

# Endolymphatic sac decompression and shunt placement



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## KEYWORDS

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Endolymphatic sac decompression and shunting is a nonablative surgical option for patients with intractable Meniere's disease. Sac surgery had a high success rate for vertigo control in this population with 75%-95% of patients experiencing improvement or resolution of their vertigo spells. The addition of high-dose steroid instillation into the sac has demonstrated improved vertigo rates. The risk of complications is low but includes hearing loss, persistent vestibular dysfunction, cerebrospinal fluid (CSF) leak or facial nerve injury. Decompression or shunt placement should be considered for all patients who fail dietary and medical therapies before considering ablative treatments. Published by Elsevier Inc.

## Introduction

The endolymphatic sac (ELS) is an anatomically and functionally intriguing structure which occupies the posterior aspect of the petrous pyramid where it is intimately associated with the layers of dura mater. It is connected via the endolymphatic duct (ELD) to the rest of the labyrinth.<sup>1</sup> It was first described by Cotugno in 1760 as a “sac” like structure which was in continuation with the ELD.<sup>2</sup> About a century later in 1869, Boettcher described the microscopic appearance of the ELS and a few years later in 1873, Carl Hasse coined the term

“saccus endolymphaticus” which has been used ever since to describe this structure.<sup>2</sup>

The purpose of this article is to review the anatomy and current understanding of the procedures aimed at decompression and drainage of the ELS for refractory Meniere disease (MD).

## Background and rationale

MD comprises a constellation of symptoms characterized by episodic vertigo, hearing loss, tinnitus, and aural fullness. It is a challenging diagnosis and a thorough history and work-up must be completed to rule out secondary causes of these symptoms.<sup>3</sup> MD was first described by Prosper Meniere in 1861.<sup>4</sup> In 1927, Georges Portmann reported on the importance of the ELS in the development of and persistence of vertigo. He proposed opening of the

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ELS as a potential nonablative procedure for MD.<sup>5</sup> This was based on numerous observations on the physiology of endolymph and the premise that blockage at the level of ELS was associated with development of MD.

Endolymphatic sac surgery has remained controversial since its first description. For example, in 1981 Thomsen et al published a “sham surgery” study showing no observable difference between patients with MD treated with endolymphatic sac decompression and shunting vs a simple mastoidectomy as control.<sup>6</sup> Both groups did report significant reduction in symptoms which the authors suggested resulted from the placebo effect. However, many studies since that time have documented the utility of ELS decompression. A critical analysis of outcomes by Sood et al showed effectiveness of ELS procedures in controlling vertiginous symptoms at both short and long term in at least 75% of patients who failed medical therapy.<sup>7</sup> In a recent survey, 80% of otologists offered ELS decompression after failure of standard medical therapy.<sup>8</sup> In our practice, ELS surgery is offered in cases with refractory vertiginous symptoms before resorting to destructive procedures.

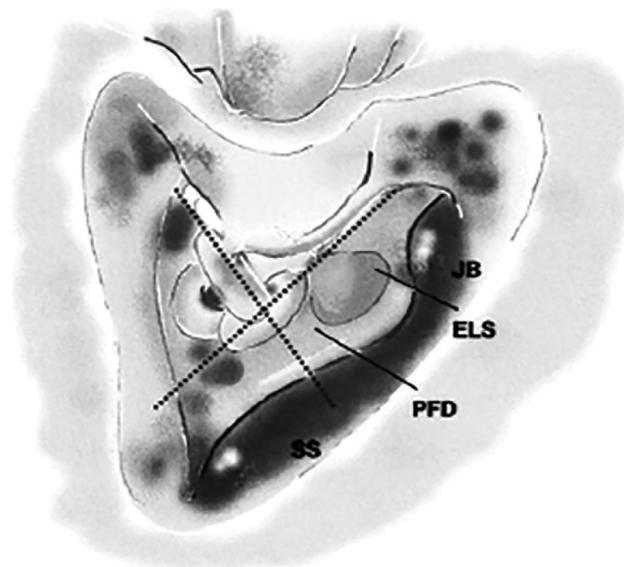
## Clinical work-up and diagnostic testing

Patients who present with symptoms related to potential MD are thoroughly assessed with clinical history, physical examination, formal audiometry and imaging of temporal bone (MRI) to rule out any secondary or retrocochlear causes. Particular attention is paid to exclude any contribution of vestibular migraine in the clinical picture. Although there is no diagnostic test highly specific for MD, balance function tests (videonystagmography, electrocochleography, and c-VEMP) and other adjunct testing are often obtained to further supplement the clinical diagnosis. These tests may aid in decision making algorithms regarding ablative vs nonablative therapy for refractory MD. The reader is referred to more comprehensive texts regarding vestibular testing and diagnosis for further reading.

## Surgical management

### Anatomy

There is considerable variation in the anatomy of the ELS and the ELD compared to rest of labyrinthine anatomy. The ED leads from the utricular and saccular ducts of the vestibule and courses via the bony vestibular aqueduct to terminate in the ELS within the operculum. The ELS has a proximal intra-osseous portion which is in continuity with the more variable extraosseous part.<sup>9</sup> ELS is a complex structure, composed of variable configuration of connecting tubules and cisterns. There is also variation in terms of size and location in relation to the Donaldson's line (imaginary line bisecting the posterior semicircular canal paralleling the long axis of lateral semicircular canal) (Figure 1).



**Figure 1** Anatomy of the endolymphatic sac (ELS). The ELS lays in the dura of the posterior fossa (PFD). It can be found anywhere below Donaldson's line (dashed horizontal line). JB, jugular bulb; SS, sigmoid sinus. Image by MTS.

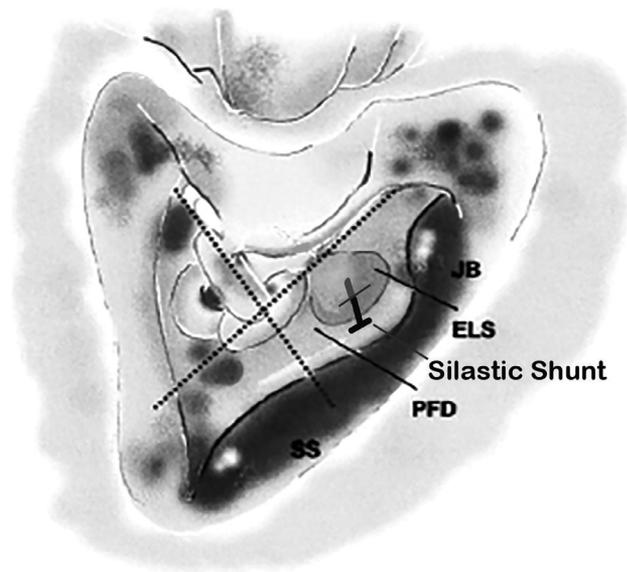
There is some evidence that a subset of MD patients have a narrow space between the posterior semicircular canal (SC) and the sigmoid sinus thus resulting in a relatively smaller ELS.<sup>10</sup> However, there is not a known causative anatomic variant for this disorder. A small posterior fossa plate may be anticipated preoperatively on a CT scan of the temporal bone.

### Informed consent

Differentiation from ablative therapy (labyrinthectomy, vestibular nerve section) needs to be clearly made. The informed consent process should include a detailed discussion of general risks of otologic procedures as well as specific risks such as hearing loss (risk of profound hearing loss in cases of superior semicircular canal (SCC) violation), spinal fluid leak, facial nerve injury, and sigmoid sinus/jugular bulb injury.

### Operative technique

After induction of general anesthesia, the patient is prepped and draped in the standard sterile otologic fashion. Facial nerve monitoring is performed during surgery with use of standard penetrating electrode electromyography. A postauricular incision is designed approximately 1 cm from the postauricular sulcus. The mastoid cortex is exposed and a cortical mastoidectomy is performed with the high speed drill. The sigmoid sinus is identified from the transverse sinus to the jugular bulb. The descending portion of the facial nerve is identified to the stylomastoid foramen. The posterior fossa bony plate is identified and exposed from the level of the horizontal SC inferiorly to the jugular bulb. This requires dissection of the retrofacial aircell track. The otic capsule around the posterior SC is



**Figure 2** The endolymphatic shunt. Silastic sheeting (0.40 mm thickness) is used to create the shunt. (A) The “T” shaped design prevents the shaft from migrating further in over time. (B) The shaft is gently inserted into the lumen of the sac after any adhesions are broken up with a Gimmick (portion inserted into the sac is outlined in grey).

exposed but the SC is not blue lined. Once the posterior fossa plate is fully exposed it can be decorticated with care being taken to not violate the dura. The junction of the ELD and ELS can generally be palpated as a band of dura extending toward the otic capsule that cannot be easily dissected from the bone. This junction is sometimes difficult to visualize directly as it is underneath the posterior semicircular canal (PSC) but can be palpated with an annulus elevator or other blunt dissecting tool. Because the extent of the ELS is extremely variable, the authors advocate decompressing the dura from the medial side of the sigmoid to the jugular bulb.

The ELS is a pearly white, thickened area in the dura inferior to the operculum. The sac can be opened for placement of a shunt at this point. To do this, an #11 blade or a Beaver blade can be used to incise the exterior leaflet of the sac. The interior of the sac is distinguished from merely splitting the dura by entering into a potential space which is lined by a very shiny tissue representing the rugae of the sac. This sac can be probed with an annulus elevator to break up any adhesions. The silastic shunt can then be fashioned. A “T” shaped silastic piece can then be inserted with the shaft of the “T” in the sac (Figure 2). The authors then infiltrate high-dose steroids (10 mg/ml dexamethasone) into the sac and allow the medication to sit in the cavity for 10 minutes before closing the wound. The wounds are closed in the standard fashion and the patient can generally be discharged the same day to home.

## Potential complications

As is the case for all surgery of the mastoid, complications such as facial nerve paralysis, hearing loss, dizziness, and CSF leak are all inherent risks. In most cases of endolymphatic sac decompression, the mastoid anatomy is pristine and not obscured by inflammatory disease making the risk for these complications lower than many other surgeries of the mastoid. The major complications of ELS decompression include CSF leak, sensorineural hearing loss, and exacerbation of vestibular dysfunction. Interestingly, one study actually revealed that a low frequency air-bone gap postoperatively correlated with a better resolution of vertigo.<sup>11</sup> Other less common complications include violation of the posterior semicircular canal and facial nerve injury. Sensorineural hearing loss may occur when the lumen of the ELS is opened or if there is inadvertent injury to the posterior or lateral SCC. Third window-related symptoms have been reported even when the posterior SCC is blue lined to fully decompress the ELS.<sup>12</sup> Injury to the facial nerve can happen specifically near the posterior SCC and proximal descending segment region. Vascular injuries include sigmoid sinus and jugular bulb injuries may be prevented with the use of diamond burs and meticulous attention to detail. In case of injury, extraluminal pressure with large pieces of gel foam is usually adequate to obtain hemostasis. Spinal fluid leak is a rare but serious complication which may need additional procedures. If a CSF leak is identified intraoperatively, repair should be attempted at that time. Small dural injuries can be plugged with muscle, fascia, and dural sealant. Layered repair, dural substitutes or mastoid obliteration should be considered if there is a large durotomy or high-volume leak.

## Surgical outcomes

Endolymphatic sac decompression has been shown to be effective in treating medically refractory MD in both the short and long term (>2 years) in greater than 75% of patients.<sup>7,13,14</sup> Patients may experience decreased intensity in the vertigo, prolonged intervals between the vertigo attacks or a complete resolution of vertigo attacks. In one prospective study, the mean number of vertigo events per month was decreased from 8.3 to 2.6.<sup>15</sup> The intraoperative use of local steroid administration, both intratympanic and direct endolymphatic sac instillation have been shown to improve vertigo control with better long-term hearing results.<sup>16,17</sup>

Approximately 5%-20% of patients do not benefit from ELS surgery. One possible reason would be that the sac has not been appropriately cannulated. The complex anatomy of the sac and its variable location is postulated to be the reason behind some surgical failures due to inability to fully decompress as well as inability to open the dominant lumen in cases of shunt procedures. Chung et al reviewed the histopathology of temporal bone specimens which had undergone shunt surgery. Of the 15 of bones reviewed, only 2 demonstrated actual cannulation of the sac although

many patients experienced improvement in their symptoms without accurate shunt placement.<sup>18</sup> Kitahara reported that accurate identification of the operculum, and consequently the ELS, improved hearing outcomes after sac surgery but did not change the vertigo control results.<sup>13</sup>

Although ELD/shunting is an invasive procedure it is considered nondestructive. The risk of immediate significant sensorineural hearing loss is low at 4%-5%.<sup>19</sup> Worsening of vertigo spells, either in intensity or shortened time between spells, is unusual with evidence in the literature that most patients experience no change in vestibular function or even a modest improvement in objective vestibular function.<sup>17,11</sup> The procedure may be offered to patients who have failed diet modification, diuretic therapy, and oral and intratympanic steroid therapy. ELS can be used for patients who have both serviceable and nonserviceable hearing as the goal of the procedure is vertigo control. The ELS procedures are a reasonable “next step” for patients who have failed conservative therapies before moving to ablative therapies such as chemical labyrinthectomy, vestibular neurectomy, or surgical labyrinthectomy. The reader is referred to more comprehensive resources for work-up and complex decision making regarding management of MD patients.

It is worthwhile to note that the natural progression of MD is a gradual improvement in symptoms with time. The threshold for surgical intervention is therefore high and the risks, benefits, and alternatives should be thoroughly discussed with each patient.

## Conclusion

Endolymphatic sac decompression with shunt placement is a safe and effective means to treat intractable vertigo associated with medically refractory MD. The surgery allows for preservation of hearing and is a nonablative treatment for MD. Additionally, steroid application to the ELS at the time of surgery improves vertigo control in the long term. Although the ELS decompression and shunt surgery is not 100% effective, many patients do experience improvement and thus ELS surgery should be included in the algorithm for treatment of medically refractory MD.

## Disclosure

Authors declare no conflict of interest.

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