International Journal of Pediatric Otorhinolaryngology 106 (2018) 80–84

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Challenges and outcomes of cholesteatoma management in children with Down syndrome☆

Saied Ghadersohi[b], Bharat Bhushan[a,b], Kathleen R. Billings[a,b,*]

[a] Ann & Robert H. Lurie Children's Hospital of Chicago, Division of Pediatric Otolaryngology-Head and Neck Surgery, Chicago, IL, United States
[b] Northwestern University Feinberg School of Medicine, Department of Otolaryngology-Head and Neck Surgery, Chicago, IL, United States

ARTICLE INFO

Keywords:
Cholesteatoma
Pediatric cholesteatoma
Down syndrome
Tympanomastoidectomy

ABSTRACT

Introduction: The high incidence of chronic otitis media with effusion and Eustachian tube dysfunction in children with Down syndrome (DS) may predispose them to cholesteatoma formation. Establishing the diagnosis, choosing the appropriate operative intervention, and post-operative care can be challenging.
Objective: To describe management strategies for cholesteatoma diagnosis, surgical treatment, and post-operative management in children with Down syndrome.
Methods: Retrospective case series of 14 patients (17 total ears) with Down syndrome diagnosed with cholesteatoma over a 9-year period.
Results: A total of 14 patients with cholesteatoma (3 with bilateral disease) were analyzed. Thirteen ears (76.5%) had ≥2 tympanostomy tubes insertions prior to cholesteatoma diagnosis, and otorrhea and hearing loss were the most common presenting symptoms. Common pre-operative CT scan findings included mastoid sclerosis and ossicular erosion. The average age at first surgery was 9.8 years, and the average follow-up was 4.3 years. For acquired cholesteatoma, most ears were managed with canal wall up (CWU) approaches, but ultimately 6/15 (40.0%) required canal wall down (CWD) approaches. Postoperatively, 3 (20.0%) ears developed new tympanic membrane retraction pockets, but no recurrent cholesteatoma. Four (26.7%) ears developed recurrent disease, and 3 (20.0%) had residual disease at secondary procedures. Ossiculoplasty was performed in 4 ears. Twelve (70.6%) ears were rehabilitated with hearing aids or FM systems.
Conclusions: The diagnosis of cholesteatoma in Down syndrome was associated with otorrhea, hearing loss, and CT scan findings of ossicular erosion and mastoid sclerosis. Most cases were managed with CWU surgical approaches. Hearing aid use was common post-operatively.

1. Introduction

The prevalence of chronic otitis media with effusion (COME) in children with down syndrome (DS) has been reported to be 93% by age 1 and 68% by age 5 [1,2]. Eustachian tube (ET) dysfunction, impairing the ability of the middle ear to clear fluid and equalize middle ear pressure, is thought to be more severe and prolonged in children with DS. This is likely due to multiple factors including craniofacial abnormalities, reduced density of ET cartilage predisposing to collapse, and generalized hypotonia affecting palatal muscles that open the ET [2,3]. ET dysfunction predisposes DS children to fluctuating and persistent conductive hearing loss due to middle ear effusion. It is estimated that chronic otitis media with effusion (COME) with conductive loss occurs in 80% of children with DS [4]. Placement of tympanostomy tubes (TT), is standard treatment for COME and has been demonstrated to improve hearing levels in 93% of children with DS by one year after TT placement [5]. Given the propensity and persistence of COME in DS, some affected children will require repeated TT insertions. DS children requiring 3 or more sets of TT are reported to have increased rates of chronic perforation, retraction pockets, atelectasis, and cholesteatoma [4].

Although the exact incidence of cholesteatoma in children with DS is unknown, it has been suggested that acquired cholesteatoma may occur more commonly and may be more extensive at time of diagnosis than in children without multiple risk factors for ET dysfunction [6,7]. Ear examinations in those with DS can be more challenging for clinicians due to narrow external auditory canals and poor patient cooperation, potentially delaying diagnosis. Surgery to excise

☆ Presented as a poster at the American Academy of Otolaryngology-Head and Neck Surgery Annual Meeting, September 2017, Chicago, IL.

* Corresponding author. Division of Otolaryngology-Head and Neck Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, 225 East Chicago Ave, Box #25, Chicago, IL 60611, United States.
E-mail address: kbillings@luriechildrens.org (K.R. Billings).

https://doi.org/10.1016/j.ijporl.2018.01.016
Received 7 December 2017; Accepted 17 January 2018
Available online 02 February 2018
0165-5876/

cholesteatomas may also be difficult due to narrow ear canals and poorly developed, sclerotic mastoid anatomy. The goal of this retrospective review is to analyze our experience with cholesteatoma management in children with DS seen at a tertiary care children's hospital over the last decade. The analysis will focus on diagnostic challenges and findings, surgical challenges and outcomes, recurrence rates, post-operative hearing outcomes, and complications associated with the cholesteatoma or surgical interventions.

2. Methods

This study was approved by the Institutional Review Board at Ann & Robert H. Lurie Children's Hospital of Chicago (IRB# 2017-730). This is a retrospective case series of children age 1–18 years, treated for cholesteatoma between January 1, 2008–May 31, 2017. An electronic medical record search of patients with the *Internal Classification of Disease, Ninth or Tenth Revision, Clinical Modification* (ICD9/10-CM) diagnosis codes 758.0/Q90.9 for Down syndrome and cholesteatoma 385.32, 385.30/H71.90–93, H71.00, H71.23 was completed through Bio Integration Suite and Clarity Databases. A total of 23 patients met the search criteria. Seven patients were excluded because they did not have confirmed diagnosis of cholesteatoma, and two were lost to follow-up prior to undergoing surgical management. A total of 14 patients were identified with cholesteatoma involving the middle ear and/or mastoid who were surgically managed at our hospital. Three patients had bilateral disease.

The electronic medical records were reviewed, and data recorded including gender, age at first diagnosis/surgery, affected side, presenting signs and symptoms, and prior history of and number of tympanostomy tube (TT) placements. Computed tomography (CT) findings were evaluated including overall mastoid pneumatization, extent of opacification, scutal erosion, ossicular erosion, facial nerve exposure, and tegmen erosion.

The type of surgical procedure (canal wall up or canal wall down mastoidectomy, endoscopic transcanal), and the intraoperative findings including cholesteatoma location, involvement of the ossicular chain, facial nerve, and/or tegmen were noted. Surgical complications (i.e., cerebral spinal fluid leak) were noted. Surgical outcomes including presence of new retraction pockets, persistent otorrhea, recurrent disease, and/or residual disease were recorded. Recurrence, defined as a new retraction pocket cholesteatoma, or residual disease, defined as cholesteatoma detected in the middle ear/mastoid at the same site as previous surgery, was noted in those having secondary procedures. The total number of surgeries needed to treat disease, including eventual need for a canal wall down (CWD) procedure, was recorded.

Post-operative pure tone averages (PTA) were recorded when available. Sound field (SF) or auditory brainstem response (ABR) testing results were noted. The incidence of auditory amplification postoperatively was noted.

2.1. Statistics

Demographic and clinical characteristics were reported as frequencies and percentages for categorical variables. Continuous variables are presented as means ± standard deviations. Significance was determined at $p < 0.05$. All statistical analyses were performed using Stata 14.1 (Statacorp, College Station, TX).

3. Results

3.1. Patient characteristics

Fourteen DS patients (17 ears) with cholesteatoma were analyzed. One patient had bilateral congenital cholesteatomas, and the remaining 15 were acquired. Characteristics of cholesteatoma patients are shown on Table 1. The average age at initial cholesteatoma surgery was 9.8 ± 4.3 years. Most patients (n = 13, 76.5%) had a history of ≥2 sets of TTs prior to the diagnosis of their cholesteatoma, and 8 (47.1%) had ≥3 sets placed (Table 1).

Table 1
Characteristics features of Down syndrome patients diagnosed with cholesteatoma.

Characteristic Features	n (%)
Gender	
Male	11 (78.6)
Female	2 (21.4)
Ear Affected	
Right	7 (41.2)
Left	10 (58.8)
Presenting Symptoms	
Otorrhea	10 (58.8)
Hearing Loss	7 (41.2)
Facial Nerve Paresis	1 (5.9)
Number of Prior TT Insertions	
≥3 TTs placed	8 (47.1)
≥2 TTs placed	13 (76.5)

TT, tympanostomy tube.

3.2. Diagnosis

The diagnosis of cholesteatoma was made on office-based otoscopic examination in 10 (58.8%) ears. The remaining 7 cholesteatomas (41.2%) were diagnosed in the operating room, either during a scheduled TT placement or during otoscopic exam in the operating room (done due to poor tolerance for exam in the clinical setting). The common presenting symptoms of otorrhea and hearing loss are outlined on Table 1. One patient presented with unilateral facial paresis, and a dehiscent facial nerve was noted at surgery. Preoperative CT reports were available in 15 (88.2%) cases. Poor mastoid pneumatization, complete opacification of the mastoid, and ossicular erosion were the most common findings seen on CT scanning (Table 2, Fig. 1).

3.3. Surgical intervention

Of the 15 cases of acquired cholesteatoma (Table 3), 11 (73.3%) had a CWU procedure, 2 (13.3%) were managed via a transcanal endoscopic approach (disease was not extending beyond the antrum), and 2 (13.3%) had a primary CWD procedure. One CWD tympanomastoidectomy, in the patient presenting with facial paresis, included facial nerve decompression, and the other was a radical tympanomastoidectomy with ear canal closure and obliteration due to extensive cholesteatoma with tegmen erosion (a cerebrospinal fluid leak occurred on cholesteatoma resection). Neither of these children developed a recurrence. All 15 (100.0%) acquired cholesteatomas involved the mesotympanum, 10 (66.7%) the epitympanum, and 8 (53.3%) extended into the antrum. Thirteen (86.7%) ears had associated ossicular erosion. The incidence of post-operative otorrhea and retraction pocket formation

Table 2
Pre-operative computed tomography (CT) scans findings for 15 of the Down syndrome patients diagnosed with cholesteatoma.

CT findings	n (%)
Mastoid Sclerosis	6 (40.0)
Opacification	
Diffuse	9 (60.0)
Focal	6 (40.0)
Erosion	
Scutum	5 (33.3)
Ossicular	9 (60.0)
Facial Nerve Dehiscence	3 (20.0)
Tegmen	2 (13.3)