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Historical Article

Menière's disease: evolution of a definition

N. J. P. Beasley, F.R.C.S.*, N. S. Jones, F.R.C.S.†

Abstract
In 1861 Prosper Menière separated patients with episodic vertigo, hearing loss and tinnitus from a group previously described as having apoplectiform cerebral congestion. He suggested the cause was disease within the semicircular canals (Menière, 1861). Over the years it became apparent that within this group there were a number of patients with characteristic signs and symptoms and in 1938 a pathological correlate was found in the form of endolymphatic hydrops. Descriptions such as Menière's 'disease', Menière's 'syndrome' and Menière's 'symptom complex' led to a confusing array of terms for this condition and monitoring of treatment results became difficult. In response to this in 1972 the American Academy of Ophthalmology and Otolaryngology Committee on Hearing and Equilibrium published a clear definition of Menière's disease and criteria for the reporting of treatment results, it was updated in 1985 and again in 1995. We describe the changes that have taken place as the definition of Menière's disease has evolved.

Key words: History of Medicine, 19th century, 20th century; Menière's disease

Historical background

Episodes of paroxysmal vertigo have been recorded since Roman times. Plutarch a Roman historian reported in his work *Lives* that Julius Caesar suffered from a 'falling sickness' characterised in part by 'giddiness'. This was referred to by Shakespeare who in addition made Caesar hard of hearing in the left ear (Cawthorne, 1958).

Other more recent historical figures also suffered from severe disequilibrium. Martin Luther once wrote to a man who complained of the itch, 'I would give ten florins to change with you; you know not how distressing vertigo is. At this very moment I am unable to read a letter through at once, and indeed I cannot read more than two or three lines of my Psalter; for when I make the attempt such a buzzing comes on my ears that I am often on the point of falling off my seat. On the other hand the itch is a useful thing.' (Cawthorne, 1947). He suspected that Satan was the cause of his symptoms and is reported to have been so distressed that on one occasion he threw his inkwell at the devil who he said was violently attacking his ear (Dandy, 1941).

Dean Swift, the 18th century satirist, was another figure tormented by dizziness. He ended a letter to the Pope in 1725 'This wildness you must allow because I am so giddy and deaf'. He became a recluse and was eventually committed to a madhouse (Cawthorne, 1947). Controversy surrounds a possible diagnosis of Menière's disease in Vincent van Gogh. His 'madness' was characterised on occasions by disabling attacks of 'le vertige', nausea and vomiting, however, it is more likely that he suffered from epilepsy or psychosis (Arenberg, 1990).

In the 18th century paroxysmal vertigo although well recognised was not well defined and its origin was thought to be central. The term 'apoplectiform cerebral congestion' covered a rather vague group of disorders where people were suddenly struck down to the ground and rendered helpless. This would have included people suffering from a diverse range of diseases such as subarachnoid haemorrhage, epilepsy and viral labyrinthitis.

By the early part of the 19th century it was recognised that many of these patients could be separated into distinct groups and that diseases often followed a predictable course. In 1821, Itard described a number of patients with 'cerebral apoplexy' who recovered from their attacks without subsequent neurological deficit. This included patients with what was probably epilepsy, hysteria or vestibular disorders. He identified that the dizziness may have in some cases have been

From the Departments of Otorhinolaryngology, Walton Hospital*, Liverpool, and Queen's Medical Centre†, Nottingham, UK.
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connected with ear disease rather than a central nervous system disorder and described in detail a patient with symptoms characteristic of Menière's disease (Itard, 1821).

Much of the otological work at the time was centred in Paris and Flourens in the mid 1800's described some physiological experiments on the semicircular canals of pigeons. By destroying selective canals he was able to produce rhythmic head movements that corresponded with the canal destroyed. He proposed that the semicircular canals were involved in the maintenance of posture and balance (Flourens, 1842).

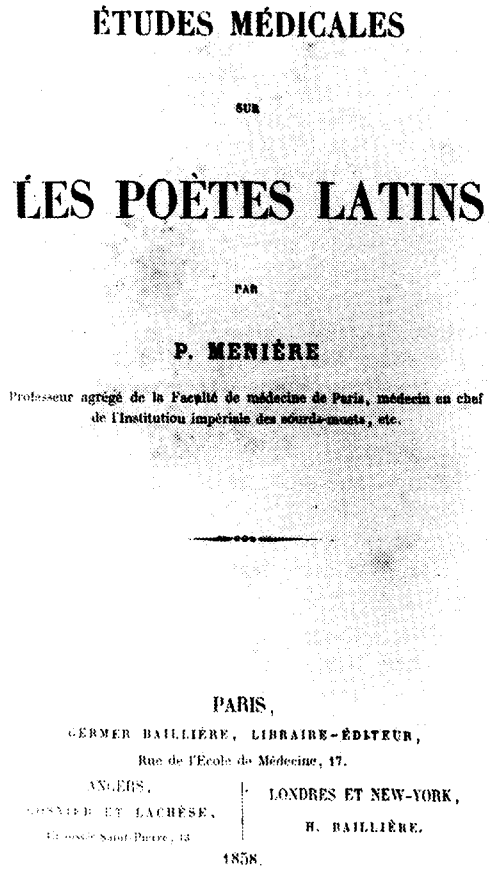
Prosper Menière (1799–1862) (Figure 1)

Prosper Menière was born in Angers on the Loire in south west France in 1799. He was educated at the Lycée in Angers and later at the University. He was recognised as a brilliant student of the classics and humanities, publishing a book of Latin poems in 1858 (Figure 2). He completed his medical studies at the Hotel-Dieu in Paris (Figure 3) receiving a gold medal in 1826. He gained his M.D. in 1828 and worked as an assistant to Baron Dupuytren. He was then appointed assistant professor in the faculty of Medicine and Hygiene but political upheaval interrupted his career and he was later sent to control the spread of cholera in the departments of Aude and Haute-taronne. For this he received the Chevalier de Légion d'Honneur but despite this his work failed to secure the professorship in medicine and hygiene in Paris that he desired, and he was instead appointed physician-in-chief at the Institute for deaf-mutes. From then on his interests centred mainly on diseases of the ear (Birch, 1974; Editorial, 1969).

His description of the disease that now bears his name was his last and most important contribution to medicine. Drawing on his experience at the Institute for deaf-mutes and on the work by Flourens, Prosper Menière first appears to have made the connection between disease arising in the labyrinth and vertigo in 1848. He described the case of a young girl who while suffering an 'influenzal' illness became suddenly completely deaf and experienced continuous vertigo. She died a few days later and at autopsy was found to have a 'plastic' red matter in the semicircular canals; there was no evidence of intracerebral disease. Despite subsequent confusion there is no suggestion that this case formed the basis for his later description of the disease that bears his name, but rather it added weight to the idea that inner ear disease might cause severe vertigo in the absence of central nervous system disease.

In 1861 he presented a paper *On a particular kind of hearing loss resulting from lesions of the inner ear* to the Imperial Academy of Medicine in Paris. The title was improved on publication to *A report on lesions of the inner ear giving rise to symptoms of cerebral congestion of apoplectic type* (Menière,

FIG. 1
Prosper Menière (1799–1862).


ÉTUDES MÉDICALES

SUR

LES POÈTES LATINS

PAR

P. MENIÈRE

Professeur agrégé de la Faculté de médecine de Paris, médecin en chef de l'Institution impériale des sourds-muets, etc.

PARIS,
GERMER BAILLIÈRE, LIBRAIRE-ÉDITEUR,
Rue de l'École de Médecine, 17.

ANGERS,
LONDRES ET NEW-YORK,
H. BAILLIÈRE.

1858.

FIG. 2
Latin Poems, The Wellcome Institute Library, London.

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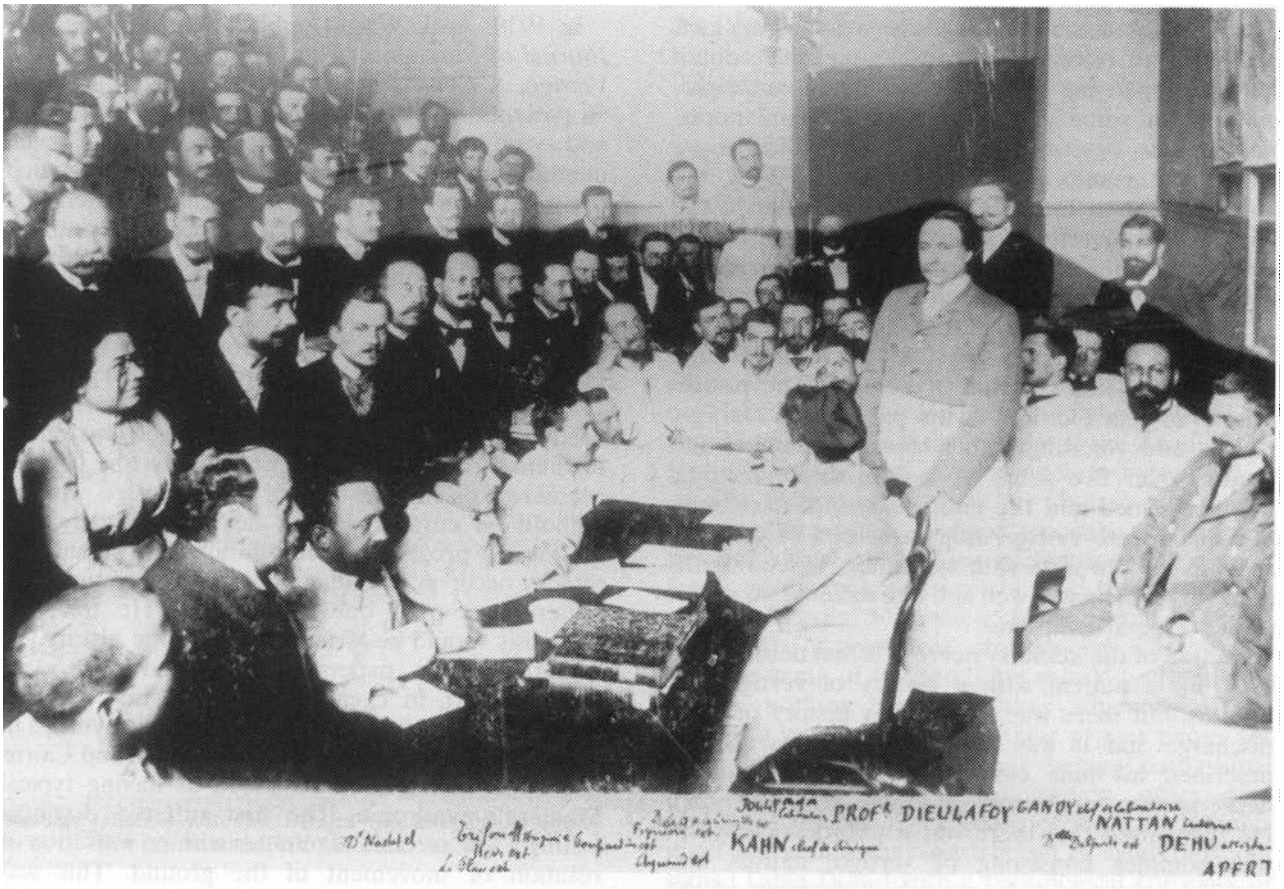

FIG. 3
Hôtel-Dieu, Paris, The Wellcome Institute Library, London.

1861). He described a number of patients suffering from episodic vertigo, deafness and tinnitus and concluded in his paper that:

- the auditory apparatus may be suddenly affected causing tinnitus, and diminution of hearing;
- the inner ear is the site which can suddenly be affected causing attacks of vertigo, dizziness, uncertain gait, staggering and falling, and may be accompanied by nausea, vomiting and syncope;
- attacks are intermittent followed by hearing loss of increasing severity;
- the lesion is likely to be in the semicircular canals.

He recognised that diseases of central origin, such as epilepsy, were distinct from episodic aural vertigo as patients with epilepsy did not experience deafness and those with aural vertigo did not lose consciousness during an attack.

1874–1938

In 1874, Charcot spoke on the description of vertigo, deafness and tinnitus by Menière and called it for the first time 'Maladie de Menière' (Charcot, 1874). He proposed that all forms of treatment were hopeless but noted that when deafness became complete the disease stopped. He wondered if surgery to divide the auditory nerve might be of some benefit, (this was before the development of asepsis and neurosurgery).

Six years later, McBride and James in the *Edinburgh Medical Journal* discussed what they termed Menière's disease, describing paroxysmal or constant vertigo with coincident deafness with or without other nervous phenomena (McBride and James, 1880). They stated that Menière's disease was an organic disease of the labyrinth caused by either an organic lesion of the semicircular canals, inflammation of the middle ear or increased pressure in the labyrinth. Shortly afterwards McBride revised this stating that the name Menière's disease should be reserved for primary lesions of the labyrinth, more particularly cases where haemorrhage into the semicircular canals was the cause of the symptoms (McBride, 1881). He was concerned with the trend of using the term Menière's disease for all deaf giddy patients, stating that 'if it be desired to perpetuate the name by all means let us speak of Menière's syndrome'. He particularly emphasised the need for a thorough neurological and otological examination to exclude non-labyrinthine diseases.

In 1902 Professor Politzer of Vienna in his textbook on '*Diseases of the Ear*' discussed Menière's findings. He stated that a number of pathological processes could cause Menière's 'type symptoms' but felt the term Menière's disease should be reserved for the apoplectiform type of sudden deafness and vertigo arising from the labyrinth. He argued that dizziness, tinnitus and deafness arising from other acute or chronic afflictions of the ear should be called 'aural vertigo'. His description of Menière's disease is characterised by dizziness,

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tinnitus, nausea and vomiting, with a staggering gait, deafness and brief loss of consciousness. He noted that accompanying signs included nystagmus, diplopia and in some cases a transient haemianopia. Politzer also described a number of medical treatments, the results which were measured by the improvement in hearing and the reduction in the number and severity of the attacks. This appears to be the first attempt to quantify treatment results for patients with Menière's disease.

The first operation for Menière's disease, labyrinthectomy, was described in 1904 by Richard Lake at the Royal Ear Hospital in London. The patient was a 21-year-old lady with paroxysmal vertigo, nausea and vomiting with increasing deafness and tinnitus over five years. A radical mastoidectomy was performed and the entire labyrinth destroyed. She had severe vertigo afterwards but by the 10th day was able to walk with assistance. Three months after surgery she was well and had suffered no return of her original symptoms.

Section of the auditory nerve was first described in 1904 for a patient with a history of vertigo and tinnitus, but there was also a clear history of aural discharge and it was not until 1928 that Dandy described his nine cases of intracranial auditory nerve section for Menière's disease (Dandy, 1928). He recognised that there was a well-defined symptom complex consisting of vertigo, nausea and vomiting, tinnitus and progressive deafness of the affected ear. The attacks he reported lasted from days to weeks with the patients well between attacks. All his patients survived and all were relieved of their symptoms. He felt that as the patients suffered loss of both hearing and vestibular dysfunction it was more likely that the underlying lesion was in the eighth nerve itself rather than the semicircular canals.

A year earlier Portmann had described the endolymphatic sac incision for severe vertigo in what he called Menière's syndrome (Portmann, 1927). The rationale for this was based on the work of Knapp in 1871 who proposed that Menière's disease was a form of aural glaucoma. He described in his paper the *Triad of Menière; vertigo, deafness and tinnitus* (Knapp, 1871).

In 1934, Furstenberg published a paper on the medical treatment of 'Menière's symptom complex', a more accurate term in his view. He pointed out that many otologists had made their own classification of the Menière's symptom complex 'whimsically' and without consideration of the scientific facts upon which a clinical entity is based, 'it has been a term loosely applied and much abused'. He was particularly troubled by the fact that this did not allow comparable groups of patients and their treatments to be evaluated. He stated that Menière's disease had already been well defined in the medical literature as violent attacks of vertigo with nystagmus in a patient with deafness, associated with nausea and vomiting and tinnitus. He felt that it was due to retention of sodium in the body and devised a sodium-exclusive diet, claiming good results.

In 1938, A. J. Wright published a paper in the *Journal of Laryngology and Otology* entitled *'Aural Vertigo, A Clinical Study'*. He described a group of 66 patients with characteristic hearing loss, tinnitus and vertigo of variable intensity, without even mentioning the name of Menière. He noted that the middle ear was normal in all cases but felt that the symptoms were due to a 'focal labyrinthitis' secondary to associated infection in the head and neck region such as dental caries or chronic tonsillitus.

In the same year, S. J. Crowe from the John Hopkins University in Baltimore wrote up a group of 117 patients (Crowe, 1938). He had performed vestibular or complete eighth nerve sections on 94 of them. From his personal observations he defined Menière's disease as sudden onset vertigo with or without an aura, inner ear deafness (usually unilateral and progressive) and tinnitus. He stated that attacks occur at irregular intervals at any time and patients are well between attacks. He felt that diagnosis should be based mainly on the history.

Another two papers published in 1938, two months apart, in London and Japan first describe the pathological finding of endolymphatic hydrops in patients with Menière's disease. Hallpike and Cairns had two patients they described as having typical Menière's syndrome. The first suffered dizziness lasting a few seconds to minutes with no sensation of rotation or movement of the ground. This was associated with loss of consciousness, progressive deafness and tinnitus. The second was a patient with rotational vertigo lasting minutes to hours and deafness. Kyoshiro Yamakawa in Osaka described his findings in the temporal bone of a colleague, Juuemon Ogata, professor of Obstetrics and Gynaecology, who had recently died (Paparella, 1992).

He had suffered from episodic vertigo with nausea and vomiting lasting days early in the disease, but this progressed to almost continuous disequilibrium. He called it a 'status of Menière's'. The Japanese paper did not come to light until some years later.

1941–1972

In 1941, Dandy wrote again on section of the audiovestibular nerve drawing attention to the continuing confusion over the terminology in use at the time. He now preferred the term Menière's syndrome to Menière's disease or 'aural vertigo' but continued to use the term Menière's disease throughout his paper. He also used the term 'Menière's attacks' to describe the episodic vertigo, hearing loss and tinnitus that occur. For patients with characteristic vertigo but no deafness or tinnitus he introduced the term 'pseudo-Menière's'.

In *Diseases of the Nose, Throat and Ear* by Jackson and Jackson (1946) it was acknowledged that there was still much controversy as to what constituted a case of what they termed Menière's syndrome. 'Under this heading are placed most of the cases of deafness, tinnitus, and dizziness occurring with non-suppurative disease of the labyrinth and without

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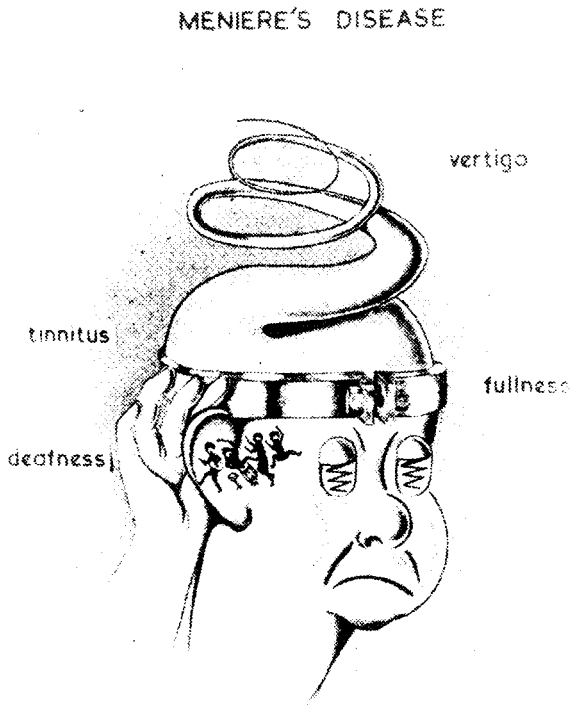

FIG. 4
Menière's disease, note the 'Beatles' representing the sensation of tinnitus! (Fick, 1966 JLO 80:289).

evidence of organic disease in the central nervous system'. They avoided the term Menière's disease as they felt this implied a knowledge of the pathological basis of the condition. Their definition included: dizziness with or without nystagmus, tinnitus (usually) and deafness. If one or two symptoms were absent they felt it should be called atypical Menière's syndrome.

In 1948, Garnett Passe and Seymour described sympathectomy for 'Menière's syndrome'. He gave a clear definition of the condition and described the results of surgery on 12 patients, based on the relief of vertigo, deafness and tinnitus, for which he claimed success. In 1964, Fick described the sacculotomy for Menière's disease. Pre- and post-operative symptoms were listed for 50 patients. All had vertigo and deafness, most tinnitus and some also had aural fullness (Figure 4). In the introduction he was careful to state that other causes of the symptoms had been excluded. In 1966, Tumarkin described the insertion of a grommet for 'Menière's syndrome', he stated that the term was used far too loosely and that the generic term 'labyrinthopathy' should be used for all syndromes involving the inner ear. He suggested that Eustachian tube dysfunction was in part responsible for the symptoms and was successful in 18 out of 20 patients treated. In 1969, Angell-James described ultrasonic destruction of the labyrinth, dividing patients in two groups, those with primary and those with secondary Menière's disease. Primary was idiopathic and called Menière's disease and he attributed this to autonomic instability which occurred during times of stress. The secondary form developed after middle ear infections or inner ear trauma and he called this Menière's syndrome. Patients had so called 'Menière's attacks' in both.

1972 onwards

Although the definition of Menière's disease as a specific entity was recognised, confusion in the terminology and range of conditions covered remained. With the advent of new treatments both medical and surgical results needed careful monitoring and the improvement in symptoms needed to be defined. In 1972, in response to this the Committee on Hearing and the Equilibrium Subcommittee on Equilibrium and its Measurement at the Academy of Ophthalmology and Otolaryngology proposed a specific definition of Menière's disease and guidelines for the evaluation of and reporting of treatment results (Alford, 1972). They recognised that 'criteria for diagnosis may vary greatly', and that 'a uniform set of criteria for diagnosis and judging and reporting the results of treatment' was needed. They stated that the name of the condition should be Menière's disease nothing else. All other afflictions of the labyrinth must be called by their proper name, for example, viral labyrinthitis or benign paroxysmal positional vertigo. Their definition states that Menière's disease is a disease of the membranous inner ear with a characteristic set of symptoms and signs (Table I) and with a pathological correlate of endolymphatic hydrops. They also defined two subvarieties of Menière's disease, cochlear Menière's disease (without vertigo) and vestibular Menière's disease (without deafness).

There have been two updates from the Committee on Hearing and Equilibrium since 1972. In 1985, it was felt that the definition of Menière's disease needed restricting to those cases with a full complement of classical symptoms and signs, as there was

TABLE I
1972 CRITERIA FOR THE DIAGNOSIS OF MENIÈRE'S DISEASE

1. Fluctuating, progressive, sensorineural deafness.
2. Episodic, characteristic definitive spells of vertigo lasting 20 minutes to 24 hrs. The patient remains fully conscious with no neurological accompaniments or sequelae; vestibular nystagmus is always present.
3. Usually tinnitus.

The attacks are characterised by periods of remission and exacerbation.

TABLE II
1985 CRITERIA FOR THE DIAGNOSIS OF MENIÈRE'S DISEASE

1. A fluctuating, sensorineural hearing loss associated with tinnitus – the deficit is characteristically of low frequency or flat type.
2. Vertigo – spontaneously occurring sensation of movement that is accompanied by unsteadiness and lasts from minutes to hours. More than one attack needed to establish diagnosis.

Definitive spell – often prostrating, often accompanied by nausea and vomiting. Patient oriented and conscious, no neurological sequelae. Horizontal or horizontal rotatory nystagmus is always present during the definitive spell.

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TABLE III
1995 CRITERIA FOR THE DIAGNOSIS OF MENIÈRE'S DISEASE

1. Recurrent, spontaneous episodic vertigo.
   definitive spell – spontaneous rotational vertigo lasting at least 20 minutes (commonly several hours), often prostrating, accompanied by disequilibrium that may last several days, usually nausea, commonly vomiting or retching, no loss of consciousness. Horizontal or horizontal rotatory nystagmus is always present.
2. Hearing loss – (not necessarily fluctuating).
3. Either aural fullness or tinnitus (or both).

'Certain' Menière's disease is 'definite' disease with histopathological confirmation.

'Definite' Menière's disease requires two or more definitive episodes of vertigo with hearing loss plus tinnitus and/or aural fullness.

'Probable' Menière's disease needs only one definitive episode of vertigo and the other symptoms and signs.

'Possible' Menière's disease if defined as definitive vertigo with no associated hearing loss or hearing loss with non-definitive disequilibrium.

no evidence that 'cochlear' or 'vestibular' Menière's disease was based on the same pathological process (Table II). Their aim was 'objectivity and clarification in the reporting of clinical observations' giving new guidelines for the reporting of treatment results and assessment of disability (Pearson and Brackmann, 1985).

The 1995 criteria aim to simplify the definition of Menière's disease and allow more flexibility, making it more usable in a wide range of settings. They clearly distinguish between recording and the analysis and interpretation of observations allowing existing data to be looked at in new ways. Menière's disease is re-defined as the idiopathic syndrome of endolymphatic hydrops and the importance of excluding other possible causes are discussed. A minimum set of signs and symptoms must be fulfilled and the degree of certainty of the diagnosis can be established (Table III). Patient functional level is assessed in a more accurate way, taking into account the problems that the previous disability scale had posed. This allows more accurate assessment of treatment progress.

In 1991, McKee *et al.* published a review of all the significant trials relating to the surgical treatment of Menière's disease. They found that of those trials published after 1972 only 65 per cent used the guidelines in reporting results and even up to 1989 trials were still not adhering to these criteria.

Conclusion

The definition of Menière's disease has evolved as more about the disease is understood. The initial breakthrough by Prosper Menière was in recognising the source of the intermittent vertigo, deafness and tinnitus as in the labyrinth rather than a condition arising centrally. It is a definition of exclusion as many diseases can almost duplicate the signs and symptoms of Menière's disease. Even today there is very little known about the aetiology of the condition and the only known pathological correlate is that of endolymphatic hydrops which can be found in a number of other labyrinthine disorders.

Initially patients with Menière's disease were a fairly well-defined group but with time a broader group became included and the definition varied between authors. This was not helped by the wide variety of terms used such as aural vertigo, labyrinthopathy and Menière's syndrome. Many patients appear to have been labelled as having Menière's disease simply because they suffered from intermittent disequilibrium without more rigorous criteria being applied. This led to the introduction of tighter inclusion and exclusion criteria in 1972 to give a degree of uniformity and allow comparison of studies to be made.

Today the criteria set down in 1995 should be used when reporting the results of treatment whether medical or surgical and reasons for going beyond these given. However, as with any definition, it should remain in discussion and be allowed to evolve as medical knowledge progresses.

Acknowledgements

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Address for correspondence:
Mr N. J. P. Beasley,
6, Sandlea Park,
West Kirby,
Wirral L48 0QF.

Fax: 0151-529-4033