Outcomes Following Primary and Revision Auditory Brainstem Implant Surgery in Children

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ABSTRACT

Objectives: The auditory brainstem implant (ABI) was developed for patients with Neurofibromatosis Type 2 (NF2). Emerging data suggest a role for ABIs in deaf non-NF2 children who are not candidates for the cochlear implant (CI). However, experience in the U.S. with pediatric ABI surgery is limited. Here, we review outcomes following three primary ABI surgeries and one revision ABI surgery for device failure.

Study design: Retrospective single institution case series

Methods: Infants with congenital deafness who 1) were not candidates for CI due to cochlear or auditory nerve hypoplasia/aplasia or 2) failed CI surgery and 3) underwent ABI surgery via retrosigmoid craniectomy were reviewed. Outcome measures included perioperative complications, electrophysiologic and behavioral audiological responses, and speech development.

Results: Five pediatric ABI surgeries were performed (4 primary, 1 revision) in children with profound hearing loss associated with cochlear and auditory nerve hypoplasia. Mean age at primary ABI surgery was 15.8 months. Intraoperatively, multi-focal Auditory Brainstem Responses (ABRs) were obtained on multiple electrodes. There were no intraoperative complications, with an average length of stay of four days. ABRs used to guide placement of the ABI electrode were variable. Behavioral thresholds of 30-40 dB were attained in all cases, including one patient who required revision surgery after device failure.

Conclusions: Based on our early experience, primary and revision ABI surgery can be a safe and effective means to provide auditory perception to infants who are not candidates for CI. Long-term follow up is needed to determine the speech and language outcomes with the ABI in this pediatric cohort.

INTRODUCTION

A major goal of clinical hearing research is the provision of meaningful sound to individuals with hearing loss. This involves assessment of auditory brainstem implant (ABI) candidacy, perioperative evaluation, and postoperative follow-up. ABI surgery for device failure.

METHODS

Patient selection: Our clinical trial was reviewed and approved by the Institutional Review Board as well as the FDA through an Investigational Device Exemption (IDE). Inclusion criteria for our study include both pre- and postlingually deafchildren with severe to profound hearing loss. Pre-lingually deafened children include those under age 5 with deafness from cochlear nerve deficiency, cochlear aplasia or severe hypoplasia, severe inner ear malformations, or post-infectious hearing loss. Patients with a cochlear that is present and patent with a normal auditory brainstem response, and normal cochlear and cochlear nerves or NF2 surgery.

RESULTS

Pre-operative evaluation:
- Full developmental and birth history
- Obstetric history
- Physical exam
- Audiological evaluation
- Review of imaging, with repeat imaging if needed

Patient characteristics:
- Age at implantation ranged from 11 mo to 30 months, average age of 13 mo old F
- 30 mo old M
- 13 mo old F
- Four primary ABI surgeries, one revision; all right sided
- No facial nerve complications

Post-operative device activation:
All patients were awake at 0-2 days post-operatively for otologcal evaluation, established EABR and awake live activation 24 hours later.

DISCUSSION

Our experience with ABI surgery in four pediatric ABI candidates with ages ranging from a 11-30 months old and hearing loss associated with cochlear nerve deficiency and/or cochlear hypoplasia/aplasia suggests that ABI surgery is wel-tolerated at this age, with preliminary data suggesting benefits in terms of hearing rehabilitation. Our experience with one revision surgery due to device failure demonstrates that such a procedure can also be done safely. Although additional studies are required, we can say that the ABI technology represents a detailed description of pediatric ABI surgery in the United States. Based on our experience in combination with the work of others internationally, it appears that ABI surgery is feasible and safe in children under the age of five.

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