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Does canal wall down mastoidectomy benefit syndromic children with congenital aural stenosis?

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ABSTRACT

Objectives: To determine whether a canal wall down mastoidectomy can provide long-term benefit for children with aural stenosis.
Methods: Retrospective case series of children with congenital aural stenosis having undergone a canal wall down mastoidectomy over a twelve-year period at a tertiary children's hospital.
Results: Data from thirteen children who underwent a total of twenty canal wall down mastoidectomies for aural stenosis were reviewed. The mean age at surgery was 7.1 years (range, 3.3–12.3 years). All patients had genetic syndromes including Trisomy 21 (n = 7), Trisomy 21 and Pierre Robin sequence (n = 1), Angelmann (n = 1), Cri-du-chat (n = 1), Branchio-oto-renal syndrome (n = 1), Spina bifida (n = 1) and Nager syndrome (n = 1). Seven (54%) children underwent bilateral canal wall down mastoidectomies. All thirteen ears that could not be visualized preoperatively had improved ease of office examination following surgery. Only one patient required revision surgery and all canals were patent at the last clinic visit. The mean follow-up was 4.9 years. There were no cases of facial nerve injury or cerebrospinal fluid leak.
Conclusion: Syndromic children with congenital aural stenosis with poorly pneumatized mastoids may benefit from canal wall down mastoidectomy to improve ease of office examinations.

1. Introduction

Congenital aural stenosis (CAS) arises from failure of recanalization of the external auditory canal (EAC) during embryologic development. Malformations of the EAC occur in 1:10,000–20,000 births [1]. Congenital aural stenosis is part of the spectrum of developmental EAC anomalies and differs from complete aural atresia due to the presence of a canal lumen. Few classification systems describe the purely stenotic canal. Schuknecht [2] described a four-level grading system that includes two types of EAC stenosis with a patent dermal tract; type A refers to narrowing limited to the fibrocartilaginous canal with normal middle ear structures and type B describes stenosis of the fibrocartilaginous and bony canals with or without dysmorphic ossicles. Middle ear anomalies in patients with CAS vary considerably and there is typically a lesser degree of conductive hearing loss than observed with complete aural atresia [3]. Conductive hearing loss in CAS may be secondary to debris occluding the canal or chronic otitis externa. Assessing the middle ear in an uncooperative child with severe EAC stenosis in the clinic is extremely challenging. This is an important consideration in the nonverbal child and can lead to inaccurate treatment of otologic infections.

There remains a paucity of data regarding management of the purely stenotic ear. Atresiaplasty outcomes have been described for patients with no detectable lumen and have excluded patients with craniofacial anomalies. These series have demonstrated that restenosis is the most common complication and indication for revision surgery, which occurs in up to 25% of cases. The tympanic membrane may become lateralized and require revision surgery for conductive hearing loss in 0–18% of cases [1,4]. A small series of five patients described a minimally invasive procedure with an endaural incision from the root of the helix down to the superior annulus with allograft to line the defect [5]. Chen et al. [6] described an endaural-conchal incision using two local rotation flaps and a transposition split-thickness scalp flap. A transmastoid approach in

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children with complete aural atresia with pneumatized mastoids showed a high restenosis rate of 17.5% ears [7]. To our knowledge, canal wall down mastoidectomy has not been described as a treatment option for congenital aural stenosis in children with syndromes and poorly pneumatized mastoids. We present our experience using this surgical technique over a twelve-year period and discuss patient factors in which this intervention would be most beneficial.

2. Materials and methods

Following Institutional Review Board approval by the University of Utah and Primary Children's Hospital, medical records were reviewed retrospectively. We identified children via the *Current Procedural Terminology* codes 69641, 69642, 69645, or 69505 and included children with a diagnosis of external canal stenosis who underwent canal wall down mastoidectomy between March 20, 2003 and July 30, 2015. We included children older than 3 years of age with <2 mm narrowing of the lateral portion of the soft issue ear canal. Patient data including age, gender, date of operation, laterality of procedure, genetic syndromes and duration of follow-up were collected.

The demographic and clinical results are summarized in Table 1. A total of thirteen children and twenty ears were included in the study. Five (38%) of the subjects were male. The mean age at surgery was 7.1 years (range, 3.3–12.3 years). All patients had genetic syndromes with the most common being Trisomy 21 (62%). Preoperative computed tomographies (CT) with Schuknecht type, EAC diameter and middle ear findings were recorded. All mastoids were poorly pneumatized. Audiometric data included preoperative and postoperative pure-tone average (PTA: 0.5, 1, 2 and 3 kHz) from audiometric brainstem response (ABR) or audiogram when available. The main outcome measure was defined as maintenance of a patent canal and improvement in ease of office examination.

2.1. Surgical technique

General anesthesia and intraoperative facial nerve monitoring were used during all procedures. Canal incisions were made adjacent to the annulus extending 12 and 6 o'clock with respect to the malleus handle and inferior canal wall to maximize the canal skin flap. The 12 and 6 o'clock incision was extended laterally. This was followed by a postauricular incision. A Palva flap was made over the periosteum, elevated and connected with the previous transcanal incision. Temporalis fascia was harvested in the standard fashion via a postauricular incision if a tympanoplasty was performed. A mastoidectomy was then performed with identification of the tegmen, sigmoid sinus, and antrum. Excluding one case in which an ossicular prosthesis was placed, the pars tensa and ossicles were left intact and therefore the procedure was completed similar to a modified Bondy procedure. A full exenteration of mastoid air cells was performed. The mastoid cavities remained small due to arrested pneumatization and did not require use of bone pate. A meatoplasty was completed by extending the two previously performed canal cuts, which were stented open with sutures. The authors did not attempt an endaural approach for the meatoplasty although this would be a reasonable option. A mastoidectomy or bony canalplasty can also be achieved via the endaural approach, however its superior orientation can pose a challenge to remove sufficient cartilage inferiorly to create a wide meatoplasty. Available EAC skin was used when available to cover the mastoid bone. The antral and sinodural angle region were left uncovered, with a minimal amount of exposed bone left to undergo epithelization from surrounding skin. Gelfoam was placed in the newly enlarged meatus. Patients wore a mastoid dressing for 24 h after surgery.

3. Results

The mean follow-up was 4.9 years. Seven (54%) children underwent bilateral canal wall down mastoidectomies. Six of the twenty ears had tympanoplasties at the time of mastoidectomy, seven had myringotomy with tympanostomy tube placement and one patient had an ossiculoplasty with prosthesis. Patient 9 had bilateral middle ear cholesteatomas with post-operative resolution of disease. Patient 12 who had undergone the CWD procedure had restenosis at four weeks following the surgery due to a post-operative infection. He underwent revision surgery and had a patent canal with improved visualization of the tympanic membrane at the 9-month follow-up visit. Thirteen ears had preoperative PTA with an average threshold of 47.4 dB and post-operative average PTA of 39.3. The mean threshold improved 6.2 dB (SD 16.4 dB). Nineteen preoperative computed tomographies were available and used to document the EAC diameter with a mean of 2.3 mm (SD 0.8 mm).

Patient 2 formed a retracted TM and cholesteatoma 53 months after surgery and subsequently underwent a transcanal tympanoplasty with stabilization with cartilage and was free of disease at the last clinic visit. Patient 5 with a failed previous canaloplasty from conventional atresiaplasty had a patent canal at follow-up (64 months) after canal wall down mastoidectomy. Eleven children had recurrent or chronic otorrhea, of which 91% had decreased episodes or resolution of infection. All children exhibited EAC stenosis that hindered the physical exam; nine (69%) patients were extremely uncooperative, of which all had improved ease of office examination after the procedure. The postoperative exam allowed visualization of the tympanic membrane with a 4 mm or wider speculum. There were no major complications such as facial nerve injury or cerebrospinal fluid leak.

4. Discussion

Canal wall down mastoidectomy entails removal of the posterior bony canal and exteriorization of the mastoid. The ideal CWD cavity is effectively an enlarged ear canal that is wide laterally and cones down to the tympanic membrane and middle ear as seen in patient 13 (Fig. 1). The technique can serve to not only create a widely patent canal and meatus but also addresses the chronically draining ear. A mastoidectomy potentially decreases middle ear fluid by improving communication between the aditus ad antrum and middle ear.

Congenital aural stenosis can exist as an independent anomaly but more frequently occurs in syndromic children. It is often associated with microtia and children with Down syndrome, Treacher Collins and Branchio-Oto-Renal syndrome [5]. In our cohort, all patients had syndromes, with more than half having Trisomy 21. These children are often the most uncooperative to examine in the office; the presence of craniofacial anomalies and Eustachian tube dysfunction often require regular ventilation tube monitoring for obstruction or extrusion. Syndromic patients are at risk for developing chronic ear disease and given their inability to report symptoms, an accurate exam is essential to evaluate for otitis media and formation of cholesteatoma [8].

The prevalence of canal cholesteatoma in patients with congenital aural stenosis ranges from 20 to 48% [3,9,10]. Nearly half of patients with a canal diameter of less than 4 mm can develop canal cholesteatoma by the age of 20 years [11]. The average preoperative diameter based on CT imaging was 2.3 mm in our cohort and the postoperative EAC accommodated a 4 mm or wider speculum. One child in our cohort had bilateral cholesteatomas that were removed at the time of surgery and has had no evidence of recurrence at 4.3 and 9.3 years from surgery. Unfortunately, one