Case Report

International Journal of Tumor Research

Bilateral Congenital Aural Atresia (Type III) with Anotia and Microtia: Case Report and Review of Literature

Bheru Dan Charan[1]*, Amreen[2], Navdeep Kaur[3] and Samanyu[1]

[1]*Department of Radiology, All India Institute of Medical Sciences, Rishikesh, India.*

[2]*Department of ENT, All India Institute of Medical Sciences, Bathinda, India.*

[3]*Department of Radiology, All India institute of medical sciences, Bathinda, India.*

***Correspondence:**

Dr. Bheru Dan Charan, Assistant Professor, All India institute of medical sciences, Rishikesh, India, Mobile: 09680699678.

Received: 10 Oct 2025; **Accepted:** 16 Nov 2025; **Published:** 30 Nov 2025

Citation: Bheru Dan Charan, Amreen, Navdeep Kaur, et al. Bilateral Congenital Aural Atresia (Type III) with Anotia and Microtia: Case Report and Review of Literature. Int J Tumor Res. 2025; 1(1): 1-3.

ABSTRACT

Congenial aural atresia and microtia commonly coexist due to the common embryological development of the external ear canal and pinna. In congenital aural atresia, the middle ear may be completely normal or have a malformation. High-resolution CT (HRCT) is necessary to find various associated anomalies in the middle ear. Here, we report a rare bilateral congenital aural atresia with bilateral anotia and microtia. The patient was advised on the bone-anchored hearing aid. This case report aims to raise awareness among clinicians about HRCT for patient treatment and outcomes.

Keyword

Atresia, Ear, Anotia.

Abbreviations

CAA: Congenital aural atresia, HRCT: High-resolution computed tomography, DB: Desible, PTA: Pure tone audiometry.

Introduction

Congenital aural atresia (CAA) is an anomaly of the external auditory canal that occurs due to failure in the recanalisation of the epithelial plate of the first branchial arch groove and is commonly associated with malformed (microtia) or absent (anotia) external ear [1]. The auricles develop from the 1st and 2nd branchial arches, while the external canal and tympanic membrane develop from the first branchial cleft. It has an incidence of 1 per 10,000–20,000 live births [2], but bilateral field is only in 25–30% [3] of these cases and is more common in males. Unilateral atresia is five times more common and commonly affects the right side [4]. It can occur with Trisomy 18 and syndromes such as Treacher-Collins, Pierre Robin, and Goldenhar. Bilateral CAA can be seen in 60% of cases of CHARGE syndrome [5]. Early diagnosis and intervention by 6 months of age are necessary to prevent permanent hearing disability [6,7]. Here we described a case of bilateral CAA with left side anotia and right2020 side grade III microtia.

Case Presentations

A 22-year-old male presented to the department of ENT with decreased hearing since birth. He had minimal hearing through his right ear compared to his left ear. There is no history of ear discharge and no other systemic illness. He had everyday language. Their family history is negative for similar anomalies. On examinations, he has right-side grade III (Marx classification) microtia (Figure 1a) with left-side anotia (Figure 1 b) and bilateral ear atresia. Audiometry shows a bilateral profound mixed hearing loss. During the BERA examination, EEG activity was fair. The patient was awake throughout the test. Threshold estimation was done at 250 Hz, 500 Hz, 1000 Hz, 2000 Hz, 4000 Hz, and 8000 Hz. Estimated pure tone audiometry (PTA) > 118.75 dB HL in the left ear and 101.25 dB HL in the right ear. HRCT temporal bone (Figure 2a to 2f in caudo-cranial direction) revealed complete atresia of the bilateral external auditory canal. No external bony depression or pit is seen. The bilateral groove for the superior petrosal sinus, Koerner septum, aditus, and mastoid antrum appears normal. The middle ear cavity was reduced in size with loss of Prussak space bilaterally (Figure 2c and 2d). Soft tissue density was seen in the left middle ear cavity, likely cholesteatoma, which is attributed to severe hearing loss in the left ear (Figure 2b, 2c, 2e, and 2f). The horizontal segment of the left facial canal was eroded in the middle ear (Figure 2 b). The Malleus head

was dysplastic bilaterally, and no scutum was visible. We have summarized Yeakley and Jahrsdoerfer's score in Table 1. Normal stapes are seen bilaterally attached to the oval window—absent/scanty pneumatisation on the right side, with sclerosis and reduced pneumatisation on the left side. Bilateral inner ear structures appear normal. The right side middle ear bones were dysplastic and inseparable from the bony atretic wall (Figure 2 b).

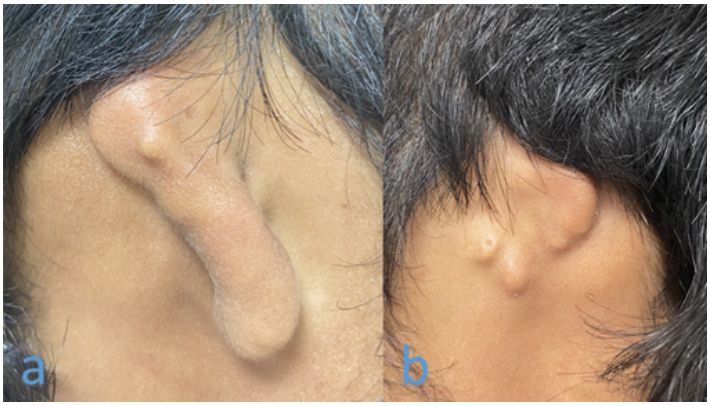

Figure 1: Picture of the right ear (1a) showing a small, malformed pinna (microtia grade III) and left ear (1b) showing only a residual skin tag (anotia) with non-visualisation of the bilateral external auditory orifice.

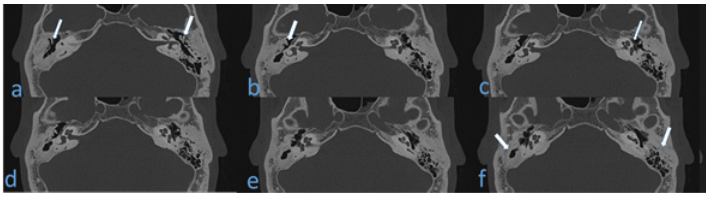

Figure 2: HRCT axial scan of the temporal bone. (These images are sequentially acquired from caudal to cranial direction from 2a to 2f.) Bilateral deformed malleus bone (arrow in Figure 1a and 1b) with reduced size of the middle ear cavity. The middle ear ossicles on the right side are inseparable from the wall of the middle ear cavity. The bilateral inner ear structure and semi-circular canals are standard. Soft tissue density was noted in the left middle ear cavity, which caused erosion of the facial canal horizontal segment (arrow in Figure 1c) on the left side, s/o cholesteatoma—on the mastoid sinus (arrow in Figure 1f), showing sclerosis with reduced pneumatisation on the left with absent/scanty pneumatisation on the right side (Figure 1d-f).

Table 1: Demonstrating Yeakley and Jahrsdoerfer's score based on HRCT temporal bone.

Yeakley and Jahrsdoerfer Score	Right Ear (5) (Grade III microtia)	Left Ear (4) (Anotia)
Normal Stapes	2	2
Open oval window	1	1
Middle ear space	0	0
Facial nerve position	1	0
Malleus-incus complex	0	0
Mastoid pneumatization	0	0
Incus-stapes connection	0	0
Open round window	1	1
Total	5	4

The patient was planned and counselled for a bone-anchored hearing aid (BAHA). To date, the patient has not visited the OPD on follow-up.

Discussion

The severity of congenital atrial atresia (CAA) deformities can range from narrowing (stenosis) to the total absence of the external auditory canal. Most cases are sporadic, often associated with microtia, and less frequently with inner ear malformation. The auricle and external auditory canal have a common embryological origin, with the auricle developing from the first and second pharyngeal arches, and the external auditory canal arising from the first pharyngeal cleft, so these two entities are commonly associated. External auditory canal atresia is more common than stenosis, and among atretic ears, bony atresia is more common than membranous atresia. To thoroughly assess the ear and determine which patients might benefit from reconstructive surgery or canaloplasty, high-resolution computed tomography (HRCT) of the temporal bone is essential [8,9].

Only a few case reports of bilateral CAA with anotia or microtia have been described in the literature [10,11]. Our case is classified as type III CAA (occluded auditory canal with a malformed middle ear cavity) based on Altmann ‘s-Cremers classification [3,12] and type D according to the Schuknecht [13] classification, which is the most severe type. These cases are usually associated with isolated moderate-severe conductive hearing loss [14], and our patient had bilateral profound mixed hearing loss with a threshold of > 110 dB. CAA can be accompanied by DiGeorge, VATER, Klippel–Feil, Fanconi, Pierre Robin, and CHARGE syndromes with concomitant anomalies including cleft palate, hemifacial microsomia, posterior cranial hypoplasia, hydrocephalus, and genitourinary abnormalities [15]; however, in our case, no other anomalies were seen. Poor mastoid pneumatisation is considered a significant anomaly and was present bilaterally in our case [16]. External auditory canal cholesteatoma can be noted in 20.8 per 48 cases of congenital aural atresia [17,18]. Jahrsdoerfer's scoring system is used to determine the candidacy for surgery. A score of 5 or less disqualifies the patients from surgery, as in our case. Bilateral atresia is corrected at 4 or 5 years of age, and unilateral atresia, surgery can be postponed until young adulthood. Surgery like auriculoplasty is challenging because of the altered anatomy of surgical landmarks and the chance of restenosis. A bone-anchored hearing aid is preferred.

Conclusion

Congenital aural atresia can be associated with various middle-ear and occasional inner-ear anomalies. Microtia can be associated with CAA. HRCT is essential for detailed ear evaluation and surgery planning.

References

1. Nicholas BD, Kesser BW. Unilateral aural atresia: current management issues and results. Curr Otorhinolaryngol Rep. 2013; 1: 92-98.

2. Kelley PE, Scholes MA. Microtia and congenital aural atresia. Otolaryngol Clin North Am. 2007; 40: 61-80.
3. Hol MK, Cremers CW, Coppens Schellekens W, et al. A new treatment for young children with bilateral congenital aural atresia. Int J Pediatr Otorhinolaryngol. 2005; 69: 973-980.
4. Aggarwal P, Goyal JP, Gupta A, et al. Congenital cholesteatoma with canal atresia: three case reports. Indian J Otol. 2013; 19: 146-148.
5. Tsang WSS, Tong MCF, Ku PKM, et al. Contemporary solutions for patients with microtia and congenital aural atresia – Hong Kong experience. J Otol. 2016; 11: 157-164.
6. Lim R, Abdullah A, Wan Hashim WF, et al. Hearing rehabilitation in patients with congenital aural atresia: an observational study in a tertiary center. Egypt J Otolaryngol. 2023; 39: 90.
7. Declau F, Cremers C, Van De Heyning P, et al. Diagnosis and management strategies in congenital atresia of the external auditory canal. Br J Audiol. 1999; 33: 313-327.
8. Jacob R, Gupta S, Isaacson B, et al. High-resolution CT findings in children with a normal pinna or grade I microtia and unilateral mild stenosis of the external auditory canal. AJNR Am J Neuroradiol. 2015; 36: 17680.
9. Takegoshi H, Kaga K, Kikuchi S, et al. Mandibulo facial dysostosis: CT evaluation of the temporal bones for surgical risk assessment in patients of bilateral aural atresia. Int J Pediatr Otorhinolaryngol. 2000; 54: 33-40.
10. Solmaz E, Öztürk M, Fazl oullar› Z, et al. Bilateral atresia of the external acoustic meatus: a case report. Anatomy. 2020; 14: 72-75.
11. Amine AM, Rabii L, Zineb EK, et al. Bilateral Microtia and Congenital Aural Atresia: About a Case. Clin Res Pediatr. 2018; 1: 1-3.
12. Cremers CW, Teunissen E, Marres EH. Classification of congenital aural atresia and results of reconstructive surgery. Adv Otorhinolaryngol. 1988; 40: 9-14.
13. Schuknecht HF. Congenital aural atresia. Laryngoscope. 1989; 99: 908-917.
14. Denoyelle F, Coudert C, Thierry B, et al. Hearing rehabilitation with the closed skin bone-anchored implant Sophono Alpha1: Results of a prospective study in 15 children with ear atresia. Int J Pediatr Otorhinolaryngol. 2015; 79: 382-387.
15. Tubbs RS, Shoja MM, Loukas M. Bergman's comprehensive encyclopedia of human anatomic variation. Hoboken (NJ): John Wiley & Sons. 2016; 1173-1175.
16. De la Cruz A, Teufert KB. Reconstruction of the auditory canal and tympanum. Cummings otolaryngology head and neck surgery. Philadelphia (PA): Elsevier Mosby. 2010. 2752-2760.
17. Cole RR, Jahrsdoerfer RA. The risk of cholesteatoma in congenital aural stenosis. Laryngoscope. 1990;100: 576-578.
18. Casale G, Nicholas BD, Kesser BW. Acquired ear canal cholesteatoma in congenital aural atresia/stenosis. Otol Neurotol. 2014; 35:1474-1479.