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Original Article

Common Causes of Congenital Deafness Diagnosed on Computed Tomography and Magnetic Resonance Imaging in a Tertiary Care Diagnostic Center in Southern Punjab

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ABSTRACT

Objective: To determine the causes of congenital deafness in children presenting to a tertiary care diagnostic center in Southern Punjab.

Study Design: Analytical cross-sectional study.

Place and Duration of Study: Department of Radiology, Combined Military Hospital, Multan, Pakistan, from Aug 2021 to Jul 2023.

Methodology: A total of 56 children, <12 years of age, belonging to either gender, who presented with the complaints of congenital deafness were included in the study. Demographic data of all patients and further necessary audiological tests were noted; they were referred for high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) of temporal bone and findings were interpreted by consultant radiologist.

Results: Out of 56 patients, 30(53.6%) patients were males and 26(46.4%) were females. Mean age was 2.80±0.83 years with consanguinity noted in 43(76.8%) cases. The prevalence of inner ear malformations was found to be present in 7(12.5%) cases. Most common anomaly noted was Cystic Hypoplastic Cochlea (CH)-III in 4 out of 56 cases (7.1%), followed by CH-II (n=2, 3.6%) while enlarged vestibular aqueduct was found only in 1 case. Furthermore, out of 7 patients, 14.3% anomalies were detected in right ear, 28.6% in left ear and 57.1% in both ears.

Conclusion: These findings underscore the value of high-resolution temporal bone HRCT and MRI in the etiological workup of pediatric congenital deafness and highlight the need for early imaging-based evaluation to guide appropriate counselling, rehabilitation planning, and future preventive strategies in populations with a high rate of consanguinity.

Keywords: Cochlear implant, congenital deafness, high resolution computed tomography.

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INTRODUCTION

Absence of hearing has social and developmental impacts on a child's life, including inability to develop language skills and communicate.[1] Etiology of congenital deafness may be hereditary or due to any antenatal or natal insult.[2] The aim for these infants is to achieve maximum speech perception and attainment of all linguistic-based or sign skills in future. Data regarding prevalence of congenital deafness in the population vary, as congenital deafness may be mixed, conductive or sensorineural, unilateral or bilateral.[3] In developed countries, the incidence of congenital bilateral hearing loss was,2–4 per 1000 live births, it was higher in developing countries, at ≤6 per 1000 live births.[4] Early diagnosis by the aid of accurate diagnostic imaging tool is fundamental such as high resolution computed tomography (HRCT) scan of temporal bone,[5] as this imaging modality has added advantage of post processing multiplanar 2D and 3D reconstruction.[6] Further, multiplanar mutisequential magnetic resonance imaging (MRI) of temporal bone can augment the findings of HRCT,[8] so it is routinely used in preoperative assessment of all feasible cases. This study was designed to determine the common causes of congenital deafness in a tertiary care diagnostic center in Southern Punjab with the aid of two imaging modalities, HRCT and MRI, to collect local data regarding prevalence of diseases responsible for causing congenital deafness and also help in patient selection that would be suitable for cochlear implantation in future.

METHODOLOGY

This analytical cross-sectional study was conducted in by collaboration between Department of

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Otolaryngology & Auditory Implant, Pediatrics and Diagnostic Radiology at Combined Military Hospital (CMH), Multan, Pakistan, from August 2021 to July 2023. As a prerequisite protocol, approval from the Ethics Review Committee of the hospital was obtained via ERC approval letter no.100/2023. Sample size was calculated using World Health Organization (WHO) sample size calculator taking confidence level 95%, margin of error 5%, reported prevalence of congenital deafness 1.5 per 1,000 newborns.[9] The estimated sample size came out to be 23 patients; however we included a total of 56 patients who reported at CMH Multan with diagnosed congenital deafness.

Inclusion Criteria: Children less than 12 years of age belonging to either gender, diagnosed with congenital hearing loss on clinical examination and audiological tests.

Exclusion Criteria: Patients who developed deafness after any trauma or chronic illness, not declared fit after pre-anesthesia assessment, or those having any contraindication to MRI.

Data of all the patients was recorded on a pre-designed proforma. After taking informed written consent, MRI of every patient was performed on 1.5 Tesla MR Toshiba Unit after proper sedation as per age with a body phased array coil. Sequences used included brain axial and coronal T2 weighted sequence, Brain axial FLAIR, Brain axial -T1, Brain DWI including temporal bones, axial and sagittal oblique CISS sequences to get additional information about any disease process involving the brain that could be contributory for congenital deafness. HRCT was done on a multi-detector 120 slice Toshiba CT scan machine (Canon Medical systems), initially done with axial cuts parallel to the long axis of the lateral semicircular canals. Post processing sagittal and coronal reformats along with 3-D images were obtained. To minimize radiation exposure to eye lens, protective eye shield was used for all patients. HRCT and MRI findings of temporal bones were interpreted by one or more consultant radiologists. Presence or absence of inner ear malformations and any other incidental brain pathologies were recorded. Data was analyzed by using Statistical Package for the Social Sciences (SPSS) version 23.0 and MS Excel 2016 software. Mean±SD was calculated for continuous variable. Frequency and percentage were calculated for categorical variables. Chi square test/Fisher exact (for qualitative variables was applied and *p*-value of ≤0.05 was considered as statistically significant.

RESULT

Out of 56 patients, 30(53.6%) patients were males and 26(46.4%) were females with mean age being 2.80±0.83 years, and consanguinity noted in 43(76.8%) cases. The prevalence of inner ear malformations was found to be present in 7(12.5%) patients in our study, with most common anomaly noted being CH-III in 4 out of 56 cases (7.1%), followed by CH-II (2, 3.6%) and enlarged vestibular aqueduct found only in 1 case as shown Figure-1.

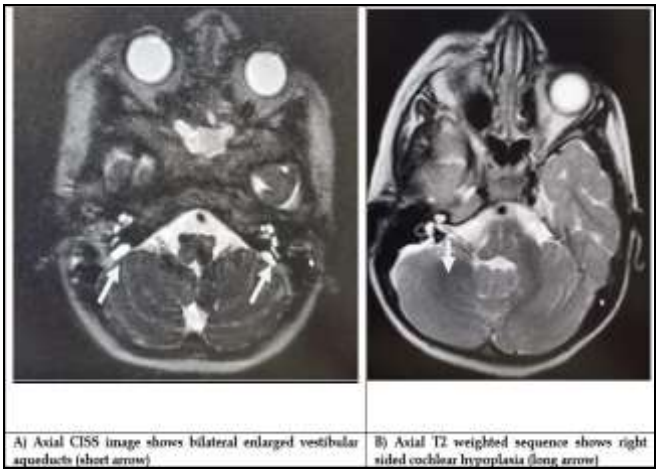

Figure-1: Enlarged Vestibular Aqueduct, (n=1)

Furthermore, out of 7 patients, 14.3% anomalies were detected in right ear, 28.6% in left ear and 57.1% in both ears, as shown in Figure-2.

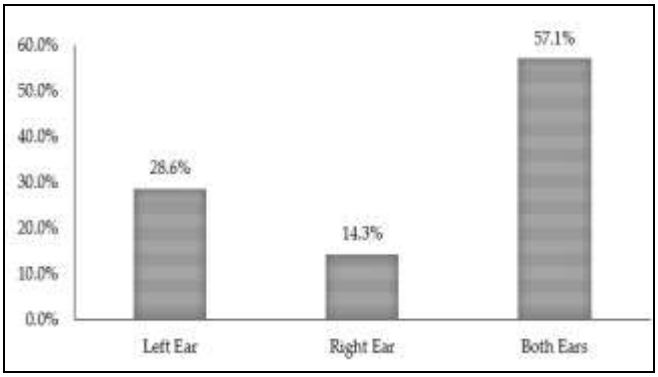

Figure-2: Frequency of Anomalies (n=7)

All Demographic and clinical variables are enumerated in detail as shown in Table-I.

No significant association of gender with type of anomaly was noted (*p*-value=0.825) while a statistically significant association was seen in Consanguinity with type of anomaly (*p*-value=0.043) shown in Table-II.

Table-I: Demographic and Clinical Variables (n=56)

Study Parameters	Values
Age (Mean±SD)	2.80±0.83 years
Gender	
Male	30(53.6%)
Female	26(46.4%)
Inner Ear Malformations	
Present	7(12.5%)
Absent	49(87.5%)
Diagnosis	
Normal study	49(87.5%)
Cystic Hypoplastic Cochlea (CH-II)	2(3.6%)
Cochlea With Less Than 2 Turns (CH-III)	4(7.1%)
Enlarge	1(1.8%)

Table-II: Association of Type of Anomaly with Gender and Consanguinity (n=56)

Parameters	Normal Study (n=49)	(CH-II) (n=2)	(CH-III) (n=4)	Enlarge (n=1)	*p*-value (≤ 0.05)
Gender					
Male	26(53.1%)	1(50.0%)	2(50.0%)	1(100.0%)	0.825
Female	23(46.9%)	1(50.0%)	2(50.0%)	0(0%)	
Consanguinity					
Present	38(77.6%)	0(0%)	4(100.0%)	1(100.0%)	0.043
Absent	11(22.4%)	2(100.0%)	0(0%)	0(0%)	

DISCUSSION

Congenital hearing loss, if not diagnosed and treated early, has multiprong effects,[10-12] due to which all efforts are directed towards detecting it as early as possible. Hearing loss is mainly of two major types: conductive and sensorineuronal with causes of conductive hearing loss including congenital aural atresia, membranous atresia or microtia, with common causes of sensory neuronal hearing loss depending upon the structure involved, classified as: complete labyrinthine aplasia, cochlear hypoplasia, incomplete cochlear partition, enlarged vestibular aqueduct or cochlear aperture abnormalities, with the most commonly found one being cochlear hypoplasia, which has further four subtypes: bud –like cochlea (CH-I), cystic hypoplastic cochlea (CH-II), Cochlea with less than 2 turns (CH-III) and cochlea with hypoplastic middle and apical turns (CH-IV).[13-18] In case of congenital aural atresia or any other external deformity, HRCT & MRI temporal bone is done to evaluate the middle or inner ear structures to diagnose any concurrent abnormality.[19] Ideal treatment modality is cochlear implantation which can only be implanted in patients with an intact cochlear nerve, but presence or absence of cochlear nerve can be assessed by highly resolution CT scan or MRI of temporal bone.[20] A narrow internal auditory canal assessed as having diameter less than 1-2 mm is highly suggestive of cochlear nerve aplasia which is an absolute contraindication for cochlear implant.[8] Studies show that some of the patients found to have normal caliber internal auditory canal on CT scan showed cochlear nerve hypoplasia or aplasia on MRI, likely due to the normal existence of other nervous components of internal auditory canal so a detailed HRCT report should also address all middle ear abnormalities.[21-23] Similarly, detailed evaluation of course of skull base vessels and identification of their aberrant course is mandatory to prevent iatrogenic injury to major vessels.[7] and utilizing diagnostic accuracies of both HRCT and MRI must be an essential preoperative evaluation tool, for detecting the cause of congenital deafness,[24] to further refine management strategies for rehabilitation of affected patients.

LIMITATION OF STUDY

Due to limited resources and affordability issues, many patients with congenital deafness from underdeveloped areas of Southern Punjab could not be included in the study. Moreover, HRCT and MRI machine were not in reach of all the deserving cases and could not be done in patients who were not fit for anesthesia due to other comorbidities.

CONCLUSION

Inner ear malformations constitute an identifiable cause of congenital deafness in a subset of children presenting to a tertiary care diagnostic center in Southern Punjab with our findings underscoring the value of HRCT and MRI in the etiological workup of pediatric congenital deafness and highlight the need for early imaging-based evaluation to guide appropriate counselling, rehabilitation planning, and future preventive strategies in populations with a high rate of consanguinity.

Conflict of Interest: None.

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Authors' Contribution

Following authors have made substantial contributions to the manuscript as under:

MA & AL: Study design, data interpretation, drafting the manuscript, critical review, approval of the final version to be published.

NS & MA: Conception, data analysis, drafting the manuscript, approval of the final version to be published.

AH & NK: Data acquisition, critical review, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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