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Early experience and health related quality of life outcomes following auditory brainstem implantation in children☆

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ABSTRACT

Objective: To assess auditory brainstem implant (ABI) outcomes in children with a prospective study.
Methods: Twelve patients with cochlear nerve deficiency received an auditory brainstem implant. Patients were evaluated with age appropriate speech perception and production assessments, and health related quality of life (HRQoL) surveys for parents of subjects and for subjects if age appropriate.
Results: Twelve patients received an ABI without major complications. Eleven out of twelve received some auditory benefit from their ABI. Parental HRQoL ratings were positive for all domains with the exception of communication. Self reported overall HRQoL metrics from two subjects were also positive.
Conclusions: ABI is a good option for patients who are not eligible for or fail CI. Our findings show that despite varying degrees of postoperative performance, HRQoL ratings were positive. The presence of additional disabilities and health problems resulted in less positive HRQoL outcomes. Our results emphasize the need to assess outcomes in these patients beyond speech perception and communication.

1. Introduction

Over the last several decades, cochlear implants (CI) have been used as a treatment for individuals with profound hearing loss often with tremendous benefit in speech and communication abilities [1]. Some with congenital profound hearing loss are not candidates for cochlear implantation because of absent cochleae or deficient cochlear nerves. The auditory brainstem implant (ABI) was initially designed for use in Neurofibromatosis 2 (NF2) patients with bilateral vestibular schwannomas and loss of their cochlear nerves [1]. Of patients undergoing ABI surgery in the US, 81% acquire auditory sensations [1]. At our center, we have implanted 40 deafened NF2 adults with an ABI. Adequate data from 31 patients, reveals 21 patients have sound detection, of which 9 have closed set discrimination and 2 have some open set speech perception.

More recently, the ABI is being explored as a treatment option for pediatric patients with cochlear nerve deficiency, bilateral cochlear ossification, and absent or severely malformed cochleae; conditions that preclude benefit from a CI. Centers with approval for investigational use of this device have published data concluding that ABI surgery is safe although outcomes have been variable [2–6]. In one study, 29 out of 35 children who received ABI had closed set word discrimination and 12 had open set discrimination above 50% [7]. A study by Colletti et al. reported that all 21 patients with cochlear nerve deficiency who initially failed CI and went on to ABI achieved environmental awareness and responded to speech sounds. Of the 21, 41% achieved open set speech perception [8].

In one United States institution, four pediatric patients implanted with an ABI achieved environmental sound awareness. One patient had spontaneous device failure. Another patient had device failure due to blunt trauma, a revision ABI and device failure a second time due to blunt trauma [3]. At another center in the United States, only one out five patients is frequently responding to environmental stimuli at the one-year post implantation stage [2].

While speech and hearing outcomes are a core part of ABI evaluation, they give a limited picture of a subject's outcomes. Hearing loss impacts psychosocial aspects of a person's life, such as communication, self-esteem and social relationships [9]. CI literature has explored these domains by creating CI specific tools to measure health related quality of life (HRQoL). HRQoL is defined as an individual's perceived mental and physical health and has become an increasingly important way of measuring outcomes and value of health interventions.

☆ This study was reviewed and approved by the institutional review board of New York University School of Medicine.

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In attempt to better encapsulate outcomes in other domains in ABI pediatric patients, we collected data from the subjects' families regarding the HRQoL of their child following ABI. We used a validated CI parental survey to collect the data to report HRQoL data on this population.

2. Materials and methods

This study was reviewed and approved by the institutional review board of New York University School of Medicine. Informed consent was obtained from the parents of all subjects.

2.1. Inclusion criteria

Patients ranged in age from 18 months to 18 years. Preoperative evaluation included MRI with or without a CT that demonstrated at least one of the following: cochlear nerve deficiency, cochlear aplasia or severe hypoplasia, severe inner ear malformation or post-meningitis ossification precluding CI. If the patients had a cochlea, a CI was placed first, given that imaging modalities cannot adequately predict CI performance in patients with cochlear nerve deficiency [10]. Patients that did not derive benefit from or progress with the CI after six months of consistent use were evaluated for ABI. Patients without a cochlea bilaterally did not need to receive a CI first.

2.2. Subject demographics

Patients were evaluated from other institutional referrals or from patients at our center with suspected cochlear nerve deficiency who did not benefit from their CI. Subjects were evaluated by a multidisciplinary ABI team to determine their baseline capabilities and eligibility. Twelve pediatric patients were determined to be eligible for an ABI and their families underwent extensive counseling. Six patients were male and six were female. Their ages at implantation ranged from 22 months to 17 years, with the mean age at five years. All patients had bilateral profound hearing loss. Patent demographics are outlined in Table 1.

2.3. Surgical implantation

The pre operative evaluation consisted of a thorough medical evaluation, a high resolution MRI of the brain and internal auditory canals. An electrically evoked auditory brainstem response (EABR) exam under anesthesia was done in cases where an absent cranial nerve was suspected and there were no auditory responses on behavioral testing.

A retrosigmoid approach was used in all pediatric subjects and the surgical team consisted of our multi-disciplinary skull base team (neurosurgery and neurotology) as well as a pediatric neurosurgeon through a technique previously published [11]. Intraoperatively, facial and lower cranial nerves were monitored. The Nucleus ABI541 was used in all patients. If the ABI was to be implanted on the same side as a previous CI, the CI was explanted during the same operation. The cochlear nucleus was identified and the device was placed. An EABR was used to optimize the position of the electrodes and assess for non-auditory responses.

Postoperatively, all patients were admitted to the pediatric intensive care unit (PICU) for at least 24 h and were transferred to the regular floor when appropriate. A non-contrast CT scan was obtained prior to discharge to evaluate device placement, and screen for postoperative hydrocephalus or intracranial hematomas. Patients were followed closely for any complications.

Initial device activation was performed under general anesthesia in the operating room with cranial nerve monitoring between 3 and 7 weeks following implantation. Electrodes that produced an EABR without non-auditory side effects were noted. Additional stimulation sessions were performed in subsequent days in our clinic. The devices were programmed to provide optimal auditory stimulation without non-auditory side effects.

2.4. Hearing and speech and language outcomes

Post-operative auditory function was evaluated using behavioral audiometry and commonly used, developmentally appropriate measures of speech perception, speech production and language. Subjects were evaluated at 1, 3, 6, 12, 18, 24, 30, and 36-month intervals using one or more of the following measures when appropriate. (1) The Infant-Toddler Meaningful Auditory Integration of Sound (IT-MAIS) is a parent report scale for children under the age of four years where parents report their child's communication abilities [12]. (2) The Ling Six Sound test presents speech sounds (ah, oo, ee, sh, s, m) via live voice and assesses the subject's ability to detect and discriminate the sounds [13]. (3) The Early Speech Perception (ESP) test assesses pattern perception, spondaic word identification and closed-set monosyllabic word identification. Subjects are placed in one of four categories, ranging from detection to consistent word identification [14]. (4) The Glendonald Auditory Screening Procedure (GASP) uses phonemes, words or sentences to assess identification and comprehension in a closed set [15]. (5) The Common Phrases test assesses the ability of subjects to repeat a phrase or at least the key word in a phrase given to them in an open set [16]. (6) The Minimal Auditory Capabilities (MAC) battery is a modified open set speech perception test that evaluates the subject's ability to distinguish the difference between noise/voice, male/female and same/different sounds [17]. (7) The Oral and Written Language Scales (OWLS) is a speech and language evaluation that assesses both comprehension and expression of language for subjects aged 3–21 years [18]. Results are compared to normative results in age-matched peers without hearing loss.

2.5. HRQoL measures from parents/caretakers

To collect data on HRQoL, we adapted the *Children with Cochlear Implants: Parental Perspectives* survey, a validated HRQoL tool for pediatric CI patients [19–21]. The survey consists of 74 questions and is completed by the parents of CI recipients. Some questions address one of eight HRQoL domains: communication, general functioning, self-reliance, well-being, social interactions, education, effects of implantation and supporting the child. The rest of the questions address the process of implantation and decision-making. The survey questions are answered on a 5 point Likert scale (ie Strongly agree, Agree, Neither agree nor disagree, Disagree, Strongly Disagree).

Although the survey was designed for parents of CI recipients, the majority of the questions were relevant to families of ABI recipients. Some questions were determined to be not suitable to our population or relevant to our implant center and were excluded; they are listed in Appendix 1. The final survey was 65 questions, reproduced in Appendix 2. The survey were distributed at a single point in time, such that parents filled out the surveys based on differing periods of device use and experience since their child's implantation. To analyze the surveys, responses to positive statements were ranked 1–5 and responses to negative statements were ranked on a reverse scale. The average response to questions within a domain was determined for each subject.

2.6. HRQoL measures from pediatric subjects

To collect self-reported HRQoL data from the subjects, the *Kid KINDLR Questionnaire for Measuring Health-Related Quality of Life* for children aged 8–11-year olds was used. It is an established HRQoL questionnaire that has been tested for reliability, validity and internal consistency [22]. It has 24 questions and covers six subscales: physical well-being, emotional well-being, self-esteem, family, friends, and school. The questions are answered on a 5 point Likert scale (i.e. very often, often, sometimes, seldom, never). An overall score and a score for