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An Update on the Surgical Treatment of Ménière's Diseases

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Abstract

Various surgical procedures have evolved for the treatment of Ménière's disease patients who are refractory to medical therapy. These can be in the form of conservative surgeries as in endolymphatic sac surgery or in the form of destructive procedures. The latter include labyrinthectomy and vestibular nerve section, which result in deafferentation of the vestibular end organ. In the past decade, intratympanic therapy with gentamycin and steroids has evolved as new methods for the treatment of Ménière's disease. The literature witnesses controversies in the effectiveness of endolymphatic sac surgery in the treatment of Ménière's disease. The choice of the surgical procedure depends on the degree of serviceable hearing, the severity of the spells, and the condition of the contralateral ear. The most commonly used procedures in the treatment of Ménière's disease and their indications are reviewed.

Key Words: Endolymphatic sac surgery, gentamycin, intractable vertigo, intratympanic therapy, Ménière's disease, steroids, vestibular nerve section

Sumario

Se han desarrollado varios procedimientos quirúrgicos para pacientes con enfermedad de Ménière quienes son refractarios a la terapia médica. Estos pueden ser enfoques conservadores como en la cirugía del saco endolinfático, o en la forma de procedimientos destructivos. Estos últimos incluyen la laberintectomía y la sección del nervio vestibular, lo que produce una interrupción en la aferencia del órgano periférico vestibular. En la última década, la terapia intra-timpánica con gentamicina y esteroides ha evolucionado como un nuevo método de tratamiento de la enfermedad de Ménière. La literatura muestra controversias en la efectividad de la cirugía del saco endolinfático para el tratamiento de la enfermedad de Ménière. La escogencia del procedimiento quirúrgico depende del grado de audición útil, de la severidad de los ataques, y de las condiciones del oído contralateral. Se revisan los procedimientos más comúnmente utilizados en el tratamiento de la enfermedad de Ménière y sus indicaciones.

Palabras Clave: Cirugía del saco endolinfático, gentamicina, vértigo intratable, terapia intra-timpánica, enfermedad de Ménière, esteroides, sección del nervio vestibular

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To date, there is no specific cure for Ménière's disease. This can be attributed to the fact that in only a few cases can the etiology of this disease be determined. One of the well-accepted theories behind the underlying pathophysiology of Ménière's disease, however, is endolymphatic hydrops, which can only be proved histopathologically in temporal bone studies. There are mainly two proposed mechanisms underlying endolymphatic hydrops, (1) obstruction of the endolymphatic duct and (2) sac system and dysregulation of endolymph production.

The classical symptom complex in Ménière's disease consists of recurrent episodic attacks of vertigo, tinnitus, and a low-frequency fluctuating sensorineural hearing loss with aural fullness. In nonclassical forms of Ménière's disease, patients may initially present with vertigo and no hearing loss or with fluctuation in hearing in the absence of vertigo spells. Most patients respond to conservative medical management; however, in about 10–20% of patients, surgical treatment is necessary (Brown, 1983; Glasscock et al, 1984).

The surgical management of Ménière's patients who are refractory to medical therapy has evolved in the past decade due to the introduction of less invasive office procedures, mainly the intratympanic therapies. The other classical surgical procedures used in the treatment of patients with intractable Ménière's disease include endolymphatic sac decompression and shunt, vestibular nerve section and labyrinthectomy. The choice depends on the degree of serviceable hearing, the severity of the spells, and the condition of the contralateral ear. The patient's age and health status may also affect the compensatory potential and should be taken into consideration when ablative procedures such as a chemical or surgical labyrinthectomy or a vestibular neurectomy are entertained.

Surgical Therapy for Vertigo Control

Ménière's disease results in various degrees of damage to the vestibular end organ, causing asymmetric input to the brain. This will disrupt the resting tone of the vestibular system and will manifest itself as vertigo, dizziness, and disequilibrium. Normally, the brain tends to compensate for, or adjust to, this asymmetry, which results in symptom relief. In Ménière's disease, the vestibular end organ is subject to recurrent insults, which will make central compensation difficult to achieve. Surgical procedures aim at controlling the vertigo spells so that central compensation can take place.

Surgical approaches can be divided into destructive and nondestructive procedures. Nondestructive procedures, such as endolymphatic sac decompression, aim at reversing the pathophysiological hydrops and preserving the hearing. Destructive procedures on the other hand result in vestibular deafferentation of the inner ear. These include labyrinthectomy, either chemical or surgical, and vestibular nerve section. Hearing preservation is a goal with vestibular nerve section and could be achieved to a certain extent with chemical labyrinthectomy; however, in surgical labyrinthectomy, loss of residual hearing is definite.

Surgical treatment in Ménière's disease patients who have no serviceable hearing is straightforward and consists of labyrinthectomy either chemical or surgical. Chemical labyrinthectomy is achieved by the use of intratympanic gentamicin, which is known for its vestibulotoxic effects. Inner ear perfusion with gentamicin, titrated to the patient's response and monitored for early signs of hearing loss, is now widely used in patients with serviceable hearing.

In patients with bilateral Ménière's disease, destructive surgeries that result in complete deafferentation of the vestibular end organ should be avoided since bilateral ablation of the labyrinths could result in some serious consequences like oscillopsia and permanent imbalance. Oscillopsia is a perception of bouncing of the visual field with walking. Although the true prevalence of bilateral Ménière's disease is not clear (ranging from 2–78% [Balkany et al, 1980]), its existence should be taken into consideration during patient counseling and choice of surgical procedure.

Endolymphatic Sac Surgery

Endolymphatic sac surgery is a conservative approach that involves decompression of the endolymphatic sac from overlying bone and drainage of its endolymph while keeping the vestibular neuroepithelium and its innervation intact. Endolymphatic sac surgery in theory has a direct effect on the pathophysiology of the inner ear by restoring normal endolymphatic pressure and thus

addresses both cochlear and vestibular dysfunctions. However, its actual ability to relieve symptoms in Ménière's disease has been very controversial in the literature. Opinions range from enthusiastic support (Paparella and Fina, 2002) to the belief that it is no more effective than placebo (Thomsen et al, 1981; Pillsbury, 1983; Bretlau et al, 1989; Thomsen et al, 1998). Despite this controversy, endolymphatic sac surgery is still a commonly performed procedure (Silverstein et al, 2003).

The most common indication of endolymphatic sac surgery is intractable vertigo in patients with classic Ménière's disease where the patient presents with vestibular complaints, cochlear complaints, and aural fullness. Vertigo spells are usually debilitating, nonresponsive to medical treatment, and associated with nausea and vomiting.

Perhaps the most widely accepted indication of endolymphatic sac surgery is in patients with bilateral Ménière's disease due to the conservative nature of this procedure and the preservation of end organ anatomy and physiology. In patients with bilateral Ménière's disease, most clinicians are hesitant to perform destructive surgery if the remaining only-hearing ear may worsen in the future.

Proponents of endolymphatic sac surgery believe in the role of revision sac surgery in patients who had good primary results from endolymphatic sac surgery but experienced recurrence of symptoms after a period of time (Paparella and Sajjadi, 1988). The theory behind this is that iatrogenic osteogenesis and formation of scar tissue reestablishes the pathogenic state (Paparella and Sajjadi, 1988; Huang and Lin, 1991; Huang and Lin, 1994). Correction of these resulting lesions will provide symptom relief (Paparella and Sajjadi, 1988).

The technique of endolymphatic sac decompression surgery has evolved over time from endolymphatic subarachnoid shunt to the more commonly used and equally effective endolymphatic mastoid shunt (Brackmann and Nissen 1987). In the latter, nanoliters of endolymph are longitudinally drained from the sac into the mastoid cavity in an attempt to decrease the endolymphatic pressure. Various types of shunting materials have been tried for this purpose including the use of Gelatin sponge, Teflon wick, Arenberg inner ear valve, Silastic strips, and many others. It has been observed that, in patients in whom the endolymphatic sac could not be localized, decompression of surrounding dura gave positive results, which led to the thinking that endolymphatic sac decompression is a crucial part of the surgery. It is possible that the removal of the overlying bone reduces the external pressure and allows for better flow of endolymph into the sac.

Postoperative perisaccular fibrosis (fibrous tissue formation around the area of the sac) and osteogenesis (new bone formation) can occur, compromising endolymphatic drainage and symptoms control. Huang (2002) noticed that the incidence of relatively severe perisaccular fibrosis is strongly correlated with the recurrence of symptoms of Ménière's disease after primary surgery. In an attempt to prevent perisaccular fibrosis there have been some reports using intraoperative topical Mitomycin C to the endolymphatic sac area.

The reported vertigo control rates after endolymphatic sac surgery range from better than 90% (Huang, 1987; Huang, 1999; Paparella and Fina, 2002) to not being different from placebo or the natural history of the disease. Other studies reported 50 to 75% vertigo control rate after endolymphatic sac surgery (Glasscock et al, 1984; Brackmann and Nissen, 1987; Monsell and Wiet, 1988; Moffat, 1994; Quaranta, 1998). Despite this uncertainty in efficacy, endolymphatic sac surgery remains one of the commonly performed procedures that is nonablative with hearing preservation properties.

Endolymphatic sac surgery has a small risk to residual hearing, up to 2% (Paparella and Sajjadi, 1987). Bleeding from the lateral sigmoid sinus and cerebrospinal fluid leak are other potential complications in endolymphatic sac surgery. Conductive hearing loss after endolymphatic sac surgery has been reported and is thought to be secondary to bone dust making its way to the middle ear (Paparella and Sajjadi, 1987).

Vestibular Nerve Section

Vestibular nerve section is performed in patients with Ménière's disease who are refractory to medical therapy. The two main objectives of this procedure are the elimination of vertigo and preservation of hearing. These can be achieved by selectively cutting the vestibular portion of the eighth nerve (CN VIII), keeping the cochlear portion intact, thus preventing the vestibular afferent stimuli from reaching the brain. Even though this surgery

does not act on the pathophysiology of Ménière's disease, it eliminates its most disturbing symptom, the vertigo.

Vestibular nerve section is indicated in patients with unilateral Ménière's disease who have intractable vertigo resistant to medical or other conservative measures. Proponents of endolymphatic sac surgery consider vestibular nerve section as a last resort. Others will perform vestibular nerve section after failure of medical therapy or intratympanic therapy in patients with serviceable hearing without the consideration of endolymphatic sac surgery.

Objective confirmation of unilateral disease by complete audiological examination, electronystagmography and electrocochleography are crucial prior to vestibular nerve section. Bilateral peripheral vestibular disease is a contraindication due to the disability resulting from bilateral loss of vestibular input to the brain, mainly oscillopsia and permanent imbalance. Central compensation after vestibular neurectomy is key for postoperative recovery of balance. This makes any indication of central nervous system disease such as cerebellar dysfunction, multiple sclerosis, physiologic old age, and poor medical condition a relative contraindication for vestibular nerve section. Preoperative patient selection in the elderly population is critical because of a higher frequency of associated disorders (e.g., lower extremity neuropathies, diabetes mellitus, ischemic brain disease), which can have an effect on the postoperative balance function and compensation. However, there have been recent reports of the benefits and safety of vestibular nerve section in elderly patients with satisfactory medical condition (Ballester et al, 2002).

Vestibular nerve section involves a craniotomy to gain access to the cochleovestibular nerve. A variety of surgical approaches are available. The most commonly used ones are the retrolabyrinthine approach, or the retrosigmoid approach. Delineation of the vestibulocochlear cleavage plane (plane between the vestibular part of the eighth nerve and the cochlear one) in the posterior fossa is critical to the selective sectioning of the vestibular nerve fibers.

Vestibular nerve sectioning is one of the most effective procedures for treating intractable vertigo in patients with unilateral Ménière's disease and serviceable hearing. Vertigo control rates between 78% and over 90% have been reported in the literature (Silverstein et al, 1990; Glasscock et al, 1991; McKenna, 1996; Molony, 1996; Rosenberg, 1999). Failure of response has been attributed to incomplete sectioning of the vestibular nerve fibers to avoid injury to the cochlear part of the eighth (Rosenberg et al, 1991), and inability to identify vestibulocochlear cleavage plane. Persistent or recurrent vertigo after vestibular nerve section can be treated by intratympanic gentamicin.

Potential complications, although uncommon, do exist with vestibular nerve section. They include hearing loss, facial nerve paralysis, cerebrospinal fluid leak, and headache.

Labyrinthectomy

Labyrinthectomy involves surgical exenteration of the labyrinth neuroepithelium in an attempt to eliminate vertigo and allow the central compensation process to take place. Hearing loss is inherent in this procedure and, as such, it is indicated in patients with severe-to-profound hearing loss, very poor word recognition, and intractable vertigo. Because of its destructive nature, labyrinthectomy is performed in unilateral cases only. Similar to patients undergoing vestibular nerve section, objective confirmation of unilateral disease by complete audiological examination, electronystagmography, and electrocochleography is crucial. As with vestibular nerve section, bilateral peripheral vestibular disease is a contraindication to labyrinthectomy due to the disability resulting from bilateral loss of vestibular input to the brain mainly oscillopsia and permanent imbalance. Central compensation after labyrinthectomy is therefore a key for postoperative recovery of balance. This makes patients with central nervous system disease, physiologic old age, and poor medical condition a contraindication for labyrinthectomy. There have, however, been reports of the benefits and safety of surgical labyrinthectomy in elderly patients when carefully selected (Langman and Lindeman, 1998; Schwaber et al, 1995). Excellent vertigo control rates up to 97% have been reported in the literature after labyrinthectomy (Hammerschlag and Schuknecht, 1981; Benecke et al, 1986; Levine et al, 1990). Potential but infrequent complications after a labyrinthectomy are facial nerve injury (2%) and CSF leak in 3% (Graham and Colton,

1980). Due to the possible bilateral involvement in patients with Ménière's disease, concerns exist that labyrinthectomy might compromise the results of cochlear implants if needed in the future (Paparella and Sajjadi, 1987). However, some reports have demonstrated that there is improvement in sound awareness, speech recognition, and communication after cochlear implantation in previously labyrinthectomized ears (Lustig et al, 2003).

Intratympanic Gentamicin Therapy

There has been considerable recent interest in the role of intratympanic therapy in the treatment of inner ear diseases. Medication instilled into the middle ear is absorbed via the round window into the fluid system of the inner ear. This mode of treatment has the advantage of targeting the inner ear system without exposing the body to the systemic side effects of the medication in use. Gentamicin, being toxic to the vestibular part of the inner ear, achieves "chemical" labyrinthectomy when used intratympanically and could be an alternative to surgery in patients with intractable vertigo, thus avoiding the morbidity associated with the other surgical procedures. The advantages of intratympanic gentamicin therapy include the fact that it can be performed as an office procedure and that symptom control can occur before total ablation of the vestibular part of the inner ear. On the other hand, intratympanic gentamicin therapy has the risk of hearing loss, which most of the time depends on the cumulative dose used. This can be minimized by meticulously titrating the dose of gentamicin to vertigo control and cessation of therapy to the earliest signs of hearing loss (subjectively and objectively by repeated hearing tests).

Intratympanic gentamicin, however, has its own controversies concerning the dose, duration, and frequency of injections. The authors use a gentamicin dose of 10 or 40 mg/ml once weekly and titrate the frequency and number of injections to the patient's response or to the earliest signs or symptoms of hearing loss, whichever occurs first. Patients should be followed up closely by careful history and physical examination to detect the presence or absence of hearing loss, disequilibrium, motion intolerance, and nystagmus. Repeated hearing tests to detect early signs of hearing loss are recommended. In patients with no serviceable hearing, higher doses of gentamicin can be used (40 mg/ml) since hearing preservation is not of considerable concern

Intratympanic gentamicin therapy is the least invasive surgical intervention in the treatment of intractable vertigo in Ménière's disease. If successful, it will avoid the morbidity of craniotomy and other surgical procedures. In patients with no serviceable hearing, intratympanic therapy will be an ideal alternative. Although it still can be used in patients with serviceable hearing, the patient should be aware of the risk of further sensorineural hearing loss associated with this technique. Intratympanic gentamicin therapy is an option in patients with refractory Ménière's disease after failed endolymphatic sac surgery in an attempt to avoid the need for vestibular neurectomy.

Vertigo control was reported in the range of 71.4–100% with hearing loss in the range of 0–81.8% (Assimakopoulos and Patrikakos, 2003; Hoffmann and Silverstein, 2003). Hearing loss occurs in 30% of patients (Hoffmann and Silverstein, 2003). When successful, intratympanic gentamicin therapy will decrease the number of patients requiring more invasive surgical procedures. However, the gentamicin concentration, route, and frequency of administration have varied in the literature. In addition, the frequency of secondary hearing loss has ranged from 7% to 81.8% of patients (Assimakopoulos and Patrikakos, 2003).

Surgical Therapy for Hearing Loss

To date, neither medical nor surgical therapy has provided a long-term effect on reversing or preventing hearing loss in patients with Ménière's disease (Kinney et al, 1997; Quaranta et al, 1997). Currently, the only available surgical options that can potentially address hearing loss in patients with Ménière's disease are the endolymphatic sac surgery and the intratympanic steroid therapy. Endolymphatic sac decompression surgery addresses the pathophysiology of the inner ear, and thus, its positive effect on cochlear function is expected. Some believe that this can be best achieved when the inner ear is still in a phase of temporary shift (Morrison, 1983). The effect of endolymphatic sac surgery on hearing stabilization or improvement varies in the literature. Some studies reported 55–85% hearing stabilization or improvement rates after two years' follow-up (Goldenberg and Justus, 1983; Monsell and Wiet, 1988; Huang

and Lin, 1995). Others think that endolymphatic sac surgery does not modify the natural course of Ménière's disease with respect to hearing and that preoperative hearing fluctuation does not improve the outcome (Goin et al, 1992).

Steroids are anti-inflammatory agents that have been employed in the regulation of immune disorders, which are thought to be an etiologic factor in some patients with Ménière's disease. Intratympanic steroid injections (dexamethasone or methylprednisolone) are being currently used in patients with Ménière's disease where systemic steroids are contraindicated or in patients who fail to respond to oral steroids. In this procedure, steroids are perfused or injected into the middle ear and absorbed into the perilymph via the round window membrane. Steroids were found in higher concentration in inner ear fluids when injected into the middle ear than when administered systematically (Parnes et al, 1999). Clinically, the success rate of intratympanic steroids is variable in the literature, which can be attributed to the wide variation in the dose, frequency of administration, and the fluctuating nature of the hearing loss in Ménière's disease. In a randomized, double-blind, crossover study in patients with Ménière's disease who underwent intratympanic dexamethasone therapy, no significant improvement in hearing was reported (Silverstein et al, 1998). Shea and Ge (1996) reported 67.9% rate of hearing improvement with intratympanic steroid injection and systemic administration, which was later decreased to 35% in patients with long-term follow-up (Shea, 1997). Barrs et al (2001) reported no benefits on hearing with intratympanic dexamethsone (4 mg/ml) in patient with Ménière's disease. Hillman et al (2004) reported 40% short-term improvement in hearing after intratympanic dexamethasone (16 mg/ml) in patients with Ménière's disease. Randomized placebo control studies are required in order to determine the role of intratympanic therapy in patients with Ménière's disease.

SUMMARY

Vertigo remains the most disabling symptom for most patients with Ménière's disease. Vestibular nerve section and labyrinthectomy have been associated with the highest control of vertigo. Intratympanic gentamicin therapy is commonly used for vertigo control with good sucess rate. Success rates with endolymphatic sac decompression and shunt surgery has been variable in the literature. Unfortunately, no treatment to date has proven to alter the natural course of disease with respect to cochlear symptoms. Prospective studies are needed to better evaluate the effect of various surgical techniques on the natural history of the disease.

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