

Transcanal Cochleosacculotomy^{☆,☆☆}



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Meniere's disease (MD) is a clinical syndrome consisting of fluctuating hearing loss, episodic vertigo, and aural fullness related to endolymphatic hydrops. When MD is refractory to maximal medical management, surgical therapies can be highly effective. Operations for MD are classified into 2 types: one that ablates the vestibular system such as labyrinthectomy and the other relieves the pressure in the endolymphatic system by fistulization of the labyrinth and decompression of the endolymphatic sac. Transcanal cochleosacculotomy is an internal shunt procedure aimed to drain excessive endolymph. This article reviews the disease pathophysiology, as well as the history, indications, technical aspects, and outcomes associated with the operation. In sum, cochleosacculotomy is moderately successful in relieving vertigo symptoms with low overall morbidity and can serve as a useful alternative to other surgical modalities in carefully selected patients.

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Introduction

Initially described by Prosper Meniere in 1891, Meniere's Disease (MD) was classified as a clinical entity consisted of tinnitus, aural fullness, and fluctuating hearing loss associated with episodic vertigo. Today, the diagnostic criteria for MD as defined by the American Academy of Otolaryngology include: (1) two or more episodes of vertigo lasting 20 minutes-2 hours, (2) unilateral low-to-mid frequency sensorineural hearing loss associated with episodes of vertigo attacks, and (3) fluctuating aural symptoms including hearing loss, tinnitus, and aural fullness.¹ The presentation and clinical course of MD can be highly variable. Patients typically present in the third-seventh

decades of life with a slight female predominance, with an incidence of approximately 1 in 500 in the United States.² This article aims to focus on the history, indications, techniques, and outcomes for surgical treatments for MD, specifically transcanal cochleosacculotomy.

Pathophysiology

Despite variability in the symptomatology of patients with MD, the unifying underlying pathology has been attributed to progressive endolymphatic hydrops. Two principal patterns of pathologic change in the temporal bones of patients with MD emerge: (1) the distention and rupture of the endolymphatic system, and (2) the atrophy and change in the cyto-architecture of sensory organs and supporting cells. Originally proposed by Schuknecht, the rupture of Reissner membrane allows the influx of endolymph into perilymph fluid, where high endolymphatic concentration of potassium ions (140 mEq of K⁺) in the endolymph is neurotoxic to hair cells and the cochlear nerve.^{3,4} It

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has been postulated that the sudden increase in potassium ion concentration paralyzes the sensory organs which then manifests as episodic vertigo, aural symptoms, and hearing loss. As the disease progresses, permanent pathologic changes in the cyto-architecture such as distortion and atrophy of the sensory and supporting cells occur, which affects the motion mechanics of the sense organs. These histopathologic changes are correlated to audiovestibular symptoms including progressive loss of hearing and balance⁵.

History

The overall goals of management of MD are to alleviate symptoms and arrest the progression of disease. Conservative strategies include diet modifications such as low salt diet and diuretic therapy.^{6,7} Acute MD attacks is first managed with systemic steroid therapy and vestibular suppressants.⁸ Intratympanic injection of gentamicin is reserved for treatment of intractable vertigo through chemical ablation of the labyrinth. Surgical management for MD is usually considered if the patient fails noninvasive therapy after 3-6 months. Broadly speaking, surgical treatment can be classified as nondestructive where the goal is to enhance drainage of endolymph and decompress the endolymphatic sac, vs destructive where neural inputs from the affected ear is surgically ablated. Endolymphatic drainage surgeries are further subclassified into external shunt operations that drain endolymph into the mastoid cavity or subarachnoid space, and internal shunt procedures that drain endolymph into the perilymphatic space.

Historically, internal shunt procedures include sacculotomy and tack operations as described by Fick and Cody, respectively,^{9,10} the otic-perotic shunt described by Pulec and House¹¹ and cochleosacculotomy. In the original description of the sac and tack procedures, the saccule is punctured through the stapes footplate by a pick to produce a fistula connecting endolymph and perilymph. However, in MD patients the saccular wall is often adherent to the footplate and fistulization can be challenging.¹² By contrast, the cochleosacculotomy involves impaling the osseous spiral lamina and cochlear duct to create a permanent fracture-disruption. This has been supported historically by evidence from human and animal temporal bone studies.¹³⁻¹⁵

Patient selection

Candidates for cochleosacculotomy must have clear unequivocal evidence of unilateral vestibular dysfunction secondary to MD. As such, other etiologies that can cause vertigo associated with fluctuating hearing loss such as autoimmune inner ear disease, perilymphatic fistulas, and intracranial tumors should be excluded. There should be no evidence of MD in the contralateral ear, as bilateral MD can develop in up to one-third of patients.¹⁶

In general, cochleosacculotomy is generally better tolerated compared to other surgical operations that definitively ablate vestibular function. Therefore, it can be the procedure of choice for patients who are at risk for the stress of postoperative vertigo, who are unfit to undergo general anesthesia, or elderly patients who otherwise would compensate poorly postoperatively. While labyrinthectomy, either via a transcanal or transmastoid approach, definitively ablates the neuroepithelium of all 5 sense organs for the control of vertigo and drop attacks, it has the disadvantage of producing severe postoperative vertigo and permanent hearing loss in the operated ear. Therefore, cochleosacculotomy may be a reasonable first surgical step towards the correction of vertigo in select patients with MD.

Surgical technique

In transcanal cochleosacculotomy, the procedure can be performed entirely under local anesthesia. The ear is prepped and draped in a standard fashion. Canal injections with lidocaine mixed with 1:100,000 epinephrine are made in 4 quadrants and the cartilaginous canal is dilated with a nasal speculum. The posterior bony canal is then exposed and injected with 1% lidocaine with 1:1000 epinephrine to achieve infiltration of anesthetic from 6 to 12 o'clock positions around the bony annulus.

An anteriorly-based tympanomeatal flap is then raised and the tympanic annulus is elevated from its bony sulcus. The ossicular chain and the chorda tympani nerve are preserved. An adequate surgical exposure includes visualization of the round window niche (RWN) and the posterior hypotympanum. In rare cases where the bony canal overhang partially obstructs the RWN, a small burr or curette may be used to remove the bony canal to improve visualization and surgical access.

A 3-mm right angled pick is then introduced through the round window membrane in the direction of the oval window (**Figure 1**). To ensure the cochlear duct is traversed, the pick is guided by hugging the lateral wall of the inner ear. If the niche is too small to accommodate the full width of the pick, the bony overhang of the RWN is carefully removed with a 2-mm diamond burr. When the pick has been fully introduced at a depth of 3 mm, the end of the pick is located deep to the stapes footplate. Occasionally, a slightly shorter (2 mm) pick may be used to avoid penetration into the vestibule and injury to the macula of the utricle. In rare circumstances where a high-riding jugular bulb is abutting the RWN, the procedure may need to be aborted due to the lack of complete access.

A slight loss of resistance can be palpated when the pick passes through the bony cochlear partition to result in fracture-disruption of the osseous spiral lamina and the cochlear duct (**Figure 2**). If the patient is awake, s/he may report momentary vertigo or hearing a "click" sound. Since the sense organs are not mechanically ablated or disturbed, the procedure does not result in new onset vertigo. The pick is then withdrawn and the round window membrane

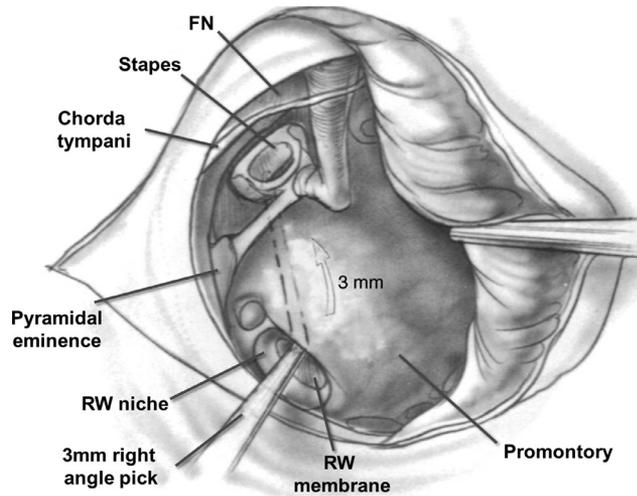


Figure 1 Transcanal cochleosacculotomy. A tympanomeatal flap is elevated and a 3-mm right angle pick is advanced through the round window membrane towards the oval window. The tip of the pick depicted in the figure is located just deep to the stapes foot plate. FN, facial nerve; RW, round window. Reprinted with permission from Brackmann D.E. et al *Otologic Surgery*, 2016.²¹

is sealed with either fascia graft, perichondrium, or adipose tissue. The tympanomeatal flap is then returned into position and the ear canal is packed with silk cloth or Gelfoam.

Postoperative complications are generally rare but include perforation of the tympanic membrane, sensorineural hearing loss, perilymphatic fistula, and otitis media.

Surgical outcomes

Initial results were reported by Schuknecht et al in a series of 51 patients, where 70% reported complete relief of symptoms and 88% reported significant symptomatic improvement at 4-month follow-up¹⁷. A follow-up study demonstrated stable relief of vertigo in 72% of patients at 22-month follow-up¹⁸. However, nearly a quarter (22 of 90, or 24%) of patients required revision surgery and 12% had profound hearing loss¹⁸. The largest series on outcomes following transcanal cochleosacculotomies consisted of 142 consecutive patients with a follow-up time ranged from 1 month to 7.4 years¹⁹. Definitive vertigo relief was achieved in 68.3% of the cases. Hearing was made worse in 35% of cases (as defined by the American Academy of Otolaryngology (AAO) criteria) at the time of most recent audiogram, and profound sensorineural hearing loss occurred in 11% of patients¹⁹. In comparison, in patients who undergo surgical labyrinthectomy, the cure rate for vertigo approaches 97%, although there is perhaps a greater portion of patients with postoperative

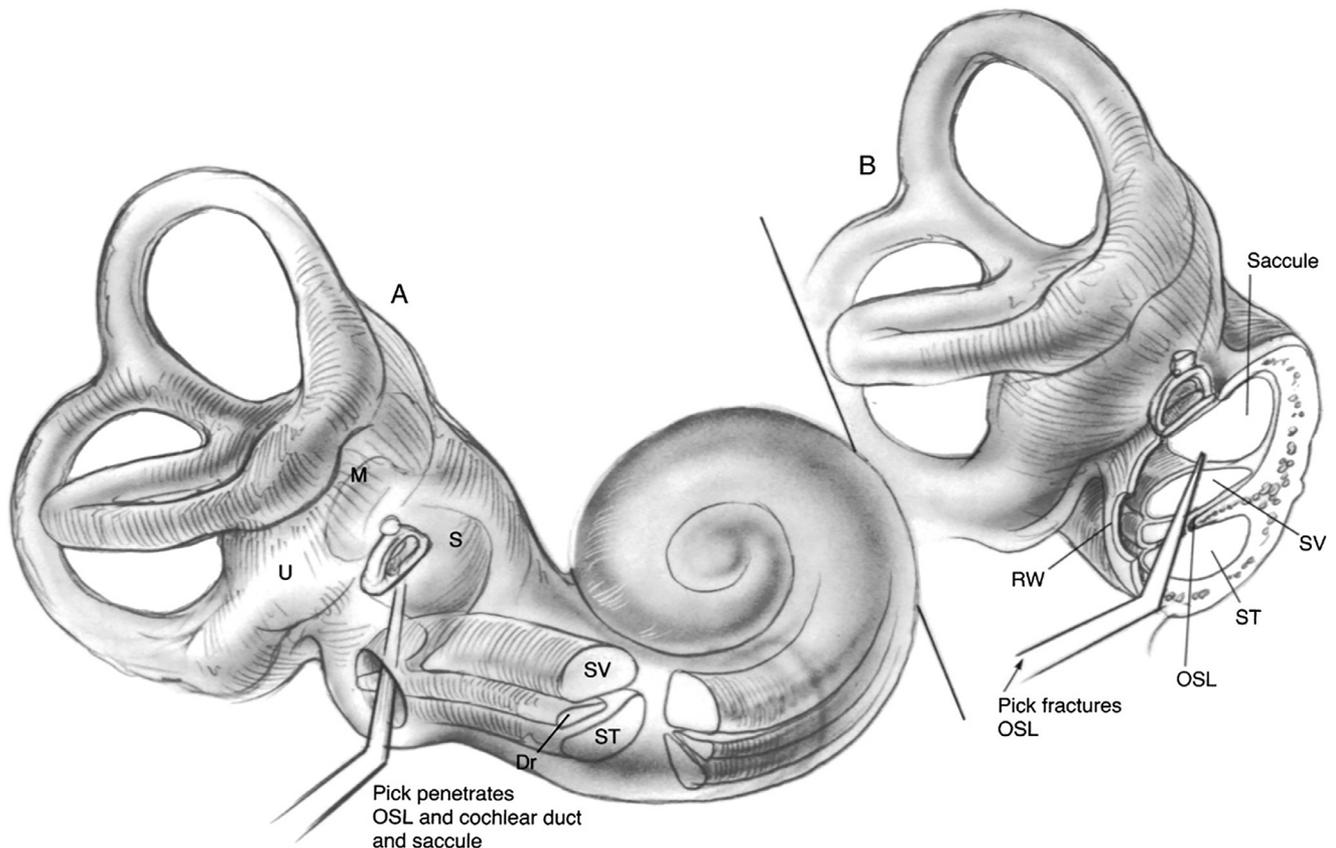


Figure 2 The 3-mm right angle pick is positioned to penetrate the osseous spiral lamina (OSL), cochlear duct, and saccule. Dr, ductus reunians; M, macula; RW, round window; S, sacule; ST, scala tympani; SV, scala vestibuli; U, utricle. Reprinted with permission from Brackmann C. et al *Otologic Surgery*, 2016.²¹

imbalance²⁰. Nevertheless, such interpretations should be made with caution as there are no large studies directly comparing the various surgical techniques.

Conclusions

Transcanal cochleosacculotomy is an effective surgical strategy alternative to vestibular neurectomy and labyrinthectomy in relieving vertigo symptoms in patients with disabling MD. Although rarely performed nowadays, the procedure presents low risk for significant postoperative complications such as Cerebral Spinal Fluid (CSF) leak or facial nerve injury and is generally well-tolerated.

Disclosures

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

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