Endoscopic Management of Primary Acquired Cholesteatoma

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KEYWORDS

• Primary acquired cholesteatoma • Endoscopic ear surgery • Cholesteatoma
• Middle ear surgery • Mastoidectomy • Otologic surgery • Surgical outcomes

KEY POINTS

- Endoscopic surgery for primary acquired cholesteatoma provides superior visualization of the attic and sinus tympani, the 2 sites at highest risk for residual cholesteatoma.
- In primary acquired cholesteatoma with limited attic extension, transcanal endoscopic ear surgery can allow for complete disease removal with mastoid preservation.
- For extensive cholesteatoma disease or anatomic factors such as a small mastoid with a low-lying tegmen and a prominent anterior canal wall, a canal wall down mastoidectomy may still be required to fully remove disease.
- Selective mastoid obliteration obliterates the mastoid space while preserving the middle ear space. Advantages include a smaller mastoid cavity with easier long-term care and a more cosmetic meatoplasty
- Outcome data for endoscopic ear surgery demonstrate at least equivalent rates of residual and recurrent disease, comparable hearing outcomes, but with decreased pain and shorter healing times compared to microscopic surgery.

INTRODUCTION

For the surgeon who is experienced in managing cholesteatoma with microscopic ear surgery, the transition to endoscopic surgical management may present initial challenges. Endoscopic middle ear surgery provides distinct advantages, such as visualizing traditionally challenging regions including the attic and sinus tympani.[1] The objective of this article was to provide a guide for surgeons who are transitioning to endoscopic ear surgery by describing surgical techniques, maneuvers, and reviewing surgical steps to perform safe endoscopic ear surgery and prevent complications in the management of cholesteatoma.

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PATHOGENESIS OF CHOLESTEATOMA

Cholesteatoma is a disease of the middle ear and mastoid characterized by the accumulation of keratinizing stratified squamous epithelium that results in local inflammation and destruction. The estimated annual incidence of cholesteatoma is 6 to 15 cases per 100,000 people.[2–4] There are 2 types of aural cholesteatoma: congenital and acquired. Congenital cholesteatoma originates from ectopic rests of squamous epithelium that form in the middle ear under an intact tympanic membrane. Acquired cholesteatoma originates from migration of the epithelium in the middle ear from a retraction or a perforation of the tympanic membrane. Acquired cholesteatoma comprises the vast majority of cholesteatoma cases and is further divided into primary and secondary acquired.[5] Primary acquired cholesteatoma results from tympanic membrane retraction. The retraction may involve the pars flaccida, the pars tensa, or both. Secondary acquired cholesteatoma results from a direct injury or perforation of the tympanic membrane due to infection or iatrogenic causes.

There are several theories on the pathogenesis of primary acquired cholesteatoma. These include (1) invagination, (2) basal cell hyperplasia, (3) metaplasia, and (4) epithelial invasion.[6] More recently, 2 new theories have been proposed: mucosal traction and selective epitympanic dysventilation theory. In the theory of mucosal traction, adhesions of opposing mucosal surfaces exert traction on the tympanic membrane, stimulating cytokine production and keratinocyte proliferation.[7] The theory of selective epitympanic dysventilation postulates that retraction of the pars flaccida occurs secondary to a block of ventilation pathways between the mesotympanic space and the epitympanic compartment. This results from a complete tensor fold that obstructs the epitympanic compartment while the remaining mesotympanic space is ventilated by the eustachian tube. The selective epitympanic dysventilation theory attempts to explain how epitympanic cholesteatoma can be associated with a well-ventilated and normal appearing pars tensa.[8]

Primary acquired cholesteatoma can occur at any age. Within the pediatric population, the estimated incidence of cholesteatoma is 3 cases per 100,000 individuals, with the majority of these attributed to acquired cholesteatoma.[9,10] In the pediatric acquired cholesteatoma population, the average age of presentation is approximately 10 years.[11,12] Pediatric primary acquired cholesteatoma typically has a more aggressive growth pattern compared to its adult counterpart.[13,14] This is attributed to more active keratinocyte proliferation and more rapid spread in widely pneumatized pediatric mastoids compared to a slower spread in adult osteitic mastoids with dense bone.[15]

ADVANTAGES OF THE ENDOSCOPIC APPROACH

Endoscopy provides superior visualization of portions of the middle ear that are not well-visualized with a microscope; these regions include the attic, sinus tympani, anterior epitympanic rim, protympanum, and hypotympanum.[1] Initially, endoscopes were used as an adjunct tool to inspect the middle ear and mastoid following microscopic work.[16] With improvements in optics and instrumentation, more surgeons have adopted endoscopes as the primary tool for repair of tympanic perforations and management of cholesteatoma with limited attic extension.[16,17] Outcomes for microscopic and endoscopic tympanoplasty show equivalent closure rates of 85% to 97%, while endoscopy obviates the need for a postauricular incision, resulting in less postoperative pain and morbidity.[18,19] Endoscopy is uniquely suited for management of attic cholesteatoma as it confers improved visualization of the attic and sinus tympani areas, the 2 most common sites of residual disease.[20,21] The improved endoscopic

view allows the surgeon to follow the pathway of the disease from the middle ear into the attic. This creates a tailored, less invasive approach that extends up to the limits of the disease and may preserve the ossicular chain and the mastoid.

IN-OFFICE ENDOSCOPIC EXAMINATION

Surgical planning for cholesteatoma begins in the office with a combined microscopic and endoscopic ear examination. Microscopy is used first to clean the ear and inspect the tympanic membrane under high magnification. The ear is then examined with a rigid endoscope. In the clinic we use 4-mm rigid sinus endoscopes that are readily available. Pediatric 2.7-mm sinus endoscopes are an alternative for pediatric and narrow ear canals. The endoscopic examination supplies a panoramic view of the entire tympanic membrane with improved visualization of the anterior tympanic rim and pars flaccida. Photo documentation is obtained and uploaded to the medical chart to help with surgical planning. Endoscopic assessment of the size of the ear canal, prominence of the anterior and/or posterior canal wall are all important details that are documented for preoperative surgical planning.

PREOPERATIVE IMAGING

We routinely order a computed tomography (CT) scan for surgical planning. In certain circumstances, such as cholesteatoma with significant tegmen erosion and a poorly defined dural plane, an MRI of the brain with gadolinium and diffusion-weighted images (DWI) is ordered to rule out a meningocele or encephalocele. Another indication for an MRI is concern for petrous apex extension. Some investigators have reported on the utility of obtaining a routine preoperative non–echo planar DWI MRI to differentiate cholesteatoma from mastoid fluid or granulation tissue to predict the feasibility of an exclusive transcanal endoscopic approach.[22]

IMAGING CONSIDERATIONS FOR ENDOSCOPIC MANAGEMENT OF CHOLESTEATOMA

Ear Canal

Although the size of the ear canal is assessed by physical examination, the CT scan supplies details on the size of the ear canal, tortuosity, possible exostosis or prominent scutum. Depending on these findings, the surgeon may anticipate drilling the canal to enlarge transcanal access or removing an exostosis before proceeding with a transcanal endoscopic approach. Tortuous or narrow ear canals may require smaller and angled endoscopes for access. In ears with pronounced exostosis, an exclusive transcanal endoscopic approach may not be feasible and the surgeon must discuss with the patient that a posterior auricular or an endaural incision may be required to remove the prominent exostosis and gain access to the middle ear.

Blunting of the scutum is a characteristic radiologic finding consistent with attic erosion and is pathognomonic for epitympanic cholesteatoma. However, a mesotympanic cholesteatoma may not present with scutal blunting as the pars flaccida is frequently spared.

Middle Ear Involvement

A high-resolution temporal bone CT provides detailed images of middle ear anatomy. Poorly defined ossicles, ossicular erosion, and soft tissue located medial to the ossicular chain suggest that cholesteatoma has involved the ossicles or is lodged medial to the ossicular chain. In addition, the CT scan offers details on the depth and

involvement of the sinus tympani, which is often the limiting factor in a transcanal microscopic approach. Shallower sinus tympani conformations are often amenable to an exclusive transcanal endoscopic approach, while a deeper sinus tympani may require a mastoidectomy with a retrofacial approach.[23] However, angled endoscopes may increase visualization and access to a deep sinus tympani.

Mastoid Involvement

It is essential to evaluate the extent of mastoid involvement, as most surgeons agree that cholesteatoma is endoscopically accessible as long as it does not extend beyond the dome of the horizontal semicircular canal. Mastoid opacification on CT imaging may represent cholesteatoma or trapped mucus from a blocked tympanic isthmus. Cholesteatoma is suspected if there is erosion of the mastoid trabeculae or other bony structures, or if the opacification has a discrete, rounded shape. However, the surgeon should be prepared for both possibilities, as the sensitivity and specificity of CT scan in detecting mastoid cholesteatoma when compared to intraoperative assessment has been reported to be as low as 65% and 87%, respectively.[24]

When cholesteatoma is present in a small, contracted mastoid, the entire mastoid may only extend a few millimeters past the horizontal semicircular canal. In such cases an exclusive transcanal atticotomy may be adequate to remove the disease (**Fig. 1**A). In other circumstances the mastoid is small, but with greater antral extension past the horizontal canal. The endoscopic approach cannot reach this space despite an extensive atticotomy and utilization of curved instruments; thus, a mastoidectomy is necessary (see **Fig 1**B).

Occasionally, cholesteatoma with involvement beyond the dome of the horizontal semicircular canal can be removed from the antrum if it possesses a well-defined sac structure. Conversely, a cholesteatoma with lesser involvement of the antrum but with a loose matrix quality may not be amenable to transcanal removal and may require a mastoidectomy.

Mastoid Pneumatization

The extent of mastoid development is an important factor in surgical planning. The degree of mastoid pneumatization is an important predictor of middle ear ventilation.[25] When a widely pneumatized mastoid is partially drilled, residual mastoid air cells

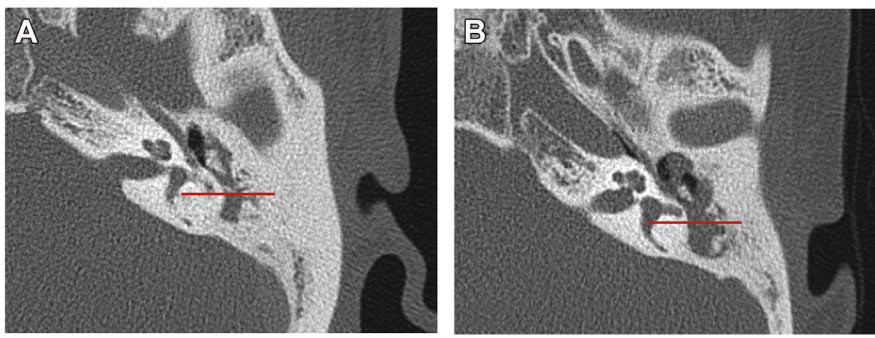

Fig. 1. CT temporal bone axial cross-sections of two left contracted mastoids with cholesteatoma. (*A*) In this case with limited mastoid extension beyond the horizontal semicircular canal, a transcanal endoscopic atticotomy was possible. (*B*) In this case with greater mastoid extension beyond the dome of the horizontal semicircular canal, a canal wall down mastoidectomy was necessary.

continue to produce nitrous gas and contribute to middle ear homeostasis. In these circumstances, it is favorable to perform a canal wall up mastoidectomy, as the mastoid is partially functional. Conversely, a small sclerotic mastoid cavity has poor gas exchange and does not significantly contribute to middle ear pressure homeostasis, especially once it has been opened and drilled. This small mastoid is more likely to self-obliterate with fibrous tissue postoperatively. We have a lower threshold to perform a canal wall down in smaller mastoids as these small drilled mastoids essentially represent a dead space without gas exchange.

Tegmen Condition and Orientation

The tegmen is evaluated for bony dehiscence or disease eroding into the tegmen. In addition, there are few intraoperative landmarks for the tegmen when performing a transcanal atticotomy. Thus, the height and thickness of the tegmen tympani must be evaluated preoperatively to prevent an iatrogenic tegmen defect.

The orientation of the tegmen plane is also assessed. If cholesteatoma extends into the antrum and the tegmen lies in a horizontal plane, the anterior aspect of the attic and aditus ad antrum are easily accessible through a mastoidectomy (**Fig. 2**A). Conversely, a low-lying or sloping tegmen obstructs transmastoid access to the attic. In this circumstance a transcanal endoscopic extended atticotomy is preferred to access the anterior epitympanic space (see **Fig. 2**B).

SURGICAL GUIDE

Prepping the Ear for Endoscopic Approach

The ear is first anesthetized by injecting local anesthetic with a diluted epinephrine solution in the cartilaginous portion of the ear canal, tragus, and posterior auricular sulcus prior to prepping. This allows the anesthetic to infiltrate in the ear canal while prepping and draping is performed. Monitored hypotension and reverse Trendelenburg positioning may help reduce bleeding. The bed is turned 180° and the screen monitor is positioned at eye level approximately 6 feet away from the surgeon. Sitting or standing while performing endoscopic ear surgery is per surgeon preference. The senior author prefers a standing position as it is ergonomically favorable with respect to the arm and shoulder girdle, avoiding neck and shoulder strain. The ear canal hair is trimmed with scissors and the canal is injected just beyond the hair-bearing skin, making sure the entire canal skin blanches and hydrodissects off the canal bone.

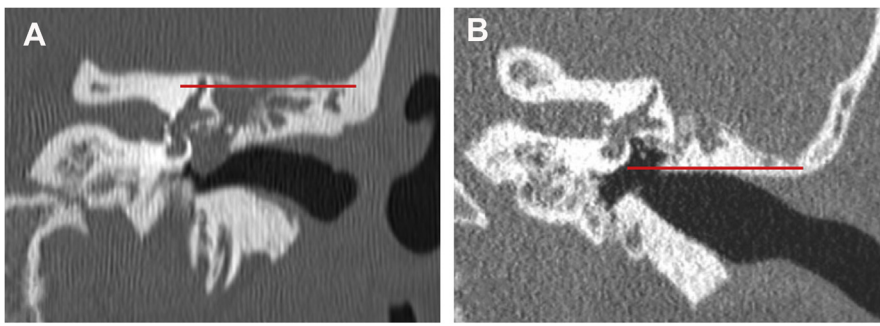

Fig. 2. CT temporal bone coronal cross-sections of 2 left ears with different tegmen conformations. (*A*) The tegmen lies in a horizontal plane allowing transmastoid access to the attic. (*B*) The tegmen is low-lying and overhanging, obstructing transmastoid access to the attic.

We always start with a transcanal endoscopic approach, even when a mastoidectomy is contemplated. This has several advantages. The initial endoscopic inspection of the tympanic membrane may reveal details about the pathophysiology of the disease that may affect the surgical approach. Additionally, starting in the middle ear not only maximizes the vasoconstrictive effects of the local anesthetic, but more importantly, allows removal of disease beginning at its origin and continuing along the pathway of spread.

Endoscopic Management of Epitympanic Cholesteatoma with Limited Attic Extension

Epitympanic cholesteatoma limited to the attic represents the ideal indication for a transcanal endoscopic approach. In epitympanic cholesteatoma, the most common pattern of disease spread is posterior and medial to the ossicular chain. Less frequently, cholesteatoma can migrate posteriorly in the attic and remain lateral to the ossicular chain. Rarely, cholesteatoma spreads toward the anterior attic without direct ossicular chain involvement. Under microscopy, the transcanal view of the attic is limited by the narrow segment of the ear canal. In contrast, endoscopes provide a wide visual field and an angled view of the posterior aspect of the attic.

The tympanomeatal incision is started anterior to the neck of the malleus to facilitate exposure of the entire attic. The flap should be 8 to 10 mm long, as significant curettage of the scutum is often necessary to gain adequate exposure and a longer tympanomeatal flap is required to cover the resultant defect (Fig. 3B).

It is safest to elevate the tympanic annulus in an area that is uninvolved by disease. In epitympanic cholesteatoma, this is most often the posteroinferior aspect. The sac frequently extends into the middle ear space and obscures the ossicles. Curettage

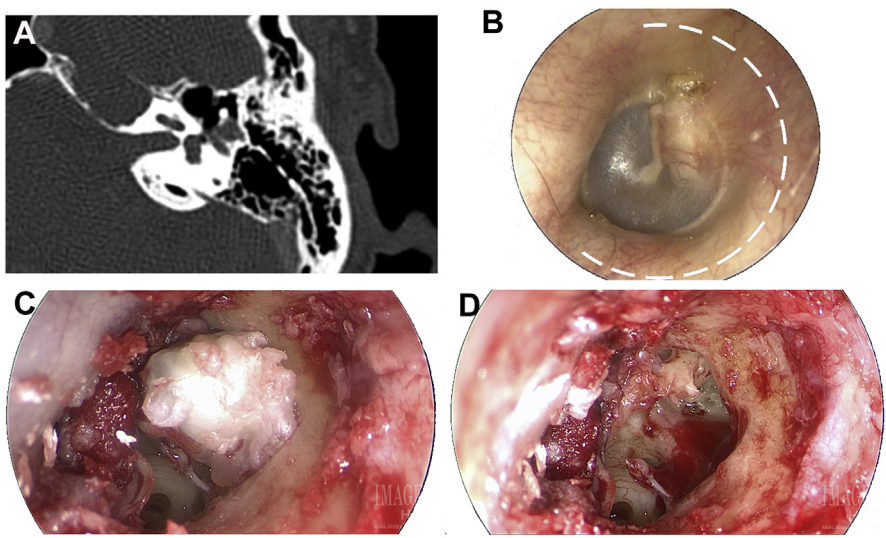

Fig. 3. A 59-year-old woman with left limited attic cholesteatoma. (*A*) CT temporal bone, axial cross-section shows the most common growth pattern of cholesteatoma, posterior and medial to the ossicular chain. (*B*) Endoscopic examination with planned incision marked by the dashed line. (*C*) Surgical exposure after curetting of the scutum to reach the lateral extent of the cholesteatoma sac. (*D*) Endoscopic inspection following removal of disease and incus.

of the posterior ear canal may facilitate identification of the lateral borders of the cholesteatoma sac as well as the bodies of the incus and malleus.

Once the middle ear is entered and the ossicular chain is identified, the surgeon must decide if the ossicular chain must be disrupted in order to eradicate the disease. We advocate removing enough of the scutum to expose the lateral edge of the cholesteatoma. This enables identification of whether the cholesteatoma lies lateral or medial to the ossicular chain.

If the cholesteatoma involves the medial aspect of the ossicular chain, it is necessary to remove the body of the incus, and depending on extension, the head of the malleus. If the malleus is transected, it is best to do so at the neck so that the manubrium remains attached to the tensor tympani tendon. This provides better stability for tympanic membrane reconstruction.

After disease removal, the posterior wall of the epitympanic space is inspected with 30-degree and 45-degree angled endoscopes. Special endoscopic instruments such as curved suctions, curved microcurettes and attic dissectors are used to remove residual squamous debris adherent to the bone. The attic defect is then reconstructed with a cartilage graft (**Fig. 4**). One of the most challenging steps of the surgery is shaping a cartilage graft to match the size of the attic defect and to lay it as a smooth surface adjacent to the tympanic membrane remnant. If the graft is too small, it will fall medially into the attic defect and allow a postoperative retraction to develop. If the graft is too large, it will obstruct visualization and postoperative monitoring of the anterior epitympanic rim. Inadequate juxtaposition of the graft with the native tympanic membrane will allow epithelium to infiltrate medial to the graft, resulting in cholesteatoma formation. It is helpful to fashion a template made of pressed Gelfoam or sterile suture metal packaging to approximate the atticotomy defect prior to shaping the cartilage graft.

Combined Endoscopic and Microscopic Management of Primary Acquired Cholesteatoma with Mastoid Extension

When cholesteatoma extends into the antrum, the crucial question is whether or not an endoscopic transcanal atticotomy is sufficient to reach the antral disease or if a mastoidectomy is required. A limited endoscopic atticotomy can be easily performed with a stapes curette. However, a wider endoscopic atticotomy requires a small diamond otologic burr or a piezoelectric drill for more expedient bone removal. The piezoelectric drill differs from a traditional otologic drill because it uses a blade that oscillates at ultrasonic frequency. Piezoelectric drilling is performed under a constant flow of saline, which reduces heat transmission. In addition, the ultrasonic frequency reduces

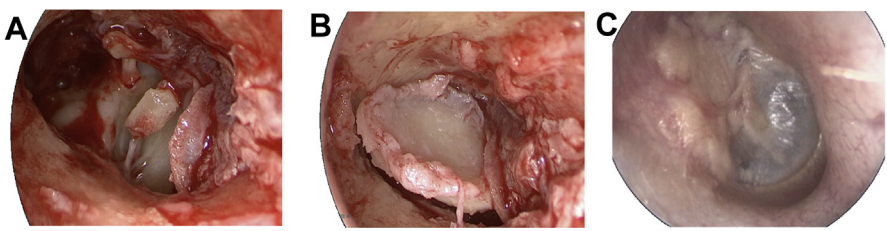

Fig. 4. Right ear cartilage graft placement after cholesteatoma resection. (*A*) Atticotomy defect and autologous incus interposed between the stapes capitulum and malleus manubrium. (*B*) The cartilage graft is sized and positioned to cover the atticotomy defect. (*C*) Well-healed attic cartilage graft.

potential for soft tissue injury. Results of revision mastoidectomy for chronic otitis media and cholesteatoma using the piezoelectric drill have reported no injury to the facial nerve, lateral sinus, or dura.[26]

If the disease is beyond the reach of a transcanal atticotomy, a mastoidectomy is performed to gain full access. Once a mastoidectomy is completed, we inspect the mastoidectomy defect with 0-degree and 30-degree endoscopes to visualize potential blind areas of disease under microscopy such as the medial surface of the skeletonized external auditory canal and the anterior and lateral attic wall.

Endoscopic Management of Mesotympanic Cholesteatoma

Whereas epitympanic cholesteatoma originates from a pars flaccida retraction, mesotympanic cholesteatoma originates from a pars tensa retraction. Tos[27] characterized mesotympanic cholesteatoma into 2 subtypes: (1) sinus cholesteatoma, which involves only the posterior mesotympanum with a normal anterior tympanic membrane (**Fig. 5**A), and (2) tensa retraction cholesteatoma which involves the entirety of the tympanic cavity. At times cholesteatoma may involve both the pars tensa and pars flaccida, making classification challenging. Rosito and colleagues[28] noted in a cohort of 356 patients that 14% of cases of cholesteatoma involved both the pars flaccida and pars tensa, and in 16% of cases no pattern of growth could be identified on otoscopy.

Of note, mesotympanic cholesteatoma occurs more frequently than epitympanic cholesteatoma in children.[28] Eustachian tube function is not fully matured in the pediatric population, predisposing them to otitis media and tympanic membrane retraction, leading to mesotympanic cholesteatoma formation.

The tympanic segment of the facial nerve, the stapes, and retrotympanic space are involved more often in mesotympanic cholesteatoma than in epitympanic

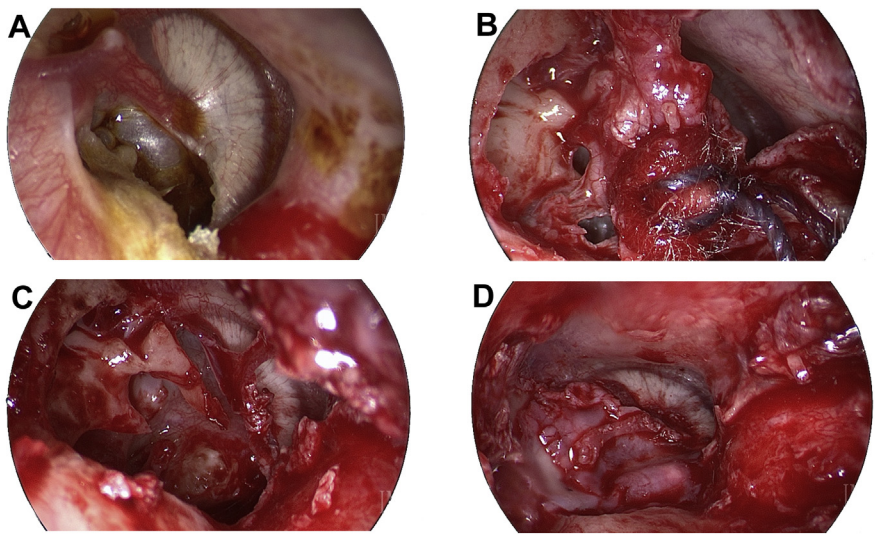

Fig. 5. A 22-year-old man with right mesotympanic cholesteatoma. (*A*) Right mesotympanic cholesteatoma with normal anterior portion of tympanic membrane. (*B*) A cottonoid pledget is used to dissect the atrophic tympanic membrane off the retrotympanic space. (*C*) After disease removal, only a small portion of the anterior tympanic membrane remains. (*D*) A cartilage graft is placed lateral to the manubrium of the malleus.

cholesteatoma.[28] These structures are difficult to reach with transcanal microscopy and often require a transmastoid facial recess approach. Endoscopy affords improved visualization which facilitates an exclusive transcanal approach.

A critical point in the approach to mesotympanic cholesteatoma is to gain wide exposure of the posterior tympanic cavity. A wide semi-circumferential incision is made to create a long tympanomeatal flap as there is often scutal erosion. To accomplish this, the incision is started anterior to the notch of Rivinus and extended 180°, taking care to preserve an 8-mm to 10-mm-wide flap. Elevation of the tympanomeatal flap may be challenging, as the tympanic membrane is often atrophic and adherent to the floor of the middle ear space. The epithelium is also often adherent to the capitulum of the stapes or the tympanic segment of the facial nerve, which may have bony dehiscence. Cottonoids soaked in epinephrine solution (1:1000) are used to dissect the flap anteriorly and away from its adhesions to the promontory (see Fig. 5B). Once the middle ear is opened, the anatomy may be distorted because of scutal erosion or granulation tissue in the location of the facial nerve and stapes. Elevation of the tympanomeatal flap continues inferiorly to identify the round window. This may be the only landmark when the rest of the middle ear is filled with disease.

When the incus and stapes are not readily identifiable, curetting the posterior superior aspect of the scutum can allow identification of the neck of the malleus, ensuring a safe anatomic landmark to start the middle ear dissection. If cholesteatoma occupies the retrotympanic space, there may be erosion of the posterior canal bone and pyramidal eminence, distorting the landmarks for the second genu of the facial nerve. The stapes may have partial or total erosion of its suprastructure and matrix may cover the footplate. Squamous debris extending into the sinus tympani and retrotympanic space can be removed under endoscopy with curved endoscopic instruments or attic dissectors, avoiding the need for a mastoidectomy with a posterior tympanotomy. Access to the retrotympanic space may be improved by standing on the opposite side of the patient, rotating the patient away from the operative ear, and using a 30-degree endoscope. This direct endoscopic retrotympanic approach requires experience in endoscopy to maintain orientation (Fig. 6).

After removing disease from the mesotympanum and retrotympanic space, often only a small portion of the anterior half of the tympanic membrane is preserved (see Fig. 5C). A cartilage graft is preferred for tympanic membrane reconstruction as it is stiffer and resists postoperative retraction. Placement of the underlay cartilage graft lateral to the malleus helps prevent postoperative retraction, a frequent sequela observed when placing the cartilage graft medial to the manubrium of the malleus (see Fig. 5D).[29]

Endoscopic Management of Mesotympanic Cholesteatoma with Infracochlear Extension

When there is extensive retrotympanic involvement, disease can extend into the petrous apex via the infracochlear and infracarotid air cell tracts (Fig. 7). If the preoperative CT shows indistinct opacification between the middle ear and the infracochlear air cell tract, a non–echo planar DWI MRI can differentiate cholesteatoma from effusion, cholesterol granuloma, or normal bone marrow. Microscopic access to the infracochlear region requires significant removal of the inferior rim of the external ear canal which places the mastoid segment of the facial nerve at risk of injury. Endoscopic approaches to this area may offer better visualization without significant removal of the inferior canal wall. When cholesteatoma has involved the infracochlear air cell tract, the anatomy can be significantly distorted. The round window niche may be eroded and the bony boundaries widened (Fig. 8). Matrix in the hypotympanic region must

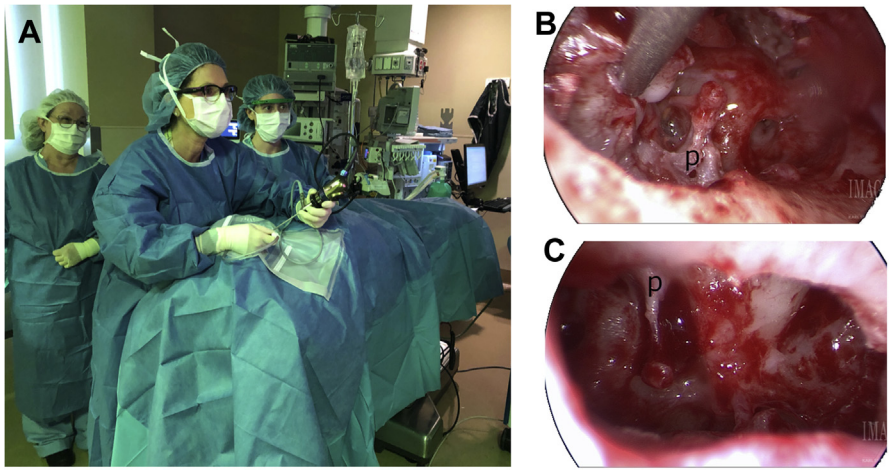

Fig. 6. Direct endoscopic retrotympanic inspection of right ear. (*A*) The surgeon stands on the opposite side of the operative ear using a 30-degree endoscope. (*B*) Right ear. Traditional endoscopic view of sinus tympani. Cholesteatoma is present along the stapedial tendon and pyramidal eminence. (*C*) Direct retrotympanic inspection of the same ear illustrates an upside-down view of the retrotympanic space after cholesteatoma removal. p, pyramidal eminence.

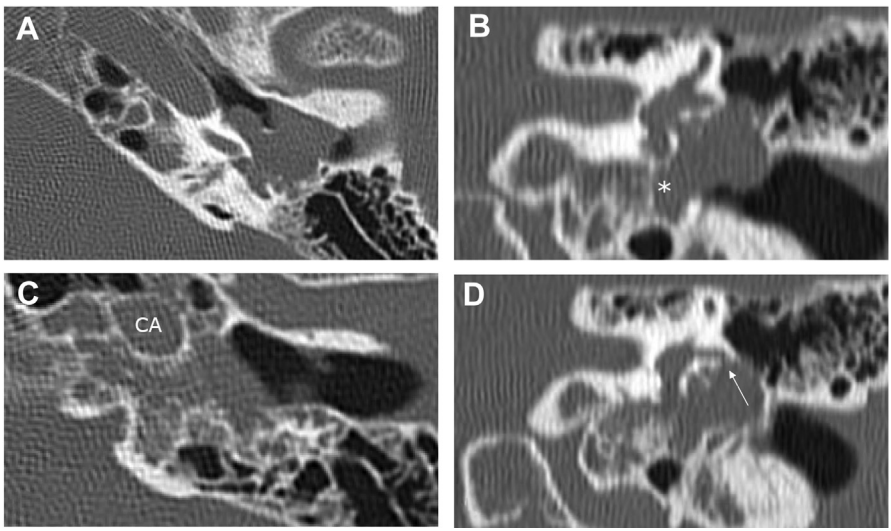

Fig. 7. CT scan of a 47-year-old man with left mesotympanic cholesteatoma. (*A*) Axial cross-section showing opacification of the entire middle ear and sinus tympani. (*B*) Coronal cross-section showing opacification extending in the infracochlear air cell tract. (*C*) Axial cross-section showing opacification in the infracarotid air cell tract. (*D*) Coronal cross-section showing possible lateral semicircular canal fistula. arrow, labyrinthine fistula; *, infracochlear air cell tract; CA, carotid artery.

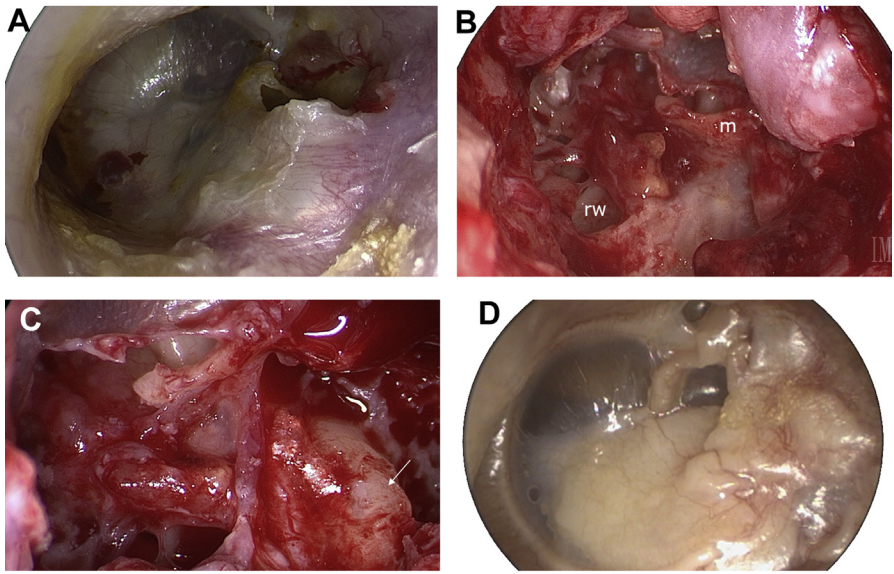

Fig. 8. Intraoperative images of the same patient from Fig. 7. (*A*) Endoscopic exam of left ear. (*B*) Cholesteatoma has eroded a significant portion of the posterior scutum with distorted anatomy. The malleus manubrium is the sole landmark to help with orientation. The round window anatomy is distorted due to niche erosion and enlargement of the bony boundaries. (*C*) After cholesteatoma removal, there is a blue lining of the dome of the horizontal semicircular canal. (*D*) Postoperative examination at 6 months. m, malleus manubrium; rw, round window; arrow, blue lining of horizontal semicircular canal.

be carefully lifted and structures palpated to assess for bony dehiscence over the jugular bulb and carotid region (Fig. 9).

Endoscopic Management of Cholesteatoma with Labyrinthine Fistula

Perilymphatic fistulas due to invasion by cholesteatoma is not an uncommon occurrence. Labyrinthine fistulas are classified according to degree of erosion. In type I,

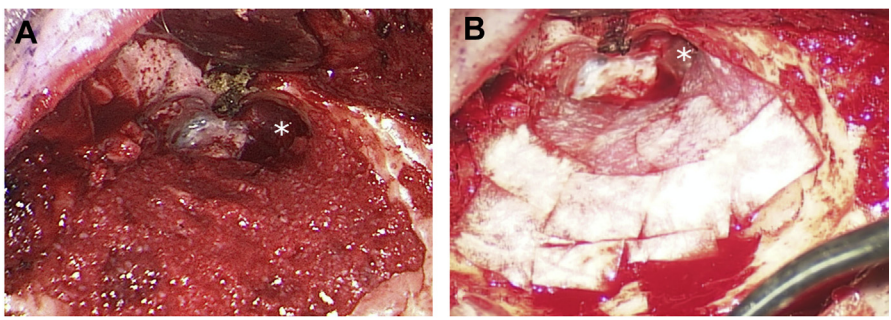

Fig. 9. Left ear with canal wall down mastoidectomy with selective mastoid obliteration. (*A*) The mastoid defect is obliterated with DBX and bone pate. (*B*) The mastoid cavity is lined with Biodesign graft as there was insufficient temporalis fascia to line the defect secondary to multiple prior surgeries. *, exteriorized attic.

the perilymphatic membrane is still covered with bone (blue lining); type II occurs when the perilymphatic membrane is exposed; and type III occurs when cholesteatoma has eroded the perilymphatic membrane or invaded into the labyrinth.[30] The estimated incidence of labyrinthine fistula is 6% to 8% in all cholesteatoma cases, with 90% or more occurring at the lateral semicircular canal.[31,32] Portier and colleagues[31] further reported a 9% incidence of a second fistula identified intraoperatively. Schmidt Rosito and colleagues[33] identified 9 labyrinthine fistulas in their cohort of 333 patients with cholesteatoma, all of which were in the setting of a posterior epitympanic cholesteatoma.

The surgical management of labyrinthine fistulas depends on several factors, including size and depth of the fistula, location, hearing status of the operative and contralateral ear, as well as the surgeon's level of comfort. A fistula can be managed conservatively by leaving cholesteatoma matrix over the fistula and performing a canal wall down mastoidectomy to exteriorize the disease. However, this approach leaves the patient with persistent symptoms as the labyrinthine fistula is not repaired but simply exteriorized into a large mastoid cavity.

More advanced techniques include complete removal of cholesteatoma matrix from perilymphatic fistulas while preserving labyrinthine function and enabling a canal wall up approach.[34] Removal of cholesteatoma matrix from the fistula is performed by carefully lifting the matrix and avoiding direct suction over the fistula. Continuous irrigation allows removal of the debris without using suction; however, the irrigation stream may obscure the field of view. Yamauchi and colleagues[35] described underwater endoscopic repair of labyrinthine fistulas. The mastoid is filled with saline and the endoscope is submerged to perform the repair, thus avoiding the refractive effects of the traditional stream of irrigation. Once the matrix is completely removed, the fistula can be repaired with temporalis fascia, perichondrium, and bone pate.

Indications for Canal Wall Down Mastoidectomy

The utilization of endoscopy as an adjunct tool in cases of cholesteatoma with mastoid involvement can facilitate the choice of a canal wall up mastoidectomy.[16] However, there are still circumstances when a canal wall down mastoidectomy is unavoidable. A canal wall down mastoidectomy is often performed to eradicate recidivistic disease after multiple canal wall up procedures have failed. There are however, situations in which a canal wall down mastoidectomy is discussed with the patient at the time of primary surgery for cholesteatoma.

Indications for a primary canal wall down mastoidectomy in the contemporary era of endoscopic ear surgery can be divided into patient characteristics, disease extent, and anatomic factors. Patient characteristics include patients with multiple comorbidities who are poor candidates for multiple ear surgeries or if the cholesteatoma is present in the patient's only hearing ear. A single, canal wall down procedure may also be optimal if there is concern for follow-up.

Disease-related indications for a canal wall down mastoidectomy can be further divided by disease extent and patient anatomy. Cholesteatoma with more than 50% bony erosion of the posterior canal wall, extensive labyrinthine fistula, or presence of a large tegmen defect with dural attachment to cholesteatoma is an indication for a canal wall down mastoidectomy. The surgeon should also take into consideration their own experience, comfort level and likelihood of completely eradicating disease.

Anatomic factors that may require canal wall down mastoidectomy as a primary surgery include cholesteatoma in a small mastoid with a low-lying tegmen. When the tegmen is low-lying, it may obstruct visualization of the anterior epitympanum

via the microscopic transmastoid route. This frequently leads to the decision to take the canal wall down to achieve an unobstructed view of the attic. In selected cases of cholesteatoma present in a small mastoid antrum and a low-lying tegmen, it is possible to perform an exclusive endoscopic transcanal retrograde inside-out mastoidectomy. This is achieved by extending the transcanal atticotomy to fully expose the mastoid antrum. In this case, a small meatoplasty may be sufficient to access the small mastoid cavity, which will require less maintenance than a traditional large mastoid cavity.

However, if a low-lying tegmen is associated with a prominent anterior canal wall, even endoscopic techniques are challenging. In this situation the prominence of the anterior canal wall precludes a satisfactory transcanal approach to the attic. In addition, when there is a prominent canal wall, reconstructive efforts to cover the attic defect with a cartilage graft may fail. The acute angle between the anterior canal wall and the anterior tympanic rim impedes good control of graft placement. Epithelium can grow under the graft at the junction between the anterior tympanic rim and the graft. In this situation a canal wall down mastoidectomy with the attic space left exteriorized and lined by fascia is a safer option for long-term disease control.

Innovations in Mastoid Obliteration Techniques

When cholesteatoma has extended in a widely pneumatized mastoid and a canal wall down mastoidectomy is performed, patients are left with a large mastoid cavity that requires long-term maintenance. One option to reduce the cavity size and minimize postoperative chronic granulations is to selectively obliterate the mastoid cavity.

Techniques for selective mastoid obliteration differ from the traditional tympanomastoid obliteration of a radical cavity, in which the entire mastoid and middle ear space, voided of ossicles, are obliterated with a large pedicled Palva flap and the canal oversewn.[36] Recently, mastoid obliteration techniques have been applied to both canal wall down mastoidectomy and canal wall up mastoidectomy with the goal of selectively obliterating the mastoid while preserving the middle ear space. Obliteration of the mastoid alone creates a small mastoid cavity that is, more manageable in terms of cleaning and debridement. In addition, a smaller mastoid cavity requires a smaller meatoplasty, avoiding the cosmetic disfigurement of a large meatoplasty. When the mastoid is selectively obliterated in a canal wall up mastoidectomy, the purpose of obliteration is to occlude the dead space and prevent cholesteatoma formation.

A recent systematic review of patients who underwent mastoid obliteration with canal wall up and canal wall down mastoidectomy found better rates of residual and recurrent cholesteatoma than the published rates for those who did not undergo obliteration. Overall rates of recurrent and residual disease were low, at 4.6% and 5.4%, respectively.[37] If the decision is made to obliterate the cavity, there are several commercially available materials. Demineralized Bone Matrix (DBX; DePuy Synthes, West Chester, PA) combined with bone dust collected during drilling has been used with stable long-term results.[38] Other materials include bioactive glass polymers,[39] titanium micromesh, hydroxyapatite, and silicone.[40] When performing mastoid obliteration techniques, postoperative monitoring with a non–echo planar DWI MRI is required to rule out occult cholesteatoma within the obliteration. A modified technique for mastoid obliteration is a partial obliteration with the epitympanum left exteriorized and lined by fascia. In this case the area at most risk for residual disease is left exposed and exteriorized.

MRI AS AN ALTERNATIVE TO SECOND-LOOK SURGERY FOR POSTOPERATIVE SURVEILLANCE

The paradigm of a second-look surgery following cholesteatoma removal has shifted with the advent of non–echo-planar DWI MRI, which was first described in identifying cholesteatoma in 2006.[41] Multiple systematic reviews comparing non–echo planar DWI MRI to intraoperative identification of cholesteatoma have reported sensitivity and specificity over 90%.[42,43] The decision to select non–echo planar DWI MRI versus a second-look surgery depends on patient factors, extent of cholesteatoma, and surgeon confidence in the resection of the cholesteatoma. MRI is appropriate for surveillance in adults with low risk of residual or recurrent disease. A second-look surgery is preferred in children, as cholesteatoma tends to be more aggressive. In addition, MRIs may require sedation for the child to participate.[42] A second-look surgery may also be preferable if there is concern for patient compliance with long-term surveillance. Finally, the surgeon must keep in consideration that MRI does not routinely detect cholesteatoma less than 3 mm in size.

Our practice has shifted from a routine second-look surgery in all cholesteatoma cases to a more tailored protocol specific for each case. In general, in cases with low concern for residual disease, a non–echo planar DWI MRI is ordered 12 to 18 months following primary surgery. In cases with increased concern for residual disease a second-look surgery is scheduled 9 to 12 months after the primary surgery.

OUTCOMES

Outcome data regarding endoscopic approaches to surgery, while limited, are promising. The endoscopic approach to cholesteatoma removal has at least equivalent outcomes with regard to residual and recurrent disease, similar rates of postoperative complications, and decreased pain and shorter healing time compared to microscopic surgery.[44–46] Hearing outcomes are also comparable. Further investigation is needed to compare operative time as well as long-term outcomes.

A primary concern following cholesteatoma resection is recidivism, which includes residual and recurrent disease. These rates are widely variable in traditional microscope otologic surgeries, with meta-analysis data reporting recurrence rates of cholesteatoma of 9% to 70% following a canal wall up procedure and lower rates of 5% to 17% after a canal wall down procedure.[47] Endoscopic ear surgery has demonstrated noninferior outcomes with regards to residual and recurrent disease rates. Presutti and colleagues[16] reported in their meta-analysis that 6.2% of patients had residual disease and 3.1% of patients had recurrence after endoscopic middle ear surgery for cholesteatoma; however, this was limited by a small number of studies and short mean follow-up period (23.4 months). Subsequent studies that compared endoscopic surgery to a microscopic control group for cholesteatoma showed equivalent rates of residual and recurrent disease, of up to 17% and 20%, respectively.[44,45] Long-term data regarding outcomes and need for revision surgery for other indications, such as perforation or graft failure, are future areas of investigation.

Differences in operative time are widely variable. One randomized controlled trial reported a mean decrease in operating time of 20 minutes with endoscopic approaches for limited attic cholesteatoma compared with microscopic approaches.[46] Other studies reported equivalent or increased time for endoscopic approaches, but with a decrease in operative time with experience.[44,45] Acute postoperative complications are rare and do not differ significantly between those undergoing microscopic versus endoscopic ear surgery. Most studies do not report facial nerve injury following either approach[17,44,46]; Killeen and colleagues[45] reported one patient with a facial nerve

palsy following endoscopic ear surgery that resolved during follow-up. Incidence of dizziness and dysgeusia following surgery did not differ by surgical approach.[44] Of note, endoscopic ear surgery appears to offer a distinct benefit in decreased pain and recovery time.[18,44]

Studies comparing endoscopic surgery with a microscopic surgery control group have not shown significant differences in hearing outcomes as measured by changes in air-bone gap closure, air conduction thresholds, median pure tone averages, and word recognition scores.[44–46]

SUMMARY

Endoscopic ear surgery is increasingly accepted as a primary modality for cholesteatoma surgery. One of its major advantages is the superior visualization of the attic and sinus tympani, the 2 sites at highest risk for residual disease in cholesteatoma surgery. Transcanal endoscopic ear surgery is particularly suited for management of limited epitympanic and mesotympanic cholesteatoma. When cholesteatoma extends in the mastoid beyond the dome of the horizontal semicircular canal, a combined approach with mastoidectomy is often necessary. Even with endoscopic assistance, certain anatomic constraints such as a low-lying tegmen associated with a prominent anterior canal wall may preclude full access to the anterior attic and complete disease removal, necessitating a canal wall down mastoidectomy. When a canal wall down mastoidectomy is necessary, partial mastoid obliteration techniques allow a selective obliteration of the mastoid cavity with preservation of the middle ear space. This has reduced the long-term sequelae of large mastoid cavities. Data comparing outcomes in endoscopic ear surgery show at least equivalent outcomes compared to microscopic ear surgery in rates of residual and recurrent disease, and are associated with decreased postoperative morbidity and shorter recovery times.

DISCLOSURE

The authors have nothing to disclose.

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