Chapter

Introductory Chapter: Ménière's Disease (MD)

Fayez Bahmad Jr

1. Introduction

Ménière's disease (MD) is probably a multifactorial disorder where the genetics and environmental factors determine the onset of the disease. This disease have been related to the accumulation of endolymph in the cochlear duct and the vestibular organs in histopathological studies, although endolymphatic hydrops (EH) per se does not explain all clinical features, including the progression of hearing loss or the frequency of attacks of vertigo [1–4].

Dizziness and vertigo are frequent symptoms in the otolaryngologist's practice, and all efforts towards the better comprehension of this system and its pathology are of fundamental importance [1, 2].

Currently, there is no universally accepted theory about the pathophysiology of the disease. Through histopathological studies, it is assumed that endolymphatic hydrops is the most descriptive pathological feature of Ménière's disease. The pathophysiology of symptoms is still widely disputed: membrane ruptures, increased pressure, and mechanical displacement of peripheral organs such as endolymph accumulation, viral infections, autoimmune disease, and several other theories that have been reported [1–3].

Great advances have been made in neuro-otology, and increasing knowledge in the field of molecular biology, genetics, and neurosciences has substantially modified the approach of the patient with balance complaints. This book studies the most interesting and controversial of these vestibular diseases, the Ménière's disease.

The Classification Committee for the International Classification of Vestibular Disorders (ICVD) nominated by the Bárány Society, 2009, standardized the nomenclature of vestibular symptoms (SV) in four groups. One of the most important is the episodic vestibular syndrome: crises of vestibular symptoms interspersed with asymptomatic periods, such as Ménière's syndrome and vestibular migraine [1, 2–6].

Ménière's disease is an inner ear alteration characterized by two groups of symptoms: vestibular and auditory symptoms. In many patients, their presentation may be unusual or different than the classical symptoms such as tinnitus, fluctuating hearing loss, aural fullness, and concomitant dizziness [1, 2].

The history of the disease may be progressive or nonprogressive, and, in addition to the typical clinical presentation of Ménière's disease, two variants of the disease were identified:

1. cochlear Ménière's disease—hearing is the predominant symptom; and

2. vestibular Ménière's disease—vestibular symptoms are predominant.

Other classifications used frequently are as follows:

1. Ménière's syndrome: known and well-established condition causing symptoms; and

2. Ménière's disease: idiopathic cause [1, 2].

Recent studies revealed that there are genotypic and phenotypic factors that influence the prevalence difference between countries [7].

In the majority of these patients, the initial presentation of the disease is often the cochlear form, which is harder to be clinically recognized and frequently is associated with another cause or is presumed to be simply due to aging.

Even after the vestibular component becomes obvious, long periods of remission may mask the complete final image of the syndrome with episodic vertigo, fluctuating autistic loss, tinnitus, and aural fullness. Therefore, generally in clinical practice, only moderate to severe cases are tabulated in the estimates so far.

A multifactorial inheritance is believed to be the best response, where the necessary conditions are met, leading to endolymph malabsorption and subsequently hydrops. Clinical and laboratory evidence supports this concept. Merchant et al. analyzed the temporal bone collection of the Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, USA, with a clinical diagnosis of Ménière's syndrome (28 cases) or a histopathological diagnosis of hydrops (79 cases).

All 28 cases with classic symptoms of Ménière's syndrome had hydrops in at least one ear. However, the reverse is not true. There were 9 cases of idiopathic hydrops and 10 cases with secondary hydrops, in which the patients did not have the classic symptoms of Ménière's syndrome. Endolymph is mainly produced in the stria vascularis. Slowly, endolymph is absorbed into the endolymphatic duct and sac, a biologically active transport structure where absorption occurs mainly and also to a lesser extent secretion occurs. Evidence strongly suggests that longitudinal flow (slow process) and radial flow (fast) occur.

In this book we will adopt the term Ménière's disease to follow the prevailing trend among most research groups and discuss the main topics, current and past ideas about etiopathogenesis, diagnosis, and treatment of Ménière's disease associated with migraine.

Some of the epidemiologically published studies to date have tended to blend different epidemiological concepts. The direction of these studies is mainly retrospective (the themes are identified after a result or illness), and they actually measure only the prevalence (existing events or the number of cases of a disease at a given moment divided by the population at risk). Only prospective studies (subjects are identified before a result or illness, future events are counted) would have the power to adequately measure this incidence. Although more reflective to real life than an artificial experiment, retrospective observational studies are susceptible to bias.

2. Diagnostic failures

There are many classifications and proposed diagnostic criteria, and it makes difficult to establish the true incidence of Ménière's disease in the general population.

The 1995 and 2015 criteria simplified the definition of Ménière's disease and allowed to be usable in all global studies and so may be able to substitute and unify all the classifications [1, 2].

3. Etiopathogeny

Almost all the researchers and histopathological works in the past presumed that endolymphatic hydrops was the pathological characteristic of Ménière's disease. There are still many questions and polemic discussions about the pathophysiology of the symptoms: increased pressure and mechanical displacement of the peripheral organs such as saccule by endolymph accumulation, ruptures of membranes, viral infections, and autoimmune disease, in addition to several other theories that have already been reported.

Recent consensus accepted that endolymphatic hydrops is no longer a central etiology but rather as one of the manifestations of the syndrome.

4. Diagnostic

Classical Ménière's disease is an excellent example of a condition that can be diagnosed on clinical grounds and simple audiometric examinations.

When it is the classic or definite form, it is characterized by recurrent and spontaneous episodes of vertigo, fluctuating sensorineural hearing loss, tinnitus, and aural fullness. In this case the diagnosis is easy even for the most naive clinician [1, 2].

In 2015, the Hearing and Balance Committee of the American Academy of Otorhinolaryngology-Head and Neck Surgery (AAO-HNS), Bárány Society, and other entities set the parameters for the clinical diagnosis of Ménière's disease.

The classification includes two categories: defined Ménière's disease and probable Ménière's disease [2].

Defined Ménière's disease is based on clinical criteria and requires the observation of an episodic vertigo syndrome associated with low- to medium-frequency sensorineural hearing loss and fluctuating auditory symptoms (tinnitus in the ear and/or fullness) in the affected ear.

The duration of vertigo episodes is limited to a period of between 20 minutes and 12 hours. Probable Ménière's disease is a broader concept defined by episodic vestibular symptoms (vertigo or dizziness) associated with fluctuating aural symptoms that occur over a period of 20 minutes to 24 hours [1, 2].

The clinical evaluation then includes the following [8, 9]:

- detailed medical history that should include all previous vertigo events;
- laboratory tests to rule out differential diagnoses of the syndrome;
- imaging tests to aid diagnosis and rule out differential diagnoses of the syndrome; and
- cochlear and vestibular, audiological, and electrophysiological examinations.

The most appropriate exams to aid in diagnosis consist of:

- glycerol dehydration test;
- electrocochleography (ECochG); and
- PEMV or VEMP test.

5. Treatment

Different treatment options for Ménière's disease exist with substantial variability between countries. None of the treatment options cure the disease. As many treatments have a significant impact on the functioning of surrounding structures, one should start with noninvasive approaches with the fewest possible side effects and proceed to more invasive steps:

- conservative;
- diet;
- diuretics;
- labyrinth suppressors;
- invasive procedures;
- intratympanic gentamicin;
- endolymphatic sac decompression surgery;
- labyrinthectomy; and
- vestibular neurectomy.

Sodium restriction diet: Low-level evidence suggests that restricting the sodium intake may help to prevent Ménière's attacks.

Betahistine: Substantial disagreement in the medical community about the use of betahistine exists. A Cochrane review found low-level evidence to support the use of betahistine with substantial variability between studies. Medical therapy in many medical centers often starts with betahistine orally.

Intratympanic steroid injections may reduce the number of vertigo attacks in patients with Ménière's disease.

Intratympanic gentamycin injections: Gentamycin has strong ablative properties towards vestibular cells. The side effects are a sensorineural hearing loss because of a certain amount of toxicity towards cochlear cells.

Surgery with vestibular nerve section or labyrinthectomy: Nerve section is a therapeutic option in patients who failed the conservative treatment options and labyrinthectomy when surgical options failed. Labyrinthectomy leads to a complete hearing loss in the affected side.

Clinically, three situations arise in which drug treatment is very helpful:

6. Acute attack drugs

Aiming at sedating the vestibule-trunk axis is particularly useful in aborting acute attacks. These include cinnarizine, promethazine, and diazepam.

Prolonged use of drugs such as cinnarizine is not advisable due to the risk of extrapyramidal side effects from prolonged use, particularly in the elderly.

6.1 Maintenance treatment

Dietary salt restriction and the use of diuretics such as furosemide, amiloride, and hydrochlorothiazide are attempts to prevent endolymphatic hydrops. The basis

for this is historical rather than scientific, as the data from the few controlled studies that exist are conflicting and the placebo effect is clinically significant.

Betahistine has been subject to some scientific scrutiny, and several controlled clinical studies have shown significant improvement in vertigo, hearing loss, and tinnitus in the short term. Betahistine, with or without a diuretic, is currently the preferred means of ensuring medical treatment.

Drugs such as cinnarizine, propranolol (particularly if the patient has a history of migraine), and corticosteroids are also used empirically by some doctors if the patient's symptoms are refractory to the above measures.

7. Ablative treatment

7.1 Intratympanic gentamicin

The toxic effects of aminoglycosides on the inner ear sensory neuroepithelium have been recognized for decades.

Chemical labyrinthectomy through intratympanic gentamicin (GIT) controls vertigo and has been helpful in mainly unilateral Ménière's disease when hearing is poor, but the vertigo presented by the patient is disabling.

The attending otologist should properly remind and advise the patient that from 3 days after the first application, fiber differentiation begins to occur and this usually leads to severe vestibular symptoms between 7 and 10 days after application. And it is a phenomenon expected by the chemical destruction of vestibular nerve afferents.

Several series have a vertigo control rate of about 90%, although a cochleotoxic effect is seen in 15–25% of cases. The future for intratympanic aminoglycosides in Ménière's disease is therefore very promising [10].

Protocol of use:

- Complete battery of vestibular tests before therapy
- Initial reference (VENG before GIT)
- Provide for college entrance compensation
- Intratympanic gentamicin (40 mg/ml)
- Weekly intervals (up to three to four applications)
- Repeat audiometry weekly
- Repeat the VENG at the end of the sessions
- Topical anesthesia
- Patient rests for 1 hour after application

7.2 Surgical treatment

Whether as a result of medical treatment or as a consequence of the clinical course of Ménière's disease, about 90% of patients have a long period of remission.

This implies that 10% of patients continue to have clinically important episodes of vertigo, and surgical treatment should be considered for them.

The various surgical procedures advocated for Ménière's disease continue to raise considerable controversy among otolaryngologists. The decision to operate and the choice of procedure are often dictated by the understanding and experience of a particular technique and the surgeon's individual threshold for surgical intervention. Generally, surgical procedures for Ménière's disease are classified as destructive or nondestructive with regard to hearing [8–10].

7.3 Endolymphatic sac surgery

Endolymphatic sac surgery was first described in 1927 by Portmann, and no other aspect of Ménière's disease has elicited further debate or controversy. Just as the exact role of the endolymphatic sac in the development of hydrops is not yet known, the precise mechanism by which surgery works remains undefined. However, endolymphatic sac decompression surgery is still widely performed [10].

7.4 Vestibular nerve section

In the vestibular nerve section, no attempt is made to modify the underlying pathophysiology. The objective is to dissociate the offensive maze from the trunk, preserving the patient's hearing.

The procedure is uniformly effective, with vertigo control in 90–95% of patients according to some series. However, it is a surgery with considerable risks inherent in any posterior fossa neurosurgical procedure [10].

7.5 Surgical labyrinthectomy

Labyrinth extirpation is indicated in patients with severe symptoms who have virtually useless hearing. Disturbance of the inner ear thus invariably leads to permanent anacusis. However, the ear on the opposite side may have subclinical hydrops, and we should be naturally concerned that the progress of the disease in the patient's contralateral ear may aggravate and make it bilaterally deaf. This is probably the reason for the widespread choice of nondestructive inner ear procedures [10].

7.6 Cochlear implant

Over the past decade, the hearing rehabilitation of certain profoundly deaf people has been transformed by cochlear implants.

Patients with severe bilateral Ménière's disease and severe to profound bilateral sensorineural deafness will end up with an indication for hearing rehabilitation with cochlear implant. Surgeons with patients with symptoms whose disease is refractory to clinical treatment have several surgical options.

We should always start with the use of intratympanic aminoglycosides as the least aggressive option.

When intratympanic gentamicin does not work, there are three management strategies: proponents of endolymphatic sac surgery as the first surgical step, reserving revision surgery, or vestibular neurectomy for patients who continue to have vertigo.

Patients who have not yet achieved clinical improvement after the endolymphatic sac decompression operation, the otologist who has no experience or staff to subject the patient to vestibular neurectomy is faced with the option of performing surgical labyrinthectomy [10].

DOI: http://dx.doi.org/10.5772/intechopen.91924

Author details

Fayez Bahmad Jr [1,2]

1 University of Brasilia, Brasilia, DF, Brazil

2 Ophthalmology and Otolaryngology Faculty, University of São Paulo (FMUSP), São Paulo, SP, Brazil

*Address all correspondence to: fayezbjr@gmail.com

References

[1] Members of the Committee on Hearing and Equilibrium. Committee on hearing and equilibrium, guidelines for the diagnosis and evaluation of therapy in Ménière's disease. Otolaryngology–Head and Neck Surgery. 1995;**113**:181-185

[2] Lopez-Escamez JA et al. Diagnostic criteria for Ménière's disease. Journal of Vestibular Research. 2015;**25**:1-7

[3] Oliveira CA, Bahmad F Jr, Sampaio AL. Ménière's Disease. In: Textbook of Vertigo - Diagnosis & Management. 1st ed. Vol. 01. New Delhi: Jaypee Brothers Medical Publishers; 2014. pp. 62-93

[4] Rauch SD, Merchant SN, Thedinger BA. Ménière's syndrome and endolymphatic hydrops: Double-blind temporal bone study. The Annals of Otology, Rhinology, and Laryngology. 1989;**98**:873-883

[5] Merchant SN, Adams JC, Nadol JB. Pathophysiology of Ménière' syndrome are symptoms caused by endolymphatic hydrops? Otology & Neurotology. 2005;**26**:74-81

[6] Agrawal Y, Minor LB. Ménière disease and other causes of episodic vertigo. In: Bronstein AM, editor. Vertigo and Imbalance. United Kingdom: Oxford; 2013. pp. 241-250

[7] Bahmad F Jr, DePalma SR, Merchant SN, Bezerra RL, Oliveira CA, Seidman CE, et al. Locus for familial migrainous vertigo disease maps to chromosome 5q35. The Annals of Otology, Rhinology, and Laryngology. 2009;**118**(9):670-676

[8] Cal R, Bahmad F Jr. Migraine associated with disfunção auditivo-vestibular auditory-vestibular dysfunction. BJORL. 2008;**74**(4):606-612

[9] Lamounier P, Gobbo DA, de Souza TSA, de Oliveira CACP, Bahmad F. Electrocochleography for Ménière's disease: Is it reliable? Brazilian Journal of Otorhinolaryngology. 2014;**80**:527-532

[10] Viana LM, Bahmad F Jr, Rauch SD. Intratympanic gentamicin as a treatment for drop attacks in patients with Ménière's disease. Laryngoscope. 2014;**124**(9):2151-2154