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Clinico-Investigational Profile and Surgical Outcomes of Pediatric Cholesteatoma: A Case Series

Paramita Saha[1], Irine A. Thomas[2], Rahul Kunkulol[3]

[1]Department of Otolaryngology, Bhagirathi Neotia Woman and Child Care Centre, Kolkata, India
[2]Department of Otorhinolaryngology, Al Azhar Medical College and Superspeciality Hospital, Thodupuzha, India
[3]Pravara Institute of Medical Sciences and Rural Medical College, Loni, India
Email: paromita2287@gmail.com, thomasirine@yahoo.in, drrahulkunkulol4@gmail.com

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Abstract

Pediatric cholesteatoma causes extensive destruction within the middle ear and adjacent structures. Despite advancements in surgical techniques, the management of pediatric cholesteatoma remains controversial due to the varied presentations and its rapid progression. This study aims to evaluate the presentation of symptoms, otoscopic findings, surgical outcomes, and functional results in a series of pediatric cholesteatoma patients at our institution. A total of 18 patients, aged 5 - 14 years, underwent canal wall down (CWD) or intact canal wall (ICW) surgery based on the extent of disease as assessed intraoperatively. The study focused on the sites involved by cholesteatoma, the surgical challenges encountered, and the techniques employed. Outcomes measured included the incidence of residual and recurrent cholesteatoma, as well as hearing function at follow-up. Our results indicated a 69% improvement in the air-bone gap (ABG) among patients, with residual perforation observed in 4% of cases and no recurrences during the follow-up period. The increased selection of the CWD technique correlated with the extensive nature of the disease presentation. This study underscores the necessity for individualized treatment plans in pediatric cholesteatoma management, considering the aggressive nature of the disease and the need for a balance between eradication and hearing preservation.

Keywords

Pediatric, Cholesteatoma, Children, Canal Wall Down

1. Introduction

Pediatric cholesteatoma is a rare but serious condition, with an estimated incidence

of 3 - 15 cases per 100,000 children [1]. The disease is characterized by the presence of keratinizing squamous epithelium within the middle ear, leading to chronic inflammation and bone erosion. Unlike adult cholesteatoma, pediatric cholesteatoma is often more aggressive, likely due to differences in immune response and anatomical development [2]. The rapid proliferation and extensive nature of cholesteatoma in children necessitate early diagnosis and a strategic surgical approach to prevent complications, such as hearing loss, facial nerve paralysis, and intracranial spread [3].

Surgical management of cholesteatoma aims to achieve a safe, dry, and hearing ear. The two primary surgical techniques employed are the canal wall down (CWD) and the intact canal wall (ICW) procedures. CWD surgery involves removing the posterior bony ear canal wall, which facilitates complete disease clearance and reduces recurrence rates but may result in a large, open mastoid cavity requiring ongoing [4]. ICW surgery preserves the bony ear canal wall, leading to better cosmetic and hearing outcomes but carries a higher risk of residual disease and requires more meticulous follow-up [5].

Recent advances in surgical techniques, such as mastoid cavity obliteration, have attempted to mitigate the disadvantages of both approaches. Obliteration using autologous materials like bone dust, cartilage, and muscle flaps has been shown to reduce cavity problems and enhance self-cleansing, potentially improving long-term outcomes in CWD procedures [6]. Despite these innovations, the choice of surgical technique remains heavily influenced by the extent of disease, patient age, and surgeon experience [7].

This study presents an analysis of 18 pediatric patients who underwent surgery for cholesteatoma at our institution. We assess the clinical presentation, intraoperative findings, surgical techniques employed, and postoperative outcomes to contribute to the ongoing discussion about the optimal management of this challenging condition.

Figure 1 and **Figure 2** illustrate the HRCT temporal bone (axial view) findings of two cases (out of 18) with a soft tissue density mass in the middle ear. The first case shows erosion of the Fallopian canal on the right side, while the second case demonstrates erosion of the ossicles on the left side.

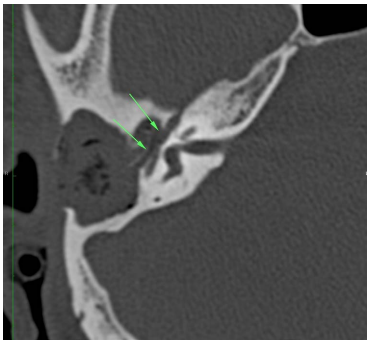

Figure 1. HRCT temporal bone Axial view (right): soft tissue density eroding the Fallopian canal.

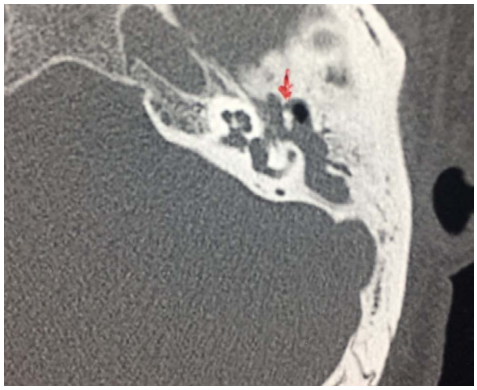

Figure 2. HRCT temporal Bone Axial View (left side): Soft tissue density mass eroding ossicles with remnant incus and malleus.

Figures 3-5 depict the preoperative otoscopic view of the tympanic membrane showing cholesteatoma.

Figure 3. Case 1—Left sided cholesteatoma with grade 4 Pars tensa retraction and granulations.

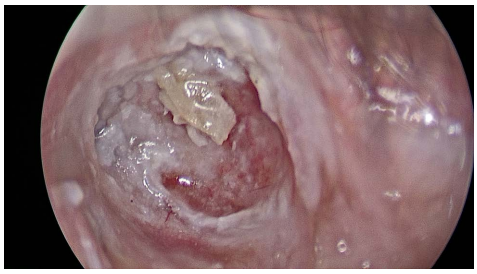

Figure 4. Case 2—Left sided Pars tensa retraction grade 4 with Cholesteatoma.

Figure 5. Case 3—Left sided Posterior superior cholesteatoma with polyp.

Figure 6 is an intraoperative image showing the CWD (Canal Wall Down) procedure, performed to remove cholesteatoma from hidden areas.

Figure 7 shows the temporalis fascia graft covering the entire mastoid cavity after the CWD procedure.

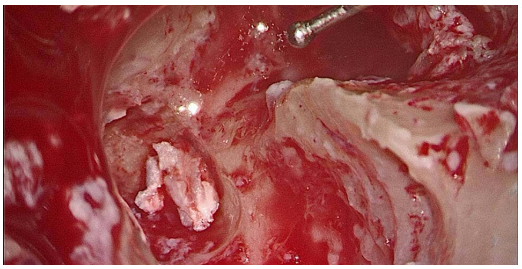

Figure 6. Canal wall down: to remove cholesteatoma from hidden areas.

Figure 7. Canal wall down with Temporalis Fascia graft placed.

2. Methodology

A total of 18 pediatric patients, aged 5 to 14 years, who underwent surgical intervention for middle ear cholesteatoma at our institution, were retrospectively analyzed. Patients were evaluated as per the age of presentation, symptomatology, clinical findings, extension of disease, surgical treatment, state of the ossicular chain, hearing results and recurrence of the disease. The patients underwent surgery for middle ear cholesteatoma at our Institution under general anesthesia.

In our analysis, we considered the surgical technique employed (ICW vs. CWD) based on HRCT grading (based on the sites involved) and after correlation with the intra-operative findings, ossicular chain involvement and the technique used for ossiculoplasty. In case of residual and recurrent cholesteatoma we have always employed a CWD technique.

3. Results

Eighteen children, aged between 5 and 14 years were treated and followed up for duration of 12 - 18 months out of which twelve patients were male and six females. While majority of patients presented to us with symptoms of foul smelling otorrhea (94%) and hearing loss (85%), two patients came with acute mastoiditis

(14%), one patient with post aural fistula (7%) and one with symptoms of URTI. Otoscopy, microscopic examination revealed the following:

Table 1. Otoscopy and microscopic findings.

Otoscopy and microscopic findings	No. of patients	Percentage
Pars Tensa Retraction	12	66%
Attic perforation/retraction	3	16%
Both (Attic + Pars tensa)	4	22%
Polyp	4	22%
Keratin flakes	16	88%

Among the patients examined, Table 1 shows that pars tensa retraction was observed in 12 cases, constituting 66% of the total, with a mean retraction depth of 7.8 mm and a standard deviation of 6.06 mm. Attic perforation or retraction was present in 3 patients, representing 16% of the cases. Both attic and pars tensa retractions were noted in 4 patients, accounting for 22%. Additionally, polyps were found in 4 patients (22%), and keratin flakes were observed in 16 patients (88%). The mean retraction depth and standard deviation for the presence of keratin flakes were 7.8 ± 6.06.

Table 2. The HRCT temporal bone grading noted for the disease extension.

Ct scan findings according to grades	%
I Disease in attic, aditus, antrum	80
II Erosion of one bony wall of antrum or disruption of the ossicular chain	100
III Erosion of two of the bony walls of antrum with ossicular chain disruption	80
IV dehiscence of facial canal	20
Intracranial extension	0
LSCC fistula	0

Table 2 illustrates the extent of the disease as observed on HRCT in pediatric cholesteatoma patients. Most patients had disease limited to Grade I (80%). Significant bony wall erosion was noted in Grades II and III, affecting the ossicular chain in 100% and 80% of cases, respectively. This underscores the importance of preoperative CT in grading the severity and guiding surgical planning.

Table 3. Intra-operative findings.

Keratin flakes	18
Ossicles engulfed by the granulation tissue	01
Incus erosion + IS joint affection	13
Stapes Suprastructure erosion	05
Facial canal dehiscence	04
LSCC dehiscence	Nil
Intracranial involvement	Nil

Table 3 shows that during surgery, keratin flakes were identified in 18 patients, with a mean count of 8.2 flakes and a standard deviation of 6.88. Ossicles engulfed by granulation were noted in 1 patient. Incus erosion, with involvement of the incudo-stapedial joint, was observed in 13 cases. Stapes suprastructure erosion occurred in 5 patients, and facial canal dehiscence was found in 4 cases. No patients exhibited LSCC dehiscence or intracranial involvement. The overall mean of findings during surgery was 8.2 ± 6.88.

The CWD technique was used as first choice in 16 patients who had CT grading of II, III and IV of which cavity obliteration was done in eight. Two patients with CT grading I underwent ICW procedure.

Table 4. Ossicular reconstruction materials.

Ossicular reconstruction materials	
Autologous ossicles	12
PORP(Teflon)	1
Cartilage strut	5

Table 4 shows the results of ossicular reconstruction. Autologous ossicles were used in 12 patients, with a mean of 6 and a standard deviation of 5.57. PORP (Teflon) was used in 1 case, and cartilage struts were employed in 5 patients. The overall mean usage of ossicular reconstruction materials was 6 ± 5.57.

Follow-up examinations were carried out at 6, 12, and 18 months using otomicroscopy. The post-operative results, tabulated below, showed no recurrence or residual disease. No major complications were noted post-operatively, except for residual perforation in a few cases. Ossiculoplasty was performed as part of a single-stage surgery using the following technique.

Table 5. Post operative findings.

	No. of patients	Percentage
Improved Hearing with ABG of 30 dB	13	69%
Grommet Inserted (severe ET dysfunction)	1	2%
Residual perforation	3	4%

Table 5 and Figure 8 show that post-operatively, hearing improvement—with an air-bone gap (ABG) of 30 dB or less—was achieved in 13 patients, accounting for 69% of the cases. The mean ABG improvement was 5.67 dB, with a standard deviation of 6.66 dB. Grommet insertion due to severe Eustachian tube dysfunction was performed in 1 patient (2%), and residual perforation was found in 3 patients (4%). The overall mean improvement in hearing was 5.67 ± 6.66 dB.

Figures 9-11 illustrate the post-operative otomicroscopic findings.

In Figure 9, we observe a residual perforation, which aligns with our overall findings of three residual perforations. Figure 10 displays a well-healed mastoid

cavity and tympanic membrane, indicating successful post-operative recovery. Figure 11 demonstrates a healed tympanic membrane with no visible mastoid cavity, a result of the mastoid obliteration technique employed during surgery, which effectively prevents common mastoid cavity-related complications.

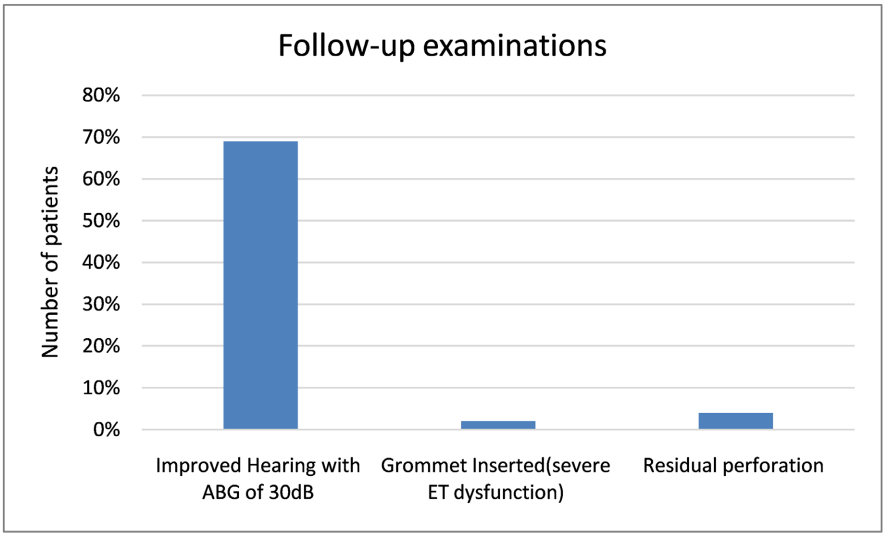

Figure 8. Showing the post operative findings.

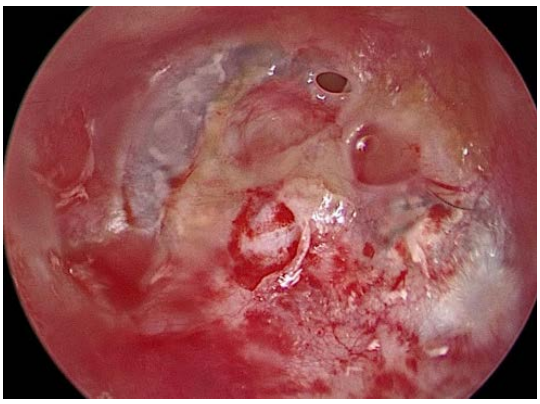

Figure 9. 6 months post operative otomicroscopic image showing a residual perforation.

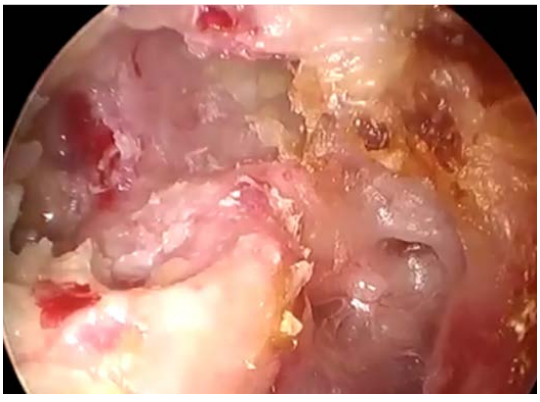

Figure 10. 12 months post operative findings of CWD without mastoid obliteration.

Figure 11. 12 months post operative CWD with mastoid obliteration.

4. Discussion

Pediatric cholesteatoma differs from its adult counterpart due to its rapid progression and higher recurrence rates [8]. This increased aggressiveness may be attributed to the unique immunobiological characteristics of cholesteatoma in children, such as heightened epithelial proliferation and increased inflammatory activity within the perimatrix [9]. Additionally, the presence of a well-pneumatized mastoid in children facilitates the spread of the disease, often necessitating more extensive surgical intervention [10].

The primary goals of cholesteatoma surgery are to eradicate the disease, preserve or improve hearing, and minimize the risk of recurrence. The choice between canal wall down (CWD) and intact canal wall (ICW) techniques is crucial and must be tailored to the individual patient based on factors such as the extent of the disease, patient age, and the surgeon's experience. In our study, the CWD approach was favored for patients with more extensive disease, as reflected by higher CT grading. This technique allowed for complete cholesteatoma clearance, reducing the risk of recurrence, albeit with the potential for cavity-related complications. However, the use of mastoid cavity obliteration techniques successfully mitigated these issues, resulting in a dry, trouble-free ear for the majority of patients [11]. Additionally, reconstructive techniques facilitated the adaptation of hearing aids where necessary.

The ICW technique was selectively employed in cases with limited disease confined to the attic and aditus. While this approach offers superior hearing outcomes and avoids complications associated with an open mastoid cavity, it carries a higher risk of residual cholesteatoma. In our series, patients undergoing ICW surgery were closely monitored with second-look procedures and follow-up MRI using diffusion-weighted imaging (DWI), which has proven effective in detecting residual disease [12].

Our findings align with previous studies, suggesting that the choice of surgical technique should be individualized based on the extent of the disease and patient-specific factors [13]. The dynamic and aggressive nature of pediatric cholesteatoma demands a comprehensive and flexible approach, balancing disease eradication with hearing preservation and quality of life.

5. Limitations

One limitation of this study is the relatively short follow-up period. A longer follow-up is required to accurately assess the recurrence rate of cholesteatoma in this age group.

6. Conclusion

The dynamic course of pediatric cholesteatoma differs from that of the adult variant. Surgical choices should be individualized, taking into account the experience of the operating surgeon. The goal of surgery is to completely eradicate cholesteatoma and related disease using the CWD technique in a single-stage procedure, with reconstruction aimed at achieving both good hearing and a dry, trouble-free ear. ICW is reserved for patients with limited disease extension who are willing to undergo second-look surgery. With the advent of MRI-DWI, recurrent or residual disease can be diagnosed more easily. However, further studies are needed to correlate recurrence rates and hearing outcomes with the extent of cholesteatoma.

Conflicts of Interest

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

References

[1] Mills, R.P. and Padgham, N.D. (1991) Management of Childhood Cholesteatoma. *The Journal of Laryngology & Otology*, **105**, 343-345. https://doi.org/10.1017/s0022215100115944

[2] Dodson, E.E., Hashisaki, G.T., Hobgood, T.C. and Lambert, P.R. (1998) Intact Canal Wall Mastoidectomy with Tympanoplasty for Cholesteatoma in Children. *The Laryngoscope*, **108**, 977-983. https://doi.org/10.1097/00005537-199807000-00005

[3] Jung, T.T., Park, S.K., Kim, Y.H., Lee, S.H. and Kim, H.J. (2004) Clinical Study on Recurrence and Hearing Results in Pediatric Cholesteatoma. *Otology & Neurotology*, **25**, 184-189.

[4] Yung, M.W. and Jacobsen, N.L. (2002) The Relative Merits of Canal Wall up and Canal Wall Down Mastoidectomy in Cholesteatoma Surgery. *The Journal of Laryngology & Otology*, **116**, 806-809.

[5] Persaud, R., Hajioff, D., Trinidade, A., Khemani, S., Bhimrao, S.K. and Jindal, M. (2017) Otologic Surgery: Canal Wall Down versus Canal Wall up Mastoidectomy. *The Journal of International Advanced Otology*, **13**, 157-166.

[6] Ayache, S., Schmerber, S., Lavieille, J.P., Gratacap, B., Duvoisin, B. and Magnan, J. (2008) Canal Wall Reconstruction in Surgery for Cholesteatoma: A Report on 152 Cases. *Auris Nasus Larynx*, **35**, 23-28.

[7] Gantz, B.J., Wilkinson, E.P. and Hansen, M.R. (2005) Canal Wall Reconstruction Tympanomastoidectomy with Mastoid Obliteration. *The Laryngoscope*, **115**, 1734-1740. https://doi.org/10.1097/01.mlg.0000187572.99335.cc

[8] Palva, T. and Ramsay, H. (1991) Cholesteatoma in Children. *Acta Oto-Laryngologica*, **482**, 154-155.

[9] Morris, D.P., Bance, M., De La Cruz, A., Lee, D. and Papsin, B.C. (2003) Pediatric Cholesteatoma: Residual and Recurrent Disease. *Otolaryngology—Head and Neck Surgery*, **129**, 378-383.

[10] Sheehy, J.L. and Brackmann, D.E. (1994) Cholesteatoma Surgery: Canal Wall Down Procedures. *The American Journal of Otology*, **15**, 15-19.

[11] Yung, M.W., Mees, K., Mulder, J.J., O'Reilly, B.F., Yung, M.W., Mees, K., *et al.* (1997) The Benefits of Mastoid Obliteration in Canal Wall Down Mastoidectomy: A Prospective Study of 100 Cases. *Clinical Otolaryngology & Allied Sciences*, **22**, 256-261.

[12] Stadler, D., Giardini, Á.C., Fisch, U. and Wichmann, G. (1998) Reliability of MRI in Detecting Recurrent Cholesteatoma after Canal Wall up Mastoidectomy. *The American Journal of Otology*, **19**, 701-706.

[13] Ludman, H. and Wright, T. (1998) Diseases of the Ear, Nose, and Throat in Children. In: Ludman, H. and Wright, T., Eds., *Diseases of the Ear*, Sixth Edition. Arnold, 37-53.