Surgical Management of Intractable Meniere’s Disease

Pietro De Luca1, Massimo Ralli2*, Claudia Cassandro3, Francesca Yoshiie Russo2, Federico Maria Gioacchini1, Matteo Cavaliere1, Marco Fiore4, Antonio Greco2, Ettore Cassandro1, Alfonso Scarpa1

ABSTRACT

Meniere’s Disease (MD) is an inner ear disorder characterized by spontaneous recurrent vertigo, fluctuating sensorineural hearing loss, aural fullness and low-pitch tinnitus. Therapeutic management of MD includes dietary restriction and medical therapy. A minority of cases is characterized by frequent vertigo attacks, progressive hearing loss and persistent tinnitus even through the continuous medical treatments; this condition is called intractable MD and requires a therapeutic escalation from non-invasive medical treatment to surgical intervention. Invasive procedures include endolymphatic sac surgery, vestibular nerve section and labyrinthectomy. These procedures have a very high success rate on symptom control but may have a severe impact on the hearing function. However, the simultaneous combined approach of demolitive surgery and cochlear implantation may be a valid approach to treat symptoms of intractable MD and preserve hearing function. In the present study, we review current literature focusing on intractable MD to describe and discuss advantages and disadvantages of established and newly proposed surgical treatments for intractable MD.

Keywords: Meniere disease, Intractable meniere, Vertigo, Hearing loss, Tinnitus.

1Department of Medicine and Surgery, University of Salerno, Salerno, Italy
2Department of Sense Organs, Sapienza Università di Roma, Rome, Italy
3Department of Otolaryngology, University of Turin, Turin, Italy
4Institute of Cell Biology and Neurobiology - IBCN CNR, Italy

*Send correspondence to
Massimo Ralli
Department of Sense Organs, Sapienza University of Rome, Italy, Viale del Policlinico 155, 00162 Roma, Italy. Email: Massimo.ralli@uniroma1.it; Tel: +39 0649976808
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INTRODUCTION

Meniere’s Disease (MD) is an idiopathic inner ear disorder, first described by Prosper Meniere in 1861, characterized by spontaneous recurrent vertigo, fluctuating Sensorineural Hearing Loss (SNHL), aural fullness and low-pitch tinnitus. One of the most debilitating aspects for MD patients is the recurrence of acute and long-term vertigo attacks that are often debilitating and may severely affect quality of life, similarly to other conditions.

The pathologic mechanism of MD is still unclear; however, Endolymphatic Hydrops (EH) of the scala media of the cochlea is one the currently known pathophysiological mechanisms that underlie MD. EH seems to be due to an overproduction of endolymph and/or a decrease in the absorption of endolymph based on histopathological studies. Several theories have been proposed to explain the cause of MD, including viral infections and immune system-mediated mechanisms. An autoimmune origin of MD based on inner ear antigens has been reported in up to one third of patients, although the immunological mechanisms involved are not clear.

MD diagnosis is based on the criteria of the Barany Society.

There are several therapeutic options for MD, but none is unanimously considered effective by the scientific community. First-line treatment commonly includes dietary modifications such as restriction of salt, caffeine and alcohol intake; however, there is no evidence from randomized controlled trials that supports the effectiveness of such approach in MD. The restriction of glucose intake for MD has been recently proposed; in a review from our group, we remarked the possible role of hyperinsulinemia in subjects affected by MD. There is actually strong evidence that the saccule, which is the main labyrinthine structure affected by pathological damage due to EH, has a large number of insulin receptors. This observation was confirmed by the examination of cadaveric subjects and in vivo analysis by cervical Vestibular Evoked Myogenic Potentials (cVEMPs).

Several drugs have been proposed for the treatment of MD. Dimenhydrinate and benzodiazepines have been proposed for acute attacks, while betahistine, beta-blockers and diuretics as prophylactic therapy, although evidence of their efficacy is lacking. Specially processed cereals (SPC) that increase endogenous antisecretory factor synthesis have been proposed to improve symptoms of MD with controversial results.

If of first-line treatment which does not guarantee a good symptom control, intratympanic administration of drugs such as corticosteroids or gentamicin has been proposed as a treatment option. Corticosteroids can lower the risk of hearing damage, but showed less efficacy to control vertigo attacks compared to gentamicin. Gentamicin has been proven as an effective treatment for vertigo in MD with a potential risk of hearing loss. Some authors suggest the use of low-dose gentamicin, in which the drug is injected once and further treatments are only performed in cases of recurrent vertigo attacks; others prefer high-dose gentamicin, titration or continuous administration in which the drug is injected until vestibular weakness is reached. Hearing loss and healthy-side vestibular dysfunction are potential risks of gentamicin treatment due to its ablative nature and ototoxicity.

A minority of cases is characterized by frequent vertigo attacks, progressive hearing loss and persistent tinnitus even through continuous medical treatments; this condition is called intractable MD and requires a different treatment approach. Management of MD should follow a therapeutic escalation from non-invasive medical treatment to surgical intervention including ablation. Invasive procedures, such as vestibular nerve section or labyrinthectomy, may be suggested in case of medical treatment failure. Surgery must be preceded by hearing assessment, ipsi and contralateral vestibular function assessment and medical imaging. Recently, an electronic questionnaire formulated by Quarta et al and sent to Italian otolaryngologists showed that refractory case of MD are treated initially with intratympanic steroids followed by gentamicin; in case of failure of intratympanic treatment, vestibular nerve section is the treatment of choice.

In the present study, we review current literature to describe and discuss advantages and disadvantages of established and newly proposed surgical treatments for intractable MD.

Surgical Management of Meniere Disease: Several surgical approaches have been described for intractable MD. Endolymphatic sac surgery was first described by Portman in 1926; and several variations on this surgical technique have been suggested. They include simple decompression, cannulation of the endolymphatic duct, endolymphatic drainage to the subarachnoid space, wide decompression that includes the sigmoid sinus, drainage to the mastoid, and removal of the extracerebellar portion of the sac. Prostheses to allow flow selectively in either the mastoid or subarachnoid direction have also been proposed, including simple silastic sheet apposition to tubes and one-way valves.

Endolymphatic Sac Surgery: The role of endolymphatic sac surgery in intractable MD cases has been widely discussed. A Brazilian retrospective study from Bent et al included 95 patients who underwent endolymphatic sac drainage. In patients with unilateral MD, the authors described a satisfactory vertigo control in 94.3% of patients, a significant improvement in cochlear function in 14%, and hearing preservation or even improvement in 88% of patients with intractable MD. In patients with bilateral MD, vertigo control was obtained in 85.7% of patients, cochlear function improved in 28% and hearing preservation or even improvement was reported in 71% of patients. The authors concluded that endolymphatic sac drainage can be considered a good surgical option.
for patients with intractable MD, with a high percentage of vertigo control and hearing preservation. The results were also supported by a study from Flores Garcia et al.60; the authors confirmed that endolymphatic sac surgery including its variants can be a good option for patients with incapacitating endolymphatic hydrops.

Nevertheless, the efficacy of this procedure remains controversial, and the evidence to support this surgery is low. Thomsen et al.61 conducted a double-blind, placebo-controlled study which compared mastoidectomy alone and endolymphatic shunt; results showed no significant differences between the two surgical approaches. Recently, Gibson et al.62 compared endolymphatic shunt surgery and intratympanic gentamicin in patients with MD: the authors demonstrated that endolymphatic shunt surgery had a successful vertigo control comparable with intratympanic gentamicin, with a lower incidence of audio-vestibular complications. A Cochrane review by Pullens et al.63 over two randomized controlled studies showed no significant effects of endolymphatic sac surgery on symptom control, and concluded that there was no sufficient evidence for the beneficial effect of this treatment. A systematic review by Devantier et al.64 concluded that there is still a lack of high-quality research that supports the role of endolymphatic sac surgery in providing a significant amount of symptomatic relief for patients with MD. In addition, Taeko et al.65 studied 21 patients who received endolymphatic sac surgery and found no correlation between the changes in hearing function and the volume of endolymphatic hydrops after surgery. Furthermore, intra-sac steroid injection during surgery did not result in further improvement in patient outcomes66. In a study performed by Higashi-Shingai et al.67 two years after sac surgery, the authors showed that sac surgery could reduce vestibular endolymphatic hydrops68. However, histologic evidence revealed that hydrops is not relieved after shunt placement69 and that there was no relationship between changes in hearing function and volume of endolymphatic hydrops after endolymphatic sac drainage.

Mattingly et al. evaluated the use of intraoperative Electrocochleography (ECochG) in patients with MD undergoing endolymphatic sac surgery. The authors detected only small objective changes in the low-frequency SP magnitude (500 Hz) immediately after surgery, but not in other frequencies or measures tested, suggesting that only minimal electrophysiological changes occurred in the cochlea as result of endolymphatic sac surgery70. Vestibular Nerve Section: Vestibular nerve section is widely used for symptom control in patients with intractable MD and is the fifth line of management for MD according to the European Statement on Meniere’s disease71.

Several studies focused on the efficacy and safety of this technique, demonstrating that vestibular nerve section, with differences based on the surgical approach, can reach a complete vertigo control in 85% to 95% of patients and hearing preservation in 80% to 90% of patients after the procedure72, 73. Although the procedure showed a higher vertigo control rate compared to endolymphatic shunt, it was more invasive and technically challenging procedure. Two retrospective studies from Lemnos et al.73 and Chen et al.74 showed that vestibular nerve section can be considered an effective treatment in the case of intractable MD, with good functional results and low failure rate.

The risk of hearing loss in vestibular nerve section procedure has been shown to be lower compared to gentamicin injection75, although the risk of hearing loss following gentamicin treatment seems to be most significant with high-dose protocols.

Labyrinthectomy: Labyrinthectomy is the most destructive procedure for treatment of MD because of the total damage of hearing and vestibular function. Indications of this procedure include patients without residual hearing or subjects that did not benefit from conservative treatment options. The procedure has a higher rate of vertigo improvement compared to vestibular nerve section76 and has been reported to improve quality of life in the majority of patients77. The procedure is mainly performed using a transmastoid exposure, although a transcanal approach has been described and is rarely used.

To treat the total hearing loss following the procedure, Heywood and Atlas proposed a simultaneous cochlear implantation and labyrinthectomy. They performed the surgery on two female patients with advanced MD and described several benefits of the simultaneous procedure such as the prevention of implantation of a fibrous or ossified cochlea, the decrease in the duration of deafness, and the use of a single operative procedure78.

Todt et al. reported a labyrinthectomy performed after cochlear implantation79; to date this is the only report in the literature.

Other Surgical Treatments: Some authors suggest the insertion of a ventilation tube in the tympanic membrane as first choice of treatment for refractory MD80. A British work by Kanegaonkar et al, treated 33 patients with early grommet insertion with intratympanic steroid injection, combined with customized vestibular rehabilitation; they suggested that this combination may provide an alternative first-line strategy for MD, preventing further true MD attacks. Compared to established surgical treatments of MD such as endolymphatic shunt surgery and vestibular neurectomy, transtympanic ventilation tube placement is a procedure with extremely low risks and comorbidities. In selected patients, ventilation tube placement might anticipate and postpone more invasive treatments81.

In the recent years, new surgery techniques for intractable MD have been proposed. Endolymphatic duct blockage has been first proposed in 201582 and consists in a non-ablative surgical technique. The initial results of this surgical option showed no clinical cochlear...
and vestibular damage and a significantly better control of the vertigo attacks in comparison to the traditional endolymphatic sac surgery\textsuperscript{83}. In addition, there were no significant complications or side effects, as confirmed by some recent works\textsuperscript{83,84}.

Another novel technique that has been proposed is tenotomy of the tendons of the stapedius and tensor tympani muscles. The use of this technique, proposed by a Belgian Otolaryngology Unit leaded by De Valck\textsuperscript{85}, had some benefits in intractable MD patients as demonstrated by studies from Reichmayr et al\textsuperscript{86}, Albu et al\textsuperscript{87} and Loader et al\textsuperscript{88}. Results included an improvement not only of vestibular symptoms, but also of hearing thresholds.

Zhang et al proposed a triple semicircular canal plugging for the treatment of intractable MD; according to the authors, this surgery offered a total control rate of vertigo for the treatment of intractable MD; according to the authors, this surgery offered a total control rate of vertigo and could represent an effective therapy for the advanced state of this disorder\textsuperscript{89}.

An interesting and recent study from Attanasio et al speculated the association between MD and chronic cerebrospinal venous insufficiency and evaluated the efficacy of bilateral percutaneous transluminal angioplasty of the jugular/azygos veins compared to medical therapy. The encouraging responses to vascular interventional therapy on MD symptoms suggest that this may be a promising treatment option for interpretation and treatment of this complex disease.

CONCLUSION

According to International Guidelines, surgical therapy for MD is recommended only for refractory disease, and therefore represents the third (or even the fifth) line of management.

Currently, the most popular surgical procedures used to control vertigo attacks are also the most invasive and have a significant impact on auditory function. However, the simultaneous combined approach of demolitive surgery and cochlear implantation may be a valid approach to treat symptoms of intractable MD and preserve hearing function.

STATEMENT DISCLOSURE

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