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Outcomes of cochlear implantation in children with inner ear malformations

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Abstract

Purpose To evaluate the surgical experience and auditory functions and progress of speech development of cochlear implantation in malformed ears.

Materials and methods Between November 1995 and July 2017, thirty-seven patients (26 females and 11 males; mean age: 138.275 ± 96.24 months) with diverse anomalies of the inner ear were retrospectively examined for surgical and audiological results. Speech Intelligibility Rating (SIR), Categories of Auditory Perception (CAP), Pure Tone Average (PTA), Speech Intelligibility Rating (SRT), and Word Discrimination Score (WDS) were the audiological tests used to evaluate the efficacy of CI in the malformed inner ears.

Results CSF gusher was experienced by six patients (three with LVA (large vestibular aqueduct), one with IP (incomplete partition) I, and two with both IP II Mondini malformations and LVA). Two patients had transient facial paresis after surgery. All patients fully recovered within 6 months. The postoperative PTA, SRT, and WDS test results showed significant differences between progressive and congenital sensorineural hearing loss (p values < 0.05 for all). On the other hand, the postoperative CAP and SIR test results revealed no significant differences between the two groups. According to etiology, the PTA and SRT values were significantly lower in common cavity patients than the LVA patients (p values < 0.01); no significant differences were found among the other etiological groups.

Conclusions Cochlear implantation is safe in children with inner ear malformations. However, the success rate is low compared to patients with normal anatomy in terms of audiological results; the most successful group of patients with inner ear malformation is large vestibular aqueduct.

Keywords Cochlear implantation · Congenital inner ear anomaly · Sensorineural hearing loss · Auditory performance · Speech development

Introduction

Cochlear implantation (CI) has become a routine procedure for the management of sensorineural hearing loss (SNHL). Over the past several decades, developments in the field of microelectronics and progress in signal processing techniques have led to an increase in the performance and effectiveness of CI devices. As otologists often have apprehensions regarding the surgery for and performance of CI, children with anomalous inner ears are still challenging cases for cochlear implantation [1]. Unexpected results after cochlear implantation cannot be predicted in cases of anomalous inner ear since the neural tissue distribution and abnormal locations of anatomical landmarks such as the lateral semicircular canal and facial nerve can make surgery difficult. Further, surgeons are reluctant to perform CI in

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patients with inner ear abnormalities due to surgical complications such as facial nerve injury, labyrinthine fistula, and cerebrospinal fluid (CSF) gushers [2]. Despite the risks and potential surgical difficulties, the results of cochlear implant surgery are similar to those in congenital sufferers without malformation [3]. The purpose of this study was to evaluate the surgical experience, auditory function, and progress of speech development following CI in malformed ears.

Materials and methods

Between November 1995 and July 2017, 2647 cochlear implant procedures were performed in the Department of Otorhinolaryngology at our university. The Ethical Committee of our University Faculty of Medicine approved the study with the number of 09.2017.703. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. During this period, we examined 37 patients with diverse anomalies of the inner ear who underwent cochlear implantation. Twenty-six patients were excluded from the study due to the presence of Michele deformity, cochlear aplasia, or absent cochlear nerves, as well as late prelingual individuals with no experience with hearing aids or rehabilitation. The inclusion criteria for the study included patients with inner ear malformations with the history of bilateral profound or severe-to-profound sensorineural hearing loss and limited benefit from binaural amplification. Preoperative radiologic findings were obtained from both magnetic resonance imaging (MRI) and high-resolution computed tomography (HRCT) scans of the temporal bone, which were acquired with a slice thickness < 1 mm. The cochleovestibular malformations in this study were classified according to the Sennaroglu and Saatci classification [4]. The average follow-up was about 108.7 months, ranging from 15 to 204 months.

One surgeon performed the surgeries using a soft insertion technique. Facial nerve monitoring was first introduced in 2004 and has since been used routinely in CI except patients with common cavity. A monopolar stimulator (constant current pulses, 0.1–3.0 mA, 4 Hz, 100 μs) was used to monitor the facial nerve; an incident threshold was established at 100 mV. The monitor alerted the surgeon to unforeseen facial contractions. CI was performed using a lasy S incision, mastoidectomy, posterior tympanotomy, and oval window or cochleostomy approach under general anesthesia in all patients except for five. Abnormally located facial nerve can be more expected in cases with common cavity than LVA or IP type II anomaly. However, the transmastoid labyrinthotomy approach helps surgeons to avoid facial nerve damage in these cases. This technique does not require posterior tympanotomy and the electrode was not inserted to the scale tympani. It is possible to work in a surgical area away from the facial nerve with the help of this technique. Thus, transmastoid labyrinthotomy approach was used for the five patients who had common cavity and we did not use facial monitoring in patients with common cavities.

First, a retroauricular incision was made, and then the temporal occipital periosteum was elevated. A deep bone groove was made for the implant based on the thickness of the dura mater. Subsequent to the cortical mastoidectomy and visualization of the short process of the incus, the facial recess was detected and enlarged to approach the round window niche. In some cases, finding the location of round window might not be easy and we preferred cochleostomy approach. The internal receiver was placed in the bone groove and covered with temporalis fascia, and then the electrodes were implanted in the scala tympani except common cavities. The surgical approach is via a transmastoid labyrinthotomy with a straight electrode in patients with common cavities. Electrodes will have a position on the periphery of the common cavity with better contact with the neural tissue. The facial recess was sealed with small pieces of muscle to keep the electrodes away from the external auditory canal and preserve the original anatomy. Complete insertion was achieved in all cases. The processor of the cochlear implant was placed into its bed on the temporal bone and fixed with Prolene sutures. The ball electrode of the cochlear implant (Cochlear Ltd., Lane Cove NSW, Australia) was placed under the temporal muscle. However, Medel Cochlear Implant Device (Medel Pulsar Cl 100, Austria) electrode does not have a a separate ball electrode. Except ball electrode, The Medel Cochlear Implant Device (Medel Pulsar Cl 100, Austria) was technically fitted in the same way as the Nucleus Cochlear Implant device (Cochlear Ltd., Lane Cove NSW, Australia). The whole periosteal flap was closed with absorbable sutures and skin with Prolene sutures. The electrical compound action potential (ECAP) and electrical stapedius reflexes were measured intraoperatively after insertion. A Stenvers and transorbital petrous skiagram was acquired during the early postoperative period to assess and record the appropriate placement of the implant. In some patients, a C-arm was used intraoperatively.

Fifteen patients were implanted with the Nucleus Freedom Contour® Advance (Cochlear Ltd., Lane Cove NSW, Australia), nine with the Nucleus CI422 Straight (Cochlear Ltd.), six with the Nucleus CI422 Slim Straight (Cochlear Ltd.), five with the Med-El Standard (Medel Medical Electronics, Innsbruck, Austria), and two with the Med-El Medium (Medel Medical Electronics).

Pure tone/free field audiometry (PTA/FFA), speech audiometry (Speech Recognition Test-SRT, and Word Discrimination Scores, WDS), tympanometry, acoustic reflex measurements, auditory brainstem response (ABR), and

otoacoustic emission (OAE) tests were performed prior to CI. CI candidates were referred to the departments of pediatric psychiatry and pediatric neurology as necessary. The first implant tune-up was performed 3 weeks postoperatively. The patients were followed up monthly for 3–4 months and then scheduled for follow-up every 6 months.

The subjects' age at onset of deafness, duration of deafness, primary mode of communication, type of malformation, preimplantation speech discrimination, age at activation of implant, educational setting, and device type were recorded. Postoperative implanted hearing thresholds were measured at 250, 500, 1000, 2000, 4000, and 6000 Hz in the free field after each mapping session. Speech perception outcomes were acquired using open set monosyllabic and trisyllabic word recognition tests. All CI patients were evaluated with the Categories of Auditory Perception (CAP) [5] and Speech Intelligibility Rating (SIR) tests [6]. Young patients with limited language abilities and understanding of the open set words were only evaluated using the CAP and SIR scales.

The CAP is an eight-point hierarchical scale that evaluates receptive auditory abilities and ranges from no awareness of environmental sound to telephone use with a familiar talker. The SIR is a five-point hierarchical scale that evaluates expressive abilities and ranges from no intelligible speech to connected speech intelligible to all listeners. The word lists were delivered in the free field at 40 dBSL and the patients were tested in an optimally aided condition with their implants. The speech reception scores obtained at the most recent audiometric session were included in the analysis.

Statistical analyses

Normality tests were performed to assess the distribution of the continuous random valuables to determine if nonparametric tests should be used. The Wilcoxon signed rank test was used for paired observations.

The Mann–Whitney *U* test was used for unpaired group comparisons and the Kruskal–Wallis test was used for comparisons between more than two groups. In cases where p was < 0.05, Dunn's test of multiple comparisons was used for correlation analyses. Spearman's rank coefficients were calculated and tested. $p < 0.05$ was considered to indicate statistical significance. Statistical analyses were carried out using the Statistical Package for the Social Sciences 15.0 for Windows (SPSS Inc., Chicago, IL, USA).

Results

The study included 37 patients [19 female (55.9%) and 15 male (44.1%) patients (mean age, 10.6 6 13.7 years; range 0.4–59.8 years)]. Of the 37 children, 16 (43.2%) presented with large vestibular aqueduct (LVA), 10 (27%) with incomplete partition (IP) Type II (IP II), 5 (13.5%) with a common cavity, 4 (10.8%) with IP Type I (IP I) and 2 (5.4%) with cochlear hypoplasia (Table 1). CSF gusher was experienced by six patients (three with LVA, one with IP I, and two with both IP II Mondini malformations and LVA). A wide cochleostomy was created for full insertion of electrode in patients with the Medel Cochlear Implant Device (Medel Pulsar Cl 100, Austria). CSF oozing was encountered in two patients (one with IP II and one with LVA). Cases with CSF gusher required 15–20 min of waiting and ceased completely only after cochlear implant insertion to the cochleostomy site. The cochleostomy site was packed with a piece of muscle and Tissel fibrin glue was used for support. Oozing stopped spontaneously after 5–10 min. Full electrode insertion was achieved in all cases with CSF gusher and oozing. The selection of the appropriate electrode has played an important role in achieving this success. Cases with common cavity or hypoplastic cochlea were implanted with the Nucleus CI422 Straight (Cochlear Ltd.) and the Nucleus CI422 Slim Straight (Cochlear Ltd.). Aside from complete bed rest, no additional precautions such as changing the patient position, hyperosmotic agents, or lumbar puncture were required during or following surgery. None of the children with CSF gusher or oozing experienced dizziness, excessive vomiting, or CSF leakage in the postoperative period. There were no

Table 1 Patients characteristics and their respective populations and selected electrodes

		Selected electrodes
LVA	15 (37.5%)	Nucleus Freedom Contour Advance, Med-El Standard, Med-El Medium
Incomplete partition type-2 (Mondini deformity)	12 (30%)	Nucleus Freedom Contour Advance, Nucleus CI422 Straight
Common cavity	7 (17.5%)	Nucleus CI422 Straight, Nucleus CI422 Slim Straight
Incomplete partition type-1	4 (10%)	Nucleus CI422 Straight, Nucleus CI422 Slim Straight
Cochlear hypoplasia	1 (2.5%)	Nucleus CI422 Straight, Nucleus CI422 Slim Straight
Cochlear nerve abnormality	1 (2.5%)	Nucleus CI422 Slim Straight
Total	40 (100%)	

cases of CSF otorhinorrhea or meningitis during the follow-up period.

Two patients had transient facial paralysis after surgery. One patient had an IP II anomaly (House–Brackmann score of 4) and one patient had cochlear nerve hypoplasia (House–Brackmann score of 3). Oral steroids were given to these two patients and tapered in the second postoperative week. All patients fully recovered within six months. The possible etiological factor was neural edema in these two patients. The burr made edema by working close to the nerve due to the narrowness of the window and caused the dissipated heat to reach the nerve. Two patients with LVA exhibited dehiscence of the tympanic part of the facial nerve; however, no early or late facial paralysis was observed in these patients.

The preoperative cochlear implantation assessment showed that every patient had severe-to-profound sensorineural hearing loss. Some of these patients had progressive hearing loss and partially benefited from well-fitting hearing aids, while some of them had congenital severe hearing loss and did not have any residual hearing. The age at cochlear implantation ranged from 2 and 34 years (average, 11.9 years). Congenital and progressive hearing loss was present in 26 and 11 patients, respectively. All patients received auditory rehabilitation services, 4 received education at a school for the deaf, and 19 attended mainstream schools.

Preoperative ABR waves were obtained from 11 of the 37 patients. For the rest of the patients, no ABR waves were present at the maximum intensity levels for clicks, tonal chirps, or tone bursts.

Table 2 Preoperative and postoperative measurements in all malformation groups for SIR, CAP, PTA, SRT, and WDS tests

Variables	Range	*p* value
SIR		
Preoperative	0–5	0.0001
Postoperative	1–5	
CAP		
Preoperative	0–7	0.0001
Postoperative	1–7	
PTA		
Preoperative	120–75	0.0001
Postoperative	20–120	
SRT		
Preoperative	55–120	0.0001
Postoperative	25–120	
WDS		
Preoperative	0–40	0.0001
Postoperative	0–100	

SIR, CAP, PTA, SRT and WDS were the audiological tests used to evaluate the efficacy of CI in the malformed inner ears. There were statistically significant differences between preoperative and postoperative measurements in all malformation grou for SIR, CAP, PTA, SRT, and WDS tests (p values < 0.0001 for all; Table 2).

The postoperative PTA, SRT, and WDS test results showed significant differences between progressive and congenital sensorineural hearing loss (p values < 0.05 for all) (Table 3). On the other hand, the postoperative CAP and SIR test results revealed no significant differences between these two groups (p values > 0.05 for all).

When the postoperative PTA and SRT values were compared among the different etiological groups according to etiology, the PTA and SRT values were significantly lower in common cavity patients than the LVA patients (p values < 0.01); no significant differences were found among the other etiological groups.

When the relationship between the age of cochlear implant and the PTA, SRT, and WDS values were examined, it was found that the PTA, SRT, and WDS scores increased significantly as the patient's age at cochlear implantation decreased (Table 4).

Sixteen patients had LVA and ten of them had progressive hearing loss. The age at implantation ranged from 1 year 6 months to 22 years 9 months, with a mean age of 9.2 years.

Table 3 PTA, SRT, and WDS test results according to progressive and congenital sensorineural hearing loss

			p value
PTA			
Congenital	40	21	0.005
Progressive	30	8	
SRT			
Congenital	40	100	0.004
Progressive	30	5	
WDS			
Congenital	28	72	0.003
Progressive	72	32	
CAP			
Congenital	7	2	0.492
Progressive	7	1	
SIR			
Congenital	4	2	0.308
Progressive	5	1	

Table 4 The relationship between the age of cochlear implant and the PTA, SRT, and WDS values

	Postoperative PTA	Postoperative SRT	Postoperative WDS
CI age	$R = -0.511$	R: −0.408	R: −0.411
	$p = 0.001$	p: 0.001	p : 0.001

Preoperative hearing thresholds were between 78 and 120+ dBHL (mean: 102 dBHL). Five patients had an air–bone gap at lower frequencies. The mean postoperative PTA was 37 dBHL. Thirteen LVA patients had an understanding of open set speech postoperatively. Open set monosyllabic word recognition scores ranged from 42 to 100%. Almost all LVA patients had increased CAP and SIR scores after implantation.

Ten patients in the study had an IP II anomaly. They received their implants at a mean age of 41 months. Preoperative ABR thresholds were absent for click, 1000 and 4000 Hz tonal stimuli in seven patients. The remaining three patients with an IP II anomaly had preoperative ABR thresholds of 80–100 dBnHL for click and 4000 Hz for tonal chirp. The postoperative mean free field PTA was 38 dBHL. Four patients did not have open set word recognition after the implantation. However, their CAP and SIR scores increased following implantation. Six patients demonstrated open set monosyllabic word recognition with a mean recognition score of 52%.

There were four patients with an IP I anomaly. Two of them had bilateral absent ABR responses to clicks and tonal chirps, others had wave V at high-intensity levels in one ear. The postoperative mean PTA with CI was 38 dBHL. All patients that benefited from implantation had increased CAP and SIR scores and three patients had open-set monosyllabic word recognition.

The five patients with a common cavity received their implants at a mean age of 33.42 months. All five patients had absent ABR waves at the maximum intensity levels. All patients had benefit from their CI. The CAP and SIR scores increased for all patients; only one patient had not an understanding of open set monosyllabic words.

Two patient had a cochlear hypoplasia who received their implants at 2 years and 17 months of age. These patients performed poorly with their implants. Although they received intensive auditory oral rehabilitation, their CAP, SIR scores and speech recognition performance remained low. None of them had open set monosyllabic word recognition. They were able to discriminate two-syllable and three-syllable open set words, but the scores were quite low.

Discussion

Cochlear implant surgery is challenging for surgeons in patients with inner ear anomalies. Despite the surgical difficulties, all of the CI cases reviewed in this study were uneventful. A standard facial recess approach was used for all of the anomalies studied. Nevertheless, facial nerve abnormalities and cerebrospinal fluid gusher may accompany inner ear malformation. Although we used a standard approach, surgeon should be aware of these complications and be able to change techniques as required. A diversity of approaches, including the total trans-canal approach by open [7] or closed tunnel [8], suprameatal approach [9], combined transmeatal approach with posterior tympanotomy [10], and combined transaditus approach [11], have been suggested for use in challenging cases of cochlear implantation. In addition to these approaches, the transmastoid labyrinthotomy approach has been used in patients with a common cavity to find the abnormally located facial nerve [12]. We performed transmastoid labyrinthectomy approaches in our common cavity patients. Although facial nerve monitoring provides significant assistance to the surgeon, we believe that knowledge of the anatomical course of the facial nerve and the surgeon's experience are more important.

The opening to the cochlea for the electrode insertion was conducted via the round window in 26 cases (70.3%), via cochleostomy in 6 cases (16.2%) and via the transmastoid labyrinthotomy approach in 5 cases (13.5%). Sun et al. assessed whether CI using the round window approach ensured better preservation of residual hearing than the cochleostomy approach and found no differences between the two techniques [13]. In addition to the cochleostomy and round window approaches, electrode selection is very important in patients with cochlear anomalies. The surgeon should not aim to insert all of the electrodes in patients with cochlear anomalies [14]. The surgeon should aim to insert as much electrode as possible.

The audiological data collected in our study showed that children with inner ear malformations may receive remarkable benefit from CI. In our study, a significant improvement was found postoperatively in all cochlear implant groups in all audiological tests. These postoperative improvements are consistent with the literature [15, 16]. When within-group statistical tests were performed, the success rate of the cochlear implants was lower in patients with cochlear hypoplasia than in those without. In the cochlear hypoplasia group, the CAP and SIR scores, as well as the speech discrimination scores, remained low in comparison with the other malformation groups.

A direct comparison of all data was not possible because there were too many cochlear anomaly subgroups in our study.

It is well known that the ability to distinguish speech is one of the most important auditory skills demonstrating the success of the implant [17, 18]. Therefore, when we compared the postoperative speech discrimination skills of the groups in terms of revealing the success of the cochlear implants, it was observed that the group with the highest postoperative speech discrimination score was the LVA group, followed by the IP 1, IP 2, common cavity and cochlear hypoplasia groups. When we performed a similar comparison for the auditory performance categories, it was observed that the group with the highest postoperative CAP

score was the LVA group, followed by the common cavity, IP 1, IP 2 and cochlear hypoplasia groups. Based on these results, it can be concluded that the group that benefitted the most from CI was the LVA group. Most subjects in the LVA group had postlingual progressive hearing loss, but the subjects with other inner ear anomalies had congenital prelingual severe-to-profound hearing loss. Therefore, it is not surprising that the postoperative speech and auditory performance of the subjects in the LVA group were higher than in the other groups.

There are some important limitations to consider when interpreting the results of our study. In retrospective studies, only the results of the patients whose data are collected during a specific time period are considered. However, since the patients in our study did not have the same age at implantation, the evaluation time and age at which they were evaluated differed. In addition, because there was an insufficient number of patients with inner ear anomalies who underwent CI, we were unable to form groups with equal numbers of patients, thus limiting interpretation of the results. In a similar study of cochlear implant patients with different inner ear anomaly etiologies, it was found that groups could not be formed from heterogeneous groups due to similar limitations. As a result, the results were difficult to interpret. However, unlike our study, the evaluation intervals in the previous study were fixed, and short-, medium- and long-term results were evaluated. Despite the limitations of interpreting the results, it was concluded that the postoperative results of speech testing in cochlear implant users with inner ear anomalies were significantly improved [19]. Similarly, in another study, it was determined that cochlear implants were successful in children with inner ear malformations and that the implants provided important auditory benefits, but that the various IPs of the internal ear malformations can result in a different prognosis in terms of auditory performance [1]. We know that there are many other factors that affect the success of cochlear implants. Onset of deafness, degree of residual hearing, duration of hearing loss, age at CI, duration of cochlear implant use, and preoperative auditory performance are some of the factors that affect the success of cochlear implants [17, 18]. Since the groups included in the present study were not homogeneous in terms of these factors, it was not possible to base implant success on a single etiological factor. Therefore, when we examined the preoperative auditory skills of the LVA and common cavity groups, which we noted had the most benefit from the implant, it was found that these groups had also better scores in comparison with other groups in terms of speech discrimination and auditory performance in the preoperative period. In particular, the LVA subgroup with progressive hearing loss had better results as expected. To better understand the potential for success and benefit of the implant, it is hypothesized that the difference between the preoperative and the postoperative test scores could provide insight into the success of the implant. When we consider the results from this point of view, the groups with the highest improvement in terms of SIR in the postoperative period were the IP I and common cavity groups, followed by the IP II, cochlear hypoplasia, and LVA groups. When a similar comparison was made in terms of the CAP score, it was observed that the most improvement was found in the IP I group, followed by the common cavity, IP II, cochlear hypoplasia, and LVA groups. As a result, when the difference between pre-implant and post-implant performance was considered, the groups with the most improvement were the IP I and common cavity groups; when only the postoperative performance was considered, the groups that showed the most improvement were the LVA and common cavity groups. Therefore, it can be concluded that cochlear implants can be successful in patients with almost all inner ear anomalies and etiologies. Despite concerns about electrode insertion and abnormal electrical stimulation, it can be concluded that implantation in subjects with inner ear malformations can be implemented successfully. In another study that assessed clinical experiences such as surgical treatments and techniques, imaging findings and outcomes, electrode placement, and hearing following multichannel CI in children with internal ear malformations, children with an incomplete partition, enlarged vestibule, or membranous anomaly were found to have similar outcomes as children with normal cochleas. Additionally, it has been suggested that specific surgical techniques can be effective for children with a common cavity, though the results are less pronounced [20].

In conclusion, inner ear anomalies are not a contraindication for cochlear implants, and patients in this group can experience significant improvement in terms of auditory development and recipient expressive language development in comparison with the pre-implant state. To make a more accurate and reliable assessment of the findings, it is recommended that studies with larger sample sizes and more homogeneous patient groups be conducted.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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