
OTOSCLEROSIS

Otosclerosis describes a condition of abnormal bone growth around one of the three small bones in the middle ear space called the stapes. When bone around the stapes hardens, the bone cannot move freely, which limits the ability to properly transmit sound. This results in hearing loss; the less movement of the bone, the greater the degree of hearing loss.

Otosclerosis usually causes conductive hearing loss, which means there is a problem with how the ear transmits sound vibrations. In a normal ear, sound vibrations are funneled by the outer ear onto the ear drum, or “tympanic membrane.” The ear drum sends these vibrations to the small bones in the middle ear: the malleus (hammer), incus (anvil), and stapes (stirrup). When the stapes bone moves, fluid in the inner ear moves and stimulates hair cells in the inner ear. These cells turn sound vibrations into electrical signals that are sent to the brain. Hearing loss can occur if any part of this process is damaged.

WHAT ARE THE SYMPTOMS OF OTOSCLEROSIS?

The symptoms of otosclerosis include:

- Progressive/worsening hearing loss
- Difficulty hearing low-pitched sounds or whispers
- Ringing, roaring, buzzing, or hissing in the ears or head, known as tinnitus

WHAT CAUSES OTOSCLEROSIS?

An estimated 2.5 to 10 percent of adults have some otosclerosis in their middle ear. Hearing loss can happen anytime, but it often begins in young adults, those in their twenties or thirties. The condition can develop in both women and men, although it has a higher incidence in woman and has been linked to rapid onset during pregnancy.

Otosclerosis affects both ears approximately 70 percent of the time; however, the amount of hearing loss may not be the same in each ear.

Approximately 60 percent of otosclerosis has an underlying genetic cause. Transmission of the gene that causes otosclerosis is complex and not everyone who has the gene will develop the condition.

Because many symptoms seen with otosclerosis can be caused by other medical or ear-related conditions, it's important to be examined by an ENT (ear, nose, and throat) specialist, or otolaryngologist. After an examination, your ENT specialist may order a hearing test, or audiogram. A common finding is a conductive hearing loss that is worse in low-frequency tones. Patients with otosclerosis can also have inner ear hearing loss, called sensorineural hearing loss.

The hearing test can also help determine if the stapes is not vibrating correctly, and your ENT specialist may request imaging scans of the ear. Based on the exam findings and test results, your doctor can make an accurate diagnosis and recommend treatment options that are best for you.

WHAT ARE THE TREATMENT OPTIONS?

The main options for otosclerosis include observation with repeated hearing tests, a hearing aid(s), or surgery. Observation is recommended only for mild hearing loss. Some medicines such as sodium fluoride or bisphosphonate supplements have been reported to limit the worsening of otosclerosis, but there is no definitive evidence of preventing its progression.

Hearing aids can help overcome a fixed stapes by increasing the sound volume. Cost, style, fit, and possible benefits of a hearing aid should be discussed with your ENT specialist. In some states, a hearing aid trial may also allow you to sample a device prior to purchase.

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If a patient is prone to dizziness or has a diagnosis of Ménière's disease, then trying a hearing aid before surgery may be advised.

Surgery for otosclerosis is called a *stapedectomy* or *stapedotomy*. In general, the stapes surgery is an outpatient procedure that can be done under local or general anesthesia. The surgery is done through the ear canal with an operating microscope, or endoscope. It involves removing part (*stapedotomy*) or all (*stapedectomy*) of the fixed stapes bone and replacing it with a prosthetic device or implant. The prosthetic device allows the bones of the middle ear to move again, correcting the conductive hearing loss. This surgery will not address any inner ear (sensorineural) hearing loss. The stapes prostheses are very small and will not set off metal detectors. Modern prostheses are also MRI compatible.

Pain is usually minimal but can vary. Some patients may experience dizziness the first few days after surgery. Taste sensation may also be altered for several weeks or months following surgery, but usually returns to normal. Risks of injuring the ear drum or causing additional hearing loss are rare but should be discussed. The ear canal is generally packed at the time of surgery to allow the ear drum to heal properly. Hearing improvement may not be noticed until it is removed. Discuss specific instructions on ear drops and other post-operative directions with your ENT specialist.

If otosclerosis affects both ears, typically the surgeon will choose to operate on the worst-hearing ear first and will not operate on both ears at the same time. The surgeon usually waits a minimum of six months before performing surgery on the second ear.

Following surgery, patients may be asked to refrain from nose blowing, swimming, or other activities that may get water in the operated ear or cause pressure within the

middle ear space. Normal activities (including air travel) are usually resumed two to four weeks after surgery. Notify your ENT specialist immediately if any of the following occurs:

- Sudden hearing loss
- Intense pain
- Facial weakness
- Prolonged or intense dizziness, especially room-spinning sensation (vertigo)
- Any new symptom related to the operated ear

In advanced cases of otosclerosis, in which the disease has caused additional inner ear (sensorineural) hearing loss, a stapedectomy procedure may not be enough to restore useful hearing. In these situations, a cochlear implant may be considered.

WHAT QUESTIONS SHOULD I ASK MY DOCTOR?

1. Is my hearing loss conductive, sensorineural, or both?
2. What are the pros and cons of using a hearing aid?
3. Are hearing aids covered by insurance?
4. What are the pros and cons, including the surgical risks, of undergoing stapes surgery?
5. When could I expect my hearing to improve after surgery?
6. If my hearing improves after surgery, would I still need a hearing aid?
7. What if I do nothing?