



ENDOLYMPHATIC HYDROPS MENIERE'S DISEASE

Introduction:

Meniere's Disease, or endolymphatic hydrops, is a disorder of the inner ear. This condition occurs because of abnormal fluctuations in the inner ear fluid called endolymph. A system of membranes, called the membranous labyrinth, contains a fluid called endolymph. This fluid bathes the inner ear balance and hearing system sensory cells and allows them to function normally. The membranes can become dilated like a balloon when pressure increases. This is called "hydrops". The amount of fluid is normally kept constant by altering the production and absorption of the fluid. Endolymph also contains a specific concentration of sodium, potassium, chloride, and other electrolytes.

If the inner ear is damaged by disease, injury, or other causes, the volume and composition of the inner ear fluid can fluctuate with changes in the body's fluid and electrolyte levels. This fluctuation in inner ear fluid can cause the symptoms of hydrops, including pressure or fullness of the affected ear, tinnitus, hearing loss, and imbalance or dizziness. Treatment of this condition is geared towards stabilizing the body fluid levels so that fluctuations in the endolymph volume can be avoided.

Meniere's episodes may occur in clusters in which several attacks may occur within a short period of time. On the other hand, years may pass between episodes. Between the acute attacks, most people are free of symptoms or note mild imbalance, tinnitus, and/or hearing loss. Meniere's affects roughly 0.2% of the population, about 200 out of 100,000 people (or in other words, 2/1000). The majority of people with Meniere's disease are over 40 years of age, with equal distribution between males and females. Symptoms start in one ear, but in about 17% of patients the other ear may become involved over many years. About 75% of patients destined to have both ears affected present within 5 years.

Causes:

Meniere's Disease is thought to occur because of obstruction of endolymphatic outflow at the endolymphatic duct level, increased production of endolymph, or reduced absorption of endolymph caused by a dysfunctional endolymphatic sac. Causes of endolymphatic hydrops include viral source, hereditary disposition, autoimmune disorders, or traumatic injury. Most commonly, the cause is unknown.

Testing:

Diagnosis can frequently be made by historical information and physical examination but several tests can be useful in not only establishing the diagnosis but also in eliminating other possible disorders. Standard audiometric testing will show the characteristic hearing loss when the patient is experiencing aural symptoms. Basic and complete balance tests such as videonystagmography (VNG) typically are abnormal but can show a variety of abnormalities.

A specific test for Meniere's disease is electrocochleography (ECoG). In this test electrical information is recorded from the inner ear as sound is presented. Normal patients have a characteristic shaped electrical response and in Meniere's disease this response is altered.

Treatment of Meniere's Disease:

The goal of treatment is to stabilize your bodily fluids so that changes in volume of the endolymphatic space will not occur. The amount and composition of the inner ear fluid is affected by the salt and sugar concentrations of your blood and other body fluids. An altered diet is required as part of treatment for this condition.

Hydrops Diet:

Inner ear fluid is usually independent of the body's overall fluid/blood system. When endolymphatic hydrops affects an inner ear, independent control is lost, and the volume and concentration of the inner ear fluid fluctuates with changes in the body's fluid/blood. This fluctuation causes the symptoms of hydrops--pressure or fullness in the ears, tinnitus (ringing in the ears), hearing loss, dizziness and imbalance.

Your inner ear fluid is influenced by certain substances in your blood and other body fluids. For instance, when you eat foods that are high in salt or sugar, your blood level concentration of salt or sugar increases, and this, in turn, will affect the concentration of substances in your inner ear.

People with endolymphatic hydrops must control the amount of salt and sugar that is added to food. You must also become aware of the hidden salts and sugars that foods contain. Limiting or eliminating your use of caffeine and alcohol will also help to reduce symptoms of dizziness and ringing in the ears.

Dietary goals:

The goal of treatment is to provide stable body fluid/blood levels so that secondary fluctuations in the inner ear fluid can be avoided. We recommend that patients avoid skipping meals and attempt to eat similar amounts during each meal during the day. It is critical that you drink plenty of water, especially if you are started on a diuretic (water pill). High salt foods should be avoided. Clearly, no salt should be added to meals, however it is of utmost importance that you select foods with low sodium content. High salt intake results in fluctuations in the inner ear fluid and may increase the symptoms

that you are experiencing. We recommend a diet with less than 1.5 gram sodium intake per day.

Caffeine-containing food and drinks, such as coffee, tea, and chocolate, should be avoided. The stimulant effects of caffeine may make symptoms worse and may increase the tinnitus. Monosodium Glutamate (MSG), often present in Chinese food and prepackaged products, should not be consumed. Alcoholic drinks should also be avoided.

Drug Considerations:

As mentioned above, caffeine should be avoided and this is found in certain medical formulations. Aspirin and non-steroidal anti-inflammatory (NSAIDs) agents should not be consumed when this is possible, as they may exacerbate tinnitus. Nicotine should not be used in any form, including cigarettes and chewing tobacco, as this may cause decrease blood flow to the inner ear and may worsen symptoms. Also, you must pay attention to the content of all over the counter medications as some may increase your symptoms.

Medical and Surgical Treatment:

Medical treatment of endolymphatic hydrops usually begins with a diuretic. These medications help to regulate the fluid pressure in your inner ear, thus reducing the severity and frequency of the Meniere's episodes. There are several different classes of diuretics, and your physician will select the type that is most suited to your condition. Dyazide is the most common medication used for this purpose, along with Lasix and others. Prednisone or other steroids (e.g. Medrol) are occasionally helpful in short bursts to control symptoms. In addition, Antivert (meclizine) may be used at times to control attacks.

Surgical treatment is reserved for patients that do not have their disease controlled with diet and medication. Some patients may be candidates for intratympanic steroids—steroid medication injected through the eardrum into the middle ear and absorbed directly into the inner ear. This treatment allows delivery of the steroids into the inner ear without absorption into the rest of the body and at higher concentrations compared with oral intake.

In patients where it has not been possible to control the vertigo generated by the affected ear, certain antibiotics (aminoglycosides) can be used to weaken the inner ear so that it is incapable of generating dizziness. One of these aminoglycosides, gentamicin, has been used to affect the inner ear directly. It can be placed in the middle ear (behind the eardrum) and be allowed to diffuse into the inner ear where it generally destroys some or all of the balance cells in the one ear. This treatment is highly successful at eliminating the remaining balance function of the affected ear while preserving the hearing—there is a risk of further hearing loss with this treatment, though this risk is slight.

In patients with persistent dizzy symptoms, or in those with special situations, surgical intervention is considered. Surgical treatment consists of endolymphatic sac

decompression, in which removal of the mastoid bone is achieved and the sac that holds the endolymph is decompressed or opened to relieve pressure in the inner ear and enhance its function. This procedure is an option in patients with normal hearing as it does not affect hearing. It is an outpatient procedure.

The remaining two procedures, vestibular neurectomy and labyrinthectomy, are ways of eliminating the balance function of the faulty ear. It is known that individuals will function better with one normal balance system than with one normal and one faulty system. The labyrinthectomy is a procedure in which the mastoid bone is removed and the inner ear is eliminated. This procedure is for patients that have lost usable hearing in the affected ear, as it entails removing all function of the inner ear, including hearing and balance. The change from having two balance systems to having one balance system alone does require a recovery or "compensation" period. It takes the brain a period of weeks to figure out that only one system is active and that it is no longer receiving information from the faulty system which it had come to expect. The second procedure, the vestibular neurectomy, is a good option if the hearing is good in the ear with the failing balance system. In this surgical procedure, the balance nerve (vestibular nerve) is cut between the inner ear and the brain. The inner ear is completely preserved but the faulty balance information is not able to reach the brain and cause the vertigo. Like the labyrinthectomy, this procedure requires a recovery period while the brain "figures out" the new situation.

Summary:

Meniere's Disease is an extremely complex condition with many potential treatments. The symptoms associated with this disease are extremely variable between patients and treatment planning is an individualized process in which many factors are taken into account when discussing options.

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