The Endoscopic Management of Congenital Cholesteatoma

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KEYWORDS

• Endoscopic ear surgery • Transcanal • Congenital cholesteatoma • Cholesteatoma • Pediatric otology

KEY POINTS

- Transcanal endoscopic ear surgery is particularly suited for the management of congenital cholesteatoma given the enhanced visualization of protympanic and epitympanic recesses, areas that are more challenging to visualize using a microscopic-assisted transcanal approach.
- Congenital cholesteatoma is primarily a pediatric disease, with an average age of diagnosis of 5 years to 6 years. In adults, congenital cholesteatoma presents at an advanced stage.
- Although the typical presentation of congenital cholesteatoma is a pearly white mass seen behind the anterosuperior aspect of the tympanic membrane, a majority of cases present with extension to multiple quadrants of the middle ear.
- When congenital cholesteatoma is contained within a sac-like structure, it is more likely to be entirely eradicated in a single-stage surgery. Infiltrative disease warrants a second-look procedure, and a transmastoid, endoscopic-assisted approach can be utilized when mastoid extension is seen.
- Overall outcomes of transcanal endoscopic ear surgery for congenital cholesteatoma have been shown to be similar to traditional microscopic ear surgery in terms of risk of residual disease.

INTRODUCTION

As technological advancements and novel applications of endoscopy have proliferated over the past decade, particular interest has been placed in the role of endoscopy

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in middle ear surgery. Surgical management of congenital cholesteatoma is especially suited for an endoscopic approach, because congenital cholesteatoma is located within the middle ear space in most cases. This article presents a review of the etiology and clinical aspects of congenital cholesteatoma as well as a surgical guide to the endoscopic management of this primarily pediatric disease.

DEFINITION

Congenital cholesteatoma by definition consists of a sac-like mass of squamous epithelium that has formed in the middle ear space under an intact and normal tympanic membrane with no prior history of middle ear disease or perforation.[1] Previous bouts of otitis media or middle ear effusions do not constitute criteria for exclusion.[2]

Congenital cholesteatoma remains a rare disease, estimated to account for 2% to 5% of all cholesteatomas.[3] In the pediatric population, congenital cholesteatoma has an estimated incidence of 0.12 per 100,000 children. The average age of diagnosis of congenital cholesteatoma is 5 years to 6 years.[4] Increased awareness by pediatricians, implemented preschool screenings, and new clinical tools, such as otoendoscopy, have helped with earlier detection. In a large retrospective series over 24 years, 73% of surgical patients were 15 years old or younger.[5]

Although congenital cholesteatoma is considered a pediatric disease, it can be diagnosed in the adult population. The literature on adult cases of congenital cholesteatoma is limited. Misale and colleagues[6] report on 6 adult cases of congenital cholesteatoma. In their study, most cases had multiple quadrants of tympanic membrane involved secondary to the long duration of symptoms and advanced lesion at presentation. Another study by Doyle and Luxford[7] included patients ranging from 1 years to 59 years of age with congenital cholesteatoma, of whom 13.5% were over the age of 18 at the time of diagnosis.

Congenital cholesteatoma most commonly presents as a pearly white ear mass, most usually in the anterosuperior quadrant of the middle ear and behind an intact and healthy tympanic membrane. The majority of congenital cholesteatoma, however, involves multiple quadrants of the tympanic membrane at presentation due to a delay in diagnosis. One large retrospective study reported disease limited to the anterosuperior quadrant in only 29% of cases; 23% of cases involved the posterosuperior quadrant, and 46% involved both anterior and posterior superior quadrants.[8]

ETIOLOGY

The most widely accepted theory on congenital cholesteatoma formation is the nonresorption of embryonal epithelial rests. This is based on observations of epidermal structures, similar in location and composition, found in the middle ear space of the human fetus.[9] Derivative structures of the first branchial arch are the upper part of the malleus and incus, tensor tympani muscle, and tendon. The second arch derivatives are the facial and chorda tympani nerve, superstructure of the stapes and lower part of the incus and malleus. Clinical observation that congenital cholesteatoma appears at the first and second branchial junction rather than in a random location explains their origin as migration errors of epithelial remnants as well as their location in close proximity of the cochleariform process and tensor tympani tendon.[10]

CLINICAL PRESENTATION

Diagnosis of congenital cholesteatoma often is made after the incidental finding of an asymptomatic small pearly white mass limited to the anterior superior quadrant of the

middle ear behind an intact and healthy tympanic membrane. In such cases, the mass does not directly involve the ossicular chain, thus hearing loss is not an initial presenting sign. As congenital cholesteatoma enlarges, the diagnosis may not necessarily become more obvious due to intact drum. Congenital cholesteatoma may be mistaken for a large plaque of myringosclerosis. Although myringosclerosis has sharp, irregular edges, cholesteatoma presents as a white mass with smooth, curved, and well-defined edges. Cholesteatoma also can be mistaken for a mucopurulent effusion if it has grown to involve all quadrants of the tympanic membrane. As congenital cholesteatoma progresses and involves the ossicular chain, then conductive hearing loss can become a more obvious presenting symptom. Other times, congenital cholesteatoma may be present in concomitance with serous otitis media, with images showing a completely opacified middle ear and mastoid. The mass, due to its typical expansion in the anterior superior aspect of the middle ear space, may cause early occlusion of the eustachian tube and obstruction of the tympanic isthmus, resulting in a middle ear and mastoid effusion.[11]

Occasionally, congenital cholesteatoma incidentally is discovered intraoperatively. A white mass may be recognized behind an intact tympanic membrane during myringotomy for recurrent acute otitis media or chronic effusion and in some cases the matrix is violated during the procedure, opening the sac and revealing squamous debris. A myringotomy also could allow epithelial migration to the undersurface of the tympanic membrane. In some cases, a tube may be advantageous in resolving an effusion in a patient with suspected congenital cholesteatoma. On CT imaging, the presence of an effusion in concomitance with a mass may obscure the true extent of disease because of its similar density to soft tissue, hindering surgical planning. In this instance, if the surgeon deems necessary to drain the effusion in preparation for preoperative imaging, it then is advisable to insert a tympanostomy tube, taking care to place it as far as possible from the visible margins of the cholesteatoma.

STAGING

Several staging systems have been proposed for congenital cholesteatoma. The most frequently used is the Potsic staging system that defines the extension of disease, as follows (**Table 1**).

Potsic stage and risk of residual disease are directly correlated, with increasing risk of residual disease with higher stage. In stage I disease, the risk of residual disease is approximately 5%; in stage II, 24%; stage III, approximately 44%; and stage IV, 64%.[3]

More recently, in 2017, the Japan Otological Society (JOS) introduced a new staging system for cholesteatoma, with the goal of more specifically describing borders of division between the middle ear and the mastoid and difficult access sites, including the sinus tympani and the supratubal recess.[11] In addition, a staging system for congenital cholesteatoma was proposed within the JOS staging and classification criteria for middle ear cholesteatoma (**Table 2**).[12]

Table 1
Potsic staging system for congenital cholesteatoma

Stage I	Disease confined to a single quadrant
Stage II	Cholesteatoma in multiple quadrants, but without ossicular involvement or mastoid extension
Stage III	Ossicular involvement without mastoid extension
Stage IV	Mastoid disease

Table 2
Japan Otological Society staging system for congenital cholesteatoma

Stage I	Cholesteatoma localized in the tympanic cavity
Stage Ia	Cholesteatoma confined to the anterior half of the tympanic cavity
Stage Ib	Cholesteatoma confined to the posterior half of the tympanic cavity
Stage Ic	Cholesteatoma involving both of sides of the tympanic cavity
Stage II	Cholesteatoma involving 2 or more sites
Stage III	Cholesteatoma with infratemporal complications and pathologic conditions
Stage IV	Cholesteatoma with intracranial complications

ANATOMY OF THE PEDIATRIC EAR CANAL

During childhood, the external ear canal undergoes significant structural changes. Although the tympanic membrane, middle ear, and inner ear are almost the same size in infancy as in adulthood, conversely, the pediatric external ear canal is much shorter and narrower compared with its adult size. Although the adult external ear canal usually is described as one-third cartilaginous and two-thirds osseous, the cartilaginous canal of a newborn directly abuts the tympanic ring, with little osseous component. In the first 5 years of life, there is significant lateral extension of the canal, with simultaneous lateral extension of the osseous tympanic ring, forming the osseous portion of the canal. From age 5 years to 18 years, the osseous canal doubles in length, reaching adult proportions.[13] The length of the canal in pediatric patients must be considered carefully in transcanal surgery, especially when raising a tympanomeatal flap. The flap raised is shorter in length and only a few millimeters away from the tympanic annulus. The width and height of the external auditory canal also undergo significant changes during childhood. The neonatal canal is approximately 4.4 mm to 6.3 mm in width and approximately 5.4 mm in height, whereas the canal of the adult is approximately 6.1 mm to 10.4 mm in width and 6.9 mm in height.[14] This is a n important consideration particularly in endoscopic ear surgery, because the bony portion of the canal is a limiting factor in accommodating the endoscope as well as instrumentation. In a young child with a very narrow ear canal, a 3-mm or 2.7-mm endoscope may be required for surgical approach. Larger ear canals in older children may accommodate a 4-mm telescope. Additionally, the angle of the tympanic annulus relative to the bony external ear canal is more acute in children, changing from 34° from horizontal in the infant, to 63° from horizontal in adulthood; this change is due to the growth of the temporal lobe and skull base rather than changes to middle ear morphology.[14] The more acute angle of the tympanic membrane relative to the plane of the bony external ear canal in younger children may make endoscopic or microscope visualization of the epitympanum and retrotympanum more challenging.

PREOPERATIVE EXAMINATION

Depending on the cooperation of the child, the in-office ear examination is performed under microscopy as well as endoscopy. Under microscopy, the ear canal is cleaned from cerumen. Although endoscopy offers a panoramic view of the tympanic membrane with improved visualization of the anterior tympanic rim, microscopic examination offers the possibility of inspecting selected portions of the tympanic membrane under high magnification. In the authors' office, a screen monitor with recording capability (Telepack, Storz, El Segundo, California) is used in order to obtain endoscopic

photo documentation that then is uploaded to the medical chart. This allows for the images to be reviewed prior to surgery as well as during follow-up visits. This is useful especially for educational purposes in an academic program. In addition to documenting the appearance of the tympanic membrane, the endoscopic examination is useful to assess the size of the ear canal. In young children with a very narrow ear canal, a 3-mm or a 2.7-mm endoscope may be required for surgical approach.[15] The preoperative endoscopic examination also is important to assess whether there is a prominent anterior canal wall; if the endoscope is unable to capture a view of the entire drum due to a prominent overhang, the surgical team expects greater difficulty with a transcanal approach. An angled scope can help improve visual access, but most dissection instruments do not have a secondary bend to accommodate the curved canal in tight cases.

PREOPERATIVE IMAGING

Temporal bone computed tomography (CT) imaging is obtained to determine extent of the disease in the middle ear, degree of ossicular involvement, and presence of mastoid disease. The authors believe that imaging is important to optimize surgical planning even in smaller congenital cholesteatomas confined to 1 quadrant. Preoperative CT scanning may not be as useful in cases of concomitant effusion, because opacification of the mastoid and middle ear space may mask the extent of disease. In these cases, tympanostomy tube placement may be necessary to resolve the effusion prior to imaging. Although a magnetic resonance image (MRI) with diffusion-weighted imaging may help with differentiating effusion from disease, the authors would not put a child under general anesthesia to obtain a preoperative MRI unless there was a concern for petrous apex disease or intracranial involvement.

SURGICAL MANAGEMENT AND ENDOSCOPIC ADVANTAGES

The primary goals of surgery for cholesteatoma are eradication of disease and preservation of hearing. Despite the fact that congenital cholesteatoma is a benign lesion, prompt surgical intervention is advocated. The disease expands over time, progressively involving multiple compartments of the tympanic cavity, the ossicular chain, with potential to eventually spread to the mastoid. As disease expands and erodes the middle ear anatomy, hearing preservation and disease eradication become more difficult. The introduction of endoscopy in ear surgery has provided improved visualization of previously inaccessible areas in the tympanic cavity and the epitympanic space. Endoscopy also offers advantages in mastoidectomy; after removal of disease, endoscopy grants the ability to inspect areas in the mastoid that are difficult to visualize under microscopy alone.

Without endoscopy, most congenital cholesteatomas can be removed using 2-handed transcanal microscopy with a similarly minimally invasive approach, which brings into question the necessity of endoscopy. Although congenital cholesteatoma of small size can be removed easily under microscopy, full exposure under microscopy becomes difficult when disease is present anterior to the manubrium of the malleus. The manubrium of the malleus, because of its slight tilt toward the promontory, obstructs the visualization of the cochleariform process and tensor tympani tendon. This explains why, under microscopy, surgical maneuvers, such as transection of the tensor tympani tendon to lift the malleus or removal of the malleus, are performed to improve the visualization of the tympanic cavity. Utilization of a 30° angle endoscope and degloving of the manubrium enables a wide-angle view of the anterior

mesotympanic and protympanic recesses without any manipulation of the malleus or transection of the tendon.

SURGICAL CONSIDERATIONS

The surgical management of congenital cholesteatoma is different from that of primary acquired cholesteatoma due to the underlying mechanism of disease formation. Patients with congenital cholesteatoma typically have normal middle ear anatomy, an intact tympanic membrane, and normal eustachian tube function. Therefore, when congenital cholesteatoma can be removed while keeping the tympanic membrane intact, the ear is expected to heal uneventfully, with an intact transparent membrane, which also allows for surveillance of residual disease. Because of the normal function of the eustachian tube and middle ear, the occurrence of postoperative tympanic or attic retraction is not expected, which is an unfortunate frequent occurrence after surgery for acquired cholesteatoma. Nevertheless, postoperative atelectasis and attic retraction sometimes are observed after surgery for congenital cholesteatoma, leading to secondary development of acquired cholesteatoma from pars tensa or pars flaccida retraction. This unusual circumstance can be secondary to poorer mastoid ventilation. Studies have shown that the mastoid cells in ears of children with congenital cholesteatoma are poorly pneumatized compared with those of children without middle ear disease, and the degree of pneumatization is significantly poorer than in the opposite side.[16] Decreased gas exchange in a reduced mastoid or sclerotic inflammatory change of the middle ear mucosa after surgery could be responsible for a tendency for delayed tympanic and attic retraction.

The structure of the cholesteatoma is an important determining factor for prognosis and reconstruction. Congenital cholesteatoma, due to its cystic origin from epithelial remnants, is more likely to be collected in a sac-like structure that facilitates complete excision, thus favoring consideration for an immediate ossicular chain reconstruction (OCR) during primary surgery. Conversely, primary acquired cholesteatoma frequently is disorganized in structure with loose or adherent matrix and is more likely to warrant a second-look surgery with staged ossicular reconstruction.

The surgical approach for congenital cholesteatoma is dependent on the extent of the disease. The transcanal approach provides adequate exposure for most congenital cholesteatoma confined to the middle ear, which facilitates the use of endoscopy. When imaging demonstrates mastoid opacification concerning for extent of disease to the mastoid, preoperative discussion about the possible need for a posterior auricular approach with mastoidectomy is discussed with the family.

CHOICE OF ENDOSCOPES

A 3-mm diameter (14-cm length) rigid Hopkins rod telescope is suitable for the majority of pediatric endoscopic cases. Older children with a larger ear canal may accommodate 4-mm endoscopes, with the advantage of providing an enhanced wide-angle view and improved illumination but come at the expense of restricted space with dissection instruments. In very young children or those with especially narrow external canals, a 2.7-mm pediatric rigid sinus endoscope can be utilized. Although this scope may be necessary to access a smaller canal, the 2.7-mm endoscope is disadvantaged by a longer, more fragile shaft (17.5 cm) and a smaller diameter, resulting in reduced wide field view of the surgical area. The authors advocate starting with a 0° 3-mm endoscope. In cases of a prominent anterior canal wall, a 30° 3-mm endoscope may be necessary to improve anterior visualization.

SURGICAL APPROACH

Endoscopic Approach to Congenital Cholesteatoma Limited to the Anterior Superior Quadrant (Stage I)

Endoscopy is particularly advantageous in the approach of a congenital cholesteatoma limited to the anterior superior quadrant. The superior visualization of the anterior rim of the tympanic membrane and protympanic area, compared with microscopy, enables removal of the sac with intact preservation of the ossicular chain. Congenital cholesteatoma limited to the anterosuperior quadrant can be approached in 2 ways: with an anterior tympanomeatal flap designed to expose the anterior portion of the tympanic space and with a posterior tympanomeatal flap that exposes the entire mesotympanum and protympanum after degloving of the manubrium. The latter approach technically is more straightforward and provides improved surgical access to disease.

Anterior tympanomeatal exposure

The anterior tympanomeatal incision is made starting at the anterior rim, anterior to the notch of Rivinus. The incision is made approximately 6 mm away from the tympanic annulus and extended from the 3-o'clock to 9-o'clock position (Fig. 1A). The flap is dissected off the anterior tympanic annulus to expose the anterior aspect of the tympanic cavity (Fig. 1B). The tympanic membrane is pulled back and detached from the neck and handle of the malleus. Full surgical exposure of the sac is obtained by dissecting the fibrous attachment of the tympanic membrane from the manubrium of the malleus, while maintaining attachment to the umbo (Fig. 1C). The reflection of the tympanic membrane exposes the small cholesteatoma and allows retrieval of the sac without exposing the ossicular chain or disrupting the posterior aspect of the tympanic membrane (Fig. 1D). This exposure is suitable for small, limited masses, or pearls, that

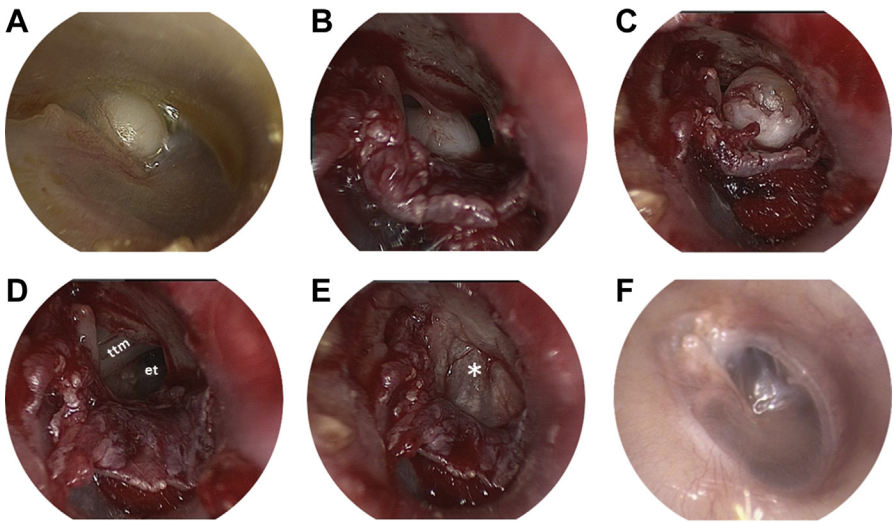

Fig. 1. A 5-year-old girl with congenital cholesteatoma of the right ear. (*A*) Pearly white mass located to the anterior superior quadrant of the tympani muscle. (*B*) Anterior tympanomeatal incision, anterior to the lateral process of the malleus. (*C*) Dissection of the tympanic membrane off the manubrium for surgical exposure of the mass. (*D*) Inspection of the protympanic space after cholesteatoma resection. (*E*) A graft of porcine extracellular mucosa is applied as an underlay. (*F*) Appearance 3.5 years postoperatively. et, eustachian tube orifice; ttm, tensor tympani muscle; *, porcine extracellular mucosal graft.

do not extend past the manubrium of the malleus and do not entirely fill the superior anterior quadrant. One disadvantage of elevating an entirely anteriorly based flap is that dissection may be challenging because the tympanic membrane can be firmly adherent to the tympanic annulus. Other investigators have performed a transtympanic approach with tympanotomy for very small congenital cholesteatoma.[17]

Posterior tympanomeatal exposure

A posterior tympanomeatal incision is made from the 1-o'clock position, anterior to the neck of the malleus, extending posteriorly to the 6-o'clock position, 6 mm to 8 mm in length from the tympanic annulus. Once the tympanic annulus is lifted, the middle ear is entered, and the tympanic membrane is dissected off the manubrium of the malleus to expose the anterior aspect of the tympanic membrane. Endoscopy provides a high degree of magnification, improving visualization of the fibrous attachment of the tympanic membrane over the malleus manubrium. Dissection of this tough fibrous attachment is performed with the sharp edge of a sickle knife or a joint knife to gently dissect the fibrous attachment of the tympanic membrane off the periosteum of the malleus manubrium. The tympanic membrane then is detached from the manubrium, except for a small portion that is left attached to the umbo to avoid postoperative lateralization. Ensuring complete removal of the cholesteatoma from the malleus, tensor tympani tendon, and cochleariform process is crucial because these areas have high risk of harboring residual disease. Curved, angled instruments, such as a Thomassin attic dissector and a Rosen needle, are helpful in removing disease from these structures. Often, the sac appears to be lodged into or protruding over the eustachian tube opening. An endoscopic curved suction is helpful in gently pulling and extracting the sac away from the opening of the eustachian tube.

After sac removal, a 30° or 45° 3-mm scope is used to inspect the protympanic space to assure that no residual disease is left behind. The anatomic structures in this area, the hemicanal of the tensor tympani muscle, the eustachian tube orifice, and the subtensor recess (SR) are inspected for any residual disease (see **Fig. 1**D). The elevated tympanic membrane then is repositioned in place. If any tear occurred during the elevation or the elevated tympanic membrane is particularly thin and fragile, an underlay graft is placed. The authors favor tragal or conchal perichondrium or heterologous porcine extracellular mucosa for its thicker consistency and postoperative transparent appearance (**Fig. 1**E). By keeping the tympanic membrane attached to the umbo, preserving the tympanic rim attachment of the anterior inferior aspect of the tympanic membrane, postoperative blunting is avoided (**Fig. 1**F).

Endoscopic Approach to Congenital Cholesteatoma Involving Multiple Quadrants Without Ossicular Erosion (Stage II)

A congenital cholesteatoma involving multiple quadrants may present as a mass medial to the malleus in a dumbbell shape (**Fig. 2**A). The bulk of the mass extends into the attic and mesotympanic areas, with the narrow portion of the dumbbell medial to the malleus (**Fig. 2**B). This is a challenging situation, because the ossicular chain is in continuity, but the mass is medial in respect to the head of the malleus (**Fig. 2**C). Consequently, it is necessary to disrupt the ossicular chain in order to extract the mass (**Fig. 2**D). In this case, in order to fully expose the attic, the incus is separated from the stapes, and removed, while the head of the malleus is transected at the neck with a malleus nipper and removed (see **Fig. 2**D). Although it may be tempting to remove the entire malleus with the aim of ensuring complete eradication of the disease, the removal of the entire malleus has profound consequences on hearing reconstruction. The tympanic membrane, no longer anchored to the malleus, lateralizes with

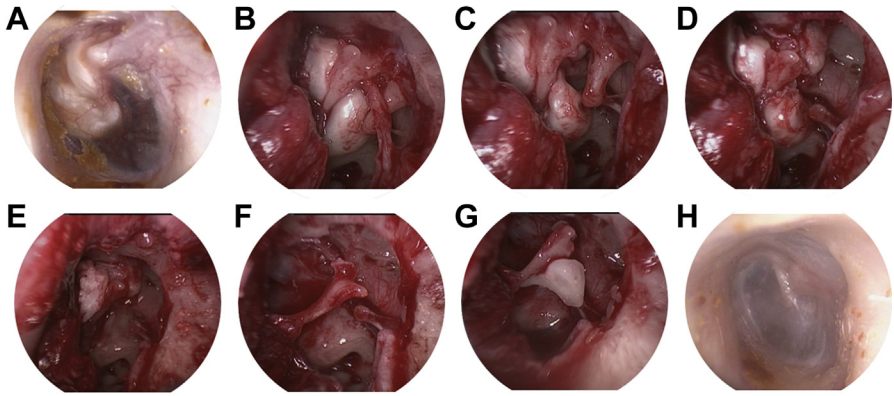

Fig. 2. A 6-year-old boy with congenital cholesteatoma of the left ear. (*A*) Mass involves multiple quadrants of the tympanic membrane. (*B*) Cholesteatoma conformed as a dumbbell-shaped cyst with the narrow portion kinked medial to the malleus. (*C*) Gentle dissection of the mass reveals an intact ossicular chain. (*D*) Incus and malleus head are removed to expose the medial component of the mass. (*E*) Residual mass in protympanic and anterior attic space. (*F*) Attic inspection with 30° endoscopy. (*G*) Autologous incus interposed between stapes capitulum and malleus manubrium. (*H*) Well-healed tympanic membrane 18 months postoperatively.

time. As the tympanic membrane lateralizes, it becomes discontinuous with any form of ossicular reconstruction (prosthesis or autologous incus) with subsequent postoperative conductive hearing loss. To avoid this, the manubrium of the malleus is left attached to the tensor tympani tendon and then lifted to inspect the area of the cochleariform process Endoscopy provides improved visualization of the attic, allowing full inspection for residual disease (Fig. 2F). The incus then is sculpted with a 2-mm diamond burr, interposed between the stapes capitulum, and notched under the malleus; this favors a more physiologic OCR than that of a partial ossicular replacement prosthesis (Fig. 2G).

Endoscopic Approach to Congenital Cholesteatoma Involving Multiple Quadrants with Ossicular Involvement (Stage III)

When a congenital cholesteatoma occupies a large portion of the middle ear space, the ossicles are enveloped in the large mass, which obscures the degree of erosion (Fig. 3A). A posterior incision is made, starting just anterior to the notch of Rivinus, and extending to the inferior aspect of the canal. The tympanic membrane is reflected, and exposure of the middle ear reveals either one large sac or multiple sac-like formations that fill the entire middle ear space. In this particular case, the ossicles are buried under multiple sacs of disease (Fig. 3B). At this point, probing or attempting to lift the sac blindly is hazardous, because the degree of involvement with the stapes or facial nerve is unknown. The most prudent course of action is to continue to lift the tympanic membrane and identify the malleus, because this is the ossicle least likely to be eroded (see Fig. 3B). Identification of the malleus facilitates palpation of the incus, which can be performed with a blunt instrument, such as a Rosen needle or a short attic dissector. Curetting the scutum with a stapes curette helps to identify the lateral aspect of the body of the incus as well as determine the posterior extension of the cholesteatoma sac. Once the sac of disease is lifted off the mesotympanum and the incus is identified, the stapes and the degree of erosion can be evaluated. If erosion of the

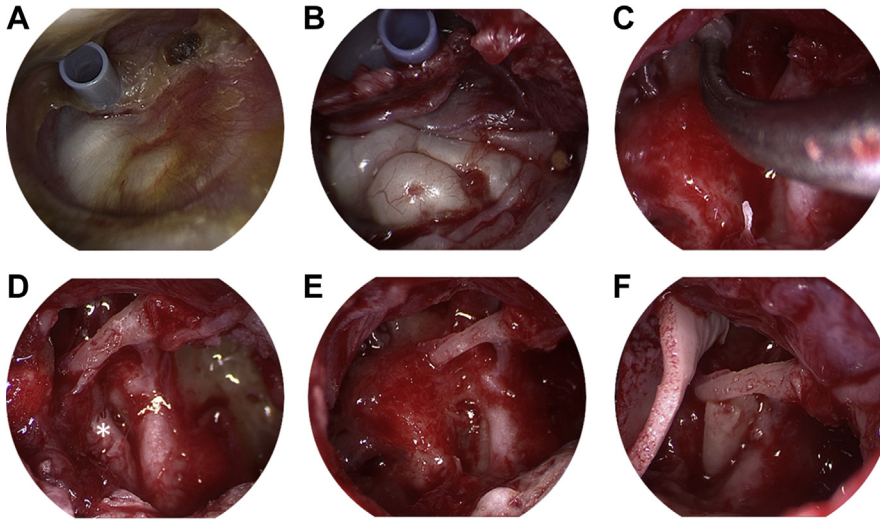

Fig. 3. A 3-year-old boy with congenital cholesteatoma of the left ear. (*A*) Cholesteatoma involves multiple quadrants of the tympanic membrane and a tube had been placed by the referring physician. (*B*) Multiple sacs of cholesteatoma obscure the visualization of incus and stapes. The malleus is identified as first anatomic landmark in the middle ear space. (*C*) A curved suction is utilized to extract a portion of cholesteatoma lodged into the eustachian tube. (*D*) A portion of cholesteatoma deeply lodged over the footplate (*) is removed last. (*E*) After removal of cholesteatoma, the footplate is exposed as stapes superstructure was completely eroded. (*F*) Autologous incus interposition graft from stapes footplate to malleus.

stapes superstructure is present, cholesteatoma matrix can be adherent to the footplate as well as over the tympanic segment of the facial nerve (Fig. 3D). The high magnification provided by endoscopy is useful particularly when working delicately in this area. Gentle elevation of matrix from the footplate is performed with a blunt instrument; extreme care is taken to avoid fracturing a thinned footplate or exposing a perilymphatic fistula under an eroded footplate. If necessary, a wider atticotomy is performed with a stapes curette or with a piezoelectric drill, which allows sharp bone curettage with an oscillating blade under a constant flow of saline, providing visualization under water.

Combined Approach for Congenital Cholesteatoma with Mastoid Involvement (Stage IV)

When preoperative imaging shows mastoid opacification, extension of disease to the mastoid should be considered. Congenital cholesteatoma with mastoid extension is present in approximately 13% of all cases.[3] Radiologic signs suggestive of disease extension to the mastoid include loss of air cell trabeculation, blunting of the scutum, and the presence of soft tissue mass with rounded edges in the mastoid. Even when a mastoidectomy appears inevitable, there are several advantages to starting the surgery with a transcanal endoscopic approach. Primarily, a posterior auricular approach or a mastoidectomy would not be committed to until it is evident that the cholesteatoma has extended to the mastoid antrum, beyond the limits of transcanal endoscopy. Additionally, starting the surgery through a transcanal approach allows the

prioritization of work time in the middle ear and attic shortly after transcanal injection and delivery of anesthetic and vasoconstrictors.

This brings to question, how far can an atticotomy be extended to retrieve disease that extends into the aditus? The limit of transcanal endoscopy is considered the posterior aspect of the lateral semicircular canal. Although angled endoscopes may visualize the disease, lack of curved instrumentation with grasping capability frequently limits disease removal. The rising popularity of endoscopic ear surgery is inspiring the development of endoscopic ear instruments; curved suctions, angled dissectors, and flexible steering tips are being developed with the goal of improving accessibility of all areas visualized by endoscopy.

When extension of disease to the mastoid is unclear, the decision whether to perseverate with transcanal attempts at removing disease or convert to mastoidectomy may be challenging. If disease still is visualized with angled endoscopy but cannot be removed entirely with curved suctions and angled dissectors, the authors favor avoiding aggressive scutum removal to reduce time and frustration in the operating room and commence with canal-up mastoidectomy. The authors' experience is that cholesteatoma extending past the prominence of the lateral canal can only be removed through an extended atticotomy if it is collected in a well-defined sac. If cholesteatoma is disorganized with loose or infiltrative matrix, it is advisable to move forward with mastoidectomy because small remnants or pedunculated extensions of cholesteatoma may be out of endoscopic view. After completion of a limited canal-up mastoidectomy, 0° and 30° endoscopes are utilized to inspect the recesses of the middle ear and mastoid that are poorly visualized transcanal endoscopically as well as transmastoid microscopically (such as the medial aspect of the posterior auditory canal and the aditus). Using a moist gauze draped over the edge of the mastoid cavity stabilizes the telescope and utilizing a curved suction in the other hand completes débridement of the cholesteatoma.

PRIMARY VERSUS SECONDARY OSSICULAR CHAIN RECONSTRUCTION

It has been shown that resection of an intact sac of congenital cholesteatoma is associated with significantly reduced risk of residual disease.[3] This is due to congenital cholesteatoma's tendency to be collected in a well-defined sac, making it more likely to be completely excised. Therefore, cases in which congenital cholesteatoma is resected are more amenable to primary OCR compared with acquired cholesteatoma. There are certain situations, however, in which primary OCR after congenital cholesteatoma resection is not recommended. Primary OCR is inadvisable when there is concern about possible residual squamous debris in challenging areas, such as the anterior crus, stapes footplate, facial nerve, sinus tympani, and subpyramidal areas. In these circumstances, second-look surgery with secondary OCR should be performed. The incus can be banked in the attic to be used for reconstruction at a later time. In cases of small congenital cholesteatomas resected in entirety, with clear and well-defined borders, primary OCR is advantageous in providing the child with an immediate form of hearing rehabilitation compared with waiting for a planned, staged OCR.

OUTCOMES

Risk of residual disease directly correlates with higher stage. In a review of 82 cases of microscopic surgery for congenital cholesteatoma, cases were stratified according to Potsic staging: 24% of cases were stage I, 21% were stage II, 41% were stage III, and 13% were stage IV. Higher stage also correlated with increased preoperative hearing

loss. Residual disease was encountered in 33% of all cases, with rates of residual disease as 5% in stage 1%, 24% in stage 2%, 44% in stage 3%, and 64% in stage 4. In addition to Potsic stage, aspects of disease presentation that were significantly correlated with higher risk of residual disease include congenital cholesteatoma located medial to the malleus or incus, disease abutting the incus or stapes, disease enveloping or eroding the stapes, and disease requiring the removal of ossicles on initial surgery. Having an intact congenital cholesteatoma pearl was associated with greatly reduced rates of residual disease.[3]

Because endoscopic ear surgery is a more recent field, studies on the outcomes of endoscopic ear surgery for congenital cholesteatoma are fewer. These early, encouraging studies, however, have shown similarities in surgical outcomes in the treatment of congenital cholesteatoma using the endoscopic approach compared with using a microscopic technique. In a study of 25 children who underwent endoscopic ear surgery for congenital cholesteatoma of the middle ear, 52% were stage I, 28% were stage II, and 20% were stage III. A majority of the stage I cholesteatomas were located in the anterior superior quadrant (12/13). After an average follow-up period of 24 months $\pm$ 8.5 months, 24 of the 25 had no evidence of recurrence.[17] Additionally, James and colleagues[15] reviewed 235 ears in 220 children who had had intact canal wall surgery; 108 underwent microscopic dissection with only endoscopic inspection, and 127 underwent increasing use of endoscopes for dissection. The investigators found a 12% risk reduction in residual disease at 2.5 years when endoscopes were used for dissection, especially in the middle ear (from 22% to 11%).[15]

SUMMARY

Congenital cholesteatoma is a rare disease that comprises approximately 2% to 5% of all cases of cholesteatoma. It primarily is a pediatric disease, with peak age of diagnosis of 5 years to 6 years. Congenital cholesteatoma, as opposed to primary acquired cholesteatoma, often presents in healthy ears with no prior history of disease and with normal eustachian tube function. Transcanal endoscopic ear surgery is particularly suited for congenital cholesteatoma, because this is primarily a disease of the middle ear, and thus the majority of cases can be managed with a transcanal approach. The underlying lack of chronic ear disease in most ears with congenital cholesteatoma, combined with its tendency to be contained in a sac-like structure, increases the chances of complete removal during primary surgery. The wide-angled view provided by transcanal endoscopy allows a superior visualization of areas, such as the protympanum and attic, compared with transcanal microscopic-assisted approaches. This further reduces the risk residual disease. In both surgical and clinical practice, the use of endoscopy to document disease and postoperative results is useful not only for surgical planning and family counseling but also as a teaching tool. Initial outcomes on endoscopic management of congenital cholesteatoma show similar residual and recurrent rates compared with microscopic ear surgery. As more surgeons adopt endoscopic-assisted transcanal and transmastoid techniques to manage pediatric ear disease, larger cohort studies can be expected on outcomes of endoscopic ear surgery for congenital cholesteatoma.

DISCLOSURE

Dr M. Fina and Dr R. McCabe have no relevant conflicts of interest or financial ties to disclose.

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