DTAS Dansk Teknisk Audiologisk Selskab

Auditory Neuropathy
What do we know at this time
Current best practice for diagnosis

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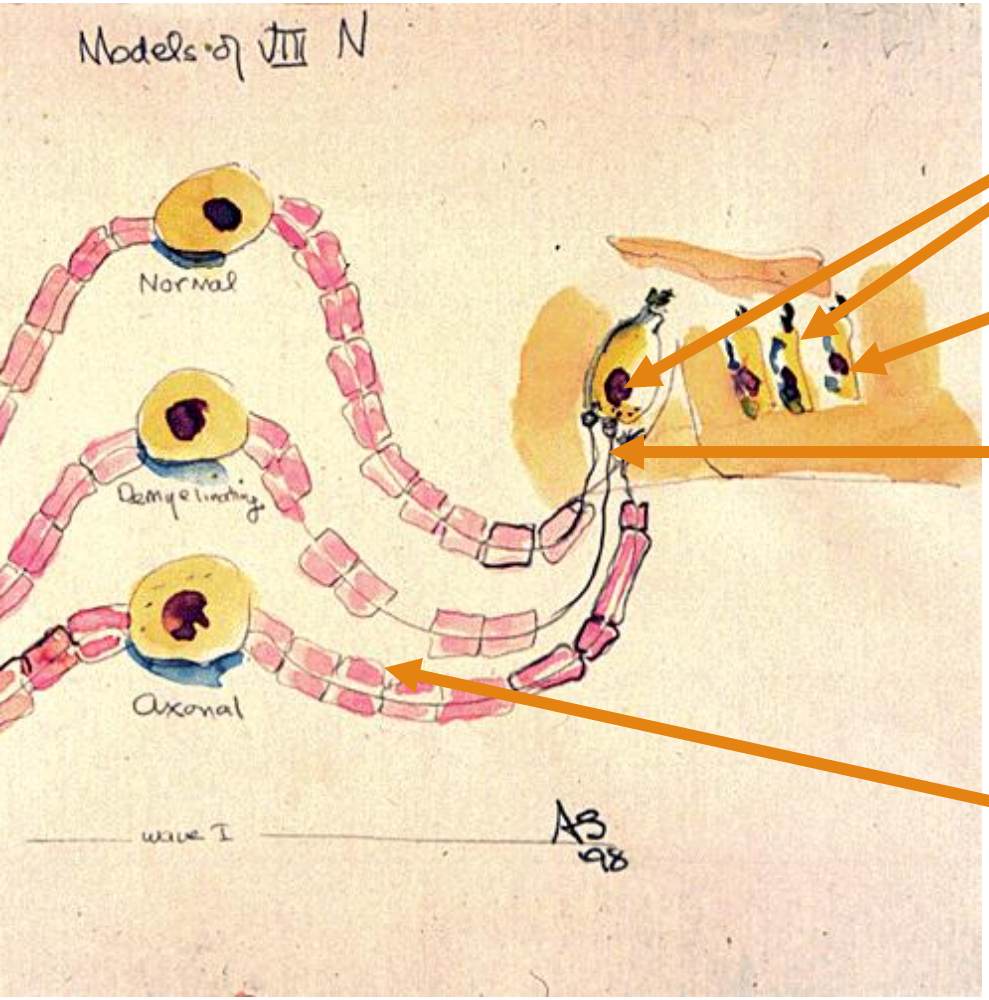
"Auditory Neuropathy"

Typical Audiologic Profile

- Hearing Loss (normal-profound) - sensorineural pattern
- Absent or severely abnormal ABR (regardless of hearing)
- Cochlear Microphonic present in ABR recording (larger than normal)
- Otoacoustic Emissions present but sometimes disappear
- Poor speech perception (relative to sensory loss of the same degree)
- No Acoustic (middle ear muscle) Reflex

Potential Sites of Lesion in AN Based on Symptoms



CM present from either IHC OHC or both

OAE (outer hair cells) Functioning

Inner hair cell function or synapse could be involved.

ABR Wave I Abnormal- (Peripheral Auditory Nerve Involved (?)

10% of Children with Hearing Loss Identified in Newborn Period will have an Auditory Neuropathy Pattern

Study	Hearing Loss	# AN	% AN
Kraus et al 1984	48	7	14.6
Cone-Wesson et al 2000	56	3	5.3
Rance et al., 1999	109	12	11
New South Wales 2004	52	7	13.5
Marion Downs Center 04	49	5	9
TOTAL	314	34	10.8

What is the Incidence of AN in Infants and Children?

Effectiveness of Population-Based Newborn Hearing Screening in England: Ages of Interventions and Profile of Cases. Kai Uus, MD, PhD, John Bamford, PhD
PEDIATRICS Volume 117, Number 5, May 2006

17 of 169 children found with permanent sensorineural hearing loss had a pattern of ANSD.

10.05%

Etiology

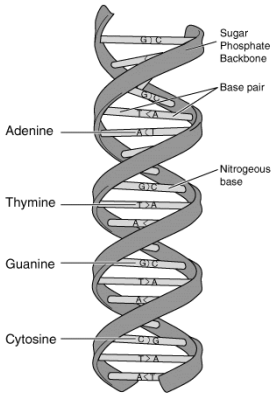
- **50-60% of children with ANSD will have significant birth histories.**
- **The remaining 40-50% of cases should be explained by a genetic disorder.**

Associated Medical Conditions

Study	N	Family Hx	Premature/ LBW	Hyper-bilirubin	Hypoxia/ Vent	Ototoxic Meds
Sininger	26	42% (11)	8% (2)	12%(3)	?	?
Madden	22	36%(8)	45%(10)	50%(11)	36% (8)	41% (9)
Raveh	26	15%(4)	31% (8)	27%(7)	?	23% (6)
Dowley	12	?	Mean GA 33 wks	33%(4)	83%(10)	75% (9)
Berlin	260	16%(41)	28% (74)			?

Also Noted: Meningitis, Cerebral Palsy, IVF

Genetics of Auditory Neuropathy Spectrum Disorder



Syndromic

- **Autosomal Dominant: Spino-Cerebellar Ataxia (Friedreich's)**
- **Hereditary Motor and Sensory Neuropathy (HMSN)**
- **Charcot-Marie-Tooth CMT-I (myelin) CMT-II (axonal)**
- **OPA1 (Dominant Optic Atrophy)**
- **Autosomal Recessive or X-linked Refsum's Disease**

Non-Syndromic

- **Autosomal Dominant /AUNA-1/**
- **Autosomal Recessive /DFNB-9/OTOF /DFNB59/Pejvakin**

OTOFERLIN GENE

Yasunaga S et al (1999) A mutation in OTOF, encoding otoferlin, a FER-1-like protein, causes DFNB9, a nonsyndromic form of deafness. *Nature Genetics* 21:363-369.

Varga et al., (2003) Non-syndromic recessive auditory neuropathy is the result of mutations in the otoferlin (OTOF) gene. *J Med Genet* 40: 45-50.

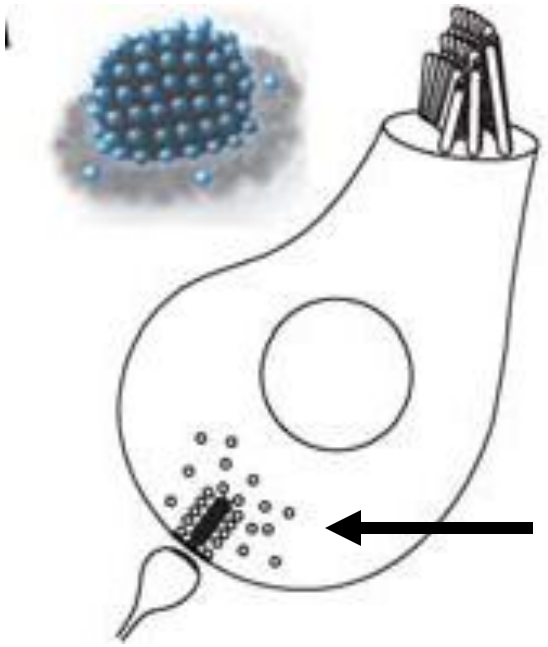
Phenotype:
Prelingual deafness, severe to profound, OAE usually present, vestibular function normal.

Called Non-syndromic, recessive auditory neuropathy (NSRAN)

Location: Chromosome 2p 22-23 gene: OTOF

Protein Otoferlin: Found in IHC, aids in vesicle transport.

Rodriguez-Ballesteros M et al (2003) Auditory Neuropathy in Patients Carrying Mutations in the Otoferlin Gene (OTOF). *Human Mutation* **22:451-456**.



OTOF mutations can explain 3.5% of non-syndromic deafness.

Otoferlin involved in vesicle-membrane fusion at inner hair cell ribbon synapse.

T. Moser et al *J Physiol* 2006

Rodriguez-Ballesteros M et al (2003) Auditory Neuropathy in Patients Carrying Mutations in the Otoferlin Gene (OTOF). *Human Mutation* **22:451-456.**

37 Subjects with two mutations of the OTOF Gene

- **24 cases were familial and 13 sporadic**
- **All hearing losses were onset < 1 year**
- **Flat audiograms >90 dB No ABR**
- **10 subjects implanted with good success**
- **21 cases were evaluated by TEOAE.**
  - **Clear, bilateral response in 6**
  - **Unilateral TEOAE in 4**
  - **One with bilateral response at 19 months gone by 26 months**
  - **10 with No OAEs.**

What are the implications of AN on the screening process?

Screening protocols that allow a pass with OAEs will miss AN and delay identification.

JCIH 2000 has mentioned AN while waiting for more data and clinical experience.

JCIH 2007 "The definition has been expanded … to include neural hearing loss (eg, "auditory neuropathy/dys-synchrony") in infants admitted to the neonatal intensive care unit (NICU)" "Separate protocols are recommended for NICU and well-baby nurseries. NICU babies admitted for greater than 5 days are to have auditory brainstem response (ABR) included as part of their screening so that neural hearing loss will not be missed."

Survey of Parents of Children with AN (N=12)

	Screening Technology	
	OAE (5)	ABR (5)
Pass	3	0
Refer	2	5
Age at Diagnosis	8.6 months 11 months (Pass)	4.4 months 2 months (omit 1 OAE FU)

AN
Diagnotic Procedures

Auditory Neuropathy Consensus Group

Kai Uus **Yvonne Sininger** **Deborah Hayes** **Ferdi Grandori**

Barbara Cone **Pat Roush** **Chuck Berlin** **Jon Shallop**

Not pictured: Arnie Starr, Christine Petit, Gary Rance

Consensus

Diagnostic Criteria

1. Tests of cochlear hair cell (sensory) function:

   a) Otoacoustic Emissions

Use either DPOAE or TEOAE in standard diagnostic protocol

OAEs in Auditory Neuropathy

- When PRESENT along with significant hearing loss and/or abnormal ABR = positive sign for AN.
- Are known to fade in some young children with AN. The reason is unclear and CM is not affected.
- Absent OAE does not rule out AN.

Consensus

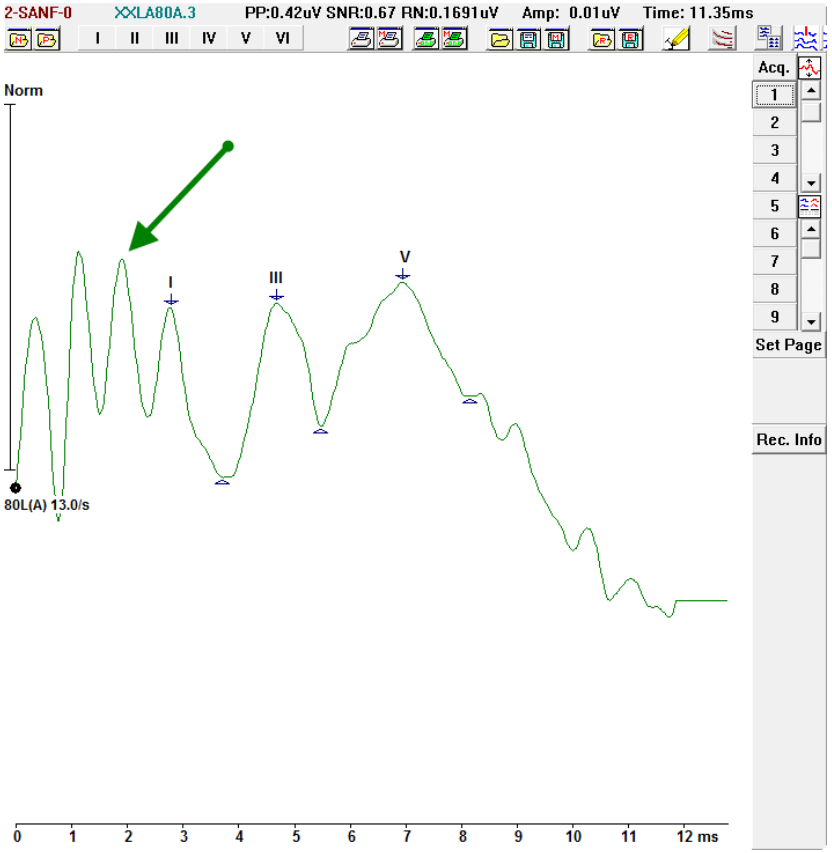
Diagnostic Criteria

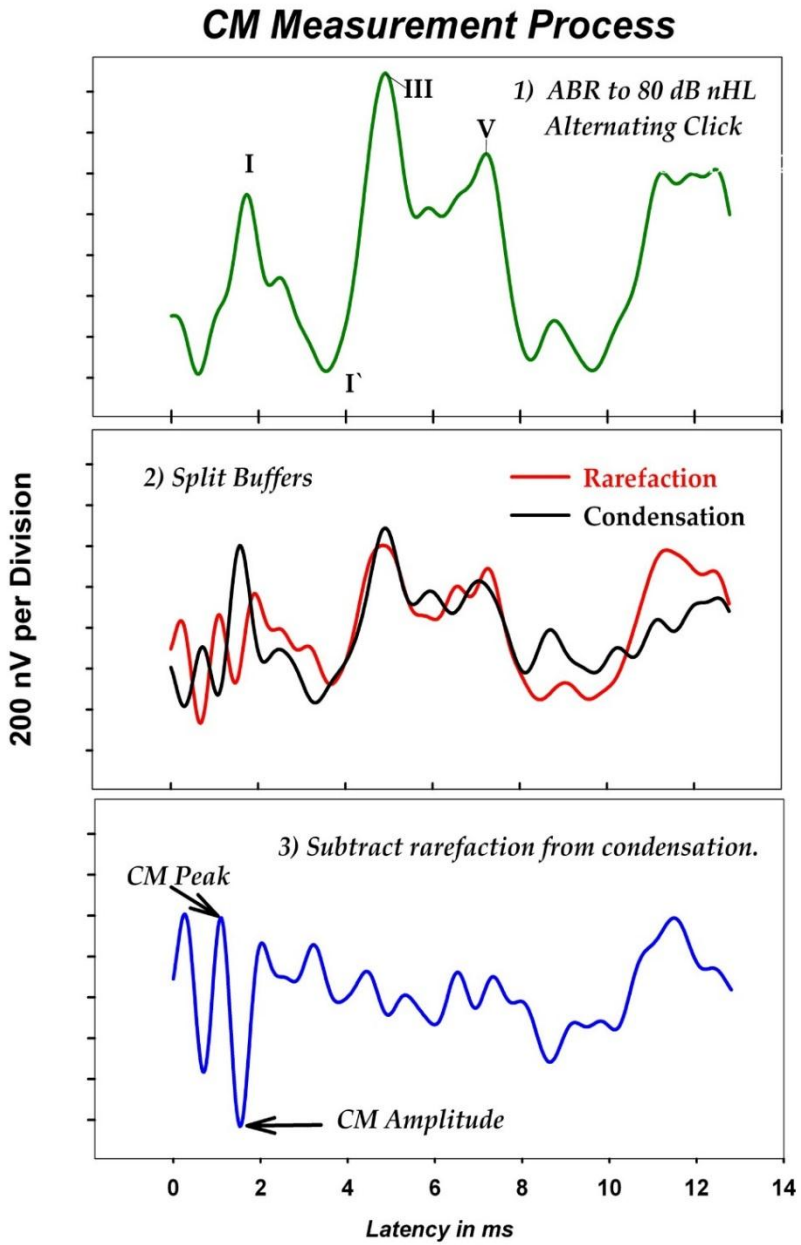
1. Tests of cochlear hair cell (sensory) function:

   b) Cochlear Microphonic

- 80-90 dB click response
- One rarefaction and one condensation average
- Insert earphones
- Clamp tubing to distinguish stimulus artifact

Normal Newborn ABR with CM

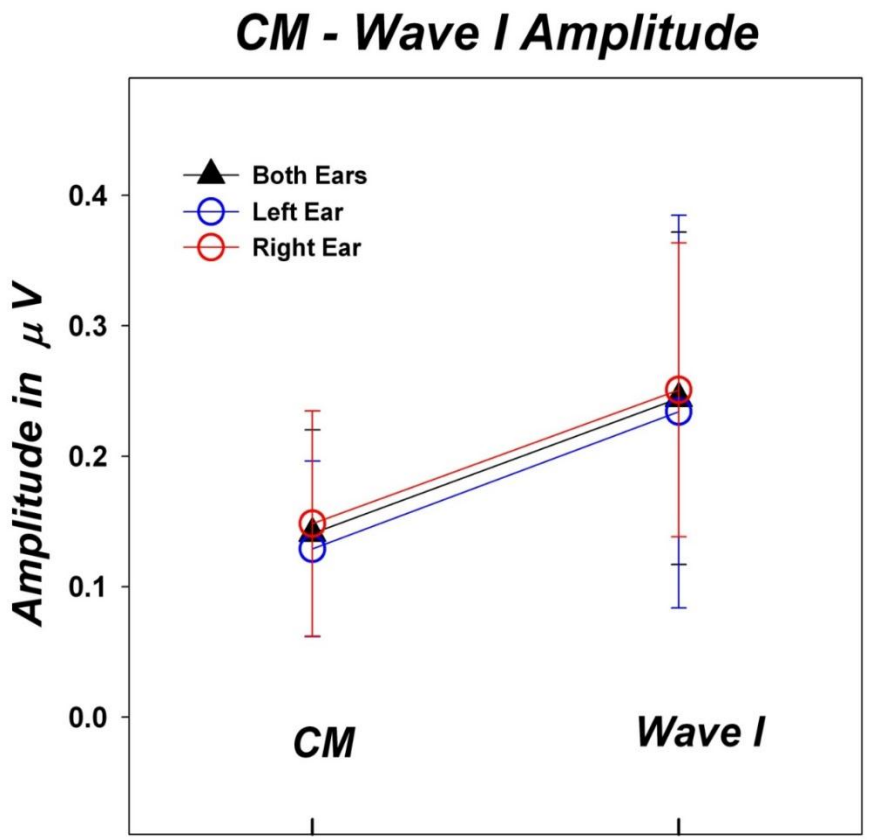


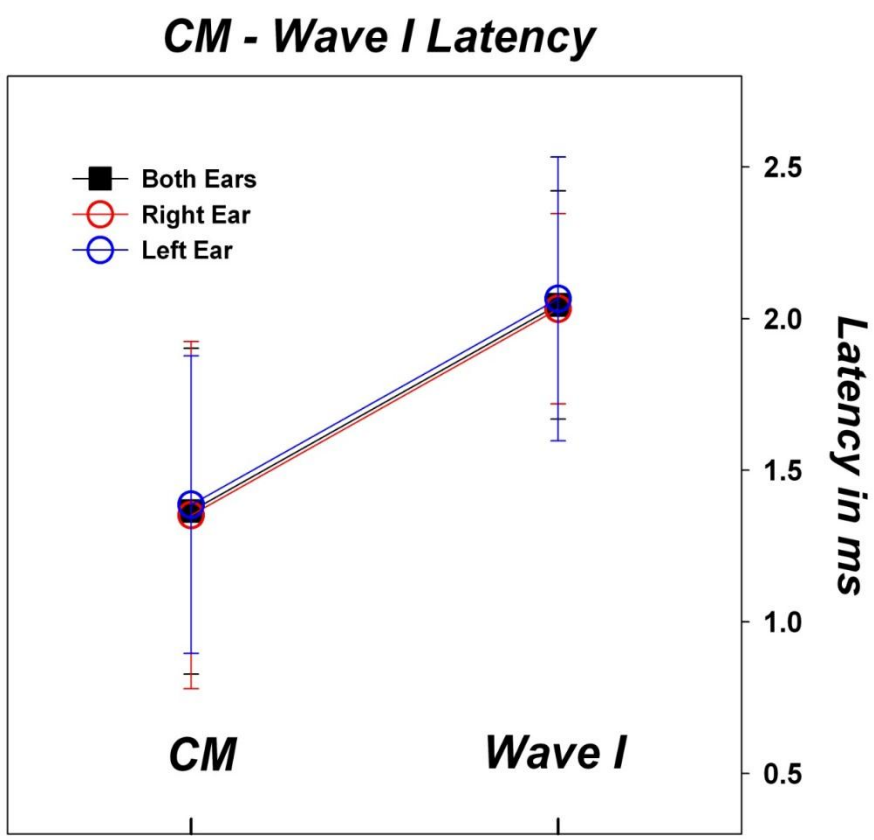


Clean average is obtained and latencies and amplitudes of peaks marked.

This response is split to condensation and rarefaction buffers.

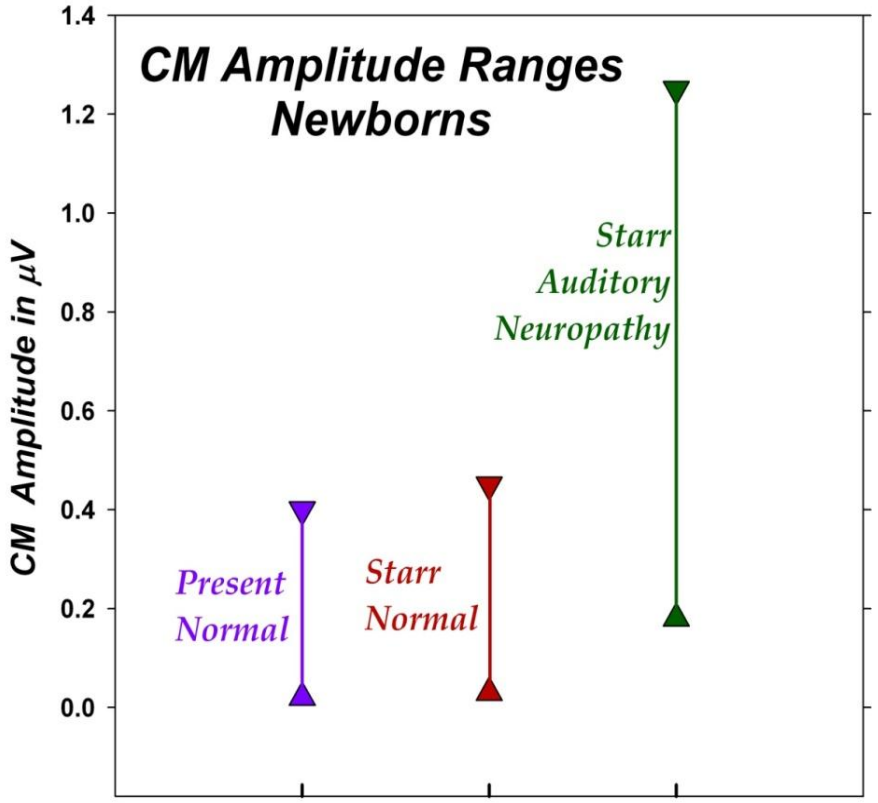
The rarefaction is subtracted from the condensation revealing the CM. The peak latency and amplitude of the CM component are computed.





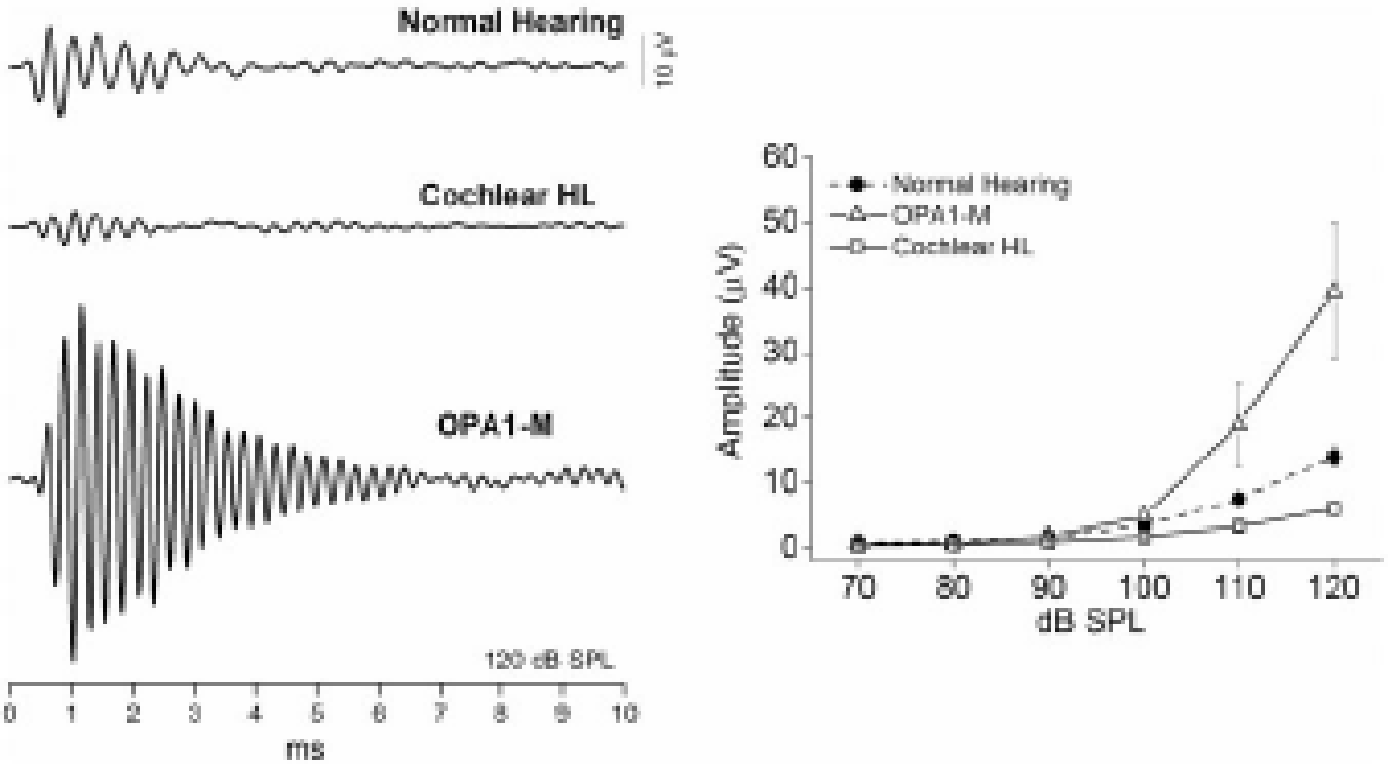
The peak amplitude and latency of the CM are plotted relative to the same measures on wave I of the ABR. Although overlap exists (error bars denote 1 standard deviation) the **CM is smaller (t= -5.095, 48 DF p< 0.0001) and earlier (t= -7.269, 48 DF, p< 0.0001) than wave I when measured in this fashion.**

CM Amplitudes from infants with Auditory Neuropathy are significantly larger than those from typical neonates.



Starr *et al.*, 2001.

CM in OPA1 Patients



Santarelli et al, Brain 2015 138:563.

Consensus
Diagnostic Criteria

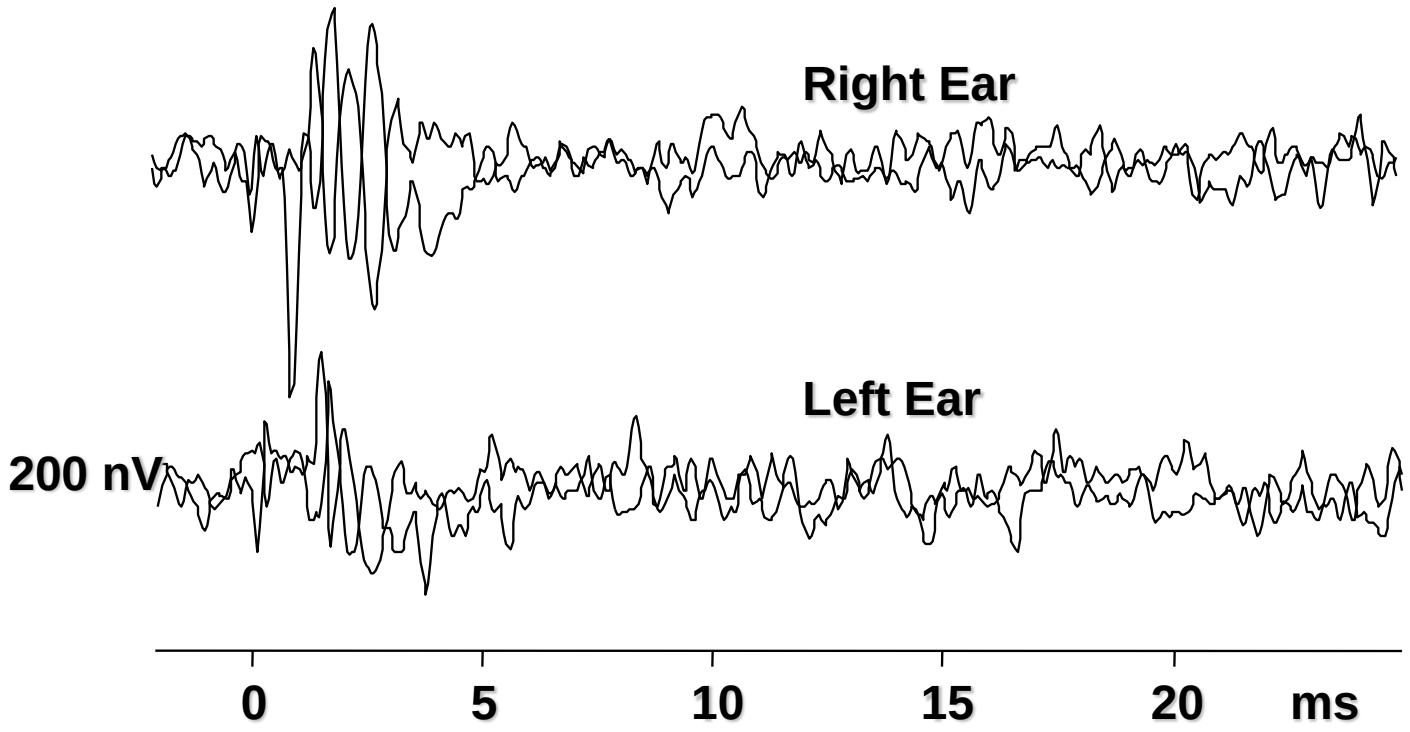
2. Tests of auditory nerve function:

   a) Auditory Brainstem Response (ABR)

- 80-90 dB click response

Auditory Brainstem Response Absent or Abnormal

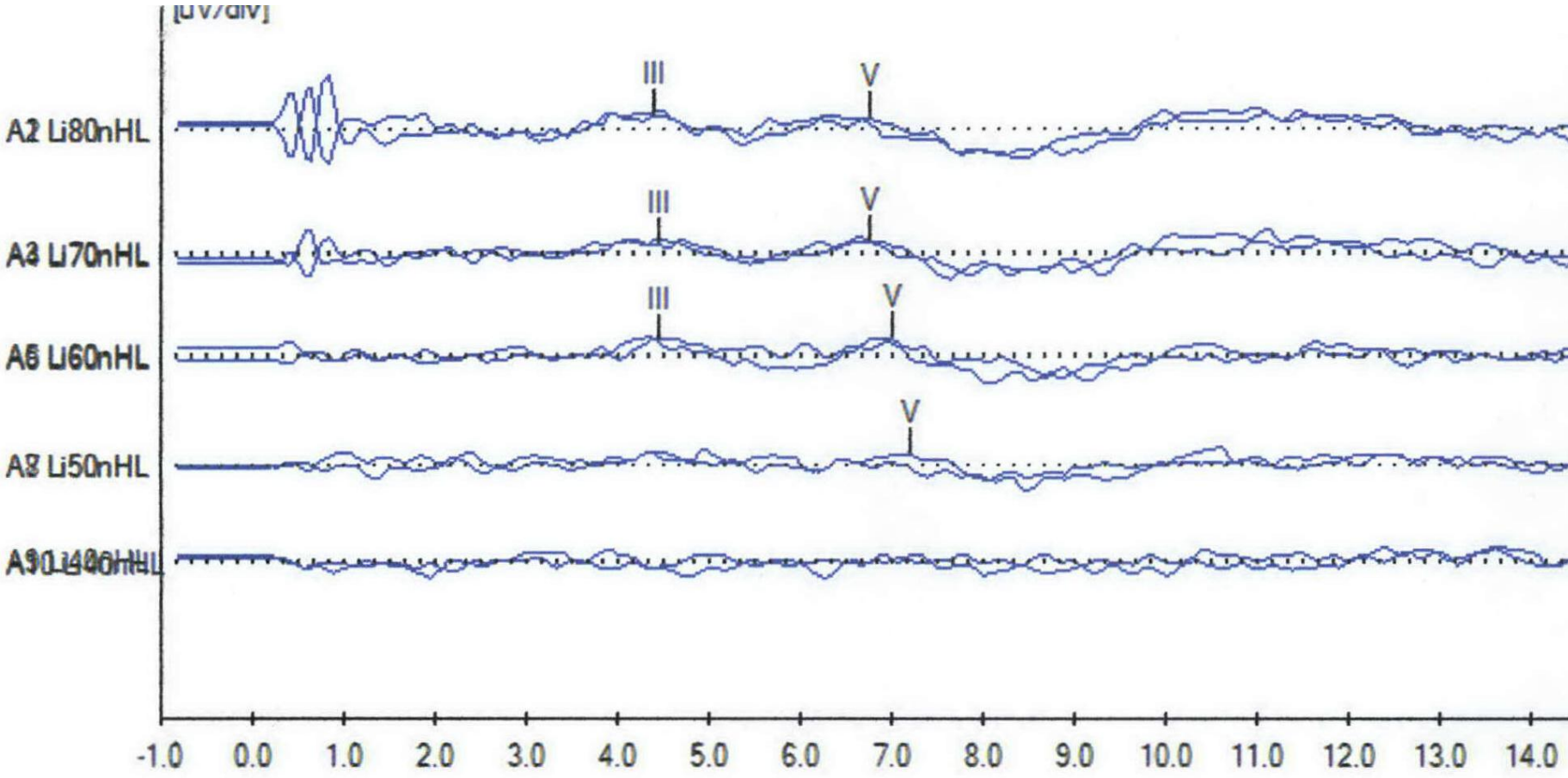
Clicks 11/s 82 dBnHL Insert earphones
Rarefaction & Condensation Overlayed



No Non-inverting (neural) waves

Large Cochlear Microphonic-
Some Waveform with Elevated Threshold

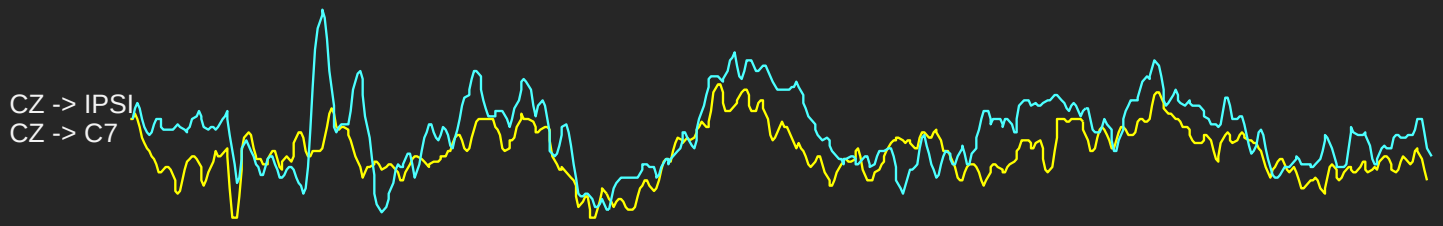
9 months old; behavioral thresholds WNL



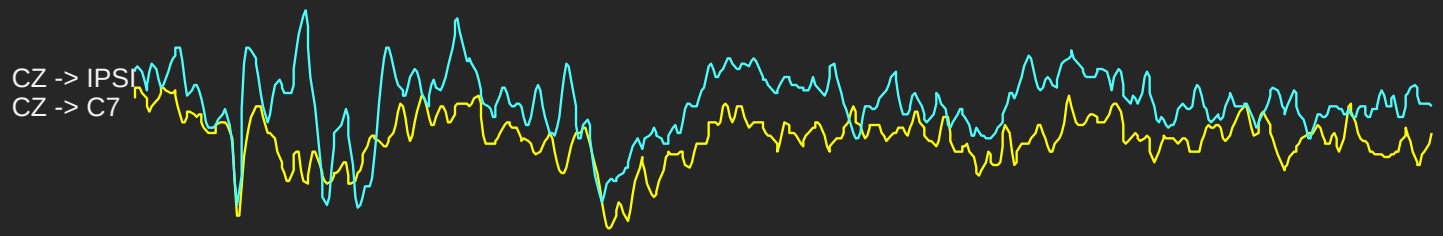
Auditory Brainstem Response

Clicks 80 dBnHL 25/s Insert Earphones
Right Ear

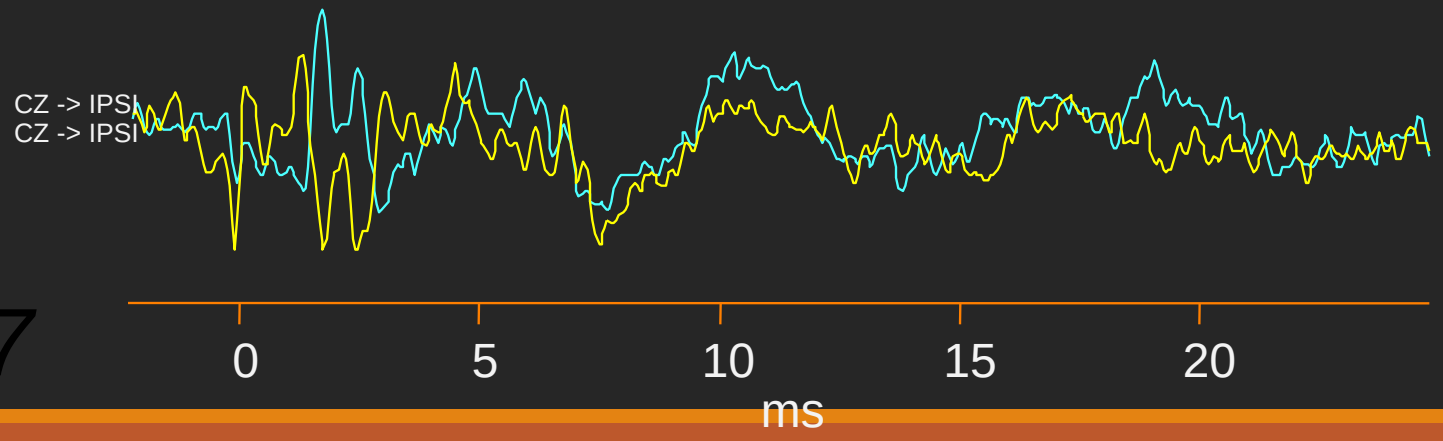
Rarefaction



Condensation



Rarefaction & Condensation Overlayed



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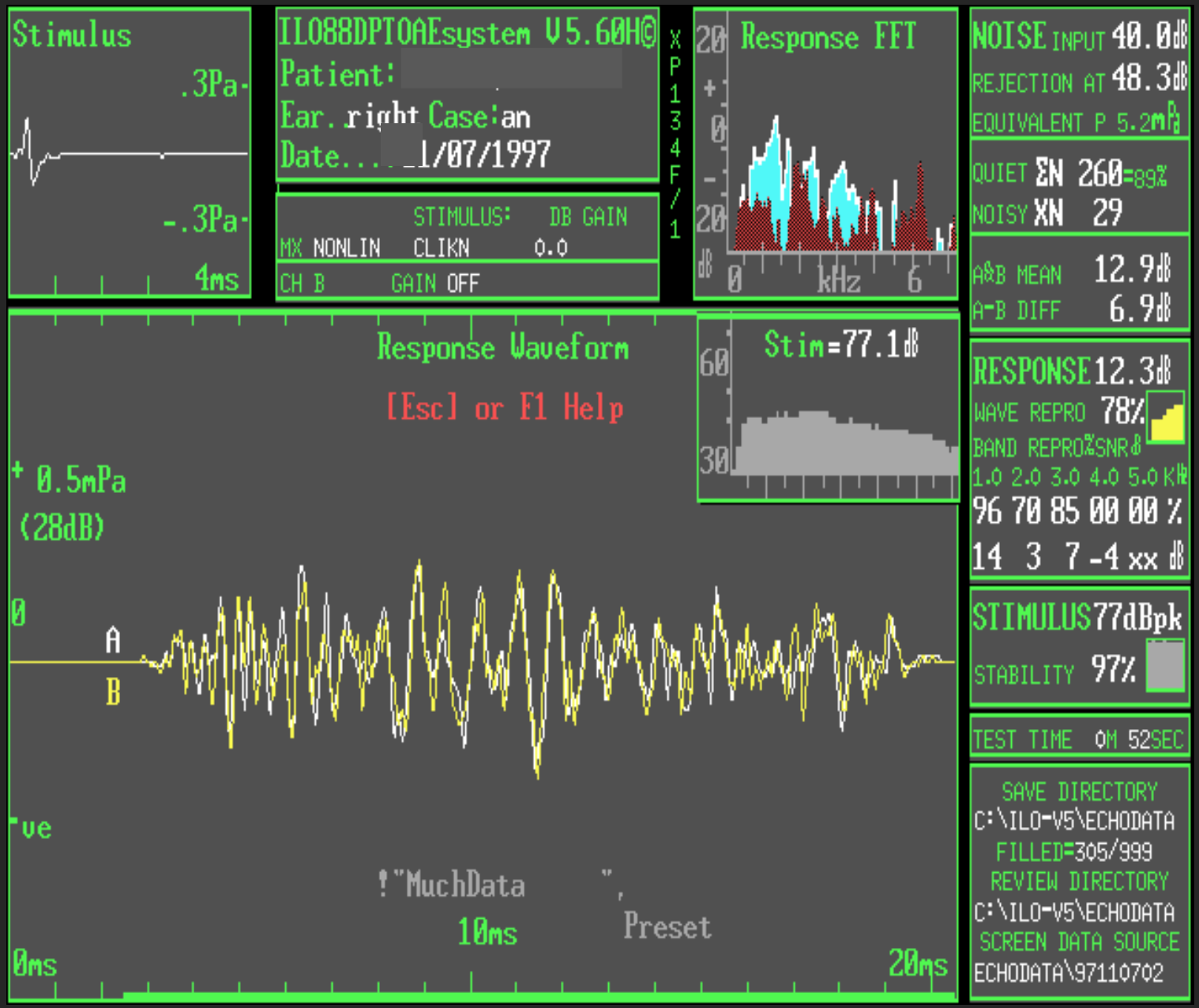
Stimulus
.3Pa-
-.3Pa-
4ms
ILO88DPTOAEsystem V5.60H©
Patient:
Ear. right Case:an
Date... 1/07/1997
STIMULUS: DB GAIN
MX NONLIN CLIKN 0.0
CH B GAIN OFF
XP134F/1
Response FFT
20
+
0
-
20
dB
0
kHz
6
NOISE INPUT 40.0dB
REJECTION AT 48.3dB
EQUIVALENT P 5.2mPa
QUIET EN 260=89%
NOISY XN 29
A&B MEAN 12.9dB
A-B DIFF 6.9dB
Response Waveform
[Esc] or F1 Help
Stim=77.1dB
60
30
RESPONSE 12.3dB
WAVE REPRO 78%
BAND REPRO%SNRdB
1.0 2.0 3.0 4.0 5.0 KHz
96 70 85 00 00 %
14 3 7 -4 xx dB
+ 0.5mPa
(28dB)
0
A
B
STIMULUS 77dBpk
STABILITY 97%
TEST TIME 0M 52SEC
SAVE DIRECTORY
C:\ILO-V5\ECHODATA
FILLED=305/999
REVIEW DIRECTORY
C:\ILO-V5\ECHODATA
SCREEN DATA SOURCE
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10ms
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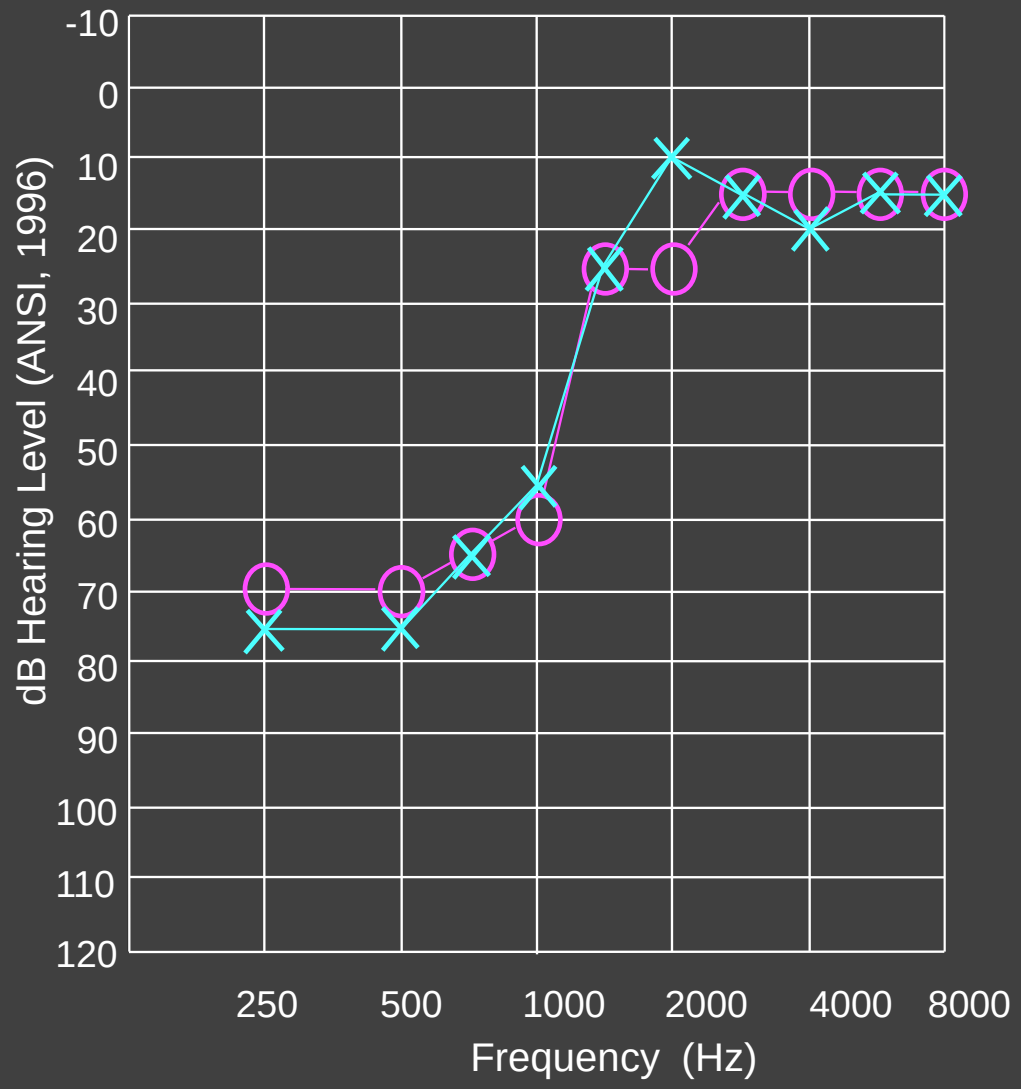
20ms

28

AUDIOGRAM



Age: 27 years

Speech Discrimination:
Right Ear = 40%
Left Ear = 64%

Tympanometry:
WNL Bilaterally

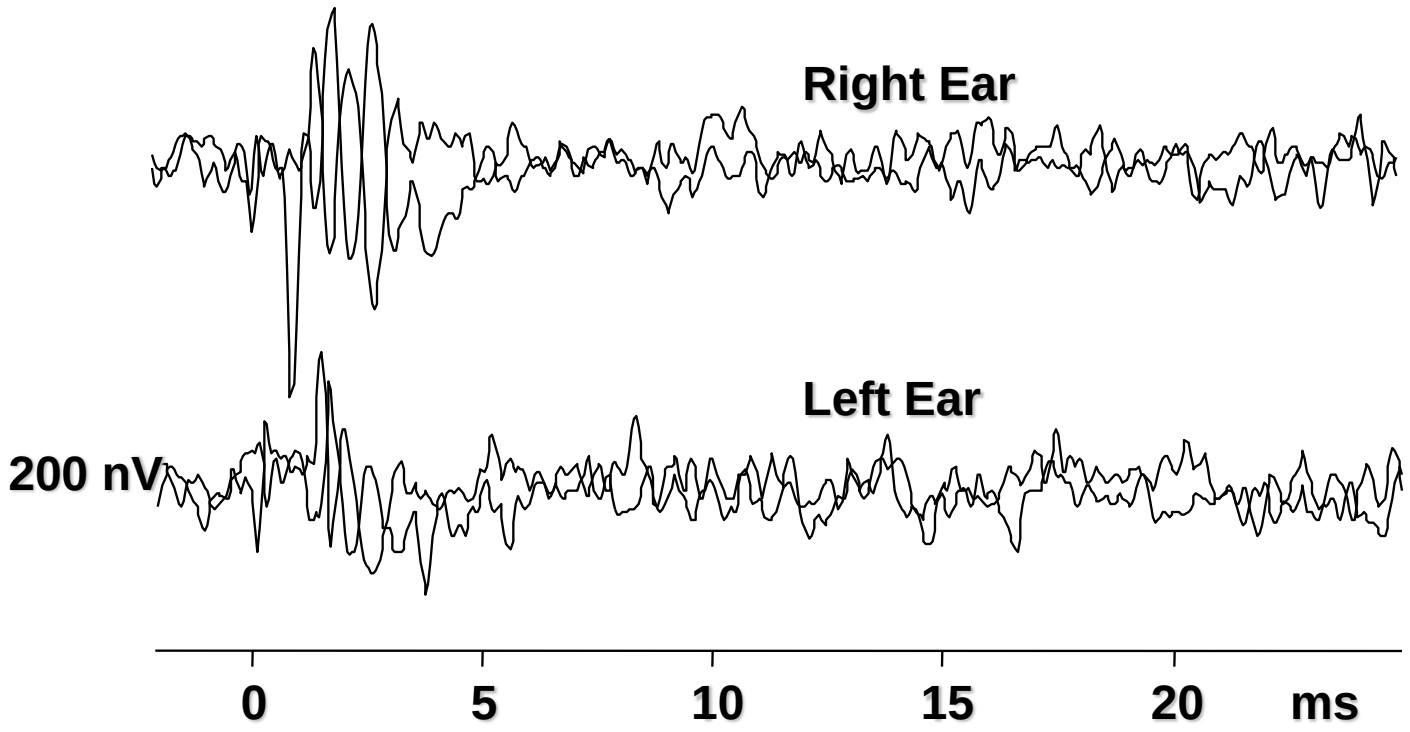
Ipsilateral Acoustic Reflex Thresholds:
Absent or Elevated Bilaterally

O Right Ear
X Left Ear

29

Auditory Brainstem Response Absent or Abnormal

Clicks 11/s 82 dBnHL Insert earphones
Rarefaction & Condensation Overlayed



Most but not all cases of AN have this ABR pattern

Some Neonates Will Show ABR Improvement Over Time!

- Madden (2002) found as many as half of infants with hyperbilirubinemia and AN improve (?) within 15 months.
- Psarommatis found 13 of 20 neonates who were followed recovered (?), most were low birthweight.
- "Improvement" is poorly documented and not well defined.

"Because "transient" ANSD has been reported in a some infants (Madden et al., 2002; Psarommatis et al., 2006; Attias and Raveh, 2007), frequent monitoring by the ANSD test battery is recommended to establish the stability of test results, especially in the first two years of life."

Consensus

Additional Tests Useful for Diagnosing Individuals with ANSD

Middle ear muscle reflexes (acoustic reflexes) are absent or elevated in individuals with ANSD (Berlin et al., 2005). Because normative data on acoustic reflex thresholds in very young infants using high probe-tone frequencies (1000 Hz) have not been established, this procedure is not required to diagnose ANSD. Nevertheless, a complete test battery for ANSD should include middle ear muscle reflex testing whenever possible.

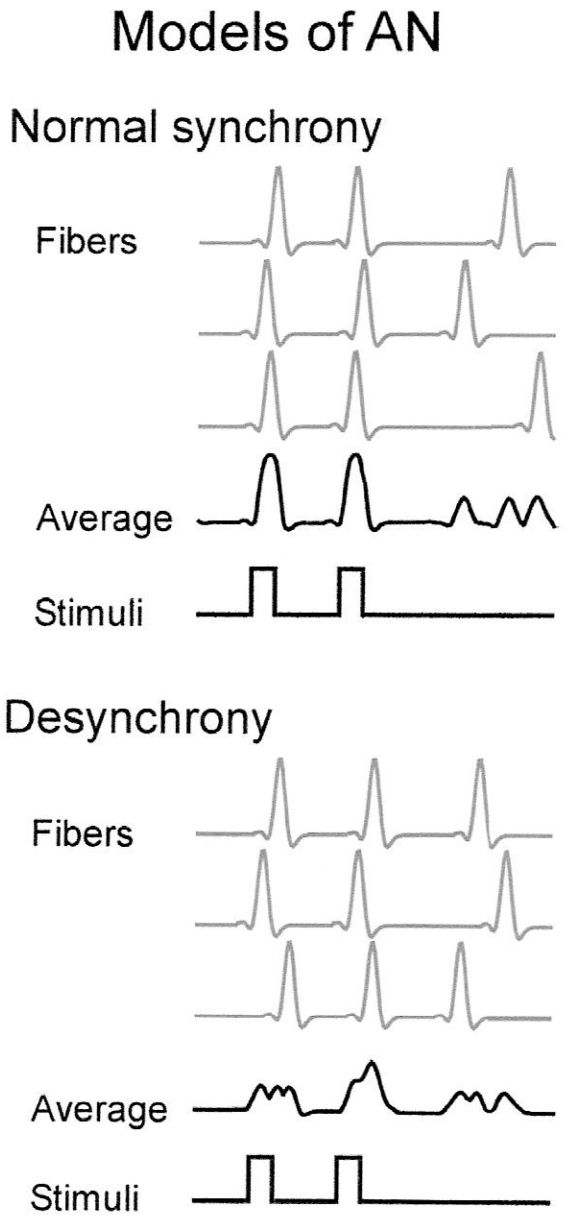
Norms for 1k probe AR measures
In infants & Children

TEMPORAL PROCESSING

PRIMARY AUDITORY DISORDER IN AN

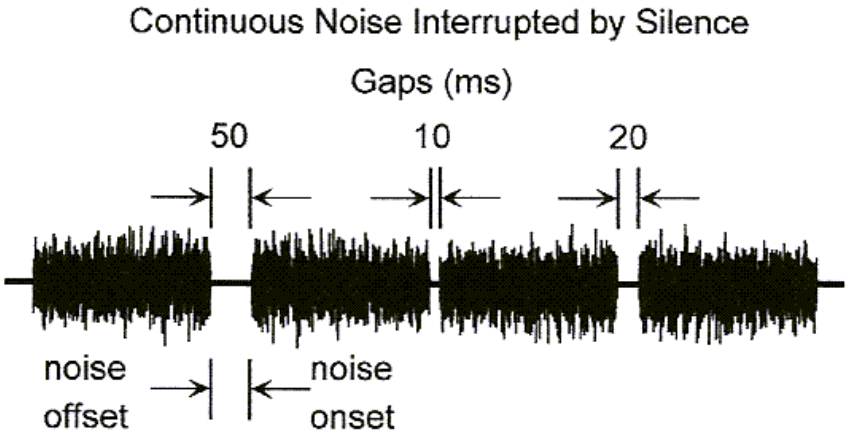
Neural disease changes the conduction properties of the nerve such that the timing is unpredictable. Individual fibers may have differing or reduced spike patterns disrupting the composite signal reaching the CANS.

This is the basis for the disruption of the ABR and speech and general perceptual tasks involving timing including localization that is experienced by patients with ANSD.

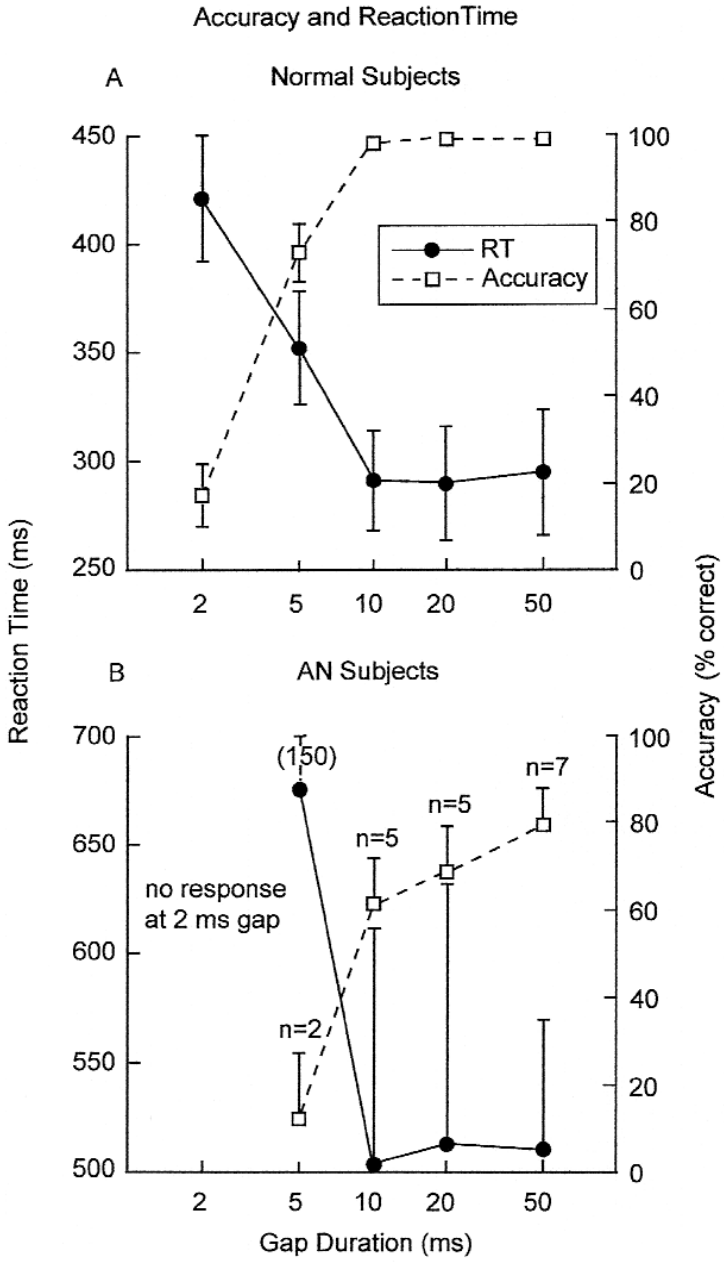


Psychoacoustics of Auditory Neuropathy

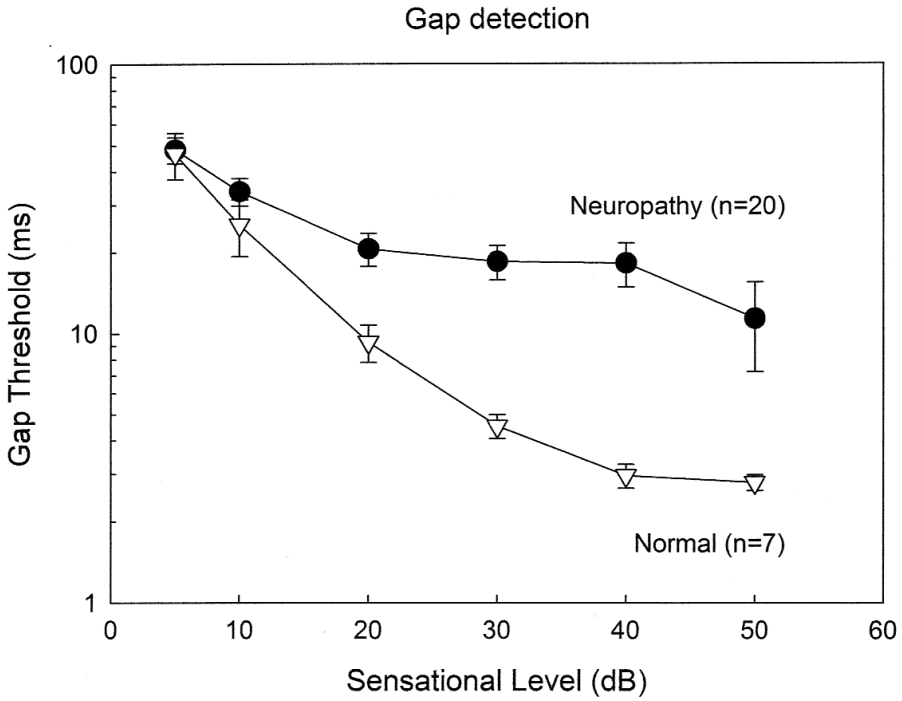
Gap Detection

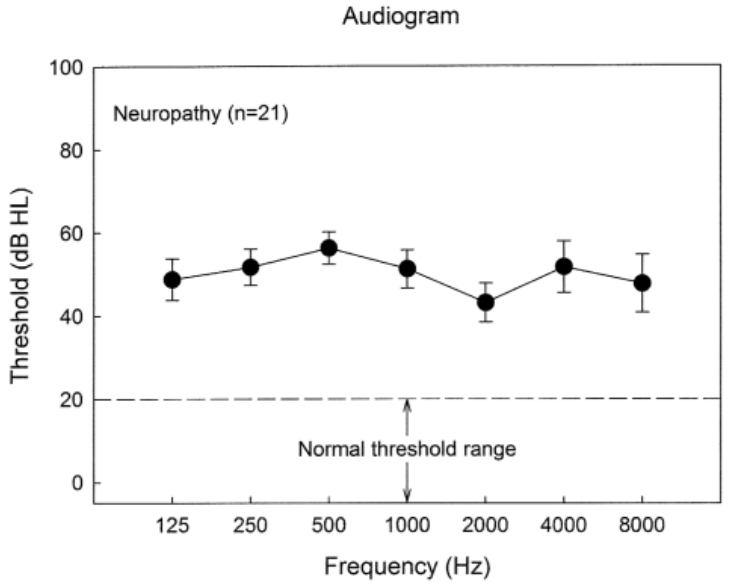


Michalewski et al 2005



Psychoacoustic Measures Indicate a Primary Temporal Processing Disorder

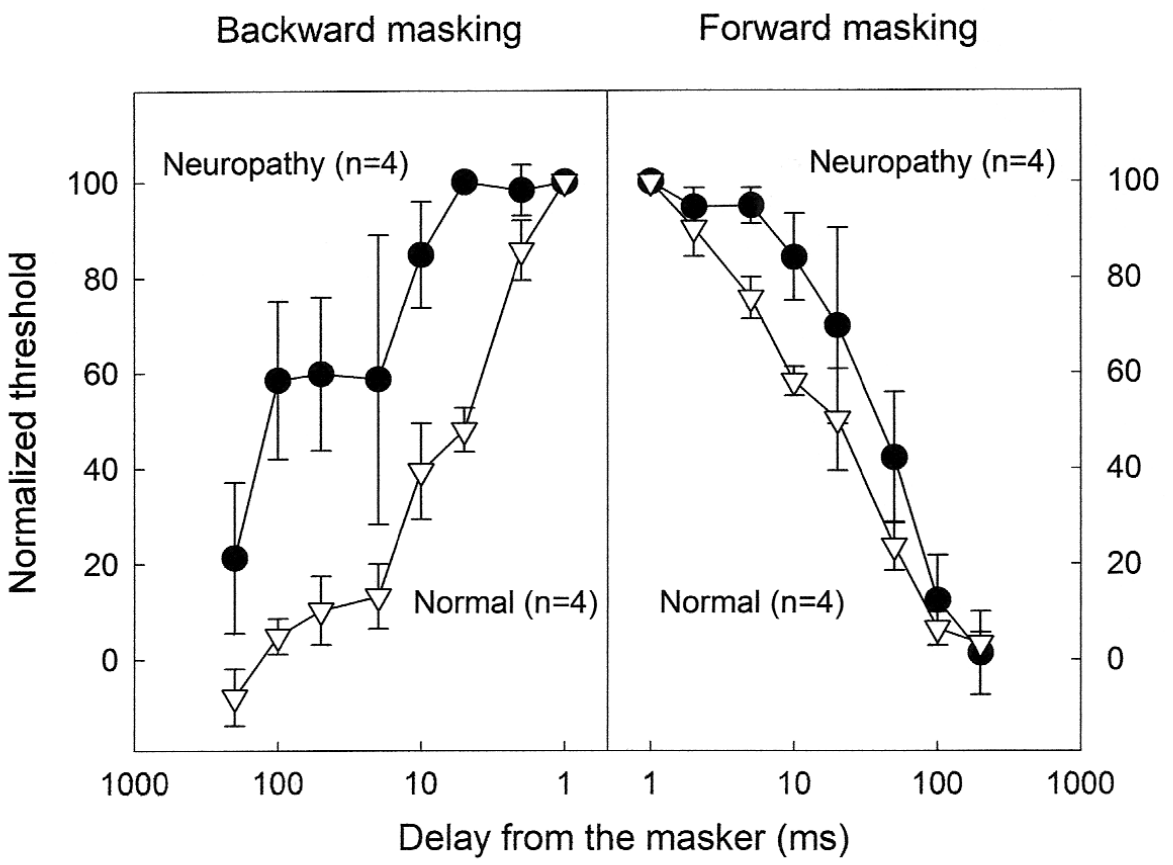




Zeng FG et al (2005) Perceptual Consequences of Disrupted Auditory Nerve Activity. *J Neurophysiol* 93 (6):3050-3063

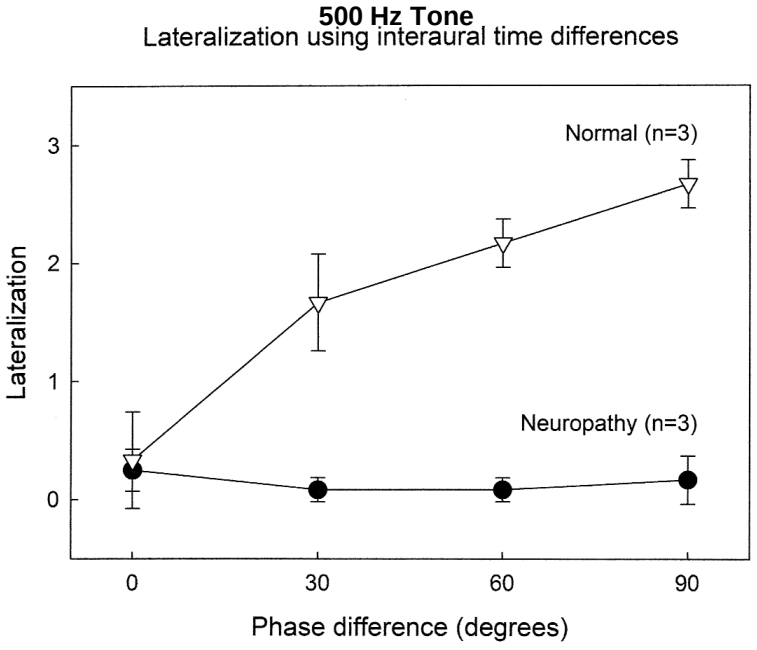
AN Patients show exaggerated effects of forward and backward masking

F.G. ZENG et al. PERCEPTUAL CONSEQUENCES OF DISRUPTED NEURAL ACTIVITIES FIG. 7

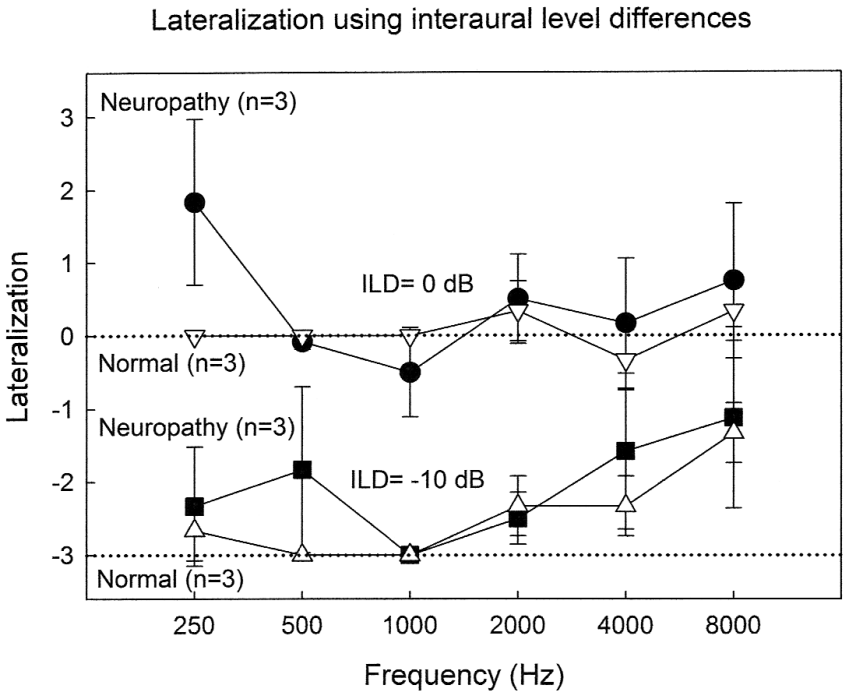


Lateralization from Timing Cues is Disrupted

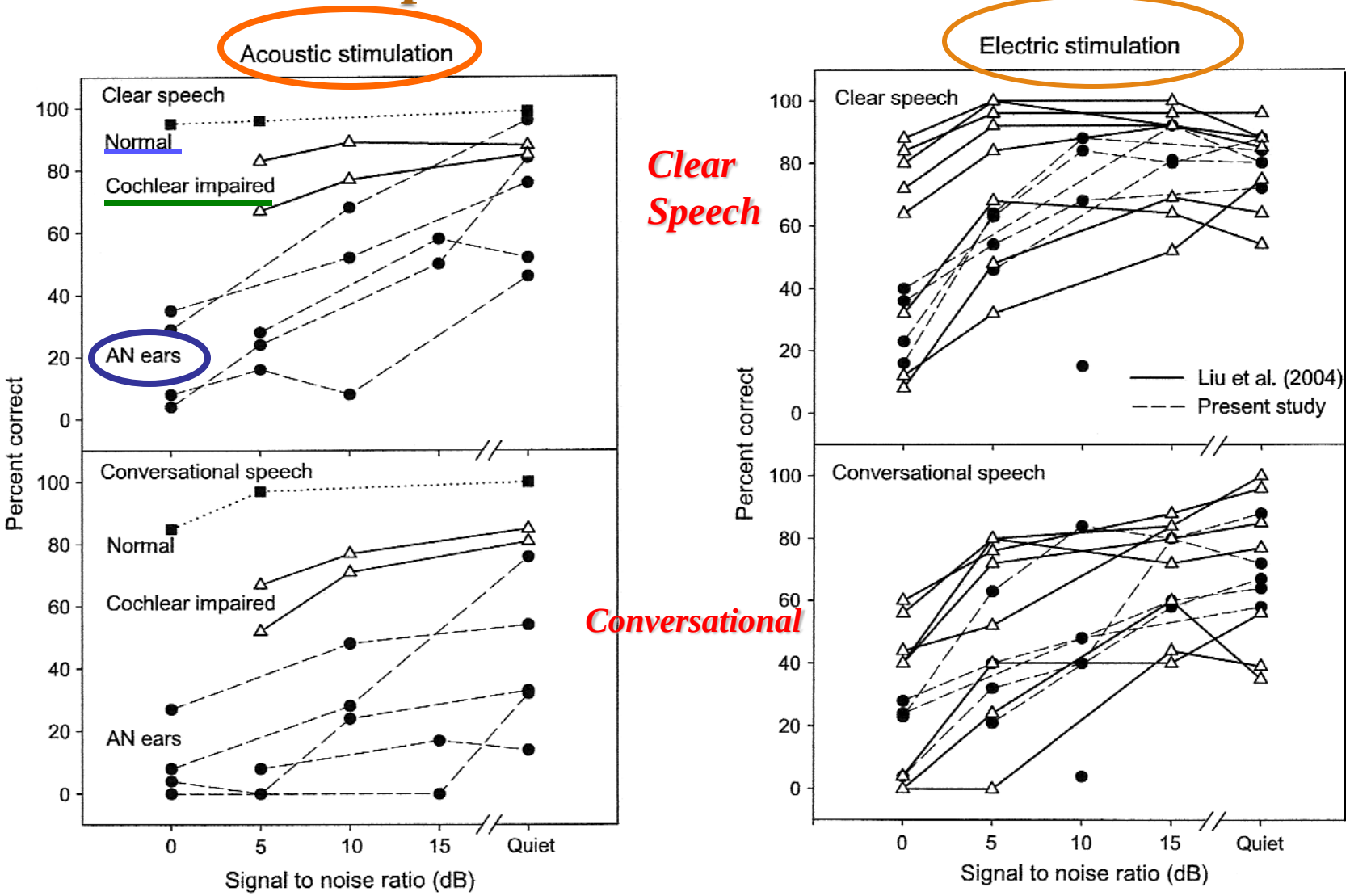
F.G. ZENG et al. PERCEPTUAL CONSEQUENCES OF DISRUPTED NEURAL ACTIVITIES FIG. 12



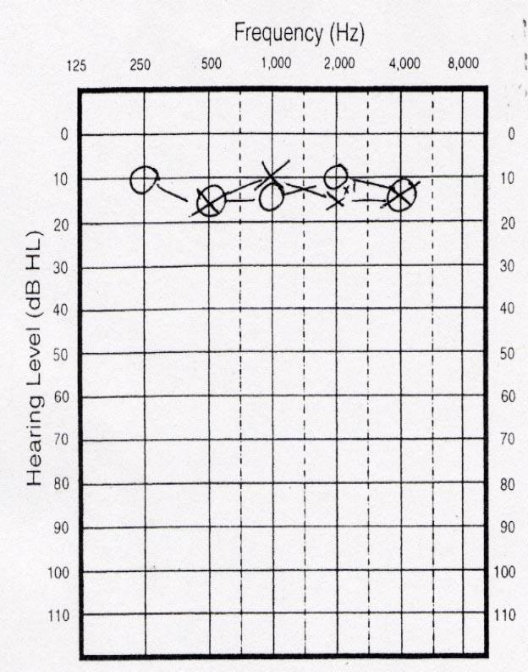
F.G. ZENG et al. PERCEPTUAL CONSEQUENCES OF DISRUPTED NEURAL ACTIVITIES FIG. 11



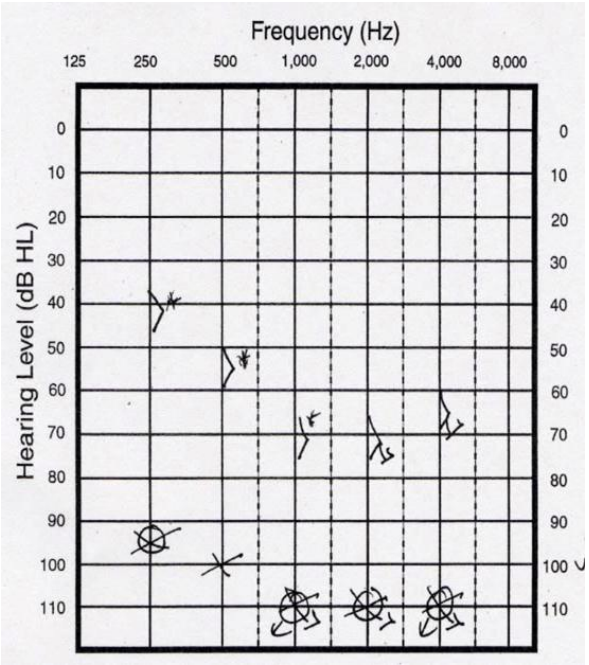
Speech Perception in Noise is Impaired in AN Subjects re Cochlear Impaired but Distinction is less with CI



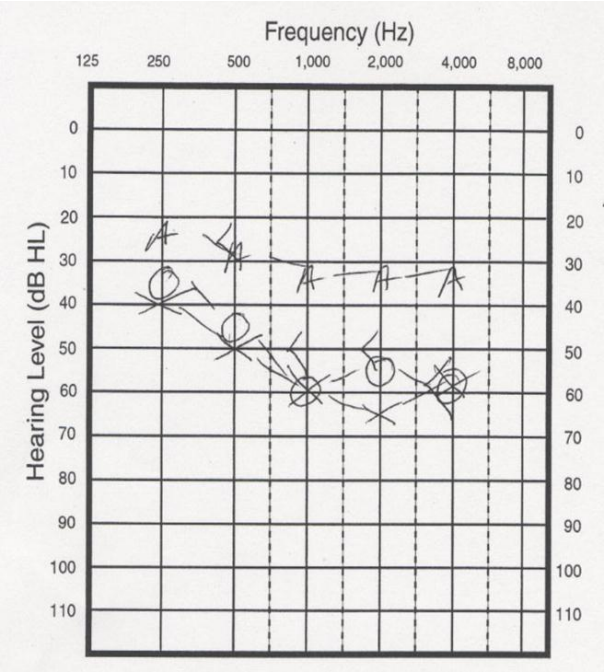
Hearing Levels Cannot Be Predicted from ABR or OAE



OAEs Present



OAEs Present



OAEs Absent

All have NO ABR, Present CM

Consensus

RECOMMENDED AMPLIFICATION STRATEGIES

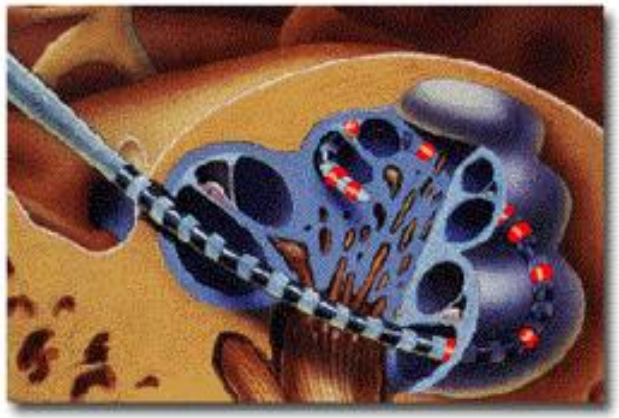
•For infants with ANSD electrophysiological methods do not predict auditory detection thresholds.

• Clinicians and parents must rely upon the infant's or young child's behavioral response to sound to guide the hearing aid fitting decision.

•If an infant or young child with ANSD demonstrates elevated pure-tone and speech detection thresholds with consistent test-retest reliability, ***hearing aid fitting should be considered*** and a trial use of hearing aids should be offered to families.

Consensus

Cochlear implants offer the possibility of **improving auditory temporal processing** by stimulating synchronous discharge of the auditory nerve. (ABR, which requires neural synchrony, can be electrically evoked in many individuals with cochlear implants)



For families who wish to consider cochlear implantation for their child with ANSD, **referral to a center with experience with managing children** with this diagnosis is strongly encouraged**.**

Cochlear Nerve Deficiency?

Buchman, CA, Roush, PA, Teagle, HF, et al. (2006). Auditory neuropathy characteristics in children with cochlear nerve deficiency. Ear and hearing, 27(4), 399-408.

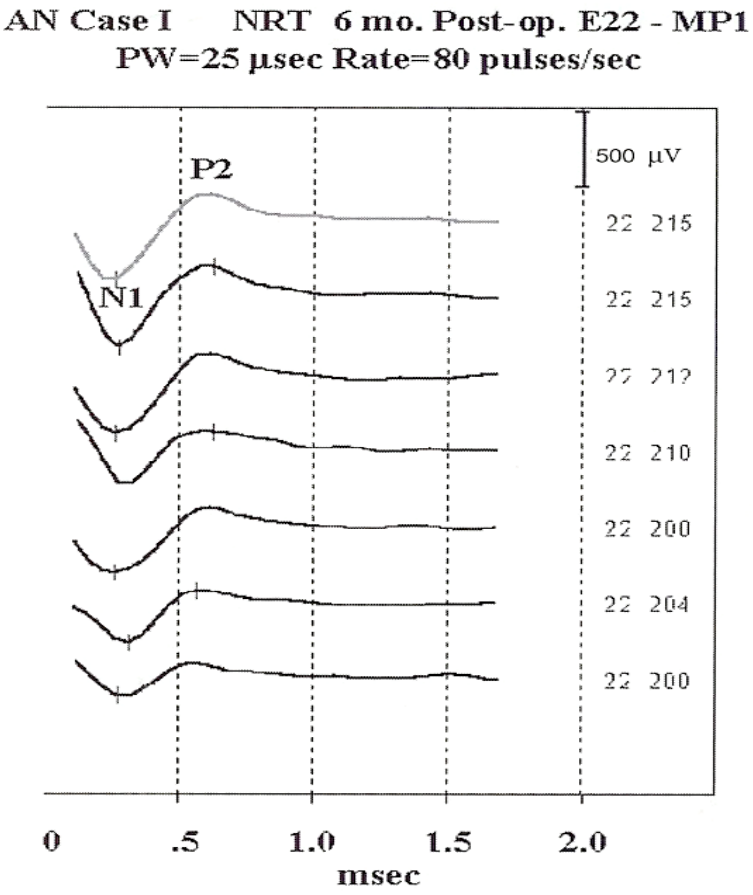
Nine (18%) of these 51 children with ABR characteristic of AN have been identified as having small (N = 2; 4%) or absent (N = 7; 14%) cochlear nerves on MRI. RESULTS: Of the nine children with cochlear nerve deficiency, five (56%) were affected unilaterally and four (44%) bilaterally.

Cochlear Implantation in Children with Auditory Neuropathy Spectrum Disorder

Teagle et al., 2010 Ear & Hearing

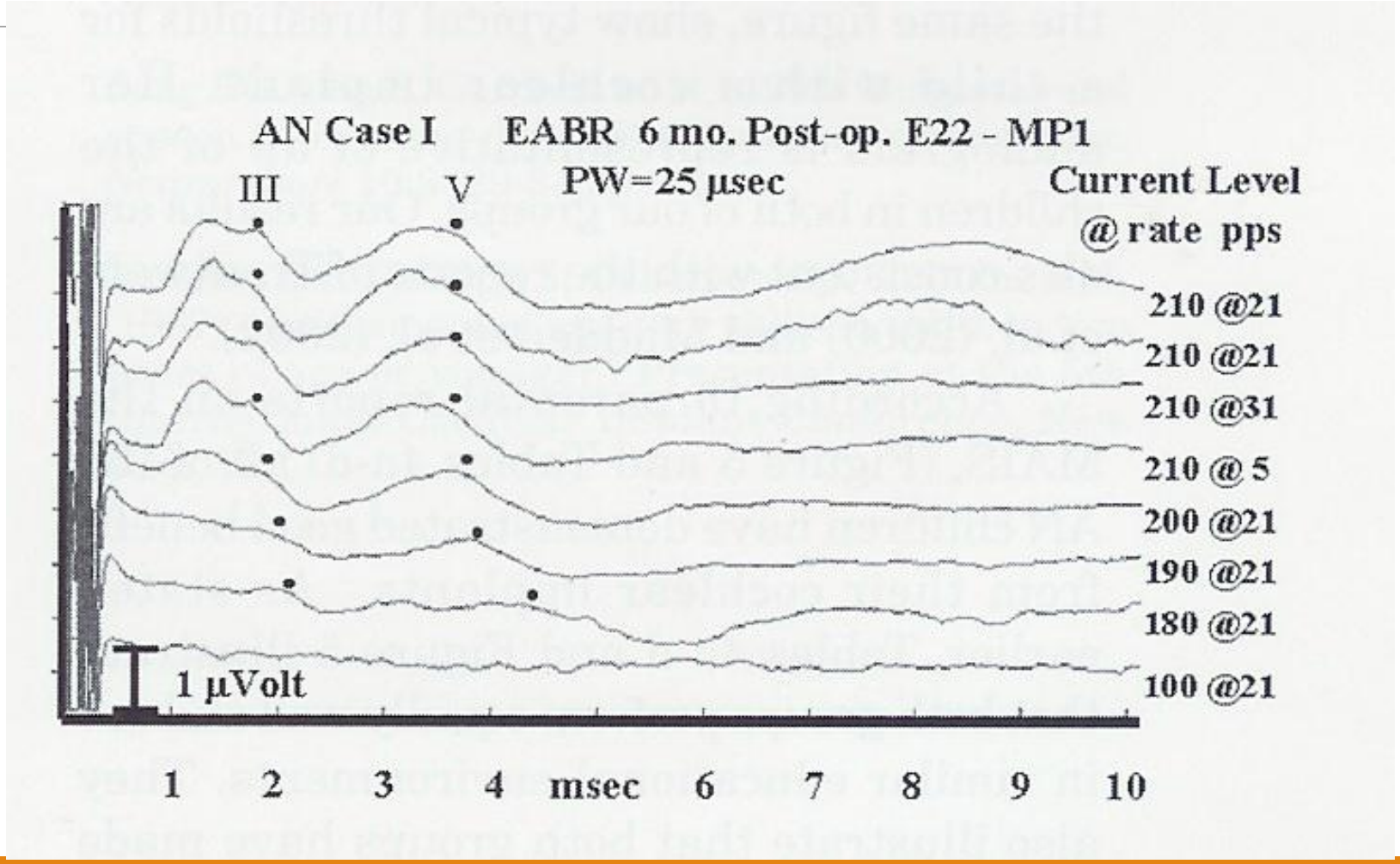
Although 50% of the implanted children with ANSD demonstrated open-set speech perception abilities after implantation, nearly 30% of them with 6 months of implant experience were unable to participate in this type of testing because of their young age or developmental delays. No child with cochlear nerve deficiency (CND) in their implanted ear achieved open-set speech perception abilities. In a subgroup of children, good open-set speech perception skills were associated with robust responses elicited on electrical-evoked intra-cochlear compound action potential testing when this assessment was possible.

Peterson, Shallop, et al.



Electrical stimulation provides timing precision and synchrony, demonstrated in an electrical ABR

Peterson, Shallop, et al.



Cortical Evoked Potentials Evaluate Auditory Processing Capacity

CAEP has been used with clinical populations (ANSD and others) in the following capacities

1. To predict auditory capacity of individual children
2. To measure potential speech perception using standard amplification (thus the need for implantation)
3. To determine the efficacy of implantation.
4. To determine neural development.

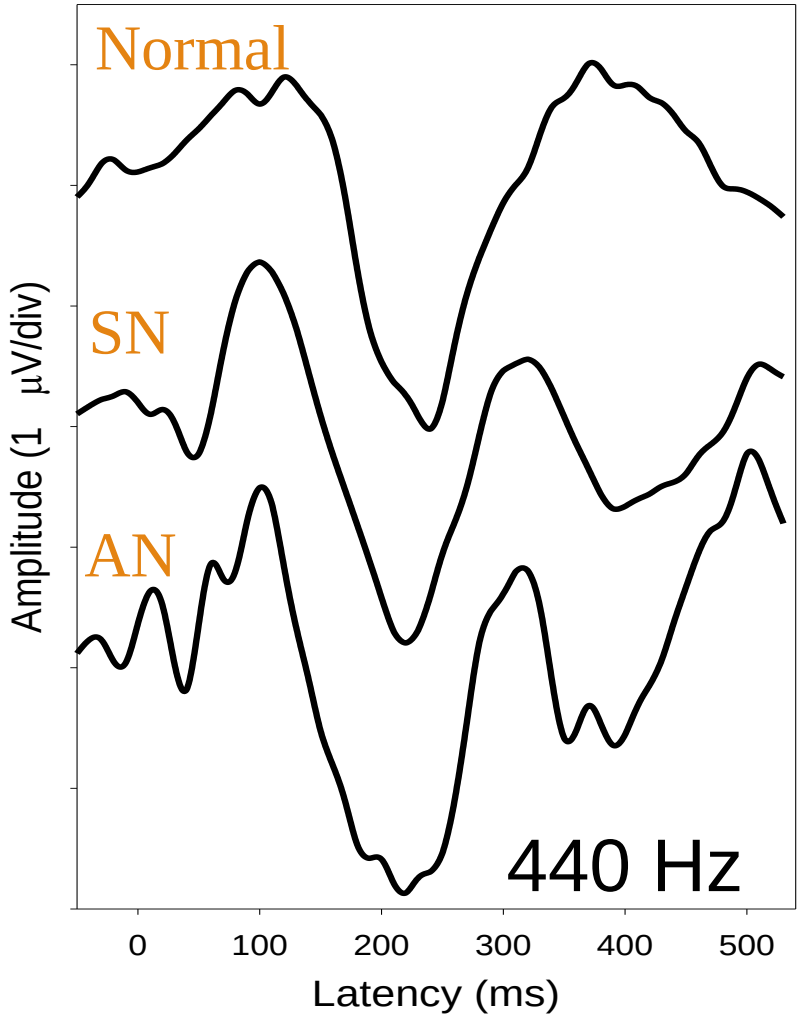
Cortical Evoked Potentials

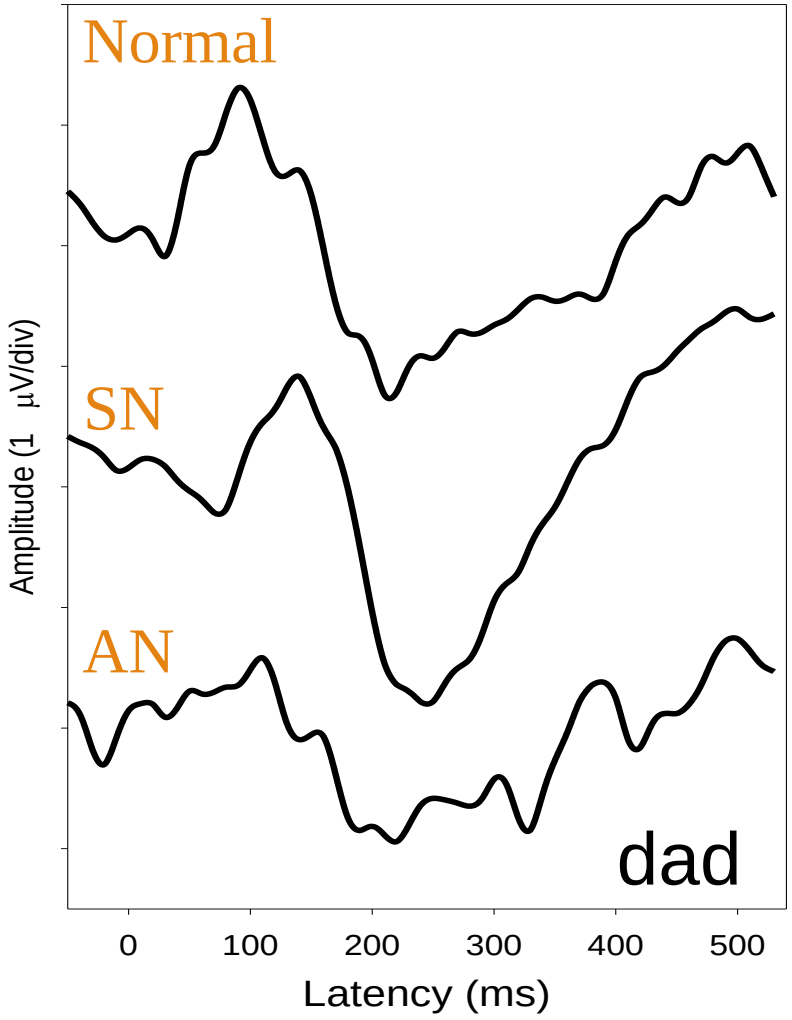
ABR is absent/abnormal in persons with AN but cortical evoked potentials (CAEP) may be present.

Rance, Cone-Wesson, Wunderlich and Dowell (2002) found that the presence of CAEP for tones or speech was positively associated with speech perception scores (and benefit from amplification).

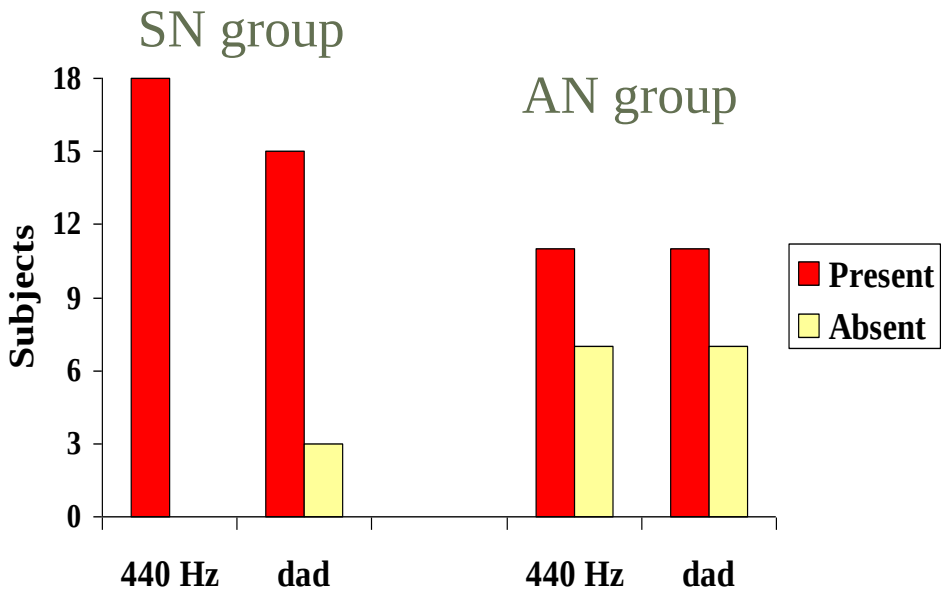
Cortical Auditory Evoked Potentials

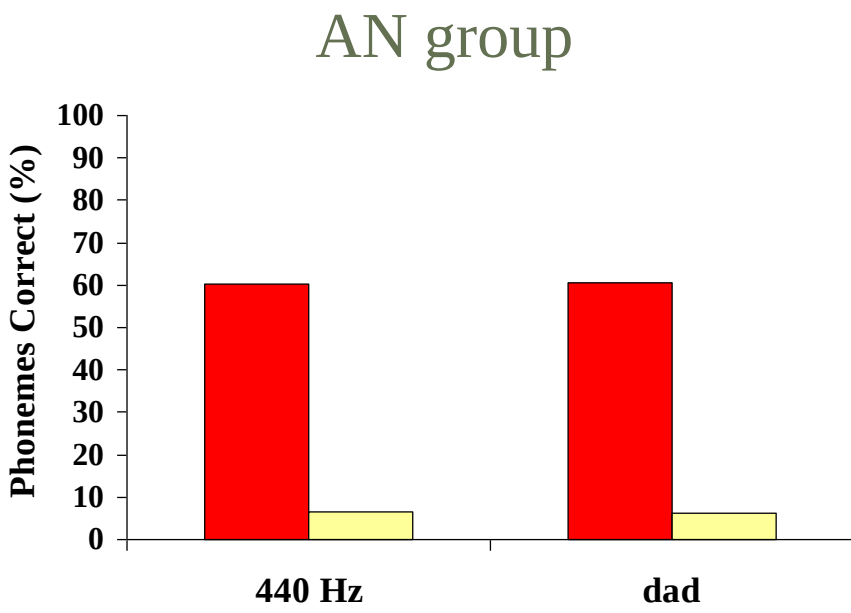
Rance, G., Cone-Wesson, B., Wunderlich, J., Dowell, R., 2002. Speech perception and cortical event related potentials in children with auditory neuropathy. Ear Hear 23 (3)

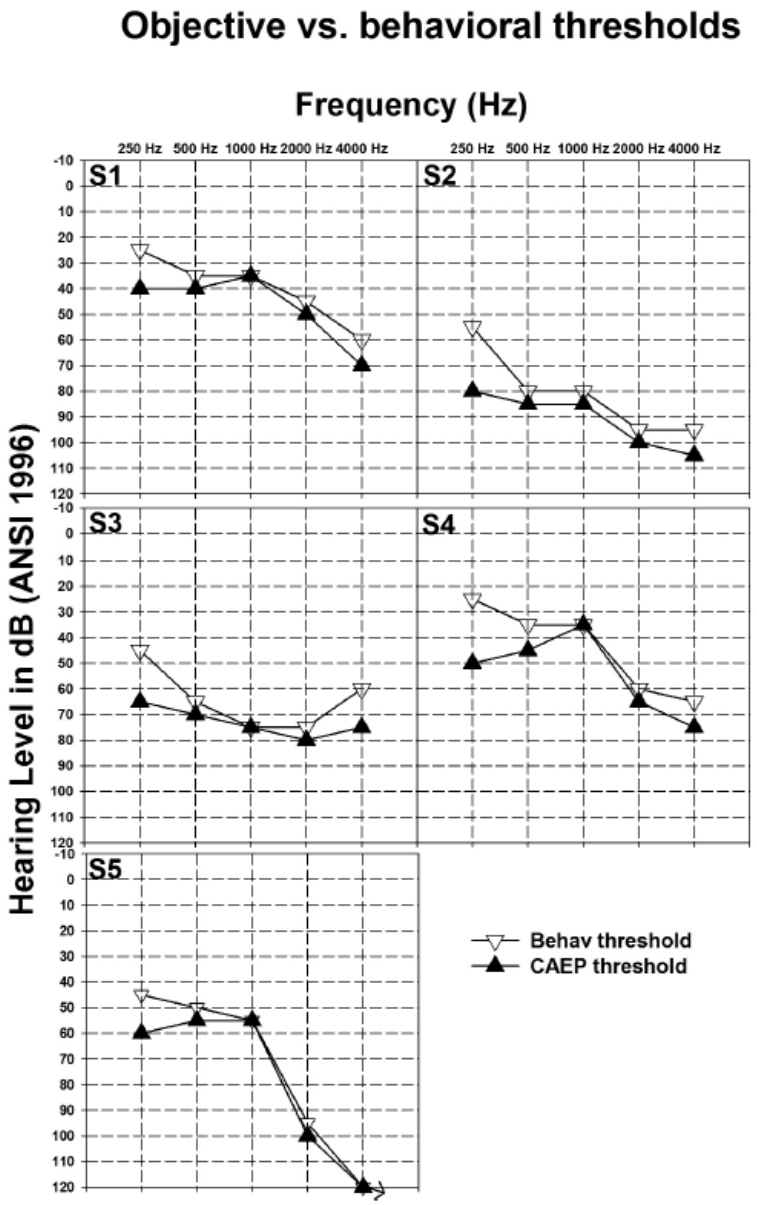




CAEP Presence vs. Absence







Objective Hearing Threshold Estimation in Children with Auditory Neuropathy Spectrum Disorder

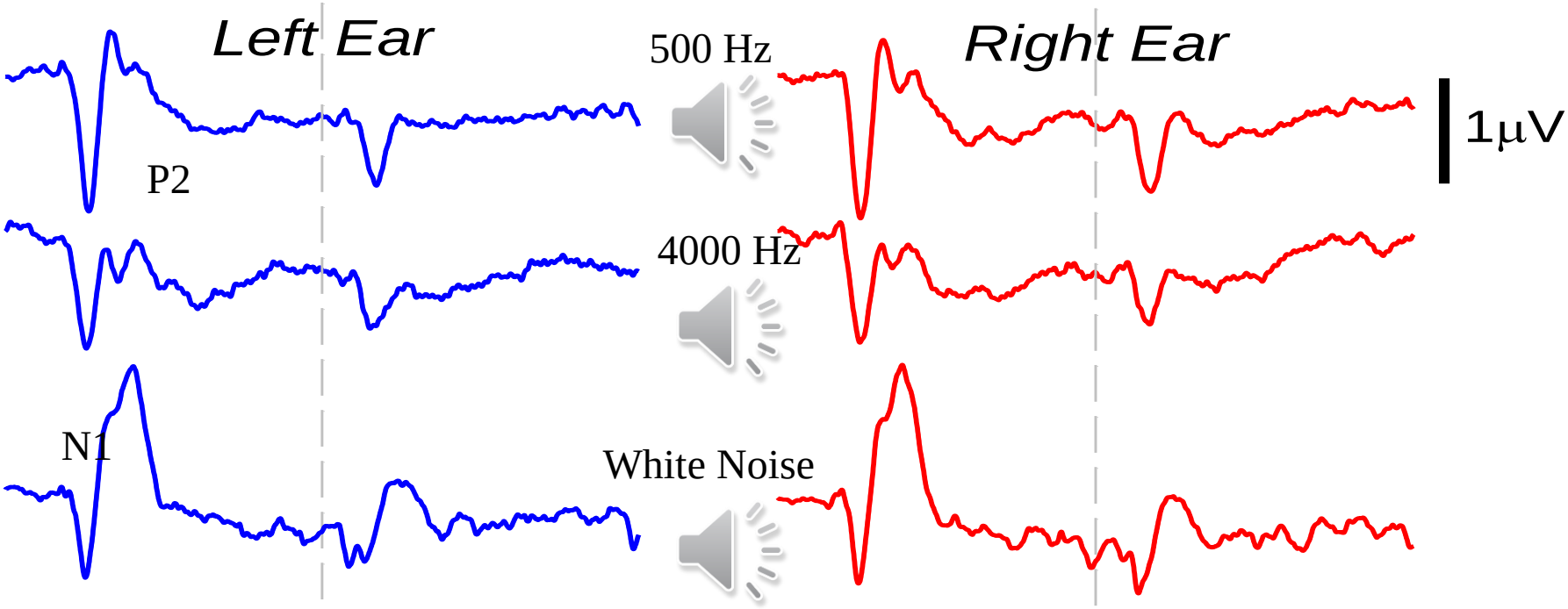
Shuman He, MD, PhD, Holly F.B. Teagle, AuD, Patricia Roush, AuD, John H. Grose, PhD, and Craig A. Buchman, MD

Department Otolaryngology – Head and Neck Surgery The University of North Carolina at Chapel Hill Chapel Hill, North Carolina

Laryngoscope. 2013 November ; 123(11): 2859–2861.

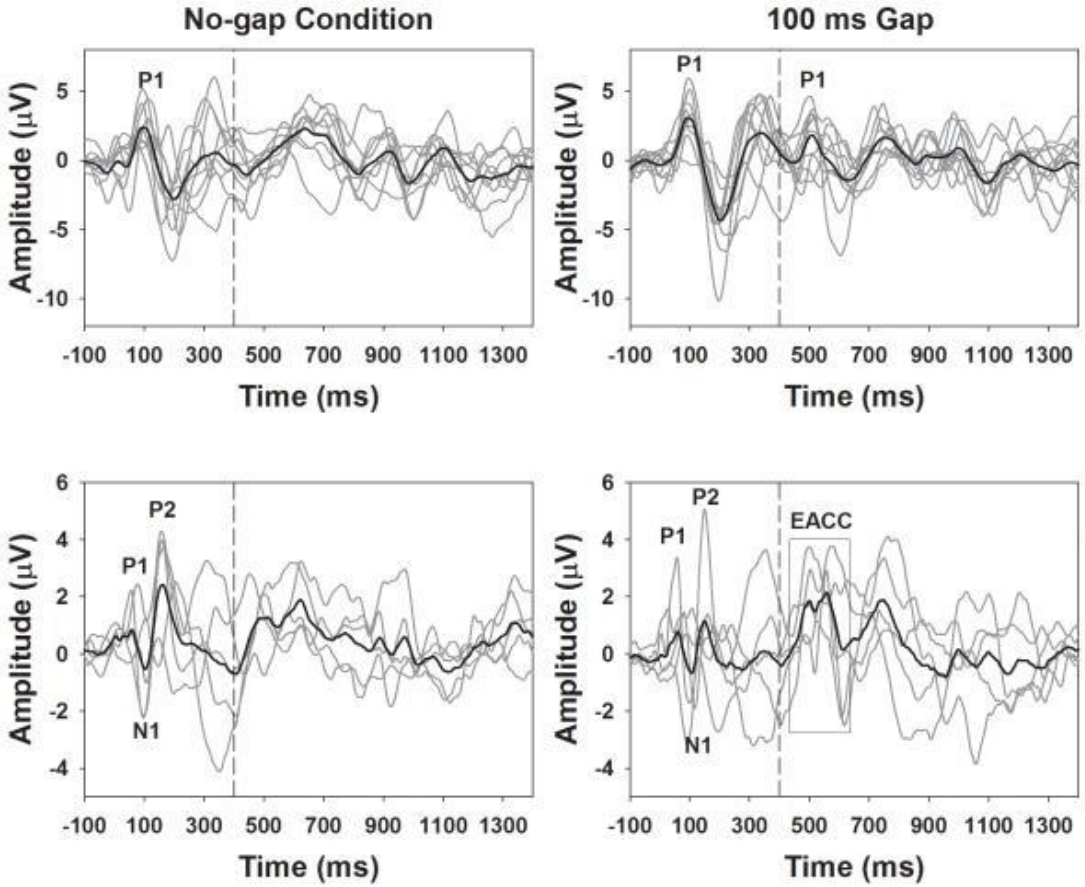
ACOUSTIC CHANGE RESPONSE

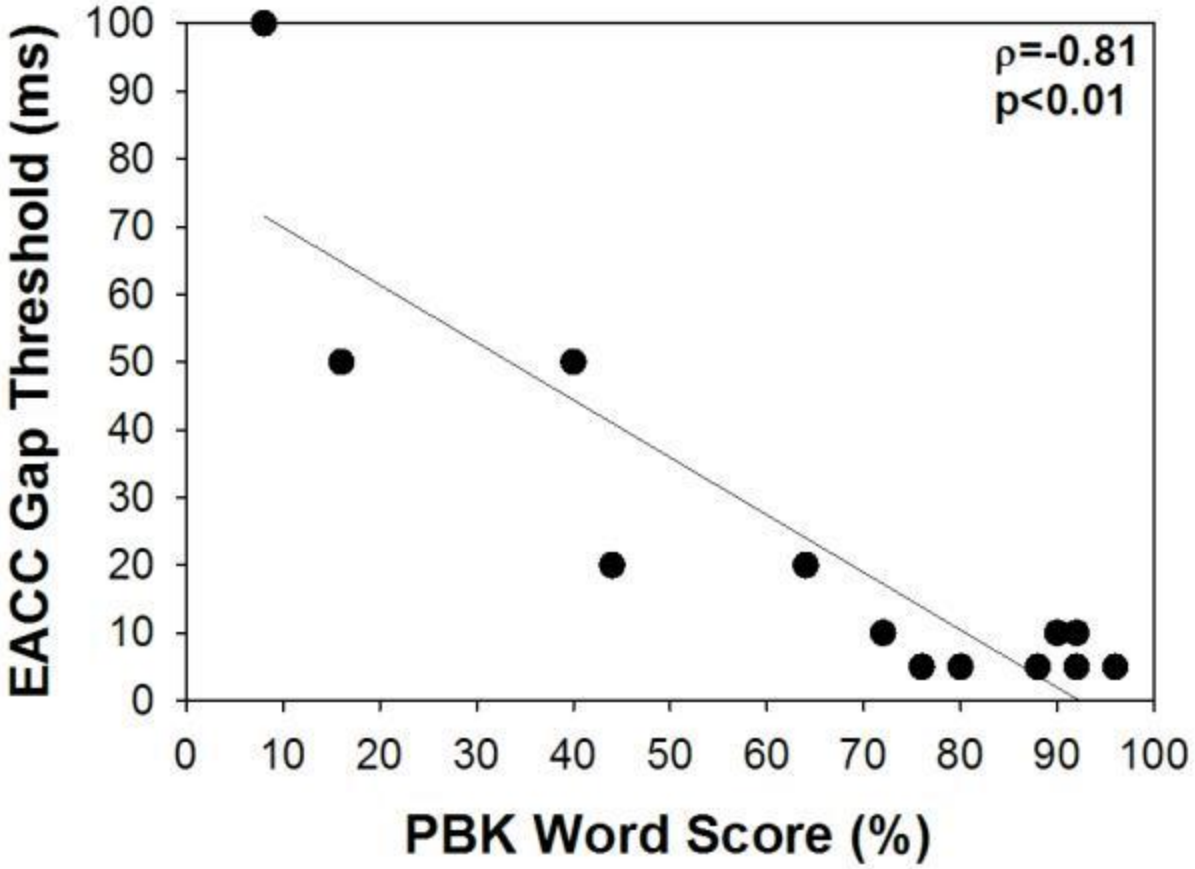
Grand Average Gap Responses at Cz



Gap Detection Measured with Electrically-Evoked Auditory Event-Related Potentials and Speech Perception Abilities in Children with Auditory Neuropathy Spectrum Disorder

Shuman He, PhD, John H. Grose, PhD, Holly F.B. Teagle, AuD, Jennifer Woodard, AuD, Lisa R. Park, AuD, Debora R. Hatch, AuD, and Craig A. Buchman, MD Ear Hear 2013 34:6.

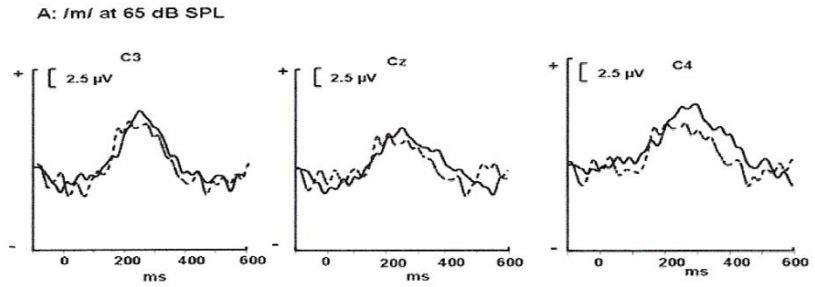



EACC Gap Threshold (ms)
PBK Word Score (%)
ρ=-0.81
p<0.01
0
10
20
30
40
50
60
70
80
90
100

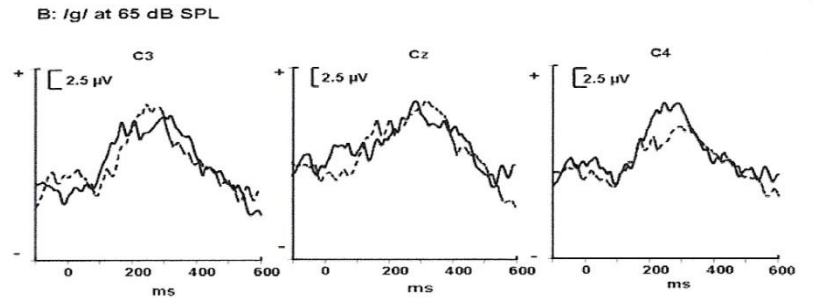
Aided Responses from One Patient with AN

Speech stimuli @ 65 dB

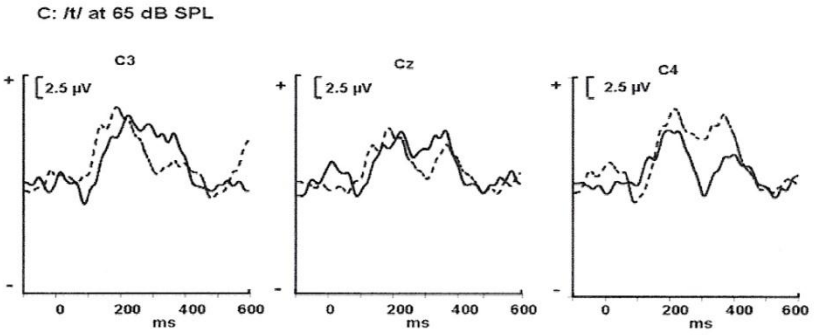
Response to /m/



Response to /g/



Response to /t/



Pearce, Golding & Dillon "Cortical Auditory Evoked Potentials in the Assessment of Auditory Neuropathy JAAA 18:380 (2007)

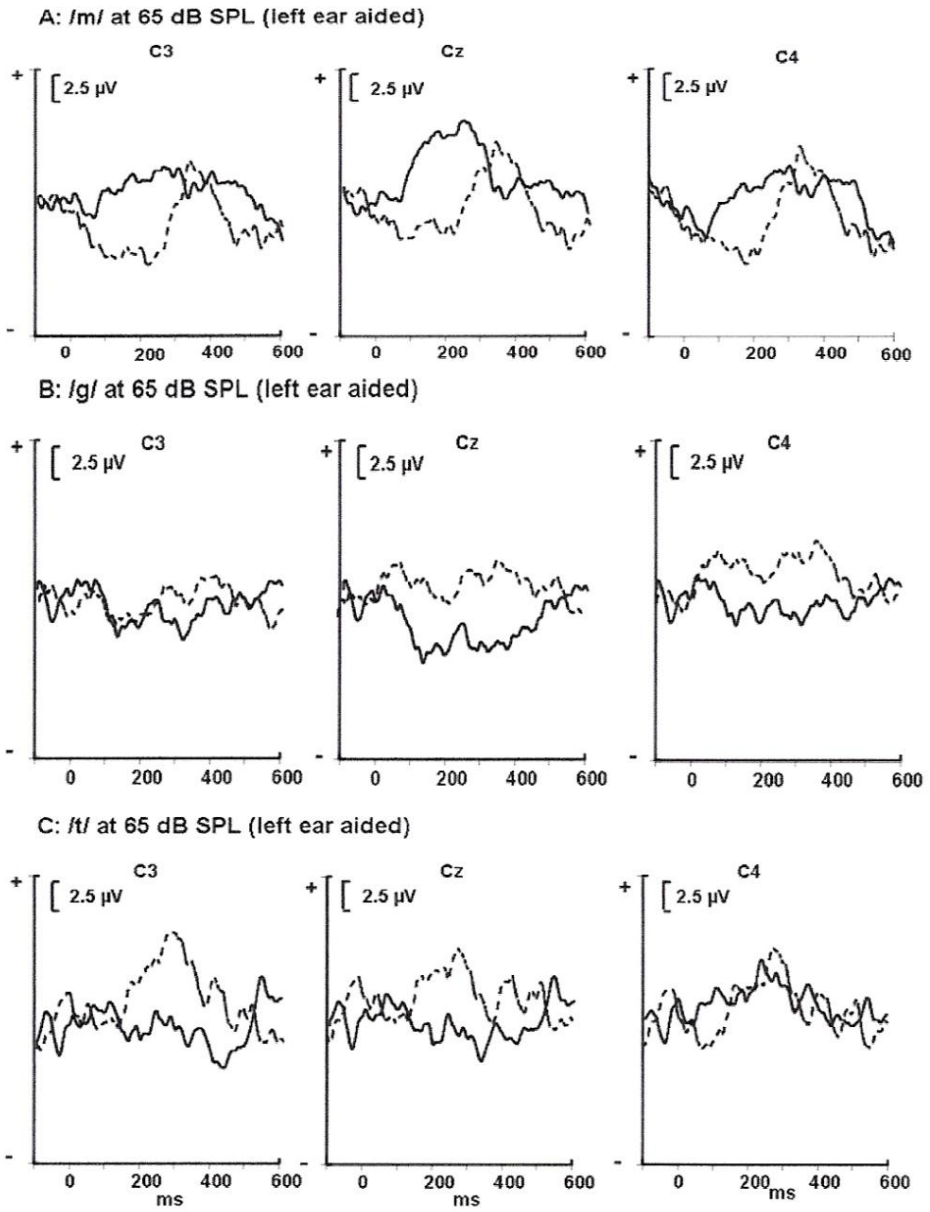
Poor Aided Responses from One Patient with AN

Response to /m/

Response to /g/

Response to /t/

Speech stimuli @ 65 dB



Thank you for listening

Questions??

Auditory neuropathy: clinical characteristics and therapeutic approach

Eyal Raveh MD , Nora Buller MD, Ola Badrana MA and Joseph Attias DSc

American Journal of Otolaryngology
Volume 28, Issue 5, September-October 2007, Pages 302-308

AN in 26 children over 5 years

HX: Prematurity (8) Hypebillirubinemia (7)
Ototoxic (6) Asphyxia (2) Family Hx (3)
Parental consanguinity (4) Meningitis (1)
IVF (6)

Seven with associated medical pathologies

Eight had no risk factors or associated path (31%)

Predicting audiogram

Hearing Aid Fitting

Cochlear Implantation

Cortical EPS

Central Auditory Maturation and Behavioral Outcome in Children with Auditory Neuropathy Spectrum Disorder who Use Cochlear Implants

Garrett Cardon and Anu Sharma

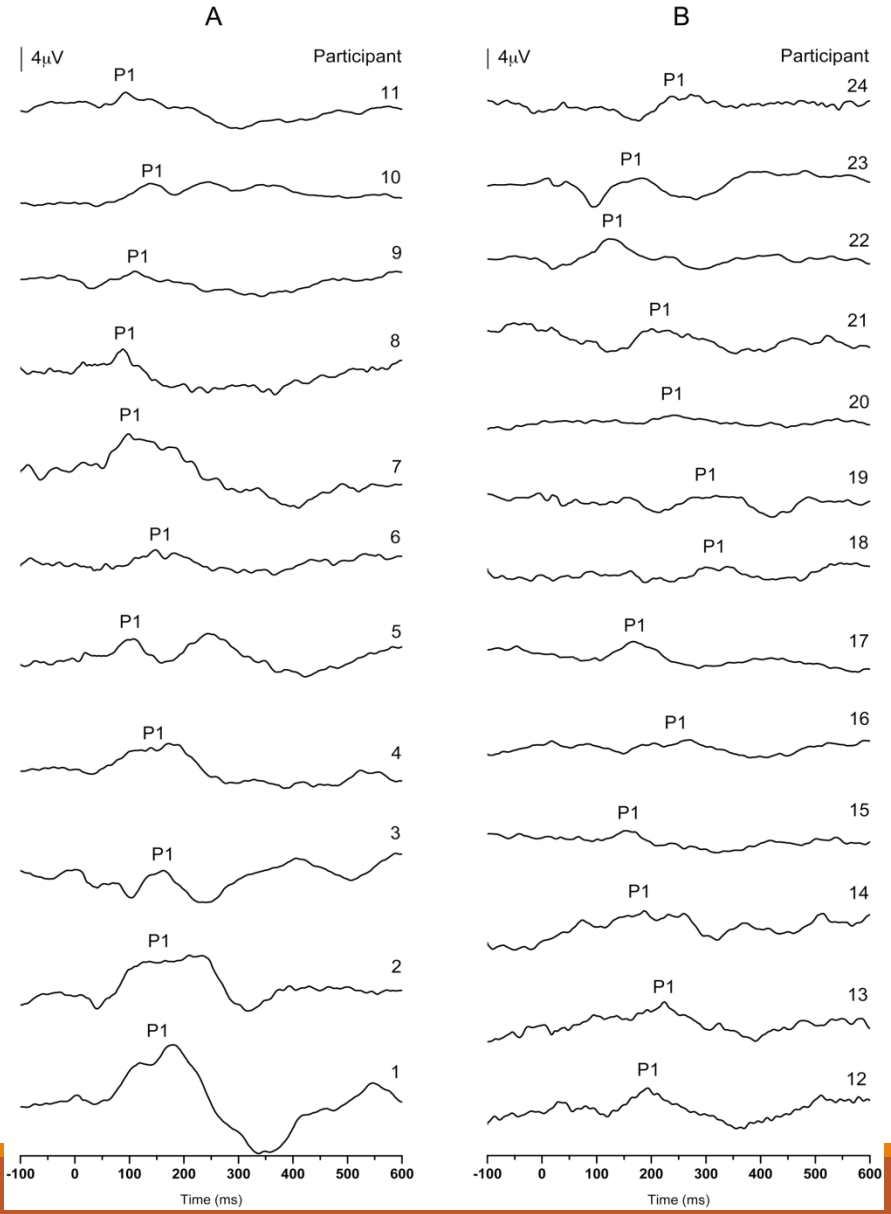
Int J Audiol. 2013 September ; 52(9): 577–586

Department of Speech, Language, and Hearing Sciences, University of Colorado, Boulder, USA

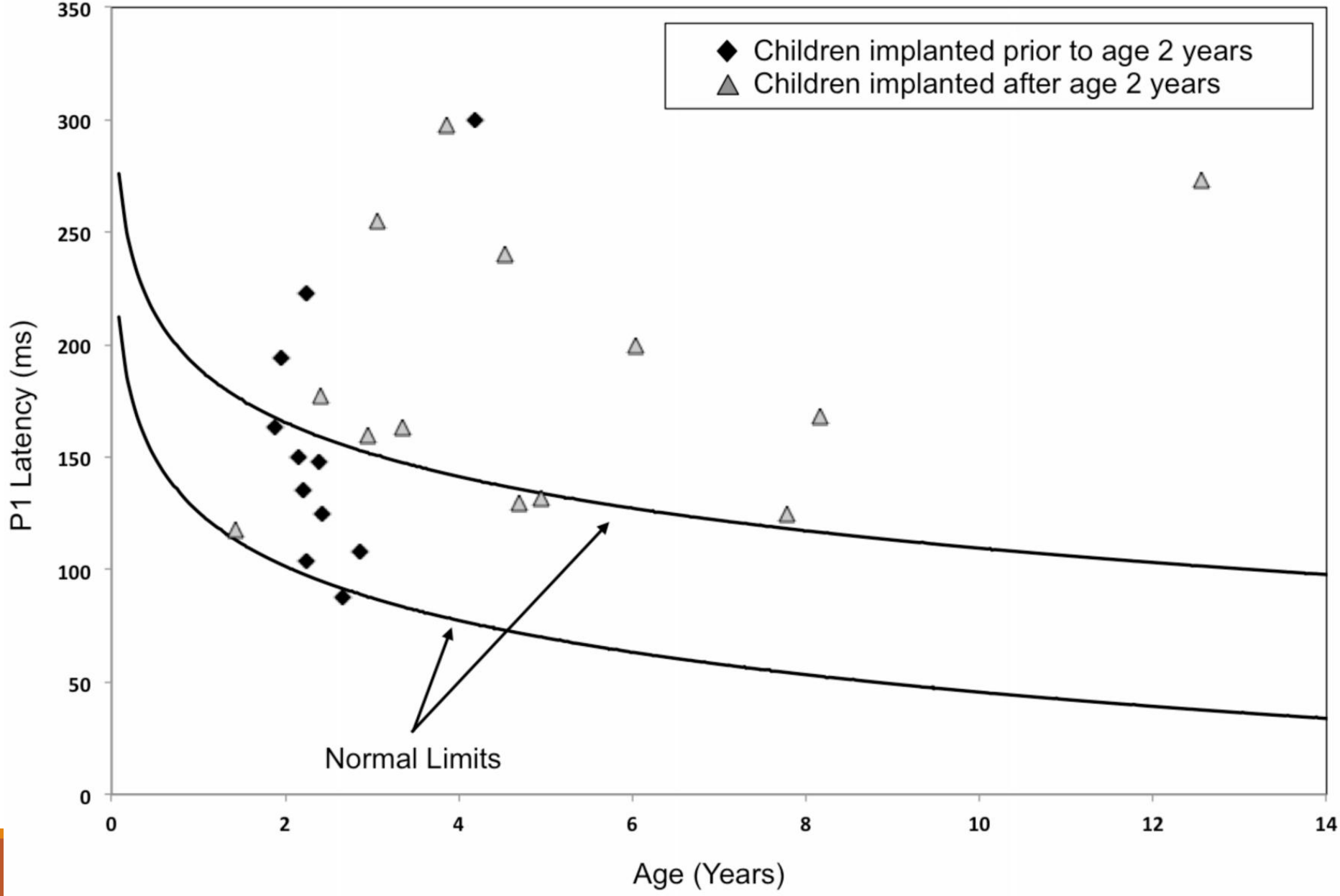
24 Children with ANSD using Cochlear Implants
Standard Cortical EPs measured with /ba/ stimulus presented in sound field at 75 dB.

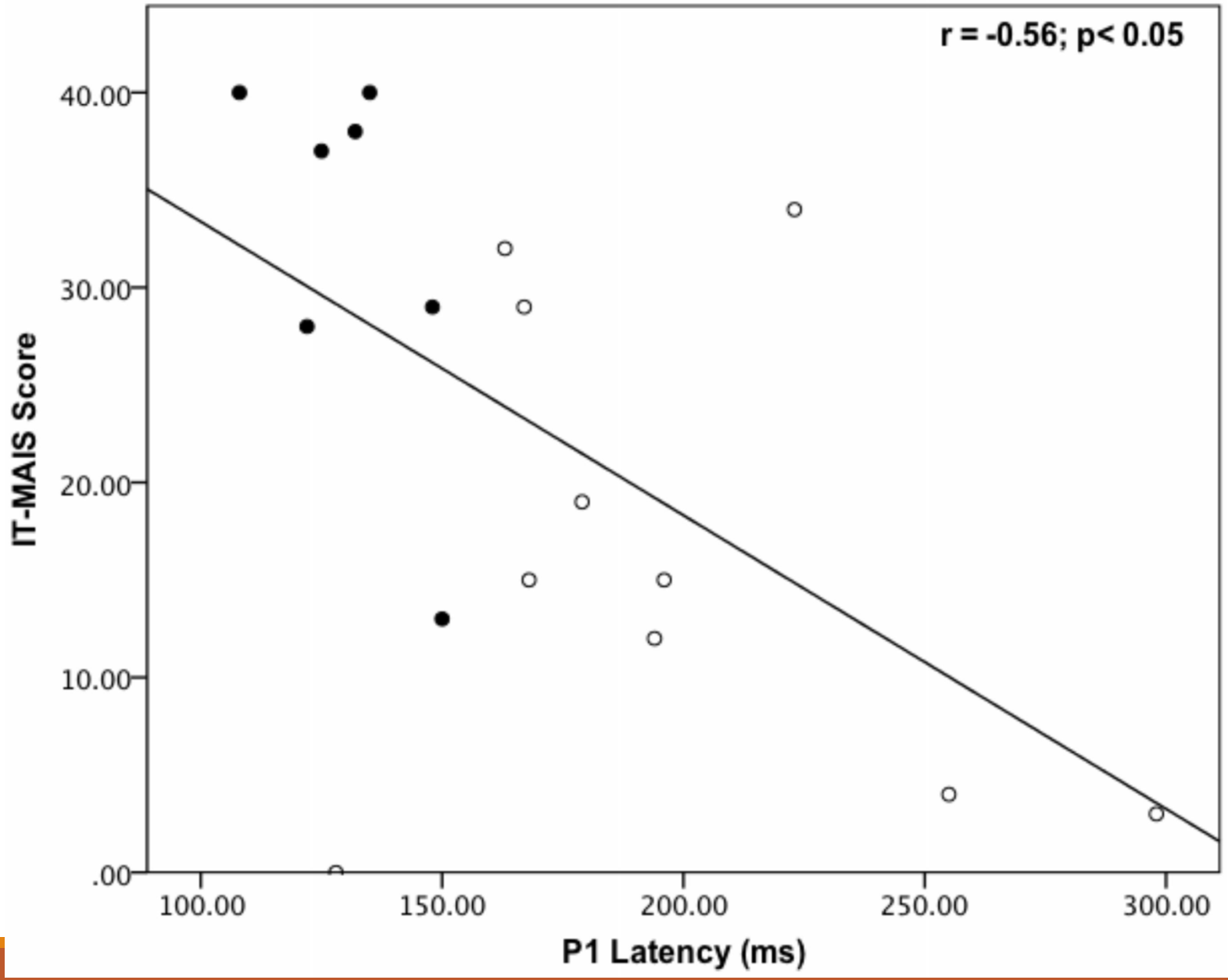
Normal P1 Latencies

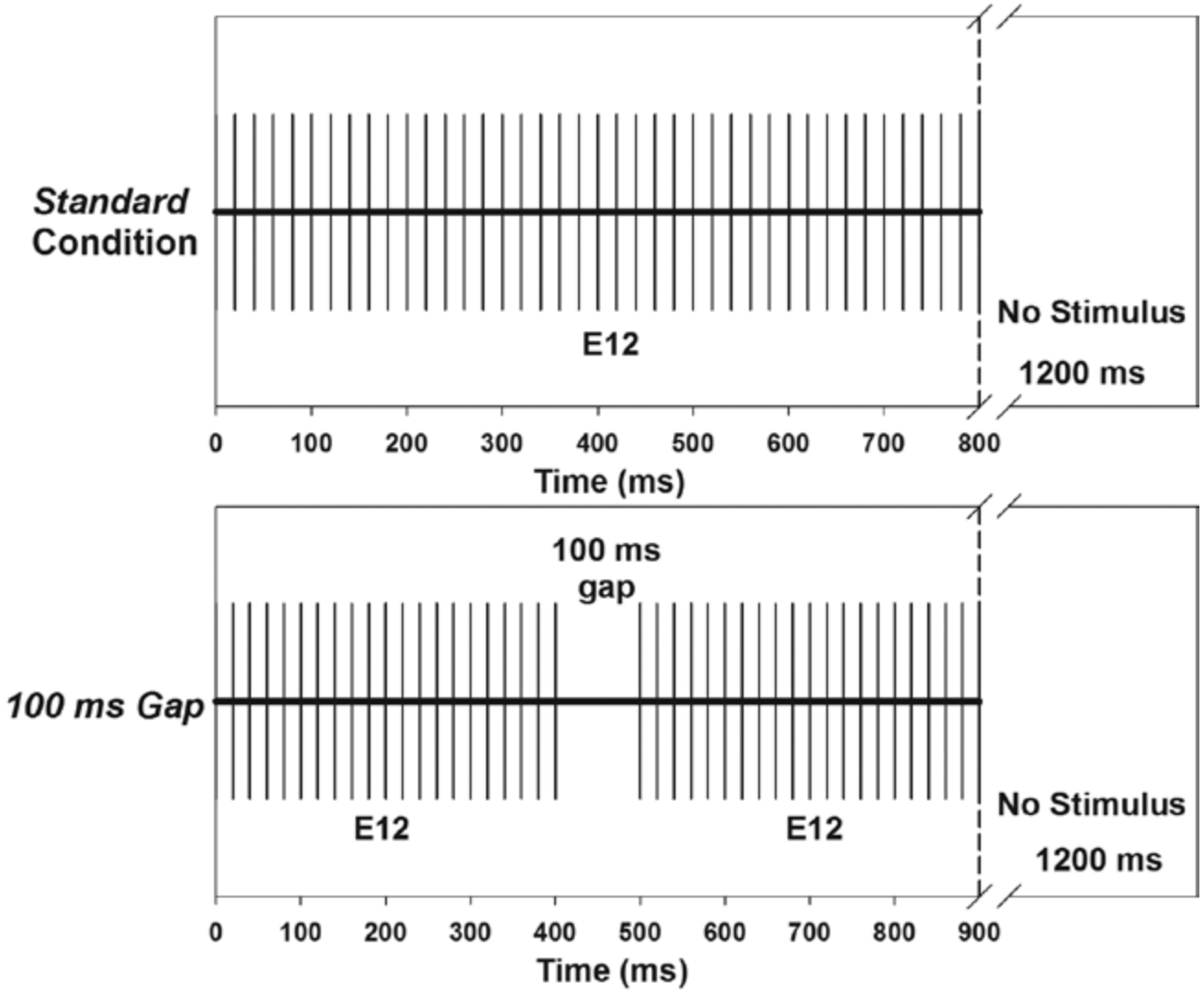
Delayed P1 Latencies

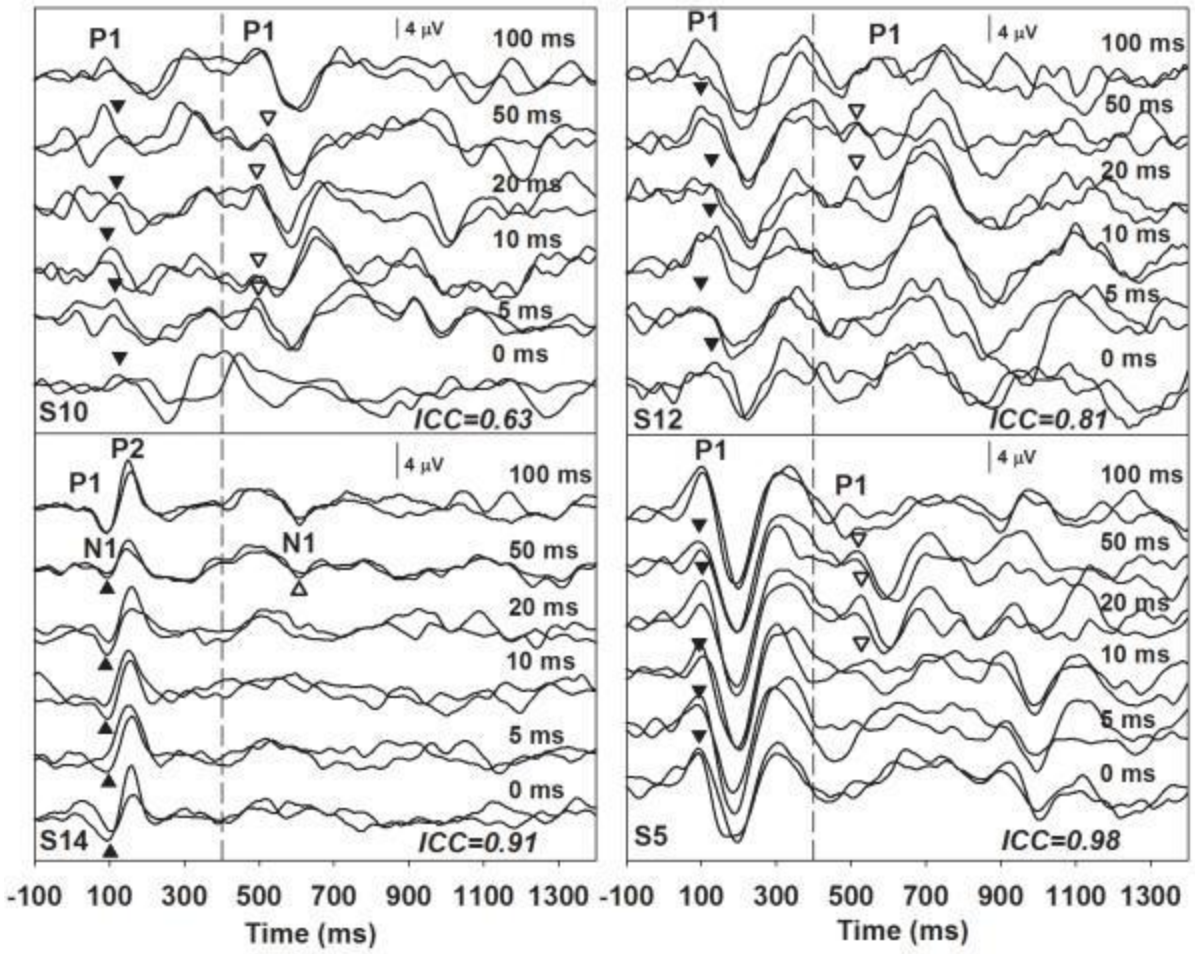


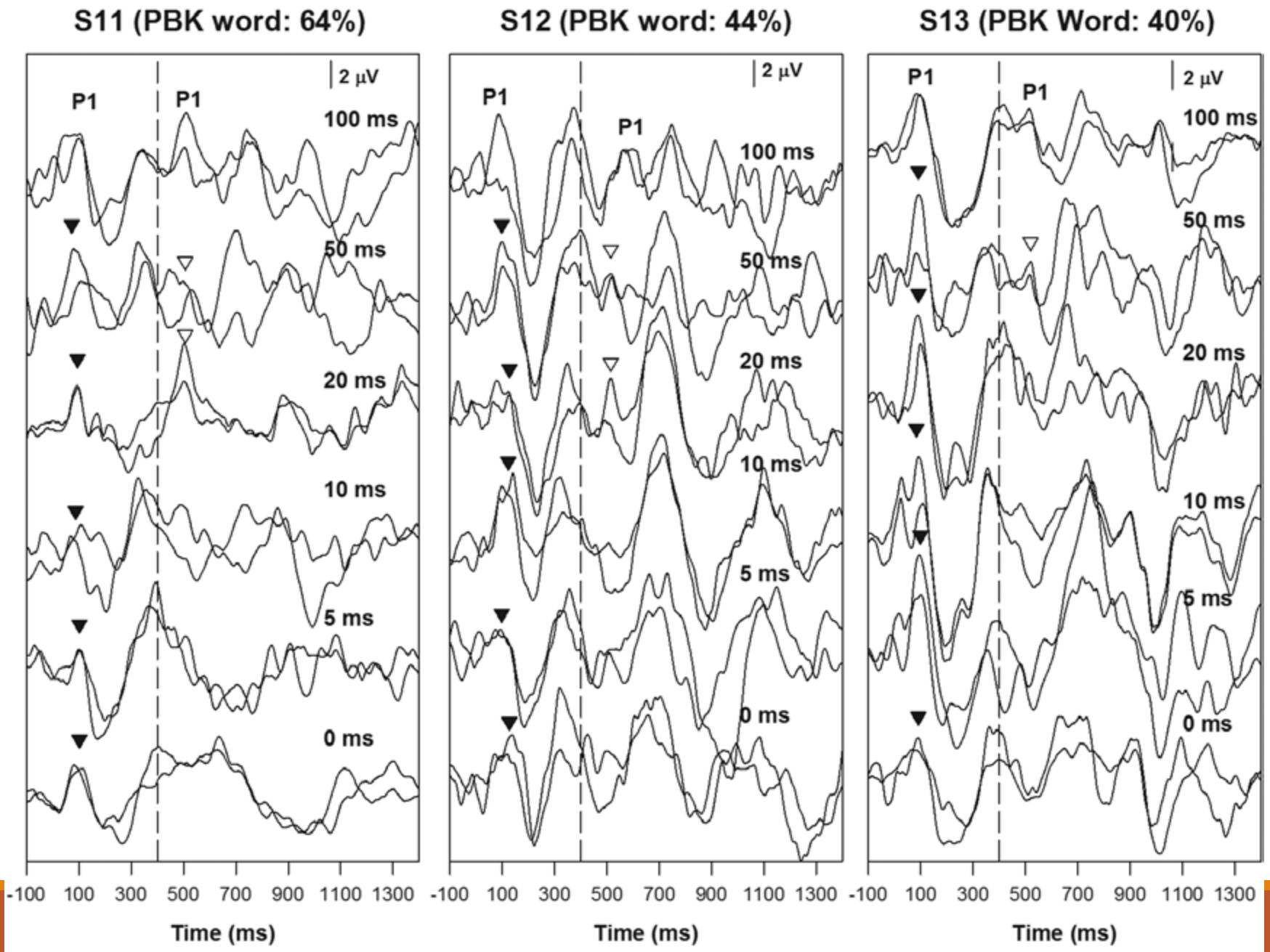
Central Auditory Maturation and Behavioral Outcome in Children with Auditory Neuropathy Spectrum Disorder who Use Cochlear Implants **Condon & Sharma**



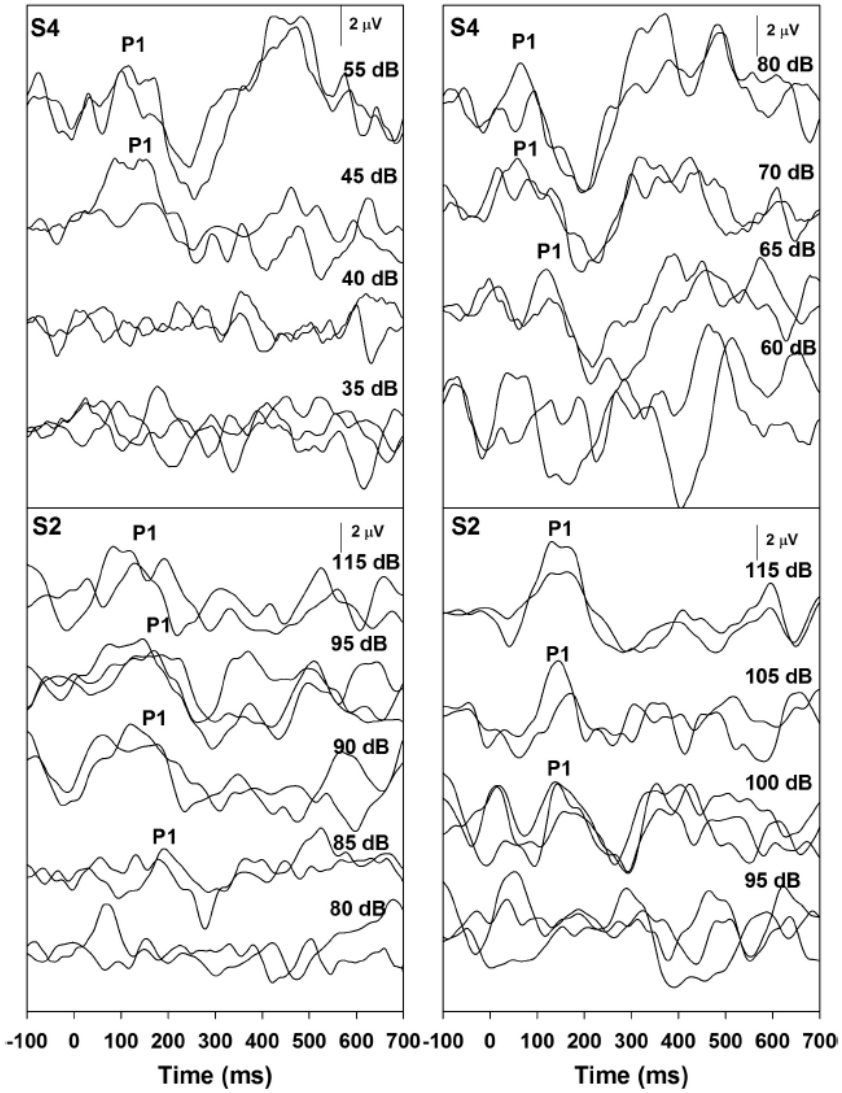

r = -0.56; p< 0.05
IT-MAIS Score
40.00
30.00
20.00
10.00
.00
100.00
150.00
200.00
250.00
300.00
P1 Latency (ms)


Standard
Condition
E12
No Stimulus
1200 ms
0
100
200
300
400
500
600
700
800
Time (ms)
100 ms
gap
100 ms Gap
E12
E12
No Stimulus
1200 ms
0
100
200
300
400
500
600
700
800
900
Time (ms)


P1
P1
4 μV
100 ms
50 ms
20 ms
10 ms
5 ms
0 ms
S10
ICC=0.63
P1
P1
4 μV
100 ms
50 ms
20 ms
10 ms
5 ms
0 ms
S12
ICC=0.81
P2
P1
N1
N1
4 μV
100 ms
50 ms
20 ms
10 ms
5 ms
0 ms
S14
ICC=0.91
P1
P1
4 μV
100 ms
50 ms
20 ms
10 ms
5 ms
0 ms
S5
ICC=0.98
-100 100 300 500 700 900 1100 1300
Time (ms)
-100 100 300 500 700 900 1100 1300
Time (ms)


S11 (PBK word: 64%)
S12 (PBK word: 44%)
S13 (PBK Word: 40%)
2 μV
P1
P1
100 ms
50 ms
20 ms
10 ms
5 ms
0 ms
-100 100 300 500 700 900 1100 1300
Time (ms)

He et al. *Laryngoscope*. 2013 November ; 123(11): 2859–2861.



Raveh et al., 2007

Degree of Loss: Mild (3) Moderate (6) Severe to Profound (17)

One of 26 was unilateral (4%)

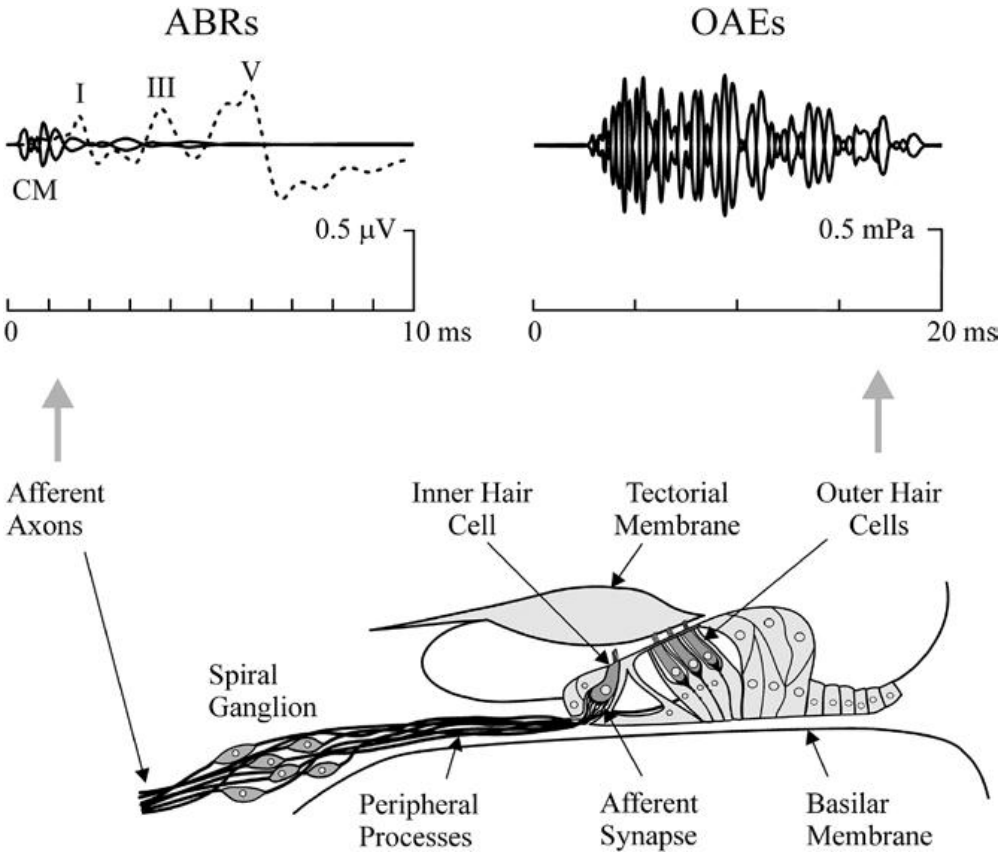
4 of 26 showed spontaneous improvement (?) by 8 months of age (16%)

18 of 26 (69%) had OAEs- All showed CM

Of 19 eligible 14 are implanted or pending

Hearing in Time: Evoked Potential Studies of Temporal Processing

Terence Picton *Ear & Hearing* Vol 34, #6, 2013



Potential Sites of Lesion in AN Based on Symptoms
(Normal OAEs or CM and No ABR Wave I)

- **Isolated inner hair cell disorder (Harrison,1998 *Ear and Hearing, 19,* 355-361). *Not likely*.**
- **Inner hair cell synaptic junction. *Proven***
- **Peripheral portion of auditory nerve (demyelinating or axonal neuropathy of the auditory nerve). *Proven***

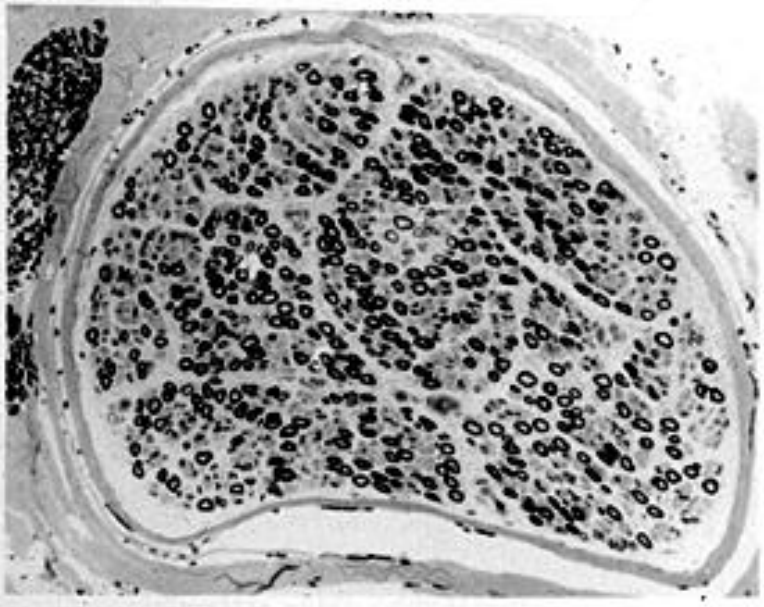
Inner Hair Cell is Usually Not the Site of Lesion in Most Cases of AN

- **No way to adequately distinguish IHC disease from primary auditory nerve disease in humans.**
- **Animal models of IHC disorder are not proven to represent human physiology and do not show disorders of timing (Phillips et al., 2001)**
- **Only one study has shown human temporal bones with isolated IHC lesions in premature infants with unclear auditory responses. (Amatuzzi et al., 2001)**

Evidence of Auditory Nerve Involvement in Auditory Neuropathy

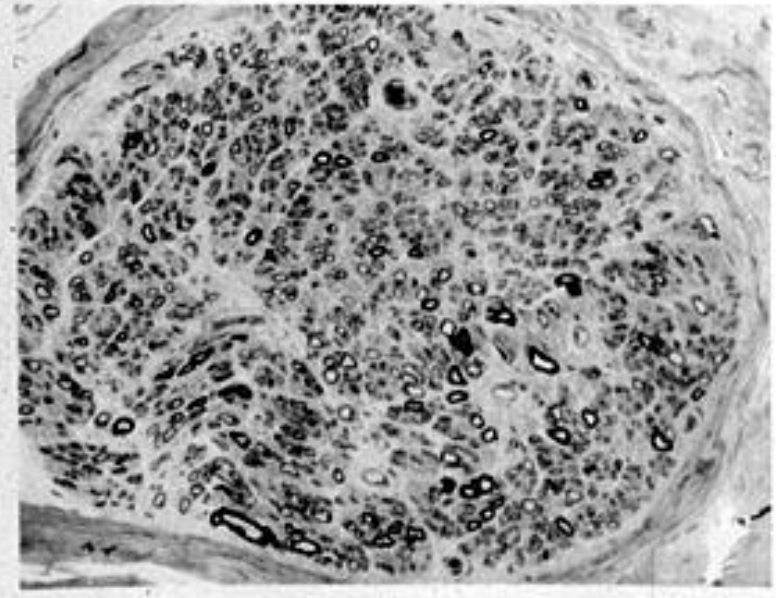
- **Starr et al., found 30-40% of patients have evidence of other peripheral nerves disorders (absent deep tendon reflexes or poor nerve conduction)**
- **As many as 80% of adult AN patients have subtle or pronounced peripheral neuropathy (HSMN, Friedreich's Ataxia).**
- **Sural nerve biopsy on 4 patients with AN shows peripheral nerve disease, primarily axonal.**

Sural Nerve From Patient With Auditory Neuropathy



Normal

280×



Patient #11

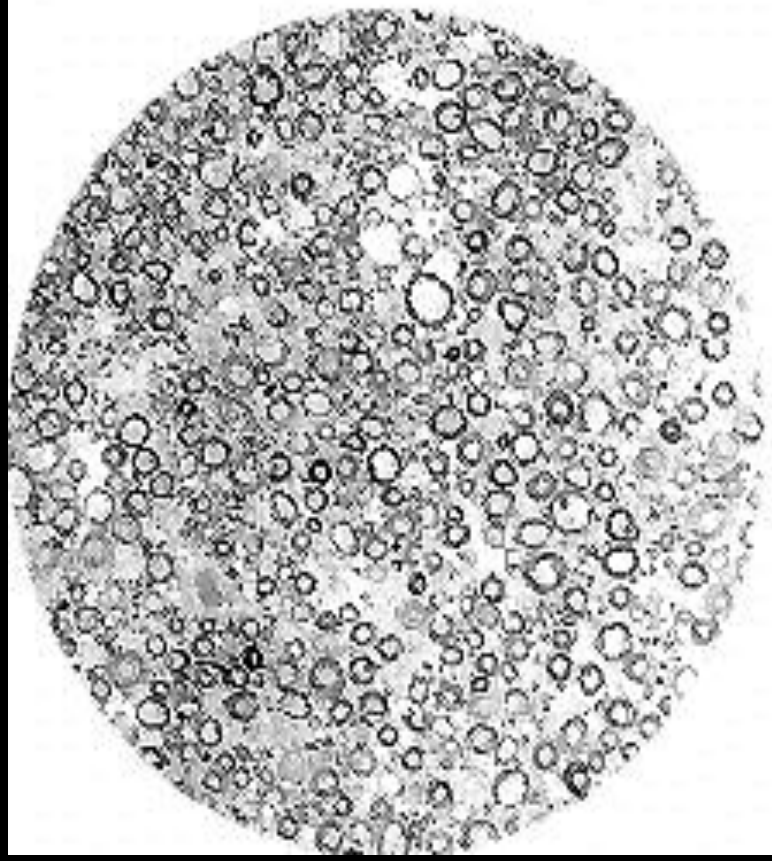
280×

Evidence of Auditory Nerve Involvement in Auditory Neuropathy

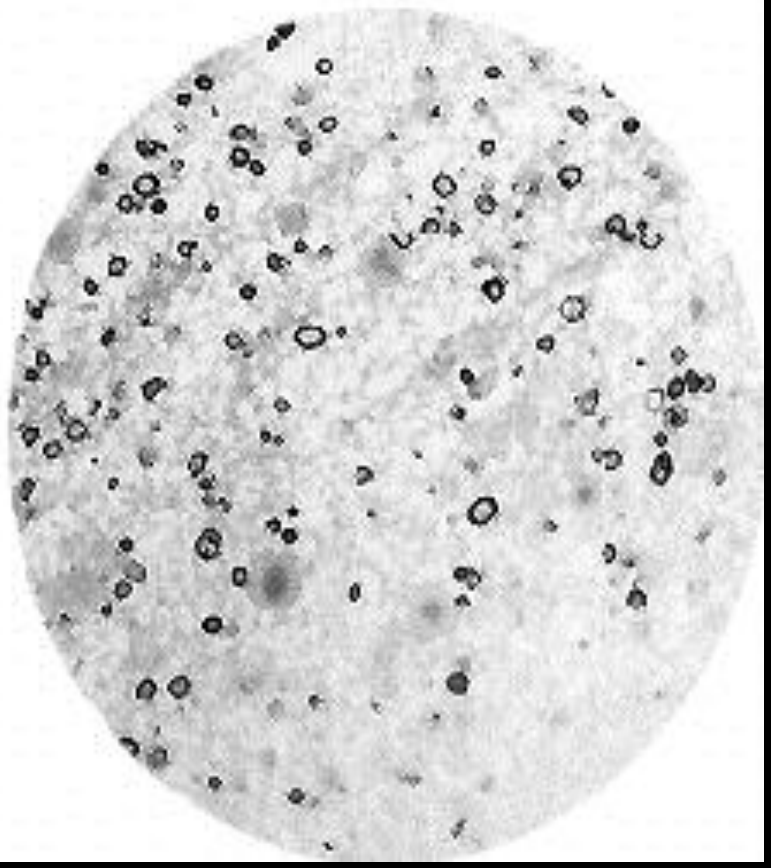
- **Human temporal bone histology showing normal complement of hair cells with poor ganglion cell survival (Spoendlin, Nadol).**
- **Temporal bone histology on patient with documented AN shows axonal degeneration (95% loss of ganglion cells), with a normal complement of hair cells (30% loss of OHC in apical turn). Starr et al., Brain 2003**

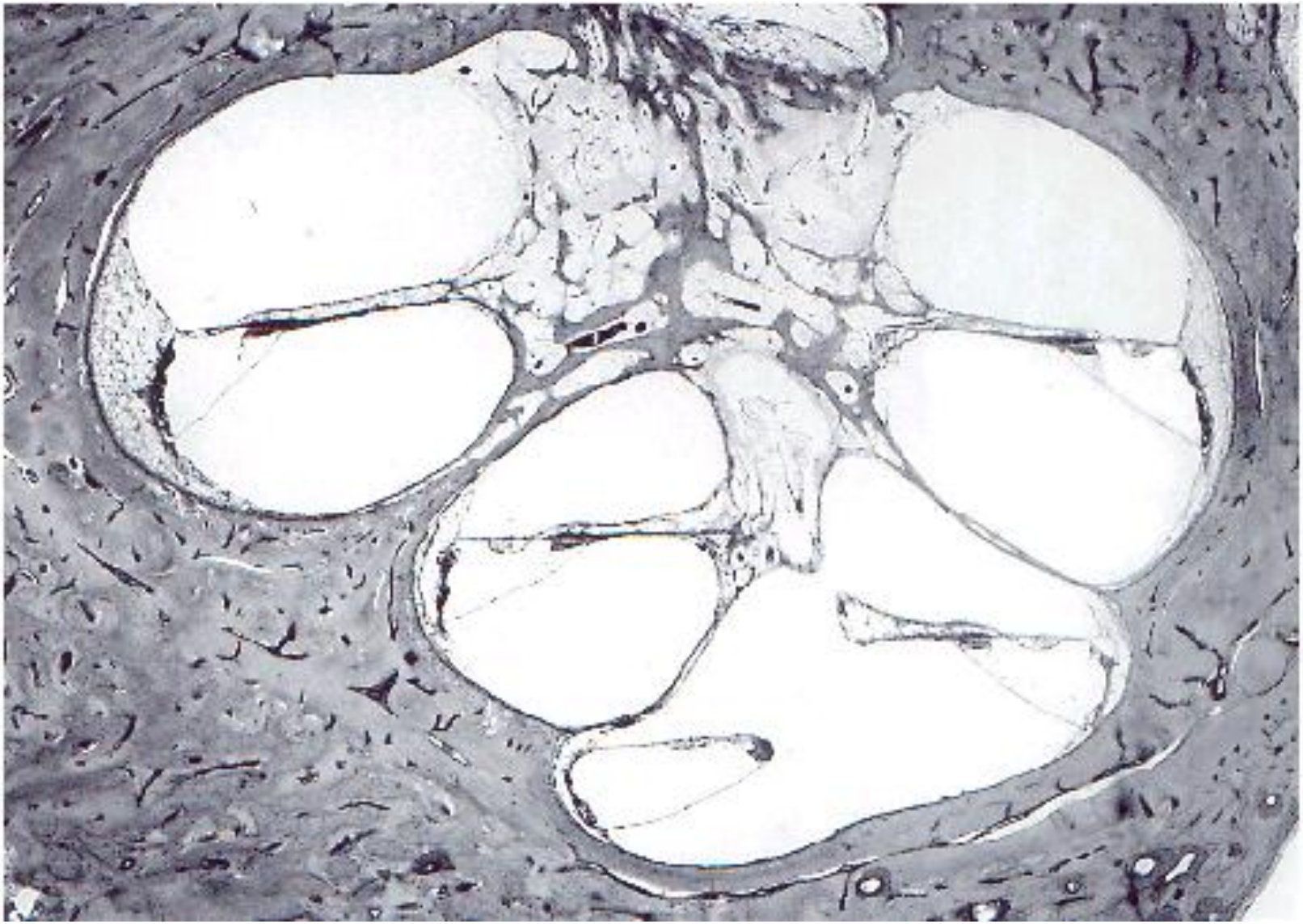
Auditory Nerve

Normal



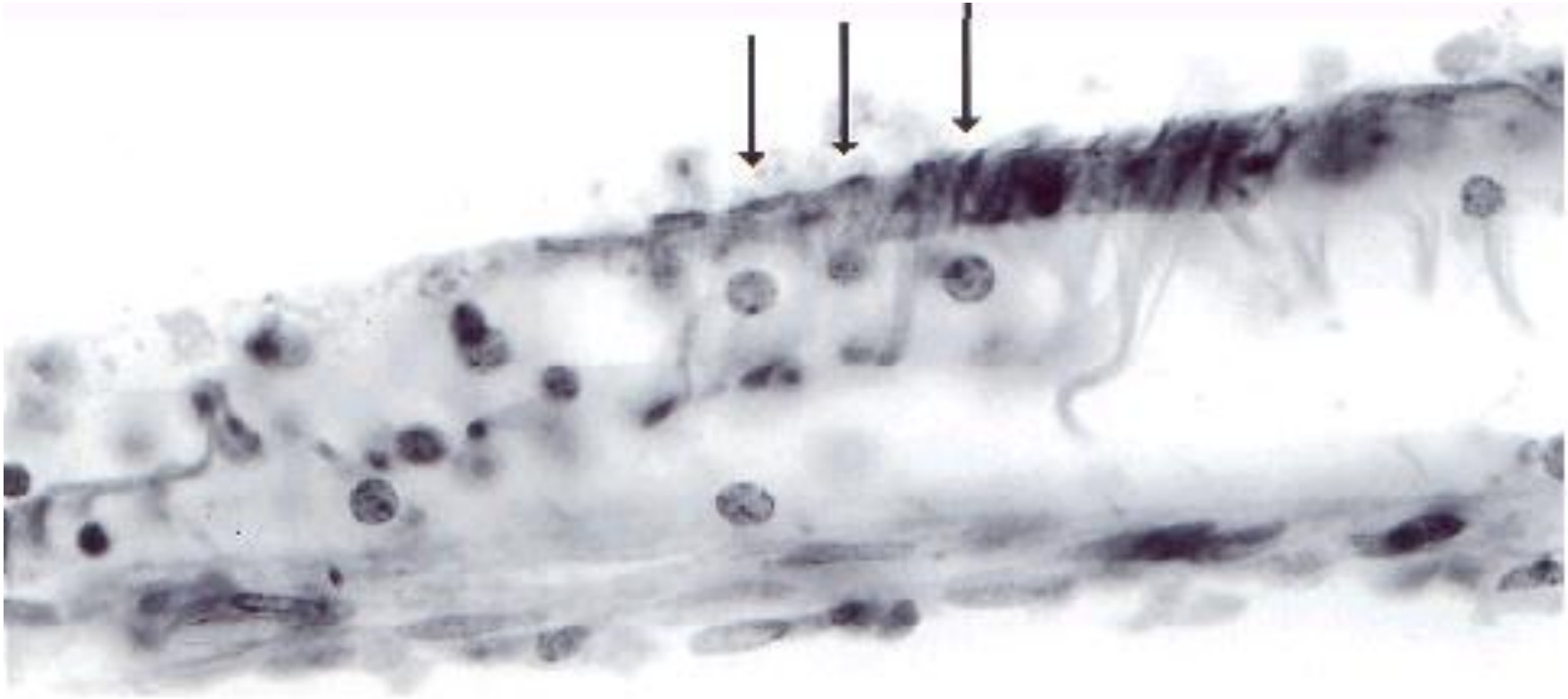
Auditory Neuropathy





Starr et al., Brain 2003

Inner Hair Cells from Temporal Bone of Auditory Neuropathy Patient



Starr et al., Brain 2003

Bilirubin and the auditory system.
Shapiro, SM and Nakamura H
J Perinatol. 2001 Dec;21 Suppl 1:S52-5; discussion S59-62.

The auditory system is highly sensitive to bilirubin toxicity. Damage to the auditory nervous system includes auditory neuropathy or auditory dyssynchrony and auditory processing problems which may occur with or without deafness, hearing loss. Auditory dysfunction may occur in children with or without other signs of classical kernicterus. Bilirubin selectively damages the brainstem auditory nuclei, and may also damage the auditory nerve and spiral ganglion containing cell bodies of primary auditory neurons. The inner ear, thalamic and cortical auditory pathways appear to be spared. Noninvasive auditory neurophysiological tests such as the auditory brainstem response (ABR) or brainstem auditory response (BAER) play an important role in the early detection of bilirubin-induced auditory and central nervous system dysfunction in the neonate.

The Jaundiced Gunn Rat Model of Auditory Neuropathy/Dyssynchrony

Shania, Shapiro & Spencer

***Laryngoscope,* 115:2167–2173, 2005**

***Conclusions:* Our findings of abnormal spiral ganglion cells and selective loss of large, myelinated auditory nerve fibers with *no abnormalities in cochlear hair cells*, support the sulfa-treated jj Gunn rat as a model for bilirubin induced AN. The paucity of large caliber neurons undermines temporal coding of auditory information and neural synchrony and demonstrates that in addition to brainstem auditory nuclei, spiral ganglion neurons are selectively vulnerable to bilirubin toxicity.**

Data From Colorado (Marion Downs Center)

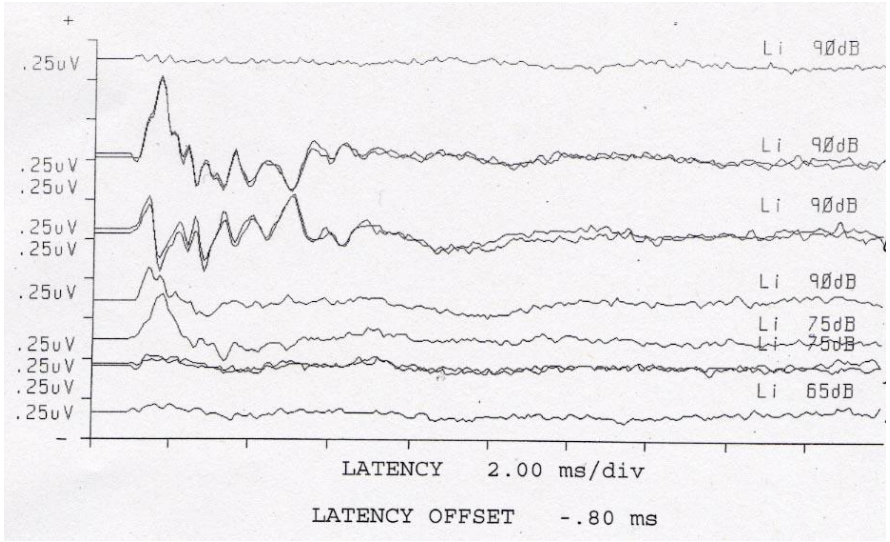
21% of childhood hearing loss detected from infants in the NICU was AN

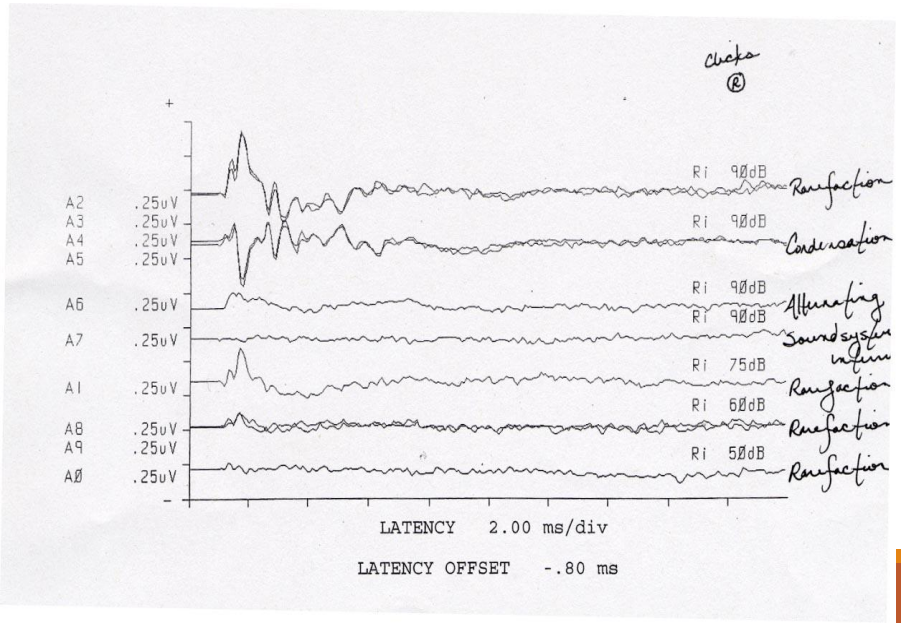
Presented at Am Acad Audiology, 2004

NHS 2008 Poster by Owen et al., Parkland Hospital, Dallas

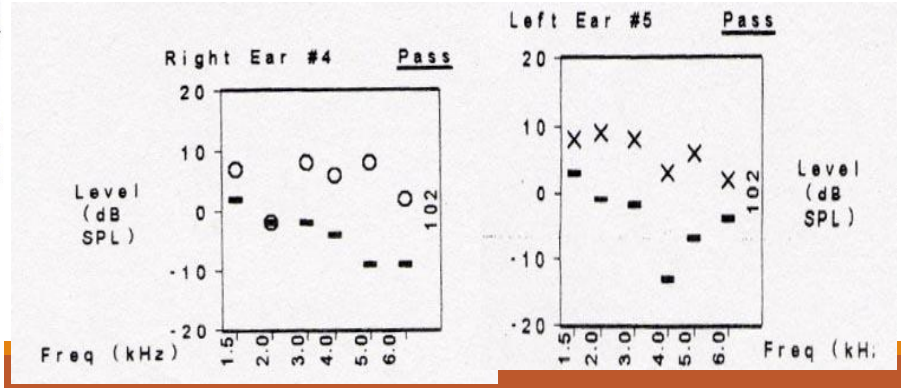
- **392 Infants identified with auditory disorder, 35 (8.9%) have an ANSD type pattern.**
- **14 of the AN babies were identified in the NICU and 21 were admitted to the well-baby nursery. This is the same pattern seen in identification of hearing loss in general, higher percentages in the NICU but equal numbers in each nursery!**
- **Adequate safeguards must be in place for infants who pass OAE in the WBN?**

Variable Presentation of ANSD Case #1
Present CM and OAEs

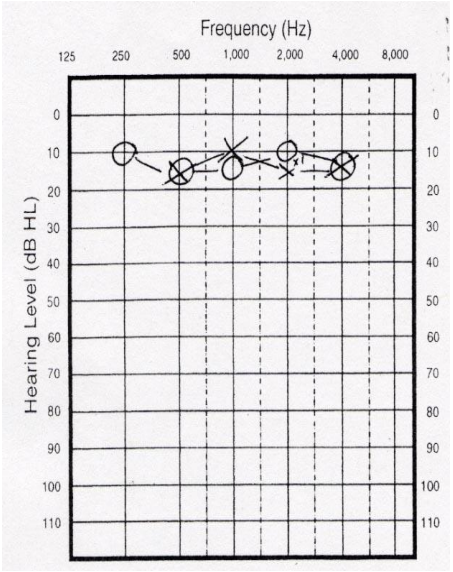




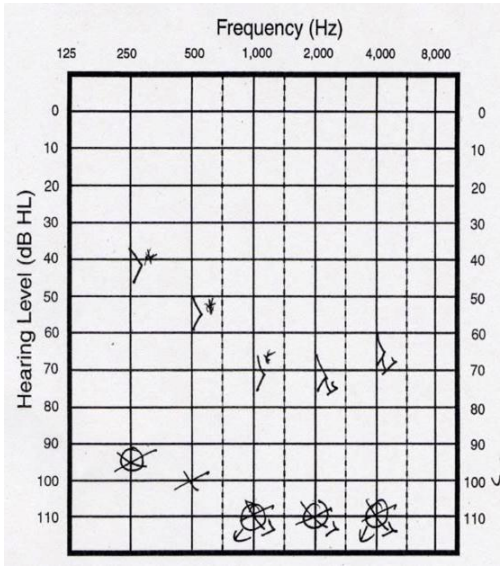
- 24 week preemie
- Intensive care nursery 4 months
- Ventilated 2 months
- ABR repeated at 18 months-no change



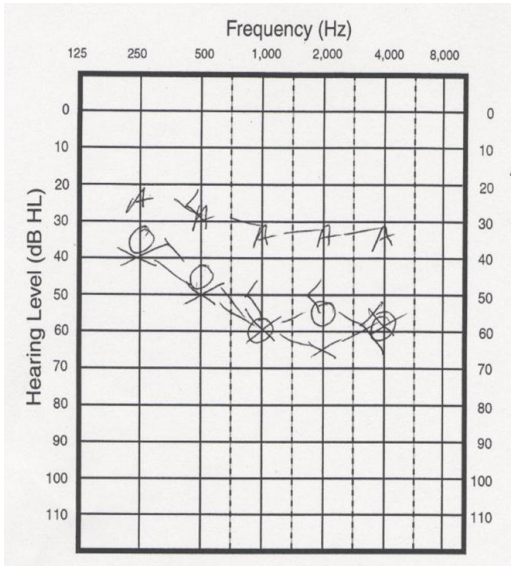
Hearing Levels Cannot Be Predicted from ABR or OAE



OAEs Present



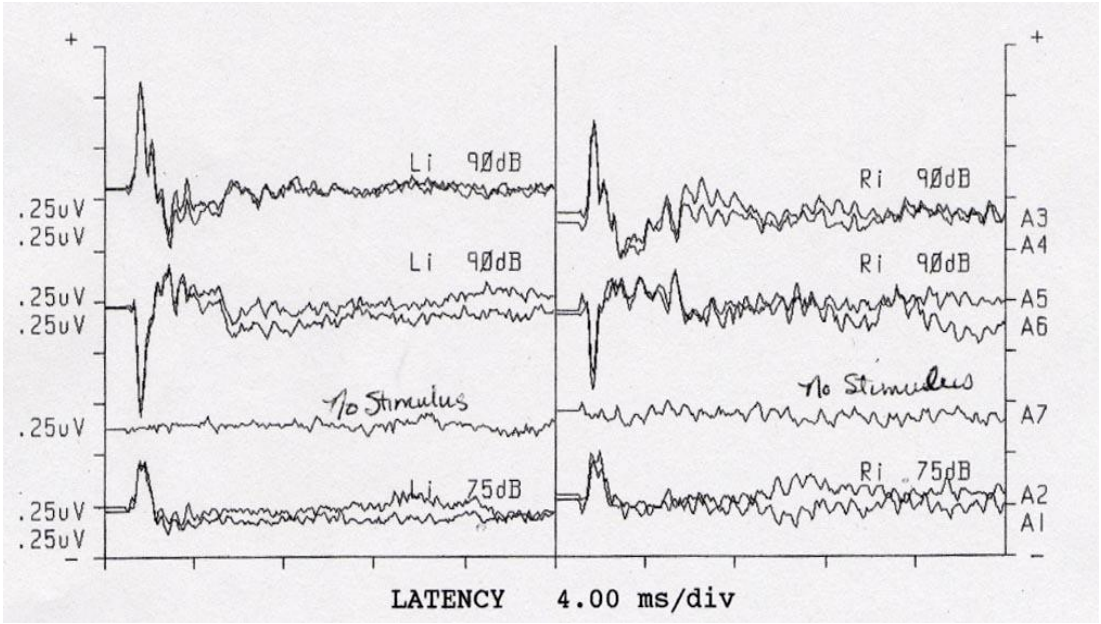
OAEs Present

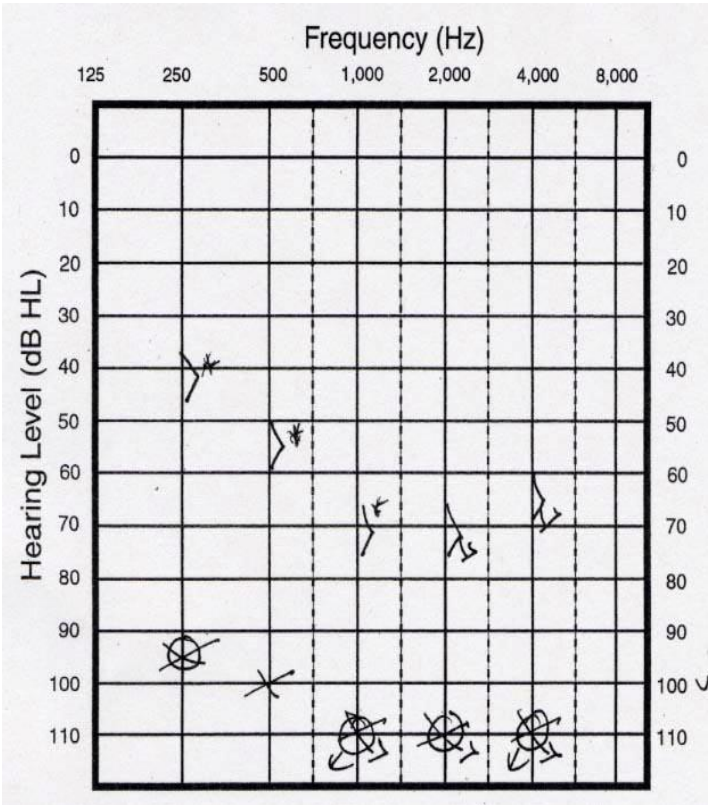


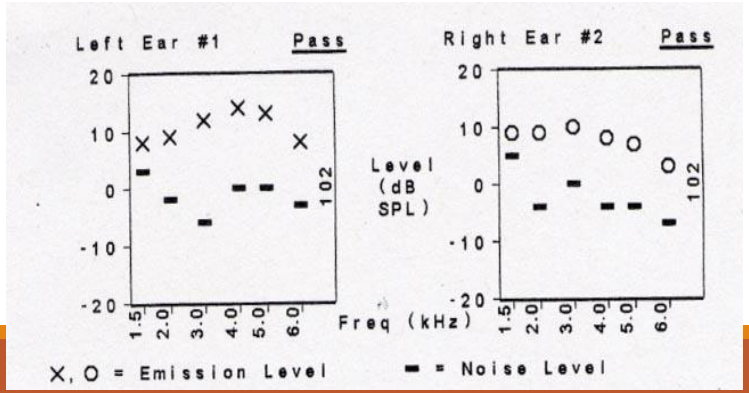
OAEs Absent

All have NO ABR, Present CM

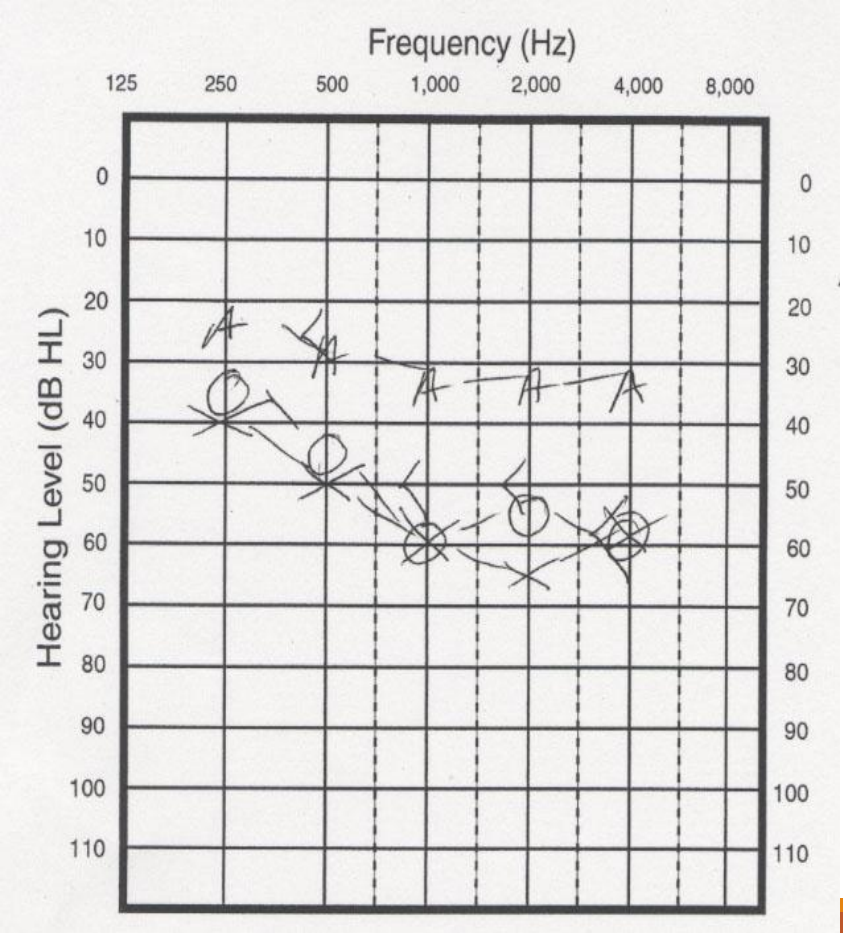
Variable Presentation of ANSD Case #2
Child with Profound Bilateral HL
Present CM and OAEs

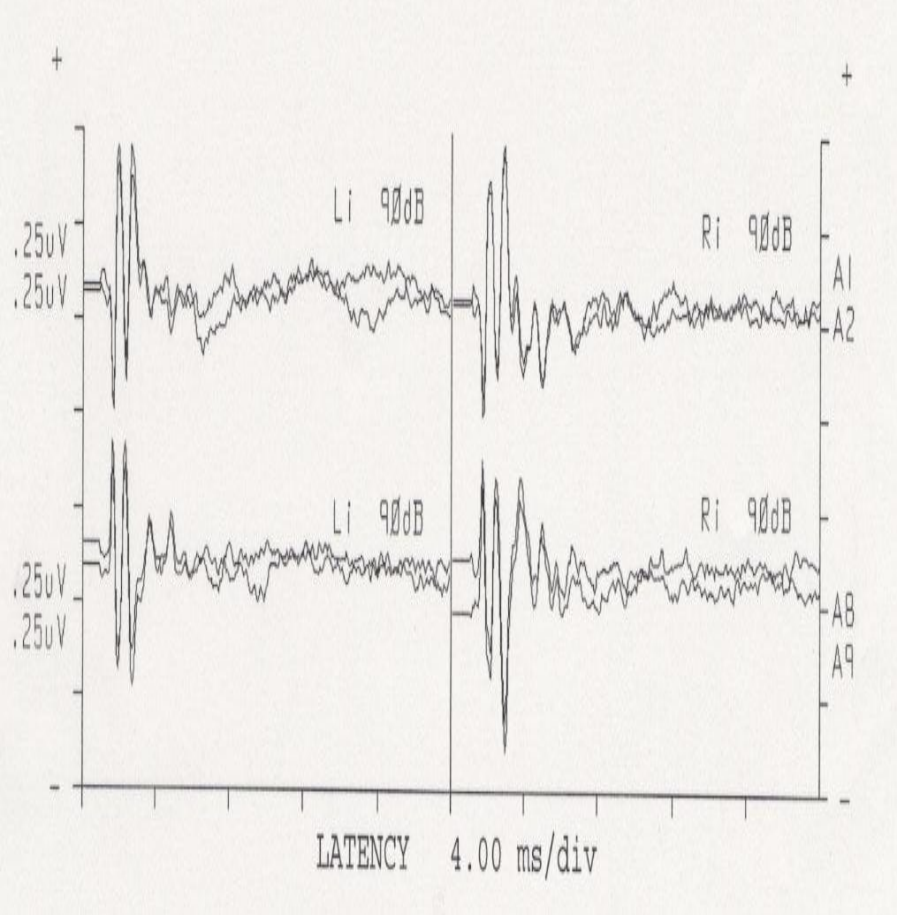




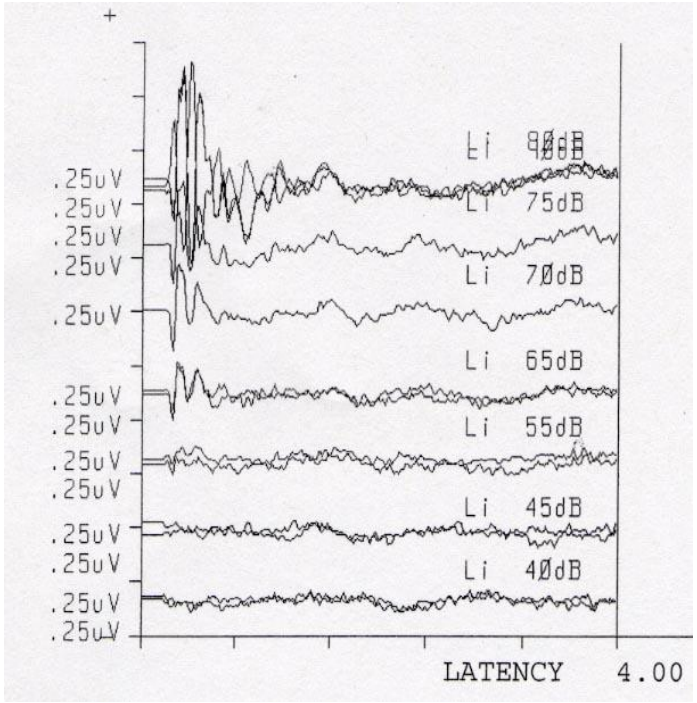


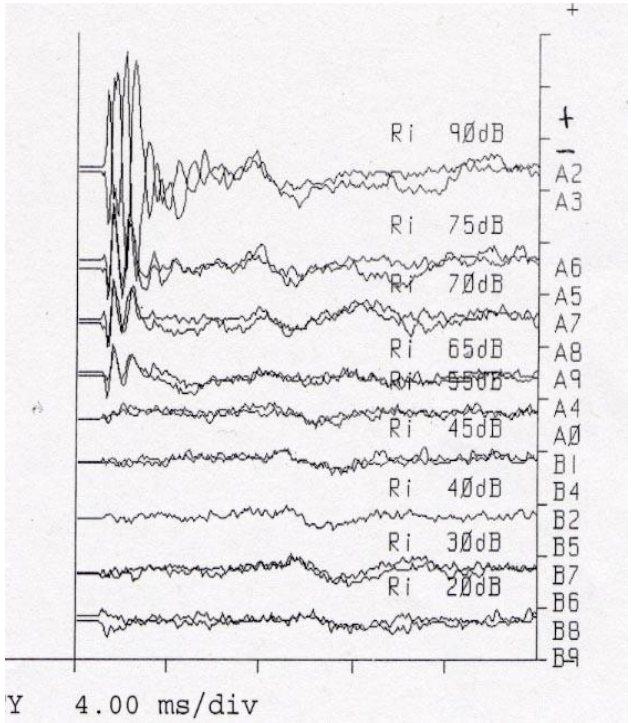
Variable Presentation of ANSD #3
Child with “moderate loss”
CM present, absent OAEs

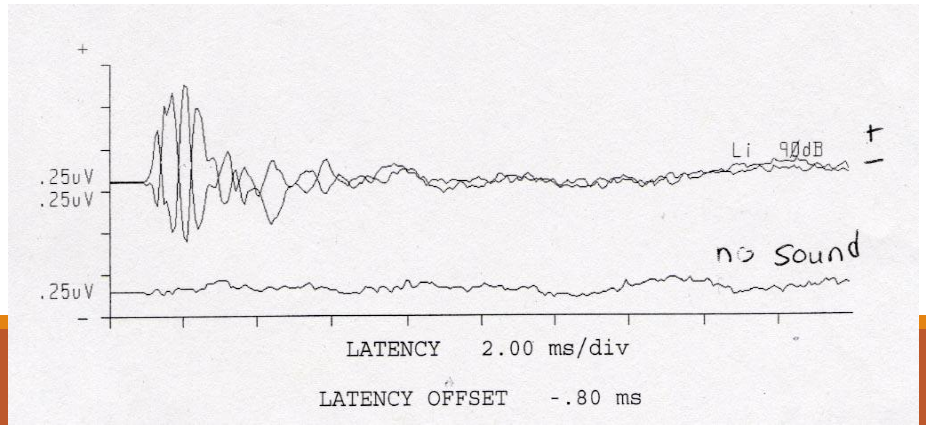


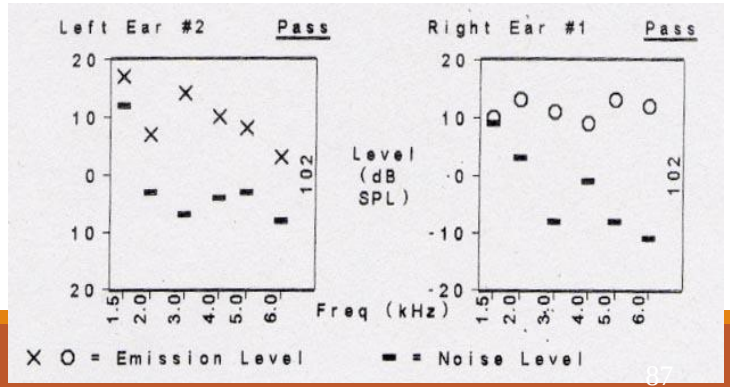


Variable Presentation of ANSD Case #4
Large CM, present OAEs but distal waveforms









Behavioral Audiometry Case #4
VRA with insert earphones
Age14 months