

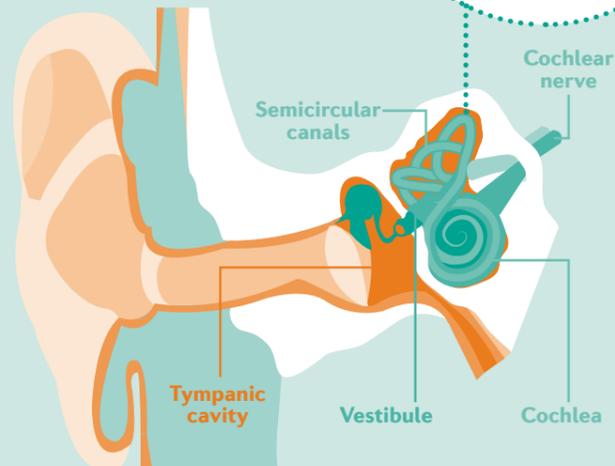
For the Primer, visit [doi:10.1038/nrdp.2016.28](https://doi.org/10.1038/nrdp.2016.28)

→ Meniere's disease (MD) is a multifactorial disorder of the inner ear that causes spontaneous episodes of vertigo attacks, fluctuating hearing loss, tinnitus and aural fullness.

## MECHANISMS

The aetiology of MD involves several factors. One characteristic sign of MD is the presence of endolymphatic hydrops (EH), which might cause neural damage. Interestingly, nerves, specifically the ganglion cells — rather than sensory hair cells — are affected by MD. Hearing loss typically starts with low-frequency tones, consistent with the location of the basilar membrane (involved in hearing), which is first affected by EH (at the apex of the cochlea). Vertigo is thought to arise from pathological firing of the auditory nerves. A possible explanation for this misfiring is leakage of potassium-rich perilymph into the endolymphatic space due to the rupture of the membrane separating these two spaces (Reissner's membrane). Alternatively, the misfiring can be explained by abnormal movements of endolymph.

EH is abnormal, transient fluctuations of endolymph in the cochlea (involved in hearing) and vestibular system (involved in balance).



## DIAGNOSIS

MD is a heterogeneous disease; patients do not necessarily have all symptoms and symptoms vary over time.

### TINNITUS

Continuous ringing noise in the ears



### AURAL FULLNESS

Feeling of pressure in the ears



### FLUCTUATING HEARING LOSS



### VERTIGO

A spinning sensation



Diagnosis of MD is based on medical history, sometimes complemented with inner ear tests such as audiometry, vestibular-evoked myogenic potential testing, caloric testing, electrocochleography and head impulse tests. Direct visualization of EH has recently been introduced, but is not yet routinely used in the clinic.

## Rx MANAGEMENT

Few evidence-based management options are available for MD. Drugs that can be used to bring relief during acute vertigo attacks are centrally acting antihistamines with anticholinergic action and benzodiazepines. Managing risk factors (such as migraine, high blood pressure and vascular conditions) and medical treatment with drugs that improve fluid regulation (for example, diuretics) and steroids might reduce the frequency of attacks. Neurectomy or labyrinthectomy can be performed for refractory disease.

When medical treatment fails to suppress vertigo attacks, intratympanic gentamicin therapy or endolymphatic sac decompression surgery is usually considered.



## EPIDEMIOLOGY

The estimated prevalence of MD is in the range of 0.02–0.5%; this high variability is probably explained by the complexity of diagnosing MD. The age of onset peaks at 40–50 years, with the highest prevalence in those

60–70 years of age. Paediatric cases are relatively rare. Women are more prone to developing MD than men. Several genetic risk factors have been

! The criteria that define MD have evolved over time. MD is classified as possible, probable, definite and certain MD. However, updated guidelines published in 2015 only take definite and probable MD into account. Classification depends on the presence of the characteristic symptoms and their severity; only the classification of certain MD requires confirmation of the presence of EH.

identified. Importantly, MD is often found in combination with migraine, allergy and immune (or autoimmune) conditions.

## QUALITY OF LIFE

Patients with MD have a poorer quality of life than people without the condition, especially in the health-related domains. The effect of hearing loss has been rated as more severe than vertigo, tinnitus and aural fullness. The unpredictability of the symptoms can lead to anxiety and social withdrawal. Social support and coherence (ability to cope with stress) can greatly improve quality of life.