

Diagnosis and treatment of Meniere's disease

Often frightening and debilitating, Meniere's disease sends patients on a desperate search for relief—and for an explanation for their condition. This article helps you to offer both.

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Meniere's disease (MD) is a common reason for referrals to otolaryngology.¹ Although it is more poorly understood than many other disease processes, enough is known to make more frequent recognition and more appropriate treatment of MD possible in the primary care setting.

Etiology

There is some indication that endolymphatic hydrops (EH), an overabundance of endolymphatic fluid in the inner ear, may cause MD.² By itself, EH can cause hearing loss, aural fullness, and tinnitus—but not dizziness.

Opinions differ about the efficacy of various treatments for MD and the relationship between MD and migrainous vertigo. The expert consensus is that the two conditions are probably closely related.³⁻⁵ According to the diagnostic criteria created by the American Academy of Otolaryngology—Head and Neck Surgery, MD involves a triad of symptoms: tinnitus, fluctuant or

permanently impaired hearing, and episodes of vertigo.⁶ The tinnitus may be replaced or supplemented by aural fullness or pressure. The presence of episodic vertigo in the classic Meniere's triad is the element that distinguishes MD from EH (see Figure 1). Without dizziness, EH can be diagnosed but MD cannot.

Epidemiology

MD is estimated to affect 15 to 46 of 100,000 Americans yearly, with a prevalence of 218 per 100,000 persons. Onset is most often between 40 and 60 years.⁷ Precipitating events include upper respiratory tract infections, other infections, pregnancy, surgery, and other conditions that cause changes in fluid levels, which can affect the inner ear. Examples are head trauma (including whiplash injury), acoustic trauma, allergies, concurrent autoimmune disease, emotional stress, and fatigue.⁸ Some studies show a genetic component, which is difficult to assess because MD was less likely to have been accurately diagnosed in the past than it is now.⁹ Positive correlations have been noted between MD and hypothyroidism, migraine, and seropositive status for herpesvirus.^{5,10,11} Recent studies indicate that late-onset MD (with symptoms beginning as late as the eighth or ninth decade) is more common than had been suspected. Some of these cases are reactivations of long-standing disease.⁸

History

A thorough history is essential when MD is suspected. Since EH and MD differ only with respect to dizziness, be suspicious of inner ear disease when faced with a chief complaint of aural fullness or otalgia, of tinnitus

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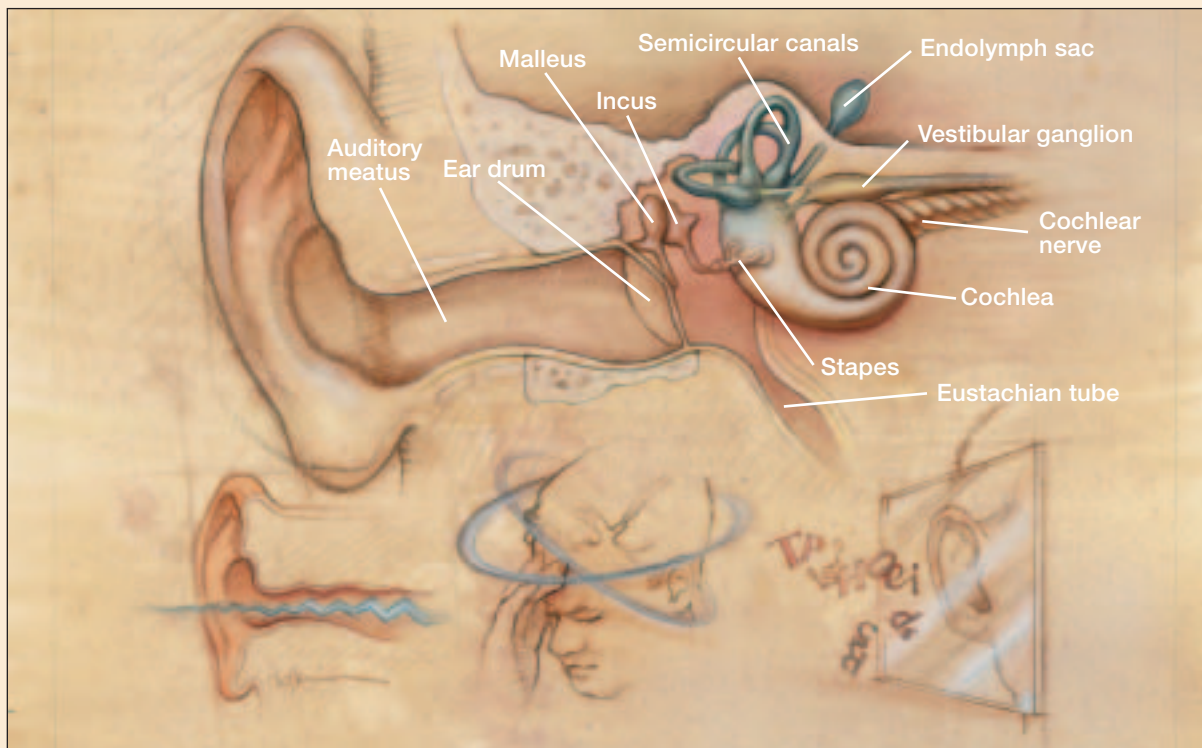
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Learning objectives

- Describe how Meniere's disease manifests and how it differs from endolymphatic hydrops
- Outline aspects of the physical assessment and diagnostic tests
- List pharmacologic and surgical options used to treat MD

FIGURE 1

Understanding Meniere's disease



Some evidence suggests that an overabundance of endolymphatic fluid in the inner ear may cause Meniere's disease, which is defined by a classic triad of symptoms according to diagnostic criteria created by the American Academy of Otolaryngology—Head and Neck Surgery: tinnitus, episodes of vertigo, and fluctuant or permanently impaired hearing.

Here and on the cover: Bonnie Hofkin

that seems unusually severe for the patient's age and ability to understand speech, or of fluctuant hearing. Since treatments for EH and MD are similar, distinguishing between the two diseases is of minor importance; if not treated, EH often progresses to MD.

Dizziness Ask the patient to determine how long the actual sensation of spinning or false sense of movement lasts, exclusive of the subsequent lingering imbalance and lightheadedness. It can be helpful to sketch a broken line and ask whether the patient has a few seconds of spinning sensation with each change of head or body position, with the entire episode comprising 20 minutes or more (the entire broken line is considered as a whole). MD usually involves vertiginous episodes lasting minutes to hours. If such an episode lasts longer than a day, a diagnosis of MD is less likely. Unless the onset of MD is very recent, the patient will almost definitely have experienced at least one episode of vertigo lasting 15 to 20 minutes.

Questions to ask Do otologic symptoms (possibly including temporary hearing loss) accompany the episodic dizziness? Do any two of the nonvertiginous

symptoms often occur together? Is there a family history of significant hearing loss (not due to noise exposure) and/or moderate to severe disequilibrium within the first six decades?

Note that people with MD often experience great discomfort during and after air travel and car rides over hilly terrain. Patients who experience sudden-onset hearing loss may describe things as sounding "distant." Ask whether the patient was upset, stressed, or fatigued before an episode since stress and fatigue are triggers in MD. Ask if the patient has ever suddenly fallen without loss of consciousness or other apparent cause—possibly even from a seated position. Such "drop attacks," also known as *Tumarkin otolithitic crises*, occur more commonly in advanced cases of MD than in early ones.^{12,13} Frequent tripping or stumbling may occur. In the later stages of MD, the patient may have the feeling of being on a rocking boat. This symptom may be chronic.

Tinnitus or aural fullness Tinnitus may be pulsatile or nonpulsatile, pitched high or low, narrow-band or broad-spectrum. Patients with MD often describe a

IN THIS ARTICLE

Key Points

- Meniere's disease (MD) most often develops between the ages of 40 and 60 years.
- A variety of events can precipitate the onset, including upper respiratory tract infections, other infections, pregnancy, surgery, and other conditions that cause changes in fluid levels.
- MD involves a triad of symptoms: tinnitus, fluctuant or permanently impaired hearing, and episodes of vertigo.
- The patient with MD should avoid risk factors as much as possible and may benefit from medications and from vestibular rehabilitation.

Competencies

Medical knowledge	◆◆◆◆◆
Interpersonal & communication skills	◆
Patient care	◆◆
Professionalism	◆
Practice-based learning and improvement	◆
Systems-based practice	◆

For an explanation of competencies ratings, see the table of contents.

sound like roaring of the ocean. Surprisingly, few papers on the topic of MD describe the quality of the tinnitus.

The author has found that roaring and buzzing are the most common descriptions offered by patients, followed by comparisons to rushing water or the chirping of crickets. Tinnitus of this kind should raise your index of suspicion. Aural fullness, pressure, and a sharp, stabbing pain are among the classic symptoms. Ask whether the otologic symptoms always involve the same ear, both ears simultaneously, or alternate ears. In one study, 29% of patients with MD reported they could influence the severity of their tinnitus by moving their mandible a certain way. Many others had cervical pain.¹⁴ Inquire about these possibilities, too.

Physical examination

The otoscope should be used to examine all patients who complain of aural fullness or otalgia. An otolaryngologist may have to perform a thorough examination of the external auditory canal and tympanic membrane with an operating microscope to help rule out a cholesteatoma or small retraction pocket. In some cases, CT of the temporal bone may be necessary, even after microscopy. Check for temporomandibular joint disease, which can cause aural fullness or pain.

Clinical signs of vestibular weakness or asymmetry in MD

Nystagmus, a rhythmic, involuntary movement of the eyes, is usually biphasic and consists of the eyes drifting in a horizontal, vertical, or rotary direction, followed by a fast component in the opposite direction. Check for spontaneous nystagmus by having the patient look straight ahead. Next, look for gaze-evoked nystagmus with the patient's gaze 30° to one side and then to the other. Do the same with the gaze in extreme left, right, up, and down positions. Look for noticeable asymmetry in the magnitude and speed of nystagmus with the gaze left or right.

You can diagnose congenital nystagmus, which is generally benign, by finding a specific direction of gaze that eliminates the nystagmus. In distinguishing between peripheral and CNS-related nystagmus, look for disconjugate gaze (nystagmus that beats vertically or changes direction). Patients with multiple sclerosis (MS) or diseases involving the medial longitudinal fasciculus may manifest this symptom.¹⁵

Smooth and saccadic pursuit Have the patient watch your index finger as you move it at a moderate, constant speed from side to side. Jerky, irregular movement of one or both eyes is indicative of poor smooth pursuit and, while not a sign of MD, is often secondary to CNS pathology. Also test saccadic pursuit by holding up your thumbs slightly to the patient's left and right and asking him to shift his gaze quickly from one side to the other. Signs of poor smooth pursuit also apply here.

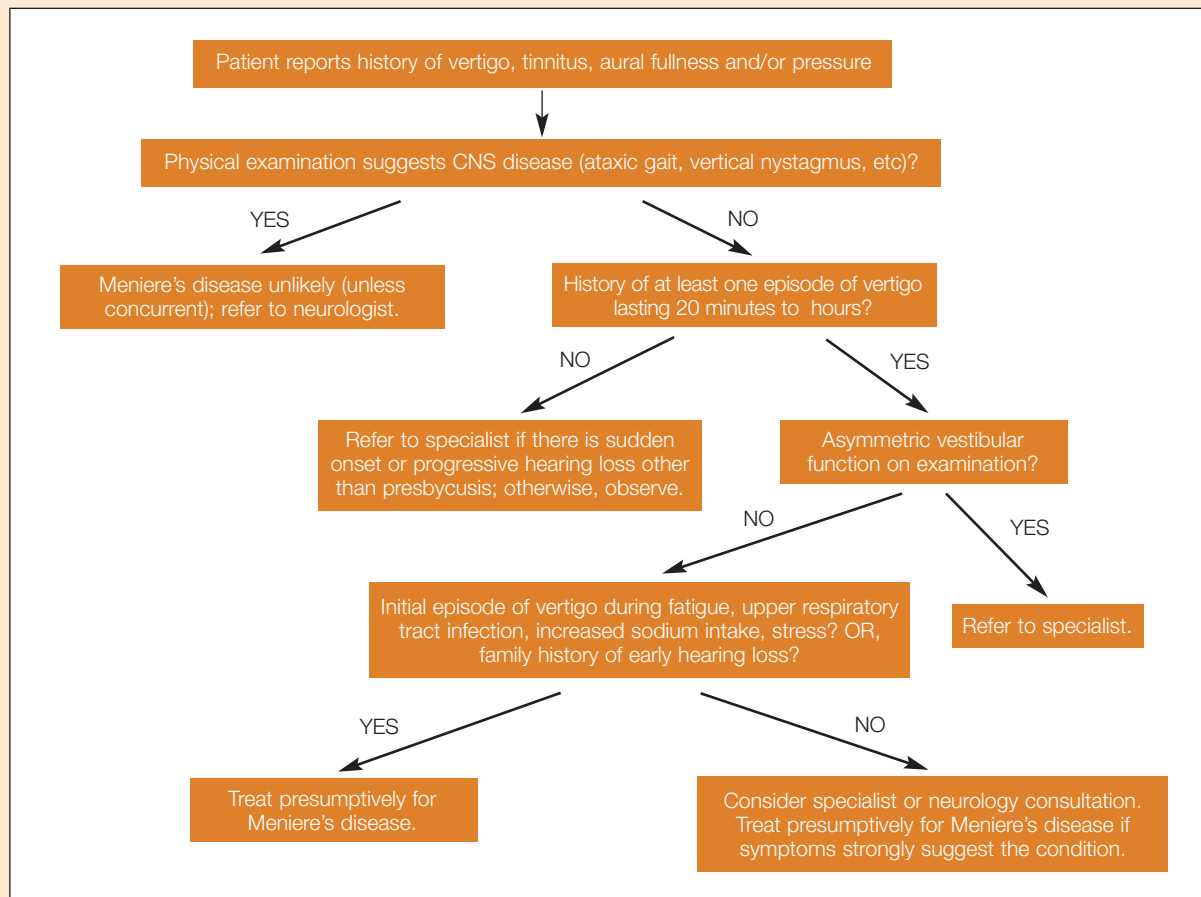
Vestibulo-ocular reflex (VOR)

Normally, when a person tries to maintain the gaze straight ahead during either active or passive head rotation, the VOR causes the eyes to rotate in an equal and opposite manner from the head. The VOR may be intact at one velocity of head rotation yet be impaired at another. Two simple tests allow the clinician to estimate the functional status of the VOR at high velocity and medium velocity, respectively:

To perform the Halmagyi head-thrust maneuver, hold the patient's head as you sit facing each other. Have the patient focus on the center of your forehead as you first slowly rotate her head about 30 degrees to the left; then rapidly bring it to midline with a jerking movement. Watch for a corrective saccade in which her eyes quickly jump to their target after the head thrust. If, after such a head-thrust to the right, you see her eyes jerk from the right to the left, you should suspect a weakness of the right vestibular system or a hyperactive left inner ear. The stimulation of the right vestibular system caused by the head rotation to the right was not sufficient to effect an equal and opposite rotation of the eyes to the left. Conversely, suspect a hyperactive right vestibular system if you see a catch-up saccade from left to right.¹⁶

ALGORITHM

Management of suspected Meniere's Disease



The Fukuda step test checks for a unilateral vestibular weakness. Have the patient stand with his feet close together and his arms folded across his chest while marching in place. A rotation of 15° or more after 20 steps suggests that there is a possible vestibular asymmetry.¹⁷

Other clinical assessments

Post-headshake test This simple test for asymmetric vestibular function is best done with the use of Frenzel's goggles if they are available. After 15 to 20 seconds of active, vigorous head-shaking (as in an emphatic shake, No!) with eyes closed, have the patient suddenly stay still and immediately open her eyes, with gaze straight ahead. Look for biphasic nystagmus. The overloading of the velocity storage center of the brain usually causes a nonlocalizing jerk nystagmus in the presence of unilateral vestibular weakness.¹⁶

Romberg's test This well-known test is primarily used to assess proprioception and somatosensory func-

tion. In the presence of acute loss of vestibular function, however, there may be a positive (abnormal) result.¹⁷

Past-pointing Sit opposite the patient with your index finger extended forward. Ask the patient to place his dominant index finger on yours (eg, the patient's right index finger on your left). Have the patient attempt, with eyes closed, to raise his hand up to ear level, and then lower his arm to touch your finger as before. If necessary, move your finger to let the patient hit the target. This will disguise the direction in which the patient was off so that you can better check for repeated inaccuracy.¹⁸ Poor performance on this test, on the nose-finger-nose test, and on the heel-shin test suggests cerebellar disease, as does dysdiadochokinesia seen during repeated opposite movements (such as slapping the palm and then the back of one hand at a time on the thigh).

Gait test Have the patient walk slowly forward down a hallway with eyes closed (unless the gait is already ataxic when the patient's eyes are open). An ataxic gait suggests possible CNS disease.¹⁷ The gait of a person with

TABLE 1

Differential diagnosis for Meniere's disease

Benign paroxysmal positional vertigo	Occurs more often in patients with Meniere's disease. May be concomitant. Benign paroxysmal positional vertigo is not associated with tinnitus, aural fullness.
Labyrinthitis/vestibulitis	Both conditions involve disabling vertigo, severe for 2-3d with gradual resolution over 2-3wk. These episodes rarely recur, unlike episodes of Meniere's disease.
Migrainous vertigo	Vertiginous episodes can last >1d, unlike those in Meniere's disease. Cephalgia need not coincide with vertiginous episodes. Ask about aura/prodrome, family history of migraines.

TABLE 2

Medications used in Meniere's disease

Medication	Dosage	Precautions, contraindications	Effectiveness	Adverse reactions
Triamterene-hydrochlorothiazide	Available in various dosage combinations	Avoid if patient has sulfa sensitivity, prediabetes, or hyperkalemia. Do not use potassium supplements.	Some patients do well on such a diuretic and dietary precautions alone, but this is not common.	Electrolyte disorders, jaundice, muscle cramps
Hydrochlorothiazide-spironolactone	25/25 mg, one daily	Avoid if patient has sulfa sensitivity, prediabetes, or hyperkalemia. Do not use potassium supplements.	Similar to triamterene/hydrochlorothiazide but can also help lower sodium.	Nausea, abdominal pain, diarrhea, headache
Lorazepam	0.5-2 mg One at onset of vertigo, repeat in 30 min if needed	Do not use with other CNS depressants. Has addictive potential so use during flare-ups and severe episodes only. Have patient test effect before need arises, at home.	Very effective for shortening episodes and decreasing severity. Provides more relief when used early in episode.	Sedation, weakness, ataxia
Clonazepam	0.5-1 mg twice or three times daily	Do not use with other CNS depressants. Has addictive potential. Patient must be weaned from drug slowly after extended use.	A less desirable choice. Can be used if dizziness is significant between episodes and other medications fail.	Drowsiness, ataxia, confusion, constipation
Betahistine	8-16 mg twice or three times daily	Contraindicated in pregnancy, recent or current peptic ulcer, pheochromocytoma, or uncontrolled hypertension.	A first-line choice but expensive if insurance does not cover it. Needs compounding. Often effective at reducing aural pressure, tinnitus, frequency and severity of vertigo.	GI upset, headache, possible fatigue in elderly, rash (rare)
Acetazolamide	125-250 mg total daily dosage, once to three times daily	Contraindicated in hypokalemia, hyponatremia, sulfa sensitivity, diabetes, others.	A second-line choice if betahistine is unavailable or not tolerated. Start low and titrate upward slowly.	Fatigue, paresthesias, GI upset

MD may be normal if there has been CNS compensation for a unilateral vestibular weakness, slow progressive symmetric bilateral loss, or no abnormal vestibular loss. Alternatively, veering to one side, or an unexpected degree of apparent uncertainty in a young or middle-aged patient, may be associated with MD. Actual ataxia may be due to such CNS disease as MS or a previous stroke.

Dix-Hallpike test This is useful for detecting benign paroxysmal positional vertigo (BPPV). A moderate to strong nystagmus with subjective dizziness is sufficient to diagnose BPPV. Be aware, though, that BPPV often occurs concurrently with MD.¹⁹

Laboratory and other diagnostic tests

Audiography is recommended for patients who have dizziness with associated otologic symptoms (tinnitus, aural fullness or pain, hearing loss). While an acoustic neuroma is an uncommon lesion, it can cause unilateral hearing loss with or without dizziness and/or tinnitus. An unexplained moderate or greater degree of asymmetry of pure tone thresholds—or of word discrimination, despite symmetric pure tone thresholds—calls for an otolaryngologic referral. In about 20% of cases of MD, there will be a rising slope in pure tone thresholds from 250 Hz to a peak (best hearing) at 2,000 or 3,000 Hz. The steeper the incline from the lower to the higher frequencies, the more important a thorough workup for MD becomes.

Electrocochleography (ECoG) indirectly measures endolymphatic pressure.²⁰⁻²² Unfortunately, it is not available in all areas. If there are audiologic findings like the rising configuration described above or a flat hearing loss in one ear (roughly the same threshold at many consecutive frequencies), a glycerol test can be used in place of ECoG. An initial audiogram is obtained, followed by immediate ingestion of 0.6 mL/lb of glycerol plus an equal volume of grapefruit juice. After three hours of fasting, audiography is repeated. Improvement by 5 to 10 decibels in two or more consecutive frequencies or a significant increase in the word discrimination score makes MD a very likely diagnosis.²³

Diagnosis

The diagnosis of MD is less straightforward than that of diseases that have definitive diagnostic parameters. The algorithm (page 37) illustrates a simple approach to diagnosis. Many researchers employ ECoG to confirm the clinical diagnosis or to measure response of the disease to treatment.^{24,25} Unfortunately, ECoG is not widely available or familiar to the primary care practitioner. The presence of such typical symptoms as moderate to severe aural fullness, tinnitus, otalgia, or hearing loss during an episode of dizziness increases the certainty of the diagnosis. An acute loss of low-frequency hearing may be expe-

rienced as aural fullness. Table 1 lists some signs and symptoms helpful in narrowing the differential diagnosis.

Treatment

Have the patient limit sodium intake to no more than 2,000 mg daily and avoid other risk factors, including caffeine, sugar, fatigue, and excessive stress when possible. Previous failure of a diuretic to alleviate symptoms should not deter the clinician from starting the patient on one or more of the medications listed in Table 2.

Other treatments include perfusions of the middle ear with dexamethasone, chemical ablation of the affected vestibule with transtympanic gentamycin, an endolymphatic sac shunt, and neurosection of the vestibular nerve. These treatment methods are reserved for particularly recalcitrant cases. The patient with MD often benefits from vestibular rehabilitation. □

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