Article published online: 2023-06-28

What a Neurosurgeon Should Know About the Endolymphatic Sac: Part 3 – Ménière Disease

O que um Neurocirurgião Deve Saber Sobre o Saco Endolinfático: Parte 3 – Doença de Ménière

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Arq Bras Neurocir 2023;42(2):e160–e164.

Abstract

Objective To elucidate all the aspects that neurosurgeons should know about the Ménière disease (MD).

Methods Review of guidelines, books, and studies from 1933 to 2021, from basic to translational research, using human and animal endolymphatic sac (ES) tissue or cells, as well as reviews, case reports, and papers about surgical experience. This article is divided into three parts. In this last part, we review the MD.

Results The MD is one of the most common pathologies in the ES. It was first described by Prosper Ménière in 1861 with its clinical triad: dizziness, tinnitus, and hearing loss. A lot of theories relating ES to the MD have been proposed. Some of them postulate that it is caused by a narrowing and shortening in the endolymphatic duct, and others relate it to severe inflammation on the ES. Mostly due to the lack of understanding of this pathology, the diagnosis is mainly clinical, despite histopathology being helpful to confirm the diagnosis. The treatment of the MD can be done in 3 different ways: pharmacological, nonpharmacological, and surgical.

Conclusion The MD is one of the most common pathologies in the inner ear and has been largely studied over the years. The latest diagnosis guidelines must help in the classification and give better basis for diagnosis and treatment, which, despite not

Keywords
- endolymphatic sac
- Ménière disease
- neurosurgery

received February 8, 2022
accepted April 6, 2022

DOI https://doi.org/10.1055/s-0042-1749087.
ISSN 0103-5355.

Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

being curative yet, has improved over the years. Pharmacological treatment based on the possible etiologies, allied with proper diet and routine exercise, is showing promising results.

Resumo

Objetivo Elucidar todos os aspectos que neurocirurgiões devem saber sobre a doença de Ménière (DM).

Métodos Revisão das diretrizes, livros e estudos de 1933 até 2021, de pesquisa básica até translacional, usando tecidos ou células do saco endolinfático (SE) humanas e animais, além de revisões, relatos de caso e artigos sobre experiencia cirúrgica. Este artigo é dividido em três partes. Nesta última nós revisamos a DM.

Resultados A DM é uma das patologias mais comuns do SE. Ela foi inicialmente descrita por Prosper Ménière em 1861 com a tríade clínica: tontura, zumbido e diminuição da audição. Muitas teorias têm relacionado o SE com a DM. Algumas delas postulam que esta é causada por uma diminuição e estreitamento do ducto endolinfático e outras a relacionam com uma inflamação grave do SE. Principalmente devido à falta de entendimento sobre a patologia, o diagnóstico é primariamente clínico, apesar da Histopatologia ajudar na confirmação diagnóstica. O tratamento da DM pode ser feito de três diferentes formas: farmacológico, não farmacológico e cirúrgico.

Conclusão A DM é um dos distúrbios mais comuns da orelha interna e tem sido muito estudada nos últimos anos. As diretrizes mais recentes devem ajudar na classificação e fornecer mais bases para o diagnóstico e tratamento, que, apesar de ainda não ser curativo, teve grandes avanços ao longo dos anos. O tratamento farmacológico baseado nas teorias etiológicas, aliado com dieta apropriada e exercícios físicos rotineiros, tem mostrado excelentes resultados.

Palavras-chave
- saco endolinfático
- doença de Ménière
- neurocirurgia

Introduction

The endolymphatic sac (ES) is a structure situated in the inner ear, together with the cochlea and the semicircular canals, or vestibular system.[1] The ES may be responsible for homeostatic regulation of the inner ear, endolymphatic fluid volume, immune response, elimination of inner ear cellular debris and floating otoconia, membranous labyrinth pressure, acid/basic transport, and the secretion of substances.[1–11]

Despite being only 3 mm long in diameter, the ES does not have a very variable location inside the inner ear.[8] Almost every alteration in this structure can cause a massive problem to the hearing, including its loss. The Ménière disease (MD) is one of the most common pathologies of the ES,[12] and it was first described by Prosper Ménière in 1861, with its clinical triad: dizziness, tinnitus, and hearing deafness, with or without aural fullness. Over the years, a lot has changed regarding the definition of this disease but some aspects, like the etiology, are still in debate.[13]

The MD can be treated surgically. Two of the possible procedures are ES decompression and vestibular neurectomy. Pharmacological and nonpharmacological treatments can also help.[12,14]

In this review, our aim is to elucidate all the aspects that neurosurgeons should know about MD.

Methodology

This article is divided into three parts. In the third part we review the MD's basic aspects, clinical diagnosis and treatment. We focused on evidence of guidelines, books, and PubMed (from 1933 to 2021) basic and translational research, using human and animal ES tissue or cells, previous reviews about the subject, case reports, and papers about surgical experience. The terms used individually and combined were: *Endolymphatic sac*; *Ménière disease*; *Neurosurgery*. Literature inclusion criteria were: English and Portuguese language only; individual case studies and long-term follow-up studies were included, and duplicate studies were excluded.

First, we briefly approached the basic aspects of MD and its relation to the ES, followed by a review of the clinical diagnosis and treatment. This study may provide a basis to guide neurosurgeons in the evaluation and treatment of this disease.

Results

Basic Aspects and Relation to the ES

The MD was first described by Prosper Ménière in 1861 with its clinical triad: dizziness, tinnitus, and hearing deafness, with or without aural fullness.[13] However, although some of

these original aspects are still present in guidelines, a lot more has been discovered about this disease.

The MD commonly affects adults from 20 to 50 years[13] and is more likely to occur in women.[15] Both ears are affected with the same frequency. Despite the large number of recent studies about the MD, the epidemiology is still unclear, bearing in mind how difficult it is to make an early diagnosis.[13] Although challenging to measure, Alexander and Harris[15] estimate that the prevalence of MS in the United States is 190 per 100,000 habitants.

Part of the trouble in making an early diagnosis of MD lies in its unclear etiology.[13] Studies have raised many theories on its possible causes, but none of them were confirmed. Some of the theories are related to the ES, such as the possibility of a narrowing and shortening in the endolymphatic duct that could hamper the reabsorption of endolymph in the ES, since a smaller duct would allow less endolymph to pass through the duct. Thus, the retained endolymph would elevate the pressure in the endolymphatic space.[13] Another possibility is a reduction of the ES itself, again hampering the reabsorption properties of the ES since the contact surface is smaller.[13] In agreement with that, an experimental obliteration of the ES caused MD symptoms.[16]

It's also possible that due to the immunological properties of the ES,[2,5,6] the occurrence of severe inflammation on the ES contributes to tissue fibrosis, further damaging the structure and its function, and generating MD.[13] Another theory, described by Cahali et al.,[13] is related to the stria vascularis, and claims that patients with MD have a significant decrease in the number of vessels, with its transversal section also being smaller. Thus, the vascular deficiency of the stria vascularis represents a possible etiology, mostly because of its importance for the endocochlear potential and endolymph secretion.[13]

There are other explanations for the etiology of MD that are not related to the ES. Glycemic and insulin disorders have been described as possible causes since 90% of the patients with MD have alterations in the glucose and/or insulin levels. Besides that, diet and exercise can diminish the symptomatology of the patients.[13] Hypothyroidism and estrogen insufficiency are among other possible causes.[13]

With fewer observational studies, some authors defend the hypothesis of food allergies (meat, corn, wheat etc.) and stress causing the MD.[13]

Clinical Diagnosis

The diagnosis of the MD is mainly clinical. Cochlear and vestibular symptoms with aural fullness, in the absence of neurological symptoms, characterize the disease. The MD attacks typically last from minutes to hours, and 96% of the cases also present neurovegetative symptoms. Bilateral tinnitus and hyperacusis are also very common, and can persist during and after the crises, in some cases becoming chronic. Other symptoms are: unilateral loud noise intolerance and sound frequencies distortion.[13]

Despite diagnosis criteria being mainly clinical, some exams can be done to confirm the suspicion.[13] Magnetic resonance imaging (MRI),[16] pure tone audiometry, glycerol test,[17] otoacoustic transport, standard vestibular test, retrocochlear tests, and glycemic and insulin curves[13] can be very helpful. Nevertheless, none of those exams are pathognomonic.[13]

The histopathological exam is the only one that comes close to confirming the diagnosis. Paparella determined that the most important histopathology finding in the MD is the endolymphatic hydrops in the cochlea and saccule. Also, saccule membrane bulging is a common finding.[18]

Using all this information, in 2015, the Barany Society made a guideline for the diagnosis of MD (►**Fig. 1**) and created simple and easily applicable diagnosis criteria.[19]

Treatment

The MD has 3 different types of treatment: pharmacological, nonpharmacological, and surgical.[13] The pharmacological aim is to do a symptomatologic aid, since none of the drug trials were proven to contribute to the healing process. Some of the choices include: hydrochlorothiazide, which increases the potassium concentration in the endolymph, labyrinth depressors, which have anticholinergics, antiemetics, and sedatives. Corticosteroids and vasodilators can also be used, based on the etiology hypothesis of, respectively, fibrosis due to severe inflammation of the ES or stria vascularis ischemia.[13]

Concerning the nonpharmacological treatment, it's possible to interfere in the patient's quality of life with dietary recommendations and routine exercise. A very similar diet for diabetic patients can be used to treat MD, since the high glucose levels can be an etiological factor. Accordingly, a diet rich in potassium and with lower sodium levels may also help. Alcohol consumption and smoking must stop, as well as caffeine, of which can be consumed a maximum of 250 mg per day.[13]

Regarding surgical treatment, two procedures stand out: ES decompression and vestibular neurectomy.[14] The ES procedure for MD was described for the first time by Portmann[20] and it is still used to treat patients' impairment with refractory MD. Hearing impairment and vertigo are the symptoms that have the largest improvement: 19% and 81%, respectively.[14] The operation starts with a bone incision at the retroauricular sulcus level, followed by mastoid opening to expose the lateral sinus. The bony wall of the lateral sinus is separated from the adjacent dura mater (DM), which is then elevated to until it reaches the adherence with the bone. After the vestibular aqueduct and the wall of the fossa are on the same level, it is possible to open the ES. First, the endolymphatic fossa is exposed and pierced in 2 to 3 mm on the ES wall. Afterwards, the opening of the ES must be done in a very delicate way in the connecting point between the dura mater and the rear wall of the petrosal bone.[20]

In 1933, Dandy[21] made the vestibular neurectomy common, but because of the collateral effects and postoperative complications, the procedure was not widely done. In 1991, 58 years later, Silverstein and Rosenberg[22] modified the previous technique and proposed what they called the "combined retrosigmoid/retrolabyrinthine vestibular nerve section". This technique demonstrated an 85% cure rate, with

Definitive Meniere disease[•]	
A.	Two or more spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours
B.	Audiometrically documented low to medium frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during, or after one of the episodes of vertigo
C.	Fluctuating aural symptoms (hearing, tinnitus, and fullness) in the affected ear
D.	Not better accounted for by another vestibular diagnosis
Probable Meniere disease	
A.	Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours
B.	Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear
C.	Not better accounted for by another vestibular diagnosis

[•]No serological or genetic marker is considered for the diagnosis of MD.

Fig. 1 Ménière disease classification. Adapted from Lopez-Escamez JA et al.[19]

a 7% chance of enhancing the vertigo. Regarding the hearing loss, Silverstein and Rosenberg identified that 20% of the patients had a change on their audiometry compared to before the surgery and 4% showed substantial hearing loss.[12]

The "combined retrosigmoid/retrolabyrinthine vestibular nerve section" makes a U-shape incision in the postauricular area. Then, a mastoidectomy is required to expose the posterior fossa and the lateral venous sinus. Following, a dural incision is made to reach the cerebellum. The next step is to open the cerebellopontine angle (CPA) arachnoid over the ninth cranial nerve, with the intention of releasing CPA liquid, and releasing the pressure. With the decompression, the cerebellum disconnects itself from the temporal bone allowing for CPA exposure without damaging the cerebellar retraction. At this point, the facial nerve must be identified to prevent damaging it, and the eighth cranial nerve must be examined. After all precautions are taken, the vestibular nerve separation is performed in a cleavage plane between the cochlear and vestibular fibers. If the identification of the cleavage plane fails, the dura mater is reflected off the temporal bone, and the opening of the internal auditory canal is performed using a diamond burr, to enable the division of the superior vestibular and posterior ampullary nerves. To close, the mastoid air cells are sealed with bone wax and the dura with watertight fashion suture. The bony defect is corrected with adipose tissue to present fistulas.[22]

Conclusion

The MD is one of the most common pathologies of the inner ear and has been largely studied over the years. Its diagnosis is mainly clinical, which allows a bigger autonomy for medical professionals, but also demands a lot of experience. The latest guidelines must help in the classification of the disease and offer a better basis for diagnosis and treatment options.

The treatment, despite not being curative yet, has improved a lot recently. The pharmacological treatment based on the possible etiologies, allied with proper diet and exercise, is showing promising results. Surgical treatments, especially ES decompression, are procedures that must be taken into consideration for patients who don't respond well to noninvasive therapeutics, and for those who fill the necessary criteria. Knowing this, we highlight the importance of a well-trained doctor to identify and offer the best treatment possible. This study may provide a basis to acquire these skills.

Conflict of Interests
The authors have no conflict of interests to declare.

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