Hearing at Birth to Cochlear Implantation
Progressive Hearing Loss

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Financial Disclosures

- Katie Sawaya
  - None
- Stacy Payne
- Steve Andreoli
  - None
- Drew Horlbeck
  - None
Introduction – progressive hearing loss

- The incidence of progressive or late-onset hearing loss (HL) in children varies from 4-30% \(^1-^3\).

- Risk factors attributed include (but not limited too): family history, syndromic hearing loss, in-utero infections, NICU stay over 5 days, and postnatal infections\(^2,^4\).

- Studies indicate that the prevalence of hearing loss increases in school aged children, compared to children failing their newborn hearing screening \(^5-^6\).
Introduction – progressive and cochlear implants

- Children who are delayed in receiving a cochlear implant are 52% more likely to have a progressive HL
  - Children who are implanted early or have residual hearing outperform their later implanted peers
- FDA guidelines are stricter for candidacy between ages 12-24 months: therefore children who have residual hearing are not always straightforward candidates
  - Pediatric: Hearing loss greater than 90dB and Less than 20-30% on simple open-set (MLNT, LNT, HINT)
  - Adult: Hearing loss greater than 70dB and score less than 40-60% on open-set sentence testing
- Limited data available on performance of children with progressive hearing loss implanted later in life
AIM

The purpose of this study is to review audiometric outcomes in children who passed newborn hearing screening and were later identified with hearing loss that ultimately required cochlear implantation.

We compared children who were implanted within 12 months of identifying their hearing loss to those who were implanted after 12 months.
Methods

- Retrospective review of CI patients at a tertiary care children's hospital from 2000-2016
  - Inclusion criteria:
    - Documented passed newborn hearing screening
    - Received a cochlear implant
  - Exclusion criteria:
    - Sudden SNHL (Profound in nature)
    - Failed NBHS
    - No information re: NBHS
    - No post implantation audiological testing information available
Methods

- Data collected:
  - Demographics
  - Hearing screening results
  - Diagnosis age
  - Etiology of hearing loss
  - Time to implantation
  - Cochlear implant Speech Reception and discrimination

- Unpaired T-test performed on Speech reception and discrimination
Results - Demographics

- 30/56 Children with progressive hearing loss and cochlear implants were included with a prevalence of 10% among CI patients

<table>
<thead>
<tr>
<th>Gender</th>
<th>N</th>
<th>Range</th>
<th>MEAN ±SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>15</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>15</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>30</td>
<td>7-17 years</td>
<td>11.5 ± 2.8</td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Caucasian</td>
<td>19</td>
<td></td>
<td></td>
</tr>
<tr>
<td>African/American</td>
<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spanish/Latino</td>
<td>5</td>
<td></td>
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</table>
## Results - Etiology

<table>
<thead>
<tr>
<th>Etiologies</th>
<th>NUMBER OF CHILDREN</th>
</tr>
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<tbody>
<tr>
<td>Unknown</td>
<td>9</td>
</tr>
<tr>
<td>Auditory Neuropathy Spectrum Disorder</td>
<td>4</td>
</tr>
<tr>
<td>NICU</td>
<td>4</td>
</tr>
<tr>
<td>Enlarged Vestibular Aqueduct</td>
<td>4</td>
</tr>
<tr>
<td>Family History of hearing loss</td>
<td>3</td>
</tr>
<tr>
<td>Noonan Syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Failure to Thrive</td>
<td>2</td>
</tr>
<tr>
<td>Mondini/dysmorphic cochlea</td>
<td>2</td>
</tr>
<tr>
<td>Microcephaly</td>
<td>1</td>
</tr>
</tbody>
</table>
Results

- Average age for Diagnosis was 2.35 ± 1.5 years

- 43.3% children received their implant within 12 months of HL diagnosis
  - 92% of these children were identified with a severe-profound HL at the time of diagnosis

- 56.7% children were implanted more than 12 months after diagnosis with a mean lag time of 3.11 ± 1.87 years
  - 25% of this population were found to have severe to profound HL at time of diagnosis
Results – speech outcomes

- The speech reception thresholds in the early and late implanted groups were comparable at 16.9 ± 2.5dB and 17.9 ± 3.6 dB respectively (p>0.05).

- Speech discrimination results from the early and late implantation show a trend towards improved results with early implantation.

<table>
<thead>
<tr>
<th></th>
<th>&lt;12 months</th>
<th>&gt;12 months</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>13</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>SRT (dBHL)</td>
<td>16.92 ± 2.5</td>
<td>17.94 ± 3.6</td>
<td>P &gt; 0.05</td>
</tr>
<tr>
<td>Discrimination</td>
<td>87.2 ± 13.4</td>
<td>77.37 ± 14.0</td>
<td>P = 0.06</td>
</tr>
</tbody>
</table>
Conclusions

- Children with progressive hearing loss that leads to cochlear implantation trend towards performing better on auditory speech testing when receiving their implants within 12 months of diagnosis however further investigation is needed.

- Limited by
  - Florida state NBHS database limited to 2005 for verifying documented NBHS
  - Retrospective cohort – limited by past test results available

- Future pursuits
  - Review speech and language testing pre- and post-implants for progressive hearing loss
  - Review preservation of hearing in these children – if their pre-implant thresholds are similar to post-implant
  - Comparison to congenitally deaf children that have received cochlear implants
References