Facial nerve stimulation necessitating auditory brainstem implantation: 8 years follow-up a case report

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ABSTRACT

Inner ear malformations are one of the common causes of hearing loss. Common cavity is extremely rare among the types of inner ear anomalies. In this study, 8-years development period of a boy with common cavity is presented. In case of side effect such as facial nerve stimulation or insufficient hearing and language development with cochlear implant use, auditory brainstem implant may be indicated on the contralateral side. Our case had a re-implantation due to facial nerve stimulation after cochlear implant. After the second cochlear implant activation, everything seemed fine, facial nerve stimulation was seen again in the patient and the child refused to wear cochlear implant. This suddenly stopped his auditory perception and language development. Our patient started to regress. As a team, we decided that our case needed an auditory brainstem implant. Improvements in auditory perception and language development were observed in our patient after auditory brainstem implant.

ARTICLE HISTORY

Received 21 May 2021
Revised 20 August 2021
Accepted 27 August 2021

KEYWORDS

Cochlear implantation; facial nerve stimulation; auditory brainstem implantation; common cavity

Introduction

Common cavity (CC) deformity, which is a type of inner ear malformation, is defined as a single, ovoid or round chamber, representing cochlea and vestibule. There may be accompanying semicircular canals (SCCs) or their rudimentary parts. The internal acoustic canal (IAC) usually enters the cavity at its center. In addition, it is expected to have neural structures [1]. However, these neural structures could be sparse or even absent [2]. Cases with CC generally have severe to profound hearing loss [1].

Cochlear implant (CI) is considered as an effective device to restore hearing for individuals if there is a functional cochlear nerve. Cochlear implantation has been successfully applied and has proven beneficial for children with CC. Various studies show good auditory perception results in children with CC [3,4].

The standard transmastoid facial recess approach is a common surgical technique for CI. The surgical approach to CI surgery with CC cases is through a transmastoid labyrinthotomy with a straight (non-modiolar hugging) electrode, as defined by McElveen et al. [5]. It would have a location of greater interaction with the neural tissue on the periphery of the CC. This technique is intended to be an effective approach for inserting the electrode array with minimal risk for facial nerve disturbances [2].

Facial nerve stimulation (FNS) is one of the causes of potential postoperative cochlear implant surgery complications. Stimulation of non-auditory structures such as the facial nerve can cause significant discomfort and impair the potential of auditory perception by effectively limiting the dynamic range of CI [6]. It could be effective to use some methods to reduce the effects of FNS. Some of these might include changing current levels, turning off the electrode contacts causing FNS, careful remapping, or using triphasic pulses instead of biphasic pulses. However, these strategies may not provide an ideal fitting for some CI users. The next method is to require re-implantation with a new device [7].

The auditory brainstem implant (ABI) is a neuroprosthetic system that offers hearing stimuli to deaf patients who are unable to receive a cochlear implant due to anatomic anomalies. In patients with cochlear and retrocochlear pathologies, the ABI uses a
multichannel surface array to electrically activate second order neurons in the cochlear nucleus (CN). In case of side effects such as FNS or insufficient hearing and language development with the use of CI, ABI can be applied on the contralateral side [8]. Indications for ABI have increased for pediatric patients with inner ear malformations, congenital deficiency of cochlear nerve, cochlear trauma, NF2 cases, post meningitis cochlear ossifications [9]. Auditory brainstem implant electrodes are embedded in the straight and rigid silastic paddle in the brainstem that runs along the strongly curved surface of the CN, unlike thin CI electrodes that adopt the curved spatial arrangement of tonotopic axis selectivity in the cochlea [10].

The aim of the present study was to report the unusual causes of ABI placement in the contralateral ear of a CI user with CC and the observed long-term audiological progression and speech development.

Case report

Demographic data

A male patient was born at term after a normal pregnancy and delivered c/section. His birth weight was 3100 g and he had no hyperbilirubinemia. Family history had consanguineous marriage. There was no family history of a genetic disorder and hearing loss. He had no diagnosed disease, no medication use.

Preoperative audiological, radiological and auditory perception assessment

He did not pass the neonatal hearing screening bilaterally. Nevertheless, the family applied to our clinic when he was around two years of age, and an audiological test battery applied. On behavioral test with insert phones, he responded to speech stimuli at 85–90 dB on the right ear but there was no response on the left ear. There were no waves and cochlear microphonics in the Auditory Brainstem Response (ABR) test and Transient evoked otoacoustic emissions (TEOAE) was absent. Therefore, bilateral profound sensorineural hearing loss was confirmed. Bilateral hearing aids and an auditory rehabilitation program were recommended until surgery time.

High Resolution Computed Tomography (HRCT) showed bilateral common cavity with normal internal auditory canal (IAC) on the right and narrow IAC on the left (Figure 1). In addition, Magnetic Resonance Imaging (MRI) demonstrated a common cochleoves-tibuler nerve (CVN), which was normal on the right side but there was only one nerve entering the left narrow IAC (possibly facial nerve) (Figure 2).

Denver Developmental Screening Test-II was applied to screen his general development. Denver II includes four categories: fine motor, gross motor, personal-social and language. There was only a delay in language skills; other areas were normal. He had
behavioral reactions to live voice, however, there was no sound localization. He was often screaming and repeating syllables, e.g. ‘ba-ba,’ ‘da-da.’

In children with hearing loss, The Meaningful Auditory Integration Scale (MAIS) is a parent-report scale used to assess functional listening abilities. Each item is scored between 0 and 4 (0 = never, 1 = rarely, 2 = occasionally, 3 = frequently, and 4 = always). The total score can range between zero and 40. The first item of MAIS, ‘Does the child wear the device all waking hours without resistance?’ question was scored as 2-occasionally. He could not score from other items. His preoperative MAIS score was 2/40.

His receptive and expressive language was evaluated with Test of Early Language Development Third Edition (TELD-3). However, he could not adapt to the TELD-3, and it was not possible to determine his receptive and expressive language. This is because the norm values are typically collected from developing Turkish children. A child who does not have enough experience with a hearing aid has difficulty performing this test.

Cochlear implant surgery procedure, post-operative audiology and auditory perception procedure and assessment

During behavioral testing there was a response to loud sound in his right ear, and common CVN was more clearly visible on MRI. No response was observed on the left side. In March 2011, he was implanted with a Med-El FORM 24 prototype electrode to the right ear with a transmastoid labyrinthotomy approach (Figure 3(A)). He was 2 years and 7 months old at that time.

Facial nerve stimulation was not observed during the initial activation of the CI. Six months after initial activation, the number of active electrodes decreased from 12 to 8 due to facial nerve stimulation, which caused a twitch around the eye, chin and lips on the right side when the implant was used. In order to manage FNS, amplitude of the electrical stimulation was decreased and duration increased. However, FNS continued. Therefore, four electrodes on basal part were inactivated. With time, FNS was observed in the medial and apical electrodes as well.
After initial activation, he started to respond to environmental sounds. Outcomes with CI were evaluated with MAIS, Categories of Auditory Performance (CAP)-II scale and Speech Intelligibility Rating (SIR) scale and Children’s Auditory Perception Skills Test (CIAT). Auditory Performance Categories II (CAP II) was used to assess the performance of children with implants in speech perception and was completed by either parents or clinicians. The Test of Early Language Development and Speech Intelligibility Rating (SIR) were developed to measure speech intelligibility of children with implants by quantifying their spontaneous speech for clinical comparisons. Children’s Auditory Perception Skills Test (CIAT), which included the closed-set Pattern Perception Test (PPT) and the closed-set and open-set Sentence Recognition Tests (SRT), were used to evaluate the daily life functional auditory perception skills. The closed-set SRT was performed to children scoring 18/24 or above.

According to the evaluation, in his post-operative first year were MAIS 10/40, CAP-II 2, and SIR 1. Tests of PPT and SRT were presented in an auditory-verbal condition. However, he could not obtain a score from PPT, SRT and TELD-3 because he did not have enough vocabulary acquisition.

Revision CI surgery and ABI decision
As the benefit from CI diminished considerably, a revision CI surgery was planned. The CT scan demonstrated that the electrode was positioned very close to the labyrinthine segment without going into the internal auditory canal (Figure 3(B)). In May 2012 a double labyrinthotomy approach was used and the electrode was repositioned away from the labyrinthine segment Figure 4.

During re-implantation, eABR testing was done and there was no wave-V even though this child had responded to environmental sounds with his CI (Figure 5). Electrical dynamic range was very limited after re-implantation, even though there was no FNS (Figure 6). After a while, auditory development showed no more progress. The MAIS score was 0/40 and he did not want to wear his CI. He started to show angry behavior. He stopped repeating the few words he had learned before.

Auditory brainstem surgery and auditory perception assessment
On the left side there was a common cavity with a very narrow internal auditory canal containing only
one nerve, which was most probably the facial nerve. As a result, our CI team decided to implant him with an ABI on the left side. In January 2013 he underwent an ABI surgery on the left side (Figure 7). During ABI surgery, eABR measurement also was conducted and clear waves were observed with double peak (Figure 8). After activation of the ABI, four electrodes out of twelve were made inactive due to twitch on the left arm. With the remaining eight electrodes, he started to show better auditory performance. His free field thresholds varied between 40 and 45 dB at 250–4000 Hz.

In the meantime he started using his CI again. He used CI and ABI together for 3 years. After that he stopped using CI, but used ABI regularly. When assessed with CI and ABI together post-operative in 3 years, he had 70% closed-set pattern perception and 20% open-set sentence recognition score in auditory verbal condition. MAIS increased 25/40, CAP score was 4 and SIR was 2 with both CI and ABI. The receptive language test score was 30 months and the expressive language test score was 24 months.

**Eight years follow-up visit with the CI and ABI**

Behavioral responses of the patient with the left ABI in the eight-years follow-up visit are given in Figure 9. His eight-years follow-up evaluation with only ABI, CAP improved to 5 and SIR improved to 3. In auditory verbal condition, closed-set PPT and closed-set SRT were 100%. Open-set SRT was 65%. Also, he still could not talk on the phone. His language development showed improvement with ABI. The receptive
language test score was 72 months and the expressive language test score was 36 months.

He was enrolled in mainstream school. He has good reading and writing skills, but has difficulty in understanding what he reads. He obeys the rules at school. He adapted to his teacher and friends. He has a sister who loves spending time with her. It was found that the quality of life had improved because of its presence in social life.

Discussion

The case presented here demonstrates the benefit of a contralateral ABI in a patient who had decreased performance from CI due to FNS.

Beltrame et al. reported that the audiological skills of children in cases with CC improved over time [11]. However, these developments will change according to the situation of the individual. Factors such as the chronological age of the child, the duration of using CI, early intervention, capacity of the family to take care of the child, additional disorders of the child, and access to support services reflect differences. In cases with inner ear malformations, additional disability is likely to be present. There was no additional disability diagnosed in our case. However, it was noticed that there was a decrease in the speed of the child’s learning skills over time. It is thought that the reason for this may have resulted from the periods of auditory deprivation of the child as a result of FNS.

Parents should be counseled to establish realistic expectations after implantation. Conditions such as gusher, electrode displacement, or FNS are common in CC cases and are a major problem. Surgeons are experienced in controlling these situations. Cushing and Papsin et al. reported a high incidence of FNS at normal stimulation levels in cases with IEMs [12]. In the case we presented, the hearing levels of the child had to be changed as a result of FNS and the family had to be informed about it. We know that CI provides significant audiological benefit as long as the CVN is seen for cases with common cavity. However, although serious, it should be effectively handled that complications may occur during or after surgery.

The transmastoid labyrinthotomy technique has been developed to place the electrode directly into the common cavity without opening the facial recess. Beltrame et al. reported, the double transmastoid labyrinthotomy technique, which is a method that avoids electrode misplacement into the internal auditory canal. Both methods are used in cases with common cavity when necessary [13]. Double labyrinthotomy was used in our patient and the electrode was positioned away from the facial nerve but it still resulted in FNS. Our case did not have a gusher during surgery.

Eisenman et al. presented that cases with malformed cochlea showed a high incidence of facial nerve activity with the stimulation of certain electrodes [14]. In several studies, it has been emphasized that the electrodes of the patients should be removed and revision due to FNS [5,15].

CI mapping strategy also changes with the appearance of side effects in patients. Changing the stimulation modes to eliminate FNS, decreasing the current amplitude, increasing the pulse width are some of the possible solutions. Shea and Domico reported that if these suggestions do not provide effective results, offending electrodes should be turned off [16]. In our case, we tried all mapping strategies and eventually closed the electrodes causing FNS. But, our case did not improve and started to suffer from auditory deprivation.

It is known that CI provides improvement in hearing and auditory language skills not only to pediatric users with normal cochleovestibular structures, but also to users with malformed cochlea. Common cavity malformation is not an exception regarding this. Zhang et al. reported improvement in CAP, SIR and IT-MAIS scores after implantation among CC cases; but scores were lower than CI users with normal cochlear anatomy [17]. Likewise; thresholds with CI were higher in CC group. In this study, our case was
not compared with a CI user nor an ABI user with a normal cochlea. Ozkan et al. reported that CI users with CC showed poorer results in terms of auditory perception and language skills compared to cases with normal cochlea, but this progressed over time [18]. In line with these findings, Pradhananga et al. reported, improvement in CAP and SIR scores at 1-year post-operative among 5 pediatric CI users with CC [2]. Moreover, CAP score improved further in more than three years, contrary to SIR scores. Similarly; Shintani et al. reported increased environmental sound awareness and vocalization in a pediatric case with CC after implantation; along with improvement in number of words used and recognition in closed set word tests [19]. Furthermore; thresholds with CI were obtained at about 40–50 dB SPL at 8 months post-op. Ahn et al. found a relatively steady improvement after CI [15]. Although our case showed differences, it has improved at the end of 10 years.

As stated above, the progress in terms of hearing and language skills may be suboptimal in pediatric CI users with CC. An option may be bilateral implantation, which would not be applicable to the case discussed in the current work due to absence of CVN on the left side. For this reason, ABI was applied to the left ear, resulting in improvement in thresholds with the devices and auditory-language skills. The improvements were observed in MAIS, closed-set sentence recognition in auditory-verbal condition as well as CAP, SIR and aided thresholds. Colletti and Zoccante also reported that failed CI cases with CC could benefit from ABI [20]. Keeping in mind the improvement in auditory and language areas, it is important to provide bilateral hearing to cases with CC; either with bilateral CI or bimodal option CI and ABI. Friedman et al., in their study evaluating bimodal auditory performance in children with CI and contralateral ABI, four patients with cochlear nerve deficiency initially underwent cochlear implantation. However, the cases did not benefit from the device, and they performed ABI on the contralateral ear. They found that three cases performed better on auditory perception assessments using both devices than alone [3]. However, despite the absence of FNS in our case, the electrical dynamic range was very limited after CI reimplantation. Accordingly, after a while, auditory development did not progress any further. He refused to wear it when he realized he wasn’t benefiting from CI. Due to the rarity of common cavity cases and the limited experience of these cases, it is necessary to determine a strategy suitable for the team approach. The meticulous evaluation and intensive follow-up of cases with malformed cochlea are difficult, but it is essential for early intervention in the presence of a problem.

**Conclusion**

Functional assessment of hearing, auditory perception and language development, and intensive follow-ups play an important role in making the decision of CI in common cavity cases. Auditory brainstem implant is a good option for patients who are not eligible for or fail CI. Severe facial nerve stimulation limits mapping and may prevent benefit from CI and contralateral ABI may be a solution in this situation. It is possible to achieve speech development in children with ABI even if they stop using CI in their other ears.

**Ethical approval**

Informed consent was obtained from the participant presented in the study.

**Disclosure statement**

No potential conflict of interest was reported by the author(s).

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