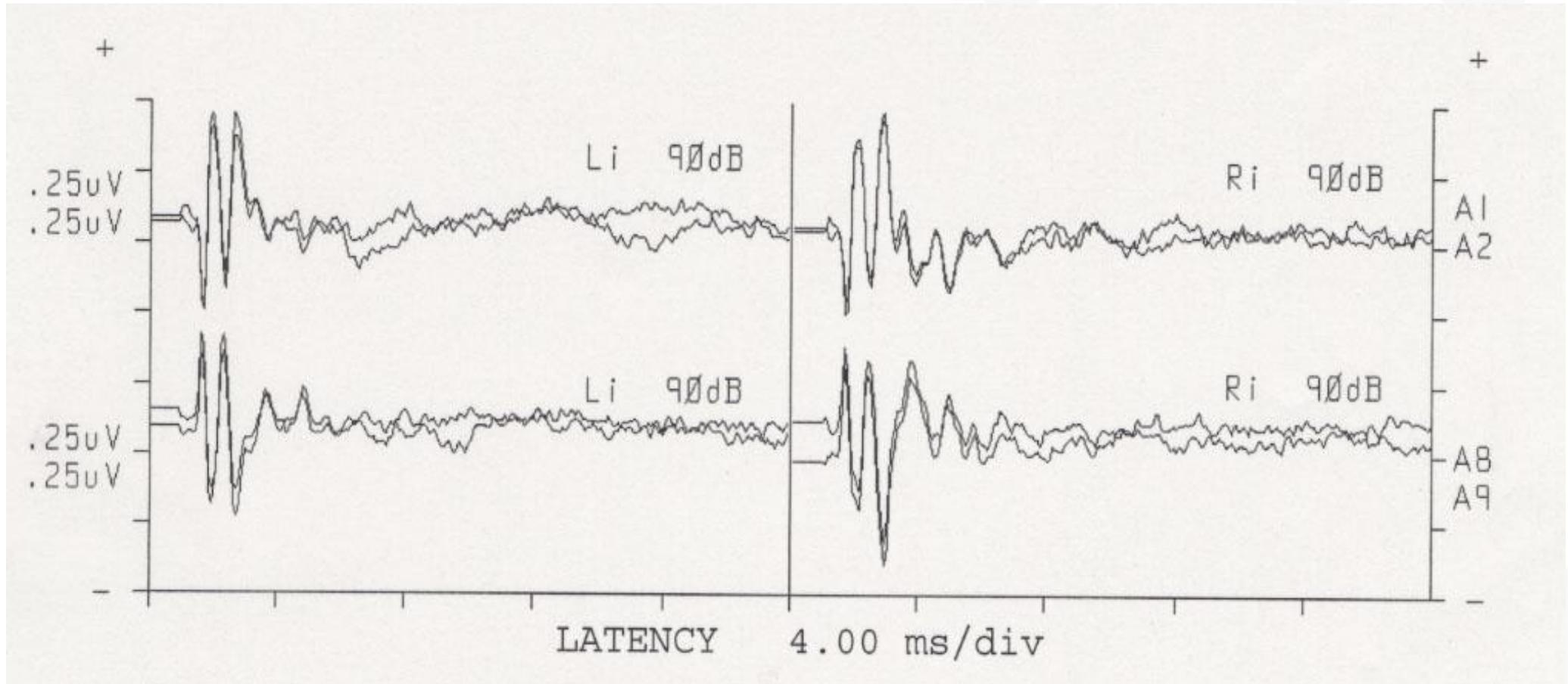
CASE #2

Child using Hearing Aids

Case # 2 Background

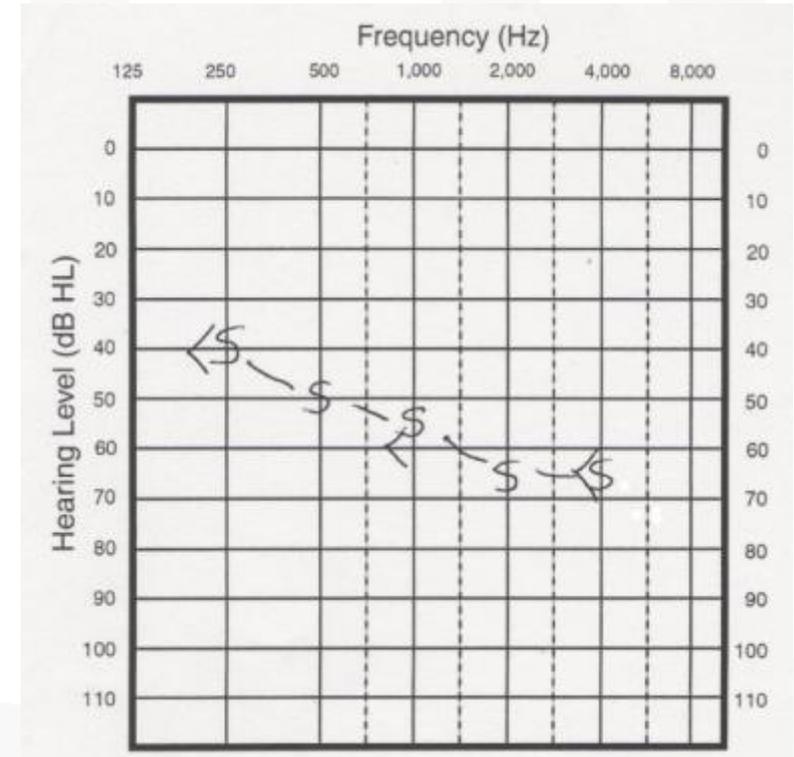
- Born at 25 weeks gestation
 - » Hospitalized 4 ½ months
 - » Ventilated for 6 weeks
 - » Oxygen 3 ½ months
 - » Hyperbilirubinemia
 - Treated with lights, exchange transfusion
 - » Treated with antibiotics and diuretics
- No family history of hearing loss
- Did not pass NB hearing screen at hospital 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)



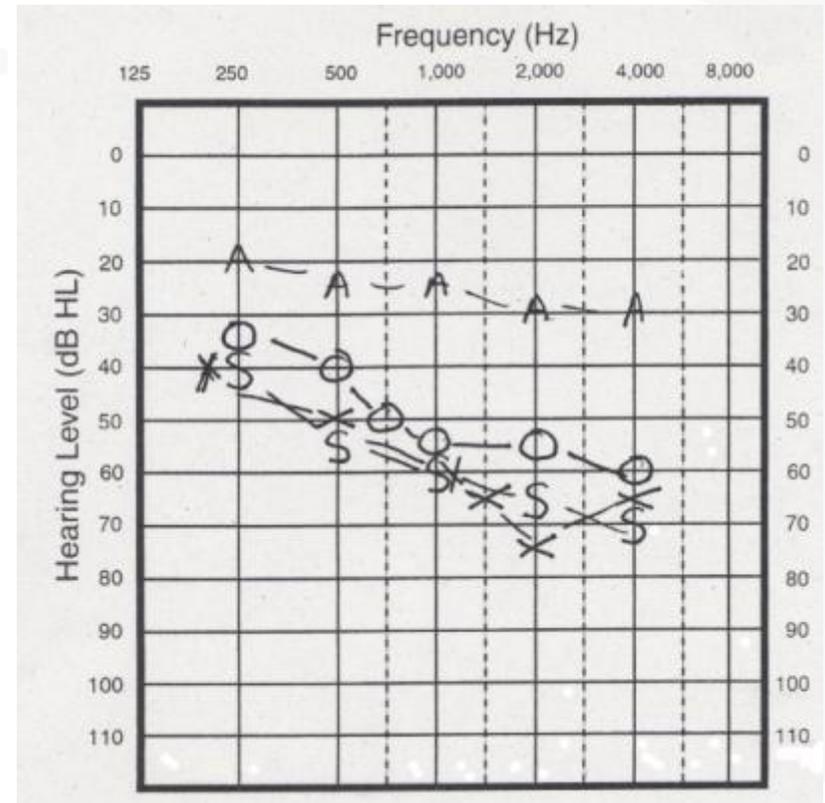
Age 10 Months (6 1/2 Months Adjusted Age): Behavioral Audiometry with VRA

- **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



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):

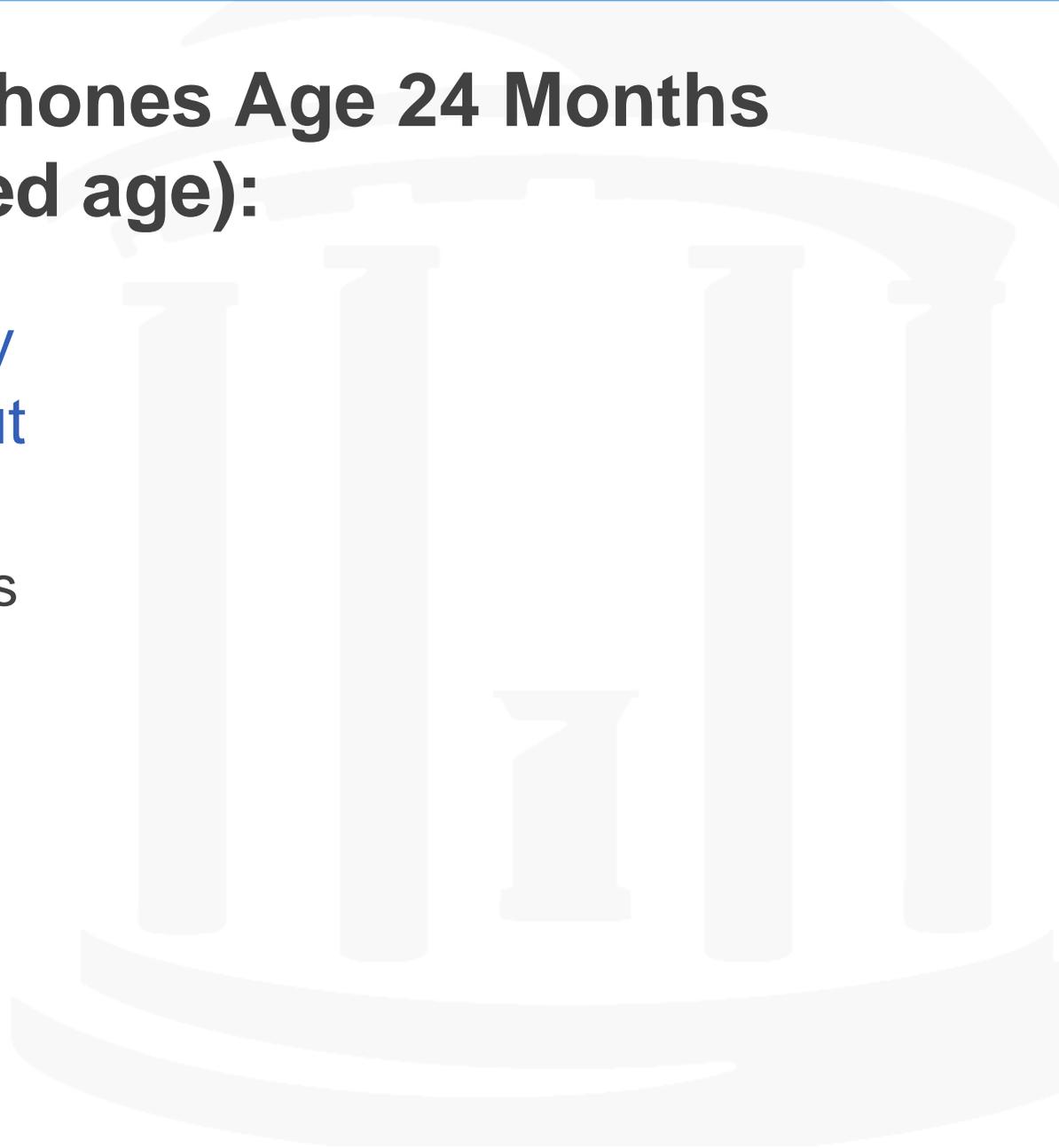
Child conditioned for play
audiometry procedure but
limited attention span

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

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



CASE #3

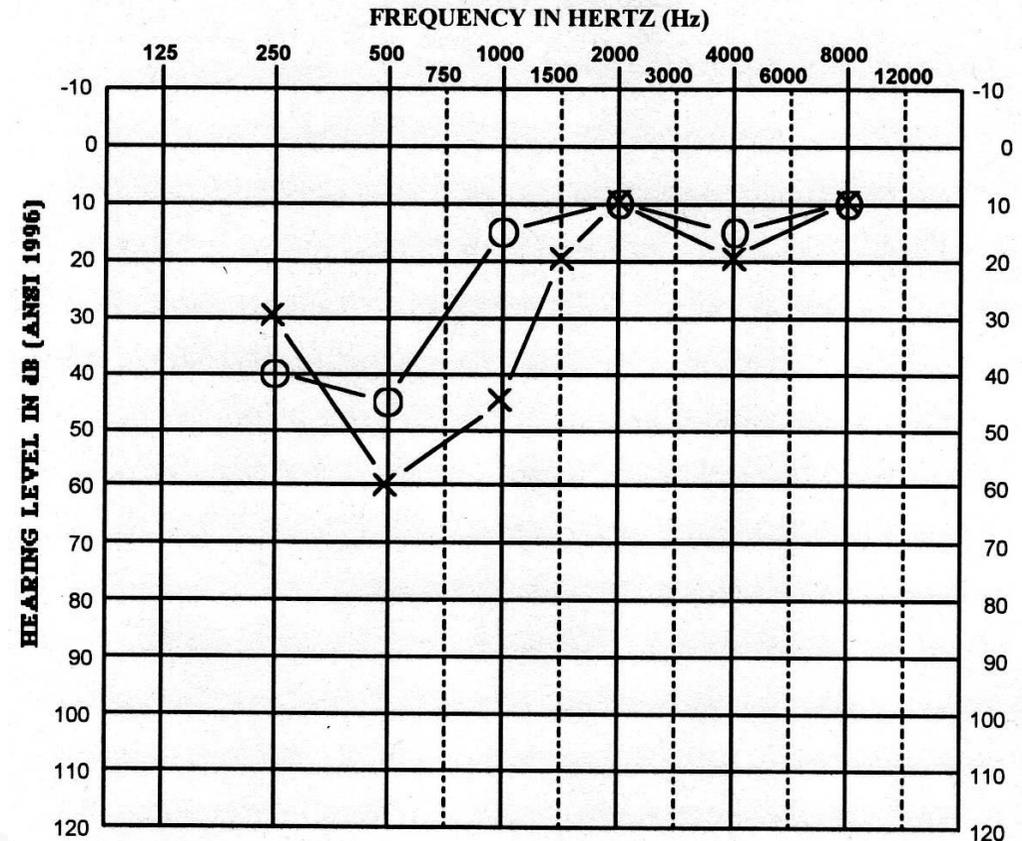
**Child with HA in one ear CI in other
Hx of progressive neurologic disease**

Case # 3 Background

- Born at full term
- Developmental milestones normal until two years of age
- At age two, child developed peripheral neuropathies including optic neuropathy
- Hospitalized at age 3 and underwent several diagnostic studies including:
 - » Electromyography
 - » Muscle Biopsies
- Numerous medical consultations including:
 - » Otolaryngology, Genetics, Neurology, Ophthalmology and Infectious Disease
- Etiology for her medical problems never determined
 - » Guillain-Barre, Charcot-Marie-Tooth and mitochondrial disease were all ruled out

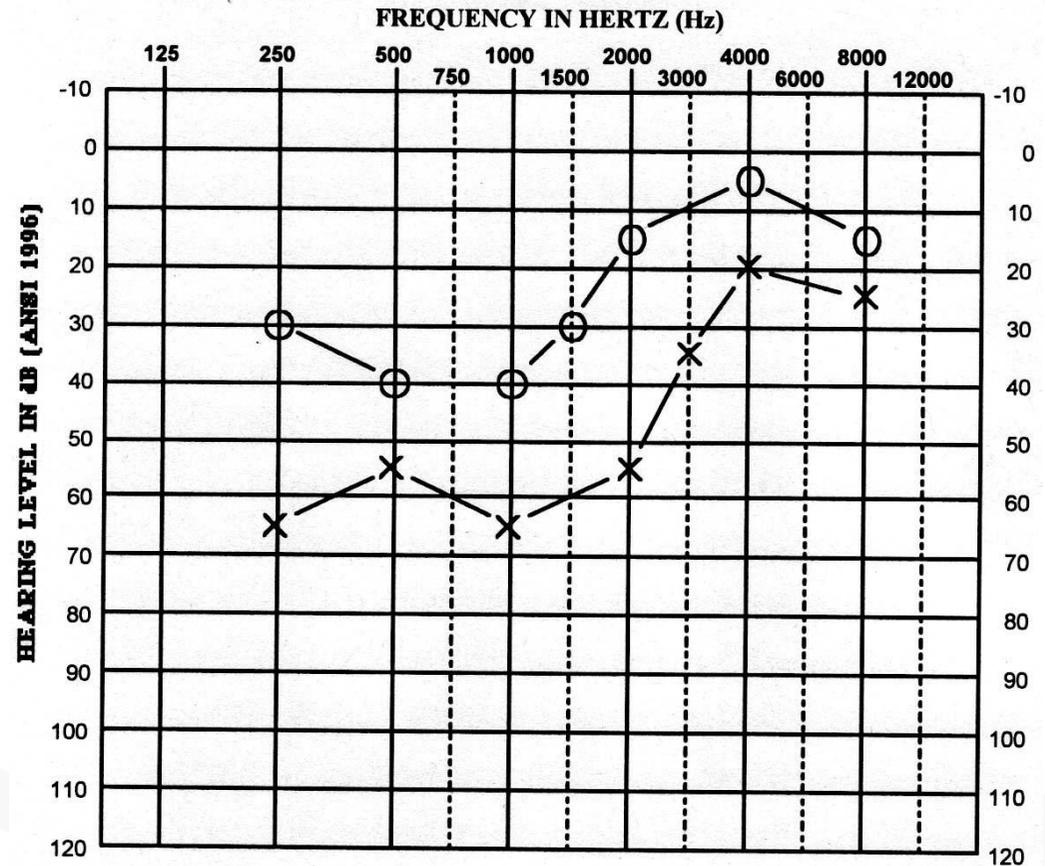
Audiogram at Age 6 years

- Bilateral rising pattern
- Unaided speech recognition using monosyllabic words:
 - » 100% right
 - » 84% left



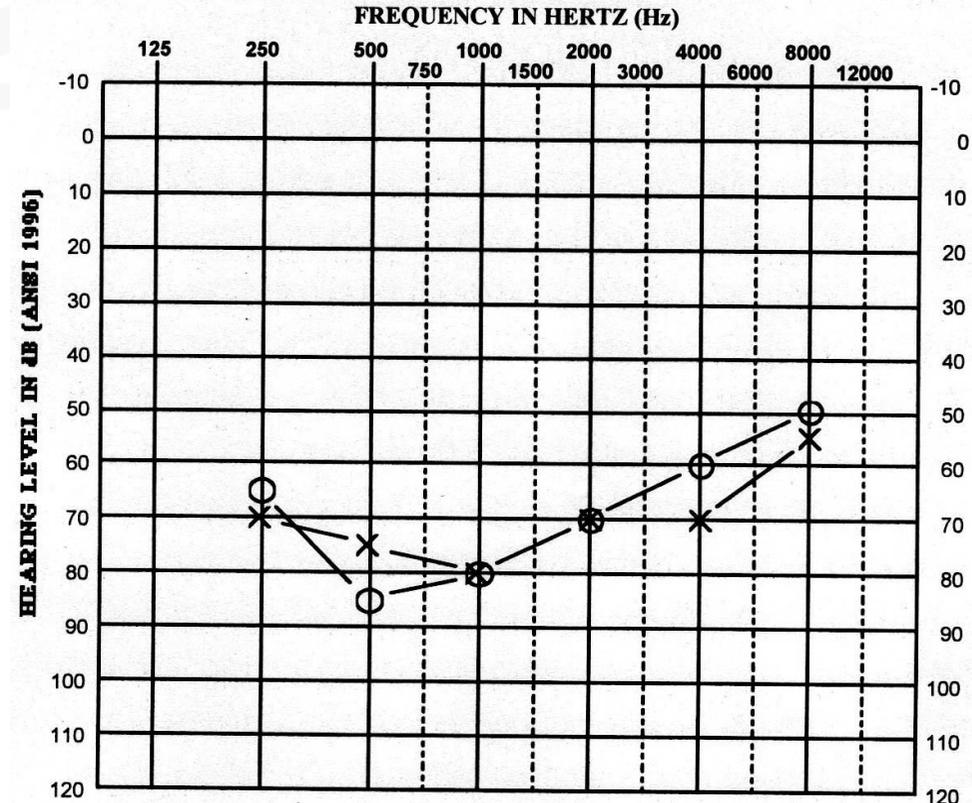
Audiogram Age 10 years

- Bilateral rising pattern: mild on right, moderate left
- Unaided speech recognition using monosyllabic words:
 - » 40% right
 - » 24% left

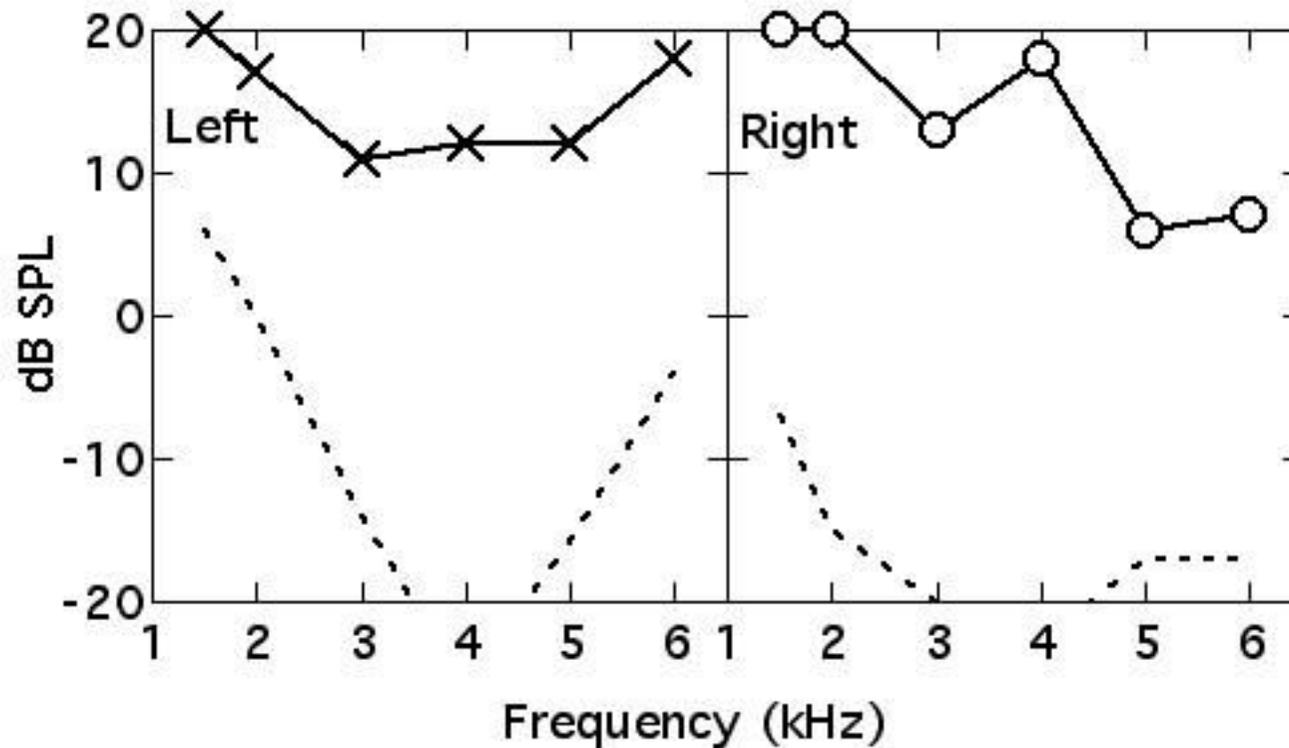


Audiogram at Age 11 years

- Child continued to have fluctuating speech recognition scores
- ABR showed AN pattern
- Hearing aids tried but child and family reported minimal benefit
- Despite significant residual hearing, child was no longer able to repeat any words on monosyllabic word test
- Successful communication could only be accomplished at close range with speech reading



Otoacoustic Emissions Age 11 years



Follow up

- Family counseled extensively regarding potential benefits and limitations of cochlear implantation, particularly in view of multiple peripheral neuropathies
- After careful consideration, family decided to proceed with left CI
- After one year of use with CI:
 - » Monosyllabic words: 32% words, 66% phonemes
 - » Parents reported that while she continued to have significant communication difficulty, they felt need for repetitions was reduced with device on

Follow up

- After 4 years of device use:
 - » Monosyllabic words: 20%
 - » Continued deterioration in motor abilities
 - » Parents report significant difficulty understanding anyone other than family members
 - » Since child still had significant residual hearing in her right ear; decision made to attempt hearing aid use age.
- At age 17 and after six years of CI use and with a hearing aid in contralateral ear:
 - » Monosyllabic word score only 20%



CASE #4

Child with Kernicterus and Multiple Challenges



Background

- First child; born at 36 weeks gestation
- Passed initial AABR screen
- Sent home with initial bilirubin of 15 mg/dL
- Became lethargic; parents brought child to ER; abnormal neurologic exam with bilirubin level of 45 mg/dL
- Admitted to PICU; underwent double volume exchange transfusion for severe hyperbilirubinemia, seizure activity



Background

- **Admitting Diagnosis:**
 - » Severe hyperbilirubinemia
 - » Encephalopathy
 - » Respiratory Failure
- **Discharge Diagnosis:**
 - » Kernicterus
 - » Resolving encephalopathy
 - » Resolved respiratory failure
 - » Risk of developmental delay
 - » Possible seizure activity
 - » Reflux
- 5 subsequent hospitalizations
- G-tube and J-tube placed

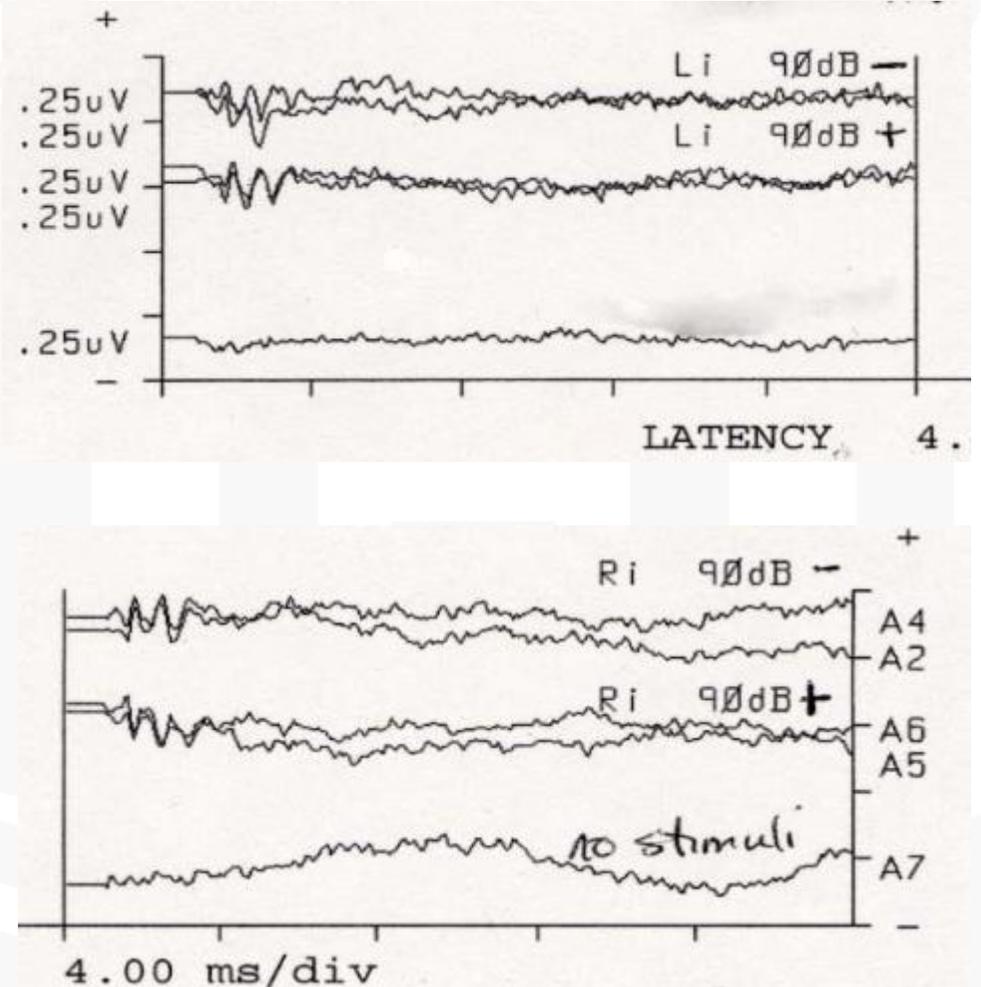


Background

- AABR repeated following exchange transfusion; child failed, bilaterally
- Diagnostic ABR at 4 weeks consistent with possible ANSD
- Repeat ABR recommended after one month
- Family contacted UNC for second opinion re diagnosis and management

Age 2 ½ months UNC diagnostic ABR

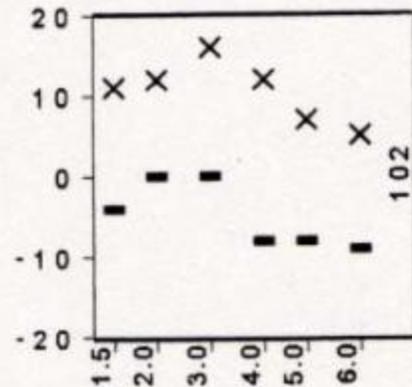
- Seen by ENT and Audiology
- ABR:
 - » Single polarity clicks
 - » No neural response at maximum intensity levels
 - » Only a cochlear microphonic present



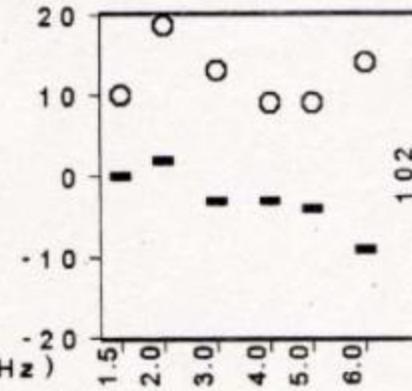


Age 2 1/2 months

Left Ear #2 Pass



Right Ear #4 Pass



X, O = Emission Level

■ = Noise Level

1.5	2.0	3.0	4.0	5.0	6.0
11	12	16	12	7	5
-4	0	0	-8	-8	-9
15	12	16	20	15	14

Freq (kHz)	1.5	2.0	3.0	4.0	5.0	6.0
DP	10	21	13	9	9	14
NF	0	2	-3	-3	-4	-9
DP-NF	10	19	16	12	13	23

OAEs present
Acoustic reflexes absent



Discussion with family

- Results consistent with ANSD type hearing loss
- ABR does not allow us to estimate hearing thresholds or predict degree of communication difficulty
- Children with AN/AD exhibit thresholds ranging from WNL to profound
- Behavioral testing with VRA will be needed when child is developmentally able



Discussion with family

- Studies have shown that some children with ANSD benefit from conventional amplification while others, with similar thresholds, do not (Rance et al 2002, 2004, 2007, 2008)
- Further recommendations regarding audiological management will be made once additional information regarding child's auditory status obtained
- Referred to *Beginnings* and Early Intervention;
 - » Weekly home visits from HI teacher, SLP, OT and PT
 - » Home health nurse with child 40 hours/week

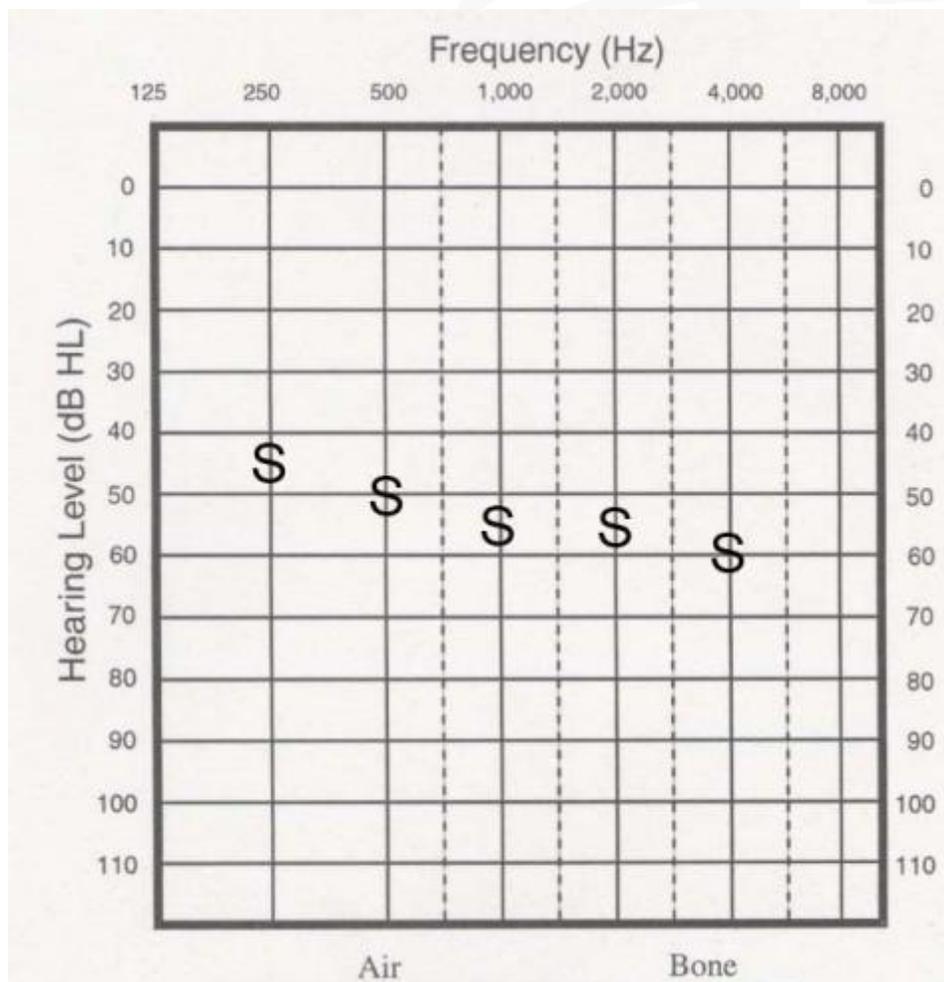


Evaluation Timeline

- 10 months: VRA attempted without success but child gave startle to NBN at moderate levels
 - » OAEs reduced as compared to test at 2 ½ months
- 11 months: CAEP completed with no response at 500Hz or 1000Hz. (Rance et al, 2002; Pearce et al, 2007)
 - » OAES absent left ear, reduced but present 4000-6000Hz)
- 12 months: VRA attempted; child irritable due to pain associated with recent abdominal surgery
- 13 months: VRA unsuccessful but child startled for NBN (250-2000Hz) at 90dBHL with awareness at moderate levels; child responding to variety of soft sounds at home
- Neurologist tells family he does not think child will ever be able to perform behavioral audiometry

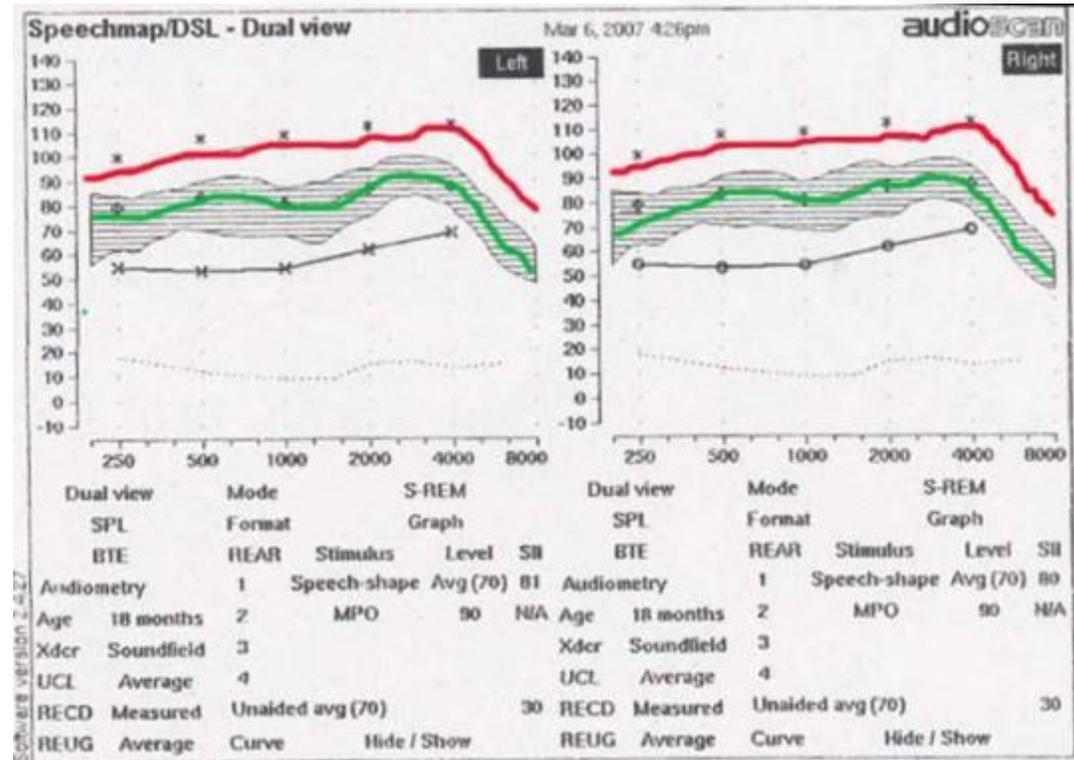


VRA in sound field 14 months



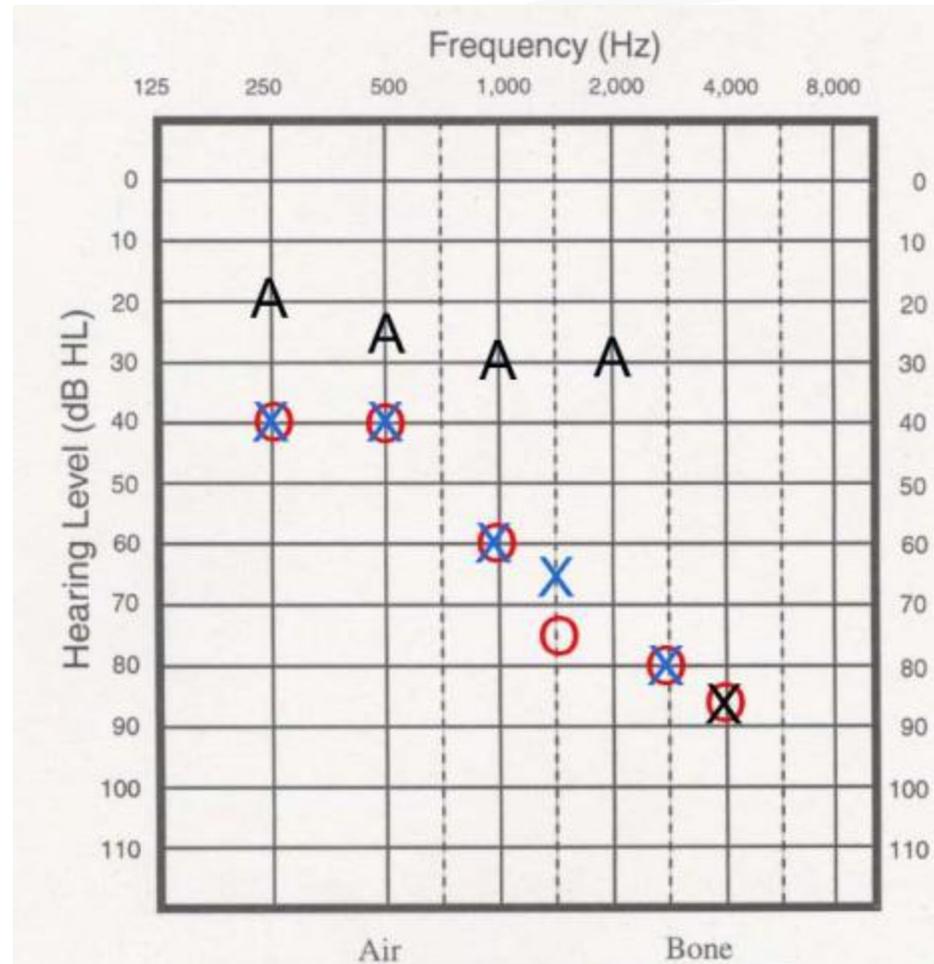
Hearing Aid Fitting Age 19 months

- Binaural digital hearing instruments dispensed
- RECDs measured
- DSL targets used
- Verified with simulated real ear measures
- Personal FM system dispensed for use at home
- Parents report increase in vocalizations following initiation of full-time hearing aid use





VRA at 20 months



Communication Options Tried or Considered

- Family's initial choice auditory/oral with cued speech support
- Signing not an option due to severe motor problems and athetoid CP
- Mom began to explore options for electronic communication board
- Arranged for meeting with SLP specialist who made useful recommendations
- Child also seen by SLP on UNC CI team



Challenges/Decisions

- Determination of what the child is able to hear and his ability to communicate what he perceives extremely challenging
- Receptive and expressive language skills at 18 months severely impaired
 - » Even with visual supplement and use of picture board, vocabulary unlikely to develop at fast enough rate to allow timely decision re CI
- After extensive discussion with family, teacher of the HI, SLPs, and CI team, decision made to proceed with monaural CI at 26 months of age,
 - » ABR prior to surgery unchanged (CM only with no neural response)

Summary of UNC Protocol for Management of Infants with ANSD

- Diagnose ANSD using ABR with single polarity clicks
- Counsel family about recommended steps in first year of life
- Enroll in early intervention
- Complete otologic exam including imaging with MRI (and CT if needed)
- Attempt behavioral audiometry with VRA beginning at 6-7 months developmental age
- Fit child with hearing aids as soon as behavioral thresholds have been established

Summary of UNC Protocol for Management of Infants with ANSD (continued)

- Set hearing aids to match targets for gain and output using prescriptive formula
- Perform hierarchical battery of speech perception tests as soon as possible
- Regularly communicate with early intervention teacher and parent re communication progress
- Consider CI if benefit from amplification insufficient for continued progress in communication skill development
- Use cortical evoked potentials to aid with management when needed
- Refer for comprehensive developmental evaluation when child has complex needs

Challenges/Unresolved Issues

- Decisions regarding use of hearing technologies is delayed until behavioral thresholds can be established
- Management of children who are able to provide behavioral thresholds but whose cognitive problems prevent the evaluation of outcomes in timely manner
- Management of children who have such significant developmental delays that it will not be possible to establish behavioral thresholds

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.
- Better ways to predict who will benefit from amplification vs cochlear implantation
- Continued research needed into the role of CAEP and other electrophysiologic tests in evaluation and management

Selected References and Resources

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THANK YOU!
Questions??

Patricia Roush, AuD

Holly Teagle, AuD

Department of Otolaryngology

University of North Carolina

School of Medicine

email:

proush@unch.unc.edu

Holly_teagle.@med.unc.edu

