



Otosclerosis

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What is Otosclerosis?

Otosclerosis is the commonest cause of progressive deafness in young adults. Otosclerosis is often inherited, although isolated cases do occur. Both ears may be affected, although in men it is commoner for one ear to be worse than the other. Untreated, the deafness gradually worsens and can cause a profound hearing loss.

In someone with normal hearing the sound passes from the 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 inner most 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 these too may be involved.

This means that freeing the stapes in some way can restore the hearing by removing the conductive block.

How to tell if it is Otosclerosis?

The diagnosis will be made by the specialist, but there are some clues which may be noticed by the hearing impaired person or his friends and family. The speech is usually quiet, while those suffering from cochlear (or nerve) deafness usually speak loudly.

The presence of extra background noise usually adds to the confusion 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 more than the high frequencies. In cochlear deafness the opposite usually occurs.

Diagnosis of otosclerosis by an ear specialist is not usually difficult. 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.

Are there any medications which can help?

Fluoride treatment has been used to good effect 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.

Are there any medications to avoid?

Otosclerosis can react to female hormones. Hence pregnancy can accelerate the condition. There have been suggestions that the Oral Contraceptive Pill and Hormone Replacement Therapy (HRT) can also worsen the condition. However, many doctors dispute this. Any woman with otosclerosis who is contemplating using the Pill or HRT should discuss the matter thoroughly with her doctor.

What about a hearing aid?

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

Why consider surgery?

Because 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 the patient does not have to wear a hearing aid at all. Also surgery seems to have a stabilising effect on the otosclerotic process and offers some

degree of protection against otosclerosis advancing to the inner ear.

What does surgery entail?

Surgical interest in otosclerosis started in the nineteenth century but it was not until the late 1950's that the modern operation of stapedectomy was developed. The operation can be performed under local or general anaesthesia but in the UK a general anaesthetic is more commonly used.

The surgeon works down the ear canal using an operating microscope so there are no external scars afterwards. The eardrum is turned forward, and the upper part of the stapes is removed with 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 many forms and can be made from a wide variety 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 an hour. The patient is usually allowed home with in a day or two though will need to avoid strenuous exercise and will probably need two to three weeks off work. The dressing in the ear canal is easily removed in the outpatient clinic, usually about a week

after the operation. However, because of the bruising and swelling inside the ear from the surgery, it may be awhile 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 with the aim to reducing the risk of damage to the inner ear.

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, 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 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 (similar to an X-ray) of the head in the future.

What happens later in life?

There is 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 the disease.

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, 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 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.

Do's 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

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This information is not a substitute for medical advice. You should always see your GP / medical professional

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