



# OTOSCLEROSIS

This factsheet has been written for anybody who is interested in the condition known as otosclerosis. It may be particularly useful if you have been diagnosed with otosclerosis, or think that you might have it. The factsheet covers the following topics:

- What is otosclerosis?
- What causes otosclerosis?
- What treatments are available?
- What is cochlear otosclerosis?

## WHAT IS OTOSCLEROSIS?

Inside the middle ear are the three smallest bones in the human body. These bones, called the ossicles, are hinged together and rock back and forth in response to sound vibrations from the eardrum. This rocking motion passes the vibrations through the middle ear and into the cochlea (or inner ear) where they are processed and sent along the auditory (hearing) nerve to the brain.

The term otosclerosis refers to a hardening of the bone surrounding the stapes, one of the three ossicles. This becomes progressively fixed in the 'oval window', which divides the air in the middle ear from the fluids in the inner ear. The resulting stiffening of the whole ossicular chain inhibits the transmission of sound to the inner ear. When sound is inhibited in this way it is known as a conductive hearing loss.

Otosclerosis is one of the more common causes of deafness amongst people in their 20s, possibly occurring more frequently in women than men. If left untreated, the hearing loss will become progressively worse.

## WHAT CAUSES OTOSCLEROSIS?

In many cases otosclerosis runs in the family. People in the same family who have otosclerosis usually have similar symptoms; that is, it will start at about the same age and progress at about the same rate, but these features can be quite different when different families are compared.

It is possible that there are a number of different genetic forms of the condition and that there are other, non-genetic, causes. In genetic cases, otosclerosis is carried by mistakes in the 'autosomal dominant' gene. Children of those affected by genetic otosclerosis will therefore have roughly a one in six chance of developing the condition.

The underlying causes of the condition may not always be the same, but all seem to involve a fault in the normal process of rebuilding bone, which happens continually throughout the body. Finding the gene or genes responsible for otosclerosis could help identify the chemicals which control the building process and lead to ways of preventing or treating the condition.

## **WHAT TREATMENTS ARE AVAILABLE?**

Surgery has been available for a long time. The first operations were carried out in the 1870s, although these early attempts (without microscopes or antibiotics) sometimes had unfortunate results for both the ear and the patient. However, by the late 1930s an effective technique called 'fenestration' had become very popular. Although the immediate results were good, over time the condition would return.

Until recently the standard procedure was a 'stapedectomy' (i.e. the removal of the stapes). This is a relatively simple operation lasting about an hour, and is performed down the ear canal with the aid of an operating microscope. There are no visible signs of surgery.

During this procedure the eardrum is turned forward allowing the surgeon access to the ossicles. The stapes, a stirrup shaped bone, is removed and replaced with a small prosthesis. This is placed between the oval window of the cochlea (or inner ear) and the incus, the middle bone in the ossicular chain, restoring the means of sound transmission.

The operation has a high success rate - up to 90% of those undergoing stapedectomies report noticeable improvements. However there is a small chance that damage to the cochlea may cause hearing loss or tinnitus. Directly after the operation there may be some temporary giddiness as a result of the leakage of inner ear fluid. Patients may also perceive a metallic flavour in their mouths due to the bruising of the chorda tympani nerve. Situated just under the eardrum, this nerve is concerned with the sensation of taste.

A modification to this procedure, 'stapedotomy', seems to have reduced the chances of inner ear damage and post-operative giddiness. In this procedure the surgeon drills a tiny hole in the stapes footplate with a microdrill or laser. A very fine vein graft covers the hole and a piston is placed into this and attached to the

Incus, this seals off the inner ear fluids immediately after the operation, reducing the risk of leak, which is one of the causes of stapedectomy failure.

However, surgery is not the only treatment option and may be inappropriate in many cases. Patients should consult their specialist for more information on the options available and should not presume surgery will be the best or only option.

Hearing aids, while not curing the deafness, are always helpful and if they are not, it is unlikely that surgery will help. Hearing aids have improved greatly since the time that stapes surgery was first practiced. Modern hearing aids are a much more satisfactory alternative to surgery than they used to be, particularly in the early stages of otosclerosis. However because otosclerosis is progressive, stronger, more powerful aids may be needed as time goes by.

## **WHAT IS COCHLEAR OTOSCLEROSIS?**

On rare occasions, the spongy bone growth associated with otosclerosis can spread from the ossicular chain into the cochlea. Here, damage to the tiny 'hair' cells can result in a sensorineural deafness. Cochlear otosclerosis is permanent and cannot be helped by surgery. However some medications may slow down the progress of the condition. Otosclerosis may also spread to the vestibular canals causing episodes of unsteadiness.

## **WHAT RESEARCH IS BEING CONDUCTED INTO HEREDITARY CONDITIONS WHICH CAUSE HEARING LOSS?**

Deafness Research UK scientists are currently looking into the genetic causes of conditions such as Usher Syndrome, Branchio-Oto-Renal Syndrome and Ménière's Disease.

Research specifically into the causes and treatment of otosclerosis remains seriously under-funded. In coming years, Deafness Research UK aims to ensure that the results of its work in developing an understanding of the genetic causes of deafness can pave the way for research to identify the causes of otosclerosis and, eventually, develop new and more effective treatments.

## FURTHER INFORMATION

Contact the Deafness Research UK Information Service for further information about research into otosclerosis.

If any of your questions concerning otosclerosis have not been answered by reading this factsheet, contact the Deafness Research UK Information Service for further assistance. Our Information team will either answer your enquiry directly or refer it to one of our scientific or medical advisers.

Open: 9.00 a.m. to 5.00 p.m., Monday to Friday (a message can be left at other times).

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Deafness Research UK is the only national medical research charity dedicated to helping people with deafness, tinnitus or other hearing problems.

Scientists are now predicting that within the next ten to fifteen years there could be a cure for some forms of deafness and much more effective treatments for tinnitus. Deafness Research UK is at the forefront of this work.

You can support us by making a donation or joining the Deafness Research UK League of Friends. For more information call us on 0207833 1733 or write to:

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