Menière's disease: myths and realities

Many of the theories of the aetiology and pathogenesis of Menière's disease are often taught as being fact. This article explores how flimsy the evidence is for these theories along with the evidence of the effectiveness of the many treatments available.

Menière's disease is not a single disease but is a name for a collection of symptoms of which deafness, true rotatory vertigo and tinnitus in the affected ear are essential to make the diagnosis. This collection of symptoms are found in a range of diseases including tertiary syphilis, Marfan's syndrome, otosclerosis and vestibular schwannoma. It is a convention to call the disorder Menière's syndrome where the collection of symptoms are believed to be caused by another otological disease and Menière's disease where the aetiology is unknown.

Migraine

Menière's disease is not a migraine variant, although it is clear that about 40% of patients with Menière's disease do have migraine as well.

Incidence

A glance at referral letters to a balance clinic in an otology department would give the impression that Menière's disease was the commonest cause of true rotatory vertigo in the UK. This is probably not the case with many published series showing benign paroxysmal positional vertigo (BPPV) having a higher incidence (Barber, 1982). In the author's own department previous audit has shown an incidence of 9% of all referrals to the balance clinic. In Japan, however, Menière's disease appears to be more common or to have a similar incidence to BPPV.

Racial influence

The incidence of 46 per 100 000 population in Sweden (Stahle et al, 1978) places it as the country with the highest incidence. It has been hypothesized that the incidence mirrors the Viking diaspora. Some support is found for this with an incidence of 15.3 per 100 000 reported from Rochester USA (Wladislavosky-Waserman et al, 1984), Siena in Italy with an incidence of 2.75 per 100 000 (Celestino and Ralli, 1991), Cantabria with an incidence of 3.0 per 100 000 (Morales Angulo et al, 2003) and Finland with 4.3 per 100 000. One may ask why Finland, a country just next door to Sweden, has such a low incidence. This may be explained by looking at the languages of the two countries. Finnish and Swedish do not appear to share a common root so presumably the peoples also do not share a common root.

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The theory, however, is blown out of the water when the incidence in Japan (Shojaku and Watanabe, 1997) is noted. The Hida district has an incidence of 36.6, the Toyama district 17.0 and the Nishikubiki district of 21.4 per 100 000 population. With an incidence higher than the USA it seems extremely unlikely that the American occupation could account for such an incidence.

The condition is unusual in Africans, Afro-Caribbeans, and those from the Indian subcontinent.

In the author's own practice about 90% of the patients have blue, grey or green eyes which is higher than the local incidence of these eye colours combined.

Genetics

It was thought at one time that the mutated COCH gene was responsible for Menière's disease. The mutated COCH gene is responsible for familial autosomal dominant deafness with vestibular dysfunction.

Familial Menière's disease is heterogenetic with possible mutations on chromosome 6 and 14. It is autosomal dominant with 60% penetrance and anticipation.

Mental ill health

It has long been held that not only is there such an entity as the Menière's patient personality but that the incidence of mental illness is very high among this group of patients. The conditions commonly seen are agoraphobia, anxiety and depression (Yardley, 1994). Studies vary but overall the prevalence is probably not as high as it appears. The condition is a mild indolent condition in a large number of patients. This group of patients may only get a few attacks in their lifetime and are often managed solely by the GP. Therefore the incidence of mental illness is only assessed in those patients referred to hospital. It can also be difficult to decide when some symptoms cease to be a natural response to circumstances and become a mental illness. A good example is the anxiety experienced by the elderly when they have to go out at night, especially if they live on an inner city housing estate.

Nevertheless it is often the fear of having an attack that causes the greatest change in lifestyle rather than actually having attacks. It is much more common to hear that a patient won't go out for fear of embarrassing him-/herself in public rather than learning of an embarrassing attack that actually occurred in a public place.

Pathogenesis

It has long been believed that endolymphatic hydrops is uniformly found in all patients with Menière's disease. It

has been postulated that failure of the stria vascularis to act as an effective ionic pump leads to increasing pressure in the endolymphatic space. When the endolymphatic space ruptures then there is a massive electrical discharge in the vestibular nerves which the patient feels as an acute attack of the disease.

However, autopsy studies of petrous temporal bones have found that endolymphatic hydrops is present in many who have never had symptoms of Menière's disease. Hydrops is also found in many cases of acoustic neuroma, otosclerosis and tertiary syphilis.

The original theories put forward to explain how endolymphatic sac surgery worked evolved around the incorrect concept that the endolymphatic sac was exactly that. It was hypothesized that the sac could not expand at times when endolymphatic pressure was high as a result of fibrosis and the surrounding bone. It was felt that by removing the bone from around the sac or incising the sac the pressure could be relieved and attacks of vertigo in the Menière's patient avoided. Although there is evidence that the surgery has some beneficial effect, the method by which the operation has this effect remains elusive. Electron microscopy has shown that it is not a sac at all.

The sac certainly does have a role in the normal functioning of the ear. Chief cells within the sac have been shown to produce a hormone (Qvortrup et al, 1996) with a natriuretic effect, called saccin. It is possible that problems with the production of this hormone have a role to play in this disease.

Medical treatment

In the UK, betahistine is the first-line drug in the management of this disease. However, there has long been doubt about its efficacy. A double blind placebo-controlled trial at 11 Italian centres, with 52 patients completing the study, showed that betahistine is more effective than flunarizine in overall vertigo control in Menière's disease (Albera et al, 2003). A Cochrane review (James and Burton, 2001) of other trials concluded that there is no concrete evidence that the drug is effective.

It has been thought that the vasodilator effect of betahistine is the method of therapeutic action; however, it acts on the central vestibular system as a H1 agonist and a strong H3 antagonist, improving the process of vestibular compensation. This central action (Tighilet et al, 2002) is a strong contender for the actual mode of action.

Diuretics are usually a second-line management drug in the Menière's patient. It has been thought that they are effective in restoring hearing as well as having a positive effect on the tinnitus associated with Menière's disease. This is not the case – a double blind crossover study (van Deelen and Huizing, 1986) has shown that dyazide does have a positive effect on the attacks of vertigo but not on hearing or tinnitus.

There are virtually no published controlled trials on the use of diuretics in the management of Menière's disease. Osmotic diuretics such as urea, isosorbide (Nozawa et al, 1995) and glycerol improve hearing but can have detrimental effects on renal function when used in the long term. Evidence that other diuretics work is poor.

Gentamicin labyrinthectomy

Two meta-analyses of methods of administering intratympanic gentamicin have been published. The Toronto study (Cohen-Kerem et al, 2004) concluded that there was no difference in the risks no matter what technique was used and the Ontario (Chia et al, 2004) study found the titration method to be best. On balance it is probably wiser to give the gentamicin using a weekly regimen as there is evidence that the risk of severe sensorineural deafness occurring may be less.

Steroid therapy

Meta-analysis has shown that intratympanic steroids have no effect on Menière's disease. However, matters are not conclusive and more well-constructed studies are needed in this therapy.

Surgery

There are proponents of endolymphatic sac excision, stating that it is more effective than simple decompression. One controlled study with ten patients in each arm showed no difference in the efficacy of sac resection *vs* shunt surgery.

The elegant and much quoted study of Thomsen and Tos (Bretlau et al, 1984) showed that endolymphatic surgery and cortical mastoidectomy alone achieved the same result. However, this study did not show that endolymphatic surgery worked through a placebo effect as there is no evidence that a cortical mastoidectomy is a placebo operation. It is possible that the surgery had a natriuretic effect or that during the procedure the chief cells were 'stimulated to work more effectively'. In short the study raised more questions than it answered.

Over the years, there have been proponents for the different types of labyrinthectomy. Ultrasonic labyrinthectomy is no longer undertaken in the UK with surgeons in Bristol ceasing to use the technique in the mid-1980s. Cryo-labyrinthectomy was practiced in Middlesbrough until 1988 when Martin Horowitz (Horowitz et al, 1989) retired but disease control was poor with a rate of only 71%. Membranous labyrinthectomy (Cawthorne, 1960) appears to have a success rate in excess of 90%. There are many variations of this operation in which the osseous labyrinth is opened at various points and the membranes pulled out with a range of specially-designed hooks. After this is done various substances have been injected into the inner ear including alcohol, gentamicin and streptomycin. The highest success rate appears to be with the total three canal drill out of the osseous labyrinth (Yazdi and Rutka, 1996), with figures quoted in excess of 95% by some authors.

Vestibular neurectomy has also had its proponents with two approaches commonly used – retrosigmoid

(Silverstein and Jackson, 2002) and middle fossa (de Diego et al, 2001). The middle fossa approach has the advantage of sectioning the inferior and superior vestibular nerves in the lateral internal auditory meatus but the disadvantage of retraction to some extent on the temporal lobe. The posterior fossa approach requires splitting of the combined vestibulo-cochlear nerve and thus runs the risk of leaving vestibular fibres intact and loss of hearing.

However, all forms of labyrinthectomy can have long-term sequelae, including tinnitus, chronic imbalance and deafness. In choosing labyrinthectomy the patient may be swopping the acute episodes of vertigo interspersed with periods of normality for other chronic problems which never leave.

Psychological management

In recent years the importance of psychological management has been realized. Now that the morbidity associated with the fear of having a vertigo attack, especially in public, is recognized to be as great if not greater than the actual disease itself, new steps to help the patient are being developed. Such psychological techniques that have been tried need to be tested as rigorously as the drug and other therapies. Once there is evidence that psychological intervention works then there is a chance of persuading primary care trusts to fund this currently scarce modality of management.

Vestibular rehabilitation

In the patient with active Menière's disease vestibular rehabilitation has a very limited benefit as the status of the balance system is in a constant state of change with each attack. However, once a labyrinthectomy has taken place vestibular rehabilitation (Dowdal-Osborn, 2002) aids a faster return to normal daily living.

Quack remedies

By its very nature of frequent remissions, this disease is a boon to those who propound quack remedies. A high percentage of patients will improve while having any form of therapy. The vulnerable patient who is desperate for anything to improve his/her life must be educated in how not to fall victim to the unscrupulous or the ignorant well meaning. This can only come about through well-designed medical research studies, and active dissemination of the results of such work to all those that need to know. In this way we should have fewer myths and more knowledge of the reality. **BJHM**

KEY POINTS

- Menière's disease is not one pathological entity but is a symptom complex.
- Endolymphatic hydrops is linked with the pathogenesis but is also found in many without the symptoms of Menière's disease.
- Saccin, a naturetic hormone produced by the endolymphatic sac (which is not a sac), may have a role in the pathogenesis.
- There is some evidence that betahistine is effective in treating Menière's disease but the evidence is not conclusive.
- Attention to the patient's psychological health is likely to play dividends in successfully managing this disorder.
- There is a high incidence of psychological co-morbidity.

Conflict of interest: none.

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