Auditory Brainstem Implantation for Children with Cochlear Nerve Deficiency

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Cochlear Nerve “Aplasia”

MRI Evidence of Cochlear Nerve Deficiency

Cochlear Implant or Not?
Speech Perception (SRI-Q) by Malformation

Buchman et al. *Laryngoscope* 2011
Auditory Brainstem Implant (ABI)

Possible Indications

• Absent Cochlea or Cochlear Nerves
  » NF2
  » Congenital absence (Colletti et al 2002, ……)
  » Total ossification
  » Traumatic transection

• Unable to or failed benefit from CI
  » Severe malformations, progressive ossification, other?

• Committed Parents or Patients
• Cognitively normal or near normal
Auditory Brainstem Implant
ABI Device

- Developed by William House
- Similar to Cochlear Implant
- Foramen of Luschka
- Removable Magnet
FDA Approved Protocol

- Investigator Initiated Investigational Device Exemption (IDEs)
- IRB Approval
- Safety and Feasibility Study (N=10 children)
- Inclusion Criteria
  - Indications:
    - Cochlear Nerve Deficiency+/-severe inner ear malformation
    - Post-meningitis ossification (PMO)
  - Previous CI (if possible) → No benefit
  - Cognitively Normal
  - Good Parents
  - Reasonable expectations
- Outcomes
  - Surgical Complications
  - Sound detection, Speech perception, Speech Production, Language
Protocol

• Retrosigmoid Craniotomy
  » Nucleus 24 ABI (Cochlear Corp)
  » Monitor CN 7, 9, 10, 11
  » Implant evoked ABR
• Postop CT
• Pediatric ICU
• OR Stim prior to activate
• Activation under monitoring
• Speech perception hierarchy similar to cochlear implantation
  » IT-MAIS, ESP, PBK
Postoperative CT Scans
## Demographics

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<thead>
<tr>
<th></th>
<th>UNC1</th>
<th>UNC2</th>
<th>UNC3</th>
<th>UNC4</th>
<th>UNC5</th>
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<tr>
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<tr>
<td>Age at ABI</td>
<td>3.33</td>
<td>2.50</td>
<td>3.50</td>
<td>5.50</td>
<td>2.17</td>
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<tr>
<td>Gender</td>
<td>M</td>
<td>F</td>
<td>M</td>
<td>F</td>
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<tr>
<td>Side</td>
<td>L</td>
<td>L</td>
<td>R</td>
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<td>Etiology</td>
<td>CND</td>
<td>CND-CC</td>
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<tr>
<td>Syndrome</td>
<td>CHARGE</td>
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1/15/15
## Results

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<tr>
<td><strong>eABR +</strong></td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
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<tr>
<td><strong>Non-Auditory OR</strong></td>
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<td>No</td>
<td>No</td>
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<tr>
<td><strong>Active Electrodes</strong></td>
<td>9</td>
<td>11</td>
<td>7</td>
<td>13</td>
<td>15</td>
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<td><strong>Non-Auditory PO</strong></td>
<td>Cough</td>
<td>Swallow</td>
<td>No</td>
<td>Vestibular</td>
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<td><strong>Complications</strong></td>
<td>CSF Leak</td>
<td>Aseptic Meningitis</td>
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<tr>
<td><strong>Sequelae</strong></td>
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## Performance

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<tbody>
<tr>
<td><strong>Age at Implant (yr)</strong></td>
<td>3.33</td>
<td>2.50</td>
<td>3.50</td>
<td>5.50</td>
<td>2.17</td>
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<tr>
<td><strong>Duration of Use (yr)</strong></td>
<td>1.62</td>
<td>1.34</td>
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<td><strong>PTA</strong></td>
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<td><strong>SDT</strong></td>
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<td>80</td>
<td>40</td>
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<td><strong>IT-MAIS</strong></td>
<td>43%</td>
<td>3%</td>
<td>30%</td>
<td>50%</td>
<td>3%</td>
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<td><strong>ESP-W</strong></td>
<td>58%</td>
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<td>NA</td>
<td>50</td>
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Conclusions

• ABI in Young Children is in very early stages in US

• Safe so far
  » CSF Leaks
  » Aseptic meningitis

• Early results
  » Sound detection in all
    • 1 with poor detection → good location, no cochleas
  » Limited speech perception thus far (very early)

• Objective Measures