Otosclerosis
By Aaron G. Benson, MD

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Otosclerosis is an abnormal microscopic growth and repair of bone in the walls of the inner ear. Frequently this causes the stapes bone to become frozen in place or fixed. Normally the stapes, the smallest bone in the body, vibrates freely to allow the transmission of sound into the inner ear. When it becomes cemented to the surrounding bone by the disease, it prevents sound waves from reaching the inner fluids, and hearing is impaired.

The disease is a little mysterious. It is a disease that runs in families. While it is thought to have a genetic basis, no gene defect has been identified yet. Studies in Scandinavia have suggested that fluoridation of the water may reduce the development of this disease. As a result of these studies, some doctors recommend that their patients take fluoride pills to slow progression in those patients who have the disease. Fluoride containing pills are an over the counter medication in the US.

Another theory is that early exposure to measles might cause the disease to progress or develop. There is little doubt that the disease is more common in women of childbearing age than it is in men. Pregnancy seems to cause the disease to progress. Women who have the disease may experience increased hearing loss with each pregnancy. The disease usually begins in the teens or early 20's.

Otosclerosis affects the ears alone and does not involve other parts of the body. When this condition is present, both ears are usually involved but frequently it is not symmetric.

Normally, sound is funneled by the auricle or outer ear through the ear canal to the eardrum. Movement of the eardrum is then transferred via a chain of bones. These three small bones end with the stapes bone moving in and out of the oval window in the inner ear. The movement of the stapes sets up a wave motion in the fluids of the inner ear. This in turn stimulates small hair cells in the inner ear that signal the nerve of hearing.
The nerve then carries the sound information to the brain.

Occasionally, the otosclerotic bone involves other structures so that the inner ear itself is damaged. But otosclerosis can also involve the other bones of hearing so that they cannot move properly. If the inner ear is involved, the hearing loss may be more than a conductive hearing loss. This means that a mixed loss of both conductive and sensorineural loss exists in these individuals. Surgery will only correct the conductive component of the hearing loss. The sensorineural hearing loss cannot be surgically corrected.

**How is it treated?**

There is no known medication available for treating otosclerosis. Some individuals have recommended the use of fluoride to prevent extension of the disease into the inner ear. Most individuals obtain fluoride through their drinking water. Those individuals who consume filtered or bottled water may consider the use of additional fluoride in their diet or as a dietary supplement. Most patients can wear a hearing aid successfully, but it is only helpful when worn. If a patient takes off the hearing aid at night, they may be unable to hear important sounds like children, alarms, or telephones. Stapes surgery can correct the conductive component of the hearing loss.

Stapes surgery is done using an operating microscope. The surgeon usually performs the operation under general anesthesia. Some individuals will tolerate local anesthesia and can have the procedure done using a technique called twilight sleep. The ear canal is anesthetized while the patient is sedated. Some surgeons prefer the use of general anesthesia. The surgeon will fold the eardrum forward looking into the middle ear space. A laser is usually used to remove a segment of the stapes and a metal prosthesis is inserted into an opening in the inner ear. The prosthesis is usually crimped into position on to the incus bone.

Individuals who have successful surgery sometimes awake and immediately notice improvement in hearing. Most individuals however will wait 2 months before blood and fluid is clear from the middle ear and packing is removed from the ear canal. Final results of surgery can take several months to fully develop.
Risks
No operation is foolproof. While stapes surgery is usually short, taking less than one hour to complete, it has some risks. There is a risk that there will be greater hearing loss. There are other risks such as vertigo, taste changes, facial weakness, tinnitus and infection. While complications are infrequent, they still occur. In addition to the risks of the operation itself there are risks from having a foreign material in the body.

Conclusion
The chances of obtaining a good result from this operation are quite high. A small number of patients may not obtain ideal results. The risks of this operation are the same as any other operation of the ear.