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Case Report

Congenital Aural Atresia Presenting with Long - Standing Conductive Hearing Loss: A Case Report

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Abstract

Congenital aural atresia is a rare anomaly causing conductive hearing loss. We report a patient with long-standing bilateral hearing impairment, more severe on the left, confirmed by HRCT to have external auditory canal atresia [1]. The patient underwent atresiaplasty with ossicular reconstruction, resulting in improved hearing and communication [2]. Early diagnosis and tailored surgical management are essential to restore auditory function and quality of life.

Keywords: Congenital Aural Atresia; External Auditory Canal Atresia; Conductive Hearing Loss; Atresiaplasty; HRCT Temporal Bone; Hearing Rehabilitation

Introduction

Congenital Aural Atresia (CAA) is a rare developmental anomaly of the external auditory canal (EAC) with variable involvement of middle ear structures, formed when the external auditory canal (EAC) fails to fully develop (recanalize), leaving a plate of bone separating the undeveloped canal from the middle ear, frequently leading to conductive hearing loss [1]. This case study explores the clinical presentation, diagnostic process, management strategies, and outcomes of a patient with congenital aural atresia, highlighting multidisciplinary approaches and long-term considerations.

Background

Congenital Aural Atresia is characterized by partial or complete absence of the external auditory canal, often associated with auricular malformations [1]. Congenital external auditory canal (EAC) stenosis is defined by Cole and Jahrsdoerfer as an external auditory canal with a diameter of less than 4 mm [3]. EAC stenosis

occurs as a result of partial failure of canalization of the EAC from the usual medial to lateral direction [4]. Problems associated with severe congenital EAC stenosis include conductive hearing loss, cerumen impactions, canal cholesteatomas, and the inability to directly see the tympanic membrane for the diagnosis of problems.

Patient information

Age/Sex

18-year-old Male.

Chief complaints

The patient presented with an eight-year history of bilateral hearing impairment, predominantly affecting the left ear (left ear more affected than the right), resulting in difficulty in hearing normal conversational speech.

Figure 1: Patient presentation to the OPD.

History of presenting illness

The patient presented with an eight-year history of bilateral hearing impairment, more pronounced in the left ear. The hearing difficulty was gradually progressive in nature. The patient reported increasing difficulty in perceiving normal conversational speech. There was no history of otalgia, otorrhea, tinnitus, vertigo, ear trauma, or sudden worsening of hearing. No episodic fluctuations in hearing were noted. There was no history suggestive of recurrent ear infections, prolonged upper respiratory tract infections, or exposure to ototoxic medications. The patient did not report any significant occupational or recreational noise exposure. No associated neurological symptoms, including facial weakness or imbalance, were present. The hearing impairment had begun to interfere with daily communication and social interactions, prompting medical evaluation.

Differential diagnosis

- Congenital Aural Atresia [1]
- Congenital External Auditory Canal Stenosis [1]
- Acquired External Auditory Canal Atresia [5]
- Congenital Cholesteatoma [1]
- Middle Ear Ossicular Malformation [1]
- Otitis Media with Effusion [6]
- Syndromic Microtia–Atresia (e.g., Treacher Collins, Goldenhar syndrome) [7]

Investigations/Diagnostic work up

Clinical examination of the ear

- Pre auricular, pinna and post auricular regions were found to be normal.
- Pinkish membrane like structure was placed laterally and did not move with Siegelization.
- Prominent feature of the tympanic membrane could not be made out.
- Fistula Test – Negative
- Mastoid Tenderness – Negative
- The facial nerve was intact, bilaterally after performing clinical examination of the nerve.

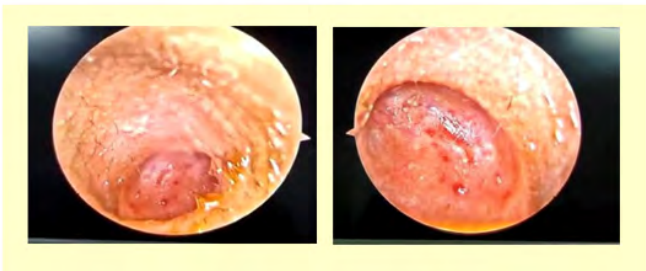

Figure 2: Otoscopic view of both the ears showing the pinkish membrane.

Tuning fork test

	Right	Left
Rinne's Test	BC>AC (Negative)	BC>AC (Negative)
Weber Test	Lateralized to Left Ear	
ABC	Not Reduced	Not Reduced

Table 1

Pure tone audiometry [8]:

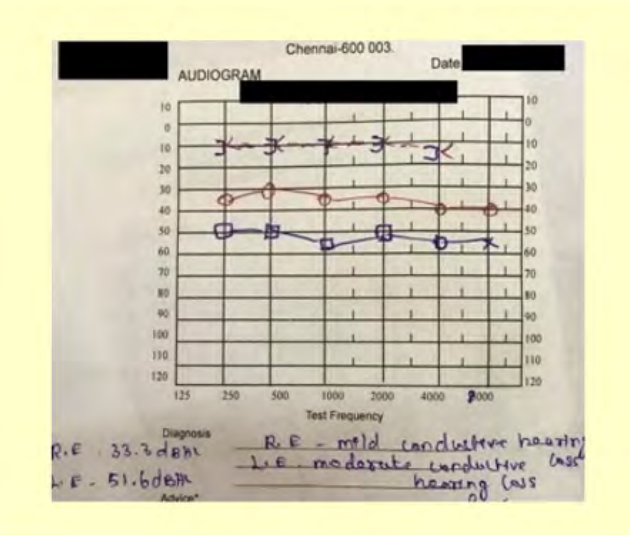

Figure 3: Pure Tone Audiometry Preoperatively.

Right Ear - 33.3 dB – Mild Conductive Hearing Loss

Left Ear - 51.6 dB – Moderate Conductive Hearing Loss.

Radiologic investigations

High Resolution Computed Tomography (HRCT) Head : HRCT of the temporal bone revealed atresia of the external auditory canal and poorly developed tympanic cavity, while the inner ear structures were preserved.

Jahrsdoerfer Grading Scale [9] (To determine appropriateness for congenital aural atresia reconstruction).

A score of less than 7 indicates that the patient is not a a favourable surgical candidate.

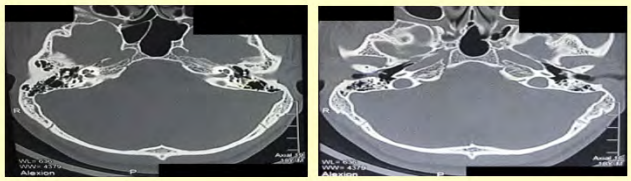

Figure 4: HRCT section of ear and its structures showing the Aural Atresia with atretic plate.

Temporal Bone Parameters	Maximum Points	Points Awarded
Stapes Present	2	2
Oval Window Normal	1	1
Round Window Normal	1	1
Malleo-incudal Complex Normal	1	1
Incudo-stapedial Connection Normal	1	1
Middle Ear Space Pneumatized	1	1
Mastoid Cavity Pneumatized	1	1
Facial Nerve Course Normal	1	1
Appearance of External Ear	1	1
Total Points	10	10
Recommended Score for Atresiaplasty	If the score is ≥7 then the patient is considered for surgery with good prognosis	

Table 2

A score that is 7 or greater indicates that the patient may be considered a candidate for atresiaplasty.

The higher the total number in the grading scale the more likely a good hearing outcome after surgery [9].

Diagnosis

Based on the clinical features and the diagnostic workup done, the patient is diagnosed with "Congenital aural atresia causing long-standing conductive hearing loss".

Management

The patient was managed with atresiaplasty following the comprehensive clinical, audiological, and radiological evaluation. After appropriate counseling regarding potential benefits, risks, and postoperative expectations, the patient was deemed a suitable candidate for surgical intervention.

Atresiaplasty involved the creation of a neo-external auditory canal, removal of the atretic plate, and reconstruction of the middle ear to establish a functional sound conduction pathway. Ossiculoplasty was performed where required, and a temporalis fascia graft was used for tympanic membrane reconstruction.

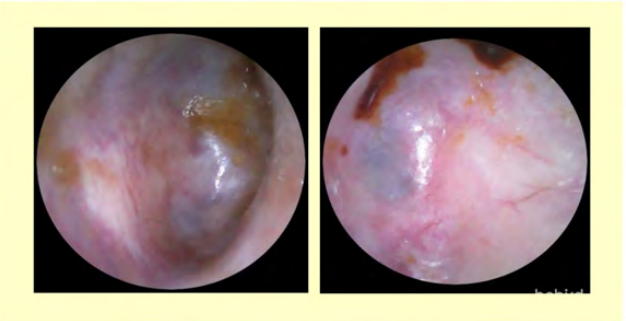

Figure 5: Otoscopic view of both the ears postoperatively.

Postoperatively, a canal stent was placed temporarily to prevent restenosis, and the patient was advised regular follow-up for aural toileting and audiological assessment.

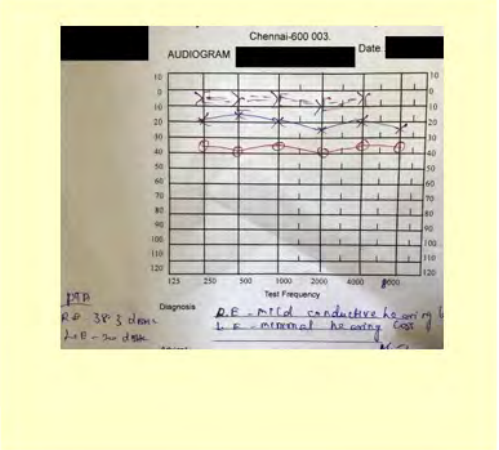

Figure 6: Pure Tone Audiometry Postoperatively.

The procedure resulted in improved hearing thresholds from 51.6 dB (moderate conductive hearing loss) to 20dB (Minimal Hearing Loss) and enhanced functional communication, emphasizing the role of atresiaplasty as an effective management option in selected cases of congenital aural atresia.

Discussion

Congenital aural atresia (CAA) presents significant surgical challenges due to altered anatomy, especially involving the facial nerve and middle ear structures, which contribute to variable outcomes and risks such as facial palsy and cholesteatoma [10-12]. Surgical success is often defined by achieving a hearing threshold of 20 dB or better, which is met in a minority of cases (8–30%), reflecting the complexity of restoring normal hearing [10]. Hearing loss in CAA is typically conductive and does not improve over time, with no significant difference in audiometric progression between complete atresia and external auditory canal stenosis [13]. Cholesteatoma risk is low in true atresia (1.7%) but markedly higher in canal stenosis (43%), guiding recommendations against routine imaging in asymptomatic CAA but supporting surveillance in stenosis cases, especially before age 12 [12]. Treatment options include atresiaplasty and osseointegrated bone conduction devices (OBCD); while OBCD shows greater pure-tone threshold improvement, atresiaplasty may provide better overall auditory access due to continuous canal use, highlighting the importance of patient selection and surgical expertise.

In the present case, the patient had all the features suggestive of CAA, which were hard of hearing for the past eight years and difficulty in hearing normal conversations. Then the examination of the ear came into play were we visualized a pinkish membrane which did not move with siegalization and then the complex yet prominent tympanic membrane anatomy was not being visualized, this further took us to undertake some investigations which was Pure Tone Audiometry which showed he had an Conductive Hearing Loss and the HRCT aiding in the diagnosis and with the use of Jahrsdoerfer Scale, it clearly gave a green signal towards proceeding with the surgery as the definitive management for the betterment of the patient in the long run.

Conclusion

Congenital aural atresia is a rare developmental anomaly that can result in long-standing conductive hearing loss if not addressed

appropriately. Early recognition, detailed audiological assessment, and high-resolution imaging are essential for accurate diagnosis and treatment planning. Atresiaplasty, when performed in carefully selected patients, can effectively restore the sound conduction pathway and significantly improve hearing outcomes. This case highlights the importance of individualized management and a multidisciplinary approach to optimize functional hearing and overall quality of life.

Future Directions

- Scope out newer perspectives in the treatment and functional outcomes of Congenital Aural Atresia
- Any possibility for minimally invasive surgery for Congenital Aural Atresia
- Use 3D Bioprinting for the customized surgical plan in Congenital Aural Atresia.

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