Unexpected Findings:

ANSD & A CASE FOR BILATERAL COCHLEAR IMPLANTATION

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UNIVERSITY OF VIRGINIA
COCHLEAR IMPLANT PROGRAM

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The presenters have no disclosures.
Introduction
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- Cochlear Implantation
  - Decision to move toward cochlear implantation is a big decision for most families, even when candidacy is straight-forward.

- ANSD: Hearing Aids versus Cochlear Implants
  - Challenging for these families because their child “can hear”;
  - Assessing speech perception potential in a child who is developing speech and language is challenging in children who do not automatically qualify for CI candidacy based on hearing thresholds, but essential;
  - Trajectory of development
Introduction

- **ANSD and Bilateral Implantation**
  - Decisions: unilateral versus bilateral
  - Families are hesitant to take hearing away from the non-implanted ear when children have “aidable” hearing due to detection ability when CI is off;
  - Child is doing really well since implantation with one CI, how much more will two provide?
  - Much research on the benefits of binaural hearing, but can we provide objective support for this particular population?
  - Why don’t we document speech perception scores prior to second surgery, immediately after surgery but pre-activation, and with two CIs...
Max
Background
Background

• Newborn Screen: pass right side / fail left
• Audiologist #1:
  - Failed opposite side / Pass on previously failed ear.
  - Three visits with “inconclusive” results
• ENT #1:
  - “Probably just fluid”...no history of otitis media
• ENT #2:
  - Two visits: First-“inconclusive” / Second- “passed both sides”
  - Told to return for recheck and Max did not detect sound in the sound booth / sound field at 25 dB.
  - Recommend sedated ABR, but need to see 3rd ENT
Background

- ENT #3: Sedated ABR
  - “This is not what I expected to tell you...”
  - Moderate to severe hearing loss
  - “Possible neuropathic component”
- Journey Continued
  - Mom called and initiated Infant Toddler services, which took 3-4 months due to “lengthy eligibility process”
  - Family had connections within the Partnership for People with Disabilities at VCU in Richmond.
  - Connected with faculty member at Longwood University who had a background working with children with hearing loss, who subsequently referred to UVA.
Audiologic Management at UVA
Typical Test Battery

- ABR with polarity reversal
  - Cochlear Microphonic
  - ? Neural potential
- OAEs
  - Present (ME status may impact results)
- Acoustic stapedial reflexes
  - Absent/abnormal (ME status may impact results)
- Behavioral audiometric testing
  - Normal to profound HL
- Speech perception measures
- Initial Audiogram at UVA in conjunction with visit to UVA ENT

8/3/2012
SAT 65
Type B tymps – large ECV, AU
Visit with UVA ENT

- 4th ENT family has seen
- Review of records including CT (dilated jugular bulbs & dilated IACs with normal ossicles)
- Scheduled for sedated testing:
  - ABR
  - MRI
  - EKG, UA & TSH – syndromic causes of hearing loss
- Recommended SoundGene panel
Sedated ABR 8/23/12
Normal appearance of 7th & 8th cranial nerves along course through brainstem and IAC

No masses noted

Cochlea, vestibules & SSCs – normal appearance

No evidence of ELVA

Patulous/enlarged appearance of bilateral IACs without other associated abnormalities
SoundGene Results

- Connexin Panel
  - Connexin 26 (Cx26) GJB2 Mutations
    - 2 copies of the 35delG mutation were detected
- Pendred Panel: No mutation detected
- Mitochondrial Panel: No mutation detected
- Cytomegalovirus DNA Detection: Negative
- Fit with binaural Oticon Safari 300 BTEs approximately 2 weeks after sedated testing

- Wears his HAs 8+hrs/day

- No measurable improvements in speech and language despite meeting weekly with private SLP and bi-weekly with UVA AVT
- Unaided audiogram completed during scheduled HA follow-up visit.

** Note asymmetry between pure tone thresholds and SATs

- LNT & MLNT attempted Lori Bobsin, Ph.D & it was noted that he would not attempt any responses without visual cues

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4/2/13
SATs: RE: 45dB, LE: 40dB
Tymps: Flat, large ECV, AU
- CI evaluation (left HA and earmold damaged several weeks prior to CI evaluation, only R HA)

- Determination made to move forward with left-sided CI based on poorer audiometric thresholds / no usable left-sided hearing aid

4/19/13
SRT: 25dB (AR)
- 8 months post LE CI
- No right sided SRT possible even with picture board despite presentation level (up to 90dB)
- Determination made that returning a HA to the right ear would not be beneficial.
- Parents to decide between unilateral Left CI (no Right HA) or proceeding with R CI
- Family elected to proceed with R CI – surgery 4/17/14

2/11/14
SAT: 45dB (RE – no SRT possible)
Bilateral CI Audiogram

7/8/15
SRT: 20dB

7/8/15
SRT: 15dB
Aural Habilitation
## Test Results

<table>
<thead>
<tr>
<th>Date</th>
<th># phonemes correct</th>
<th>% correct</th>
<th>Device</th>
</tr>
</thead>
<tbody>
<tr>
<td>04/02/13</td>
<td>0/74</td>
<td>0%</td>
<td>hearing aids</td>
</tr>
<tr>
<td>Date</td>
<td># phonemes correct</td>
<td>Correct</td>
<td>Accuracy</td>
</tr>
<tr>
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<tr>
<td>08/27/13</td>
<td>44/74</td>
<td>60%</td>
<td>1 CI</td>
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## Lexical Neighborhood Test (LNT)

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<td>1 CI post-surgery</td>
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Conclusion

- What supports optimal speech perception outcomes after initial implantation?
  - Aid the non-implanted ear?
  - Leave non-implanted ear unaided?
  - Implant non-implanted ear?

- For those with ANSD and moderate-severe hearing loss:
  - Bilateral implantation not only provides the benefits of binaural hearing, but also...
  - Eliminates the interference of a neuropathic signal from the non-implanted ear.
Conclusion

- Best outcomes for children with ANSD, and we believe all children with hearing loss, are achieved through a collaborative effort between audiologists and speech language pathologists;

- The need for speech perception measures to document trajectory of growth is essential;

- This highlights the need for qualified professionals to work with these children.
Thank You!