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ABR: An Illustration of Auditory Dysfunction through Clinical Cases

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IT'S HOW MEDICINE SHOULD BE

ABR: An Illustration of Auditory Dysfunction through Clinical Cases

Robin B. Stoner, Au.D.
Amy K. Winston, Au.D.

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Our Goal

To present real-world ABR cases from our clinic, and through these illustrations, to reinforce understanding of the auditory pathway and ABR.

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Learning Outcomes

- Describe the characteristics of a normal click-evoked ABR relative to the waveform neural generator sites
- Discuss how observed changes in the click-evoked ABR waveform can assist in characterizing the nature and degree of identified hearing loss
- Explain the effects of disruptions at specific points along the auditory pathway on the ABR waveform

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What is ABR?

- Auditory brainstem response testing (ABR) is an objective test of auditory system function that is an invaluable diagnostic tool for testing infants, young children, and difficult-to-test older children and adults.
- ABR is a test of synchronous neural function and it is not a direct test of hearing; however, it can be used to ***estimate*** hearing sensitivity.

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What is ABR?

- ABR stimuli include the broadband click stimulus (2-4 kHz) and frequency-centered toneburst stimuli (500Hz, 1000Hz, 2000Hz, 4000Hz).
- ABR stimuli are of brief duration and trigger a rapid onset of the neural impulse, click more so than toneburst.
- The ABR stimuli can be conducted by air conduction or bone conduction.

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What is ABR?

- The ABR waveform has five primary components, waves I through V, that arise from known neural generator sites along the auditory pathway from the distal portion of c.n. VIII to the lower brainstem.
- Waveform latencies from the onset of the evoking stimulus are predictable, as are the interwave latency measures.

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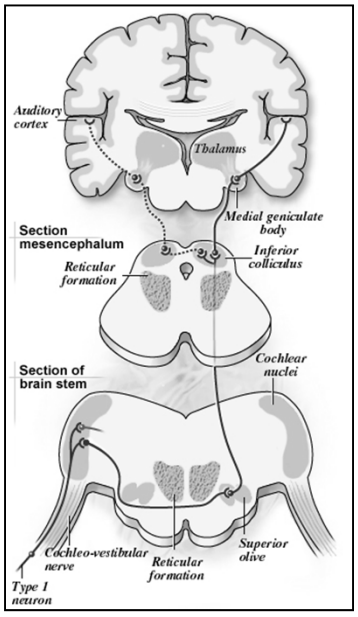
What is ABR?



Wave I: Peripheral (distal) portion of c.n. VIII **(1.5ms)**

Wave II: Central (proximal) portion of c.n. VIII **(2.5ms)**

Wave III: Cochlear nucleus **(3.5ms)**

Wave IV: Superior olivary complex/lateral lemniscus **(4.5ms)**

Wave V: Lateral lemniscus/inferior colliculus **(5.5ms)**

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What is ABR?

- Predictable changes in waveform characteristics in response to changes in auditory stimulus, including intensity and polarity, provide diagnostically significant information about the presence and type hearing loss.
  - Presence/absence of wave V to air-conducted stimuli
  - Presence/absence of wave V to bone-conducted stimuli
  - Absolute and interwave latency measures
  - Changes in waveform in response to stimulus polarity
  - Waveform morphology and amplitude

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What is ABR?

- Auditory thresholds can be estimated from the lowest intensity level where wave V is present and replicable.
- Latency information and results from bone-conducted stimuli can provide insight to the nature of the hearing loss.
- Polarity effects on the waveform can determine the presence or absence of auditory neuropathy/dyssynchrony (AN/AD).

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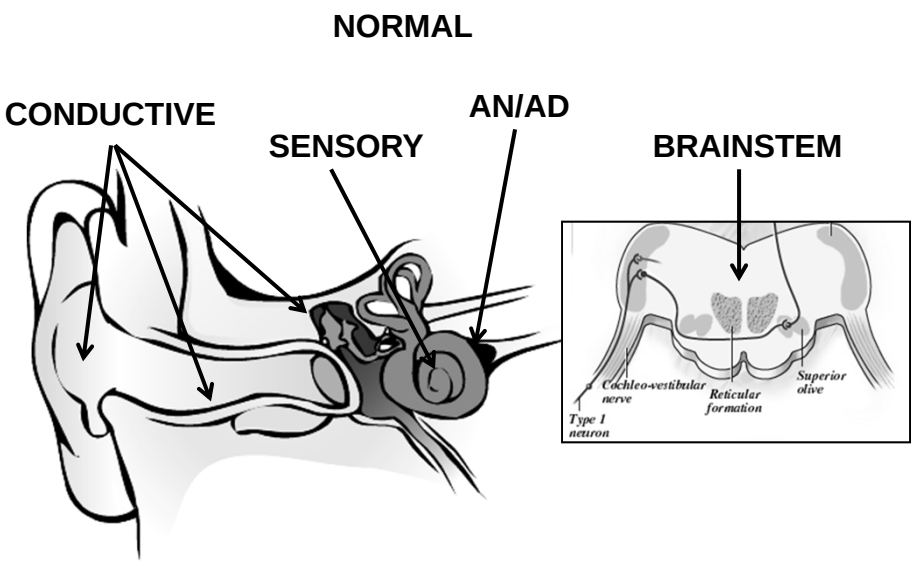
The Cases!



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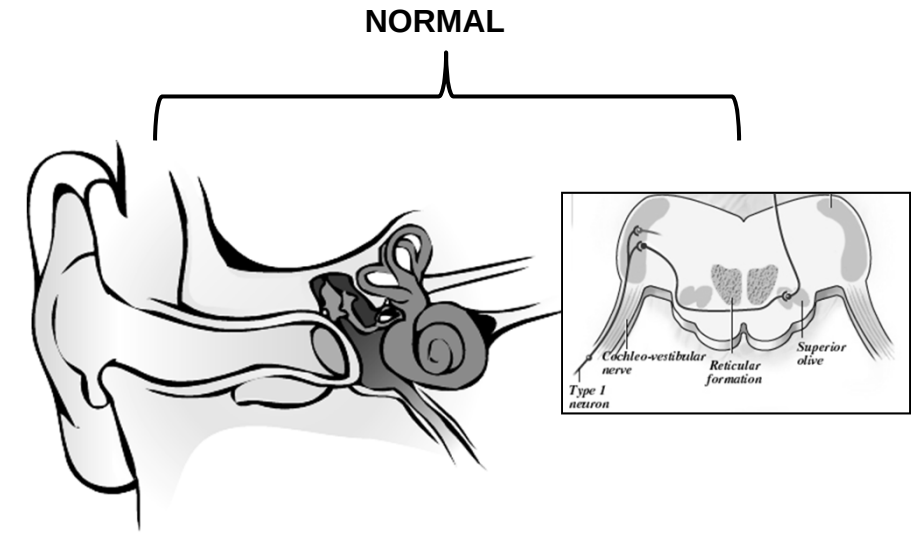
Case #1



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Case #1: History

- The patient was a three-year-old male.
- The parents stated that their son only responds to certain sounds and has a very limited expressive vocabulary.
- A psychological evaluation one month prior indicated that the patient may have a mild degree of autism.

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Case #1: History

- Pregnancy and birth histories were essentially normal.
- His parents denied a patient history of ear infections and a family history of hearing loss.

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Case #1: History

- Behavioral Audiometry (VRA): A Minimal Response Level (MRL) was noted at 30 dB HL at 2000 Hz in the sound field. SAT was obtained at 0 dB HL.
- Tympanometry (226 Hz): Normal mobility, ear canal volume, and middle ear pressure bilaterally.
- DPOAEs: CNT due to probe intolerance.
- A sedated ABR in the PICU was scheduled.

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Case #1: ABR Results

- Testing with a rarefacting and condensating air-conducted broadband click stimulus, a robust waveform was observed at 80 dBnHL; no inversion of the waveform was noted.
- Absolute wave latencies and interwave latencies were within normal limits. Wave V remained intact and replicable to 20 dBnHL in both ears.

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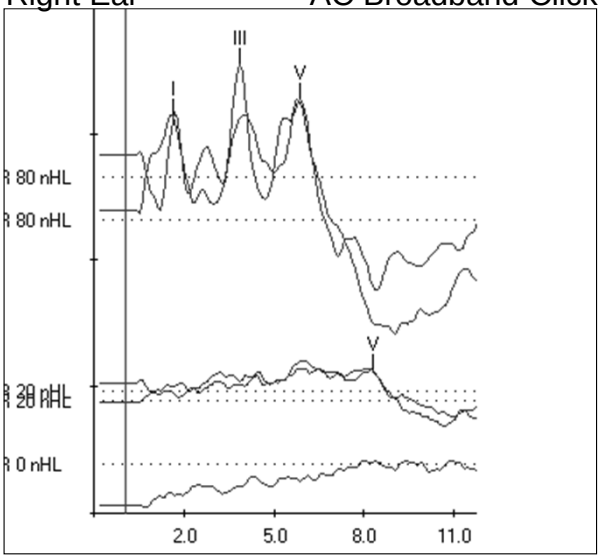
Case #1: ABR Results

Right Ear AC Broadband Click



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Case #1: Discussion

- A robust waveform that does not invert with a change in stimulus polarity rules out concern for auditory neuropathy/dyssynchrony.
- Absolute wave latencies and interwave latencies were within normal limits.
- The presence of an intact wave V down to 20 dBnHL is consistent with normal hearing sensitivity for at least a portion of the frequency range of the click stimulus (2-4kHz).

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Case #1: Discussion

- Testing with tone burst stimuli revealed results within normal limits, suggesting normal hearing sensitivity across the primary speech frequencies.
- Overall interpretation: ABR results were consistent with normal auditory system function and sensitivity within normal limits across the primary speech frequencies (500-4000 Hz).

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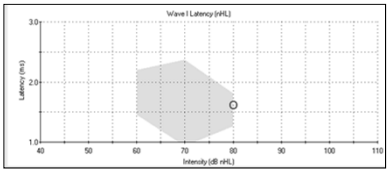
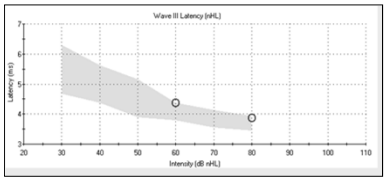
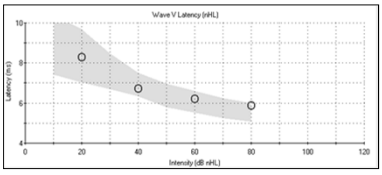
Characteristic ABR Findings

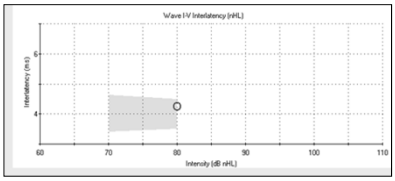
NORMAL

- Nature of Loss
  - Absolute wave I, III, and V latencies WNL
  - Interwave latencies WNL









- Degree of Loss
  - Wave V threshold WNL

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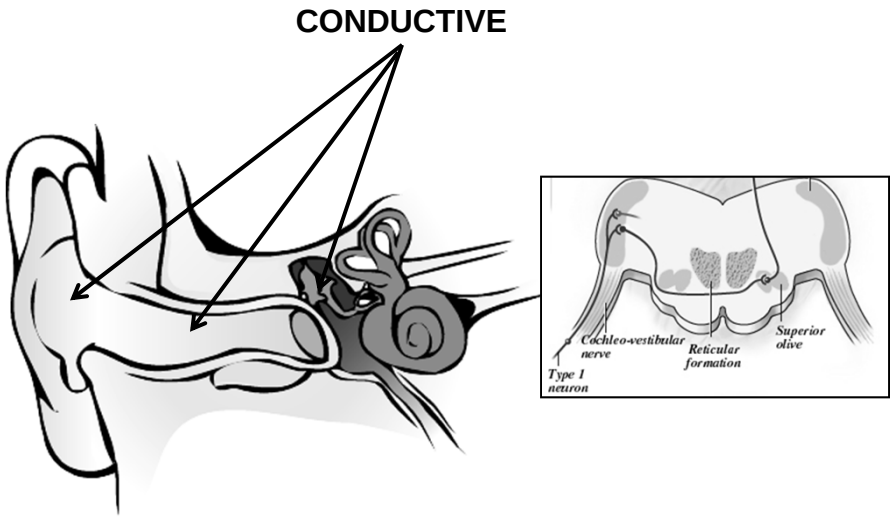
Case #2



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Case #2: History

- 3-week-old female
- Failed NBHS bilaterally
- Patient born at 36 weeks gestation; maternal health during pregnancy compromised by prenatal diabetes
- Medical history significant for bilateral microtia with left external auditory canal atresia and stenosis of the right external auditory canal

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Case #2: History

- Absence of left external auditory canal was confirmed on CT scan
- It was also noted on CT that the malleus and incus appeared to be fused bilaterally
- Other test findings:
  - Tympanometry (1000 Hz): Grossly abnormal for the right ear; CNT left ear.
  - DPOAEs (1500-6000 Hz): Absent in the right ear; CNT left ear.

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Case #2: ABR Results

Right Ear:

- Waves I, III, and V were present at a high intensity level. No inversion of the waveform was noted with a change in stimulus polarity.
- Absolute wave latencies were prolonged; interwave latencies were within normal limits.
- Wave V remained intact and replicable to 70 dBnHL.
- Testing with a 500 Hz tone burst stimulus revealed wave V intact and replicable to 60 dBnHL.

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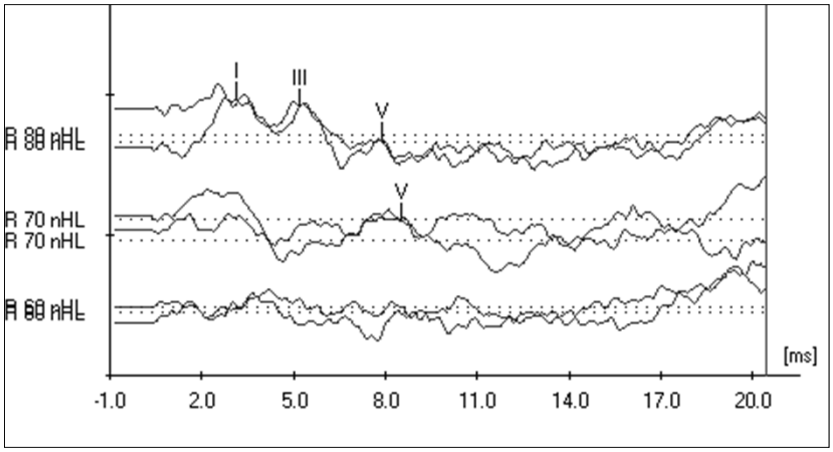
Case #2: ABR Results

Right Ear AC Broadband Click



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Case #2: ABR Results

<u>Left Ear</u>:

- Unmasked bone conduction testing with a click stimulus revealed wave V present and replicable down to 30 dBnHL.
- Bone conduction testing with a 500 Hz tone burst stimulus revealed wave V present and replicable to 25 dBnHL.
- Wave latencies were judged to be within normal limits; therefore, response is considered to be likely from the left ear. *(Hall, 1992)*

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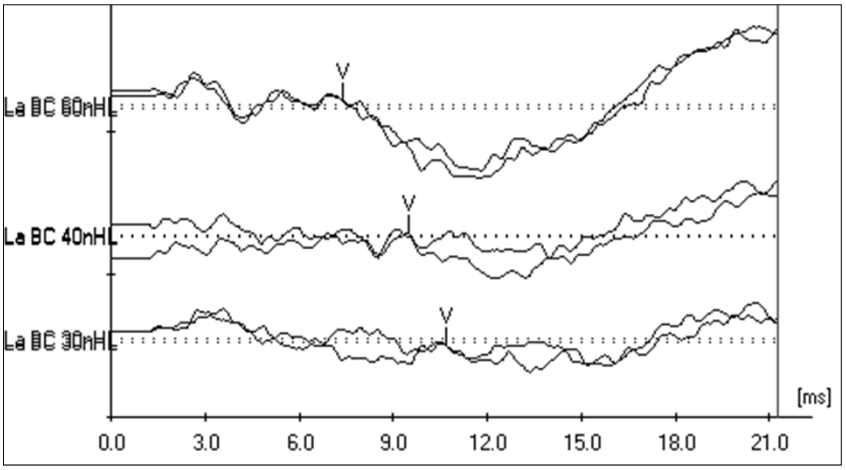
Case #2: ABR Results

Left Ear BC Broadband Click (unmasked)



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Case #2: Discussion

- Presence of prolonged absolute wave latencies with normal interwave latencies on ABR is indicative of conductive hearing loss.
- Right ear results suggest the presence of at least a moderate hearing loss across the test frequencies that is likely conductive in nature.
- Ossicular chain fusion reduces the efficiency of the middle ear transfer function, resulting in a reduction in pressure at the oval window.

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Case #2: Discussion

- Findings from left bone conduction testing suggest normal cochlear function across the test frequencies in the left ear.
- The absence of a patent external auditory canal would result in a maximum conductive hearing loss in the left ear.
- Patient ultimately fit with a Baha softband amplification device at an outside facility.

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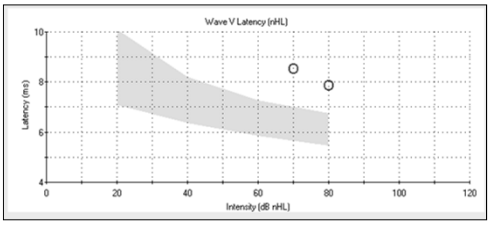
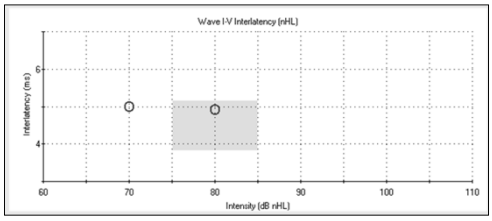
Characteristic ABR Findings

CONDUCTIVE

- Nature of Loss
  - Absolute wave latencies are prolonged
  - Interwave latencies are WNL
  - Bone conduction results are WNL
- Degree of Loss
  - AC Wave V threshold elevated





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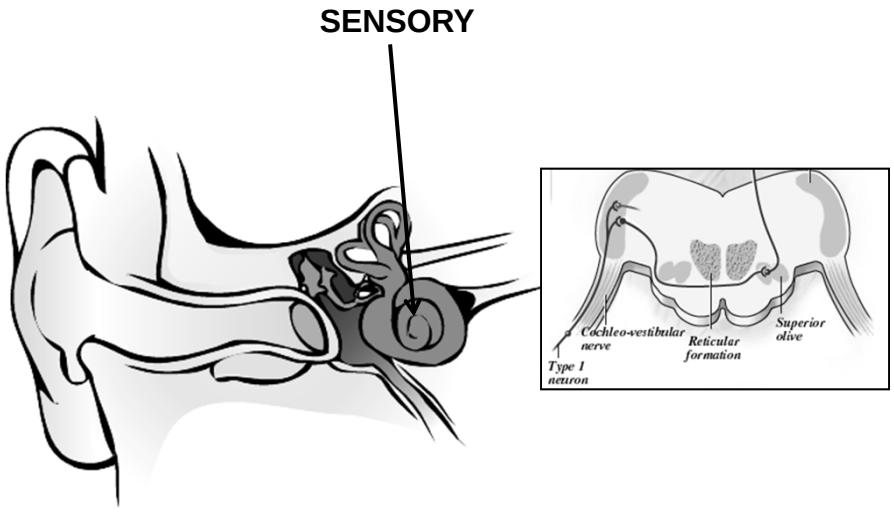
Case #3



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Case #3: History

- Nine-month-old female, referred to us due to a failed newborn hearing screening at an outside facility.
- She was born at 38 weeks gestation; maternal health during pregnancy was good.
- There were no risk factors identified, and a family history of hearing loss and patient history of ear infections were denied by the patient's mother.

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Case #3: History

- VRA: SAT was noted with fair reliability at 30 dB HL in the right ear and 50 dB in the left ear using insert earphones.
- Tympanometry (226 Hz): Normal mobility, ECV, and middle ear pressure bilaterally. Acoustic reflex screen revealed an intact reflex at 1000 Hz bilaterally.
- DPOAEs: CNT due to patient movement and noise.

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Case #3: ABR Results

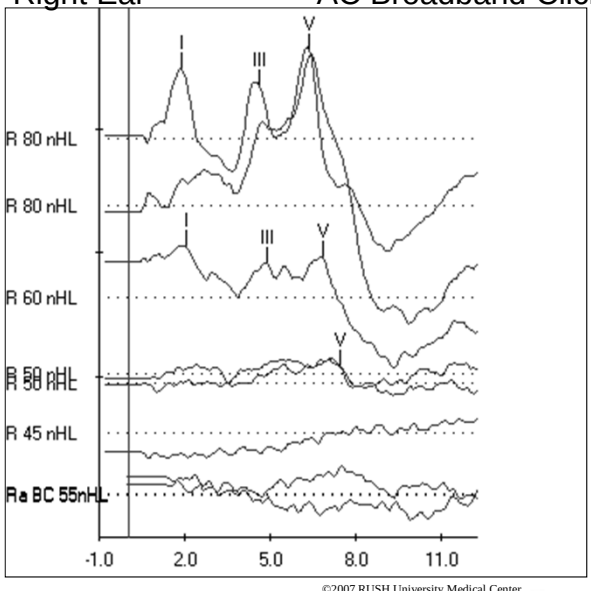
- Testing with a rarefacting and condensating air-conducted broadband click stimulus, a robust waveform was observed at 80 dBnHL.
- No inversion of the waveform was noted.
- Wave V remained intact and replicable to 50 dBnHL. No response was noted at 45 dBnHL.

Case #3: ABR Results

- Absolute wave V latency was within normal limits at a high intensity.
- Interwave latencies were within normal limits at 80 dBnHL.
- No repeatable response was noted at the output limits of the bone oscillator.

Case #3: ABR Results

Right Ear AC Broadband Click



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Case #3: Discussion

- Air conduction and bone conduction ABR results were consistent with a moderate sensory (cochlear) hearing loss for at least a portion of the frequency range of the click stimulus.
- Results using tone burst stimuli (500-4000 Hz) indicated a mild sloping to moderate sensory hearing loss.

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Case #3: Discussion

- The sensory nature of this hearing loss is supported by:

1. Normal absolute wave and interwave latencies at a high intensity,
2. Elevated wave V threshold,
3. The absence of a response to the bone-conducted stimulus.

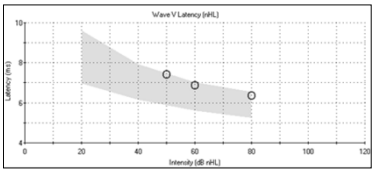
Case #3: Discussion

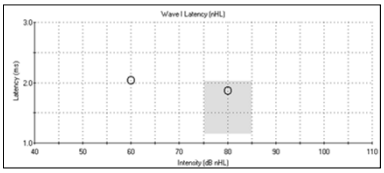
- The patient was ultimately fit with binaural amplification at an outside facility.
- Results from behavioral testing (VRA) after the ABR were consistent with the ABR results for the right ear.

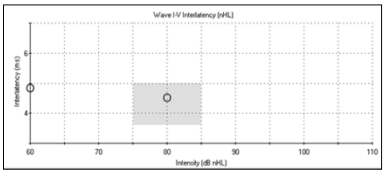
Characteristic ABR Findings

SENSORY

- Nature of Loss
  - Absolute wave I WNL or prolonged
  - Absolute wave V latency WNL at high intensities, but longer than normal at reduced intensities (L-shaped)
  - Interwave latencies WNL or shortened







- Degree of Loss
  - Wave V threshold elevated

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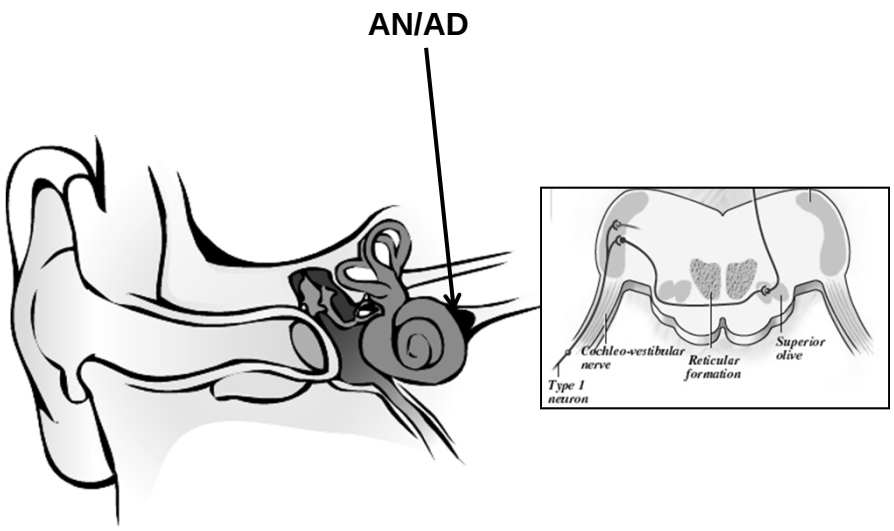
Case #4



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Case #4: History

- 3-month-old female
- Failed NBHS bilaterally at Cook County Hospital
- Medical history significant for G6PD deficiency
- G6PD is an inherited X-linked deficiency in the enzyme glucose-6-phosphate dehydrogenase.
- This defect causes red blood cells to break down prematurely, resulting in anemia. *(Genetics Home Reference, 2013)*

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Case #4: History

- G6PD deficiency can result in severe anemia and jaundice, particularly in newborns. *(Genetics Home Reference, May 2013)*
- Bilirubin in a byproduct of red blood cell breakdown.
- When present at high levels in the blood, bilirubin crosses the blood-brain barrier into the brain tissues, leading to a condition called kernicterus. (*Kernicterus and Newborn Jaundice Online)*

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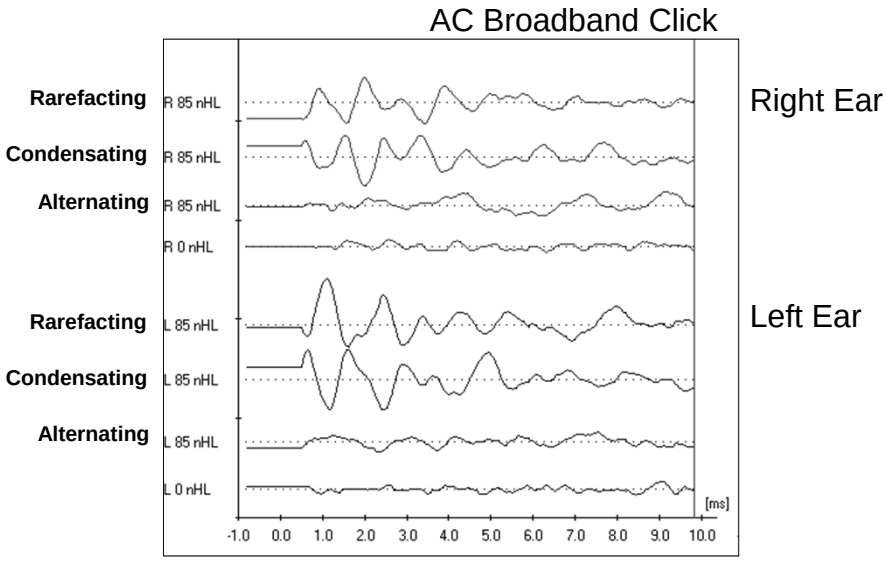
Case #4: History

- Kernicterus often includes damage to brainstem nuclei and the cerebellum, resulting in hearing loss and balance problems. (*Kernicterus and Newborn Jaundice Online)*
- When bilirubin levels are high, an exchange transfusion may be used to rapidly remove it from the blood.
- Per physician report, the patient developed severe hyperbilirubinemia that required exchange transfusion.

Case #4: ABR Results

- Other test findings:
  - Tympanometry (1000 Hz): Grossly normal bilaterally.
  - DPOAEs (1500-8000 Hz): Absent in both ears.
- Testing with rarefacting and condensating click stimuli revealed a complete inversion of the waveform in both ears.
- Testing with an alternating click stimulus eliminated the waveform entirely.

Case #4: ABR Results



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Case #4: Discussion

- The inversion of the ABR waveform in response to a change in stimulus polarity indicates that the generator site is cochlear rather than neural.
- The presence of a cochlear microphonic with no neural-based waveform components is consistent with auditory neuropathy/dyssynchrony (AN/AD)—in this case, bilateral AN/AD.
- The site of lesion for AN/AD is not definitively known. Research suggests that the issue is either presynaptic or postsynaptic. *(Picton, 2011)*

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Case #4: Discussion

- Presynaptic issues would include:
  - Abnormality in the inner hair cell (IHC)
  - Disruption between the IHC and tectorial membrane
  - Problem in the synapse between the IHC and the dendrite of the afferent nerve fiber
- Postsynaptic sites of lesion would include:
  - Abnormalities in dendrites of afferent nerve fiber or axon leading from the spiral ganglion to the brainstem
  - Loss of myelin surrounding the afferent nerve fiber

(Picton, 2011)

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Case #4: Discussion

- Hyperbilirubinemia is a known risk factor for auditory neuropathy/dyssynchrony.
- In the classic case of AN/AD, OAEs are typically present, particularly in a young child, although they may disappear over time.
- The absence of DPOAEs in this case suggests the presence of cochlear OHC dysfunction and a possible concomitant sensory loss in both ears.

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Case #4: Discussion

- Given patient's history, there is also concern for possible brain damage that has not yet been identified.
- The patient has not been seen for follow up at RUMC.

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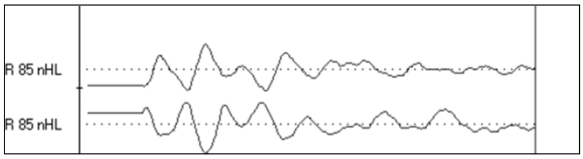
Characteristic ABR Findings

AN/AD

- Nature of Loss
  - Inversion of the waveform with reversal of stimulus polarity is indicative of AN/AD



- Degree of Loss
  - Because the response is not neural, the degree of hearing loss, if any, cannot be determined from ABR.

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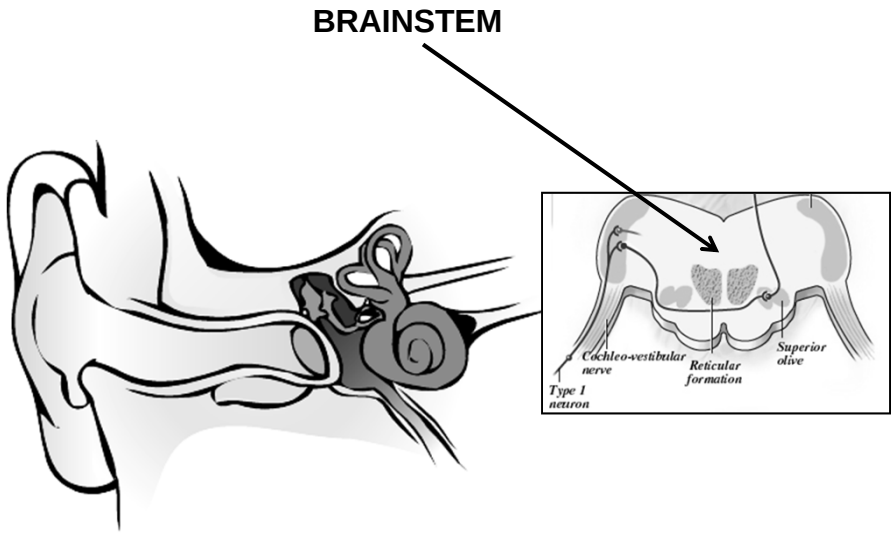
Case 5



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Case #5: History

- The patient, a two-month-old female, was seen for a natural sleep ABR due to a failed newborn hearing screening in the NICU.
- She was born at 38 weeks gestation with normal birth weight; the patient's mother was under the care of the high-risk fetal neonatal center due to a sacral defect on an ultrasound.

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Case #5: History

- Patient's medical history was significant for:
  - Myelomeningocele
  - Chiari II malformation
  - Perinatal intraventricular hemorrhage
  - VP shunt
  - G-tube
  - Central apnea

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Case #5: History

- MRI of the brain revealed no recognizable fourth ventricle.
- Per physician note, cerebellar tissue was herniated into the upper cervical spinal canal and cerebellar tissue appeared to wrap around the medulla and upper cervical cord.
- The patient has central apnea requiring long-term ventilation and trach.

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Case #5: ABR Results

- Using an air conducted click stimulus, **wave I** was clearly identified at high intensity. There was no inversion of **wave I** with a change in stimulus polarity bilaterally.
- The absolute latency of **wave I** was within normal limits at all tested intensities, and remained intact to 20 dBnHL bilaterally.
- Waves II through V were **ABSENT** at all test intensities in both ears

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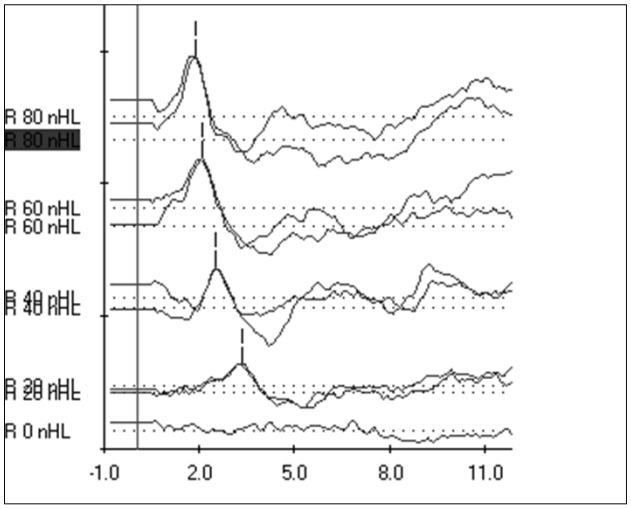
Case #5: ABR Results

Right Ear AC Broadband Click



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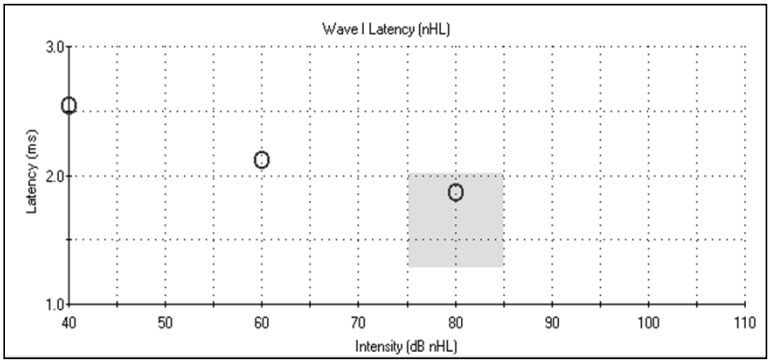
Latency Intensity Function



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Case #5: History

- Tympanometry (1000 Hz): Grossly normal bilaterally.
- OAEs: TEOAEs and DPOAEs revealed intact emissions 2000-8000 Hz bilaterally.

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Case #5: Discussion

- Myelomeningocele, a neural tube defect and the most common type of spina bifida, is a condition in which the backbone and spinal canal do not close before birth.

(Foster, Kolaski, & Riley, 2012)

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Case #5: Discussion

- Chiari II malformation is a congenital malformation of the brain, and it is associated with myelomeningocele.
- Chiari II malformation includes structural changes to the pons and 4th ventricle and downward displacement of the medulla, 4th ventricle, and cerebellum into the cervical spinal canal.

(Incesu, Khosla, & Aiello, 2011)

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Case #5: Discussion

- Neurologic examinations are typically conducted to determine the status of nerve related functions below the defect.
- Similarly, an ABR can be utilized to assess the functional integrity of the structures along the auditory pathway.

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Case #5: Discussion

- This patient's ABR revealed a clearly identified wave I only, with no clearly identifiable waves II-V in either ear.
- Wave I corresponds to recordings from the peripheral (distal) portion of cranial nerve VIII.
- Results from OAE testing are consistent with normal OHC (cochlear) function and ABR results.

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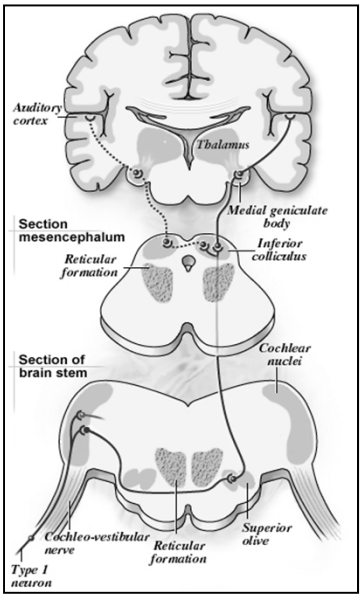
ABR Generator Sites



Wave I: Peripheral (distal) portion of c.n. VIII

Wave II: Central (proximal) portion of c.n. VIII

Wave III: Cochlear nucleus

Wave IV: Superior olivary complex/lateral lemniscus

Wave V: Lateral lemniscus/inferior colliculus

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Case #5: Discussion

- The absence of waveforms beyond wave I suggests abnormal auditory function of the central portion of the cranial nerve VIII, the cochlear nucleus, superior olivary complex, lateral lemniscus, and inferior colliculus.
- The fourth ventricle is located within the pons or in the upper part of the medulla. The superior olivary complex, lateral lemniscus and the inferior colliculus are also located in the pons.

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Case #5: Discussion

- "…gross ABR abnormalities (e.g., disappearance of wave III and/or V) were usually found in patients with mesencephalic/pontine or lower-CNS clinical signs." *(Hall, 1992)*
- Recommendations for this patient included referral to neurology, Early Intervention, ophthalmology, and possibly the use of visual and/or tactile communication strategies pending the outcome of other sensory evaluations.

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That's it folks!

Thank you for attending our presentation!

Questions?

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