Current Developments in our Understanding of Auditory Neuropathy ANSD

[Auditory Neuropathy Spectrum Disorder]

Jon K. Shallop, Ph.D.
Professor Emeritus
Mayo Clinic Rochester, MN

Institut Raymond-Dewar Montreal, Canada June 25, 2009

BACKGROUND INFORMATION

- **Starr et al. (1991) identified auditory neuropathy (AN) as a timing disorder**
- **Zeng et al. (1999) demonstrated temporal processing problems in adults with auditory neuropathy.**

http://www.com.uci.edu/hesp/home.html

NeuroReport 10, 3429-3435 (1999)

Temporal and speech processing deficits in auditory neuropathy

Fan-Gang Zeng,[CA] Sandy Oba,[1] Smita Garde,[2] Yvonne Sininger[1] and Arnold Starr[3]

Department of Hearing and Speech Sciences and Center for Comparative Neuroscience, 0100 Lefrak Hall, University of Maryland, College Park, MD 20742; [1]House Ear Institute, Los Angeles, CA 90057; [2]University of Southern California, Los Angeles, CA 90089; [3]Department of Neurology, University of California, Irvine, CA 92717, USA

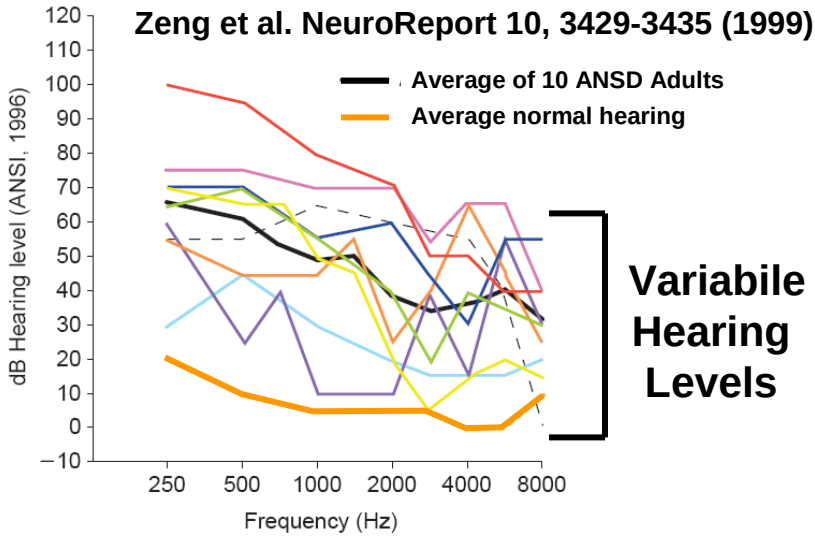
[CA]Corresponding Author

AUDITORY neuropathy affects the normal synchronous activity in the auditory nerve, without affecting the amplification function in the inner ear. Patients with auditory neuropathy often complain that they can hear sounds, but cannot understand speech. Here we report psychophysical tests indicating that these patients' poor speech recognition is due to a severe impairment in their temporal processing abilities. We also simulate this temporal processing impairment in normally hearing listeners and produce similar speech recognition deficits. This study demonstrates the importance of neural synchrony for auditory perceptions including speech recognition in humans. The results should contribute to better diagnosis and treatment of auditory neuropathy. *NeuroReport* 10:3429–3435 © 1999 Lippincott Williams & Wilkins.

Key words: Acoustic simulation; Auditory neuropathy; Hearing disorder; Human; Neural synchronization; Speech perception; Temporal processing

The characteristics of Hearing Loss in ANSD

Zeng et al. NeuroReport 10, 3429-3435 (1999)



Variabile Hearing Levels

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BACKGROUND INFORMATION

- Lesinski-Schiedat A. Frohne C. Hemmaouil I. Battmer RD. Lenarz T. Funktionelle Taubheit bei peri-synaptischer Audiopathie - Isolierte Storungen der inneren Haarzellen? Laryngo- rhino-otologie. 80(10):601-4, 2001
- Subjective deafness in case of **peri-synaptic audiopathy**. Isolated defects of the inner haircells?
- "Hypoxia, carboplatin, ototoxicity and metabolic disorders are possible etiologies for damage to the inner hair cells or synapsis."

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BACKGROUND INFORMATION

- **Rodriguez-Ballesteros M. del Castillo FJ. Martin Y. Moreno-Pelayo MA. Morera C. Prieto F. Marco J. Morant A. Gallo-Teran J. Morales-Angulo C. Navas C. Trinidad G. Tapia MC. Moreno F. del Castillo I.**
- **Unidad de Genetica Molecular, Hospital Ramon y Cajal, Madrid, Spain.**
- Auditory neuropathy **in patients carrying mutations in the otoferlin gene (OTOF).**
- ***Human Mutation*. 22(6):451-6, 2003 Dec.**

Varga R, Kelley PM, Keats BJ, et al. Non-syndromic Recessive Auditory Neuropathy is the Result of Mutations in the Otoferlin (OTOF) Gene. J Med Genet 40:45-50, 2003.

BACKGROUND INFORMATION

- Berlin et al. (2001), and Hood (2002) proposed auditory dys-synchrony as a more descriptive term. Typical findings include:
  - abnormal evoked potentials
  - otoacoustic emissions (usually present)
  - absent middle ear reflexes
  - lack of benefit from amplification

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Prevalence

There are no known data on how many children with ANSD have been implanted but if we assume that there are approximately 140,000 cochlear implants worldwide at the present time (Wilson and Dorman, 2007) and that 40% of them are children, then there are at least 56,000 children worldwide with cochlear implants.

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Prevalence

A conservative estimate of 10% for ANSD (Uus and Bamford, 2005) can be argued which suggests that more than 5,000 children with cochlear implants worldwide are likely to have ANSD as a component of their hearing loss etiology.

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Table 1: Prevalence of ANSD in "at-risk" infant populations

Study	Population	# Subjects	No. of ANSD	subjects % of Total
Stein et al. (1996)	Special Care Nursery	100	4	4.00
Psarommatis et al. (1997)	Intensive Care Unit	102	2	1.96
Rance et al. (1999)	"at-risk" infants	5199	12	0.23

Rance 2005 AN/AD and it's Perceptual Consequences

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Table 2: Prevalence of ANSD in children with permanent hearing loss.

Study	Population	No. of Cases Permanent Hg. Loss	No. of AN/AD Cases	% of Total
Kraus et al. (1984)	Hg. impaired children	48	7	14.58
Park & Lee (1998)	Hg. impaired children	139	7	5.04
Vohr et al. (1998)	Universal screening	111	2	1.80
Rance et al. (1999)	"at-risk" infants	109	12	11.01
Berlin et al.	Hg. impaired	1000	87	8.70

The characteristics and co-morbidities of ANSD

Children with ANSD can present with some unique characteristics that may initially be misunderstood by parents teachers and clinicians.

This also implies that some children presently in classrooms may in fact have this disorder.

The characteristics and co-morbidities of ANSD

In general, various physical, sensory or cognitive issues can be observed in addition to hearing loss and <u>all of these will be educationally significant</u>.

As these "issues" arise, there is an increase in the probability of auditory neuropathy [ANSD] as an additional problem.

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The characteristics and co-morbidities of ANSD

- **Developmental delays**
- **Learning disabilities, ADD, ADHD**
- **Autism spectrum disorders**
- **Emotional or behavioral problems**
- **Uncorrected visual problems, blindness**

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The characteristics and co-morbidities of ANSD

- Other health impairment
- Cerebral palsy, motor disorders
- Apraxia
- Inner ear malformations, Atretic or absent auditory nerve
- Seizures
- Syndromes
- Other

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Lessons Learned:
The characteristics and co-morbidities of ANSD

- Hearing is usually abnormal in these children
- However they may exhibit good sound “detection”
- And very poor speech recognition in most cases
- But some will benefit from hearing aids
- …. And why ? Sound demonstration

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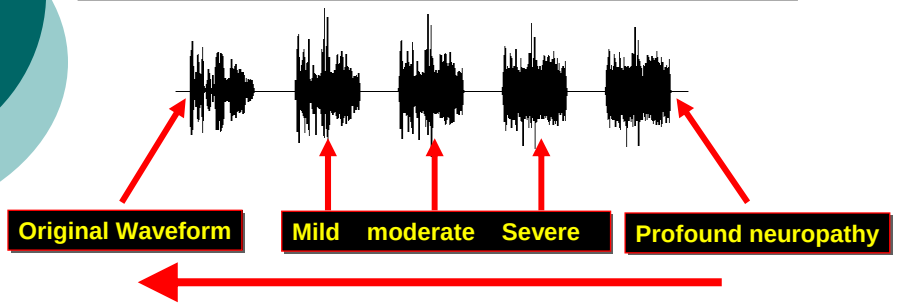
Acoustic Simulations of ANSD

Notice the acoustic 'smearing'



Simulation : from "profound" to "original" speech segments progressively overlap resulting in a "smearing" effect.

Fan-Gang Zeng et al. (1999)

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Gap Detection

Speech is a series of sequential events with "gaps" between words

My house is your- house
ma maison est votre maison

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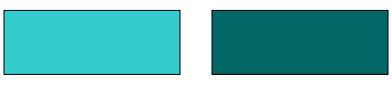
Gap Detection

However, what if the words overlap ?

ma maison est votre maison

Gap Detection

What is gap detection and why is it important for speech recognition ?



Gap Detection

What is gap detection and why is it important for speech recognition ?

Speech is segmented

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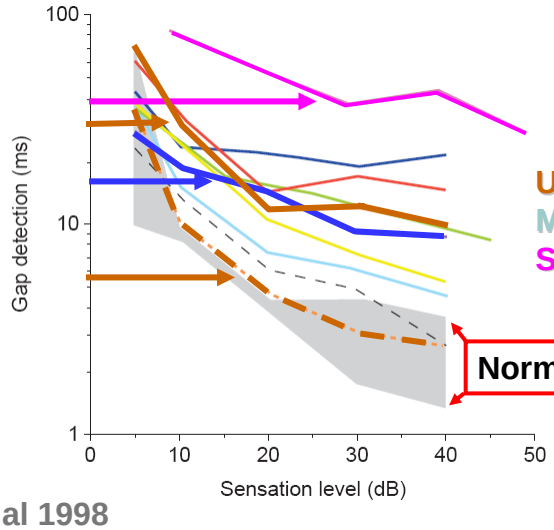
Gap Detection is abnormal for persons with ANSD



Zeng et al 1998

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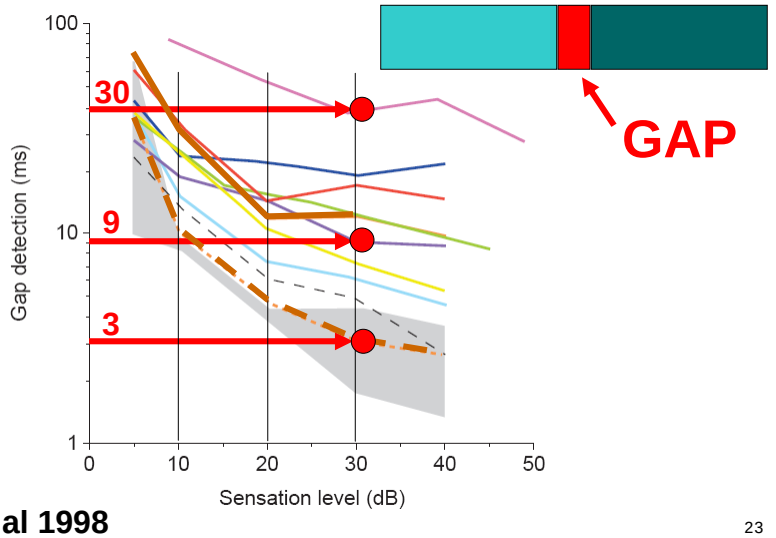
Gap Detection is abnormal for persons with ANSD



Zeng et al 1998

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Lessons Learned

Speech recognition is very dependant on the ability to make fine timing discriminations in the speech signal which has small temporal gaps and transitions

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Gap Detection

Some examples of the importance of gap detection

"cue" vs "coo" /ku/ vs. /kiu/

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Gap Detection

Some examples of the importance of gap detection

"stop" vs "shop"

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Case Study #1

Profound Hearing Loss in the Right ear ?

* Passed OAE screening in both ears
* Absent ABR in **Right** ear, Normal ABR in **Left** ear

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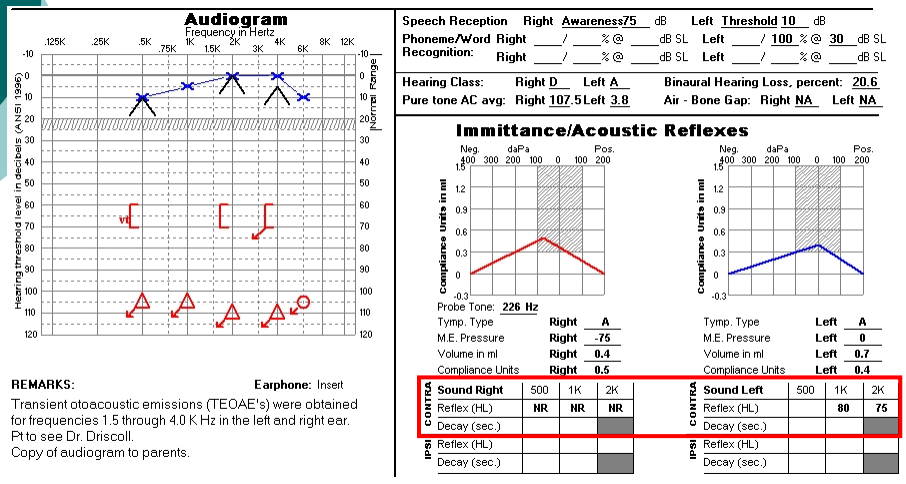
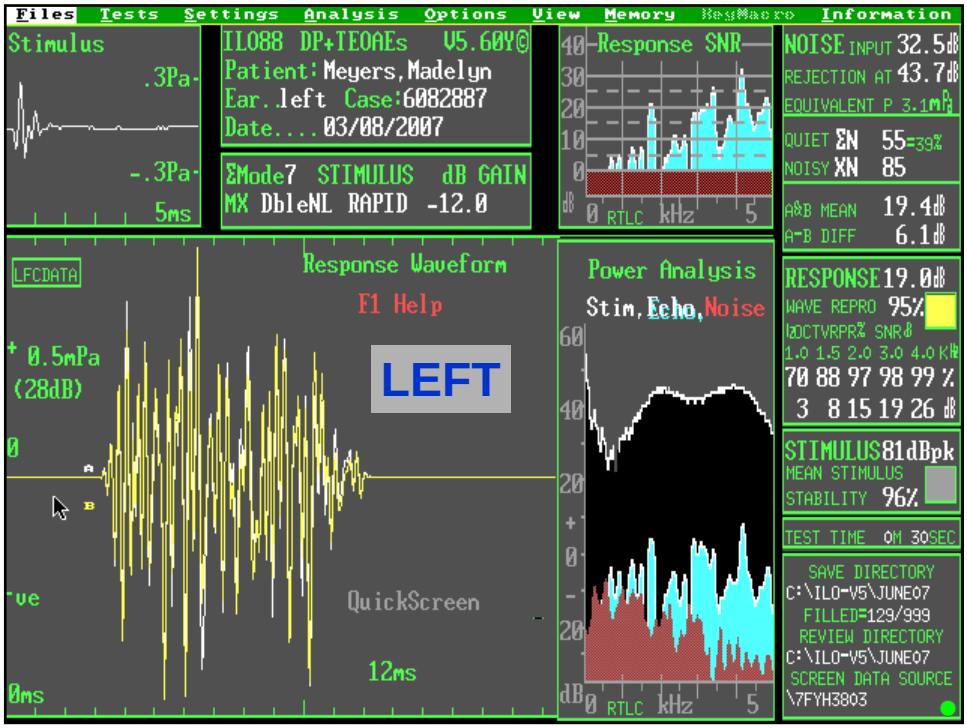
Otoacoustic Emissions Present In Both Ears



Contra-lateral acoustic reflexes present: sound in LEFT ear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.3Pa
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Patient: Meyers,Madelyn
Ear. left Case:6082887
Date.... 03/08/2007
EMode7 STIMULUS dB GAIN
MX DbleNL RAPID -12.0
Response SNR
NOISE INPUT 32.5dB
REJECTION AT 43.7dB
EQUIVALENT P 3.1mPa
QUIET ΣN 55=39%
NOISY XN 85
A&B MEAN 19.4dB
A-B DIFF 6.1dB
LFCDATA
Response Waveform
F1 Help
+ 0.5mPa
(28dB)
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QuickScreen
12ms
0ms
LEFT
Power Analysis
Stim,Echo,Noise
RESPONSE 19.0dB
WAVE REPRO 95%
OCTVRPR% SNR dB
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70 88 97 98 99 %
3 8 15 19 26 dB
STIMULUS 81dBpk
MEAN STIMULUS
STABILITY 96%
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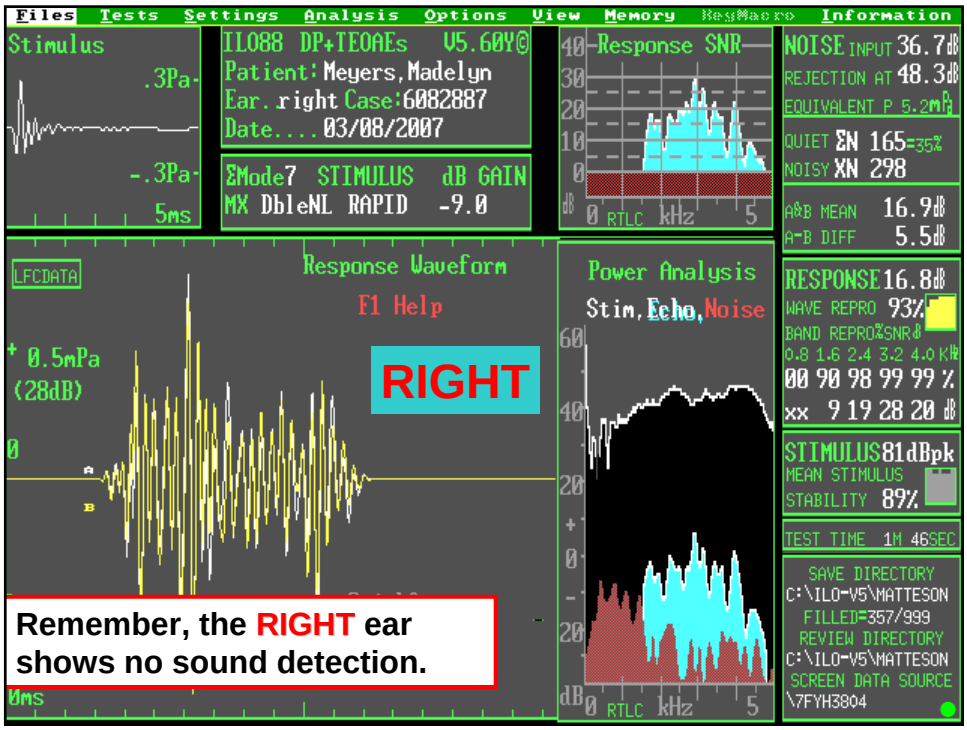
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Ear. right Case:6082887
Date.... 03/08/2007
EMode7 STIMULUS dB GAIN
MX DbleNL RAPID -9.0
Response SNR
NOISE INPUT 36.7dB
REJECTION AT 48.3dB
EQUIVALENT P 5.2mPa
QUIET ΣN 165=35%
NOISY XN 298
A&B MEAN 16.9dB
A-B DIFF 5.5dB
LFCDATA
Response Waveform
F1 Help
+ 0.5mPa
(28dB)
0
RIGHT
0ms
Power Analysis
Stim,Echo,Noise
RESPONSE 16.8dB
WAVE REPRO 93%
BAND REPRO% SNR dB
0.8 1.6 2.4 3.2 4.0 kHz
00 90 98 99 99 %
xx 9 19 28 20 dB
STIMULUS 81dBpk
MEAN STIMULUS
STABILITY 89%
TEST TIME 1M 46SEC
SAVE DIRECTORY
C:\ILO-V5\MATTESON
FILLED=357/999
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\7FYH3804
Remember, the RIGHT ear shows no sound detection.

Lessons Learned

A profound unilateral hearing loss can be caused by the absence of an auditory nerve and cochlear function could be normal in such cases.

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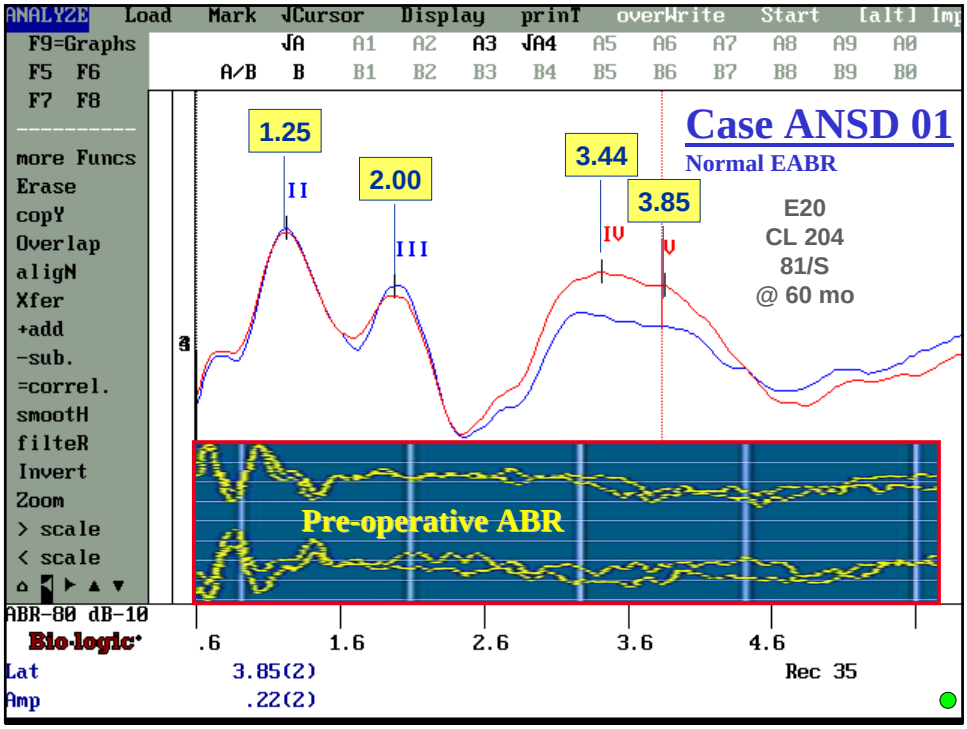
Temporal Bone Study of Premature Infants

(Courtesy of Mass. Eye and Ear, published June 2001 Archives Otolaryngology)

Abnormal anatomical findings in autopsies of a group of pre-mature babies

Note missing inner hair cell, normal nerve fiber count inside the habenula perforata, and normal outer hair cells which would lead to normal emissions and NO ABR.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II
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Case ANSD 01
Normal EABR
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@ 60 mo
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3.6
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Lat 3.85(2)
Amp .22(2)
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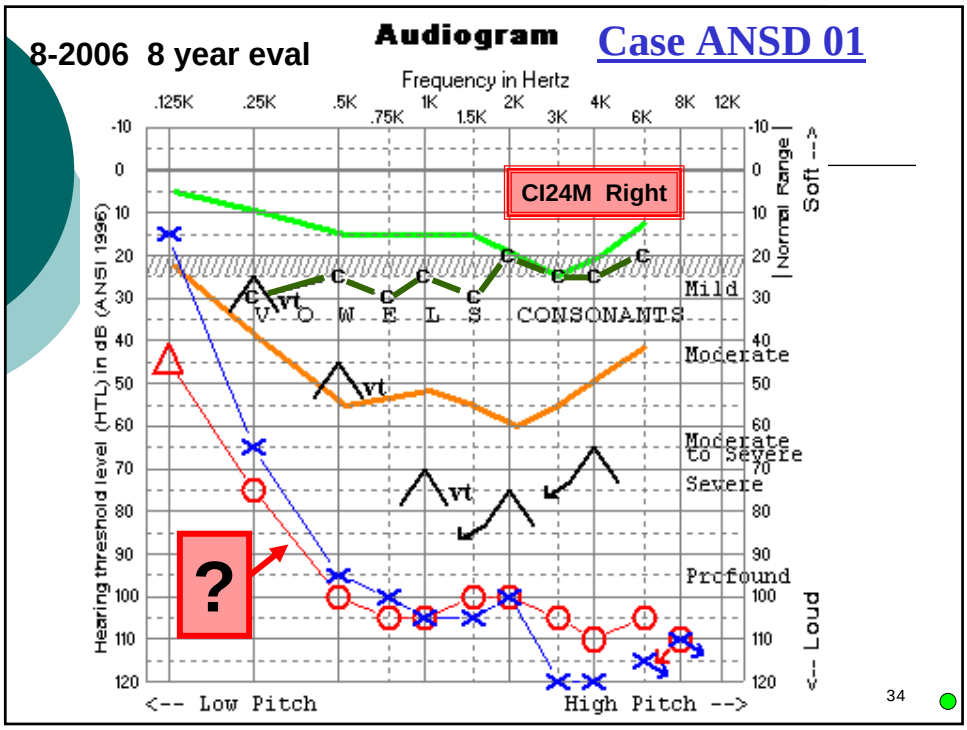
8-2006 8 year eval
Audiogram
Case ANSD 01
Frequency in Hertz
.125K .25K .5K .75K 1K 1.5K 2K 3K 4K 6K 8K 12K
CI24M Right
Hearing threshold level (HTL) in dB (ANSI 1996)
Normal Range
Soft -->
<-- Loud
Mild
Moderate
Moderate to Severe
Severe
Profound
VOWELS CONSONANTS
?
<-- Low Pitch
High Pitch -->
34

ANSD: C1 - 8 year test results 60 dBA SPL

- CNC words: 60% correct for words & 78% phonemes
- HINT sentences in quiet: 97% correct
- BKB-SIN 4 list average: 10.25 dB SNR for 50% correct
- She performs at the average speech recognition levels as our adults.

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Summary Points

- Hearing loss is typically a loss of **sensitivity** in hearing levels as demonstrated on an audiogram
- But hearing loss can also be a loss of the ability to make fine **temporal** and **frequency** discriminations
- These factors will reduce the ability of children to "understand" spoken language

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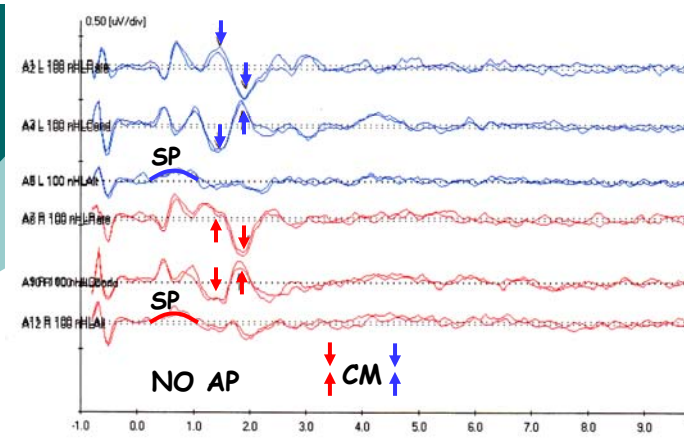
Case Study X01MK DOB June 06

This child was not screened for hearing at birth and her diagnosis has been delayed.

Audiology- February 2008 – Wearing Spirit3 SP digital hearing aids since November 2007. Not wearing HAs consistently but has begun to wear them since Christmas. She is beginning to say “dada”.

		1kHz	2kHz	4kHz
VRA aided sound field dB HL		35	40	50
VRA inserts	Right	85	80	80
	Left 85	80	85	
Unmasked bone conduction		50	70	NR

Beaumont June 08 – ABR LEFT & RIGHT and OAEs under GA ABR tracings for rare vs. condensation clicks only show very large cochlear microphonics of .8 to 1.0 uV in each ear at 100 dB nHL. OAEs are absent on this date. Child has patent groments in situ R/L today.



Left
100 rare
100 con
100 alt
Right
100 rare
100 con
100 alt

MRI February 09

T2 axial imaging shows ventriculomegaly involving the 4th, 3rd and lateral ventricles. Most prominent in the left lateral ventricle. There is no transpendymal

oedema. Significant white matter loss bilaterally, more left than right. ***Appearances are suggestive of peri ventricular leuko malacia with almost complete loss of white matter on the left. 7th and 8th nerves and inner ear structures appear to be normal bilaterally***.

Lessons Learned

- **ALL children being considered for a cochlear implant should have MRI of the brain and auditory nerve cross section to visualize the 4 nerves within the internal auditory canal and verification of brain structures.**

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Lessons Learned

- Hearing loss is typically a loss of sensitivity in hearing levels as demonstrated on an audiogram
- But hearing loss can also be a loss of the ability to make temporal discriminations
- Both factors will reduce the ability of children to "understand" spoken language

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