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Otolaryngology: Menieres Disease

Ménière's disease is a disorder of the inner ear which causes spells of vertigo, fluctuating hearing loss, tinnitus (noises) and fullness in the ear. Ménière's disease usually affects one ear, but it may affect both. Ménière's disease affects women more than men, and usually affects adults.

SPECIALISTS WHO TREAT:

Richard A. Chole, M.D., Ph.D. Joel A. Goebel, M.D., FACS Timothy E. Huller, M.D.

Symptoms of Ménière's disease

Symptoms may vary from person to person, but they are generally in four categories:

- 1. Vertigo Spells: The typical spells of vertigo in Ménière's disease are the rapid onset of a sensation of movement, very often a spinning movement. These spells may last from moments to hours, and sometimes a feeling of imbalance persists for hours or days. The vertigo spells may be mild or severe. Some spells are so severe that the patient can fall to the ground without loss of consciousness. (These are "drop attacks"). The vertigo of Ménière's disease may also cause nausea and vomiting.
- 2. Fluctuating Hearing Loss: Typically, Ménière's disease will cause fluctuation of the hearing in one ear. In some cases, the hearing may fluctuate at the time of a Ménière's attack, but it may also fluctuate at other times. Most fluctuation of hearing in patients with Ménière's disease is noticed for low sounds of frequency. Low frequency sensorineural (nerve type) hearing loss is typical of Ménière's disease.

An ear affected by Ménière's disease also may exhibit another rather peculiar symptom. The ear may not be sensitive to the quietest sounds, but be overly sensitive to loud sounds (auditory recruitment). Some people with Ménière's perceive a different tone in each ear when listening to a single tone (diplacusis). Many patients with Ménière's have dizziness when exposed to loud noise (Tullio's phenomenon).

3. Tinnitus: Tinnitus (inner ear or head noise) is common in patients with Ménière's disease. The perceived sound of the tinnitus can vary in pitch and intensity, and can vary from time to time. Sometimes changes in tinnitus warn of an upcoming attack of vertigo. Tinnitus can be high-pitched like a siren, or a low-pitched rumble.

Tinnitus is not a real sound, and cannot be heard by other people, nor can it be measured. The sound sensation seems to come from a hypersensitivity or "irritation" of the auditory nerve, which is perceived as sound. In extreme cases, tinnitus may continue after the nerve cells are permanently damaged, comparable to sensations of a "phantom limb" after amputation.

4. Ear Fullness: Most patients with Ménière's disease experience fullness in the affected ear. This fullness is sometimes described as a blocked feeling, or even pressure or pain. The fullness seems to fluctuate from time to time, and may even correspond to a vertigo attack. Altitude changes are sometimes aggravating.

Cause of Ménière's Disease

The cause of Ménière's disease is unknown. A doctor named Prosper Ménière first noticed a case and wrote about it in the 1870's. He recognized that it came from the inner ear. We now recognize several more things about Ménière's disease.

- Genetic origin. In about 20% of the cases, another member of the family has the disease. Therefore, we think that there is a genetic factor in the disease.
- Fluid inbalance. We also know that certain things happen in the inner ear with Ménière's disease that give us some insight into the cause. There are two different clear fluids in the inner ear, perilymph and endolymph. They are separated from one another by a membrane (Reissner's membrane). In patients who have had Ménière's disease, we find that the endolymph space is much larger than normal. This is called endolymphatic hydrops. Therefore, we assume

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that Ménière's disease is an abnormality of the fluids and fluid balance within the inner ear, much as glaucoma is an abnormality of the fluids of the eye.

The overabundance of endolymph could be caused by over-production, or a defect in absorption of the fluid. Most scientific evidence points to a failure of absorption of the fluid. Ménière's disease attacks are often more severe when fluid and salt retention occurs, further supporting the idea that this is a fluid balance disorder.

There is also some evidence that the fluid balance in the inner ear may be controlled by immune factors in the inner ear. Inner ear antibodies have been detected in some patients with Ménière's disease, and some doctors believe there is an association with allergy.

Treatment for Meneire's Disease

Meneire's disease can be treated by non-surgical and by surgical methods, depending on the individual situation.

Nonsurgical methods of treating Ménière's disease

- 1. A Low Salt Diet is recommended for Ménière's disease. Sodium intake should be restricted to 1500 mg per day. To achieve this level of salt restriction, one must avoid adding salt to foods and read the labels of prepared foods carefully. Many commercially prepared foods contain large amounts of sodium.
- 2. Avoidance of Caffeine and Nicotine. Both of these substances are thought to worsen Ménière's disease by an effect on blood vessels and circulation.
- **3. A Diuretic** will often be recommended for Ménière's patients. Diuretics help by increasing sodium loss from the body, changing salt-regulating hormone levels which may affect fluid balance in the ear. Some diuretics may also affect inner ear fluids directly.

The first line of diuretics are those containing hydrochlorothiazide, and the most commonly prescribed is Dyazide®. The next line of diuretics are the carbonic anhydrase inhibitors Diamox® and Neptazane®. These are drugs that are commonly used for glaucoma of the eye, and may be effective in some cases of Ménière's disease. These drugs have some side effects, including tingling of the fingers and toes and changes in electrolyte balance of the blood. There are also some other rare complications of this class of drugs.

- **4. Intra-Tympanic Steroids:** Steroids can be oral or injected behind the eardrum.
- **5. Intra-Tympanic Treatment with Gentamicin.** In recent years it has been found that putting the antibiotic gentamicin into the middle ear of Ménière's patients can help alleviate their vertigo.

Gentamicin acts by entering the inner ear and damaging some of the balance detectors (vestibular hair cells), reducing the sensitivity of the balance system. Gentamicin is applied to the ear that is causing the vertigo by injecting it through a small hole in the eardrum with a tiny needle. The hole heals up rapidly after the injection. Within a day or two, the patient may experience vertigo which gradually improves. The treatment may be repeated several times at intervals to completely stop the Ménière's attacks.

A difficulty with this procedure is that each individual responds differently, and every injection of gentamicin may lead to a new vertigo attack. The vertigo may take weeks or months to completely subside. There is also a risk to the hearing with Gentamicin injections which increases with the number of injections given.

Surgical methods of treating Ménière's disease

1. Endolymphatic Shunt. The endolymphatic shunt procedure is a surgical procedure done from behind the ear through the mastoid bone. A small out pouching of the inner ear, the endolymphatic sac, is opened and a small drain tube is put in place. The idea of this operation is that any buildup of fluid of the endolymph will be relieved by this small tube. The amount of endolymph in the inner ear is far less than a drop, so the amount produced is minuscule.

The risks of the operation are low. The procedure is done as an outpatient and there is no vertigo afterwards. There is a one out of 200 chance of hearing loss from the operation.

The endolymphatic shunt operation relieves vertigo attacks in 50-60% of patients over the long term. However, there is little effect on hearing. This operation is often chosen as the first operation to do for disabling Ménière's disease since it carries such low risk.

2. Vestibular Neurectomy. Vestibular neurectomy is a procedure in which the balance nerve (vestibular nerve) in the affected ear is severed. This operation is

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done either above the ear or behind the ear in the fluid spaces around the brain (cerebrospinal fluid). The operation is an extremely delicate operation which takes three to four hours to perform.

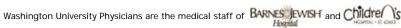
After the operation there is usually significant vertigo for a period of time, and the length of hospitalization is usually about four days. The vertigo from this operation gradually improves over weeks to months, but it is highly successful in ridding a patient of vertigo attacks.

About 95% of patients who have this operation have no more vertigo, although some may continue to experience disequilibrium or imbalance after the procedure. The hearing is not improved by this procedure, and there is a five percent risk of further hearing loss.

Additionally, since the vestibular nerve is next to the facial nerve, there is a small chance of facial nerve weakness from the procedure, which is always temporary. The facial nerve causes movement of the muscle of facial expression and eye closure.

3. Labyrinthectomy. Labyrinthectomy is a procedure in which the inner ear is completely removed. The procedure is done from behind the ear, and all the balance canals are removed.

This procedure is only done for patients who have no usable hearing in the ear, since there is no hearing left after this procedure. This procedure is 95% successful in stopping the attacks of vertigo. The procedure usually results in extreme vertigo for a few days which gradually subsides over weeks to months. This procedure requires a day or two of hospitalization, but it does not require opening the area of the spinal fluid. Additional information is available at the Department of Otolaryngology, Washington University School of Medicine at Ménière's disease.



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