Auditory Brainstem Implantation in Children

Craig Buchman, MD
Matthew G. Ewend, MD
Holly F.B. Teagle, AuD
Lillian Henderson, SLP
Shuman He, PhD
John Grose, PhD
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  • NIH/NIDCD
  • Cochlear Corporation

ABI is investigational under an IDE from the US FDA
Pediatric Hearing Loss - The Problem

- Estimates
  - 3-4/1000 have HL (450-500 children/yr)
  - 1:1000 have severe to profound HL (~120 children in NC in 2011)

- Impact
  - Sound awareness
  - Speech understanding and language development
  - Educational Impact
  - Employment opportunities and Earning potential
  - ~$1 million per child lifetime costs \textit{when untreated}
    - 35% direct and 65% indirect (lost earning potential, ...
Criteria for Cochlear Implantation in Children

- Severe to profound SNHL
- Limited benefit from hearing aids
- No active middle ear pathology
- Normal eighth nerve and present cochlea
Factors that Delay Implantation and Outcome

• Auditory
  • Delay in diagnosis
  • Significant residual hearing
  • Fluctuating hearing
  • Unreliable or conflicting test results
  • ANSD
  • Underfit amplification

• Speech development
  • Good progress despite profound HL

• Parental issues
  • Missed appointments
  • Don’t wear devices
  • No educational buy-in
  • Socioeconomic

• Medical
  • Anatomic uncertainty
    • CN deficiency
    • Severe inner ear malformation
  • Multiple Challenges
    • Cerebral palsy
    • Autism
    • Other
Inner Ear Malformations
Cochlear Nerve “Aplasia”

MRI Evidence of Cochlear Nerve Deficiency

Right Ear

Left Ear

Cochlear Nerve Deficiency

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

Buchman et al *Laryngoscope* 2011

**Figure:**
- **Axes:** Duration of Use (Months) vs. Speech Perception Score
- **Data Points:** CND, IP-EVA, Hypoplastic, CCVA, CC, SCC
- **Legend:**
  - CND
  - IP-EVA
  - Hypoplastic
  - CCVA
  - CC
  - SCC

**Source:**

**Date:** 10/13/2016
Auditory Brainstem Implant (ABI)

Possible Indications

• Absent Cochlea or Cochlear Nerves
  • NF2
  • Congenital absence
  • 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
UNC Pediatric ABI Feasibility Study

- Safety and efficacy of the Nucleus 24 Multichannel ABI:
  - to demonstrate safety of the surgical procedure
  - tolerance of device stimulation, and
  - the potential for auditory benefit beyond that experienced with their CI
- May provide the preliminary experience for a larger scale clinical trial
- Requires a team approach among surgeons, audiologists, speech/language pathologists & electrophysiologists and families
- Investigational Device Exemption from the FDA
- Institutional Review Board Approval at UNC-CH
UNC Pediatric ABI Feasibility Study

Candidates

• Subjects
  • 10 pre-linguistic young children (18 mos to 5 yrs. of age)
  • 5 post-linguistic children (<18 yrs of age)
• Failed CI OR unable to receive a CI
• No developmental/cognitive delays that would impede progress
• Appropriately motivated family
ABI Team

Surgery

- Surgery, Medical management, Assessment, Oversee study

Speech/Language Pathology

- Assessment, Therapy, Local service contact

Electrophysiology

- Device programming
- Assessment, Study coordination
- Intra & Post operative testing

Audiology

- Local service contact
Retrosigmoid Craniotomy
Retrosigmoid Craniotomy
Retrosigmoid Craniotomy
Retrosigmoid View

- **Intraop maneuvers:**
  - Cranial opening to sinus
  - Drain cisterna magna
  - Retract cerebellum
  - Widely open arachnoid
  - Identify lower CN (9-11)
  - Identify (7, 8)
  - Move vessels
  - Retract choroid plexus
  - Retract flocculus
  - Enter Foramen of Luschka
Retrosigmoid View

• Intraop maneuvers:
  • Cranial opening to sinus
  • Drain cisterna magna
  • Retract cerebellum
  • Widely open arachnoid
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  • Identify (7, 8)
  • Move vessels
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  • Enter Foramen of Luschka
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
- Ongoing device programming
- Speech perception/Speech & Language Assessments similar to cochlear implantation
Postoperative CT Scans
Intraop Electrically-Evoked ABR

Amplitude (0.5 μV/div)

Latency (mSec)

15-21

21-15

3-9

9-3

10/13/2016 27
Audiology: Device Programming

Goal is similar to CI patient programming

- Establish electrical threshold (T) and comfort (C) levels
- Allocation based on array design
  - Creating the auditory template that will develop with time
- Avoid stimulation of other cranial nerves (initial stim in OR)
  - Facial twitching,
  - Balance disturbance (head & trunk tilt),
  - Coughing, choking, sensation in mouth, throat, tongue, palate,
  - Heart rate changes
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<tr>
<th></th>
<th>UNC1</th>
<th>UNC2</th>
<th>UNC3</th>
<th>UNC4</th>
<th>UNC5</th>
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<td>Previous/Current CI</td>
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<td>No/No</td>
<td>No/No</td>
<td>Yes/Yes</td>
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<tr>
<td>Age at ABI</td>
<td>3.3</td>
<td>2.5</td>
<td>3.5</td>
<td>5.5</td>
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<td>F</td>
<td>M</td>
<td>F</td>
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<td>Side</td>
<td>L</td>
<td>L</td>
<td>R</td>
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<td>Etiology/Syndrome</td>
<td>CND/</td>
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<tr>
<td></td>
<td>CHARGE</td>
<td>Michel</td>
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Case 1

- Dx at 11 mos with HL
- CHARGE Syndrome
- Previous CI use
- Cued Speech
Case 2

- Identified at birth
- Profound HL
- Absent cochlea and cochlear nerve
- No previous CI
- SEE

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Case 3

- CHARGE syndrome
- Cleft lip & palate (repaired)
- No previous CI
- Total Communication/SEE
Case 4

- CIs at 10 & 13 months
- Received sound detection with CIs
- Total Communication/SEE
Case 5

- CI at 15 mos – no sound awareness
- ASL
Surgery Results
# Listening Skills and Speech Production

<table>
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<tr>
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<th>UNC2</th>
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<td><strong>Duration of Use</strong></td>
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<td>2.5</td>
<td>2</td>
<td>18</td>
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<td><strong>Communication mode/Speech Production</strong></td>
<td>Cued Speech 92% Word Patterns 53% Vowels</td>
<td>SEE 55%Word Patterns 29% Vowels</td>
<td>TC CNT</td>
<td>TC 48% Word Patterns 17% Vowels</td>
<td>ASL CNT</td>
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<tr>
<td><strong>OWLS Composite (85-115)</strong></td>
<td>71</td>
<td>71</td>
<td>77</td>
<td>44</td>
<td>70</td>
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<tr>
<td><strong>Learning to Listen</strong></td>
<td>100%</td>
<td>0% chance?</td>
<td>100%</td>
<td>47%</td>
<td>0%</td>
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<td><strong>Song Identification</strong></td>
<td>100%</td>
<td>50%</td>
<td>80%</td>
<td>75%</td>
<td>0%</td>
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<tr>
<td><strong>Phrase Identification</strong></td>
<td>100%</td>
<td>5%</td>
<td>75%</td>
<td>0%</td>
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</table>
Speech Skills-1 year post ABI

- 100% vocalize
- 60% use vowel variety
- 80% use initial sounds to imitate words /b, m/
- 80% have increased their quality voice for pitch and duration

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Electrophysiology (Shuman He)

• Cortical Potentials
  • Threshold Detection
  • Gap and Electrode Change
Electrophysiology

- **Objective Measures:**
  - **Standard condition:** 800-ms biphasic pulse train.
  - **Gap condition:** Two 400-ms stimulus bursts separated by a silent interval (i.e. gap).
Area of Investigation
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
  • Limited speech perception – No open set in US
  • Speech Production—Some emerging—very delayed
• Objective Measures--