Microtia and congenital atresia of the external auditory canal: a case report

Clinical Case

Authors

Pedro Marques Gomes
Unidade Local de Saúde de Matosinhos – Hospital Pedro Hispano, Portugal

Diogo Cunha Cabral
Unidade Local de Saúde de Matosinhos – Hospital Pedro Hispano, Portugal

Delfim Duarte
Unidade Local de Saúde de Matosinhos – Hospital Pedro Hispano, Portugal

Gustavo Lopes
Unidade Local de Saúde de Matosinhos – Hospital Pedro Hispano, Portugal

Correspondence:
Pedro Marques Gomes
apedrommarquesgomes@hotmail.com

Article received on March 23, 2024.
Accepted for publication on July 10, 2024.

Abstract

Microtia and congenital atresia of the external auditory canal (EAC) are congenital malformations of the external ear that are often associated.
Microtia results from a malformation of the auricle, while congenital atresia is a total or partial occlusion of the EAC.
The challenge for the otolaryngologist is the management, which may include reconstructive surgery and auditory rehabilitation.
Each case is unique and requires an individualized approach by a specialized team of otolaryngologists and plastic surgeons.
We present the case of a 62-year-old woman with microtia and congenital atresia of the right EAC, with a history of reconstructive surgery of the pinna, in which auditory rehabilitation with an osseointegrated implant was chosen.
Keywords: Microtia; Congenital atresia of the external auditory canal; Deafness; Osteointegrated hearing implant.

Introduction

Microtia is a congenital malformation of the auricle that is generally associated with congenital external auditory canal (EAC) atresia. It has a reported incidence of 1 in 10,000 individuals, with both environmental and genetic etiological factors identified in previous studies[1]. Microtia and congenital EAC atresia can be associated with ocular, cervical, cardiac, and renal anomalies, which require thorough evaluation and treatment. Currently, several systems are used to grade the level of microtia (Marx[2], Table 1) and EAC atresia (Weerda[3], Table 2) and provide information on the most appropriate therapeutic approach for each patient. The Jahrsdoerfer grading scale is based on temporal bone computed tomography (CT) and helps in determining which patients are eligible for canalplasty (Table 3)[4].

Table 1
Marx clinical staging system for microtia (adapted[2])

Grade I	Smaller ear but all the characteristics of a normal auricle are recognizable
Grade II	Some characteristics of a normal auricle are recognizable
Grade III	Rudimentary soft tissue and cartilage (upper cartilage remnants, anterosuperiorly rotated lobe)
Grade IV	Absence of the ear canal and auricle

Table 2
Weerda clinical staging system for congenital EAC atresia (adapted[3])

Type A	Marked EAC narrowing with an intact skin layer.
Type B	Partially-patent lateral EAC with an atretic middle meatus plate.
Type C	Complete EAC atresia.

EAC, external auditory canal.

Table 3
Jahrsdoerfer grading scale (adapted4)

Anatomical structure	Score
Favorable stapes	2
Open oval window	1
Pneumatized middle ear	1
Favorable facial nerve	1
Favorable incudomalleolar joint	1
Intact incudostapedial joint	1
Well-pneumatized mastoid	1
Open round window	1
Normal auricle	1
Total	10

This scale determines canalplasty indication based on ear computed tomography (CT) findings. ≥ 7: favorable; < 6: not favorable

Patients who are not eligible for or refuse canalplasty have several therapeutic options, including surveillance in case of unilateral atresia; soft or rigid band bone conduction hearing aid (pediatric age); osseointegrated bone conduction implant; or the Contralateral Routing of Signals (CROS) or Bilateral Contralateral Routing of Signals (biCROS) systems, depending on the contralateral auditory function[5].

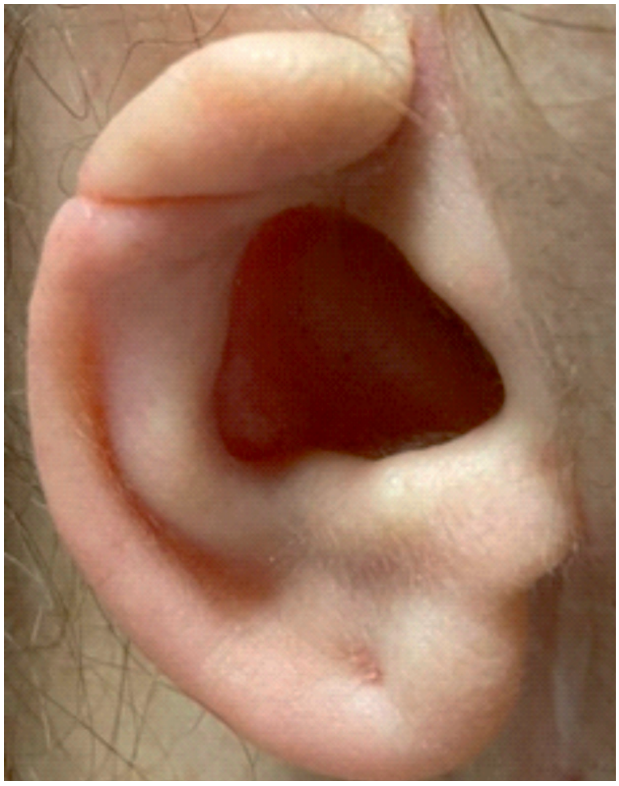
Clinical Case

A 62-year-old White woman with a history of dyslipidemia, osteoporosis, and hypothyroidism visited the Otorhinolaryngology (ORL) department at the Pedro Hispano Hospital (HPH). The patient had hearing loss in the right ear since birth and in the left ear for the last two years, which was associated with constant tinnitus, like the sound of the ocean. She had no other ORL complaints or history of auditory rehabilitation. Her surgical history included reconstructive right ear surgery in a private hospital in 2010, with no surgical records. General ORL examination revealed a normally-implanted left auricle, without any malformation, and a permeable EAC and normal tympanic membrane. The right auricle exhibited grade II microtia (Figure 1 , Table 1), with an external auditory meatus and EAC in cul-de-sac. The tympanic membrane was not visible. Audiometry revealed a negative Rinne test on the right side and positive on the left, while the Weber test showed lateralization to the right. Pure tone (Figure 2) and speech discrimination audiometry exhibited severe mixed hearing loss on the right, with an air-bone gap of 40–60 dB at conversational frequencies, Speech Recognition Threshold (SRT) of 80 dB, and 100% intelligibility at 110 dB. The patient had mild sensorineural hearing loss on the left, with an SRT of 30 dB and 100% intelligibility at 60 dB.

Ear CT (Figure 3) revealed membranous EAC atresia on the right side, middle ear dysplasia with rudimentary manubrium mallei, and undefined incudomalleolar and incudostapedial joints. Additionally, the carotid canal demonstrated protrusion and dehiscence at the level of the middle ear. The left ear showed no abnormalities.

The patient was diagnosed with grade II microtia (Table 1) and type B congenital EAC atresia (Table 2) of the right ear, with severe mixed hearing loss. Canalplasty was ruled out due to an unfavorable middle ear anatomy. We presented the available therapeutic options to the patient, which included surveillance or osseointegrated implant. The patient chose to have an osseointegrated hearing device implanted in her right ear.

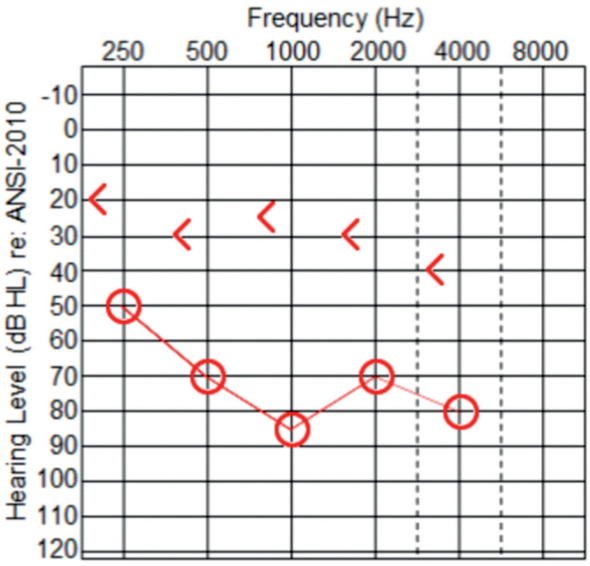
Figure 1
Grade II microtia on the right.



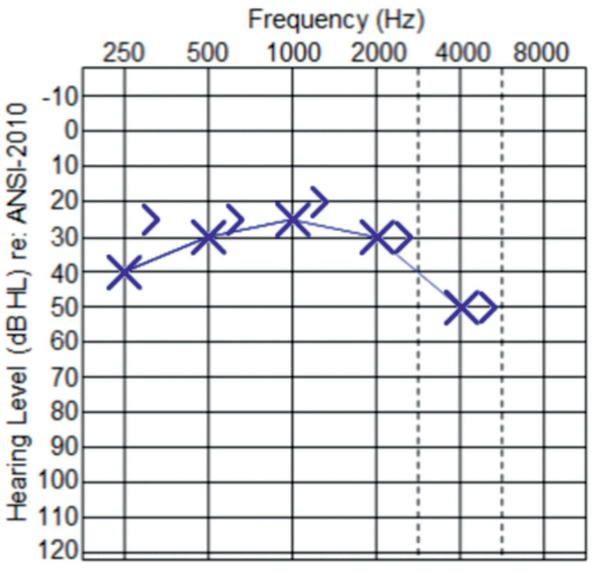
Discussion

EAC atresia refers to the acquired or congenital lack of a patent ear canal. Acquired atresia is often triggered by inflammation, trauma, or ear surgery[1]. Congenital atresia is an EAC malformation that causes conductive hearing loss in the newborn and persists into later life[1]. The middle ear can be structurally and functionally normal or exhibit concomitant malformations[1]. Congenital atresia is usually associated with microtia, an auricle malformation[6]. The case reported here had congenital membranous EAC atresia with concurrent middle ear and auricle malformations.

Figure 2
Pure tone audiometry. SRT, speech reception threshold

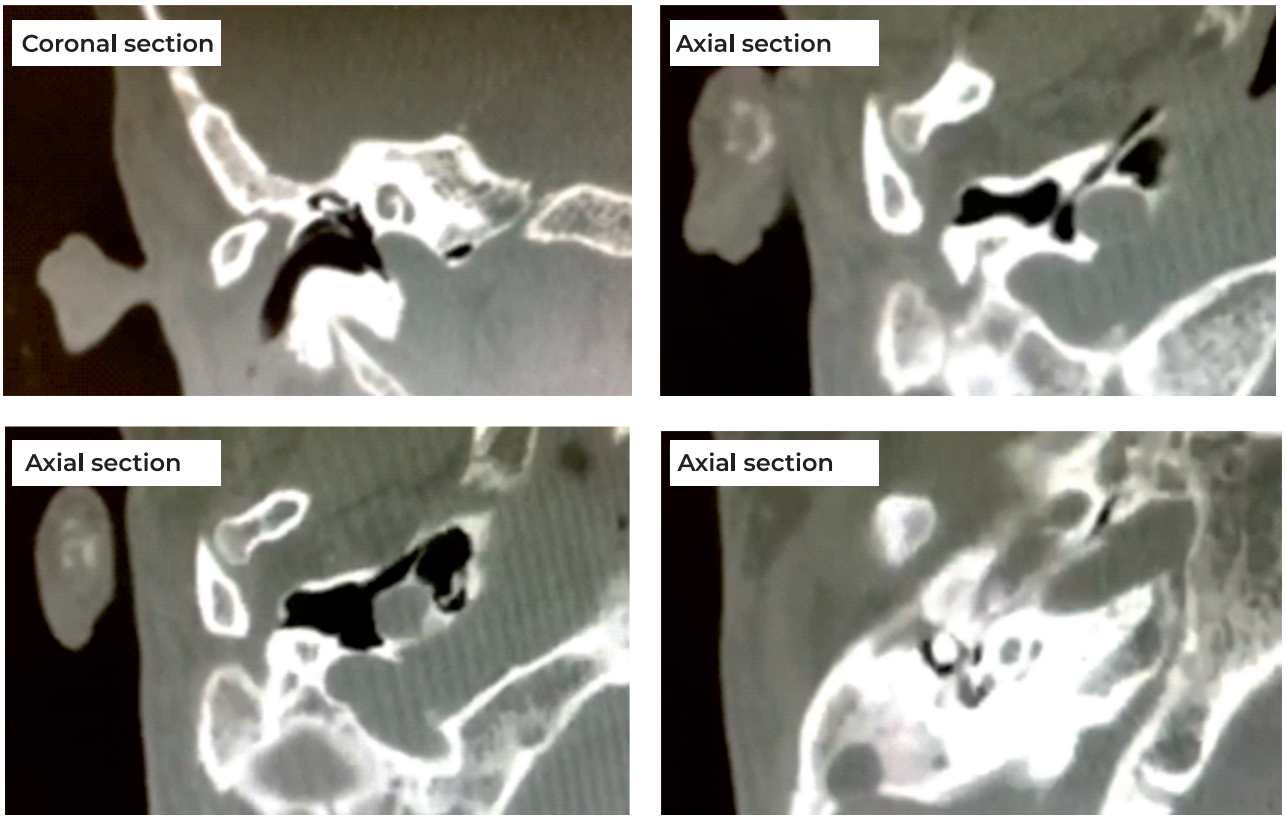


SRT of 80 dB; 100% intelligibility at 110 dB



SRT of 30 dB; 100% intelligibility at 60 dB

Figure 3
Computed tomography (CT) of the right ear (axial and coronal sections).



The Marx clinical staging system classifies microtia into four grades according to the severity of the malformation (Table 1)[2]. Our patient exhibited grade II microtia, in which some features of a normal auricle are recognizable—in this case the tragus, antitragus, lobe, concha, and external auditory meatus.

The Weerda clinical staging system classifies congenital EAC atresia into three types according to the severity of the EAC obstruction (Table 2)[3]. Our patient presented with type B atresia, with a partially-patent lateral EAC but with an atretic middle meatus plate.

These auricular and EAC malformations can lead to a significant degree of hearing loss, with a direct impact on the quality of life. Treatment options for congenital EAC atresia include canalplasty, osseointegrated hearing implant, or the CROS/biCROS system[5]. The appropriate therapy is guided by disease laterality, hearing condition, aesthetic desires, and hearing restoration feasibility. Recent studies have demonstrated that pediatric unilateral atresia can affect academic performance[7]. Patients with microtia and atresia should have the EAC atresia repaired in combination with auricle reconstruction. Although the guidelines are not definitive, hearing restoration should be prioritized over aesthetics, and many surgeons advocate repair of canal atresia before reconstructing the auricle[8]. In our case, we assumed that the patient underwent auricle reconstruction without EAC repair. This may be because of the unfavorable middle ear anatomy, with a dysplastic ossicular chain and protrusion of the carotid canal, which led to a high surgical risk.

The Jahrsdoerfer classification system[4] scores (1–10) the atretic ear according to the presence or absence of nine anatomical structures, with the stapes accounting for two points (Table 3). In addition to evaluating the patient's surgical indication, this scale helps to predict the audiometric result, with the higher the score, the greater the likelihood of postoperative hearing recovery[4,9]. A poorly-pneumatized middle ear and mastoid are the main predictors

of poor postoperative results[4]. Patients with a score greater than six are eligible for surgery. Our patient scored six points on the Jahrsdoerfer grading scale (Table 3); therefore, he was not considered a good candidate for canalplasty. This decision was based on the presence of an unfavorable ossicular chain, unconfirmed incudostapedic joint integrity, poorly pneumatized mastoid, and grade II microtia. The CROS and biCROS systems are good therapeutic options for patients with unilateral hearing loss or significant auditory asymmetry[10]. The CROS system can be prescribed for patients with normal pure tone audiometry in the unaffected ear, while the biCROS system is recommended for patients with sensorineural hearing loss in the less affected ear[10]. Since our patient exhibited auricle and EAC malformations, conventional hearing aids and the CROS/biCROS system were not indicated. Osseointegrated hearing implants are also important therapeutic options. These medical devices are implanted in a simple one- or two-step procedure, under local or general anesthesia. They represent a reversible surgical method without the risk of additional hearing loss and avoid the aesthetic concerns related to conventional hearing aids[11]. The prosthesis is implanted directly into the temporal bone and fuses with the skull in an osseointegration process. Direct implantation has several advantages over conventional hearing aids, eliminating the need for in-the-ear or in-the-canal devices, thereby reducing moisture accumulation and discomfort in the EAC[11]. Osseointegrated hearing implants are good alternatives for patients with contraindications to conventional hearing aids, such as congenital or acquired anatomical deformities and chronic middle or external ear infections[12]. Recent studies[11,12] have demonstrated the beneficial effects of osseointegrated hearing implants in deaf patients, including the fact that the implants provide better speech discrimination in noisy environments than the traditional CROS system. However, neither the osseointegrated implant nor the CROS system improved the sound localization.

Surveillance without auditory rehabilitation is also a therapeutic option, but our patient underwent a rehabilitative approach because she had severe hearing loss in the affected ear, which affected her quality of life.

Conclusion

Treating patients with microtia and congenital EAC atresia is a challenging yet rewarding experience. The choice between surgical and clinical auditory rehabilitation depends on the patient's hearing capacity, anatomy of the middle and inner ear, and the surgeon's preferences. This process requires close collaboration and discussion, and respect for the wishes of the patient and their family.

Conflict of Interests

The authors declare that they have no conflict of interest regarding this article.

Data Confidentiality

The authors declare that they followed the protocols of their work in publishing patient data.

Human and animal protection

The authors declare that the procedures followed are in accordance with the regulations established by the directors of the Commission for Clinical Research and Ethics and in accordance with the Declaration of Helsinki of the World Medical Association.

Privacy policy, informed consent and Ethics committee authorization

All the processed data were based in published reports that fulfilled privacy policy and ethical considerations.

Financial support

This work did not receive any grant contribution, funding or scholarship.

Scientific data availability

There are no publicly available datasets related to this work.

References

1. Bartel-Friedrich S, Wulke C. Classification and diagnosis of ear malformations. GMS Curr Top Otorhinolaryngol Head Neck Surg. 2007:6:Doc05.

2. Campo AA, Victor-Baldin A, Contreras-Mérida SM. Surgical-based classification for microtia. J Craniofac Surg. 2018 Sep;29(6):1651-1654. doi: 10.1097/SCS.0000000000004772.

3. Zhang TY, Bulstrode N, Chang KW, Cho YS, Frenzel H, Jiang D. et al. International consensus recommendations on microtia, aural atresia and functional ear reconstruction. J Int Adv Otol. 2019 Aug;15(2):204-208. doi: 10.5152/iao.2019.7383.

4. Shonka DC Jr, Livingston WJ 3rd, Kesser BW. The Jahrsdoerfer grading scale in surgery to repair congenital aural atresia. Arch Otolaryngol Head Neck Surg. 2008 Aug;134(8):873-7. doi: 10.1001/archotol.134.8.873.

5. Watkinson JC, Clarke R. editors. Scott-Brown's Otorhinolaryngology Head and Neck Surgery, Volume 2: Paediatrics, The Ear, and Skull Base Surgery. Boca Raton: CRC Press; 2018.

6. Abdel-Aziz M. Congenital aural atresia. J Craniofac Surg. 2013 Jul;24(4):e418-22. doi: 10.1097/SCS.0b013e3182942d11.

7. van Hövell Tot Westerflier CVA, van Heteren JAA, Breugem CC, Smit AL, Stegeman I. Impact of unilateral congenital aural atresia on academic performance: a systematic review. Int J Pediatr Otorhinolaryngol. 2018 Nov:114:175-179. doi: 10.1016/j.ijporl.2018.09.002.

8. Chang SO, Min YG, Kim CS, Koh TY. Surgical management of congenital aural atresia. Laryngoscope. 1994 May;104(5 Pt 1):606-11. doi: 10.1002/lary.5541040514.

9. Vázquez de la Iglesia F, Cervera-Paz FJ, Manrique Rodríguez M. Surgery for Atresia auris. Retrospective study of our results and correlation with Jahrsdoerfer prognostic criterium. Acta Otorrinolaringol Esp. 2004 Aug-Sep;55(7):315-9. doi: 10.1016/s0001-6519(04)78529-5.

10. Seong J, Yang SK, Jang P, Lee SY, Carandang M, Choi BY. Clinical factors influencing the trial and purchase of bilateral microphones with contralateral routing of signal in patients with asymmetric sensorineural hearing loss. J Audiol Otol. 2020 Jan;24(1):29-34. doi: 10.7874/jao.2019.00409.

11. Hagr A. BAHA: bone-anchored hearing aid. Int J Health Sci (Qassim). 2007 Jul;1(2):265-76.

12. Baguley DM, Bird J, Humphriss RL, Prevost AT. The evidence base for the application of contralateral bone anchored hearing aids in acquired unilateral sensorineural hearing loss in adults. Clin Otolaryngol. 2006 Feb;31(1):6-14. doi: 10.1111/j.1749-4486.2006.01137.x.