MÉNIÈRE'S DISEASE

Introduction

Ménière's disease (also known as *Idiopathic Endolymphatic Hydrops*) is a chronic clinical syndrome that consists of the triad of:

- **Episodic vertigo**
- **Tinnitus**
- **Sensorineural hearing loss**

The exact cause is unknown, but is possibly multifactorial, and so the symptom triad may represent the final common pathway of many different **inner ear** pathologies.

Along with vestibular migraine, Ménière's disease is one of the commonest causes of **recurrent spontaneous** vertigo.[2]

There is no specific diagnostic test for Ménière's disease and so diagnosis is currently clinical. Definitive diagnosis can only be made postmortem.

Treatment is symptomatic.

History

Ménière's disease is named for **Prosper Ménière**, (1799 - 1862) a French physician who first reported that the inner ear could be the source of a syndrome that manifested at episodic vertigo, tinnitus, and progressive hearing loss.

Epidemiology

Ménière's disease can begin at any age but patients typically present with symptoms between the ages of **20 to 40.**

Ménière's syndrome in children is most often associated with congenital malformations of the inner ear.

Pathology

The exact cause is unknown, but is possibly multifactorial, and so the symptom triad may represent the final common pathway of many different **inner ear** pathologies.

The classic *pathological lesion* seen in Ménière's disease is termed **endolymphatic hydrops** (i.e oedema). It is unclear why excess fluid builds up in the endolymphatic spaces of the inner ear. There is distortion and distention of the membranous, endolymph-containing portions of the labyrinthine system. Although most patients have no other identifiable underlying otologic disease.

The lesion can only be *definitively* diagnosed by postmortem histopathologic analysis of the temporal bone. However, hydrops has also been identified in postmortem examination of temporal bones where there was no history of Meniere symptoms.

While endolymphatic hydrops is present in *all* patients with Ménière's disease, not all patients with endolymphatic hydrops have symptoms.

Many etiologies have been proposed, including:

1. Anatomical blockage at the endolymphatic sac or duct:

    - One popular theory postulates an abnormality in the resorption of endolymph at the endolymphatic sac. Endolymphatic hydrops has been experimentally induced in guinea pigs by blocking the entrance to the endolymphatic sac.

    - Blockage can be due to a congenital cause such as hypoplasia of the vestibular aqueduct.

2. An immunologic mechanism.

3. A genetic predisposition:

    - An autosomal dominant inheritance pattern has been reported, involving 8 to 15 percent of patients with Meniere disease in two studies.

    - Patients with a family history had an earlier age of onset and the manifestations were more severe in successive generations.

    - A specific genetic marker however has not been identified.

4. A viral etiology:

    - Although a viral etiology has been proposed, DNA for suspected viruses (herpes simplex, varicella zoster, and cytomegalovirus) has not thus far been detected in endolymphatic samples taken at surgery for patients with Meniere disease.

5. A vascular aetiology:

- Migraine occurs more commonly in patients with Ménière's disease than in the general population, leading to the postulation of a common vascular pathophysiology for the two disorders.

Clinical features

Ménière's disease is a syndrome that consists of the triad of:

1. **Episodic vertigo**:

    - Frequently associated with distressing symptoms of nausea and vomiting.

2. **Tinnitus**:

    - Tinnitus is characteristically low pitched (as listening to the sea in a seashell).

3. **Sensorineural hearing loss**:

    - Hearing loss usually *fluctuates*. It often initially affects the lower frequencies.

    - Hearing loss *progresses* over time, and often results in permanent hearing loss at all frequencies in the affected ear over an 8 to 10 year period

Aural "fullness" or pressure is also described

Disease can be unilateral or bilateral. The incidence of bilateral disease is uncertain but reports range from 10 to 50 percent of cases.

Otoscopy findings are usually **normal.**

Criteria for diagnosis:

Patients usually have some variable auditory and/or vestibular symptoms for 3 -5 years before they meet the diagnostic criteria for Ménière's disease.

Diagnostic criteria proposed by the American Academy of Otolaryngology and Head and Neck Surgery (AAO-HNS) stipulate that a "definite" diagnosis of Ménière disease requires the following:

- Two spontaneous episodes of rotational vertigo lasting at least 20 minutes

- Audiometric confirmation of sensorineural hearing loss

- Tinnitus and/or a perception of aural fullness

Further investigation is also required to rule out other disorders in the differential diagnosis

Disease progression:

Symptoms tend to be episodic with intense exacerbations alternating with prolonged periods of remission.

Approximately two thirds of patients experience vertigo attacks in clusters.

Approximately one-third have sporadic attacks.

The frequency of vertigo episodes may decline over time.

The course of Ménière disease varies among individuals with different patterns of symptoms:

- Some have marked hearing fluctuation and progressive hearing loss with infrequent vestibular symptoms
- Some have severe and frequent vertigo with only mild auditory symptoms
- Some manifest both auditory and vestibular symptoms in equal measure.

Disease severity:

The American Academy of Otolaryngology-Head and Neck Foundation (AAO-HNS) has set forth criteria for Ménière's disease disability: [4]

Severity	**Symptoms**
Mild	Intermittent or continuous dizziness/unsteadiness that precludes working in a hazardous environment.
Moderate	Intermittent or continuous dizziness that results in a sedentary occupation.
Severe	Symptoms so severe as to exclude gainful employment.

Trigger Factors:

Some patients may have specific trigger factors.

These may include:

1. High salt intake:

    - Salt restriction is sometimes recommended as part of initial therapy, although data supporting its efficacy are not available.

      An appropriate salt-restricted diet will have approximately 2 to 3 grams of sodium per day. The daily sodium intake should be evenly spread across meals to avoid a large bolus at any time

2. Caffeine:

    - A vasoconstrictor that may reduce microvascular flow in the labyrinthine system. Limiting intake is often recommended.

3. Nicotine:

    - A vasoconstrictor that may reduce microvascular flow in the labyrinthine system.

4. Alcohol:

    - It has been surmised that alcohol may causes fluid and electrolyte shifts that may bring on acute attacks. Limiting intake is often recommended.

5. Stress

6. Monosodium glutamate (MSG)

7. Some allergies.

Avoidance for patients with identified triggers may alleviate or ameliorate symptoms.

Differential diagnoses:

Important considerations will include:

1. Multiple sclerosis:

    - Multiple sclerosis (MS) can present with symptoms identical to those of Ménière's disease. However, the observed nystagmus during an attack of MS is typically more severe and longer lasting, and the patients may have other neurologic complaints.

2. Recurrent vestibular neuronitis

3. Neurosyphilis

4. Acoustic neuroma:

   - Patients typically present with progressive asymmetric hearing loss but can sometimes have fluctuating hearing loss.

   - Such patients rarely have true vertigo but may complain of imbalance.

5. Migraine-associated vertigo:

   - This is an important consideration, especially those with a history of migraine headache and in young patients with new onset of episodic vertigo.

   - Headache is usually present with migrainous vertigo, either during the episode or afterwards (when vertigo or tinnitus is a migraine aura). Migrainous vertigo is often accompanied by photophobia or phonophobia, symptoms not seen in vertigo episodes associated with Ménière's disease.

   - Successful treatment with **triptans** during an attack is generally diagnostic, although migrainous vertigo is less responsive than headaches.

6. Microvascular disease:

   - Diabetes

   - Hypertension.

7. Autoimmune disease

Note that TIAs rarely cause simultaneous vestibular and cochlear symptoms, they do not cause persistent tinnitus or objective hearing loss.

Investigations

There is no specific diagnostic test for Ménière's disease and so diagnosis is currently clinical. Definitive diagnosis can only be made postmortem.

Investigations aim to document sensorineural hearing loss and to rule out differential diagnoses.

Blood tests:

1. FBE

2. CRP

3. U&Es/ glucose

4. Syphilis serology

Audiometry

Audiometry should be performed in all patients suspected of having Ménière's disease.

The most common audiometric pattern in early Ménière's disease is a low frequency or combined low and high frequency sensory loss with normal hearing in the mid frequencies.

Over time the hearing loss evens out.

Electronystagmography (ENG):

Vestibular testing may be normal early in the course, but will eventually be abnormal on the affected side.

Testing is primarily useful in determining candidacy for interventional treatments or identifying possible bilateral disease.

A standard vestibular evaluation includes electronystagmography (ENG), rotary chair testing, and computerized dynamic posturography.

With progression of Ménière's disease, both the ENG and rotary chair test should show evidence of declining peripheral vestibular function in the affected ear.

The ENG (particularly the caloric test, in which the ear canals are irrigated with warm and cool water to stimulate the inner ear) is more sensitive for inner balance dysfunction, but the rotary chair test is more specific.

Vestibular evoked myogenic potential (VEMP):

VEMP is an emerging technology that has not yet been standardized or fully validated clinically.

Cervical VEMP (cVEMP) is an inhibitory sacculocollic reflex test that shows characteristic changes in symptomatic ears of Meniere patients, and may detect early saccular hydrops before the onset of classic Meniere symptoms.

Ocular VEMP (oVEMP) engages both utricular and saccular afferent nerve fibers and may also be useful in assessment of Meniere patients.

VEMP is a newer test that shows promise for both diagnosis and monitoring of disease progression and to identify the active ear in patients with bilateral disease.

CT Scan

A useful screen to exclude intracerebral bleeds, for acute presentations of vertigo which may be due to a central cause.

MRI Scan

This is the best imaging test.

Magnetic resonance imaging (MRI) can identify features that support a diagnosis of Ménière's disease, but the findings are not diagnostic.

MRI is indicated to rule out central nervous system (CNS) lesions that can mimic Ménière's disease, including CNS tumors, aneurysms, or stenosis of the posterior circulation, Arnold-Chiari malformations, and findings suggesting multiple sclerosis.

Management

Ménière's is difficult to treat, not the least because of uncertainty over its pathophysiology, and conclusive evidence for the efficacy of various pharmacological treatments in Ménière's disease is lacking. [1]

Determining the optimal treatment for Ménière's disease is limited by the lack of randomized, controlled trials. In addition, drug therapy has been associated with a significant *placebo effect*, and the relapsing and remitting nature of the disorder has made evaluation of various treatments problematic.

Treatment is therefore largely symptomatic. It may be possible to control vertigo symptoms, but the associated tinnitus and hearing loss are generally less responsive to treatment.

Preventive agents:

Preventive agents that are sometimes used include:

1. Hydrochlorothiazide 25 mg orally, daily: [1]

    - One aim has been to reduce endolymphatic pressure by reducing the sodium and water content of the endolymph with a thiazide diuretic. [1]

    - Alternatively, combination therapy with a thiazide and a potassium-sparing diuretic (e.g. amiloride, triamterene) may be used. When using thiazide diuretics, electrolytes should be monitored. Potassium replacement may be required in the presence of hypokalaemia.

2. Betahistine 8 to 16 mg orally, 2 or 3 times daily. [1]

- Betahistine is a vasodilator that has been used on the basis that it may increase blood supply to the inner ear, though this may not be its only mechanism of action in Ménière's disease. It may be a useful preventive treatment in some patients.

Prevention is also aimed at **avoidance of any particular triggers** that the patient may demonstrate, (see above).

Symptom Relief:

Symptom control is primarily directed to episodes of vertigo and vomiting

Agents that are typically used include:

1. Prochlorperazine

2. Promethazine

3. Benzodiazepines (for more severe symptoms).

Surgical Options:

Surgical options, may be considered when pharmacological therapy fails and symptoms are severe, although there is no agreement on which procedures are first line therapy.

Options include:

Non-destructive measures:

1. Endolymphatic sac decompression and sacculotomy:

    - These procedures are associated with a low risk of sensorineural hearing loss and are commonly performed for Ménière's disease in patients with intact hearing.

      Efficacy however is uncertain.

2. Intratympanic glucocorticoids:

    - Vertigo symptoms may improve after treatment with intratympanic glucocorticoids, without change in hearing or tinnitus.

Destructive measures:

1. Vestibular nerve section

2. Intratympanic gentamicin injection:

- Although this is used to control vertigo, treatment is associated with a moderate risk of **sensorineural loss (up to 30 percent of patients) that is irreversible.** It is usually reserved for those who already have significant unilateral hearing loss.

3. Surgical labyrinthectomy:

   - Surgical destruction of the bony and membranous labyrinth by removal of all of the neuroepithelium from the treated side relieves vertigo in virtually all patients but also causes **irreversible hearing loss in all patients.**

     Thus, it is only indicated in individuals with *severe and intractable* symptoms despite medical therapy who *already have poor or complete hearing loss on the affected side.*

4. Vestibular neurectomy:

   - Vestibular neurectomy involves surgical lysis of the vestibular nerve bundle as it enters the internal auditory canal. It relieves vertigo in 90 - 95 % of patients and is associated with a 10 - 20 % risk of of sensorineural hearing loss.

     However, vestibular neurectomy requires general anesthesia with craniotomy and overnight monitoring in an intensive care unit, and is associated with the potential for **significantly** more morbidity than labyrinthectomy.

Disposition:

All patients with suspected Ménière's disease should be referred *early* to an ENT specialist.

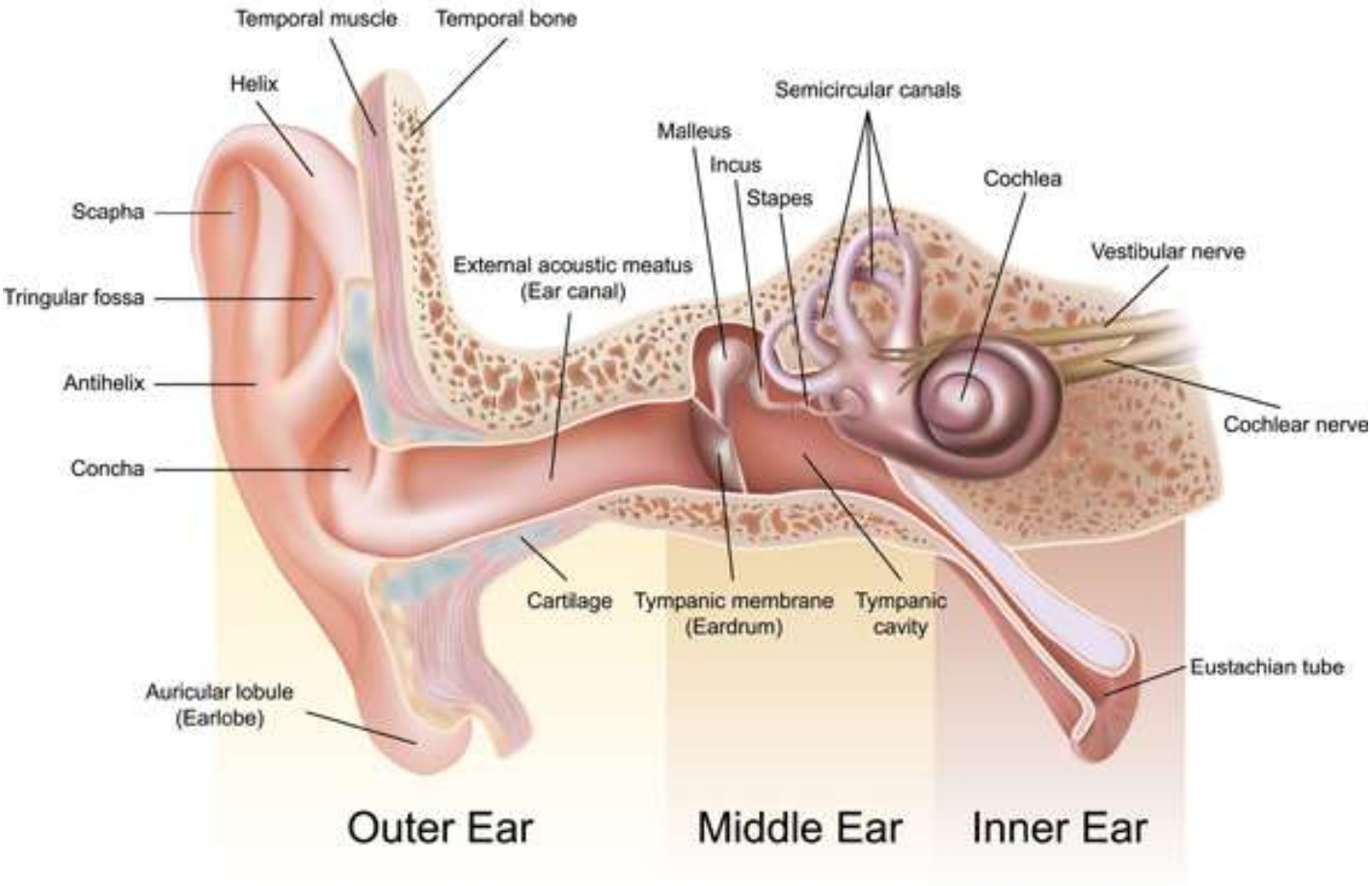
Patient Resources

Ménière's Australia:

"Ménière's Australia" is a national non-profit, non-government organization that aims to promote and facilitate the development of Australia-wide services and support for those living with Ménière's disease and other vestibular conditions

- www.menieres.org.au/

Appendix 1



Anatomy of the Ear.

Anatomy of the Inner Ear: [5]

The relevant anatomy centers on the petrous bone and the inner ear. The ear is divided into 3 sections: external, middle, and inner. The external ear consists of the auricle, external ear canal, and tympanic membrane. The tympanic membrane separates the external ear from the structures of the middle ear. The middle ear is an air-containing space that houses the 3 hearing bones: the malleus, the incus, and the stapes. The inner ear is completely encased in bone and consists of the cochlear-vestibular apparatus and its associated nerves.

The cochlear-vestibular apparatus is a complex structure arranged in a complex yet elegant spatial orientation. Because it is completely encased in bone, this structure is housed in a series of winding tunnels and interconnecting spaces. The mazelike orientation of these tunnels is appropriately named the labyrinth. The bone that encases it is the bony labyrinth.

The cochlea is a snail-shaped chamber that houses the organ of Corti. It is responsible for translating mechanical vibrations into electrical impulses and sending them to the brain through the cochlear nerve.

The vestibular system consists of a large chamber (i.e, the vestibule) from which 3 semicircular canals protrude. Within the vestibule, 2 sensors (the utricle and the saccule), detect linear acceleration, and the semicircular canals detect rotational movements in the 3 planes of rotation. The vestibular apparatus gives off 2 nerves: the superior and the inferior vestibular nerves. Together with the cochlear and facial nerves, the vestibular nerves travel through the internal auditory canal to the cerebellopontine angle.

The cochlea and the vestibular system are joined in the middle and share a dual-chambered hydraulic system. These hydraulic chambers are bathed by 2 fluids: endolymph and perilymph. Endolymph is produced primarily by the stria vascularis in the cochlea and also by the planum semilunatum and the dark cells in the vestibular labyrinth. Perilymph is protein-poor extracellular fluid. A membrane (i.e, the membranous labyrinth) separates the fluids and completely surrounds and contains the endolymph.

The system may be visualized as a water balloon floating in a pool. In this analogy, the water inside the balloon is the endolymph, and the balloon itself is the membranous labyrinth that contains the endolymph. The surrounding pool water is the perilymph, which supports the delicate nerve tissues of the membranous labyrinth. The walls of the pool represent the limits of the bony labyrinth space, and the ground encasing the pool is the bone that encases the labyrinthine space.

The endolymphatic sac is a reservoir pouch that resides on the posterior surface of the petrous bone against the posterior fossa dura. It is connected via the vestibular duct to drain into the endolymphatic space of the cochlea.

Endolymphatic flow has been described as following a "lake-river-pond" model. The endolymph flows from the endolymphatic fluid space (the lake) through the vestibular aqueduct (the river) to the endolymphatic sac (the pond). If there is obstruction, then endolymphatic hydrops will occur.

References

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5. J.C Li et al. Ménière's disease in eMedicine Website, 12 December 2014.

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