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Communication outcomes following cochlear implantation in a child with cystic cochleovestibular anomaly

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Abstract

Cochlear implantation is one of the best amongst the various management options available for children and adults with severe to profound sensorineural hearing loss. Inner ear and internal auditory canal (IAC) malformations accounts to approximately 25% of congenital sensorineural hearing loss in children. The primary goal of this report was to evaluate the communication outcomes after cochlear implantation in a child with cystic cochleovestibular anomaly (CCVA). The child was evaluated through various standardized outcome measures at regular intervals to track the progress in terms of auditory and spoken language skills. The scores on Categories of Auditory Perception (CAP), Meaningful Auditory Integration Scale (MAIS), Speech Intelligibility Rating (SIR), Meaningful Use of Speech Scale (MUSS), and listening and spoken language skills showed a significant leap in 12 months duration post implantation. The report thus highlights and correlates the significant progress in auditory and spoken language skills of the child with congenital malformations to appropriate auditory rehabilitation and intensive parental training.

Keywords: Cochlear implants; Cystic cochleovestibular anomaly; Inner ear malformation; Communication outcomes

1. Introduction

Cochlear implant surgery is considered paramount amongst the various rehabilitative choices for various age groups ranging from newborns to adults with severe to profound sensorineural hearing loss (Pakdaman et al., 2011). Amongst this group, inner ear and internal auditory canal malformations present a significant challenge even to the most experienced clinicians as they account for approximately 25% of congenital sensorineural hearing loss. Many of these patients have been denied implantation also due to the uncertainty in audiological and communication outcomes post surgery. However, recent researches have reported similar outcomes for children with inner ear malformations in general, mondini dysplasia in specific and those with normal cochlea (Zhou et al. 2014; Chen et al. 2014). Till date, there are many studies reporting the surgical outcomes (Beltrame et al., 2000; Sennaroglu and Aydin, 2002; Buchman et al., 2004) and a very few reporting the audiological outcomes descriptively (Munro et al., 1996; Luntz et al., 1997; Incesulu et al., 2002; Bauer et al., 2002). Although there are many researches reporting the outcomes in general for those with inner ear malformations, there are not any reported till date on cystic cochleovestibular anomaly per se. Herein we are reporting the audiological and communication outcomes following cochlear implantation in one such child with cystic cochleovestibular anomaly (CCVA) in the implanted ear and cochlear aplasia in the other ear, based on various standardized measures.

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2. Case report

A four and half year old female child reported with the complaint of delayed speech and language secondary to hearing loss. Baseline assessment with audiological test battery revealed bilateral profound hearing loss on pure tone audiometry, bilateral type A tympanogram on impedance measurement and absent otoacoustic emissions. Findings were correlated with electrophysiological assessment using Auditory Brainstem Response (ABR), which also revealed bilateral profound hearing loss. Radiographic investigations included Magnetic Resonance Imaging and Computerized Tomography. Their findings revealed cystic cochleovestibular anomaly, which is an Incomplete Partition type I (IP-I) in left ear and cochlear aplasia in the right ear. CCVA is defined as a malformation in which the cochlea lacks the entire modiolus and cribriform area, resulting in a cystic appearance, and an accompanying large cystic vestibule. The left ear with CCVA was implanted at the age of 5 years 4 months. The surgical report showed partial insertion with nine electrodes. This child underwent regular speech therapy for around 14 months post implantation with regular mapping. A longitudinal single case study was carried out on this child in order to report the progress made by this child at regular intervals during post implantation. Auditory verbal therapy and structured auditory training were the major treatment strategies employed by the clinician during the therapy sessions and the mother was instructed to train the child with the same target goals during home training. All the assessment tools were administered in a clinical setup by a qualified speech language pathologist with more than ten years of clinical and research experience. A written informed consent was obtained from the parent prior to testing.

The child was evaluated through various standardized outcome measures at 3, 6 and 12 months to track the progress in terms of auditory and spoken language skills. The standardized measures used to track the progress include Categories of Auditory Performance II (CAP), Meaningful Use of Speech Scale (MUSS), Speech Intelligibility Rating (SIR), Meaningful Auditory Integration Scale (MAIS), and other measures assessed were listening and spoken language skills, integration into mainstream school system. CAP II measures the speech perception performance of implanted children, with their scores reflecting everyday auditory performance in a more realistic way. It is a hierarchical scale of auditory perception ranging from 0 for "no awareness of environmental sounds" to 9 for "Use of phone with unknown speaker in unpredictable context". MAIS is a structured parental questionnaire comprising ten questions assessing three different domains, viz, confidence level of child in using the processor, awareness to different sound stimuli and child's perception and identification of various sounds. Overall, the questionnaire aims to evaluate the functional abilities of a prelingual child after aiding them with hearing aids or cochlear implants from parental view point. MUSS is also a ten item questionnaire, which focuses on assessing the child's potential in using speech meaningfully. This questionnaire is again from the parental perspective. This consists of three subsections, amongst which, questions 1 to 3 assesses the ability of child's effective control on voice, questions 4 to 8 verifies the child's ability to use spontaneous speech and rest of them explores the child's capability in changing various communication strategies contextually. In totem, MAIS and MUSS are validation instruments to track the progress in oral aural skills during the post intervention period for children with hearing impairment. SIR is a tool to evaluate speech intelligibility of children with scores ranging from 1 to 5, quantifying their everyday spontaneous speech. SIR comprises five performance categories ranging from "pre recognizable words in spoken language" to "connected speech is intelligible to all listeners".

3. Results and discussion

The child was assessed with various standardized measures mentioned above at regular intervals and results are summarized in this section. The baseline assessment was done immediately after switch on and this is depicted as '0' months. The child achieved a score of 7 (Use of phone with familiar speaker) for a maximum score of 9 on Categories of Auditory Performance II by 12 months of post implantation as seen from Fig. 1. MAIS scores progressed from 0 to 38 and MUSS scores moved to 35 from baseline score of 0 as evident from Fig. 2. The MAIS and MUSS questionnaires have a maximum score of 40 and these tools enable us to assess the progress in children's early auditory perception skills and their correlation to speech production skills relative to perception. The results reveal a proportional raise in MAIS and MUSS scores. This parallel progress in scores is suggestive of the child's ability to use spontaneous speech and different communication strategies relative to the auditory perception skills. MUSS comprised of 3 subsections, viz, the voice control, use of

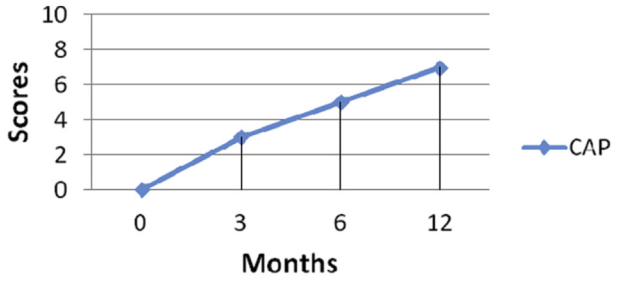

Fig. 1. Scores on Categories of Auditory Performance (CAP) at regular intervals post implantation.

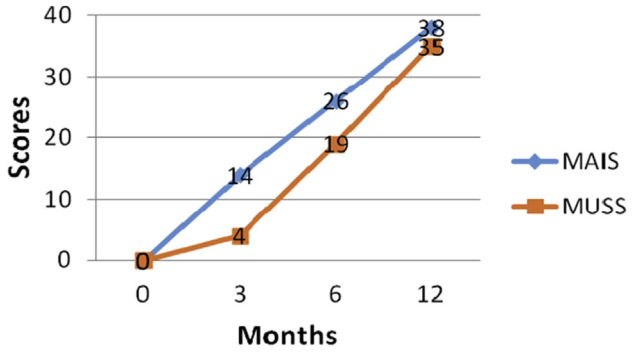

Fig. 2. Progress in MAIS and MUSS scores at regular intervals post implantation.