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## DISORDERS

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### ENDOLYMPHATIC HYDROPS

Ménière's disease results in an abnormally large buildup of fluid called endolymph in the inner ear.

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## ARTICLE

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# Ménière's Disease

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Ménière's disease is a chronic, incurable vestibular (inner ear) disorder that produces a recurring set of symptoms as a result of abnormally large amounts of a fluid called endolymph collecting in the inner ear. This buildup of endolymph fluid, also known as endolymphatic hydrops, is considered to be the pathological mechanism behind Ménière's disease. The exact cause of Ménière's disease and its symptoms are not yet known. It may start with fluctuating hearing loss, eventually progressing to attacks of vertigo and dizziness. No treatment currently exists to cure Ménière's disease. However, treatments exist that can lessen symptoms of vertigo. There are no treatments for permanent hearing loss.

In 1861 the French physician Prosper Ménière theorized that attacks of vertigo, ringing in the ear (tinnitus), and hearing loss came from the inner ear rather than from the brain, as was generally believed at the time. Once this idea was accepted, the name of Dr. Prosper Ménière began its long association with this inner ear disease and with inner ear balance disorders in general.

### WHAT IS MÉNIÈRE'S DISEASE?

Ménière's disease is a chronic, incurable vestibular (inner ear) disorder defined in 1995 by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology–Head and Neck Surgery as “the idiopathic syndrome of endolymphatic hydrops.”<sup>1</sup> This means that Ménière's disease, a form of endolymphatic hydrops, produces a recurring set of symptoms as a result of abnormally large amounts of a fluid called endolymph collecting in the inner ear.

Ménière's disease can develop at any age, but it is more likely to happen to adults between 40 and 60 years of age. The exact number of people with Ménière's disease is difficult to measure accurately because no official reporting system exists. Numbers used by researchers differ from one report to the next and from one country to the next. The UK-based Ménière's Society reports that the incidence of Ménière's disease is between 1:1000 and 1:2000 of the population; depending on the source, and about 7-10% of those affected have a family history of the condition.<sup>2</sup> The National Institutes of Health estimates that about 615,000 people in the U.S. have Ménière's disease and that 45,500 new cases are re-diagnosed each year.<sup>3</sup>



## CAUSES

The exact cause and reason why Ménière's disease starts is not yet known. Many theories have been proposed over the years. They include circulation problems, viral infection, allergies, an autoimmune reaction, migraine, and the possibility of a genetic connection.

Experts aren't sure what generates the symptoms of an acute attack of Ménière's disease. The leading theory is that they result from increased pressure of an abnormally large amount of endolymph in the inner ear and/or from the presence of potassium in an area of the inner ear where it doesn't belong. These conditions may be due to breaks in the membrane separating endolymph from the other inner ear fluid, perilymph. Some people with Ménière's disease find that certain events and situations, sometimes called triggers, can set off attacks. These triggers include stress, overwork, fatigue, emotional distress, additional illnesses, pressure changes, certain foods, and too much sodium or salt in the diet.

## DIAGNOSTIC CRITERIA

The Bárány Society established the following diagnostic criteria for Ménière's disease. The following criteria must be met for the diagnosis of Ménière's disease<sup>4</sup>:

Two or more spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours.

Audiometrically documented low- to medium frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during, or after one of the episodes of vertigo.

Fluctuating aural symptoms (hearing loss, tinnitus or fullness) in the affected ear.

Not better accounted for by another vestibular diagnosis.

## PROGRESSION OF SYMPTOMS

Common symptoms of a Ménière's disease attack do not reflect the entire picture of the disorder, because symptoms vary before, during, between, and after attacks, and also during the late stage of Ménière's disease.

Ménière's disease may start with fluctuating hearing loss, eventually progressing to attacks of vertigo and dizziness.

Oncoming attacks are often preceded by an "aura," or the specific set of warning symptoms listed below. Paying attention to these warning symptoms can allow a person to move to a safe or more comfortable situation before an attack:

balance disturbance

- dizziness, lightheadedness
- headache, increased ear pressure
- hearing loss or tinnitus increase
- sound sensitivity
- vague feeling of uneasiness

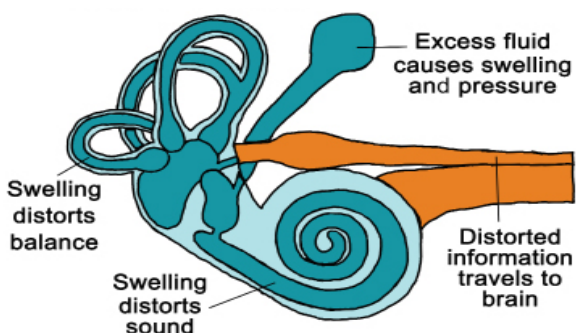
During an attack of early-stage Ménière's disease, symptoms include:

spontaneous, violent vertigo  
fluctuating hearing loss  
ear fullness (aural fullness) and/or tinnitus

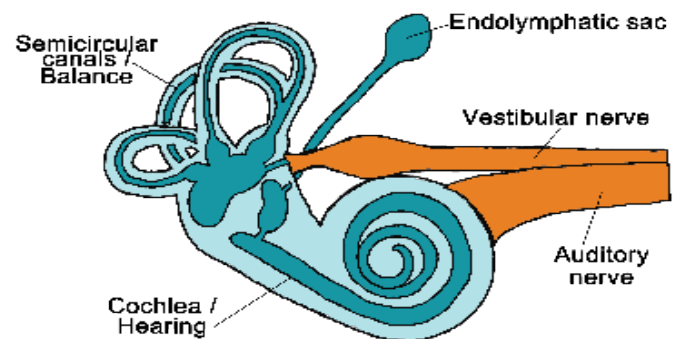
In addition to the above main symptoms, attacks can also include:

- anxiety, fear
- diarrhea
- blurry vision or eye jerking

**Ménière's Disease Ear**



**Healthy Inner Ear**



- nausea and vomiting
- cold sweat, palpitations or rapid pulse
- trembling

Following the attack, a period of extreme fatigue or exhaustion often occurs, prompting the need for hours of sleep.

The periods between attacks are symptom free for some people and symptomatic for others. Many symptoms have been reported after and between attacks:

- anger, anxiety, fear, worry
- appetite change
- clumsiness
- concentration difficulty, distractibility, tendency to grope for words
- diarrhea
- fatigue, malaise, sleepiness
- headache, heavy head sensation
- lightheadedness (faintness)
- loss of self-confidence and self-reliance
- nausea, queasiness, motion sickness
- neck ache or stiff neck
- palpitations or rapid pulse, cold sweat
- sound distortion and sensitivity
- unsteadiness (sudden falls, staggering or stumbling, difficulty turning or walking in poorly lit areas, tendency to look down or to grope for stable handholds)
- vision difficulties (problems with blurring, bouncing, depth perception, glare intensification, focusing, watching movement; difficulty looking through lenses such as binoculars or cameras)
- vomiting

Late-stage Ménière's disease refers to a set of symptoms rather than a point in time. Hearing loss is more significant and is less likely to fluctuate. Tinnitus and/or aural fullness may be stronger and more constant. Attacks of vertigo may be replaced by more constant struggles with vision and balance, including difficulty walking in the dark and occasional sudden loss of balance. Sometimes, drop attacks of vestibular origin occur in this stage of Ménière's disease and are characterized by sudden brief loss of posture without loss of consciousness. Some of these late-stage symptoms can become more problematic in conditions of low lighting, or with fatigue, or when a person is exposed to visually

stimulating situations.

### **DURATION AND FREQUENCY OF ATTACKS**

Attacks can last from 20 minutes to 24 hours. They can occur with the frequency of many attacks each week or they can be separated by weeks, months, and even years. The unpredictable nature of this disease makes managing it challenging. It also complicates the ability of scientists and physicians to study it.

### **IS THERE A CURE?**

To "cure" a disease means to eliminate the root cause of the disease and reverse the damage it has inflicted (on the inner ear, in this case). No treatment currently exists to cure Ménière's disease. However, medical treatments exist that can help manage it.

### **TREATMENT**

Existing treatments fall into two categories. Some treatments aim at reducing the severity of an attack while it is occurring, while other treatments attempt to reduce the severity and number of attacks in the long term. Experts feel these medical treatments provide some degree of improvement in 60-80% of the treated people. 5 Gentamicin is > 80% effective at controlling vertigo.

The most conservative long-term treatment for Ménière's disease in the U.S. involves adhering to a reduced-sodium diet and using medication that helps control water retention (diuretics or "water pills"). The goal of this treatment is to reduce inner-ear fluid pressure. Some physicians, more commonly outside of the U.S., also weigh the potential efficacy of using betahistine HCl (Serc) as a vestibular suppressant for Ménière's disease.<sup>6</sup>

Medications can be used during an attack to reduce the vertigo, nausea/vomiting, or both. Some drugs used for this include diazepam (Valium), lorazepam (Ativan), promethazine (Phenergan), dimen-hydrinate (Dramamine Original Formula), and meclizine hydrochloride (Antivert, Dramamine Less Drowsy Formula).

Vestibular rehabilitation therapy is sometimes used to help with the imbalance that can plague people between attacks. Its goal is to help retrain the ability of the body and brain to process balance information. When successful, this can help a person regain confidence in the ability to move about.

When conservative treatments don't work



For the 20-40% of people who do not respond to medication or diet, a physician may recommend a treatment that involves more physical risk. One such method, intratympanic gentamicin, destroys vestibular tissue with injections into the ear of the aminoglycoside antibiotic gentamicin. Recently, intratympanic steroid injections have been used with less risk of hearing loss and persistent imbalance.

Another less conservative treatment method involves surgery. Two categories of surgery are available. The goal of the first type is to relieve the pressure on the inner ear. Surgery to reduce pressure is not as widely used now as it was in the past due to questions about its long-term effectiveness.

The goal of the second type of surgery is to block the movement of information from the affected ear to the brain. The process involves either destroying the inner ear so that the ear does not generate balance information to send to the brain or destroying the vestibular nerve so that balance information is not transmitted to the brain. In either instance, physical therapy is useful to help the brain compensate for the loss of inner ear function due to surgery.

### PROGNOSIS

It is difficult to predict how Ménière's disease will affect a person's future. Symptoms can disappear one day and never return. Or they might become so severe that they are disabling.

### COPING

Coping with Ménière's disease is challenging because attacks are unpredictable, it is incurable, some of the symptoms are not obvious to others, and most people know virtually nothing about the disorder. Many people with Ménière's disease are thrust into the role of educator—they must teach themselves, their family, friends, coworkers, and sometimes even health care professionals about the disorder and how it impacts them.

Key features of communicating with family and friends include informing them about what might happen with the onset of an acute attack and how they can help. If a low-sodium diet is effective, family and friends should be informed about how important it is for them to support adherence to the diet regimen. Changes in lifelong eating patterns can be easier with the assistance of others.

Managing an acute attack involves preparation. This includes consulting with a physician about any

appropriate drugs that can be taken when an acute attack occurs and deciding ahead of time when it is appropriate to go to a hospital. During an attack, it is helpful to lie down in a safe place with a firm surface and avoid any head movement. Sometimes keeping the eyes open and fixed on a stationary object about 18 inches away is helpful. To control dehydration, a doctor should be called if fluid intake is not possible over time due to persistent vomiting.

After an acute attack subsides, it is not uncommon to want to sleep for several hours. Resting in bed for a short time is appropriate if the person is exhausted. But it is also important for the person to get up and move around as soon as possible so that the brain readjusts to the changed balance signals. Precautions need to be taken in this process to accommodate any new balance sensations.

Successfully coping with symptoms involves understanding the disease. Talking with health care providers, communicating with other people who are experiencing the same disease, and reading books and articles about the topic are all helpful methods of learning more about Ménière's disease.

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