Pediatric Auditory Brainstem Implants: New Challenges for Audiologists

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Disclosure: ABIs donated by Cochlear Americas
ABI in Children: Two Premises

1. Pediatric ABI users in most cases represent a true model of auditory deprivation by virtue of being born without a viable cochlea/VIII N

2. Pediatric ABI users receive a scrambled auditory signal through varying numbers of viable/discriminable electrodes
ABI in Children: Two Questions

In view of deprivation and the unknowns in ABI map optimization:

1. Can plasticity overcome deprivation to facilitate auditory skill development?
2. Can effective reorganization at higher auditory centers occur in ways to support spoken communication development?
Auditory Brainstem Implant (ABI)

- Indicated for patients unable to benefit from a CI
- The ABI internal electrode array is placed on the cochlear nucleus of the brainstem
Pediatric ABIs: 36 years in the making!

1979  The first adult with NF2 to receive an ABI

1980  The first child to receive a Single-channel CI

William F. House
CI Regulatory Status

1984
Single-channel device was the first CI to receive FDA approval for adults

1990
Nucleus Multichannel device the first CI to receive FDA approval for children
Nucleus ABI FDA Approval

YR 2000 for patients with NF2 (12y+)
ABI in Children
Colletti et al. (2014) *Audiol Neurotol*

Vittorio Colletti MD

First pediatric ABI
In 1999
CAP Scores
NF2, AN, 8th N aplasia + Other Disabilities

0. No awareness of environmental sounds or voice
1. Awareness of environmental sounds
2. Response to speech sounds

Pediatric ABI CAP Scores over time

- C. Ossif +Trauma (4)
- VIII In Aplasia (20)
- C. Mal (8)
- Eight In Aplasia+ (29)
- NF2 & AN (3)
CAP Scores
VIII Nerve Aplasia/Cochlear Malformation

3. Identification of environmental sounds
4. Discrimination of speech sounds without lipreading
5. Understanding of common phrases without lipreading
CAP Scores
Ossification or Trauma

5. Understanding of common phrases without lipreading
6. Understanding of conversation without lipreading
7. Use of a telephone with known speaker
What were US families to do?

- Families began traveling to Europe at great personal expense
- Families needed a center in the US for follow-up management
- Compassionate use exemption from FDA was and still is required for non-approved devices
Experiences with Children Receiving an ABI Outside of US (N=10)

- With the ABI, speech sounds are detected at or above levels of normal conversation.
- Two children have become oral communicators; others have not progressed much beyond detection.
- A significant percentage of children have additional disabilities that would likely have impacted oral communication development even had these children had normal hearing.
US Child with ABI implanted in Italy
Comparison to CI children

CDaCI Speech Recognition Hierarchy

1 yr  2 yr  3 yr  4 yr  5 yr

Open-Set Recognition:
MLNT
LNT
PBK
HINT-C

Closed-Set Identification:
ESP
PSI

Auditory Behaviors:
IT-MAIS
MAIS

CDaCI 5-yr data

Follow-Up Interval (Yrs)

Speech Recognition Index - Quiet
Los Angeles Program

2012 Phase I safety trial approved for 10 non-NF2 children (2-5 y/o)
2013 NIDCD U01
2014 First 3 ABI surgeries
L.A. Pediatric ABI Study

- Children enrolled (N=8)
  - Ages 2;3 to 4;11 (yrs;mos)
  - Sign language
- CI prior to ABI evaluation (n=6)
- Did not meet criteria (n=3)
- ABI surgery (n=4)
- Undergoing evaluation (n=1)
Results
Emerging Pattern Perception

ABI 01
12-mos post
Age at surgery: 3y
SDT – 35 dBHL
IT-MAIS - 8

ABI 02
12-mos post
Age at surgery: 3y
SDT – 30 dBHL
MAIS - 24

ABI 03
12-mos post
Age at surgery: 2y3m
SDT – 35 dBHL
IT-MAIS - 15

ABI 04
6-mos post
Age at surgery: 4y11m
SDT – 25 dBHL
MAIS - 27
Programming the ABI

- Pediatric Advanced Life Support (PALS) and emergency equipment on standby
- Electrodes stimulated individually (guided by EABR)
- Behavioral responses for possible/probable auditory stimulation
- Monitoring for signs of non-auditory stimulation (e.g., dizziness, tingling sensation, facial twitching, coughing)
It Starts with Detection

As with other sensory device fittings, the goal in programming is to provide the child with an audible signal across a wide range of frequencies while not exceeding loudness discomfort.
The Past as Prologue

Auditory skill development with the ABI resembles that of children with the single-channel CI

- Children with the single-channel CI were older at the time of surgery and many communicated through sign language
- Pattern perception and closed-set word identification were the norm with the single-channel CI
Some children with the single-channel CI developed open-set speech recognition (Geers & Moog, 1988).

Two ABI patients seen clinically developed open-set speech recognition ~4 years and communicate via spoken language.

Auditory benefit through the ABI appears to be enhanced by visual cues for most children, fostering multimodal processing of speech.
Challenges

- CI vs ABI
  - Sequential vs simultaneous
- ASL vs spoken language post ABI
  - Cued speech
- Additional disabilities/developmental delays
  - Does the benefit outweigh the risk?
Two Questions

In view of deprivation and the unknowns in ABI map optimization:

1. Can plasticity overcome deprivation to facilitate auditory skill development?

2. Can effective reorganization at higher auditory centers occur in ways to support spoken communication development?
In a word “yes”

Some children with the ABI eventually learn to make sense of the “scrambled” auditory signal in a way that facilitates auditory development and spoken language.
However, progress is slow!

- It is critical that habilitation be tailored to the child’s individual communication needs.
- This can be a delicate balancing act between:
  - supporting the child’s most effective communication modality
  - maximizing auditory skill development
Los Angeles Pediatric ABI Team

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