**Surgical Treatment of Pediatric Cholesteatomas**

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**Objective:** Management of pediatric cholesteatomas remains controversial. We reviewed our 16-year experience in the surgical treatment of cholesteatomas in children and describe a treatment paradigm. **Study Design:** The authors conducted a retrospective review. **Methods:** A total of 106 mastoidectomies (86 for an acquired cholesteatoma and 20 for a congenital cholesteatoma) were performed in children 16 years old and younger from 1988 to 2003. Follow up ranged from 2 years to 12 years with a mean follow-up period of 6 years. Hearing outcomes, cholesteatoma recidivism, and dry mastoid cavity were the main outcomes measured. **Results:** Seven (7%) patients had revision surgery for cholesteatoma recidivism. Rates of cholesteatoma recurrence for canal all up (CWU) and canal wall down (CWD) mastoidectomy groups were similar (8% vs. 6%). The percentage of patient with good serviceable hearing (pure-tone average ≤25 dB) was higher in those with a CWU mastoidectomy as compared with the CWD mastoidectomy group (81% vs. 47%) \((P < .05)\). Extent of disease and stapes superstructure erosion on presentation were significant \((P < .05)\) predictors of both cholesteatoma recidivism and poor hearing. All 106 subjects studied had a dry mastoid and 78 patients (74%) had a maintenance-free cavity at the time the study was completed. **Conclusion:** The treatment of pediatric cholesteatomas should be individualized with CWD mastoidectomy chosen for patients with recurrent or more extensive disease. We conclude that the CWU procedure is an adequate surgical option for treating most acquired and congenital cholesteatomas, preventing disease recurrence, and maintaining good hearing outcomes. **Key Words:** Pediatric cholesteatomas, canal wall up mastoidectomy, canal wall down mastoidectomy.

**INTRODUCTION**

The incidence of cholesteatoma in childhood is estimated to be three to six per 100,000 people. These rates tend to higher among groups predisposed to chronic ear infections such as Native-Americans and those with craniofacial abnormalities. Cholesteatomas in the pediatric population are defined as either acquired (AC) or congenital (CC). Nearly 70% of pediatric cholesteatomas are AC. Surgical management of pediatric cholesteatomas has continued to raise debate.

The goals of cholesteatoma surgery in children are: 1) eliminate all disease, 2) preserve or restore hearing, and 3) prevent recurrent disease. To achieve these goals, two surgical techniques exist: the canal wall up mastoidectomy (CWU) and canal wall down mastoidectomy (CWD). The CWU technique offers the advantage of having a relatively small anatomic deformity with a normal ear canal. Recurrences have been reported to be more frequent with this procedure and may be more difficult to detect. With CWD mastoidectomy, on the other hand, the removal of the canal wall allows for earlier detection of recurrent disease. As a result of the prevalence of chronic serous otitis media (CSOM) in children, rapid tissue growth, and eustachian tube dysfunction, cholesteatomas in children may have a more aggressive growth pattern relative to adults. The incidence of both residual and recurrent disease is higher in children compared with adults. These findings have been a continued reason for controversy in the management of pediatric cholesteatomas. In this present study, we retrospectively reviewed our 16-year experience with pediatric cholesteatomas and describe the long-term results. We propose a paradigm that individualizes treatment for each patient and uses a CWU procedure with facial recess approach for most pediatric cholesteatomas and reserves CWD mastoidectomy for more extensive disease.

**MATERIALS AND METHODS**

After approval from the Investigational Review Board of Loyola University Medical Center, a chart review of patients 16 years old and younger who had a mastoidectomy for cholesteatoma between 1988 to 2003 was examined retrospectively. A total 106 patients were identified. Follow up ranged from 2 years to 12 years with a mean follow-up period of 6 years. All surgeries were performed by the three senior authors (J.P.L., S.J.M., K.M.), and the criteria for choosing either an open or closed procedure re-
RESULTS

Data were available for review on 106 patients. There were a total of 68 males and 38 females with an age range from 3 to 16 years old (mean age, 8 years). There were a total of 86 patients with an AC and 20 patients with a CC (Table I). The location of the cholesteatoma was in the epitympanum in 11 patients (10%), mesotympanum in 7 patients (7%), attic and meso/hypotympanum 32 patients (30%), attic and antrum 35 patients (33%), and all tympanic and mastoid cavities 21 patients (20%) (Fig. 1). Cholesteatoma was found in the classically described anterosuperior quadrant in only five patients (25%) with CC and diffuse invasion of the mesotympanum was seen in 15 patients (75%). The decision to remove the canal wall remained constant among all surgeons and was based on preoperative imaging, surgical anatomy in the operating room, only hearing ear, reliability of follow up, and extent of disease.

Of the 86 patients with an AC, 38 patients had a CWU procedure (mean age, 10.5 years) (Table I). The procedure was staged with delayed ossiculoplasty in 17 patients (45%). Primary ossiculoplasty was done in three patients (7.5%). Eighteen patients (47%) required no ossiculoplasty as a result of limited disease. Two patients required revision surgery (one for residual cholesteatoma and one for recurrent disease) (Table II). Both of these patients underwent a CWD mastoidectomy as a result of their extensive disease. Forty-eight patients had a CWD mastoidectomy for an AC (mean age, 10 years). Twenty-one patients (44%) had a previous CWU mastoidectomy before referral to our institution. The procedure was staged with delayed ossiculoplasty in 22 patients (46%). Primary ossiculoplasty was done in seven patients (15%). Revision mastoidectomy was required in 10 patients (21%) (two for recurrent disease and eight for granulation and stenosis).

Of the 20 patients with CC, 15 underwent a CWU mastoidectomy (mean age, 4.5 years) (Table I). The procedure was staged with delayed ossiculoplasty in all patients. Revision mastoidectomy was performed in two patients (13%) (one for residual cholesteatoma and one for recurrent disease) (Table II). One patient required CWD mastoidectomy for recurrent disease. Canal wall down mastoidectomy was chosen in five patients for CC (mean age, 7 years) as a result of significant disease extending into the sinus tympani and thus making complete removal of disease difficult with an intact canal wall procedure. The surgery was staged with delayed ossicular reconstruction in three patients (60%). Primary ossiculoplasty was performed in two patients (40%). Revision mastoidectomy was needed in one patient (20%) for recurrent disease.

In patients with an AC, the most current audiometric data revealed that 34 patients (89%) who underwent a CWU mastoidectomy and 22 patients (46%) who had a CWD procedure had PTA ≤25 dB (Table III). Poor hearing outcomes were found to be significantly (P < .05) related to stapes superstructure (SS) erosion. Twenty patients (42%) in the CWD group had SS involvement as compared with five patients (13%) in the CWU group (Table IV). In patients with CC, hearing results revealed that nine patients (60%) who underwent a CWU mastoidectomy and three patients (60%) who had a CWD procedure had PTA ≤25 dB. Incidence of SS erosion was the same (40%) in both groups (Table IV).

Revision mastoidectomy was necessary in 15 patients (14%). Seven patients (7%) had revision surgery for cholesteatoma recidivism. The recidivism rate was higher (15% vs. 5%) in patients with CC as compared with those with an AC (Table II). Sixty-eight patients (64%) had serviceable hearing with PTA ≤25 dB. The percentage of patients with good serviceable hearing was higher in those with a CWU mastoidectomy as compared with the CWD mastoidectomy group (81% vs. 47%) (P < .05) (Table III). Thirty-three patients (31%) had SS erosion in this study.
which was an accurate predictor of poor hearing outcome regardless of surgical procedure performed \((P < .05)\). The CWD mastoidectomy group had a higher percentage of SS erosion \((42\% \text{ vs. } 21\%) \text{ } (P < .05)\) (Table IV). One patient \((0.8\%)\) had facial nerve paralysis, which required decompression in this series. The patients had normal facial function noted 2 months postoperatively. A dry mastoid cavity was present in all 106 patients at the conclusion of this study. Seventy-eight patients \((74\%)\) chose to do a CWD mastoidectomy \((n = 53)\).

**DISCUSSION**

The goals of cholesteatoma surgery in children are to eliminate disease, create a safe ear, and preserve or restore hearing. Cholesteatomas in children tend to be more difficult to eradicate compared with adults as a result of the more extensive and aggressive nature of the disease in children.\(^4,5\) To create a disease-free cavity and to restore hearing, two surgical procedures (CWU and CWD mastoidectomy) are available. The CWU procedure preserves the posterior canal wall, allows for more rapid healing, avoids restrictions on water exposure, and provides greater choice on hearing aids.\(^7\) The CWU procedure is therefore ideal in a well-pneumatized mastoid and middle ear. Relative contraindications to a CWU procedure include a sclerotic mastoid, labyrinthine fistula, only hearing ear, poor eustachian tube function.\(^8\) The CWD mastoidectomy, on the other hand, involves taking down the posterior canal wall to the vertical facial ridge and marsupialization of the mastoid cavity. The surgeon should therefore be provided better access to the middle ear and the epitympanum. These patients require long-term follow-up and periodic cleaning of their mastoid cavity. We chose to do a CWD mastoidectomy \((n = 53)\) in those patients who 1) had a failed previous CWU procedure \((21\%\) of patients \([40\%]\)), 2) poor follow up and therefore require a one-stage procedure for complete disease removal \((4\%\) of patients \([8\%]\)), 3) sclerotic mastoid with no aeration in the attic and antrum \((53\%\) of patients \([100\%]\)), 4) only hearing ear \((4\%\) of patients \([8\%]\)), and 5) labyrinthine fistula \((4\%\) of patients \([8\%]\)) (Table V).

In assessing outcomes in our series, we agree with previous authors that the total percentage of cholesteatoma recidivism should be the basis for evaluation of the outcome of surgery.\(^9\) We defined residual disease as persistence of disease after incomplete removal. Recurrent cholesteatoma was defined as a newly formed disease process secondary to a retraction pocket. With CWU mastoidectomy for both AC and CC, four patients \((8\%)\) required revision surgery: two patients \((4\%)\) had recurrent disease and two patients \((4\%)\) for residual disease. In analyzing the CWD group for both AC and CC, 11 patients \((21\%)\) had revision surgery: three patients \((6\%)\) for recurrent disease, eight patients \((15\%)\) for stenosis and granulation, and no patients were found to have residual disease. There was no statistical difference between the two groups in terms of recurrent disease \((\text{CWU} 8\% \text{ and CWD } 6\%)\) (Table II). In addition, these rates are lower than reported in several previous large series.\(^9,10\) Review of the literature states that extension of the disease into the epitympanum or the sinus tympani is considered one of the most common causes of failure in CWU mastoidectomy.\(^11\) Development of a deep retraction pocket results in a higher recidivism rate with a CWU mastoidectomy and detection of disease may be delayed, especially if the recurrence presents in the attic or mastoid cavity.\(^10,12\) A second-look procedure 6 to 9 months later in select patients who underwent a CWU mastoidectomy resulted in a relatively low recidivism rate. Better visualization with the CWD mastoidectomy was confirmed in this series because no residual cholesteatomas were detected in 53 patients who underwent this procedure. Nevertheless, a relatively low percentage \((4\%)\) of patients in CWU mastoidectomy group had residual disease. All seven patients who had recurrent or residual disease in this series had extensive disease and SS involvement on presentation.

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**TABLE II.**

<table>
<thead>
<tr>
<th></th>
<th>Acquired Cholesteatoma</th>
<th>Congenital Cholesteatoma</th>
<th>Recidivism Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Canal wall up</td>
<td>2</td>
<td>2</td>
<td>8% (4 of 53)</td>
</tr>
<tr>
<td>Canal wall down</td>
<td>2</td>
<td>1</td>
<td>6% (3 of 53)</td>
</tr>
<tr>
<td>Recidivism rate</td>
<td>(5% (4 of 86))</td>
<td>(15% (3 of 20))</td>
<td></td>
</tr>
</tbody>
</table>

**TABLE III.**

<table>
<thead>
<tr>
<th></th>
<th>Acquired Cholesteatoma</th>
<th>Congenital Cholesteatoma</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Canal wall up</td>
<td>(89% (34 of 38))</td>
<td>(60% (9 of 15))</td>
<td>(81% (43 of 53))*</td>
</tr>
<tr>
<td>Canal wall down</td>
<td>(46% (22 of 48))</td>
<td>(60% (3 of 5))</td>
<td>(47% (25 of 53))</td>
</tr>
<tr>
<td>Total</td>
<td>(65% (56 of 86))</td>
<td>(60% (12 of 20))</td>
<td></td>
</tr>
</tbody>
</table>

*\(P < .05\).*

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**TABLE IV.**

<table>
<thead>
<tr>
<th></th>
<th>Acquired Cholesteatoma</th>
<th>Congenital Cholesteatoma</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Canal wall up</td>
<td>(13% (5 of 38))</td>
<td>(40% (6 of 15))</td>
<td>(21% (11 of 53))</td>
</tr>
<tr>
<td>Canal wall down</td>
<td>(42% (20 of 48))</td>
<td>(40% (2 of 5))</td>
<td>(42% (22 of 53))*</td>
</tr>
<tr>
<td>Total</td>
<td>(29% (25 of 86))</td>
<td>(40% (8 of 20))</td>
<td></td>
</tr>
</tbody>
</table>

*\(P < .05\).*

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**TABLE V.**

<table>
<thead>
<tr>
<th>Reasons for Canal Wall Down Mastoidectomy</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Failed canal wall up procedure</td>
<td>40%</td>
</tr>
<tr>
<td>Poor follow up, requiring single-stage procedure</td>
<td>8%</td>
</tr>
<tr>
<td>Sclerotic mastoid</td>
<td>100%</td>
</tr>
<tr>
<td>Only hearing ear</td>
<td>8%</td>
</tr>
<tr>
<td>Labyrinthine fistula</td>
<td>8%</td>
</tr>
</tbody>
</table>

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Thus, extensive disease on presentation with ossicular chain involvement is a good predictor of patients at high risk for recurrent disease (P < .05).

As reported by Ueda et al., we staged ossiculoplasty in conjunction with a second-look procedure in those patients with significant disease in the tympanic and mastoid cavities and whose preoperative computed tomography imaging showed no aeration in the attic and antrum. If good aeration is seen on computed tomography without significant ossicular erosion, a one-stage procedure was performed. If ossicular erosion was evident during surgery, we elected to do a planned second look and staged the ossiculoplasty. Mucosal and matrix preservation techniques were used in all cases in which healthy mucosa and matrix could be preserved. In addition, use of silastic sheeting in patients at high risk for residual or recurrent disease has improved mastoid aeration and helped to decrease cholesteatoma recidivism in our experience. In patients who continue to have poor aeration of the attic and antrum and develop extensive cholesteatoma, a CWD mastoidectomy is helpful in disease eradication. We do not recommend obliteration of the mastoid cavity as a result of risk of recurrent or residual disease in this patient population. The posterior canal wall is not reconstructed because many of the patients in our study (20%) had already failed CWU procedures.

A second important outcome measure is hearing restoration or preservation. Several published reports have described that the status of the canal wall has little effect on hearing outcomes. Our data support this contention in the CC group. Among patients with an AC, 89% of patients treated with a CWU mastoidectomy had PTA ≤25 dB as compared with 46% of patients who underwent a CWD procedure (P < .05). As stated by Dodson et al., serviceable hearing depends primarily on middle ear parameters such as the mucosal status (stenosis and granulation), condition of the TM (thickness, contour), depth of the middle ear cleft, and presence or absence of SS erosion. The incidence of SS erosion was the same (40%) in patients with CC who had a CWU or CWD mastoidectomy. These findings also explain the reason why the status of the canal wall had no effect on hearing outcomes in this group. With an AC, 58% of patients in the CWD mastoidectomy group had either SS erosion or significant stenosis or granulation as compared with 13% of patients in the CWU group (P < .05). In addition, patients with CC had worse hearing compared with those with an AC. The CC group had a higher percentage of patients with SS erosion (40% vs. 27%) (Table IV). These results were not statistically significant, however, as a result of the smaller sample size in the CC group. This greater prevalence of extensive disease in the CC group explains the much higher cholesteatoma recidivism rate observed in this patient population (15% vs. 6%) (Table II). The majority of our patients with CC (75%) had extensive disease in the mesotympanum instead of disease isolated to the classically described anterosuperior quadrant, thus explaining the worse hearing results observed. Similar to the findings of Potsic et al., our findings conflict with the theory that CC arise from embryonic cell rest in the anterior superior quadrant. The pathogenesis of CC, therefore, may be multifactorial as stated by Sade et al. (metaplasia of middle ear mucosa) and Aimi (epithelial migration). Similar to previous reports, we found that hearing outcomes were closely related to preoperative hearing. Our study showed that in both AC and CC groups, patients whose preoperative PTA ≤25 dB had the best chance of maintaining good hearing (P < .05). With AC, 21 patients (24%) had PTA ≤25 dB both pre- and postoperative, 31 patients (36%) improved their PTA to ≤25 dB, 23 patients (27%) maintained their PTA >25 dB, and 11 patients (13%) had worse hearing postoperatively. With CC, five patients (25%) had PTA ≤25 dB and all maintained good serviceable hearing postoperatively, seven patients (35%) improved their PTA to ≤25 dB, and three patients (15%) had worse hearing postoperatively.

CONCLUSION

Our review of 106 pediatric patients who had cholesteatoma surgery over a 16-year period revealed some interesting conclusions about management of this disease process. Patients with an AC presented at a later age than their CC counterparts. Congenital cholesteatomas isolated to the classically described anterior superior quadrant were rare. A second type of CC group with extensive disease involving the mesotympanum and SS were more common. Despite complete disease removal and ossiculoplasty, the worst hearing outcomes in both AC and CC were seen in those patients with SS erosion and stenosis/granulation tissue in the middle ear space. In addition, our review showed that there was a significantly higher recurrence rate in patients with more extensive disease and SS resorption compared with those with disease isolated to the mesotympanum and without SS involvement. Cholesteatomas in children may be more difficult to treat with a single surgical procedure. Therefore, treatment should be individualized for each patient. We propose that a CWU mastoidectomy should be recommended in most cases of both AC and CC with a planned second look in patients with extensive disease. CWD mastoidectomy should be part of an informed consent discussion with patients and families before surgical intervention and should be used in select patients.

BIBLIOGRAPHY


