Fortnightly review

Diagnosis and treatment of Ménière's disease

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In 1861 Prosper Ménière described a syndrome characterised by deafness, tinnitus, and episodic vertigo. Contrary to the thinking at that time, he correctly thought that the condition was a disorder of the inner ear.[1] In 1938 the principal underlying pathology, endolymphatic hydrops, was described by Hallpike and Cairns (fig 1),[2] but the precise aetiology of Ménière's disease still remains elusive. This fact, coupled with the capricious behaviour of the disease and the clinically important response to placebo treatment, has aroused controversy about its management and has plagued the scientific evaluation of the results of the various treatments used.

Summary points

- The aetiology of Ménière's disease is multifactorial but the relation between these factors and the development of the disease remains unknown
- Distension of the endolymphatic compartment of the inner ear may be due to otological or systemic disorders and is the constant pathological finding
- This condition can be diagnosed on clinical grounds and simple audiometry. Secondary endolymphatic hydrops and space occupying lesions of the cerebellopontine angle need to be excluded
- The relapsing-remitting nature of the disorder, response to placebo, and undetermined aetiology have plagued scientific attempts to evaluate treatments
- Around three quarters of patients respond to medical treatment or enter a state of prolonged remission. For the rest, surgery aims to control the disabling vertigo
- Bilateral Ménière's disease remains a therapeutic challenge

Methods

Over the years, a vast literature has accumulated on Ménière's disease. I have based this article on information gained from three sources.

The first was a Medline search from 1966 to 1997, which yielded nearly 2000 references using the text word "Ménière's" and 546 citations using "endolymphatic hydrops." Combining the two sets for 1992 to 1997 gave 405 references, and combining this set with the term "surgery" yielded 58 references. I examined the abstracts of these 463 citations. Recent articles on surgery for Ménière's disease predominantly report results and various minor modifications to established surgical techniques. Articles looking at the aetiology and pathogenesis of Ménière's disease and new forms of treatment are, however, of greater interest and I examined such papers more closely.

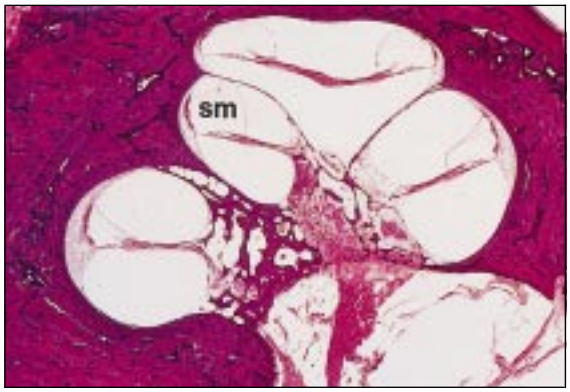

Fig 1 Histological section of the cochlea showing distension of the scala media (sm) due to endolymphatic hydrops. Note that the basal turn has been spared. Reproduced from Hawke and Jahn[3] with permission

The second source was a hand search of the main ear, nose, and throat journals in the English language in 1997 as there is a three to six month lag between the results of a Medline search and current issues of a given journal. The third source was references to textbooks and a review article written by my colleagues and me in 1994.[4]

Epidemiology

Ménière's disease predominantly affects white people, with a prevalence in the United Kingdom of 1 per 1000 of the population, both sexes being represented equally.[5] The disorder may occur in children[6] but has a peak onset between 20 and 50 years of age. The

incidence of disease affecting both ears increases to over 40% with longer follow up.[7]

Aetiology and pathogenesis

The precise aetiology of Ménière's disease remains obscure. Several theories explaining the development of endolymphatic hydrops have been put forward, although in reality the pathogenesis is probably multifactorial. Some or all of the aetiological factors listed below may be acting, but their precise relation to the sequence of events leading to the clinical picture remain unknown.

Anatomical—Ménière's disease is associated with several abnormalities of the temporal bone, including reduced pneumatisation of the mastoid and hypoplasia of the vestibular aqueduct.[8] The endolymphatic sac is small and lies in an abnormal position below the labyrinth.

Genetic—A familial predisposition to Ménière's disease has been recognised for over half a century. More recently, pedigree studies by Morrison yielded a family history in 7.7% with an autosomal dominant mode of inheritance, penetrance of around 60%, and obvious genetic anticipation.[5]

Immunological—The endolymphatic sac is osmotically and immunologically active.[9] Evidence of immune complex deposition in the endolymphatic sac[10] in patients with Ménière's disease has reinforced the belief that the disease is an immune disorder.

Viral—The role of neurotropic viruses is conflicting. Calenoff et al showed specific IgE to herpes simplex virus types I and II, Epstein-Barr virus, and cytomegalovirus in the serum of patients with Ménière's disease.[11] In contrast, Welling et al did not find more neurotropic viral DNA in patients with Ménière's disease than in a control group.[12]

Vascular—The association between migraine and the symptoms of Ménière's disease was recognised by Ménière himself.[1] This association has been substantiated in several studies and suggests a common vascular pathogenesis. Certainly, migraine may antedate the Ménière's symptoms by many years and occurs in as many as one in three patients.[13]

Metabolic—Endolymph is a potassium rich hyperosmolar fluid that is positively charged with respect to perilymph. Maintenance of this ionic milieu depends on the activity of sodium potassium ATPase in the stria vascularis of the cochlear duct. The endolymphatic sac is capable of secreting osmotically active molecules such as glycoproteins and glycosaminoglycans, which is probably how the sac regulates its fluid volume.[14] In Ménière's disease distension of the endolymphatic space either alters membrane permeability or causes rupture of Reissner's membrane, leading to potassium intoxication of the hair cells and vestibular neuroepithelium, with resultant deafness and vertigo. Repeated or protracted exposure of the hair cells to potassium results in chronic loss of hair cell motility, and the initial characteristic fluctuating deafness is replaced by progressive permanent deafness.[15]

Psychological—Several reports suggest a psychological basis for Ménière's disease. An increased prevalence of obsessional traits,[16] psychosomatic personalities, and neuroses has been noted in patients with Ménière's disease compared with control populations.[17]

Clinical features

To try to introduce uniformity into the diagnosis and treatment of Ménière's disease, the American Academy of Ophthalmology and Otolaryngology introduced guidelines in 1972, which were revised by the American Academy of Otolaryngology-Head and Neck Surgery in 1985 and 1995.[17 18] Based on control of vertigo, disability, and effects on hearing, these guidelines are now a widely accepted tool for evaluating treatment.

Clinically, three stages are generally recognised.

Stage I—In the early phase of the disease, the predominant symptom is vertigo. This is characteristically rotatory or rocking and is associated with nausea or vomiting. Signs of vagal disturbance, such as pallor and sweating, may occur, but loss of consciousness is not a feature. The episode is often preceded by an aura of fullness or pressure in the ear or side of the head and usually lasts from 20 minutes to several hours. Between the attacks hearing reverts to normal and examination of the patient during this period of remission invariably shows normal results.

Stage II—As the disease advances the hearing loss becomes established but continues to fluctuate. The deafness is sensorineural and initially affects the lower pitches (fig 2(*a*)). The paroxysms of vertigo reach their maximum severity and then tend to become less severe. The period of remission is highly variable, often lasting for several months.

Stage III—In the last stage of the disorder the hearing loss stops fluctuating and progressively worsens, both ears tending to be affected so that the prime disability is deafness (fig 2(*b*) and (*c*)). The episodes of vertigo diminish and then disappear, although the patient may be unsteady, especially in the dark.

Differential diagnosis and investigations

Classic Ménière's disease is an excellent example of a condition that can be diagnosed on clinical grounds and simple audiometric tests. Problems arise, however, because many dizzy patients have difficulty in describing their symptoms and doctors may find it difficult to construct a logical history. Overcoming this depends on detailing the symptoms and working through the differential diagnoses to reach the correct diagnosis. The box outlines the more common disorders that may give rise to vertigo.

Two investigations serve to aid clinicians when the clinical presentation is less than precise: the glycerol dehydration test and electrocochleography. The glycerol dehydration test measures the audiometric response to an oral dose of glycerol. Improvement in scores for hearing low frequency sounds and discriminating speech is diagnostic as there is no other condition apart from endolymphatic hydrops in which this change is observed. Similarly, electrocochleography gives a highly characteristic waveform in hydrops, though this test may give negative results in the early and late stages of the disease (fig 3).

Treatment

Currently, the treatment of Menière's disease is empirical. As yet, no treatment has prospectively modified the clinical course of the condition and thereby prevented

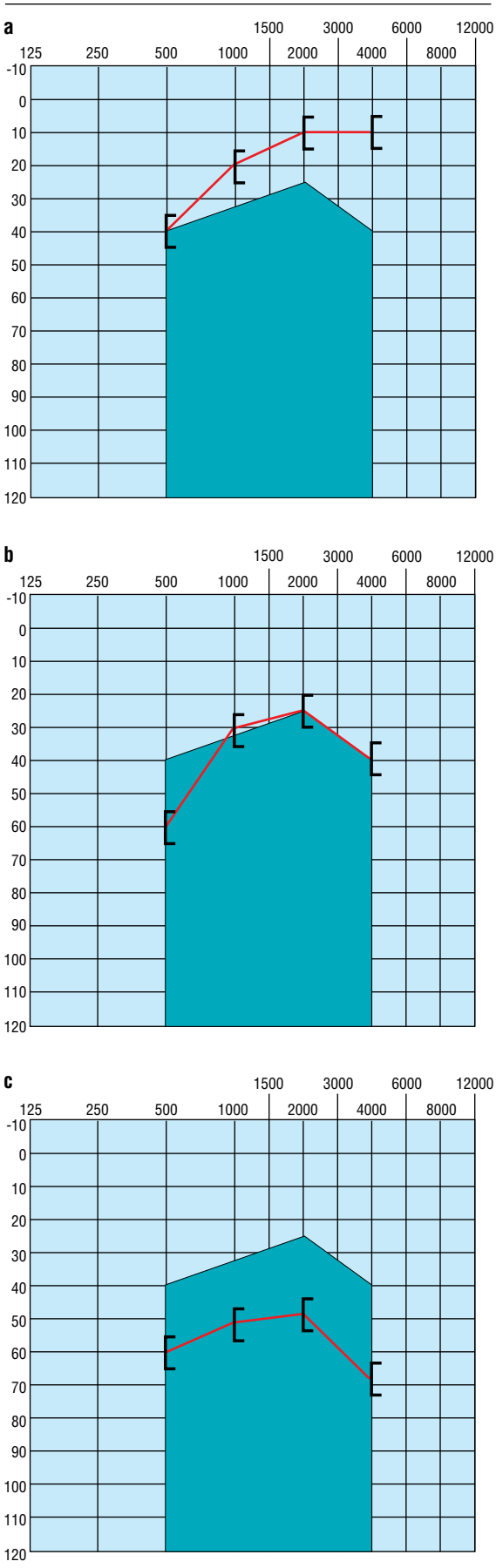

Fig 2 Masked bone conduction thresholds in right ear showing progressive sensorineural hearing loss with progression of Ménière's disease: (*a*) low frequency loss, (*b*) high frequency loss, (*c*) severe loss across whole frequency range

Differential diagnosis of vertigo

Rotatory vertigo
- Benign paroxysmal positional vertigo
- Vertebrobasilar insufficiency
- Ménière's disease
- Secondary or delayed endolymphatic hydrops
- Vestibular neuronitis
- Head injury
- Labyrinthine fistula
- Infective labyrinthitis
- Otological surgery
- Otosclerosis
- Cerebellopontine tumours or vascular lesions
- Disseminated sclerosis

Unsteadiness
- Side effects of drugs
- Lesions in central nervous system
- Active chronic suppurative otitis media
- Head injury
- Perilymph fistula
- Hyperventilation
- Functional (non-organic)

the progressive hearing loss. The reasons for this are fourfold. Firstly, the precise aetiology is unknown. Secondly, the placebo effect of drugs in this disorder is widely recognised. Thirdly, the disorder has a tendency to relapse and remit. Finally, the clinical course of the condition is such that over several years the vertigo eventually disappears in about 70% of patients.[19] Nevertheless, there are numerous conservative and surgical measures available to otolaryngologists that are aimed principally at abolishing the frightening and disabling vertigo with which patients present.

Conservative measures

Time spent giving a complete explanation of the disorder and an outline of the anticipated course of the disease is therapeutic in itself and invariably has a positive effect on the subsequent management. Clinically, three situations arise in which drug treatment may be useful.

Acute attacks—Drugs aimed at sedating the vestibulo-brain stem axis are particularly useful in aborting acute attacks. These include prochlorpera-

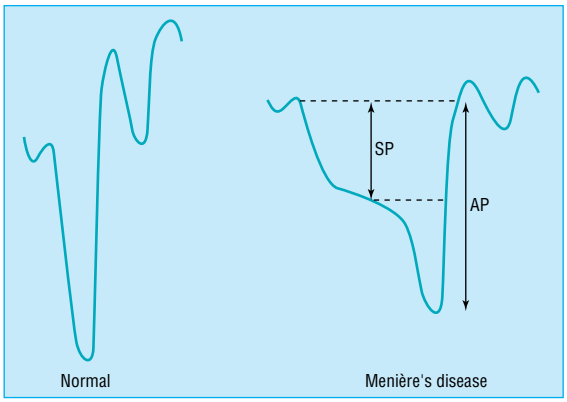

Fig 3 Transtympanic electrocochleogram. The compound action potential (AP) in Ménière's disease is characteristically widened as a result of a non-neural shift in direct current, the summating potential (SP)

zine, cinnarizine, promethazine, and diazepam. Longer term use of drugs such as prochlorperazine is inadvisable as the attendant sedation may be unacceptable, and the risk of extrapyramidal side effects from protracted use requires caution, particularly in elderly people.

Maintenance treatment—Dietary salt restriction and the use of diuretics such as frusemide, amiloride, and hydrochlorothiazide is an attempt to modify the endolymphatic hydrops itself. The basis for this is historical rather than scientific as the data from the few controlled studies that exist are conflicting and the placebo effect is clinically significant.[20 21] Vasodilators are used for prophylaxis on the basis that hydrops is due to ischaemia of the stria vascularis. The histamine analogue betahistine has been subject to some scientific scrutiny, and several controlled clinical studies have shown a significant improvement in vertigo, hearing loss, and tinnitus in the short term.[22] Currently, betahistine with or without a diuretic constitutes the favoured means of providing maintenance medical treatment. Drugs such as cinnarizine, propranolol (particularly if the patient has a history of migraine), and corticosteroids are also used empirically by some clinicians if the patient's symptoms are refractory to the above measures.

Ablative treatment—The toxic effects of aminoglycosides on the sensory neuroepithelium of the inner ear have long been recognised.[23] Medical ablation of the end organ with systemic streptomycin controls vertigo and has been useful in advanced bilateral Ménière's disease when poor but aidable hearing precludes surgical intervention. Cumulative doses of streptomycin, however, carry a risk of cochlear toxicity, and the incidence of ataxia and oscillopsia becomes unacceptable. The indications for this form of treatment are therefore now limited, particularly with the current interest in intratympanic delivery of gentamicin.[24] Several series show a rate of control of vertigo of around 90%, though a cochleotoxic effect is seen in 15-25% of cases. The future for intratympanic aminoglycosides in Ménière's disease is therefore promising.

Surgical treatment

Whether as the result of medical treatment or as a consequence of the clinical course of Ménière's disease, around 70% of patients have a sustained period of remission. This implies that a quarter of patients continue to have clinically important episodes of vertigo, and surgical treatment must be considered for them. The various surgical procedures advocated in Ménière's disease continue to arouse great controversy among otolaryngologists. The decision to operate and choice of procedure are often dictated by the individual surgeon's understanding and experience of a particular technique and of the threshold for surgical intervention. Broadly speaking, surgical procedures for Ménière's disease are classified as destructive or non-destructive with respect to hearing (box).

Surgical options in Ménière's disease

Not destructive to hearing
- Endolymphatic sac surgery
- Vestibular nerve section
- Sacculotomy
- Ultrasound treatment
- Cryosurgical treatment
- Insertion of tympanostomy tubes
- Cervical sympathectomy

Destructive to hearing
- Labyrinthectomy
- Cochleosacculotomy
- Vestibulocochlear neurectomy
- Translabyrinthine vestibular neurectomy

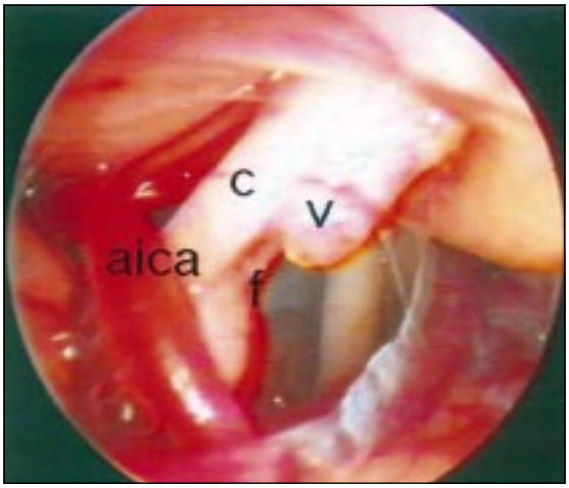

Fig 4 Endoscopic vestibular neurectomy. The sectioned vestibular nerve (v) and the cochlear nerve (c) can be seen emerging from the porus of the internal auditory meatus. The facial nerve (f) is visible anteriorly and in the distance is the trigeminal nerve. aica=anterior inferior cerebellar artery

Endolymphatic sac surgery was first described in 1927 by Portmann,[25] and no other aspect of Ménière's disease has aroused more debate or controversy. As the precise role of the sac in hydrops is still not known, the precise mechanism by which the surgery works remains elusive. Nevertheless, saccus surgery is widely performed. In a recent analysis of 100 consecutive endolymphatic mastoid shunt operations, Moffat reported complete or substantial control of vertigo in 81% of patients, with clinically important improvement in hearing in 19%, using the 1985 guidelines of the American Academy of Otolaryngology-Head and Neck Surgery.[26] Such results are consistent with findings from various centres,[17] and endolymphatic sac surgery remains the most commonly used conservative operation for Ménière's disease when hearing is still serviceable.

Vestibular nerve section—In sectioning the vestibular nerve, no attempt is made to modify the underlying pathophysiology. The objective is to dissociate the offending labyrinth from the brain stem while preserving the patient's hearing. The procedure is uniformly effective, with control of vertigo in 90-95% of patients according to the series.[17] The operation, however, is a considerable undertaking and carries the attendant risks of any neurosurgical procedure in the posterior cranial fossa (fig 4).[27]

Labyrinthectomy—Extirpation of the labyrinth is indicated in patients with symptoms who have poor or non-serviceable hearing. Violating the inner ear in this manner invariably leads to total permanent deafness. However, the opposite ear may be subclinically hydropic,[28] so doctors are naturally concerned that

should the disease progress in the other ear the patient may become totally deaf. The argument that in Ménière's disease, any hearing, however poor, should be preserved is exemplified by the widespread use of the non-destructive procedures.

Cochlear implantation—Over the past decade the auditory rehabilitation of certain profoundly deaf people has been transformed by cochlear implantation. Patients with severe bilateral Ménière's disease and non-serviceable hearing find their deafness is a greater handicap than any remaining vertigo, and end stage bilateral Ménière's disease is a recognised indication for cochlear implantation.

Selection of surgical procedure

Surgeons with patients with symptoms whose disease is refractory to medical treatment have several surgical options. Broadly, three management strategies have evolved. Proponents of endolymphatic sac surgery view this as the first surgical step, reserving either revision sac surgery or a vestibular neurectomy for patients who continue to have vertigo. Those who view sac surgery as a non-specific or placebo operation and are versed in neurotological practice will consider a vestibular neurectomy as the first line surgical procedure. The third group of surgeons, who also see little merit in sac surgery but have no experience of neurotology, are faced with the option of either undertaking a labyrinthectomy or referring the patient to a neurotologist for a vestibular neurectomy. Perhaps the use of intratympanic aminoglycosides will be a viable intermediate line of management.

Conclusion

Over 135 years after it was first described, every facet of Ménière's disease continues to evoke controversy. The precise aetiology and pathogenesis remain elusive, the diagnosis remains inaccurate, and the behaviour of the disease remains capricious. Not surprisingly, much of the treatment is empirical and provokes much debate among otolaryngologists. Although the condition cannot currently be cured in the true sense, most patients can be managed satisfactorily and an inadequate understanding of the underlying mechanisms should not detract from the prime objective—to provide symptomatic relief for the patients who have this difficult and disabling disorder.

I thank Professor Richard Ramsden for his invaluable advice during the preparation of this paper. Figure 4 is reproduced with permission of Professor Jacques Magnan (Service d' Oto-Rhino-Laryngologie, Hôpital Nord, Marseilles).

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One hundred years ago

Medical duty with the army

The Director-General of the Army Medical Staff has addressed a letter to the governing bodies of the medical schools, stating that the Secretary of State for War has approved of the employment of a certain number of young civil surgeons for duty with Her Majesty's troops in the United Kingdom, and asking for nominations. The terms of appointment are that the engagement will be for six months, subject to good behaviour, that the remuneration will be at an inclusive rate of £270 per annum, that the gentlemen selected may be detailed for duty in any part of the United Kingdom, and that their services shall be wholly devoted to such duties in connection with the medical charge of troops as may be required of them. The reasons assigned for this step are the large demand made on the strength of the Army Medical Staff at home, due to the additional troops employed in Egypt and South and West Africa, and the impossibility of obtaining the entire services of ordinary general practitioners. The need to make such applications indicates the serious strain which has been placed upon the Medical Department. (*BMJ* 1898;i:907)