RESEARCH ARTICLE

Demographic analysis of hearing impairment based on various parameters in patients with cochlear implant

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Abstract

Objective: To analyse the demographic and clinical variables in children having undergone cochlear implant surgery because of deafness.

Method: The cross-sectional study was conducted from January to November 2022 at the Centre for Research in Experimental and Applied Medicine laboratory of the Department of Biochemistry and Molecular Biology, Army Medical College, Rawalpindi, Pakistan, in collaboration with the Ear, Nose and Throat Department of Combined Military Hospital, Rawalpindi, and comprised children of eith gender aged up to 10 years who had received cochlear implant. Data was collected through questionnaire-based detailed interviews. Syndromic Hearing Loss, Non-Syndromic Hearing Loss, and Acquired Hearing Loss were identified among the subjects. Data was analysed using SPSS 22.

Results: Of the 250 cases, 147(58.8%) were boys, 146(58.4%) were aged 0-5 years, 219(87.6%) had prelingual onset of disease, and 202(80.8%) had a non-progressive disease course. In 203(81.2%) cases, normal developmental milestones were seen. Parental consanguinity was observed in 219(87.6%) cases. However, 63(25.2%) patients had a first-degree relative who had a history of deafness. In 170(68%) cases, hearing loss was hereditary, whereas in 80(32%) it was acquired. Meningitis was the most commonly identified risk factor 55(68.75%). Acquired risk factors and family history had significant association with hearing loss ($p<0.05$). Speech perception significantly improved in all 219(100%) patients with prelingual hearing loss who underwent cochlear implantation.

Conclusion: Majority of the cases were found to be male, had a prelingual disease onset and a non-progressive disease course. Family history was a significant factor, while meningitis was the most common acquired cause of hearing loss.

Keywords: Autosomal recessive hearing loss, Hereditary hearing loss, Non-syndromic hearing loss, Sensorineural hearing loss. (JPMA 74: 476; 2024) **DOI: https://doi.org/10.47391/JPMA.8708**

Introduction

Hearing loss (HL) is common in children. Their capacity to participate in different activities, their ability to develop their language and cognitive skills, and their quality of life (QOL) can all be adversely affected by HL.[1] According to one of the Global Burden of Disease studies, HL is the fourth most common cause of disability worldwide. Hearing screening programmes for newborns in majority of industrialised nations allow for early discovery, which can aid in early intervention and improved QOL.[2] In Pakistan, hereditary deafness is around 70%, and the increased rate of hereditary HL is because of a high rate of consanguineous marriages.[3] Non-syndromic hearing loss (NSHL) with autosomal recessive (AR) inheritance is seen in 75-80% cases, which is frequent in consanguineous marriages, followed by autosomal dominant (AD) 20-25%, which is postlingual, progressive and prevalent in non-consanguineous marriages, whereas X-linked or mitochondrial cases account for 1-2%.[4] Autosomal dominant inheritance related to HL are considered postlingual HL, which is progressive in nature. Whereas, phenotype for autosomal recessive HL is considered prelingual, and is non-progressive in nature.[5] There are three main types of hearing loss: mixed HL (MHL), conductive HL (CHL), and sensorineural HL (SNHL). The most common cause of CHL is problems related to the ear canal by impediment from wax, debris or foreign bodies. Perforation of the eardrum can also cause CHL.[6] Trauma or disease of cochlea can cause the loss of primary auditory neurons or damage to sensory hair cells that manifest as HL. The cause of SNHL is very complex, which can be congenital or acquired HL (AHL).[7] Hereditary HL (HHL) is classified into syndromic HL (SHL) and non-syndromic HL (NSHL).[8] SHL constitutes 30% and NSHL 70% of the hereditary causes of CHL.[2]

Severity of meningitis is one of the major risk factors for HL in children.[9] Cytomegalovirus (CMV) is the most prevalent cause of congenital infection, and CMV is a major cause of SNHL in children globally.[10]

Cochlear implant has now become a recognised means of hearing rehabilitation for severe to profound HL in

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children.[11] Cochlear implant is well-recognised for its efficacy, reliability and safety, and children appear to be the major group of patients to get benefit. Recent research suggests that healthy spiral ganglion can influence cochlear implant outcome after surgery.[12] Hence, it is a frequently used procedure to treat HL in children.[13]

The current study was planned to analyse the demographic and clinical variables in deaf children having undergone cochlear implant surgery.

Patients and Methods

The cross-sectional study was conducted from January to November 2022 at the Centre for Research in Experimental and Applied Medicine (CREAM) laboratory of the Department of Biochemistry and Molecular Biology, Army Medical College (AMC), Rawalpindi, Pakistan, in collaboration with the Ear, Nose and Throat (ENT) Department of Combined Military Hospital (CMH), Rawalpindi. After approval from the AMC ethical review committee, the sample size was calculated using the World Health Organisation (WHO) calculator.[14] with 95% confidence interval (CI) based on deafness prevalence 0.18%.[15] Since there has not been a population by nature of disability census since 1998, therefore, the sample size was adjusted to identify outliers and to reduce error margin. The sample was raised using non-probability purposive sampling technique. Those included were children of either gender aged up to 10 years who had received a cochlear implant. Children who were not deaf were excluded.

After obtaining written informed consent from children's parents/guardians, demographic and clinical data was collected through questionnaire-based detailed interview. Age, gender, parental consanguinity, HL onset, HL laterality, HL stability, acquired risk factors, family history of the disease, and post-cochlear implant speech perception were noted. The subjects were categorised into SHL, NSHL and AHL groups.

Data was analysed using SPSS 22. Descriptive statistics were used and expressed as frequencies and percentages. Linear regression analysis was used to evaluate the relationship between dependant and independent variables. P<0.05 was considered significant.

Results

Of the 285 deaf patients who received cochlear implants, 250(87.7%) matched the age criterion and were included; 147(58.8%) boys and 146(58.4%) aged 0-5 years (Table 1). Besides, 219(87.6%) had prelingual onset of disease and 31(12.4%) had postlingual disease onset, 202(80.8%) had non-progressive and 48(19.2%) had progressive disease course. All 250(100%0 cases had bilateral HL. Further, 203(81.2%) subjects had acquired development milestones at appropriate ages, while 47(18.8%) had delayed development milestones.

Parental consanguinity was observed in 219(87.6%) cases and 31(12.4%) had no parental consanguinity. There were 175(70%) children born to first-cousin parents, and 44(17.6%) from distantly related marriages. However, 63(25.2%) patients had a first-degree relative with a history

Table-1: Age and gender distribution of the subjects (n=250).

	n (%)
Age (years)	
0-5	146 (58.4)
5.1-10	103 (41.2)
Gender	
Male	147 (58.8)
Female	103 (41.2)

Table-2: Frequency of types of hearing loss (HL), relationship of disease onset and developmental milestones with HL types, consanguinity distribution, and distribution of family history of HL with disease groups (n=250).

Types of HL	n (%)	Disease-onset [n (%)]		Development milestones [n (%)]	
		Prelingual	Postlingual	Normal	Delayed motor skills
NSHL	161 (64.4)	150 (93.2)	11 (6.6)	133 (82.6)	28 (17.4)
AHL	80 (32.0)	61 (76.3)	19 (23.7)	62 (77.5)	18 (22.5)
SHL	09 (3.6)	08 (88.9)	01 (11.1)	08 (88.9)	01 (11.1)
Total	250 (100)	219 (87.6)	31 (12.4)	203 (81.2)	47 (18.8)
Consanguinity distribution					
Relation	**n (%)**				
First cousin	175 (70.0)				
Distant relative	44 (17.6)				
Unrelated	31 (12.4)				
Family history distribution among disease groups [n (%)]					
Hearing loss	**First degree relatives**	**Second degree relatives**	**Third degree relatives**	**Distant relatives**	**Insignificant family history**	**Total**
NSHL	54 (33.5)	12 (7.5)	11 (6.8)	20 (12.4)	64 (39.8)	161
AHL	03 (3.75)	01 (1.25)	04 (05)	05 (6.3)	67 (83.7)	80
SHL	06 (66.7)	0 (0)	0 (0)	0 (0)	03 (33.3)	09
Total	63	13	15	25	134	250

NSHL: Nonsyndromic hearing loss, AHL: Acquired hearing loss, SHL: Syndromic hearing loss.

Table-3: Association between outcome and exposure variables.

Parameters	Odds Ratio (95% CI) (beta-coefficients)	*p*-value
Disease Onset	0.011 (-208- 0.246)	0.869
Stability of HL	-0.086 (-0.312- 0.067)	0.205
Family history of HL	0.127 (.003- 0.080)	0.036*
Parental relation	-0.029 (-0.069-0.115)	0.628
Acquired risk factors	-0.348 (-0.060- -0.29)	0.000*

CI: Confidence interval, HL: Hearing loss. *p-value <0.05 as statistically significant.

of deafness, while 13(5.2%) had second-degree relative, 15(6%) had third-degree relative, 25(10%) had distant relatives, and 134(53.6%) had random family history of HL.

In 170(68%) cases, HL was hereditary, whereas in 80(32%) it was AHL (Table 2). Meningitis was the most commonly identified risk factor 55(68.75%) among AHL cases, followed by typhoid 6(7.5%), toxoplasmosis, others (syphilis, hepatitis B), rubella, cytomegalovirus (CMV), and herpes simplex (TORCH) 5(6.25%), drug-induced HL, long stay in neonatal intensive care unit (NICU), preterm babies and severe jaundice 3(3.75%) each, and exchange transfusion 2(2.5%) cases of AHL.

Overall, there was no significant association between the groups and study variables (p.0.05), although acquired risk factors and family history of HL had significant association with HL ($p<0.05$) (Table 3).

Speech perception significantly improved in patients with prelingual HL who underwent cochlear implantation compared to those with postlingual HL who underwent cochlear implantation ($p<0.05$).

Discussion

The study showed that the majority of cases (58.4%) were under the age of 5 years and had prelingual onset of the disease. Prelingual HL is present in children before they develop their speech.[16] In the current study, prevalence of HL was found more in male children (58.8%). One study reported similar finding.[17] HHL was found in 68% cases of NSHL. One study claimed that genetic mutations are responsible for 80% of CHL.[12] The majority of NSHL cases had prelingual disease onset (93.2%) and non-progressive disease course (88.2%), which might be because autosomal recessive NSHL frequently has a non-progressive disease course and prelingual (until 5 years of age) disease onset.[18] Among NSHL cases, majority (82.6%) of the patients had normal developmental milestones, owing to the fact that they are not associated with other malformations.[19] Parental consanguinity was a common feature in the current study, with overall 70% prevalence of first-cousin marriages. According to a recent survey, 73% of Pakistani marriages are somewhere between relatives, and the nation has the biggest population of deaf children.[20] As per a research, consanguineous marriages frequently result in NSHL with autosomal recessive inheritance, which is reported in 75-80% of cases.[4] Family history of HL was non-significant in 39.8% of the NSHL cases. This phenomenon is most likely caused by the generational inheritance pattern seen in autosomal recessive individuals.[21]

AHL was found in 32% cases. In AHL cases, the disease onset was mostly prelingual (76.3%), and the disease course was non-progressive (65%). This was most likely because prelingual HL is usually brought on by an acquired illness, most frequently a disease or trauma.[22] The majority of acquired risk factors affect children most significantly during pregnancy and the first few years of life.[23] Analysis of the acquired risk factors revealed that meningitis, which was reported in 68.75% HL cases, was the most frequent risk factor. Meningitis is the most common cause of AHL.[24] Typhoid fever was the next most prevalent acquired risk factor leading to HL. Acquired risk factors for HL highlight the importance of detecting the problem early because when children are treated earlier in life for HL, their language, speech and socioemotional development is better than in children who receive treatment later in life.[22]

Speech perception significantly improved after cochlear implantation in 219 children with postlingual HL compared to 31 children with prelingual HL. Children with good language skills prior to cochlear implantation had strong early gains in open-set speech perception skills after implantation. According to these findings, children with postlingual HL are ideal as cochlear implantation applicants.[25]

The current has a limitation related to its small sample size, which was primarily because few cochlear implant centres are functional in the city to manage deafness.

However, in the light of the findings, it is recommended that awareness regarding the need for pre-marital screening. Special genetic counselling programmes need to be initiated at the grassroots level to target the masses. This will not only help in the prevention, but also in the early detection of affected individuals.

Conclusion

Majority of the cases were found to be male, had a prelingual disease onset and a non-progressive disease course. Family history was a significant factor, while meningitis was the most common acquired cause of HL. As parental consanguinity was found to be high, programmes for genetic counselling should be established to inform families about the risk of cousin marriages.

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Author Contribution:

IR: Data collection, analysis and drafting.

AR and AM: Study design, data interpretation, review and final approval.