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Functional Outcomes from Cochlear Implants in Children with Acquired Hearing Loss: Case Studies

Sita Sreekumar[1], Shibila Sherin AP[2], Akshaya Dev[2]

[1]Senior Lecturer, Department of ASLP, NISH, Trivandrum; [2]Speech Language Pathologist, Kollam, India

Purpose: Early identification of postlingual hearing loss (HL) and knowing its probable cause is of great significance. Causes that result in irreversible profound HL require immediate radiological investigation and intervention to decide the best listening device.

Methods: Case reports of three children with acquired HL using cochlear implant (CI) are being presented to understand the functional outcomes in domains of audition and language skills after 12 months of CI use. The age of onset of deafness and implantation were not identical. Etiology of deafness was meningitis in two cases and viral infection in the other. Consent was obtained from the parents for participating in the study. Case history review was conducted to collect demographic and audiological details of the participants. Functional benefits from CI in those children were assessed in the domains of auditory and language skills using standardized assessment tools respectively.

Results: Improvements in all domains of auditory and language skills were observed. However, the degree of benefits depended on the duration of deafness, etiology of deafness, age at onset of deafness, age at implantation and effectiveness of intervention.

Conclusions: The results of the study can be used during pre-implant CI counselling to facilitate development of real expectations about the functional benefits of CI in children with acquired HL due to various reasons and showed the importance of early cochlear implantation following the diagnosis. These case presentations give an understanding of the various factors that can affect the functional outcome from CI in children with post-lingual HL.

Keywords: Cochlear implant, Acquired/post-lingual hearing loss, Functional outcomes

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Correspondence:
Sita Sreekumar

Department of Audiology and Speech Language Pathology, National Institute of Speech & Hearing, NISH Road, Sreekariyam P.O., Trivandrum 695017, Kerala, India
Tel: 08113807779
Fax: +91-471-2944646
E-mail: sitas@nish.ac.in

INTRODUCTION

Hearing loss (HL) is a widespread problem that has an impact on one's personality and socializing skills, potentially leading to isolation and seclusion [1]. It has detrimental effect on all aspects of child development; such as speech-language skills, cognition, academic and vocational outcomes; to name a few. The impact of HL on child development varies as a function of several characteristics of HL; such as the onset, degree, type, configuration, and stability of HL. Most common causes of permanent HL found in children are congenital; nevertheless, it is reported that trauma, infection, ototoxic medications, autoimmune disorders sometimes result in post-lingual HL [2]. Acquired or post-lingual deafness are terms used interchangeably, where HL develops after the acquisition of speech and language. This have shown to impact on one's daily life, having higher risks of physical damage and negative psychological consequences [3].

There are many etiologies of permanent HL, in which meningitis is one of the most prevalent causes of acquired HL [4], followed by viral infections.

In clients with sudden acquired HL where the cause is identified, the clinician is left with a dilemma as to recommend for a hearing aid (HA) trial or directly refer for radiological investigation. The latter helps to understand the extent of obliteration especially in cases of known cause of meningitis; that help clinician to fast-track clients for evaluation for cochlear implant (CI) candidacy at the earliest.

There is vast literature on outcomes of pre-lingual children after CI, but scarce on outcomes in post-lingual children. It is expected that the outcomes of post-lingual children are generally favorable, as they would have developed the foundational speech language skills before the onset of HL [5]. Studies report that postlingual children showed better equivalent levels of HL [6] and improvement in speech perception and production abilities [1,2,5].

CI is reported to show better performance in terms of improved quality of life, and overall procedure cost-effectiveness in post-lingual deafness [1]. However, surgical and functional outcomes vary significantly among implantees. The variability in outcomes is predicted better with shorter periods of hearing deprivation before implantation, cause of HL and residual hearing [5].

The process for decision making for CI is often stressful and complex for parents of children with acquired HL. It is a challenge for a CI team to make the parents go through an informed decision making process. Lack of insurance coverage and financial assistance in India for children acquiring HL above age of 5 years adds hindrance to this process. Evidence-based information on the functional skills of children with acquired HL using CI can thus be an important step towards this process. It will aid an audiologist in setting up realistic expectations and counseling prospective CI candidates. Profiling the functional skills on the domains of auditory, speech-language performance is also an important step towards planning a holistic management.

Several previous studies have reported benefit from CI for adults with acquired HL which has mostly used open-set speech perception assessments. Use of assessment tools that profile auditory skills can probe into more real-life performances and also becomes a guideline for intervention planning [7]. There is need for strong clinical data demonstrating efficacy of CI in this population [8], and hence we assume that these case reports would be helpful in contributing to evidence-based practice by understanding the various factors affecting the outcomes. Hence, the aim of this case study is to understand the functional outcomes shown by three children with acquired HL in the domains of auditory and language skills after 12 months of CI usage.

METHODS

Case reports

This case study was approved by the Review Authority for Research (NISH187529). Three case reports have been used in this study to study the aim proposed. The following section details about the 3 cases, namely A, B, and C respectively.

Case A

A 11-year-old female child was diagnosed with bilateral profound HL secondary to the attack of pyogenic meningitis at 10 years of age. HA trial was done and speech discrimination scores were very poor and client was immediately recommended for CI counseling. High resolution Computerized Tomography (HRCT) didn't reveal any obvious abnormality in the brain parenchyma, bony skull or other observed structures and Magnetic Resonance Imaging (MRI) findings showed signs of Pyogenic meningitis. HA trial was done and there was limited benefit in both ears. The child underwent surgery for CI and was fitted with Cochlear Nucleus CP802 with CI24 (RE) straight electrode in the right ear within 2 months after diagnosis of HL with the help of Government of Kerala funding. She procured Danavox LG 290-D HA for left ear after 6 months of CI use.

Case B

A 10-year-old male child was diagnosed with bilateral profound HL due to the attack of viral infection secondary to viral fever at 6 years of age. The child was fitted with Phonak Naida V UP HA at the age of 6.2 years and attended speech language intervention from 6.10 years of age. The parents were recommended for CI counseling after HA fitting but due to financial constraints, the parents had to wait for a period of 3.3 years to undergo CI surgery. The client was fitted with Advanced Bionics Naida Q30 with Hi focus slim J electrode in the right ear at 9.3 years of age. Radiological investigation (HRCT & MRI) does not reveal any obvious abnormality in the brain parenchyma, bony skull or other observed structures.

Case C

A 15-year-old male child was diagnosed with bilateral profound HL due to the attack of bacterial meningitis at 14 years of age. HRCT reports revealed ossification within the bilateral cochlear lumen and semicircular canals (SSC); suggestive of bilateral labyrinthitis ossificans. MRI findings showed reduced T2 hyperintense signal of basal turn of right cochlea and entire turns of left cochlea. Non visualized middle and apical turn of right cochlea and entire aspects of basal turn of left cochlea. Completely absent turns of bilateral SSC with reduced signal of bilateral vestibule; which confirmed the presence of bilateral labyrinthitis ossificans. HA trial was not done and was directly sent for CI procedures. The client was fitted with Cochlear Nucleus CP802 with CI 422 straight electrode in the left ear within 3 months after diagnosis of HL. Scala vestibuli insertion of the electrode array was done with the partial insertion of intracochlear electrodes; i.e., 16 active electrodes out of 22.

Procedure

The study was initially approved by the Review Authority for Research of the Institute. Informed consent was obtained from the parents of all the three children. All the participants have been attending auditory training post-implantation in telemode after CI due to COVID-19 pandemic. The functional outcomes were assessed in the domains of audition and language skills using standardized assessment tools.

Detailed profiling of auditory skills were done using Functional Auditory Performance Index (FAPI) [9]. FAPI profiles the auditory skills in seven categories in hierarchical order namely, Awareness & Meaningful (6 items), Feedback & integration (5 items), Localizing sound source (8 items), Discrimination (4 items), Comprehension (4 items), Short term auditory memory (1 item), Linguistic auditory processing (9 items) respectively. The items were scored based on the child's performance and parental reports and a percentage score was obtained for each category separately. Scoring was done as mentioned in the manual and percentage scores were computed for each auditory skill level. Skills were labeled based on the percentage of attainment of the skill, namely; not present (N) for 0-10%, emerging (E) for 11-35%, in process (P) for 36-79%, and acquired (A) for 80-100%.

For understanding the subjective benefit from implant, the HISQUI19 questionnaire was used [10]. The questionnaire consists of 19 questions and the individual has to self-rate the level of benefit on a seven point Likert scale (always to never). The score achieved total indicates the sound quality perceived with the implant, and categorized into five (ranging from <30 – poor sod quality to >111- very good sound quality).

The language skills were assessed using Linguistic Profile Test (LPT) in Malayalam [11]. The test consisted of tasks in domains of Phonology (76 items), Syntax (10 items), and Semantics (26 items) respectively. Participants' responses were scored as one for all correct answers, half for the emergent behavior which is acceptable but not listed in expected response and zero for incorrect answers. The combined receptive and expressive score was computed for each domain separately and the percentage score was calculated. Percentage score was then compared to the age-equivalent scores.

Auditory and language skills were reassessed using the same assessment tools after a duration of approximately 12 months. A pre-post comparison of the scores obtained in each test at baseline and 12 months of CI usage was used to understand the improvement. All participants underwent auditory training and speech language intervention.

RESULTS

The results are represented in domains of auditory and language skills for the case A, B, and C respectively from Figures 1-4.

Auditory skills

It can be seen in Figure 1 that Case A has achieved better auditory skills (>80% score denoting acquired) in all domains and had shown improvements after 12 months of implant use except for skill Linguistic auditory processing (79.69% after 12 months of CI use). Whereas for Case B (Figure 2), the basic skills of auditory awareness and feedback and integration was acquired, and rest of the skills were "in process" (score between 36-79%). The skills also have not improved after 12 months of CI use. In Case C (Figure 3), auditory skills were "in process" of acquisition and there was slight improvement in the skills after 12 month of CI use.

Subjective benefit also showed a similar trend. Case A reported a good sound quality (achieved total score of 95) and reported difficulty in perceiving speech and communication interference, both in the presence of background noise. Case B reported moderate sound quality (with an achieved total score of 68), and felt difficulty in discrimination of speech in terms of gender differences and difficulty following speech in noisy backgrounds. Case C reported poor sound quality (achieved

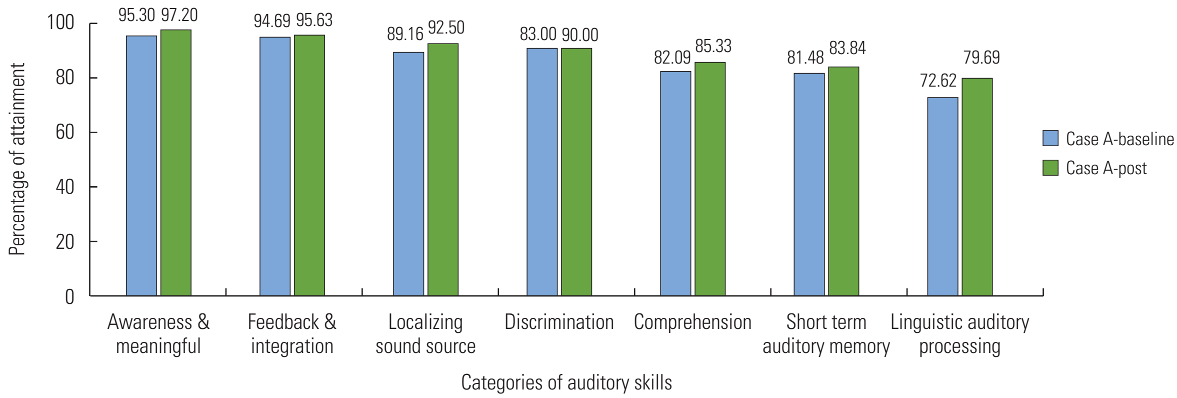

Figure 1. Auditory skills of the Case A at baseline and 12 months' post imlant (represented as "post" in figure).

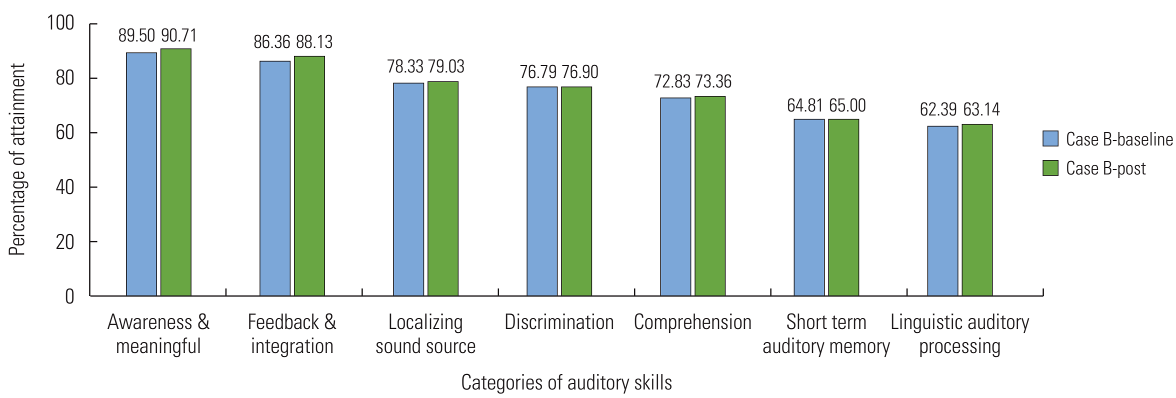

Figure 2. Auditory skills of the Case B at baseline and 12 months' post implant (represented as "post" in figure).

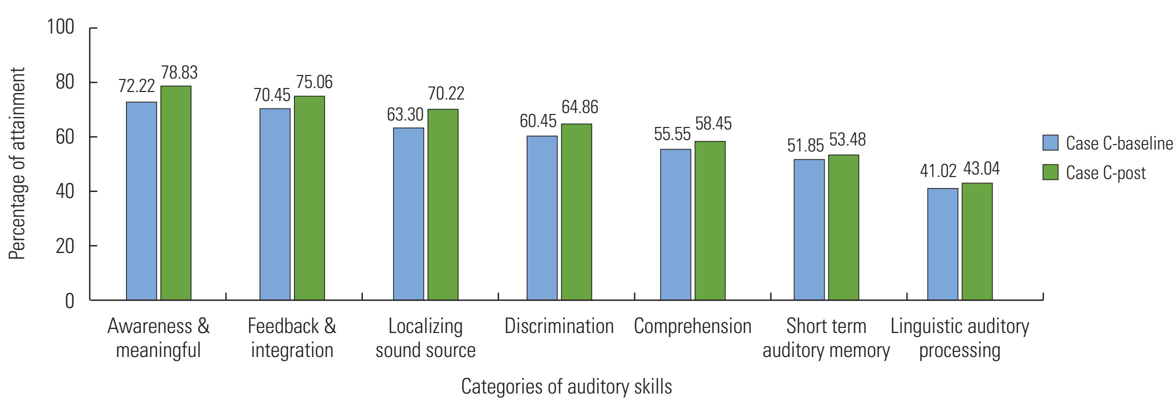

Figure 3. Auditory skills of the Case C at baseline and 12 month's post implant (represented as "post" in figure).

total score of 52) from the implanted hearing device in his personal, everyday listening situation. He found it to be very difficult to perceive speech in the presence of background noise and in the absence of visual cues, felt more difficulty to perceive speech during online classes and telephonic conversations.

Language skills

As depicted in Figure 4, the language scores were age appropriate for Case A and C in all domains of language assessed, namely phonology, semantics and syntax respectively. Case B showed inadequate skills in all the three domains at baseline, however, the skills improved in a smaller magnitude after 12 months of implant use.

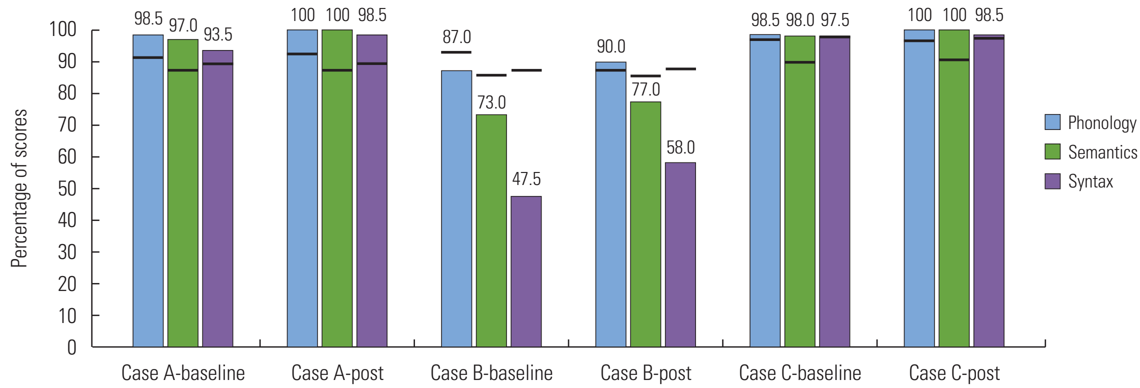

Figure 4. Language skills of the Case A, B, and C at baseline and 12 months' post implant (represented as "post" in figure). Mean score with respect to age is depicted as black line in each bar (Asha, 1997) for Case A, mean score w.r.t age as per the test is 95.5%, 91.55%, 93.9% for phonology, semantics and syntax respectively. For Case B, mean score is 93.75%, 90.95%, 92.95% and for Case C, mean score is 99.7%, 94.47%, and 99.9%, respectively for phonology, semantics and syntax.

DISCUSSION

The study aimed to understand the functional outcomes in terms of auditory and language skills after CI in three children who acquired HL. Improvements in areas of auditory and language skills was observed in all the three cases studied. Improvement in audiometry, speech perception and speech production abilities have been reported following CI in post-lingual patients [1,2,5,12,13]. Another study showed positive effects of CI on using telephone and self-confidence as well as significant improvement of quality of life in 30 post lingual deaf patients who had minimum 12 months of CI usage [13]. CI is also shown to induce a significant improvement in speech recognition in a ten-year follow up [9].

However, in this study, differences were found in the magnitude of improvement. Case A had acquired an almost higher level of performance in all the domains studied. This could be attributed to the fact that the decision making for implantation was taken soon after confirmation of HL and shorter period of auditory deprivation [13-15]. Even though the bilateral profound deafness was subsequent to meningitis in this case, she performed good after 12 months of CI use. But speech perception was reported to be difficult in noisy background. Many studies have shown that the audiological outcomes achieved in meningitis patients post-implantation were comparable to those in a wider group of implanted patients, deafened by other causes [4,16-18]. Early CI in children was associated with favorable outcomes in terms of preservation of the auditory nerve response, restoration of speech discrimination and recognition to levels comparable to patients with deafness due to other causes.

Case B was "in process" of acquisition in most of the domains of auditory performance and had exhibited delay in all domains of language skills. The auditory deprivation period was almost 3.3 years. It is shown that even though language grows across the domain of phonology, lexicon, grammar and pragmatics in 5 years of age, expansion of linguistic repertoire across multiple domains still continues and makes children proficient in reading and writing skills throughout school years [19]. Case B had missed the critical period for higher language development since he acquired HL at age of 6 years and was dependent on speech reading for spoken language comprehension until he got the CI and initiated auditory training.

Case C was "in process" of acquisition for all domains of auditory performance. He had age adequate language skills but the speech perception and comprehension were greatly affected. Subjective benefit reported also shows that he required visual cues for spoken language comprehension, and thus experienced most difficulties in telephonic conversations and online classes where visual cues were minimum. Even though he was implanted immediately following the diagnosis of HL, the cause of HL and implant history had a significant effect on the performance. Due to the presence of bilateral labyrinthitis ossification, scala vestibuli insertion of the electrode array was done with partial insertion of intracochlear electrodes. When there is a greater delay in CI, especially in cases with meningitis, significant ossification of the cochlea can preclude the possibility of hearing recovery and lead to complications/incomplete electrode insertion [20]. Thus, the factors that adversely affect the success of CI include the number of residual spiral ganglion cells, partial versus complete electrode insertion and duration of deafness prior to implantation [20].

It can be concluded that CI in post-lingual children significantly improves hearing function in terms of speech perception and speech intelligibility [9], but can show variable degrees of auditory outcomes depending on the duration of deafness, age at onset and age at implantation [21]. The case study approach used in this study helped to understand the individual variability in performance in a better way. Hence, need for accommodating speech reading and communication strategies training along with perceptual training is warranted in planning intervention in some cases.

Limitation and future directions

Due to COVID pandemic, testing was carried out in an online mode using google meet sessions for assessing auditory and language skills. Open speech perception tests would have given a better understanding of the performance, but could not be carried out as parents were not willing to report to the clinic for a post-implant evaluation. The influence of adequate home training is also a factor that needs to be investigated which cannot be tracked in these children as they were attending sessions in an online modality. Hence, further research is in these lines are warranted for better understanding the outcomes.

CONCLUSION

Early identification of postlingual HL and its probable cause is of great significance. Causes that result in irreversible profound HL require immediate radiological investigation and intervention to decide the best listening device. These case illustration gives an understanding of the various factors that can affect the functional outcome of CI in children with postlingual HL. It showed differences in the overall performance of the children due to the effect of duration of auditory deprivation and cause. Information from the results of the study can be used during pre-CI counseling to facilitate realistic expectations about the functional benefits of CI in children with acquired HL due to various reasons. The results of the study showed the importance of early CI following the diagnosis and adequate case-specific post-implant rehabilitation.

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DECLARATION OF INTEREST STATEMENT

There are no competing interests to declare.

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