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Acquired middle ear cholesteatoma in children with cleft palate: Experience from 18 surgical cases

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ABSTRACT

Objectives: To review an institutional experience with the surgical management of middle ear cholesteatoma in children with cleft palate.
Materials and methods: We analyzed retrospectively 18 children diagnosed with cleft palate who underwent surgery for acquired middle ear cholesteatoma between 2000 and 2007. The following data were recorded: age, sex, history of ventilation tube insertion, status of the contralateral ear, cholesteatoma location and extension, and surgical technique involved. Cholesteatoma recidivism, stable mastoid cavity and hearing levels were the main outcomes measured.
Results: Follow-up ranged from 5 to 12 years (mean 8 years). Twelve children underwent planned staged canal wall up mastoidectomy: a residual cholesteatoma was found and removed during the second-look procedure in 2 ears (16.6%); two children (16.6%) showed a recurrent cholesteatoma and required conversion to canal wall down mastoidectomy. A modified Bondy technique was chosen in two children with an epitympanic cholesteatoma with an intact tympano-ossicular system, while in the remaining four subjects a canal wall down mastoidectomy was performed because of an irreparable erosion of the postero-superior canal wall: no cases of recurrent cholesteatoma were observed in these 6 children; revision mastoidectomy was needed in one patient for cavity granulation. A postoperative air-bone gap result of 0–20 dB was achieved in 11 children (61.1%); in 5 cases (27.7%) postoperative air-bone gap was between 21 and 30 dB, while in 2 (11.1%) was >30 dB. Bone conduction thresholds remained unaffected in all cases.
Conclusions: Our results indicate that most cleft palate children with cholesteatoma can be managed with a canal wall up mastoidectomy with low complication rates. In extensive disease with large erosion of the canal wall as well in presence of a retraction pocket in the contralateral ear, a canal wall down mastoidectomy should be considered. In epitympanic cholesteatomas with an intact tympano-ossicular system and mesotympanum free of disease, the modified Bondy procedure is an effective surgical option. As in the general pediatric population, improvement or preservation of hearing can be obtained in most patients.

1. Introduction

The incidence of cholesteatoma in childhood has been reported at 3–6 per 100,000 [1], while in children with cleft palate (CP) is much higher, ranging from 1.8% [2] to 9.2% [3].

A poor Eustachian tube (ET) function leading to reduced middle ear pressure and tympanic membrane retraction contributes to the elevated incidence of acquired cholesteatoma in these children. Beyond the traditional explanation of ET dysfunction, potential genetic contribution to the pathogenesis of cholesteatoma has also been suggested [4]. In addition, since children with CP are more likely to undergo one or more ventilation tube insertions for persistent otitis media with effusion (OME), it has been discussed over the role of grommet insertion as an iatrogenic cause of secondary cholesteatoma. Vlastarakos et al. [5] in a study on complications associated with ventilation tube insertion reported an incidence of secondary cholesteatoma around 1%.

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If untreated, middle ear cholesteatoma can lead to facial palsy, labyrinthine fistula with sensorineural hearing loss as well as endocranial complications; until novel medical management emerges, surgical treatment is our only tool to avoid such complications. Historically, a controversy exists in the literature about the optimal management of pediatric cholesteatomas: some surgeons have advocated the canal wall up (CWU) technique [6,7], others have preferred the canal wall down (CWD) procedure [8,9], and a number of surgeons have suggested hybrid and reconstructive techniques [10,11]. Many surgeons believe that most cholesteatomas can be managed with a CWU technique and consider relative contraindications to this procedure a poor ET function, only hearing ear, labyrinthine fistula, and contracted mastoid [12]. Since ET dysfunction is an almost universal finding of CP, children with this facial malformation do not seem ideal candidates to a CWU procedure. On the other hand, it has been demonstrated that ET function improves with increasing age, facial skeletal growth and after CP repair, reaching normal levels by the age of 10 years [13]. Furthermore, there is some evidence that results of tympanoplasty in patients with CP are similar to those in patients without CP [14,15]. To our knowledge, no study has addressed specifically the results of surgical treatment of cholesteatoma in children with CP. In this study, we retrospectively reviewed our experience with acquired middle ear cholesteatoma in children with a history of CP repair and described the long-term anatomical and functional results.

2. Materials and methods

Our otological database was used to search out all children (0–16 years) diagnosed with cleft palate who underwent surgery for acquired middle ear cholesteatoma at the Department of Otolaryngology of the University of Parma between January 2000 and September 2007. The study protocol was approved by the institutional review board and an informed written consent was obtained from all patients. Cholesteatoma was diagnosed otoscopically and confirmed by high resolution computed tomography in all children. Medical reports were analyzed by age, sex, history of ventilation tube insertion, cholesteatoma location and extension, status of the contralateral ear, surgical procedure adopted, preoperative and postoperative audiograms. Cholesteatoma recidivism, ears with CWU mastoidectomy later requiring conversion to CWD mastoidectomy, postoperative complications, and hearing levels were the main outcomes measured. Hearing results were evaluated according to guidelines set forth by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology and Neck Surgery for the evaluation of results of treatment of conductive hearing loss [16]. Pure-tone average (PTA) was calculated as the mean of 500, 1000, 2000, and 3000 Hz thresholds. The air-bone gap was reported as the four-tone PTA for air-conduction and bone-conduction values determined at the same time. Hearing results were determined at the last follow-up.

In our department the surgical procedure of choice for the treatment of childhood cholesteatoma is CWU mastoidectomy; in most cases a second-look procedure is performed 12–18 months after the first surgery as it is considered an integral part of CWU mastoidectomy for the detection and removal of residual cholesteatoma. In all patients of this series, when a CWU mastoidectomy had been chosen, the first-stage operation consisted in the exposure of the bony external canal and mastoid by retroauricular approach, mastoidectomy, atticotomy and posterior tympanotomy. The removal of the cholesteatoma was performed using a combined approach (transcanal–transmastoid) with the support of chemically assisted dissection, as previously described [17,18].

At the end of the extirpation of the disease, a silastic sheet with a projection into the ET was placed through the epitympanum and posterior tympanotomy to completely cover the medial wall of the middle ear and the mastoid. Defects of the postero-superior canal wall were repaired using bone patè and myringoplasty was performed using the temporalis fascia.

The second-stage procedure was performed at least 12 months after the first-stage operation using the same facial recess approach created during the primary surgery. Following the silastic sheet removal, the exploration of all the cavities of the middle ear and mastoid was performed using the same combined technique as in the first surgical step. In all cases small micromirrors or 30° or 70° angled endoscopes were used to search for residual disease. Residual cholesteatoma was defined as a collection of squamous epithelium found in the middle ear considered to have developed from epidermal debris left inadvertently in place during the first stage of surgery.

3. Results

A total of 18 children were identified and included in this study: 11 boys and 7 girls. Ten of these 18 patients belonged to the group of 293 children treated and followed up at our Department; thus, the incidence of cholesteatoma in our group of cleft patients was 3.3%. The remaining 8 children were referred to our department from other hospitals. Twelve (60%) out of the 18 patients developed a cholesteatoma despite one or more ventilation tube insertion (Table 1), while the remaining 6 (all of these subjects belonged to the group of patients referred to us from other hospitals) had a cholesteatoma with a negative history for ventilation tube insertion

Table 1
Relevant patient demographic data, cholesteatoma extension, type of surgery and postoperative complications in patients with a positive history for ventilation tube insertion.

Patient	Age/sex	Cholesteatoma extension	Number of VTI	Surgical technique	Postoperative complications
1	6/M	R	1	Staged CWUM	None
2	11/F	E	1	Staged CWUM	None
3	13/M	E,A,M	2	Staged CWUM	RC requiring CWDM
4	9/M	E,R,A,M	1	Staged CWUM	None
5	8/M	E,A,M	1	Staged CWUM	None
7	16/F	R	2	Staged CWUM	RC requiring CWDM
9	10/M	E,R,A,M	1	Staged CWUM	None
11	10/M	E,A,M	2	Staged CWUM	None
13	11//F	E,A,M	2	CWDM	None
14	12/F	E,R,A,M	1	CWDM	Otorrhea requiring revision
15	9/M	E,R,A,M	1	CWDM	None
18	7/M	E	1	MBT	None

R: retrotympanum; E: epitympanum; A: antrum; M: mastoid; CWUM: canal wall up mastoidectomy; CWDM: canal wall down mastoidectomy; MBT: modified Bondy technique; VTI: ventilation tube insertion; RC: recurrent cholesteatoma.