Indonesian Journal of Medicine (2026), 11(02): 107-115
Masters Program in Public Health, Universitas Sebelas Maret

Case report
Open Access

When the Eustachian Tube Is Normal: Selective Epitympanic Dysventilation Syndrome in Primary Acquired Cholesteatoma – An Evidence-Based Case Report

Dewi Pratiwi[1)], Hadi Sudrajad[1)], Novi Primadewi[2)], Dienia Nop Ramliana[3)]

[1)]Sub Division Otology of Otorhinolaryngology Head and Neck Surgery Department, Medical Faculty Universitas Sebelas Maret, Dr Moewardi Hospital, Surakarta, Indonesia
[2)]Sub Division NeuroOtology of Otorhinolaryngology Head and Neck Surgery Department, Medical Faculty Universitas Sebelas Maret, Dr Moewardi Hospital, Surakarta, Indonesia
[3)]Medical Residency Program of Otorhinolaryngology Head and Neck Surgery Department, Medical Faculty Universitas Sebelas Maret, Dr Moewardi Hospital, Surakarta, Indonesia

Received: 04 February, 2026; Accepted: 12 March, 2026; Available online: 10 April, 2026

ABSTRACT

Background: Primary acquired cholesteatoma is a benign yet locally destructive middle ear lesion with an incompletely understood pathogenesis. While Eustachian tube dysfunction is commonly implicated, it does not explain cases with normal middle ear pressure. Selective epitympanic dysventilation syndrome (SEDS) has been proposed as an alternative mechanism involving localized ventilation impairment within the epitympanum. This study aims to provide a clearer understanding of the clinical features of cholesteatoma associated with SEDS, which may help clarify its etiopathogenesis, support earlier detection, limit disease progression, and guide more appropriate, individualized clinical decision-making.

Case presentation A 55-year-old female presented with one year of persistent, malodorous left otorrhea and progressive hearing loss over three months. Otoscopy revealed mucopurulent discharge with an attic perforation. Nasoendoscopy showed a patent Eustachian tube. Audiometry demonstrated mild conductive hearing loss (31.25 dB), and tympanometry showed a Type A curve, indicating normal middle ear pressure. Mastoid MSCT revealed opacification of mastoid air cells and antrum with intact ossicles and no mesotympanic involvement, suggesting a localized ventilation disorder consistent with SEDS.

Conclusion: SEDS is an important and underrecognized mechanism in primary acquired cholesteatoma. Its recognition is essential for early diagnosis and for guiding surgical strategies that restore epitympanic ventilation to prevent recurrence.

Keywords: Primary acquired cholesteatoma, selective epitympanic dysventilation syndrome, epitympanum, middle ear ventilation disorders, retraction pocket

Correspondence:
Dewi Pratiwi. Sub Division of Otology, Department of Otorhinolaryngology–Head and Neck Surgery, Faculty of Medicine, Universitas Sebelas Maret/Dr. Moewardi Hospital, Surakarta, Central Java, Indonesia. Email: pratiwidewi81@staff.uns.ac.id.

Cite this as:
Pratiwi D, Sudrajad H, Primadewi N, Ramliana DN (2026). When the Eustachian Tube Is Normal: Selective Epitympanic Dysventilation Syndrome in Primary Acquired Cholesteatoma – An Evidence-Based Case Report. Indones J Med. 11(02): 107-115. https://doi.org/10.26911/theijmed.2026.953.

BACKGROUND

Cholesteatoma is an indolent yet locally invasive keratinizing lesion of the middle ear that often requires definitive surgical intervention and long-term follow-up (Kennedy et al., 2024). Although typically unilateral, its progression may lead to significant morbidity, including progressive hearing loss, vertigo, and facial nerve dysfunction (Kennedy et al., 2024). Histopathologically, cholesteatoma is characterized by the accumulation of keratinizing squamous epithelium within the middle ear and mastoid cavity, with a well-recognized capacity for enzymatic and mechanical destruction of adjacent bony structures (NCBI MedGen, 2026). Despite advances in surgical techniques, complications and recurrence remain clinically relevant, underscoring the importance of understanding its underlying pathophysiology (Bal & Deshmukh, 2022).

The etiopathogenesis of cholesteatoma remains incompletely understood and is widely considered multifactorial (Kennedy et al., 2024). Traditional theories emphasize eustachian tube dysfunction, epithelial migration, and squamous metaplasia. However, no single mechanism sufficiently explains the wide spectrum of clinical and morphological presentations (Bölcsföldi et al., 2022). In particular, these conventional models fail to account for cases in which middle ear pressure and Eustachian tube function appear normal, suggesting that alternative mechanisms may be involved.

Historically, the attic retraction pocket theory has been the dominant paradigm, attributing disease development to chronic negative pressure within the pars flaccida, often secondary to obstruction at the tympanic isthmus. This process leads to progressive invagination of the tympanic membrane and subsequent accumulation of keratin debris (Casale et al., 2023). More recently, attention has shifted toward selective epitympanic dysventilation syndrome (SEDS), which proposes a localized ventilation defect confined to the epitympanum despite preserved global middle ear aeration (Casale et al., 2023; Poe & Kivekas, 2016). This concept introduces a clinical paradox in which significant attic pathology, including cholesteatoma, may develop in the presence of a normal tympanogram, highlighting the role of compartmentalized middle ear ventilation.

This case report aims to illustrate the role of SEDS as a potential etiopathological mechanism in primary acquired cholesteatoma, emphasizing its clinical implications for early diagnosis and targeted surgical management.

CASE PRESENTATION

A 55-year-old woman presented with a one-year history of persistent, foul-smelling yellowish discharge from the left ear, accompanied by progressive hearing loss over the preceding three months. She denied otalgia, tinnitus, vertigo, facial asymmetry, or systemic symptoms. There was no history of recurrent upper respiratory infections, ear trauma, or habitual ear manipulation.

Otolaryngological examination showed a normal right ear. The left ear showed mucopurulent discharge and an attic perforation involving the pars flaccida, consistent with TOS grade IV/Sade grade I retraction. Nasal and oropharyngeal examinations were unremarkable, and no cervical lymphadenopathy was noted.

Tuning fork tests demonstrated Weber lateralization to the left and a negative Rinne test, suggesting conductive hearing loss. Pure-tone audiometry confirmed mild conductive hearing loss in the left ear (31.25 dB), with normal hearing in the right ear. Notably, tympanometry revealed a Type A curve bilaterally, indicating normal

middle ear pressure. Eustachian tube function testing was also normal, presenting a clinical paradox in the context of suspected cholesteatoma. Facial nerve function was intact (House–Brackmann grade I).

Microbiological examination identified *Staphylococcus aureus* sensitive to ciprofloxacin. Mastoid multislice computed tomography (MSCT) demonstrated a soft-tissue density lesion occupying the epitympanum, Prussak's space, mastoid antrum, and mastoid cavity, with intact ossicular structures and no mesotympanic involvement. These findings suggested primary acquired cholesteatoma associated with localized ventilation impairment consistent with selective epitympanic dysventilation syndrome (SEDS).

The patient underwent canal wall up mastoidectomy with atticotomy, canaloplasty, and type I tympanoplasty. Intraoperatively, a pars flaccida defect with scutum erosion and cholesteatoma matrix was identified, while the incus and stapes remained intact. Endoscopic ear surgery was incorporated to evaluate and restore ventilation pathways, including clearance of the tympanic isthmus and tensor fold.

Postoperatively, the patient showed an uneventful recovery. Histopathological examination confirmed cholesteatoma. At 3-month follow-up, the patient remained symptom-free, with an intact tympanic membrane and no evidence of recurrence. Hearing levels remained stable, consistent with mild conductive hearing loss.

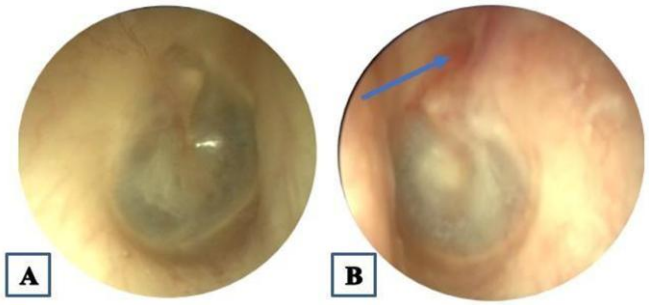

Figure 1. Otoscopic findings. (A) Right ear showing a clear external auditory canal, no discharge, intact tympanic membrane, and preserved light reflex. (B) Left ear showing mucopurulent discharge with attic (pars flaccida) perforation

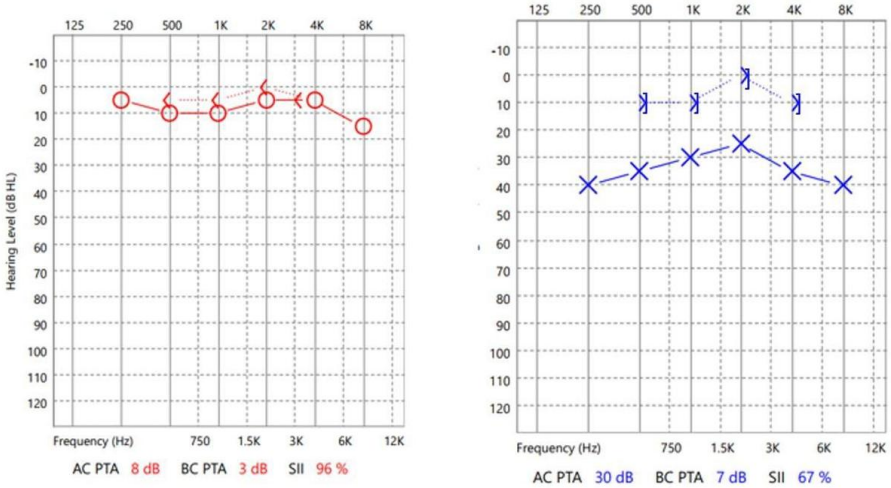

Figure 2. Pure tone audiometry. Left (AD) normal hearing 7.5 dB. Right (AS) mild conductive hearing loss 31.25 dB.

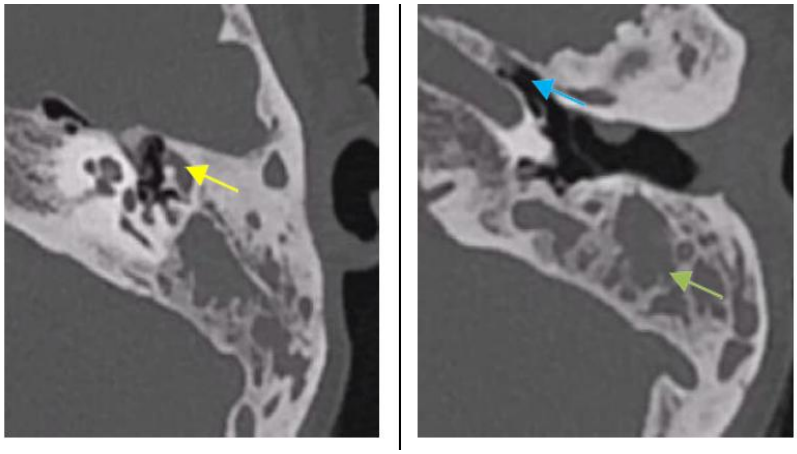

Figure 3. Mastoid MSCT without contrast shows the image of filling the epitympanum and prusack space (yellow), the mastoid cavity (green), Eustachian tube (blue).

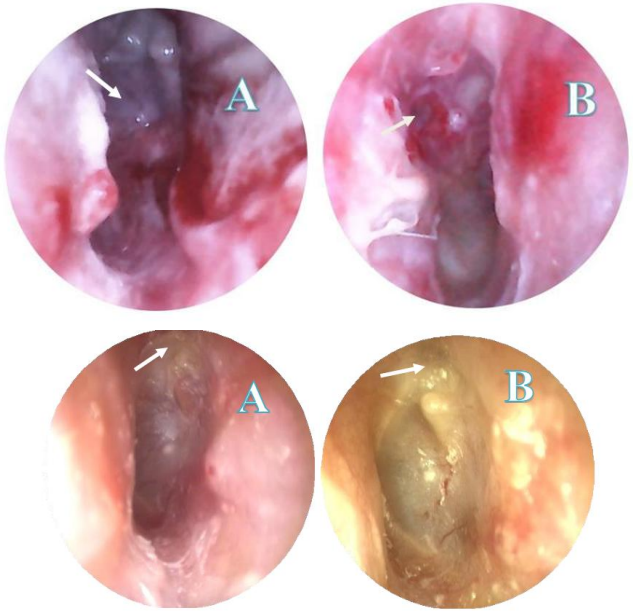

Figure 4. Post-operative otoscopy follow-up. (A) Day 14 (B) Day 31 (C) Day 43 (D) Day 62

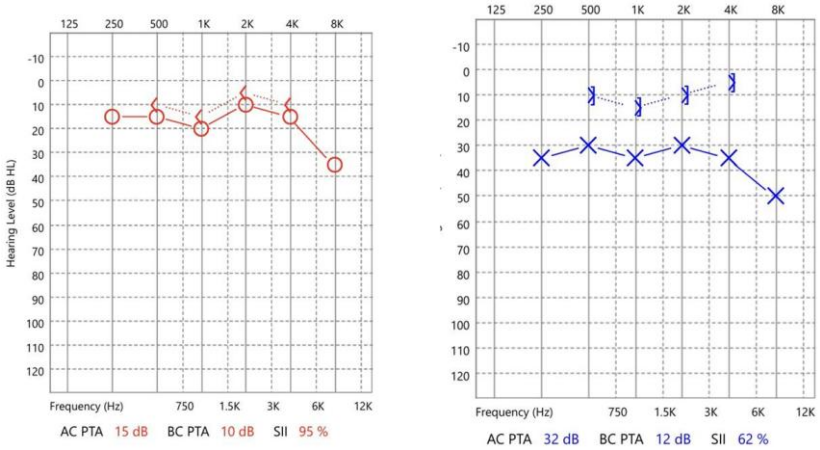

Figure 5. D+90 post operative PTA. (Left) AD normal hearing 10 dB. (Right) AS mild conductive hearing loss 32.5 dB.

DISCUSSION

The middle ear is a complex, compartmentalized system consisting of the epitympanum, mesotympanum, hypotympanum, protympanum, and retrotympanum (Isaacson, 2018; Mansour et al., 2013). The epitympanum is separated from the mesotympanum by the tympanic diaphragm, creating a semi-independent space that relies on narrow ventilation pathways, particularly the tympanic isthmus and the pouches of von Tröltsch, for pressure equilibration (Szysmanski & Geiger, 2023; Milenkovic et al., 2020). Normal middle ear aeration depends on the patency of these micro-ventilation pathways. The tympanic isthmus serves as a critical conduit between the epitympanum and mesotympanum, while mucosal folds such as the tensor tympani fold and malleal folds regulate airflow (Vasudev et al., 2022).

Obstruction or functional narrowing of these structures may isolate the epitympanum from the rest of the middle ear, resulting in localized dysventilation. This condition promotes the development of negative pressure within the attic, leading to pars flaccida retraction and retraction pocket formation (Pachpande & Singh, 2022; Matsuzawa et al., 2016).

This pathophysiological mechanism underlies selective epitympanic dysventilation syndrome (SEDS), which challenges the traditional concept that cholesteatoma primarily results from global Eustachian tube dysfunction (Gaillard et al., 2025). In SEDS, localized ventilation impairment occurs despite preserved overall middle ear pressure, explaining the clinical paradox observed in this case—namely, the presence of attic cholesteatoma in a patient with a Type A tympanogram and normal Eustachian tube function (Marchioni et al., 2013).

Expanding on this concept, a systematic review by Swain (2024), encompassing 41 studies on non-SEDS attic retraction pockets (ARP), demonstrated that SEDS and ARP are closely interrelated. In this framework, ARP is considered a manifestation of SEDS arising from obstruction of the tympanic isthmus, which leads to ventilation isolation and negative pressure within the epitympanum. This process induces pars flaccida retraction and promotes cholesteatoma formation.

The review further identified key susceptibility factors, including poorly pneumatized mastoids and impaired tympanic isthmus ventilation, both of which increase the risk of disease development. Importantly, SEDS provides a unifying explanation for the origin of primary cholesteatoma developing in the absence of prior infection or tympanic membrane perforation. In terms of disease severity, deeper ARP and increased keratin debris accumulation were associated with a higher likelihood of complications, including ossicular erosion, semicircular canal fistula, and intracranial abscess.

From a management perspective, established cholesteatoma in this context often necessitates canal wall down mastoidectomy combined with attic wall reconstruction and ossicular chain reconstruction. Unlike non-SEDS mechanisms, which typically involve diffuse middle ear dysfunction and are more frequently associated with pars tensa retraction, SEDS-related pathology is localized and often progresses silently (Rosito et al., 2018). The pars flaccida is particularly vulnerable to focal negative pressure generated by tympanic isthmus obstruction (Hamed et al., 2016). Supporting evidence suggests that reduced epitympanic volume and narrowed isthmus dimensions predispose individuals to retraction pocket formation and

cholesteatoma development, highlighting the critical role of anatomical configuration and micro-ventilatory dynamics (Hacking et al., 2024; Vasu et al., 2023).

This concept is reinforced by the retrospective cohort study conducted by Rosito et al. (2018) involving 432 patients with middle ear cholesteatoma. Their findings demonstrated a sequential pathogenic progression in which SEDS leads to pars flaccida retraction, followed by the formation of ARP, and eventual progression to posterior epitympanic cholesteatoma (PEC). Both histopathological and clinical data from this study indicate that tympanic membrane retraction, particularly involving the pars flaccida and pars tensa, represents an early and significant determinant of both susceptibility and disease severity.

Further supporting this mechanism, Hamed et al. (2016), in a comprehensive literature review, highlighted that certain cholesteatoma cases develop in the absence of a prior history of otitis media or tympanic membrane perforation. Within this context, SEDS induces localized negative pressure in the epitympanum, promoting retraction pocket formation and progressive keratin accumulation, ultimately leading to cholesteatoma development.

Additionally, anatomical features of the epitympanum, such as tensor folds and the presence of granulation tissue, may predispose individuals to obstruction and increased disease risk. Interestingly, preserved Eustachian tube function in these patients may contribute to delayed diagnosis, thereby allowing disease progression to occur silently. The authors also emphasized that while current management strategies primarily focus on surgical intervention targeting epitympanic obstruction, future research is expected to explore non-surgical therapeutic approaches aimed at modulating osteoclastogenesis, inflamemation, and cellular proliferation.

The present case demonstrates these principles clearly, with imaging and intraoperative findings revealing disease predominantly confined to the epitympanum despite preserved overall middle ear pressure. This observation supports the concept that localized dysventilation, rather than global Eustachian tube dysfunction, can act as the primary driver of disease. In line with this, Marchioni et al. (2013), in a retrospective cohort study of patients with attic retraction pockets or epitympanic cholesteatoma and normal Eustachian tube function, reported that such patients exhibit increased susceptibility to epitympanic cholesteatoma due to functional isolation of the epitympanum from mesotympanic ventilation. In this framework, ARP in non-SEDS conditions may represent the final morphological manifestation of an underlying selective dysventilation process, reflecting the progressive sequence from SEDS to ARP and ultimately to cholesteatoma formation.

Recognition of SEDS carries important therapeutic implications. While conventional management emphasizes complete eradication of cholesteatoma through mastoidectomy, increasing attention is being directed toward restoring physiological ventilation of the epitympanum. The integration of endoscopic ear surgery techniques allows for direct visualization and clearance of obstructed micro-ventilation pathways, including the tympanic isthmus and tensor tympani fold, thereby addressing the underlying valve-like mechanism responsible for selective dysventilation. By targeting the root cause rather than solely the disease manifestation, this approach has the potential to reduce recurrence rates and improve long-term functional outcomes.

In summary, SEDS represents a paradigm shift from the traditional global pressure-based model toward a localized ventilation disorder in the pathogenesis of primary acquired cholesteatoma. The accumulated evidence from systematic reviews, cohort studies, and clinical observations consistently supports the role of epitympanic ventilation failure as a key initiating factor. Awareness of this entity is essential, particularly in patients presenting with normal tympanometric findings, as early recognition and targeted intervention addressing epitympanic obstruction may prevent disease progression and reduce the risk of serious complications.

.

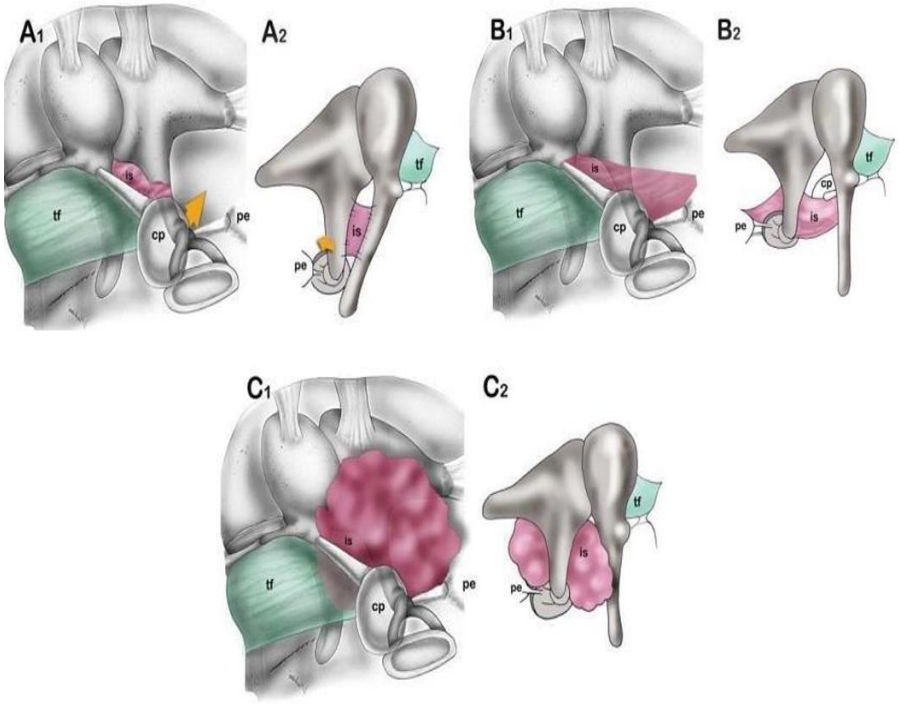

Figure 7. Anatomical findings in SEDS

Fig. 7 illustrates several patterns of obstruction at the isthmus region of the middle ear. **(A)** Partial isthmus obstruction is characterized by a narrow gap between the pyramidal eminence and the incudostapedial joint, as seen in both medial (A1) and lateral (A2) views. (B) Total isthmus obstruction results from mucosal folds that are anchored posteriorly to the pyramidal eminence and extend anteriorly into the space between the incudostapedial joint and the cochleariform process, as demonstrated in medial (B1) and lateral (B2) views. (C) Isthmus occlusion may also occur due to granulation tissue, which completely fills and obstructs the space, as observed in medial (C1) and lateral (C2) perspectives. The anatomical structures involved include the tensor fold (tf), isthmus (is), pyramidal eminence (pe), and cochleariform process (cp).

Selective epitympanic dysventilation syndrome (SEDS) represents a distinct etiopathological mechanism in primary acquired cholesteatoma, characterized by localized ventilation impairment within the epitympanum despite preserved Eustachian tube function. This condition may lead to insidious disease progression, often presenting as a clinical paradox with normal tympanometric findings.

Recognition of SEDS is essential for improving diagnostic accuracy, particularly in patients with attic retraction or cholesteatoma in the presence of normal middle ear pressure. Unlike non-SEDS mechanisms, which are typically associated with diffuse middle ear dysfunction, SEDS involves a localized process driven by obstruction at the tympanic isthmus and epitympanic compartmentalization.

From a therapeutic perspective, management should not only focus on cholesteatoma removal but also on restoring physiological ventilation of the epitympanum. Surgical strategies that include clearance of the tympanic isthmus and correction of obstructive mucosal folds may help reduce recurrence and improve long-term outcomes. Early recognition and targeted intervention are therefore crucial in preventing disease progression and complications.

AUTHORS CONTRIBUTION

Conceptualization and study design were led by Dewi Pratiwi, with significant clinical input and conceptual guidance from Hadi Sudrajad and Novi Primadewi. Data collection, including the retrospective clinical investigation and the structured literature search across multiple databases, was primarily performed by Dienia Nop Ramliana and Dewi Pratiwi. Formal analysis of the literature and clinical findings was conducted by Dienia Nop Ramliana under the supervision of Dewi Pratiwi. The original manuscript was drafted by Dienia Nop Ramliana, while Dewi Pratiwi, Hadi Sudrajad, and Novi Primadewi were responsible for the critical revision and final approval of the version to be published.

FINANCIAL SUPPORT AND SPONSORSHIP

This study received no external funding and none sponsorship.

ACKNOWLEDGEMENT

The authors would like to thank the Department of Otorhinolaryngology–Head and Neck Surgery, Faculty of Medicine, Universitas Sebelas Maret, and Dr. Moewardi Hospital, Surakarta, for their support and facilitation of this study. Appreciation is also extended to all clinical staff involved in patient care.

CONFLICT OF INTEREST

The authors declare no conflicts of interest regarding the publication of this research.

REFERENCE

Bal R, Deshmukh P (2022). Management of eustachian tube dysfunction: a review. Cureus. 14(11): e31432. doi:10.7759/-cureus.31432.

Bölcsföldi TB, Pap I, Tóth T, Tóth I, Révész P, Gerlinger I, Szanyi I (2022). A szelektív epitympanalis dysventilatiós szindróma műtéti megoldása [Surgical solution of selective epitympanic dysventilation syndrome]. Orv Hetil. 163(36): 1440–1446. doi:10.1556/-650.2022.32563.

Casale J, Shumway KR, Hatcher JD (2023). Physiology, eustachian tube function. In: StatPearls. Treasure Island (FL): StatPearls Publishing.

Gaillard F, Gajera J, Campos A (2026). Acquired cholesteatoma. Radiopaedia.org. doi:10.53347/rID-841.

Hacking C (2026). Pharynx. Radiopaedia.org. doi:10.53347/rID-53617.

Hamed MA, Nakata S, Sayed RH, Ueda H, Badawy BS, Nishimura Y, et al. (2016). Pathogenesis and bone resorption in acquired cholesteatoma:

current knowledge and future perspectives. Clin Exp Otorhinolarynxgol. 9(4): 298–308. doi:10.21053/ceo.2015.01662.

Isaacson B (2018). Anatomy and surgical approach of the ear and temporal bone. Head Neck Pathol. 12(3): 321–327. doi:10.1007/s12105-018-0926-2.

Kennedy KL, Singh AK (2024). Middle ear cholesteatoma. In: StatPearls. Treasure Island (FL): StatPearls Publishing.

Mansour S, Magnan J, Haidar H, Nicolas K, Louryan S (2013). Comprehensive and clinical anatomy of the middle ear. Berlin: Springer. doi:10.1007/978-3-642-36967-4.

Marchioni D, Grammatica A, Alicandri-Ciufelli M, Aggazzotti-Cavazza E, Genovese E, Presutti L (2012). The contribution of selective dysventilation to attical middle ear pathology. Med Hypotheses. 78(1): 116–120. doi:10.1016/j.mehy.2011.11.011.

Matsuzawa S, Iino Y, Yamamoto D, Hasegawa M, Hara M, Shinnabe A, et al (2017). Attic cholesteatoma with closure of the entrance to pars flaccida retraction pocket. Auris Nasus Larynx. 44(6): 766–770. doi:10.1016/j.anl.2016.11.014.

Milenkovic I, Schiefer U, Ebenhoch R, Ungewiss J (2020). Aufbau und funktion der hörbahn [Anatomy and physiology of the auditory pathway]. Ophthalmologe. 117(11): 1068–1073. doi:10.1007/s00347-020-01070-0.

National Center for Biotechnology Information (2023). Cholesteatoma (Concept ID: C0008373). MedGen. Bethesda (MD): National Library of Medicine.

Pachpande TG, Singh CV (2022). Diagnosis and treatment modalities of cholesteatoma: a review. Cureus. 14(11): e31153. doi:10.7759/cureus.31153.

Poe DS, Kivekäs I (2016). Anatomy and physiology of the eustachian tube. In: Lalwani AK, Sataloff RT, editors. Sataloff's comprehensive textbook of otorhinolaryngology: head and neck surgery. Vol. 1. New Delhi: Jaypee Brothers Medical Publishers. doi:10.5005/jp/books/12689_7.

Rosito LPS, Sperling NM, Teixeira AR, Selaimen FA, da Costa SS (2018). The role of tympanic membrane retractions in cholesteatoma pathogenesis. Biomed Res Int. 2018: 9817123. doi:10.1155/2018/9817123.

Swain SK (2024). Attic retraction pocket: a review. Int J Otorhinolaryngol Head Neck Surg. 10(5): 606–611. doi:10.18203/issn.2454-5929.ijohns202427-25.

Szymanski A, Geiger Z (2023). Anatomy, head and neck, ears. In: StatPearls. Treasure Island (FL): StatPearls Publishing.

Vasu PK, Nair S, Aishwarya JG, Sameema VV, Geethu KP (2023). Cog: a radiological indicator in identifying dysventilation syndrome. Indian J Otolaryngol Head Neck Surg. 75(1): 419–423. doi:10.1007/s12070-022-03308-4.

Vasudev V, Prasad KC, Babu AP, Karunasagar A, Varsha GI, Koneru P (2022). Intricacies of the epitympanum—endoscopically revisited. Indian J Otolaryngol Head Neck Surg. 74(Suppl 1): S563–S568. doi:10.1007/s12070-021-02389-z.