Chapter 19: Ménière's disease

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In 1861, Prosper Ménière published a description of the clinical entity that was soon designated 'maladie de Ménière'. The account he gave was so complete and accurate that virtually no addition could improve the picture. He described a condition characterized by sudden and recurring episodes of vertigo with nausea and vomiting, together with hearing loss and tinnitus. He stressed that the abnormality was one of the internal ear and was not related to cerebral apoplexy as had previously been thought. Confusion has arisen following Ménière's reference in his famous paper of 1861, to the post-mortem findings of a serosanguineous exudate in the semicircular canals of a young woman who died 5 days after the onset of continuous vertigo, vomiting and deafness. When Ménière first recorded this case as an addendum to the chapter on nerve deafness in his translation of Kramer's textbook of otology in 1848, he did not mention the vertigo and described the exudate as filling the labyrinth. In fact, Ménière quoted this case some 13 years after the event, merely to illustrate that the symptoms could arise from a labyrinthine lesion and his intention was *not* to identify such a haemorrhagic lesion as the cause of the non-fatal syndrome. Lack of appreciation of Ménière's objective in presenting this evidence, together with misquotation, has led to the long-lasting misbelief that he had ascribed haemorrhage into the labyrinth as the cause of the disease which bears his name.

The spelling of the eponym should be that used by Ménière himself in his publications, although curiously his family gave a second (acute) accent to his name, on his tomb in Paris.

The term Ménière's 'syndrome' could imply that the condition is merely a collection of symptoms without a pathological basis. For this reason, the term 'disease' is generally preferred since the clinical and histological features are sufficiently constant to indicate the presence of a definite abnormality. The expression 'endolymphatic hydrops' is often used and is acceptable because of the outstanding histopathological feature of the disease. Endolymphatic hydrops, however, may also occur in certain congenital malformations of the ear, in syphilitic labyrinthitis, and especially in viral labyrinthitis.

Incidence

Ménière's disease is not a particularly uncommon disorder although the criteria for diagnosis employed by different clinicians vary tremendously and so, accordingly, does its apparent frequency.

Matsunaga (1976) have an average incidence of 0.5% for the disease in patients attending ear, nose and throat clinics in various hospitals in different countries. Drachman and Hart (1972) and Wilmot (1974) both quoted an incidence of 5% of Ménière's disease in a clinic dealing with balance disorders, a figure which is similar to that of the present author.

Age of onset

The experience of most otologists is that, in the majority of patients, symptoms generally start before the age of 50 years. *Table 19.1* brings together the age of onset from several authors.

Table 19.1. Age of onset of Ménière's disease in 1054 patients

Age (years)	0-10	11-20	21-30	31-40	41-50	51-60	61-70	71-80	over 80
No of patients	14	61	204	286	246	163	64	16	nil

Bilateral involvement

The incidence of bilateral involvement is usually put at about 10%, but it appears to depend upon the length of time patients are kept under observation and on the criteria used in diagnosing involvement of the second ear. The author's experience is that second side involvement, when it occurs, is usually seen early, and within 2 years in 50% of cases. Many older patients do show hearing changes in the contralateral ear, although not necessarily associated with dysequilibrium referable to that ear. Nevertheless, Haye and Quist-Hanssen (1976) quoted 37% of their 111 patients as having bilateral involvement.

Predisposing factors

No definite predisposing factors are known. Portmann (1980) taking a 60-year retrospective look, observed that 'Ménière's disease is one of the least understood disorders in general and by specialists in particular...'

Although Ménière's disease remains a disorder of unknown origin, it is increasingly regarded as the late result of an insult to the labyrinth which may have passed unnoticed years before the onset of symptoms. Shambaugh and Wiet (1980) made the suggestion that the early damage may be viral in origin. Schuknecht and Gulya (1983) and Schuknecht (1984) in an overview of endolymphatic hydrops generally, and of Ménière's disease in particular, regarded endolymphatic hydrops as a pathological condition which is the final manifestation of a variety of otological insults. In their papers, they presented a classification on the basis of clinical and pathological data which distinguishes symptomatic and asymptomatic forms of hydrops. In symptomatic hydrops, the initiating factor may be congenital, inflammatory, traumatic, or idiopathic. It disrupts those mechanisms responsible for maintaining the proper volume of endolymph so that a progressive situation is reached which becomes symptomatic. Asymptomatic hydrops is clinically silent and found only upon routine histopathological examination of temporal bones, but again may be congenital or acquired, or idiopathic in origin. Schuknecht therefore suggested that Ménière's disease may be redefined as an idiopathic, symptomatic form and as such may be considered to be one member of a family of disorders linked by the common pathology of endolymphatic hydrops.

Clinical features

In order to improve the accuracy of the diagnosis and to establish uniform standards, the criteria to be used should be those laid down by the special committee of the American

Academy of Ophthalmology and Otolaryngology (AAOO) chaired by Alford (1972) (*see* Appendix 19.1). Likewise, especially when comparing results of treatment, it seems sensible to use a system of staging as proposed by Arenberg and Stahle (1979, 1980), Balkany, Sires and Arenberg (1980) and Watanabe (1980). Depending upon the behaviour of the hearing, three phases of the disease can be recognized characterized by early reversible hearing loss, established fluctuant hearing loss, and late non-fluctuant hearing loss.

Various positive and negative criteria should be applied to ensure accurate diagnosis of Ménière's disease as emphasized by Wilmot (1974). Repeated episodes of vestibular failure associated with cochlear and with vestibular symptoms constitute the acute phases of the disease. Between acute attacks, periods of remission of varying duration occur and during these, hearing may return to normal in the earlier stages of the disease. The cycle of activity and remission is characteristically irregular and capricious in Ménière's disease and makes the assessment of treatment difficult.

In the majority of patients, the remission period will be between 3 and 12 months. Remission is seldom less, but occasionally can be very prolonged and may be even of several years' duration. Hearing loss and dysequilibrium most commonly begin together but in about one-third of patients the dysequilibrium occurs first and only after months or sometimes a year or two may the syndrome be complete. Fluctuant hearing is less frequently the first symptom.

The acute attack

The dramatic symptom of vertigo dominates the picture in the acute phase. Occasionally the patient is so distressed that the auditory symptoms may go unnoticed, but usually the hearing loss and tinnitus become more obvious to the patient as the vertigo subsides. Unless the attack is minor, it is accompanied by nausea and often vomiting and other symptoms of vagal disturbance, such as sweating and pallor with slowing of the pulse. This may lead to slight transient faintness, but it is emphasized that loss of consciousness is *not* a feature of Ménière's disease.

Quite often the patient will get warning of an attack by discomfort or aching in the ear, an alteration in the hearing or tinnitus. In other patients, there is just a vague sense of uneasiness. The acute phase may consist of a single attack, although there is often a tendency to cluster formation before a more prolonged remission occurs.

The vertigo occurs in different forms and may be sometimes relatively mild or occasionally extremely severe. It may be rotational in nature, the patient may have a feeling of unsteadiness which causes him to veer to one or other side, sometimes there may be a to and fro, or up-and-down motion 'as on a ship at the mercy of a stormy sea'. On rare occasions the patient is thrown to the ground without warning in what is known as 'a drop attack' or 'utricular crisis'.

The paroxysms can come at any time, or may even waken the patient. The duration of the attack may be from 20 minutes to not more than 24 hours. The patient is fully oriented and conscious throughout the attack and there are no neurological symptoms or sequelae, except those referable to the labyrinth.

Nystagmus is always present during an attack and may change direction during an episode; this disappears as the vertigo subsides.

Symptoms in the remission phase

In the early stages the patient will be entirely asymptomatic in the remission phase and will have normal hearing and no tinnitus. As the disease progresses some tinnitus and hearing loss become permanently established.

The vestibular apparatus like the cochlea is also capable of varying degrees of recovery after an acute attack, but nevertheless, with the passage of time some permanent damage is done so that a proportion of patients, even in remission may get brief imbalance, for example on sudden head movement, or they may develop a tendency to motion sickness, positional vertigo, or momentary ataxia when changing direction.

In some patients there appears to be an association with typical migraine attacks with which the Ménière's attacks may 'change places'.

Clinical variations

As already mentioned, there is occasionally an interval of several years before all symptoms are present. It is to be noted that the diagnosis of Ménière's disease is not to be made until all symptoms have developed. Cochlear hydrops without vertigo is an undoubted entity. It should be suspected in the patient who has a low frequency sensorineural hearing loss that is fluctuant and in whom other aetiological factors have been excluded. Electrocochleography may be confirmatory. It may be wise to inform the family doctor that such a patient may be a future candidate for Ménière's disease.

Vestibular hydrops without hearing loss probably occurs, but it is more difficult to identify and its pathological basis has not yet been determined. Occasionally, accurate and repeated audiological examination near to an episode of vertigo may demonstrate a hearing change of which the patient was unaware, although more often, the condition can only be diagnosed in retrospect after the complete symptoms of Ménière's disease have eventually shown themselves.

The Lermoyez syndrome is a rare variant in which hearing loss and tinnitus develop over a period of hours; vertigo then occurs, often quite suddenly and with it the hearing and tinnitus improves.

Ménière's disease in combination with other ear diseases

Chronic middle ear suppuration may coexist and there may be a diagnosis of erosion of the labyrinthine capsule when in fact no such complication has occurred. Otosclerosis is a disorder in which dysequilibrium sometimes occurs and may lead to some confusion. On occasion otosclerosis and Ménière's disease may coexist. Chronic suppuration, otosclerosis, and Ménière's disease, are not rare disorders in otological practice and will therefore sometimes be seen occurring together.

Investigation

General assessment

An accurate history which includes a detailed description of each relevant symptom can often lead to a correct provisional diagnosis. A search for vertigo, hearing loss and tinnitus, even in the distant past, should be made as these may have been forgotten if recovery has apparently taken place. a past history of exposure to ototoxic drugs, head injury, or ear disease and operations may be relevant to the differential diagnosis.

Obviously the patient's general state must be assessed in detail, the cardiovascular system must be healthy, the carotid pulses equal and normal and the blood pressure within normal limits when taken on both arms. The central nervous system should be healthy and neuro-otological examination negative apart from the findings related to the eighth nerve. Neck movements should be unimpaired. In the course of assessment, the otologist will have taken note of the patient's psychological adjustment to the symptoms. As may be expected from the nature of the condition a significant number of patients develop an overlying anxiety state, but this must not be allowed to mask the organic nature of their basic disorder. Preliminary otological examination is undertaken to exclude any evidence of past or present disease in the middle ear cleft. The fistula test should always be applied, a positive Hennebert sign will be found in approximately 50% of patients according to Schuknecht (1975) who attributed this to the formation of adhesions between the distended saccule and the inner surface of the stapes footplate.

Audiological investigation

Preliminary tuning fork tests will confirm the sensorineural nature of the hearing loss, although a false negative Rinne may be present in patients with severe unilateral deafness. The tuning fork tests may also demonstrate diplacusis. Complete audiometric investigation follows and is an essential step towards diagnosis. The tests involved have been reviewed by Schuknecht (1963) and by Hedgecock (1968).

The various audiometric tests available are described in Volume 2 (Chapters 7 and 8). Characteristically, the results will be those of an end organ lesion, but unusual results are sometimes obtained which may give a mixed pattern of responses. Pure tone audiometry is of fundamental importance and should be repeated at each attendance to try to demonstrate the fluctuant nature of the hearing loss. Three main patterns of pure tone audiogram may be found: a hearing reduction maximal in the low or high tones, or a flat loss. The rising type of curve is generally found in the earlier stage of the disorder, while in older patients a pre-existing high tone loss (as may be shown in the unaffected ear) can be superimposed. As the disease progresses, the curve is seen to flatten out and the hearing loss, although it may halt its progress at any point, becomes increasingly severe.

Further evaluation will include tests for loudness recruitment, loudness discomfort level, stapedius reflex threshold, and possibly tone decay and Békésy testing. Speech audiometry is important and in patients with Ménière's disease, the speech reception threshold very closely matches the pure-tone threshold in over 90% of patients. Discrimination is impaired, although much less severely than in patients with a neural lesion.

Electrocochleography now constitutes a very important part in the diagnosis of endolymphatic hydrops and details of this are given in Volume 2.

Dehydration tests based on the use of glycerol, urea and frusemide (furosemide) have a place in the diagnosis of Ménière's disease. Glycerol was initially used by Klockhoff and Lindblom (1967), and experimental work on guinea-pigs by Angelborg and Agerup (1975) has confirmed that the diuresis after glycerol administration produces a lowering of internal ear pressures. In the presence of an endolymphatic hydrops, a temporary hearing improvement occurs. This may be useful not only in the diagnosis of Ménière's disease, but also in the assessment for the suitability of diuretic treatment and in the selection of patients for operations on the endolymphatic sac. The test, however, is not entirely dependable. Van de Water and Arenberg (1983) compared glycerol and urea and found that they were equally reliable, but that patients preferred urea because of its fewer side-effects. Imoto and Stahle (1983) have also made the same comparison, but concluded that the degree of response depended upon the severity of the hydrops and that an absent response indicated either remission or permanent neural hypofunction.

Vestibular investigation

Reference should be made to Volume 2 (Chapter 9). The method used for caloric testing is frequently that described by Fitzgerald and Hallpike (1942) and may show either a simple canal paresis or sometimes a directional preponderance, but a normal response is by no means uncommon and does not rule out the presence of Ménière's disease. A comprehensive account of the clinical, caloric and rotational test findings has been given by Wilmot (1974). A detailed analysis of electronystagmography in the diagnosis of Ménière's disease is provided by Stahle (1976a) who found that 59% of his patients had a reduced caloric response, 40% had a normal response, and 1% an exaggerated response.

Galvanic stimulation, as described by Swaak and Oosterveld (1976) provides an important means of differentiating an end organ from a neural type of vestibular lesion, although the test has not found wide acceptance in the UK.

X-ray examination

There has been some debate concerning the relationship of pneumatization to the development of Ménière's disease. Oku, Hasegawa and Watanabe (1980) concluded that there was no difference in the pneumatization of the temporal bone radiologically in Ménière's disease, various types of sensorineural deafness and the normal individual. Conversely, Stahle and Wilbrand (1983) concluded that gross anatomical changes were present in respect of decreased pneumatization, not only in the mastoid generally, but particularly around the periductile region, while Clemis and Valvassori (1968) and Valvassori (1983) found abnormalities of the vestibular aqueduct itself in patients with Ménière's disease. Dreisbach, Seibert and Arenberg (1983) also examined the question of patency and visibility of the vestibular aqueduct in Ménière's disease, but concluded that there was considerable anatomical variation, and that demonstration accordingly was often difficult. Considering the size of the duct it is perhaps hardly surprising that even with the most modern scanning techniques it is not always possible to demonstrate the structure. It has been suggested that operations on the

saccus are futile if the duct cannot be demonstrated radiologically but most otologists remain sceptical and regard the case as not-proven.

Differential diagnosis

A vast number of clinical conditions enter into the differential diagnosis of vertigo and it has to be admitted that many patients remain without an exact diagnosis. The typical case of Ménière's disease can usually be diagnosed quite readily; in other cases the diagnosis will depend on how far the otologist is willing to stretch his criteria and this clearly is of great significance in those patients who may be candidates for surgical treatment.

Other disorders which affect the labyrinth include cholesteatoma and fistula formation, dysequilibrium after head injury, viral labyrinthitis, drug toxicity, positional vertigo and perilymph fistula. Syphilis produces an endolymphatic hydrops with symptoms that simulate Ménière's disease. In Cogan's disease, the symptoms are those of Ménière's disease to which are added redness of the eyes and blurring of the vision with interstitial keratitis. Disorders of the vestibular nerve and its central connections always have to be considered and include vestibular schwannoma (neuroma) in its various atypical forms, vestibular neuronitis and multiple sclerosis. Cardiovascular disturbances, giddiness of ischaemic or neck origin when coexisting with a sensorineural hearing loss can be confusing (although less so if the sensorineural loss is bilateral and symmetrical). Craniocervical dysplasia can produce fluctuant sensorineural hearing loss with tinnitus and dysequilibrium and may mimic Ménière's disease. However, the dysequilibrium has different characteristics, as described by Ellies and Plester (1980). When in doubt, it is sometimes helpful to go back to the basic points and to remind oneself that the condition is usually unilateral, and likewise that the symptoms are unilateral, and consist of fluctuant hearing loss, fluctuant tinnitus and episodes of dysequilibrium separated by characteristic remissions, and that other neurological, cardiovascular, neck problems etc, are absent or separate.

Pathology

Although is was long speculated that endolymphatic hydrops was associated with Ménière's disease, this was only confirmed by Hallpike and Cairns in 1938. Many others have since reported similar findings. The most comprehensive description available at present is that of Schuknecht (1974, 1975) based on material in his own collection of temporal bones. The most obvious abnormality is the distension of the endolymphatic spaces. This mainly affects the cochlea and the saccule. The utricle is involved to a lesser degree and the semicircular canals only slightly (in the region of the ampullae). Dilatation of the endolymphatic sac has not been found. The distension of the scala media of the cochlea is seen as a bulging of Reissner's membrane into the scala vestibuli, often leading to obliteration of this part of the perilymphatic compartment. Reissner's membrane is frequently observed to bulge through the helicotrema into the apical part of the scala tympani.

The second feature in Ménière's disease concerns herniation or rupture of the membranous labyrinth. These herniations can occur at any site in Reissner's membrane or in the saccule, utricle or ampullae. That rupture can occur implies that healing can also take place. Accordingly, it has been suggested that these ruptures are vital phenomena, and are related in some way to the exacerbations and remissions which characterize Ménière's disease.

The incidence of these ruptures is variable. Antoli-Candela (1976) observed them to be present in 13 out of 19 temporal bones that were studied, but Fraysee, Alonso and House (1980) observed them in only three out of 23 bones they examined.

A third feature is collapse of the membranous labyrinth which presumably occurs when a break therein fails to heal. It may be that a permanent break of this kind is the situation which occurs in 'burnt-out' Ménière's disease.

The fourth point of interest concerning the membranous labyrinth, is the proliferation of fibrous tissue in the vestibule. Adhesions occur between the footplate and the walls of the utricle and saccule. They are though to account for the presence of Hennebert's sign in some patients with Ménière's disease.

The fifth feature, concerns the hair-cell and ganglion-cell population of the ear affected by Ménière's disease. Schuknecht, Benitez and Beekhuis (1962) were able to achieve early fixation of the labyrinthine structures in three temporal bones from patients with Ménière's disease and found normal hair-cell populations in the organ of Corti, the maculae of the saccule and the cristae of the three semicircular canals. The neuron population of the spiral and vestibular ganglia were also normal. Schuknecht (1968) confirmed these findings in a further paper and concluded that degenerative changes only occur in exceptional cases. Such exceptions have been presented by Lindsay, Cohurt and Sciarra (1967) and also by two cases now in the Schuknecht collection. In these cases the loss of hair cells and of ganglion cells is confined to the apical region of the cochlea.

Antoli-Candela (1976) in a further study of material in the Schuknecht collection provided a very detailed account of the appearance on light microscopy and confirmed the normality of the hair-cell population in the organ of Corti. Changes in the hair cells and ganglion cells were the same in both the affected and unaffected ear. He could find no correlation generally between the severity of the sensorineural hearing loss and the histopathological findings in the sensory and neural structures. Fraysee, Alonso and House (1980) also found that the hair-cell population was normal, although considered that there might be some loss of ganglion cells in specimens showing very severe hydrops.

Ylikoski, Collan and Palva (1979) reported on material obtained at operation from the cochlear nerve, vestibular nerve and neuroepithelial areas of the utricle and semicircular canal cristae and found no microscopic evidence of deterioration, even in long-standing advanced disease, only minor variations from normal were found in individual cells. Whatever the nature of the degenerative changes in the receptors, it is evident that they are not gross enough to be shown by light microscopy in the material which has become available for study, and that any structural changes are ultramicroscopic in character.

Electron microscopy has been carried out on material obtained at operation and from experimental material in animals by a number of researchers including Kimura and Schuknecht (1970) and by Calman, Friedmann and Wright (1975) who examined the vestibular nerve and other parts, from patients undergoing vestibular neurectomy, but it is often difficult to know whether the features described are those of Ménière's disease or are the result of ageing and other processes.

Endolymphatic distension

To appreciate the theories of causation of endolymphatic hydrops, an understanding is necessary of the normal anatomy and physiology of the inner ear and its fluids (*see* Volume 1, Chapters 1 and 2).

Briefly, it may be said that distension of the endolymphatic spaces, as seen in Ménière's disease, could in theory arise in several ways:

(1) insufficient production of perilymph, if it is accepted that perilymph production is an active process

(2) excess production of endolymph, assuming it is produced by the stria vascularis and other lesser sites

(3) by inadequate absorption in the endolymphatic sac, if it is accepted that endolymph moves towards the sac and that the latter has an absorptive function.

The various theories of the cause of endolymphatic hydrops have been reviewed by Lawrence (1968) although he emphasized that he does not necessarily accept the view that over-accumulation of endolymph *per se* is the cause of clinical symptoms. As he pointed out, the hydrops may be a concomitant and incidental occurrence, along with the other features of the disease, all arising from some basic underlying disturbance such as alteration in ionic concentrations or osmotic pressure relationships. Most otologists, however, accept the hydrops as *the* basic feature, even though there is difficulty in explaining its cause and in correlating it with the symptomatology.

Generally, the theories of causation of endolymphatic hydrops can be grouped as follows:

(1) Those based on disturbance of fluid formation. These are mainly dependent on the principle of radial flow of inner ear fluids, described by Naftalin and Harrison (1958).

(2) Those concerned with mechanical blockage and disturbed reabsorption. These are mainly dependent on the theory of longitudinal flow of endolymph as described by Guild (1927), that is with the movement of endolymph towards the endolymphatic sac, or with malfunction of the sac itself.

It should not be thought that these two theories are incompatible or conflicting and, indeed, it is probable that both types of flow occur in the normal ear, a fast radial flow and a slow longitudinal flow.

In the theory of radial flow of endolymph, Naftalin and Harrison (1958) suggested that secretion and absorption occur in the same radial area of the cochlea. Their theory deals with ionic exchanges in the inner ear, and they suggested that fluid in the inner ear passes from perilymph and that the function of the stria is to absorb perilymph. They postulated that the function of Reissner's membrane is to retain potassium ions in the endolymph and to prevent protein from entering endolymph by being impermeable to large molecules. The stria, like the

tubular cell of the kidney, extracts sodium and inserts potassium to maintain a high endolymphatic concentration of the latter. They emphasize that according to the theory, ionic transport against gradients is carried out by the stria vascularis, the only structure capable of doing so. Potassium and sodium exchange across Reissner's membrane is in the direction of concentration gradients and therefore requires no energy. At the present time (1986) it is increasingly accepted that the basic problem in Ménière's disease is one affecting Reissner's membrane (Wersäll, 1986 personal communication).

Naftalin and Harrison further stated that perilymph is formed by ultrafiltration from vascular tissues of the perilymph space but in Ménière's disease there is decreased production and this leads to an apparent increase in endolymph. This is later followed by a true increase of endolymph caused by a gradual increase of potassium as flow through Reissner's membrane diminishes.

The work of Johnstone (1975) should also be noted. He put forward sound evidence which supports the possibility of two fairly independent radial circulations, one mainly in the cochlea and saccule, the other confined to the utricle and canals. Others have also put forward evidence in favour of radial flow (combined with longitudinal flow) and suggested that radial flow provides for the energy metabolism and ion exchange and is the only satisfactory way in which the high energy demands of the organ of Corti can be met. Lawrence (1980) accepted chemical exchange as occurring all along the endolymphatic space, this occurring together with longitudinal flow to the endolymphatic sac.

The work of Kishimoto et al (1983) is of interest. Their experiments showed that the endolymphatic sac and duct can transmit physiological variations of cerebrospinal fluid pressure to the inner ear fluids and submit that this function may be significant in maintaining normal perilymphatic and endolymphatic pressures.

Rauch (1968), like Naftalin and Harrison earlier, saw the primary defect as being in the production of perilymph, but endolymphatic hydrops could equally be from overproduction of endolymph (in the stria vascularis supplemented by the planum semilunaris and dark vestibular cells) or from obstruction to longitudinal flow or malabsorption in the endolymph sac.

Experimental work by Kimura (1967, 1968) indicated that obliteration of the duct and sac can produce a gross endolymphatic hydrops in certain animals, but it must be emphasized that other procedures in experimental animals, even those merely confined to the middle ear, can also produce hydrops. Such experimental hydrops is not usually associated with dysequilibrium. The symptoms of Ménière's disease have yet to be produced in experimental animals. However, the theory that the endolymphatic sac may have a resorptive function finds further support in the work of Lundquist, Kimura and Wersall (1964), Ishii, Silverstein and Balogh (1966), and Adlington (1967).

Radiological evidence, as already mentioned, has also been submitted by Clemis and Valvassori (Clemis and Valvassori, 1968; Valvassori, 1983) to suggest that obstruction to longitudinal flow towards the endolymphatic sac may have a part in the production of Ménière's disease. Support has also been provided by Stahle and Wilbrand (1974). This work, however, has not received general acceptance, and as Dreisbach, Seibert and Arenberg 919830

concluded, the anatomy of the vestibular aqueduct is so variable that even with the best scanning techniques non-visualization readily occurs as a result of the inability to cut in exactly the required plane. It also has to be said that histological studies have failed to demonstrate definite abnormality of the endolymphatic duct in patients with Ménière's disease.

Accordingly, the view that operations designed to drain the endolymphatic sac are futile unless the duct can be shown radiologically is invalid.

Correlation of pathology with symptoms

Attempts have been made to explain the symptoms partly on a mechanical and partly on a biochemical basis. It has also been suggested that the early and variable symptoms may be caused by mechanical aspects while the later irreversible symptoms are the result of permanent biochemical factors.

The assumption that early and reversible low frequency hearing loss could be explained on a mechanical basis by the greater distortion of the broad part of the basilar membrane has been supported by the use of cochlear models constructed by Tonndorf (1957). He expanded his original theory as a result of further work (1968) and concluded that several of the auditory changes in Ménière's disease could be explained on the basis of altered responses characteristics of the cochlea.

A mechanical explanation for the sudden onset of vertigo is difficult, but Lindsay (1960) noted the frequency with which out-pouchings of the membranous labyrinth could be found near the canal ampullae and suggested that they could interfere mechanically with the cristae contained therein.

In respect of biochemical factors, it will be recalled that according to classical membrane theory, it would be physiologically impossible for action potentials to be generated in high potassium fluid surroundings, such as endolymph. It was demonstrated by Rauch (1960) that the cortilymph in the spaces of Nuel and the tunnel of Corti, has extracellular properties and closely resembles perilymph. It is poor in potassium and thus provides an appropriate medium for normal neural excitation and transmission. Leakage of potassium into the perilymphatic compartment with consequent contamination of the cortilymph through the canaliculae perforantes might be expected to produce symptoms of cochlear failure. Such contamination could occur from breakage or leakage of the histologically-proven herniations which are frequently present. Experimental alteration of potassium levels by perfusion of the cochlea in animals supports this theory. Dohlmann (1965) described how vestibular function can be affected in a similar biochemical fashion.

It must be emphasized, however, that the significance of the membranous herniations continues to be one of debate. One view is that rupture is responsible for the onset of symptoms and another, to the effect that rupture is responsible for the remissions that occur in Ménière's disease. It is also difficult to explain how multiple herniations can develop; if one area of weakness has been produced it would be expected that the same area would rupture on future occasions rather than a new herniation developing. It is also difficult to accept that potassium contamination occurring, for example, through a rupture of Reissner's membrane can produce almost simultaneous onset of both cochlear and of vestibular

symptoms, such as occurs in the majority of patients in an acute attack. It is equally awkward to image simultaneous ruptures occurring separately in the cochlear and in the vestibular parts of the labyrinth.

For further discussion of this very interesting aspect of Ménière's disease the reader should refer to the previous edition of this volume, and also to Chapter 5.

Treatment

Because our knowledge of the basic pathology is so inadequate, it is difficult to design any satisfactory medical or surgical treatment for Ménière's disease. Schuknecht (1976) summed up the situation regarding medical treatment in the following words: 'I think if we sit as a jury of honest judges looking at the results, I doubt that we could approve one single drug in the treatment of Ménière's disease'. The situation seems to have improved very little since Furstenburg, Lashmet and Lathrop (1934) commented on 'The bewildered and futile state of medical therapy in the Ménière's syndrome complex'. Indeed, one might reasonably ask oneself whether medical treatment has improved to any significant extent since Ménière first described the disorder!

The variety of surgical procedures is also bewildering. Any success claimed for different operations, like medical treatment, must always be weighed against the widely varying periods of natural remission. The only surgical exception being total destruction or denervation. Indeed, there is much evidence that suggests that most medical treatment and certain surgical procedures are effective only through a placebo effect. Fortunately, it is becoming a little easier now to compare results since more authors are using a system of staging and analysing their results on the AAOO system previously mentioned.

General management

The relief of tension and the anxiety state resulting from the unpredictable nature of the disease is of prime importance and an understanding and sympathetic approach to the patient's problems is equally essential. Strong reassurance stressing the non-fatal nature of the disorder is necessary, coupled with emphasis that it is entirely a disorder of the inner ear. Some explanation of its nature, for example 'an excess of fluid in the balance organ' is of some help. It must be explained that the condition can be ameliorated, even if it cannot be completely cured. It is also wise to tell patients that relapses can occur and, if need be, management may be altered from time to time, otherwise the patient will inevitably suffer loss of confidence sooner or later.

Pulec (1972) emphasized the need to treat any abnormality which can be discovered in thyroid function, pituitary/adrenal insufficiency, glucose tolerance and similar problems. He claimed to find a high proportion of patients in whom some basic disturbance is discovered, but this has not been confirmed by other writers, including Schmidt, Brunsting and Antvelink (1979), although syphilis, of course, must be excluded routinely.

Medication

Much medical treatment is on an empirical basis. There is little statistical evidence that medication alters the natural history of the disease or confers any specific benefit. Nevertheless, most otologists provide some kind of medication and such treatment should always be given a substantial trial before recommending surgical management. There is some evidence that regular supervision, special testing and the involvement in the machinery of a big hospital has in itself a significant placebo effect. If the patient is seen regularly it also gives the opportunity of offering suitable surgical treatment at the appropriate time if deterioration occurs.

Dietetic treatment based on low salt and fluid intake was introduced over 50 years ago and is still employed. Boles et al (1975) reported satisfactory results and few operations in a series of 500 consecutive patients treated along these lines, although there is little evidence that labyrinthine or serum electrolyte concentration can be altered by ordinary dietetic measures.

Vasodilators are frequently used, although this seems a curious way to diminish endolymph production by the stria vascularis. The experimental evidence of Snow and Suga (1975) has confirmed that cochlear blood flow was greatly increased by carbon dioxide, amyl nitrite, and betahistine, but no increase occurred after nicotinic acid administration.

Various phenothiazine drugs with antihistamine properties have been reported to be useful; cinnarizine and diphenidol, are frequently used.

Other drugs used in treatment include lithium carbonate, which was introduced in the hope that it might favourably alter the transport of fluid and ions across the membranes of the internal ear, although Thomsen et al (1976) demonstrated no more than a placebo effect. Lemon bioflavonoid derivatives and vitamin therapy seem to have a few supporters. Sedatives and tranquilizers sometimes appear helpful, especially in those patients who are aware that attacks tend to occur during periods of stress. Preparations such as prochlorperazine and thiethylperazine are useful to suppress nausea and vomiting, as well as the unsteadiness which sometimes follows an acute exacerbation.

Treatment of the acute attack

Some patients appear to control minor episodes or even prevent a major attack from coming on by retiring to a quiet corner and telling their well-wishers to stop fussing around. Minor episodes can sometimes be managed by sitting down and taking various drugs, such as promethazine, dimenhydrinate, perphenazine, or chlorpromazine but, in a severe and prolonged attack, the patient must retire to bed and be given one or other of these drugs intramuscularly in fairly high dosage.

Gejrot (1976) believed that an acute attack can also be relieved by the intravenous infusion of lignocaine in a dosage of 1 mg/kg body weight to be given at the rate of 6 mg/minutes.

Stellate ganglion block is a simple procedure which should be within the capability of an otologist and can likewise give rapid relief from a severe episode. Stellate block repeated two or three times a week can also promote a remission in those patients who are going through a bad cluster of major attacks.

Ototoxic therapy

Streptomycin therapy has been described by Fowler (1948) and by Schuknecht (1957) whose same long-term patients and more recent ones were reviewed by Singleton and Schuknecht (1968). This therapy takes advantage of the vestibulotoxic effect of the drug and may have a place in patients with bilateral disease (*see* Chapter 5). The treatment, however, is not entirely without some risk of cochlear damage, and bilateral suppression of vestibular function itself constitutes a substantial disability which requires intensive physiotherapy for adaptation. Streptomycin was injected locally into the middle ear of unilateral cases by Lange (1972). Other vestibulotoxic drugs have been similarly employed either by local injection or insertion through a ventilation tube into the middle ear.

Treatment with hearing aids

Because of the impaired discrimination and limited tolerance of amplification that is present in most patients with Ménière's disease, the benefit from a hearing aid is often limited. This matter has been discussed by Johnson and House (1979). Most patients with unilateral disease do not find an aid beneficial. There are many exceptions to this rule, and occasionally the benefit to a patient even with bilateral disease and very severe loss of hearing can be substantial.

Surgical treatment

Surgical treatment must be considered for those patients in whom disabling symptoms are continuing to occur without evidence of adequate remission. It is customary to provide medical treatment for about 6 months before making any decision, but this will depend upon individual circumstances. If a patient's condition is deteriorating rather than improving, and he is unable to follow his normal activities, then surgical treatment may be indicated. In the past, the severity of the vertigo has been the main criterion used in reaching a decision. However, with increasing realization that even the best medical treatment does little to conserve hearing, combined with the knowledge that the progressive nature of the disorder will almost inevitably lead to increasing hearing damage, there is now a greater tendency to operate earlier in an attempt to conserve hearing (albeit in the knowledge that surgical treatment is also uncertain and unproven from this point of view).

Most surgical procedures can be considered under the following main headings.

(1) Procedures designed to influence endolymph production (for example sympathectomy).

(2) Procedures designed to influence endolymph absorption (for example operations on the endolymphatic sac).

(3) Selective denervation of the vestibular labyrinth (vestibular nerve resection).

(4) Labyrinth destruction

        (a) selective destruction of the vestibular labyrinth (by ultrasound or cryosurgery)

        (b) total destruction of the labyrinth (labyrinthectomy) combined perhaps with total denervation (translabyrinthine cochleovestibular neurectomy).

The exact procedure will mainly depend on the level of the hearing present and whether the disease is unilateral or bilateral. All of the operations except the last in the list, aim to preserve hearing. Operations in the last group offer no possibility of retention of hearing after operation and accordingly are only suitable if it is felt that the residual hearing in the affected ear can safely be sacrificed and that the other ear is healthy.

Conservation operations

Cervical sympathectomy

This has been suggested on the basis that in some way, as yet undetermined, it helps to correct the microcirculatory fault in the stria vascularis. Golding-Wood (1973) reported satisfactory results from the operation and gave long-term follow-up in 247 patients. An important point to remember about sympathectomy is that it is an operation which is remove from the ear and therefore totally free from any risk to the hearing. Accordingly, it may be especially suitable for those patients having Ménière's disease in an only remaining ear. The resection must be from C3 to T3 inclusive. However, it has to be said that sympathectomy has not been widely used in the treatment of Ménière's disease.

Operations on the endolymphatic sac

There has been much interest in these operations since the sac was first opened by Georges Portmann (1927). The particular patient was still living at the age of 75 never having experienced a return of vertigo (Michel Portmann, personal communication).

Although operations on the endolymphatic sac have an important place in the surgical management of Ménière's disease it has been suggested that the results are no different from those one might expect from the natural history of the disease. This view finds some support in the controlled double-blind study reported by Thomsen et al (1981) and by Bretlau et al (1984). Some of their patients had a simple mastoidectomy carried out, in others the sac was opened. Their statistical analyses revealed no significant difference between the two groups of patients, and they concluded that the effect of surgery was purely as a placebo. However, a separate statistical analysis on the same patients made by Pillsbury et al (1983) produced the opposite conclusions.

Experience at the Radcliffe Infirmary, Oxford, shows a very substantial difference in results between patients in whom the surgeon had been unable to identify the sac with a high degree of certainty, or in whom the operation consisted of merely 'decompressing' the posterior fossa dura without a search being made for the sac, compared with a group of patients in whom the sac had been identified with a very high degree of certainty and a

Silastic strip inserted into its lumen. The remission rate was much better and more prolonged in the latter group.

The author's experience has been that the insertion of a saccus-subarachnoid shunt gave no advantage over simply inserting a Silastic drain into the sac. There is, however, renewed interest in the use of valved subarachnoid shunts. Superior results have been reported by Arenberg (1980) and by Brachmann and Anderson (1980); but it is yet another aspect of Ménière's disease which remains controversial.

The use of the glycerol test and urea test has been discussed previously in respect to diagnosis. It is also suggested by Wiet (1983) and by other authors that the test may be useful in a selection of patients for saccus surgery and for judging the prognosis (*see also* Chapter 17). Arenberg (1980) stated that a good hearing improvement after saccus surgery may be expected in the presence of a strong response to the glycerol test (although a negative test is not a contraindication for attempting relief of vertigo by the operation). Reference has already been made concerning radiological studies of the endolymphatic duct in a selection of patients for saccus surgery. Shea, Emmett and Moore (1979) indicated that superior results could be obtained if the vestibular aqueduct was readily demonstrated, but Wiet (1983), probably representing the majority view, came to the conclusion that radiological studies were interesting for demonstrating the anatomy and the extent of pneumatization, but that there was no relationship between the results of operation and of the ability or inability to demonstrate the aqueduct.

The surgical anatomy of the endolymphatic sac has been described in detail in a beautifully illustrated paper by Arenberg et al (1977) to which reference should be made. Further detailed information for the surgeon concerning the anatomy in 20 normal ears is provided by Shea, Emmett and Moore (1979).

Although operations on the endolymphatic sac are relatively straightforward and without any great risk of complications, it is important to remember that severe sensorineural hearing loss can occasionally occur. Fisch (1976) has quoted a risk of 4% of partial sensorineural hearing loss. This complication is often said to be the result of an accidental injury of the posterior semicircular canal, but the author's experience is that the complication can occur in the undoubted absence of such an injury.

The results of saccus surgery reported by various surgeons employing different techniques have been reported on many occasions and reference should be made to the current literature. Palva, Karja and Palva (1976) found their results were somewhat less satisfactory than in most published series but, nevertheless, were in accordance with the experience of many others. Many otologists with interest and experience in this field will find themselves in sympathy with Palva's observations.

Endolymph-perilymph shunts

Various ways have been suggested by which the endolymphatic space might be decompressed with consequent relief of vertigo and preservation of hearing. Decompression of the saccule was first described by Fick (1964), but the experience of nearly all otologists has been that the operation carries a dangerously high risk of profound sensorineural hearing

loss. Accordingly, the operation must be regarded as destructive, but it may have a place in the management of the older patient, who could be expected to have difficulty in adaptation after a formal labyrinthectomy. Cody (1968) inserted a stainless steel tack which was left permanently in position in the stapes footplate so that the distending saccule could be automatically and repetitively decompressed by contact against it. In a long-term follow-up (Cody, 1974) he reported on 140 such patients, but in spite of his encouraging results, the operation has not become generally accepted because of the high risk to hearing.

House (1968a) used a cryosurgical probe onto the promontory in order to try to create a small fistula while Pulec (1968) inserted a small tube through the basilar membrane using a *round* window approach. A further refinement is that of Schuknecht (1982), who described the creation of a cochlear endolymphatic shunt. His results were analysed using the AAOO criteria (Schuknecht, 1986, personal communication). With an average follow-up of 22 months, the severe episodes of vertigo were relieved in 72% of patients, 45% of patients had some worsening of hearing at the time of their most recent audiogram, but only 12% suffered a profound sensorineural hearing loss. The author's personal experience of the operation has been much better in respect of vertigo relief, but much worse in respect of hearing conservation.

Vestibular neurectomy

Division of the vestibular nerve for persistent aural vertigo was achieved by Frazier (1912), McKenzie (1932) and by Cairns and Brain (1933). Dandy (1941) was able to report a series of 401 operations with only one death, utilizing a posterior fossa approach. The middle fossa microsurgical technique of House (1968b) is now well known and has been further modified and described by Fisch (1970, 1976). The operation demands a high degree of surgical skill and it potentially carries the various complications and risks of morbidity that are associated with intracranial procedures in the middle fossa.

There has now been considerable experience with the middle fossa procedure and, in experienced hands, it has shown itself to be highly predictable in the relief of vertigo and preservation of hearing with minimal complications. However, the operation is *not* one for the occasional surgeon. The results obtained by Fisch (1976) and by Palva, Ylikoski and Paavolainen (1979) are probably representative. They quoted 90% of patients relieved of their vertigo, and 80% with preservation of hearing; 3% of their patients had temporary partial facial weakness. Vestibular neurectomy is followed by a powerful bilateral suppression of vestibular activity which was investigated by Fisch (1973) and which makes for a more rapid compensation compared to that after labyrinthectomy. This is probably a result of the divisions of the efferent fibres to the labyrinth.

It has been suggested that the natural history of the disease may be altered by this operation with consequent control of any further hearing deterioration. This too, has been attributed to the division of the efferent fibres to the labyrinth. The author's experience, however, is that with prolonged follow-up the hearing gradually continues to deteriorate, probably because the disease is still continuing, albeit in a fairly asymptomatic fashion.

At the present time, attention is again being given to the posterior fossa approach. Indeed, Bryan and Bucy as long ago as 1973 reported on 17 such patients, although with

substantially less satisfactory results than those obtained with the middle fossa approach. The method used can be either neurosurgical and retrosinus or alternatively transmastoid retrolabyrinthine. House et al (1983) using the latter approach, obtained complete relief of vertigo for 83% of their patients.

Selective destruction of the labyrinth

Ultrasonic energy was first employed to destroy the human labyrinth by Krejci (1951) and was soon followed by Arslan (1953) and by Angell-James (1973) in England. Experimentally the effectiveness of ultrasound in the destruction of the vestibular end organs in animals was demonstrated by Brain et al (1960).

The technique employed is essentially that described by Angell-James (1969a, b) who reported his results from 232 patients several years after first employing the method. Stahle (1976b) reported his experience with 356 patients over a 12-year period using fairly similar methods. Barnett and Kossoff (1977) described a special applicator for use at the round window.

Although ultrasound treatment still has its advocates and can undoubtedly obtain satisfactory results, most otologists have abandoned its use. The reason for this has probably been similar to those of the author, namely the difficulties in obtaining and maintaining a generator and applicator whose output of energy can be depended upon. It has also been his experience that in the very long term the hearing continues to deteriorate even though the patient may remain free of vertigo.

The author's view is that ultrasound treatment, like the saccus operation, is best regarded as a time-borrowing procedure; the amount of time gained can be very variable, but a substantial number of patients remain free of vertigo for 10 years or more.

Destruction operations

Labyrinthectomy

If it is decided that the residual hearing in the ear is permanently at a very low level, that dysequilibrium continues to be disabling, and that the opposite ear is undoubtedly asymptomatic, then the labyrinth can be destroyed by various methods:

(1) the postaural lateral semicircular canal approach

(2) an extension of this with complete exenteration of all three canals and of the vestibule (and perhaps with neurectomy)

(3) a permeatal transtympanic window approach (which can also include a neurectomy).

All methods of labyrinthectomy provide a high likelihood of relieving the major attacks of vertigo, but it has to be emphasized to the patient that this is at the expense of total hearing loss in the operated ear and consideration must always be given to the possibility of

disease affecting the only surviving ear. Labyrinthectomy, therefore is not a decision to be taken lightly, especially in the younger patient who has many years ahead during which second side involvement may occur. Likewise, careful consideration has to be given in the case of the older patient because the older the patient the greater the difficulty with adaptation; for this group, there is much to be said for performing either a Fick procedure or a cochleotomy in the first instance (*see above*). Sacculotomy and cochleotomy are very much less disturbing to the patient and if they fail a formal labyrinthectomy can still be considered.

Appropriate exercises help to establish full adaptation. Results are generally excellent and the loss of distorted hearing in the ear is a benefit. Labyrinthectomy through a lateral semicircular canal approach is probably the simplest operation available. It is highly reliable, although not completely so. Sometimes the destruction is incomplete and there is later a return of symptoms. It is for this reason that complete exenteration of all three semicircular canals and of the vestibule, is to be preferred. Translabyrinthine neurectomy, as described by Pulcer (1968) enables the otologist to check that no abnormality of the internal meatus has been missed, provides the patient with more complete and more rapid adaptation, and if the cochlear nerve is resected some patients obtain relief from their tinnitus. However, extension of the operation in this way makes it a more major procedure and calls for considerable skill and the possible advantages have to be balanced against the extra danger to the facial nerve, as well as to the occasional possibility of a cerebrospinal fluid leak and consequent meningitis. The permeatal transtympanic window labyrinthectomy was described by Schuknecht in 1956. The stapes is extracted, and the round window is opened. It is absolutely essential that, using a suitable pick, every attempt is made to extract each individual area of sensory epithelium. If this is not done, there is a possibility of return of some function and of symptoms. Extension of the operation has been described by Silverstein (1976) to include a cochleovestibular neurectomy by drilling away the promontory for increased access. It may be considered, however, that the access is somewhat awkward, the hazard to the facial nerve too great, and that if one wishes to undertake a neurectomy a translabyrinthine route gives better exposure and greater safety.

The question of incomplete compensation (especially in older patients) has been discussed by Palva, Karja and Palva (1976). They found that a high proportion of older patients, although relieved of their major episodes of vertigo had a fairly constant, but ill-defined mild unsteadiness. The patients of Pedersen and Sørensen (1970) were perhaps less well motivated. Half of their 32 patients, who they were able to trace with an average follow-up of 7 years, still complained of persistent imbalance that was sufficiently severe to incapacitate them and prevent their return to full-time work. Fortunately, most patients achieve much better results than this.

Other procedures

For completeness the following two modalities of treatment require a mention.

(1) Insertion of a ventilation tube through the tympanic membrane has been advocated on the assumption that, in Ménière's disease, the internal ear disturbance is a manifestation of abnormal middle ear pressures. Scientific support for this assumption is somewhat lacking, but nevertheless dramatic improvements in symptoms have been reported following this minor

operation. It is difficult to see any scientific basis for it and it seems that the effect is purely that of a placebo.

(2) Treatment in the hypobaric chamber of patients with acute attacks of symptoms has been described by Tjernström et al (1980), but is of interest from the scientific aspect rather than from the treatment point of view.

Choice of operation

Operation is to be considered when there is failure to control symptoms by so-called medical treatment, or if there is absence of prolonged spontaneous remission. It is always an individual decision and depends mainly on the degree to which attacks of vertigo are interfering with the patient's normal life. If the symptoms are disabling then operation should be offered. Usually, the severity of the vertigo has been the main factor in reaching a decision. In recent years, and in spite of the unpredictability in hearing conservation operations, there has been a tendency to operate earlier in the disease in the hope of controlling progressive hearing loss. However, the relief of tinnitus and improvement of hearing by surgery although sometimes obtained, cannot be relied upon. It seems that the best results in this last respect, occur probably in younger patients and in early cases where the degree of permanent cochlear damage is not too great.

The type of operation is generally determined by the level of hearing in the affected ear. If hearing in the affected ear is useless and the other ear is healthy then a destructive operation can be carried out. The surgeon will generally be less willing to undertake a destructive procedure in a younger patient who has many years ahead of him in which he may develop bilateral disease. If hearing in the affected ear is still at a useful level, if the patient has only one hearing ear, if the patient has bilateral disease, or if he shows any evidence whatever that might suggest early signs of trouble in the apparently healthy ear, then any operation to be offered must clearly be of the hearing conservation type.

Generally speaking, the 'hearing-destructive' operations are straightforward and give a very high probability of relief from the acute attacks. Although they may leave some imbalance they are probably the best method available to restore full working capability to the patient. The hearing-destructive operations include the various types of labyrinthectomy and translabyrinthine neurectomy, and must also include the Fick and Cody operations on the saccule.

The 'hearing-conservation' operations are generally more complex procedures. They have a high probability of doing no damage to hearing, but offer less certain relief of the vertigo compared to destructive operations (except for middle fossa vestibular neurectomy). These operations include the various procedures on the endolymphatic sac, as well as ultrasound and cryosurgical methods, and sympathectomy. Middle fossa vestibular neurectomy is also to be included in this category, as may posterior fossa neurectomy and perhaps the insertion of a ventilation (pressure-equalizing) tube.

Labyrinthectomy, in some form, will be the operation of choice when disease is clearly unilateral and the hearing is so diminished and distorted as to be useless, but it is again emphasized that apparently minor transient symptoms in the opposite ear, are sometimes the

first indication of major involvement which may only become manifest after a prolonged interval and, particularly, in the younger patient are to be regarded with apprehension. Labyrinthectomy, by either a permeatal or mastoid approach, is highly effective in the unilateral case and in younger, well-motivated patients is followed by rapid vestibular compensation. Furthermore, the patient is often relieved of hearing which is so distorted as to interfere with reception on the normal side. The advantage of proceeding to a translabyrinthine resection of the vestibular nerve and may be the cochlear nerve also must be weighed against its potential further difficulties and complications and should perhaps only be undertaken by a surgeon who is familiar with the internal auditory meatus.

The Fick and Cody transfootplate operations on the saccule must be regarded as destructive procedures; even their main protagonists have failed to demonstrate that hearing can be stabilized or improved. The majority of surgeons who have performed these operations have reported an extremely high incidence of total hearing loss in the ear. Nevertheless, they can be useful operations for the elderly, fragile, or ill patients who can only tolerate a minor operation, performed if necessary under local anaesthesia.

The author's experience with cochleotomy, as already mentioned, places it in this group of operative procedures, it carries a high risk of severe sensorineural hearing loss, but is extremely undisturbing and makes for easier adaptation for the older patient in whom a conventional labyrinthectomy would be the only alternative.

Of the operations designed for hearing conservation there has been increasing interest in operations involving the saccus endolymphaticus, but there is no satisfactory evidence to indicate that any one type of operation is superior. The best results seem to be obtained in the earlier (and therefore potentially reversible) stages of Ménière's disease in which hearing is normal or near-normal during remission. A positive test with glycerol or frusemide may be helpful in making a decision. The radiological demonstration the patency of the vestibular aqueduct is of doubtful value.

A saccus procedure can, nevertheless, sometimes give a good result in the later stages of the disease in spite of a negative glycerol test. Ultrasound treatment offers a satisfactory alternative in those departments where the necessary equipment and skills are available and the same may apply to cryosurgery, although there have been far fewer reports relating to the results. Sympathectomy, as mentioned, is an operation away from the ear and therefore is the only procedure available which carries no risk to hearing. It should be considered especially in those patients with either bilateral disease or only one functioning ear. Its exact mode of action is likely to remain uncertain until the efferent innervation to the ear is clarified.

Selective resection of the eighth nerve through a middle fossa approach, although a major procedure, has now established itself as highly predictable for the relief of vertigo. It is associated with an acceptably low risk of damage to hearing in skilled and experienced hands. Its long-term effect upon the natural history of the disease remains to be proven. The precise indications for the operation are still debated. Fisch (1976) regards it as the surgical treatment of choice in those patients with irreversible Ménière's disease, that is those who have a hearing loss which is stable or a hearing loss which fluctuates but is never normal. Palva, Karja and Palva (1976) use the operation if hearing is at a level of 70 dB or better on pure-tone testing with a discrimination score of at least 50%. They also regard it as the

treatment of choice if there is a possibility of bilateral disease. It is a major operation, which should perhaps be regarded not as a primary procedure, but one which should be utilized when an earlier operation such as the saccus operation, has failed in a particular patient. Like translabyrinthine neurectomy it is an operation which calls for an advanced degree of surgical skill.

The choice of operations for Ménière's disease remains somewhat bewildering and to some extent the choice will depend upon the experience and personal preference of the otologist concerned. Useful reviews have been provided by Snow and Kimmelman (1979) and by Kinney (1980). It is to be hoped that the criteria set down in the AAOO system will help to clarify the situation. Until these or some similar criteria for assessing results are more widely employed, it will continue to be impossible to compare directly the effects of one treatment with those of another and the management of Ménière's disease will remain somewhat haphazard. Until better understanding of the pathogenesis of Ménière's disease becomes available and provides some basis for improved medical treatment or preventative measures, future progress seems to lie along the lines of early conservation surgery before irreversible damage occurs to the inner ear.

Definitions

The *deafness* is sensorineural in type, fluctuating, usually unilateral and progressive. The deafness may recover in large measure between episodes early in the disease but each episode tends to cause some additional permanent impairment.

The *vertigo* occurs in well-defined episodes. The *definitive spell* is often prostrating, frequently accompanied by nausea and sometimes vomiting, and persists for a prolonged period of time (20 minutes to no more than 24 hours). The patient is fully oriented and conscious throughout the spell and there are no neurologic accompaniments or sequelae to the spell except those referable to the end-organ. Vestibular nystagmus is always present (of the end-organ variety: fine, rapid, quick-slow, in a single definite direction, and horizontal or horizontal-rotatory). Between definitive spells there may be various kinds of *adjunctive spells,* such as motion intolerance, positional vertigo, falling attacks, and momentary ataxia on cornering, but the diagnosis is not tenable unless definitive spells are present with good health between them. During and briefly before a definitive spell hearing in the affected ear may decrease and tinnitus increase, remaining so for a variable time after the spell. It is accepted that many patients notice no subjective change in hearing during a spell, and it may rarely occur that hearing increases after a spell.

The *tinnitus* is quite variable and always subjective. It generally varies directly with the magnitude of the deafness.

Subvarieties of Ménière's disease

There are two subvarieties:

Cochlear Ménière's disease, or Ménière's disease without vertigo, is characterized solely by a fluctuating and progressive sensorineural deafness;

Vestibular Ménière's disease, or Ménière's disease without deafness is characterized solely by the definitive spells of vertigo.

Reporting results of treatment

The following criteria are offered reporting success or failure of therapy.

Vertigo

Control means absence of definitive spells for ten times the average interval between spells before treatment. In this group, a subgroup may be defined in which both definitive and adjunctive dizzy spells have been absent for ten times the average interval between definitive spells before treatment. If unindicated, it is assumed that some or all patients continue with adjunctive spells. A statistical analysis of the data reported is desirable.

Deafness

(1) *Hearing improved and serviceable* means a sustained pure tone threshold or SRT of 30 dB or better and a discrimination score of 80% better where one (or both) was not so before

(2) *Hearing improved but nonserviceable* means either

(a) a sustained increase in the speech frequencies of an average of 15 dB or more (but an SRT of greater than 30 dB) together with a discrimination score of at least 80% or an improvement of 15%, or

(b) a discrimination score improvement of 20% or better

(3) *Hearing worse* means a 15 dB or greater loss in the average of the speech frequencies or a 15% or greater decrease in a discrimination score that was 80% or less

(4) *Hearing unchanged* means less than 15 dB change in the average of the speech frequencies, and less than a 15% change in discrimination score.

Reporting according to overall level of result

There is some value in uniformity of reporting according to overall result. It would seem to be useful if all authors and their readers could immediately know, by a simple letter designation for example, that the patients under discussion were relieved of all spells as well as the definitive or major spells, or that the patient under discussion received both improvement in hearing and relief of spells, etc. The following classified levels are suggested.

Class A

(1) Absence of definitive spells for described period (in addition, absence of adjunctive spells as well could be noted)

(2) Hearing improved (in addition, hearing improved as well as serviceable could be noted).

Class B

(1) Absence of definitive spells for described period
(2) Hearing unchanged.

Class C

(1) Absence of definitive spells for described period
(2) Hearing worse.

Class D

Failure of control of definitive spells.