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Original Article

The Results of Cochlear Implantation in the Inner Ear Malformations

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BACKGROUND: Cochlear implants are arguably the most successful neural prosthesis today. Cochlear implantation has several difficulties in patients with internal ear anomalies. This study was performed to analyze intraoperative, postoperative findings, and auditory performance of 55 patients who had inner ear malformations and were treated with cochlear implants at Otorhinolaryngology Department of Çukurova University Hospital.

METHODS: Auditory performances were analyzed in 30 of 55 patients. Patients with cochlear anomalies were evaluated as group I, patients with vestibular malformation as group II, and patients with the normal bone labyrinth as group III. Listening progress profiles and meaningful auditory integration scale tests were used to determine performances.

RESULTS: Comparison between the listening progress profiles test performance of the groups at 12th and 18th month of group I was significantly lower than other groups ($P < .05$) and reached at the same level after the 24th month. Comparison between the meaningful auditory integration scale test performance of groups at 24th and 36th month of group I was significantly lower than other groups ($P < .05$). Perilymph gusher was observed in 3 patients who had incomplete partition I malformation. Oozing was observed in 50% of incomplete partition II patients. Facial nerve traced a variant course in 4 of 55 patients and 6 patients had postoperative meningitis.

CONCLUSION: Initially patients with inner ear anomalies showed that the level of language development was worse than patients with normal bone anatomy. However, it was shown that they both reached the same point as a result. Facial nerve anomaly and meningitis risk is higher in patients with inner ear malformations.

KEYWORDS: Cochlear Implant, inner ear malformation, language development, meningitis, perilymph gusher

INTRODUCTION

Cochlear implants (CI) are arguably the most successful neural prosthesis to date. Scientific, surgical, and technological advances in CI technology have facilitated a transformative approach toward rehabilitation of hearing loss in a pediatric population. Clinical outcomes vary based on several patient-related factors such as age at implantation, onset and duration of hearing loss, cochleovestibular anatomy, physiology of the auditory nerve, presence of neurodevelopmental disorder, level of psychosocial support, and the quality of postoperative rehabilitation efforts.[1-3] The percentage of inner ear malformations in children who have undergone CI ranges between 6.9% and 35%.[4,5]

Any delay in the embryological processes of the inner ear may result in an incomplete formation of its structures or in the formation of a smaller size cochlea. Using the most commonly accepted classification of Jackler et al. Sennaroğlu and Saatçi reclassified the most commonly accepted inner ear anomalies based on state-of-art computerized tomography (CT) and magnetic resonance imaging (MRI) findings. In their study, cochlear, vestibular, semicircular canal (SCC), internal acoustic canal (IAC), and vestibular-cochlear aqueduct malformations were classified into subgroups. As a result of this examination, cochlear malformations were divided into 7 groups (Figure 1) as Michel deformity, common cavity, cochlear aplasia, hypoplasic cochlea, incomplete partition type I (IP-I), incomplete partition type II (IP-II/Mondini deformity), and incomplete partition type III (IP-III); vestibular malformations were divided into 3 groups as vestibular dilatation, SCC malformations, and IAC anomalies; and vestibular and cochlear aqueduct

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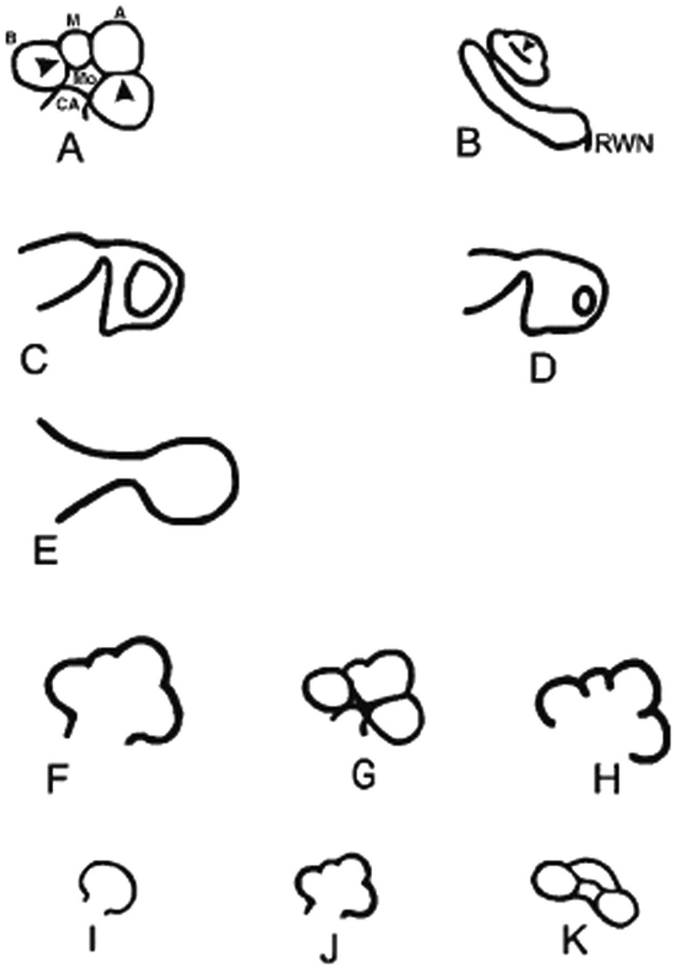

Figure 1. Schematic representation of the normal cochlea and cochlear malformations. (A) Normal cochlea, midmodiolar section. Mo, modiolus; CA, cochlear aperture; B, basal turn; M, middle turn; A, apical turn; arrowheads, interscalar septa. (B) Normal cochlea, inferior section passing through the round window niche (RWN). Arrowhead, interscalar septum between middle and apical turns. (C) Cochlear aplasia with normal vestibule. (D) Cochlear aplasia with enlarged vestibule. (E) Common cavity. (F) Incomplete partition type I. (G) Incomplete partition type II. (H) Incomplete partition type III. (I) Cochlear hypoplasia, bud type (type I). (J) Cochlear hypoplasia, cystic cochlea type (type II). (K) Cochlear hypoplasia, with less than 2 turns (type III).[9]

malformations were divided into 2 groups as vestibular aqueduct anomalies and cochlear aqueduct anomalies.

Cochlear implantation surgery can be performed in all patients with malformation except in those with cochlear aplasia, Michel deformity, and cochlear nerve agenesis. Various complications, such as cerebrospinal fluid leakage, problems in electrode placement, or facial nerve stimulation during activation of the implant, have been reported in CI surgery for patients with inner ear malformations.[6-8] The aim of this study was to analyze intraoperative findings, postoperative findings, and the auditory performance with inner ear malformations with CI due to bilateral total hearing loss.

METHODS

This study was performed to analyze intraoperative, postoperative findings, and auditory performance of 55 patients who had inner ear malformations and were treated with cochlear implants at Çukurova University Hospital.

In this retrospective study, 840 patients who underwent cochlear implantation between July 2000 and July 2014 were analyzed at Otorhinolaryngology Department of Çukurova University Hospital. Fifty-five out of 840 patients who were diagnosed with inner ear malformations were included in the study (Table 1). We implanted the Cochlear Nucleus in 28 patients, MED-EL in 22 patients, and Advanced Bionics CI system in 5 patients. Two patients who underwent brainstem implant were excluded. Computerized tomography (CT) and MRI scans of these patients were obtained from the local database. Inner ear malformations were diagnosed by radiologists. Patients were classified in accordance with the Sennaroglu and Saatci[10] classification system.

In all patients, we performed CI surgery via the classical transmastoid-facial recess approach. In 4 patients, the vertical segment of the fascial nerve was located anteromedially toward the promontory, but we did not need to modify the surgical approach. In transmastoid-facial recess approach, cochleostomy was performed through the triangular space between the fossa incudis, facial canal, and the ear canal. Cochlear implant electrode placement into the scala tympani was performed using the round window technique in 53 (96%) patients and the cochleostomy technique in 2 (4%) patients. The Cochleostomy technique was used in 2 patients because the round window could not be observed.

In addition, the auditory performances of 30 patients with inner ear malformations who regularly used CI devices and continued their training regularly for 12–36 months were evaluated. These results were compared with the control group that included 30 patients who had undergone CI surgery due to bilateral total hearing loss without internal ear anomaly according to CT and MR images. Fifteen patients with cochlear anomalies constituted group I (Table 2), 15 patients with vestibular anomalies and vestibular aqueduct anomalies as group II (Table 3), and 30 control patients with normal inner ear anatomy constituted the group III.

Evaluation of Auditory Responses to Speech test battery was utilized to analyze the auditory performance of the patients. This test battery was composed of 2-syllable open-ended words, 2-syllable closed-ended word test,[11] listening progress profile (LIP),[12,13] Monosyllable-Trochee-Polysyllable test,[14] closed-ended sentence test,[15] Glendonald Auditory Screening Procedure test,[16] Meaningful Auditory Integration Scale (MAIS), and Meaningful Use of Speech Scale.[17]

In our study, the results of only LIP and MAIS were analyzed from this battery. Listening progress profile test was applied in the preoperative period, at the first fitting, and 1st, 3rd, 6th, 12th, 18th, 24th, and 36th months. Meaningful Auditory Integration Scale test was applied in the preoperative period and 3rd, 6th, 12th, 18th, 24th, and 36th months.

Mean, standard deviation, median lowest, highest, frequency, and ratio values were used in the descriptive statistics of the data. The distribution of the variables was measured by the Kolmogorov–Smirnov test. Mann–Whitney *U* test was used in the analysis of the quantitative

Table 1. General Information of Patients with Internal Ear Anomalies

Number	Operation Age (Month)	Gender	Radiological Findings		Operation Side	Operation Findings
			Right Ear	Left Ear		
1	17	M	CC	CC	R	Oozing round window could not be seen promontorium cochleostomy
2	14	F	CC	CC	R	Oozing round window could not be seen promontorium cochleostomy
3	33	F	CC	IP-I	L	Gusher
4	168	F	IP-I	IP-I	R	Gusher
5	48	F	IP-I	IP-I	R	Anterior and lateral placement of facial nerve
6	31	F	IP-I	Michel aplasia	R	No findings
7	13	F	IP-I	IP-I	L	No findings
8	31	F	IP-I	IP-I	L	Gusher. anterior and lateral placement of the facial nerve
9	22	M	IP-II	IP-II	R	Oozing
10	48	F	IP TIP II	IP TIP II	L	No findings
11	132	F	IP TIP II	IP TIP II	R	No findings
12	126	F	IP TIP II	IP TIP II	R	Oozing
13	15	F	IP TIP II	IP TIP II	R	Oozing
14	65	M	IP TIP II	IP TIP II	R	Oozing
15	157	F	IP TIP II	IP TIP II	R	No findings
16	16	M	IP TIP II	IP TIP II	R	No findings
17	58	M	IP TIP II	IP TIP II	L	No findings
18	14	M	IP TIP II	IP TIP II	R	No findings
19	25	F	IP TIP II	IP TIP II	R	Oozing
20	402	F	MİCHEL APLASIA	IP TIP II	L	Oozing
21	190	M	LVA	LVA	R	Oozing
22	77	M	LVA	LVA	R	No findings
23	30	M	LVA	LVA	R	Oozing
24	371	M	LVA	LVA	L	No findings
25	138	F	LVA	LVA	L	No findings
26	186	F	LVA	LVA	R	Oozing
27	50	M	LVA	LVA	R	No findings
28	26	F	LVA	LVA	R	No findings
29	68	M	LVA	LVA	R	No findings
30	300	F	LVA	LVA	R	No findings
31	42	F	LVA	LVA	R	Oozing
32	221	M	LVA	LVA	R	Oozing
33	160	M	LVA	LVA	L	Oozing
34	232	F	LVA	LVA	R	No findings
35	167	M	LVA	LVA	L	No findings
36	42	M	LVA	LVA	R	No findings
37	101	F	LVA	LVA	R	No findings
38	67	F	LVA	LVA	R	No findings
39	129	F	LVA	LVA	R	Oozing
40	59	F	LVA	LVA	L	No findings
41	444	F	LVA	LVA	R	No findings
42	34	M	LVA	LVA	L	Oozing

Table 1. General Information of Patients with Internal Ear Anomalies (*Continued*)

Number	Operation Age (Month)	Gender	Radiological Findings		Operation Side	Operation Findings
			Right Ear	Left Ear		
43	15	F	LVA	LVA	R	No findings
44	57	M	LVA	LVA	R	Oozing
45	77	F	LVA	LVA	R	Oozing
46	12	M	LVA	LVA	R	No findings
47	15	M	SCC aplasia	SCC aplasia	R	No findings
48	30	F	Cochlear hypoplasia	Cochlear hypoplasia	R	No findings
49	30	F	Cochlear hypoplasia	Cochlear hypoplasia	L	No findings
50	17	F	SCC aplasia	SCC aplasia	R	No findings
51	21	F	SCC aplasia	SCC aplasia	R	No findings
52	23	F	SCC aplasia	SCC aplasia	R	Anterior and lateral placement of facial nerve
53	157	F	SCC aplasia	SCC aplasia	R	No findings
54	41	F	SCC aplasia	SCC aplasia	R	No findings
55	648	M	SCC aplasia	SCC aplasia	R	No findings

CC, common cavity; IP-I, incomplete partition type I; IP-II, incomplete partition type II; LVA, large vestibular aqueduct; SCC aplasia, semicircular canal aplasia; R, right ear; L, left ear.

Table 2. Group I Patients with Cochlear Anomalies Whose Auditory Performance Is Analyzed

Number	Gender	Operation Age (Month)	Operation Side	Cochlear Anomaly Type
1	F	31	R	IP-I
2	F	33	L	IP-I
3	F	48	R	IP-I
4	F	31	L	IP-I
5	F	14	R	CC
6	M	17	R	CC
7	F	13	R	IP-I
8	M	15	R	IP-II
9	F	157	R	IP-II
10	M	50	R	IP-II
11	F	126	R	IP-II
12	F	48	L	IP-II
13	M	65	R	IP-II
14	F	25	R	IP-II
15	F	132	R	IP-II

CC, common cavity, IP-I, incomplete partition type I; IP-II, incomplete partition type II; R, right ear; L, left ear.

data. Wilcoxon was used in the analysis of recurrent measurements. Chi-square test was used to analyze the qualitative data, and the Fischer test was used when chi-square test conditions were not met. Statistical Package for the Social Sciences Version 22.0 program was used in the analyses.

Ethics committee approval was received from Çukurova University (Approval no: 40/22, Approval date: 06.03.2015). Written informed consent was obtained from all participants who participated in this study.

RESULTS

Twenty-one (38%) males and 34 (62%) female patients participated in this study. The average age of the patients was 104.5 months. The CI was applied to the right ear of 43 patients (78%) and to the left ear of 12 patients (22%).

Computerized tomography (CT) and MRI scans of the whole participants revealed that 2 common cavities, 6 incomplete partition type

Table 3. Group II Patients with Vestibular Anomalies Whose Auditory Performance Is Analyzed

Number	Gender	Operation Age (Month)	Operation Side	Vestibular Anomaly Type
1	F	138	L	LVA
2	F	41	R	SCC aplasia
3	F	129	R	LVA
4	F	21	R	SCC aplasia
5	M	160	L	LVA
6	M	167	L	LVA
7	M	190	R	LVA
8	M	50	R	LVA
9	F	101	R	LVA
10	M	34	L	LVA
11	F	186	R	SCC aplasia
12	F	26	R	LVA
13	F	17	R	SCC aplasia
14	F	67	R	LVA
15	M	57	R	LVA

LVA, large vestibular aqueduct; SCC aplasia, semicircular canal aplasia; R, right ear; L, left ear.

I (IP-I), 12 incomplete partition type II (IP-II), 2 cochlear hypoplasia, 26 large vestibular aqueducts (LVA), 7 SCC aplasia were present.

Gusher was observed in 3 out of 55 patients (6%). All these patients displayed IP-I anomalies. While there was no gusher in patients with common cavity, cochlear hypoplasia, IP-II, large vestibular aqueduct, and SCC aplasia, oozing was observed in 50% of patients with IP-II anomalies and 39% of patients with large vestibular aqueduct.

Facial nerve paresis occurred merely in 1 of 55 patients, but it was completely resolved after surgery. In this patient, facial nerve anomaly had not been observed during perioperative period.

Meningitis occurred in 6 patients (11%) postoperatively. One of these patients had a common cavity, 2 had IP-I, 2 had IP-II, and 1 had SCC aplasia. The cause of 5 out of 6 cases was thought to be acute otitis; however, the cause of meningitis in 1 patient was the spontaneous cerebrospinal fluid leak from the nonoperated ear.

Of the 15 patients in group I who had cochlear anomalies and underwent auditory performance analysis, 11 were female (73%) and 4 were male (27%). The mean age was 52 months. Of the 15 patients in group II who had vestibular anomalies and performed auditory performance analysis, 9 were female (60%) and 6 were male (40%). The mean age of the patients was 81.4 months. Cochlear implants were applied to the right ear of 11 patients (73%) and to the left ear of 4 patients (27%). In group III, 10 (33%) of the control group who had normal inner ear anatomy were female and 20 (66%) were male. The mean age of the patients was 49.1 months. Cochlear implants were applied to the right ear of 25 patients (83%) and to the left ear of 5 patients (17%).

In all 3 groups after cochlear implantation, the performance of basic auditory skills such as patient response, voice discrimination, and voice recognition increased steadily after cochlear implantation. In the 12th month, basic skill improvement reached 90% in patients with vestibular anomalies and normal inner ear anatomic patients; however, this improvement was reached in the 18th month in cochlear anomaly patients.

In the LIP test, when the groups were compared, it has seen that the performance improvement curves of the 3 groups were continuously increasing. In group I, patients with cochlear anomalies, this development seemed to be slower compared to other groups. This improvement was not statistically significant ($P > .05$) in group I in the first fitting, postoperative 1st, 3rd, 6th, 24th, and 36th months when compared to groups II and III. However, at 12th and 18th months, the test values in group I were significantly lower than the groups II and III ($P < .05$). In groups II and III, during the postoperative 12th and 18th months, the LIP test did not differ significantly ($P > .05$) (Figure 2).

In the LIP test, we compared the performance in group I between common cavity, IP-I, and IP-II. The improvement was statistically significant at the 18th month between IP-II and common cavity and at 24th and 36th months between common cavity, IP-I, and IP-II. In common cavity patients, LIP test improvement seemed to be slower compared to other anomalies. The mean MAIS values of all groups at postoperative 3rd, 6th, 12th, and 18th months were not statistically significant ($P > .05$) but were lower in group I. However, at postoperative 24th

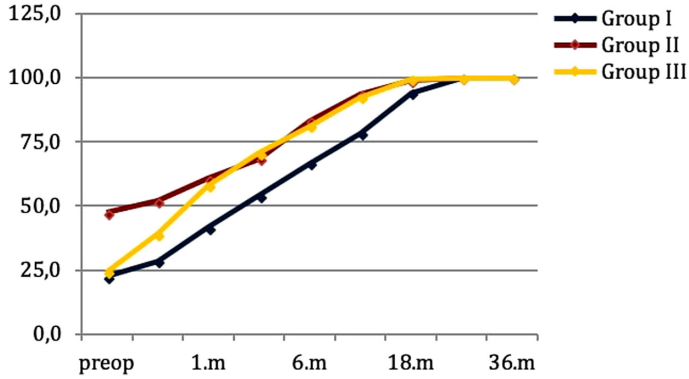

Figure 2. Change in listening progress profile test performance percentage over time before and after the operation.

and 36th months, the MAIS score was significantly lower ($P < .05$) in group I compared to groups II and III. In groups II and III, the 24th and 36th month MAIS values were not significantly different ($P > .05$). When the groups were compared, it was seen that the performance improvement curves of all 3 groups are continuously increasing. In groups II and III, the MAIS test performance increased to 90% in the 18th month. In group I, the MAIS test performance increased to 90% at the 24th month, but it was significantly lower ($P < .05$) than the groups II and III. The improvement in group I reached that in groups II and III only at 36th months. There were no significant differences ($P > .05$) between groups II and III preoperatively, postoperatively at 3rd, 6th, 12th, 18th, 24th, and 36th month MAIS values (Figure 3).

In the MAIS test, we compared the performance in group I between common cavity, IP-I, and IP-II. There were no statistically significant differences between all patients.

DISCUSSION

Recently, owing to advancing technology and imaging methods, inner ear anomalies have been identified as a common cause of congenital hearing loss. For example, inner ear anomaly in the temporal bone has been reported in 20% of patients with congenital hearing loss.[18]

Initially, the inner ear malformations were seen as a contraindication to cochlear implantation, but nowadays, this view has completely changed. Cochlear implant surgery can be performed in all patients with inner ear malformation except cochlear aplasia, Michel aplasia, and cochlear nerve agenesis. Meningitis and facial nerve anomalies have been reported more frequently in patients with inner ear

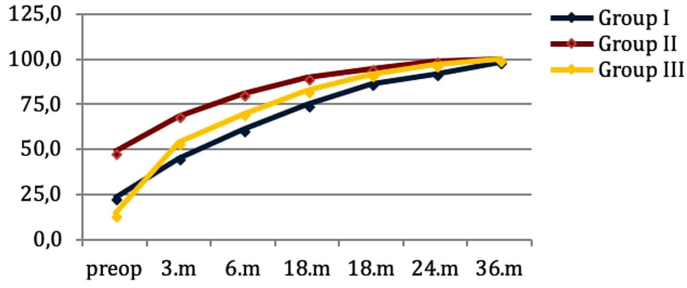

Figure 3. Change in Meaningful Auditory Integration Scale test performance percentage over time before and after the operation.

anomalies than in normal patients, and it has been seen that these patients have fewer auditory outcomes.[19-24] Four patients with cochleovestibular anomalies in our study had abnormal facial nerve anatomy (7.2%). Sennaroglu et al[19] observed abnormal facial nerve anatomy in 4 out of 20 patients in their study. In a study conducted by Çatlı et al in 2014, 4 out 21 patients had anterior and lateral placement of the facial nerve. To 2 patients with a common cavity in their study groups, they have preferred the transcanal approach instead of the standard facial recess method due to the abnormal course of the facial nerve to 2 patients.[20] In a study conducted by McElveen et al[25], it is seen that 3 patients have been reported to have abnormal facial nerve involvement, all of which were found to have common cavity deformities.

Another problem that may be encountered in patients with inner ear malformations is perilymph fistulas. This usually occurs due to defects in the lateral end of the internal auditory canal. If perilymph gusher occurs during surgery, the cochleostomy should be closed completely around the electrode to avoid the risk of meningitis. In 1995, Tucci et al[26] observed gusher in 3 out of 6 patients with inner ear malformations (1 hypoplastic cochlea, 1 IP, 1 common cavity). In our study, gusher was observed in 2 patients with common cavities and was not observed in any patient with the hypoplastic cochlea, whereas gusher was observed in 3 out of 6 patients (50%) with IP-I anomalies. Similar to Sennaroglu et al[19] and Au and Gibsons studies,[27] no gusher was observed in any of the patients with large vestibular aqueducts in our study.

Meningitis is a life-threatening complication of cochlear implantation surgery. A study by Biernath et al[28] have shown that the risk of meningitis in patients with CI was 3-fold higher than in the normal population. In a study by Phelps et al.[29] recurrent meningitis has been observed in 4 out of 20 patients who have CI and inner ear malformations. Weber et al[30] has shown that 3 patients had meningitis preoperatively in a study of 12 patients with inner ear anomalies in 1995. In addition, Suzuki et al have shown histopathologic features of a CI patient with Mondini deformity, who died of tympanogenic meningitis. In this study, they saw that the infection spread through the round window into the inner ear.[31]

In our study, meningitis was observed in 6 out of 55 patients (11%) who had inner ear malformations (1 patient with common cavity, 2 patients with IP-I, 2 patients with IP-II, and 1 patient with SCC aplasia). In our clinic, all the patients with meningitis after cochlear implantation had inner ear malformations.

Postoperative hearing and speech rehabilitation programs are of utmost importance for all patients who get cochlear implants. With these programs, patients with cochlear anomalies can also develop their auditory performance at a level observed in patients with normal inner ear anatomy.[32,33] Current studies indicate that early implantation is necessary to gain better communication skills in bilateral total hearing loss.[34] It has been shown that pediatric patients should be operated on early in order to maximize profits.[35] Özdemir et al[36] found that there was a negative correlation between age at the time of operation and development of auditory performance: test scores have increased more rapidly in the small age group. Kim et al[37] have observed that cochlear implantation is beneficial for patients with inner ear malformations and that there was a slight delay in auditory skills in the early post-implantation period compared to normal inner ear anatomic patients. Chen et al[38] have observed that auditory skills were less in the first year of patients with Mondini deformity than in normal anatomic patients and that they had acquired normal anatomic patients in the following period. Our findings are in line with these studies.

In addition, similar to our study, recent studies also have shown that patients with large vestibular aqueducts and patients with normal inner ear anatomy have similar auditory performance.[34,39] The following conditions should be taken into account to avoid mortal complications such as meningitis: informing the patient's relatives about all details and risks of the operation, interpreting radiological images carefully, vaccination before the operation, facial monitoring in the course of operation, close follow-up for rhinorrhea in the postoperative period.

CONCLUSION

As a result, auditory outcomes of cochlear implantation in patients with inner ear malformations can reach that of CI patients without inner ear anomalies in time. However, it should be known that the facial nerve anomalies and the risk of meningitis are higher in patients with inner ear malformations compared to CI patients without inner ear malformations.

Ethics Committee Approval: Ethical committee approval was received from the Ethics Committee of Çukurova University (Approval no: 40/22, Approval date: 06.03.2015).

Informed Consent: Written informed consent was obtained from all participants who participated in this study.

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