Ménière’s Disease

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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.”¹ In plain language, 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 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.²

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

ENDOLYMPHATIC HYDROPS

Menière’s disease results in an abnormally large buildup of fluid called endolymph in the inner ear.
stress, overwork, fatigue, emotional distress, additional illnesses, pressure changes, certain foods, and too much salt in the diet.

**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
- 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, quiesceness, 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 (Tumarkin’s otolithic
3 crisis) 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; some 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. Gentamicin is > 80% effective
at control of 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.

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),
dimenhydrinate (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,
destroy 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 from 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. In order 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.

**REFERENCES**


**FURTHER READING**

Some helpful documents available from the Vestibular Disorders Association include the following:

- Secondary Endolymphatic Hydrops
- Dietary Considerations with Endolymphatic Hydrops, Ménière’s Disease, and Vestibular Migraine
- Surgery for Peripheral Vestibular Disorders
- Vestibular Rehabilitation: An Effective, Evidence- Based Treatment

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