



Transmastoid labyrinthectomy

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Transmastoid labyrinthectomy is a highly effective procedure for controlling vertigo attacks in patients with refractory Meniere's disease and ipsilateral nonserviceable hearing. Performed under general anesthesia, the procedure involves a cortical mastoidectomy with subsequent opening and removal of the vestibular neuroepithelium of the 3 semicircular canal ampullae, as well as opening and ablation of the utricle and saccule of the vestibule. Transmastoid labyrinthectomy typically requires several days of hospitalization to permit acute central compensation, and a period of vestibular physical therapy is typically necessary for patients to return to normal activity levels by 2 months. Transmastoid labyrinthectomy is a safe procedure with low rates of complications. Hearing rehabilitation can be performed via cochlear implantation, when indicated. Overall, transmastoid labyrinthectomy provides high rates of relief from vertigo and remains the gold standard for surgical treatment of Meniere's disease.

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Introduction

Patients with Meniere's disease (MD) classically present with fluctuating hearing loss, vertigo, aural fullness, and tinnitus, although not all symptoms may be present. Most affected individuals harbor unilateral symptoms but up to 10% may initially present with bilateral disease, and an additional 10% of patients may go on to develop MD in the contralateral ear.¹ The definition of probable or definite MD includes 2 or more episodes of spontaneous vertigo lasting 20 minutes to 12 or 24 hours, fluctuating hearing loss, tinnitus, or aural fullness.² Other structural pathology,

such as vestibular schwannoma or central ischemia, must be excluded.

Surgical ablation, particularly via the transmastoid labyrinthectomy, is reserved for patients with unilateral, debilitating disease that has been unresponsive to less-invasive therapy along the MD treatment ladder.³⁻⁶ Labyrinthectomy is typically offered only to patients without serviceable hearing in the ipsilateral ear, as surgery will result in anacusis. Although the procedure is destructive, surgical labyrinthectomy has near-total success in controlling vertigo attacks and remains the gold standard for treatment in this subset of patients.

A discussion of other surgical treatment options for MD including endolymphatic sac decompression and vestibular nerve section are covered in other chapters of this series. Herein, we outline the preoperative evaluation, surgical

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technique, and expected postoperative course for patients undergoing transmastoid labyrinthectomy.

Preoperative evaluation

History and physical exam

The initial work-up of any dizzy patient requires differentiation of a central vs peripheral lesion based on clinical history and physical exam findings.³ Patients with persistent disequilibrium due to poor central compensation (eg, following vestibular neuritis) may present similarly to those with MD, but must be differentiated as this cohort benefits from vestibular rehabilitation.³ The most confounding diagnosis in this subset of patients is vestibular migraine, for which diagnostic criteria have recently been proposed.⁷ A trial migraine prophylaxis medication, that is, calcium channel blockers, may be initiated to expose an underlying migraine condition or to optimize migraine control prior to surgery.⁸

Further testing and imaging

Vestibular testing should be performed to confirm laterality and document the severity of vestibular hypofunction in all patients considering ablative surgery.⁹ Audiograms should also be obtained both at baseline and during an acute episode of vertigo to establish fluctuating sensorineural hearing loss and to document the hearing status of both ears.⁹ If unilateral hearing loss is identified, it is essential to obtain magnetic resonance imaging with and without contrast to rule out retrocochlear pathology, including ischemic, neoplastic, or demyelinating lesions.³

As for all surgery of the temporal bone, high-resolution computed tomography should be obtained for proper surgical planning. Anatomic structures to note during preoperative imaging review include: the location of the mastoid tegmen, sigmoid sinus, jugular bulb, and presence of any concurrent middle ear pathology.⁹ Computed tomography imaging may also be used to rule out other causes of vertigo, such as cholesteatoma, third window lesions, or cochlear otosclerosis.³

Patient selection

Typically, patient should have nonserviceable hearing in the ipsilateral ear as defined by poor word discrimination. One should also keep in mind the nature of MD, which, in most cases, will result in progressive hearing loss.⁹ The use of surgical ablation in patients with bilateral vestibular loss should be considered with care, due to expected poor postoperative central compensation. However, patients suffering from life-threatening Tumarkin drop attacks may be also candidates for surgical ablation, regardless of the status of the contralateral ear.¹⁰ Determining which ear is responsible for drop attacks in cases of bilateral disease can be challenging, but a careful review of lateralizing

symptoms and detailed analysis of vestibular test results should identify the responsible ear. Ultimately, the decision for surgery should come as a shared decision from the otologist and patient.

Patient counseling

Prior to surgery, all patients must be counseled on the risks and benefits of surgical labyrinthectomy. While highly effective, there is always a chance that surgery may not completely resolve vestibular symptoms. Patients must understand that ipsilateral anacusis will result from labyrinthectomy. However, cochlear implantation (CI), either at the time of surgery or in a delayed fashion remains an option to restore hearing.¹¹ Patients with evidence of contralateral vestibular hypofunction or with other comorbidities, such as peripheral neuropathy, proprioceptive disorders, arthritis, or impaired vision, may also have limited or prolonged postoperative central compensation.¹²

Surgical approach

Preoperative considerations and set-up

Labyrinthectomy is performed under general anesthesia. Facial nerve monitoring is essential, and the anesthesiologist must be made aware to avoid long-acting paralytics on induction or throughout the case.³ The patient is positioned supine on the operating table and positioned as for a mastoidectomy procedure, with the head rotated toward the contralateral (nonoperative) side and the neck extended until the floor of the middle fossa is positioned vertically.⁹ Hair within the surgical field is shaved and the area is sterilely prepped and draped.

Technique for ablation

A cortical mastoidectomy is performed: first, a postauricular incision is made, extending to the mastoid periosteum. A scalpel is then used to make a T-shaped incision in the mastoid periosteum along the temporal line. The flap is elevated to the posterior aspect of the external auditory canal with a Lempert Elevator to identify the spine of Henle and the root of the zygoma. A canal-wall-up mastoidectomy is performed, with care taken to thin the bone overlying the sigmoid sinus and tegmen mastoideum, for adequate access to the labyrinth ([Figure 1A](#)).⁹ Rarely, a poorly pneumatized mastoid will prevent adequate surgical access through the mastoid, and thus, decompression of the sigmoid sinus or dura of the middle cranial fossa is necessary to visualize the 3 semicircular canals.³ After the antrum is opened and the lateral canal is identified, the second genu and vertical segment of the facial nerve are skeletonized with a diamond burr using smooth motions along the direction of the nerve with copious irrigation.³

All 3 semicircular canals are subsequently identified and skeletonized, and the trabeculated bone between the canals

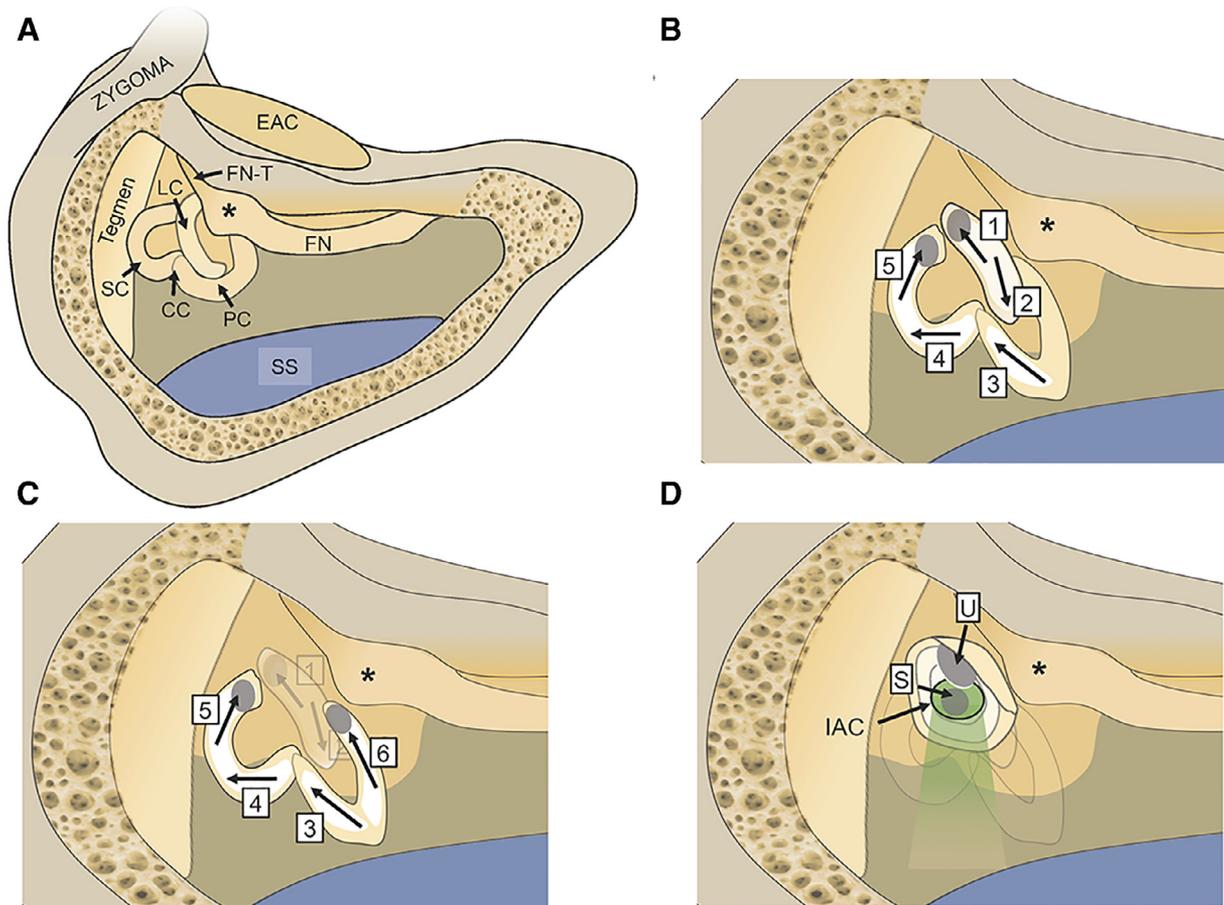


Figure 1 Illustration of surgical procedure for transmastoid labyrinthectomy. Right ear in surgical position. (A) A standard mastoidectomy is performed with care taken to bevel bone over the sigmoid sinus (SS) and tegmen mastoideum. The superior, posterior, and lateral semicircular canals (SC, PC, and LC, respectively) are identified. The tympanic segment of the facial nerve (FN-T) runs parallel and inferior to the LC and the FN-T, second genu of the FN (*) and vertical segment of the facial nerve (FN) are identified and skeletonized. (B-D) Right ear rotated anteriorly with respect to part (A) to view structures medial to the facial nerve. After the semicircular canals and facial nerve have been skeletonized, the canals are drilled open in the order shown by arrows (1→6) to identify and remove all 3 ampullae (gray ovals; B, C). The bone between the common crus (CC) and LC is then drilled away to enter into the vestibule. The utricle (U) is found on the lateral wall of the vestibule and the sacculus (S) is found on the anterior wall of the vestibule at the level of the stapes footplate. The internal auditory canal (IAC) enters the vestibule on its anteromedial surface and its trajectory, anterior to the vestibular labyrinth, is shaded in green (D). (Color version of figure is available online.)

is removed. First, the lateral semicircular canal (LSCC) is drilled open, keeping in mind that the tympanic facial nerve runs parallel and inferior to this structure. Drilling is extended anterosuperiorly to open the ampullated end of the LSCC and then posteriorly. At the posterior end of the LSCC, the dome of the posterior semicircular canal is identified and drilled open superiorly toward the deep and medial common crus. Once the superior semicircular canal has been identified, it is drilled anteriorly toward its ampullated end. The ampulla is opened and all neuroepithelium removed under direct vision. It is critical to avoid drilling through the anteromedial wall of the superior canal ampulla in order to prevent injury to the labyrinthine segment of the facial nerve that lies immediately beyond this bony landmark. Minor bleeding from the subarcuate artery, which runs through the subarcuate tunnel of the superior semicircular canal, may be encountered at this step

(Figure 1B).³ Bleeding can be controlled using a small (eg, 2.0 mm) diamond burr. Finally, the ampulla of the posterior semicircular canal is drilled open, with extreme care due to its location medial to the second genu of the facial nerve. Access to this ampulla requires adequate removal of the posterior end of the LSCC (Figure 1C).⁹ Direct visualization of all 3 ampullae allows for complete removal of neuroepithelium with either suction or a curette.

At this point, the vestibule is opened by extending drilling through the bone between the common crus and LSCC (Figure 1D). For adequate visualization, the facial nerve must be skeletonized along its medial surface using a 2- or 3-mm diamond burr.⁹ Further rotation of the patients head away from the surgeon by tilting the bed may assist in adequate visualization of the vestibule. The utricle is found on the lateral wall of the vestibule just posteromedial to the tympanic segment of the facial nerve.⁹ Removal of the

macula of the utricle may be accomplished using a stapes curette, but care should be taken as the tympanic segment of the facial nerve may be dehiscant into the vestibule at this point.⁹ The macula of the saccule lies in the spherical recess on the medial (deep) wall of the vestibule at the level of the oval window and may be removed with suction or a curette.³ To ensure complete removal of neuroepithelium, the anteromedial wall of the vestibule should be drilled-down, while taking care to not violate the thin bone of the lateral aspect of the internal auditory canal (IAC).^{3,9} If the IAC is opened, a spinal fluid leak is possible and must be repaired.

Technique for closure

Following complete removal of all vestibular end organs, the vestibule is packed with absorbable packing, such as Gelfoam, soaked in gentamicin. This maneuver is highly effective at completing ablation of the vestibular neuroepithelium. The wound is then closed in layers, with absorbable sutures in the mastoid periosteum and nylon sutures or stainless steel staples for the skin. A sterile mastoid compression dressing is applied.

Postoperative care and rehabilitation

Patients should expect to wake up with 12-24 hours of postoperative vertigo and nystagmus. The degree of postoperative vertigo is dependent on the amount of preoperative residual vestibular function.⁹ Rehabilitation begins in the hospital, with the initiation of vestibulo-ocular exercises on the first postoperative day. Twenty-four hours after surgery, the patient should be out of bed to chair. By postoperative day 2 or 3, the patient should be able to walk without assistance, and may be discharged home if there are no other medical concerns.³

Once discharged home, patients should be counseled to remain as active as is tolerable, while avoiding strenuous activities through the second postoperative week. After postauricular sutures are removed, patients may return to most daily activities. The more active patients are during this period, the quicker their recovery will be.³ Driving, however, must be avoided until patients are able to turn their heads and look behind them without becoming disoriented.⁹ By about 2-months postop, most patients should have nearly complete compensation. However, it is essential to keep in mind that certain comorbidities, such as vision loss, diminished proprioception, or other chronic illness may be associated with prolonged rehabilitation or poorer outcomes.¹² Additionally, there is always a risk for disease progression in the contralateral ear, which may be indicated by late decompensation after initial improvement.¹

Hearing rehabilitation

Following labyrinthectomy, patients are left with anacusis in the operated ear. For these patients, simultane-

ous or staged CI is the sole options for hearing rehabilitation. CI can be considered if there is fluctuating hearing in the contralateral ear, or if the patient has normal contralateral hearing but struggles with bothersome tinnitus and sound localization. Patients with unilateral hearing loss have demonstrated benefits from CI for single-sided deafness, specifically in terms of tinnitus suppression and sound localization.¹³

Recent reports of patients undergoing CI following labyrinthectomy show similar hearing results regardless of whether implantation is performed at the time of labyrinthectomy or even years later.^{11,14,15} In the case of delayed implantation, progressive intracochlear ossification may occur following labyrinthectomy and surgeons should consider placement of an intracochlear spacer at the time of labyrinthectomy.^{16,17} A recent study suggests that intracochlear ossification following labyrinthectomy may only occur when there is violation of the IAC with subsequent compromise of the labyrinthine artery, such as during surgery for vestibular schwannoma.¹⁷ In contrast to previous studies, these authors found no evidence of intracochlear ossification multiple years following labyrinthectomy for MD, suggesting that, in this patient cohort, the timing of CI may be more flexible than previously believed.¹⁷

Conclusion

Transmastoid labyrinthectomy is a highly effective surgical treatment for vertigo associated with MD, but is reserved for patients with unilateral, debilitating disease that is unresponsive to conservative treatment. Patients should have nonserviceable hearing in the ipsilateral ear, and typically have failed IT gentamicin. The surgical procedure involves a standard mastoidectomy with subsequent drilling of the semicircular canals and vestibule, which allows for removal of all 5 vestibular end organs under direct visualization. Within 2 months, most patients should have nearly complete resolution of vestibular symptoms and have achieved central compensation; however, this course may be prolonged in patients with medical comorbidities affecting vision or balance. Hearing rehabilitation in labyrinthectomy patients may be accomplished via simultaneous or sequential CI. Overall, transmastoid labyrinthectomy provides high rates of relief from vertigo and remains the gold standard for surgical treatment of MD.

Disclosure

The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

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