Speech Perception Outcomes And Hearing Preservation Following Cochlear Implantation Of Children With Steeply-Sloping Sensorineural Hearing Loss

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Disclosures

We have the following relevant financial relationship to disclose:

We are employees of Seattle Children’s Hospital. We have no relevant nonfinancial relationships to disclose.
High Frequency Hearing Loss in Children

- Challenges with hearing aids
  - Insufficient access to:
    - Consonants
    - Higher vowel formants
  - Potential distortion from cochlear dead regions
  - Gain limitations
  - Acoustic feedback
Cohort Characteristics

- N= 8. Males = 5.
- Late identification of hearing loss
  - All passed UNHS
  - Average age of identification of HL
    43 months (24-67)
  - Frequency lowering hearing aids used prior to implant
  - SII < 47
Cohort Characteristics

- Normal anatomy
- Progressive high frequency hearing loss
  - DFNB8/10 (3)
  - Unknown (5- testing in progress)
  - 2 now have bilateral cochlear implants; 2 undergoing candidacy for 2nd side
Device and Surgical Characteristics

- 6 round window approach
- 2 traditional cochleostomy
- Full insertions without complication

Devices
- Nucleus Slim Straight CI 422 (5)
- Nucleus CI 512 (1)
- Nucleus CI 24RE (CA) (4)
Pre-Implant Thresholds by Subject

Frequency (Hz)

Hearing Level dB HL

Ear to Be Implanted

Contralateral Ear
Subject 3: Complete loss in implanted ear

3 months post implant

6 months post implant
Subject 2: Both ears progressing

3 months post implant

18 months post implant
Hearing Preservation

Post-Implant Threshold Prior to Initial Stimulation

Post-Implant Threshold At 6 Months after Stimulation

Pre-Implant Hearing Level dB HL

Post-Implant Hearing Level dB HL

- 250 Hz
- 500 Hz
- 750 Hz
- 1000 Hz
Open-Set Word Recognition as a Function of Time
Post-Implant

<table>
<thead>
<tr>
<th>Time</th>
<th>Word Recognition Score (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre</td>
<td></td>
</tr>
<tr>
<td>1 Mo</td>
<td></td>
</tr>
<tr>
<td>6 Mo</td>
<td></td>
</tr>
<tr>
<td>12 Mo</td>
<td></td>
</tr>
<tr>
<td>18 Mo</td>
<td></td>
</tr>
<tr>
<td>24 Mo</td>
<td></td>
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</tbody>
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p < 0.001
Conclusions

- Hearing preservation outcomes are variable
- Speech perception improvement is rapid and significant.