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Inner Ear Anomalies among Cochlear Implant Candidates in Erbil City, Iraq

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Abstract

Background and objective: To study the frequency of each type of inner ear abnormalities in cochlear implant candidates in Erbil city, Iraq.

Methods: This is a case series study of high resolution computed tomography and magnetic resonance images of temporal bone in 111 candidates for cochlear implantation in Erbil city between June 2015 and December 2017; who had congenital bilateral profound hearing loss.

Results: In total, 111 cases of congenital sensorineural hearing loss were included. Of these, 101 patients had a normal inner ear pattern. Ten patients identified to have inner ear anomalies (9%). unilateral anomaly seen in one child, so the total number of ears with inner ear anomalies was 19. Among these 19 abnormal ears, 7 were classified as incomplete partition type II (36.8%), 4 as incomplete partition type I (21.1%), 4 as a common cavity (21.1%), 2 as cochlear aplasia (10.5%), 1 as cochlear hypoplasia (5.3%) and 1 as Michel deformity (5.3%).

Conclusions: The most common type of anomaly was incomplete partition type II “Mondini” deformity followed by common cavity and incomplete partition type I deformities. The incidence of congenital hearing loss with inner ear anomalies is similar to the results of such studies worldwide.

Keywords: Inner ear anomaly, cochlear implant, Mondini deformity.

Introduction

Cochlear implantation is indicated in cases of severe or profound sensorineural hearing loss (SNHL) in patients who do not obtain sufficient benefit from conventional hearing aids which only amplify sound and who need a partially functioning hearing. The idea of cochlear implantation arose in the 1960s, and in 1969, the first human implantation procedure was recommended by Lo. The cochlear implant is a sophisticated device which is surgically implanted and which electrically stimulates the auditory nerve fibers[1]. Since the initial recommendation, the number of devices which have been implanted has increased gradually and, in the last 20 years, there has been an obvious increase in the number of cochlear implant procedures being performed[2]. It is recommended that both axial and coronal plane thin-section HRCT should be performed for potential cochlear implant candidates. This will provide the diagnosis of

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the congenital malformation, the extent and severity of the deformity and if there are any associated anomalies. Furthermore, it will show the landmarks for mastoidectomy and will indicate whether the facial nerve is abnormally located or absent in patients with cochlear malformations. These findings are very important for a successful surgical procedure and to indicate any difficulties which might occur during surgery, and will also help with the decision on the easier ear for implantation.[3,4] The use of a 1.5- or 3-T MRI system is preferred for inner ear examinations, and it is strongly recommended that this is performed under general anesthesia. A thin-section gradient-echo sequence that is heavily T2-weighted is best suited for evaluation of the fluid-filled spaces of the membranous labyrinth and the eighth cranial nerve. Oblique sagittal reformatted images should be obtained in planes perpendicular to the course of the seventh and eighth nerves in the internal auditory canal and cerebellopontine angle [3,5]. Congenital inner ear malformations occur as a result of the arrest or aberrance of inner ear development due to heredity, gene mutations or other factors. The malformations can exist in any part of the inner ear, 20% of which are bony structure malformations. The remaining 80% are membranous malformations which cannot be detected by radiologic diagnostic tools, are hidden inside the bony structure, and the pathology lies at the cellular level making diagnosis difficult[6]. In 2002, Sennaroğlu and Saatci proposed a classification for cochleovestibular malformations and this has been modified many times by assimilating more information on inner ear anatomy based on newer and better imaging techniques[5,7]. The aim of this study was to determine the inner ear abnormalities and the frequency of each anomaly in cochlear implant candidates in Erbil city, Iraq and comparing it with the percentage of inner ear anomalies worldwide.

Patients and methods:

An observational cohort study in which we analyzed the imaging findings of 111 patients with congenital bilateral profound hearing loss who were candidates for cochlear implantation in multiple centers in Erbil city between June 2015 and December 2017. For all of the patients, radiological evaluation consisted of HRCT and MRI. All HRCT examinations were performed with contiguous 0.4–0.5 mm coronal sections and 0.4–0.5 mm axial sections obtained parallel to the orbitomeatal line. Magnetic resonance imaging examinations were performed on a 1.5-Tesla system with T2-weighted contiguous slices of 2-mm thickness through the temporal bone in axial and coronal planes. Oblique sagittal images were obtained in planes perpendicular to the course of the seventh and eighth cranial nerves in the internal auditory canal. Due to small age of the candidates, most of the imaging was performed under general anesthesia. The imaging study focused on mastoid pneumatization, the size of the internal auditory meatus, the presence of the cochlear nerve, the status of the bony labyrinth, and cochlear duct patency. Inner ear anomalies were classified according to the most recent classification by Sennaroğlu as shown in Table (1). Cases with inner ear anomalies which were not included in this classification were excluded from the study. Kurdistan Board of Medical Specialties granted ethical approval for this study. And Informed consent was obtained from the parents of the children included in the study.

Table 1: Sennaroğlu's classification for cochleovestibular malformations

Type of inner ear malformation	**Radiology**
Complete labyrinthine aplasia	Absent labyrinth
Rudimentary otocyst	Incomplete millimetric otic capsule remnant
Cochlear aplasia	Absent cochlea
Common cavity	Round or ovoid cystic structure for cochlea and vestibule
Cochlear hypoplasia	Cochlear size small; four types
Incomplete partition I	Cystic cochlea
Incomplete partition II	Cystic cochlear apex
Incomplete partition III	Modiolus absent; interscalar septa present
Enlarged vestibular aqueduct	Normal cochlea with enlarged vestibular aqueduct
Cochlear aperture abnormalities	Narrow or absent cochlear aperture

Results

In total, 111 patients with congenital hearing loss were included in this study, 59 (53%) were female and 52 (47%) were male. From the 111 cases of congenital SNHL, 101 (91%) had a normal inner ear pattern and 10 had inner ear anomalies on HRCT and MRI. The percentage of abnormal inner ears which were detected in our imaging study and which could be classified using Senneroğlu's classification was 9%. From these 10 cases, 6 (60%) were female and 4 (40%) were male, and only one had a normal inner ear on one side; in all the other cases, the deformities were bilateral. So the total number of ears with inner ear anomalies was 19. From Senneroğlu's classification, of the 19 abnormal ears, seven ears (36.8%) had an incomplete partition type II deformity. Figure (1).

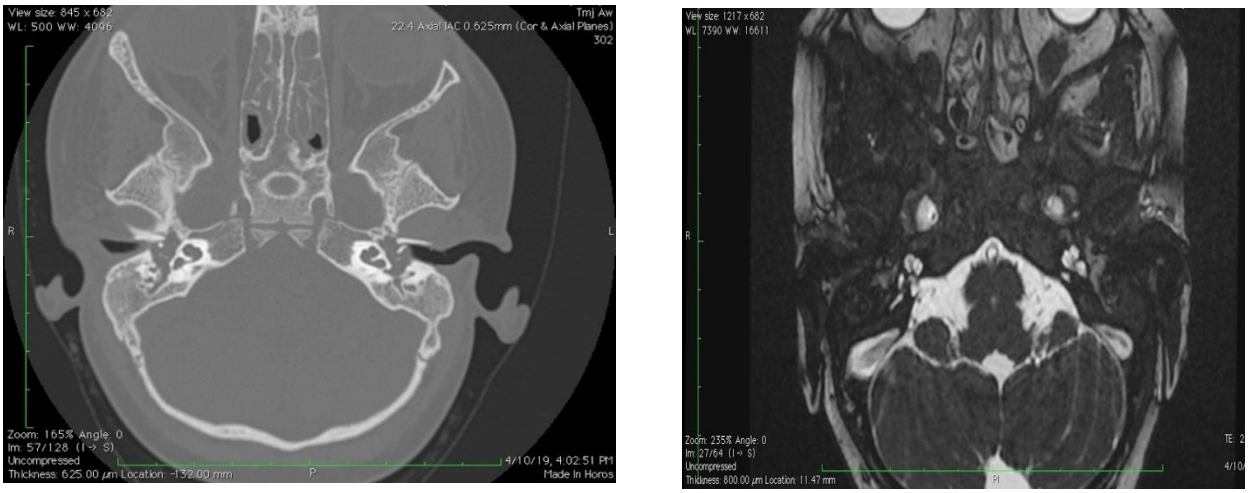

Figure 1: Left panel, CT scan of temporal bone; axial section. Right panel, MRI axial section 3D FIESTA. Both panels show bilateral incomplete partition type II

In three cases, this deformity was bilateral. Four ears (21.1%) had an incomplete partition type 1 deformity and this deformity was bilateral in one case. Figure (2).

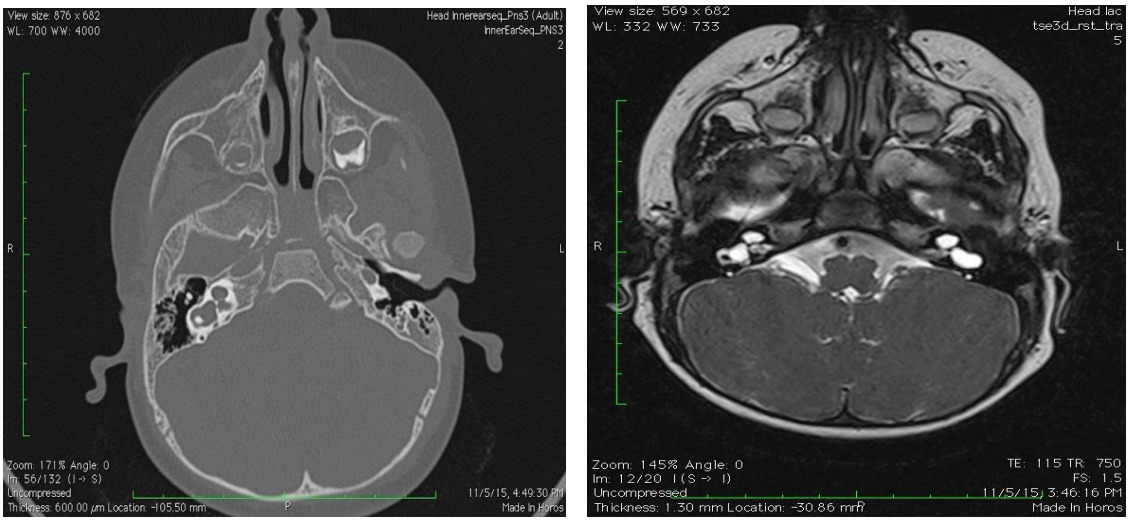

Figure 2: Left panel, CT scan of temporal bone, axial section, showing right-sided incomplete partition type 1 Right panel, MRI axial section T2-weighted image showing bilateral incomplete partition type 1

Four ears (21.1%) had a common cavity deformity Figs (3) and (4) and this deformity was bilateral in one case.

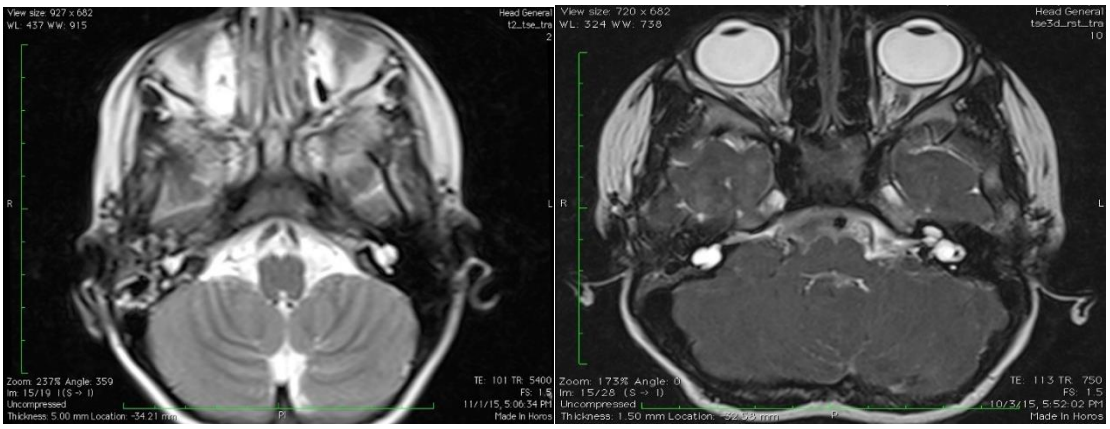

Figure 3: MRI axial section T2-weighted image showing bilateral common cavity anomaly

Figure 4: MRI axial section 3D FIESTA showing right-sided common cavity deformity and left-sided incomplete partition type 1

Two cases (10.5%) had cochlear aplasia Figure (5), one case (5.3%) had cochlear hypoplasia, and one (5.3%) had a Michel deformity Figure (6).

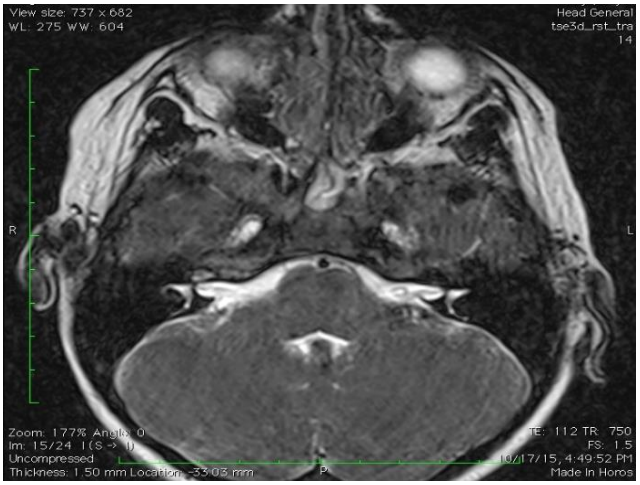

Figure 5: MRI axial section 3D FIESTA showing left-sided cochlear aplasia

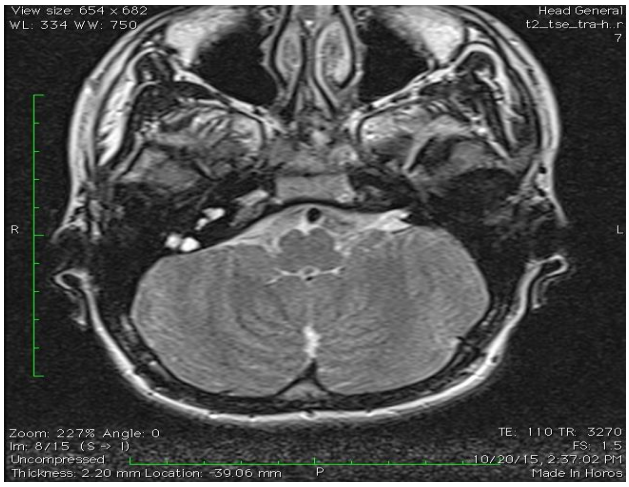

Figure 6: MRI axial section 3D FIESTA showing left-sided Michel anomaly

All Mondini cases (incomplete partition type II) were accompanied with a dilated vestibular aqueduct. Figure (7) shows the number of inner ear anomalies and their percentages

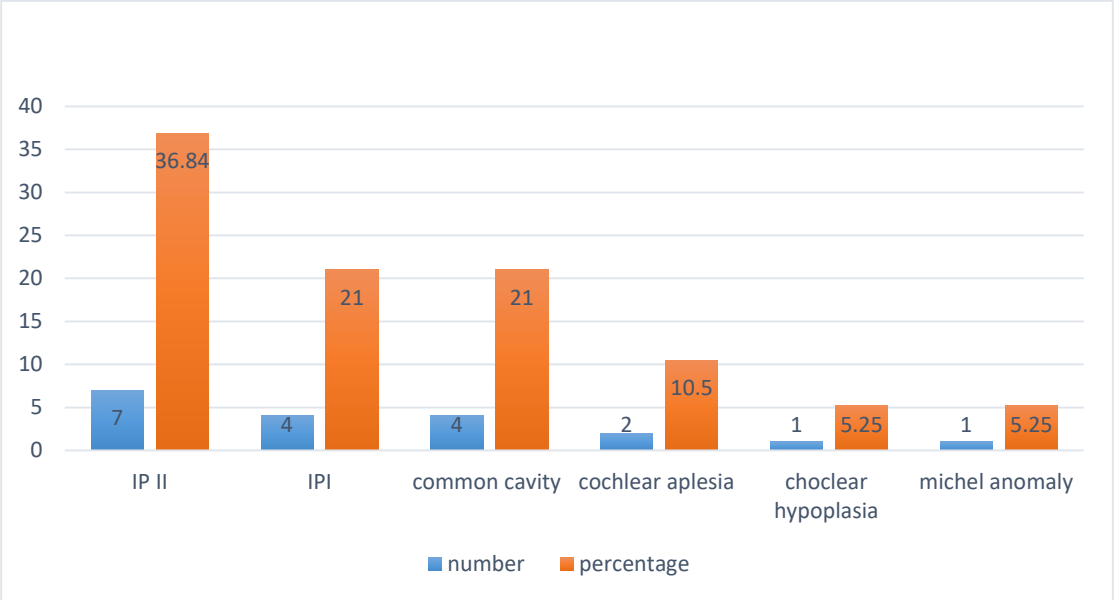

Figure 7: Frequency distribution of the different types of inner ear anomaly

Discussion

Its mandatory to assess children with congenital SNHL preoperatively by CT scan and MRI for detailed examination of the temporal bone contours, the cochlear nerve, and the auditory pathways. From this, the suitability and choice of which ear to receive an implant can be established. Some findings affect the electrode array type which is used for the patient, and occasionally, may indicate that the surgical procedure should be modified in consonance with those findings. In our practice; it's obvious that the congenital hearing loss is not an uncommon phenomenon in our community and as a fact it has a serious impact on both the child and family. Detection of congenital anomalies of the inner ear preoperatively is of great importance and remarkably assists the surgeon. Till now no available data regarding the frequency and types of inner ear malformations in our city. The high consanguinity plays a great role in inner ear malformations existence; other risk factors include improper perinatal care and poor education.

In this study, the percentage of inner ear anomalies was 9%, which is similar to values reported in other studies in other countries. Aldhafeeri reported that the percentage of inner ear anomalies was 7.5%[8], while Buchman et al. showed that the incidence of inner ear anomalies among cochlear implant candidates in North Carolina was 8.8%[8]. In 2010, in a literature review, Sennaroğlu, classified the inner ear malformations and their frequencies as follows: Michel deformity 6%; cochlear aplasia 5%; common cavity malformation 8%; cochlear hypoplasia 12%; incomplete partitions 41% (IP-I or what is called cystic cochleovestibular malformation forming 20%, IP-II [Mondini deformity] 19%, and IP-III [X-linked deafness] 2%); and large vestibular aqueduct (LVA) 15%. The same literature review indicated that the commonest malformations and their frequencies varied between studies in different areas[10]. In our study among the inner ear anomalies detected, the Mondini deformity was the most common at 36.8%. This result is in concordance with the study by Jackler et al. in which they mentioned that the Mondini deformity was the most common type of inner ear malformation accounting for over 55%[11]. While its somewhat similar to the result of Daneshi et al in which they found the Mondini deformity is 29%[12]. In this series study, two cases of cochlear aplasia (10.5%) were identified. While senneroglu in his study reported the percentage of cochlear aplasia as 5%[10]. We detected one Michel anomaly (5.3%). This is a severe malformation of the ear defined by the total absence of inner ear structures, which was first demonstrated by Michel in 1863 and has only been reported in selected case reports in the literature[13]. Our finding for the Michel deformity is similar to Senneroğlu's percentage. In most of the cases with the Mondini deformity, we detected a large vestibular aqueduct. Although an isolated dilated vestibular aqueduct has been observed and reported by Jackler[14], Lemmerling *et al*[15]. in their series, found that all ears with large vestibular aqueducts had an associated modiolar deficiency. Hence the term isolated dilated vestibular aqueduct seems obsolete. IP-II (classic Mondini) is always associated with a dilated vestibular aqueduct.[15] In his older classifications. Senneroğlu did not mention an enlarged vestibular aqueduct as an isolated malformation, but in his updated classification, he showed that the percentage was about 15%; however, in our study, no isolated vestibular aqueduct has been found but this anomaly mostly accompanies other inner ear anomalies, especially incomplete partitions.

Conclusion

We found that Mondini (incomplete partition type II) is the most common type of inner ear malformations, followed by common cavity and incomplete partition type I deformities. Generally, CT and MRI are recommended for diagnosis of such malformations

Conflict of interests

There were no conflicts of interest.

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