Meniere’s Disease (Endolymphatic Hydrops)

How the Inner Ear Works
The inner ear serves both hearing and balance functions. The snail shaped cochlea collects sounds and transmits this information via the cochlear nerve. The adjacent labyrinth collects balance information via the vestibular nerve. The labyrinth is composed of three semicircular canals (superior, horizontal and posterior) and two otolithic organs (utricle and saccule). The semicircular canals sense angular acceleration (i.e., head turning) while the otolithic organs sense linear acceleration (i.e., swingset, elevator). Signals from the inner ear are then sent to the brain and integrated in the balance center.

What are the symptoms of Meniere’s disease?
The symptoms of Meniere’s disease are caused by excessive fluid build-up of fluid in the inner ear. The common symptoms are:

1. Ear fullness/pressure
2. Fluctuating hearing loss
3. Tinnitus (ringing in the ear)
4. Vertigo (an illusion of motion) – typically lasting hours

After an attack, most people find they are exhausted and must sleep for several hours whereas others have prolonged periods of unsteadiness. Meniere’s spells often occur in clusters, with several attacks occurring within a short period of time. Between spells however, most people enjoy good balance.

What causes Meniere’s?
The inner ear is composed of two fluid spaces: perilymph and endolymph. Perilymph (seen in blue) is composed primarily of sodium, whereas endolymph (seen in red) is composed primarily of potassium.

The difference in this potassium concentration creates a biological “battery” that powers the cells of hearing and balance. The endolymphatic fluid is produced by the stria vascularis and is absorbed by the endolymphatic sac. The amount of endolymph produced is related to dietary salt (sodium) intake. Symptoms of ear fullness, hearing loss and tinnitus are related to increasing endolymphatic pressure. When this fluid pressure reaches a critical level, Reissner’s membrane bulges outwardly and ruptures causing spillage of potassium into the sodium containing perilymphatic space. This results in a “short circuit” of the inner ear battery, producing hearing and balance symptoms.

How is Meniere’s disease diagnosed?
Meniere’s affects 2 people out of 1,000 and is diagnosed primarily on your symptoms. A hearing and balance history and physical exam is performed along with an audiogram (hearing test) to check your hearing. An audiogram of a patient with Meniere’s (right) typically shows low-tone hearing loss. While this pattern of hearing loss is most common, nearly all patterns of hearing loss can be seen – including normal hearing.

In addition, an electronystagmogram (ENG) is sometimes performed to confirm the diagnosis. This is a test of your inner ear function that often produces dizziness. Occasionally, blood tests or MRI scans of the head are ordered to investigate atypical symptoms to rule out other causes of hearing loss or dizziness.

The Endolymphatic Sac
Dysfunction of the endolymphatic sac is thought to be the main culprit in Meniere’s disease. The sac becomes dysfunctional for a variety of reasons, leading to accumulation of endolymphatic fluid (i.e., endolymphatic hydrops). Compare the normal size of
endolymphatic space (left) to that of a patient with Meniere’s (right). Some of the reasons for poor sac function include fibrosis (scarring), accumulated debris (slugging) and inflammation of the sac. An attack of Meniere’s disease is believed to result from fluctuating pressure within the inner ear.

**Preventing the Dizzy spells**

The purpose of treatment between attacks is to prevent or reduce the number of episodes, and to decrease the chances of further hearing loss.

1. **Dietary restrictions**
   - a. Sodium intake less than 1600 mg/day (less than one teaspoon total). Read Labels!
   - b. Reduce caffeine and alcohol intake

2. **Diuretics** (Dyazide or Lasix). These “water pills” act on the ear like they do the kidney, reducing fluid accumulation in the inner ear.

3. **Steroids** (Prednisone). Many patients with Meniere’s have inflamed endolymphatic sacs. When significant spells of vertigo occur or sudden losses of hearing, oral and occasional steroids injected past the eardrum are very beneficial.

**What if medical treatment doesn’t work?**

The medical treatment above is helpful in controlling symptoms of vertigo in over 80% of patients. For those who continue to have symptoms of dizziness, multiple surgical options exist:

1. **Trans tympanic Steroids.** For those who are responsive to oral steroids, injectable steroids offer a safe, effective way of reducing inner ear inflammation - without the side effects of oral prednisone. This helps ~50% of patients and offers little to no risk to residual hearing.

2. **Trans tympanic Gentamycin.** This powerful antibiotic is effective in weakening the balance nerve, resulting in less severe symptoms. There are typically 3-4 injections given over 1-2 months to help eliminate dizziness. This helps ~80% of patients but carries a ~15% risk of significant hearing loss.

3. **Endolymphatic shunt.** This operation involves opening the draining the abnormal endolymphatic sac, leaving a small drain in place. Although the mechanism of vertigo control is not definitely known, improved blood supply to the endolymphatic sac after surgery probably plays a role. This helps ~75% of patients but carries only a ~1% risk of significant hearing loss.

4. **Selective Vestibular Nerve Section.** This operative involves opening the skull and cutting the nerve of balance, preserving the hearing nerve. This operation helps ~95% of patients but carries a ~25% chance of deafness and a small chance of weakness of the facial nerve. Recovery is much more prolonged than the operations above, but has excellent results in curing dizziness from Meniere’s disease.

5. **Labyrinthectomy.** When hearing is no longer usable, a labyrinthectomy is often advised to control dizziness. This surgical procedure is less invasive than a vestibular nerve section with similar chances of cure (~98%), but will leave the operated ear permanently deaf.

**Frequently Asked Questions (FAQs)**

- **How often does Meniere’s affect both ears?**
  - About 30% of patients will develop symptoms of Meniere’s disease in the opposite ear within 10 years.

- **Is Meniere’s inherited?**
  - No, although some families clearly show a tendency.

- **What medicines can I use when I am having a bad dizzy spell?**
  - Meclizine (Antivert) is available over the counter and is very effective. Other medications are available by prescription to help the vertigo and nausea.

- **Should I be taking Meclizine regularly to help prevent spells?**
  - No, the regular use of Meclizine prevents your brain from adjusting to the changing balance function of your ear. Taken over long periods (weeks-months) leads to a constant feeling of unsteadiness.

- **What happens if my spells get better but my balance remains poor?**
  - Vestibular rehabilitation can be of significant value for poorly compensated balance. This specialized form of physical therapy strengthens your balance at the brain level.

- **How long should I stay on my water pill if my dizzy spells are controlled?**
  - I like to ensure spells are well controlled for 6 months before discontinuing medication.

- **I have Meniere’s and I suddenly lost my hearing. What should I do?**
  - This is considered an emergency and should be promptly evaluated. A hearing test should be arranged through our office and medication begun (oral steroids ± other medications).

- **Will I go deaf in my Meniere’s ear?**
  - It is uncommon to go deaf as a result of Meniere’s but it happens in rare instances. Continued medical treatment is important in preventing further hearing loss.

- **Will a hearing aid help my hearing loss?**
  - Yes, a hearing aid is often very helpful in improving your hearing and can help reduce the severity of your tinnitus.