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Long term outcome of cochlear implantation in five children with common cavity deformity

Rabindra Bhakta Pradhananga [1,*], John K. Thomas, Kiran Natarajan, Mohan Kameswaran

Department of Implantation Otology, Madras ENT Research Foundation (MERF), 1, I Cross Street, Off. II Main Road, R.A. Puram, Chennai 600 028, Tamil Nadu, India

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ABSTRACT

Introduction: A common cavity (CC) deformity is a deformed inner ear in which the cochlea and vestibule are confluent forming a common rudimentary cystic cavity. The outcome of cochlear implantation (CI) in this deformity is not expected as good as in other cochlear deformity. Categories of Auditory Performance (CAP) score, Speech Intelligibility Rating (SIR) scale, Meaningful Auditory-Integration Scale (MAIS) and Meaningful Use of Speech Scale (MUSS) are scales designed to assess benefit of functional device (hearing aid, Cochlear implant, ABI) in young children. The aim of the study was to observe and report the long-term audiological progress and speech development of children with CC deformity after CI by means of CAP, SIR, MAIS, MUSS scores and CI aided threshold.

Methods and materials: The retrospective and prospective study was carried out in five Indian children with CC deformity who underwent CI from 2004 to 2010. Demographic data of onset and/or duration of deafness, age at implantation, time with hearing aids before implantation, duration with CI and co-morbidity were retrieved from a local data base. Their outcome score in audition and speech development on the basis of their CAP and SIR score at the end of 1 year of habilitation were collected. All the patients were called to follow up to obtain latest CI aided audiogram, CAP, SIR, MAIS and MUSS scores.

Results: One male and four female children were detected with common cavity deformity among 258 deaf children of age below 6 years operated during study period. The CAP and SIR Scores increased from average of 0.4 and 1 to 3.4 and 2.4, respectively, over one year of habilitation. The CAP score improved in subsequent follow up to average of 5.0 after more than 3 years of follow up but the SIR score hardly improved. We assessed the MAIS and MUSS at last visit after 3 years. The mean of MAIS and MUSS was 32.4 and 26.4, respectively.

Conclusion: Though the habilitation outcome was not up to the mark in children with CC deformity, CI provides excellent awareness of environment sound and development of few words.

1. Introduction

A common cavity (CC) deformity is a deformed inner ear in which the cochlea and vestibule are confluent forming a common rudimentary cystic cavity that usually lacks an internal architecture. It is often associated with abnormally formed semicircular canals [1]. If inner ear development is interrupted between the 4th and 5th weeks of gestation after differentiation of the otic placode into the otocyst but before differentiation of the otocyst into the primordial of the cochlea, vestibule and semicircular canals, a common cavity deformity results [2]. The entity constitutes 26% of cochlear malformations and is the second most common abnormality described [1].

It presents as an ovoid or spherical smooth-walled cystic cavity in which sensory and supporting cells may be scattered peripherally around the walls of the cyst. Neural population is usually sparse or absent. Hearing is usually, but not invariably, poor. Quantification of cochlear nerve function is critical to determine in these cases, as there are significant implications for cochlear implantation (CI). CI has been performed successfully and has

* Corresponding author. Tel.: +91 97510010564.
E-mail addresses: rabindrabp@yahoo.com, rabindra9034@gmail.com (R.B. Pradhananga).
[1] Permanent address: Department of ENT and Head–Neck surgery, TU Teaching Hospital, Maharajgunj Medical College, Maharajgunj, Kathmandu, Nepal.

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proven beneficial for children with CC deformity [3]. Several studies show good auditory perception outcomes in children with CC.

The standard transmastoid facial recess approach has become the popular technique for cochlear implantation. Although this approach has also been used for implantation in patients with CC deformity, it is not without increased risk for injury to the facial nerve, for a cerebrospinal fluid gusher and for electrode placement in the Internal Auditory Canal (IAC). McElveen and Miyamoto [4] had described a transmastoid labyrinthotomy approach which is the direct approach to the common cavity. They found it as an effective approach for placement of the electrode array with minimum risk to the facial nerve and intraoperative and postoperative cerebrospinal fluid leaks.

There are numerous methods for the assessment of functional outcome after CI. Some of the most commonly applied tools are the Categories of Auditory Performance (CAP) score and Speech Intelligibility Rating (SIR) scale. CAP score [5] consists of eight performance categories arranged in order of increasing difficulty with very high inter-user agreement making it a reliable tool in measuring the auditory capacity after CI. (Table 1) Similarly, SIR scale [6] is a time effective global outcome measure of speech production, which might be recognizable to the listener. It consists of an index 1–5 with increasingly better language (Table 2). The Meaningful Auditory-Integration Scale (MAIS) and Meaningful Use of Speech Scale (MUSS) [7] are other scales designed to assess functional device (hearing aid, Cochlear implant, ABI) benefit in young children based upon information provided by the child's parent in response to 10 probes in each. The MAIS is used to assess the child's spontaneous responses to sound in his/her everyday environment [8]. These 10 enquires assess three main areas: (a) vocalization behavior; (b) alerting to sounds and (c) deriving meaning from sound. Similarly, the MUSS is designed to assess the child's use of speech in everyday situation [9]. The probes of it assess the areas like: (a) vocal control, (b) use of speech without gesture or sign, (c) use of communication strategies in everyday situations.

The aim of the present study was to observe and report the long-term audiological progress and speech development of children with CC deformity after CI by means of CAP, SIR, MAIS, MUSS scores and CI aided threshold.

Table 1
Criteria for CAP score.

CAP score	
7	Use of telephone with a known listener
6	Understanding of conversation without lip reading
5	Understanding of common phrases without lip reading
4	Discrimination of some speech sounds without lip reading
3	Identification of environmental sounds
2	Responses to speech sounds (e.g. 'go')
1	Awareness of environmental sounds
0	No awareness of environmental sounds

Table 2
Criteria for SIR score.

SIR score	
5	Intelligible to all listeners
4	Intelligible to a listener who has little experience of a deaf person's speech
3	Intelligible to a listener who concentrates & lip-reads
2	Intelligible speech is developing in single words
1	Unintelligible

2. Materials and methods

After getting the clearance from ethical review board of the institute (NO.MERF-EC-AUG.14/008), the retrospective and prospective study was carried out in five Indian children with CC deformity who underwent CI at Madras ENT Research Foundation (Tertiary cochlear implant referral centre), Chennai, India during the period of 7 years from 2004 to 2010.

Demographic data of onset and/or duration of deafness, age at implantation, time with hearing aids before implantation, duration with CI and co-morbidity were retrieved from a local data base.

We used standard transmastoid facial recess approach with implantation through cochleostomy or round window and a direct transmastoid labyrinthotomy approach. In transmastoid facial recess approach, after cochleostomy or opening the round window membrane the electrode array was inserted directing anterosuperiorly to avoid entry into the IAC. In the transmastoid labyrinthotomy procedure, after cortical mastoidectomy and widening of aditus to visualize incus, there was no need to open the facial recess. Labyrinthine bone was clearly delineated in the area where the lateral semicircular canal (SCC) would normally be situated. A labyrinthotomy was created at this region above and posterior to the tympanic facial nerve. The opening created was designed to be only slightly larger than the electrode to be inserted. During insertion of the electrode, the tip was directed toward the outer wall of the common cavity here also to prevent inadvertent entry into the IAC. After the electrode had been placed, temporalis muscle facia was packed tightly at the insertion site. Intraoperative facial nerve monitoring was used in all cases.

The surgical findings, the various problems including CSF gusher, difficult insertion and techniques of control of CSF gusher were noted. Rates of extra-cochlear placement, intra-meatal placement and complication were also analyzed. Number of electrodes inserted, complications like facial nerve stimulation were noted from their records.

The intra-operative impedance across the electrode array and neural telemetry results were noted. Their improvements in audition and speech development were assessed at the end of 1 year of habilitation on the basis of their CAP and SIR score. All the patients were called for follow up to obtain CI aided audiogram, CAP, SIR, MAIS and MUSS. We compared the last result with previous scores. We also asked the parents for their satisfaction.

3. Results

One male and four female children were detected with common cavity deformity among 258 deaf children of age below 6 years operated for cochlear implantation from 2004 to 2010. Three of them were implanted at the age of 2 years, one at 3 year and one at 5 years. Two were operated for left ear and the rest on right side. We selected left ear in 2 cases as one case had hypoplastic cochlear nerve and another had Michel deformity on right side. This second child also had hypothyroidism (Pendred Syndrome) and was on Thyroxin (75 μg daily) since the age of 8 months. We inserted the electrode array of the implant by labyrinthotomy in 2 cases, by cochleostomy in 2 cases and through round window in 1 case. Intraoperatively it was noted that there was absence of round window and promontory in three cases in which cochleostomy was performed anteroinferior to the oval window in two cases (cases 2 & 5) and switched over to labyrinthotomy in one case (case 4). Other demographic data are highlighted in Table 3. Full insertion was achieved in four cases. In one case, last 4 electrode of nucleus 24 remained out of the cavity. The Case 2 had full insertion in first surgery but had incomplete insertion (last 2 electrodes) on revision surgery.

In both the cases of labyrinthotomy, there was no CSF gusher. But in all other cases profuse CSF gusher was noted after making