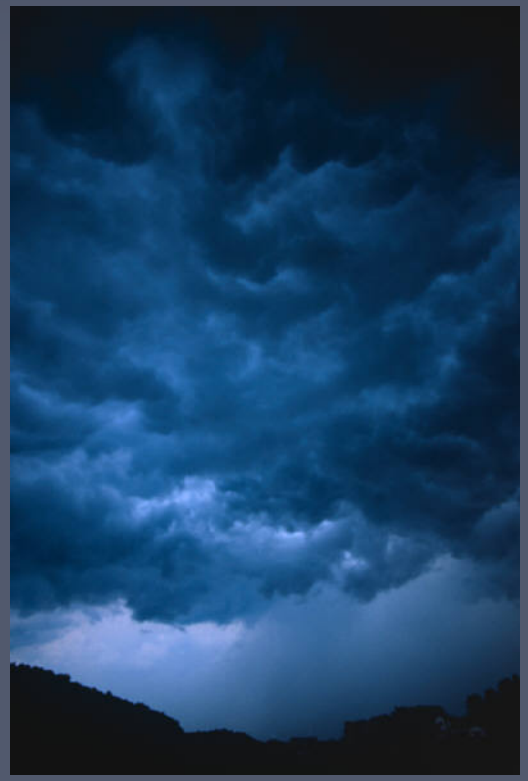
Auditory Neuropathy/ Dys-synchrony:
Shades of Gray



EHDI 2008

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Auditory Neuropathy

…term to characterize a hearing disorder in which patients presented with:

- Evidence of poor auditory neural function
- Evidence of poor auditory function
- Evidence of present hair cell function

- Incidence widely variable ranging from 1-20% of HI population

- HI = involvement of auditory nerve either as part of generalized neuropathy or isolated auditory nerve disorder

Starr et al, 1996

Auditory Neuropathy

- Berlin et al (2001) added auditory dys-synchrony
- Term "morphs": auditory neuropathy/dys-synchrony or AN/AD
- Important because the auditory nerve is not always affected always affected

Six Categories of AN/AD

- **Berlin, 2001**
  - 1. Children with absent ABR but otherwise normal hearing ability who develop speech and language
  - 2. Children with OAEs and CM initially, but disappear over time;; behavior consistent with severe-profound profound HL —hearing function may appear improved at times
  - 3. Similar to #2, but pt. functions as severely impaired majority of time; OAEs eventually disappear/CM activity remains

Six Categories of AN/AD

- **Berlin, 2001**
  - 4. Pts with no ABR and behavior consistent with profound loss
  - 5. Children "normal" at birth that develop problems w/hearing, speech, language and are later later dx with AN/AD as part of a general peripheral neuropathy (Charcot-Marie Tooth)
  - 6. Adults with no ABR but otherwise normal auditory and language function

Possible Causes of AN/AD

- May be multiple underlying causes
  - Abnormality in the synapse between primary neurons and IHC leads to temporal "jitter"; nerves are no longer phase locked to the stimulating waveform
  - Loss of function of IHCs and/or auditory neurons so that fewer or no spikes are evoked in the auditory nerve; consistent with the fact that some pts have near-normal thresholds since only a few functioning IHCs are required
  - Might be associated with "patchy" dead regions over a large part of the cochlea

Etiology of AN/AD

- Yes: Hyperbilirubinemia; perinatal asphyxia; prematurity; ototoxicity; family history; consanguinity; other neuropathies
- Maybe: IVF-6 of 26 (Raveh et al, 2007)
- No: 25-35% w/no known risk factors for AN/AD

Communication Characteristics:
Shades of Gray

- Inconsistent response to sound ***(but some exhibit consistent response to sound)***
- Speech understanding poorer than predicted by audiogram ***(but not always)***
- Speech understanding poor in presence of background noise ***(OK…almost always)***
- Often difficult to learn spoken language through listening alone ***(but can happen)***
- Range of vocal quality ***(can vary day to day)***

Sininger & Starr, 2001

AN/AD

Rapin and Gravel, 2003 & 2006

- "urge that the term auditory neuropathy be reserved for demonstrable involvement of 8th nerve as a whole or selective involvement of the spiral ganglion cells or their processes"

- " should not be used for pathologies of uncertain or mixed locations"

Anatomic Site of Pathology	Proposed Nomenclature
Hair Cells	Sensory Hearing
Spiral Ganglion Cells/VIII nerve	Auditory Neuropathy
Spiral ganglion cells/VIII nerve and/or central auditory pathway (when locus of pathology is undetermined)	Neural Hearing Loss
Hair cells and/or spiral ganglion cells/VIII nerve and/or central auditory pathway (when locus of pathology is undetermined)	Sensorineural Hearing Loss
	Rapin and Gravel, 2006

Protocol for Assessment and Management

- ABR; tymps, ART, OAEs, Case Hx
- Cochlear nerve MRI (absent/deficiency)
- Developmentally appropriate behavioral/speech perception/language assessment at frequent intervals (every 3 mos)
- Once behavioral sensitivity is established, amplification trial (DSL, etc.) w/counseling and monitoring
- Genetics, Opthalmology, Otology, EI

Protocol for Assessment and Management

Hearing Aids: Benefit or Not?

- Timeframe varies due to:
  - Developmental level
  - Consistency of amplification use
  - Clinical "wavering" of professionals
  - Progress in Speech and Language Development
  - Results of subjective evaluations (ITMAIS, ELF, etc.)

Protocol for Assessment and Management

Cochlear Implants: No, Maybe, Yes

- No: Early cases of AN thought to be due to poor function of the VIII cranial nerve
- Maybe: Due to results of pts implanted prior to OAEs
- Yes: Outcomes vary, but are similar to those w/SNHL

Cochlear Implants and AN/AD

Why does it work?

May bypass the site of lesion (IHC, synaptic junctions)

Electrical stimulation may restore synchronous firing of cochlear nerve

Post Implant: EABR and electrically evoked stapedial reflexes indicates that neural synchrony has been enhanced/achieved

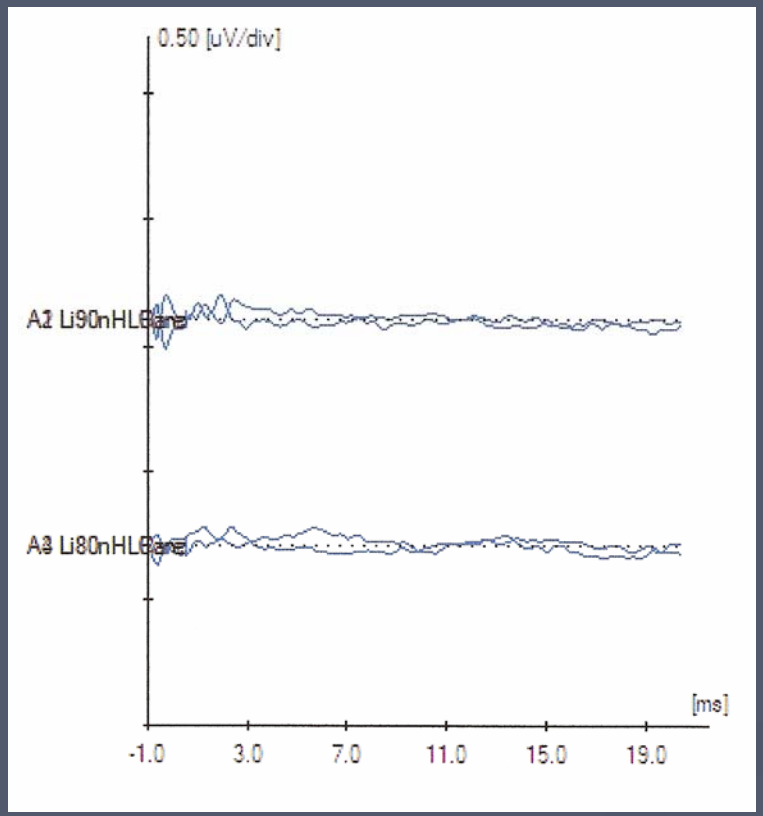
B.W.

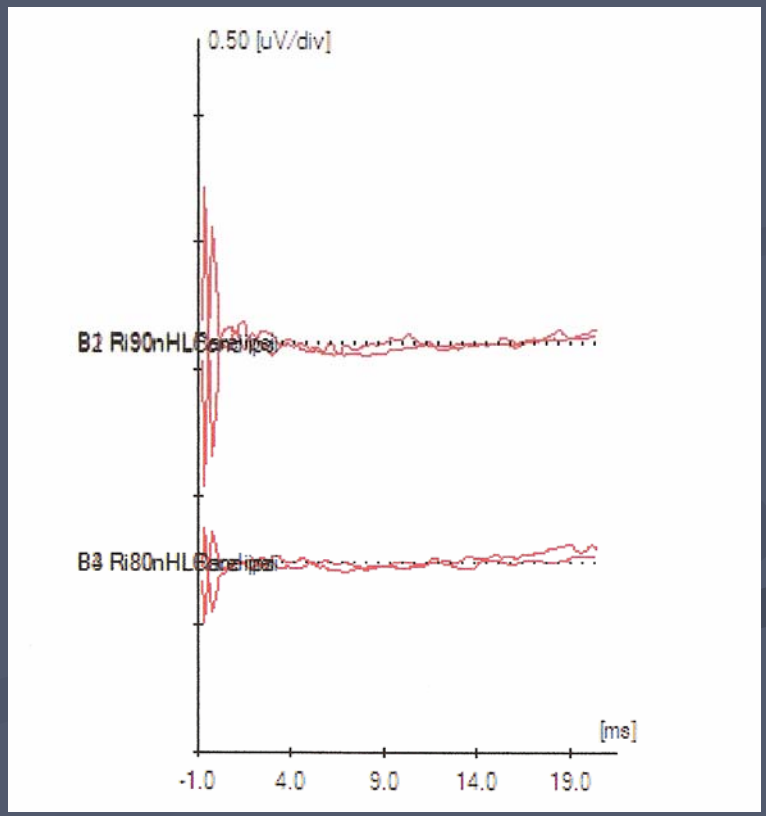
Case 1

- 8 months old w/ hx of NICU stay
- Congenital anemia, hyperbilirubinemia (double volume exchange transfusion, peak direct bili level of 24.4)
- Passed NBHS Phase I (OAEs); referred Phase II (AABR)

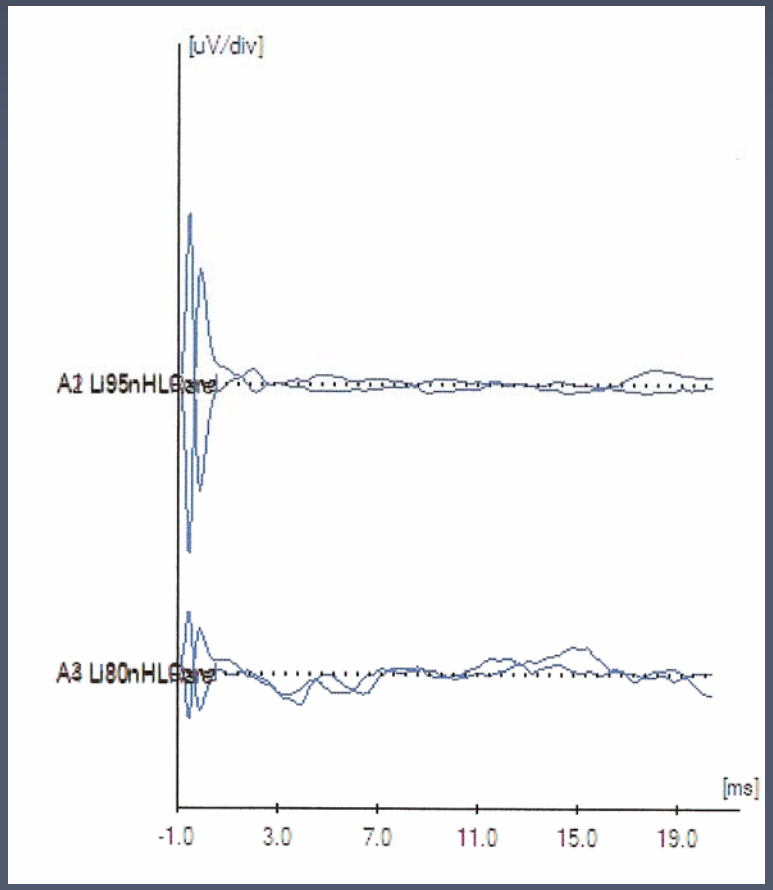
B.W. - 1st AER

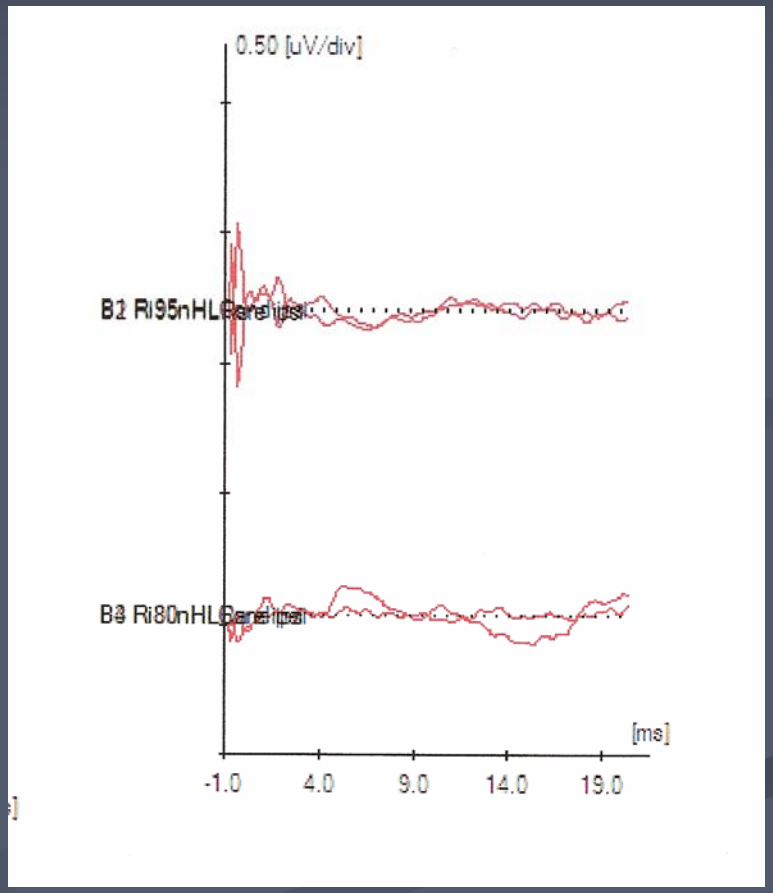
- Diagnostic eval at 3 months
  - Present OAEs, Absent Acoustic Reflexes, ABR
  - Re-eval in 3 months





B.W. - 2nd AER


[uV/div]
A1 Li95nHL
A3 Li80nHL
[ms]
-1.0
3.0
7.0
11.0
15.0
19.0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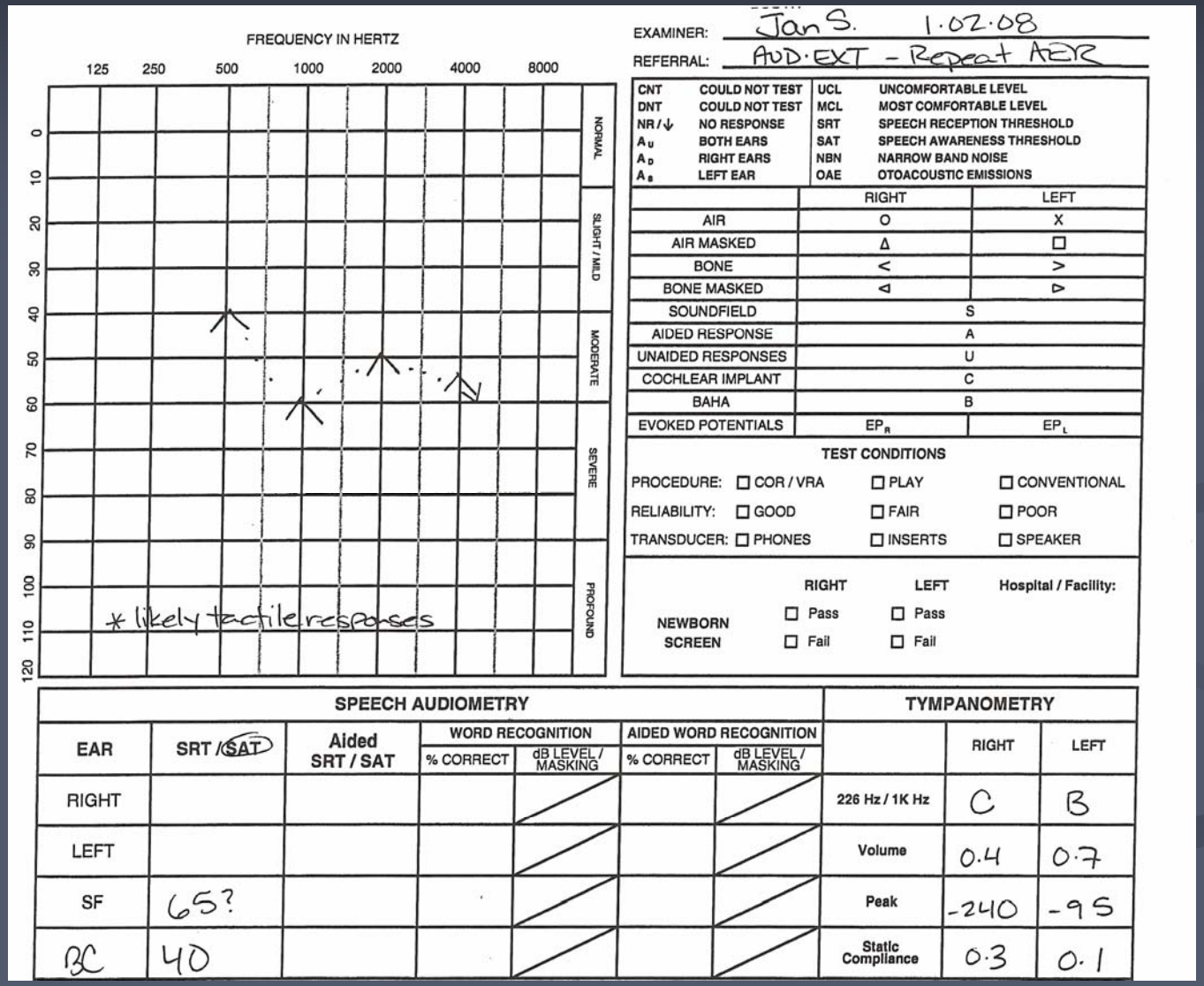
B1 Ri95nHL
B3 Ri80nHL
[ms]
-1.0
4.0
9.0
14.0
19.0

B.W. - behavioral



FREQUENCY IN HERTZ

125 250 500 1000 2000 4000 8000

0 10 20 30 40 50 60 70 80 90 100 110 120

NORMAL
SLIGHT / MILD
MODERATE
SEVERE
PROFOUND

* likely tactile responses

EXAMINER: Jan S. 1.02.08

REFERRAL: AUD·EXT - Repeat AER

CNT	COULD NOT TEST	UCL	UNCOMFORTABLE LEVEL
DNT	COULD NOT TEST	MCL	MOST COMFORTABLE LEVEL
NR / ↓	NO RESPONSE	SRT	SPEECH RECEPTION THRESHOLD
A_U	BOTH EARS	SAT	SPEECH AWARENESS THRESHOLD
A_D	RIGHT EARS	NBN	NARROW BAND NOISE
A_S	LEFT EAR	OAE	OTOACOUSTIC EMISSIONS

	RIGHT	LEFT
AIR	O	X
AIR MASKED	Δ	□
BONE	<	>
BONE MASKED	◁	▷
SOUNDFIELD	S	
AIDED RESPONSE	A	
UNAIDED RESPONSES	U	
COCHLEAR IMPLANT	C	
BAHA	B	
EVOKED POTENTIALS	EP_R	EP_L

TEST CONDITIONS

PROCEDURE: ☐ COR / VRA ☐ PLAY ☐ CONVENTIONAL

RELIABILITY: ☐ GOOD ☐ FAIR ☐ POOR

TRANSDUCER: ☐ PHONES ☐ INSERTS ☐ SPEAKER

	RIGHT	LEFT	Hospital / Facility:
NEWBORN SCREEN	☐ Pass ☐ Fail	☐ Pass ☐ Fail	

SPEECH AUDIOMETRY

EAR	SRT / (SAT)	Aided SRT / SAT	WORD RECOGNITION % CORRECT	WORD RECOGNITION dB LEVEL / MASKING	AIDED WORD RECOGNITION % CORRECT	AIDED WORD RECOGNITION dB LEVEL / MASKING
RIGHT						
LEFT						
SF	65?					
BC	40					

TYMPANOMETRY

	RIGHT	LEFT
226 Hz / 1K Hz	C	B
Volume	0.4	0.7
Peak	-240	-95
Static Compliance	0.3	0.1

B.W.

- Behavioral testing limited
- Parents note some responses to louder stimuli – but not much.
- Began trial with mild gain amplification and speech/language therapy
- Monitor and adjust!

E.C.

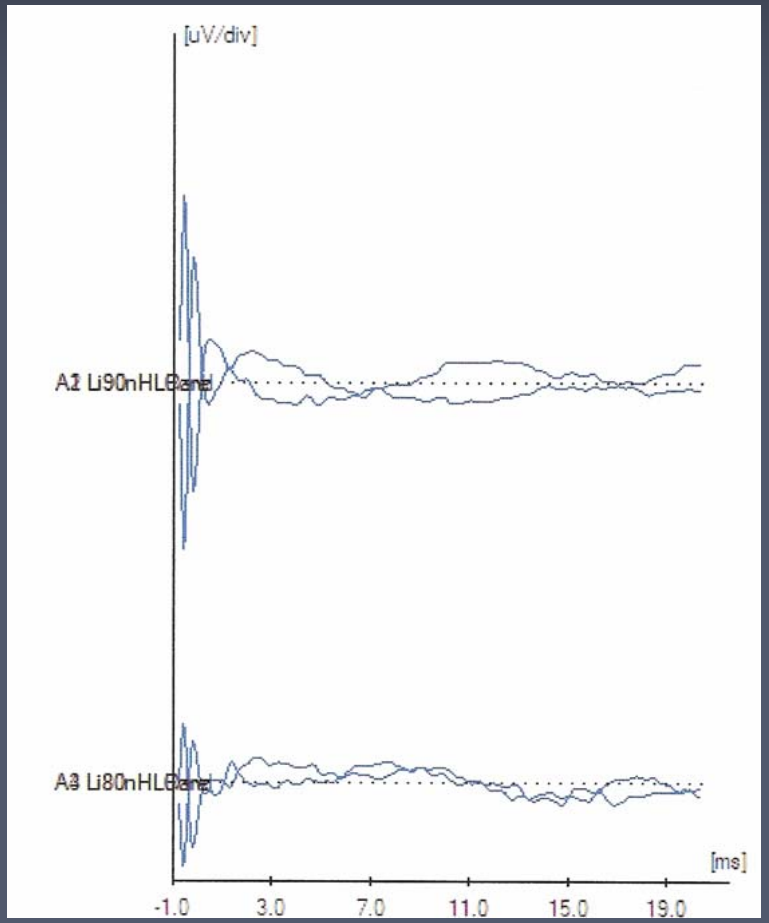
Case 2

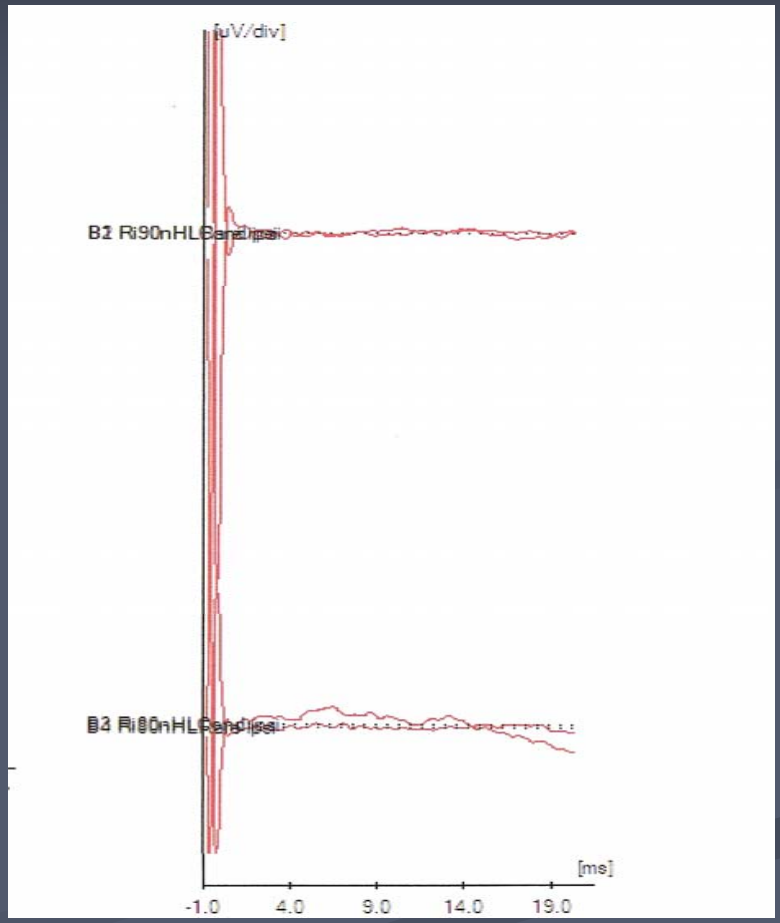
- (Foster Care) possible preterm birth, no prenatal care, exposure to cocaine/methamphetamines in utero, abnormal external ear shape
- Failed NBHS in right ear
- Previously diagnosed with suspected AN/AD in another state; mild bilateral amplification fit, inconsistent use

E.C.

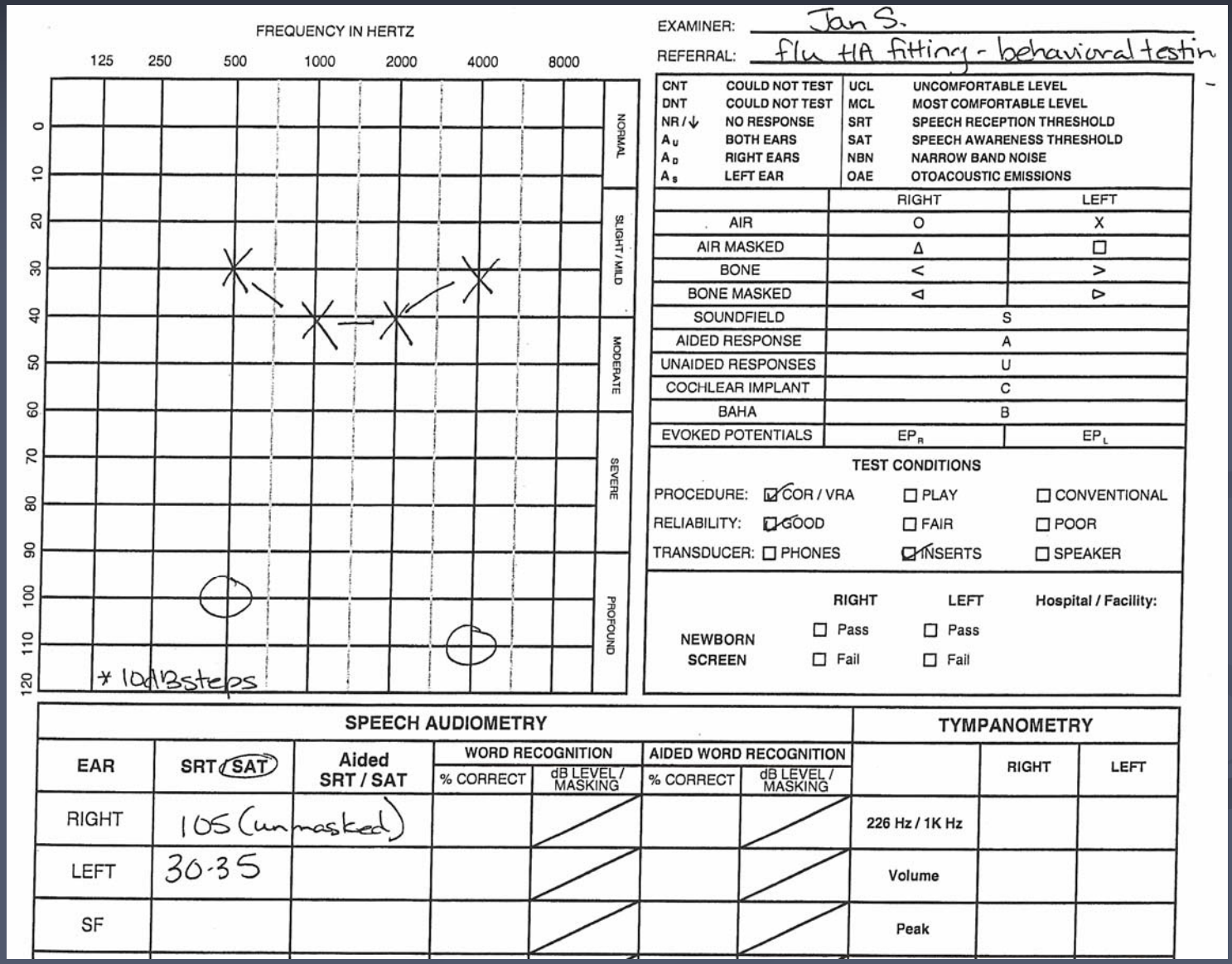
- Initial visit at ACH – 15 months:
  - CT scan, repeat AER, behavioral, speech/lang. eval
  - Present OAEs, absent ABR - left ear
  - Absent OAEs/ABR – right ear

E.C. 11/6/07

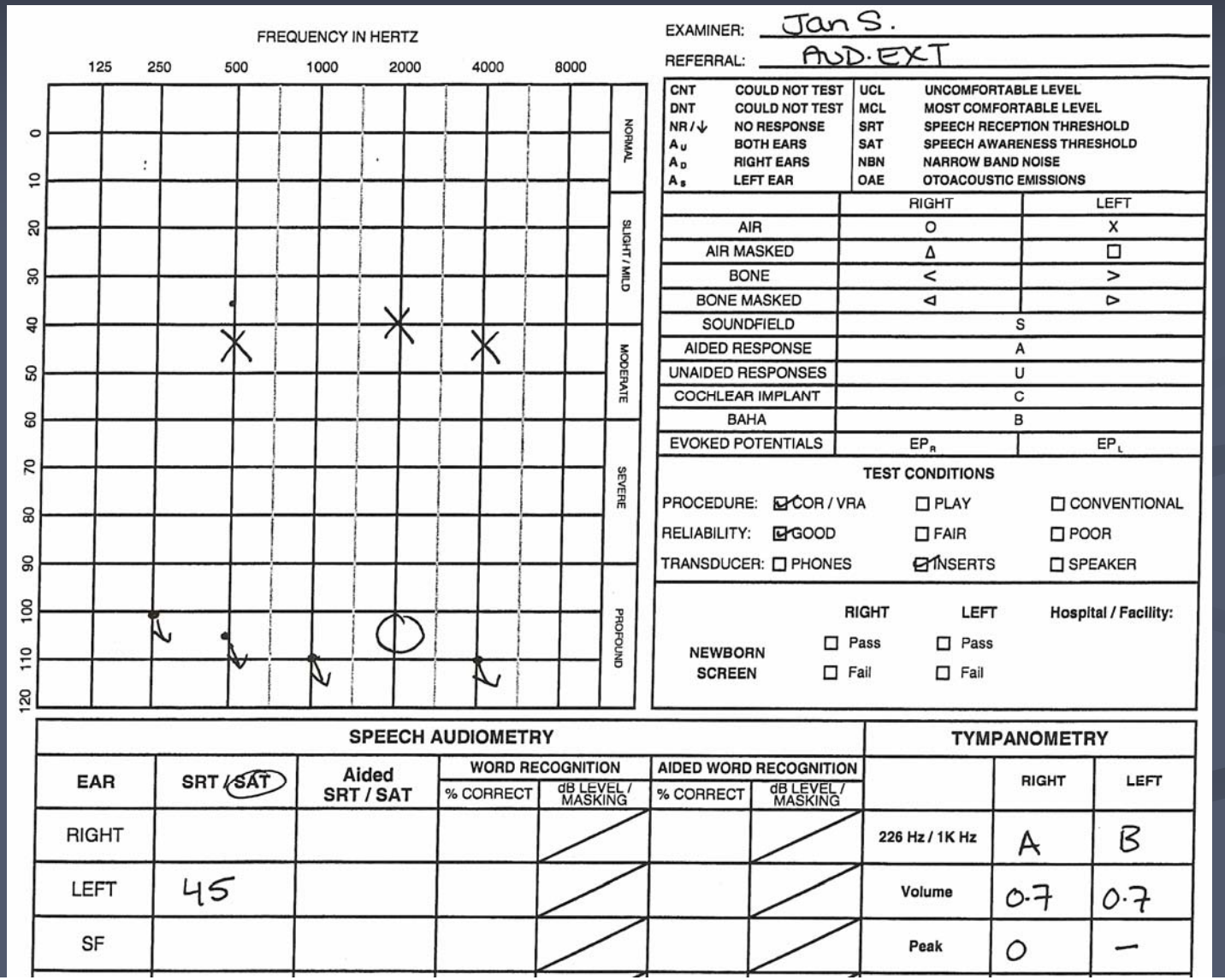




E.C. 12/7/07


FREQUENCY IN HERTZ
125 250 500 1000 2000 4000 8000
0 10 20 30 40 50 60 70 80 90 100 110 120
NORMAL
SLIGHT / MILD
MODERATE
SEVERE
PROFOUND
* 10dB steps
EXAMINER: Jan S.
REFERRAL: flu HA fitting - behavioral testin
CNT COULD NOT TEST
DNT COULD NOT TEST
NR / ↓ NO RESPONSE
A U BOTH EARS
A D RIGHT EARS
A S LEFT EAR
UCL UNCOMFORTABLE LEVEL
MCL MOST COMFORTABLE LEVEL
SRT SPEECH RECEPTION THRESHOLD
SAT SPEECH AWARENESS THRESHOLD
NBN NARROW BAND NOISE
OAE OTOACOUSTIC EMISSIONS
RIGHT LEFT
AIR O X
AIR MASKED Δ □
BONE < >
BONE MASKED ◁ ▷
SOUNDFIELD S
AIDED RESPONSE A
UNAIDED RESPONSES U
COCHLEAR IMPLANT C
BAHA B
EVOKED POTENTIALS EP R EP L
TEST CONDITIONS
PROCEDURE: ☑ COR / VRA □ PLAY □ CONVENTIONAL
RELIABILITY: ☑ GOOD □ FAIR □ POOR
TRANSDUCER: □ PHONES ☑ INSERTS □ SPEAKER
NEWBORN SCREEN
RIGHT □ Pass □ Fail
LEFT □ Pass □ Fail
Hospital / Facility:
SPEECH AUDIOMETRY
EAR
SRT (SAT)
Aided SRT / SAT
WORD RECOGNITION % CORRECT
WORD RECOGNITION dB LEVEL / MASKING
AIDED WORD RECOGNITION % CORRECT
AIDED WORD RECOGNITION dB LEVEL / MASKING
RIGHT 105 (unmasked)
LEFT 30-35
SF
TYMPANOMETRY
RIGHT
LEFT
226 Hz / 1K Hz
Volume
Peak

E.C. 1/8/08



FREQUENCY IN HERTZ

125 250 500 1000 2000 4000 8000

0 10 20 30 40 50 60 70 80 90 100 110 120

NORMAL
SLIGHT / MILD
MODERATE
SEVERE
PROFOUND

EXAMINER: Jan S.

REFERRAL: AUD·EXT

CNT	COULD NOT TEST	UCL	UNCOMFORTABLE LEVEL
DNT	COULD NOT TEST	MCL	MOST COMFORTABLE LEVEL
NR / ↓	NO RESPONSE	SRT	SPEECH RECEPTION THRESHOLD
A_U	BOTH EARS	SAT	SPEECH AWARENESS THRESHOLD
A_D	RIGHT EARS	NBN	NARROW BAND NOISE
A_S	LEFT EAR	OAE	OTOACOUSTIC EMISSIONS

	RIGHT	LEFT
AIR	O	X
AIR MASKED	Δ	□
BONE	<	>
BONE MASKED	◁	▷
SOUNDFIELD	S	
AIDED RESPONSE	A	
UNAIDED RESPONSES	U	
COCHLEAR IMPLANT	C	
BAHA	B	
EVOKED POTENTIALS	EP_R	EP_L

TEST CONDITIONS

PROCEDURE: ☑ COR / VRA ☐ PLAY ☐ CONVENTIONAL

RELIABILITY: ☑ GOOD ☐ FAIR ☐ POOR

TRANSDUCER: ☐ PHONES ☑ INSERTS ☐ SPEAKER

	RIGHT	LEFT	Hospital / Facility:
NEWBORN SCREEN	☐ Pass ☐ Fail	☐ Pass ☐ Fail	

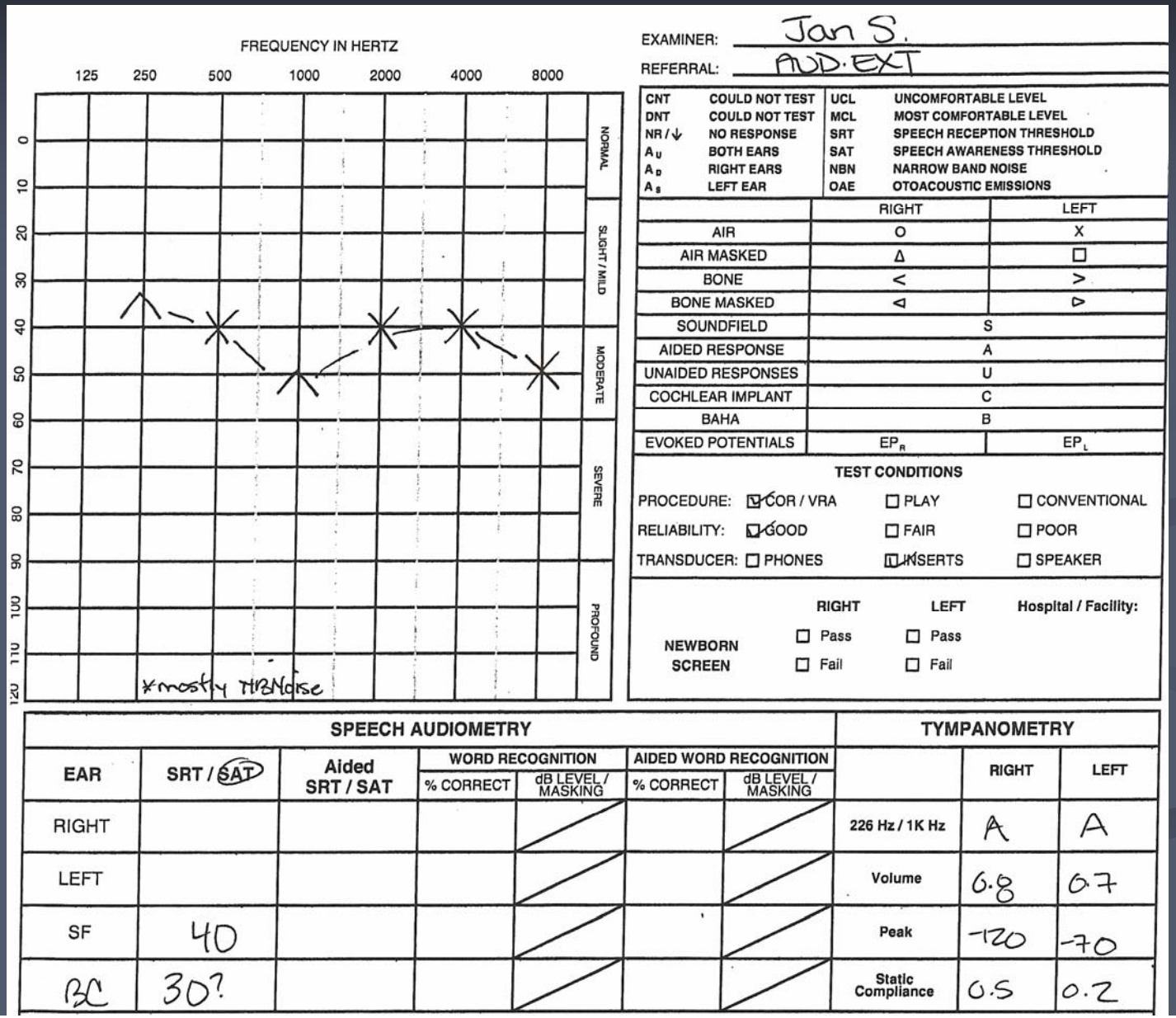
SPEECH AUDIOMETRY

EAR	SRT / SAT	Aided SRT / SAT	WORD RECOGNITION % CORRECT	WORD RECOGNITION dB LEVEL / MASKING	AIDED WORD RECOGNITION % CORRECT	AIDED WORD RECOGNITION dB LEVEL / MASKING
RIGHT						
LEFT	45					
SF						

TYMPANOMETRY

	RIGHT	LEFT
226 Hz / 1K Hz	A	B
Volume	0.7	0.7
Peak	O	—

E.C. 2/13/08



SPEECH AUDIOMETRY

EAR	SRT / SAT	Aided SRT / SAT	WORD RECOGNITION % CORRECT	WORD RECOGNITION dB LEVEL / MASKING	AIDED WORD RECOGNITION % CORRECT	AIDED WORD RECOGNITION dB LEVEL / MASKING
RIGHT						
LEFT						
SF	40					
BC	30?					

TYMPANOMETRY

	RIGHT	LEFT
226 Hz / 1K Hz	A	A
Volume	0.8	0.7
Peak	-120	-70
Static Compliance	0.5	0.2

E.C.

- Now has full-time HA use (mild gain on left; fit to loss on right)
- Currently getting speech tx 3xs month
- Recent MRI – normal eighth nerve on left; absent/deficient eighth nerve on right
- CI has been discussed, but due to MRI findings and recent acquisition of full-time HA use/tx, no decision has been made yet.

E.C.

Plans:

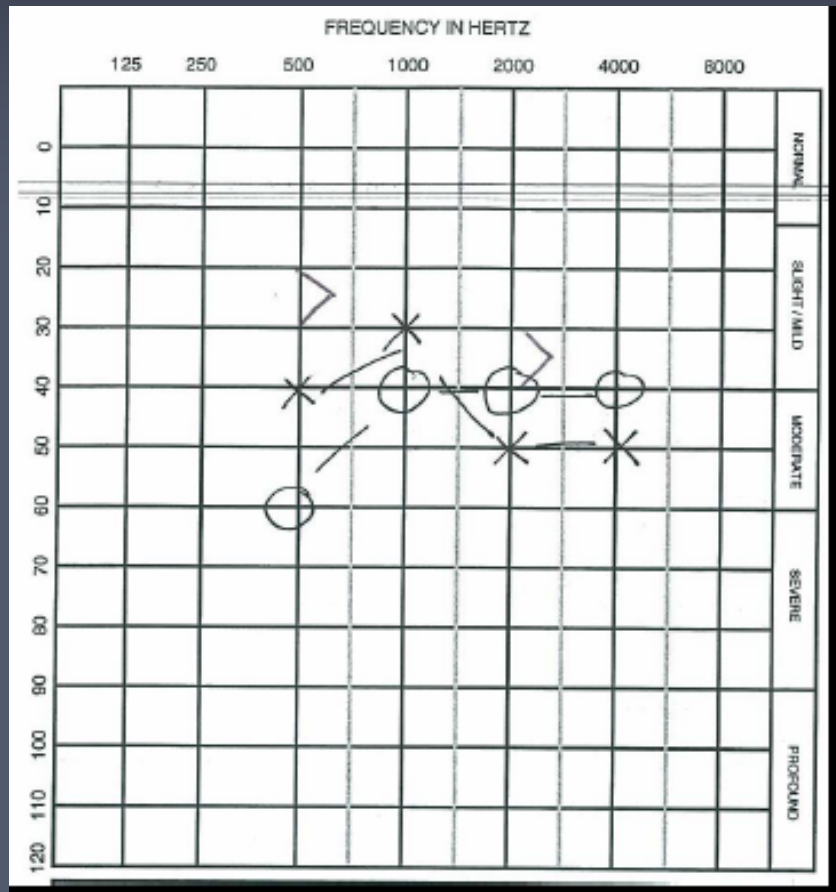
- Continue to monitor rate of speech/language development
- Possibly increase amount of speech/language therapy
- Remove right HA (no VIIIth nerve)
- Consider CI

K.M.

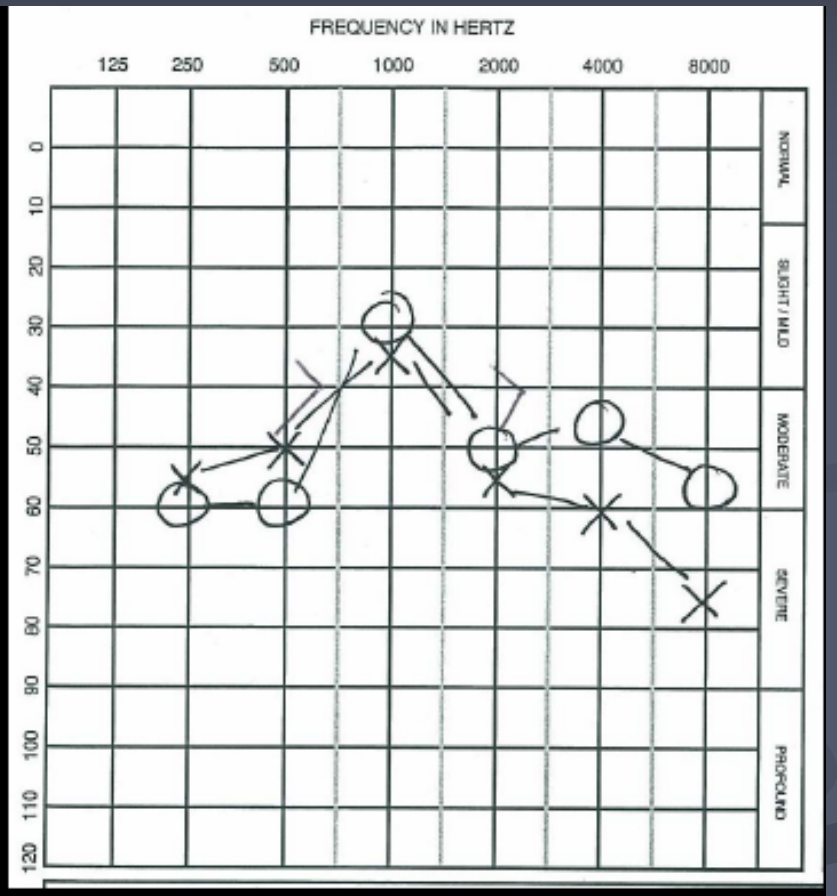
Case 3

- 6 year old female
- Normal hearing, speech/language, development
- Seen as in-pt initially with hx of viral cerebellar ataxia
- Unsteady gait
- Unable to understand parents following viral attack

K.M.



3/16/07

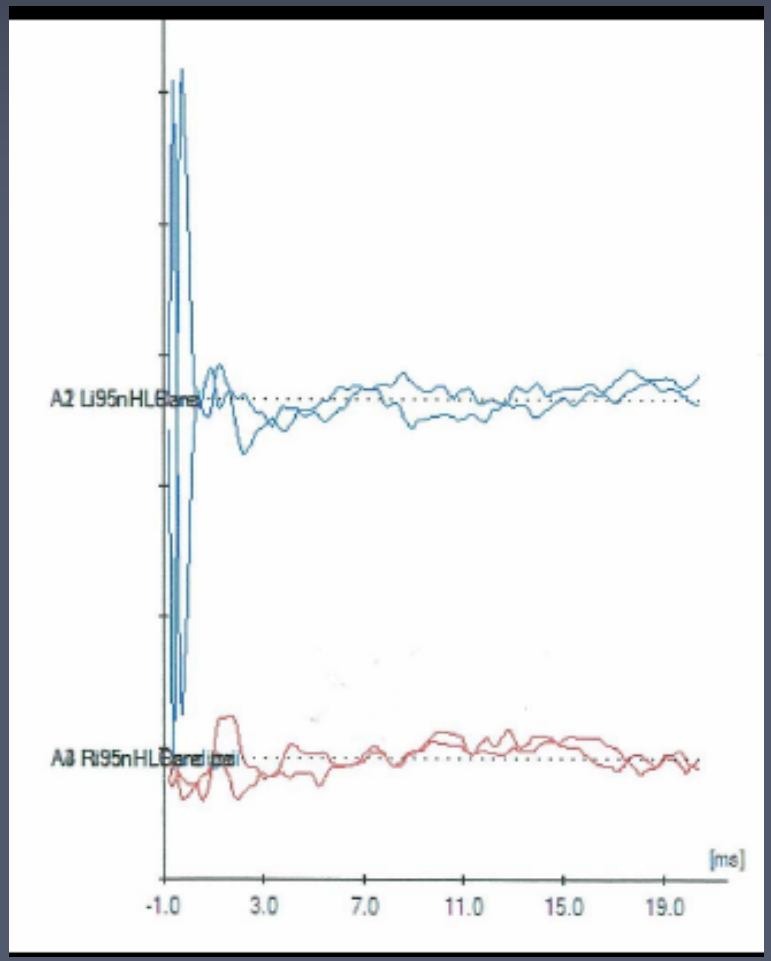


4/03/07

K.M.

- Mild/moderate behavioral hearing loss
- Can not obtain SAT/SRT
- Present OAEs bilaterally
- Absent/elevated Acoustic Reflexes

K.M.



Absent ABR - AN/AD

K.M.

- Due to sudden onset – conservative approach
- Trial with personal FM
- Repeat behavioral testing
- Speech/language tx (short-term)

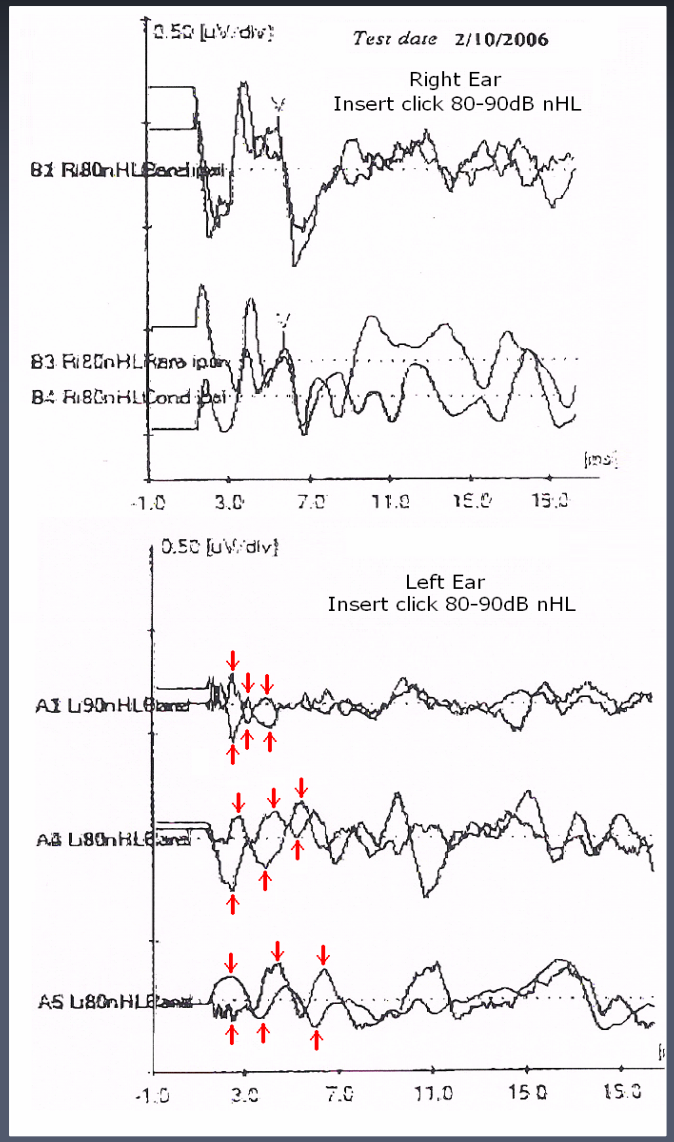
- Almost one year out – no noted improvement
  - Just began trial with bilateral amplification
  - CI has been discussed, but parents very uncertain at this time
  - Discussed further neurological/genetic testing

B.A.

Case 5

- Ten year old male who referred following hearing screening with teacher concerns
  - OAE present and WNL, AU
  - Type A tympanograms, AU
  - Audiometric results
    - Hearing within normal limits, AD
    - No reliably, obtained thresholds, AS

- "normal" "malingering"???



ABR:

- Right ear: good wave morphology and absolute latency of components in the expected range
- Left ear: poor wave morphology, inverting CM when stimulus polarity reversed

Reflexes:

- MEMR absent ipsi, elevated contra, AS
- MEMR present ipsi, absent contra, AD

B.A.

- Recommendations
  - Classroom
    - Optimal seating
    - Confirmation of understanding what has been said
    - Additional visual aids/media to supplement the spoken information of the lesson
  - Medical
    - Referral to see otologist
    - Follow-up re-evaluation
    - Trial use of an earplug
    - Lost to f/u

S.C.

Case 6

Complaints:

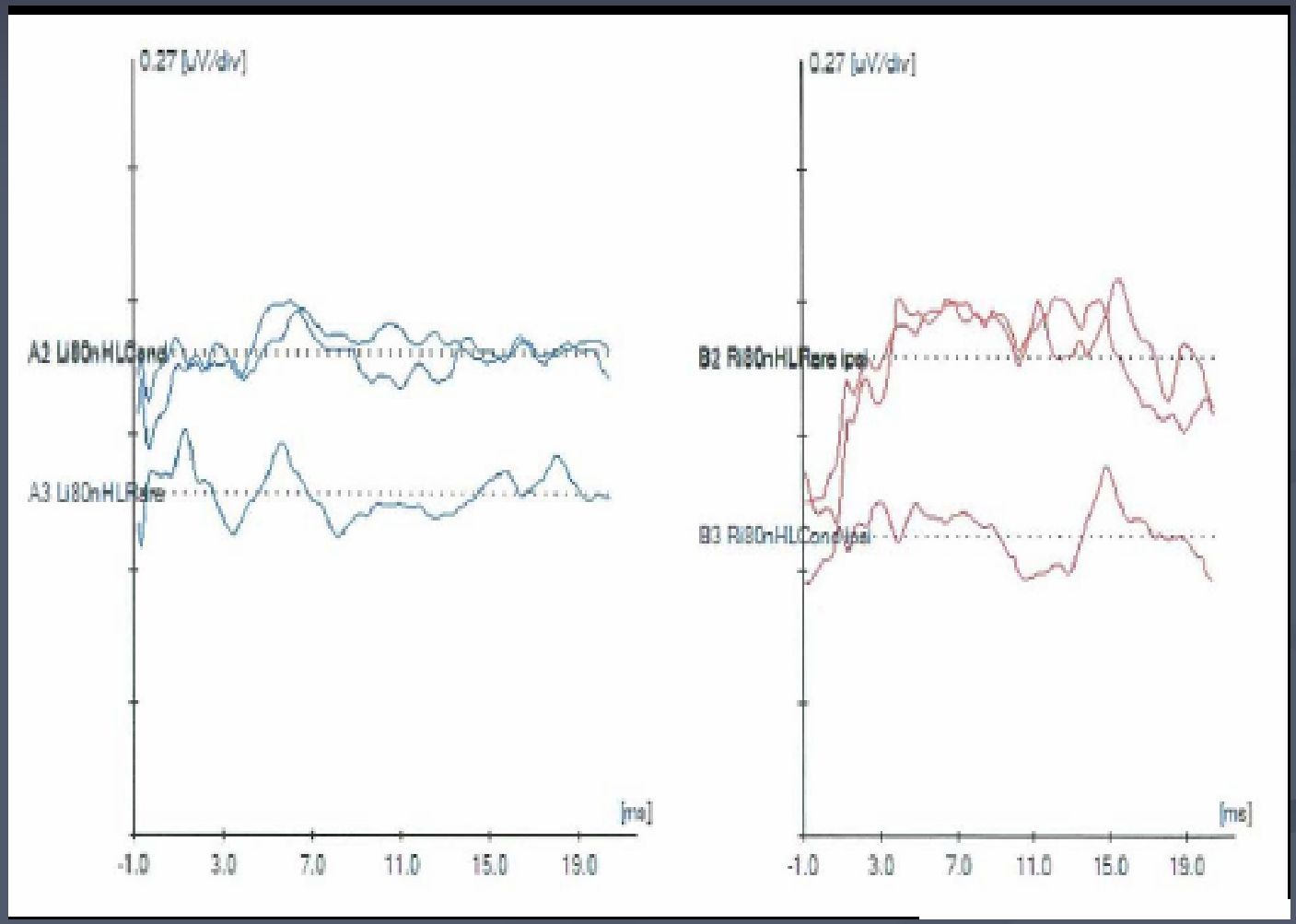
- Trouble listening
- Trouble in school
- TV up
- Some days better/worse

- Currently in a facility for teenagers with emotional disorders/hx of sexual abuse
- Initial school problems considered to be related to ESL (moved to US at 6) although bilingual
- Educated, concerned family

S.C.

- SRTS at 40-50 dB
- PT essentially WNL
- Malingering ?
- Absent/elevated reflexes
- Present OAEs
- Reports speech as “bzup bzup, bzup bzup”

S.C.


0.27 [µV/div]
A2 Li80nHLCond
A3 Li80nHLRare
[ms]
-1.0
3.0
7.0
11.0
15.0
19.0
0.27 [µV/div]
B2 Ri80nHLRare ipsi
B3 Ri80nHLCond ipsi
[ms]
-1.0
3.0
7.0
11.0
15.0
19.0

S.C.

Outcomes:

- Family/individual/staff counseling
- Trial comparing HA and personal FM
- Preferred FM
- Unanticipated environmental interference

Auditory Neuropathy/ Dys-Synchrony

Summary

- **The cross check principle continues to be the gold standard "only the rules are different"**
- **The one constant about AN/AD/?? is variability!**