Auditory Brainstem Implants in Non-NFII Pediatric Patients:

William Shapiro, AuD, Janet Green, AuD, Alison Singleton, AuD, J. Thomas Roland Jr, MD, Susan Waltzman, PhD

NYU Cochlear Implant Center
New York, New York
ABI in Children Inclusion Criteria

FDA clinical trial

• Age 18 mo to 21 years
  • Younger the better – earliest age controversial
• Bilateral profound (absent hearing) SNHL documented on physiologic and behavioral assessment
• No cochlea or cochlear nerve hypoplasia
• Inability to place a CI
• Meningitis with no benefit from CI
• Failed CI attempt
• Strong family support
• English language competency in guardians
• Reasonable expectations of family
  • Understand that child may not develop oral language
ABI in Children Exclusion Criteria

• Anomaly or pathology of brain stem or cortex
• NF2 or other brain stem or cranial nerve tumors
• Medical contraindication
• Psychological contraindication
Work Up

- Imaging
- Behavioral testing
- Communication evaluation
- Neuropsych
  - Other

Physiologic testing
  - ABR (ASSR)
  - OAE (test hair cell function)
  - EABR (via promontory stimulation with needle electrode)
    - Not commonly available
    - We developed special equipment and can do reliable testing
Electrode Design
Programming
Programming timetable

• OR

• 4 weeks post- in OR

• 1-4 days post initial activation in office

• Monthly ..........
Programming Considerations

• The possibility of stimulating **other brainstem structures** presents an added risk to the recipient
  • need to be conservative
  • careful monitoring- two person approach critical

• Aim for just a few electrode pairs at IS

• Possibility of **side effects** on many electrodes
  • not as prevalent as NFII population
  • BP vs MP stimulation

• The electrode array **numbering** is spatially different
  • no definite **tonotopic** relationship
Programming Considerations

• ECG and BP monitoring required at initial activation

• Different **pitch** perception than CI

• Sessions more **lengthy**

• Takes longer to get to ‘cruising altitude’

• Rehabilitation- key component
 ABI

• 41 NFII ABI surgeries to date

• Non NFII- 6 pediatric patients
  • 1 child at 18 months
  • 1 child at 9 years
  • 1 child at 17 years
  • 1 child at 6 years
  • 1 child at 4.5 years-3months
  • 1 child at 2.11 years-OR
Case #1

- 18 mos, no cochlea or nerves on imaging
- No promontory stim EABR
- Good insert of electrode paddle with intraoperative EABR, no non auditory stimulations
<table>
<thead>
<tr>
<th></th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>6000</th>
<th>Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>7-10-13</td>
<td>40</td>
<td>35</td>
<td>40</td>
<td>30</td>
<td>35</td>
<td>40</td>
<td>dBHL</td>
</tr>
</tbody>
</table>

IT-MAIS-
pre-op 0/40
post-op 22/40

<table>
<thead>
<tr>
<th></th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>6000</th>
<th>Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>11-10-14</td>
<td>40</td>
<td>40</td>
<td>40</td>
<td>45</td>
<td>50</td>
<td>40</td>
<td>dBHL</td>
</tr>
</tbody>
</table>

Total communication
Case # 2

- 9 yo girl, no cochlea on left, hypoplastic cochlea on the right
- Implanted with CI at age 2, moderate results
- Device failure 6 years later, reimplantation with obstruction, only one electrode and FN stim
- Regressing
- Referred to NYU
- 2nd revision unsuccessful
- 6/13 ABI
P1 = 1.03 ms

reversed polarity

dB nHL
12 months post*- variable mode

<table>
<thead>
<tr>
<th>date</th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>6000Hz</th>
<th>Speech</th>
</tr>
</thead>
<tbody>
<tr>
<td>06-25-14</td>
<td>35</td>
<td>30</td>
<td>25</td>
<td>25</td>
<td>30</td>
<td>30</td>
<td>10dBHL</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>ESP closed set</th>
<th>ESP closed set spondees</th>
<th>ESP closed set mono</th>
<th>WPII closed set words consonant recog</th>
<th>MLNT – easy open set</th>
</tr>
</thead>
<tbody>
<tr>
<td>100% rec</td>
<td>96% rec</td>
<td>63% rec **29% (CI)</td>
<td>68% MLV **28% (CI)</td>
<td>25% MLV **25% (CI)</td>
</tr>
</tbody>
</table>

These results are equal to or nearing her best CI performance

*Moog Center for Deaf Education
** score just prior to 3rd CI
Case #3

• 17 yo
• No hearing
• Educated in mainstream
• Total communication
• Speaks, speech reading, augmented with sign
• Assistance in school
• About to leave home for a famous cooking school and wants ABI (security, safety)
• Significant counseling efforts
• ABI 6/13
3 month / 1 year evaluation  
Case #3- (19 electrodes MP1+2)

<table>
<thead>
<tr>
<th>dB</th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>3000</th>
<th>4000Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date</td>
<td>25</td>
<td>25</td>
<td>25</td>
<td>35</td>
<td>45</td>
<td>60</td>
</tr>
</tbody>
</table>

/ba/-15dB; /sh/-15dB; /s/-15dB

Question/statement -40%, 40%  
Noise/voice- 63%, 58%  
Spondee same/different- 50%, 35%  
Male/female- 70%, 30%

CID visual enhancement  
without- 3%  
with- 27%
Case #4- 6 y/o F- Cochlear nerve deficiency?, small modiolus, cochlear aplasia

- Congenital deafness, late diagnosis, very bright child, no other issues
- Had pre-op prom stim EABR, left ear (positive results)
- Implanted at age 2
- Never really took off, moved from central PA to Phila for Clarke School
- We saw patient, No HEARING, vibrotactile device?
- Introduced Sign Language- began to progress
- ABI Right side desired/offered
ABI-

- No Cochlear nerve seen
- Good ABI placement
- Intra-op EABR in numerous bipolar pairs across/around the device
- Initial activation OR- successful
- 19 active electrodes
  - MP1+2
- Closed set discrimination, emerging open set
  - Better performance on ABI side than CI- best AU
Issues

• Surgeon prepared for numerous anatomic variants

• Audiologist prepared for not dealing with the “tonotopically friendly” cochlea

• Auditory progress slower than CI patients

• It’s like my daughter’s homework

• “Very complicated”
Conclusions

• Preliminary results indicate ABI placement and programming safe in children

• All 4 patients are getting various degrees of benefit

• Rehabilitation critical in this population

• Further studies are needed
NYU Cochlear Implant Center

- Co-Directors of the Cochlear Implant Center
  - J. Thomas Roland Jr., M.D.
  - Susan Waltzman, Ph.D.

- Surgeons
  - J. Thomas Roland Jr., M.D.
  - Sean McMenomey, M.D.
  - Daniel Jethanamest, M.D.
  - David Friedmann, M.D. – Fellow

- Cochlear Implant Audiologists
  - William Shapiro, Au.D. – Supervisor
  - Betsy Bromberg, M.A.
  - Janet Green, Au.D.
  - Laurel Mahoney, Au.D.
  - Carie Page, Au.D.
  - Alison Singleton, Au.D.
  - Kaitlyn Coscione, Au.D.
  - Jennifer Rhodes, B.S. – AuD Fellow

- Educational Coordinator
  - Rose Drous, M.Ed., Cert AVT

- Speech Language Pathologist
  - Nancy Geller, M.A.

- Research
  - Mario Svirsky, Ph.D.
  - Susan Waltzman, Ph.D.
  - Arlene Neuman, Ph.D.
  - David Landsberger, Ph.D.
  - Matthew Fitzgerald, Ph.D.
  - Chin-Tuan Tan, Ph.D.
  - Elad Sagi, Ph.D.
  - Mahan Avadpour, Ph.D.
  - Monica Padilla-Velez, Ph.D.
  - Annette Zeman, Au.D.
  - Maggie Miller, Au.D.
  - Natalia Stupak, Au.D.
Auditory Brainstem Implant

7 months post