

Meniere's Disease

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*This paper covers basic information about Meniere's Disease,
Its Possible Causes and Implications*

Meniere's disease is a disorder of the inner ear which causes episodes of vertigo, ringing in the ears (tinnitus), a feeling of fullness or pressure in the ear, and fluctuating hearing loss. It was first described by a French physician Dr. Prosper Meniere (illustrated above) in 1861.

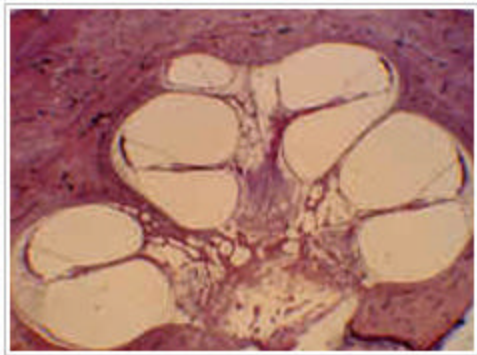
There is a great deal of disagreement regarding the actual incidence of Meniere's Disease in the population. Estimates range from 0.2 percent to as much as 4 percent. Based on a study conducted by the National Institute of Deafness and Communication Disorders (NIDCD) approximately 45,500 new cases are diagnosed each year.

There is no single known cause for Meniere's Disease. For this reason many identify this disorder as a syndrome. Some of the theories proposed:

Endolymphatic hydrops: When one looks at the inner ear of many deceased individuals who suffered from Meniere's Disease the inner ear membranes look distended. This condition is called hydrops. The theory is that progressive dilation of the inner ear structures causes micro-ruptures in the delicate membranes causing contamination of the inner ear nerve endings. What causes this dilation is unknown. Not all Meniere's patients have this condition and some normal people have it as well so this is probably not the complete story.

Autoimmune: There is a large body of evidence that the body's own immune mechanism may be responsible for this disorder. Researchers have found antibodies to inner ear proteins circulating in the bloodstreams of many patients with the disorder.

Anatomy: The endolymphatic sac is thought to be responsible for cleansing the inner ear fluids. Physical abnormalities of the bone surrounding the sac may be responsible for dysfunction of this cleansing system.



Viral: viral particles have been identified within the inner ear of many Meniere's patients

Diagnosis of Meniere's Disease

One of the main difficulties in studying this disorder is that is no "hard" medical test, such as a blood test, for the disease. The most important data is the person's symptoms. Meniere's disease is usually characterized 4 symptoms:

- Periodic episodes of rotatory vertigo or dizziness.
- Fluctuating, progressive, low-frequency hearing loss
- Tinnitus
- A sensation of "fullness" or pressure in the ear

Meniere's Disease is one of the most over-diagnosed disorders in medicine. By definition, a person does not suffer from this disorder unless all of the symptom criteria are met.

Detailed Description of Symptoms

Periodic episodes of rotatory vertigo or dizziness.

The hallmark symptom of Meniere's Disease is vertigo. Vertigo is defined as a feeling that the world is spinning around the individual. Periodic attacks of vertigo is the most disruptive of the symptoms to the patient. It is usually the vertigo attack which causes the patient to seek medical treatment. Typically, vertigo occurs in the form of a series of attacks over a period of weeks or months, interspersed by periods of remission of variable duration. Unsteadiness may persist for the following hours or days.

The onset of vertigo may be preceded by a sensation of fullness or pressure in the ear, increased hearing loss and tinnitus, as described below:

Fluctuating, progressive, low-frequency hearing loss

The hearing loss usually affects one ear, which typically loses sensitivity to low-frequency (bass) sounds the most. As well as being harder to hear, sounds may appear "tinny" or distorted. Loud sounds may cause more discomfort than normal (loudness intolerance). The hearing loss fluctuates over time but eventually affects all sound frequencies. Complete hearing loss (dead ear) is uncommon, however, the hearing loss typically worsens to a moderate to severe loss. The vertigo may even subside when this happens. Many physicians call this a "burned out Meniere's ear."

Tinnitus

The tinnitus experienced in Meniere's Disease is a low rumbling sound that is directly temporally related to the vertigo and hearing loss episodes

Aural fullness

The feeling of "fullness" in the ear is similar to that experienced by barometric pressure changes (such as when riding up or down a hill, or ascending or descending in an airplane). However, this fullness cannot be cleared by swallowing, as in the case of pressure changes.

Diagnostic Testing

Diagnostic testing is rarely helpful in the diagnosis. Historically ECoG, or electrocochleography, testing was used. This test is controversial and has been shown to be of little benefit. Audiologic evaluation (hearing test) is the most helpful test. The typical "Meniere's pattern" is a low frequency sensorineural hearing loss. As the disease progresses the hearing loss becomes more of a flat moderate to severe loss. ENG testing is often done but is also of limited usefulness. The ENG is important, however, in documenting the health of the balance system in the other ear. This is crucial to know if ablative (vestibular nerve section, gentamicin injection) therapy is contemplated.

Treatments

The analogy that is often applied to Meniere's Disease is that of a twin engine airplane flying. As long as both engines are running the plane flies straight. If one engine fails the plane starts to spin until the pilot compensates and straightens the aircraft back to level flight. The problem occurs if there is a "sputtering engine" that turns on and off intermittently. The pilot does not have a chance to correct the situation and the airplane continues to rotate. The remedy is to stop the engine completely so that a stable situation is achieved. A Meniere's ear is very similar to a sputtering engine. Normally both ears provide equal and opposite signals to the brain. A Meniere's ear sends inconsistent (sputtering) signals to the brain. One of the main treatments for Meniere's Disease is to weaken or shut down that sputtering engine.

Diet and drugs: Meniere's Disease is initially treated conservatively with a salt restriction diet and diuretics. Patients are asked to maintain an intake of about 3gms per day of salt. The absolute number does not seem to be as important as is maintaining a steady level of intake from day to day. A diuretic such as Dyazide is routinely prescribed. Seventy five percent of all patients that are treated conservatively have a significant decrease in vertigo episodes.

Menniott Device: This is a small air pump that delivers slow puffs of air against the eardrum. The patient must use the device three to four times a day in treatments that last about five minutes. A ventilation tube must be inserted through the eardrum and maintained so that the pressure waves can act on the inner ear during treatment. The Menniott has seen some success and popularity in Europe. The exact mechanism of action has not been discovered and good placebo controlled studies have not been done to document effectiveness of the treatment.

Transtympanic gentamicin injection: This procedure is also called a chemical labyrinthectomy. Gentamicin is an antibiotic that was once used extensively to treat infections. The drug was found to be toxic to the hearing and balance nerve endings and so is no longer used except for very selected situations. This procedure takes advantage of the toxic effects of the drug to specifically poison and weaken the balance nerve ending in the diseased ear (see sputtering airplane engine analogy above). The procedure is done in the office. The ear drum is first anesthetized. Then approximately 1cc of gentamicin is injected through the ear drum using a small needle. The injection may cause a brief burn and vertigo feeling and may need to be repeated several times. In some cases the injection may not work at all. This is usually due to scar tissue from previous middle ear surgery. Some people may be genetically resistant to the drug's effects. The main possible complication is further hearing loss that may be permanent and profound. Recently some studies have suggested that using a steroid instead of gentamicin may be effective without risking hearing loss.

Endolymphatic Sac Shunt

The endolymphatic shunt or decompression procedure is an ear operation that usually preserves hearing. This operation drains excess endolymph from the inner ear. It is usually performed under general anesthesia as an outpatient. An incision is made behind the ear. A mastoid operation is performed and a tube is inserted into the endolymphatic sac of the inner ear to control the abnormal fluid pressure.

A shunt operation usually is advised when hearing is relatively good in the involved ear. Further permanent loss of hearing occurs in 5 percent of patients. Total loss of hearing in the operated ear occurs in 1 percent. Attacks of vertigo are controlled in one half to two-thirds of cases, but control is not permanent in all cases. Recovery time after this procedure is short compared to the other procedures.

Vestibular Neurectomy

This is a procedure in which the balance nerve is cut as it leaves the inner ear and goes to the brain. Vertigo attacks are permanently cured in a high percentage of cases, and hearing is preserved in most cases. This procedure is performed under general anesthesia and usually requires five to seven days of hospitalization. Through an incision behind the ear a mastoidectomy is performed and the balance nerve is cut before it enters the inner ear.

Retrolabyrinthine section of the vestibular nerve may be advised when hearing is good in the involved ear. Up to two percent of patients may develop a severe hearing impairment in the operated ear following surgery. Fortunately the attacks of dizziness are eliminated in nearly every instance. Persistent unsteadiness, however, may continue for weeks or months until the opposite ear stabilizes the balance system.

Labyrinthectomy and Eighth Nerve Section

These are procedures in which the balance and hearing mechanism in the inner ear are destroyed on one side (see airplane analogy above). This is considered when the patient with Meniere's disease has poor hearing in the affected ear. Labyrinthectomy and eighth nerve section result in the highest rates for control of vertigo attacks. The operation is performed under general anesthesia and requires hospitalization for approximately five to seven days. An incision is made behind the ear and a mastoidectomy is performed. The inner ear balance chambers are removed and the balance nerve is cut. In cases selected for labyrinthectomy and section of the vestibular nerve, hearing is severely impaired.

The operation results in total loss of hearing in the operated ear and frequently a temporary increase in dizziness. Fortunately, the attacks of dizziness are eliminated in nearly every instance. Persistent unsteadiness, however, may continue for a period of weeks or months until the opposite ear stabilizes the balance system.

Conclusion

Unfortunately, there currently is no firm diagnostic tool to confirm the presence of Meniere's Disease. There is also no cure for this disease. Treatments rely on decreasing the symptoms that patients experience. For the vast majority of people Meniere's Disease can be controlled so that the individual can live a long and normal life.

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