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Auditory brainstem implant candidacy in the United States in children 0–17 years old

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ABSTRACT

Objectives: The auditory brainstem implant (ABI) is an option for hearing rehabilitation in profoundly deaf patients ineligible for cochlear implantation. Over the past decade, surgeons have begun implanting ABIs in pediatric patients who are unable to receive cochlear implants due to congenital or acquired malformations of the inner ear. No study has examined the potential population-level demand for ABIs in the United States (US). Herein, we aim to quantify the potential need for pediatric ABIs.

Methods: A systematic literature review was conducted to identify studies detailing the rates of congenital cochlear and/or cochlear nerve (CN) anomalies. Absolute indications for ABI include bilateral cochlea or CN aplasia (Group A), and relative indications for ABI include bilateral cochlea or CN hypoplasia (Group B). Data was subsequently correlated to the US Census Bureau, the National Health Interview Survey, and the Gallaudet Research Institute to provide an estimation of pediatric ABI candidates.

Results: Eleven studies documented rates of bilateral findings. Bilateral cochlea aplasia was identified in 0–8.7% of patients and bilateral CN aplasia in 0–4.8% of patients (Group A). Bilateral cochlea hypoplasia was identified in 0–8.7% of patients and bilateral CN hypoplasia in 0–5.4% of patients (Group B). Using population-level sensorineural hearing loss data, we roughly estimate 2.1% of potential implant candidates meet absolute indications for an ABI in the United States.

Conclusion: Congenital cochlear and cochlear nerve anomalies are exceedingly rare. This study provides the first preliminary estimate of cochlea and CN aplasia/hypoplasia at the population level albeit with limitations based on available data. These data suggest the need for dedicated ABI centers to focus expertise and management.

1. Introduction

Advances in neuroprosthetic devices and surgical techniques over the last 40 years have revolutionized treatment options for children born with severe to profound hearing loss. Cochlear implants (CI) have been at the forefront of this revolution, offering both sound and speech perception to pediatric patients worldwide [1]. The CI bypasses the non-functional hair cells of the inner ear to directly stimulate spiral ganglion neurons, the first order neurons of the auditory pathway. Over the past 50 years, over 300,000 individuals worldwide have received a CI [2]. Technology has been evolving during this period from a single channel implant to a multichannel auditory neurostimulator providing sound and speech perception to the majority of deaf users. The recent Lasker Award, given to developers of the CI, highlights the overwhelming success of this device and its positive impact on society [3].

There exists a small subset of deaf individuals, however, who will not benefit from the CI due to (1) a small or absent cochlea, (2) a small or absent auditory nerve, or (3) injury or scarring of the inner ear or auditory nerve secondary to meningitis, trauma, or tumor. The auditory brainstem implant (ABI) is an alternative to the cochlear implant that bypasses the damaged or absent cochlea and auditory nerve to directly stimulate the cochlear nucleus (CN) in the brainstem [4–9]. More than 1000 adult patients worldwide

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Table 1
Radiologic indications for auditory brainstem implants in pediatric population based on Sennaroglu et al. Consensus Statement, 2011 [18].

Well-defined congenital indications	Possible congenital indications	Acquired indications
(1) Complete labyrinthine aplasia (Michel aplasia) (2) Cochlear aplasia (3) Cochlear nerve aplasia (4) Cochlear aperture aplasia	(1) Hypoplastic cochlea with cochlear aperture hypoplasia (2) Common cavity and incomplete partition type I cases if the cochlear nerve is not present (3) Common cavity and incomplete partition type I cases if the cochlear nerve is present (4) The presence of an unbranched CVN, after insufficient response to CI (5) Hypoplastic cochlear nerve, if a sufficient amount of neural tissue cannot be followed into the cochlear space	(1) Postlingually deafened children due to meningitis with severe ossification of the cochlea (2) Bilateral temporal bone transverse fractures with cochlear nerve avulsion (3) Cochlear otosclerosis with gross destruction of the cochlea, which is readily diagnosed on, computed tomography and MRI [29].

have been implanted with an ABI [10]. Audiometric outcomes of ABI patients, however, have been variable for a host of reasons [11], and many patients only achieve improved lip reading scores and sound awareness [12].

Over the past decade, surgeons have begun implanting ABIs in pediatric patients who are unable to receive cochlear implants due to congenital or acquired malformations of the inner ear with the hopes of improved audiometric outcomes [13]. To date, pediatric outcomes are still being investigated and early studies have shown sound and speech perception is possible [14–16]. This relatively new indication for the ABI has increased the number of operations throughout Europe in patients with congenital anomalies who previously had no chance of achieving auditory sensation [17]. In the United States, ABIs are currently approved only for neurofibromatosis (NF2) patients ages 12 and above; however, FDA trials are now underway at four major care centers across the United States (Massachusetts Eye and Ear Infirmary, University of North Carolina-Chapel Hill, House Ear Institute, and New York University) to conduct ABI surgeries in non-NF2 children with congenital and acquired malformations of the cochlea and auditory nerve, as outlined in the 2011 consensus report [18,19].

Given that pediatric ABI implantation is still in its nascency and the patient population with bilateral congenital abnormalities is small, an accurate estimation of potential ABI patients is challenging. With new indications and patient populations for the pediatric ABI on the horizon, an important question is, "How many children are there that stand to benefit from this device?" This basic question has implications for device development, resource allocation, and surgical education. Previous studies have addressed the potential population-level needs for cochlear implants in the United States and Europe [20,21]. Using a similar methodology, we aim to quantify the overall number of potential pediatric candidates for ABIs in the United States, as well as provide commentary about the number of necessary US "ABI Centers."

2. Materials and methods

Previous studies, such as Bradham et al. and Davis et al., estimated the potential population need for cochlear implants in Europe and the United States [20,21]. Both studies employed similar population-based analyses to formulate rough estimates. Their methods included: (1) utilization of census-level data to estimate the number of deaf children in the population; (2) employment of ratios obtained from the literature to determine estimates for various exclusion criteria for cochlear implantation, e.g. neurological devastation, absence of the cochlear nerve, and presence of cognitive disorders. We employ an analogous methodology. The Massachusetts Eye and Ear Infirmary Institutional Review Board approved this study.

2.1. Indications for pediatric auditory brainstem implants

Using the 2011 consensus statement [18] as a guide (see Table 1), potential ABI candidates were divided into two groups based on radiological classifications: absolute indications for the ABI (Group A) and relative indications for the ABI (Group B). Absolute indications included (1) bilateral cochlear aplasia, which included complete labyrinthine aplasia, and (2) bilateral cochlear nerve (CN) aplasia. Relative indications included (1) bilateral cochlear hypoplasia, (2) bilateral CN hypoplasia, (3) bilateral complete cochlear otosclerosis/ossification, and (4) bilateral trauma of the temporal bone. A flowchart showing an overview of this methodology is illustrated in Fig. 1.

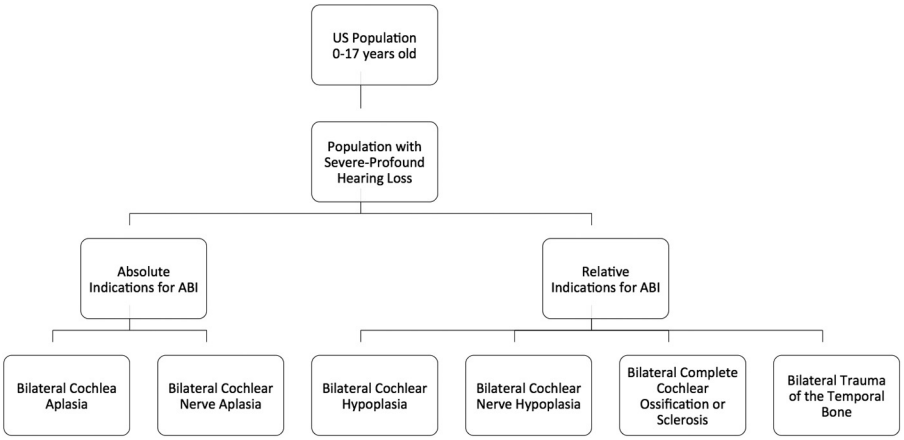

Fig. 1. Flowchart of methodology used to determine absolute and relative indications for ABI.