

Ménière's Disease

This factsheet has been written for people who have been diagnosed with Ménière's Disease or who think they have similar symptoms. It may also be useful for relatives or friends of people with Ménière's, or for anyone with a general interest in hearing-related conditions.

What is Ménière's disease?

Ménière's affects about 15 in every 10,000¹ adults in the UK population. It is a late-onset disease usually starting between the ages of 30-50 years. Onset in the 60s is less usual, and childhood and young adult cases are relatively unusual. For most the onset is sudden and frightening, the predominant symptom being vertigo, a subjective sense of intense rotation of self and/or the environment. Before Prosper Ménière drew attention to the inner ear as the site of the problem, attacks were considered to be apoplectic or epileptic. This gives a measure of the severity of severe attacks.

Ménière's is characterised by severe attacks of vertigo combined with progressive deafness, tinnitus and a feeling of pressure in the affected ear. Attacks often start without warning and lead to loss of confidence, anxiety and sometimes depression. Those affected can feel isolated and the effects can impact upon relationships and working life.

¹ Ballantyne's Deafness (6th Edition) 2001, edited by John Graham and Mike Martin, p.157.

The severity of symptoms is variable, though typically includes:

- Severe attacks of vertigo lasting up to several hours, with associated nausea, vomiting and sometimes diarrhoea.
- Pressure in the ear(s), tinnitus and a blocked feeling with fluctuating deafness.
- Active spells with attacks spread over several weeks, following by periods without attacks lasting some months.

What is the outlook for people with Ménière's?

Typically, people with Ménière's will find that, over a number of years, the attacks will become shorter, less severe and, in some cases, may cease altogether. However, as episodes of vertigo become less frequent, hearing loss often worsens. Although in 85-90% of cases, hearing loss will be restricted to one ear in the early stages, around half of those affected will eventually suffer hearing impairment in both ears. Hearing loss usually increases before, during or following vertigo attacks, though in some cases, the deterioration of hearing can progress independently of attacks.

Some 10% of people with Ménière's have severe and continuing symptoms with short remissions and they may develop serious balance problems as a result of the loss of inner ear function.

Some develop 'drop attacks', which can be very worrying.

What causes Ménière's?

The exact cause of Ménière's is unknown, though researchers do have a good understanding of the processes at work in the inner ear. Those affected experience an excessive build-up of fluid within the innermost compartment of the ear, sometimes known as idiopathic endolymphatic hydrops.

In some 10 per cent of those affected there is a family history of the disease, suggesting some individuals have a genetic predisposition. Other factors that have been implicated in triggering active spells include viral infections, allergy, abnormal responses of the immune system and, in many cases, stress.

How is Ménière's diagnosed?

If you suspect you may be, it is important to seek the advice of your family doctor (GP) immediately. The doctor will usually refer you to a Consultant.

The Consultant will build up a history of the attacks and will exclude other possible causes of the hearing loss, tinnitus, dizziness and vertigo. Routine and special hearing tests reveal the pattern of deafness, indicating if it is typical of Ménière's.

Balancing tests and electrical recordings of eye movements will also be used to evaluate the balance functions of the inner ear. These tests are not invasive. Electrical recordings from the inner ear may be indicated.

Tests such as auditory brain-stem responses (ABR), computerised axial tomography (CAT) or magnetic resonance imaging (MRI) may also be used to rule out more serious though rare causes, such as tumours or various neurological diseases.

The doctor and/or Consultant are also likely to carry out routine physical and blood tests.

What treatments are available?

Although there is no cure, there are a number of effective treatments which can help.

- Various sedative drugs can reduce the severity of vertigo during attacks.
- Medicines designed to influence the blood supply or fluid movements in the inner ear may help and are frequently prescribed.

In severe cases, if attacks persist despite medication, a Consultant may advise surgery. A number of different operations have been devised, some to reduce pressure in the inner ear, others to diminish or abolish the abnormal impulses passing from ear to brain.

What should I do if I suffer an attack?

During acute attacks, lie quietly, preferably with the eyes closed, on whichever side helps to minimise the spinning sensation. A drug called Buccastem, taken under the tongue, will reduce the effects of the attack. In the case of a prolonged attack, your doctor may administer the drug by injection.

Severe attacks are likely to result in several days of dizziness and instability. Until these attacks have passed, it is important to rest and a few days off work may be necessary.

Mild attacks can be managed by medication and a few days of rest.

Should an attack start while driving (there is usually some warning such as increased pressure in the ear or slight

dizziness), pull over and seek assistance. Road traffic accidents are remarkably rare for those with Ménière's. If you suffer from drop attacks, you should not drive until they have been stopped through surgery.

How can lifestyle factors influence Ménière's?

Many believe that a healthy lifestyle can help to alleviate the symptoms of Ménière's, making the attacks less frequent or less severe.

In particular, those affected should try to avoid:

- stress
- alcohol
- excess salt
- nicotine
- caffeine

Balancing exercises have also been designed specifically for those affected.

What research is being conducted into Ménière's?

Researchers funded by Deafness Research UK are currently conducting a major project to unravel the genetic basis of Ménière's. Most cases of Ménière's are sporadic but in 7% of cases, other family members are similarly affected. There is no clinical difference between sporadic and familial causes.

To search for genes likely to harbour DNA mutations that may cause Ménière's, a 'genome scan' is being conducted at the MRC Geneservice laboratory in Cambridge, with funding from The Ménière's Society.

In this approach, DNA from affected and unaffected individuals from 17 families with Ménière's is employed to generate data that will reveal the chromosomal

location of genes that may explain the disorder in some or all of these families.

A major grant from Deafness Research UK has enabled researchers to analyse the gene mapping data from the genome scan. So far about two thirds of the data have been analysed, and new results strongly suggest that a gene that causes Ménière's in about half the families is located on chromosome 14.

No other chromosomal regions have so far emerged as likely candidates for harbouring any other genes, but the analysis is ongoing.

Genes located on chromosome 14 are being screened for mutations in affected individuals, but so far, those screened do not appear to be involved.

Additional work funded by this grant; the researchers are examining the role of immune system genetics in predisposition to Ménière's, since there is thought to be an autoimmune component to the genetic predisposition.

Other approaches are also being taken using the grant funding from Deafness Research UK. In one such approach, researchers are screening genes located anywhere in the genome that they predict may predispose to Ménière's when they are not functioning properly. In this 'candidate gene' approach, genes are screened directly for mutations in patients with a family history of Ménière's. A number of potential genes have so far been ruled out by this process, and others are currently being examined.

Longer term plans include searching for genes that predispose to Ménière's in a large population of sporadic patients. This will use approaches complementary to those being used already, in an effort to maintain the momentum that has been built up in the search for all genes that influence Ménière's.

Unravelling the genetic basis of Ménière's will help to direct the search for new therapeutic and even preventative strategies, in the long-term. In the short term, it will speed up early diagnosis, which can sometimes be difficult, and it will provide a predictive test for relatives or descendants of affected patients, especially in familial cases.

Further information?

If any of your questions concerning sudden sensorineural hearing loss have not been answered by reading this factsheet, contact the Deafness Research UK Information Service for further assistance. Our Information team will either answer your enquiry directly or refer it to one of our scientific or medical advisers.

Open: 9.30 a.m. to 5.30 p.m., Monday to Friday (a message can be left at other times)

Freephone: 0808 808 2222

E-mail: info@deafnessresearch.org.uk

or click the 'ask question' option from our website homepage:

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NHS Choices

Has the latest information regarding treatment available on the NHS. It also has information on any clinical trials that are recruiting.

<http://www.nhs.uk/conditions/menieres-disease/Pages/Introduction.aspx>

Deafness Research UK is the only national medical research charity dedicated to helping people with deafness, tinnitus or other hearing problems.

Scientists are now predicting that within the next ten to fifteen years there could be a cure for some forms of deafness and much more effective treatments for tinnitus. Deafness Research UK is at the forefront of this work.

You can support us by making a donation. For more information call us on 0207 164 2290 or write to:

Deafness Research UK, 330-332 Gray's Inn Rd, London WC1X8EE

Charity no. 326915

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You should not act on any advice without first referring to your family doctor or another medically qualified adviser.

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