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Surgery for Menière's Disease

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Abstract Menière's disease is a chronic disease of unknown cause, with a capricious natural course. It is characterized by bouts of vertigo, accompanied by fluctuating sensorineural hearing loss, tinnitus and/or aural fullness. This disease can be invalidated. Endolymphatic hydrops of the inner ear due to an imbalance of fluid production and absorption is thought to be the pathophysiological substrate. A broad range of therapies has been suggested, due to the high variability in the expression of the disease and poor quality of the evidence to support these therapies. We suggest a clinical algorithm for treatment of the disease. If conservative management of the disease fails, then we suggest perfusion of the middle ear with dexamethasone when hearing is functional, followed by gentamicin when repeated injections have failed, or when hearing is non-functional. Selective neurectomy of the vestibular nerve is a therapy of last resort. Endolymphatic sac surgery and labyrinthectomy are not advised by this author, based on the lack of high-level evidence supporting their use.

Keywords Menière's disease · Surgery · Treatment algorithm

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Introduction

Clinical Aspects

It was Ménière [1] who in 1861 for the first time described the triad of hearing loss, tinnitus and vertigo and attributed it to dysfunction of the inner ear. It was not until 1938 that Hallpike and Cairns [2] identified endolymphatic hydrops as the causative pathophysiological phenomenon responsible for the attacks. The aetiology of the disease, however, is still not fully understood.

Patients with Menière's disease suffer from incapacitating attacks of vertigo, accompanied by nausea, vomiting, unilateral sensorineural hearing loss, tinnitus and/or aural fullness. When individuals start to suffer from Menière's disease, their lives change dramatically. Attacks of serious bouts of vertigo, accompanied by vomiting, hearing loss and a loud, uncomfortable, whizzing noise in the affected ear, make these patients feel very sick and disabled. Patients can only lie down and wait for the attack to subside. The attacks last from 20 min to several hours. The frequency of these attacks varies. Intervals of days, weeks or even months occur. Usually, the attacks slowly become less severe and after 2–8 years, 60–80 % of the patients no longer have any attacks [3, 4]. The patients end up with profound sensorineural hearing loss and often disturbing tinnitus of the affected ear. However, the natural course of the disease has a great variability.

Clinically, Three Stages are Generally Recognized [5]

Stage I In the beginning of Menière's disease, patients suffer from attacks of vertigo, lasting from 20 min to several hours. These are accompanied by nausea and even

vomiting, as well as temporary sensorineural hearing loss and an aura of fullness or pressure in the affected ear. After the attack, hearing is often normal, and examination in between attacks shows normal results.

Stage II When the disease progresses, hearing loss can be measured in between attacks, especially in the lower pitches. It still fluctuates. The bouts of vertigo can be more or less frequent. A vestibular paresis of the affected labyrinth can be established.

Stage III In this stage, the sensorineural hearing loss no longer fluctuates with the attacks, but slowly progresses and affects all pitches. Tinnitus is a frequent complaint. The attacks of vertigo subside. A vestibular paresis of the affected labyrinth may result in a balance disorder in the end.

The American Academy of Otolaryngology—Head and Neck Surgery (AAO-HNS) has tried to make evaluation of diagnosis and treatment more uniform by introducing a guideline for Menière's disease in 1985, which was updated in 1995 [6].

Epidemiology

The prevalence of the disease is reported to be between 17 per 100.000 inhabitants in Japan [7] to 46 per 100.000 in Sweden [8]. Both sexes are affected almost equal. There seem to be 1.3 more women affected as compared to men [9]. Adults in their forties and fifties are the most frequently affected [10, 11]. Involvement of the contralateral ear is reported to be from a 9.1 % bilateral involvement in the first year after the initial symptoms to 41.5 % involvement after 20 years of follow-up [12].

Pathophysiology

The underlying mechanism of the recurrent attacks of disabling vertigo and progressive hearing loss in Menière's disease is thought to be the swelling of the endolymphatic fluid compartment of the inner ear. This endolymphatic hydrops of the inner ear is hypothesized to be due to an imbalance in production and absorption of endolymphatic fluid. Kimura [13] developed an animal model in guinea pigs and showed in 1967 that blockage of the endolymphatic duct and sac causes obstruction of the outflow of endolymph, resulting in hydrops of the endolymphatic compartment of the inner ear. The aetiology-triggering hydrops in Menière's disease remains unknown. Several mechanisms have been hypothesized, like a genetic predisposition of anatomical abnormality, and also underlying inflammatory or autoimmune inner ear processes have been suggested to cause Menière's disease, at least in some patients [14].

Diagnosis

Menière's disease is foremost a clinical diagnosis, based on the classification of the subcommittee on hearing and equilibrium of the AAO-HNS of 1995 [6]. Therefore, the clinical history is of the utmost importance. Additional testing can consist of audiometric assessment and bithermal caloric evaluation. An MRI-scan of the internal auditory canal is needed to rule out retrocochlear pathological disorders. Patients with Menière's disease present with a typical low-frequency sensorineural hearing loss. When the disease progresses, a flat sensorineural hearing loss may result, affecting all frequencies. As one of the typical features of the disease is fluctuation, serial audiograms may be needed to support the diagnosis. During bithermal caloric testing, a unilateral vestibular paresis may be measured, although in the early stage of the disease, the caloric assessment might still be normal. Here , regular testing is also advised. In the past, many patients might have been wrongly diagnosed with Menière's disease, while suffering from vestibular migraine. In 2012, Lempert et al. [15] published the diagnostic criteria of vestibular migraine as established in consensus meeting between de Barany Society and the International Headache Society. Ever since, it is possible to separate Menière's disease from vestibular migraine more clearly. Controversy remains over additional haematological testing. Thyroid disease, diabetes, syphilis or autoimmune disease might have to be excluded. More general haematological testing is not needed, unless indicated for a specific reason.

Treatment

Treatment for Menière's disease consists of medication, changes in lifestyle and psychological counselling, prior to surgical therapy which should be the last resort. In the acute phase, when people feel sick and are nauseated, symptomatic medication like anti-emetics is the hallmark of therapy. Daily medication to prevent or reduce the frequency and intensity of attacks is controversial. Diuretics might be effective, although there is no evidence of sufficient quality to support the effect [16]. James and Burton [17••] came to the same conclusion for betahistine in their 2011 Cochrane review. Systemic corticosteroids have been advocated by Morales-Luckie et al. [18] and Sajjadi [19]. However, the level and quality of the evidence are insufficient to support routine use. Intratympanic-applied steroids seem to be effective, although the level of evidence is limited and derived in a Cochrane review from only one study [20••]. The most effective medical treatment for Menière's disease appears to be intratympanic gentamicin, as we have shown in a Cochrane review, recently revised [21••]. One must be aware of the possibility of

sensorineural hearing loss following this treatment. A more recent non-invasive therapy is the pressure-pulse treatment with a pressure-pulse generator. Several randomized controlled trials have been conducted with the device [22–26]. Although these studies suggest effectiveness of the device in reducing complaints of vertigo, all studies were subject to a high risk of bias. The presented evidence, therefore, is not conclusive.

Types of Surgery

As we have described in our Cochrane Review on Surgery for Meniere's disease [27••], surgery for Menière's disease can be divided into 2 types of surgery: non-destructive surgery which aims to alter the expression of the disease, and destructive surgery that aims to control vertigo by destruction of vestibular function.

Non-destructive Surgery

Non-destructive procedures target on changing the natural course of the disease by reducing the frequency and intensity of the attacks. These procedures are less invasive and do not preclude the use of conservative treatment modalities.

1. Endolymphatic sac decompression and/or shunt, which aims to restore the imbalance in the production and absorption of endolymphatic fluid.
2. Insertion of a ventilation tube and pressure-pulse treatment, which aims to regulate middle ear pressure changes, on the assumption that symptoms are caused by pressure disturbances of the middle ear (discussed earlier).

Destructive Surgery

In Menière's disease, the vestibular end organ (labyrinth) is responsible for the attacks of vertigo, and the cochlea is responsible for the hearing loss. The rationale of destructive surgery of the labyrinth is to cure the patient of episodic vertigo by destructing the vestibular end organ. The brain will eventually compensate for the loss of vestibular function on one side, provided that the contralateral vestibular organ is working properly. An imbalance may be the result. Destructive procedures of the labyrinth have a high risk of destroying the cochlea as well. These procedures are irreversible and should, where possible, be avoided in patients with bilateral involvement and in patients with adequate hearing.

1. Selective vestibular nerve section, which aims to decrease vertigo by sectioning the vestibular nerve so the input of the diseased vestibular end organ cannot reach the brain. The patient is left with unilateral vestibular function.
2. Cochleovestibular nerve section, which has the same effect as the above but in addition leads to total loss of hearing in the operated ear.
3. Labyrinthectomy, which aims to decrease vertigo by total destruction of the labyrinth but in addition leads to total loss of hearing in the operated ear.
4. Intratympanic application of gentamicin in the middle ear to accomplish a chemical ablation of the labyrinth, which aims to decrease frequency and intensity of vertigo but may result in loss of hearing (discussed earlier).

Effectiveness of Surgery

Surgery of the endolymphatic sac was first proposed by Portmann and later became a routine practice by advocates as House [28] and Paparella et al. [29]. Several case series reported a control of vertigo in about 75 % of cases, with preservation of hearing in 98 % of patients. In up to 40 % of patients, hearing appeared to be better following the endolymphatic sac surgery (ESS) as compared to hearing prior to treatment [30–32]. Bretlau et al. [33] conducted a clinical trial, comparing ESS with a sham operation. Both groups had a 70 % success rate in controlling vertigo. The results of the trial have been criticized extensively. Claimed flaws are considered to be the small study size, the influence of general anaesthesia on fluid balance in the inner ear, and the possible beneficial effect of the 'sham' mastoidectomy [11]. Also, there was criticism concerning the methodology of the study. Randomization was unclear, and 7 out of 30 patients were lost to follow-up [27••]. Thomson et al. [34] compared ESS with grommet placement, hereby addressing the criticism of the possible efficacy of a mastoidectomy. Also, the risk of bias in this study was lower as compared to the Bretlau study [27••]. Again, no significant difference was found between the intervention and control group in terms of control of vertigo. The study reported 2 out of 30 patients with severe sensorineural hearing loss as a complication of surgery. Although subjected to criticism, both randomized controlled trials constitute the best available evidence at this moment. It allows more evidence-based conclusions as compared to case series published [31, 32, 35, 36]. As efficacy could not be established of ESS for Menière's disease and there is a risk of severe sensorineural hearing loss, ESS is not advised as routine treatment, until a clinical trial, addressing all the criticisms mentioned above, has been conducted.

Neurectomy of the cochleovestibular nerve for treatment of Menière's disease was described already in 1933 by Dandy [37]. Several authors modified his initial technique

and suggested a selective neurectomy of the vestibular nerve [38–40]. Silverstein [41] proposed a retrosigmoid/retrolabyrinthine technique, resulting in control of vertigo or at least substantial improvement in 92 % of patients (1989). 20 % of patients experienced limited hearing loss, whereas in only 4 %, significant hearing loss was reported.

Labyrinthectomy of the affected labyrinth always ends up in total deafness and is, therefore, reserved for patients with non-functional hearing. As other treatment options are less destructive, labyrinthectomy is a last resort operation.

Both selective neurectomy of the vestibular nerve, as well as labyrinthectomy, will be executed far less nowadays. Adequate medical treatment, combined with perfusion of the middle ear with either corticosteroids or gentamicin, has reduced the number of patients with Menière's disease that need these destructive procedures.

Conclusion and a Clinical Algorithm for Treatment of Menière's Disease

When a patient visiting your outpatient clinic is diagnosed with Menière's disease, the first step is to ensure that the patient fully understands the pathophysiology and the variability of the natural course of the disease. Medication to suppress the nausea during an attack as well as medication to diminish both the frequency and the intensity of the attacks is prescribed. Dietary measures and lifestyle advises can be given. It must be clear to the patient that most of the suggested medication, as well as dietary and lifestyle advises, although suggested to be effective in many lower levels of evidence research reports, have not yet been proven to be effective in robust trials. The doctor and his patient should find out what fits best for the patient in a process of shared decision making. When the bouts of vertigo do not subside, other measures should be considered. If the patient still has functional hearing, then intratympanic treatment with corticosteroids can be advised. When this repeatedly does not work, or the patient already has non-functional hearing, the middle ear can be perfused with intratympanic gentamicin. If, following this treatment regimen, intractable Menière's disease still exists, then destructive surgery can be considered. Then, selective neurectomy of the vestibular nerve is the treatment of choice. Procedures like endolymphatic sac surgery or labyrinthectomy are not advised based on the lack of high-level evidence supporting their use. Furthermore, it is important to keep in mind that at any stage of the disease, vestibular rehabilitation and psychological counselling can be of great support to the patient.

Complicance with Ethics Guidelines

Conflict of Interest Peter Paul G. van Benthem declares that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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