MÉNIÈRE'S DISEASE **MEDICAL APPENDIX**

DEFINITION

1. MÉNIÈRE'S disease was first described by Prosper Meniere in 1861. It is a disease of the labyrinth of the ear characterized by progressive fluctuating deafness and paroxysmal vertigo, with tinnitus or aural fullness or both. Some authorities add loudness intolerance and diplacusis (the hearing of identical sound differently by the two ears) to the definition.

    1.1. Confusion has arisen from the use of the term "Ménière's syndrome", where the symptom complex had an identifiable extrinsic cause, as opposed to the "disease" in which the condition was regarded as idiopathic. However, the former term has now been largely abandoned.

    1.2. The use of Ménière's disease as an umbrella term for vertigo is also erroneous and confusing.

2. **Pathology**
    Ménière's disease has, as its pathological correlate, gross distension of the endolymphatic spaces of the labyrinth. This is called endolymphatic hydrops and may result from either overproduction or impaired absorption of the fluid, with distortion of the membranous labyrinth. The disturbance is essentially vascular. The presence of endolymphatic hydrops can be demonstrated by transtympanic electrocochleography.

CLINICAL MANIFESTATIONS

3. Estimates of the incidence of Ménière's vary widely. In Britain, the incidence is about 0.1%. No age group is immune, but it is very rare below the age of 10 years and in most cases, the disease begins before the age of 60 years. The peak incidence of the onset of vertigo is in the fourth and fifth decades.

4. **Vertigo** is the most distressing symptom. It usually occurs suddenly and spontaneously. There may be a premonitory feeling of fullness in the affected ear, or an increase in tinnitus, but there is usually no warning. It may occur during sleep. The vertigo is typically rotational in type although linear movements sometimes occur. The patient is usually prostrated with nausea and vomiting. Typically, each attack lasts for between half an hour and a few hours. A history of attacks lasting for days on end without remission throws doubt on the diagnosis of Ménière's. Occasionally, the attacks occur singly with intervals possibly of months or years between but often there is a series of attacks occurring over a period of weeks or months, followed by a long period of complete freedom. The vertigo of Ménière's is not associated with impairment of consciousness or orientation and there is no focal neurological symptom.

5. Generally, the pattern of vertigo in Ménière's disease is a gradual increase in the frequency of attacks over a period of years until a maximum is reached, followed by decreasing frequency as the disease runs its course, with irreversible damage to the inner ear.

6. Loss of vestibular function is often bilateral. **Nystagmus**, which may cause visual blurring, is present during an attack, and may be detectable between attacks.

7. **Deafness** is usually the first symptom to appear. It may precede the onset of vertigo by some months. Characteristically, it is initially unilateral, of the perceptive type, predominantly affecting the lower frequencies and recovering between attacks. In some cases, recruitment is a prominent symptom. As the disease progresses, the deafness becomes severe, with a sensorineural loss of 60 dB or more extending over the whole frequency range in both ears, and is permanent.

8. **Tinnitus** accompanies the deafness in nearly all cases of Ménière's disease, typically increasing in volume and character during and sometimes prior to, the episodes of vertigo. Its pitch tends to correspond to that of the most severe hearing loss.

AETIOLOGY

9. Controversy remains as to whether Ménière's disease is a disease in its own right or merely a symptom complex produced by many different entities. Ménière's disease can be either primary or secondary in type. The cause of Ménière's disease remains unknown in many cases.

Aetiological factors in primary Ménière's Disease

10. Genetic factors.

    10.1. There is evidence of a **familial** predisposition. In between 14% and 20% of patients there is a positive family history of the disease, in some cases with a pattern suggestive of autosomal dominant transmission.

    10.2. Evidence of **racial variation** has been hard to find, because of difficulty in obtaining reliable figures of incidence, and even more so regarding prevalence. However, the rarity of Ménière's in some populations, such as southwestern American Indians, suggests a genetic factor.

    10.3. There is no good evidence of gender preponderance in the incidence of Ménière's.

11. **Anatomical factors.** High-resolution magnetic resonance imaging of the inner ear normally gives good visualization of the endolymphatic duct and sac, but not so in ears affected by Ménière's disease. This suggests that anatomical malformation may be, at least, a contributory factor in a proportion of cases. Some studies support the theory that an abnormally narrow vestibular aqueduct can cause Ménière's disease.

12. **Viral infection.** Evidence implicating latent viral infection as a cause of Ménière's comes also from studies which have found raised circulating levels of viral antibodies or specific viral proteins in active cases. Other studies have found viral DNA in the endolymph.

13. **Autoimmune Process.** In some cases of Ménière's disease, notably where the disease is bilateral, there is serological evidence of an autoimmune factor.

14. **Allergy** has been discounted as an aetiological factor in Ménière's, because IgE changes have not been found.

15. A primary **psychosomatic** aetiology has been suggested, but there is good evidence that psychiatric morbidity associated with Ménière's is a result, rather than a cause, of the disease.

Aetiological factors in secondary Ménière's Disease

16. **Trauma**

    16.1. Direct physical or acoustic trauma. Trauma, in the form of direct head injury, surgical damage to the inner ear, severe explosion blast injury, or barotrauma (such as occurs in deep-sea diving) may produce any or all of the manifestations of Ménière's disease.

    16.2. Damage due to trauma is usually immediate and hearing loss first noticed 6 months or more after the injury cannot reasonably be attributed to that injury.

    16.3. An injury insufficiently severe as to cause loss of consciousness is very unlikely to damage the hearing.

    16.4. There is no good evidence that continuous noise exposure, with consequent damage to the cochlear cells, causes Ménière's Disease.

    16.5. Vertigo is common following head injuries and, as with deafness, can occur without a skull fracture. The unsteadiness is most commonly associated with post-concussion syndrome, usually settles in a matter of 6-12 months and, if prolonged beyond this time, the question of a post-concussional neurosis must be considered.

    16.6. In any **severe** head injury involving the vestibular aqueduct, apart from the immediate symptoms, delayed endolymphatic hydrops may develop. There is likely to be profound deafness. Only if there is any remaining vestibular function, which is very unlikely following such an injury, can episodic rotatory vertigo ensue.

17. **Abnormal fluid balance.** The Ménière's triad of symptoms may be associated with changes in fluid balance in metabolic and endocrine disorders. Examples are hyperglycaemia, hypoglycaemia, haemodialysis, hypothyroidism and pituitary insufficiency. In a sub-group of female patients with Ménière's, episodes were found to occur in the premenstrual phase, when there is altered fluid balance. The disturbed fluid balance is thought to precipitate symptoms in predisposed ears, rather than causing the disease. The symptoms of endolymphatic hydrops will not persist after return to normal fluid balance.

18. **Chronic otitis media** has been found in association with endolymphatic hydrops in a high proportion of temporal bone studies, in both animals and humans. Its effect on the inner ear is probably due to migration of toxins and associated enzymes through the round window membrane into the fluids of the inner ear. Mastoiditis with consequent osteitis may also interfere with the blood supply to the inner ear. Where Ménière's follows such infections, it is reasonable to accept a causal relationship.

19. **Syphilis**, either congenital or acquired, is a known cause of endolymphatic hydrops and the Ménière's triad of symptoms.

20. **Viral infection,** there is evidence that endolymphatic hydrops may occur as a delayed sequel of inner ear damage sustained during a subclinical attack of viral labyrinthitis occurring many years previously, for example in measles or mumps.

21. Ménière's Disease can occur particularly in middle aged patients with pre-existing **systemic autoimmune disorders** such as rheumatoid arthritis who develop the Ménière's triad of symptoms.

22. Patients with **otosclerosis** occasionally develop symptoms similar to Ménière's. This may be due either to impingement of otosclerotic bone on the vestibular aqueduct or to biochemical alteration of the inner ear fluids. It is sometimes referred to as otosclerotic inner ear syndrome. Any **tumour** of, or impinging on, the inner ear can give rise to a similar picture.

23. **Other possible causes**
Leukaemia, Shy-Drager syndrome and temporal arteritis have been found to be associated with the development of endolymphatic hydrops.

CONCLUSION

24. Ménière's disease is a relatively common entity recognised by a characteristic triad of symptoms. The diagnosis should not normally be made without the presence of all three elements of the triad. Whilst the aetiology in the individual case may not be identifiable, certain environmental and other factors are known to lead to its development.

25. There is no evidence that adverse climatic conditions, psychological or physical stress (other than the infections and injuries mentioned above), are causative factors.

REFERENCES

Kerr A G, Smyth G D L. Ear Trauma. In: Booth J B (Ed). Scott-Brown's Otolaryngology. 5th Ed. Vol 3. Otology. London. Butterworth. 1987. p444-464.

Kerr A G (Gen Ed). 1997. Scott-Brown's Otolaryngology. 6th Ed. Vols 1–6. Oxford. Butterworth Heinemann.

Moffatt D A and Ballagh R H. In: Booth J B (Ed). Scott-Brown's Otolaryngology. 6th Ed. 1997. London. Butterworth Heinemann. 3:19:1-38.

Paparella M M and Mancini F. Trauma and Ménière's Syndrome. In: Larngoscope 93. August 1993. p1004-12.

Rudge P. in Weatherall D J. et al (Eds). Oxford Textbook of Medicine. 3rd Ed. 1996. Oxford. p3785.

Schuknecht H F. Classification of Endolymphatic Hydrops. Ann J of Otology. 1984;5:528.

Schuknecht H F. Delayed Endolymphatic Hydrops. Ann Otol Rhinol Laryngol 1978;87:743-748.

Shambaugh & Wiet. The Endolymph sac and Ménière's Disease. Otolaryngolic Clinics of North America. 1980;13:585-588.

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