



Sudden Sensorineural Hearing Loss

This factsheet has been written for people who have experienced a sudden loss of hearing and who want guidance on what action to take or who have been diagnosed with the condition and want to understand it better. It may also inform people with relatives or friends who have lost their hearing suddenly. The factsheet covers the following areas:

- What is Sudden Sensorineural Hearing Loss?
- Some of the possible causes
- What to do if you lose your hearing suddenly
- Some of the possible treatments
- How to cope with losing your hearing
- Who to contact for more information and support

WHAT IS SUDDEN SENSORINEURAL HEARING LOSS?

Sensorineural hearing loss may be either sensory (affecting the sense organ of hearing in the cochlea) or, less commonly, neural (affecting the nervous pathways which connect the cochlea to the hearing centres in the brain).

Generally, sensorineural hearing loss is described as sudden if you notice a drop in your hearing instantaneously or over a short period not exceeding three days. You may notice a popping sound when it happens, or you may detect it when you first wake up or use the ear in a testing situation, like making a phone call. Sudden sensorineural hearing loss (SSHL) is considered a medical emergency and you will need to contact your doctor immediately. It currently affects about 20 in every 100,000 people.

As middle ear infection or accumulation of fluid behind the tympanic membrane (eardrum) can also occur rapidly, causing a temporary conductive hearing loss, and these conditions are more common than SSHL, it is important to obtain a specialist examination with hearing tests, to distinguish them from SSHL.

WHAT CAUSES SUDDEN SENSORINEURAL HEARING LOSS?

In 85 per cent of cases of sudden sensorineural hearing loss the cause is unknown. These cases are referred to as idiopathic. In the majority, the hearing is affected in only one ear (unilateral) and may return to normal or near normal in 70 per cent of cases.

Scientists are focusing on two main theories as to why sudden hearing loss occurs in these unknown cases. The first is that it may be of viral origin, as some viruses are known to damage the hearing, and a viral infection seems to precede the hearing loss in some people. The second theory is that it may be due to some vascular defect (relating to blood vessels), but there is no conclusive proof at present to support either view.

In the 15 per cent of cases where it is possible to establish a firm cause of SSHL the cause may be due to the following:

Infections

Meningitis is one of the commonest causes of severe or profound acquired deafness in babies and children, though cases are falling due to the availability of vaccines. Hearing loss due to meningitis usually affects both ears (bilateral). Anybody who has suffered from meningitis, especially a child, should have their hearing tested as soon as possible after recovery.

Measles and mumps are also associated with SSHL. In measles, the loss is usually moderate to profound and affects both ears, whereas in mumps it usually affects only one ear. The herpes zoster virus may, on rather rare occasions, cause sudden hearing loss accompanied by weakness of the facial muscles and vertigo. This so-called Ramsay Hunt syndrome may affect adults who have been close to children with chicken pox, which is caused by the same virus.

Some infections are confined to the ear itself. Labyrinthitis is an inflammation of the inner ear, caused by bacterial or viral infection, which can cause dizziness, tinnitus (noise heard in the ear or head with no external source), and can lead to sensorineural hearing loss, but usually only in one ear. Bacterial labyrinthitis often results in permanent hearing loss. One common cause of bacterial labyrinthitis is an existing chronic middle ear infection, often with cholesteatoma (invasion of skin into the middle ear) invading the cochlea.

Head injuries

Head injuries, especially those associated with a fractured skull, may produce profound and often permanent hearing loss. Even where there is no fracture, sensorineural hearing loss may occur, caused by damage to the central nervous system or the inner ear itself.

Noise

Hearing loss caused by exposure to loud noise is usually gradual in onset and can be prevented by avoiding noisy situations or wearing appropriate ear protection. However, sudden hearing loss may occur from exposure to excessively loud

noises, for example from blast injuries (from a nearby explosion) or from firearms or fireworks, especially in enclosed spaces. The degree of hearing loss can range from total, permanent deafness in one or both ears to a relatively minor high frequency loss. Where minor, hearing may recover spontaneously in time. For more information about the effects of noise, see the Deafness Research UK factsheets on this topic.

Ear surgery

Sensorineural hearing loss may occur after any surgical procedure on the ear, and the degree of risk depends on many factors. These include the nature of the procedure, the underlying disease and the skill of the surgeon. The hearing loss may occur immediately, in the few days following the operation, or even many years later. It is important to balance the potential benefits of surgery against the risks to hearing, and these issues should be discussed with a specialist.

Barotrauma

Barotrauma may occur when the ear is exposed to sudden pressure changes, possible when flying or diving. Most commonly, the middle ear is affected, causing a conductive hearing loss. It is rarer for the inner ear to be involved, but sensorineural hearing loss can occur if the membrane of the round window (one of the two windows which separate fluid in the inner ear from air in the middle ear) ruptures, causing a leakage of perilymph (one of the inner ear fluids).

Ototoxicity

Sudden sensorineural hearing loss is sometimes attributed to ototoxic drugs (drugs that may damage the inner ear) however, the deafness related to ototoxicity is more often gradual than sudden, and is often preceded by tinnitus. Drugs that are known to cause permanent hearing loss are usually given only when no other alternative exists for treating a life-threatening disease. If the prescription of such a drug is necessary, the potential effect on hearing should be explained by the patient's doctor and the hearing monitored regularly during treatment.

Ménière's Disease

Ménière's Disease affects about one in every two thousand adults in the UK. It is a disease associated with later life, usually starting between the ages of 30-50 years. Ménière's Disease is characterised by severe attacks of vertigo combined with fluctuating deafness, tinnitus and a feeling of pressure in the affected ear. Attacks often start without warning and can lead to loss of confidence, anxiety and sometimes depression.

Other causes

Less common causes of sudden sensorineural hearing loss include an acoustic neuroma, which is a benign (non-malignant) tumour. The hearing loss associated with acoustic neuroma is usually gradual and often unnoticed, but may be sudden in as many as 10% of cases. Even rarer is the neurological condition of multiple sclerosis, in which 'plaques' of the disease may affect parts of the brainstem associated with hearing. The diagnosis of this condition is made by electrical tests of brainstem function and a magnetic resonance imaging (MRI) scan, and fortunately the hearing often recovers spontaneously.

Immunological disorders

Disorders affecting the immune system may have a role in causing sudden sensorineural hearing loss but the link is, at best, uncertain. There is a certain amount of debate over the diagnosis of the condition known as Autoimmune Inner Ear Disease (AIED), its relationship with sudden deafness, and potential treatments, but it probably accounts for less than one per cent of all cases of hearing loss.

WHAT SHOULD YOU DO IF YOU LOSE YOUR HEARING SUDDENLY?

As a sudden hearing loss can return to normal or near normal in some cases, it may be tempting to adopt a 'wait-and-see' policy. However, if you think you have a sudden hearing loss in one or both ears, you should be seen by a specialist as soon as possible because even where a cause is unknown any treatment may be more successful the earlier it is given.

You should go to your GP (family doctor) and ask for an urgent referral to an ear, nose and throat (ENT) specialist. It is especially important to make your GP aware that the hearing loss was sudden to ensure you receive prompt medical attention. Family doctors may not have the facilities to distinguish SSHL from the commoner conductive, middle ear hearing loss, and if an incorrect assumption is made that the deafness is caused by a fluid build up or a middle ear infection; decongestants may be prescribed and some weeks may pass before a correct diagnosis is made. If you can't see your GP, and you have a severe sudden hearing loss, you should go to your nearest hospital accident and emergency department.

HOW CAN SUDDEN SENSORINEURAL HEARING LOSS BE TREATED?

As with any medical condition, treatment depends upon the cause, when the cause is known. As the cause is not known in so many cases of Sudden Sensorineural Hearing Loss, the treatment strategies may vary, being dependent on the doctor's observation and experience.

Treatment strategies used can include steroids, vasodilation (treatment to relax blood vessels), antiviral agents, diuretics and low-salt diets. The most often used of these is high-dosage steroid therapy either given orally or intravenously (into the bloodstream). However, after a course of treatment between 30 and 50% of patients show no response. New research suggests that better results are obtained if a course of intravenous steroids is followed up by a course of intratympanic steroids (steroids administered directly into the middle ear), but further trials are required to establish a firm evidence base.

Some viral infections respond to anti-viral agents like acyclovir, and for the Ramsay Hunt syndrome short-term treatment with steroids and acyclovir is justified if started early enough, as it may lead to a reversal of the hearing loss to near-normal thresholds.

There is no evidence of cardiovascular disease in the majority of patients suffering from SSHL, but there are certain 'vascular' conditions which may occasionally

cause sudden deafness. These include reduced blood flow in the cochlea and disorders of coagulation (clotting) of the blood. Research is being carried out on the potential value of carbogen inhalation in such cases, carbogen being a mixture of 95% oxygen and 5% carbon dioxide.

Sudden Sensorineural Hearing Loss with a known cause

In many cases of Sudden Sensorineural Hearing Loss, there is no effective treatment for the hearing loss itself, but further loss may be reduced and occasionally hearing may even be restored by preventive measures. For example, the adverse effects of drugs that can cause hearing loss may be halted and to some extent reversed if the patient stops taking that particular drug (in consultation with a medical adviser) and replaces it with a different agent. Similarly, the risk of noise-induced hearing loss can be greatly reduced by appropriate ear defenders, and the use of helmets in certain sports and by motor cyclists undoubtedly reduces the risk of deafness in skull fractures due to head injuries. Finally, timely surgery for the 'dangerous' type of middle ear infection will prevent what is called suppurative labyrinthitis, which occurs if the infection spreads to the inner ear.

When a perilymph leak (a leak of one of the inner ear fluids) is suspected, bed rest is advised for up to five days. If there is no improvement in the hearing after that period, surgical exploration of the ear is needed, with repair of the leak when possible.

In cases of Ménière's Disease, most forms of medical treatment are aimed at controlling the crippling attacks of vertigo, while preserving the hearing if at all possible. In severe cases, if attacks persist despite medication, a consultant may advise surgery. A number of different operations have been devised, some to reduce pressure in the inner ear, others to diminish or abolish the abnormal impulses passing from ear to brain. For more information about Ménière's Disease, see the Deafness Research UK factsheet on this topic.

COPING WITH SUDDEN SENSORINEURAL HEARING LOSS

When a hearing loss occurs suddenly, the shock and sense of loss can be overwhelming.

If you experience a sudden hearing loss, you may go through a range of emotions including denial, hope (for a cure or improvement), anger, isolation, acceptance, embarrassment and resolve. Other people may react to your hearing loss by feeling awkward, embarrassed or unsympathetic. You may feel embarrassed about it yourself. It is important to be open and honest with the people around you, and explain what you want them to do. With time, and with the right sort of professional support, as well as the support of your friends and family, you should find that you become more skilled at being aware of your environment and communicating. As your skills grow you will feel more in control and you might find that you are able to adapt your leisure interests, social life and hobbies to your hearing loss.

There are many sources of information and advice available to help adjust if it seems that the hearing loss is permanent. This includes guidance on equipment

and devices available, and implications for employment and benefits. A wide range of services and support, including rehabilitation courses can also be accessed. Hearing Therapists and lipreading classes can be valuable sources of support. Details of relevant organisations to contact are given below.

COPING WITH PERMANENT HEARING LOSS

Hearing Aids

When treatment is impossible or ineffective, most patients whose hearing loss is diagnosed as permanent can be helped by a hearing aid or aids and/or by other assistive devices. Hearing aids are available free of charge through the NHS and to receive one, patients are generally referred within their local hospital to an audiologist.

The type of hearing aid prescribed will depend on the severity of the hearing loss, the frequencies affected and whether one or both ears are affected. On the NHS, the main type of hearing aid is worn behind the ear. For people with one-sided deafness there is a particular type of hearing aid known as a CROS (Contralateral Routing of Signals) aid which can be of benefit in certain situations where the microphone is located on the side of the worse ear and the signal from it is fed to the better ear. There is also a BICROS aid in which the better ear receives an additional signal from a second microphone placed on the better-hearing side. These systems can mean that users are able to hear sounds directed to the impaired ear more easily, without turning the better ear towards the sound, and they also provide extra cues for locating the direction of sound sources.

Another hearing aid suitable for single sided sensorineural hearing loss is a bone anchored hearing aid (BAHA), which transfers sound through bone conduction. It works by stimulating the cochlea of the normally hearing ear giving users the sensation of hearing sounds from the deaf side. An operation is required which takes approximately thirty minutes to insert a small titanium implant into the mastoid bone behind the ear. The site is then left for three months allowing the implant to fuse with the skull bone in a process known as osseointegration before the hearing aid (a small rectangular box) can be attached and easily removed for sleeping and bathing.

There are a number of hospitals in the UK where the BAHA procedure is currently performed and where a person would need to be referred for assessments. Part of the assessment involves a trial of the device on a headband which can give a good idea of whether this type of aid will be helpful.

Assistive Devices

Even when a hearing loss is relatively mild, people with sudden sensorineural hearing loss often have difficulty hearing high frequency sounds such as telephones, doorbells and alarm clocks, but may find it unnecessary to wear a hearing aid all the time. To help, there is a wide range of special equipment available, such as amplified doorbells, vibrating alert systems, loop systems and headphones. Social services may help to pay for, or provide, equipment. For more information contact your local social services department. The Government's

Access to Work scheme may help to pay for equipment you need for work or for job interviews.

There are also a number of suppliers from whom you can purchase equipment. See contact details below.

Lipreading

In combination with an efficient hearing aid, lipreading plays an important part in helping people with acquired hearing loss to communicate better and regain confidence. In terms of understanding speech, the most important sounds are the consonants. They are formed by the lips, tongue and teeth and so can be seen. Therefore, reading the lips can provide vital clues for the understanding of speech.

Learning to lipread is challenging, but it is worth persevering, as the improvement in communication can be significant. Lipreading classes can also be very good places to meet other people who have had similar experiences. Lipreading classes are usually offered at local adult education centres and/or colleges. However, if you have difficulty finding a class, contact the Association of Teachers of Lipreading to Adults (ATLA) whose address is given below.

Hearing Therapy

A qualified Hearing Therapist can play a vital role in helping someone with an acquired hearing loss. They use a variety of methods to work out an individual programme for each patient to assist them in managing their hearing loss, improving their communication skills and enabling them to participate fully in their daily activities. Hearing Therapists can offer:

- help with hearing aids;
- auditory training (working with the remaining hearing);
- counselling both for the deafened person and their family
- advice about environmental aids;
- lipreading instruction; and
- tinnitus management.

Some Hearing Therapists may also specialise in cochlear implants, helping with balance problems or working with patients with learning disabilities or dual sensory loss. Although not all NHS Trusts employ Hearing Therapists, most are situated in either the Audiology or Ear, Nose and Throat departments of hospitals.

Cochlear Implants

The cochlear implant is one of the major scientific triumphs of the last few decades. They are increasingly being used to help adults with acquired total or profound hearing loss in both ears, as well as in children born profoundly deaf or acquiring deafness before the development of speech. They work by electrically stimulating the auditory nerve (nerve of hearing) to give the user a sense of sound, and, in the case of a child born profoundly deaf, the potential to understand speech and to learn to speak.

Although cochlear implants are available through the NHS, rigorous assessment is undertaken to ensure that each recipient meets certain referral criteria. These criteria change over time and may vary between cochlear implant centres.

For more information about cochlear implants, see the Deafness Research UK factsheets on this topic.

FURTHER INFORMATION

If any of your questions concerning sudden sensorineural hearing loss 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)

Freephone: 0808 808 2222

Textphone: 020 7915 1412

E-mail: info@deafnessresearch.org.uk

or click the 'ask question' option from our website homepage:
www.deafnessresearch.org.uk

You can also get information from other organisations including:

Association of Teachers of Lipreading to Adults (ATLA)

C/o Hearing Concern LINK, 19 Hartfield Road, Eastbourne, East Sussex, BN21 2AR

Website: www.lipreading.org.uk Email: ATLA@lipreading.org.uk

Hearing Concern LINK

19 Hartfield Road, Eastbourne, East Sussex, BN21 2AR

Tel: 01323 638230 / Textphone: 01323 739998

Website: www.hearingconcernlink.org Email: info@hearingconcernlink.org

National Association for Deafened People

PO Box 50, Amersham, Bucks, HP6 6XB

Tel: 01227 379 538 / Textphone: 01227 762 879

Website: www.nadp.org.uk Email: enquiries@nadp.org.uk

Royal National Institute for Deaf People (RNID)

19-23 Featherstone Street, London, EC1Y 8SL

Tel: 0808 808 6666 / Textphone: 0808 808 007

Website: www.rnid.org.uk Email: informationline@rnid.org.uk

RNID Shop

1 Haddonbrook Business Centre, Orton Southgate, Peterborough PE2 6YX

Tel: 0870 789 8855 / Textphone: 01733 238020
Website: www.rnid.org.uk/shop Email: solutions@rnid.org.uk

Connevans Limited

54 Albert Road North, Reigate, Surrey, RH2 9YR
Tel: 01737 247571/ Textphone: 01737 243134
Website: www.connevans.org.uk

Gordon Morris Ltd.

Unit 21, Wessex Park, Somerton Business Park, Somerton, Somerset, TA11 6SB
Telephone: 01458 272121
Website: www.gordonmorris.co.uk Email: online@gordonmorris.co.uk

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Tel: 01642 247789, Textphone: 01642 251310, Fax: 01642 230827
Website: www.sarabec.co.uk Email: mail@sarabec.co.uk

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 0207 833 1733 or write to:

Deafness Research UK, 330-332 Gray's Inn Rd, London WC1X 8EE
Charity no. 326915

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