Approach to Ménière disease management

Vincent Wu MD  Edward A. Sykes PhD  Michael M. Beyea MD PhD
Matthew T.W. Simpson MD MSc CD CCFP  Jason A. Beyea MD PhD FRCSC

Abstract

Objective  To provide family physicians with an updated approach to the diagnosis and management of Ménière disease (MD), detailing the natural course of MD and describing how to initiate medical therapy while awaiting consultation with otolaryngology–head and neck surgery.

Sources of information  The approach is based on the authors’ clinical practices and review articles from 1989 to 2018. Most of the cited studies provided level II or III evidence.

Main message  Ménière disease is an uncommon disorder of the inner ear causing vertigo attacks with associated unilateral hearing loss, tinnitus, and aural fullness. It has a degenerative course that often results in permanent sensorineural hearing loss. On average, MD stabilizes with no further vestibular attacks by about 8 years after the onset of symptoms; however, this is highly variable. Vertigo symptoms can be controlled through a combination of dietary salt restriction, stress reduction, and medical therapy (betahistine, diuretics, or both). These can be initiated by family physicians before consultation with otolaryngology–head and neck surgery. Symptoms refractory to such strategies can be treated using nonablative, and occasionally ablative, therapies.

Conclusion  A thorough history is key to the approach to and management of MD and permits differentiating MD from other vestibular and nonvestibular conditions.

Ménière disease (MD) is a disorder of the inner ear, with hallmark symptoms consisting of spontaneous and episodic vertigo, unilateral sensorineural hearing loss (SNHL), tinnitus, and aural fullness. The presentation of MD can be highly variable between patients. In Canada, no population-based study has explored the incidence and prevalence of MD. International studies have reported an incidence of 8.2 to 13.1 per 100 000 person-years and a point prevalence between 120 and 513 per 100 000 persons.\(^1\)\(^-\)\(^5\) Ménière disease has an equal sex distribution, is more common among white patients, and has a peak age of onset between the fourth and fifth decade.\(^5\)\(^-\)\(^7\)

The development of endolymphatic hydrops—increased fluid within the inner ear organ—is observed (or presumed) in all individuals with MD.\(^6\)\(^,\)\(^9\) However, not all patients with endolymphatic hydrops have a history of MD.\(^8\)

Numerous factors including trauma, infection, ischemia, autoimmunity, and genetics have been implicated as causative of MD.\(^10\)\(^-\)\(^12\) Together, this suggests a multifactorial cause of the disease but also continued uncertainty surrounding the origin of MD.

Case description

Ms A. is a 48-year-old woman presenting to the family medicine clinic with a 6-month history of vertigo attacks. She reports that during the attacks, she sees the room spinning (true vertigo). The onset of the attacks is sudden, occurring 1 to 3 times per month, with vertigo lasting on average 30 minutes. She always gets warnings before the attacks with intensification of tinnitus in her left ear.

Editor’s key points

- Ménière disease (MD) is an uncommon cause of dizziness and vertigo. The first step in the approach to patients with dizziness or vertigo is delineating between true vertigo and other causes of dizziness. A thorough history is key.

- Treatment of MD ranges from conservative through nonablative to occasionally ablative therapies. Conservative therapy begins with diet modification and betahistine, which can be initiated by the family physician. Referral to an otolaryngologist is recommended for all patients suspected of having MD.

- While therapy does not exist that can halt or slow the progression of the hearing loss that accompanies MD, most patients obtain good control of their symptoms with personalized management.
She feels unsteady and nauseated during these episodes, often vomiting, with nearly constant ringing and a fullness sensation in her left ear. Her hearing in her left ear has been fluctuating over the past 6 months, but is overall worsening. She has no history of migraines or headaches. She is otherwise healthy, takes no medications, denies any tobacco or alcohol use, but does report consuming 2 to 3 large coffees per day.

Given the history of Ms A.’s true vertigo attacks lasting at least 20 minutes and her unilateral ear symptoms, MD is suspected.

Sources of information
The approach described is a nonsystematic review based on the authors’ clinical practices, along with research and clinical review articles from 1989 to 2018. Most of the cited studies provided level II or III evidence.

Main message
History of presentation. The symptoms of MD can overlap with those of other disorders, some of which can be life threatening. In a new patient with acute-onset vertigo, it is critical to first rule out stroke and other emergent intracranial pathologies (including intracranial tumours, brain infections, and traumatic brain injury). Revised MD diagnostic guidelines have been developed by the American Academy of Otolaryngology–Head and Neck Surgery, which will also be releasing an updated MD clinical practice guideline. A diagnostic approach is illustrated in Figure 1.

Characteristics of MD attacks: Classically, patients with MD experience recurrent perceptions of rotational movement of their environment. This is considered true vertigo. An important first step in the approach to MD is to delineate whether the sensation experienced by the patient is true vertigo or some other form of dizziness. Patients with MD experience vertigo attacks lasting 20 minutes to 12 hours (most lasting 2 to 3 hours) and have unilateral hearing loss and tinnitus, with or without aural fullness or pressure. Nausea, vomiting, sweating, and possibly diarrhea occur during these attacks, with onset typically preceded by a sudden intensification of unilateral nonpulsatile tinnitus.

Natural history of disease: Symptoms of MD might also fluctuate after initial presentation, with attacks varying in frequency and severity. Some authors suggest that vertigo can be triggered by excessive caffeine or sodium intake and changes in barometric pressure. Further, hearing loss typically becomes permanent as MD progresses. Patients who have bilateral MD might have fluctuations of symptoms in either ear and might have substantially reduced quality of life given bilaterally affected vestibular function and hearing.

Differential diagnosis: Unlike benign paroxysmal positional vertigo, which is the most common cause of peripheral vertigo, MD attacks last longer and are not reproducible by specific head movements. Similarly, dizziness without the illusion of spinning that is triggered by sudden changes in vertical head height are likely a result of orthostatic hypotension, commonly secondary...
to dehydration or cardiogenic causes. Vestibular migraines (VM) can present almost identically to MD, but patients with VM typically have a history of migraines. The vertigo episodes of VM might not be temporally associated with headaches, with the onset of vertigo sometimes appearing years after with headache-free intervals in between. Vertigo attacks in VM might last from minutes to days, and occasionally involve visual auras or allodynia. Viral infections of the inner ear, including vestibular neuritis and viral labyrinthitis, can mimic MD, with vertigo episodes lasting days to weeks. Viral labyrinthitis presents additionally with hearing loss. Another viral infection, Ramsay Hunt syndrome, secondary to varicella zoster virus reactivation within the vestibular ganglion, typically presents with the combination of vertigo and facial nerve paralysis. Superior canal dehiscence syndrome can present with vertigo that lasts seconds at a time and is associated with hyperacusis, whereby patients hear exaggerated sounds of body movements. Patients with acoustic neuroma (vestibular schwannoma) do not typically present with symptoms similar to MD. However, it should be considered in any patient who presents with asymmetric SNHL. Multiple sclerosis differs from MD by the presence of motor weakness, neuropathic pain, and sexual dysfunction. Moreover, symptoms present in MD can overlap with symptoms of cerebrovascular disease, but in the latter there are characteristically symptoms of visual disturbances, diplopia, peripheral weakness, and headaches.

**Physical examination.** Physical examination should be directed at excluding other causes of the patient’s symptoms, particularly those that might be life threatening (such as stroke). The HINTS (head-impulse, nystagmus, test-of-skew) examination can be performed to differentiate between peripheral and central causes of vertigo. Orthostatic vital signs should always be measured to rule out dizziness secondary to dehydration or cardiogenic instability that is frequently mistaken as vertigo. Once nonvestibular causes have been excluded, gait, tandem gait, Romberg, cranial nerve, and cerebellar testing are performed. Otoscopic examination is focused on evaluating for structural abnormalities within the external and middle ear that might be contributing to the patient’s symptoms. Pinna vesicles or tympanic membrane (TM) retraction pockets can help rule out conditions related to herpes zoster infections (such as Ramsay Hunt syndrome) or cholesteatomas, respectively. For patients with MD, otoscopy examination findings will typically be normal. Hearing can be initially assessed using a combination of Weber and Rinne tuning fork tests. The Dix-Hallpike maneuver can be used to identify patients with benign paroxysmal positional vertigo.

**Investigations**

**Bloodwork:** No biomarkers exist for the diagnosis of MD. Bloodwork is indicated for ruling out other conditions that might present similarly to MD including hyperthyroidism or hypothyroidism, diabetes, autoimmune disease, autoimmune inner ear disease or Cogan syndrome, and neurosyphilis.

**Ear-specific testing:** Complete diagnostic audiometric testing is necessary in all patients in whom MD is suspected. Audiometry typically shows low or combined low- and high-frequency SNHL with normal mid-frequency hearing. Vestibular testing through videonystagmography is always performed in patients suspected of having MD. It will often show a weakened caloric response in the affected ear. This testing can be organized by family physicians through the otolaryngology–head and neck surgery (OHNS) service, or by the consulting otolaryngologist.

**Imaging:** In patients with unilateral tinnitus, SNHL, or both, magnetic resonance imaging of the head should be considered. Magnetic resonance imaging should be performed if the physician suspects intracranial diseases that might mimic MD, including acoustic neuroma, aneurysms, Arnold-Chiari malformations, and multiple sclerosis.

**Management.** Once other disorders have been ruled out and the diagnosis of MD has been made (Box 1), the family physician is well positioned to initiate conservative therapy. A referral to OHNS should also be initiated. Treatment of MD is individualized based on the patient’s symptom control and is directed at minimizing the frequency, duration, and severity of vertigo attacks. While personalized MD treatment is successful in controlling vertigo symptoms, there is, unfortunately, no existing therapy that can halt or slow the progression of hearing loss. We acknowledge that management of MD can vary from centre to centre. The following is the approach we support and find to be successful in managing patients with MD while minimizing risk.

**Conservative therapy:** Counseling to reduce stress and lifestyle modifications such as dietary changes to minimize caffeine and alcohol intake are recommended. Restriction of sodium and monosodium

---

**Box 1. Diagnostic criteria for Ménière disease**

**Definitive criteria**
- Two or more spontaneous episodes of vertigo each lasting 20 minutes to 12 hours
- Documented sensorineural hearing loss greater than 30 dB below and above 2 kHz
- Fluctuating aural symptoms (hearing, tinnitus, and fullness) in the affected ear
- Not better accounted for by another vestibular disease

**Probable criteria**
- Two or more episodes of vertigo or dizziness each lasting 20 minutes to 12 hours
- Fluctuating aural symptoms in the affected ear
- Not better accounted for by another vestibular disease
glutamate intake has been associated with a reduction in vertigo attacks, by physiologically acting to decrease pressures in the hydropic ear.\textsuperscript{39,41} Recommended daily intake of sodium for adults in Canada should not exceed 2300 mg.\textsuperscript{42} Betahistine is recommended in MD and has been shown to improve vertigo, but only when taken regularly and prophylactically.\textsuperscript{43} Diuretics such as hydrochlorothiazide and triamterene have anecdotally been suggested to slow hearing loss by reducing fluid pressures in the hydropic ear, but evidence of their efficacy remains limited.\textsuperscript{44} Short-term use of oral prednisone can reduce the severity of vestibular symptoms by minimizing inflammation and autoimmune reactions that affect the vestibular nucleus.\textsuperscript{45} However, given their considerable systemic risks, such therapy is not typically recommended in MD. Benzodiazepines can be used judiciously to suppress vestibular symptoms during acute attacks.\textsuperscript{41} Nonablative and ablative therapies discussed below can be initiated by OHNS. 

**Nonablative therapies:** For the subset of patients whose symptoms are not well controlled by conservative therapy, intratympanic steroids can be offered for control of vertigo episodes. These are typically administered by OHNS in the clinic under local anesthesia. Studies support their usefulness in this respect, with suggested mechanisms being overall decreased inflammation and autoimmune reactions.\textsuperscript{41,46,47} There is a low risk of persistent TM perforation associated with this procedure.\textsuperscript{48} Commercial local overpressure devices have been introduced in recent years, but evidence is lacking showing their effectiveness in controlling MD symptoms.\textsuperscript{49}

Endolymphatic sac shunt surgery is another option for control of vertigo attacks. The effectiveness of this procedure has been debated.\textsuperscript{50} The physiologic rationale for this procedure is to drain excess endolymph, reducing the possibility of endolymphatic hydrops, and thus reducing the possibility of a vertigo attack. This surgery is typically only offered to patients whose symptoms are debilitating and who have failed to have adequate control of vertigo episodes after conservative therapy and intratympanic steroid injections.

**Ablative therapies:** Ablative therapies include intratympanic gentamicin, vestibular neuroectomy, or labyrinthectomy. Although these treatments are beyond the scope of this article, they are typically curative, but are not commonly needed as MD in most patients will be well controlled using the treatments discussed above.

**Driving restrictions:** Most patients with MD can continue to drive, provided they have sufficient warning before attacks. This needs to be determined in each case by the treating physician. The exception is patients with Tumarkin otolithic crises, also known as sudden drop attacks, which occur without warning. This subset of patients need their driver’s licences suspended by the treating physician through the provincial ministry of transportation. Once the patient has been free of drop attacks for at least 6 months, consideration should be given to reinstating the licence.

**Case resolution**

Recognizing Ms A.’s symptoms as consistent with MD, the physician performed a directed history and physical examination to rule out other potential causes of her symptoms. The clinical history of dizziness is clarified, and it is established that Ms A. was experiencing episodic vertigo. Otoscopy examination revealed normal TM with no fluid or masses within the middle ear space. Tuning fork testing revealed right-lateralizing Weber findings, and a Rinne test found air conduction was greater than bone conduction bilaterally, together suggestive of left-sided SNHL. Orthostatic vital signs and findings of a screening neurologic examination were normal.

A referral was made to an audiologist for hearing assessment and to OHNS. In the interim, Ms A.’s family physician counseled her on decreasing her caffeine and sodium intake. Ms A. was also started on oral betahistine (24 mg twice daily) for symptom management.\textsuperscript{40} Her otolaryngologist confirmed the diagnosis of left-ear MD after audiologic and vestibular testing. As her vertigo attacks persisted despite the conservative and medical therapy, she decided to undergo an intratympanic steroid injection. She found this to improve the frequency and severity of her attacks, in conjunction with continuing her conservative and medical therapy.

**Conclusion**

Ménière disease is an uncommon cause of dizziness and vertigo. However, the classic features are readily provided by patients during history taking. Migraine-associated dizziness and vertigo is a common mimicker of MD, which can also be managed initially by the family physician, with consideration given to a referral to neurology or neurotology (subspecialty of otolaryngology). As peripheral vestibular disorders are uncommon causes of dizziness and patient-reported vertigo, the first step in the approach to patients with dizziness or vertigo is delineating between true vertigo and other causes of dizziness. Treatment of MD ranges from conservative through nonablative to occasionally ablative therapies. Conservative therapy begins with diet modification and betahistine, which can be initiated by the family physician. Referral to an otolaryngologist is recommended for all patients suspected of having MD. Most patients with MD obtain good control of their symptoms with personalized management.

Dr Wu is a first-year resident in the Department of Otolaryngology—Head and Neck Surgery at the University of Toronto in Ontario. Dr Sykes is a medical student in the Department of Otolaryngology at Queen’s University in Kingston, Ont. Dr M.J. Beyea is a fourth-year resident in the Department of Emergency Medicine at Western University in London, Ont. Dr Simpson is a family physician in the Department of Family Medicine at Queen’s University. Dr J.A. Beyea is an otologist, neurotologist, and cranial base surgeon, Clinician Scientist, and Assistant Professor in the Department of Otolaryngology at Queen’s University, and Adjunct Scientist at ICES Queen’s.


