Annals of Clinical Otolaryngology

Research Article
Published: 18 Jul, 2018

Cochlear Implantation in Patients Afflicted with Inner Ear Malformations - The Pecs Experience

Bako P[1], Nemeth A[1], Molnar K[2], Toth T[1], Harmat K[1], Rath G[3] and Gerlinger I[1]*

[1]*Department of Otorhinolaryngology, Head and Neck Surgery, University of Pécs, Hungary*

[2]*Department of Radiology, University of Pécs, Hungary*

[3]*Department of Pediatrics, University of Pécs, Hungary*

Abstract

Aim: To successfully analyze the surgical and audiological results of cochlear implantation in patients afflicted with inner ear malformations. This study briefly presents our surgical strategies, the selected implants and the audiological results.

Methodology: Objective hearing threshold was measured using an auditory brainstem response and middle latency response audiometry. Structural imaging of high resolution temporal bone computer tomography and inner ear MR scan was performed in support of all patients. Images revealed bilateral cochlear malformation in 7 patients. 3 ears with common cavity deformity, 2 ears with incomplete partition Type I, and 3 ears suffering from cochlear hypoplasia, were all rehabilitated using cochlear implantation. The type of the electrode and the surgical technique were individually determined reflective of the type of malformation. The success of the surgery and potential postoperative complications were duly noted and fully registered. Postoperative audiological measurements were performed and the results were analyzed.

Results: The average audiological follow-up was 40.3 months. The average free field, pure-tone threshold of speech frequencies (0.5 kHz to 3 kHz) gained postoperatively were within the range of 32.5 dB to 41.6 dB. In two of the procedures, a cerebrospinal fluid gusher did occur and was successfully managed intra-operatively, all without any postoperative complications.

Conclusion: Temporal bone high resolution CT and inner ear MRI imaging are mandatory to detect and characterize the inner ear malformation. Early implantation of malformed cochlea is crucial towards achieving ideal postoperative audiological results. Notably, an intra-operative gusher is not regarded as a contraindication of the cochlear implantation.

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Correspondence:
Bako P, Department of Otorhinolaryngology, Head and Neck Surgery, University of Pécs, 2 Munkácsy Street, 7621 Pécs, Hungary, E-mail: drbakopeter@gmail.com

Received Date: *18 Jun 2018*
Accepted Date: *11 Jul 2018*
Published Date: *18 Jul 2018*

Citation:
Bako P, Nemeth A, Molnar K, Toth T, Harmat K, Rath G, et al. Cochlear Implantation in Patients Afflicted with Inner Ear Malformations - The Pecs Experience. Ann Clin Otolaryngol. 2018; 3(1): 1033.

Introduction

Developmental anomalies of the bony labyrinth are found within 20% of patients diagnosed with congenital sensorineural hearing loss [1]. Causes include both genetic and environmental factors, such as infection, trauma or teratogen drugs [2].

The first classification of Inner Ear Malformations (IEM) was published by Jackler et al. In 1987, using polytomographic images [1]. The authors suggested how an arrest in any developmental phase leads to a varied appearance of the inner ear. Today, obvious, medical progress in radiology techniques has resulted in a more comprehensive understanding of the labyrinth, thus, the classification of Jackler was modified in accordance to our extended breadth of knowledge. The current classification is based on publications of Sennaroglu, et al. [3-5] and summarized in Table 1.

The development of the bony cochlea is complete at the end of the 8th gestation week. All of the malformations are the result of developmental failure appearing during this time-period. The severity of the deformity is dependent upon the phase in which development has ceased and fundamentally influences varied options in support of hearing rehabilitation. Therefore, a High Resolution CT (HRCT) scan of the temporal bone in consideration of the anatomical structures, such as the bony labyrinth, internal auditory canal and the Fallopian canal, and the sagittal T2 weighted MR scan in support of the evaluation of the auditory nerve, proves to be crucial and is the recommended form of radiological diagnostics in all patients prior to implantation [6]. Cochlear implantation is contraindicated in Michel deformity, the complete absence of cochlear and vestibular elements. In these cases only the Auditory Brainstem Implantation (ABI) can be

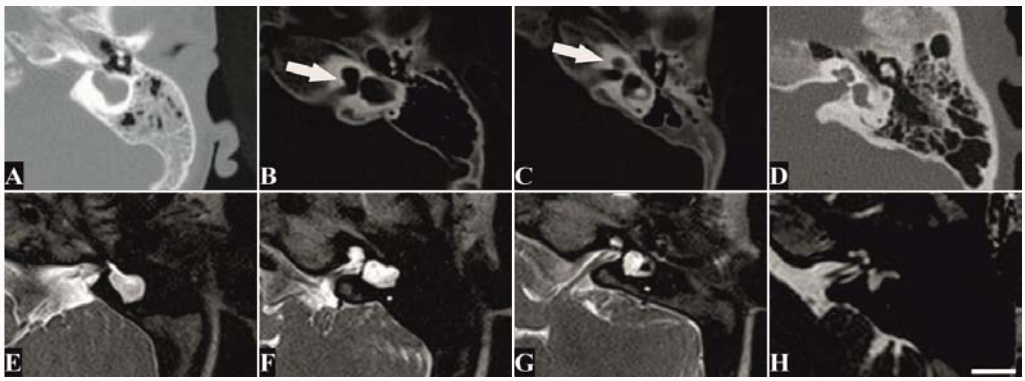

Figure 1: Axial temporal bone HRCT (1A-1C) and T2-weighted MRI (1E-1G) images of different cochlear malformations compared to a normal developed cochlea (1D, 1H). 1A, 1E: common cavity; 1B, 1F: incomplete partition type I; 1C, 1G: cochlear hypoplasia type I; 1D, 1H: normal cochlea. The white arrow on 1B and 1C indicates the enlarged communication between the cochlea and the internal auditory canal. All images are from our patients. Scale bar: 1 cm.

reasonably considered an option suitable for rehabilitation [7]. In other cochlear malformations, cochlear implantation is theoretically possible. The detailed preoperative radiological evaluation serves to avoid potential surgical complications and operative risks, such as Cerebrospinal Fluid (CSF) leakage and/or facial nerve damage.

This study presents patients diagnosed and rehabilitated with cochlear deformities within our department. We summarized the implanted electrodes and surgical techniques, potential intra-operatively managed complications and demonstrated promising audiological results.

Material and Methods

Patients

Between 2009 and 2015 8 cochlear implantations on 7 patients suffering from varying levels of cochlear deformity (6 females and1 male) were performed. The average age of the patients at time of operation was 9.7 years (from a spread of 2.8 to 19). Sequential bilateral implantation was performed in 1 patient (Patient No. 5 in Table 2) separated by a time span of 5.4 years. The remaining patients were operated on the left ear.

Six patients highlighted an uneventful perinatal history without any other congenital deformities. In the case of Patient No. 5, a serious herpes simplex infection was diagnosed during the pregnancy and, as a result, the patient was born suffering from multiple congenital deformities, including cochlear hypoplasia, tracheal stenosis, eye atrophy and patent ductus arteriosus.

Patient No. 2 had an unsuccessful bilateral implantation performed in another department and suffered intra-operative facial nerve palsy on the left side (House-Brackmann grade III), which eventually improved but some facial contracture remained.

Preoperatively, no Auditory Brainstem Response (ABR) waves could be detected in any of the selected ears of 6 patients. Patient No. 7 showed an ABR threshold of 80 dB in the selected ear. In the contralateral ear, a threshold of 70 dB was found in 3 patients (Patient No. 3, 6 and 7).

The Middle Latency Response (MLR) audiometry detected a threshold of 50 dB in both ears in Patient No. 4, and a threshold of 80 dB in the contralateral ear in Patient No. 1 and 6.

Preoperative pure-tone audiometry was performed in patients older than 6 years of age (Patient 3, 5, 6, 7). In Patient No. 6, a pancochlear hearing remnant at the threshold of 100-110 dB could be detected in both ears, as well as the same pattern discovered in Patient No. 3 in the selected ear with a higher threshold (threshold of 80 db to 95 dB in the lower frequencies) in the contralateral ear. No threshold could be established in Patients No. 5 and 7.

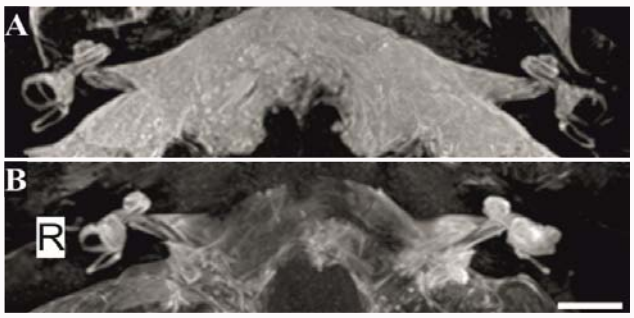

Figure 2: 3D reconstructed T2-weighted inner ear MRI images of normal and malformed cochleae.2A. Normal cochlear morphology. 2B. Asymmetric cochlear morphology. Right ear: cochlear hypoplasia, left ear: incomplete partition type I. R indicates the right side. Scale bar: 1 cm.

All patients were fitted with a hearing aid prior to the cochlear implantation; however, no measurable speech development was achieved.

Cochlear malformations

Distinctively, as a vital role in preoperative assessments, in all cases, temporal bone HRCT and inner ear MRI were requested. On the side of the operation, a common cavity deformity was found in three patients, cochlear hypoplasia occurred in three patients, and incomplete partition Type I found in two patients. In all cases, the deformities exhibited a symmetric appearance, except in the case of one patient, which featured an incomplete partition Type I on the operated side, and cochlear hypoplasia on the contralateral side.

Figure 1 shows axial HRCT and T2-weighted inner ear MRI images of the different malformations found in our cases: common cavity (Figure 1A and 1E), incomplete partition type I (Figure 1B and 1F) and cochlear hypoplasia (Figure 1C, 1G). Normal cochlear and inner ear morphology is shown in Figure 1D and 1H. In common cavity deformity, the cochlea and the vestibule appeared as a common cystic cavity. The opening of the internal auditory canal is typically located in the middle part of the cyst. Incomplete partition Type I deformity shows a separated cochlea and vestibule. The cochlea is

Table 1: The latest classification of bony inner ear malformations.

I. Cochlear malformations	Michel deformity Cochlear aplasia Common cavity deformity Incomplete partition type I. Cochlear hypoplasia Incomplete partition type II. Incomplete partition type III.	Type I. Type II. Type III.
II. Vestibular deformities	Michel deformity Common cavity deformity Absent Hypoplastic Dilated	
III. Semicircular canal deformities	Absent Hypoplastic Enlarged	
IV. Inner ear canal deformities	Absent Narrow Widened	
V. Aqueductus cochleae, aqueductus vestibuli	Normal Enlarged	

normal in size, however, lacks any modiolus or interscalar septa. The internal auditory canal is dilated and the bony separation to the cochlea is not always present (The white arrows in Figure 1B and 1C indicate the wide communication between the inner ear canal and the cochlea). The hypoplastic cochlea in Figure 1C is comparably smaller than the normal one shown in Figure 1D.

Figure 2 shows a 3D reconstruction of the T2-weighted inner ear MRI images. A normal cochlea (shown in Figure 2A) is compared with the image of the patient with cochlear hypoplasia on the right and incomplete partition Type I on the left side (Figure 2B). On the right side, the basal turn of the cochlea as well as the semicircular canals can be distinguished, but on the left side only two cystic structures are observed.

Peri-operative care

All surgical operations were carried out under general anesthesia. In all cases, Stenvers view was performed on postoperative Day 1 to demonstrate the proper electrode position. During the post-operative period, amoxicillin, in combination with clavulanic acid, were administered for one week.

Audiological measurements

The initial fitting of the speech processor was carried out during post-operative weeks, 4 to 8. The regular audiological examination and installation of the implant were performed at our department. Free field (1-meter distance from the loudspeaker) pure-tone audiometry, in addition to Speech Reception Test (SRT) and Word Recognition Score (WRS) was performed on every visit using a Danplex DA 45 Audiometer (GN Otometrics, Taastrup, Denmark). The pure-tone audiometry measurements were performed at eight different frequencies, between 125 Hz and 8000 Hz. In consideration of the SRT, the official Hungarian test of Gőtze was used [8]. Twenty 2-digit numbers were presented and the correctly repeated numbers were tallied (5% awarded to each correct answer). In view of the WRS, monosyllabic words were presented, and the correctly repeated answers were later compiled and evaluated.

Results

Surgical results

All surgeries were performed using transmastoid approach. In three patients suffering from common cavity malformation (Patients No. 1, 2, 3), the electrode was inserted following a labyrinthotomy (out of 8; 37,5%). In the remaining five operations, a cochleostomy was used in support of implantation following posterior tympanotomy. In Patient No. 4, 6, 7 and during the first operation of Patient No. 5 (a Type I incomplete partition and a cochlear hypoplasia), complementary access to the inner ear through anterior tympanotomy was required (out of 8, 50%). In three cases, the facial nerve was routed in an abnormal, anteromedial position, and in one case, the promontory did not bulge enough into the tympanic cavity to perform a cochleostomy from the facial recess. Five normal sized and three short electrodes were used.

The intra-operative gusher occurred in two cases afflicted with incomplete partition Type I (Patient No. 4, 6). The insertion site was covered using Tachosil (Baxter Healthcare Corporation, Deerfield, USA) and segments of temporal muscle and fascia. No further intra or postoperative procedure was necessary. No meningitis or newly developed facial palsy was observed. Table 2 summarizes patients' data, electrodes used, complications and follow-up periods.

Audiological results

Pure-tone audiometry: Pure-tone audiometry measurements were successfully performed in all patients. The average audiological follow-up examination was 40.3 months (2 to 84). Figure 3 shows the most current, free field pure-tone audiometric results of the seven patients. Each audiogram is visualized separately in Figure 3A, and the average audiogram including standard deviation is shown in Figure 3B.

The finest thresholds were measured on Patient No. 4, a case with incomplete partition deformity Type I. The average threshold value at speech frequencies (500, 1000, 2000, 3000 Hz) was 21.25 dB, following a 4-year follow-up period. This patient was operated on at the age of 3.5 years. Patient No. 6, suffering from the same deformity, showed the second poorest audiological results, scoring an average of 47.5 dB. This patient underwent implantation at 19 years of age.

Among the three patients suffering from common cavity deformity (Patients No. 1, 2 and 3), Patient No. 3 (operated at 14.7 years of age) showed poorer threshold values than Patient No. 1 and 2, operated at the age of 2.8, and 5.1 years, respectively. Patient No. 5 underwent bilateral implantation in the effective treatment of cochlear hypoplasia on both ears. The pure-tone threshold showed a symmetric hearing of up to 2000 Hz. At high frequencies, 4000 and 8000 Hz, the left ear, which was operated upon later, showed higher threshold values. The average threshold values concerning speech frequencies on the left ear showed similar results with 36.25 dB in the

Table 2: Data of our implanted cochlear malformed patients.

Patient	Sex	Age at surgery	Malformation	side	Implant	Electrode	Sound-processor	Complication	Wearing time (month)
1	female	2.8	Common cavity	Left	Pulsar (MedEl)	Standard	OPUS 1	-	59
2	female	5.1	Common cavity	Left	Sonata (MedEl)	Standard	OPUS 2	-	52
3	male	14.7	Common cavity	Left	Sonata (MedEl)	Flex 24	OPUS 2	-	6
4	female	3.5	Incomplete partition type I.	Left	Pulsar (MedEl)	Standard	OPUS 1	strong gusher	48
5	female	10.5	Cochl. hypoplasia	Right	Combi (MedEl)	Standard	OPUS 2	-	84
		15.9	Cochl. hypoplasia	Left	Concerto (MedEl)	Flex 24	OPUS 2	-	20
6	female	19	Incomplete partition type I.	Left	Nucleus 5 (Cochlear)	CI24RE CA	FREEDOM	slight gusher	52
7	female	6.1	Cochl. hypoplasia	Right	Concerto (MedEl)	Standard Flex 28	RONDO	-	2

right and 40 dB in the left ear.

The poorest values were measured in Patient 7, with an average of 55 dB regarding speech frequencies. The average audiogram (Figure 3B) showed slightly deteriorated values at higher frequencies, although the values were constant regarding speech frequencies (32.5 dB at 1000 Hz, 41.6 dB at 3000 Hz). The poorest average threshold value was measured at 8000 Hz (58.75 dB).

SRT and WRS: Despite successful pure-tone measurements, regarding SRT and WRS tests, the results are inconclusive, in consideration of Patients No. 1, 2, 4 and 7. The finest performance in SRT was measured in Patient No. 5, with 70% in 40 dB (right ear) and 90% in 50 dB (left ear). Patient No. 3 featured the highest result in 60 dB, 50%, and Patient No. 6 performed 20% of the correct reception at 60dB. In view of WRS, Patient No. 3 showed the finest result with 50% of correctly repeating monosyllabic words at 70 dB. Patient No. 5 scored 40% in 50dB (right ear) and 25% in 60 dB (left ear). No correct answer in Patient No. 6 could be detected.

Discussion

Cochlear implantation in a patient suffering from cochlear malformation was first performed in 1983 [9]. Since then, the field of medical diagnostics and technology has witnessed a remarkable development. Cochlear implantation inevitably became the gold standard and a successful method in hearing rehabilitation for patients suffering from inner ear deformities [5].

Due to the abnormal anatomical circumstances and the possibility of enlarged connections between the inner ear structures, inner ear canal and CSF spaces, the most frequent complications involving surgery are gusher and concomitant meningitis. Gusher is defined as a robust CSF leakage, immediately following the opening of the inner ear structures. The likely hood of a gusher during cochlear implantation on malformed cochlea is 40% to 50% [4,10]. In our cases, an intra-operative gusher occurred in two of the eight operations, or 25%. Distinctively, our figure is considerably lower to what is unveiled in the published literature; however, the number of cases we studied is not proportionately comparable with those in the cited articles [4,10].

In our cases, intra-operative management of the gusher was sufficient enough in both cases to prevent postoperative meningitis and no insertion of lumbal drainage was deemed necessary. In the cases of recurrent liquorrhea and meningitis, revision surgery and subtotal petrosectomy are to be considered, for which our team is suitably prepared and regularly performs these procedures in revised cholesteatoma cases. Performing cochleostomy or finding the round window for implantation often proves challenging, due to anomalies of facial nerve anatomy, which, when considering the majority, are likely diagnosed preoperatively. The abnormal, mainly anteromedial position of the nerve is generally associated with cochlear deformity [11], but facial nerve bifurcation was also reported in a patient suffering from hypoplastic cochlea [12]. Nerve monitoring is a useful tool towards avoiding potential damage of the facial nerve. No peripheral facial nerve palsy was observed during our procedures. The tympanic cavity is usually opened through the transmastoideal approach, together with posterior tympanotomy; however, in several cases a combined approach in association to anterior tympanotomy is required for safe, effective cochleostomy [13]. In the case of cochlear hypoplasia, for example, there is the distinct possibility in which no promontory is detectable through posterior tympanotomy, or cochleostomy cannot be performed due to the abnormal position of the facial nerve. In consideration of common cavity deformities, transmastoideal labyrinthotomy can be implemented to reach the inner ear afflicted with a cystic appearance [14].

The success of the cochlear implantation in malformed inner ears is also influenced by the type of electrode used, which should also be selected as part of the preoperative assessment. The availability of varied electrodes differ in size (shortened, full, & elongated), in shape (straight, &curved), and in terms of rigidity. Electrodes with a cork-type stopper possess a distinct role in the depth of the cochleostomy and may prevent the incidence of gusher and subsequent meningitis [15].

The audiological performance of implanted deformed cochlea in pure-tone audiometry is comparable to the results of deaf cochlea with a radiologically normal appearance [16]. Although the limited number of patients does not allow the statistical analysis of the audiological results, tendencies suggest the age at which the surgery was performed has a greater influence on the later audiological performance than the severity of the deformity. A clear difference in the audiological performance is observed in ears with the same deformity, yet a different age at surgery. A common cavity deformity can be rehabilitated resulting in an average threshold value of 22.5 dB, if the operation is performed early enough.

Conclusion

Prior to cochlear implantation, temporal bone HRCT and inner ear MRI imaging are mandatory towards effectively detecting and classifying the inner ear malformation. In consideration of proper preoperative planning, characteristically hinges upon the most appropriate surgical approach and the best fitting electrode. Early implantation of malformed cochlea is crucial towards achieving

ideally superb postoperative audiological results.

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