

MÉNIÈRE

an ear disease

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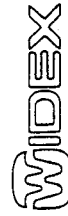
Ménière's disease

Ménière's disease is a disease of the ear, combining hearing loss, tinnitus (ringing, buzzing), severe attacks of vertigo and sound distortion. By definition all Ménière patients present these symptoms, but in the beginning of the disease most individuals, later on characterized as suffering from Ménière's disease, may show only one or two of these symptoms. There are mild and severe cases. In severe cases, the disease may lead to disablement to such an extent that the person afflicted must give up working.

Knowledge about the disease

Little is known about the causes of the disease. Unfortunately, knowledge about efficient treatment of the disease also leaves much to be desired. In most cases, however, there can be some relief for the symptoms, which typically vary considerably from one person to another.

It is not only doctors that know too little about Ménière's disease. Those who suffer from the disease, their families and friends also lack information. Our aim with this brochure is to improve this situation.



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Ménière's disease

Ménière's [mān yārzh] disease is a disease of the inner ear, described for the first time by the French otologist Prosper Ménière in 1861.

The disease is characterized by the development of increasing *hearing loss* and *tinnitus* in a formerly normal ear, combined with *severe attacks of dizziness*.

Even though the disease has been the object of extensive research during the last 135 years, the actual cause of the disease is still not known. This may be explained by the presumption that it is not the same cause or incident that leads to two different persons contracting the disease. In about 20% of all cases, however, the disease is believed to be caused by hereditary factors. The course of the disease may also vary from one patient to another.

Even though only little is known about the possible cause of Ménière's disease, we do know much about what happens in the inner ear of those who contract the disease.

Ménière's disease is not rare. It is estimated that about one out of every two thousand people suffer from its symptoms. It affects men and women equally. It may begin in childhood or late in life. Typically, however, it affects persons around the age of 40 to 50, but it is not unusual to see it begin in people above the age of 60.

The Symptoms of Ménière's disease

Attacks of Dizziness

The most characteristic symptom is the patient's suffering from *attacks of dizziness*. It is important to stress that we are talking about *attacks*, which means that the patient feels very dizzy within a clearly limited period of time. The patient may experience a random case of unsteady balance or a slight dizziness (like everybody else does on occasion), but this "discomfort" will in no way resemble the character and the intensity of the balance problems and the dizziness experienced during the attacks.

Dizziness attacks can be of short or long duration, but if they last for only a few seconds or minutes, it is *not* Ménière's disease. If they last for more than 2 or 3 days, it is also not believed to be Ménière's disease. The typical Ménière attack will last for 2 to 4 hours, but this does not prevent individual patients from having attacks of different duration. It is a characteristic of the disease that the patient either feels (relatively) fine, or really bad - the latter characterising an attack.

For most patients it is easy to define when an attack starts, but not so easy to describe when it ends.

Many patients have a kind of forewarning revealing that an attack is to be expected very soon. That forewarning may be a feeling of fullness in the ear or an experience of increasing tinnitus in the diseased ear. During a typical attack, the patient has a *feeling of being tumbled around*. This experience makes it impossible to keep one's balance - often for several hours. The patient is most comfortable lying down, quite still. During the attack there will be involuntary, jerking eye movements (called nystagmus) which disturb the patient's eyesight - often to the extent that it is impossible for the patient to see clearly during the attack.

Nystagmus (involuntary jerking eye movements), is caused by a reflex from the balance organ in the inner ear. Normally when a person turns his head, the eyes react by automatically moving in the opposite direction at exactly the same speed as the rotation. This compensatory eye movement enables us to fixate on the same spot even while rotating the head. When eventually the eyes have turned 10 to 15 degrees in the opposite direction of the rotation, they make a quick movement in the direction of the rotation and thus find a new fixation point. When the balance nerve in the inner ear for some reason is paralysed, spontaneous nystagmus will occur, i.e. eye movements which apparently try to compensate for "imagined" rotation, which is what the dizziness really is. During the Ménière attack the balance nerve in the afflicted ear seems to be paralysed. This explains why the Ménière attack is always accompanied by nystagmus.

During an attack of Ménière's disease, patients feel exactly as if they were seasick. They are nauseous and may vomit. It is characteristic of seasick persons that they feel *very tired* and often also *very depressed*. Headaches are also a frequent symptom. The same is true for Ménière attacks. The fatigue and the depression are hard to get rid of after the attack. Therefore, a Ménière's patient who is going through a tough period, will be tired and depressed. This, consequently, is not a mental reaction to being ill, but part of the disease itself. Many find these symptoms to be just as much of a strain as the attacks themselves. On top of the fatigue and depression, many patients also experience *impairment of memory*. Some patients think that their brain has been damaged and that they are growing senile; but again this is *part of the disease*. When patients recover from the episode, they discover that they have regained their memory, their cheerfulness and their energy.

Unknowing observers who see a person during a Ménière attack quite often believe that he or she is drunk. If this happens at the patient's place of work, and if it happens several times per month, colleagues may turn their backs on him, treating him with a kind of "understanding" which is quite inappropriate. Only very few people know about the disease, so it can be difficult to explain that it is a disease like so many others, brought about through no fault of one's own.

The Hearing of Ménière Patients

In Ménière's disease, the inner ear develops symptoms other than the attacks of dizziness.

In order for a disease to be classified as Ménière's disease there must be a progressive deterioration in the hearing of the sick ear accompanied by *buzzing or ringing in the ear* (tinnitus).

In the early stages of the disease, most Ménière patients only suffer from hearing loss during the attacks of dizziness. But after an accumulation of attacks, the hearing loss gradually makes its presence felt also between attacks. Patients who have had the disease for quite some time may have rather significant hearing losses, creating trouble for them in their daily lives.

Many patients do not understand the change in their hearing function. On one hand, it is obvious that a hearing loss is present - listening on the telephone becomes more difficult and it is hard to perceive sounds coming from the side of the afflicted ear. But at the same time the patient also becomes oversensitive to loud sounds and noise. So it is not only a question of "turning down the sound volume", but also a sense of "turning up the noise volume." The explanation to why an ear can have reduced hearing and yet be oversensitive to noise is that the inner ear does not function merely as a microphone. Some of the sensory cells in the cochlea (the outer

rows of hair cells) actually do not perceive sound, rather they have the special function of amplifying weak sound signals and damping loud sound signals. To use a metaphor, they work like a person who keeps a swing moving. When the swing moves too slowly, the person gives it a push, and when the swing moves too much, the person reduces its movements. These sensory cells in the inner ear are precisely the ones that are destroyed first when the inner ear is afflicted by disease.

The abnormal sensation of loud sounds can be verified at the hearing examination. The change in the ear's amplifying function leads to sound distortion which make it difficult for the patient to understand many words, mainly because the difference between unstressed and stressed syllables in a word is dramatically increased. In medical terminology the abnormal amplification phenomenon is called *recruitment of loudness* and the difficulty in understanding words is called *reduced intelligibility*.

As the Ménière patient's hearing changes another unpleasant phenomenon occurs in the ear, tinnitus. All sounds in the ear which do not originate from outside sound sources in the surrounding environment, are referred to by doctors and others as tinnitus, whether there is a buzzing, roaring, hissing, ringing or whatever it sounds like.

In the long run, tinnitus is very often the symptom which causes the Ménière patient the most discomfort. For people who do not suffer from tinnitus, it can be hard to imagine precisely how much of a mental strain it is to constantly have a foreign sound in one's ear. The sensation may be compared to a tap which is dripping. If this becomes an annoyance when one is trying to fall asleep, the noise of the drop hitting the sink gradually assumes unrealistic proportions. The same is true for tinnitus - the difference being that you cannot replace a defective washer in the inner ear and it gives no relief to

place a pillow over one's head. It takes a very strong psyche to learn to live with one's tinnitus.

We have no medicine or other treatment which can reduce tinnitus. We only know of one thing that works: "drowning" it. Consequently, many tinnitus patients like to listen to soft music. Using a hearing aid can also help - of course this is especially useful if the patient also has a hearing loss. It may be quite difficult to get used to wearing a hearing aid if the other ear has normal hearing. Most important of all, tinnitus patients must learn to live with their disease, just like others must learn to live with having lost a leg.

With many Ménière patients the attack of dizziness starts with the occurrence and increase of tinnitus, or the hearing loss getting worse. It is sometimes helpful to get such a "warning" before the attack of dizziness. In the good periods between attacks, tinnitus is often reduced and hearing is better than during bad periods.

Other Ménière Symptoms

Many Ménière patients have a sensation of fullness, sometimes even pain, in the sick ear previous to the attacks of dizziness. They feel that something is "brewing" in the sick ear. This may also occur during the attack.

In connection with attacks, some Ménière patients have symptoms similar to migraine. Prosper Ménière believed that the disease, which later came to bear his name, could be related to migraine. Sometimes the doctor may be in doubt as to whether a patient suffers from a special type of migraine (basilar migraine) or from Ménière's disease. Sometimes we even see that a migraine patient suddenly stops having attacks of headache and instead has attacks of dizziness and hearing symptoms (this is often the case with women having their menopause). There is no doubt that some of the patients with

symptoms similar to the ones we find with Ménière's disease (especially those who do not develop a chronic hearing loss) in actual fact suffer from some kind of migraine of the inner ear. There is a possibility that we are drawing too fine a line between the two diseases.

The Course of Ménière's disease

The disease often begins with attacks of dizziness, as described above. With other patients, the disease starts with changes in the patient's hearing, and with tinnitus. We only know for sure that we are dealing with a case of Ménière's disease when a patient has attacks of dizziness, hearing loss (of the type described above) and tinnitus (perhaps only during attacks). As the disease starts in different ways with different patients, it may take quite some time before the diagnosis is certain. Great effort is being put into developing methods of investigation which should enable doctors to arrive at the final diagnosis much quicker than today. Even though the disease itself is very unpleasant, it may be just as unpleasant not to have an explanation, e.g. of severe attacks of dizziness. It is also of great importance for treatment that the doctor is absolutely sure that the diagnosis is correct.

When a patient has had Ménière's disease for some time, the hearing loss will increase in the sick ear and there is a tendency for the attacks of dizziness to become less frequent and less severe. With most patients the attacks of dizziness disappear completely after a number of years, as the balance nerve function in the sick ear degenerates, but also does the auditory nerve function. Most people get along fine without the balance nerve function in one ear and are therefore mainly bothered by the hearing loss and tinnitus. Unfortunately, it is impossible to predict the course of Ménière's disease for an individual patient. Often the disease starts "at full tilt" and

then suddenly disappears - only to return perhaps 5 or 10 years later. While the dizziness gradually "fades away" as far as the attacks are concerned, the hearing loss has usually become so severe that the patient considers the ear to be deaf (even though there may be considerable tinnitus still experienced.)

Formerly, it was believed to be an exception if patients developed the disease in both ears. This was thought only to happen to about 5 or 10% of all Ménière patients. Unfortunately this is not the full truth. When we follow a group of Ménière patients for many years, we see that almost half of the patients also develop a hearing impairment, and very often tinnitus, in the other ear. In some of those patients the other ear seems to cause attacks of dizziness too. Only in those cases do the patients suffer from bilateral Ménière's disease.

Often, when patients get the disease in the other ear, the attacks of dizziness are not as severe. For these patients it is characteristic that they have quite severe balance problems between attacks. In order to keep their balance, these patients end up walking almost like a person who is drunk. They spread their legs in an uncertain gait, needing now and then an extra little step to one or the other side, in order to keep their balance. Some patients have the terrible experience of seeing other people "coming to understand what's wrong" (What a shame! I knew Jones had a glass or two once in a while, but an alcoholic! And then a hearing loss on top of it - what a pity! Too bad about his wife and children...etc.).

Ménière's disease and stress

Most Ménière patients report having their attacks - and having them more frequently and more severely - whenever they are under stress at work or are having personal problems. There is no doubt that this is true.

But the opposite is also true. Having Ménière's disease means having to live with the fact that you can without warning feel severely dizzy - and perhaps especially when something exciting is about to happen. The fact that you sometimes cannot stand on your feet for hours, you get sick and throw up, you must live with a hearing loss and at the same time have a constantly hissing, buzzing or roaring devil inside your ear - probably constitutes the worst kind of stress to which a mentally sound person can be subjected. If one is going through a bad period, it can be difficult to keep pace at work and to fulfil one's family duties. All this very easily develops into a vicious circle: you get stress from your disease and you become ill from your stress.

Treatment of Ménière's disease

The disease can safely be cured by no known medicine and no known operation. But this does not mean that a doctor cannot do a great deal for his Ménière patients.

It is my personal opinion that the doctor's point of departure should be an attempt to break the vicious circle of stress. Such a process begins by allocating the necessary time to listen to the patient's complaints, examining the patient's hearing and balance function thoroughly, and explaining to the patient the exact nature of Ménière's disease. Both parties must accept that it may take time (sometimes many months) to diagnose correctly and to find the type of treatment, which helps this specific patient.

The ideal treatment of any disease, of course, is to remove the cause of the disease. But unfortunately, it is quite rare that we are able to establish for the individual Ménière patient a direct cause, and it is even more rare that we can remove the cause. (In rare cases, a patient is able to tell us that he or she has an attack after having eaten specific types of food.) Usual-

ally we are in the situation where the aim of the treatment is to reduce or eliminate a certain symptom. The most important symptom to target is almost always the attacks of dizziness. If we succeed here we have also broken the vicious stress circle, and thus intervened on an important disease mechanism.

In the treatment of Ménière's disease we can also try to affect another well-known disease mechanism. For many years it has been known that with Ménière patients there is an increased pressure in the inner ear fluid (labyrinthine hydrops).

The structure of the inner ear is highly complex (see figure 1).

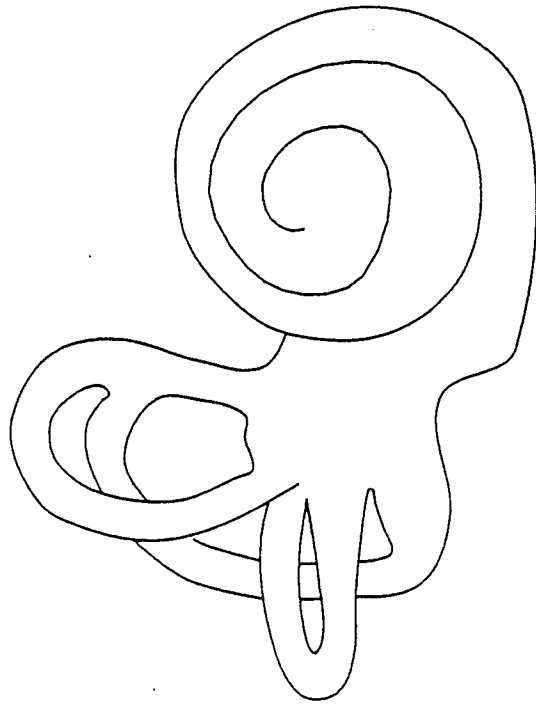


Figure 1. The inner ear, its semicircular canals and the cochlea.

In principle, the sensory cells for hearing and balance are located inside a membrane which contains a fluid (the endolymph) which is rich in potassium chloride. This structure is called the *membranous labyrinth*. The membranous labyrinth, like the Chinese boxes, is located inside the *bony labyrinth* which is a complex structured cavity in the temporal bone (with the cochlea (the hearing organ), the semicircular canals and the cavity (vestibulum) which contains the otolithic organs utricle and saccule). Between the membranous labyrinth and the bony wall in the bony labyrinth there is another fluid (the perilymph) which is rich in sodium chloride (see figure 2). If we examine the temporal bone from the body of a person who has suffered from Ménière's disease, we find a dilation of the membranous labyrinth as if there has been increased pressure in the endolymph. (In reality, it may be due to insufficient pressure in the perilymph as well.)

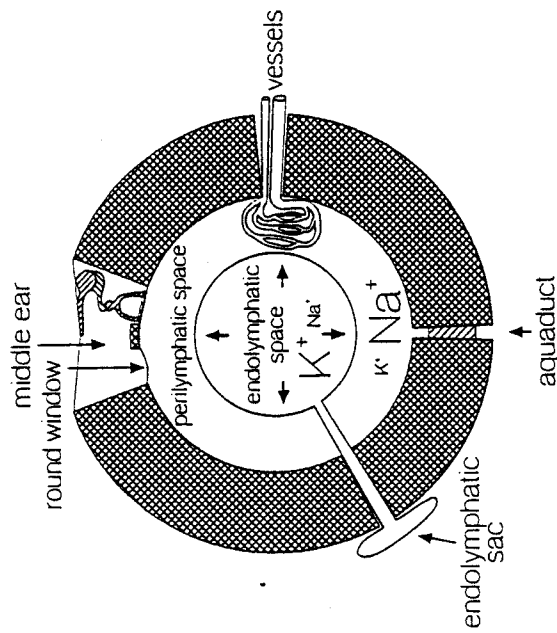


Figure 2. The system of membranes and fluids of the inner ear and its bony surroundings.

It is believed that the attacks of dizziness in connection with Ménière's disease are caused by the membranous labyrinth bursting from time to time, so that potassium chloride and sodium chloride mix and cause paralysis of the sensory cells in the semicircular canals, the otolithic organs and in the cochlea. Part of the changes in the patient's hearing and the occurrence of tinnitus may be due to the dilation of the membranes in the cochlea; the more permanent changes are due to a degeneration of the sensory cells in the hearing organ and the balance organ.

Most of the treatments suggested for Ménière patients aim at reducing the inner ear pressure. The simplest way of doing that is to reduce the consumption of salt and take diuretic medicine. This will help some patients, but far from all.

Other patients can be helped by taking a drug, betahistina (SercTM). It is believed that this drug increases the blood circulation in the labyrinth, thereby reducing the pressure in the ear.

The most widespread surgical method in the treatment of Ménière's disease also has the aim of relieving the pressure in the inner ear. Doctors refer to this operation as *sacotomy* or *sac-mastoid shunt surgery*.

The membranous labyrinth has a small bulge (the endolymphatic sac, see figure 2) situated behind the temporal bone between the bone and the thick brain membrane (dura mater). By drilling a cavity in the temporal bone behind the ear (which is frequently done in cases of chronic middle ear disease) it is possible to access the bulge and open it in order to drain the endolymph out into the bared part of the temporal bone. This reduces the pressure in the membranous labyrinth and will prevent increases in the endolymph pressure from bursting the membranes.

For all of the methods of treatment mentioned above, we cannot be *sure* they work on *all* Ménière patients.

If we are very skeptical and carefully deliberate the various methods and their presumed effects in a very scientific way, we easily arrive at the conclusion that we do not know for certain why the methods work, even though the immediate explanations may seem logical.

If a strategy of treatment is selected, the sole purpose of which is to treat the symptoms of Ménière's disease in order to break the vicious stress circle, the first thing to concentrate on is the elimination or alleviation of the attacks of dizziness. It is often possible to alleviate these attacks by using some of the drugs normally used against seasickness.

The most widespread of these drugs are those with minimal sedative effect, such as cinnarizine, cyclizine and meclizine. These drugs are more or less equally efficient, but the sedative effect as well as the effect on the dizziness can vary considerably from one patient to the next. It is, therefore, very wise to consult one's doctor before using any of these drugs. Furthermore, it is noteworthy that the new non-sedative drugs in this group of so-called antihistamines have no effect whatsoever on seasickness and Ménière's disease.

Another method to prevent attacks of dizziness is simply to cut the balance nerve in the sick ear. This is a difficult operation to perform. In some cases the same effect can be achieved by carefully injecting a toxic substance (streptomycin or gentamycin) into the middle ear. Especially for this operation, but also for the sacotomy procedure, we usually insist that all known drug treatments have been attempted without success before such a procedure is considered.

Formerly surgery was performed whereby the Ménière patient's entire labyrinth was destroyed. This implied destroying both the sense of balance and of hearing on that ear. When doctors came to appreciate the risk of contracting the disease also on the other ear, most surgeons ceased to perform this type of operation, in order to make possible a later use of a hearing aid on the operated ear.

The surgical methods used today take all this into account to the extent that it is an exception that the operated ear becomes deaf as a result of an operation.

An important element of the Ménière treatment is *hearing rehabilitation using hearing aids*. In this connection there are two difficulties.

As the typical Ménière's disease ear is very sensitive to noise due to the recruitment of loudness phenomenon, it can be difficult to use a hearing aid which not only amplifies useful sound signals (such as speech), but unfortunately, also amplifies background noise. Many modern hearing aids have an integrated limiter which is able to regulate the maximum output from the hearing aid. This solution is not ideal, but it has enabled many Ménière patients to benefit from a hearing aid. In the near future computerized hearing aids may solve this problem.

Another difficulty is the fact that the recruitment phenomenon also implies a significant sound distortion - and the hearing aid can do nothing about this. If the good ear hears normally, amplification of the distorted sound in the sick ear will affect total hearing ability negatively. Many patients actually prefer using an earplug in the sick ear (for example when listening to music) in order to reduce the distortion.

For most patients, the use of a hearing aid has the advantage of reducing the inconveniences of tinnitus.

All in all, the use of a hearing aid is a benefit to Ménière patients in most cases. At the same time however, just like drugs and surgical treatment, it must be individualized because the effect can vary from one patient to the other - and from time to time with the individual patient.

Many Ménière patients feel they need "nerve medicine", for example diazepam (Valium™ and similar drugs). These drugs can break the vicious stress circle, but they have the adverse effect of sedation and possible habituation of the patient to the drug. Therefore, such treatment requires a continued and trustful relationship between the patient and the doctor in order for both parties to be able to decide on the right time for ending the treatment.

As can be seen from the previous pages, authorized medical treatment is not always satisfactory and is always difficult. Inevitably, this leads many patients to seek alternative treatment - a field having its main success in areas where doctors cannot offer satisfactory treatment.

Although it cannot be dismissed that some patients (at least for short periods of time) have experienced benefits from alternative treatment - perhaps because the treatment has succeeded in breaking the vicious stress circle - no one today will reasonably claim that the solution to the treatment of Ménière can be found in this field.

The same is true for all the health maxims and health recommendations emanating from all corners of society. Obviously the Ménière patient must also find a healthy lifestyle. It is always wise to avoid the misuse of all kinds of intoxicants; but it must be up to the individual patient to define the notion of "misuse" based on his or her own experience and attitude to life.

What is true of intoxicants is equally true of food; an unbalanced diet over-emphasizing certain types of food (regardless of whether these are being classified as healthy or detrimental by the present health gurus) is a question of the patient's attitude to life and is beyond any form of medical science, no matter how much science is invoked in the public debate.

Epilogue:

It is the author's hope that the preceding pages will be read by the largest possible number of Ménière patients, their families and friends and others who - at work or elsewhere - are in contact with people who suffer from this troublesome disease.

It is my wish to contribute to the educational work being carried out by Ménière Patient Associations towards patients, relatives and public authorities. It is of great importance that especially social authorities come to understand that there is a large group of apparently healthy and able-bodied people who suffer from a disease which implies considerable problems in their social, professional and private lives.