Menière's disease

Lance Hollis, Ian Bottrill

Menière's disease is an idiopathic condition classically described as a triad of vertigo, deafness and tinnitus. A wide differential diagnosis exists, but acoustic neuroma must be actively excluded. Various medical and surgical treatment options are available, although two-thirds of patients spontaneously resolve with time.

Mr Lance Hollis is Specialist Registrar and **Mr Ian Bottrill** is a Consultant Ear, Nose and Throat Surgeon in the Department of Ear, Nose and Throat, Head and Neck Surgery, Radcliffe Infirmary, Oxford OX2 6HE

Correspondence to: Mr L Hollis

The triad of tinnitus, vertigo and deafness was first described in 1861 by Prosper Menière and reported to the Paris Academy of Medicine. Despite the passage of time, no clear aetiological factor has ever been found to be the sole cause of the disease. It is recognized, however, that 'endolymphatic hydrops' is the prime pathology. This refers to distension of the endolymphatic compartment of the inner ear channels either as a result of endolymph over-production or impaired reabsorbtion (*Figure 1*). When this condition is truly idiopathic it is known as Menière's disease and, when secondary to a known cause, it is known as Menière's syndrome.

INTRODUCTION

As stated above no clear aetiological factor of Menière's disease has been found but numerous theories abound and these will be discussed.

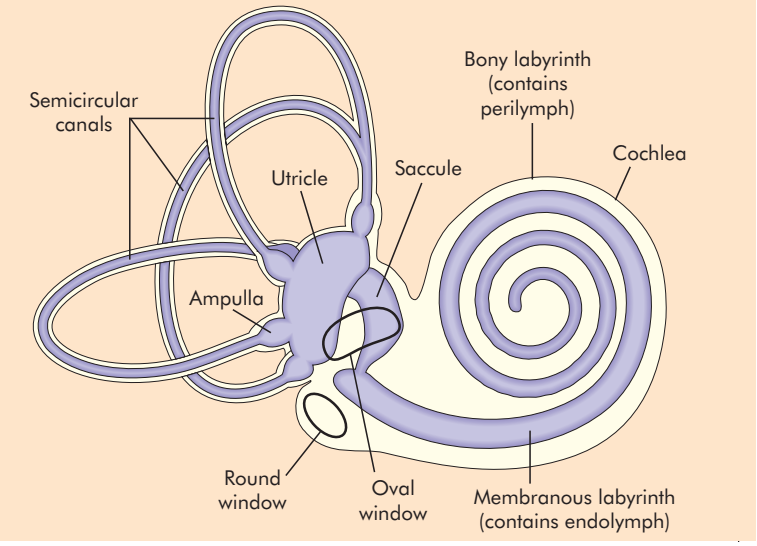

Figure 1. Schematic view of the labyrinth.

Diagnosis of vestibular disorders is often confusing; this is especially true in Menière's disease as a wide spectrum of symptomatology may be present. This said, diagnosis often rests on the history rather than on any investigations being performed. However, there are important differential diagnoses to bear in mind, and those such as acoustic neuroma must be assiduously excluded.

It might be expected that any disease without clear cause must be a challenge to treat. This has certainly proven to be the case with Menière's disease, with many medical and surgical treatments having been advocated, ranging from the ingenious to at best optimistic.

HISTORICAL BACKGROUND

Before Prosper Menière's description of the labyrinthine pathology, patients suffering from symptoms including vertigo, nausea and vomiting were grouped under the diagnostic umbrella of 'apoplectiform cerebral congestion' or 'nervous deafness'. It was not until Menière's work that such patients were recognized as suffering from a distinct disorder whose underlying pathology resided in the labyrinth. The idea of elevated intracochlear pressure originated with Knapp in 1871 which he described as 'aural glaucoma'. This idea, like Menière's hypotheses, was ahead of its time and was not confirmed histologically for another 50 years. It was not until 1938 that Hallpike and Cairns first described the histological changes which are now so widely recognized. They described gross dilatation of the saccule and scala media with obliteration of the perilymph spaces of the vestibule and scala vestibuli (*Figures 2* and *3*).

CLINICAL FEATURES

The American Academy of Otolaryngology-Head and Neck Surgery (Committee on Hearing and

Equilibrium, 1985) (AAO-HNS) states that deafness, vertigo, tinnitus and aural fullness are the cardinal features of Menière's, although they may not all be present in each patient. Adjunctive vertiginous features are also recognized, such as long-lasting feelings of unsteadiness or floating. The deafness is characteristically a fluctuating sensorineural loss that worsens during a vertigo attack. In the early stages the hearing loss is mainly in the low frequencies. Patients may also notice that the same frequency of sound is heard as a different pitch in each ear (diplacusis).

The tinnitus may be of any frequency, but is often low pitched. It may increase in volume in conjunction with the fullness, before the onset of a vertigo attack. Attacks are usually spontaneous and often occur at rest. The vertigo of Menière's classically lasts from 24 minutes to 24 hours, but not less than 5 minutes. This helps in the differential diagnosis. Rarely, sudden falling attacks occur in which the patient may fall as if pole-axed. This is known as Tumarkin's utricular crisis. A rare variant of Menière's is Lermoyez's syndrome in which progressive deterioration of the hearing is followed by vertigo, after which the hearing improves.

DIFFERENTIAL DIAGNOSES

Central causes of vertigo

Lesions affecting the CNS (such as multiple sclerosis) tend not to produce paroxysmal attacks of vertigo. Spontaneous nystagmus and other neurological signs may be present.

Infective labyrinthitis

Middle ear disease causing labyrinthitis should be easily visible on otoscopy.

Benign positional vertigo

Auditory symptoms are absent and the positionally induced vertigo only lasts for a matter of seconds in benign positional vertigo.

Sudden vestibular failure

Vertigo often lasts for more than a week with gradual improvement as central compensation occurs.

Acoustic neuroma

Approximately 5% of acoustic neuromas present with Menière's-type symptoms and so patients presenting for the first time must have this serious condition excluded with a magnetic resonance imaging (MRI) scan of the internal auditory meati and cerebellopontine angles.

AETIOLOGY

Virtually all parts of the 'surgical sieve' have been described in the aetiology of Menière's disease (*Figure 4*) and evidence for each will be described in turn.

Autoimmune

This has a female preponderance, with onset in the third decade. Immunoglobulins and T lymphocytes have been found in association with Menière's disease. An HLA association has been found. Of cases seen, 30% are bilateral. Some patients respond to steroids and plasmapharesis.

Chronic inflammation

Perisaccular and vestibular fibrosis has been found in the temporal bones of some Menière's patients at prosection.

Metabolic

Abnormal glycoprotein metabolism may occur in the endolymphatic sacs of Menière's patients.

Viral infection

Herpes simplex, cytomegalovirus, Epstein–Barr virus and enteroviruses have been implicated in the aetiology of Menière's disease.

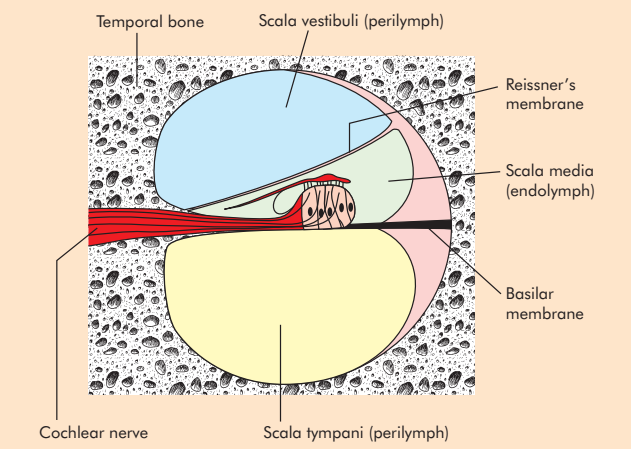

Figure 2. Pathological specimen of the normal cochlea fluid compartments.

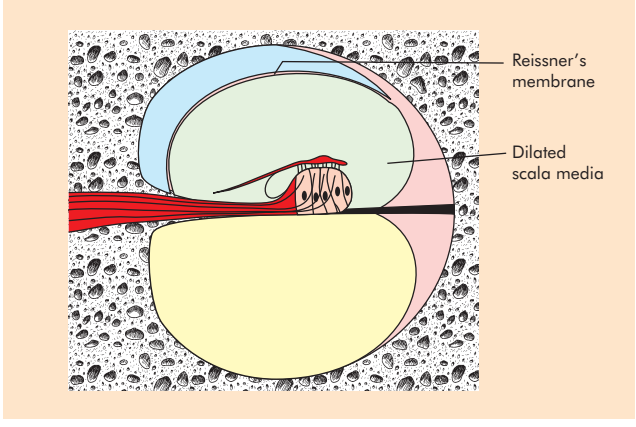

Figure 3. Pathological specimen of a patient with Menière's disease showing the dilatation of the scala media.

Genetic link

A Menière's locus was postulated between the HLA-C and HLA-A loci on the short arm of chromosome 6.

Vascular

Migraine prevalence may be higher in patients with Menière's disease.

Secondary hydrops (Menière's syndrome)

There are a number of causes of Menière's syndrome which include acoustic or temporal bone trauma, chronic suppurative ear disease, leukaemia, otosclerosis and Paget's disease.

INVESTIGATIONS

Pure tone audiometry

Low tone fluctuating sensorineural loss seen in early stages with more severe and permanent loss in 'burnt out' end-stage Menière's disease.

Electrocochleography

This is an investigation in which electrical potentials within the cochlea are measured by means of a transtympanic electrode. Characteristic changes to the summating potential (SP)/action potential (AP) waveform are seen (*Figure 5*). These changes may be reversed in the glycerol dehydration test in which plasma osmolality is increased by at least 10 mOsm/kg after the oral administration of flavoured glycerol.

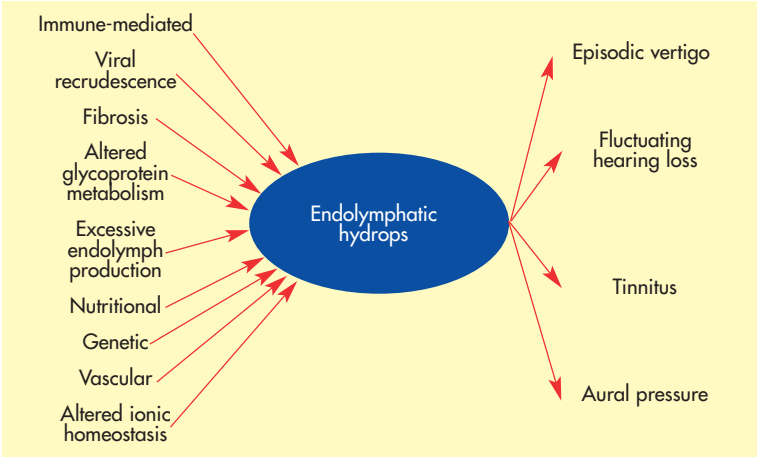

Figure 4. Schematic demonstration of the postulated aetiological factors in Menière's disease.

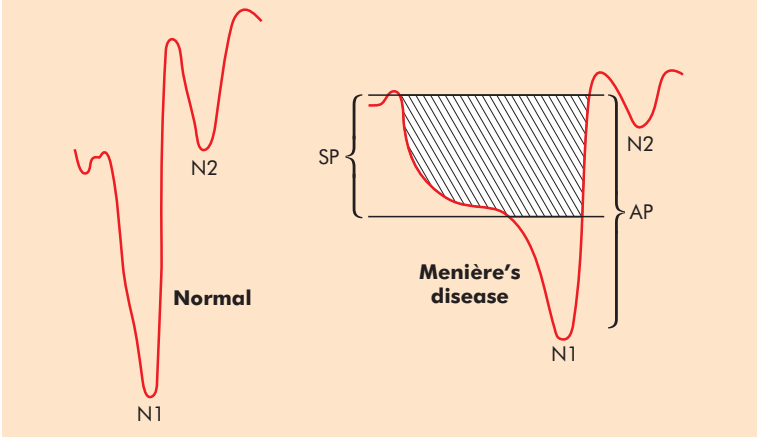

Figure 5. Electrocochleogram of a normal ear and one affected by Menière's disease.

Caloric testing

This may demonstrate a canal paresis. An MRI scan of internal auditory meatus and cerebellopontine angle should be performed.

TREATMENT

As the cause of Menière's is not known, and is almost certainly multifactorial, research into effective treatments is dogged by triallists' inability to include a homogeneous patient group (Brookes, 1996), and the fact that Menière's has a tendency to spontaneously resolve: Silverstein et al (1989) showed a 71% complete resolution in untreated patients who were followed up for 8 years.

It is also difficult to recruit enough patients for statistical validity and until recently standardization of reporting was not widespread. In addition, the placebo effect of any modality of treatment is well recognized.

These factors have resulted in limited double-blind, placebo-controlled trials of medication and virtually none involving surgery.

Treatment involves several steps:

- Management of the acute vertigo attack
- Attempts to reduce the frequency of the attacks
- Ablation of the vestibular organ/nerve.

Medical treatment

Patients experiencing acute rotatory vertigo may be helped by the use of vestibular suppressants such as prochlorperazine or cinnarizine. If the attacks are accompanied by vomiting, the medication can be given either sublingually, rectally or intramuscularly. If the patient is very distressed, it may be helpful to use a benzodiazepine for sedation, assuming there are no contraindications.

This type of medication should not be continued once the acute event is over and the patient is starting to mobilize. The patient should be actively encouraged to mobilize to promote central compensation; the patient's natural tendency is to sit still to prevent any dysequilibrium.

Therapies aimed at reducing the frequency of attacks can be divided into two groups: vasodilators, to encourage reabsorbtion of endolymph, and diuretics, to actively eliminate endolymph.

Betahistine has been shown in some controlled studies to be beneficial in prophylaxis. Combined with the treatment's safety profile, this makes betahistine the first line of treatment.

The usual starting dose is 16 mg three times daily, and this can be continued for many years if the patient appears to be benefitting.

There are also trials showing benefit from using low-dose bendrofluazide, 5 mg once daily, but the effect is marginal at best. Bendrofluazide should only be added if betahistine alone is ineffective; however, the patient's potassium levels will need to be checked before commencing treatment (Bottrill, 1998).

Dietary restriction of salt and vasoactive foodstuffs may provide improvement, but is not supported by scientific evaluation.

Aspects other than the vertigo might need to be evaluated. These may include a hearing loss, for which a hearing aid may be appropriate. Tinnitus can be very distressing for some patients, and patient information literature or tinnitus retraining therapy may be appropriate.

If patients continue to get troublesome attacks following a good trial of medical therapy, further surgical treatment may be an option.

Surgical treatment

The available surgical options have either of two possible effects: reducing endolymphatic hydrops or reducing over-stimulation of the vestibular nuclei by the labyrinth.

It is noteworthy that numerous procedures to treat Menière's have been described over time, this fact is often an indicator of failure of efficacy of such procedures. At present endolymphatic sac decompression is the most popular procedure to reduce endolymphatic hydrops. The reasons for this are that it aims to correct the main functional defect of the disease, i.e. endolymphatic hydrops, and also preserves hearing which would otherwise decline.

Nowadays, vestibular nuclei stimulation can be reduced by three main methods: intratympanic gentamicin injection, osseous labyrinthectomy and vestibular nerve section.

Intratympanic gentamicin injection is toxic to the vestibular sensory cells and prevents the effects of the hydropic changes reaching the brainstem and being perceived as vertigo (Nedzelsski et al, 1992). This is only effective for vertigo control and does not alter the underlying pathology. In addition total hearing loss in the treated ear is a possible complication and so the procedure is generally only used in patients with pre-existing profound hearing loss in the affected ear. Similarly, steroids such as dexamethasone may be administered by this route in the attempt to reduce immune damage to the vestibular organs. However, there is little evidence of this method's efficacy.

Osseous labyrinthectomy removes all inner ear neuroepithelium, but leaves a permanent total hearing loss on the affected side and is reserved for patients with no serviceable hearing.

Vestibular nerve section is an intracranial procedure in which the vestibular nerve is divided as it enters the brain. In unilateral cases it has the highest success rate of about 95% complete control at 2 years postoperation. However, because it involves an intracranial procedure with possible high postoperative morbidity, patients are generally more reluctant to accept it. Following surgery patients generally require vestibular rehabilitation to facilitate compensation.

CONCLUSIONS

The management of Menière's disease remains a challenge. The unknown aetiology means that treatment remains empirical, but our inadequate understanding should not detract from our efforts to provide symptomatic treatment for this disabling and distressing condition. Patients need to develop a good relationship with their doctors and to feel that they understand their condition. They should be accurately informed of the choices available and their limitations, and psychological support should always be made available. HM

Bottrill I (1998) Diagnosing and treating Menière's disease. *The Practitioner* **242:** 482–4

Brookes GB (1996) The pharmacological treatment of Menière's disease. *Clin Otolaryngol* **21:** 3–11

Committee on Hearing and Equilibrium (1985) Menière's disease: criteria for diagnosis and evaluation of therapy for reporting. *AAO-HNS Bulletin* **5** (July): 6–7

Hallpike CS, Cairns H (1938) Observations on the pathology of Menière's syndrome. *J Laryngol Otol* **53:** 625–54

Knapp H (1871) A clinical analysis of the inflammatory affections of the inner ear. *Arch Ophthalmol* **2:** 204–83

Menière P (1861) Observations de maladies de l'oreille interne caracaterisees par des symptomes de congestion cerebrale apoplectiforme. *Gaz Med de Paris* **167:** 379–81

Nedzelsski JM, Schessel DA, Bryce GE, Phleiderer AG (1992) Chemical labyrinthectomy: local application of gentamicin for the treatment of unilateral Menière's disease. *Am J Otol* **13:** 18–22

Silverstein H, Smouha E, Jones R (1989) Natural history *vs* surgery for Menière's disease. *Otolaryngol Head Neck Surg* **13:** 18–22

KEY POINTS

- Menière's disease or idiopathic endolymphatic hydrops is characterized by the classical features of vertigo, deafness, tinnitus and aural fullness but adjunctive symptoms may also be present, such as persistent dysequilibrium.
- Diagnosis is generally from a well taken history but important differential diagnoses must be excluded with a magnetic resonance imaging scan in every new patient who presents with a history suggestive of Menière's disease.
- Treatment involves careful counselling of the patient and explanation of the pathology and initial medical therapy with vestibular sedatives such as betahistine. In refractory cases potassium-sparing diuretics may be prescribed and advice on a low salt diet given.
- Surgical treatments are generally only used as a last resort and nowadays involve surgical decompression of the endolymphatic sac and intratympanic gentamicin injection.