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When a Cochlear Implant Is No Longer An Option: ABI Management And Outcomes Of a 11-year Old Child At 16 Months Post
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Topic: Other Implantable Devices

Keywords: Auditory Brainstem Implants

Introduction: Cochlear re-implantation typically results in positive outcomes for children who experience either a hard or soft failure. In cases where re-implantation fails, an opportunity often exists to provide an implant at the opposite ear. However, when a child has only one viable cochlea, which no longer benefits from cochlear implantation, FDA approved options to restore hearing cease to exist. For children whose primary mode of communication includes aural input, the auditory brainstem implant offers the only alternative for which hearing may continue to be part of their communication modality.

Methods: Case report of surgical, audiologic, and rehabilitative management of a child with a single “bulbous” cochlea who, after 5 yrs of cochlear implant (CI) use, experienced two re-implantation failures and eventually received an auditory brainstem implant (ABI), off-label.

Results: At 16 mos post failed CI revision attempts, the child received an ABI. ABI eABR results indicated good neural responses. Initial activation revealed long tract non-auditory side effects (NASE) that were managed through programming. Continued programming focused on use of channels that provided good sound quality, reasonable loudness growth and absence of NASE. Challenges included sudden non-responsiveness on channels with concomitant report of quality change, NASE requiring re-routing of indifferent electrodes, extreme sensitivity to distant soft sounds requiring assessment of alternate parameter settings (loudness growth, ASC breakpoints, T-SPL) and assessment of FM/DM systems with limited speech perception materials. Initial child-centered fear of change, due to longer adaptation periods, resulted in new methods for implementing program changes. Auditory skills therapy was initiated 2 weeks post activation of the child’s ABI. Weekly, individual sessions occurred, and a typical auditory skills hierarchy followed in relation to detection, identification, and comprehension. Additional time was focused on development of skills required for programming sessions, and assessment of performance with specific MAPs. Auditory skills progressively improved with periods of gain, plateau, and regression. Detailed discussion of this child’s performance in therapy occurred with the team in order to optimize programming and maximize progress. To date (10 mos post ABI), auditory skills development and performance in a therapeutic setting is approaching prior CI performance 5 yrs post implant, with the exception of connected speech skills. Future performance at 16 mos post ABI will be shared.

Conclusion: When re-implantation with a CI is no longer a viable option, candidacy for an ABI should be considered. Alternate approaches to programming and therapy may be necessary to achieve desired outcomes. Positive, open collaboration on the part of professionals at various centers serving the child is paramount and results in the best outcomes for the child and family.

Cochlear Americas\textsuperscript{1}, Advanced Bionics\textsuperscript{2}, Cochlear Americas\textsuperscript{3}, Advanced Bionics\textsuperscript{4} Cochlear Americas\textsuperscript{5}
Auditory Brainstem Implant Surgery in Infants: Our Experience at a Major Tertiary Care Center in the United States

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Topic: Surgery/Medical

Keywords: Auditory Brainstem Implants , New Indications , Young and Very Young Children

Introduction: The auditory brainstem implant (ABI) was originally developed for older children and adults with Neurofibromatosis Type 2 (NF2). Emerging data from Italy and Turkey suggest a role for the ABI in deaf non-NF2 infants who are not candidates for the cochlear implant (CI) due to anatomic considerations. To date, the experience in the United States with the ABI in the pediatric population is limited.

Objective: To determine the safety and feasibility of ABI surgery in deaf infants who are not candidates for the CI.

Methods: Infants with congenital deafness who 1) were not candidates for the CI due to cochlear or auditory nerve aplasia or 2) failed CI surgery and 3) underwent ABI surgery were included in the study. Main outcome measures included assessment of perioperative complications, electrophysiologic and behavioral audiologic responses, and speech development.

Results: ABI surgery was performed using a retrosigmoid craniotomy approach in a series of infants with profound hearing loss associated with cochlear and auditory nerve hypoplasia. Intraoperatively, multiphasic Evoked Auditory Brainstem Responses (EABRs) were obtained on multiple electrodes. Detailed surgical, audiologic, and speech outcomes will be presented. Preliminary data suggest that 1) perioperative complications rates are low, 2) EABRs used to guide placement of the ABI electrode are variable and 3) behavioral thresholds of 30-40 dB in the ABI only condition are possible.

Conclusions: Based on our experience, ABI surgery in infants is a safe and effective means to habilitate deaf pediatric patients who are not candidates for the CI due to anatomic considerations.
Auditory and Non-Auditory Responses in Children with Auditory Brainstem Implants

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Keywords: Auditory Brainstem Implants

Introduction: The criteria for auditory brainstem implants (ABI) used to be restricted to adults with neurofibromatosis type II (NFII). Recently the criteria have been extended and the ABIs can be applied in cases of children who cannot benefit from cochlear implants. Objective of the study was to analyze type of responses (auditory versus non auditory) to electric stimulation in children users of Auditory Brainstem Implant (ABI) systems.

Methods: The behavioural assessment was performed in 6 children (aged 2 to 16 years) users of ABI system. The charge level was gradually increased on each electrode with standard fitting software.

Results: The mean number of electrodes with the responses qualified as: auditory and non auditory were 11 and 3 respectively. Stimulation 11 out of 12 electrodes did not produce any responses in one child.

Conclusion: Responses qualified as auditory prevailed in all but one children. The work was supported by Polish National Science Centre, decision no. DEC-2013/09/B/ST7/04213 COI: Med-El
**The UNC Pediatric Auditory Brainstem Implant Feasibility Study: Team Approach to Patient Management and Review of Outcomes to Date**

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**Topic:** Other Implantable Devices

**Keywords:** Auditory Brainstem Implants

**Introduction:** A clinical trial of the use of the Cochlear Nucleus 24 Auditory Brainstem Implant (ABI) to demonstrate safety and efficacy for children who are unable to use a cochlear implant due to cochlear anatomy disorders has been undertaken at the University of North Carolina. Team members include neurootologists, a neurosurgeon, audiologists, speech-language pathologists and auditory physiologists. Five children have undergone surgery and had devices activated. All aspects of patient care, including surgery, electrophysiologic and behavioral assessment, device fitting and post implant intervention will be reviewed in the context of the team approach to management and decision making.

**Methods:** A repeated measures, single subject design has been used to quantify outcomes of individual children.

**Results:** Each child is a case study with unique outcomes.

**Conclusion:** The ABI provides most children with sound awareness and increased potential to incorporate sound for communication. Medical/surgical considerations are significant and require special attention to avoid complications. Collaboration of electrophysiologic and behavioral testing has been critical for device programming. At this early stage of the study it appears that visual support for communication is essential to progress.

Cochlear Corporation¹, Cochlear Corporation²
Pediatric Auditory Brainstem Implant Clinical Trial: Initial Surgical and Audiological Results

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Topic: Other Implantable Devices

Keywords: Auditory Brainstem Implants , Future of Implantable Devices

\textbf{Introduction}: Pediatric auditory brainstem implantation (ABI) is in the early stage of investigation in the United States under an FDA investigational device exemption and support from the National Institute on Deafness and Other Communication Disorders (NIDCD) to assess safety, feasibility, and early efficacy. Objective: The goal of this pediatric ABI clinical trial is to determine the safety and feasibility of ABI implantation in 10 children, ages 2 to 5 years, presenting with cochlear aplasia/severe malformation or cochlear nerve hypoplasia with no demonstrable benefit from cochlear implants. We report results from the first year of this 5-year study.

\textbf{Methods}: Eligible pediatric patients who meet inclusion criteria undergo extensive audiological, medical, speech-language, and radiological assessment, along with educational habilitation counseling. Patients are evaluated with their cochlear implant (if applicable) prior to ABI to determine whether or not responses are present to sustained auditory neural stimulation. Patients undergo ABI surgery using surgical protocols that have been developed and modified during the trial to document techniques that will emphasize surgical safety and minimize non-auditory side effects. The intent is to ensure maximum number of useable electrodes for optimal auditory stimulation of the cochlear nucleus at subsequent audiological mapping.

\textbf{Results}: To date, one 3-year-old patient has undergone ABI surgery and the speech processor has been activated using electrophysiological and behavioral techniques. Nine electrodes have been programmed and the child demonstrated consistent behavioral responses during initial stimulation. We anticipate an additional three to five ABIs surgeries in 2014.

\textbf{Conclusion}: A pediatric ABI clinical trial has recently been initiated under FDA and NIDCD guidelines. An important objective of this trial will be to elucidate proper surgical protocols and to document preliminary audiological results in a select group of pediatric patients who are suitable candidates for ABI surgery. [NIH grant # U01 DC13031-01]

Med-EL Corporation\textsuperscript{1}, Med-EL Corporation\textsuperscript{2}, Cochlear Americas\textsuperscript{3} Cochlear Americas\textsuperscript{4}
Auditory Perception After Auditory Brainstem Implantation: Early Experience

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Topic: Other Implantable Devices

Keywords: Auditory Brainstem Implants

Introduction: Children with total hearing loss due to severe cochlear anomalies or absence of the cochlear nerve most often do not benefit from conventional amplification or cochlear implants leaving them without access to sound. To date, the auditory brainstem implant (ABI) is a possible option to provide auditory access.

Methods: Subjects included 4 patients implanted between the ages of 23 months and 17 years. All subjects presented with bilateral profound sensorineural hearing loss. Radiographic studies indicated etiologies consistent with bilateral cochlear aplasia without cochlear nerves, bilateral cochlear nerve deficiency or bilateral absence of cochlea. Two subjects with cochlear nerve deficiency were previously implanted with cochlear implants without success. None of the children had responses to auditory stimuli preoperatively. All four patients were implanted with the Nucleus 24 ABI device via a retrosigmoid approach. Intraoperative monitoring was performed to assist in optimal device placement to effectively stimulate the brainstem without stimulating adjacent cranial nerves. Cranial nerves V, VII, IX and X were monitored as well. Initial activation was performed approximately 6 weeks post-surgery in the OR under sedation using eABR to determine which electrode pairs produce an auditory response without non-auditory side-effects. For the second day of stimulation, the children were awake with vital signs monitored and behavioral responses were obtained. The measurement of T’s and C’s was continued without monitoring on the third day and a program was created. The children were scheduled for monthly programming sessions and age appropriate auditory speech perception measures were administered post-implant to assess performance.

Results: The surgery was uneventful in all children. One had a delayed CSF leak after a fall 7 days post-surgery which resolved with revision surgery. Following surgery all had access to normal conversational speech at levels varying between 25-40 dB. Age appropriate speech perception tests administered 6 months post-stimulation revealed improved IT-MAIS scores, detection and discrimination of the Ling sounds, improvement in closed set discrimination auditory alone and with visual cues and one child exhibited some open-set speech understanding. Three of the 4 communicate using lipreading, speech and manual language while the 4th is an oral communicator/lipreader.

Conclusion: Preliminary results indicate that children with compromised auditory nerves who cannot benefit from a cochlear implant can obtain access to sound accompanied mainly by closed set speech understanding and minimal open-set speech recognition using auditory brainstem implants. The baseline characteristics and other factors that might correlate with development of open-set speech discrimination are under investigation.

COI: Advanced Bionics, Cochlear Americas¹
Auditory, Speech and Language Outcomes Post Auditory Brain Stem Implant in a Child with Down’s Syndrome
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Topic: Other Implantable Devices

Keywords: Auditory Brainstem Implants

Introduction: Reports are emerging on speech perception outcomes following auditory brain stem implants (ABI) in young children. However, there are little-to-no data on speech and language outcomes in this group. This presentation details auditory, speech and language outcomes in a child with Down’s syndrome, who received a cochlear implant at 1-year of age and then an ABI at the age of 3 years, 4 months.

Methods: Therapy session notes, progress reports, parent reports, the Reynell Developmental Language scales (RDLS) and Cottage Acquisition Scales for Listening, Language and Speech (CASLLS) were used to document auditory, speech and language skills at 1-year post activation of the ABI (Time A) and 1-year, 8-months post activation (Time B). In addition, audio recordings were collected at Time B. Participant received auditory training without visual cues for the first year of ABI use at one facility followed by a multisensory approach with continuous emphasis on listening and speaking during the next 8 months. The latter was delivered by a Certified Auditory-Verbal Therapist.

Results: At Time A, the participant exhibited auditory awareness of and conditioned response to the Ling 6 sounds. Occasionally she responded to her name, phone ringing and door knocks. At time B, she attempts imitation of all of the 6 Ling sounds delivered through auditory modality alone. She was also able to identify at least 5 animal sounds without visual cues. In terms of language skills, RDLS scores revealed a standard score of <63 and a percentile rank of <1 at both time points. Speech production skills at Time A included spontaneous vocalization to indicate wants and approximation of some pitch and durational changes of others, but no spontaneous words. At Time B, she produced emerging word shapes, a variety of vowels and consonants as well as a few diphthongs. Session notes and audio recordings indicated vocalizations that consisted of variegated babbling, word production and emerging melodic patterns in spontaneous utterances. Session notes and the CASLLS indicated a growing expressive and receptive spoken vocabulary.

Conclusion: Aural rehabilitation involved a multisensory approach, emphasizing auditory processing of speech and language, speech imitation and aggressive home training program. Findings suggest that within 22 months of ABI use, a child with Down’s syndrome is beginning to use auditory processes for acquisition of receptive and expressive spoken language. Together, these resulted in diverse early speech behaviors, vocabulary growth and partial transition from sign to use of spoken language.
Long-term hearing preservation in Electric-Acoustic Stimulation patients, up to 10 years.

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Background
Hearing preservation (HP) surgery was initiated more than ten years ago for combined Electric and Acoustic Stimulation (EAS). Preserved residual low-frequency hearing has been demonstrated to improve speech reception in noise as well as music appreciation in EAS users up to 2 years. Multiple study groups aimed to evaluate initial loss of residual hearing (RH) as a consequence of HP surgery. However at one year and 2 year follow-up further decline was reported. Therefore, the study aims to focus on long-term RH, speech reception and subjective benefit after first fitting up to 10 years in EAS users who underwent HP surgery.

Subjects and Methods
9 post-lingual partially deaf patients who underwent HP surgery in the Antwerp University Hospital were included in the study (11 implanted ears). HP (0%= Loss of hearing; >0 - 25%= Minimal HP; >25 - 75%= Partial HP; >75%= Complete HP), speech reception and subjective benefit (APHAB) were evaluated on a long-term.

Results
Complete HP was obtained in 3/11 ears, partial HP in 5/11 ears, minimal in 2/11 ears and one subject lost his RH completely over time. Mean HP rate was 48% (ranging from 6 up to 10 years post- first implantation). Speech reception analysis up to 10 years showed a continuous statistically significant improvement. The maximum subjective benefit was found already 3 months after the implantation, the subsequent stable period remain statistically significant for the following 10 years.

Conclusion
Long-term HP in EAS users after soft surgery turned out to be possible, although there is a small continuous decline of HP rate of 3% per year (measured from first fitting up to 6 years post-operative). Nevertheless, a continuous improvement was found in the speech reception results of the EAS users. Moreover, the positive subjective benefit, assessed 3 months post-operative, remained stable up to 10 years.