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Exploring Cochlear and Vestibular Abnormalities in Children with Hearing Impairment; a Tertiary Care Experience

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ABSTRACT

Inner ear malformations have a great impact on hearing. Diagnosing them accurately through advanced imaging modalities can help plan management strategies. **Objectives:** To determine anatomical defects of inner ear malformations in children requiring hearing aids and cochlear implants coming to the tertiary care hospital. **Methods:** A cross-sectional research design was used with a sample of 104 hearing-impaired children, aged 1 to 14 years, who had moderate to profound hearing loss, collected through a purposive sampling technique. Demographic information was collected through a specially designed questionnaire, which included age, gender, developmental delay, etc. were also assessed. **Results:** 7 (6.7%; 95% CI (0.019, 0.115)) cases had anatomical defects of the inner ear, including the temporal bone. Among 7 cases, all had bilateral cochlear hypoplasia CH-III, semicircular canal malformations, 3 (42.9%) bilateral hypoplasia, 3 (42.9%) bilateral dysplasia, 1 (14.4%) had bilateral aplasia of anterior and lateral semicircular canals, 3 (42.9%) bilateral dilated vestibule, and 1 (14.3%) narrowing of the internal auditory canal left side. In these 7 cases, 5 (71.4%) with profound degree hearing loss were candidates for cochlear implant and 2 (28.6%) with severe degree hearing loss were using hearing aids. Results of chi-square showed that age at the time of diagnosis ($p<0.05$), family history of hearing impairment ($p<0.001$), and severity of hearing loss ($p<0.001$) had a significant association with the type of hearing impairment. **Conclusions:** The frequency of inner ear malformations in hearing-impaired children using hearing aids or candidates for cochlear implant was 6.7%. The most common malformations were CH-III and semicircular canal malformations.

INTRODUCTION

Hearing impairment is the fourth major cause of disability occurring globally [1]. More than 34 million children are suffering from deafness or hearing loss worldwide. Developing and underprivileged countries harbor a great proportion of children affected by hearing loss [2]. In Pakistan, one study reported 1.97% prevalence of hearing impairment in school-going children [3]. The most common type of permanent hearing loss is sensorineural hearing loss. It happens after damage to the inner ear, vestibulocochlear nerve, or brain. It is estimated that every 1 in 1,000 children is born with either severe or profound deafness, and congenital sensorineural hearing loss is present in 90 percent of cases [4]. Half of the cases of congenital sensorineural hearing loss are related to a genetic cause [5]. Syndromic cases are approximated to 30%, and the remaining 70% attributed to non-syndromic cases, often presenting with inner ear malformations [6]. Acquired causes of sensorineural hearing loss include birth asphyxia, teratogens, inner ear infections, autoimmune disorders, trauma, or posterior fossa tumors, which usually

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present later in childhood. Hearing-impaired children with inner ear malformations (IEMs) have a high prevalence of 20% worldwide [7, 8]. In Pakistan, overall congenital malformations were reported to be 7%. Ear, eye, face, neck malformation constitutes (20%) [9, 10]. Inner ear malformations constitute a major contributor to sensorineural hearing loss. Inner ear malformations can be classified into eight categories as proposed by Sennaroglu [8, 11]. In these anomalies, 80% are membranous malformations causing pathology of inner ear hair cells. The other 20% include various bony labyrinth malformations diagnosed on imaging studies. In a study conducted internationally,13% of children had inner ear malformation among cochlear implant candidates [12]. Early diagnosis plays a key role in the rehabilitation of hearing-impaired children with sensorineural hearing loss. Audiological investigation, along with imaging techniques such as High-resolution computed tomography (CT) scan and Magnetic resonance imaging (MRI) studies provide valuable details regarding the temporal bone and membranous structures like 8th cranial nerve (CN), inner ear (IE) structures, middle ear (ME), and outer ear (OE). The findings of these studies will help otologists to plan surgery, guide regarding prognosis, and assess the risk of complications. Although most cases may be managed by hearing aids, cochlear implantation and auditory brainstem implantation (ABI) are also options, depending on the degree of hearing loss. Mild to moderate degree hearing loss can be managed by hearing aids, while for children with severe to profound degree hearing loss, a cochlear implant is the best option. Surgical risks and benefits must be weighed before considering this choice. Parents and families should be counselled, and their preferences will guide the best course regarding rehabilitative services.

Despite the global burden of hearing impairment and the significant role of inner ear malformations in sensorineural hearing loss, there is a paucity of local data in Pakistan regarding their prevalence and impact. This study is of immense importance as studies lack local data, which can guide us in planning rehabilitative strategy for such children for better outcomes. Statistical data will help in future research. This study aims to find relation of inner ear malformations and the degree of hearing loss in children rehabilitated by hearing aids or cochlear implants.

METHODS

This cross-sectional study was conducted at the Department of Developmental and Behavioral Pediatrics, The Children's Hospital, and the University of Child Health Sciences, from April 2025 to June 2025. A sample of 104 hearing-impaired children was recruited through convenience sampling. Sample size was collected through G power analysis using a medium effect size and alpha 0.05. After taking approval from the Institutional Review Board (reference no. 1089/CH-UCHS), data were collected. Both unilateral and bilateral cases of hearing impairment were initially screened. Hearing impairment was defined as hearing loss greater than 25 dB in either ear. Severity of hearing loss was categorized as moderate (56–70 dB), severe (71–90 dB), and profound (>90 dB) [2]. The degree of hearing loss was assessed using the most recent audiological assessment conducted in the Department OF Audiology, where diagnostic procedures such as Otoacoustic Emissions (OAE), Auditory Brainstem Response (ABR), or pure tone audiometry had already been performed. Audiological findings were interpreted by 2 audiologists before confirmation by the consultant audiologist. Only non-syndromic cases of congenital sensorineural hearing loss were included in the study, while children with syndromic features were excluded. Hearing-impaired children who had any history of otitis media, ototoxic drug exposure, or neonatal jaundice were excluded. Afterwards, those participants who were enrolled in the study were sent to the Department of Radiology, where High-resolution computed tomography (HRCT) of the temporal bone and magnetic resonance imaging (MRI) of the inner ear were performed for every participant. Imaging findings were interpreted by 2 radiologists and finalized by a consultant radiologist to ensure consistency. Written informed consent was obtained from parents or guardians, and confidentiality was ensured. Demographic data, including age, gender, urban/rural residence, and gestational age, were recorded. Other clinical factors such as history of ear discharge, birth asphyxia, family history of using hearing aids or cochlear implant and developmental delay were assessed. The ShaMaq developmental screening tool (SDST) was used as a tool for screening the development of a child. It screens children aged 6 months to 5.5 years. Each group has four domains: Gross motor and locomotion, vision and manipulation, hearing and speech, and socialization. Any child having a total of four unsatisfactory scores in all domains or one in each domain was considered as having developmental delay [13]. Children were assessed by a clinical psychologist for developmental delay through SDST or informal IQ testing. Data were analyzed using statistical package for the Social Sciences software (SPSS 25), where demographics were analyzed using frequency and percentages. A chi-square test of independence was performed to examine the association between the type of hearing impairment (hearing aid vs cochlear implant) and sociodemographic characteristics. The p-value<0.05 was considered statistically significant.

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RESULTS

Out of 104 hearing-impaired children with bilateral sensorineural hearing loss, 55 (52.9%) were male, and 49 (47.1%) were female. The mean age of the participants was 68.84 ± 45.39 months. Most of the cases (69.2%) belonged to an urban area. The majority of cases (55.8%) had their audiological testing done between age 1 and 3 years, and predominantly (59.6%), Auditory Brainstem Response was done. The majority of participants (62.5%) had profound degree hearing loss (Table 1).

Table 1: Demographic and Clinical Characteristics of Participants (n=104)

Medical Factors	n (%)
Gender	
Male	55 (52.9%)
Female	49 (47.1%)
Place of Residence	
Urban	72 (69.2%)
Rural	32 (30.8%)
Natal or Post-Natal Complication	
Yes	3 (2.9%)
No	101 (97.1%)
History of Ear Discharge	
Yes	10
No	94
Age of Audiological Testing	
<1 Year	1 (14.3%)
1 to 4 Years	6 (86.7%)
Specific Audiological Test	
Oto Acoustic Emission Test	1 (14.3%)
Auditory Brain Stem Response	6 (85.7%)
Severity of Hearing Loss	
Severe	2 (28.6%)
Profound	5 (71.4%)

Out of the total 104 participants, 7 (6.7%; 95% CI (0.019, 0.115)) cases had structural abnormalities of the inner ear, including the temporal bone. In these 7 cases, 5 (71.4%) participants with profound degree hearing loss were candidates for cochlear implant and 2 (28.6%) participants with severe degree hearing loss were using hearing aids. Among the 7 cases, all had bilateral cochlear hypoplasia with less than 2 turns (CH-III). Semicircular canal malformations were reported in all 7 cases (3 (42.9%) participants had bilateral hypoplasia, 3 (42.9%) had bilateral dysplasia, and 1 (14.4%) had bilateral aplasia of anterior and lateral semicircular canals). Among associated structural defects, 3 (42.9%) participants had bilateral dilated vestibule,1 (14.3%) had bilateral non-visualization of vestibulocochlear complex, and 1 (14.3%) had narrowing of internal auditory canal on the left side. All of the 7 cases had no history of prenatal, natal, or postnatal complications. Among the 7 cases, none of the participants had the newborn audiological screening. 6 (85.7%) cases were diagnosed at an age between 1 and 4 years after audiological testing. Only 1 (14.3%) participant had a developmental delay, and none had a family history of hearing impairment (Table 2).

Table 2: Inner Ear Anomalies (n=7)

Medical Factors	n (%)
Gender	
Male	2 (28.6%)
Female	5 (71.4%)
Age of Audiological Testing	
<1 Year	1 (14.3%)
1 to 4 Years	6 (86.7%)
Specific Audiological Test	
Otoacoustic Emission Test	1 (14.3%)
Auditory Brain Stem Response	6 (85.7%)
Severity of Hearing Loss	
Severe	2 (28.6%)
Profound	5 (71.4%)
Non-visualization of the Left Vestibulocochlear Complex	
Yes	1 (14.3%)
No	6 (85.7%)
Bilateral Dilated Vestibule	
Yes	3 (42.9%)
No	4 (57.1%)
Bilateral Semicircular Canal Malformations	
Hypoplastic	3 (42.9%)
Dysplastic	3 (42.9%)
Aplasia of Anterior and Lateral Semicircular Canal	1 (14.3%)
Narrowing of the Internal Auditory Canal on the Left Side	
Yes	1 (14.3%)
No	6 (85.7%)
History of Developmental Delay	
Yes	1 (14.3%)
No	6 (85.7%)

Results of chi square test of independence showed that the association between age at the time of diagnosis and type of hearing impairment was significant, $\chi^2(3) = 8.74$, ($p<0.05$), but no significant association was found between gender and hearing type. There was a significant association between developmental delay and hearing type, $\chi^2(1) = 6.37$, ($p<0.01$). Family history of hearing impairment $\chi^2(1) = 20.15$, ($p<0.001$), history of ear discharge χ^2 (1, n=104) = 11.06, ($p<0.001$), specific audiological test χ^2 (2, n=104) = 70.45, ($p<0.001$) and severity of hearing loss χ^2 (2, n=104) = 62.4, ($p<0.001$) are associated with type of hearing impairment (Table 3).

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Table 3: Descriptive Statistics and Chi-Square Results for Types of Hearing Impairment in Relationship with Sociodemographic Characteristics(n=104)

Variables	Cochlear Implant Planned (n=52)	Hearing Impaired Using Hearing Aids (n=52)	χ^2	df	p-value	Cramer's V
	F (%)	F (%)				
Gender						
Male	29 (55.8%)	26 (50.0%)	0.35	1	0.560	0.04
Female	23 (44.2%)	26 (50.0%)				
Age at the Time of Diagnosis						
<1 Year	3 (5.8%)	2 (3.8%)	8.74	3	0.030	0.13
1 to 4 Years	49 (94.2%)	42 (80.8%)				
5 to 9 Years	0 (0%)	6 (11.5%)				
10 to 14 Years	0 (0%)	2 (3.8%)				
History of Developmental Delay						
Yes	6 (11.5%)	0 (0%)	6.37	1	0.010	0.14
No	46 (88.5%)	52 (100%)				
Family History of Hearing Impairment						
Yes	2 (3.8%)	21 (40.4%)	20.15	1	<0.001	0.25
No	50 (96.2%)	31 (59.6%)				
History of Ear Discharge						
Yes	0 (0%)	10 (19.2%)	11.06	1	<0.001	0.18
No	52 (100%)	42 (80.8%)				
Specific Audiological Test						
Pure Tone Audiometry	0 (0%)	37 (71.2%)	70.45	2	<0.001	0.37
OAE	0 (0%)	5 (9.6%)				
ABR	52 (100%)	10 (19.2%)				
Severity of Hearing Loss						
Moderate	0 (0%)	10 (19.2%)	62.4	2	<0.001	0.35
Severe	0 (0%)	29 (55.8%)				
Profound	52 (100%)	13 (25%)				

DISCUSSION

The present study aimed to find out various inner ear malformations in hearing-impaired cases occurring frequently in this region. In one study in Pakistan, Ahmad *et al.* reported 10% prevalence of inner ear malformation [14]. Another study reported that cochleovestibular anomalies had a prevalence of 11.1% [15]. In another study, it was reported that 5.8% patients presented with inner ear malformations [10]. Our study findings revealed the prevalence of cochleovestibular anomalies to be 6.7%. CT scan of the temporal bone and MRI are fundamental to identify inner ear malformations [16]. Ahmad *et al.* showed that complete labyrinthine aplasia (CLA) and cochlear hypoplasia (CH-III, CH-I) were the most prevalent inner ear malformations [14]. In another study, it was reported that anomalies of the cochlea (45/100) and semicircular canal (20/100) were most frequently occurring [16]. Incomplete partition followed by cochlear hypoplasia was most common in another study carried out by Young [17]. In the current study, the most prevalent IEMs were cochlear hypoplasia (CH-III) and semicircular canal malformation. The majority of cases with IEM had a profound degree of hearing loss and were candidates for cochlear implants [18]. However, being a public sector hospital, Cochlear implant is not performed in our center due to the greater financial burden and less favorable outcomes as compared to cases with normal inner ear anatomy [19]. Hearing-impaired children with cochleovestibular anomalies can present with various associated disabilities [20]. In one study, 13 out of 31 hearing-impaired individuals with anomalies of the inner ear had global developmental delay [21]. In the present study, it was found that 1 deaf child had bilateral cochlear vestibular anomalies and concurrent developmental delay. There was no significant correlation between gender and type of anomaly in the study by Asha [22]. The results of our study are consistent with this pattern. However, inner ear malformations were predominant in the female population (71%) in the current study. Many inner ear malformations can have familial occurrence and a genetic basis of inheritance. Brotto *et al.* described familial occurrence and genetic inheritance of various anomalies [7]. This contrasts with findings in our study, as no familial occurrence was reported in our study. Diagnosis of genetic mutation causing inner ear malformation helps in predicting recurrence risk and the

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need for further investigations to diagnose associated complications [7]. Unfortunately, in resource limited country, cost and poor access to genetic testing create a constraint to ascertain this benefit. Newborn screening plays a pivotal role in early diagnosis of hearing impairment, leading to early intervention and favorable developmental and language outcomes [23]. Being one of the underdeveloped countries, none of the children underwent a newborn screening test in our study. Inadequate early screening practices in the community lead to delayed diagnosis of hearing impairment. Thus, timely evaluation for cochlear implantation cannot be carried out. Subsequently, many patients with inner ear malformations remain undiagnosed. It can make it difficult to estimate the true prevalence of cochleovestibular anomalies and to decide about appropriate management course for such cases that can lead to better outcomes. More public awareness regarding the significance of this challenge is needed at the hour.

This was a single-center study with a limited sample size, which may restrict the generalizability of the findings. Additionally, the absence of newborn hearing screening and genetic testing limited accurate estimation of true prevalence and evaluation of hereditary associations. A multicenter trial should be conducted to ascertain the prevalence of various other types of anomalies. This will enable us to relate the result to a broader population group and enhance credibility. Given limited access to cochlear implantation, super-sonic hearing aids were recommended as an alternative rehabilitation strategy for children with profound hearing loss and inner ear malformations.

CONCLUSIONS

The frequency of inner ear malformations in hearing-impaired children using hearing aids or candidates for cochlear implants was 6.7%. The most common malformations were CH-III and semicircular canal malformations.

Authors' Contribution

Conceptualization: MAJ
Methodology: MAJ, AF, AA, AAQ, IS, WA
Formal analysis: MAJ, AA
Writing and Drafting: MAJ, AF, AAQ
Review and Editing: MAJ, AF, AA, AAQ, IS, WA

All authors approved the final manuscript and take responsibility for the integrity of the work.

Conflicts of Interest

All the authors declare no conflict of interest.

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