Early identification of hearing loss in infants and young children using objective electrophysiological measures: Best practices and new research

PART 1a (8:30 am - 12:00 pm)

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University of British Columbia
Hamber Professor of Clinical Audiology

CASLPM, Winnipeg, MN
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Disclosure statement
BC Early Hearing Program (consultant) & Hamber Chair position: contribute to my research program

Other funding
UBC Faculty of Medicine
LEARNING OUTCOMES

As a result of this Continuing Education Activity, participants will be able to:

1) Conduct air- and bone-conduction auditory brainstem response testing in infants

2) Interpret auditory brainstem response findings for infants with normal hearing and conductive and sensory/neural hearing loss

3) Explain potential clinical applications of auditory steady-state responses and cortical auditory-evoked potentials for the infant population
TOPIC AREAS TO BE ADDRESSED

PART 1 (a & b):
Estimation of infant hearing thresholds using brief-tone auditory brainstem response (ABR)

PART 2:
Clinical application of auditory steady-state responses (ASSRs) & cortical auditory evoked potentials (CAEPs):
a brief overview of new research
PART 1:
Estimation of infant hearing thresholds using brief-tone auditory brainstem response (ABR) & identification of hearing loss type

8:30-10:00 am: a.1) Overview of methodology & clinical protocols
10:00 am - 12:00 pm: a.2) Case Studies
1:00-2:45 pm: b) Auditory neuropathy spectrum disorder (ANSD) -- Protocols + Case Studies
"The audiological assessment should include:

... A frequency-specific assessment of the ABR using air-conducted tone bursts and bone-conducted tone bursts when indicated. When permanent hearing loss is detected, frequency-specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices.”
Air-conduction (AC) ABR to brief tones—preamble

Clinical goal for AC ABR testing?

- Accurate estimation of behavioural hearing thresholds
  AC thresholds within normal limits?
  OR
  AC thresholds elevated?

- Standard practice for pure-tone audiometry & diagnostic ABR testing
Clinical goal for BC ABR testing?

- Accurate estimation of BC thresholds to determine type of hearing loss responsible for elevated air-conduction (AC) thresholds

  Conductive?  Sensorineural?  Mixed?
  - How much is conductive?

- Standard practice for pure-tone audiometry

- Should be standard practice for infant ABR testing
Very Brief History of BC ABR testing:

- In the late 1970s and 1980s, BC ABR research emerged (brief tones and clicks) – some technical issues arose but research continued

Examples of early studies:
- Mauldin & Jerger (1979) found that adult wave V latencies to BC clicks were longer than AC clicks
- Boezeman et al. (1983) found the same for 2000-Hz brief tones
- Cornacchia et al. (1983) compared AC & BC ABR wave V latencies in infants & adults; found that infant wave V latencies to BC stimuli were prolonged relative to adults

-- Differences in AC vs BC ABR results and maturational effects emerging in ABR research
Examples of early studies that focussed on clinical use of brief-tone BC ABR testing in infants:

**Clicks:**

**Brief-tones:**

Examples of more recent infant brief-tone BC ABR research:

(i) feasible to record brief-tone BC ABRs clinically
(ii) frequency- and mode- (AC vs BC) dependent infant-adult differences to be accounted for in their interpretation
Overview of methodology
Typical stimuli used to elicit the ABR (e.g., Davis et al., 1984)

Under investigation
**Air- and bone-conducted brief tones**

*Stimulus parameters*

"2-1-2" (cycles) linearly-gated tones; 5-cycle Blackman tones (no plateau)

500 Hz: AC/BC 4 ms rise/fall, 2 ms plateau; 10 ms
1000 Hz: AC/BC 2 ms rise/fall, 1 ms plateau; 5 ms
2000 Hz: AC/BC 1 ms rise/fall, 0.5 ms plateau; 2.5 ms
4000 Hz: AC 0.5 ms rise/fall, 0.25 ms plateau; 1.25 ms

* BC 1 ms rise/fall, 0.25 ms plateau, 2.25 ms

* to reduce ringing

(Small & Stapells, 2003)
Calibration
Supra-aural TDH49/ER3-A insert earphones/B71 transducer

**UNITS:** $\text{dB peak (peak hold) minus } 3 \text{ dB} = \text{dB ppe}$

**AC:** $\text{dB ppe SPL}$

**BC:** $\text{dB ppe re: } 1 \mu\text{N}$

### Acoustic calibration for 0 dB nHL

<table>
<thead>
<tr>
<th></th>
<th>500 Hz</th>
<th>1000 Hz</th>
<th>2000 Hz</th>
<th>4000 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>AC TDH</td>
<td>25</td>
<td>22</td>
<td>67</td>
<td>23</td>
</tr>
<tr>
<td>AC ER3-A</td>
<td>22</td>
<td>25</td>
<td>54</td>
<td>26</td>
</tr>
</tbody>
</table>

(Small & Stapells, 2003)
**ABR testing: What we aim for in the clinic!**

**AC testing**

- insert earphones preferred for comfort & more accurate hearing-aid fittings
- supra-aural earphones can be used when insert earphones not appropriate
Bone oscillator coupling method in infants

Small, Hatton & Stapells, 2007

- No significant differences 500-4000 Hz

Caveat: used trained individuals for both methods
-- We do not recommend that a parent couple the bone oscillator to their child’s skull

- BC EHP: clinicians often use hand-held method
  – least likely to wake up infant
Bone oscillator placement

- Small, Hatton & Stapells (2007) compared infant ASSR thresholds at different positions on the skull

No difference for T versus M position

Significantly poorer for F versus T or M position

- BC EHP: clinicians use the T position method
  - greatest range of intensities available
  - easier to maintain firm consistent placement than M position
**EEG recording parameters**

- AC: 1-channel recording

Consider two-channel AC recordings if large asymmetry between ears

Adapted from Stapells & Oates (1997)
**EEG recording parameters**

- **Cz**
- **M1**
- **M2**
- **ground**
- **Stimulus left**

**BC**: 2-channel recording
EEG recording parameters

- **Always use two-channel BC recordings**
EEG recording parameters

EEG filter:
- High Pass: 20-30 Hz
- Low Pass: 1500-3000 Hz (Slope: 6 or 12 dB/octave, analog)

Artifact reject:
- Trials exceeding ± 25 µV (equals Nicolet "50 µV")
  -- can reduce this to optimize recording (balance with rejection rate)

# of trials:
- Typically 2000 per average

# of sweeps:
- Two or more as needed to obtain good signal-to-noise ratio

Display Scale:
- Avoid too large a scale close to threshold

Adapted from Stapells & Oates (1997)
**Air- and bone-conduction brief tones**

Setting latency window for signal-to-noise ratio (SNR) & residual noise (RN)

<table>
<thead>
<tr>
<th>500 Hz</th>
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<th>2000 Hz</th>
<th>4000 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>AC &amp; BC</td>
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</tr>
</tbody>
</table>

*Start*  
10.5  

*End*  
20.5

*Note: should shift 10 ms window later for higher presentation levels to avoid stimulus artifact*

500 Hz BC stimuli > 30 dB re: 1 µN  → 14 – 24 ms

(BCEHP, 2012)
Interpretation of waveforms

Response present:

- SNR > 1
- Wave V visually replicates

Response absent:

- SNR << 1
- Visually flat
- RN < 0.05-0.08 µV

Cannot evaluate:

- SNR << 1
- RN > 0.08 µV
- No repeatable peaks
- Not visually flat

(BCESHP, 2012)
SNR latency windows

Infant: 12 months
BC left mastoid

2000 Hz
500 Hz

dB nHL
6.5 16.5
10.5 20.5

Ipsi
Contra

Ipsi
Contra

Ipsi
Contra

Ipsi
Contra

Ipsi
Contra

Green check marks:
- 2000 Hz: 60 dB nHL, latency 6.5 ms
- 500 Hz: 50 dB nHL, latency 10.5 ms

Red X:
- 2000 Hz: 40 dB nHL, latency 16.5 ms
- 500 Hz: 20 dB nHL, latency 20.5 ms
Infant: 12 months
BC left mastoid

SNR latency windows

All SNRs < 0.54
Cannot evaluate:
- SNR <<1
- RN > 0.08 µV
- No repeatable peaks
- Not visually flat

- SNR = 0.51 µV
- RN = 0.12 µV
- Not visually flat

- SNR = 0.47 µV
- RN = 0.15 µV
- Not visually flat

- SNR = 0.78 µV
- RN = 0.09 µV
- Not visually flat

- SNR = 0.32 µV
- RN = 0.05 µV
- Visually flat
What does the absence or presence of a response mean re: the infant’s hearing?

• need to relate these results to what is “normal” or “near normal” for AC & BC stimuli for infants

• need to know how these elevated responses predict the degree & type of hearing loss
Definition of terms (BCEHP, 2012)

**Normal behavioural threshold:**
- 25 dB HL

**Normal ABR maximum level:**
- ABR presentation level at which the majority of normal-hearing infants have a response present

**eHL correction:**
- Correction factor used to estimate behavioural hearing threshold (dB HL) from the ABR threshold (dB nHL)

\[
\text{ABR threshold (dB nHL)} - \text{eHL correction (dB)} = \text{estimated behavioural threshold (dB HL)}
\]
Normal ABR maximum levels & eHL correction for infants

*Air- and bone-conduction ABR*

<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>BC EHP</td>
<td>AC</td>
<td>AC</td>
<td>AC</td>
<td>AC</td>
</tr>
<tr>
<td>Normal ABR Max (dB nHL)</td>
<td>35</td>
<td>35</td>
<td>30</td>
<td>25</td>
</tr>
<tr>
<td>Range in literature</td>
<td>30-35</td>
<td>30-35</td>
<td>20-30</td>
<td>20-25</td>
</tr>
<tr>
<td>BC EHP eHL correction (dB)</td>
<td>10</td>
<td>10</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Range in literature</td>
<td>10-15</td>
<td>5-10</td>
<td>0-5</td>
<td>-5-0</td>
</tr>
</tbody>
</table>

*(BC-EHP 2012, 2015; Small & Stapells, Ch. 21, 2017)*
# Normal ABR maximum levels & eHL correction for infants

## Air- and bone-conduction ABR

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(BC-EHP 2012, 2015; Small & Stapells, Ch. 21, 2017)
Estimation of infant hearing thresholds
BABY X
AIR-CONDUCTION, 2000 Hz

ELEVATED THRESHOLD @ 60 dB nHL

Flat tympanogram
Absent OAE

Conductive?
BABY X
AIR-CONDUCTION, 2000 Hz

80 dBnHL

60 dBnHL

50 dBnHL

30 dBnHL

NORMAL

Flat tympanogram
Absent OAE

Conductive?
Could be sensorineural or mixed!
Need BC ABR testing to be sure!
BABY X
AIR-CONDUCTION @ 2000 Hz

2-channel BC @ 30 dB nHL

ELEVATED THRESHOLD @ 60 dB nHL

Flat tympanogram
Absent OAE

BC ABR ABSENT @ 30 dB nHL

sensorineural (or mixed) hearing loss
• If audiologist conducts only AC ABR testing and tympanometry & otoacoustic emissions (OAEs) to identify a conductive component
  ➢ *May lead to error*

• Tympanometry in very young infants:
  - may fail to identify middle-ear involvement
  - flat tympanogram does not assess amount of hearing loss attributed to the conductive component

• OAEs:
  - sensitive to middle-ear involvement but only helpful if present

Only BC thresholds can distinguish between sensorineural, conductive and mixed losses

**AND**
determine magnitude of conductive loss
How well do BC ABR results predict the nature of the hearing loss (conductive versus sensorineural loss?)

Data collected from BC EHP diagnostic follow up:

<table>
<thead>
<tr>
<th></th>
<th>Nature of loss is certain</th>
<th>All data (includes cases where assumptions made)</th>
</tr>
</thead>
<tbody>
<tr>
<td>500 Hz</td>
<td>91.9% (65 cases)</td>
<td>81.2% (126 cases)</td>
</tr>
<tr>
<td>2000 Hz</td>
<td>94.2% (37 cases)</td>
<td>93.7% (49 cases)</td>
</tr>
</tbody>
</table>

(Hatton, Janssen & Stapells, 2012)
Where does BC ABR testing fit in the diagnostic protocol?

- After AC thresholds are established for both ears?
- After AC threshold search, tympanometry & OAEs?
- Before AC testing?
- As soon as AC thresholds are determined to be elevated?

➤ Want BC ABR thresholds early in diagnostic testing to avoid delays in medical follow-up or intervention

✓ BC testing occurs after AC thresholds are shown to be elevated in at least one ear at 2000 Hz (> normal max) -- before AC threshold search
BC EHP ABR Test Protocol (2012)

Tone-ABR Test Sequence (partial)

Start
- EAR#1
  - AC 2k
  - 30 dB nHL

Go to 500 Hz @ normal max Ear#1
-- 30 dB nHL

Begin AC at 2000 Hz at normal max Ear#1
-- 30 dB nHL

AC normal @ 2k both ears

Go to 500 Hz @ normal max -- 35 dB nHL
Case 1: Diagnostic ABR following “refer” from BC EHP

NORMAL BILATERALLY
BC EHP ABR Test Protocol (2012)

Tone-ABR Test Sequence (partial)

Start

Present

EAR#1
AC 2k
30dB

Absent

Conductive hearing loss

Present

EAR#2
AC 2k
30dB

Absent

BC normal level

Present

EAR#2
BC 2k
30dB

Absent

Present

EAR#2
BC 500
20dB

Absent

Present

EAR#2
BC 2k
60dB & thresh

Absent

EAR#1
BC 2k
30dB

Present

EAR#1
BC 500
60dB & thresh

Absent

go to

EAR#1
AC 2k
60-80dB & thresh

AC threshold search

BC normal level

60-80dB, thresh

600 Hz
(Each ear @ 30-40 dB)
BC EHP ABR Test Protocol (2012)
Tone-ABR Test Sequence (partial)

Start

Present

EAR#1
AC 2k
30 dB

Absent

SNHL hearing loss

Present

EAR#1
AC 2k
30 dB

Absent

BC elevated level
(60 dB nHL)

Present

EAR#1
BC 2k
30 dB

Absent

AC threshold search

Present

EAR#1
BC 500
20 dB

Absent

AC 500 Hz
[Each ear @
30-40 dB]

Present

EAR#2
BC 500
20 dB

Absent

EAR#2
AC 2k
30 dB

Present

EAR#2
BC 2k
60 dB & thresh

Absent

EAR#1
BC 500
20 dB

Present

EAR#1
BC 2k
60 dB & thresh

Absent

EAR#2
AC 2k
60-80 dB, thresh

Present

EAR#1
AC 2k
60-80 dB, thresh

Absent

500 Hz
AC & BC
Each ear

Present

A

B

C

go to

A

C

D

E
Features of BC EHP protocol:

- Start at “normal levels” for AC testing to save time if infant has hearing within normal limits
- Go to BC testing early in test sequence when AC is elevated
- Go to maximum BC testing levels first if BC is not within normal limits
- Use large steps when searching for threshold; small step sizes as time allows

- Want to determine type then degree of loss as efficiently as possible
Isolation of test cochlea
**BC ABR: Isolation of test cochlea**

**INFANTS**
- Clinical masking?
  -- IA for AC stimuli are not known
  -- IA for BC have been approximated with indirect measures (ABR & ASSR data)
  -- effective masking levels for BC not known for ABR (BC ASSR data available)
  -- are corrections for occlusion effect needed? (BC ASSR data available)

**ADULTS**
- Use masking to isolate test ear as needed
  - interaural attenuation (IA) & effective masking levels for AC & BC stimuli are well established
  - corrections for occlusion effect are known
<table>
<thead>
<tr>
<th>Study</th>
<th>Method</th>
<th>Indirect measure</th>
<th>Age</th>
<th>Interaural Attenuation (dB)</th>
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<tr>
<td>Yang &amp; Stuart 1987</td>
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<td>Neonate</td>
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<td></td>
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<td>12 months</td>
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## Interaural attenuation of BC stimuli – indirect measures

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<td>Small &amp; Stapells 2008</td>
<td>ASSR- AM/FM 500-1000 Hz Fc</td>
<td>Ipsi/contra asymmetries</td>
<td>Adult</td>
<td>0-10</td>
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<td></td>
<td></td>
<td></td>
<td>0-6 months</td>
<td>10-30</td>
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<td></td>
<td></td>
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<td>10-30</td>
</tr>
<tr>
<td>Hansen 2010 (M.Sc. Thesis)</td>
<td>ASSR- AM/FM 1000 Hz</td>
<td>Effective masking levels (Binaural AC)</td>
<td>Adult</td>
<td>0</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>0-7 months</td>
<td>10-15</td>
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</table>

- Interaural attenuation for BC stimuli in infants is a minimum of 10-35 dB depending on the age
Utilize ipsilateral/contralateral asymmetries?

> Expected pattern for normal cochleae up to 1 to 2 years of age -- normal hearing or conductive loss (e.g., aural atresia)

[e.g., Foxe & Stapells, 1993; Stapells & Ruben, 1989; Stapells & Mosseri, 1991]

<table>
<thead>
<tr>
<th>Amplitude:</th>
<th>contra <strong>smaller than</strong> ipsi</th>
<th>Latency:</th>
<th>contra <strong>later than</strong> ipsi</th>
</tr>
</thead>
</table>

BC left mastoid

2000 Hz @ 40 dB nHL
Utilize ipsilateral/contralateral asymmetries?

- Expected pattern for normal cochleae up to 1 to 2 years of age -- normal hearing or conductive loss (e.g., aural atresia) [e.g., Foxe & Stapells, 1993; Stapells & Ruben, 1989; Stapells & Mosseri, 1991]

2000 Hz @ 40 dB nHL

**Amplitude:** contra **smaller than** ipsi  
**Latency:** contra **later than** ipsi
Bone-conduction ABR

Contra >> IPSI

SEVERE UNILATERAL (RIGHT EAR) SNHL

(Stapells, personal communication)
Case 2: 13 months – referred from NHS

2000 Hz BC Right Mastoid

Ch A = right
Ch B = left

Which ear is responding? Right ear!

30 & 40 dB nHL: Ipsi > contra

30 dB repeated
13 months – referred from NHS

2000 Hz BC **Left** Mastoid

**Ch A = right**
**Ch B = left**

Which ear is responding? **Left ear!**
Factors contributing to BC ipsi/contra asymmetries?

1. Infant-adult differences in positioning of neural generators

2. Greater IA compared to adults due to unfused cranial sutures

**Evidence:** infant AC ABR/ASSRs show consistent ipsi/contra asymmetries; adult AC ABR/ASSRs do not show these patterns

(Reviewed in Small & Stapells, 2017)

- Two-channel recordings are routinely used by our provincial program (BCEHP) for BC brief-tone ABRs

**NOTE:** Can also use ipsi and contra EEG channel for AC if a large difference in thresholds between ears exists (and contra masking not used)
What if ipsi/contra asymmetries in BC ABRs are ambiguous?

- Need clinical masking

Main reason masking not routinely used clinically for infant BC ABRs:
-- effective masking levels (EMLs) for BC ABR stimuli in young infants have not been measured directly

- currently under investigation in my lab (Lau, M.Sc. thesis)

- What do we know about EMLs for BC auditory evoked potentials?

-- EMLs for infant BC ASSR stimuli were estimated for 500-4000 Hz using binaural AC masking (Hansen & Small, 2012; Small, Smyth & Leon, 2014)
Recommended EMLs (dB SPL) for BC ASSR stimuli presented at 35 dB HL

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infant</td>
<td>81</td>
<td>68</td>
<td>59</td>
<td>45</td>
</tr>
<tr>
<td>Adult</td>
<td>66</td>
<td>63</td>
<td>59</td>
<td>55</td>
</tr>
</tbody>
</table>

* Significant infant minus adult EML difference (dB)

- Frequency-dependent infant-adult differences in EMLs except at 2000 Hz

(Hansen & Small, 2012; Small, Smyth & Leon, 2014)
Is there an occlusion effect (OE) in infants?

- Adults with normal hearing or a sensorineural hearing loss: occluding the ear canal results in a significant improvement in pure-tone BC thresholds.

- Do we need to correct for an OE in infants when we obtain BC thresholds with earphones in place?

We investigated this phenomenon in infants (2 studies):

(i) Small, Hatton & Stapells, 2007
   - *no occlusion effect for BC ASSR thresholds 500-4000 Hz*

(ii) Small & Hu, 2011
   - *Sound pressure ‡ in ear canal when occluded: infants >> adults*
   - % occurrence of OE (+ mean amplitude/threshold data):
     - Older infants: OE emerging at 500 & 1000 Hz
     - Young infants: OE absent at 1000 Hz (very small at 500 Hz)
Earphones in or out during BC testing?

Recommendations (conservative):
(i) Young infants: leave earphones in place
(ii) Older infants: remove earphones
Case Study
Case 2: Diagnostic ABR following “refer” from BC EHP

1st ABR

AC 2000 Hz

BC 2000 Hz – Left mastoid

<table>
<thead>
<tr>
<th>dB nHL</th>
<th>Ipsi</th>
<th>Contra</th>
</tr>
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<tbody>
<tr>
<td>30</td>
<td>✗</td>
<td>✗</td>
</tr>
<tr>
<td>30</td>
<td>✗</td>
<td>✗</td>
</tr>
<tr>
<td>30</td>
<td>✗</td>
<td>✗</td>
</tr>
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</table>

ipsi > contra
1st ABR

dB nHL

BC 500 Hz

dB nHL

20

Ipsi

Contra

20
1st ABR

**dB nHL**

- **80**: Green check mark
- **75**: Red x
- **45**: Red x

**AC 500 Hz**

**AC 4000 Hz**

- **dB nHL**: 45
What we know so far: Elevated AC & BC both ears

- **LEFT**: severe SNHL at 500 Hz (threshold @ 80 dB nHL) rising to no worse than mild/moderate SNHL at 2000 & 4000 Hz
- **RIGHT**: at least a moderate SNHL at 2000 Hz

- need further ABR testing to fill in gaps – summarized in table on next slide
3rd ABR (2nd ABR not shown)

AC 2000 Hz @ 110, 100 & 90 dB nHL

Larger amp

110

contra

ipsi

110

100

90

dB nHL

dB nHL

dB nHL
<table>
<thead>
<tr>
<th>Stimulus</th>
<th>ABR (dB nHL)</th>
<th>Estimated Behavioural Threshold (dB eHL)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RIGHT</td>
<td>LEFT</td>
</tr>
<tr>
<td>AC – 500 Hz</td>
<td>&gt; 100</td>
<td>80</td>
</tr>
<tr>
<td>BC – 500 Hz</td>
<td>&gt; 20</td>
<td>&gt; 20</td>
</tr>
<tr>
<td>AC – 1000 Hz</td>
<td>&gt; 100</td>
<td>≤ 55</td>
</tr>
<tr>
<td>AC – 2000 Hz</td>
<td>&gt; 100</td>
<td>40</td>
</tr>
<tr>
<td>BC – 2000 Hz</td>
<td>&gt; 60</td>
<td>35-60</td>
</tr>
<tr>
<td>AC – 4000 Hz</td>
<td>&gt; 90</td>
<td>25</td>
</tr>
</tbody>
</table>

+ ipsi/contra asymmetries (BC & AC) support left ear responding

1st appointment: BC ABR established nature & severity of loss L & R

2nd and 3rd appointment completed AC ABR testing:
-- L: thresholds at 500, 1000, 2000 & 4000 Hz (reverse slope SNHL)
-- R: established profound loss

MRI/CT: confirmed absence of cochlear nerve on the R (click ABR—no clear signs of ANSD)
Thank you for your attention!

Thoughts?
Case Discussion
10:15 AM to 12:00 PM

Cases 1 & 2: marked waveforms & interpretation provided
Case 3: unmarked waveforms provided (annotated version provided after discussion)

Questions to ponder when reviewing cases:
• Did the ABR assessment(s) meet all EDHI goals?
• Did you identify any issues with waveform interpretation, test sequence, elements of protocol?
• What would you have done differently (or the same)?