The Impact of Cochlear Implantation on Speech and Language Outcomes in Children with Asymmetric Sensorineural Hearing Loss

Prashant S. Malhotra, MD, Oliver Adunka, MD, Jaron Densky, MD, Manasa Melachuri, MS2 Samyuktha Melachuri, MS3, Amanda Onwuka, PhD, Krista Winner, AuD, Shana Lucius, CCC-SLP/AVT, Ursula Findlen, PhD
Disclosures

• Ursula Findlen, PhD- Research support from Advanced Bionics, Inc.
• Prashant Malhotra, MD- Has served on Pediatric Advisory Board, Med-El
• Oliver Adunka, MD:
  – Consultant for:
    • Advanced Bionics Corporation
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    • Spiral Therapeutics
  – Research Support:
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    • Cochlear Corporation
    • Advanced Bionics Corporation
  – Ownership:
    • Advanced Cochlear Diagnostics, LLC
NCH Hearing and Implant Program

Surgeons:
- Oliver Adunka, MD (Medical Director)
- Ed Dodson, MD
- Prashant Malhotra, MD

Pediatric Nursing/Otolaryngology:
- Kelly Brothers, CPNP
- Emily Seitz, RN

Speech Manager: Lindsey Pauline, MA, CCC-SLP

Pediatric Speech and Language Therapy:
- Janelle Huefner, M.A. CCC-SLP
- Shana Lucius, MA, CCC-SLP, LSLS Cert. AVT
- Erin Stefanik. CCC-SLP
- Lauren Wills, CCC-SLP
- Lauren Yoshihiro, M.S., CF-SLP

Neuropsychology:
- Jennifer Cass, PhD

Audiology Manager: Gina Hounam, Ph.D.-CCC-A

Pediatric Audiologists:
- Sandra Alston, AuD
- Rebecca Belt, Au.D
- Virginia Bolster, Au.D
- Nikia L. Bridges, Au.D
- Nicole Schuller, Au.D
- Ursula Findlen, PhD
- Jamie Hadley Godsey, Au.D
- Alecia Jayne, Au.D
- Devon McIlvaine Springer, Au.D
- Rebecca Matsche, Au.D
- Lauren Durinka, Au.D
- Christine Schafer, AuD
- Holly Gerth, , AuD
- Lindsey Cameron, Au.D
- Michelle Shannon, AuD
- Cindi Warner, AuD
- Krista Winner, AuD
Expanding Indications…

Significant benefit is observed for children not meeting typical candidacy:

- **Asymmetric hearing loss**
- Less severe hearing losses (*e.g.* moderate to profound)
- Partial deafness (*e.g.* ototoxic/high frequency)
- Auditory Neuropathy Spectrum Disorder
- Younger implantation, < 12 months
- **Unilateral** Profound Sensorineural Hearing Loss
- Complex medical and developmental comorbidities
Expanding Indications

Bilateral Severe to Profound

Asymmetric SNHL

SSD
Asymmetric Sensorineural Hearing Loss

Definition:

Worse ear: severe to profound ($\geq$70 dB)
Better ear: $>20$-30dB and $<60$-70dB

PTA-4
Interaural difference 30dB

Some variability in definitions
CI in ASNHL

• CI in children with asymmetric sensorineural hearing loss (SNHL) has been described
  – **Primarily audiologic outcomes** *(Tzifa 2013, Cadieux 2013)*
    • Speech Perception in the implanted ear improves (not surprising…)
    • Improved speech recognition in noise *(adults… Arndt 2017)*
    • Improved sound localization
    • Acceptance of CI is excellent *(Sadadcharam 2016)*
    • Acceptance of Acoustic + Electric signal generally high (>90%)
      o Benefit may be task dependent *(Crew et al 2015)*
  
• Little is reported about impact on **speech and language outcomes**
Our Study

• IRB approved, Retrospective review
• Free-standing, tertiary children’s hospital

Aim:

To determine if children with bilateral asymmetric sensorineural hearing loss had improvements in speech and language outcomes after cochlear implantation

− ASNHL in this study: audiometric thresholds < 70 dB HL at any frequency in the better ear and thresholds ≥ 70 dB HL at most frequencies in the poorer ear
Methods

• Inclusion criteria:
  – <18 years old
  – Implanted with CI for ASNHL in poorer ear, 2014-2017
  – Maintained HA in better ear

• Excluded:
  – Single Sided Deafness
  – Auditory Neuropathy or cochlear nerve disorder in either ear
  – Multiply involved or cognitively impaired (other reasons for delays)
  – Poor CI use after surgery
  – Non-native English speakers
Outcomes

• Preoperative and postoperative, ear-specific audiometry
• (Speech Perception)
• Speech and language outcomes (every 6 months)
  – When available, measures of **speech articulation, expressive and receptive language, and vocabulary**
    • Goldman-Fristoe Test of Articulation (GFTA-3)
    • Clinical Evaluation of Language Fundamentals (CELF-P or CELF-5)
    • Receptive-Expressive Emergent Language Test-Third Edition (REEL-3) battery
    • Receptive One-Word Picture Vocabulary Test (REWOPVT)
    • Expressive One Word Picture Vocabulary Test (EOWPVT)
## Results

### Demographic Characteristics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N=26</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Length of follow up in months (Mean, SD)</strong></td>
<td>30.1</td>
<td>11.8</td>
</tr>
<tr>
<td><strong>Female</strong></td>
<td>10</td>
<td>38%</td>
</tr>
<tr>
<td><strong>Etiology</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bilateral EVA</td>
<td>7</td>
<td>27%</td>
</tr>
<tr>
<td>Bilateral EVA/Pendred’s</td>
<td>4</td>
<td>15%</td>
</tr>
<tr>
<td>Congenital CMV</td>
<td>2</td>
<td>8%</td>
</tr>
<tr>
<td>Connexin</td>
<td>2</td>
<td>8%</td>
</tr>
<tr>
<td>DFNB59 gene</td>
<td>1</td>
<td>4%</td>
</tr>
<tr>
<td>Stroke/meningitis</td>
<td>1</td>
<td>4%</td>
</tr>
<tr>
<td>Unknown</td>
<td>8</td>
<td>31%</td>
</tr>
<tr>
<td>Usher/Von Willebrand</td>
<td>1</td>
<td>4%</td>
</tr>
<tr>
<td><strong>Age at Cochlear Implantation (Mean, SD)</strong></td>
<td>7.9</td>
<td>4.6</td>
</tr>
<tr>
<td><strong>Implanted Ear: Right</strong></td>
<td>13</td>
<td>50%</td>
</tr>
<tr>
<td><strong>Manufacturer</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>AB</td>
<td>8</td>
<td>31%</td>
</tr>
<tr>
<td>Cochlear</td>
<td>14</td>
<td>54%</td>
</tr>
<tr>
<td>Med El</td>
<td>4</td>
<td>15%</td>
</tr>
<tr>
<td><strong>Progression of 2nd ear to CI</strong></td>
<td>8</td>
<td>31%</td>
</tr>
</tbody>
</table>

### Etiology

- **EVA** 27%
- **EVA/Pendred’s** 15%
- **Unknown** 31%
- **Congenital CMV** 7%
- **DFNB59 gene** 4%
- **Connexin** 8%
- **Stroke/Meningitis** 4%
- **Usher/Von Willebrand** 4%
Unaided PTA-4 at time of CI:
   Better ear: 62dB HL
   Worse ear: 92dB HL
# Preoperative Speech and Language Deficits

<table>
<thead>
<tr>
<th>Main Speech Indication for Surgery</th>
<th>N=26</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Audition</td>
<td>10</td>
<td>38%</td>
</tr>
<tr>
<td>Articulation</td>
<td>14</td>
<td>54%</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>15</td>
<td>58%</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>17</td>
<td>65%</td>
</tr>
<tr>
<td>High Frequency access (i.e plurals)</td>
<td>2</td>
<td>8%</td>
</tr>
<tr>
<td>None</td>
<td>2</td>
<td>8%</td>
</tr>
<tr>
<td>Auditory Closure</td>
<td>1</td>
<td>4%</td>
</tr>
<tr>
<td>Global Issues</td>
<td>1</td>
<td>4%</td>
</tr>
</tbody>
</table>
Speech and Language Evaluations
Same Test Preop => Postop

• (Speech perception improved, not surprisingly)
• N = 13/14 (96%) who had same test, improved

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>Standard/Scaled Score</th>
<th>Difference in Standard Score</th>
<th>Time of Eval after Surgery (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Goldman Fristoe -2</strong></td>
<td>5</td>
<td>74.8 ± 21.9</td>
<td>11.2 ± 8.0</td>
<td>14.8 ± 5.4</td>
</tr>
<tr>
<td><strong>Receptive One Word Picture Vocabulary</strong></td>
<td>5</td>
<td>85.0 ± 16.3</td>
<td>2.8 ± 7.9</td>
<td>15.6 ± 10.5</td>
</tr>
<tr>
<td><strong>Expressive One Word Picture Vocabulary</strong></td>
<td>4</td>
<td>87.8 ± 20.6</td>
<td>4.5 ± 2.9</td>
<td>13.5 ± 5.7</td>
</tr>
<tr>
<td><strong>CELF P-2 - Core Language</strong></td>
<td>4</td>
<td>73.0 ± 16.3</td>
<td>10.3 ± 17.9</td>
<td>13.3 ± 5.9</td>
</tr>
<tr>
<td><strong>CELF 5 - Core Language</strong></td>
<td>7</td>
<td>67.6 ± 15.1</td>
<td>7.0 ± 11.3</td>
<td>8.3 ± 4.7</td>
</tr>
</tbody>
</table>
Speech and Language – Change Over Time
Articulation and Core language
Speech and Language – Change Over Time

Vocabulary

Changes in Receptive One Word Picture Vocab Over Time (n=5)

Changes in Expressive One Word Picture Vocab Over Time (n=4)
Limitations

• Retrospective
  – biased towards poor performers?
• Inconsistent follow up and timing of speech and language re-evaluations
• Small number
Conclusions

- Age of CI is later (7.8 years) in ASNHL
- 42% of kids implanted had bilateral EVA EVA/Pendred’s
- Children had most difficulty preoperatively with *audition*, *articulation*, *receptive language*, *expressive language*
- After CI, most improvement seen in Goldman-Fristoe 2 (articulation), CELF P2, and CELF 5 Core language tests
Clinical Implications

• Children with bilateral ASNHL **do** suffer from speech and language deficits
• Improvement in speech and language measures can be demonstrated in children undergoing unilateral cochlear implantation for asymmetric sensorineural hearing losses.
• These children, who are not typical CI candidates, can benefit from a CI in the poorer ear.
Thank You!