Ménière's disease (also known as endolymphatic hydrops) is a disorder of the inner ear where there is over-accumulation of fluid within the inner ear. This leads to the following symptoms:

1. fluctuating low frequency hearing loss, and
2. tinnitus (ringing in the ear),
3. episodic vertigo (a sensation of the world spinning), and
4. aural fullness (the sensation of pressure in the ear).

Classically, patients sense an attack coming when pressure builds up in the ear, which may be accompanied by tinnitus and/or hearing loss. The process typically culminates in a vertiginous (dizzy) attack, which may last hours. Patients usually feel fatigued after an attack. Attacks may occur daily, weekly, monthly, or yearly in often unpredictable patterns. The good news is that there are effective treatment options to decrease the frequency and severity of these attacks.

DIAGNOSIS

There are many causes of vertigo beyond Ménière's disease; therefore a complete otolaryngologic work-up is necessary to confirm the diagnosis. By far, the most useful information is the patient's history of symptoms. An audiogram is usually ordered to assess hearing. Additional testing includes electronystagmography (ENG), rotational testing, vestibular evoked myogenic potential (VEMP), and electrocochleography (ECOG). ENG assesses a patient's sense of balance by measuring eye motion after stimulating the semicircular canals (balance organs located within the inner ear). ECOG and VEMP assess the nerve impulses sent from the inner ear to the brain; a specific pattern of these impulses is associated with Ménière's disease. Rotational testing tests the degree of compensation of the brain related to a vestibular weakness.

PATHOGENESIS

The cause of Ménière's disease is unknown. The accumulation of fluid in the inner ear is most likely the final pathway of numerous insults to the body. Trauma to the ear (i.e. motor vehicle crashes or after ear surgery), infections (both to the entire body and those localized to the ear), and autoimmune disorders have all been implicated as causative events. It is well-known that the final end-point of these insults is accumulation of fluid (endolymph) in the inner ear. As the fluid accumulates, pressure builds up, contributing to a sense of fullness in the ear and precipitating tinnitus and hearing loss; vertigo usually follows. Therapies are directed at decreasing this accumulation of fluid in the inner ear.
TREATMENT

Treatment options follow a continuum of increasing invasiveness. Over 90% of Ménière’s patients have control of their symptoms with medications and few require surgical intervention.

LIFESTYLE ALTERATIONS

(1) **Caffeine** - Stop ingesting caffeine in all forms (coffee, tea, soda, chocolate, etc...) as it has been shown to exacerbate Ménière’s attacks!

(2) **Salt** - Begin to have a low salt diet. This means more than just not adding salt at the dinner table. Many prepared foods and restaurant meals are very high in sodium (especially Chinese food). A diet limiting intake of sodium to 1500 mg is strongly suggested.

(3) **Nicotine and Alcohol** – Nicotine (in any form) and alcohol have both been shown to make dizziness worse in patients with Ménière’s disease.

(4) **Stress and Fatigue** - One of the worst things about Ménière’s is the unpredictability of the attacks. Physical and emotional stress and fatigue can aggravate this process. Effective stress reduction including psychological counseling is often helpful in learning to deal with this chronic problem. Get plenty of rest!

MEDICAL THERAPY

(1) **Diuretics** - Diuretics (also known as water pills) act to decrease the accumulation of fluid in the inner ear and help to control the disease process. Be forewarned that these medications increase urinary frequency and take a number of weeks to get used to.

(2) **Anti-Vertigo Medications** - Various medications suppress the inner ear's balance function. The first group is the antihistamines, including meclizine (Antivert®) and glycopyrolate (Robinul®). These medications may be accompanied by mouth dryness and urinary retention. The second group is the benzodiazepines of which diazepam (Valium®) and alprazalam (Xanax®) are the most recognized. These drugs directly suppress the balance organs, but may cause patients to feel "drunk" or “out of it”.

(3) **Anti-Emetics** – Anti-emetics (anti-nausea) can be helpful when a vertiginous attack flares. Promethazine (Phenergan®) is an excellent anti-emetic which has the side effect of inducing sleep which may be helpful in the throws of an attack.
(4) **Oral Steroids** – Steroids are anti-inflammatory medications which squash flare-ups of Ménière’s disease. They may be given orally, in which case medications which protect the stomach are also indicated (i.e. Nexium® or Xantac®). A side-effect of oral steroid use may be a jittery feeling. Alternately, steroids could be injected into the middle through the tympanic membrane avoiding many of the systemic side-effects.

(5) **Injectable Steroids** – Trans-tympanic perfusion of Dexamethasone is a relatively new treatment for Ménière’s disease that we are very excited about at Vanderbilt. This is an almost painless procedure that can easily be done in the clinic. Typically, several drops of a topical anesthetic (numbing medicine) are inserted into the affected ear canal. Then a powerful steroid (Dexamethasone) is gently injected with a needle through the eardrum, into the middle ear space. This procedure is repeated a couple of times within about 30 minutes time. Within several days to a couple of weeks, patients very frequently notice significant improvement in the frequency and severity of their vertigo attacks. If the treatment is beneficial, this procedure can be repeated every 4-6 months.

(5) **Aminoglycosides** - These antibiotics are known to be toxic to the inner ear. They can be used to selectively damage the balance organs of the inner ear to relieve vertigo. A side effect of these drugs is hearing loss. There are two ways to administer the drugs – trans-tympanically or systemically. Trans-tympanic administration is given after numbing the eardrum following which a needle is used to inject an aminoglycoside (usually gentamycin) into the middle ear space. Repeat injections may be necessary. Hearing loss from trans-tympanic administration of gentamycin ranges from 5-15%. Systemic administration is used only for bilateral Ménière’s (15% of patients) and consists of intramuscular shots of streptomycin.

(6) **Low-Pressure Pulse Generator** – A treatment option now available is showing promise. The Meniett® is a device that delivers low-pressure pulses to the middle ear through a tube connected to an earpiece placed in the outer ear. It is believed that the energy of the pressure pulses causes displacement of the inner ear fluids, which relieves the symptoms of Ménière’s disease.

**SURGICAL THERAPY**

(1) **Endolymphatic Sac Surgery** - Part of the plumbing system in the inner ear includes a reservoir for the inner ear fluid; this reservoir is called the endolymphatic sac. By relieving pressure at the sac, vertigo is controlled in up to 80% of patients. To access the sac, a middle ear surgery called a mastoidectomy is performed. The sac is then identified and incised and often times a small drain may be placed which remains in place draining tiny (<<1ml) amounts of fluid from the inner ear to the mastoid cavity. Very infrequently, this surgery may be complicated by hearing loss, paralysis of the facial nerve, bleeding, infection, and/or leaking of cerebrospinal fluid. This surgery is routinely performed as outpatient surgery.
(2) **Vestibular Nerve Section Surgery** - For patients with intractable vertigo who have useable hearing, vestibular nerve section is effective (90+%) in relieving acute vertiginous attacks; low-grade disequilibrium may persist. This surgical procedure involves intracranial access to the vestibular nerve via a craniectomy. Owing to the larger scale of this operation, a couple days recuperation in the hospital is usually necessary. Infrequent risks to the procedure included chronic headaches, leakage of cerebrospinal fluid, facial nerve paralysis, and hearing loss.

(3) **Labyrinthectomy Surgery** - For patients with intractable vertigo and no useable hearing, the inner ear can be surgically destroyed via a labyrinthectomy. This surgical procedure usually involves a mastoidectomy to approach the inner ear followed by surgical removal of both the organs of balance and the organs of hearing. This procedure is very effective in controlling vertigo (>95%) but has as it downside the loss of hearing. Acutely, vertigo may be more intense as one's body gets used to lack of input from the inner ear. A couple day hospitalization may be necessary to accommodate to this process. Other risks of the surgery are infrequent CSF leak and facial nerve paralysis.

**A Word about Alternate Therapy**

The scientific community does not know precisely what causes Ménière’s disease. Various other therapies have been advocated including anti-oxidants, hysterectomy, etc... Our therapeutic options are based on the model of Ménière’s from accumulation of inner ear fluid. We do not discourage alternative therapy. If you have success from such therapy please let us know so that we can discuss whether it might work for others.

**Support**

Support for your Ménière’s disease can be found by contacting Suzanne Wyatt with The Ear Foundation at (615) 627-2724. She can also be reached at suzanne@earfoundation.org. Other resources are available through The Ear Foundation at www.earfoundation.org.

**HOPE ! 😊**

We counsel all of our patients to never let this disease beat them. Ménière’s is a cruel disease which seems to strike at unsuspecting times. It makes social interactions difficult at best. Remember the continuum of therapy outlined above and always remember that there is a next step. Find a healthcare provider that you can effectively communicate with so that together you may manage this disease process and not let it manage you.