The Detection and Management of Auditory Neuropathy/Dys-synchrony

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Re-name the observations:

- AUDITORY DYS-SYNCHRONY or
- Auditory Neuropathy Spectrum Disorders

Recommended Test Order

- Tympanometry
- Middle Ear Muscle reflexes
- Otoacoustic Emissions
- ABR/ EcochG
- WHY??

Auditory Neuropathy: Review of What it is and how we find it?

- Patients who have normal Otoacoustic Emissions but absent ABRs and Absent Middle Ear Muscle Reflexes almost always have some form of "Auditory Neuropathy".
- It is diagnosed with combined use of ABR with a positive and negative polarity click, and otoacoustic emission testing, which should be preceded by Immittance testing.
- See Hood, 1999; Sininger et al., 1995; Starr et al. 1993,1996; Berlin et al.,1993, 1998, and many others.

Remember that they also often have...

- Inverting CMs which masquerade as ABRs or at least Wave I of the ABR to the unwary.
- Absent efferent suppression of emissions.
- Absent MLDs.
- Poor sound localization and speech perception.
- All of which point to....

A disorder of neural synchrony which can be accompanied by pure tone behavioral audiograms ranging from near normal to “total deafness”

Tests for Making the Diagnosis

- Otoacoustic Emissions — Present
- MEMRs — Absent
- ABR — Absent
- Cochlear microphonics — Present
- Pure tone thresholds — Variable
- Speech audiometry — Usually poor, especially in noise
- Other measures...

Two Demonstrations

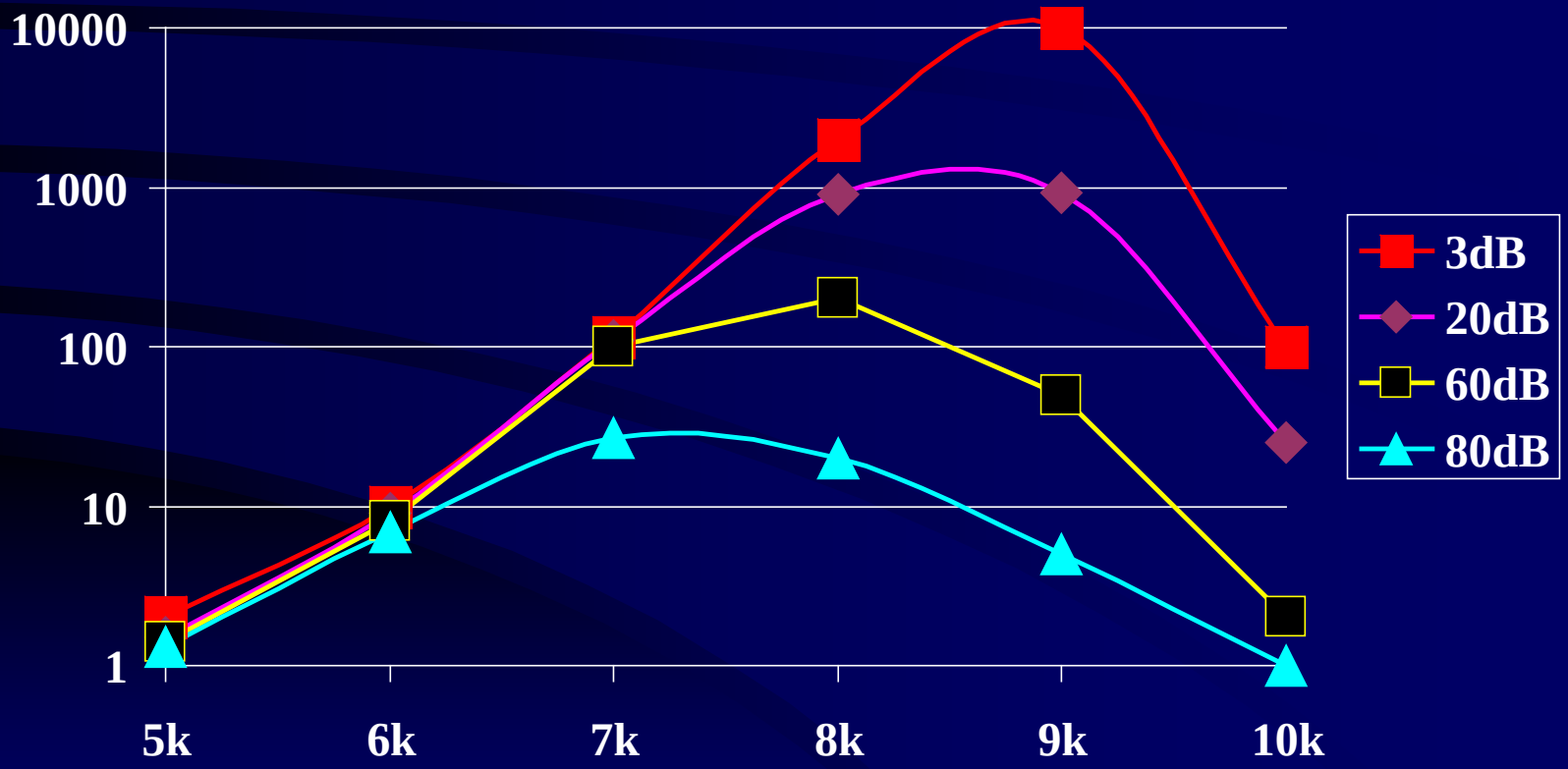
- A moving organ of Corti to describe to parents and other interested professionals how we can study the organ clinically with emissions and ABR or EcochG and why these patients are not “garden variety” deaf children. (www.neurophys.wisc.edu).
- A sample of digital speech whose timing has been disrupted in a way suggestive of what might be happening with these patients. Courtesy of Fang Geng Zheng, Voice Arnold Starr MD.

Why not put hearing aids on children like this who have poor audiograms?

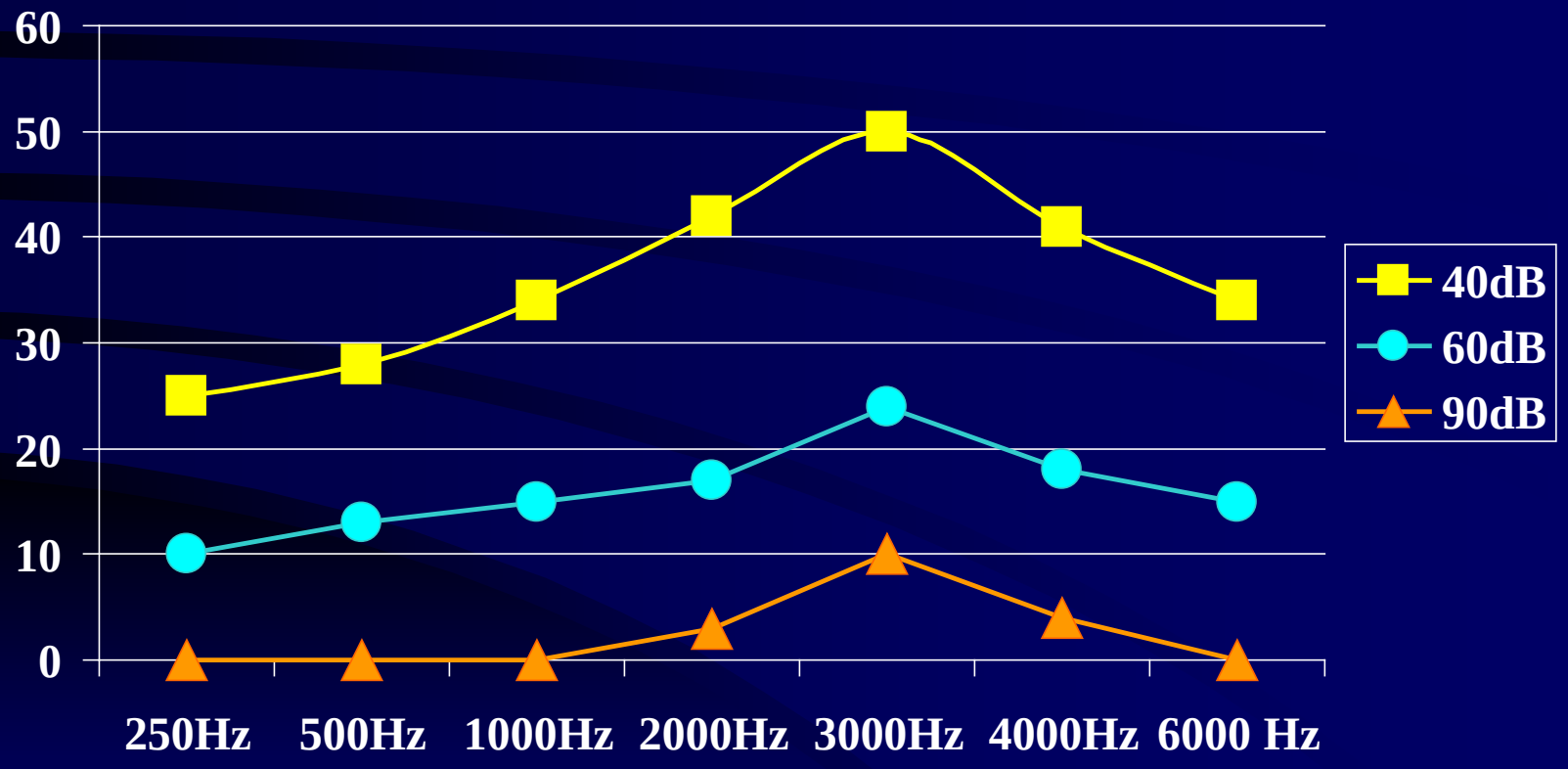
- In order to follow the rationale, we have to first review cochlear physiology and otoacoustic emissions.
- There are 5 electroacoustic events commonly measured in the cochlea.
- EP, CM, CAP and ABR, SP and OAE's.
- How do outer hair cells work?

Displacement of the Chinchilla Basilar Membrane Relative to the Stapes

adapted from Ruggero in Berlin, 1996



Idealized Gain Function of a hearing aid which would do somewhat the same thing in the intensity domain and whose compression knee begins at 40 dB input adapted from Berlin, 1996



How can we explain what's happening in their auditory systems

- Some of our patients get better, and some others show progressive loss of cochlear microphonic and emissions.
- Few seem to do well with hearing aids in terms of learning language auditorily.
- Some show remarkable progress after cochlear implantation becoming quite auditory and verbal.
- Some develop peripheral neuropathies in other systems.

Therefore it is likely that...

- “Auditory Neuropathy” is a description of unexpected test results *(absent or abnormal ABR, present emissions and cochlear microphonics, absent MEMR, absent efferent suppression, absent MLD etc.)* with multiple etiologies and time courses.
- Thus, the disorder may be in part a misnomer but for now a convenient descriptor until we learn how to separate the various prognoses and underlying mechanisms.

Theoretical Animal Models

- The Bronx Waltzer Mouse (bv/bv).
- The chinchilla and/or mouse whose inner hair cells have been chemically compromised with carbo-platin *(Harrison et al., Salvi et al.)*
- The Tetrodotoxin treated animal where transmitter actions have been blocked but structures remain?

This is sometimes a genetic disorder

- Siblings have been recorded.
- Entire families have been recorded (Starr et al. Slovenian families).
- A few putative genes have been isolated (Vargas et al.;)
- There may be a genetic hypersensitivity to clinically low levels of bilirubin in people who recover.

The final common path probably involves

- Failure of inner hair cells to communicate synchronously to primary single units of the auditory nerve.

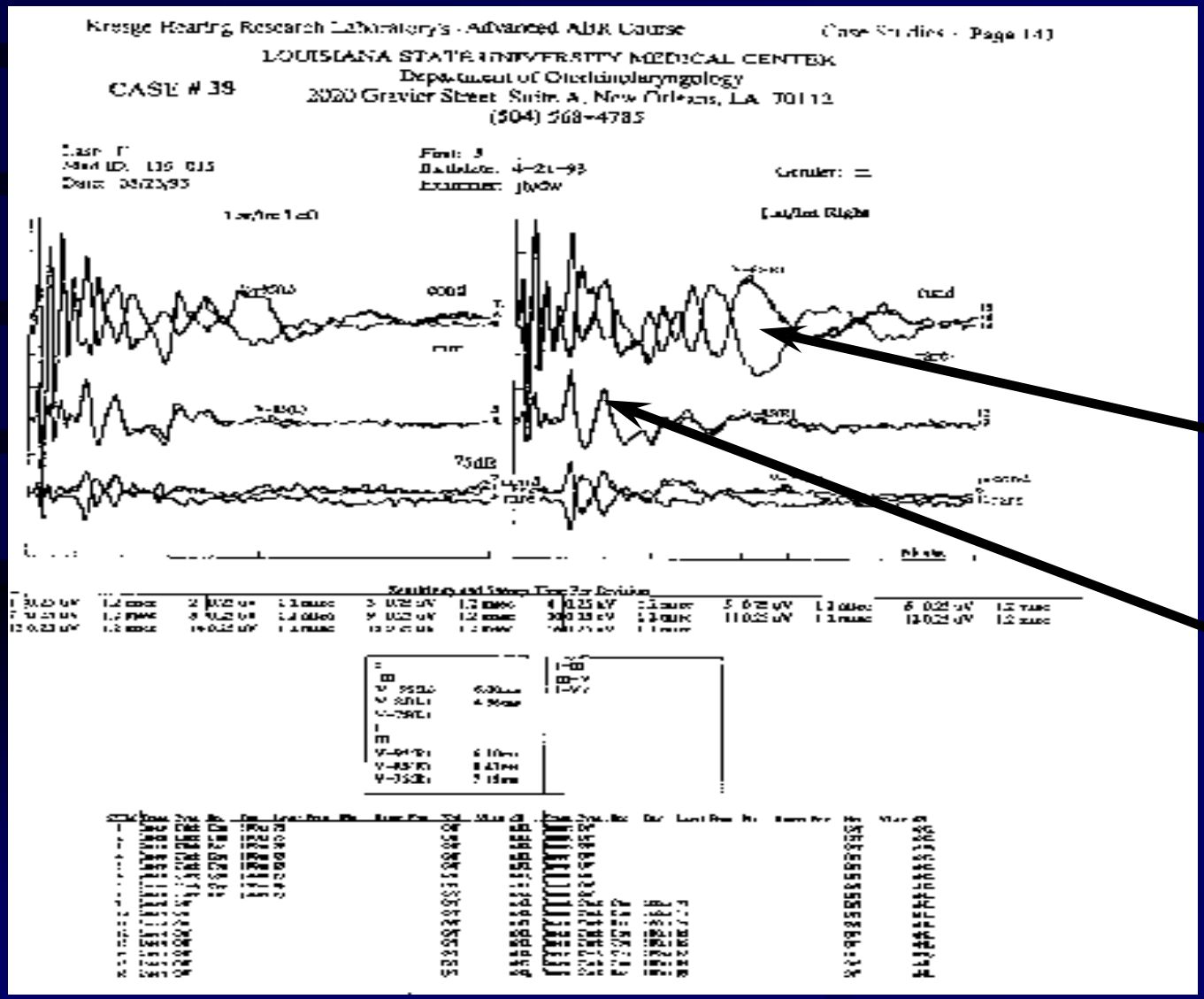
Some Putative Mechanisms which may explain success of CIs

- The Inner Hair Cells may be missing or compromised
- They may not be mechanically accessible (compromised or ineffective tectorial membrane).
- They may be chemically or mechanically prevented from controlling neural elements (kernicterus deposits).
- There may be myelin dysfunction or transmission dysfunction as in MS ...axonal vs. Demyelinating disorder which will not respond well to CIs.

Time Course for Auditory Neuropathy Patients: Type PL

- Some Patients show a retrograde loss of cochlear microphonics, loss of otoacoustic emissions, and become audiologically almost indistinguishable from garden-variety deaf children. One such child in our practice has been successfully implanted after acquisition of Cued Speech and is doing extremely well. Labeled here Type PL for ***P***rogressive ***L***oss of peripheral auditory integrity.

Twin JO's ABR at LSU in 1993 shows all waves are cochlear microphonics

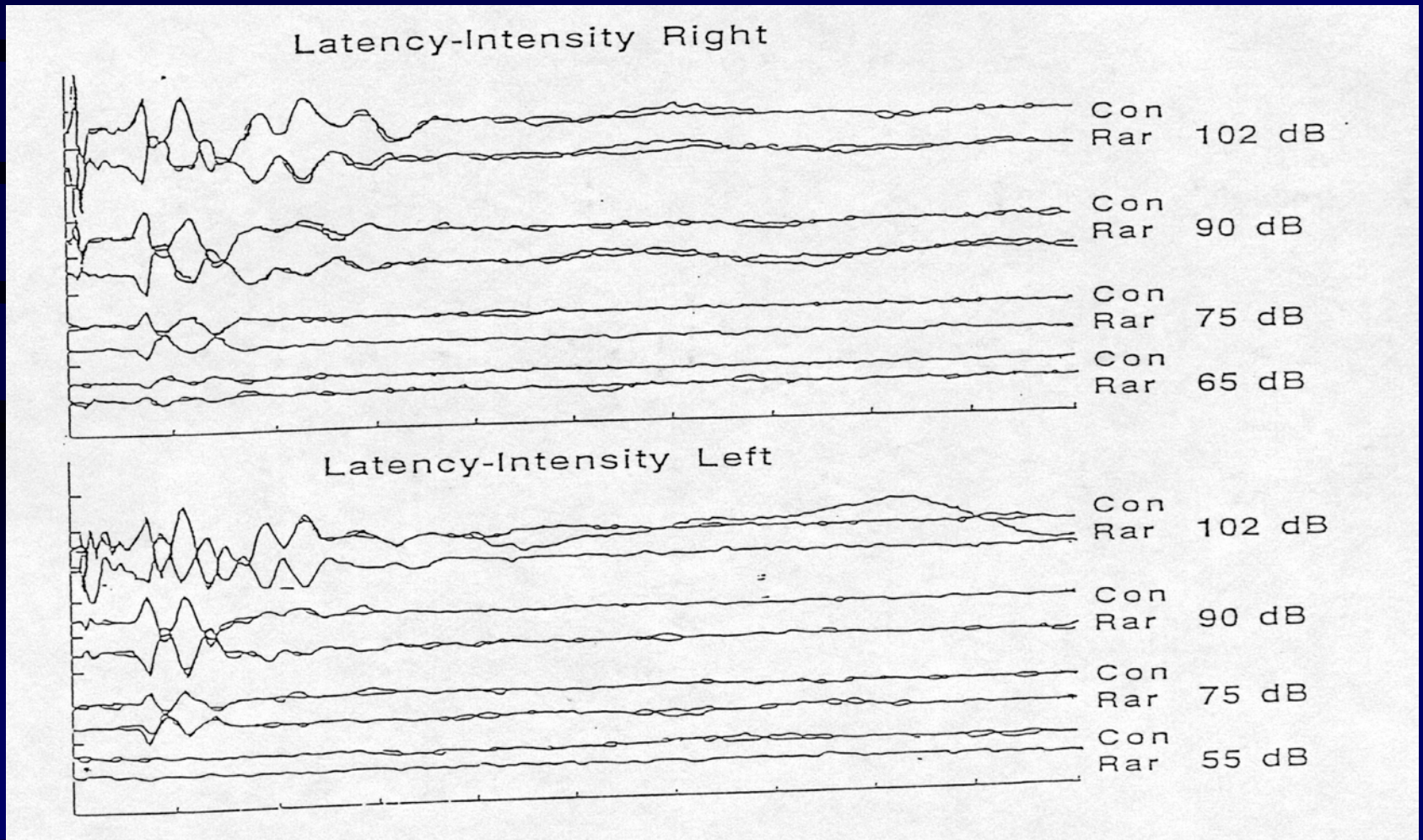


Note polarity inversion and no latency shift showing this to be a CM.

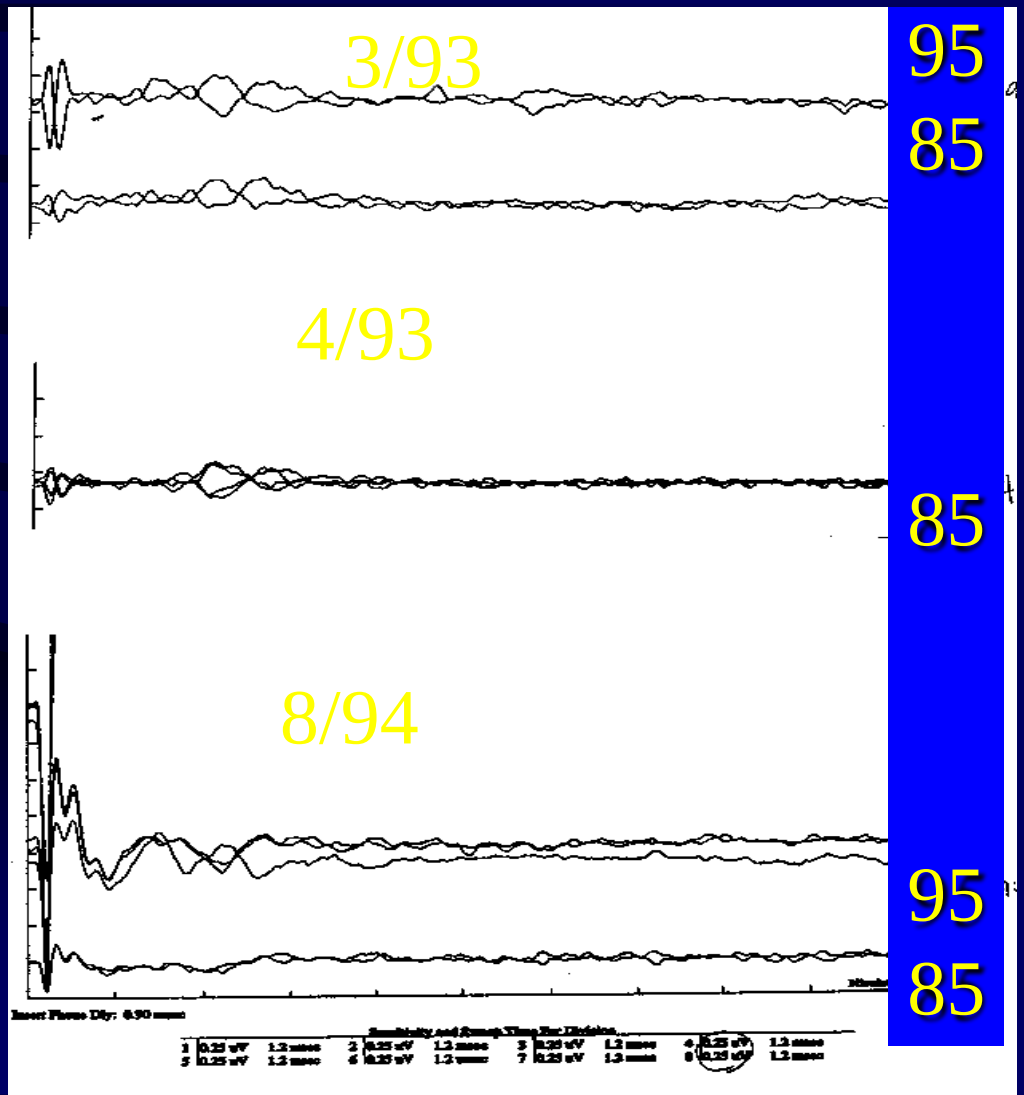
Masking Level Difference

- Low frequency pulsed tone, 40-50 dB in both ears.
- Find level at which narrow bands of noise in each ear masks the pulsed tone.
- Then...
- Reverse the phase of the tones (or the noise).
- Normal MLD release from masking will be 10-13 dB.
- Neuropathy patients show no release from masking

Type PL patient whose Cochlear Microphonic Masqueraded as an ABR and gradually disappeared



Type PL...Loss of CM over time



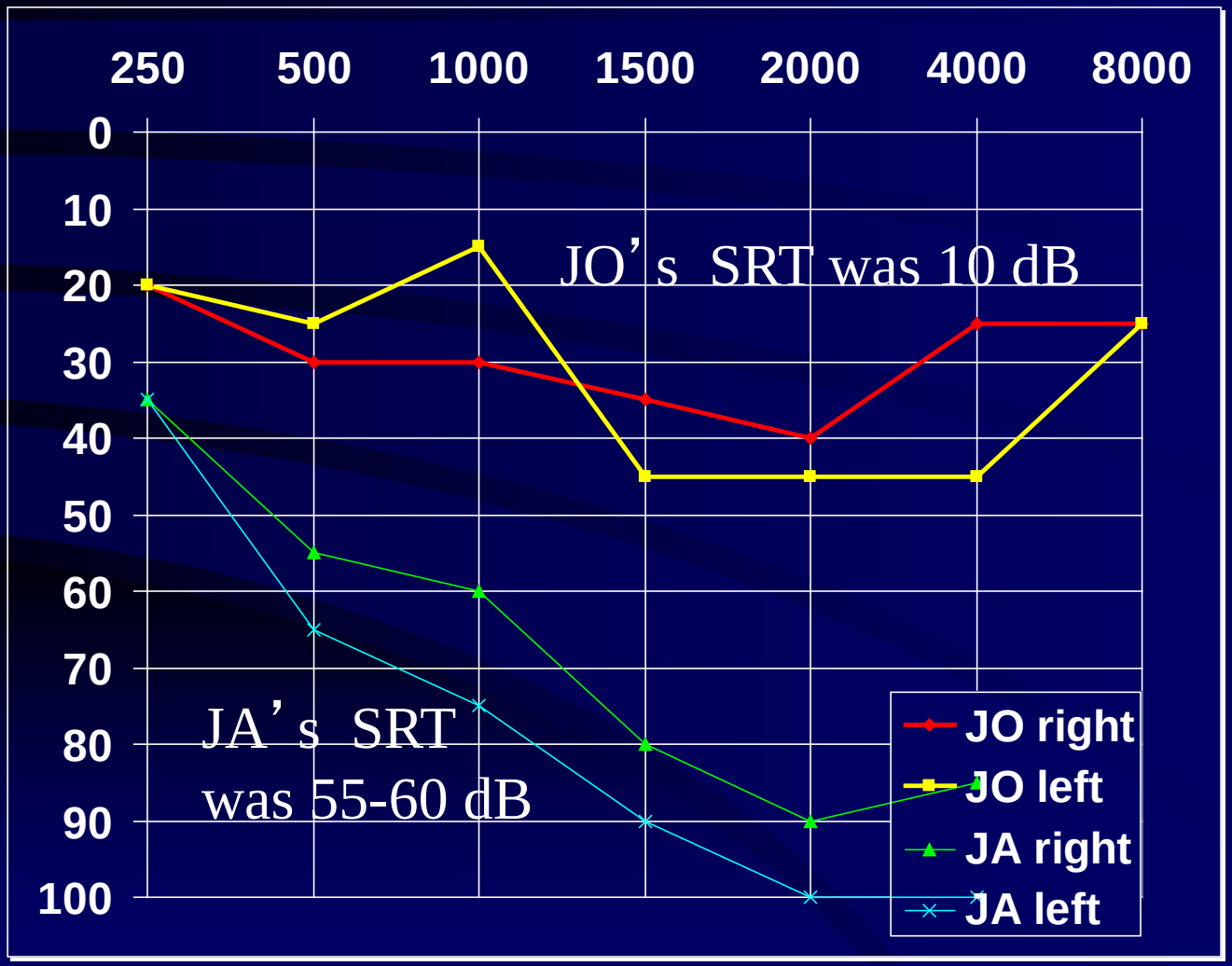
Clinical Suggestions to aid in identification

- Do emissions at least once on every new patient even for hearing aids, cochlear implants, and general diagnostic intake. We have seen normal emissions in “dead ears” and in unilateral hearing losses.
- Do ABRs with one positive and one negative polarity click to separate CM from AP.
- Do MEMR on every patient at least once. A paradoxically absent MEMR is neither normal nor always due to absent stapedius muscle. A non-acoustic reflex test will help rule out the latter.

Time Course for Auditory Neuropathy Patients: Type PR

- Still others seem to recover pure tone sensitivity and awareness of sound, but continue to show de-synchronized ABRs, robust cochlear microphonics and normal otoacoustic emissions. Called here Type PR for partial recovery. Speech and Language are delayed but develop.

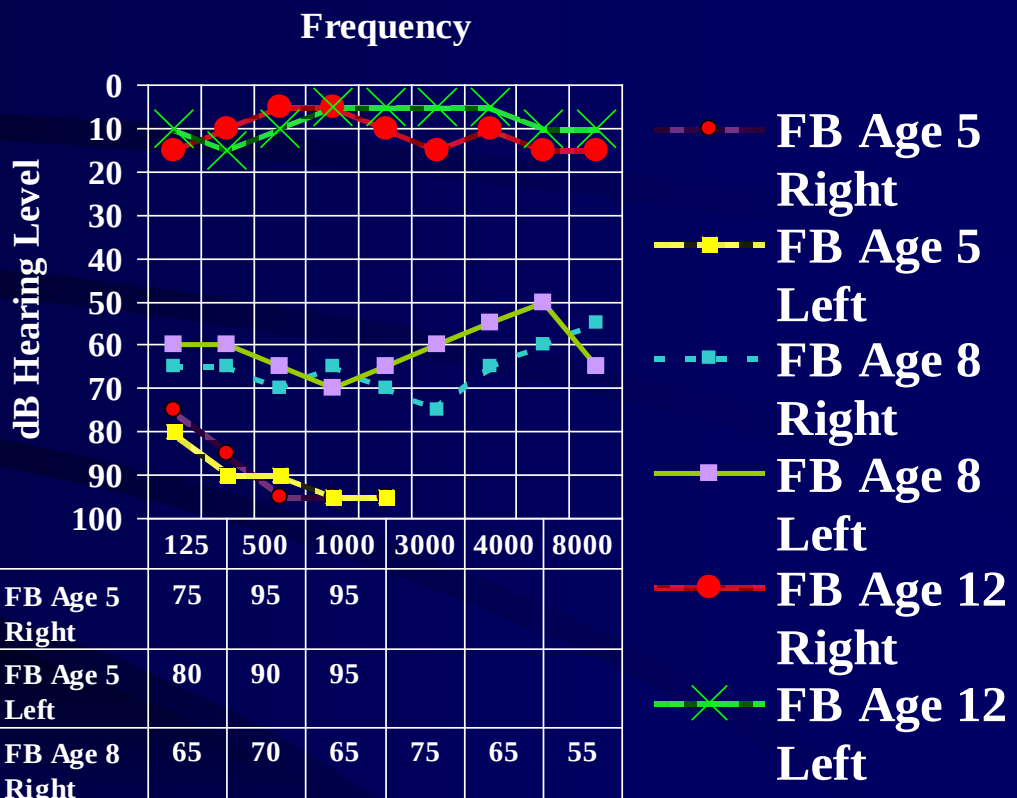
Twin JO (Type PR) compared to his brother JA (Type PL)



Igm.nlm.nih.gov

Patient FB: ABR absent at birth, Emissions not available. Diagnosed as Deaf and reportedly raised at a school for the Deaf without speech and with ASL only. No ABR at age 12, but a normal emissions, immittance, pure tone audiogram and awareness of many sounds. Limited speech, language and reading skills.

Type PR



	125	500	1000	3000	4000	8000
FB Age 5 Right	75	95	95			
FB Age 5 Left	80	90	95			
FB Age 8 Right	65	70	65	75	65	55
FB Age 8 Left	60	65	70	60	55	65
FB Age 12	15	5	5	15	10	15

Latest Data courtesy of parents and Jack Katz Ph.D. With the University of Buffalo Audiology Service

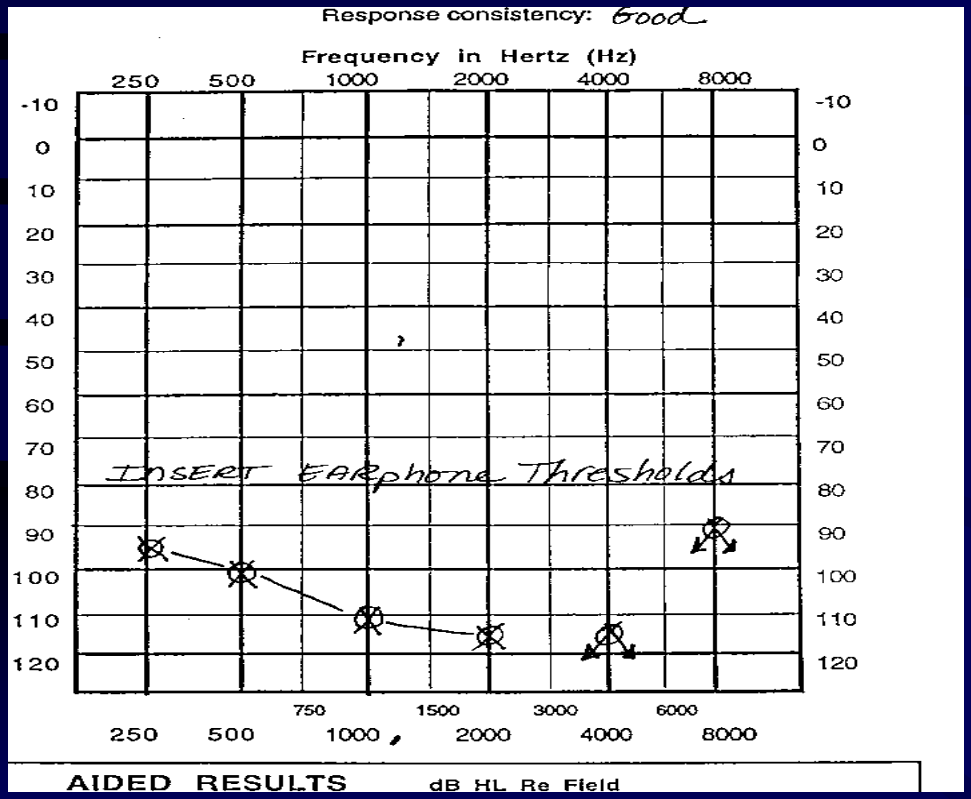
www.kregelab.com

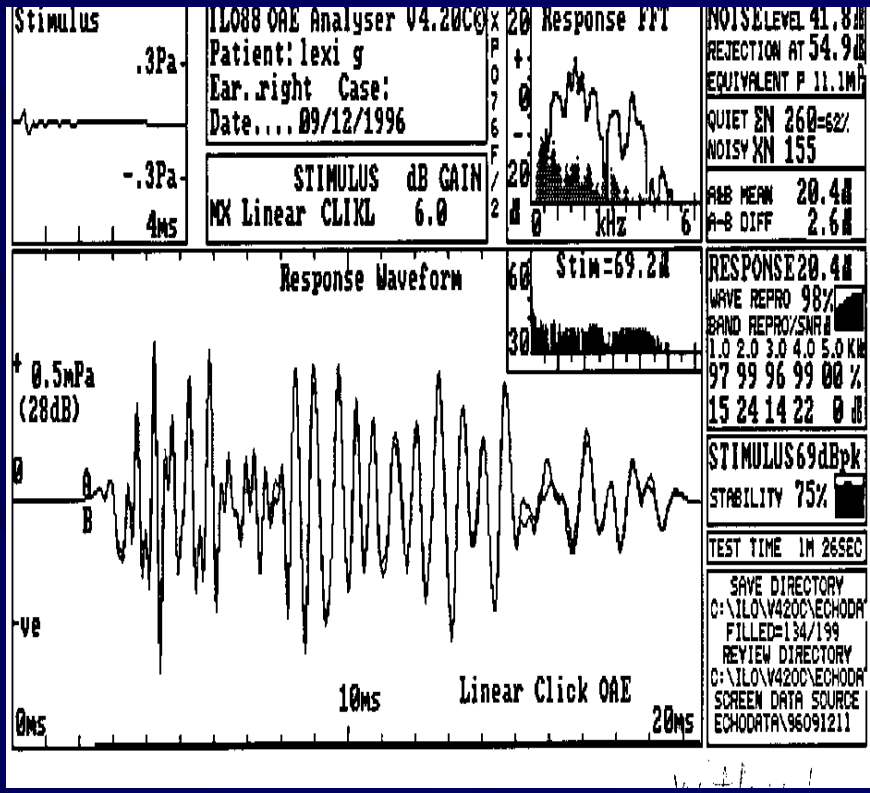
Www.auditoryneuropathy@yahoo.com

Time course for Auditory Neuropathy Patients...Type M

- Some maintain cochlear microphonics and otoacoustic emissions, but do not learn fluent speech without cochlear implantation. Labeled here Type M for “Maintain”. Four patients like this in our consultancy, all of whom were using Cued Speech at time of cochlear implantation, have already been implanted, three with reportedly successful results at Mayo Clinic, one in North Carolina.

Type M patient who maintains emissions but behaves Deaf



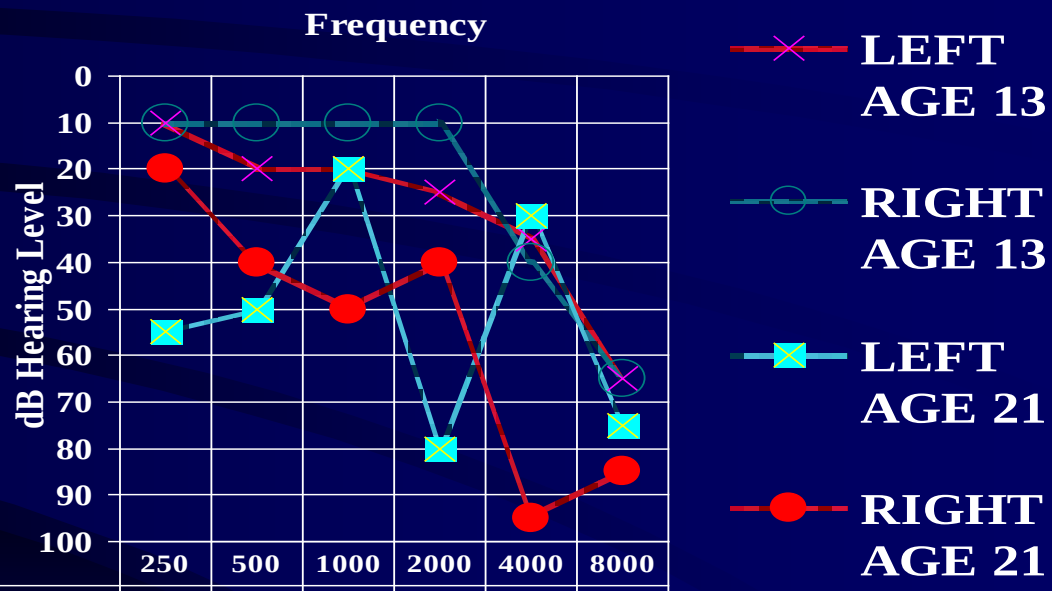


Time Course in Children...Type WPN

- Finally, some show a worsening of symptoms and develop other peripheral neuropathies. Called here Type WPN for “Worsening with Peripheral Neuropathy”.

Patient DV: No ABR, Normal Otoacoustic Emissions, No Middle Ear Muscle Reflexes. Audiogram shifts but emissions remain normal and ABR remains absent. Patient Developed Charcot-Marie Tooth Disease; sister has normal audiogram, abnormal ABR, no efferent suppression, but no symptoms as yet.

Type WPN.



	250	500	1000	2000	4000	8000
LEFT AGE 13	10	20	20	25	35	65
RIGHT AGE 13	10	10	10	10	40	65
LEFT AGE 21	55	50	20	80	30	75
RIGHT AGE 21	20	40	50	40	95	85

How to manage clinically and guide families through such an unpredictable course?

- Our goal is to create a “literate taxpayer”.
- Auditory Verbal therapy by itself or with hearing aids has not worked well for us with a few notable exceptions.
- Sign Language is adequate but is unrelated to English Phonology and often impedes literacy (Case FB for example) and is difficult for hearing parents to learn.
- Cued Speech simpler, allows you to cue in your own native language, and is an ideal place “in the middle of the chessboard” which offers maximal flexibility.

But...in the a recent screening visit to a Florida Oral School...

- We found an implanted child with normal emissions in her untouched ear. She was implanted at her mother's suggestion because hearing aids weren't working and she wanted a hearing and speaking child.
- The child's voice was virtually normal and her speech and language were exceptional.
- Is she using her implant or normal ear? In informal experiments mother confirms that without the implant she behaves "deaf".

So our final take home messages are...

- Screen all patients with emissions the first time (or go back and recheck the unoperated ears of your CI patients who didn't have emissions done). Note: Emissions sometimes disappear in ANSD.
- Monitor emissions and/or CMs regularly to watch for degeneration. One positive one negative polarity click.
- Use Cued Speech so that if the child's synchrony improves, they are language literate. If synchrony worsens, they are still language literate and prepared for self-support.

Acknowledgments

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