

Diagnostic Auditory Evaluations with Infants, Toddlers, and Older Children who have Developmental Disabilities

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Agenda



- Principles of Pediatric Audiology
- Gold Standards
- Clinical Guidelines
- Tools (Applications and Limitations)
- Stimuli (Effectiveness and Limitations)
- Special Considerations (re: Diagnostic Categories)
- Clinical Examples

Introduction



- 1) Everything we do in pediatric testing is an estimate – a very good estimate – but an estimate
- 2) You must work with what the child gives you
- 3) The mental age of the individual is more important than the chronological age
- 4) When you encounter dynamic changes in hearing sensitivity, go slowly and increase frequency of repeat visits

Introduction



Overarching Principle of Pediatric Audiology

Three factors must agree:

Parental report and observations

Clinical observations

Clinical test results

Disagreement by any one factor with the other two means something is missing – the search must continue until all three agree

Gold Standard ABR



Auditory Brainstem Response

Auditory brainstem response is the gold standard test for threshold estimation for infants and children who cannot complete behavioral audiologic assessment. ABR provides ear- and frequency-specific threshold estimates that are necessary for the diagnosis of the type, degree, and configuration of hearing loss and provision of amplification (Gorga et al., 2006).

Gold Standard Tympanometry



Tympanometry

Measures of middle ear function should be completed as part of the diagnostic audiology process for infants and young children. Either tympanometry or wideband reflectance can be used to characterize middle ear function (Hunter et al., 2013).

Gold Standard

Acoustic Reflexes



Acoustic Reflexes

Acoustic reflexes are an important test of middle ear function and the integrity of auditory brainstem pathways (de Lyra-Silva et al., 2015).

Gold Standards

Otoacoustic Emissions



Otoacoustic Reflexes

Otoacoustic emissions provide important information about the integrity of the outer hair cells of the cochlea and provide critical information about the differential diagnosis of auditory neuropathy spectrum disorder and sensorineural hearing loss (Gorga et al., 2000).

Gold Standard Behavioral Assessment



Behavioral Assessment

Behavioral assessment of hearing is the gold standard for estimation of hearing thresholds. Visual reinforcement audiometry (VRA; for infants 6–24 months; Widen et al., 2005) and condition play audiometry (CPA; for toddlers 24+ months; Norrix, 2015) are established methods based on conditioned responses to sound.

Developed based on typically developing children:

- Focus on the chronological age of the child
- Ear specific data, especially for hearing aids

With Developmental Disorder Children:

- You may encounter situations involving ethical dilemmas
- There is great dependence on both intra-test and inter-test cross-checks combined with replication/validation

Tools and Limitations

OAE

ABR

Cortical Evoked Potentials

BOA

VRA

Conditioned Play

Standard

Stimuli

Warble tone

Narrow band noise

Pediatric noise

White noise

Pulsed tones

Tone bursts

Clicks

Voice

Music

Mother's voice

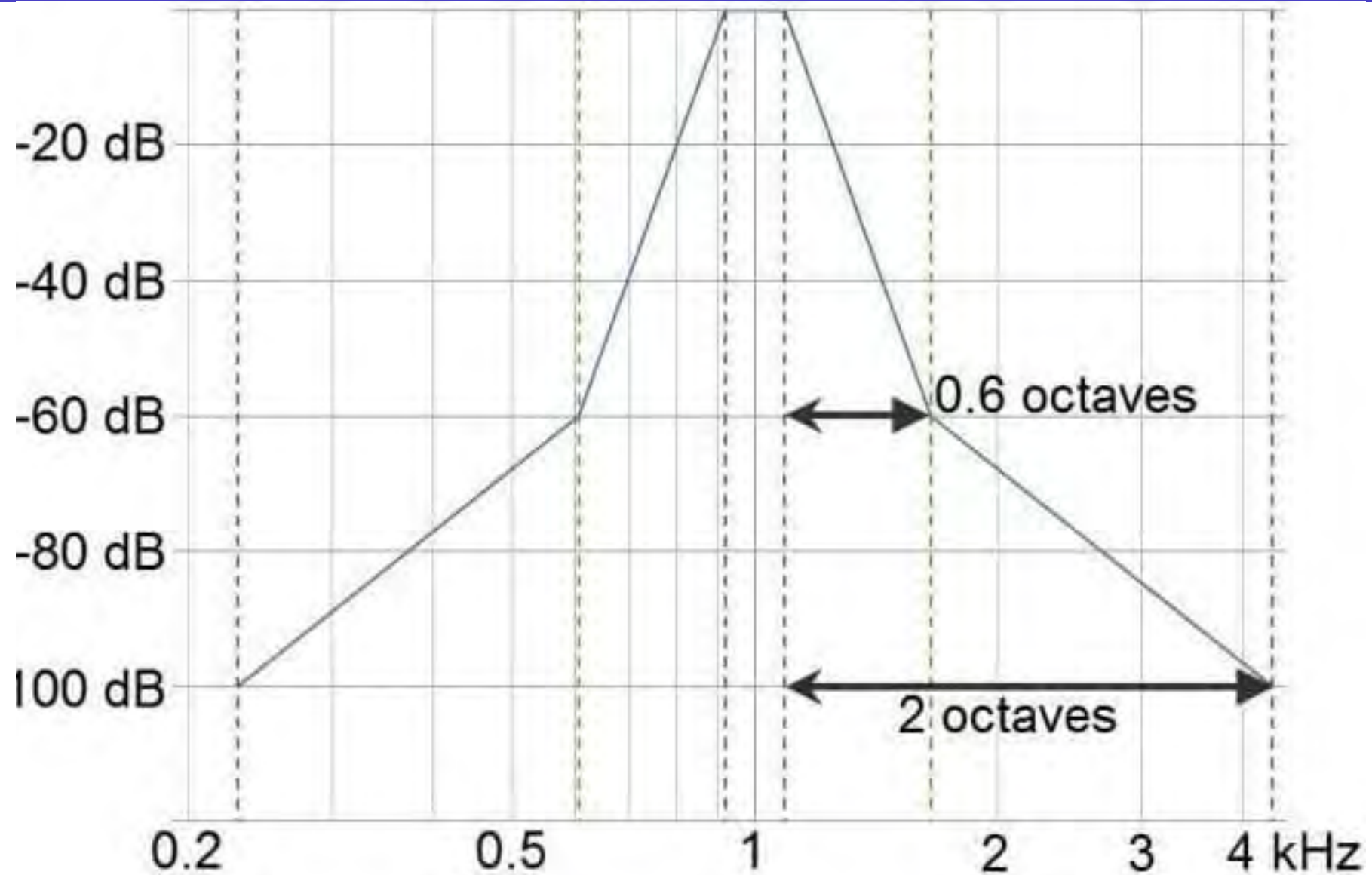
Non-tonal stimuli:

(e.g., frequency analyzed toys or
frequency filtered toys)

Narrow Band Noise

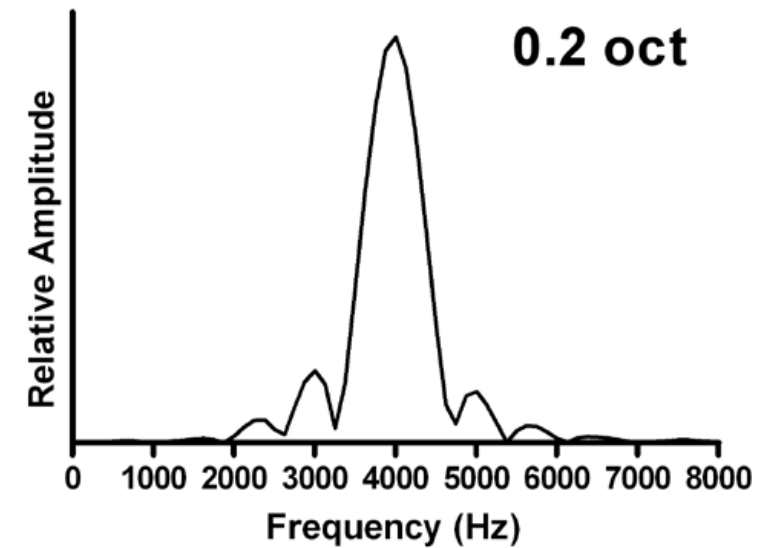
Center Frequency	Lower Frequency	Upper Frequency
12.5	11.2	14.1
16	14.1	17.8
20	17.8	22.4
25	22.4	28.2
31.5	28.2	35.5
40	35.5	44.7
50	44.7	56.2
63	56.2	70.8
80	70.8	89.1
100	89.1	112
125	112	141
160	141	178
200	178	224
250	224	282
315	282	355
400	355	447
500	447	562
630	562	708
800	708	891
1000	891	1122
1250	1122	1413
1600	1413	1778
2000	1778	2239
2500	2239	2818
3150	2818	3548
4000	3548	4467
5000	4467	5623
6300	5623	7079
8000	7079	8913
10000	8913	11220
12500	11220	14130
16000	14130	17780
20000	17780	22390

Pediatric Noise

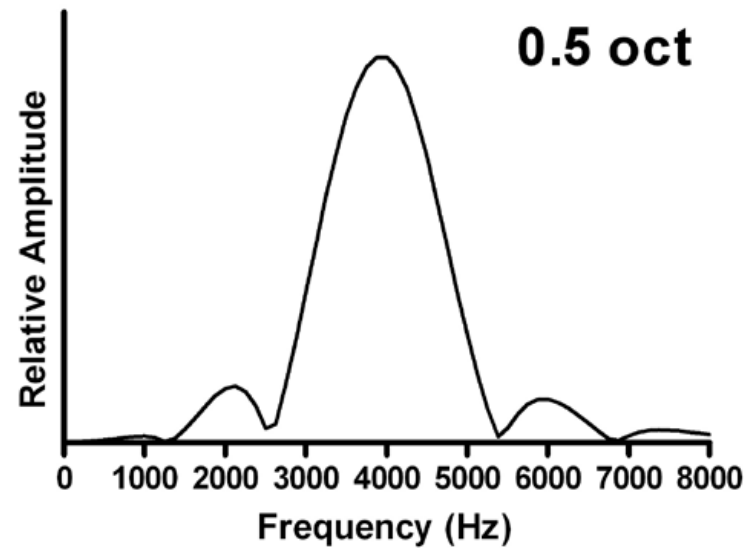


Tone Bursts

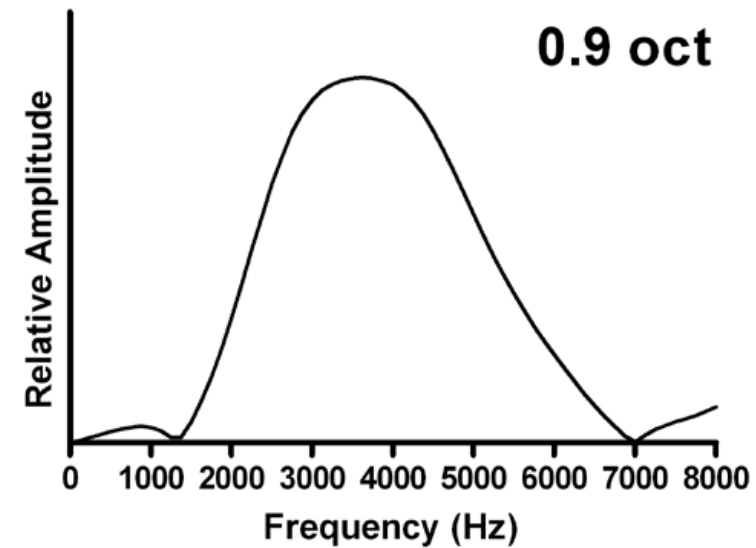
A



B



C



Stimulus effectiveness influenced by:

- Onset time of stimulus
- Bandwidth of stimulus
 - Warble tone
 - NBN
 - Frequency analyzed toys
 - Music
- Listener Experience

Goal

Establish estimate of auditory sensitivity
through:

Minimum response levels

VRA

ABR

Stimulus

Role of BOA

Beware of bias from increased bandwidth

- Underestimation of sloping hearing loss thresholds
- Tone burst ABR underestimation of thresholds for sloping SNHL;
 - Tone burst ABR may exaggerate HL levels for low frequencies

Special Considerations: Autism Spectrum Disorder

Position on the spectrum

- Information processing

- Do NOT use cold running speech

- /bup/bup/bup/bup/bup/bup/bup/

- Sensory defensiveness

- “One and done” response pattern

- Do NOT use standard conditioning paradigm

- Familiarity of physical setting

 - Resistant behavior

 - Crying behavior

 - Use of novel stimulus

Special Considerations: Auditory Neuropathy

Single or dual disorder

By definition: Neural transmission issue

Various classification of ANSD

May have concomitant cochlear hearing loss or neural transmission only

Classic diagnosis: abnormal ABR with cochlear microphonic

Abnormal ABR, significant behavioral hearing loss and present OAEs

Other diagnostic indicators:

ABR suggesting severe to profound HL with behavioral responses 30 dB to 80 dB better

Fluctuating SNHL with disproportionately poor discrimination and present OAEs

Dynamic fluctuations and improvement of hearing sensitivity: do not be overly aggressive regarding CI referral

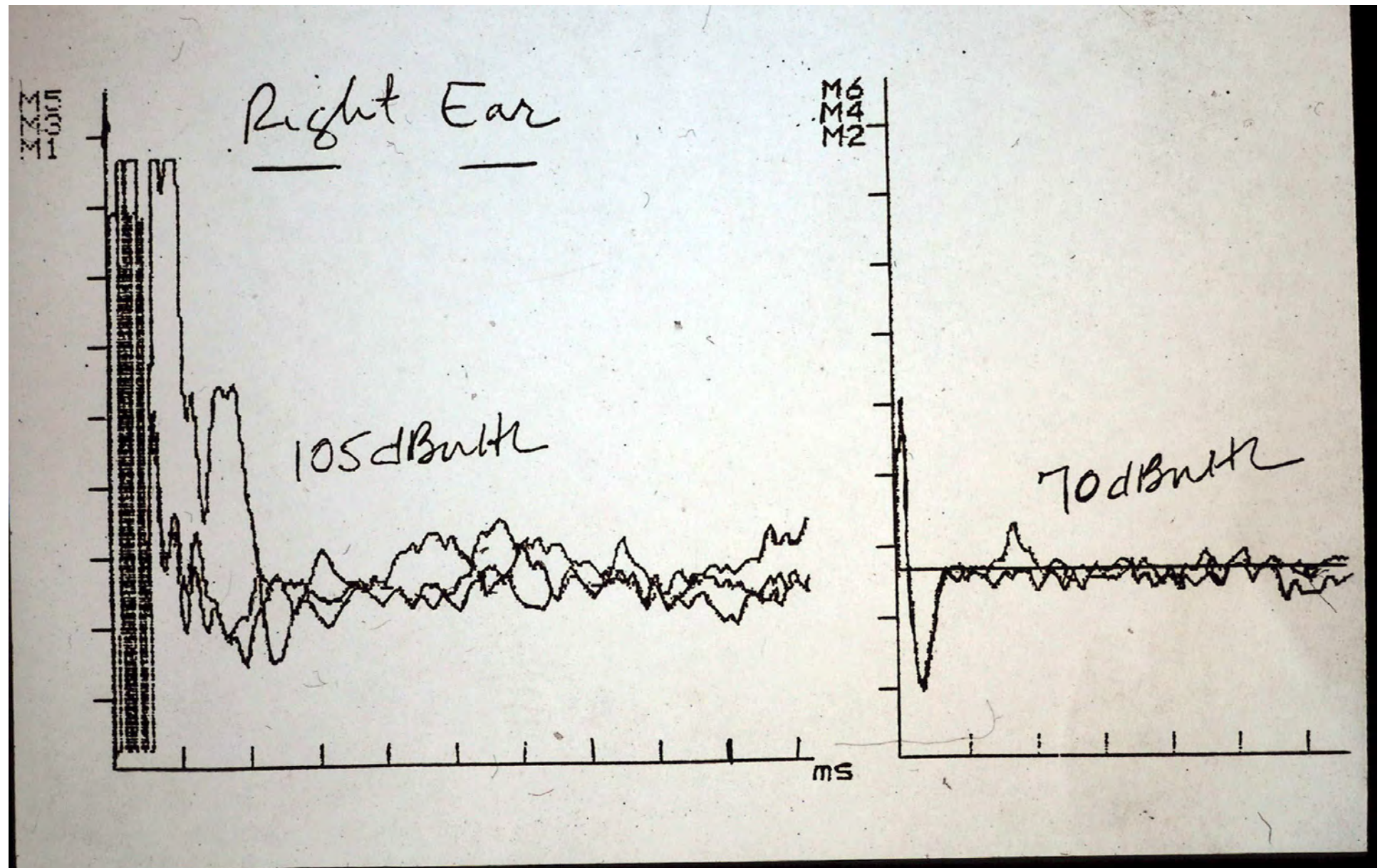
Special Considerations: Combined Diagnoses – Autism and SNHL

- Hearing loss diagnosed before 6 months via ABR, OAE, VRA, BOA
- Do not respond to auditory amplification with development of listening and alerting behaviors, sporadic responses to auditory stimuli
- Do not increase vocalizations and vocal play
- Confirmed appearance of ASD typically around 18-24 months
- Intervention must go beyond AVT and auditory habilitation – must include autism focused interventions
- Monitor closely to ensure validity of estimated minimum response levels and absence of hearing loss progression

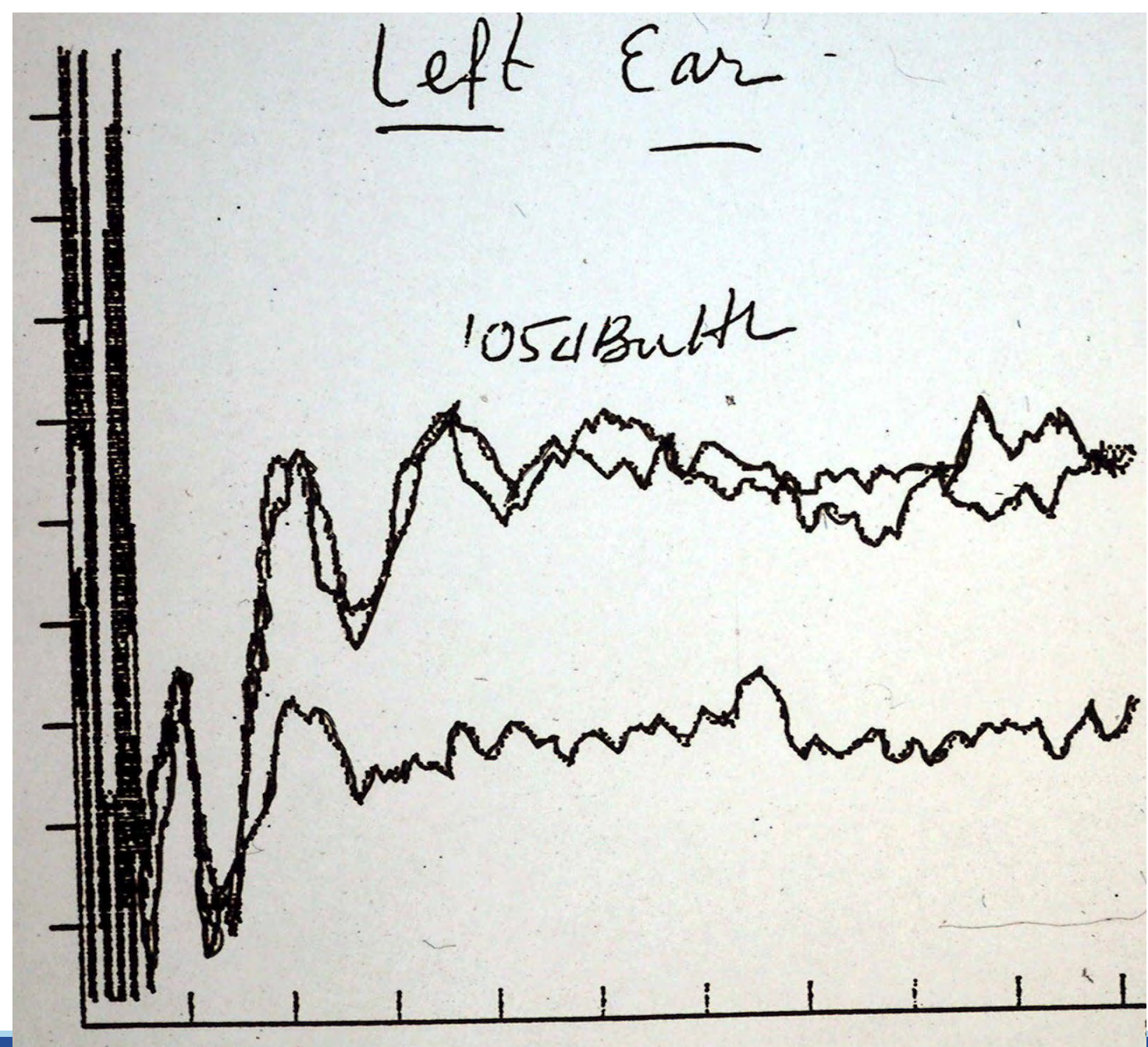
Clinical Example: FG

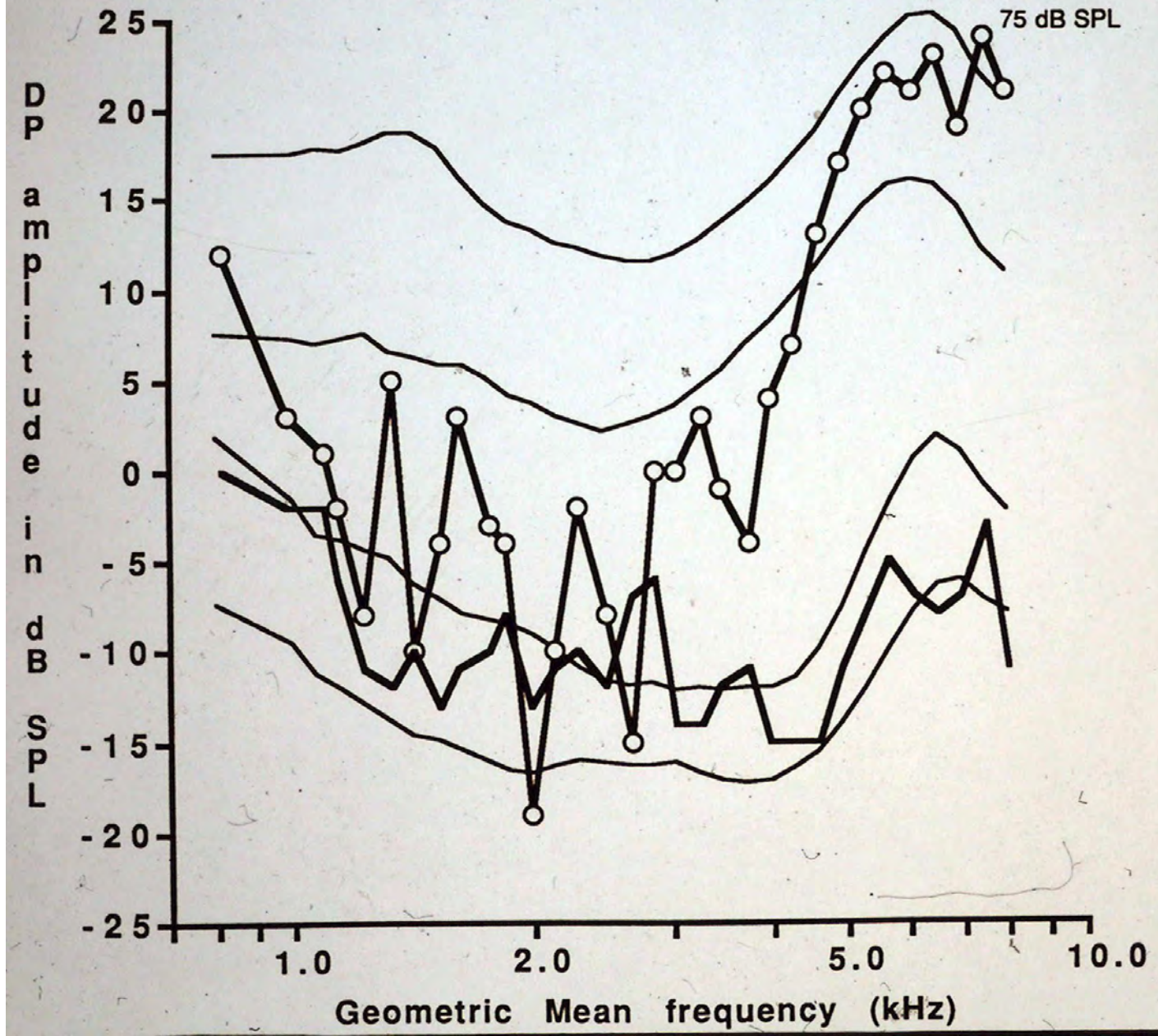
- Born early 1990s at 26 weeks GA
 - Hyperbilirubinemia
 - Hypoxia
- Flat line ABR with microphonic
- Father asked: “Does hearing loss ever get better?”
- Audiograms progressively better
- Final audiogram: high frequency SNHL
- Residual diagnosis of ASD

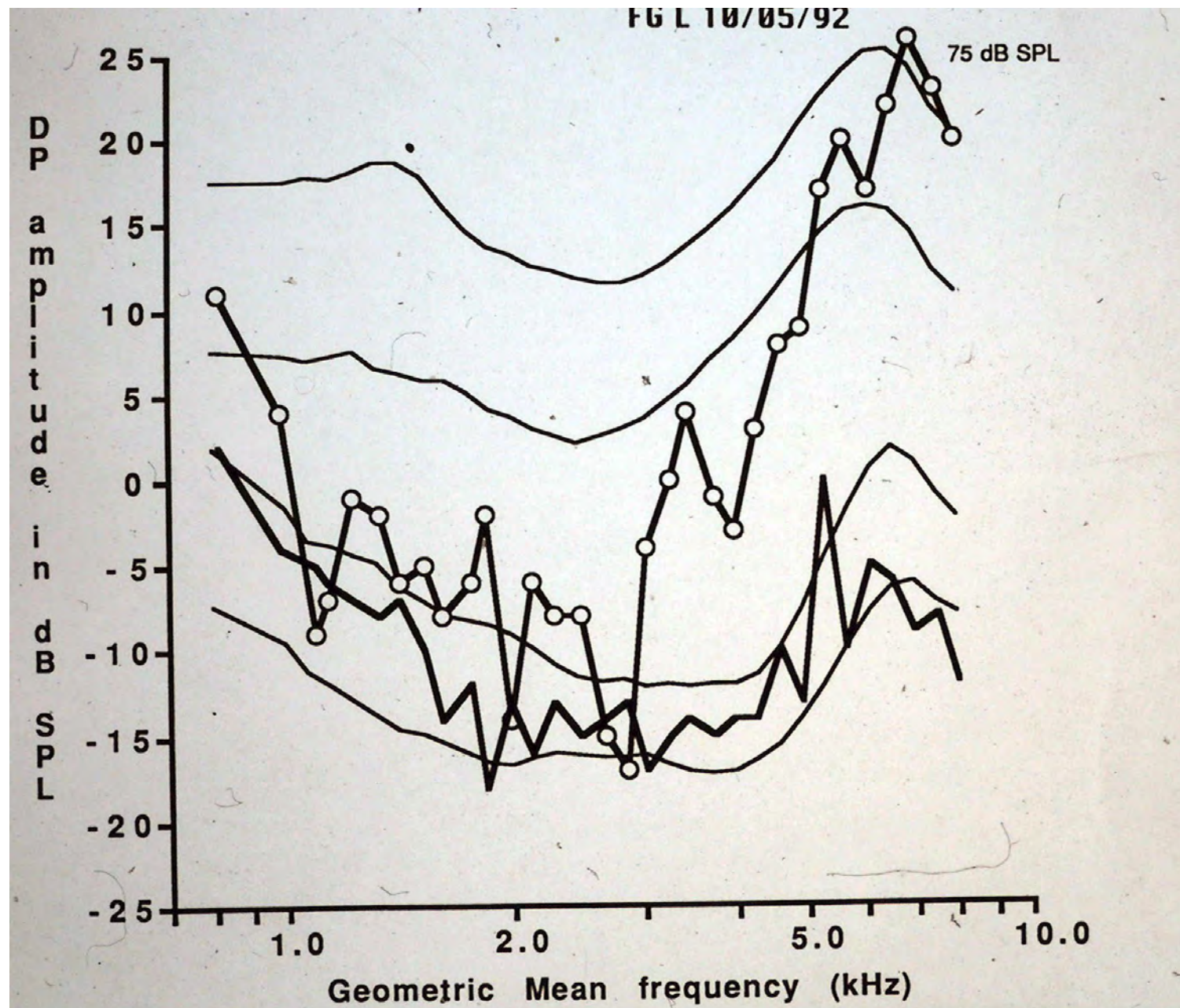
Patient: FG

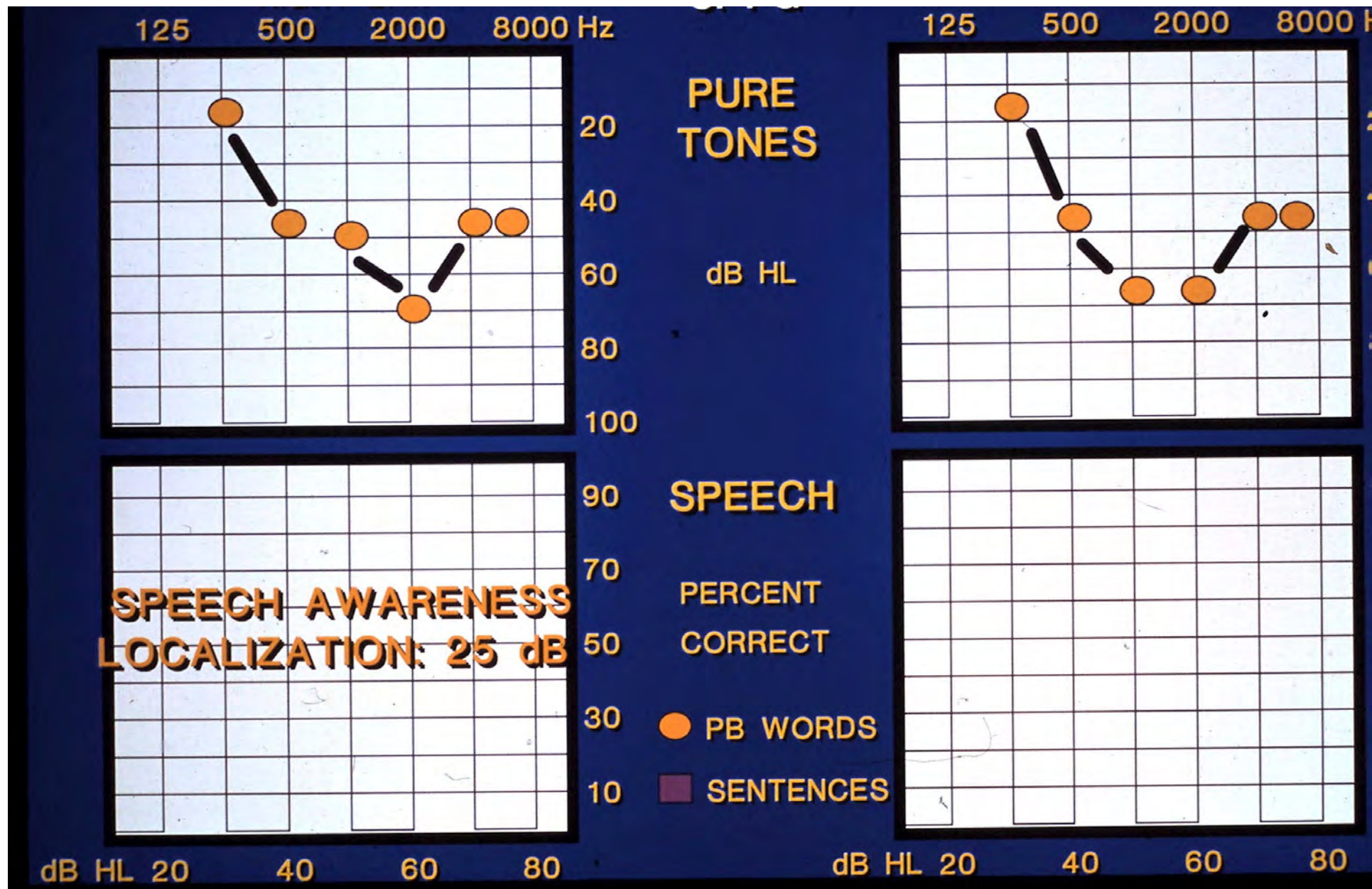


Patient: FG









Clinical Example: CS

In vitro fertilization

Emergency C-section 2° pre-eclampsia

5 weeks in NICU – mild jaundice

Dx of auditory neuropathy from another facility

Frequent falls as a toddler – more than expected

Abnormal ABR: +cochlear microphonic, no neural response

No response to sound

*Programmed hearing aids based on best estimate

Clinical Example: CS (cont'd)

Progressive improvement in audiogram requiring HA adjustments

At age 3 years, Tx SLP/ OT/ PT over prior year

Increased vocabulary and use of spoke language

Audio: conditioned play – 35 dB at 500 Hz and 4000 Hz

SAT: 35 dB

Less frequency of falling

Clinical Example: CS (cont'd)

Age 3 years 9 months

Still receiving ST, OT, PT

Pulling out hearing aids and “tossing” them

CPA: Right – 25 dB @ 1000 Hz; 40 dB @ 6000 Hz

Left – 25 dB @ 500 Hz; 40 dB @ 4000 Hz

Putting 2-3 words together in meaningful sentences

Interacting and responding well w/o hearing aids

Discontinued hearing aid use

Clinical Example: CS (cont'd)

Age 5 years 4 months

Autism symptoms much more apparent

Still receiving ST, OT, PT

Falling issue resolved

Audiogram

EAR	500 Hz	1000 Hz	2000 Hz	4000 Hz	6000 Hz
Right	20	15	10	35	35
Left	15	15	10	35	40

Secondary diagnosis of ASD became primary diagnosis

Clinical Example: CS (cont'd)

Age 7 years 6 months

Autism symptoms clearly apparent

Still receiving ST, OT, PT

Audiogram

EAR	500 Hz	1000 Hz	2000 Hz	4000 Hz	6000 Hz
Right	10	0	0	10	10
Left	10	0	0	10	15

Doing well in school with supports. Using carryover IEP.

Clinical Example: CS (cont'd)

Age 12 years 6 months

Autism related behaviors self-managed

No longer needs classroom supports

Excellent grades => Honor Roll

ST, OT, PT discontinued

Failed school hearing screening

Audiogram

EAR	500 Hz	1000 Hz	2000 Hz	4000 Hz	6000 Hz
Right	10	25	40	45	40
Left	10	20	35	40	40

Hearing aids programmed and dispensed October 2022

Clinical Example: NS 10/18/21

- Born a twin at 38 weeks GA. Displayed hypotonia leading to OT and PT therapy from post-natal to present day.
- We first saw him at 14 months of age
- He had a previous hearing test that was inconclusive.

Clinical Example: NS 10/18-25/2021

- ABR on 10/18 indicated click threshold at 70 dB, 60 dB at 500 Hz, and 90 dB at 2000 Hz for left ear.
- ABR on 10/25 had tone burst threshold of 70 dB at 2000 Hz, then he woke up.
- Behavioral testing same date:

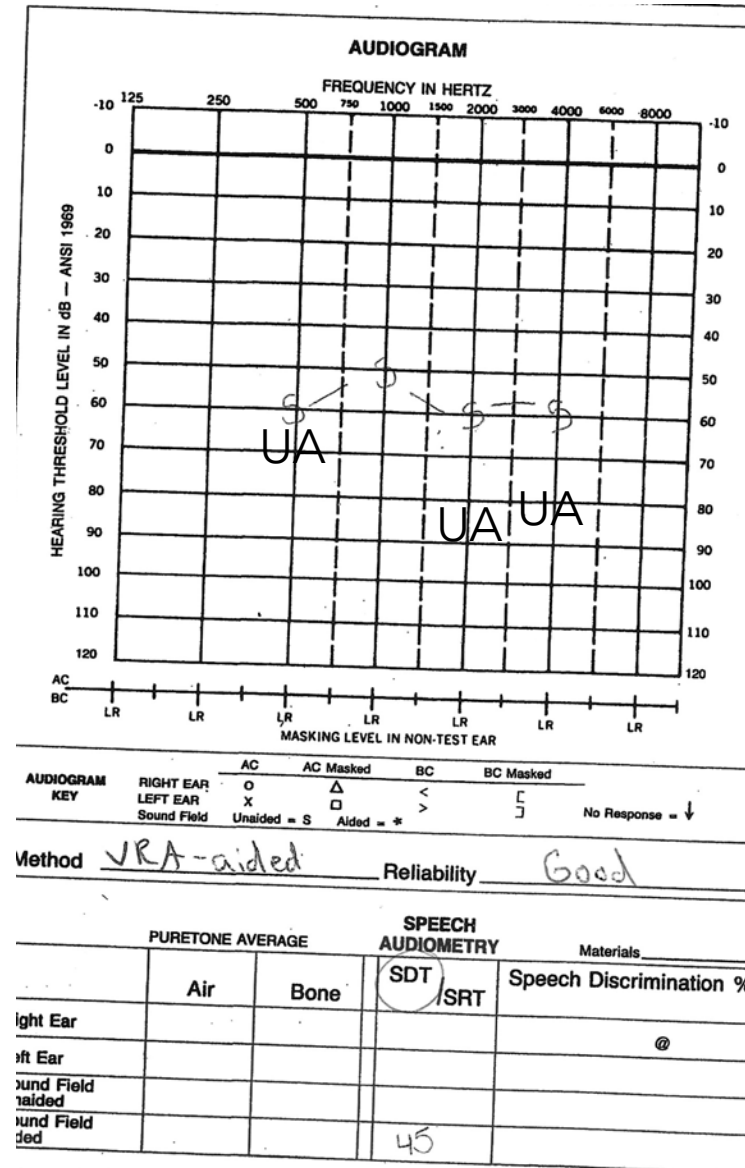
	500 Hz	1000 Hz	2000 Hz	4000 Hz
Sounds Field	70 dB	DNT	85 dB	80 dB

- Following this outcome, hearing aids were ordered and dispensed based on his response levels.

Clinical Example: NS 1/24/2022

- He accepted the hearing aids after the first two weeks and did not resist.
- He did not show the typical response, even with a possible diagnosis of ANSD of increased vocalization/self-stimulation or change in listening behaviors. Mother reported slight differences but no truly significant change in auditory behaviors.
- In late March, he started swiping the hearing aids off and resisting having them on his ears.

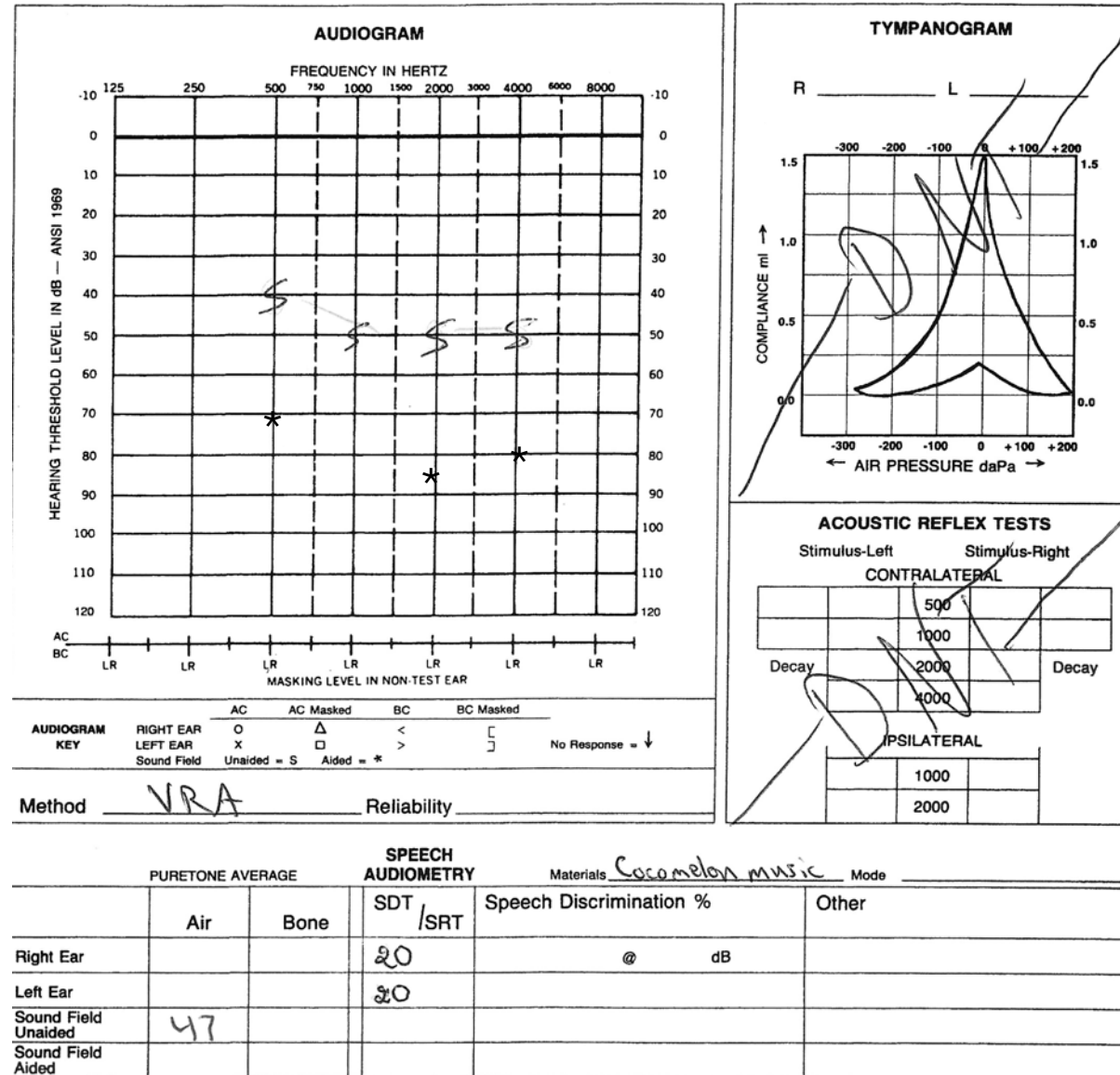
Clinical Example: NS 1/24/2022



These are aided responses using VRA in sound field

Clinical Example: NS 4/27/2022

4/27/2022



Excellent localization.
Response to music
(Coco-melon) had ear
specific responses
(started with insert
earphones and after
the music, he pulled
them out and fought
putting them back in.
Remainder of
assessment was in
sound field.
A month before this
visit, he started
resisting his hearing
aids.

ABA therapy started.

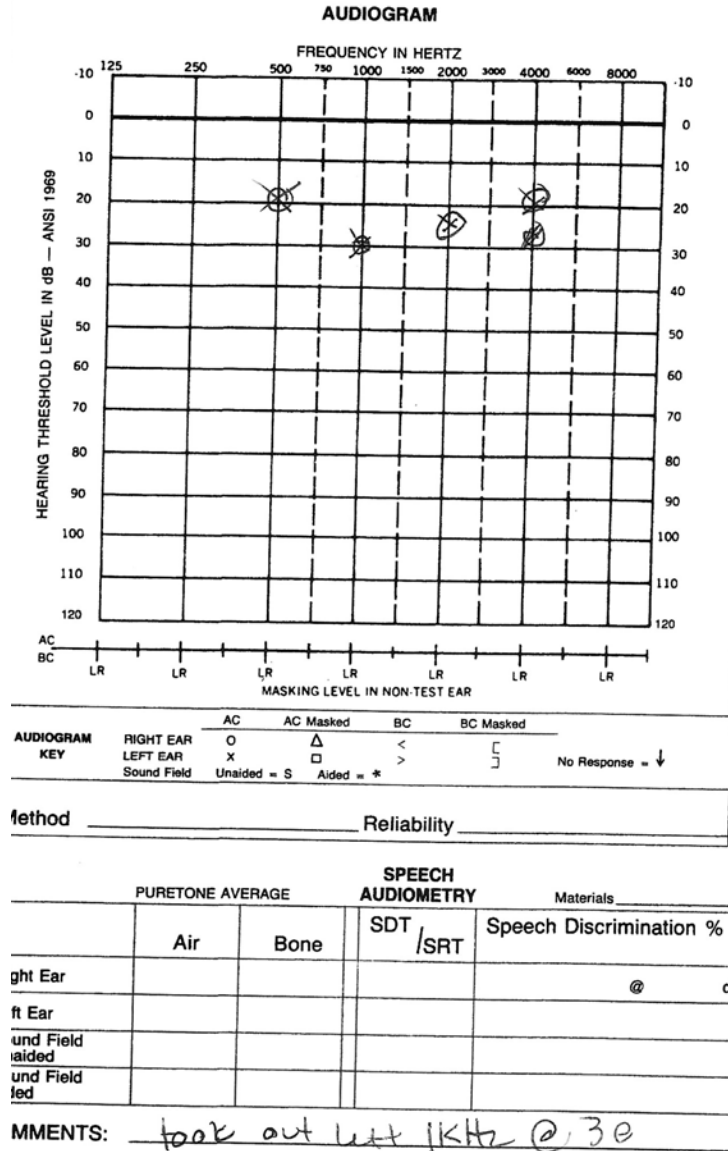
Clinical Example: NS 4/27/2022

- We were able to also performed ABR testing for the right ear on this date:

Stimulus	Intensities Tested (dB nHL)	Wave V Threshold (dB nHL)
2000 Hz	90, 80, 70, 0	NR
1000 Hz	80	NR
500 Hz	70, 60, 50, 40	60

Then the child woke up.

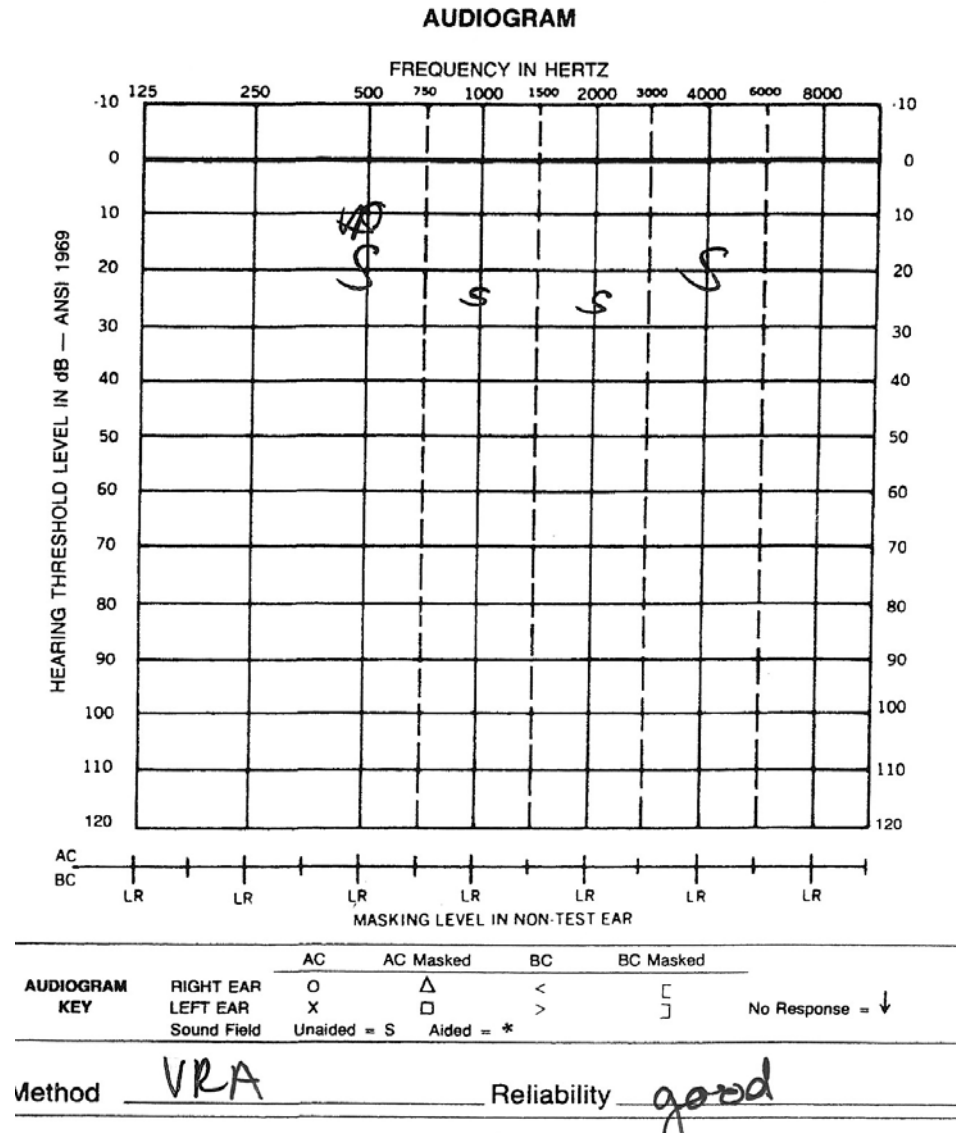
Clinical Example: NS 8/8/2022



Prior to this visit, he had a sedated ABR done at another facility which showed no response to a click at 105 dB nHL

Since March 2022, more symptoms of autism spectrum disorder were appearing. E.g., off to a "different world".
Alerting only to unexpected sounds.
Sporadic auditory attention. Little eye contact.

Clinical Example: NS 10/4/2022



SDT sound field: 20 dB
with excellent
localization.

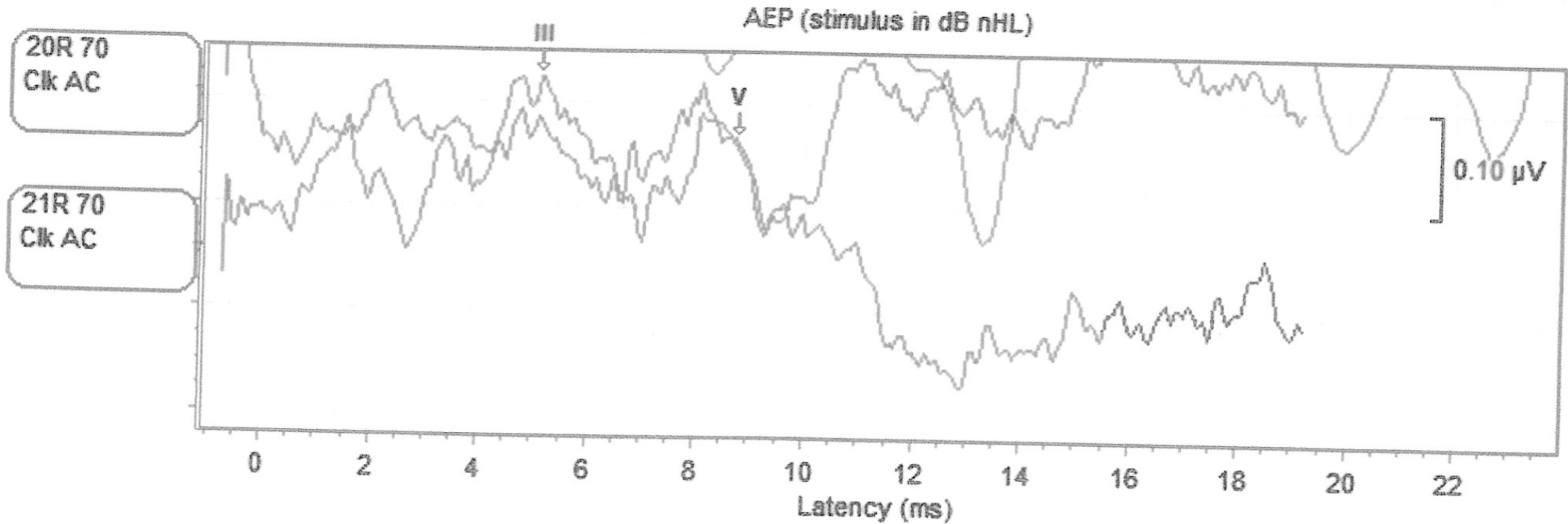
Evaluated by a
diagnostic team with a
final diagnosis of Level
4 Autism Spectrum
Disorder. Ended
auditory verbal therapy
and began search for
autism focused therapy.

Clinical Example: NS 1/9/2023

Behavioral Audiometry: Hearing sensitivity was assessed via visual reinforcement audiometry using insert earphones. Stimuli consisted of pulsed pure tones and pulsed narrow band noise, and children's songs presented through the audiometer speech circuit. Responses were judged by time-locked changes in head movement (e.g., searching for the sound) to the right or the left in response to a stimulus. Minimum response levels were within normal limits at 500 Hz, 1000 Hz and 4000 Hz before Nico fatigued to testing. A minimum response level to speech was obtained at 15 dB HL.

	500 Hz	1000 Hz	4000 Hz
LEFT EAR	25 dB HL	25 dB HL	20 dB HL
RIGHT EAR	25 dB HL	25 dB HL	25 dB HL

Clinical Example: NS 5/23/2023



ABR is still abnormal with elevated thresholds and degraded morphology. But this response illustrated greater synchrony along the auditory pathway than was shown in previous ABR results.

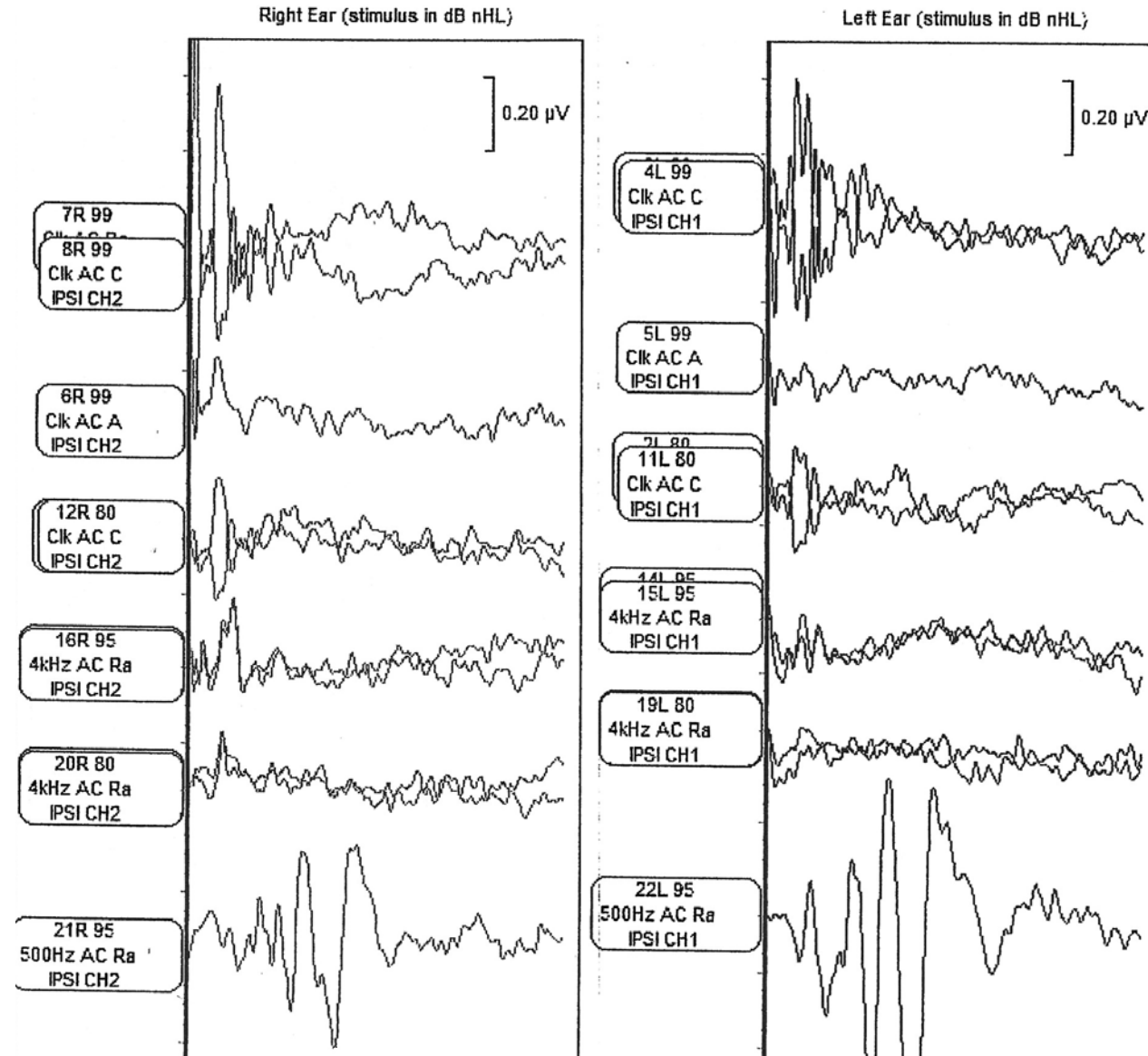
Discussion: NS, CS, FG

- Three children with early signs of auditory neuropathy who demonstrated improved hearing sensitivity over the first two years of life. Each child's final audiogram showed thresholds that were either normal or, in the case of FG, significantly improved compared to first contact.
- The literature mentions this phenomenon briefly but without discussion other than there is no explanation at this time underlying these auditory improvements.

Clinical Example: TW

- Born at 26 weeks GA due to premature rupture of membranes
- NICU admission for 3 months
 - No history of IVH or seizures
 - Dx of pulmonary hypoplasia – d/c with oxygen supplement
 - General milestones delayed
 - No speech development – enjoys loud music
 - ABR at 9 months (~6 months corrected)
 - Severe to profound SNHL

Clinical Example: TW



Waveforms "flip"
Some OAEs
Dx: auditory neuropathy

Clinical Example: TW

- Referred for CI evaluation at 23 months of age; also referred to Mailman Center for audiological evaluation
- Parents concerned about CI. Discussed initial trial with hearing aids
- Parents considered options and decided to go with hearing aids first and CI following HA trial with therapy
- Child rejected insert earphones; sound field testing completed for 3 frequencies only in severe to profound range - good localization
- Earmolds made at 23 months of age; hearing aids dispense 3 weeks later

Clinical Example: TW

- After one month of use, she chewed the hearing aid case and bit off a piece of her earmold
- We continued unaided and aided estimates of minimum response levels every few weeks with varying results depending on whether she attended to the sounds.
- Unaided continued in sound field; she would flail her arms, kick, and remove the insert earphones if we had success in getting them in her ear canals.
- On many trials, she would not attend to sound despite being above her MRL estimates

Clinical Example: TW

- She demonstrated PICA, an eating disorder whereby inappropriate objects are swallowed.
- Over 18 months, we replaced 17 earmolds and 10 hearing aids
- Referred for ASD evaluation; did not meet criteria for ASD but behavior therapy was recommended.
- Behavior therapy begun at 37 months of age and continued for 9 months.
- Audiometric testing through sound field continued due to strong resistance to earphones; 1-4 frequencies tested for each ear based on responsive behavior; responses varied from 50 dB through 80 dB

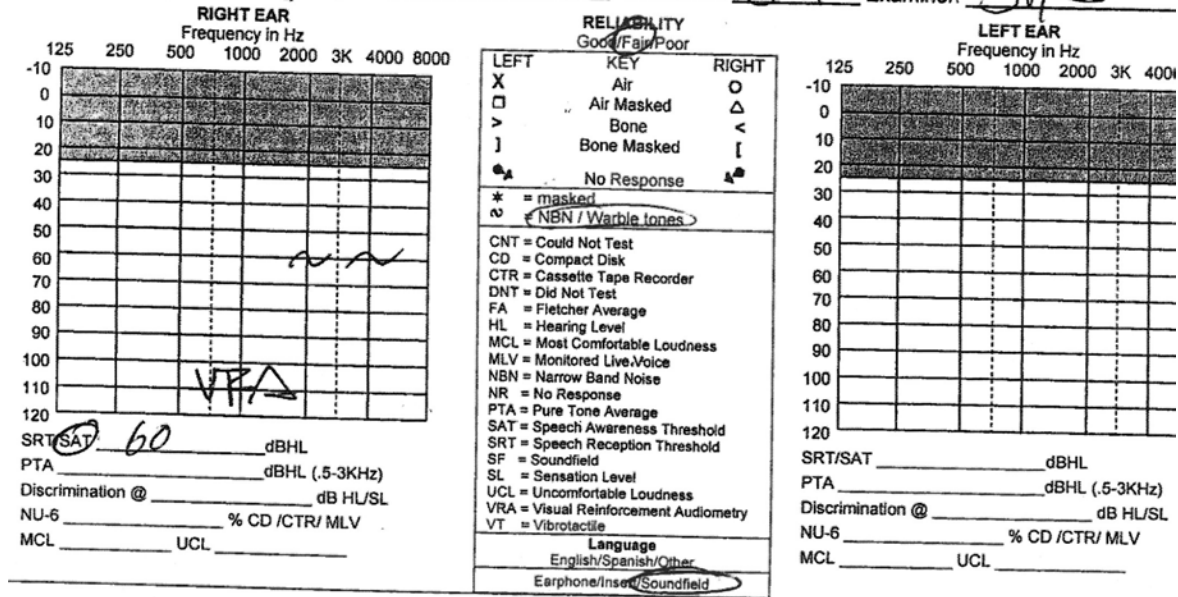
Clinical Example: TW

- At 46 months of age, she was calm enough to allow insert earphones and an ear specific audiogram. From her behavior therapy, she was using hearing aids more and beginning to develop spoken vocabulary in addition to sign.
- At that time, mother decided that she desired a referral to ENT for a CI evaluation. Arrangements were made for bilateral Cis.
- Final diagnosis: auditory neuropathy
Oppositional defiant behavior disorder
PICA
Severe to profound SNHL (based on behavioral)

Clinical Example: VI

- Born at 23 weeks G.A.
- NICU stay for ~7 months
- Failed newborn hearing screen but did not receive a follow-up diagnostic evaluation
- NICU Tx: transfusions, grade III IVH, PPHN
- Initial audio at 22 months of age
- Two SF frequencies plus SAT at ~60 dB

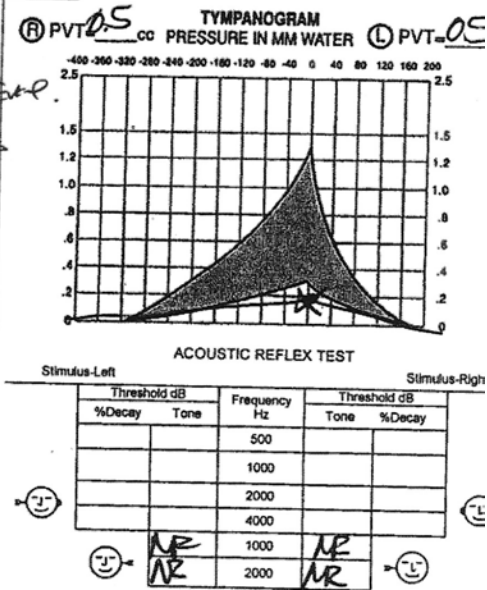
11/4/2013



INTERPRETATION

5x 34/Week, PT/OT
Did not pass the MBHS, AU, did not receive follow up GEP.
Delivered @ 23 weeks GA. NICU stay 6-7 months

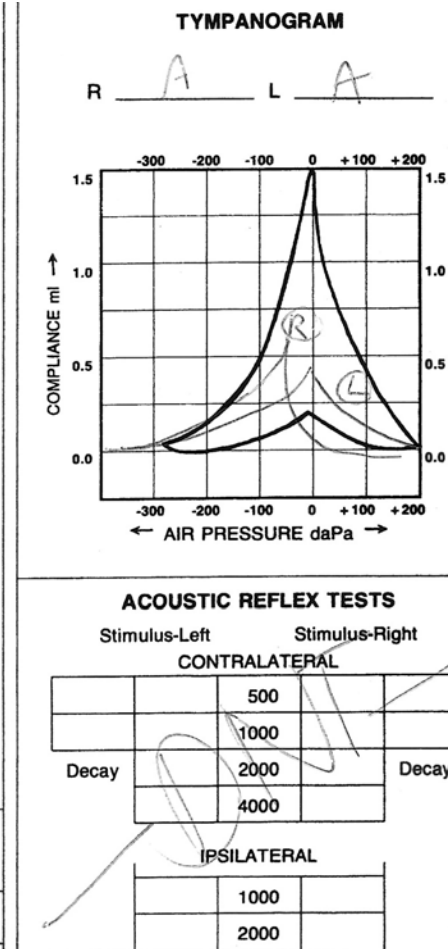
Plan
- ~~ABZ~~ GME or ZTester Audio



Clinical Example: VI

- Did not return for follow-up for one year
- Child obtained hearing aids from another practice in the Miami area
- Mother commented that she did not see much improvement with Has
- Data logging indicated only 15 minutes of use per day over the previous 240 days
- Mother reported that she only wore them during therapy sessions

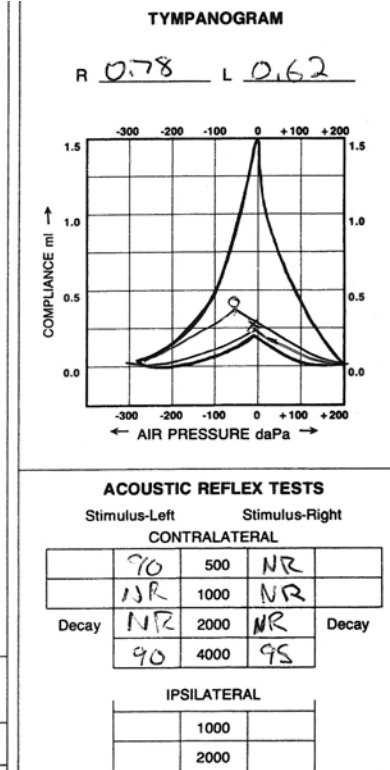
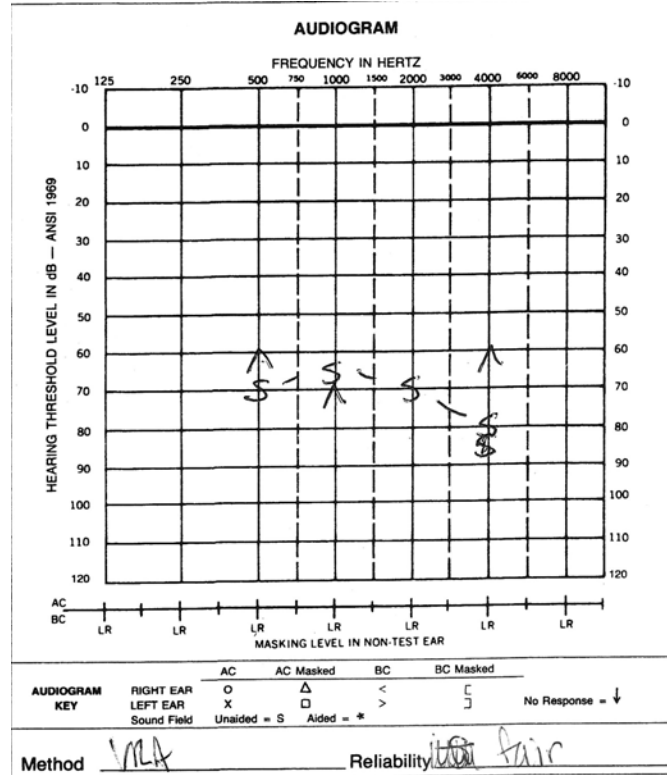
11/17/14



Would not respond to tonal stimuli with insert earphones but did respond to NBN.

PURETONE AVERAGE			SPEECH AUDIOMETRY		Materials	Mode
	Air	Bone	SDT / SRT	Speech Discrimination %		
Right Ear			CNT	insec @		
Left Ear			CNT	insec		

12/11/2014



PURETONE AVERAGE		SPEECH AUDIOMETRY		Materials	Mode
Air	Bone	SDT	SRT	Speech Discrimination %	Other
Right Ear		60		@ dB	
Left Ear		65			
Sound Field Unaided		55			
Sound Field Aided					

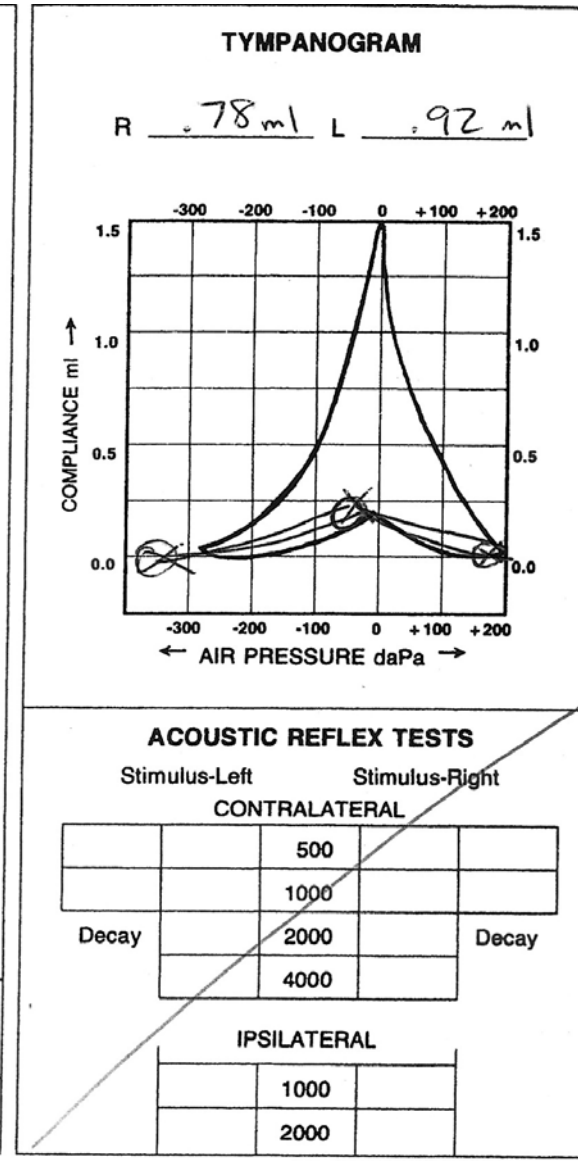
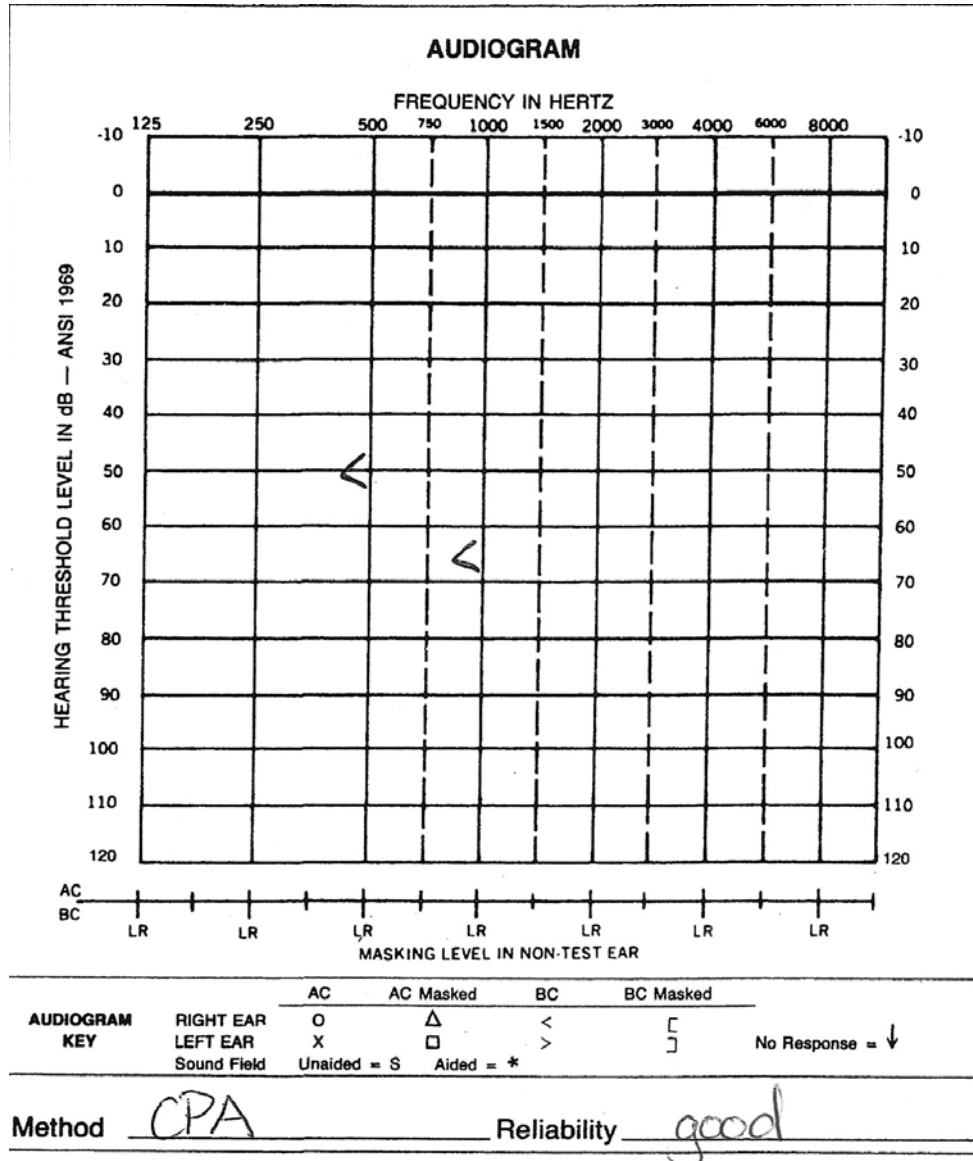
COMMENTS: Otoscopy: left had minimal wax unable to get a full visual of the ear drum. The right ear canal minimal wax ear drum visible

Clinical Example: VI

- Following the December 2014 evaluation, I noted that the child was responding to her mother's voice without hearing aids.
- She was also producing phonemes in vocal play without hearing aids.
- Mother said that both observations were common which is why she did not enforce hearing aid use at home.
- This led to the possibility of auditory neuropathy whereby responses can be sporadic including unaided behavioral responses

Clinical Example: VI

1/15/2016

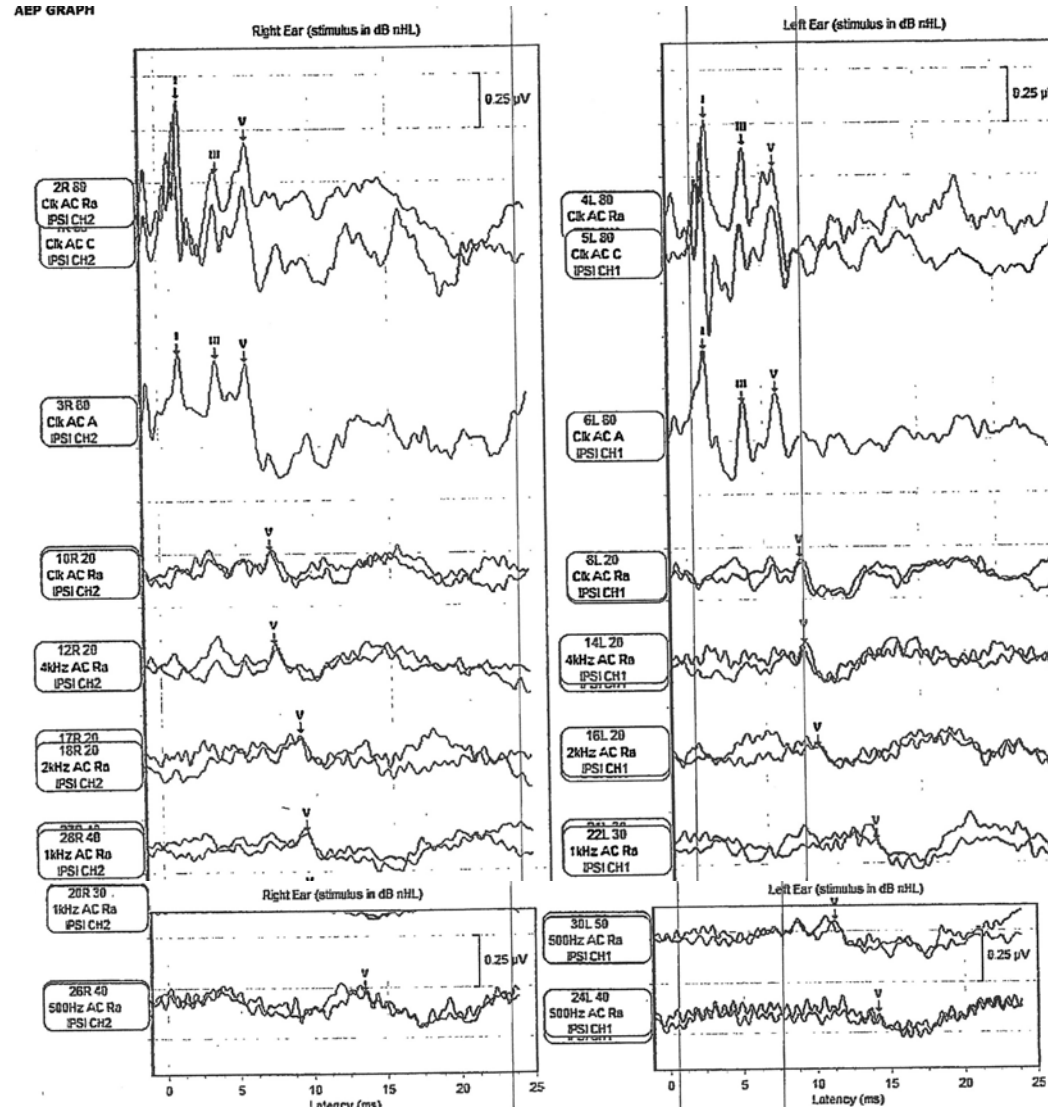


Clinical Example: VI

- Following the January 2016 appointment, ENT once again took over her care.
- Their initial evaluation could not go beyond sound field measurements.
- When they eventually did obtain ear-specific thresholds using pulsed pure tones, it was discovered that the NBN that we had to use slightly underestimated the degree of hearing loss because of the threshold slope across frequencies.
- The child was evaluated for and received cochlear implants.
- The question of possible ANSD was never answered.

Clinical Examples: AP 3/17/22

Normal sound field audiogram when he is alerted to pay attention. Audiogram has normal hearing sensitivity at all frequencies.



Referred from another facility for hearing aids with a diagnosis of auditory neuropathy.

Child will not respond to speech but will respond readily to music and sporadically to environmental sounds.

Hx of chronic, long-lasting seizures.

Final diagnosis: Cortical deafness



PET scans by Lawrence Parsons, Peter Fox, and Donald Hodges show how the brain responds to music. Some of these activations are shown here in color, overlaid onto horizontal magnetic resonance images of subject's brains.

Left panel: *the harmony condition activated the left side of the brain more than the right. It also activated inferior (or lower) regions of the temporal cortex as compared to the melody condition.*

Center panel: *the melody condition activated both sides of the area called the temporal cortex (which is known to represent sound) to a much greater extent than did the rhythm and harmony conditions.*

Right panel: *much of the brain activation observed during the rhythm condition was in the cerebellum.*

Clinical Examples: DO

- 32 year old with Down's Syndrome
- Followed for more than 18 years due to chronic otitis media
- Has tactile defensiveness that will not allow supra-aural or insert earphones – nothing that enters the ear canal.
- Will allow tympanometry
- He would permit sound field testing and tympanometry for each visit and bone conduction on some visits.

**Well, the day is almost
done, and it is a time for
rest...**



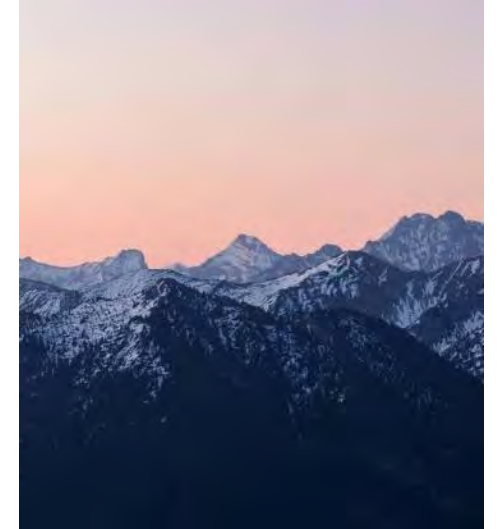
As we begin to wind down...

- Children with developmental delays and disabilities to follow the “typical” trajectory of responsiveness compared to “typically” developing children.
- There are days when you have obtained one or two minimum response levels, you declare a victory for that day!
- In many ways, pediatric audiology is like solving a “Who Done It” mystery. One must put the clues together in order to determine the final audiometric status and auditory diagnosis.
- BE AWARE! When you inherit the child, you also inherit the family. Your responsibility is much broader than with adults. Your job is to educate the family to a level where they become full decision-making partners for the sake of their child.
- REMEMBER: Professionals come and go from that child’s life, but the family is the only constant that child has for many years..

Summary

This presentation gave examples that may seem extreme, but they are not that uncommon. Always be alert that something like this may exist, especially for children of very early births and very low birth weights.

Patience and perseverance are the keys to success. And don't be in too much of a rush to implant. If the child shows stable or worsening thresholds and behaviors over a period of several months, make the CI referral. But if there are signs of improvement – behavior first – go slowly and don't be too aggressive.

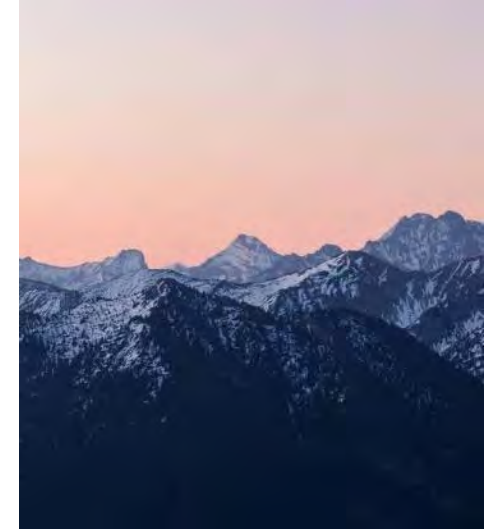


Remember:

In solving the mystery, three things must agree:

1. Parental history and observations
2. Clinical observations
3. Behavioral and/or objective test results

Any disagreement among these elements means that something is missing from this picture. KEEP SEARCHING FOR THE TRUTH!





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Thank you

