

# Otosclerosis

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## What is otosclerosis?

Otosclerosis is the abnormal growth of bone of the middle ear. This bone prevents structures within the ear from working properly and causes hearing loss. For some people with otosclerosis, the hearing loss may become severe.

## How do we hear?

Hearing is a series of events in which the ear converts sound waves into electrical signals and causes nerve impulses to be sent to the brain where they are interpreted as sound. The ear has three main parts: the outer, middle, and inner ear. Sound waves enter through the outer ear and reach the middle ear, where they cause the ear drum to vibrate.

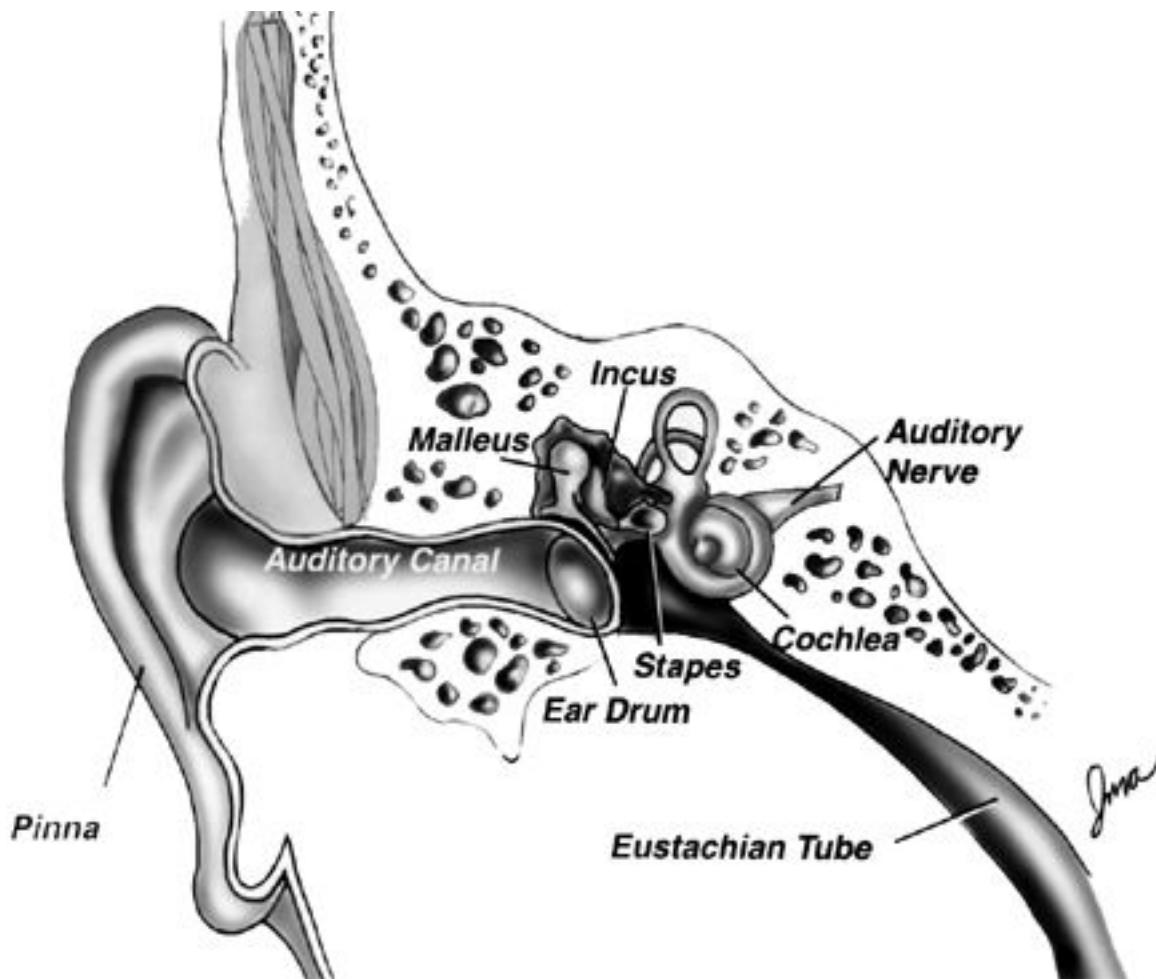
The vibrations are transmitted through three tiny bones in the middle ear called the ossicles. These three bones are named the malleus, incus, and stapes (and are also known as the hammer, anvil, and

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stirrup). The ear drum and ossicles carry the vibrations to the inner ear. The stirrup transmits the vibrations through the oval window and into the fluid that fills the inner ear.

The vibrations move through fluid in the snail-shaped hearing part of the inner ear (cochlea) that contains the hair cells. The fluid in the cochlea moves the top of the hair cells, which initiates the changes that lead to the production of the nerve impulses. These nerve impulses are carried to the brain, where they are interpreted as sound. Different sounds stimulate different parts of the inner ear, allowing the brain to distinguish among various sounds, for example, different vowel and consonant sounds.



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Image of the inner ear.

## **How does otosclerosis cause hearing impairment?**

Otosclerosis can cause different types of hearing loss, depending on which structure within the ear is affected. Otosclerosis usually affects the last bone in the chain, the stapes, which rests in the entrance to the inner ear (the oval window). The abnormal bone fixates the stapes in the oval window and interferes with sound passing waves to the inner ear.

Otosclerosis usually causes a conductive hearing loss, a hearing loss caused by a problem in the outer or middle ear. Less frequently, otosclerosis may cause a sensorineural hearing loss (damaged sensory cells and/or nerve fibers of the inner ear), as well as a conductive hearing loss.

## **What causes otosclerosis?**

The cause of otosclerosis is not fully understood, although research has shown that otosclerosis tends to run in families and may be hereditary, or passed down from parent to child. People who have a family history of otosclerosis are more likely to develop the disorder.

On average, a person who has one parent with otosclerosis has a 25 percent chance of developing the disorder. If both parents have otosclerosis, the risk goes up to 50 percent. Research shows that white, middle-aged women are most at risk.

Some research suggests a relationship between otosclerosis and the hormonal changes associated with pregnancy. While the exact cause

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remains unknown, there is some evidence associating viral infections (such as measles) and otosclerosis.

## **What are the symptoms of otosclerosis?**

Hearing loss is the most frequent symptom of otosclerosis. The loss may appear very gradually. Many people with otosclerosis first notice that they cannot hear low-pitched sounds or that they can no longer hear a whisper.

In addition to hearing loss, some people with otosclerosis may experience dizziness, balance problems, or tinnitus. Tinnitus is a sensation of ringing, roaring, buzzing, or hissing in the ears or head that accompanies many forms of hearing loss.

## **How is otosclerosis diagnosed?**

An examination by a physician is needed to rule out other diseases or health problems that may cause these same symptoms.

A hearing healthcare professional will administer a hearing test. The test should include an assessment of the middle ear function. The results will be used to help determine if the hearing loss is the results of otosclerosis.

## **How is otosclerosis treated?**

In many cases surgery is an option for treatment of otosclerosis. In an operation called a stapedectomy, a surgeon (otolaryngologist or otologist) bypasses the diseased bone with a prosthetic device that allows sound waves to be passed to the inner ear. It is important to discuss the risks and possible complications of this procedure, as

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well as the benefits, with the surgeon. In rare cases, surgery can worsen the hearing loss.

If the hearing loss is mild, surgery may not be an option. Also, on occasion, some hearing loss persists after surgery. A properly fitted hearing aid may help some people with otosclerosis in situations that include persistent hearing loss. A hearing aid is designed to compensate for a hearing loss by amplifying sound. A hearing healthcare professional can discuss the various types of hearing aids available and make a recommendation based on the specific needs of an individual.

## **What research is being done on otosclerosis?**

Scientists are conducting research to improve understanding of otosclerosis. Genetic studies continue in order to identify the gene or genes that may lead to this disorder. Other researchers are studying the effectiveness of lasers currently used in surgery, of amplification devices, and of various stapes prostheses. Improved diagnostic techniques are also being examined and developed.