



Case Series

Surgical treatment of external auditory canal cholesteatoma in congenital malformation of the ear: A case series

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ABSTRACT

Background: External auditory canal (EAC) cholesteatoma is a lesion lined with stratified squamous epithelium containing proliferative keratin with bony erosion in EAC which can spread to the tympanic cavity, mastoid, and surrounding organ. External cholesteatoma can occur in patients with congenital abnormalities such as congenital aural atresia (CAA).

Method: This case series was reported using the 2020 PROCESS Guideline. The design of this study used a retrospective study during the 2015–2020 period.

Result: 3 participants aged 10.67 ± 2.31 years with CAA had other complaints of ear infections. All participants experienced sensorineural hearing loss with an average threshold of 59.33 ± 36.68 dB and suspicious cholesteatoma from a CT scan. Canal wall down, meatoplasty, and/or canaloplasty were performed based on the findings.

Conclusion: Surgical procedure in CAA with cholesteatoma aimed on preventing further complications and recurrence.

1. Introduction

Congenital aural atresia (CAA) is a condition in which a patent external ear auditory canal (EAC) does not develop [1]. CAA is a malformation of the EAC and varies depending on the size of the EAC, stenosis when the diameter is less than 4 mm to complete atresia when there is no ear canal. CAA usually occurs in congenital malformations of microtia and occurs in 1 : 10,000–20,000 births [2,3]. Patients with CAA have a risk factor of up to 20% of external cholesteatoma [4]. This article will discuss several cases of external cholesteatoma that occur in this disorder. Congenital ear malformation either in the form of complete or incomplete CAA and microtia performed different management based on the clinical condition.

2. Method

The design of this study is a case series reported using the Preferred Reporting of Case Series in Surgery (PROCESS) 2020 Guideline [5]. Data collection was carried out in the period January 2015–December 2020 at Dr. Hasan Sadikin General Hospital. All participants were examined

by HRCT (high-resolution Computer Tomography) and pure tone audiometry. HRCT was used to identify CAA stages based on Weerda [6,7]. Participants underwent detailed examination of their ears for signs of microtia and cholesteatoma which grade of cholesteatoma was assessed based on Naim staging [8]. In addition, participants were also evaluated for signs of Bell's palsy using the House Brackman scale [9]. After a complete examination, the participants were prepared for several operations such as Canal Wall down, meatoplasty, and canaloplasty [10, 11].

3. Result

The average age of the participants was 10.67 ± 2.31 years with a median value of 12 (10–12) years. Most of the participants experienced CAA problems in the left ear, all of them were male. All participants experienced severe sensorineural hearing loss with an average participant hearing threshold of 59.33 ± 36.68 dB. The details of this case series are described in Table 1. Participant 1 has a peripheral facial nerve paralysis grade IV since birth (House Brackman grading system; Fig. 1). The cholesteatoma was found in the medial 1/3 of the EAC, and

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Table 1
Detailed data of Indonesia children with congenital aural atresia.

No	Age	Ear	HB	PTA	Audio	CSOM	Naim Stage	Microtia	CAA	Surgery	Note
1	8 years	Right	IV	81.5 dB	SNHL	✓	II	III	IV	Meatoplasty + Canaloplasty	stent installer
2	12 years	Left	-	70 dB	SNHL	✓	IVM	II	III	Canal Wall Down + Meatoplasty	-
3	12 years	Left	-	89.5 dB	SNHL	✓	IVM	I	I	Canal Wall Down + Meatoplasty	Mass Biopsy

Note: HB = House Brackman scale; PTA = pure tone audiometry; CAA = congenital aural atresia; SNHL = sensorineural hearing loss; CSOM = chronic suppurative otitis media.

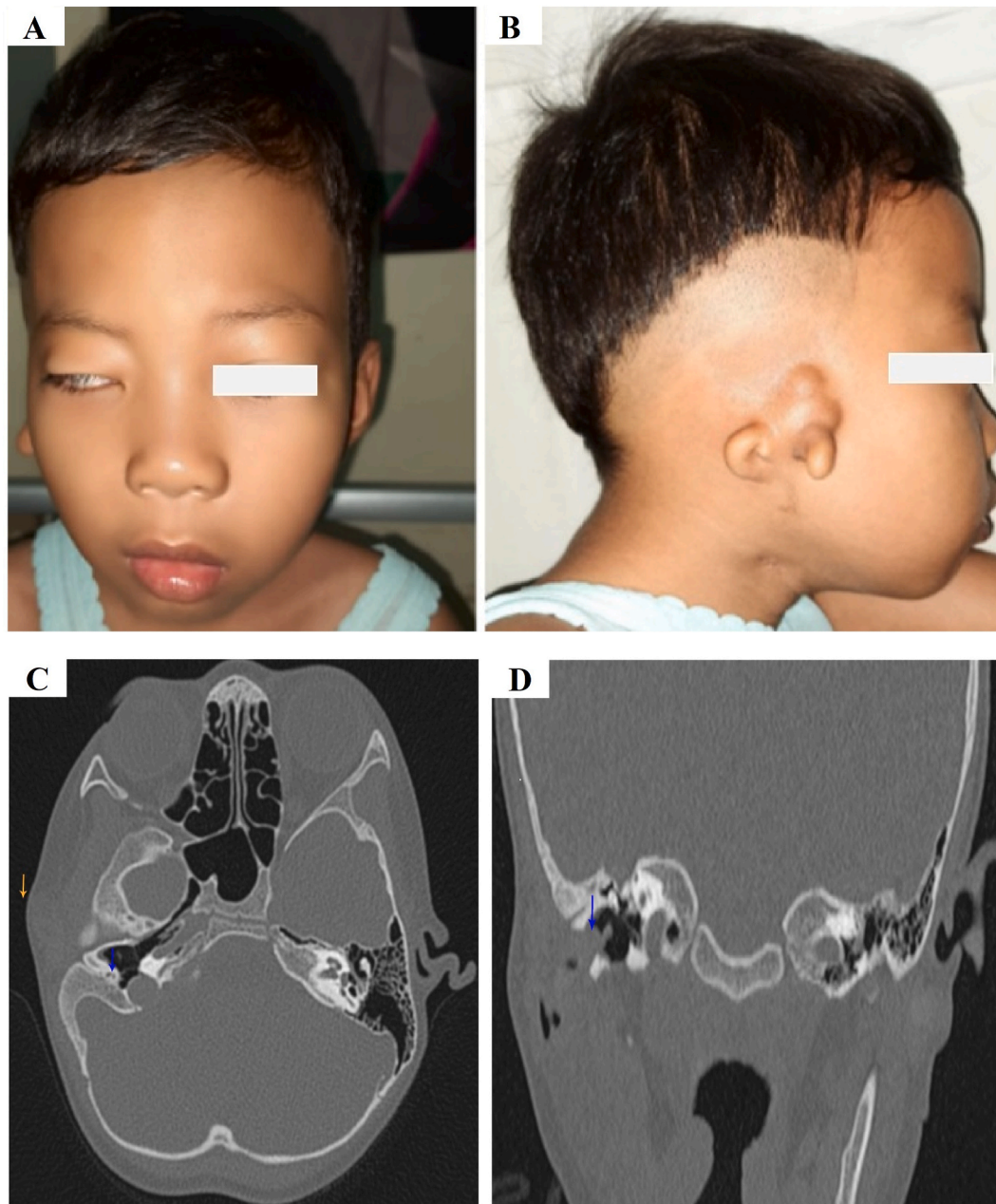


Fig. 1. A-B) Patient with right peripheral facial nerve paralysis House Brackman IV (left) and grade III microtia right; C) auricle of right side smaller than the left side (yellow arrow), facial nerve mastoid segment is more lateral than left, and there is no mastoid air cell (blue arrow); D) isodense lesion in EAC (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

the cholesteatoma was removed. On participant 2 from CT-scan found that the EAC narrowed in the left ear (Fig. 2). The patient underwent a canal wall down procedure to evacuate the cholesteatoma in the mastoid air cavity, as well as meatoplasty by removing the cartilage on the conchae. In participant 3 on examination of the left ear, it was found that

the auricle was smaller than the right ear and EAC atresia. There is a mass measuring 5x4x3 cm inferior to the left auricle that is mobile, soft, well-defined, and there was no tenderness (Fig. 3). CT scan showing an isodense lesion filling all over the left EAC and the left mastoid air cells. The patient underwent surgery in the form of canal wall down Auris

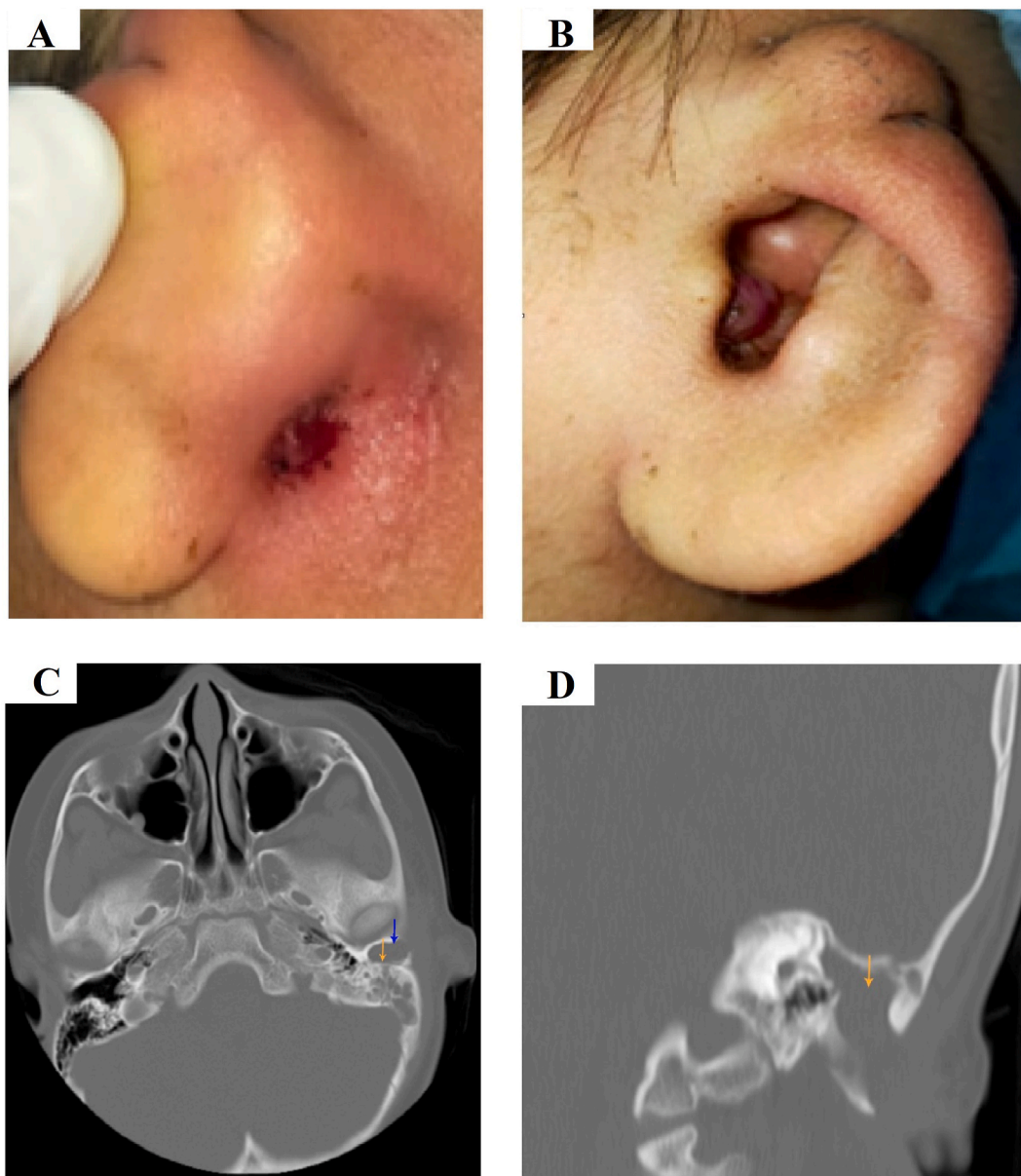


Fig. 2. A-B) Fistula retro auricula and granulation tissue over the EAC; C) isodense lesion filling EAC (blue arrow) and the posterior wall partially destroyed (yellow arrow); D) isodense lesion filling mastoid air cell. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Sinistra to evacuate the cholesteatoma, meatoplasty by removing cartilage on the concha, and extirpation and biopsy of the tumor. The result of the biopsy is an epidermal cyst.

4. Discussion

CAA occurs during malformations of ear formation. The external ear is formed from the first and second pharyngeal arches and the first pharyngeal cleft. In the fourth week, the first pharyngeal cleft will form an EAC. These structures will migrate to the endoderm pharyngeal pouch until it reaches the mesoderm. These migrating structures form three layers of the tympanic membrane. The auricle is formed at the beginning of the sixth week of pregnancy. Six hillock which is the six mesenchymal buds will be responsible for the formation of the auricle. The auricle concha will invaginate inward at the eighth week and form the fibrocartilage portion of the EAC. The canal will be fully formed at 28 weeks. CAA and microtia will form if there are problems with the

embryological process [2–4]. We used external cholesteatoma classification based on Naim et al, microtia classification based on Aguilar et al, and CAA based on Weerda et al [7,12].

In case 1, the patient was diagnosed with House Brackman IV facial nerve paralysis, chronic suppurative otitis media with II degree external cholesteatoma with III degrees microtia, and IV degree CAA. In grade IV CAA, there is complete atresia with decreased mastoid pneumatization and facial nerve abnormalities. From the physical examination, House Brackmann IV facial nerve paralysis was found since birth and a CT scan showed the facial nerve which is more lateral than normal and does not have mastoid air cells [13]. The diagnosis of grade 3 microtia was made because in this patient the ears are shaped like nuts, multiple ridges, deformed tissue containing some cartilage [7]. CT scan showed the presence of a grade 2 external cholesteatoma, which means there was a cholesteatoma in the EAC without any pars osseous destruction [12]. The treatment in case 1 was a retro auricular incision and anterior dissection to expose the EAC. Canaloplasty was performed on the

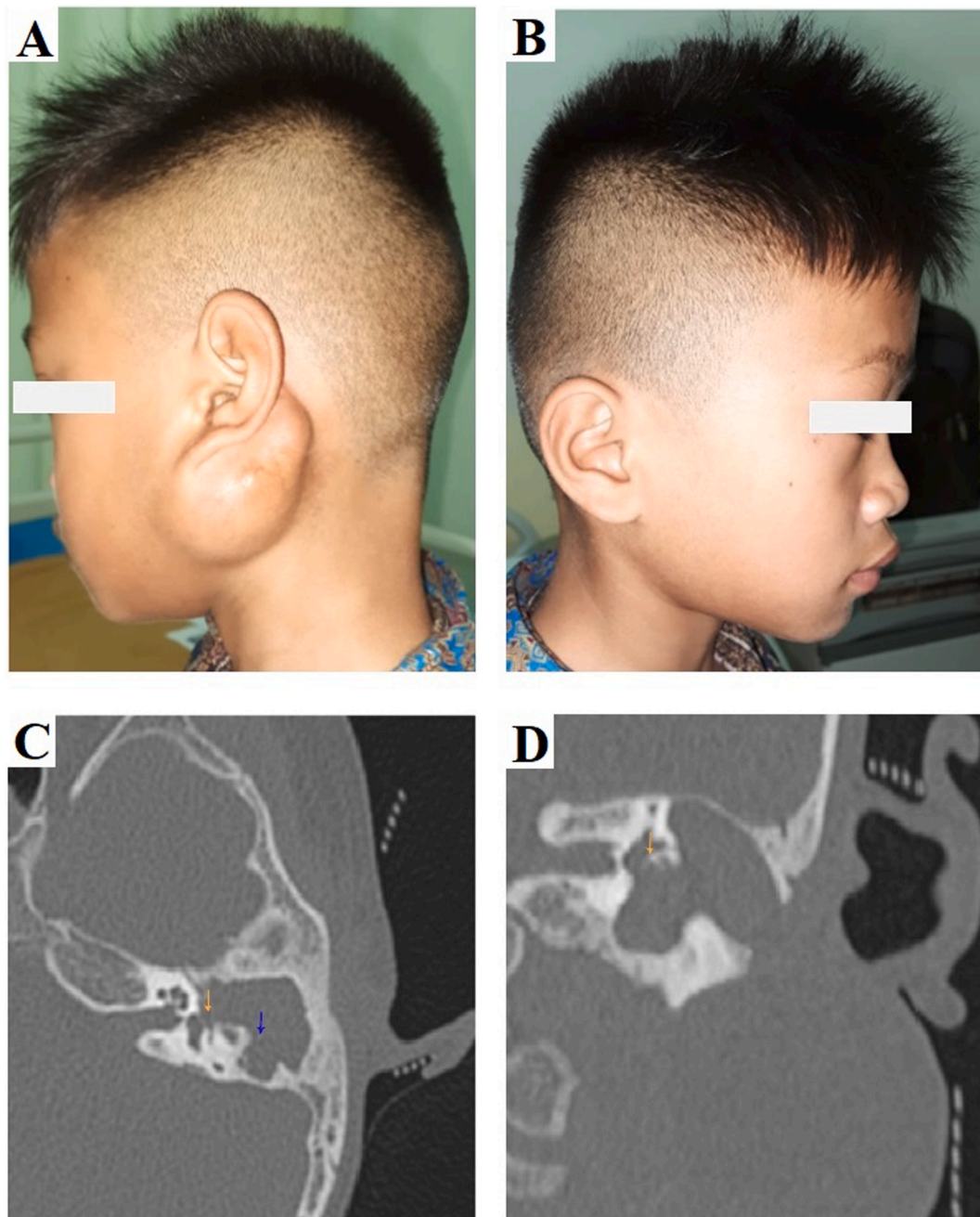


Fig. 3. A-B) There is a mass inferior to the auricle; C) isodense lesion filling mastoid air cell (blue arrow) and facial nerve dehiscence at tympanic and mastoid segments (yellow arrow); D) isodense lesion filling mastoid air cell and dehiscence of the lateral semicircular canal (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

posterior and superior pars osseous to get access to the anterior epitympanum with a target EAC diameter >10 mm [14–16]. Meatoplasty was performed to prevent recurrent stenosis [1,11]. Mastoidectomy was not performed because the cholesteatoma was only in the EAC [10,17].

In case 2, the patient was diagnosed with grade IVM external cholesteatoma and grade 2 microtia, and grade 3 CAA. In grade 3 CAA, complete atresia in EAC with pneumatization of the mastoid air cells is formed and there are no problems with the facial nerve. Microtia grade 2, the pinna is rudimentary and deformed but contains several recognizable components [7]. Grade IVM external cholesteatoma is a cholesteatoma in the EAC that has destroyed the surrounding anatomic structures of the mastoid air cells [4,12]. Canal wall down was performed because the posterior canal wall was partially destroyed and

cholesteatoma filling all over the mastoid air cell. The purpose is to unite the EAC, the tympanic cavity, the mastoid antrum, and prevent recurrence of cholesteatoma. Meatoplasty is performed with a V-shape incision from the concha to proximal EAC [18]. The cartilage of the concha is removed completely to widen the meatus and prevent recurrent stenosis [10,17].

In Case 3, the patient was diagnosed with grade IVM external cholesteatoma with grade I microtia and grade II CAA and left infra-auricular tumor. In grade I CAA, there is a narrowing of the fibrocartilage [13]. Grade I microtia have a smaller size in the pinna than the opposite ear [7]. Infra-auricular excision of the tumor was performed to prevent compression from the inferior. Canal wall down was performed with retro auricular access. The cholesteatoma and granulation tissue

were completely evacuated [18]. Meatoplasty is performed with a V-shape incision in the conchae to proximal to the EAC, with the removal of the conchae cartilage to widen the meatus [4].

5. Conclusion

Good results are obtained from maximum eradication of the focus infection. Cholesteatoma should not be left because it has a high chance of recurrence. Canaloplasty is intended to widen the canal by more than 10 mm. Meatoplasty is recommended by removing the cartilage on the conchae to prevent restenosis. Canal wall down was performed in external cholesteatoma grade IVM because has a lower recurrence rate of cholesteatoma than canal wall up.

Consent

We have explained the aim and benefits of our reporting to parents or guardians which they are willing to fill out the consent form consciously.

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Ethical approval

We have conducted an ethical approval base on the Declaration of Helsinki at the Health Research Ethics Committee in Dr. Hasan Sadikin General Hospital, Bandung, Indonesia.

Author contribution

All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

Guarantor

Lina Lasminingrum is the person in charge for the publication of our manuscript.

Declaration of competing interest

The authors declare that they have no conflict of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2021.102880>.

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References

- [1] S. Bouhabel, P. Arcand, I. Saliba, Congenital aural atresia: bone-anchored hearing aid vs. external auditory canal reconstruction, *Int. J. Pediatr. Otorhinolaryngol.* 76 (2) (2012) 272–277, <https://doi.org/10.1016/j.ijporl.2011.11.020>.
- [2] A. McFarren, S. Jayabose, M. Fevzi Ozkaynak, O. Tugal, C. Sandoval, Cleft palate, bilateral external auditory canal atresia, and other midline defects associated with Diamond-Blackfan anemia: case report, *J. Pediatr. Hematol./Oncol.* 29 (5) (2007) 338–340, <https://doi.org/10.1097/MPH.0b013e31805d8f45>.
- [3] G.J. Service, J.B. Roberson Jr., Current concepts in repair of aural atresia, *Curr. Opin. Otolaryngol. Head Neck Surg.* 18 (6) (2010) 536–538, <https://doi.org/10.1097/MOO.0b013e32833feb26>.
- [4] M.Y. Lee, Y.S. Cho, G.C. Han, J.H. Oh, Current treatments for congenital aural atresia, *J. Audiol. & Otol.* 24 (4) (2020) 161–166, <https://doi.org/10.7874/jao.2020.00325>.
- [5] R.A. Agha, C. Sohrabi, G. Mathew, T. Franchi, A. Kerwan, N. O'Neill, The PROCESS 2020 guideline: updating consensus preferred reporting of CasEseries in surgery (PROCESS) guidelines, *Int. J. Surg.* 84 (2020) 231–235, <https://doi.org/10.1016/j.ijsu.2020.11.005>.
- [6] S. Bartel-Friedrich, C. Wulke, Classification and diagnosis of ear malformations, *GMS Curr. Top. Otorhinolaryngol., Head Neck Surg.* 6 (2007) Doc05.
- [7] T.Y. Zhang, N. Bulstrode, K.W. Chang, Y.S. Cho, H. Frenzel, D. Jiang, et al., International consensus recommendations on microtia, aural atresia and functional ear reconstruction, *J. Int. Adv. Otol.* 15 (2) (2019) 204–208, <https://doi.org/10.5152/iao.2019.7383>.
- [8] R. Naim, F. Linthicum Jr., T. Shen, G. Bran, K. Hormann, Classification of the external auditory canal cholesteatoma, *Laryngoscope* 115 (3) (2005) 455–460, <https://doi.org/10.1097/01.mlg.0000157847.70907.42>.
- [9] H. Goldsztein, J.B. Roberson Jr., Anatomical facial nerve findings in 209 consecutive atresia cases, *Otolaryngol. –Head and Neck Surg. : Off. J. Am. Acad. Otolaryngol. Head Neck Surg.* 148 (4) (2013) 648–652, <https://doi.org/10.1177/0194599812473430>.
- [10] S. Mahdiani, L. Lasminingrum, B. Purwanto, R. Handayani, Canal wall down in tuberculosis matoiditis: 4 case series, *Int. J. Surg. Case Rep.* 83 (2021) 106029, <https://doi.org/10.1016/j.ijscr.2021.106029>.
- [11] F. Memari, M. Maleki Delarestaghi, P. Mir, M. GolMohammadi, E. Shams Koushki, Meatoplasty in canal wall down surgery: our experience and literature review, *Iran. J. Otorhinolaryngol.* 29 (90) (2017) 11–17.
- [12] K.Y. Ho, T.Y. Huang, S.M. Tsai, H.M. Wang, C.Y. Chien, N.C. Chang, Surgical treatment of external auditory canal cholesteatoma - ten years of clinical experience, *J. Int. Adv. Otol.* 13 (1) (2017) 9–13, <https://doi.org/10.5152/iao.2017.2342>.
- [13] C. Fuchsmann, S. Tringali, F. Disant, G. Buires, C. Dubreuil, P. Froehlich, et al., Hearing rehabilitation in congenital aural atresia using the bone-anchored hearing aid: audiological and satisfaction results, *Acta Otolaryngol.* 130 (12) (2010) 1343–1351, <https://doi.org/10.3109/00016489.2010.499879>.
- [14] B.A. Jennings, P. Prinsley, C. Philpott, G. Willis, M.F. Bhutta, The genetics of cholesteatoma. A systematic review using narrative synthesis, *Clin. Otolaryngol. : Off. J. ENT-UK ; Off. J. Netherlands Soc. Oto-Rhino-Laryngol. & Cervico-Facial Surg.* 43 (1) (2018) 55–67, <https://doi.org/10.1111/coa.12900>.
- [15] L.M.R. Moxham, N.K. Chadha, D.J. Courtemanche, Is there a role for computed tomography scanning in microtia with complete aural atresia to rule out cholesteatoma? *Int. J. Pediatr. Otorhinolaryngol.* 126 (2019) 109610, <https://doi.org/10.1016/j.ijporl.2019.109610>.
- [16] R. Gautam, J. Kumar, G.S. Pradhan, J.C. Passey, R. Meher, A. Mehndiratta, High-resolution computed tomography evaluation of congenital aural atresia - how useful is this? *J. Laryngol. Otol.* 134 (7) (2020) 610–622, <https://doi.org/10.1017/s002221512000136x>.
- [17] S. Mahdiani, L. Lasminingrum, D. Anugrah, Management evaluation of patients with chronic suppurative otitis media: a retrospective study, *Ann. Med. Surg.* 67 (2012) 102492, <https://doi.org/10.1016/j.amsu.2021.102492>, 2021.
- [18] A.M. Windsor, R. Ruiz, R.C. O'Reilly, Congenital soft tissue stenosis of the external auditory canal with canal cholesteatoma: case report and literature review, *Int. J. Pediatr. Otorhinolaryngol.* 134 (2020) 110053, <https://doi.org/10.1016/j.ijporl.2020.110053>.