FACTSHEET
Hearing health

ACTION ON HEARING LOSS
A national charity since 1911

Otosclerosis

This factsheet tells you about otosclerosis, a common cause of gradual hearing loss in young adults. It explains what the condition is, and how it's diagnosed and treated.

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Medical disclaimer

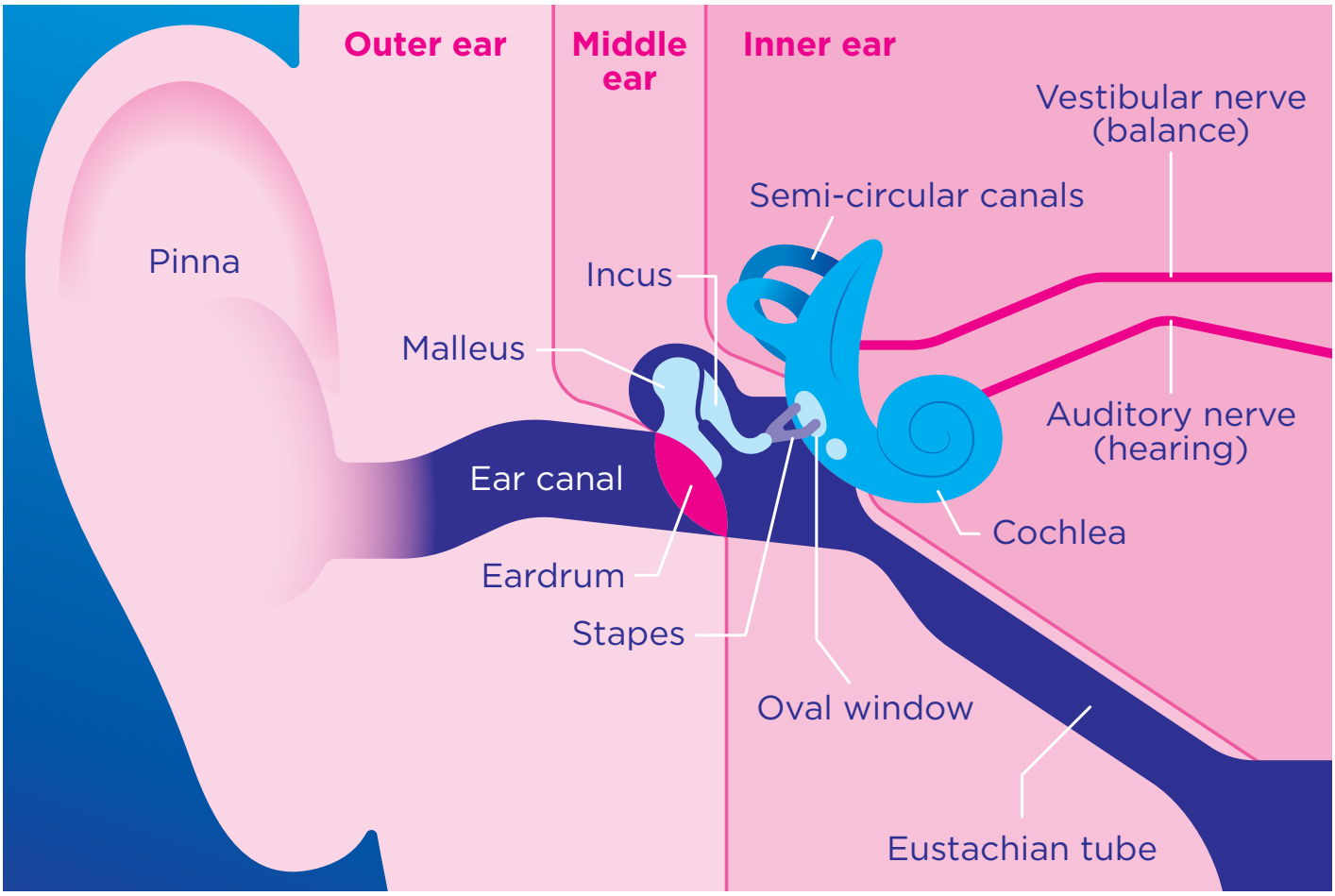
The information given in this factsheet is not medical advice and, by providing it, Action on Hearing Loss does not undertake any responsibility for your medical care, or accept you as a patient. Before acting on the information contained in this factsheet, or deciding on a course of treatment, you should discuss the matter with your GP or other medical professional.

How do we hear?

Our ears have three parts: the outer, middle and inner ear (see diagram below).

The **outer ear** consists of the pinna, which is the part you can see on the side of your head, and the external ear canal. Sound waves (vibrations) are gathered by the pinna and travel down the external ear canal until they reach the eardrum, causing it to vibrate.

The **middle ear** is an air-filled space behind the eardrum that contains a chain of three tiny bones called the ossicles (the malleus, incus and stapes). Vibrations pass from the eardrum to the ossicles, and the ossicles move back and forth to pass the sound waves into the inner ear.



The **inner ear** contains the cochlea, which is our organ of hearing. It's lined with thousands of tiny sensory cells known as hair cells. When sound waves enter the cochlea, the hair cells trigger electrical signals in the hearing nerve. The hearing nerve sends the electrical signals to the brain, which recognises them as different sounds – for example, people talking or footsteps.

What is otosclerosis?

Otosclerosis is a condition that mainly affects the stapes, one of the tiny ossicles (bones) in the middle ear. For you to hear properly, the ossicles need to be able to move freely, so they can pass sound waves into the inner ear. In otosclerosis, abnormal bone gradually grows around, and onto, the stapes, which reduces its movement. This causes hearing loss, because the ossicles can't pass sound waves into the inner ear as efficiently as they used to. Eventually, the stapes becomes fixed so it can't move at all – this can cause severe hearing loss.

In most cases, otosclerosis just affects the stapes. But sometimes the cochlea is affected too.

Otosclerosis usually affects both ears, but sometimes just one.

What causes otosclerosis?

Bone is a living tissue that is continually being broken down and remade. In otosclerosis, it seems that the process for making new bone doesn't work properly and abnormal bone forms. The reason why this only affects the stapes, and sometimes the cochlea, isn't entirely clear.

While the exact cause of otosclerosis is unclear, between a quarter and a half of cases are thought to be caused by a faulty gene being inherited from a parent. Research that we've funded has recently identified the first gene to be linked to otosclerosis (we tell you more about this on page 6).

In people who don't inherit a faulty gene, it's been suggested that the condition may be linked to a number of factors, including the measles virus. But it's still unclear what role – if any – these factors actually play in otosclerosis.

Who gets otosclerosis?

As otosclerosis tends to run in families, people who have a family history of the condition are more likely to develop it. But some people with otosclerosis have no family history of the condition.

Otosclerosis usually develops in people in their late 20s or their 30s, but it can develop in younger people too.

Twice as many women as men are diagnosed with otosclerosis. Some women report that the condition gets worse during pregnancy; it's thought that this may be due to the high concentration of the hormone oestrogen during pregnancy.

If you have otosclerosis and are worried that pregnancy may affect your hearing, see your doctor. You may have to have a hearing test from time to time to monitor your hearing.

Otosclerosis can also worsen for some women during menopause, perhaps due to changing hormone levels.

How can I tell if I have otosclerosis?

The main symptom of otosclerosis is hearing loss. It can take a few months to a few years for the stapes to become fixed, so it may take a while for you to notice that your hearing isn't what it used to be. Your hearing loss may stay relatively mild, but in most cases it becomes worse – either quickly or over many years.

Hearing loss usually makes people speak loudly and struggle to hear when there's background noise. But if you have hearing loss caused by otosclerosis, in the early stages you may hear *better* when you're chatting to someone in a busy, noisy place. This may be because the hearing loss caused by otosclerosis can initially make it difficult to hear deeper or lower-pitched sounds – voices usually become raised in volume and pitch in noisy environments, so you may find them easier to hear. As the condition progresses, however, it's likely that you'll start to notice other sounds being difficult to hear too, and you may start to struggle to hear in noisy places.

Other symptoms of otosclerosis include:

- **Tinnitus** – this is the name for sounds that you hear in your ear(s) or head that don't have an external source. You may hear different sounds including ringing, hissing, buzzing, roaring or machine-type noises. Four out of five people with otosclerosis have tinnitus.
- **Vertigo** (the sensation of spinning or moving) – this is a rare symptom, but you may have it if otosclerosis affects the balance system in your inner ear.

If you have any of these symptoms, see your GP.

How is otosclerosis diagnosed?

If you notice that you can't hear as well as you used to, or if you have tinnitus, see your GP. They will ask you about your symptoms and check your ears – they may also carry out some simple hearing tests. Your GP should then refer you to a hearing specialist for a full hearing assessment. The specialist may look at the movement of the stapes within your ear – this test is very quick and isn't painful.

How is otosclerosis treated?

As the hearing loss caused by otosclerosis worsens, you may find hearing aids very helpful. You also have the option of having an operation to replace the stapes with an artificial bone – this is usually very successful.

Hearing aids

You'll usually be given the chance to try hearing aids before surgery is considered. You can get digital hearing aids for free from the NHS, or you can buy them privately. A hearing specialist will tell you about the different types of hearing aid available and recommend a type to meet your needs.

Surgery

The operation, called a stapedectomy, aims to improve your hearing by replacing the stapes with an artificial bone made of plastic or metal. If both of your ears are affected by otosclerosis, you'll usually have the operation on the ear that has the most hearing loss. Further down the line, it may be possible to have surgery on your other ear.

The operation usually takes about an hour. You'll either have a general anaesthetic (to put you to sleep) or a local anaesthetic (to numb your ear), so you won't feel anything. The surgeon will make a cut inside your ear canal and remove the top part of the stapes. They will make a small opening at the base of the stapes and insert the artificial bone, so that it can transfer sound waves from the remaining ossicles into the inner ear.

Eight out of 10 people find the operation a success and report a good improvement in their hearing. The operation may not cure tinnitus, however, and it won't improve your hearing if otosclerosis has affected the hair cells in the cochlea.

Are there any risks to having surgery?

Although rare, there are some complications that can arise if you have a stapedectomy. Your surgeon should discuss with you the risks and benefits of the operation before you have the surgery.

The potential complications include:

- losing more or all of your hearing – this may happen if your inner ear is accidentally damaged during the operation
- vertigo (spinning dizziness) – this is usually temporary
- a hole in the eardrum
- altered sense of taste – this is usually temporary
- tinnitus (new or worsened)
- weakness of the muscles in your face – this is rare and usually only short-lived.

What happens after the operation?

It's likely that you'll be able to go home the same day as the operation, or the day after. You may have some earache but the hospital can give you painkillers. You may also get a small amount of discharge from your ear canal.

For the first few days after surgery, you may feel dizzy when you make sudden head movements or stand up quickly. Rarely, this dizziness can last several weeks. Speak to your surgeon if you experience this or any other problems after you leave the hospital.

You may need to take up to three weeks off work, depending on how you feel.

After the operation, you should:

- keep your ear dry for a few weeks
- avoid straining and lifting anything heavy for a few weeks
- only blow your nose gently
- avoid air travel for two months
- avoid diving or flying when you have a cold, if possible.

You'll receive an appointment to go back to the hospital two to three weeks after the operation to have the stitches and the dressing in your ear canal removed.

Your hearing won't return to normal straight away, as your inner ear, including your eardrum, will need time to recover – this can take up to three months.

Once your eardrum has healed, it's likely that you'll notice an improvement in your hearing.

What otosclerosis research is Action on Hearing Loss funding?

Identifying the genetic causes of otosclerosis is crucial to finding effective treatments for it. Until recently, no genes had been definitively linked to otosclerosis, but now, in research funded by us, a team led by Dr Sally Dawson at the UCL Ear Institute has discovered the first gene to be linked to the ear condition.

Dr Dawson and her collaborators collected DNA samples from people with inherited otosclerosis and studied them, looking for 'changes' in their DNA that may be linked to the condition. In doing so, they identified a gene that causes otosclerosis in some families.

Knowing the genes that are involved in causing a condition means that the underlying processes can be better understood. This in turn can lead to the development of treatments that target these processes and correct them when they go wrong.

We are continuing to fund Dr Dawson and her team as they search for more genes involved in otosclerosis. To find out more about this project, and our other biomedical research, visit actiononhearingloss.org.uk/your-hearing/biomedical-research/projects-and-research/projects.aspx

Where can I find out more about otosclerosis?

British Tinnitus Association

The BTA campaigns for better services for people with tinnitus. It supports a network of tinnitus groups, provides a range of publications and produces a quarterly magazine, *Quiet*.

Ground Floor
Unit 5, Acorn Business Park
Woodseats Close
Sheffield
S8 0TB

Telephone: **0800 018 0527**
Textphone: **0114 258 5694**
Email: info@tinnitus.org.uk
Website: tinnitus.org.uk

ENT UK

A professional body representing clinicians working in ear, nose and throat (ENT) medicine. Patient information is available on a wide range of ENT conditions.

Telephone: **020 7404 8373**
Fax: **020 7404 4200**
Email: entuk@entuk.org
Website: www.entuk.org

Information you can trust

The Information Standard certifies us as producers of high-quality, evidence-based information.

Thank you to **Dr Sally Dawson, Lecturer at University College London Ear Institute**, for reviewing this factsheet for us and making sure that our information is accurate and based on the latest evidence.

For a list of references for this factsheet, please email references@hearingloss.org.uk

Did you find this factsheet helpful?

We'd love to know what you think of this factsheet – please email us at reviewpanel@hearingloss.org.uk

If you'd like to join our Readers' Panel, to help us create new publications and improve existing ones, please let us know.

Further information from Action on Hearing Loss

Our expert information covers everything you need to know about:

- hearing loss and deafness
- tinnitus
- ear problems and treatments
- hearing aids and cochlear implants
- useful products and technology
- communication tactics and support
- benefits and grants
- your rights.

Visit our website actiononhearingloss.org.uk or call our Information Line (see last page) for information, support and publications. You can also find out about services in your area, our hearing research, and how you can get involved.

Please help us support others

We provide our leaflets, factsheets and Information Line service free of charge to anyone affected by deafness, tinnitus or hearing loss in the UK. We rely on the generosity of our supporters to help us do this. We would be very grateful if you would consider making a donation – of as little or as much as you can afford.

Please send a cheque, payable to Action on Hearing Loss, to:

Freepost RTLX-CZKX-BTTZ
Action on Hearing Loss
1–3 Highbury Station Road
London N1 1SE
(No stamp needed)

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0203 227 6185

Our purpose is to help people confronting deafness, tinnitus and hearing loss to live the life they choose. We enable them to take control of their lives and remove the barriers in their way.

To find out more about what we do and how you can support us, go to **actiononhearingloss.org.uk**

Action on Hearing Loss Information Line

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