Outcomes of Cochlear Implantation in Children with CHARGE Syndrome

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Authors & Disclosures

• Meredith Holcomb, AuD
  – Advanced Bionics Audiology Advisory Board
  – Med El Corporation: 2 Research Grants

• Ted Meyer, MD, PhD
  – President Elect, National AG Bell

• David White, MD
  – Med El Pediatric Surgical Advisory Board

• Ted McRackan, MD
Objective

• To describe the surgical, audiometric, and language outcomes of cochlear implantation in children with CHARGE syndrome
Study Design / Methods

• IRB approved retrospective chart review of children with CHARGE syndrome who received a cochlear implant at the Medical University of South Carolina between 2001-2016

• Data analyzed:
  – Diagnostic ABR results
  – MRI and/or CT scan results
  – Pre-operative audiogram
  – Intra-operative ECAP (NRT/NRI/ART)
  – Post-operative ECAP (NRT/NRI/ART)
  – Post-operative CI audiogram
  – Language outcomes

• 5 children (10 ears) identified for inclusion in this study
### Subject #1

<table>
<thead>
<tr>
<th>Ear</th>
<th>ABR</th>
<th>Audio</th>
<th>Aided Audio</th>
<th>Abnormal CT</th>
<th>MRI</th>
<th>CI</th>
<th>OR NRT</th>
<th>Post-op NRT</th>
<th>Post-op Audio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td></td>
<td>Deficient 8th</td>
<td>X</td>
<td>NR</td>
</tr>
</tbody>
</table>

Age at CI: 22 mos  
Device: CA N5  
Surgical Complications: None  
CI Outcome: Non-user  
Language Outcome: ASL
# Subject #2

<table>
<thead>
<tr>
<th>Ear</th>
<th>ABR</th>
<th>Audio</th>
<th>Aided Audio</th>
<th>Abnormal CT</th>
<th>MRI</th>
<th>CI</th>
<th>OR NRI</th>
<th>Post-op NRI</th>
<th>Post-op Audio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right</td>
<td>90 dB @4kHz</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td>Deficient 8th</td>
<td>X</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>Left</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td>Absent 8th</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Age at CI: 51 mos  
Device: AB HR90k  
Surgical Complications: Yes  
CI Outcome: Consistent User  
Language Outcome: ASL (Autism/ADHD)
# Subject #3

<table>
<thead>
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<th>ABR</th>
<th>Audio</th>
<th>Aided Audio</th>
<th>Abnormal CT</th>
<th>MRI</th>
<th>CI</th>
<th>OR ART</th>
<th>Post-op ART</th>
<th>Post-op Audio</th>
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</thead>
<tbody>
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<td>Right</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td>X</td>
<td>NR</td>
<td>NR</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Deficient 8th</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td>Absent 8th</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

Age at CI: 21 mos  
Device: ME Concert Flex 24  
Surgical Complications: None  
CI Outcome: ?  
Language Outcome: ?
# Subject #4

<table>
<thead>
<tr>
<th>Ear</th>
<th>ABR</th>
<th>Audio</th>
<th>Aided Audio</th>
<th>Abnormal CT</th>
<th>MRI</th>
<th>CI</th>
<th>OR NRT</th>
<th>Post-op NRT</th>
<th>Post-op Audio</th>
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</thead>
<tbody>
<tr>
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<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>X</td>
<td></td>
<td>X</td>
<td>NR</td>
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<td>Left</td>
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<td>NR</td>
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<td>X</td>
<td>Deficient 8th</td>
<td>X</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

Age at CI: 27 mos  
Device: CA CI24RE-CA  
Surgical Complications: None  
CI Outcome: Non-user  
Language Outcome: ASL
### Subject #5

<table>
<thead>
<tr>
<th>Ear</th>
<th>ABR</th>
<th>Audio</th>
<th>Aided Audio</th>
<th>Abnormal CT</th>
<th>MRI</th>
<th>CI</th>
<th>OR NRT</th>
<th>Post-op NRT</th>
<th>Post-op Audio</th>
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</thead>
<tbody>
<tr>
<td>Right</td>
<td>Mod to prof SNHL</td>
<td>Mod to prof SNHL</td>
<td>20-60 dB HL</td>
<td>X</td>
<td>Present 8th</td>
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<td>present</td>
<td>15-20 dB HL</td>
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<td>X</td>
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</tbody>
</table>

Age at CI: 60 mos  
Device: CA CI24RE-CA  
Surgical Complications: None  
CI Outcome: Consistent user  
Language Outcome: Oral
Overall Results

• 8 of 10 total ears (80%) –
  – No response on ABR testing
  – hypoplastic or absent auditory nerves on MRI scans
  – Of these, 4/4 chosen for implantation did not stimulate with the CI

• Full insertion of internal CI device was achieved in all five implantations, although 1 was complicated.
Overall Results

• 4 of the 5 patients (80%) used ASL as their main mode of communication, and one child (20%) was an oral communicator with speech and language scores in the normal range.

• 2 patients are non-users of CI, 2 patients are consistent users, and 1 is lost to follow-up.
  – For the non-users, pre-op ABR was no response, MRI revealed hypoplastic or absent auditory nerves, and intra-operative and post-operative ECAP responses were absent.
Conclusions

• Cochlear implantation is an appropriate option of treatment of profound SNHL in children with CHARGE syndrome.
  – MRI is necessary to evaluate status of auditory nerves for this population.
  – Outcomes limited by status of cochlear nerve.

• A no response ABR may predict poor outcomes with CI for children with CHARGE syndrome.
Thank You!