

# Otosclerosis

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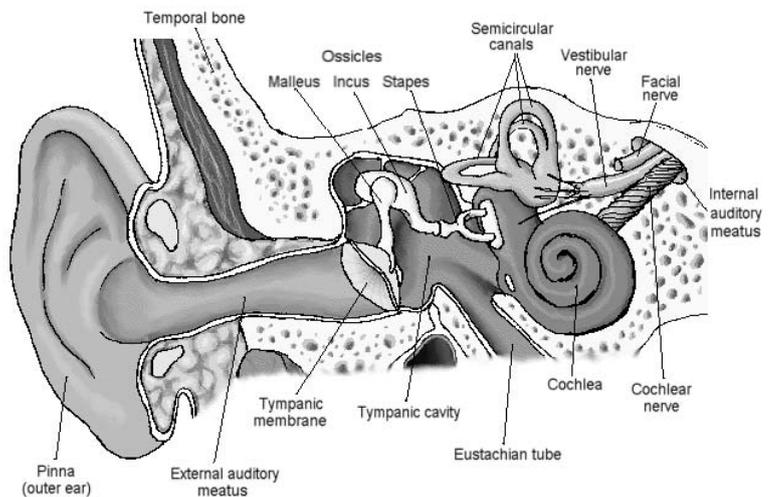
Issued August 2010. Version 1.4 revised March 2011. To be reviewed August 2013

This information has been written to help you understand more about otosclerosis; the most common cause of progressive deafness in young adults.

## What is otosclerosis?

Otosclerosis is the commonest cause of progressive deafness in young adults although the numbers of incidences seems to be reducing. The exact cause of otosclerosis is not fully understood. Genetic factors are involved, so the condition often (but not always) runs in families. Other factors thought to play a role include viral infections and in particular, infection with the measles virus, as otosclerosis appears to be less common amongst people who have been vaccinated against measles. It has also been suggested that otosclerosis may be affected by hormonal changes or that it may be a form of autoimmune condition in which the body's defence mechanisms attack the body's own tissues.

In someone with normal hearing the sound passes from the *tympanic membrane* (ear drum) to three small bones, or *ossicles*, which transmit the sound to the inner ear or *cochlea*. The *stapes*, or stirrup bone, is the innermost of these ossicles. It is the smallest bone in the body and sits in a hole or "window" into the cochlea. It is free to vibrate within the window, allowing transmission of sound. In otosclerosis the bone around the base of the stapes becomes thickened and eventually fuses with the bone of the cochlea. This reduces normal sound transmission resulting in a conductive deafness. In the early stages of otosclerosis, the cochlea and the nerve of hearing are not affected, though eventually they can be.



Both ears may be affected, although in men it is more common for one ear to be worse than the other. Untreated, the deafness gradually worsens and in a small percentage of people it can cause profound hearing loss. Other symptoms of otosclerosis can include tinnitus and balance problems. Pain is not usually a symptom of otosclerosis.

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## How is otosclerosis diagnosed?

The diagnosis will be made by a specialist, but there are some signs which may be noticed by you or your friends and family. A person with otosclerosis usually has speech which is quiet, while people with cochlear (or nerve) deafness usually speak loudly. The presence of extra background noise usually adds to the hearing difficulty of people with cochlear deafness, but in otosclerosis this confusion does not occur. A person with otosclerosis may even hear better in noisy surroundings, possibly because other people's voices are raised in frequency and loudness. Otosclerosis tends to affect the low frequencies as much as the high frequencies. In cochlear deafness the high frequencies are generally much more affected than low frequencies.

Diagnosis of otosclerosis by an ear specialist is usually straightforward. Examination of the ear will reveal a normal, healthy looking ear drum. Hearing tests with tuning forks and audiometric tests will show a conductive deafness. The specialist may also study the movement of the ossicles using a machine called a *tympanometer* to perform a test called the *stapedial reflex test*. This is non-invasive, quick and painless. Occasionally, scans of the ears may be performed though generally they are not required.

## Are there any medications which can help?

Fluoride treatment has been used with good results in certain forms of otosclerosis but is generally not favoured in the UK. If fluoride treatment is considered it is essential to know the concentration of fluoride in the patient's drinking water to ensure a correct dosage.

## What about a hearing aid?

Hearing aids are helpful with all kinds of conductive deafness, including otosclerosis. Indeed, they are usually more beneficial for otosclerosis than with nerve deafness.

## Why consider surgery?

As hearing aids work well and are completely safe, many patients with otosclerosis decide not to undergo surgery. However, surgery does offer the chance of returning the hearing to normal so that a hearing aid is not required. Surgery can also have a stabilising effect on the otosclerotic process and can offer some degree of protection against otosclerosis advancing to the inner ear.

## What does surgery entail?

Surgery for otosclerosis started in the nineteenth century but it was not until the late 1950s that the modern operation *stapedectomy* was developed. Although the term stapedectomy is still in widespread use, some surgeons prefer to call the operation *stapedotomy*. The original stapedectomy operation involved removal of the stapes whereas a stapedotomy is a more delicate operation that leaves part of the stapes in position. The operation can be performed under local or general anaesthesia; in the UK a general anaesthetic is more commonly used.

During a stapedotomy, the surgeon operates down the ear canal using an operating microscope, so usually there are no external scars afterwards. The eardrum is turned forward, and the upper part of the stapes is removed using extremely fine instruments. A small hole is then made through the lower part of the stapes - the part that is fused with the bone of the cochlea. This very precise hole is made in the stapes footplate with a micro drill or laser. A prosthetic stapes is then placed between the *incus* (the middle of the three ossicles, also known as the anvil) and the hole that has just been created. The prosthesis is available in a wide variety

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of materials: Teflon, polyethylene and stainless steel are among those commonly used. The ear drum is then returned to its normal position and a small dressing is placed in the ear canal. The operation generally takes about one hour.

The patient is usually allowed home within a day or two, and generally needs two or three weeks off work; strenuous exercise must also be avoided. The dressing in the ear canal about a week after the operation and this can be performed within an outpatient clinic. However, because of the bruising and swelling inside the ear from the surgery, it may be a while before the full improvement in hearing is noticed, sometimes as long as 3-6 weeks. Most surgeons advise their patients that they should not travel by airplane for several weeks or even months after surgery.

Many surgeons modify the basic technique. For example, some surgeons remove a piece of very fine vein from the patient's hand and place this as a graft over the hole in the stapes before positioning the prosthetic stapes. This aims to reduce the risk of damage to the inner ear.

### Does surgery help the tinnitus?

Several research studies have looked at the effect of stapedectomy on tinnitus, and the majority of tinnitus patients are helped by the surgery. However, there is a small risk that the surgery will not help and an even smaller risk that it might make tinnitus worse. Figures for improving or eliminating tinnitus are quoted at 78% or better. The risk of making the tinnitus worse is quoted at 6% or less.

### Are there any risks?

No operation is entirely free from risk, despite modern anaesthetics and surgical skills.

- Hearing - the original stapedectomy operation involved making a relatively large hole through the base of the stapes. This operation produced good hearing in 85%, slight improvement in 10% and worsening in 5%. About 2% had severe hearing loss due to damage to the cochlea. Modern techniques have improved this figure but there is still a risk of worse hearing after the surgery.

It is important for patients to ask their surgeon for his or her success rates. In any surgery, it is wise to select a surgeon who has a special interest in the field, and undertakes the procedure on a regular basis with good results. Any surgeon should be prepared to discuss these issues, and if their interests and expertise are in another area of otolaryngology (ear, nose and throat surgery), they may wish to refer the patient to a colleague who specialises in this kind of operation.

- Balance - giddiness or unsteadiness is common immediately after the operation but usually clears within a few days.
- Taste - sometimes bruising can occur to the small nerve concerned with taste which runs just under the ear drum. As a result, some patients experience a metallic taste on the side of the tongue for a month or two after the operation. It generally settles down in the long term.
- Others - if a metallic prosthesis is used the patient may not be able to have a Magnetic Resonance Scan (MRI) of the head in the future. Anyone who has had a stapedectomy should mention this to the radiology department before undergoing MRI scanning. Plastic prostheses are not affected by MRI scans.

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Other forms of scanning such as Computed Tomography (CT) scans are completely safe after stapedectomy, whatever the type of prosthesis.

## What happens later in life?

There are a small number of patients (less than 1%) who have undergone a successful stapedectomy and then experience sudden hearing loss many years after the operation. However, these are very much a minority. There are many people who had stapes operations thirty five years ago or more and the vast majority still have useful hearing. It does seem that in some respects the operation halts the course of otosclerosis.

A certain degree of "old age hearing loss" affects everyone eventually and patients with otosclerosis are no different from the rest of the population in this respect. It is usually possible to correct this later loss of hearing with low powered hearing aids.

## Second ear surgery

One of the other arguments is whether or not the operation should be done on the second ear, in situations where both ears are affected. Some authorities feel that the second ear should not be operated on. If anything should go wrong with the first operated ear at a later date, it is still usually possible to hear something using a hearing aid in the un-operated ear. If the taste nerve was damaged in the first operation, a second operation carries the risk of a total loss of taste. However, increasingly, many experienced surgeons are prepared to operate on the second ear. It is a decision that needs to be made with great care, and the patient must be aware of the risks involved.

## Dos and Don'ts

- Do try a hearing aid if you are not sure about surgery and you have a relatively slight hearing loss.
- Do ask your General Practitioner for a second opinion if you are not completely happy about the advice you have received.
- Do discuss the technique that will be used and the results that may be expected with the surgeon who is going to perform the operation. Make sure that any figures being quoted are those of your surgeon, not figures from some internationally famous stapedectomy expert. If your surgeon cannot tell you about his or her figures, go somewhere else!
- Don't consider any surgery unless you have a very clear idea of the risks and benefits that are involved.
- Do go ahead and have the operation once you are entirely happy: the most likely outcome is restoration of normal hearing which can dramatically improve your life quality.

## References:

Full references for this article can be obtained from the address below.

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