Ménière's disease and cochlear hydrops

Interview with
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Q What is Ménière's disease and why does it cause vertigo?

Ménière's disease is a common cause of repeated attacks of vertigo. It is characterized by episodes of room-spinning vertigo that vary in duration from a few minutes to several hours. Hearing loss and head noise (tinnitus) usually accompany these attacks, and a sensation of pressure and fullness in the ear is usually present. The attacks of vertigo may occur suddenly. Violent spinning, whirling and falling, associated with nausea and vomiting, are common symptoms. Clearly, an active case of Ménière's can be quite debilitating for patients.

The episodes of vertigo generally are due to increased pressure of the inner ear fluids. Fluids in the inner ear chambers are constantly being produced and absorbed by the inner ear. Any disruption to this delicate process results in over-production or under-absorption of the fluids. This leads to increased fluid pressure (hydrops) that in turn produces vertigo and hearing loss. This usually affects only one ear, but research at the Institute shows it affects both ears in approximately 20% of patients.

Q Can you further explain the symptoms associated with Ménière's and related disorders?

Attacks of vertigo may recur at irregular intervals. Ménière's is unpredictable. The individual may be free of vertigo symptoms for years at a time, and between attacks the individual tends to remain free of symptoms, though hearing loss may continue.

Occasionally hearing impairment, head noise and ear pressure occur without vertigo. This type of Ménière's disease is called *cochlear hydrops.* Similarly, episodic vertigo and ear pressure may occur without hearing loss and tinnitus; this is called *vestibular hydrops.* Treatment of both of these conditions is the same as for Ménière's disease.

Q Is it possible that these symptoms may have other causes?

A thorough evaluation is necessary to determine if the patient is suffering from Ménière's, cochlear hydrops or another disorder. The four hallmark symptoms of Ménière's are episodic vertigo, tinnitus, fluctuating hearing loss and ear fullness, though many Ménière's patients may have two or fewer hallmark symptoms. It's important to note that according to the American Academy of Otolaryngology – Head and Neck Surgery, a true definition of Ménière's requires that three of the hallmark symptoms be present. If Ménière's is suspected, it is important to try to determine the cause of the increased fluid pressure. It's possible that metabolic, toxic, allergic, circulatory or stress and emotional factors play a part in some cases.

Q How are Ménière's or cochlear hydrops diagnosed?

The diagnosis of Ménière's disease in patients is based on the history from the patient. The history includes episodic vertigo, fluctuating hearing loss, tinnitus and fullness in the involved ear. Most patients will report an increase in the tinnitus and fullness prior to and during the vertigo attacks. In addition, between attacks the patients are symptom free other than a possible hearing loss.

Diagnosis of Ménière's disease can be challenging since its symptoms are also seen in a range of other disorders. The presence of symptoms alone does not guarantee the presence of Ménière's disease, and there is not a homogenous population of Ménière's patients, which makes unequivocal diagnosis difficult. However, a thorough examination of all possible causes of symptoms enables us to diagnose with a fairly high level of confidence. The diagnostic test developed by House Ear Institute

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researcher Manny Don, Ph.D., which involves monitoring auditory brainstem response (ABR) in electrophysiology, has been helpful in further confirming a diagnosis of Ménière's for those patients who display 3-4 of the classic symptoms.

Q How do you treat Ménière's disease?

Treatment of Ménière's disease may be medical or surgical, though the House Clinic generally favors a medical approach as the first course of treatment. Treatment is aimed at improving the inner ear circulation and controlling the fluid pressure changes of the inner ear chambers. At times it is necessary to proceed to a surgical intervention.

Medical treatment of Ménière's disease varies with the individual patient according to suspected cause, magnitude and frequency of symptoms. It is effective in decreasing the frequency and severity of attacks in about 80% of patients. Treatment may consist of medication to stimulate the inner ear circulation, decrease the inner ear fluid pressure or prevent inner ear allergic reactions.

Various vasodilating drugs are used to stimulate the inner ear circulation and are prescribed together with anti-dizziness medication. Vasoconstricting substances (such as caffeine or nicotine) have an opposite effect and, therefore, should be avoided. Diuretics ("water pills") and a low sodium diet are usually prescribed to decrease the inner ear fluid pressure.

Ménière's disease may be caused or aggravated by metabolic or allergic disorders. Special diets or drug therapy are indicated at times to control these problems.

On rare occasions, intra-tympanic injections of steroids are used to help reduce hydrops in the inner ear.

Q What are some of the surgical treatments recommended to treat Ménière's?

Surgery is indicated when medical treatment fails to relieve the disabling attacks of vertigo. The type of operation selected depends upon the degree of hearing impairment in the affected ear since every effort is made to preserve this hearing. In some cases the hearing may be improved following surgery and in others it may become worse; usually it remains the same. Head noise (tinnitus) may or may not be improved and in some cases may even become more marked.

Surgery is successful in relieving acute attacks of vertigo in the majority of patients. In the event that a conservative operative procedure does not relieve the attacks of vertigo, a second operation may be recommended.

Q What is the "shunt procedure" and is it the most successful surgical treatment for Ménière's?

The shunt procedure has a 70% success rate, with astronaut Alan Shepard being the most famous Ménière's patient to receive relief from symptoms with this operation.

Endolymphatic Shunt

This operation alters the function of the endolymphatic sac such that its abnormally low resorption of inner ear fluids and/or secretion of molecules disrupting normal fluid balance is positively changed. It is usually performed under general anesthesia as an outpatient.

An incision is made behind the ear. A mastoid operation is performed and a drain is inserted into the endolymphatic sac of the inner ear in an attempt to control the abnormal inner ear fluid pressure.

A shunt operation usually is advised when hearing is relatively good in the involved ear. Further permanent loss of hearing occurs in less than 5% of patients. Total loss of hearing in the operated ear occurs in 1%. This procedure controls vertigo attacks in seven out of ten patients.

There are several other surgical procedures that treat balance (vestibular) problems and these can sometimes be employed in Ménière's cases where a shunt would not be indicated. These other surgical procedures include cochleosacculotomy, transcanal (oval window) labyrinthectomy, translabyrinthine labyrinthectomy and section of the vestibular (balance) nerve, retrolabyrinthine section of the vestibular (balance) nerve, middle

fossa section of the vestibular (balance) nerve, and retrosigmoid microvascular vestibular nerve decompression. I'll outline those that can be employed for Ménière's cases.

Transcanal Mastoid (Oval Window) Labyrinthectomy

This procedure is performed under general anesthesia usually as an outpatient. The semicircular canals and secreting membranes are removed from the inner ear chambers.

Labyrinthectomy usually is advised only when the hearing is poor; it results in total loss of residual hearing in the operated ear and a temporary increase in vertigo.

Translabyrinthine Labyrinthectomy and Section of the Vestibular (Balance) Nerve

The operation is performed under general anesthesia and requires hospitalization for approximately three to four days. Through an incision behind the ear a mastoidectomy is performed, the inner ear balance chambers are removed and the balance nerve is cut.

In labyrinthectomy cases and section of the vestibular nerve, hearing is already severely impaired. The operation results in total loss of hearing in the operated ear and frequently a temporary increase in vertigo. Fortunately, the attacks of spinning vertigo 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.

When necessary, this operation can be performed if other surgery is not successful.

Retrolabyrinthine Craniotomy Section of the Vestibular (Balance) Nerve

This procedure is performed under general anesthesia and usually requires four to five days of hospitalization. Through an incision behind the ear a mastoidectomy is performed and the balance nerve is separated from the hearing nerve and 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 2% of patients may develop a severe hearing impairment in the operated ear following surgery. Fortunately, the attacks of vertigo are eliminated in nearly every instance. Persistent unsteadiness, however, may continue for weeks or months until the opposite ear stabilizes the balance system.

Middle Fossa Craniotomy Section of the Vestibular (Balance) Nerve

This procedure is performed under general anesthesia and usually requires four days of hospitalization. Through an incision above the ear, the balance nerve is cut before it enters the inner chamber.

Middle fossa section of the vestibular nerve may be advised when hearing is good in the involved ear. Up to 5% of patients may develop a severe hearing impairment in the operated ear. Fortunately, the attacks of vertigo 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. Complications of all craniotomies are extremely rare but can include the risk of meningitis, CSF leak, temporary facial weakness, and stroke.

Jonathan Swift

Literary figure and Ménière's sufferer

The author of *Gulliver's Travels* and *The Modest Proposal,* Jonathan Swift was a renowned satirist. Born in Dublin in 1667, Swift attended Trinity College and Oxford University. Although best known for his writings, Swift spent much of his life in the service of the church, including as dean of St. Patrick's Cathedral in Dublin.

Beginning in his youth, Swift suffered from periodic bouts of deafness, sometimes combined with illness or 'giddiness'. It is thought that these bouts were caused by Ménière's disease, also known as idiopathic endolymphatic hydrops, which is an inner ear disorder.

(Copy courtesy of the Adams Center, AAO-HNSF. Image courtesy of the National Portrait Gallery, London.)

Q Are there any risks or complications from surgery for vertigo?

Hearing loss can be a risk, that is, further hearing impairment in the

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operated ear may occur following any of the procedures I mentioned and is the expected result following some. Tinnitus (head noise) usually remains the same as before surgery. If the hearing is worse following surgery, tinnitus may likewise be more noticeable. Some patients experience taste disturbance and mouth dryness for a few weeks following surgery. In some patients this disturbance is prolonged. There may be some temporary facial nerve sagging post-surgery. The facial nerve travels through the ear bone in close association with the hearing and balance nerves, the inner ear and mastoid. Temporary weakness of one side of the face is an uncommon postoperative complication of ear surgery that may occur as the result of an abnormality or a swelling of the nerve. Permanent paralysis of the face is extremely rare. Should it occur, however, eye complications could develop, requiring treatment by an eye specialist.

There are some additional potential risks that occur in very rare cases that may require prolonged hospitalization and healing including post-surgery infection or hematoma – collection of blood under the skin incision. In most instances the distressing symptoms of vertigo can be greatly benefited or eliminated by medical or surgical management.

Q **Recurring episodes of vertigo and other symptoms of Ménière's can be scary and debilitating, leading to loss of work, anxiety and frustration for many patients. What is the general treatment outlook and prognosis for improvement for these patients?**

The general treatment outlook is good and patients can be optimistic that with a treatment approach that is tailored to their individual case, they will find relief. In most instances the distressing symptoms of dizziness can be greatly benefited or eliminated by medical treatment. Again, 80% of patients with Ménière's and related disorders have found relief from bouts of dizziness with certain prescribed medications and diuretics, sometimes in combination with dietary changes or treatment for allergies. For some patients, surgical management of the symptoms is beneficial.

Q **Isn't there a fairly new device that can be implanted in the eardrum to relieve vertigo from Ménière's?**

Yes, early studies conducted in the U.S. and overseas with the Meniett® device have shown some success in reducing the severity and frequency of vertigo episodes for Ménière's patients. The Meniett is a low-pressure pulse generator that is designed to help restore the balance in the hydrodynamic system of the inner ear by transmitting low-pressure pulses to the middle ear. It is believed that the energy of the pressure pulses causes a displacement of inner ear fluids, which may relieve endolymphatic hydrops and symptoms of Ménière's disease. However, this device still has limited availability and long-term results are not yet known.

Q **Are there any additional treatments or interventions in the pipeline?**

There don't seem to be new treatments in development outside of a study of delivery of gentamicin (an antibiotic) to the intra-tympanic membrane of the inner ear for management of Ménière's. That study is still in its preliminary phase in the House Ear Institute's Clinical Studies Department, but gentamicin treatment has a 25% chance of causing irreversible sensorineural hearing loss, so early results suggest that the shunt surgery may preserve hearing better. ❖