Ménière's Disease

This is a disorder of the inner ear caused by a change in fluid volume in the labyrinth. If the cause is known, the condition is more properly called Ménière’s syndrome.

In the inner ear are the cochlea (for hearing) and the vestibular apparatus (for balance). The vestibular apparatus is a set of tubes enclosed by the membranous labyrinth. The membranous labyrinth contains fluid called endolymph.

In Ménière’s disease there is a progressive distension of the membranous labyrinth, which is called 'endolymphatic hydrops'. This may injure the vestibular system, causing vertigo; or the cochlea, causing hearing loss. [1]

The condition is diagnosed on the basis of the following three factors:
- Clinical features.
- Audiometric findings.
- Exclusion of other causes.

Aetiology [1]

The exact cause is unknown but is probably multifactorial. Possible risk factors include:

- Allergy - eg, food allergy.
- Autoimmunity, including antiphospholipid antibodies, rheumatoid arthritis and lupus.
- Genetic susceptibility.
- Metabolic disturbances involving the balance of sodium and potassium in the fluid of the inner ear.
- Vascular factors (there is an association between migraine and Ménière’s disease).
- Viral infection, otosyphilis, Cogan's syndrome (non-syphilitic interstitial keratitis and bilateral audiovestibular deficits).

Epidemiology [2]

- One estimate reported a UK prevalence of 157 per 100,000.
- The condition can occur at any age but the peak incidence is 40-60 years.
- The condition is probably rare in children but has been reported in this age group. [3]

Presentation [2, 4]

Symptoms

Core symptoms are vertigo, tinnitus and fluctuating hearing loss with a sensation of aural pressure. The hallmark of the disease is its fluctuating and episodic pattern of symptoms:

- Acute attacks typically last minutes-hours, often 2-3 hours.
- Acute episodes may occur in clusters of about 6-11 per year.
- Remission of symptoms may last several months.
- Most patients develop unilateral symptoms initially. Bilateral symptoms may develop, often many years later.

Other symptoms:
Some patients have 'drop attacks', ie sudden unexplained falls without loss of consciousness or associated vertigo. This is reported in about 4% of Ménière's disease patients.

Imbalance is sometimes reported. This tends to occur once the spinning sensation has decreased.

Three stages of disease are described, although patients do not necessarily progress through all these:[5]

- Early-stage - predominantly vertigo attacks which are sudden and unpredictable. Hearing worsens and tinnitus increases. There is good recovery between attacks; these remissions can last days-years.
- Middle-stage - continuing episodes of vertigo; there may be giddiness before and after attacks. Sensorineural hearing loss develops. Tinnitus also progresses. Periods of remission vary; they may last several months.
- Late-stage - hearing loss increases. Vertigo lessens; balance may be difficult, especially in the dark. Tinnitus persists.

Diagnostic criteria[1, 2, 4]

For a firm diagnosis, the following symptoms should be present:

- Vertigo - at least two spontaneous episodes lasting at least 20 minutes within a single attack of Ménière's disease.
- Tinnitus and/or perception of aural fullness.
- Hearing loss confirmed by audiometry to be sensorineural in nature.

Examination

There are no diagnostic signs. Examination of cardiovascular, neurological and ENT systems is advisable to look for other causes of similar symptoms. Examine for:

- Anaemia, blood pressure (lying and standing), arrhythmias, carotid bruits.
- Cranial nerves (including nystagmus), gait and co-ordination (Romberg's test and finger-nose test).
- Ears for wax; hearing tests (Weber's test and Rinne's test).
- Cervical spine for vertigo associated with cervical spondylosis and neck pain.
- The Hallpike manoeuvre is used to diagnose benign paroxysmal positional vertigo (BPPV).

Differential diagnosis[1]

Many other conditions can present with vertigo, tinnitus or deafness. (It is the combination that helps to diagnose Ménière's disease.) In primary care, common causes of vertigo are BPPV, acute vestibular neuronitis and Ménière's disease.

Other ENT causes:

- Exclude acoustic neuroma in anyone with unilateral deafness, tinnitus and/or facial nerve palsy.
- Otitis media.
- Earwax.
- Ototoxic drugs.

Intracranial pathology - for example:

- Vertebrobasilar insufficiency, transient ischaemic attack (TIA), stroke, thrombosis of labyrinthine artery.
- Intracranial tumours.
- Migraine. Migraine-associated dizziness may present like Ménière's disease.

Systemic illness:

- Anaemia
- Hypothyroidism
- Diabetes mellitus
- Autoimmune disease
- Syphilis
Investigations\textsuperscript{[1, 2, 6]}

Blood tests to exclude systemic illness - for example:

- FBC, ESR, thyroid function, syphilis screen, fasting glucose, renal function, lipids.

Audiometry is recommended:

- This helps to diagnose Ménière's disease if sensorineural hearing loss is found.
- A test for loudness recruitment (if feasible) is sensitive but not specific for Ménière's disease.
- During the early stages, hearing loss can be transient, making it difficult to confirm hearing impairment by audiometry. Serial audiograms may help.
- Further details of possible audiometric findings are available.\textsuperscript{[4]}

Diagnosis may be aided by:

- Video nystagmography or electronystagmographic testing with bithermal caloric evaluation.
- Electrocochleography.
- Brainstem auditory evoked potentials.

Radiology:

- MRI brain scan - is advised for unilateral cases of Ménière's disease, to exclude other causes of unilateral vertigo and hearing loss - eg, acoustic neuroma. This should include views of the internal auditory canal, with and without contrast.
- Standard lateral mastoid radiographs - can aid diagnosis by documenting the forward location of the sigmoid sinus, seen in almost all patients with Ménière's disease.\textsuperscript{[4]}

Management\textsuperscript{[2]}

Driving\textsuperscript{[7]}

Drivers of any type of vehicle are required to send a form to the Driver and Vehicle Licensing Agency (DVLA) - available from their website - if they have vertigo, irrespective of the cause. Each case is taken on its own merits. Factors taken into consideration include whether attacks are preceded by a warning, how disabling they are, whether medication has been started and whether the condition is under control.

Treatment

The aim of therapy is to:

- Alleviate acute attacks.
- Reduce severity and frequency of attacks.
- Improve hearing and reduce the impact of tinnitus.

Acute attacks

- Vertigo and nausea can be alleviated by prochlorperazine, cinnarizine, cyclizine, or promethazine.
- If there is vomiting, buccal or intramuscular doses may be needed.
- Consider patient preference for both choice of drug and route of administration.
- For severe symptoms, hospital admission may be needed to maintain hydration.
- Intramuscular steroid injection followed by a tapering dose of oral steroids has also been recommended.\textsuperscript{[2]}
Prophylaxis[1]

- Lifestyle measures may be helpful. Low-salt diet and avoiding caffeine, chocolate, alcohol and tobacco are often advised. Excessive fatigue appears to be a trigger factor in some patients and should be avoided. Evidence to support these measures is, however, lacking.
- Drug prophylaxis:
  - Consider a trial of betahistine (initially 16 mg three times a day) to reduce the frequency and severity of attacks.
  - Diuretics may be helpful but are not usually recommended for use in primary care.

Supportive measures

- Safety - if prone to sudden vertigo, consider safety and risks with activities involving heights, dangerous machinery, swimming, etc. See 'Management', above, for driving regulations.
- Vestibular rehabilitation programmes:[8]
  - These seem to be effective in some situations - eg, for stable vestibular loss or for stable, unilateral vestibular disease.
  - The programmes involve exercises such as learning to bring on the symptoms to 'desensitise' the vestibular system; learning to improve balance, co-ordination and coping skills.
- Maintain mobility:
  - After an acute attack of vertigo, patients naturally tend to sit still. Encourage them to move around to promote central compensation, where the brain uses vision and other senses to compensate for the loss of vestibular function.
- Hearing support:
  - Hearing aids tailored to pattern of hearing loss.
  - For tinnitus - masking devices, sound therapy and relaxation/distraction techniques. See 'Further reading & references', below, for information.
  - Avoid loud noise, if intolerant to it.
- Acupuncture may be beneficial.[9]

Further treatments[2, 4]

- Local gentamicin treatment:
  - This is known as transtympanic gentamicin perfusion, transtympanic gentamicin injection or intratympanic gentamicin injection.
  - The aim is to use the damaging action of gentamicin on the sensorineural epithelium and labyrinthine cells to reduce vertigo, while preserving hearing (although there may be a risk of sensorineural hearing loss).
  - Recent studies suggest that endolymphatic surgery is superior in terms of preserving hearing loss and relieving vertigo.[10]
- Local steroid injection - this is transtympanic or intratympanic dexamethasone injection.
- Pressure pulse treatment (Meniett® device):
  - This is a non-invasive treatment for intractable vertigo, comprising positive pressure provided through a pulse generator into the ear canal.
  - Reports from small studies found it to be effective, although long-term efficacy may be poor.
Surgical treatments:
- Endolymphatic sac surgery - this involves decompression of the endolymphatic sac and sigmoid sinus. The effectiveness of this treatment has been confirmed by a recent Cochrane review. [11]
- Vestibular nerve section - aims to cure vertigo while preserving hearing.
- Micropressure therapy has been endorsed by the National Institute for Health and Care Excellence (NICE). This involves inserting a grommet via the eardrum into the middle ear and blowing air at low pressure into the inner ear. The aim is to reduce pressure in the ear, thus obviating the need for more invasive surgery. [12]
- Labyrinthectomy - this is a last option, as hearing in that ear would also be lost.

Complications [5]
- Loss of driving licence, if symptoms are sudden and disabling. [7]
- Quality of life can be severely affected in some people.
- Depression or anxiety.

Prognosis [5]
- There is no cure but most patients can be helped by the above treatments. The Ménière's Society suggests that 80% of patients will have their symptoms alleviated by non-invasive treatments.
- The disease course has three stages (as above) but does not necessarily progress in all patients.
- In about 50% of patients, the disease eventually affects both ears.

Further reading & references
- The British Tinnitus Association
- Action on Hearing Loss

1. Meniere’s disease; NICE CKS, September 2012 (UK access only)
5. Ménière's disease; Ménière's Society
7. Assessing fitness to drive: guide for medical professionals; Driver and Vehicle Licensing Agency
12. Micropressure therapy for refractory Ménière's disease; NICE Interventional Procedure Guideline, April 2012

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