



# Sudden Sensorineural Hearing Loss

This factsheet has been written for people who have experienced a sudden loss of hearing and who wish to know more about this type of hearing loss. It may also be useful for the relatives and friends of somebody who has 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
- Research into Sudden Sensorineural Hearing Loss
- 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 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 try to use the impaired ear. Sudden Sensorineural Hearing Loss (SSHL) is a medical emergency and you will need to contact your doctor immediately.

People with Sudden Sensorineural Hearing Loss often describe themselves as 'deafened'. They may also say they have an 'acquired hearing loss'. If you lose your hearing after learning to speak, you are said to have 'post-lingual hearing loss'.

## WHAT CAUSES SUDDEN SENSORINEURAL HEARING LOSS?

### **Sudden Sensorineural Hearing Loss with no known cause**

Most of the time the cause is unknown – it is only possible to establish a firm cause of Sudden Sensorineural Hearing Loss in about 15% of cases. The remaining 85% are referred to as idiopathic (with no known cause). In the majority of these idiopathic cases, the hearing is affected in only one ear (unilateral) and will improve spontaneously. It can be tempting, therefore, to adopt a 'wait-and-see' policy, but you should be seen by a specialist as soon as possible if you experience Sudden Sensorineural Hearing Loss, because it is important to establish a diagnosis since treatment may be needed if a specific cause is found.

In an attempt to find a reason for these idiopathic cases of hearing loss, two main theories have been advanced. The first theory 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 cases. The second theory is that it may be due to some vascular defect (relating to blood vessels). There is no conclusive proof to support either view.

### **Infections**

Meningitis is one of the commonest causes of severe or profound acquired deafness in infants and children. 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 Sudden Sensorineural Hearing Loss. In measles, the loss is usually moderate to profound and bilateral, whereas in mumps it usually affects only one ear. The herpes zoster virus may on rather rare occasions produce a Sudden Sensorineural 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 make you feel dizzy, give you tinnitus (ringing in the ears), and can lead to sensorineural hearing loss, but usually only in one ear. Bacterial labyrinthitis often results in permanent hearing loss.

### **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**

Noise induced hearing loss (NIHL) is usually gradual in onset and can be prevented by 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 and fireworks, especially in enclosed spaces.

This sudden hearing loss can range from total deafness in one or both ears to a relatively minor high-frequency loss. In these latter cases, the hearing may recover spontaneously in time.

### **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 your specialist.

### **Barotrauma**

Barotrauma may occur when the ear is exposed to sudden pressure changes, as in 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).

### **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 Sensorineural Hearing Loss, and potential treatments, but it probably accounts for less than one per cent of all cases of hearing loss. Sudden deafness has been known to occur in established autoimmune diseases such as rheumatoid arthritis and diabetes. However, there is no evidence that rheumatoid arthritis or diabetes cause sudden deafness; they are common conditions and it is likely that some people who have them also happen to develop idiopathic Sudden Sensorineural Hearing Loss.

### **Ototoxicity**

Sudden Sensorineural Hearing Loss is sometimes attributed to ototoxic drugs (drugs that may damage the inner ear) and rare cases have been reported following the use of (for example) gentamicin (an antibiotic). 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 your doctor prescribes ototoxic drugs, you should discuss with them how this might affect your hearing.

### **Ménière's Disease**

Ménière's Disease affects about one in every two thousand adults in the UK population. It is a late-onset disease 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 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.

## **WHAT SHOULD YOU DO IF YOU LOSE YOUR HEARING SUDDENLY?**

If you think you have a sudden hearing loss in one or both ears, you need to see your GP (family doctor) and ask for an urgent referral to an ear, nose and throat (ENT) specialist. 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.

### **Sudden Sensorineural Hearing Loss where the cause is unknown**

Although hearing returns to normal or near-normal in about 70% of these cases, 'failure to investigate patients will inevitably lead to a missed diagnosis and a missed opportunity for treatment' (Booth<sup>1</sup>). Hence, in all such cases, urgent investigation is required.

Where cases of Sudden Sensorineural Hearing Loss may be due to a virus or vascular disorder, a wide range of treatments have been recommended, but none have had convincingly beneficial results. However, some viral infections respond to anti-viral agents like acyclovir, and in 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 Sudden Sensorineural Hearing Loss, 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. So far no value has been proven.

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<sup>1</sup> John B Booth, in Scott-Brown's *Otolaryngology*, 5<sup>th</sup> edition (1987), Vol. 3, p.424.

## **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 the hearing may even be restored by preventive measures. For example, the adverse effects of ototoxic drugs may be halted and to some extent reversed if the patient stops taking that particular drug (in consultation with a medical advisor) 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 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, bedrest 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**

Deafness is without doubt one of the most poorly understood of all disabilities and it attracts little sympathy from the hearing majority. In a 2002 survey by Deafness Research UK, 40 per cent of people questioned about the impact of hearing loss on their relationships said that communication with their partners had become more difficult. One in four said they now had to miss out on social events. This sense of isolation is often compounded by the need to wear a visible hearing device. When the 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 you adjust to your hearing loss. Deafness Research UK, Hearing Concern and RNID all provide a range of factsheets, and there may be a local support group you can join. The Link Centre for Deafened People provides a wide range of resources, services and support for deafened people. Hearing Therapists and lipreading classes can also be valuable sources of support.

## **COPING WITH PERMANENT HEARING LOSS**

### **Hearing Aids and Assistive Devices**

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.

There are many different types of hearing aid and they can be worn behind the ear, in the bowl of the ear or in the ear canal itself. The type of aid prescribed in each individual case will depend on the severity of the hearing loss, and whether one or both ears are affected. In the case of one-sided deafness, a CROS (Contralateral Routing of Signals) aid can be of benefit in certain situations. The microphone is located on the side of the worse ear and the signal from it is fed to the better ear. In a BICROS aid, 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.

Even when the hearing loss is relatively mild, people with Sudden Sensorineural Hearing Loss often have difficulty in hearing telephones and doorbells, alarm clocks and other environmental sounds, but it may be unnecessary to wear a hearing aid all the time. 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 social worker for deaf people or 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.

The Royal National Institute for Deaf People (RNID) produces some helpful information on the subject of assistive devices (contact details below). There are a number of suppliers from whom you can purchase equipment. For example, RNID Sound Advantage sells a wide range of individual and environmental aids to hearing.

### **Lipreading**

In combination with an efficient hearing aid, lipreading plays an important part in the rehabilitation of deafened people. In terms of understanding speech, the most important sounds are the consonants. As these are formed by the lips, tongue and teeth, they can be seen, and therefore reading the lips can provide vital clues for the understanding of speech.

Many deafened people find it very difficult to learn to lipread, 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 to yourself. The Association of Teachers of Lipreading to Adults (ATLA) is the body which regulates the profession in the United Kingdom.

### **Hearing Therapy**

Getting a hearing aid or aids is only the beginning of a long and often difficult process of rehabilitation for deafened people, particularly when both ears are affected. A qualified Hearing Therapist can play a vital role in this process. A Hearing Therapist uses 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).

In the great majority of cases of Sudden Sensorineural Hearing Loss it is the 'hair cells' in the cochlea which are affected. However, even when the very last of these tiny hair cells have been lost, there remain nearly always a number (perhaps 10 per cent) of auditory nerve fibres, which can be stimulated by the electrodes implanted into the cochlea in cochlear implantation.

The implantation of the electrodes and associated 'hardware' is only the beginning of a long process of intense rehabilitation. It may take as long as two years, or even more, before optimum results are obtained. Cochlear implantation is essentially a team effort, involving otologists, audiologists, speech therapists, psychologists and, with children, teachers of the deaf.

## **WHAT RESEARCH IS BEING DONE INTO SUDDEN SENSORINEURAL HEARING LOSS?**

Much of the recent research on Sudden Sensorineural Hearing Loss has been targeted, not surprisingly, on those idiopathic cases in which no firm cause can be established.

### **Steroids and anti-viral agents**

As mentioned above, the anti-viral agent acyclovir is occasionally used to treat Sudden Sensorineural Hearing Loss, on the assumption that some cases may have a viral origin. However, researchers are still investigating how effective acyclovir is as a treatment for hearing loss.

### **Reducing clotting in the artery supplying the cochlea**

It is still unclear whether vascular factors play a part in Sudden Sensorineural Hearing Loss, but on the assumption that they have a role, many attempts have been made to reduce the risk of clotting in the very small artery supplying the cochlea. A study in Germany<sup>2</sup> on patients with Sudden Sensorineural Hearing Loss found that some of them had excesses in the blood of fibrinogen (which is involved in the process of clotting) and of the 'dangerous' type of cholesterol. The researchers then tried to find out whether partially removing these harmful substances – an established procedure in patients suffering from coronary artery disease – could produce better results than standard treatments of Sudden Sensorineural Hearing Loss. The results showed small, but not significant, improvements, but the author concluded that this procedure could reasonably be used as an alternative to conventional therapy.

### **Establishing an immune disorder connection**

Many authorities include immune disorders in their lists of causes of Sudden Sensorineural Hearing Loss, although the hearing loss caused by autoimmune inner ear disease (AIED) is more commonly gradual than sudden. Researchers are working on developing a test which could determine whether an immune disorder has led to Sudden Sensorineural Hearing Loss.

### **Technology**

In addition to research into the causes of Sudden Sensorineural Hearing Loss, there is a lot of work going on to improve hearing aids and cochlear implants. Research on hearing aids includes the development of digital technology, improving evaluation and fitting procedures and the development of more effective sound processing technologies to improve the perception of music and the human voice. For more information about research into hearing aids, see the Deafness Research UK factsheet on Choosing a Hearing Aid. Research into improving cochlear implants is advancing all the time. Areas of research include improving the quality of sound, improving tuning procedures, investigating the advantages of bilateral implants (implants in both ears) and techniques which can provide high-frequency hearing via an implant whilst allowing conventional amplification (e.g. via a hearing aid) to provide low-frequency hearing. For more information see the Deafness Research UK factsheet on Cochlear Implants.

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<sup>2</sup> *The Lancet*, 7 December 2002.

## **Ménière's Disease**

Scientists funded by Deafness Research UK are currently conducting a major project to improve understanding of the causes of Ménière's Disease. Many cases of Ménière's are sporadic but in 7% other family members are similarly affected. This, together with the predominantly Caucasian/Eurasian distribution, is strong evidence of a genetic predisposition.

Preliminary genetic studies have pinpointed one area of the human genome which could be involved. Deafness Research UK has funded the initial molecular genetic study of this site. In due course other candidate genes will be examined. The identification of the Ménière's disease gene(s) made possible by this funding will lead to a boost to the hypotheses as to the cellular pathways involved in the underlying cause. This should in turn help to direct the search for new therapeutic and even preventative strategies, though in the long-term. Identifying the gene(s) predisposing to Ménière's disease will bring two immediate clinical benefits; it will speed up early diagnosis, which can sometimes be difficult, and it will provide a predictive test for relatives or descendants of affected patients, especially in familial cases.

## **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](mailto:info@deafnessresearch.org.uk)

or click the 'ask question' option from our website homepage:  
[www.deafnessresearch.org.uk](http://www.deafnessresearch.org.uk)

You can also get information from other organisations including:

### **Association of Teachers of Lipreading to Adults (ATLA)**

PO Box 506, Henley, Stoke-on-Trent, ST2 9RE

email: [atla@lipreading.org.uk](mailto:atla@lipreading.org.uk)

### **Hearing Concern Link**

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

Tel: 01323 638230 / Textphone: 01323 739998

Email: [info@hearingconcernlink.org](mailto:info@hearingconcernlink.org)

## **National Association for Deafened People**

PO Box 50, Amersham, Bucks, HP6 6XB

Tel: 01227 379 538 / Textphone: 01227 762 879

email: [enquiries@nadp.org.uk](mailto: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

email: [informationline@rnid.org.uk](mailto:informationline@rnid.org.uk)

## **RNID Sound Advantage**

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

Tel: 0870 789 8855 / Textphone: 01733 238020

email: [solutions@rnid.org.uk](mailto:solutions@rnid.org.uk) / Website: [www.rnidshop.com](http://www.rnidshop.com)

## **FURTHER READING**

More detailed medical information about Sudden Sensorineural Hearing Loss can be found in Ballantyne's Deafness (6<sup>th</sup> Edition), edited by John Graham and Mike Martin, and published by Whurr Publishers Ltd, 196 Compton Terrace, London N1 2UN.

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 WC1X8EE  
Charity no. 326915

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This factsheet has been produced by Deafness Research UK, in consultation with a team of medical and scientific experts who form part of our Communications Advisory Panel. Whilst all reasonable efforts have been made to ensure the information and advice given is taken from reputable sources and passed to the public in good faith, no responsibility can be taken on the part of Deafness Research UK or its advisors for any error or omission. You should not act on any advice without first referring to your family doctor or another medically qualified adviser.

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