RESEARCH ARTICLE

Frequency of congenital inner ear malformations among cochlear implant candidates at a tertiary care Hospital, Pakistan

Ali Hussain[1], Sohail Awan[2], Wasim Memon[3]

Abstract

Objective: To determine the frequency and factors associated with congenital inner ear anomalies among cochlear implant candidates.

Method: The cross-sectional study was conducted at the Department of Otolaryngology, Aga Khan University Hospital, Karachi, from October 2021 to March 2022, and comprised patients of either gender aged <18 years who received cochlear implants. All subjects underwent computed tomography and magnetic resonance imaging of the temporal bone to identify any inner ear anomaly. The data gathered included implantation age, gender, computed tomography and magnetic resonance imaging findings, and history of congenital anomaly, neurodevelopmental disorder, family hearing loss, consanguinity and brain-evoked response auditory result. Data was analysed using SPSS 23.

Results: Of the 120 patients, 66(55%) were females and 54(45%) were males. The median age of the patients was 3 years (interquartile range: 2-5 years). Of the total, 7(5.8%) patients presented with inner ear malformations; 3(2.5%) each with enlarged vestibular aqueduct, Mondini malformation type II, and 1(0.8%) with semi-circular canal dysplasia.

Conclusion: The frequency of inner ear anomalies amongst cochlear implant candidates was 5.8%, and the most common anomalies were enlarged vestibular aqueduct and Mondini malformation type II.

Keywords: Cochlear implant, Inner ear anomalies, Inner ear malformations, Hearing loss, Frequency, Consanguinity, loss, Temporal bone, Tomography, Magnetic resonance, Neurodevelopmental disorders, Otolaryngology.
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Introduction

Hearing loss management by means of cochlear implants in patients with inner ear anomalies has been a topic of discussion for a long time.[1] Hearing loss in children is a recurrent clinical phenomenon in Pakistan, which takes a toll on patients and their families. The benefits that can be provided by cochlear implants can vary from substantial word understanding to simple sound detection. Cochlear implantation (CI) has proven to improve and develop the percentage of language acquisition, aiding in the development of advance literacy and spoken language in deaf children.[2] Pre-lingual or post-lingual categories range from severe to profound hearing loss, according to its establishment before or after language acquisition.[3,4] Cochlear implants give the most promising results as a prosthesis of modern times.[5] Approximately 20% of all congenital hearing loss is related with inner ear malformations (IEMs).[6] Imaging scans, like magnetic resonance imaging (MRI) and high-resolution computed tomography (HRCT), are useful in identifying inner ear malformations while also facilitating the correct judgement whether to or not perform the implantation procedure. These abnormalities may affect a patient's prognosis, and, therefore, may cause the risk of complications as well as auditory improvement.[7]

The current study was planned to determine the frequency and factors associated with congenital inner ear anomalies among cochlear implant candidates.

Patients and Methods

The cross-sectional study was conducted at the Department of Otolaryngology, Aga Khan University Hospital (AKUH), Karachi, from October 2021 to March 2022. After approval from the institutional ethics review committee, the sample size was calculated using Open Epi version 3.01 with level of significance 5%, precision 5 and anticipated frequency 7.5%.[1] The sample was raised using non-probability consecutive sampling technique. Those included were patients of either gender aged <18 years who received cochlear implants. Patients who had a history of ear surgery or head trauma were excluded.

All the subjects underwent CT and MRI scans of the temporal bone to identify any inner ear anomalies, which was confirmed by a senior radiologist using the Cincinnati criteria to assess enlarged vestibular aqueduct (EVA), and the cochleovestibular classification system.[1] The collected data included implantation age, gender, CT and MRI

[1,2]Department of Otolaryngology, Aga Khan University Hospital, Karachi, Pakistan; [3]Department of Radiology, Aga Khan University Hospital, Karachi, Pakistan.

Correspondence: Ali Hussain. e-mail: alidrhussain@gmail.com
ORCID ID. 0009-0007-2717-6558

findings, history of congenital anomaly, neuro-developmental disorder, and family history of hearing loss, consanguinity and audiology data/ brain-evoked response auditory (BERA) results. Data was analysed using SPSS 23.

Results

Of the 120 patients, 66 (55%) were females and 54 (45%) were males. The median age of the patients was 3 years (interquartile range [IQR]: 2-5 years) (Table 1). There was no history of neurodevelopment disorder in 111 (92.5%) cases and no history of congenital anomalies in 114(95%) cases (Table 2).

Table-1: Age and gender distribution.

	n (%)	*p*-value
Age (years) Median (IQR)	3 (2-5)	
Gender		
Male	54 (45)	0.313
Female	66 (55)	0.241

IQR: Interquartile range.

Table-2 Patient history.

	n (%)	*p*-value
History of Neurodevelopment Disorder		
No	111 (92.5)	<0.999
Yes/Autistic	1 (0.8)	
Delayed Developmental Milestone	7 (5.8)	
Down Syndrome	1 (0.8)	
Total	120 (100.0)	
History of Congenital Anomalies		
No	114 (95.0)	<0.999
G6PD	3 (2.5)	
Pierre Robin Sequele	1 (0.8)	
Patent ductus arteriosus	1 (0.8)	
Down syndrome	1 (0.8)	
Total	120 (100.0)	0.602
BERA		
No Response	80 (66.7)	
Profound	34 (28.3)	
Severe	6 (5.0)	
Total	120 (100.0)	
Family History of Hearing Loss		
Yes	3 (42.9)	0.365
No	4 (57.1)	
Total	7 (100.0)	
History of Consanguinity		
Yes	4 (57.1)	0.699
No	3 (42.9)	
Total	7 (100.0)	

BERA: Brain-evoked response auditory

Table-3: Inner ear anomalies (n=7).

Enlarged Vestibular Aqueduct	Mondini Malformation Type II	Semi Circular Canal Dysplasia
3	3	1

Of the total, 7(5.8%) patients presented with inner ear malformations (Table 3); 3(42.85%) each with EVA

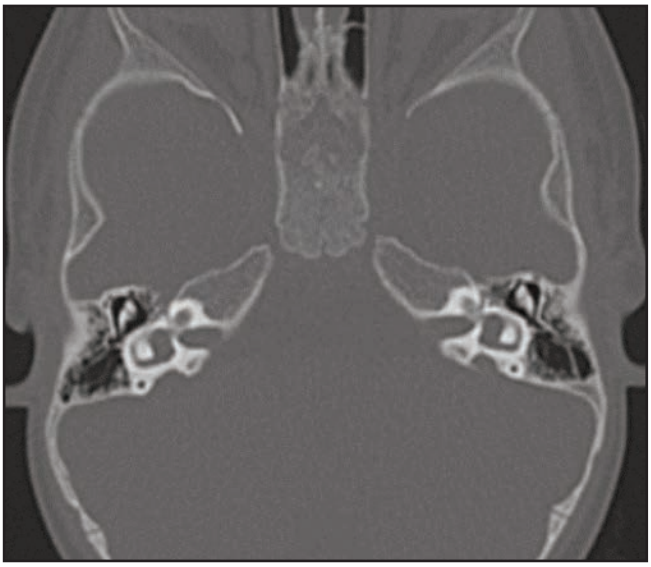

Figure-1: Bilaterally enlarged vestibular aqueduct (EVA).

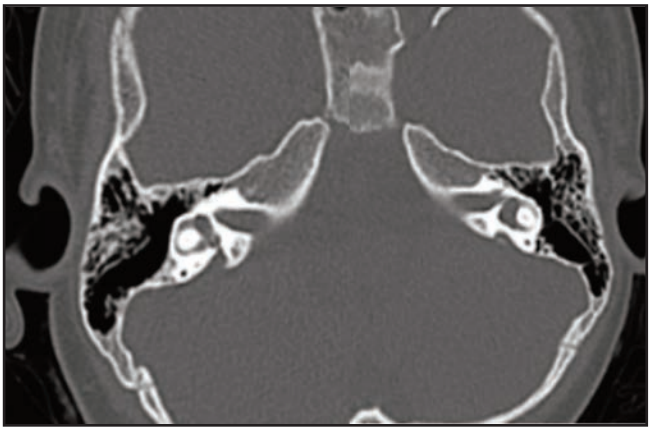

Figure-2: Mondini malformation type II.

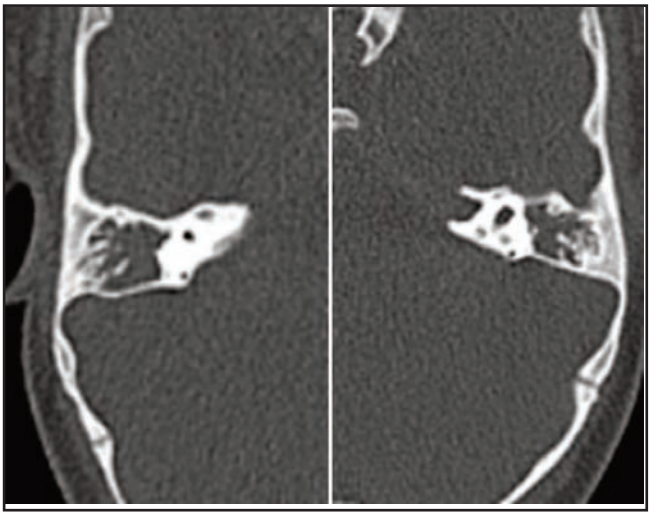

Figure-3: Semi-circular canal dysplasia.

(Figure 1), Mondini malformation type II (Figure 2), and 1(14.3%) with semi-circular canal dysplasia (Figure 3). Among the 7 cases, family history for hearing loss was positive in 3(42.9%), and history of consanguinity in 4(57.1%). Also, 5(71.4%) of these cases were males and 2(28.6%) were females.

Discussion

An earlier study showed that congenital inner ear malformations were based on their radiographic appearance.[8] In the current study, CT/MRI was done on all 120 patients, and 7 of them were found to have congenital anomalies, with Mondini type II and EVA being the most common. Mondini dysplasia is believed to represent the arrest of embryonic development at about 7 weeks of gestation.[9]

It can be challenging for an Ear-Nose-Throat (ENT) consultant to confirm the diagnosis for inner ear malformations at a glance. A radiologist works hand in hand in such a case with HRCT and MRI examinations to determine the final outcome. CT enables accurate surgical planning to help avoid anatomical variants that may influence the surgery by visualising the bony structures of the inner, middle and external ear. On the other hand, MRI is imperative for the assessment of the cochlear nerve.[10]

A study reported the frequency of congenital hearing loss in Pakistan to be 15% profound and 85% moderate to severe, which is rather high compared to findings elsewhere.[11] The current study found the frequency of inner ear malformations in cochlear implant candidates to be 5.8%. Pakistan's first cochlear implant in a case with inner ear anomalies was reported in 2013.[12]

Majority of the patients have bilateral severe to profound hearing loss and are contenders for a cochlear implant.[13] More than half the patients in the current study presented with severe hearing loss.

In the current study, there was no correlation with middle and external ear deformities for patients that had inner ear anomalies. On the other hand, consanguineous marriages showed an association with those having hearing loss and anomalies, while family history did not show any correlation. Neurodevelopmental disorders or congenital anomalies did not show much of an association in such patients. No significant association or difference in relation to these variables was found with either those with inner ear anomalies or those without any anomaly.

A study showed no significant link between the type of anomaly and gender.[5] In the present study, among those with inner ear anomalies 71% were males.

The most common complication for those with inner ear malformations having cochlear implants is cerebrospinal fluid (CSF) leak which can be gushing or oozing, depending on the intensity of the flow. A CSF leak usually can arise if there is dehiscence between the lateral fundus of the internal auditory canal and the cochlea.[14] In the current study, two patients with inner ear anomalies had perilymph gusher.

Conclusion

The frequency of congenital inner ear anomalies was 5.8%, with the most frequent anomalies being EVA and Mondini malformation type II. In the present study, we observed no statistically significant correlation among the variables under consideration. This absence of correlation can be construed as a favorable outcome, as it indicates that the factors did not have any adverse effects on patients with SNHL who presented with inner ear anomalies and subsequently underwent cochlear implantation procedures.

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