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OTHER ARTICLES

Auditory Brainstem Implants in Children with Inner Ear Anomalies: An Indian Perspective

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Abstract Auditory brainstem implantation (ABI) is a viable option for individuals with inner ear anomalies, or other retro-cochlear pathologies, where cochlear implantation is not a suitable option. Although ABI has its advantages in those populations, most often, ABI is not recommended or accepted by the patients because of its limitations and shortcoming such as open-set word recognition in auditory mode, limited infrastructures for carrying out ABI surgery, and the high cost of implantation. This review highlights the benefits in non-tumor patients with ABI surgery and possible reasons for the limited acceptance of the ABI device from an Indian perspective.

Keywords Auditory Brainstem Implants · Cochlear nerve aplasia · Neurofibromatosis type-2 · Speech perception

Introduction

Auditory Brainstem Implant (ABI) is the surgically implantable device that directly stimulates the cochlear nucleus of the auditory system bypassing the cochlea and cochlear nerve [1]. ABI consists of an externally located sound processor and an electrode on a titanium plate internally stimulating the second-order neurons in the cochlear nucleus [2]. Hitselberger and House performed the first ABI surgery at the House Ear Institute in a female patient with Neurofibromatosis type 2 (NF2) in 1979 [3]. Colletti and colleagues did the first pediatric ABI surgery in 2001 for an auditory nerve aplasia, a single case study [4]. In India, the first ABI surgery was done in a 15-year-old girl with NF2 in 2003 [5]. Initially, ABI was the mode of treatment option for individuals with NF2. However, over the years, ABI has become a treatment option for non-tumor patients such as inner ear anomalies and other retro-cochlear pathologies, especially when they are not suitable candidate for cochlear implants (CI). The congenital inner ear anomalies in which the cochlea are dysplastic or aplastic as well as those individuals having auditory nerves aplastic or hypoplastic are also the most probable candidate for ABI. Some of the acquired conditions such as neurofibromatosis type 2 and post-meningitis cochlear ossifications do not make cochlear implants a suitable option most often in the later stage. Studies indicated more benefit with ABI in non-tumor patients than those with NF2 tumor patients [6–8]. Although ABI can provide detection and discrimination of auditory information, it provides limited information in terms of identification and comprehension of auditory tasks as with cochlear implants, probably because of the site of the stimulation i.e., the cochlear nucleus lacks tonotopicity as in cochlea. Besides, ABI programming is more challenging than CI due to variation in electrode placement and the stimulation pattern [2]. Although globally, there are significant improvements in the ABI device development and expanding the candidacy criteria for the possible treatment option in non-tumor inner ear anomalies patients. There is a limitation for the patients looking for an ABI device as a potential treatment option in inner ear anomalies cases in India. The present review focuses on the possible reason for the poor acceptance of the ABI device globally, including in developing countries like India. Furthermore, the limitation with the ABI device in terms of speech

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perception benefit outcome, acceptance of the patients with ABI device, limited infrastructure to undergo ABI surgery, and cost of the device are the major possible reasons for the poor acceptance in India.

Method

A comprehensive systematic literature search was carried out targeting Indian studies on pediatrics populations who underwent auditory brainstem implants. The Literature search was carried in major databases such as Google Scholar, PubMed Scopus, and Web of Science. The search consisted of the following keywords; auditory brainstem implant, children and India, and their derivatives. Studies including pediatric ABI patients, speech perception measurements in those participants, and studies stating the time of implantation and assessment were included for the review. Studies with the unavailable full manuscript, insufficient information for analysis, and studies not published in English and not relevant to research question were excluded. Titles and abstracts were screened independently by two authors, and the third authors reviewed any discrepancies regarding the selection of articles.

Results

A total of 8369 articles were identified using database searches, which excluded duplicates. A total of 8347 articles were selected for the title and abstract screening. From those, 10 articles were selected for full-text screening (Fig 1). Four articles that met the inclusion criteria were selected for the study. The selection process was validated by inter-judge selection and discussion of disputes. Two articles have shown to have similar patient data, period of study and are from same institute with same authors, they are reviewed as one study (Table 1).

Patient Characteristics

From the included study in the review the total patient population constitutes 39 children. Study by Raghunandhan et al. (2019) mentioned the gender of the children as 12 males and 12 Females. Other studies have not included the gender of participants. All of the 39 patients were prelingually deaf. Figure 2 shows the percentage of inner ear deformities among the review population which comprises 31% (10 patients) with cochlear aplasia, 31% (10 patients) with cochlear nerve hypoplasia, 28% (9 patients) with cochlear nerve aplasia and 10% (3 patients) with cochlear malformations.

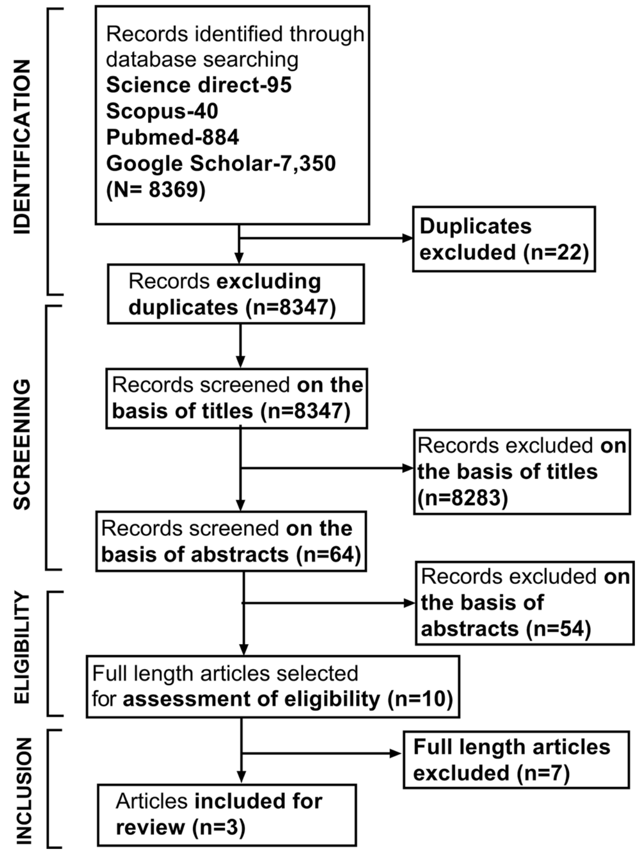

Fig. 1 Flowchart of the selection of articles

None of the patients in the studies has undergone previous cochlear implantation and none of the patients had any additional disabilities or non-auditory conditions except one child had refractive error in vision which was corrected through use of spectacles [9]. Retrosigmoid approach was adopted in all of the studies. ABI in 12.8% (5/39) were implanted on the right side, for the rest 87.1% the implanted side was not mentioned. The post-surgical complication includes facial nerve twitching in 4 patients, head/neck twitching in 1 patient, high impedance in electrodes in 2 of the patients, problems in balancing for 2 patients and bradycardia for 2 patients.

Speech Perception Benefits using Auditory Brainstem Implants

Four of the studies reported in this review has used different speech perception tests to measure speech perception abilities in ABI recipients, such as Categories of Auditory Performance (CAP), Speech Intelligibility Rating (SIR), Meaningful Auditory Integration Scale (MAIS)/(ITMAIS), and Meaningful Use of Speech Scale (MUSS).

Table 1 Characteristics of studies selected for the review

Authors/Years	Patient Demographics	Implant Characteristics	Tests Used	Methods	Outcomes
S. Raghunandhan, K. Madhav, A. Senthilvadivu, K. Natarajan, M. Kameswaran, 2019	24 Children, mean age- 4.1yrs	MED-EL (Innsbruck, Austria) ABI implant system	CAP, SIR	Tested at different time intervals. (3,6,9 and 12 months)	All patients had improvement in audiological and verbal outcomes after the ABI surgery. The mean CAP and SIR scores after 6 months of AVHT were 2.07 and 1.37 which improved to 3.42 and 2.33 after 1 year of auditory verbal therapy
Ranjith Rajeswaran & Mohan Kameswaran 2019	10 children,age range- 18 m-18yrs	MED-EL ABI Pulsar or Concert & Opus 2 processor MED-EL (Innsbruck, Austria) ABI implant system	LIP, MAIS, MUSS, MTP, CAP, SIR, Little EARS Auditory Questionnaire (LEAQ), Checklist of Auditory Communication skills, Ling six sound test	Communication performance was assessed via a battery of tests up to 24 months after first fitting	The mean communication skills on all eight tests improved significantly from preoperative to one year post-op and either increased or remained stable from one year to 2 year after surgery. Individual variation in communication development was observed
Janani Jeyaraman, Charlet Grace Rebecca, Padmasri Pokala, Rajalakshmi Ramamoorthy, Premlatha Punniyaraj, Priyadharshni Dhinakaran, Ranjith Rajeswaran and Mohan Kameswaran 2017	5 children, age range- 3–94 months	MED-EL (Innsbruck, Austria) ABI implant system	CAP, SIR, REELS, AusPlan	Auditory, speech, and language development assessed using formal and informal assessment tools up to 36 months after surgery	There was an increment in auditory perception, speech intelligibility, and receptive and expressive language scores over time in all the participants. However no participants achieved maximum scores

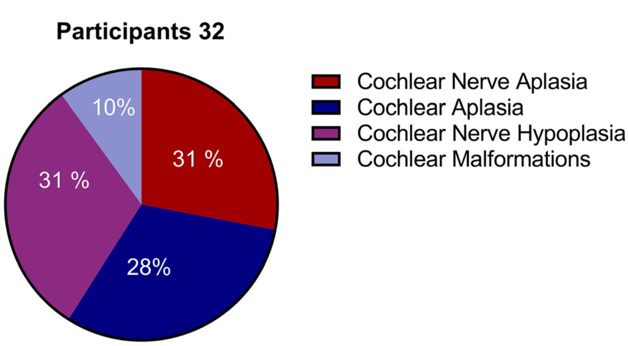

Fig. 2 Distribution of inner ear malformations in study population

In a study by Raghunandan et al. for the 24 participants implanted, the average CAP scores and SIR scores were 3.42 and 2.33, respectively. When the results were further analyzed, 12 participants with anatomical variation had a maximum number of poorer performances than those without anatomical variation, although the results were not significantly different.

In another study by Rajeswaram and Kameswaram (2020), the children were followed up for 24 months. All the children had superior scores in various tests used to assess their speech and language performance skills. The range for preoperative scores and postoperative scores after 24 months has been given below (Table 2).

In the study by Jeyaraman et al. (2017), participants auditory, speech, and language development was assessed using CAP, SIR, REELS, AusPlan, at regular intervals and were followed up till 36 months after switch on. There was a steady improvement in all the participants in auditory perception, speech intelligibility, and receptive and expressive language scores over time. However, none achieved maximum scores on any test till 36 months.

Table 2 Speech Perception benefits in different tests performed

Tests used	Preoperative range	Postoperative range after 24 months
LIP	0–4.8	14.3–100
MAIS	0–10	30–96
MUSS	0–17.5	12.5–80
MTP test		
3 words	0%	50–100
6 words	0%	22.5–100
12 words	0%	16.7–96.8
CAP	0–1	2–5
SIR	1–3	2–4
LEAQ	0–3	5–34
Checklist of auditory communication skills	0–2	9–52
Ling 6 sounds		
Detected	0	4–6
Identified	0	2–6

LIP Listening Progress Profile, *MAIS* Meaningful Auditory Integration Scale, *MUSS* Meaningful Use of Speech Scale, *MTP* Monosyllabic Trochee Polysyllabic Word Test, *CAP* Category of Auditory Performance, *SIR* Speech Intelligibility Rating, *LEAQ* The littlEars Auditory Questionnaire

Although all participants attended the therapy session, the maximum achievement was still not possible to depend solely on auditory mode of communication. Hence there are some limitations of the ABI, but all in all, speech performance as a supplementary measure to lip-reading will eventually help children maximize their communication abilities.

ABI professional team realizes that all ABI patients are not candidates for an auditory and oral-verbal communication approach. They will also need visual assistance or support (speech reading) to assist in language learning. However, it is not appropriate to ignore the auditory skill development among these ABI recipients.

Although evidence suggests good speech perception scores after ABI, these are not recommended routinely because of the variable outcomes seen compared to CI uses and surgical complications associated with it. There is concordance in findings between studies regarding complications (both auditory & non-auditory) [10]. Findings advocate that ABI in children without NF2 is a safe option and can significantly improve auditory abilities in many children, sometimes even up to the level of children with CI of 24 months of hearing age. Though ABI is often safe, complications can be fatal, even more so than in cochlear implants, and must always be addressed cautiously. Similarly, in most cases with ABI, an audiovisual communication mode is preferred because, with only the ABI, good open-set performance is not always achieved.

Discussion

There is a dearth of the published articles on auditory brainstem implant compared to cochlear implant on children. Different studies showed the benefit of the auditory brainstem implant in children when cochlear implants were not viable. Along with proper habilitation, the ABI children have shown improvement in facilitating the speech perception scores when used along with lip reading. The published studies are majorly from a few centers globally, and best of our knowledge, only a few centers are involved in India. It is generally thought that ABI has limited benefits and would only assist in lip reading. But ABI has improved speech perception with open set stimuli and intelligible speech [7].

The differences in the development of speech perception scores in different individuals can be attributed to various factors. For example, the cochlea and the auditory nerve structures, cause and duration of hearing loss, age of surgery, and language skills before implantation, implant technology, habilitation after implantation, intervention quality, and home training affect the postoperative results [9].

In a country like India, where 88 million of the total population have low income, the cost associated with the surgery, device, and maintenance will take a financial toll on family members. With most of the CI surgeries going on under the government's schemes, we have seen many patients dropping out of therapy in our personal experience.

Hence, unless the ABI will be considered for those individuals free of cost, the probability of prospective patients going for the surgery with all the risks associated with the brainstem surgery is very low. There are government schemes for funding to ABI surgery available at present under the Tamil Nadu chief minister's comprehensive health insurance scheme depending on the need of patients and recommendation from the medical team. A similar model could be incorporated in other schemes from the central government agencies under the schemes such as the Assistance to Disabled persons for purchasing/fitting of aids and appliances (ADIP) scheme, or Rashtriya Bal Swasthya Karyakram (RBSK), which might increase the beneficiaries number for ABI surgery. Similarly, there are very limited centers where ABI surgery team and the necessary infrastructure are even available as an option. In a country like India, the unavailability of surgery options near the people's feasibility might have caused a reduction in surgeries. Most of the time, the audiologist's proper referrals and counseling may help those clients decide about ABI.

The decision for the Auditory Brainstem Implantation and the rehabilitations for the same is a team approach. The team of Neurosurgeons, ENT surgeons, Audiologists, Speech-language pathologists, and Special educators are responsible for candidacy selection. Since the outcomes from ABI are limited compared to the cochlear implantation, only those who do not improve from CI or where CI is not an option would be suggested to go for auditory brainstem implantation. All the team members are responsible for providing appropriate recommendations about the approaches, risks, and benefits associated with ABI.

Due to the limited benefit with the only auditory-verbal approach, these auditory habilitation programs for the ABI children should focus on both the communication approaches, i.e., auditory and oral/verbal approaches, with and without a visual cue. Similarly, equal emphasis should be given to both the Bottom-up and Top-down procedures. The bottom-up process is a structured auditory training for enhancing listening abilities moving from simple-to-complex level, while the top-down approach uses connected speech/ conversations and provides a natural and holistic language and listening stimulation [9]. Studies have suggested that outcomes of ABI improve even after a year of implantation. Therefore, pediatric ABI recipients' habilitation programs should encourage long-term parent-clinic contact, regular visits, and train parents to be supportive by giving them home training tips.

However, there is still a lack of published literature in the Indian scenario. Moreover, most of the surgeries and publications are from the limited team of ABI. The studies indicate ABI as a safe and viable option in children with such congenital cochlear anomalies. Though the development of auditory abilities in children with ABI is at the infancy stage in comparison to the cochlear implantation, there is evidence of the children with ABI achieving test scores as CI users.

The limited numbers of ABI recipients may account for many factors, primarily not very significant open-set speech perception scores. Furthermore, for proper positioning of the ABI electrodes, the functional MRI or PET/CT of the cerebellar region (flocculus) and pons would be beneficial [11]. Three-dimensional volumetric analysis of the cochlear nucleus is being studied to understand the onion-peel like tonotopicity, a phenomenon not tapped yet by surface electrodes [2]. Similarly, more referrals for candidacy from all the team would help the client reach an informed decision regarding ABI. Like for Cochlear Implantation, if funding is available from different schemes from central Government like ADIP and RBSK, the numbers of individuals undergoing ABI can be expected to increase.

Conclusion

Overall, the benefits of ABI in non-tumor patients are encouraging though there are limited recipients in India. The possible reasons for the limited recipients and other compounding factors are highlighted in this review based on available literature. This scenario might be possible to improve if some of the corrective measures as suggested are taken into consideration by the policymakers/government policy.

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Conflicts of interest Authors have no conflict of interest to disclose.

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