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The value of intraoperative EABRs in auditory brainstem implantation

Abbas Anwar*, Alison Singleton, Yixin Fang, Binhuan Wang, William Shapiro, J. Thomas Roland Jr., Susan B. Waltzman

New York University School of Medicine, NYU Langone Medical Center, United States

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ABSTRACT

Objective: To compare the intraoperative electrically evoked auditory brainstem response (EABR) morphologies between neurofibromatosis II (NF2) adult auditory brainstem implant (ABI) recipients who had auditory percepts post-operatively and those who did not and between NF2 adult ABI recipients and non-NF2 pediatric ABI recipients.
Methods: This was a retrospective case series at a single tertiary academic referral center examining all ABI recipients from 1994 to 2016, which included 34 NF2 adults and 11 non-NF2 children. The morphologies of intraoperative EABRs were evaluated for the number of waveforms showing a response, the number of positive peaks in those responses, and the latencies of each of these peaks.
Results: 27/34 adult NF2 patients and 9/10 children had EABR waveforms. 20/27 (74.0%) of the adult patients and all of the children had ABI devices that stimulated post-operatively. When comparing the waveforms between adults who stimulated and those who did not stimulate, the proportion of total number of intraoperative EABR peaks to total possible peaks was significantly higher for the adults who stimulated than for those who did not ($p < 0.05$). Children had a significantly higher proportion of total number of peaks to total possible peaks when compared to adults who stimulated ($p < 0.02$). Additionally, there were more likely to be EABR responses at the initial stimulation than intraoperatively in the pediatric ABI population ($p = 0.065$).
Conclusions: The value of intraoperative EABR tracing may lie in its ability to predict post-operative auditory percepts based on the placement of the array providing the highest number of total peaks.

1. Introduction

Cochlear implants (CI) are considered the gold standard of treatment for patients with severe to profound sensorineural hearing loss. These devices use a multiple electrode array that is inserted directly into the cochlea to electrically stimulate the cochlear nerve (cranial nerve VIII). A major contraindication to a CI is an absent cochlea or cochlear nerve, as both of these structures are required for a CI to effectively transmit electrical signals to the central auditory cortex.

There is a subset of patients who are eligible for cochlear implantation but may actually be at risk for worse audiologic outcomes after implantation. For example, patients with neurofibromatosis type 2 (NF2), a genetic disease characterized by a defect on the long arm of chromosome 22 and the presence of bilateral vestibular schwannomas that often results in total deafness, may not receive any benefit from a CI. This is because the growth of the benign tumors themselves or the surgery to remove them may affect the cochlear nerve to such an extent that a CI is no longer a feasible option. For these patients, an auditory brainstem implant (ABI) may be the only remaining alternative for auditory rehabilitation.

An ABI is an electronic device designed to bypass the cochlea and the cochlear nerve in order to transmit sound directly to the cochlear nucleus in the brainstem. The cochlear nucleus, which is situated in the lateral recess of the fourth ventricle, receives signals from the ascending fibers of the cochlear nerve and transmits these signals to higher levels of auditory processing within the brain. An ABI is inserted directly onto the surface of this cochlear nucleus, and in this way, may provide a sense of hearing by stimulating the auditory pathway at a level higher than the damaged cochlear nerve (Fig. 1).

Since the approval of the ABI in 2000, adults and children older

* Corresponding author. NYU Langone Medical Center, Department of Otolaryngology – Head & Neck Surgery, 550 First Avenue, New York, NY 10016, United States.
E-mail address: abbas.a.anwar@gmail.com (A. Anwar).

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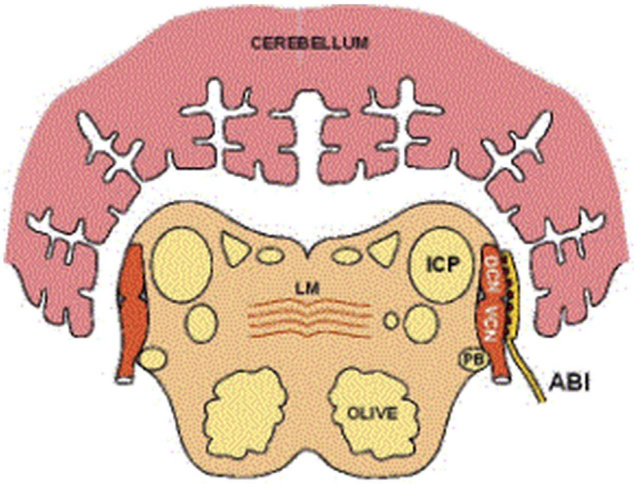

Fig. 1. Location of ABI on cochlear nucleus in brainstem.

than age 12 with hearing loss secondary to NF2 with bilateral acoustic tumors have been the primary group of patients implanted with ABIs. Audiologic outcomes in these patients with NF2 have been encouraging, although in general, speech perception outcomes are poor when compared with those of CI users. Nevertheless, most NF2 patients that receive an ABI can detect and often discriminate sounds that can be useful in everyday life and can help to enhance lip-reading [1,2].

More recently, ABIs have begun to be utilized in adult patients who do not have NF2. These patients have a variety of non-tumor etiologies for their deafness that make CIs impractical including bilateral temporal bone fractures and cochlear ossification secondary to meningitis. Published studies involving this subset of non-tumor adult patients have shown dramatic levels of speech recognition as well as significantly better speech recognition scores compared to their NF2 counterparts [3–7].

An exciting area of new research is examining whether these promising results will translate to non-NF2 congenitally deaf children who cannot benefit from CIs. This subset of children are born with profound sensorineural hearing loss due to cochlear or retrocochlear pathologies other than NF2. These anomalies range from cochlear nerve aplasia to severe cochlear deformities.

Several recent studies have evaluated this specific subset of non-NF2 congenitally deaf children who have received ABIs. For example, in a consensus statement on the long-term results of ABIs in children with complex inner ear malformations, several centers in Europe were able to obtain a pure tone average with an ABI between 30 and 60 dB HL in many patients [8]. In another study by Colletti et al., 64 children who received ABIs were reviewed up to 12 years after their surgery. Auditory perceptual abilities were evaluated on the Categories of Auditory Performance (CAP) scale. Seven children (11%) were able to achieve the highest score on the CAP test, twenty (31.3%) achieved open set speech recognition (CAP $>$ 5) and 30 (46.9%) achieved a CAP level of 4 or higher [9]. In another study, Colletti compared children with bilateral sensorineural hearing loss due to cochlear nerve deficiency who received CIs versus those who received ABIs and showed that those implanted with ABIs did significantly better with regard to open set speech perception and verbal language competence [10].

While it appears clear that ABIs can provide significant benefit to both children and adults both with and without NF2, accurate placement of the device intraoperatively onto the cochlear nucleus is paramount to obtaining a satisfactory result. As such, finding ways to assist in precise device placement to optimize device activation/stimulation is an area of immediate interest in both non-NF2 and NF2 patients.

Electrically evoked auditory brainstem response (EABR) tracings have been used intraoperatively for several years in order to aid with precise positioning of the ABI on the cochlear nucleus. Waring et al. was the first to describe the morphology of EABR waveforms that occurred when stimulating an ABI at the cochlear nucleus [11]. Waveforms containing two to four peaks were described in his research and were each attributed to stimulation of potential anatomic landmarks along the auditory pathway. Nevertheless, although intraoperative EABRs are still routinely recorded in most centers performing ABI surgery mostly to ensure appropriate device placement, studies examining the correlation between EABR waveforms and clinical outcome have been limited.

In this study, our objective was to compare the intraoperative EABR morphologies of NF2 adult ABI recipients whose device provided auditory percepts post-operatively with those NF2 adults whose device did not provide auditory percepts. In this way, we hoped to find certain characteristics of intraoperative EABR waveforms that may be predictive of whether an ABI can provide any meaningful auditory stimulation post-operatively. In addition, we also examined the EABR morphologies of non-NF2, prelingually deaf children who received ABIs and compared them with the EABRs of NF2 adults who received ABIs. We hypothesized that there would be significant differences in EABRs between these two groups not only because of age but also due to differences in pathology. Finally, because the non-NF2 children had EABRs recorded both intraoperatively and again post-operatively at initial device stimulation, we also investigated whether there were any changes in the waveforms between these two recordings and whether the nature of the recordings could be correlated to postoperative speech perception performance.

2. Materials and methods

A total of 34 patients with NF2 who received ABIs from 1994 to 2014 were retrospectively reviewed. Of these 34 patients, twenty-seven (15 female and 12 male) had intra-operative EABRs that were available and included in the analysis. The mean age of this group of patients was 31.0 years with a range from 15 to 57 years old (Table 1).

In addition, under an approved research protocol, a total of 11 non-NF2 children who received ABIs were also analyzed. Of these 11 patients, 10 had intra-operative EABR data available and 7 had post-operative, initial stimulation EABR data available. The mean age of this group was 5.3 years with a range of 1.9–17.8 years old. Six of the patients had bilateral cochlear nerve aplasia or hypoplasia and the other five had either hypoplastic or absent cochlea bilaterally. All those children who had a present and patent appearing cochlea on imaging, had previously received a CI from which they received very limited to no benefit on closed set or open set speech perception prior to being considered for an ABI. Those without a cochlea proceeded directly to an ABI.

The ABI itself consists of a radio receiver-stimulator that is implanted into a well that is drilled into the temporal bone, a ground electrode which is inserted under the temporalis muscle, and the multichannel brainstem implant that is inserted onto the cochlear nucleus within the lateral recess of the fourth ventricle via a translabyrinthine approach. Sound is picked up by the microphones on the external speech processor which sits on the pinna. The speech processor transmits the signal via a transmitter coil to the receiver-stimulator that is in the temporal bone which then sends the signal to the brainstem implant thus causing stimulation of the cochlear nucleus.

Between July 1994 and September 2000 an early generation, 8-