

OTOLOGY

Dealing with paediatric cholesteatoma: how we changed our management

Il colesteatoma in età pediatrica: come si è modificato il trattamento nella nostra esperienza

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SUMMARY

We reviewed our series of surgeries for paediatric cholesteatoma to assess outcomes and functional results considering the extension of disease and surgical techniques. Between January 2003 and December 2009, 36 patients (range 6-14 years) were operated on for cholesteatoma. We considered the sites involved by the cholesteatoma (mastoid, antrum, attic, middle ear, Eustachian tube), surgical techniques used (intact canal wall, canal wall down) and how our habits changed over the years; moreover, we evaluated ossicular chain conditions and how we managed the ossiculoplasty. As outcomes, we considered the percentage of residual and recurrent cholesteatoma for each technique and hearing function (air bone gap closure, high frequencies bone conduction hearing loss) at follow-up. Intact canal wall was performed in 20 patients and canal wall down in 13 patients, in 9 as first surgery. In both groups, we observed improvement of the air bone gap; in the intact canal wall group, a residual cholesteatoma was observed in 6 patients whereas, during follow-up, 2 patients who underwent a canal wall down showed a recurrent cholesteatoma that was treated in an outpatient setting. Eradication of cholesteatoma and restoration of hearing function in paediatric patients present unique surgical challenges. Our experience shows an increased choice of intact canal wall over the years. Therefore, it is important for the surgeon to counsel parents about the probable need for multiple surgeries, especially if an intact canal wall mastoidectomy is performed.

KEY WORDS: Paediatric cholesteatoma • Intact canal wall technique • Canal wall down technique • Ossiculoplasty • Middle ear endoscopy • Recurrence • Hearing threshold

RIASSUNTO

Abbiamo analizzato i risultati nella nostra casistica riguardo al trattamento chirurgico del colesteatoma in età pediatrica considerando l'estensione della patologia e la tecnica chirurgica impiegata. Nel periodo compreso tra gennaio 2003 e dicembre 2009 abbiamo trattato 36 bambini e nell'analisi abbiamo valutato le sedi coinvolte dal colesteatoma (mastoidi, antra, attico, cassa del timpano, tuba di Eustachio), la tecnica chirurgica impiegata (timpanoplastica chiusa e aperta) e come il loro impiego si sia modificato nel corso degli anni; inoltre abbiamo valutato le condizioni della catena ossiculare e come abbiamo effettuato l'ossiculoplastica. Nell'analisi dei risultati abbiamo valutato il colesteatoma residuo e ricorrente per ciascuna tecnica e la funzionalità uditiva (riduzione dell'ipoacusia trasmissiva, ipoacusia neurosensoriale per le alte frequenze). La timpanoplastica chiusa è stata effettuata in 20 pazienti mentre quella aperta in 13 e per 9 di questi come primo intervento. Con entrambe le tecniche abbiamo ottenuto una riduzione dell'ipoacusia trasmissiva; un colesteatoma residuo è stato individuato in 6 pazienti trattati con timpanoplastica chiusa, mentre 2 pazienti sottoposti a timpanoplastica aperta hanno presentato durante il follow-up un colesteatoma ricorrente. L'eradicazione del colesteatoma e il ripristino della funzionalità uditiva presentano maggiori problematiche nell'età pediatrica. La nostra esperienza fa emergere un maggior impiego della timpanoplastica chiusa nel corso degli anni. Perciò è importante che il chirurgo informi i genitori della possibilità di dover ricorrere a più interventi chirurgici durante la crescita del bambino, qualora si opti per una tecnica chiusa.

PAROLE CHIAVE: Colesteatoma pediatrico • Timpanoplastica chiusa • Timpanoplastica aperta • Ossiculoplastica • Endoscopia dell'orecchio medio • Recidiva • Funzionalità uditiva

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Introduction

Middle ear cholesteatoma can be defined as abnormal collections of squamous epithelium and keratin debris that usually involves the middle ear and mastoid, and can be congenital or acquired; in the latter, it may be primary or secondary. Retraction pockets in the tympanic membrane,

resulting from an atelectatic process, after a long standing Eustachian tube dysfunction, is the most common cause of acquired cholesteatoma in children. These pockets may form in the pars flaccida or, more frequently, in the upper posterior portion of the pars tensa¹. Well pneumatized mastoids allow for more extensive disease compared with more sclerotic mastoid bones in adults^{2,3}. Cholesteatoma

in children often involves the entire mastoid and mesotympanum. For this reason, surgery in children is more difficult than in adults and results are commonly considered to be poorer.

Early surgical treatment is necessary for paediatric cholesteatoma to thoroughly eliminate epithelial and bone disease, to prevent recurrent disease, to produce a dry and safe ear and to restore serviceable hearing. The two main techniques employed to reach these goals are the canal wall-down (CWD) technique and the intact canal wall (ICW) technique⁴. The CWD technique provides lower recurrence rates, but it often requires regular cavity cleaning and is associated with recurrent infection, water intolerance, caloric-induced vertigo and the diminished ability to wear a hearing aid. The ICW technique preserves the normal bony anatomy, avoids the disadvantages associated with cavities, and has shown better hearing results, although it has a significantly higher recurrence rate^{5,6}. More recently, other authors have proposed a CWD technique in conjunction with a mastoid obliteration using material such as fascia, vascularised musculoperiosteal flaps, cartilage and bone pate, reducing the final cavity volume and maintaining the cavity shape to render the ear free of discharge and maximise self-cleansing^{7,8}. Accordingly, there are no universally accepted opinions about the choice of surgical technique for cholesteatoma, especially in children.

In spite of reported data, the evaluation of the outcome of middle ear cholesteatoma surgery in children is difficult because the literature studies differ in terms of age range, patient selection criteria, expertise of surgeons, follow-up times and hearing documentation⁹.

We reviewed the charts of our paediatric cholesteatoma patients to evaluate clinical findings, extension of disease, surgical treatment, recurrences, state of the ossicular chain and hearing results. We focused our analysis on how technological improvements have changed our management philosophy and surgical technique.

Materials and methods

Thirty-six children underwent surgery for middle ear cholesteatoma between January 2003 and December 2009 at

our Institution. In our analysis, we considered the surgical technique employed (ICW vs. CWD), in how many cases an ICW was converted in a CWD and how, over the years, we employed the two techniques.

Based on intraoperative findings, we evaluated the number of sites involved by the cholesteatoma (mastoid cavity, antrum, attic, mesotympanum, Eustachian tube), and the ossicular chain involvement in terms of erosion or absence of the ossicles. We also considered the technique used for ossiculoplasty, which was often delayed at second look in staged ICW procedures (after 9-12 months). CWD procedures were never staged.

We also analysed the number of residual and recurrent cholesteatomas: in case of residual cholesteatoma larger than a small pearl involving an insidious site, such as sinus tympani, we employed a CWD technique.

Follow-up examinations were carried out by otomicroscopic checks and audiometric tests. Considering hearing function, we evaluated the variation between pre-operative and post-operative air-bone gap (ABG) for frequencies between 250 and 4000 Hz. We also considered the variation of high frequency bone conduction (BC) at 2 and 4 kHz to exclude sensorineural damage due to surgical procedures.

Data was processed using GraphPad Software Prism for Macintosh (Version 4.0c). Statistical analysis was done using the Student t-test to compare differences between pre-operative and post-operative ABG and BC thresholds. A p value < 0.05 was considered statistically significant.

Results

Thirty-six children, aged between 6 and 14 years (mean: 10 years; standard deviation: 3 years), were treated; the average follow-up was 38 months (range 13-96 months); during this period three children were lost to follow-up and their data was not included. Nineteen patients were male and 14 female; the left ear was involved by the pathology in 20 cases and the right ear in the remaining 13 cases.

Pre-operative ABG of all patients was 29 dB on average, and during follow-up it showed a mean improvement of 8 dB. High frequency BC decreased by 3 dB (15 dB pre-op and 18 dB post-op).

We observed that, during the period considered in the analysis, ICW techniques increased at the expense of CWD techniques (Fig. 1).

Pre-operative ABG was 31.69 dB (± 3.525) in the CWD population and 26.80 dB (± 2.662) in ICW-treated patients; differences between the two groups did not show statistical significance ($p = 0.2699$), suggesting no differences in preoperative audiological function.

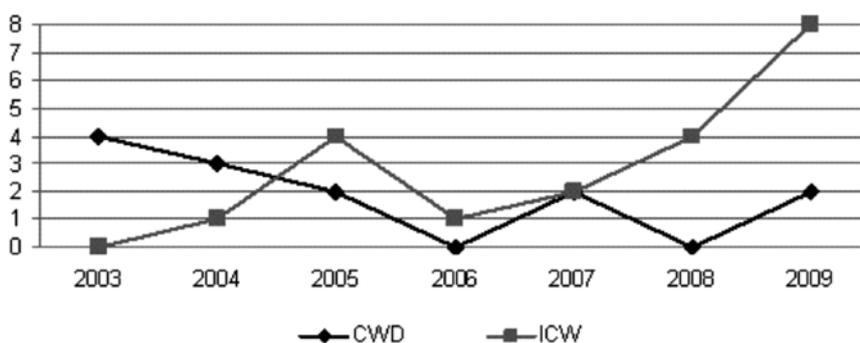


Fig. 1. Number of CWD and ICW techniques over the years.

Table I. Clinical intraoperative findings with cholesteatoma sites of involvement and ossicular chain conditions.

Patients	Sites	ICW			CWD			
		Malleus	Incus	Stapes	Sites	Malleus	Incus	Stapes
1	2	p	e	p	2	e	e	e
2	4	e	e	e	4	e	e	e
3	3	p	e	e	4	p	e	e
4	2	p	e	e	4	e	e	p
5	1	e	e	p	3	p	e	e
6	1	p	e	p	2	p	e	e
7	1	p	e	p	3	p	e	e
8	2	p	e	p	1	p	e	p
9	3	p	e	p	2	p	e	p
10	2	p	e	p	3	p	e	p
11	2	p	e	p	5	p	e	e
12	2	p	e	p	4	e	e	e
13	3	e	e	e	3	e	e	e
14	3	e	e	e				
15	5	p	e	e				
16	2	p	e	e				
17	4	p	e	p				
18	1	p	p	p				
19	2	p	p	p				
20	5	p	p	p				

p: present; e: eroded.

Clinical intraoperative findings with sites of involvement and ossicular chain conditions are reported in Table I. The ICW technique was used as first choice in 20 patients. The cholesteatoma had grown in 2.5 sites on average; the malleus was eroded/absent in 4 cases, the incus in 17 cases and the stapes in 7 cases. The ossicular chain was completely normal and not involved by the cholesteatoma in 3 patients. We observed only one case of erosion of the bony canal of the facial nerve. We performed ossiculoplasty during the first procedure in 11 cases using the body incus remodelled; we delayed ossiculoplasty at the second stage in 6 patients, and we used tragal and conchal cartilage in 2 patients, and a titanium total ossicular replacement prosthesis (TORP) in 4 cases. During the second look staged procedure, there were 6 cases (30%) of residual cholesteatoma; during follow-up we did not observe any recurrent cholesteatoma. Median pre-operative ABG was 26.80 dB (± 2.662) and post-operative ABG was 18.60 dB (± 2.947) showing a significant median improvement of 8.200 ± 3.972 dB (0.1567 to 16.24, IC95%, $p = 0,0458$): at the last audiometric test, 85% of patients showed an ABG between 0 and 20 dB (Fig. 2). The decrease of post-operative high-frequency BC was 3 dB (14 dB pre-op vs. 17 dB post-op), which was not statistically significant ($p = 0.0636$).

The CWD technique was employed as first choice in 9 patients; in remaining 4 it was performed after a CWU for the presence of a dangerous residual cholesteatoma. The cholesteatoma had grown in 3 sites on average; the malleus was eroded/absent in 5 cases, the incus in all cases and the stapes in 9 cases. In two cases, a facial nerve bony canal dehiscence was noted and the second intra-tym-

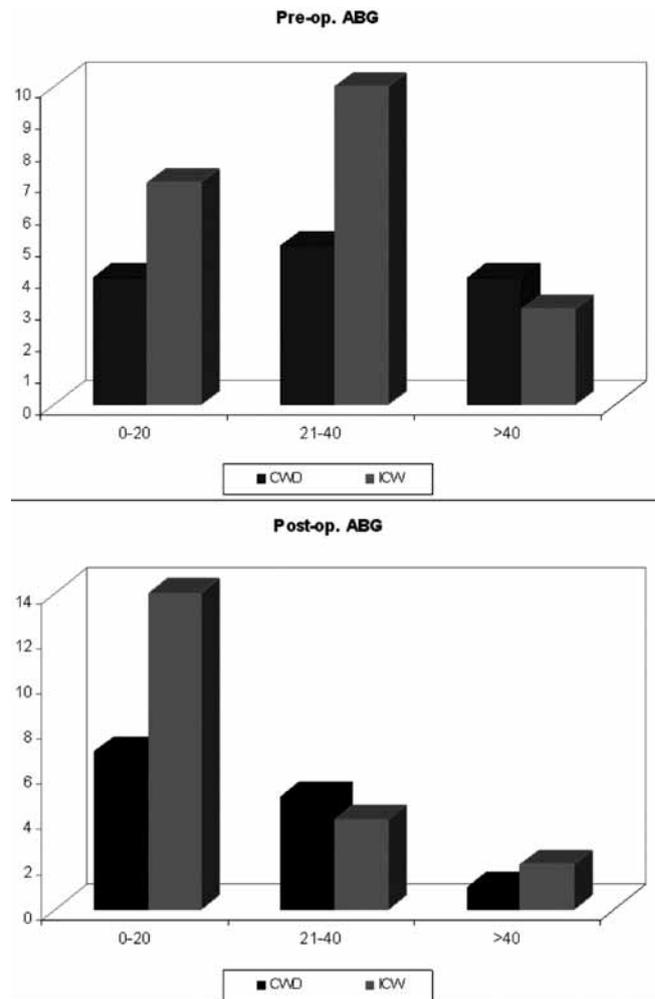


Fig. 2. Pre- and post-operative ABG in the CWD and ICW groups.

panic part was involved. Ossiculoplasty was performed with the remodelled incus remnant in 6 cases, with tragal/conchal cartilage in 4 cases, with a titanium TORP in 2 cases and with the remodelled malleus in 1 case. During follow-up, two recurrent cholesteatomas were observed that were treated in an outpatient setting. Median pre-operative ABG was 31.69 dB (± 3.525) and post operative ABG was 24.38 dB (± 2.807), with a median improvement of 7.308 ± 4.506 dB that was not statistically significant (-1.993 to 16.61, IC95%, $p = 0.1179$): 54% of patients showed an ABG between 0 and 20 dB at the last audiometric test (Fig. 2). The decrease of post-operative high-frequency BC was 4 dB (15 dB vs. 19 dB) that was not statistically significant ($p = 0.1320$).

Fig. 3 shows the median variation of AC and BC for each frequency in both ICW and CWD patients.

Discussion

The most debated topics about paediatric cholesteatoma surgery concern the timing for surgery, choice of surgical technique and outcomes. Some authors believe that paediatric cholesteatoma is more aggressive than in adults, influencing the therapeutic approach and leading to worse clinical outcomes¹⁰⁻¹⁴, whereas others have reported less osteolysis and fewer complications¹⁵⁻¹⁷.

Previously, we reported on a series of 60 children (median age 9 years) treated between 1997 and 2002. We obtained good hearing function and excellent disease eradication employing the CWD technique: indeed, recurrent cholesteatoma was observed in 27% of patients and 57% of patients presented a post-operative ABG of 20 dB or less⁶.

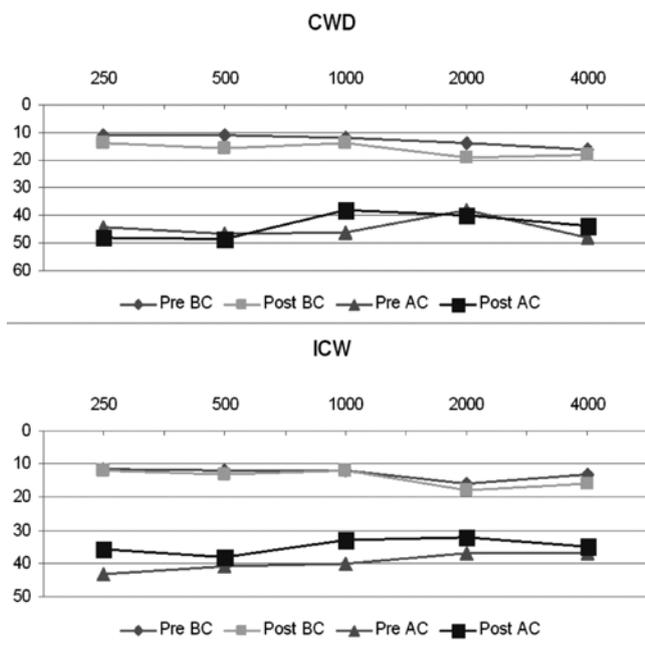


Fig. 3. Median pre- and postoperative hearing threshold in the CWD and ICW groups.

During the following years, the number of CWD procedures decreased and the ICW procedures increased, without a worsening of outcomes. A recurrent cholesteatoma was observed in 2 cases (15%) with the CWD technique, whereas a residual cholesteatoma in 4 cases (20%) using ICW. These results can also be related to a more conservative behaviour: if at the second look surgery a residual cholesteatoma showed a dimension larger than a pearl or involved hidden middle ear sites, such as the sinus tympani, we performed a CWD. We also performed CWD in cases of recurrent cholesteatoma after ICW. This allowed for better control of disease and spared patients from undergoing multiple surgeries. Moreover, we never obliterated the cavities in paediatric patients as we felt that this technique provides few long term advantages and can delay diagnosis of a recurrence. Technological advances have allowed the use of endoscopes even in ear surgery (Fig. 4). Initially, the endoscope was used only to visualise anatomical abnormalities or disease-related anatomical changes during a classical microscopic procedure¹⁸.

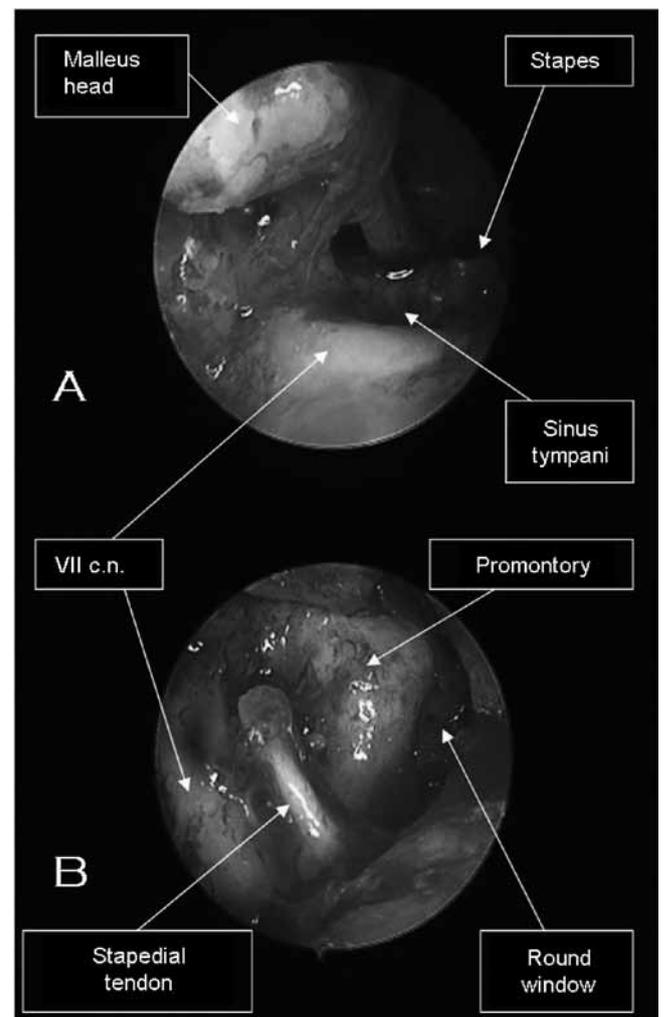


Fig. 4. Endoscopic pictures through the mastoid antrum (A) and the external auditory canal (B); such high quality images allow exploring hidden sites such as the sinus tympani.

Nowadays, endoscopes are taking an important role during certain stages of the surgical procedure itself by reducing the risk of leaving tissue remnants. Moreover, they allow exposure of hidden sites that are difficult to explore only with the microscope¹⁹⁻²¹.

Moreover, during paediatric surgical procedures the use of endoscopes has expanded: Rosenberg²² explored the middle ear after an ICW procedure and concluded that an open second-look mastoidectomy may be avoided if minimal or no recurrent cholesteatoma is found during endoscopic exploration. Good and Isaacson²³ used a rigid endoscope to evaluate the middle ears of a paediatric population during a surgical procedure after all visible cholesteatoma had been removed under the operating microscope, and the removal continued until all disease visualised with the endoscope was removed. Using the endoscope, they detected incompletely removed cholesteatoma in 24% of cases. At planned exploratory procedures, residual disease was found in 18% of ears judged free of cholesteatoma by both otomicroscopy and otoendoscopy at the first surgical procedure. They concluded that, even if otoendoscopy is clearly useful in detecting incompletely removed cholesteatoma, a planned exploratory procedure is required.

However, factors that should be kept in mind for choosing the best surgical technique for paediatric cholesteatoma include: intraoperative view of disease extension, mastoid pneumatization, mucosal and ossicular chain conditions, which can be only supposed with the preoperative radiological scans, and Eustachian tube function.

The ICW technique shows unquestionable advantages compared to CWD such as the need for less postoperative therapy and better hearing function; on the other hand, these advantages cannot justify the need for a child to undergo multiple surgical procedures, due to the higher percentage of residual and recurrent cholesteatoma reported with the ICW technique.

CWD is now reserved for larger cholesteatoma or for unfavourable anatomic conditions. The choice to perform a CWD is often intraoperative and related to anatomical conditions that do not allow safe cholesteatoma removal (a low-lying middle cranial fossa dura and an anteriorly positioned sigmoid sinus which create a small mastoid and limit access into the anterior epitympanum) or for the presence of a perilymphatic fistula.

Patients submitted to CWD did not improve hearing loss in a statistically significant manner, but showed an ABG between 0 and 20 in 54% of cases and a sensorineural hearing loss similar to patients submitted to ICW (4 vs. 3 dB). We reserved CWD for large cholesteatoma as revealed by the number of anatomical sites involved and by greater ossicular chain involvement.

Considering functional outcomes, there is no agreement about the need to preserve the posterior canal wall to obtain a good hearing function²⁴, and preoperative hearing

function seems to be more important, which may be a sign of either smaller or larger ossicular chain involvement^{25,26}. Indeed, pre-operative status and the surgical technique for the ossiculoplasty are the most important factors influencing postoperative hearing function. Even today, incus transposition, if not totally involved by cholesteatoma, is the technique of choice; in the remaining cases different solutions, such as cartilage and biocompatible materials, are needed.

Conclusions

Eradication of cholesteatoma and restoration of hearing function in paediatric patients present unique surgical challenges. The balance between these two goals is related to the incidence of recidivism, the degree of ossicular damage and the experimental evidence that this disease exhibits a more aggressive behaviour than in adults. Fortunately, intratemporal and intracranial complications, such as inner ear fistula, facial nerve paralysis and epidural or intracerebral abscess, are rare in children.

Therefore, the surgeon must counsel parents on the probable need for multiple surgeries, especially if an ICW mastoidectomy is performed. The more intriguing prospects of paediatric cholesteatoma should focus on standardisation of the endoscopic approach and on its real advantages in removal of remnant cholesteatoma with ICW; experimental studies have the possibility to detect new histological markers that could help in predicting recurrences. In the light of the above-mentioned data, an individualised approach is needed for the treatment of paediatric cholesteatoma, and the choice of surgical technique should be based on anatomical, biological, radiological and social factors.

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