



OTOSCLEROSIS

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



Key points

- A complex disorder of abnormal bone growth in the middle ear.
- Tiny bones that conduct sound knit with surrounding bone.
- Usually affects both ears.
- One of the most common causes of conductive hearing loss in young adults.
- Speed and extent of hearing loss is unpredictable.
- Causes dizziness and balance problems if bone growth extends into the inner ear.
- Hearing aids are usually the first treatment choice.
- Hearing improves for 80% of patients after minimally invasive surgery.
- Following a healthy lifestyle generally helps better manage the challenges of adjusting to unexpected hearing loss.

What is otosclerosis?

Otosclerosis (oh-toe-skler-OH-suhs) a complex disorder of abnormal bone growth in the middle ear. It most often happens when the tiny stapes (“STAY-peeZ”) bone knits with surrounding bone.

Otosclerosis usually results in slow, progressive conductive hearing loss.

When the stapes is unable to vibrate, hearing becomes impaired. Soundwaves no longer move efficiently from the eardrum to the inner ear. Far advanced otosclerosis can cause dizziness and affect balance, likely when abnormal bone growth extends into the inner ear.

Sometimes hearing loss in otosclerosis is relatively mild and stays that way. Usually, it worsens progressively. This may happen quickly or over many years. While otosclerosis can lead to severe hearing loss, it rarely results in total deafness.

Otosclerosis affects both ears (bilateral) in about 70 to 80% of cases. Patients often report hearing loss only in one ear (unilateral) in early stages of the disease.

Otosclerosis is one of the most common causes of hearing impairment in young adults. Most often, hearing loss is first noticed during the teens and twenties. Otosclerosis can develop in children and older people too. White people are most affected. Women are about twice as likely as men to be diagnosed with otosclerosis. Pregnancy can cause otosclerosis to advance more quickly.

Otosclerosis is rare, affecting about 3 in 1,000 people. Research suggests between 25 to 50% of people with otosclerosis have a family history of the condition.

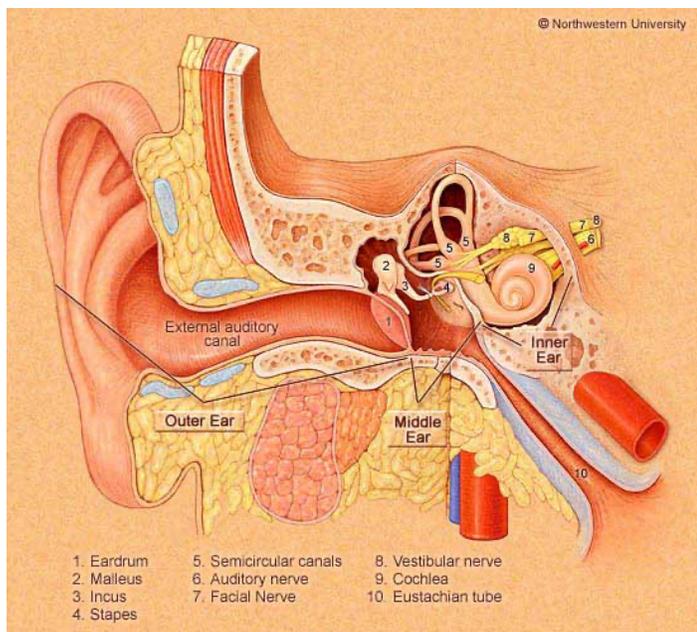
The word otosclerosis comes from Greek. It means abnormal hardening of body tissue (sclerosis) of the ear (oto).

How do we hear?

To understand why otosclerosis causes hearing loss, it is important to have a basic understanding of how we hear. For hearing to function normally a sound has to travel through all three parts of the ear: outer, middle, and inner. The first two are air filled; the latter is fluid filled.

The outer ear is made up of the part you can see on the side of your head (pinna) and the funnel-shaped external ear canal. The pinna gathers sound waves (vibrations) and channels them through the ear canal to the eardrum (tympanic membrane). This makes the eardrum vibrate, changing the sound waves into mechanical vibrations.

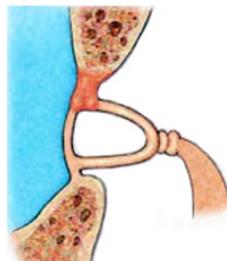
These vibrations are transferred to three tiny bones in the middle ear (maleus, incus and stapes – the Latin names for hammer, anvil, and stirrup) in the air-filled space of the middle ear. These bones, collectively known as the ossicles, are arranged to augment the vibration.



Cross section of the ear. Otosclerosis involves the small bones of the middle ear - the malleus (2), the incus (3) and the stapes (4), as well as the bone that surrounds the inner ear.

The stapes, the innermost bone, is attached to the snail-like cochlea (hearing organ) in the inner ear. There are tiny hairs and fluid within the cochlea.

When the stapes vibrates against a “window” in the cochlea’s wall, the fluid moves. This causes the hairs to bend and sway and produce electrical signals. These signals are sent to the brain through the vestibular nerve (eighth cranial nerve) to be interpreted as sound.



Hearing becomes impaired in otosclerosis when the stapes joins with surrounding bone of the middle ear. The stapes can no longer vibrate, and sound is not moved efficiently from the eardrum into the inner ear.

Types of hearing loss in otosclerosis

There are several types of hearing loss in otosclerosis:

- **Conductive hearing loss** happens in the early course of otosclerosis when bone remodeling impairs the mechanical transfer of sound by any or all of the bones in the middle ear. Most people with otosclerosis have conductive hearing loss. This process usually reaches its peak when people are in their 30s.
- **Sensorineural hearing loss** happens later on when remodeling happens in the bone surrounding the inner ear (otic capsule). This process is called otospongiosis. Mineral density decreases and the bone becomes sponge-like as it is remodeled. It is theorized that enzymes leak from the bone into the inner ear and cause sensorineural hearing loss. Wasting away (atrophy) of the outer wall of the cochlear duct (called the spiral ligament) and deterioration of its blood vessels (called the stria vascularis) also contribute to sensorineural hearing loss in otosclerosis.
- **Mixed conductive and sensorineural** Mixed hearing loss in all frequencies results in the last far advanced stage when otosclerosis extends into bone surrounding the inner ear (otic capsule) or the bony labyrinth of the inner ear.

What are the causes?

Bone is living tissue that is continually being broken down and remade. In otosclerosis, the process for remodeling bone does not work properly and abnormal bone forms. Scientists do not fully understand why this happens.

Contributing reasons may include:

- A faulty gene inherited from a parent. Researchers have recently identified a range of changes in the SERPINF1 gene that can cause otosclerosis.
- Exposure to the measles virus may play an important role in activating a gene responsible for otosclerosis. The exact role of measles in the development and progression of otosclerosis is not yet understood. Decreased cases of measles due to vaccination may be contributing to a continuing decline in the number of people with otosclerosis.
- Stress fractures to bones in the ear and the bony tissue surrounding the inner ear.
- Some studies suggest an autoimmune response in the body, possibly related to environmental or genetic factors, may be linked to otosclerosis.

Changes in levels of the hormone estrogen during pregnancy may play a role in worsening otosclerosis. In one study, women with otosclerosis in both ears experienced a 33% decline in hearing after a single pregnancy.

Otosclerosis often occurs along with a rare disorder called brittle bone disease (osteogenesis imperfecta).

What are the symptoms?

Symptoms of otosclerosis include:

- Gradually progressive hearing loss.
- Some people report particular difficulty hearing deeper or lower-pitched sounds and whispers.
- Some people report finding it easier to hear when there is background noise. This is called Willis paracusis or false paracusis.

- Speaking quietly because your voice seems too loud to you.
- Tinnitus (ringing in the ears) affects over 50% of people with otosclerosis.
- Vertigo (sensation of spinning or moving) and imbalance affects about 30% of patients. Vertigo develops when otosclerosis has moved into the inner ear, affecting the otolith organs and/or semicircular canals.

The symptoms of otosclerosis can be hard to tell apart from other causes of hearing loss. These include several mechanical disorders of the middle ear bones (ossicles), fluid within the middle ear (otitis media with effusion or serous otitis media), and even other inner ear disorders such as semicircular canal dehiscence (SCD).

How is it diagnosed?

See a general practitioner (family doctor) if you are concerned about your hearing. The doctor will ask about your symptoms, medical and family history, and do some basic hearing tests.

If otosclerosis is suspected, you will be referred to an otolaryngologist (an ear, nose and throat or ENT doctor) who will do a bedside examination and go over your family history. ENTs work closely with audiologists (hearing health professionals). You will likely have some of these diagnostic tests:

- **Full hearing assessment** including tests that measure hearing sensitivity (audiogram) and middle-ear sound conduction (tympanogram).
- **Acoustic reflex (AR) testing** to analyse the movement of the stapes within your ear. This test is done by presenting loud tones to the ear – it is quick and painless.
- **High-resolution CT** is beginning to be used in diagnosis and surgical planning for otosclerosis. It reveals variants in patient anatomy and severity of the disease, including cochlear involvement.

Some of the diagnostic tests are done to rule out other diseases or health problems. Tumours

(including acoustic neuroma), semicircular canal dehiscence and Ménière's disease are among the conditions with similar symptoms. Your doctor needs to consider all the possibilities before making a diagnosis and coming up with a treatment plan.

How is it treated and managed?

Treatment for otosclerosis depends on the severity and type of hearing loss, age and overall health, and patient preferences. Your doctor will discuss the risks and benefits of each treatment.

Approaches to treatment and management of otosclerosis include:

Observation

The progression of otosclerosis differs in extent and speed for different people. Some people never have more than minor hearing loss. It takes many years for hearing to worsen in others. Your doctor may first suggest monitoring your condition by having your hearing tested at least once a year. If your hearing continues to worsen, use of a hearing aid may be suggested.

Hearing aids

If otosclerosis has not progressed past the middle ear, hearing aids are usually the preferred first treatment choice to correct conductive hearing loss. They are a less risky alternative to surgery.

If you choose to try hearing aids, you will work with an audiologist to select and adjust to a suitable device. Hearing aids can be customized to amplify only the frequencies you need based on your hearing tests. As the disease progresses, adjustments can be made to increase amplification as needed.

Bone conduction hearing aids or bone anchored hearing aids (BAHA) are options for patients who:

- cannot or will not have surgery for otosclerosis
- have difficulty with, or no benefit from, wearing conventional hearing aids

Medical treatment

No medicines have been proven to improve hearing in persons with otosclerosis. Fluoride therapy was used in the past but is no longer recommended because of its negative effect on the hip and other bones. Evidence that sodium fluoride supplementation is helpful in otosclerosis is either limited or conflicting.

Surgery

Stapes surgery corrects conductive hearing loss by restoring the mechanical transfer of sound through the middle ear. It cannot, however, correct sensorineural hearing loss. Stapes surgery is increasingly uncommon and is not usually considered until less invasive treatments have been shown to be unsuccessful for a patient.

Stapes surgery is considered a minimally invasive procedure. There are two surgical variations:

- **Stapedotomy** (“stay-puh-DAW-tuh-mee”)

The surgeon makes a small incision above the ear canal (endaural incision). The eardrum is lifted up to access the middle ear. The top half of the stapes is removed. A small hole is made at the bottom (footplate) of the stapes and a prosthesis (man-made replacement) is put in place of the defective bone.
- **Stapedectomy** (“stay-puh-DEK-tuh-mee”)

This procedure is similar to a stapedotomy. The main differences are that the incision is larger and made inside the ear canal, and the entire stapes is removed.

Of the two procedures, the stapedotomy is generally preferred. It is less invasive, has fewer complications, and gives better hearing results at high frequencies.

80% of patients have significantly better hearing after surgery. Hearing improvement may not be noticeable immediately after surgery. Hearing usually improves as swelling decreases in the first three weeks after surgery. It can take up to three

months for the inner ear and eardrum to recover. Maximum hearing is reached in about six months.

Surgery may not, however, get rid of tinnitus. Nor will it improve hearing if otosclerosis has affected the hair cells in the cochlea (organ of hearing).

To be considered for stapes surgery, patients need good inner-ear function in both ears (bilateral) as well as in range conductive hearing tests.

“Red flags” likely to rule out surgery include:

- poor physical condition
- fluctuating hearing loss with vertigo
- ruptured eardrum
- infection
- out-of-range results on certain hearing tests

Surgical risks and complications are rare but can include:

- Further hearing impairment happens in about 2% of cases due to a variety of causes.
- Perforation (hole) in the eardrum happens in only about 1% of cases. It is usually due to an infection and heals on its own. If not, the hole can be repaired by a surgical procedure (myringoplasty).
- Injury to the facial nerve, usually temporary. Inner-ear fluid leakage (known as a “perilymph gusher”) happens when brain fluid escapes through the surgical opening because of an abnormal connection with the brain cavity. This is very rare and can be corrected with a lumbar drain (a small flexible tube inserted in the lower spine).
- Taste disturbance and mouth dryness are common in the first weeks after surgery. This disturbance persists in about 5% of cases.
- Tinnitus may develop or worsen.
- Vertigo and imbalance, usually resolvable over time and helped by vestibular rehabilitation, a type of exercise-based therapy. Its goal is to help train your brain to relearn how to balance

and how to respond to signals from the vestibular and visual systems.

- The prosthesis may shift position over time. It may “float” into the central part of the bony labyrinth of the inner ear (vestibule), causing immediate loss of hearing.

In most patients, otosclerosis usually continues to progress after surgery. For this reason, 10 to 20% of patients will need a second surgery (revision). Success rates for surgical revision are about 75%.

Cochlear implant

Cochlear implantation is rarely a treatment option and often comes only after unsuccessful otosclerosis surgeries. A cochlear implant (CI) is an electronic device that partly brings back sensorineural hearing loss. It comes with a risk of impaired balance.

Living with otosclerosis

It can be challenging to accept and live with otosclerosis, especially for young adults. Following a healthy lifestyle generally helps people better manage the challenges of adjusting to unexpected hearing loss. Helpful strategies include:

- learning how to cope better with a chronic condition
- practicing relaxation techniques
- eating well
- getting enough sleep
- keeping physically active

What to expect in the future

The progression of otosclerosis varies from person to person. How quickly, or to what extent, hearing will be affected is unpredictable. Regular hearing tests and consultations on how best to treat and manage your condition are likely to continue in the years ahead.

Researchers have recently succeeded in identifying the first gene involved in causing otosclerosis by using a new DNA-sequencing technique called “whole exome sequencing”. The research team is

continuing to work on identifying more otosclerosis genes. They are also looking into how mutations in identified genes cause otosclerosis.

Separate research studies include looking at:

- the role of inflammatory or autoimmune conditions in otosclerosis
- bisphosphonates (a class of drugs that stops the loss of bone density) and vitamin D as possible treatments

Hopefully research will lead to the development of drugs to treat – or prevent – otosclerosis.

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In addition, find information about how the balance system works, the journey from diagnosis to treatment, building a wellness toolkit, and more.

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